PCAD Project

Working Resources List on Dementia Care Management and Intellectual Disability

Preparing Community Agencies for Adults Affected by Dementia - "PCAD" Project

v.26d

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To find resource or content of interest, search by keyword (e.g., assessment, carer, decline, etc.) in PDF reader.

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Acton, D., Duncan, C., & Jaydeokar
Co-production of post-diagnostic psychosocial intervention with carers of people with intellectual disability and dementia
Advances in Mental Health and Intellectual Disabilities, 2022, 16(3), 169-178. https://doi.org/10.1108/AMHID-01-2022-0006
Abstract: This paper aims to underline the importance of using a collaborative approach when designing and adapting a post diagnostic psychosocial intervention of cognitive stimulation therapy (CST) for people with intellectual disability and dementia. As part of a service improvement, a manual of CST was adapted, for delivery in clinical practice. A qualitative co-production method allowed participants with a lived experience to provide regular feedback relating to the development of the adapted CST manual and intervention program. This feedback was used to make continual development changes to the CST manual. The study demonstrated co-production with those who provide care is valuable in adapting psychosocial therapies for people with an intellectual disability and dementia. Additional findings identified the need for carer education in ageing, dementia care and the physical health needs for older people with intellectual disability. To the best of the authors' knowledge, this is the first study that has used a co-production approach with families and carers in adapting a group therapy program for people with an intellectual disability. This paper underlines the need for post diagnostic clinical interventions for people with dementia and those who provide care.

Adams, D., Oliver,C., Kalsy, S., Peters, S., Broquard, M., Basra, T., Konstandinidi, E., & McQuillan, S.
Behavioural characteristics associated with dementia assessment referrals in adults with Down syndrome.
Journal of Intellectual Disability Research. 2008 Apr;52(Pt 4):358-68. Epub 2008 Jan 22.
Abstract: Behavioral changes associated with dementia in Down syndrome are well documented, yet little is known about the effect of such behaviors on carers and referral. By comparing the behavioral and cognitive profiles of individuals referred for a dementia assessment with those of individuals not referred, some insight can be gained into behavioral characteristics that initiate referral for specialist support or interventions. Forty-six adults with Down syndrome were divided into two groups dependent upon method of entry into the study; post-referral to a specialist service for older adults with intellectual disabilities and Down syndrome for a dementia assessment (n = 17) or after receiving information sent out to day centers and residential homes (n = 29). These groups were compared on established measures of dementia alongside two informant measures of behavior. Those referred for a dementia assessment evidenced scores indicative of cognitive decline on both informant and direct Neuropsychological Assessments and showed more behavioral excesses, but not deficits, and lower socialization and coping skills than those in the comparison group. Carers of those referred for a dementia assessment reported a greater impact of behavioral excesses on staff than on the individual showing the behavior in contrast to the comparison group. The behavioral differences between those referred and the comparison group suggest that two factors are involved in the instigation of a referral for a dementia assessment: the nature of the behavioral presentation (excesses rather than deficits) and the effect of that behavioral change upon the care staff.

Adam, E., Sleeman, K.E., Brearley, S., Hunt, K., & Tuffrey-Wijne, I.
The palliative care needs of adults with intellectual disabilities and their access to palliative care services: A systematic review.
Palliative Medicine, 2020 Sep, 34(8), 1006-1018. doi: 10.1177/0269216320932774. Epub 2020 Jun 17.
Abstract: There is evidence that people with intellectual disabilities experience healthcare inequalities, including access to specialist palliative care, but to date, there has not been a systematic review of empirical evidence. To identify the palliative care needs of adults with intellectual disabilities and the barriers and facilitators they face in accessing palliative care. Systematic review using a narrative synthesis approach (International prospective register of systematic reviews (PROSPERO) registration number: CRD42019138974). Five databases were searched in June 2019 (MEDLINE, Embase, PsycINFO, the Cochrane library and CINAHL) along with hand searches and a search of the grey literature. All study designs were included. A total of 52 studies were identified, all of which were conducted in high-income countries, the majority in the United Kingdom (n = 28). From a total of 2970 participants across all studies, only 1% were people with intellectual disabilities and 1.3% were family members; the majority (97%) were health/social care professionals. Identified needs included physical needs, psychosocial and spiritual needs, and information and communication needs. Barriers and facilitators were associated with education (e.g. staff knowledge, training and experience), communication (e.g. staff skill in assessing and addressing needs of people with communication difficulties), collaboration (e.g. importance of sustained multidisciplinary approach) and health and social care delivery (e.g. staffing levels, funding and management support). This review highlights the specific problems in providing equitable palliative care for adults with intellectual disabilities, but there is a lack of research into strategies to improve practice. This should be prioritised using methods that include people with intellectual disabilities and families.

Ahlström, G., Axmon, A., Sandberg, M., & Flygare Wallén, E.
Health care utilisation among older people with Down syndrome compared to specific medical guidelines for health surveillance: a Swedish national register study
BMC Health Services Research, 2020, Oct 15, 20(1), 949. doi:10.1186/s12913-020-05800-7.
Abstract: Specific medical guidelines for health surveillance exist for people with Down syndrome (DS) since 25 years but knowledge of adherence to the guidelines is lacking. The guidelines were developed to avoid unnecessary suffering from preventable conditions. The aims of the study were to investigate 1) planned health care visits in relation to the co-morbidities described in specific medical guidelines as a measure of adherence, 2) unplanned health care visits as a measure of potentially unmet health care needs and 3) gender differences in health care utilisation among older people with DS. This register-based study includes people with DS (n = 472) from a Swedish national cohort of people with intellectual disability (n = 7936), aged 55 years or more, and with at least one support according to the disability law, in 2012. Data on inpatient and outpatient specialist health care utilization were collected from the National Patient Register for 2002-2012. A total of 3854 inpatient and outpatient specialist health care visits were recorded during the 11 years, of which 54.6% (n = 2103) were planned, 44.0% (n = 1695) unplanned and 1.4% (n = 56) lacked information. More than half of the visits, 67.0% (n = 2582) were outpatient health care thus inpatient 33% (n = 1272). Most planned visits (29.4%, n = 618) were to an ophthalmology clinic, and most unplanned visits to an internal medicine clinic (36.6%, n = 621). The most common cause for planned visits was cataract, found at least once for 32.8% in this cohort, followed by arthrosis (8.9%), epilepsy (8.9%) and **dementia** (6.6%). Pneumonia, pain, fractures and epilepsy each accounted for at least one unplanned visit for approximately one-fourth of the population (27.1, 26.9, 26.3 and 19.7% respectively). Men and women had similar numbers of unplanned visits. However, women were more likely to have visits for epilepsy or fractures, and men more likely for pneumonia. Increased awareness of existing specific medical guidelines for people with DS is vital for

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preventive measures. The relatively few planned health care visits according to the medical guidelines together with a high number of unplanned visits caused by conditions which potentially can be prevented suggest a need of improved adherence to medical guidelines.

Alftberg, Å., Johansson, M., & Ahlström, G.
Ambivalence among staff regarding ageing with intellectual disabilities: Experiences and reflections.
Journal of Intellectual Disabilities, 2021 Jun;25(2):192-209. doi: 10.1177/1744629519874997. Epub 2019 Sep 30.
Abstract: This study explores the experiences and reflections of staff in intellectual disability (ID) services concerning ageing with ID. Qualitative interviews were conducted with 24 staff members in group homes and daily activity centres. The findings showed that the staff were uncertain about the signs of ageing in people with intellectual disabilities; they compared the life conditions of these people with conditions in older people without intellectual disabilities. Their emphasis on an active lifestyle was very strong. The staff members also mentioned uncertainty about how to facilitate assistive devices and whether 'ageing in place' was the best solution. The overall theme was manifested as ambivalence where notions of older people with intellectual disabilities seemed incompatible with notions of old age in general and could be explained by the theoretical concept of age coding. The findings of this study indicate the need to provide education about ageing to staff working in ID services.

Ali, A., Brown, E., Tsang, W., Spector, A., Aguirre, E., Hoare, S., Hassiotis, A.
Individual cognitive stimulation therapy (iCST) for people with intellectual disability and dementia: a feasibility randomised controlled trial.
Aging & Mental *Health. 2022 Apr;26(4):698-708. doi: 10.1080/13607863.2020.1869180.* Epub 2021 Jan 4. PMID: 33393364.
Abstract: To examine the feasibility, acceptability and fidelity of individual Cognitive Stimulation Therapy (iCST) in people with intellectual disability (ID) and dementia. We aimed to recruit forty dyads (carer and individual with dementia and ID) who were randomised to iCST or a waiting list control group. Both groups received treatment as usual. Family and paid carers delivered the manualised intervention (40 sessions over 20 weeks). Recruitment and retention of participants, intervention adherence, fidelity and acceptability were assessed. Outcome measures of cognition, adaptive functioning, quality of life (QoL) and carer outcomes were collected at baseline, midpoint (11 weeks) and at 21 weeks. Qualitative interviews were conducted with six carers about their experience of iCST. Forty dyads were recruited over 10 months from 12 National Health Service trusts. One dyad dropped out and 87.5% and 97.5% completed the midpoint and end-point assessments respectively. Assessment of fidelity indicated that the correct session structure was not followed; 70% completed at least 20 sessions and there was a high level of satisfaction with iCST. QoL was significantly higher in the iCST arm at 21 weeks (adjusted mean difference: 3.11; 95% CI: 0.64 to 5.58). There were no differences in the other outcome measures. The intervention was feasible and acceptable. A full-scale trial is warranted but some modifications are needed, including improved training and supervision for carers to improve fidelity

Altuna, M., Giménez, S., & Fortea, J.
Epilepsy in Down syndrome: A highly prevalent comorbidity
Journal of Clinical Medicine, 2021 (June 24), 10(13). 2776- [e-print]. doi: 10.3390/jcm10132776.
Abstract: Individuals with Down syndrome (DS) have an increased risk for epilepsy during the whole lifespan, but especially after age 40 years. The increase in the number of individuals with DS living into late middle age due to improved health care is resulting in an increase in epilepsy prevalence in this population. However, these epileptic seizures are probably underdiagnosed and inadequately treated. This late onset epilepsy is linked to the development of symptomatic Alzheimer's disease (AD), which is the main comorbidity in adults with DS with a cumulative incidence of more than 90% of adults by the seventh decade. More than 50% of patients with DS and AD **dementia** will most likely develop epilepsy, which in this context has a specific clinical presentation in the form of generalized myoclonic epilepsy. This epilepsy, named late onset myoclonic epilepsy (LOMEDS) affects the quality of life, might be associated with worse cognitive and functional outcomes in patients with AD dementia and has an impact on mortality. This review summarizes the current knowledge about the clinical and electrophysiological characteristics, diagnosis and treatment of epileptic seizures in the DS population, with a special emphasis on LOMEDS. Raised awareness and a better understanding of epilepsy in DS from families, caregivers and clinicians could enable earlier diagnoses and better treatments for individuals with DS.

Alzheimer's Association
Guidelines for dignity: Goals of specialized Alzheimer/dementia care in residential settings
47 pp.
Chicago: The Alzheimer's Association [919 North Michigan Avenue, Suite 1000, Chicago, IL 60611-1676] (1992)
Abstract: Standards for care and structure of care settings housing persons affected by Alzheimer's disease. Includes sections on philosophy, pre-admission activities, admission, care planning and implementation, adapting to changes in condition, staffing and training, physical environment and "success indicators."

Alzheimer's Australia
Down syndrome and Alzheimer's disease
12 pp.
[Place of publication not provided] (no date)
Source: http://www.cddh.monash.org/assets/dsad-booklet-final.pdf
Abstract: Informational booklet on dementia and people with Down syndrome jointly issued by Alzheimer's Australia, Down Syndrome Victoria, and Centre for Developmental Disability Health Victoria. Contains three main sections: (1) About Alzheimer's disease and Down syndrome, (2) Diagnosis, and (3) Support, as well as a section on local resources.

Alzheimer's Disease International
Planning and design guide for community based day care centres
21 pp.
London: Alzheimer's Disease International [45/46 Lower Marsh, London SE1 7RG, United Kingdom (www.alz.co.uk)] (1999)
Abstract: An illustrated 21-page booklet highlighting main design issues and suggestions for organizing an effective environment for adults with dementia - with applications for residential environment.

Alzheimer's Disease Society
Safe as houses -- Living alone with dementia (A resource booklet to aid risk management)
London: Alzheimer's Disease Society [Gordon House, 10 Greencoat Place, London SW1P 1PH, United Kingdom] (1994)
30 pp.
Abstract: A 30-page booklet designed for the carer who is concerned about an older person with early to mid-stage dementia who may be living on their own. The booklet examines risks that the older adult may encounter and suggests how they could be minimized. The intent of the booklet is to aid the older person remain functional at home, with as minimal risk, for as long as possible. Covers personal care, finances, wandering, security, medication, utilities, and household safety. Whilst information is generic, resource information is geared toward the UK.

Alzheimer's Society
Learning disabilities and dementia
Alzheimer's Society UK, (2022), Alzheimer's Society, 43-44 Crutched Friars, London, EC3N 2AE
https://www.alzheimers.org.uk/about-dementia/types-dementia/learning-disabilities-dementia
Abstract: Web-based information produced in the UK on the topic of intellectual disabilities and dementia. Contains background information, as well as diagnosis, identificatiion of symptoms and support and care services.

Alzheimer's Society
Supporting a person with dementia who also has a learning disability
Alzheimer's Society UK, (2023), Alzheimer's Society, 43-44 Crutched Friars, London, EC3N 2AE
https://www.alzheimers.org.uk/get-support/publications-and-factsheets/dementia-together/supporting-person-dementia-learning-disability
Abstract: Informational webpage on how to approach helping a relative or friend with an intellectual disability who's diagnosed with dementia.

Anderson-Mooney, A.J., Schmitt, F.A., Head, E., Lott, I.T., & Heilman, K.M.
Gait dyspraxia as a clinical marker of cognitive decline in Down syndrome: A review of theory and proposed mechanisms

Brain and Cognition, 2016, Apr, 104, 48-57. doi: 10.1016/j.bandc.2016.02.007. Epub 2016 Feb 27.
Abstract: Down syndrome (DS) is the most common genetic cause of intellectual disability in children. With aging, DS is associated with an increased risk for Alzheimer's disease (AD). The development of AD neuropathology in individuals with DS can result in further disturbances in cognition and behavior and may significantly exacerbate caregiver burden. Early detection may allow for appropriate preparation by caregivers. Recent literature suggests that declines in gait may serve as an early marker of AD-related cognitive disorders; however, this relationship has not been examined in individuals with DS. The theory regarding gait dyspraxia and cognitive decline in the general population is reviewed, and potential applications to the population with individuals with DS are highlighted. Challenges and benefits in the line of inquiry are discussed. In particular, it appears that gait declines in aging individuals with DS may be associated with known declines in frontoparietal gray matter, development of AD-related pathology, and white matter losses in tracts critical to motor control. These changes are also potentially related to the cognitive and functional changes often observed during the same chronological period as gait declines in adults with DS. Gait declines may be an early marker of cognitive change, related to the development of underlying AD-related pathology, in individuals with DS. Future investigations in this area may provide insight into the clinical changes associated with development of AD pathology in both the population with DS and the general population, enhancing efforts for optimal patient and caregiver support and propelling investigations regarding safety/quality of life interventions and disease-modifying interventions.

Antonangeli, J.M.
Of two minds: A guide to the care of people with the dual diagnosis of Alzheimer's Disease and mental retardation.
167 pp.
Malden, Mass.: Cooperative for Human Services [110 Pleasant Street, Malden, MA 02148] (1995)
Abstract: Written in training manual format, this text covers a range of topics related to dementia among persons with intellectual disabilities, including the notions behind dementia, structuring physical environments, safety and control issues, communication strategies, assessing and aiding with activities of daily living, behavior management strategies, medical concerns, and aiding carers. Much of the text is drawn from general practice in the Alzheimer's field with reference to application for settings with persons with intellectual disabilities.

Antonangeli. J.M.
The Alzheimer project: formulating a model of care for persons with Alzheimer's disease and mental retardation
American Journal of Alzheimer's Disease, 1995, 10(4), 13-16.
Abstract: Article speaks to a pilot project conducted in Massachusetts to increase staffing, education and Alzheimer case management supports. Special supports were designed and offered to a number of adults with Down syndrome affected by dementia, including specialize assessments, team care planning meetings, home adaptations and behavior loss supports.

Arvio, M., & Bjelogrlic-Laakso ,N.
Screening of dementia indicating signs in adults with intellectual disabilities
Journal of Applied Research in Intellectual Disabilities, 2021 (May 01), https://doi.org/10.1111/jar.12888
Abstract: In intellectual disability, the cognitive delay is observed during developmental age, whereas in dementia, cognitive decline occurs during post-developmental period. So far, the risk of dementia in people with intellectual disability, excluding those with Down syndrome, is poorly known.
We screened dementia signs in a study group of 230 adults (34–80 years of age) with the help of the British Present Psychiatric State—Learning Disabilities assessment. Of the study members, 42% showed two or more signs. The overall frequency of symptoms did not differ between age groups. The number of individuals with a genetic syndrome or disease manifesting with a shortened lifespan was greater in the younger age groups when compared to the older age groups. People with an intellectual disability represent numerous rare syndromes with comorbidities. It seems that dementia signs may affect any age groups of adults with intellectual disability.

Arvio, M., & Bjelogrlic-Laakso, N.
Down syndrome - Onset age of dementia.
Journal of Alzheimer's Disease & Parkinsonism, 2017, 7(3), 1000329. doi: 10.4172/2161-0460
Abstract: Alzheimer's disease is the most common cause of death in people who have Down syndrome (DS).This prospective, population-based, 15-year follow-up study aimed to define the onset age of dementia. At baseline 98 adults were screened for the first time by using the Present Psychiatric State-Learning Disabilities assessment;19 also participated a second parallel follow up survey. These screenings were repeated twice more during the study. The number of study subjects during the 15 year follow-up period decreased. Another study found that adaptive skills in people with DS start to deteriorate at the age of 35 and our current findings indicate that also the first signs of dementia appear at the same age. The indicative signs for dementia increased rapidly after the age of 35 and appeared most frequently as reduced self-care skills, loss of energy, impaired understanding and forgetfulness. Regular follow-up of people who have Down syndrome from the age of 30 onward enables appropriate interventions to delay the progression of dementia. Early recognition of dementia is of uttermost importance, because any deterioration in daily performance due to some treatable but undiagnosed condition may result in more restrictive residential care. Once memory changes have been observed, they may not reflect the onset of AD (depression, sleep disorders, B12-vitamin deficiency, hypothyroidism, urinary tract infections or polypharmacy may be the underlying causes behind the first dementia signs. When periodic assessments are completed at regular intervals by a carer (who is familiar with the person for a long time) explains why the first dementia signs were recognized at clearly younger age in our study than in other reported surveys (i.e., 35 versus 48-56 years).

Aschenbrenner, A.J,, Baksh, R.A., Benejam, B., Beresford-Webb, J.A., Coppus. A., Fortea, J., Handen, B.L., Hartley, S., Head, E., Jaeger, J., Levin, J., Loos;i. S.V., Rebillat, A-S., Sacco, S., Schmitt, F.A., Thurlow, K.E., Zaman, S., Hassenstab, J., & Strydom, A.
Markers of early changes in cognition across cohorts of adults with Down syndrome at risk of Alzheimer's disease
Alzheimer's and Dementia (Amsterdam, Netherlands), 2021,13(1), e12184. doi: 10.1002/dad2.12184
Abstract: Down syndrome (DS), a genetic variant of early onset Alzheimer's disease (AD), lacks a suitable outcome measure for prevention trials targeting pre-dementia stages. We used cognitive test data collected in several longitudinal aging studies internationally from 312 participants with DS without dementia to identify composites that were sensitive to change over time. We then conducted additional analyses to provide support for the utility of the composites. The composites were presented to an expert panel to determine the most optimal cognitive battery based on predetermined criteria. There were common cognitive domains across site composites, which were sensitive to early decline. The final composite consisted of memory, language/executive functioning, selective attention, orientation, and praxis tests. We have identified a composite that is sensitive to early decline and thus may have utility as an outcome measure in trials to prevent or delay symptoms of AD in DS.

Auty, E., & Scior, K.
Psychologists' clinical practices in assessing dementia in individuals with Down syndrome
Journal of Policy and Practice in Intellectual Disabilities, 2008, 5(4), 259-268. DOI:10.1111/j.1741-1130.2008.00187.x
Abstract: There are now ample guidelines for the assessment and diagnosis of possible dementia in individuals with intellectual disabilities (ID) and Down syndrome. However, little is known about their implementation in clinical practice. This study set out to examine the clinical practice of one key professional group, namely clinical psychologists. A national survey of clinical psychologistsin ID services in the United Kingdom was undertaken. Detailed descriptions of clinical practice were obtained from 64 psychologists. Responses were further explored in focus groups. The results suggest marked variability in practice, assessment methods, and explanations clinicians give to service users and carers. Clinicians described struggling with the ethics and practicalities of how to present dementia assessments to individuals with ID and highlighted a need for more research, debate, and guidance. They also noted numerous shortfalls in service provision once aging individuals with ID show signs of dementia. Further research and clearer consensus guidelines are needed to improve assessment and diagnosis for this group.

Axmon, A., Björne, P., Nylander, L., & Ahlström, G.
Psychiatric diagnoses in older people with intellectual disability in comparison with the general population: a register study
Epidemiology and Psychiatric Sciencies, 2018 Oct, 27(5), 479-491. doi: 10.1017/S2045796017000051

Abstract: To describe the occurrence of psychiatric diagnoses in a specialist care setting in older people with intellectual disability (ID) in relation to those found in the same age group in the general population. A cohort of people with ID (n = 7936), aged 55 years or more in 2012, was identified, as was an age and sex-matched cohort from the general population (n = 7936). Information regarding psychiatric diagnoses during 2002–2012 was collected from the National Patient Register, which contains records from all inpatient care episodes and outpatient specialist visits in Sweden. The mean age at the start of data collection (i.e. January 1st, 2002) was 53 years (range 44–85 years). Seventeen per cent (n = 1382) of the people in the ID cohort had at least one psychiatric diagnosis recorded during the study period. The corresponding number in the general population cohort was 10% (n = 817), which translates to an odds ratio (OR) of 1.84. The diagnoses recorded for the largest number of people in the ID cohort were 'other' (i.e. not included in any of the diagnostic groups) psychiatric diagnoses (10% of the cohort had at least one such diagnosis recorded) and affective disorders (7%). In the general population cohort, the most common diagnoses were affective disorders (4%) and alcohol/substance-abuse-related disorders (4%). An increased odds of having at least one diagnosis was found for all investigated diagnoses except for alcohol/substance-abuse-related disorders (OR = 0.56). The highest odds for the ID cohort was found for diagnosis of psychotic disorder (OR = 10.4) followed by attention deficit/hyperactive disorder (OR = 3.81), **dementia** (OR = 2.71), personality disorder (OR = 2.67), affective disorder (OR = 1.74) and anxiety disorder (OR = 1.36). People with ID also had an increased odds of psychiatric diagnoses not included in any of these groups (OR = 8.02). The percentage of people with ID who had at least one diagnosis recorded during the study period decreased from more than 30% among those aged 55–59 years in 2012 (i.e. born 1953–1957) to approximately 20% among those aged 75+ years in 2012 (i.e. born in or before 1937). Older people with ID seem to be more likely to have psychiatric diagnoses in inpatient or outpatient specialist care than their peers in the general population. If this is an effect of different disorder prevalence, diagnostic difficulties or differences in health care availability remains unknown. More research is needed to understand the diagnostic and treatment challenges of psychiatric disorders in this vulnerable group.

Axmon, A., Karlsson, B., & Ahlström, G.
Health care utilisation among older persons with intellectual disability and dementia: a registry study
Journal of Intellectual Disability Research, 2016, Dec, 60(12), 1165-1177. doi 10.1111/jir.12338. Epub 2016 Oct 11.
Abstract: Both persons with intellectual disability (ID) and persons with dementia have high disease burdens, and consequently also high health care needs. As life expectancy increases for persons with ID, the group of persons with the dual diagnosis of ID and dementia will become larger. Through national registries, we identified 7936 persons who had received support directed to persons with ID during 2012, and an age- and gender-matched sample from the general population. A national registry was also used to collect information on health care utilisation (excluding primary care) for the period 2002-2012. Health care utilization was measured as presence and number of planned and unplanned in-patient and out-patient visits, as well as length of stay. In comparison with persons with ID but without dementia, persons with ID and dementia were more likely to have at least one planned out-patient visit (odds ratio [OR] 8.07), unplanned out-patient visit (OR 2.41), planned in-patient visit (OR 2.76) or unplanned in-patient visit (OR 4.19). However, among those with at least one of each respective outcome, the average number of visits did not differ between those with and without dementia. Persons with ID and dementia were less likely to have at least one planned out-patient visit than persons with dementia in the general population sample (OR 0.40), but more likely to have at least one unplanned in-patient visit (OR 1.90). No statistically significant differences were found for having at least one unplanned out-patient or planned in-patient visit. Nevertheless, among those with at least one unplanned out-patient visit, the number of visits was higher in the general population sample. Persons with ID and **dementia** are less likely to receive planned health care than persons with dementia in the general population. They have, however, higher levels of unplanned health care utilization. This may be an indication that the current support system is not sufficient to meet the challenges of increased longevity among persons with ID.

Aylward, E., Burt, D., Thorpe, L., Lai. & Dalton, A.J.
Diagnosis of dementia in individuals with intellectual disability: report of the task force for development of criteria for diagnosis of dementia in individuals with mental retardation
Journal of Intellectual Disability Research, 1997, 41, 152-164
Abstract: The foremost impediment to progress in the understanding and treatment of dementia in adults with intellectual disability is the lack of standardized criteria and diagnostic procedures. Standardized criteria for the diagnosis of dementia in individuals with intellectual disability are proposed, and their application is discussed. In addition, procedures for determining whether or not criteria are met in individual cases are outlined. It is the intention of the authors, who were participants of an International Colloquium on Alzheimer Disease and Mental Retardation, that these criteria be appropriate for use by both clinicians and researchers. Their use will improve communication among clinicians and researchers, and will allow researchers to test hypotheses concerning discrepancies in findings among research groups (e.g. dementia prevalence ranges and age of onset). [This report is available also on www.aamr.org at the following URL: http://161.58.153.187/Bookstore/Downloadables/index.shtml]

Ball, S.L., Holland, A.J., Hon, J., Huppert, F.A., Treppner, P., & Watson, P.C.
Personality and behaviour changes mark the early stages of Alzheimer's disease in adults with Down's syndrome: findings from a prospective population-based study.
International Journal of Geriatric Psychiatry, 2006, 21(7), 661-673
Abstract: Research based on retrospective reports by carers suggests that the presentation of dementia in people with Down syndrome may differ from that typical of Alzheimer's disease (AD) in the general population, with the earliest changes tending to be in personality or behavior rather than in memory. This is the first long-term prospective study to test the hypothesis that such changes, which are more typical of dementia of frontal type (DFT) in the general population, mark the preclinical stage of AD in DS. A previously identified population sample of older people with DS, first assessed in 1994 and followed-up 18 months later, were reassessed after a further 5 years. This study focuses on the 55 individuals who took part in the second follow-up. Dementia diagnosis was made using the modified CAMDEX informant interview and neuropsychological assessment was undertaken using the CAMCOG. Progression in clinical presentation was examined and degree of cognitive decline over time (on the CAMCOG and derived measures of executive function (EF) and memory) was compared across groups based on diagnosis and age: AD, DFT, personality/behavior changes insufficient for a diagnosis of DFT (PBC), no diagnosis <50 years and no diagnosis 50 + years. Progression was observed from early changes in personality and behavior to an increase in characteristics associated with frontal lobe dysfunction and/or a deterioration in memory, prior to the development of full AD. Individuals who met criteria for DFT were significantly more likely to progress to a diagnosis of AD over the following 5 years than those who did not and those with PBC were significantly more likely to progress to a more severe diagnosis (DFT or AD) than those without. In the 5 years prior to diagnosis, participants with PBC and DFT had shown a degree of global cognitive decline intermediate between those with no dementia and those with AD. Both these groups had shown a significant decline in EF but not in memory, while the AD group had shown significant decline on both measures, with a significantly greater degree of decline in memory. Older participants without informant reported changes showed a more generalized pattern of decline. These findings confirm that the early presentation of AD in DS is characterized by prominent personality and behavior changes, associated with executive dysfunction, providing support for the notion that the functions of the frontal lobes may be compromised early in the course of the disease in this population. This has important implications for the diagnosis, treatment and management of dementia in people with DS.

Ball, S.L., Holland, A.J., Huppert, F.A. Treppner, P., Watson, P.C., & Hon, J.
The modified CAMDEX informant interview is a valid and reliable tool for use in the diagnosis of dementia in adults with Down's syndrome
Journal of Intellectual Disability Research, 2004 Sep;48(Pt 6):611-20. doi:10.1111/j.1365-2788.2004.00630.x.
Abstract: Dementia because of Alzheimer's disease (AD) commonly affects older adults with Down's syndrome (DS). Methods are needed, with established concurrent and predictive validity, to facilitate the diagnostic assessment of dementia, when it is complicated by pre-existing intellectual disabilities (ID). We report on the reliability and validity of a modified version of the Cambridge Examination for Mental Disorders of the Elderly (CAMDEX) informant interview, for use when assessing people with DS suspected as having dementia. As part of a previous epidemiological study of older people with DS, the CAMDEX informant interview was used to determine the prevalence of dementia. The 74 people with DS included at that time (Time 1) had also completed the Cambridge

Cognitive Examination (CAMCOG), the neuropsychological assessment from the CAMDEX schedule. Fifty-six were assessed again 6 years later (Time 2). Based on the CAMDEX informant interview, nine of the 74 at Time 1, and 11 of the 56 at Time 2, were found to meet clinical criteria for AD. Forty-one scored above floor on the CAMCOG at Time 1 and were included in the analysis of cognitive decline. Concurrent validity was established by comparing diagnosis at Time 2 with independent evidence of objective decline on cognitive tasks since Time 1. Predictive validity was established by examining how accurately diagnosis at Time 1 predicted both cognitive decline and future diagnosis. Inter-rater reliability was determined by comparing the level of agreement between two raters. CAMDEX-based diagnosis of AD was shown to be consistent with objectively observed cognitive decline (good concurrent validity) and to be a good predictor of future diagnosis. Although numbers are small, some support is also provided for the accuracy with which diagnosis predicts cognitive decline. Inter-rater reliability was good with Kappa > 0.8 for 91% of items and > 0.6 for all items. The use of the modified CAMDEX informant interview enables the structured collection of diagnostic information, so that a valid and a reliable diagnosis of dementia can be made in those with pre-existing ID, using established diagnostic criteria.

Ball, S.L., Holland, A.J., Treppner, P., Watson, P.C., & Huppert, F.A.
Executive dysfunction and its association with personality and behaviour changes in the development of Alzheimer's disease in adults with Down syndrome and mild to moderate learning disabilities.
British Journal of Clinical Psychology, 2008, 47(Pt 1), 1-29. doi: 10.1348/014466507X230967.
Abstract: Recent research suggests that preclinical Alzheimer's disease (AD) in people with Down syndrome (DS) is characterized by changes in personality/behavior and executive dysfunction that are more prominent than deterioration in episodic memory. This study examines the relationship between executive dysfunction and the clinical and preclinical features of AD in DS. To determine the specificity of this relationship, performance on executive function (EF) measures is contrasted with performance on memory measures. One hundred and three people with DS (mean age 49 years, range 36-72) with mild to moderate learning disabilities (LD) took part. Dementia diagnosis was based on the CAMDEX informant interview conducted with each participant's main carer. Reported changes in personality/behavior and memory were recorded. Participants completed six EF and six memory measures (two of which also had a strong executive component) and the BPVS (as a measure of general intellectual ability). First, performance was compared between those with and without established dementia of Alzheimer's type (DAT), controlling for age and LD severity using ANCOVA. Next, the degree to which informant-reported changes predicted cognitive test performance was examined within the non-DAT group using multiple regression analyses. The DAT group (N=25) showed a consistent pattern of impaired performance relative to the non-DAT group (N=78), across all measures. Within the non-DAT group, number of informant-reported personality/behavior changes was a significant predictor of performance on two EF and two 'executive memory' tests (but not on episodic memory tests). Informant-reported memory changes, however, were associated with impaired performance on a delayed recall task only. These findings provide further evidence for a specific impairment in frontal-lobe functioning in the preclinical stages of AD in DS.

Ballard C, Mobley W, Hardy J, Williams G, Corbett A.
Dementia in Down's syndrome.
Lancet Neurology, 2016 May;15(6):622-36. doi: 10.1016/S1474-4422(16)00063-6. Epub 2016 Apr 11.
Abstract: Down syndrome is the most common genetic cause of learning difficulties, and individuals with this condition represent the largest group of people with dementia under the age of 50 years. Genetic drivers result in a high frequency of Alzheimer's pathology in these individuals, evident from neuroimaging, biomarker, and neuropathological findings, and a high incidence of cognitive decline and dementia. However, cognitive assessment is challenging, and diagnostic methods have not been fully validated for use in these patients; hence, early diagnosis remains difficult. Evidence regarding the benefits of cholinesterase inhibitors and other therapeutic options to treat or delay progressive cognitive decline or dementia is very scarce. Despite close similarities with late-onset Alzheimer's disease, individuals with Down syndrome respond differently to treatment, and a targeted approach to drug development is thus necessary. Genetic and preclinical studies offer opportunities for treatment development, and potential therapies have been identified using these approaches.

Bauer, A.M., & Shea, T.M.
Alzheimer's disease and Down syndrome: A review and implications for adult services
Education and Training of the Mentally Retarded, 1986, 21, 144-150.
https://eric.ed.gov/?id=EJ341367
Abstract: In this article, the diagnosis of Alzheimer's disease and its progressive behavioral impact on persons with Down syndrome is discussed. Several implications and suggestions for care and service provision for adults with Down syndrome are presented, including that Alzheimer's disease in an adult with Down syndrome has an impact on the carer, adjusting communication strategies to correspond to the stage of dementia, aiding families to seek assistance from social agencies, stressing the remaining abilities and skills, aiding families and carers to develop realistic methods of providing care, and adapting the persons care and environment to help them cope with losses stemming from dementia. The authors also suggest proactive strategies for anticipating decline among adults with Down syndrome associated with dementia.

Baumer NT, Becker ML, Capone GT, Egan K, Fortea J, Handen BL, Head E, Hendrix JE, Litovsky RY, Strydom A, Tapia IE, Rafii MS.
Conducting clinical trials in persons with Down syndrome: summary from the NIH INCLUDE Down syndrome clinical trials readiness working group.
J Neurodev Disord. 2022 Mar 23;14(1):22. doi: 10.1186/s11689-022-09435-z.
Abstract: The recent National Institute of Health (NIH) INCLUDE (INvestigation of Co-occurring conditions across the Lifespan to Understand Down syndromE) initiative has bolstered capacity for the current increase in clinical trials involving individuals with Down syndrome (DS). This new NIH funding mechanism offers new opportunities to expand and develop novel approaches in engaging and effectively enrolling a broader representation of clinical trials participants addressing current medical issues faced by individuals with DS. To address this opportunity, the NIH assembled leading clinicians, scientists, and representatives of advocacy groups to review existing methods and to identify those areas where new approaches are needed to engage and prepare DS populations for participation in clinical trial research. This paper summarizes the results of the Clinical Trial Readiness Working Group that was part of the INCLUDE Project Workshop: Planning a Virtual Down Syndrome Cohort Across the Lifespan Workshop held virtually September 23 and 24, 2019.

Bayen, E., Possin, K.L., Chen, Y., Ckeret de Langavant, L., & Yaffe, K.
Prevalence of aging, dementia, and multimorbidity in older adults with Down syndrome
JAMA Neurology, 2018, Nov 1, 75(11), 1399-1406.
doi:10.1001/jamaneurol.2018.2210.
Abstract: As the life expectancy of people with Down syndrome (DS) has markedly increased over the past decades, older adults with DS may be experiencing a higher incidence of aging conditions. In addition to longevity, the amyloid precursor protein gene located on chromosome 21 places individuals with DS at a high risk for developing Alzheimer disease. Yet, few studies have determined prevalence of dementia and comorbidities among older people with DS. To determine the prevalence of dementia and aging-related comorbidities in older adult individuals with DS. Cross-sectional analysis of 2015 California Medicare claims data. We examined 1 year of cross-sectional Medicare claims data that included 100% of Californian Medicare beneficiaries enrolled in both Medicare Part A and B in 2015. Of these 3 001 977 Califomian Medicare beneficiaries 45 years or older, 878 individuals were identified as having a diagnosis of DS. Data were analyzed between April 2017 and February 2018. The frequency of DS dementia was assessed across different age categories. The number and frequency of 27 comorbidities were compared among individuals with DS with and without dementia and by age and sex groups. A total of 353 DS individuals (40%) were identified as having dementia diagnoses (mean, 58.7 years; 173 women [49%]) and 525 without dementia diagnoses (mean, 55.9 years; 250 women [48%]). The frequency of DS dementia among those 65 years or older rose to 49%. The mean number of comorbidities per individual increased with age in general. Comorbid conditions were more numerous among those with dementia compared with those with DS without dementia (mean, 3.4 vs 2.5, respectively), especially among those younger than 65 years. In particular, 4 treatable conditions, hypothyroidism, epilepsy, anemia, and weight loss, were much more frequent in DS dementia. Older Medicare beneficiaries in California with DS, especially those with dementia, have a high level of multimorbidity including several treatable conditions. While DS follow-up

has long been confined to the pediatric sphere, we found that longevity in individuals with DS will necessitate complex adult and geriatric care. More evidenced-based and standardized follow-up could support better long-term comorbidity management and dementia care among aging adults with DS.

Bejanin, A., Iulita, M.F., Vilaplana, E., Carmona-Iragui, M., Benejam, B,, Videlam, L., Barroeta, I, et al.
Association of apolipoprotein E ε4 allele with clinical and multimodal biomarker changes of Alzheimer disease in adults with Down syndrome.
JAMA Neurology, doi:10.1001/jamaneurol.2021.1893. Published online July 6, 2021.
Abstract: Alzheimer disease (AD) is the leading cause of death in individuals with Down syndrome (DS). Previous studies have suggested that the APOE ε4 allele plays a role in the risk and age at onset of dementia in DS; however, data on in vivo biomarkers remain scarce. To investigate the association of the APOE ε4 allele with clinical and multimodal biomarkers of AD in adults with DS.
his dual-center cohort study recruited adults with DS in Barcelona, Spain, and in Cambridge, UK, between June 1, 2009, and February 28, 2020. Included individuals had been genotyped for APOE and had at least 1 clinical or AD biomarker measurement; 2 individuals were excluded because of the absence of trisomy 21. Participants were either APOE ε4 allele carriers or noncarriers. Participants underwent a neurological and neuropsychological assessment. A subset of participants had biomarker measurements: Aß1-42, Aß1-40, phosphorylated tau 181 (pTau181) and neurofilament light chain (NfL) in cerebrospinal fluid (CSF), pTau181, and NfL in plasma; amyloid positron emission tomography (PET); fluorine 18-labeled-fluorodeoxyglucose PET; and/or magnetic resonance imaging. Age at symptom onset was compared between APOE ε4 allele carriers and noncarriers, and within-group local regression models were used to compare the association of biomarkers with age. Voxelwise analyses were performed to assess topographical differences in gray matter metabolism and volume. Of the 464 adults with DS included in the study, 97 (20.9%) were APOE ε4 allele carriers and 367 (79.1%) were noncarriers. No differences between the 2 groups were found by age (median [interquartile range], 45.9 [36.4-50.2] years vs 43.7 [34.9-50.2] years; P = .56) or sex (51 male carriers [52.6%] vs 199 male noncarriers [54.2%]). APOE ε4 allele carriers compared with noncarriers presented with AD symptoms at a younger age (mean [SD] age, 50.7 [4.4] years vs 52.7 [5.8] years; P = .02) and showed earlier cognitive decline. Locally estimated scatterplot smoothing curves further showed between-group differences in biomarker trajectories with age as reflected by nonoverlapping CIs. Specifically, carriers showed lower levels of the CSF Aß1-42 to Aß1-40 ratio until age 40 years, earlier increases in amyloid PET and plasma pTau181, and earlier loss of cortical metabolism and hippocampal volume. No differences were found in NfL biomarkers or CSF total tau and pTau181. Voxelwise analyses showed lower metabolism in subcortical and parieto-occipital structures and lower medial temporal volume in APOE ε4 allele carriers. In this study, the APOE ε4 allele was associated with earlier clinical and biomarker changes of AD in DS. These results provide insights into the mechanisms by which APOE increases the risk of AD, emphasizing the importance of APOE genotype for future clinical trials in DS.

Benejam, B., Videla, L., Vilaplana, E., Barroeta, I., Carmona-Iragui ,M,. Altuna ,M., Valldeneu, S., Fernandez, S., Giménez, S., Iulita, F., Garzón, D., Bejanin, A., Bartrés-Faz, D., Videla, S., Alcolea, D., Blesa, R., Lleó, A., & Fortea, J.
Diagnosis of prodromal and Alzheimer's disease dementia in adults with Down syndrome using neuropsychological tests.
Alzheimers Dementia (Amst), 2020 Jun 28,12(1), e12047. doi: 10.1002/dad2.12047.
Abstract: We aimed to define prodromal Alzheimer's disease (AD) and AD dementia using normative neuropsychological data in a large population-based cohort of adults with Down syndrome (DS). We employed a cross-sectional study. DS participants were classified into asymptomatic, prodromal AD and AD dementia, based on neurologist's judgment blinded to neuropsychological data (Cambridge Cognitive Examination for Older Adults with Down's syndrome [CAMCOG-DS] and modified Cued Recall Test [mCRT]). We compared the cutoffs derived from the normative data in young adults with DS to those from receiver-operating characteristic curve (ROC) analysis. Diagnostic performance of the CAMCOG-DS and modified Cued Recall Test (mCRT) in subjects with mild and moderate levels of intellectual disability (ID) was high, both for diagnosing prodromal AD and AD dementia (area under the curve [AUC] 0.73-0.83 and 0.90-1, respectively). The cutoffs derived from the normative data were similar to those derived from the ROC analyses. Diagnosing prodromal AD and AD dementia in DS with mild and moderate ID using population norms for neuropsychological tests is possible with high diagnostic accuracy.

Bevens, S., Dawes, S., Kenshole, A., & Gaussen, K.
Staff views of a music therapy group for people with intellectual disabilities and dementia: A pilot study
Advances in Mental Health and Intellectual Disabilities, 2015, 9(1), 40-48. https://doi.org/10.1108/AMHID-04-2014-0005
Abstract: Despite the longstanding use of music therapy with people with intellectual disabilities and the growing evidence base for using music therapy as a tool to aid behavioural and psychological symptoms of dementia in the general population, there is little work published which details the use of music therapy groups for people with intellectual disabilities who have a diagnosis of dementia. The purpose of this paper is to report a qualitative evaluation of staff views of a music therapy group for people with intellectual disabilities and dementia. Carers of service users attending the group were interviewed either individually or through a focus group in order to ascertain their views about the music therapy group. The interview transcripts were then analysed using thematic analysis. Two core themes and eight sub themes emerged from the data. These themes show that the group was felt to be pleasurable and enjoyable for the service users and that some tangible benefits of attending the group were observed by staff members. Notwithstanding the positive feedback, the results also suggested that more work is needed to inform carers of the goals and purpose of such groups. Further psycho-education for carers is suggested as a strategy to support future groups to run successfully. There is little published research into the use of music therapy for people with intellectual disabilities who also have dementia. The current paper provides a starting point for future work in the area and further recommendations for future practice and research are considered.

Bigby, C., & Beadle-Brown, J.
Culture in better group homes for people with intellectual disability at severe levels
Intellectual and Developmental Disabilities, 2016 Oct, 54(5), 316-331. doi: 10.1352/1934-9556-54.5.316.
Abstract: Building on cultural dimensions of underperforming group homes this study analyzes culture in better performing services. In depth qualitative case studies were conducted in 3 better group homes using participant observation and interviews. The culture in these homes, reflected in patterns of staff practice and talk, as well as artefacts differed from that found in underperforming services. Formal power holders were undisputed leaders, their values aligned with those of other staff and the organization, responsibility for practice quality was shared enabling teamwork, staff perceived their purpose as "making the life each person wants it to be," working practices were person centered, and new ideas and outsiders were embraced. The culture was characterized as coherent, respectful, "enabling" for residents, and "motivating" for staff. Though it is unclear whether good group homes have a similar culture to better ones the insights from this study provide knowledge to guide service development and evaluation.

Bigby, C., Bowers, B., & Webber, R.
Planning and decision making about the future care of older group home residents and transition to residentialaged care
Journal of Intellectual Disability Research, 2011 Aug, 55(8), 777-789. doi: 10.1111/j.1365-2788.2010.01297.x
Abstract: Planning for future care after the death of parental caregivers and adapting disability support systems to achieve the best possible quality of life for people with intellectual disability as they age have been important issues for more than two decades.This study examined perceptions held by family members, group home staff, and organizational managers about the future of older residents and the decisions made that a move to residential aged care was necessary. Grounded Dimensional Analysis was used to guide data collection and analysis by an interdisciplinary research team. Three sets of interviews over a period of 18 months were conducted with a family member, house supervisor and the program manager for each of seventeen older group home residents in Victoria, Australia. For the eight people for whom it was decided a move was necessary and the six who eventually moved focussed questions were asked about the decision-making process. While plans for lifelong accommodation in a group home proved unfounded, key person succession plans were effective. However, decisions amove to a residential aged care facility was necessary were made in haste and seen as a fait accompli to involved family members. Although family members take seriously their mandate to oversee well-being of their older

relative, they have little knowledge about their rights or avenues to safeguard untimely or inappropriate decisions being made by professionals.

Bigby, C., Knox, M., Beadle-Brown, J., Clement, T., & Mansell, J.
Uncovering dimensions of culture in underperforming group homes for people with severe intellectual disability
Intellectual and Developmenal Disabilities, 2012 Dec, 50(6), 452-467. doi: 10.1352/1934-9556-50.06.452.
Abstract: Culture recurs as an important but under-investigated variable associated with resident outcomes in supported accommodation for people with intellectual disability. This study aimed to conceptualize the potential dimensions of culture in all group homes and describe the culture in underperforming group homes. A secondary analysis, using an inductive interpretative approach, was undertaken of a large qualitative data set from a study that had used ethnographic and action research methods to explore the quality of life outcomes for residents in 5 small group homes. Five categories were developed: misalignment of power-holder values with organizations espoused values, otherness, doing for not with, staff centered, and resistance. Differences from institutional culture are discussed, and the potential of the findings as a starting point to consider culture in high performing group homes and develop a quantitative measure of culture.

Bishop, K.M., Hogan, M., Janicki, M.P., Keller, S.M., Lucchino, R., Mughal, D.T., Perkins, E.A., Singh, B.K., Service, K., Wolfson, S. & Health Planning Work Group of the National Task Group on Intellectual Disabilities and Dementia Practices.
Guidelines for dementia-related health advocacy for adults with intellectual disability and dementia: national task group on intellectual disabilities and dementia practices.
Intellectual and Developmental Disabilities, 2015 Feb, 53(1), 22-29. doi: 10.1352/1934-9556-53.1.2
Abstract: Increasing numbers of adults with intellectual disabilities (ID) are living into old age. Though this indicates the positive effects of improved health care and quality of life, the end result is that more adults with ID are and will be experiencing age-related health problems and also exhibiting symptoms of cognitive impairment and decline, some attributable to dementia. Early symptoms of dementia can be subtle and in adults with ID are often masked by their lifelong cognitive impairment, combined with the benign effects of aging. A challenge for caregivers is to recognize and communicate symptoms, as well as find appropriate practitioners familiar with the medical issues presented by aging adults with lifelong disabilities. Noting changes in behavior and function and raising suspicions with a healthcare practitioner, during routine or ad hoc visits, can help focus the examination and potentially validate that the decline is the result of the onset or progression of dementia. It can also help in ruling out reversible conditions that may have similar presentation of symptoms typical for Alzheimer's disease and related dementias. To enable caregivers, whether family members or staff, to prepare for and advocate during health visits, the National Task Group on Intellectual Disabilities and Dementia Practices has developed guidelines and recommendations for dementia-related health advocacy preparation and assistance that can be undertaken by provider and advocacy organizations.

Bittles, A.H., & Glasson, E.J.
Clinical, social, and ethical implications of changing life expectancy in Down syndrome
Developmental Medicine & Child Neurology, 2004, 46, 282-286.
Abstract: Increased life expectancy generates greater ethical and legal dilemmas in the treatment of people with Down syndrome. Assumptions that younger cohorts of people with DS will experience healthier lives when compared to previous generations may not be realized as specific health issues associated with DS are genetically encoded and thus contemporary generations may face the same adverse health issues. With respect to dementia, authors note that by age 60 years, dementia involving memory loss, cognitive decline, and changes in adaptive behavior may be present in at least 56% of adults with DS and that some the neuropathological features of Alzheimer disease may be evident as early as age 40.

Blesa, R., Trias, C., Fortea J., & Videla. S.
Alzheimer's disease in adults with Down syndrome: a challenge.
T21 Res Sci Soc Bull. 2015; 2: 4
Abstract: None provided.

Bowers, B., Webber, R., & Bigby, C.
Aging and health related changes of people with intellectual disabilities living in group homes in Australia.
Journal of Policy and Practice in Intellectual Disabilities, 2009, 6(2), 98. [SINGLE PAGE]
Abstract: Group homes for people with ID are based on social models, emphasizing inclusion, engagement in community, and quality of life. As age related changes occur, group home staff members are faced with decisions about how to respond, how to support people experiencing health problems, and whether or for how long people can remain in the group homes. This study explored how group home staff members respond to aging and age related health conditions in group home residents and to identify factors that put people at risk of premature or inappropriate relocation. Using a longitudinal design in order to observe, over time, the onset of health problems, the initial responses of housing staff to health, the development of health conditions, the consequences of their initial responses, and the outcomes for both staff and residents were considered. In-depth interviews were conducted—at three 6-month intervals with 18 clusters of the housing manager, family member, the person with the disability, and in some cases, healthcare providers. A total of 91 interviews were completed, transcribed, and analyzed and in keeping with the theory-generating approach, early interviews were open and exploratory, evolving over time to facilitate comparative analysis across groups, strategies, conditions, and care issues. Staff and family members agreed that aging and the development of associated health conditions was increasingly becoming an issue for them. Significantly, there was wide variation among housing staff in terms of philosophy of care, with some believing that people should be supported to remain at the group homes for as long as possible. This, however, required the acquisition of new resources, a range of organizational changes to support staff and residents, changes to staffing patterns and levels, and a change in recruiting as a strategy to alter skill mix of house workers. Authors concluded that problems identified by most housing staff included: (a) inability of residents to retire despite age and health status; (b) risk of premature moves to aged care; and © disruption to general house activities and routines of other residents. Staff members' experienced altered work routines, concerns about the safety of residents and themselves, and frequent turnover. Availability of resources, such as equipment and home modifications, flexibility of staffing to accommodate changing resident needs, and philosophy of care all had a significant impact on residents' ability to "stay home."

Bowey, L. & McGlaughlin, A.
Adults with a learning disability living with elderly carers talk about planning for the future: Aspirations and concerns.
British Journal of Social Work, 2005, 35(8), 1377-1392. DOI: https://doi.org/10.1093/bjsw/bch241
Abstract: The majority of adults with an intellectual disability live with family carers, many of whom are ageing and have support needs of their own. Planning for the future thus becomes the key to preventing a crisis situation when family care is no longer viable because of death or ill health. Existing knowledge and practice are largely based upon the perspective of professionals and carers. This study explored the views, aspirations and concerns of adults with an intellectual disability, about living at home and planning for the future. Findings show that participants were very aware of the need for alternative housing or support in the future and had clear preferences about their future options. However, they also showed extensive concern for their family carers and this often impacted on their willingness to plan for the future or to move to alternative housing. Their demonstrable awareness of the inevitable death or ill health of family carers, and willingness to engage with the implications, emphasize the importance of involving adults with intellectual disability in planning for their future, as well as providing them with bereavement support.

Bratek, A., Krysta, K., & Kucia, K.
Psychiatric comorbidity in older adults with intellectual disability
Psychiatria Danubina, 2017, 29(Suppl 3), 590-593. PMID: 28953835
Abstact: The population of older adults with intellectual disability (ID) is large and growing due to a significant increase of life expectancy caused by improvements in health and social care. Multimorbidity is highly prevalent in this population and co-morbid psychiatric disorders are especially frequent. This article provides a review of the prevalence and consequences of psychiatric comorbidity in the population of older adults with ID. We therefore performed a literature search of studies relevant to adults with ID, published since January 2006, using the following keywords: intellectual disability and comorbidity, intellectual disability and mental disorders, intellectual disability and polypharmacy. Psychiatric

comorbidity is frequent among patients with ID and correlates with older age. Mental disorders are present in up to 40% of older adults with ID and the most prevalent are challenging behaviour, depression, anxiety and dementia. Patients with ID and at least one co-morbid mental disorder are at a high risk of polypharmacy. Importantly, psychiatric comorbidity was found to significantly increase service use and costs of care.
Further investigation of the population of older adults with ID is needed, with special attention to development of clear treatment guidelines in order to effectively manage co-morbid mental illnesses and physical health problems.

Brawley, E.C.
Designing for Alzheimer's disease - Strategies for creating better care environments.
313 pp.
New York: Wiley (1997)
Abstract: 20 chapter general text on adapting homes and living environments for persons with dementia; applicable to home and other residential situations for adults with intellectual disabilities and dementia. Chapter sections include Aging and Alzheimer's disease, Sensory environment (light and aging vision, lighting, impact of color, patterns and texture, acoustical changes, and wayfinding guidelines), Special care settings (creating a home feeling, designing spaces, therapeutic gardens and outdoor spaces), Implementing effective interior design (furniture and fabrics, floor-covering, wall and ceiling finishes, windows and window treatments), and the Design process. Contains a directory of resources and a glossary of terms.

Brodaty, H., Seeher,K., & Gibson, L.
Dementia time to death: A systematic literature review on survival time and years of life lost in people with dementia.
International Psychogeriatrics, 2012, 24(7),1034-1045. Published online on 13 February 2012. doi: 10.1017/S1041610211002924.
Abstract: Life expectancy with dementia directly influences rates of prevalence and service needs and is a common question posed by families and patients. As well as years of survival, it is useful to consider years of life lost after a diagnosis of dementia. Authors systematically reviewed the literature on mortality and survival with dementia which were compared to estimated life expectancies in the general population. Both were then compared by age (under 65 years vs. 65+ years), gender, dementia type, severity, and two epochs (prior to and after introduction of cholinesterase inhibitors in 1997). Survival after a diagnosis of dementia varies considerably and depends on numerous factors and their complex interaction. Relative loss of life expectancy decreases with age at diagnosis across varying sex, dementia subtypes (except for frontotemporal dementia and dementia with Lewy bodies), and severity stages. Numerous study deficiencies precluded a meta-analysis of survival in dementia. Authors concluded that estimates of years of life lost through dementia may be helpful for patients and their families.

Burt, D.B., & Aylward, E.
Assessment methods of diagnosis of dementia
In M.P. Janicki & A.J. Dalton (Eds.), Dementia, Aging, and Intellectual Disabilities.
pp. 141-156
Philadelphia: Brunner-Mazel (1999)
Abstract: Standardized diagnostic criteria and procedures are proposed to further progress in the understanding and treatment of dementia in adults with intellectual disabilities. This book chapter is a revised summary of previous reports prepared by participants of an international working group, which was conducted under the auspices of the International Association on Intellectual Disability and the American Association on Mental Retardation. Similarities in diagnostic issues between adults with intellectual disability and those in the general population are discussed, followed by a summary of issues unique to adults with intellectual disability. A brief overview of the application of ICD-10 diagnostic criteria to adults with intellectual disability is presented, including a description of procedures for determining whether criteria are met in individual cases. Finally, clinical and research recommendations are made.

Burt, D.B., Loveland, K.A., & Lewis, K.R.
Depression and the onset of dementia n adults with mental retardation.
American Journal of Mental Retardation, 1992, 96(5), 502-511.
Abstract: The relation between dementia and depression in 61 adults with Down syndrome or 43 adults with mental retardation due to other causes was examined. Age-matched participants, ranging in age from 20 to 60 years, received a neuropsychological battery to assess declines in functioning and caregiver report measures to assess adaptive behavior and depression. Eight adults with Down syndrome had both depression and declines in functioning. No adults with mental retardation due to other causes had declines. Greater severity of depression was related to lower MA, poorer memory, and lower adaptive functioning in adults with Down syndrome only. Results suggest that dementia and depression are associated in Down syndrome but not in mental retardation due to other causes.

Burt, D.B., Primeaux-Hart, S., Loveland, K.A., Cleveland, L.A., Lewis, K.R., Lesser, J., & Pearson, P.L.
Tests and medical conditions associated with dementia diagnosis
Journal of Policy and Practice in Intellectual Disabilities, 2005, 2(1), 47-56. https://doi.org/10.1111/j.1741-1130.2005.00007.x
Abstract: Diagnosis of dementia in adults with intellectual disabilities requires documentation of clinically significant declines in memory and other cognitive skills, as well as changes in everyday and emotional functioning. To improve diagnostic accuracy in adults with Down syndrome, the authors examined conditions often associated with dementia, as well as tests useful for documentation of decline. Specific aims were to identify psychiatric disorders or medical conditions that increased the odds of a dementia diagnosis; to evaluate the sensitivity and specificity of widely used dementia scales; and to determine which tests, used singly or in combination, most accurately supported the presence of dementia. Participants were 78 adults with Down syndrome. Two methods based on a large test battery and one method based on clinical judgment were used to diagnose dementia. It was found that combinations of tests lead to increased levels of diagnostic sensitivity compared with single tests. When taken in combination with other investigations, our results suggest that assessment for **psychiatric disorders, delayed memory decline, adaptive behavior decline, and the presence of seizures** would be useful for the diagnosis of dementia and that dementia scales would provide additional useful information. The authors conclude that combinations of tests and scales will be most useful for diagnosing dementia in adults with intellectual disabilities. The authors suggest that further research is needed to promote rapid progress, with studies that focus on common diagnostic methodology, identification of screening instruments, and amounts of decline indicative of dementia.

Burt, D.B., Primeaux-Hart, S., Loveland, K.A, Cleveland,L.A., Lewis, K.R., Lesser, J., & Pearson, P.L
Comparing dementia diagnostic methods used with people with intellectual disabilites
Journal of Policy and Practice in Intellectual Disabilities, 2005, 2(2), 94-115. https://doi.org/10.1111/j.1741-1130.2005.00022.x
Abstract: Accurate detection of dementia in adults with intellectual disabilities is important for clinical care, program planning, andclinical research. This paper reports on a study that examined two major diagnostic methods that varied in the following ways: (1)the extent to which they relied on clinical judgment; (2) the statistical method used to detect declines; and (3) the sensitivity to declines in functioning. Two methods based on testing were compared with one based on clinical judgment. Data were drawn from annual sequential assessments of 168 adults with intellectual disabilities (78 with Down syndrome and 90 with other etiologies).Agreement between testing and clinical judgment methods was 72-75% depending on testing method used. Clinical judgment produced a higher rate of dementia diagnosis for adults with Down syndrome compared with testing methods, suggesting a possible bias. The authors found that diagnostic criteria were useful both for identifying dementia and for describing its characteristics. Our results suggest that clinical judgment could result in a higher number of adults with Down syndrome diagnosed with dementia than methods based on test batteries. Common results across research studies indicate that combinations of sources of information(interviews/direct testing) would be most useful for dementia diagnosis. Future collaboration across research sites is needed to promote rapid progress in this important area, with emphasis on differential diagnosis.

Bush, A., & Beail, N.
Risk factors for dementia in people with Down syndrome: Issues in assessment and diagnosis
American Journal of Mental Retardation, 2004 Mar,109(2), 83-97. doi:10.1352/0895-8017(2004)109<83:RFFDIP>2.0.CO;2.

Abstract: It has been clearly established that there is an increased incidence of early onset dementia of the Alzheimer type (DAT) in people who have Down syndrome. There are variations in the age of onset of the clinical signs of DAT, which may be accounted for by different risk factors. In this review we examined the evidence that different biological and psychological factors may influence the risk for DAT. Limitations in design of early studies, the need for consistent diagnostic criteria for DAT in individuals with Down syndrome, and the lack of adequate psychometric tools to detect cognitive change are highlighted. Implications for research and clinical practice are considered in order to assess potential risk factors.

Cairns, D., Kerr, D., Chapman, A.
Difference realities: a training guide for people with Down's syndrome and Alzheimer's disease
pp. 54
University of Stirling (Dementia Services Development Centre), Stirling, Scotland FK9 4LA
A working guide for staff who are working with people with intellectual disabilities affected by Alzheimer's disease. Topical sections cover the definitions of dementia and deal with diagnostic suggestions, as well as dealing with communication, helping maintenance of skills, dealing with challenging behaviors, structuring activities, and overall management of dementia. Written in an easy style, this guide is a very useful addition to any materials given to staff to help them understand and related to people affected by dementia.

Cairns, V., Lamb, I., & Smith, E.
Reflections upon the development of a dementia screening service for individuals with Down's syndrome across the Hyndburn and Ribble Valley area.
British Journal of Learning Disabilities, 2011, 39(3), 198-208.
https://doi.org/10.1111/j.1468-3156.2010.00651.x
Abstract: The high prevalence of dementia in individuals with Down's syndrome has led intellectual disability services in the Hyndburn and Ribble Valley (HRV) area of England to develop a screening service to address this need. The authors offer reflections upon this process by its members after the first 12 months of operation. A multidisciplinary team, comprising professionals from intellectual disability psychology, intellectual disability speech and language therapy, intellectual disability community nursing and older adults psychiatry, has developed, and begun to implement, screening care pathways. The service conducts routine screening assessments, provides intervention for individuals where concerns arise and delivers training to carers. At the point of writing, 27 service users have received screening assessments and six have been identified as at moderate–high risk of developing dementia. Reflection and feedback has highlighted issues for consideration throughout the service development process, and an evaluation of the training provided by the service has found this to be effective in increasing carers understanding about dementia and intellectual disabilities.

Carfi, A., Antociccio, M., Brandi, V., Cipriani, C., Fiore, F., Mascia, D., Vetrano, D.L., Onder, G..
Down syndrome in adulthood: A disease for geriatricians
European Geriatric Medicine, 2014, 5(Supp 1), 549.
Abstract: Authors evaluated 89 adults with Down syndrome at a clinic in Rome, Italy, using a range of physiological and neurological methods, including nutritional and sensory assessments. The S's mean age was 42 years (range 18 to 72); 51% were females. Authors found behavioral disorders (53%), mood disorders (43%), seizures (22%), osteoporosis (40%), hypothyroidism (53%), diastolic dysfunction (80%), OSAS (90%), and hearing impairment (82%). Authors noted severe cognitive impairments in 67%, BMI greater than 25 in 66%, and low scores on physical performance measures (50%). Authors conclude that the pattern of diseases and conditions noted resemble those of other older adults and recommend that a mandatory geriatric evaluation be undertaken in older adults with Down.

Carling-Jenkins, R., Torr, J., Iacono, T., & Bigby, C.
Experiences of supporting people with Down syndrome and Alzheimer's disease in aged care and family environments.
Journal of Intellectual and Developmental Disability, 2012, 37(1), 54-60. doi: 10.3109/13668250.2011.645473. Epub 2012 Jan 3
Abstract: Australian research addressing the experiences of families of adults with Down syndrome and Alzheimer's disease in seeking diagnosis and gaining support is limited. The aim of this study was to gain a greater understanding of these processes by exploring the experiences of families and carers in supporting people with Down syndrome and Alzheimer's disease who had lived most or all of their lives with family. Three detailed case studies were created from multiple data sources, and then analyzed thematically. Families of adults with Down syndrome experienced stress and confusion as they negotiated a service system poorly equipped to meet their needs and professionals more focused on longstanding disability than the recent diagnosis of Alzheimer's disease. Such overshadowing led to mismanagement by services. Authors conclude that this research advances understandings of the support needs of people with Down syndrome and Alzheimer's disease and their families and exposes gaps in the service system.

Carmeli E, Ariav C, Bar-Yossef T, Levy R, Imam B.
Movement skills of younger versus older adults with and without Down syndrome.
Res Dev Disabil. 2012 Jan-Feb;33(1):165-71. doi: 10.1016/j.ridd.2011.09.008. Epub 2011 Oct 4. Erratum in: Res Dev Disabil. 2012 Mar-Apr;33(2):781. PMID: 22093661.
Abstract: Adults with Down syndrome (DS) are often physically inactive, which may accelerate the onset of disease and aging symptoms. Eight older persons with DS (aged 54-61), and 10 younger persons with DS (aged 26-35) living in a residential care center were examined. Eighteen age- and gender-matched individuals without DS served as control groups. Sensory-motor tasks and Posture Scale Analyzer (PSA) were used to examine coordination and standing stability. The isokinetic muscle strength test was used for muscle strength investigation. The functional performance, coordination, and leg muscle strength of the older adults with DS were more impaired than both the younger DS and the control groups. The older DS group showed lower sway rate and more symmetrical weight-bearing distribution during quiet standing than both the younger DS and the control groups. Our observations may have significant implications for understanding movement dysfunction in older adults with DS.

Carmeli, E., Ariav, C., Bar-Yossef, T., & Levy, R.
Movement skills in persons with Down syndrome decrease with aging
International Journal on Disability and Human Development, 2010, 9(1), 29–34.
https://www.degruyter.com/document/doi/10.1515/IJDHD.2010.005/html
Abstract: Persons with Down syndrome (DS) are comparatively physically inactive, which could accelerate the onset of disease, resulting in symptoms associated with aging that are detrimental to health. The aim was to evaluate movement abilities across the life span in persons with DS. Eleven persons with DS (>50 years, mean age 58 years), and 10 younger persons with DS (<49 years, mean age 28 years) who resided in a residential living center were included in the study. Age- and gender-matched people without DS (n=22) served as control group. Five sensory-motor tasks that involved the integration of hand movements with visual information were used, as well as the posture scale analyzer system to examine postural stability. Results showed that the older persons with DS had more medical problems than the young persons with and without DS. The hand coordination and postural stability of the older adults with DS were more impaired in comparison with the young group and both control groups. It is postulated that their poor motor function and slower responses might be explained by a less active lifestyle, that could accelerate the onset of disease, resulting in symptoms associated with aging that are detrimental to health. Our observations could have significant implications for understanding the mechanisms underlying movement dysfunction in older adults with DS and might offer new approaches for possible prevention.

Carr, J.
Stability and change in cognitive ability over the life span: a comparison of populations with and without Down's syndrome
Journal of Intellectual Disability Research, 2005 Dec;49(Pt 12):915-28. doi: 10.1111/j.1365-2788.2005.00735.x.
Abstract: Longitudinal studies show that in the general population IQ declines with age: early and rapidly in the case of performance IQ, later and more slowly in the case of verbal IQ. These populations have not apparently included people with intellectual disabilities (ID). A literature search identified 11 studies, some cross-sectional and others longitudinal, which provided data on a variety of verbal and performance tests, over periods from 3 to 19 years, on older people with ID. Following statistical advice the results from the different tests were converted into the equivalent of, for verbal scores, British Picture Vocabulary Scale, and for performance scores, Leiter International Performance Scale, raw scores. Percentage change between earlier and later scores was then calculated. With

one exception the studies considered tend to show verbal ability declining relatively more, and performance ability declining relatively less, than has been shown to occur in the general population. Potential confounding factors, such as population attrition, cohort effects, etc., are thought not to have affected these results. The pattern of change with age in verbal and performance ability appears different in people with ID from that seen in the general population. Some possible reasons for this difference are discussed.

Carr, J., & Collins, S.
Ageing and dementia in a longitudinal study of a cohort with Down syndrome.
Journal of Applied Research in Intellectual Disabilities, 2014, 27(6), 555-563.
doi: 10.1111/jar.12093. Epub 2014 Mar 29.
Abstract: A population sample of people with Down syndrome has been studied from infancy and has now been followed up again at age 47 years. Intelligence and language skills were tested and daily living skills assessed. Memory/cognitive deterioration was examined using two test instruments. Scores on verbal tests of intelligence changed little. Those on a non-verbal test, on self-help skills and on both memory tests showed some decline, even when the scores of those already suffering from dementia were discounted. At age 47, scores on most tests of even the majority of the cohort (i.e. those not definitely diagnosed with dementia) showed some decline. While this includes the scores of people who may subsequently develop dementia, it may also reflect the normal ageing process in this population.

Castro, P., Zaman, S., & Holland, A.
Alzheimer's disease in people with Down's syndrome: the prospects for and the challenges of developing preventative treatments.
Journal of Neurology, 2017 Apr, 264(4), 804-813.
doi:10.1007/s00415-016-8308-8. Epub 2016 Oct 24.
Abstract: People with Down's syndrome (DS) are at high risk for developing Alzheimer's disease (AD) at a relatively young age. This increased risk is not observed in people with intellectual disabilities for reasons other than DS and for this reason it is unlikely to be due to non-specific effects of having a neurodevelopmental disorder but, instead, a direct consequence of the genetics of DS (trisomy 21). Given the location of the amyloid precursor protein (APP) gene on chromosome 21, the amyloid cascade hypothesis is the dominant theory accounting for this risk, with other genetic and environmental factors modifying the age of onset and the course of the disease. Several potential therapies targeting the amyloid pathway and aiming to modify the course of AD are currently being investigated, which may also be useful for treating AD in DS. However, given that the neuropathology associated with AD starts many years before dementia manifests, any preventative treatment must start well before the onset of symptoms. To enable trials of such interventions, plasma, CSF, brain, and retinal biomarkers are being studied as proxy early diagnostic and outcome measures for AD. In this systematic review, we consider the prospects for the development of potential preventative treatments of AD in the DS population and their evaluation.

Centre for Developmental Disability Health Victoria
Dementia and Intellectual Disability – A guide to supporting people with intellectual disabilities through their journey with dementia: Online Learning for Disability Support Workers
http://www.cddh.monash.org/online-learning/
Abstract: These are on-line learning modules for disability staff supporting people who were at risk of developing, or had already been identified as having, dementia. There are four 16 minute nodules in the series addressing key questions you may have when supporting someone with dementia. They cover helpful information related to dementia and ID. Module 1: Understanding dementia and intellectual disability; Module 2: Taking action – The role of the support worker in assessment; Module 3: Supporting someone with intellectual disability and dementia; and Module 4: Supporting people through environment and activity. There are also a series self-taking test questions.

Chao, S.F., McCallion, P., & Nickle, T.
Factorial validity and consistency of the Maslach Burnout Inventory among staff working with persons with intellectual disability and dementia
*Journal of Intellectual Disability Research***,** 2011, 55(5), 529-536.
https://doi.org/10.1111/j.1365-2788.2011.01413.x
Abstract: Burnout has been considered important to understanding the well-being of workers in the intellectual disabilities (ID) field and the quality of services delivered to clients/consumers. However, little research has examined the psychometric properties and applicability to staff in ID services of one of the most widely used burnout measurements – the Human Services Survey version of the Maslach Burnout Inventory (MBI-HSS). Data were gathered using a mailed questionnaire comprising the MBI-HSS and demographic information. The sample consisted of 435 staff delivering direct care and working in out-of-home community placements for persons with ID in New York state. The factorial structure of the scale was examined using confirmatory and exploratory factor analysis. Internal consistency estimates of reliability of the MBI-HSS were determined using Cronbach's alpha. Confirmatory factor analysis supported the MBI-HSS as an acceptable measure to evaluate burnout in ID services staff. However, the reliability statistics obtained for the Depersonalization (DP) sub-scale was much lower than what has been reported in studies with other staff populations. An exploratory factor analysis suggested that a four-factor solution, dividing the DP sub-scale into two factors, provided a somewhat better fit for the sample. The use of the MBI-HHS as an instrument for measuring burnout among ID workers has its attraction but also its limitations. In particular, the DP sub-scale should be used with caution because there appear to be wording issues for staff in ID settings that may lead to inconsistent responses.

Chapman, M., Lacey, H., & Jervis, N.
Improving services for people with learning disabilities and dementia: Findings from a service evaluation exploring the perspectives of health and social care professionals.
British Journal of Learning Disabilities, 2017, 45(1), 33-44. Doi: 10.1111/bld.12210.
Abstract: Dementia prevalence rates are higher amongst people with intellectual [learning] disabilities than the general population. People with Down's syndrome are at even greater risk of developing dementia and of developing dementia at an earlier age. This study, conducted as part of a wider service evaluation, explored community learning disability team perspectives on screening, pathways, training, information and supports developed to improve services for people with learning disabilities and dementia. A focus group was held with health and social care professionals working in community learning disability services. Thematic analysis was used to analyze the data. The dementia screening, pathways and processes had become embedded in practice, leading to a common framework, an efficient, multidisciplinary, proactive approach, earlier detection and diagnosis of dementia and identification of other health needs and issues. This avoided crisis situations supporting people to remain at home longer. Training and information were felt to improve care quality and reduce caregiver anxiety. People with intellectual disabilities and caregivers were involved to varying extents. External influences impacting on support included the availability, appropriateness, cost and effectiveness of different models of service provision. Service developments have been made as a result of the findings which suggest that dementia pathways and supports improve service provision and outcomes for people with intellectual disabilities. It is important to develop the evidence base on the effectiveness of different service models for people with intellectual disabilities and dementia.

Chaput, J.L.
Housing people with Alzheimer disease as a result of Down syndrome: a quality of life comparison between group homes and special care units in long term care facilities.
Master's thesis, Department of City Planning, University of Manitoba (1998)
Abstract: Report of study to determine which form of housing, group homes or special care units (SCUs), provided an enhanced quality of life for individuals with Down syndrome (DS) and Alzheimer disease (AD). Ten long term care (LTC) facilities with SCUs for people with AD in the Winnipeg, Canada area and ten group homes for people with DS and AD across Canada participated in the study. Results indicated that the group homes seemed to provide an enhanced quality of life for adults with DS and AD because they provided a home-like environment and they operated according to a therapeutic philosophy of care. In addition, costs for caregiving seemed to be more economical in group homes than in SCUs because group homes utilized lower staff wages and medical costs. Report provides information on practices and costs.

Chaput, J.L., & Udell, L.
Housing people with Alzheimer disease as a result of Down syndrome: a quality of life comparison between group homes and special care units in long term care facilities.
Journal of Intellectual Disability Research, 2000, 44, 236 (abstract No. 186)
[Paper presented at the 11th World Congress of the International Association for

the Scientific Study of Intellectual Disabilities, Seattle, Washington (USA), August 1-6, 2000]
Abstract: The purpose of the study was to determine which form of housing, i.e., group homes or special care units (SCUs), provided a better quality of life for individuals with Alzheimer disease (AD) as a result of Down syndrome (DS). The study also provided Winnserv Inc. (a non-profit housing organization that houses people with mental disabilities) with important information. Using the study results, Winnserv Inc. was able to determine that their group homes were suitable to maintain individuals with DS and AD and that their group homes were more cost-effective than SCUs in terms of caregiving. Twenty caregivers from both group homes and SCUs were selected to participate in this study. Ten long term care (LTC) facilities with SCUs for people with AD were selected in the Winnipeg area and ten group homes for people with Down syndrome and AD were chosen in Winnipeg and across Canada. The results indicated that the group homes seemed to provide the best quality of life for people with AD as a result of Down syndrome because they provided a home-like environment and they operated according to a therapeutic philosophy of care. In addition, costs for caregiving seemed to be more economical in group homes than in SCUs because group homes utilized lower staff wages and medical costs. Based on the results, it was recommended that Winnserv Inc. continue to house people with DS and AD.

Chaput, J.L.
Adults with Down syndrome and Alzheimer's disease: Comparisons of services received in group homes and in special care units
Journal of Gerontological Social Work, 2002, 38, 197-211
https://doi.org/10.1300/J083v38n01_05
Abstract: An increasing number of people with Down syndrome are at risk of dementia resulting from Alzheimer's disease. Many reside in community group homes. When they are affected by dementia, the challenge to agencies providing group homes is how to best provide continued housing and provide effective dementia-related care management. In the general population, long term care is typically provided in nursing facilities, often in special care units (SCUs). This study evaluated select factors found in group homes and SCUs to determine which is able to provide a better quality of life for people with Down syndrome affected by dementia. Interviews, using quality of life indicators, were conducted at 20 sites, equally selected from group homes and SCUs, on the basis of their experience with people with dementia. Results indicate that group homes can provide conditions associated with better quality of life and, additionally, operate with lower staffing costs due to the non-utilization of medical staff.

Chicoine, B., Rivelli, A., Fitzpatrick, V., Chicoine, L., Jia, G., Rzhetsky, A.
Prevalence of common disease conditions in a large cohort of individuals with Down syndrome in the United States.
Journal of Patient-Centered Research and Reviews, 2021, 8(2), 86-97. doi: 10.17294/2330-0698.1824
Abstract: Given the current life expectancy and number of individuals living with Down syndrome (DS), it is important to learn common occurrences of disease conditions across the developmental lifespan. This study analyzed data from a large cohort of individuals with DS in an effort to better understand these disease conditions, inform future screening practices, tailor medical care guidelines, and improve utilization of health care resources. This retrospective, descriptive study incorporated up to 28 years of data, compiled from 6078 individuals with DS and 30,326 controls matched on age and sex. Data were abstracted from electronic medical records within a large Midwestern health system. In general, individuals with DS experienced higher prevalence of testicular cancer, leukemias, moyamoya disease, mental health conditions, bronchitis and pneumonia, gastrointestinal conditions, thyroid disorder, neurological conditions, atlantoaxial subluxation, osteoporosis, dysphagia, diseases of the eyes/adnexa and of the ears/mastoid process, and sleep apnea, relative to matched controls. Individuals with DS experienced lower prevalence of solid tumors, heart disease conditions, sexually transmitted diseases, HIV, influenza, sinusitis, urinary tract infections, and diabetes. Similar rates of prevalence were seen for lymphomas, skin melanomas, stroke, acute myocardial infarction, hepatitis, cellulitis, and osteoarthritis. While it is challenging to draw a widespread conclusion about comorbidities in individuals with Down syndrome, it is safe to conclude that care for individuals with DS should not automatically mirror screening, prevention, or treatment guidelines for the general U.S. population. Rather, care for those with DS should reflect the unique needs and common comorbidities of this population.

Choi, P., Motl, R.W., & Agiovlasitis, S.
Feasibility of social cognitive theory-based fall prevention intervention for people with intellectual disabilities living in group-home.
Journal of Intellectual Disability Research, 2022 Dec 18. doi: 10.1111/jir.13001. Epub ahead of print.
Abstract: Adults with intellectual disability (ID) have a higher rate of fall events than the general population. Consequently, interventions for reducing fall events and improving health are highly required for individuals with ID. One essential step towards effectively delivering fall prevention interventions among adults with ID involves evaluating their feasibility. This study examined the feasibility of a home-based exercise intervention, supplemented with behavioural change strategies, among individuals with ID living in residential settings. This study provided an 8-week intervention, consisting of a workshop for support workers and sessions for participants with ID, focusing on behavioural reward/s, education regarding fall prevention/exercise and exercise training. One week prior to and 1 week following such an intervention, such participants underwent measurements for (1) physical performance, (2) fall efficacy, (3) self-efficacy for activity and (4) social support. Participants having ID (n = 33), support workers (n = 11) and one administrator participated in this study. There were no adverse events during the intervention, and the mean adherence rate was 70.8 ± 19.5%. Two participants with ID dropped out of the programme due to a lack of interest. The participants with ID significantly improved individual physical performance, self-efficacy for activity, fall efficacy and support from friends and support workers. Fall prevention interventions for adults with ID living in group-homes were highly promising for eventual large-scale implementation within such communities.

Cipriani, G., Danti, S., Carlesi, C., & DiFiorino, M.
Aging with Down syndrome: the dual diagnosis - Alzheimer's disease and Down syndrome
American Journal of Alzheimer's Disease & Other Dementias, 2018, 33(4), 253-262. doi: 10.1177/1533317518761093. Epub 2018 Mar 5.
Abstact: People with Down syndrome (DS) enjoy a longer life expectancy now than they ever have before and are therefore at greater risk of developing conditions associated with aging, including dementia. Authors undertook at review to explore the phenomenon of dementia in DS. Medline and Google Scholar searches were conducted for relevant articles, chapters, and books published until 2017. Search terms included Alzheimer's disease, cognitive impairment, dementia, DS, and trisomy 21. Publications found through this indexed search were reviewed for further references. Authors concluded that virtually, all subject aged 35 to 40 show key neuropathologic changes characteristic of Alzheimer's disease, but only a part of them show clinical signs of dementia, usually around the age of 50 years. Early signs of dementia in people with DS may be different from those experienced by the general population. Failure to recognize this can delay diagnosis and subsequent interventions.

Cipriani, G., Picchi, L., Dolciotti, C., & Bonucecelli, U.
Alzheimer's disease and Down's syndrome: An unhappy union [La malattia di Alzheimer e la sindrome di Down: un infelice connubio]
Quaderni Italiani di Psichiatria, 2011 March, 30(1), 26-32.
https://www.researchgate.net/publication/235977633_La_malattia_di_Alzheimer_e_la_sindrome_di_Down_un_infelice_connubio_Alzheimer's_disease_and_Down's_syndrome_an_unhappy_union
Abstract: People with Down's syndrome (DS) have an increased risk of developing Alzheimer's disease (AD) during middle age. Both disorders can present with a decline in cognitive skills and behavioral symptoms. Therefore, dementia, particularly in its early stages, can be difficult to diagnose in this population. We conducted a search of electronic databases for literature on the relationship between AD and DS. The key words used were: ''Down syndrome'', ''Alzheimer's disease'', ''dementia'', and ''mental retardation''. AD onset has been reported as early as age 30 in individuals with DS, and there is a dramatic increase in prevalence rates in older age groups. This trend reflects increased survival of persons with DS probably as a result of advances in medical treatment and improved living conditions. Even with careful clinical assessment, it can be very difficult to identify early symptoms of dementia when it is superimposed on a background of intellectual disability. The reasons include the wide intra-individual variability in cognitive functioning and difficulties involved in establishing baseline levels of the premorbid condition. Many frontal lobe-related symptoms usually associated with later stages of dementia in the general population are commonly seen in the early stage of the dementia that develops in adults with DS. After

onset, the clinical symptoms of dementia progress rapidly in all subjects with DS. Research suggests that the presentation of dementia in people with DS may differ from that typical of AD in the general population. Early changes tend to involve personality and behavior rather than memory. DS can be best understood as a complex syndrome of genetic origin that has protean neurobiological consequences and numerous clinical characteristics. [Note: article text in Italian]

Cleary, J., & Doody, O.
Professional carers' experiences of caring for individuals with intellectual disability and dementia: A review of the literature.
Journal of Intellectual Disabilities, 2017 Mar, 21(1), 68-86. doi:10.1177/1744629516638245.
Abstract: The number of people with intellectual disability living into old age and developing dementia continues to increase. Dementia presents a wide range of challenges for staff due to progressive deterioration. This article presents the findings from a narrative literature review of professional caregivers' experiences of caring for individuals with intellectual disability and dementia. Seven electronic databases were searched using Boolean operators and truncation to identify relevant literature. Search results were combined and narrowed to articles relevant to staff working with individuals with intellectual disability and dementia, and 14 articles met the criteria for review. Themes outlined in the review include staff knowledge of dementia, staff training in dementia, caregiving, challenging behavior, pain management, mealtime support and coping strategies. Overall carers must review and adjust their care delivery and support to people with intellectual disability and dementia, not only in terms of identifying and responding to their health needs but also through collaborative team working within and across services.

Cleary J, & Doody O.
Nurses' experience of caring for people with intellectual disability and dementia.
Journal of Clinical Nursing, 2017, 26(5-6), 620-631. doi: 10.1111/jocn.13431. Epub 2016 Nov 14.
Abstract: The authors endeavored to explore nurses' experiences of caring for older people with intellectual disability and dementia. Ageing and dementia prevalence is increasing along with the life expectancy of people with intellectual disability. As a population group, people with intellectual disability have a high prevalence of dementia, which is higher within the subpopulation of Down syndrome. People with intellectual disability live in residential care, community or residential settings, and nurses are required to adapt their practices to meet the changed needs of the individual. A qualitative Husserlian descriptive phenomenological methodology was undertaken by the researchers so as to be able to become absorbed in the quintessence of meaning and explore nurses' experience of working with older people with intellectual disability and dementia. Ethical approval was obtained, and data were collected utilizing semistructured interviews (n = 11). Interviews were transcribed and analyzed using Colaizzi's framework for data analysis. The authors extracted three key themes were identified: 'knowledge of dementia', 'person-centred care' and 'transitioning within the service'. The study highlights the need for proactive planning, life story books of the patient, and funding to support client and staff. The authors concluded that overall, the study highlights the importance of knowing the person, supporting the individual and recognizing presenting behaviors as outside the control of the individual. The article presents the experiences of nurses caring for the older person with intellectual disability and dementia. Transitions are often very difficult for both the person and their peers, and they experience benefit from the efforts of a multidisciplinary team facilitating a person-centered approach.

Clifford, C., & Lauer, E.
Evaluating dementia capability of service systems for people with intellectual and developmental disorders and dementia
Journal of Applied Research in Intellectual Disabilities, 2021, 34(5), 1215-1216. https://onlinelibrary.wiley.com/toc/14683148/2021/34/5
Abstract: People with an intellectual or developmental disability (IDD) experience complex age-related issues, including dementia-related disorders, at higher rates and earlier ages than the general population. Increased support needs of this subpopulation can strain caregivers and existing community supports. Patterns of resource awareness and utilization and unmet needs are not well understood for this subpopulation. A collaboration of the Massachusetts Council on Aging (MCOA), the Massachusetts Department of Developmental Services (DDS), and the Center for Developmental Disability Evaluation and Research (CDDER) at University of Massachusetts Medical School conducted a needs assessment with caregivers of people with intellectual and developmental disabilities to assess awareness and utilization of community-based resources, and unmet needs including for caregiver supports. Home visits, including an environmental assessment, and interviews with caregivers of 95 adults with dementia-related diagnoses were conducted. Interviews asked about changes in the adult's condition since diagnosis including a needs assessment about the dementia-related knowledge and training, care confidence levels, perceived barriers and/or concerns to care provision and resource use. About half of the caregivers reported significant changes in the person's skills, function, and memory since diagnosis, as well as worsening of the person's gait, continence, and swallowing. 78% of caregivers reported feeling confident providing care currently and 68% were confident about providing future care. 100% of the respondents found outreach provided by a nurse practitioner helpful. Caregiver concerns included lack of suitable day programming, future planning resources, and caregiver burnout/stress. Most caregivers were currently aware of some local resources but with scattered use. Environmental assessments indicated most homes, while accessible, warranted additional lighting and clutter removal. Caregivers requested additional training in addressing the behavioral and mental health needs. Findings suggest a need for increased collaboration across the intellectual and developmental disability and aging/human services systems, and additional training and resource navigation guides for caregivers.

Cohen, U., & Wiesman, G.D.
Holding on to home: Designing environments for people with dementia.
181 pp.
Baltimore: Johns Hopkins University Press (1991)
Abstract: General text on adapting homes and living environments for persons with dementia; applicable to home and other residential situations for adults with intellectual disabilities and dementia.

Collacott R.A.
Epilepsy, dementia and adaptive behaviour in Down's syndrome.
Journal of Intellectual Disability Research, 1993, 37(2), 153-60. doi: 10.1111/j.1365-2788.1993.tb00582.x.
Abstract: Widespread inquiry identified 378 adults with Down's syndrome resident in Leicestershire, England. The immediate carer of 351 of these (92.8%) was interviewed for the purpose of establishing a past history of seizures, including the age at which the seizures began. The immediate carer was also invited to provide information to enable the completion of an Adaptive Behaviour Scale (A.B.S.) rating. Individuals with a history of seizures were divided into two groups on the basis of whether or not seizures commenced prior to or after age 35 years. Two control groups of individuals with Down's syndrome, but without a history of seizures were selected. Adaptive Behaviour Scale scores for those in whom seizures commenced at a younger age were similar to those who had no recorded history of seizures. However, in those in whom seizures began in later life, scores on all domains of the A.B.S. were significantly reduced compared to both young epileptic patients and their controls. Adaptive Behaviour Scale scores for the older control group held an intermediate position, suggesting that late-onset epilepsy may be a late manifestation of a dementing process. A clinical diagnosis of dementia recorded in the case records was significantly associated with the presence of late-onset epilepsy. This is supportive of the hypothesis that late-onset epilepsy in individuals with Down's syndrome is associated with Alzheimer's disease.

Coppus, A.M.W
People wtih intellectual disabiltiy: What do we know about adulthood and life expectancy?
Developmental Disabilities Research Review, 2013, 18(1), 6-16. doi: 10.1002/ddrr.1123.
Abstract: Increases in the life expectancy of people with Intellectual Disability have followed similar trends to those found in the general population. With the exception of people with severe and multiple disabilities or Down syndrome, the life expectancy of this group now closely approximates with that of the general population. Middle and old age, which until 30 years ago were not recognized in this population, are now important parts of the life course of these individuals. Older adults with Intellectual Disabilities form a small, but significant and growing proportion of older people in the community. How these persons grow older and how symptoms and complications of the underlying cause of the Intellectual Disability will influence their life expectancy is of the utmost importance.

Coppus, A. , Evenhuis, H., Verberne, G.J., Visser, F., van Gool, P., Eikelenboom, P., & van Duijin, C.
Dementia and mortality in persons with Down's syndrome.
Journal of Intellectual Disability Research, 2006, 50(10), 768-77. doi:10.1111/j.1365-2788.2006.00842.x.
Abstract: Numerous studies have documented that persons with Down's syndrome (DS) are at an increased risk of Alzheimer's disease (AD). However, at present it is still not clear whether or not all persons with DS will develop dementia as they reach old age. We studied 506 people with DS, aged 45 years and above. A standardized assessment of cognitive, functional and physical status was repeated annually. If deterioration occurred, the patients were examined and the differential diagnosis of dementia was made according to the revised Dutch consensus protocol and according to the ICD-10 Symptom Checklist for Mental Disorders. We compared our findings with those reported in the literature. The overall prevalence of dementia was 16.8%. Up to the age of 60, the prevalence of dementia doubled with each 5-year interval. Up to the age of 49, the prevalence is 8.9%, from 50 to 54, it is 17.7%, and from 55 to 59, it is 32.1%. In the age category of 60 and above, there is a small decrease in prevalence of dementia to 25.6%. The lack of increase after the age of 60 may be explained by the increased mortality among elderly demented DS patients (44.4%) in comparison with non-demented patients (10.7%) who we observed during a 3.3-year follow-up. There was no decrease in incidence of dementia in the age group of 60 and above. Our findings are very similar to those published in the literature. Patients with dementia were more frequently treated with antiepileptic, antipsychotic and antidepressant drugs. The history of depression was strongly associated with dementia. Our study is one of the largest population-based studies to date. We found that despite the exponential increase in prevalence with age, the prevalence of dementia in the oldest persons with DS was not higher than 25.6%.

Coppus, A.M.W , Evenhuis, H.M, Verberne, G-J., Visser, F.E., Oostra, B.A. Eikelenboom, P., van Gool, W.A., Cecile, A., Janssens, J.W. , van Duijn, C.M.
Survival in elderly persons with Down syndrome.
Journal of the American Geriatrics Society, 2008, 56(12), 2311-2316. doi: 10.1111/j.1532-5415.2008.01999.x.
Abstract: The longer life expectancy now experienced by persons with Down syndrome (DS) makes it necessary to know the factors influencing survival in older persons with this syndrome. In a prospective longitudinal cohort study of dementia and mortality, 506 persons with DS aged 45 and older were followed for a mean of 4.5 years (range 0.0–7.6 years). Cognitive and social functioning were tested at baseline and annual follow-up. The diagnosis of dementia was determined according to a standardized protocol. Cox proportional hazards modeling was used for survival analysis. Relative preservation of cognitive and functional ability is associated with better survival in this study population. Clinically, the most important disorders in persons with DS that are related to mortality are dementia, mobility restrictions, visual impairment, and epilepsy -- but not cardiovascular diseases. Also, level of intellectual disability and institutionalization were associated with mortality.

Coppus, A., Evenhuis, H., Verberne, G.J., Visser, F., van Gool, P., Eikelenboom, P., & van Duijin, C.
Dementia and mortality in persons with Down's syndrome.
Journal of Intellectual Disability Research, 2006, Oct;50(Pt 10):768-77. doi: 10.1111/j.1365-2788.2006.00842.x.
Abstract: Numerous studies have documented that persons with Down syndrome (DS) are at an increased risk of Alzheimer's disease (AD). However, at present it is still not clear whether or not all persons with DS will develop dementia as they reach old age. The authors studied 506 people with DS, aged 45 years and above. A standardized assessment of cognitive, functional and physical status was repeated annually. If deterioration occurred, the patients were examined and the differential diagnosis of dementia was made according to the revised Dutch consensus protocol and according to the ICD-10 Symptom Checklist for Mental Disorders. We compared our findings with those reported in the literature. The overall prevalence of dementia was 16.8%. Up to the age of 60, the prevalence of dementia doubled with each 5-year interval. Up to the age of 49, the prevalence is 8.9%, from 50 to 54, it is 17.7%, and from 55 to 59, it is 32.1%. In the age category of 60 and above, there is a small decrease in prevalence of dementia to 25.6%. The lack of increase after the age of 60 may be explained by the increased mortality among elderly demented DS patients (44.4%) in comparison with non-demented patients (10.7%) who we observed during a 3.3-year follow-up. There was no decrease in incidence of dementia in the age group of 60 and above. Our findings are very similar to those published in the literature. Patients with dementia were more frequently treated with antiepileptic, antipsychotic and antidepressant drugs. The history of depression was strongly associated with dementia. The authors concluded that their study is one of the largest population-based studies to date. We found that despite the exponential increase in prevalence with age, the prevalence of dementia in the oldest persons with DS was not higher than 25.6%.

Coppus, A.M., Evenhuis, H.M., Verberne, G.J., Visser, F.E., Eikelenboom, P., van Gool, W.A., Janssens, A.C., & van Duijn, C.M.
Early age at menopause is associated with increased risk of dementia and mortality in women with Down syndrome.
Journa of Alzheimer's Disease, 2010;19(2):545-50. doi: 10.3233/JAD-2010-1247.
Abstract: In a prospective longitudinal cohort study of dementia and mortality in persons with Down syndrome aged 45 years and older, 85 postmenopausal women were followed for a mean follow-up time of 4.3 years (range 0.0 to 7.4 years). The effect of age at menopause on age at diagnosis of dementia and survival was estimated using correlation analysis and Cox Proportional Hazard Model. We found a significant correlation between age at menopause and age at diagnosis of dementia (rho=0.52; p< 0.001), and between age at menopause and age at death (rho=0.49; p=0.01). Early age at menopause is associated with a 1.8 fold increased risk of dementia: Hazard Ratio (HR): 1.82 (95%Confidence Interval (CI): 1.31-2.52) and with risk of death: HR: 2.05 (95%CI: 1.33-3.16). Our study suggests that age at menopause in women with Down syndrome is a determinant of age at onset of dementia and mortality.

Cooper, S-A.
High prevalence of dementia among people with leaming disabilities not attributable to Down's syndrome.
Psychological Medicine, 1997, 27(3), 609-616. doi: 10.1017/s0033291796004655.
Abstract: For many years, it has been known that dementia can occur in people with learning disabilities, but there have been few research studies. Studies that do quote rates for dementia show these to be high, but this important fact has received remarkably little attention. Comprehensive psychiatric and medical assessments were undertaken on the whole population (ascertained as far as is possible) of people with learning disabilities aged 65 years and over living in Leicestershire, UK (N=134), in order to ascertain rates of DCR defined dementia, and associated factors. Dementia was diagnosed in 21·6%, against an expected prevalence of 5·7%, for a group with this age structure. The rate of dementia increased in successive age cohorts: 15·6% aged 65-74 years; 23·5% aged 65-84 years; and 70·0% aged 85-94 years. People with dementia tended to be older, female, with more poorly controlled epilepsy, a larger number of additional physical disorders, less likely to be smokers and had lower adaptive behavior scores than did the elderly people without dementia. They were more likely to live in health service accommodation. Dementia occurs at a much higher rate among elderly people with learning disabilities than it does among the general population; this is independent of the association between dementia and Down's syndrome. Whether this relates etiologically to genetics, lack of brain 'reserve' or history of brain damage is yet to be determined.

Cooper, S-A., & Prasher, V.
Maladaptive behaviours and symptoms of dementia in adults with Down's syndrome compared with adults with intellectual disability of other aetiologies
Journal of Intellectual Disability Research, 1998, 42(4), 293-300. https://doi.org/10.1046/j.1365-2788.1998.00135.x
Abstract: Dementia commonly occurs in elderly people with intellectual disability, especially those with Down's syndrome. The non-cognitive symptoms of dementia can be of greater significance to individuals and carers than the cognitive changes caused by this condition. It is not known whether there are differences between people with Down's syndrome and those with intellectual disability of other causes with regard to the prevalence of such symptoms. The present study was undertaken to draw a comparison between a group with Down's syndrome and dementia (n= 19), and a group with intellectual disability of other causes and dementia (n= 26). Maladaptive behaviours and psychiatric symptomatology were assessed in both groups. The group with Down's syndrome had a higher prevalence of low mood, restlessness/excessive overactivity, disturbed sleep, being excessively uncooperative and auditory hallucinations. Aggression occurred with greater frequency in those subjects with

intellectual disability of other causes. These findings are of epidemiological importance in terms of service planning and understanding psychiatric presentation.

Cooper, S-A., & van der Speck
Epidemiology of mental ill health in adults with intellectual disabilities
Current Opinion in Psychiatry, 2009, Sep, 22(5), 431-436. doi:10.1097/YCO.0b013e32832e2a1e.
Abstact: Adults with intellectual disabilities experience higher rates of mental ill health than the general population. Despite this, the epidemiological knowledge base remains limited. The purpose of this article is to review mental health epidemiological studies relevant to adults with intellectual disabilities, published since January 2008. Several studies have aimed to build the epidemiological evidence base, particularly with regards to problem behaviours, which appear to be remitting-relapsing conditions rather than necessarily being chronic. Most of such work confirms prevalence and incidence rates, and conducts exploratory analyses to determine factors independently related to mental ill health. Down syndrome protects against problem behaviours and mental ill health (except dementia that occurs at a higher rate), whereas epilepsy does not appear to affect risk for mental ill health. Dementia is four times more common in older persons with intellectual disabilities without Down syndrome than in the general population. Persons with borderline intellectual disabilities also experience higher rates of mental ill health than the general population, but receive fewer treatments.

Cordell, C.B., Borson, S., Boustani, M., Chodosh, J., Reuben, D., Verghese, J., Thies,W., Fried, L.B.,& Medicare Detection of Cognitive Impairment Workgroup
Alzheimer's Association recommendations for operationalizing the detection of cognitive impairment during the Medicare Annual Wellness Visit in a primary care setting
Alzheimers & Dementia, 2013 Mar, 9(2),141-150. doi: 10.1016/j.jalz.2012.09.011. Epub 2012 Dec 20.
Abstract: The Patient Protection and Affordable Care Act added a new Medicare benefit, the Annual Wellness Visit (AWV), effective January 1, 2011. The AWV requires an assessment to detect cognitive impairment. The Centers for Medicare and Medicaid Services (CMS) elected not to recommend a specific assessment tool because there is no single, universally accepted screen that satisfies all needs in the detection of cognitive impairment. To provide primary care physicians with guidance on cognitive assessment during the AWV, and when referral or further testing is needed, the Alzheimer's Association convened a group of experts to develop recommendations. The resulting Alzheimer's Association Medicare Annual Wellness Visit Algorithm for Assessment of Cognition includes review of patient Health Risk Assessment (HRA) information, patient observation, unstructured queries during the AWV, and use of structured cognitive assessment tools for both patients and informants. Widespread implementation of this algorithm could be the first step in reducing the prevalence of missed or delayed dementia diagnosis, thus allowing for better healthcare management and more favorable outcomes for affected patients and their families and caregivers.

Cosgrave, M.P., Tyrrell, J., McCarron, M., Gill, M., & Lawlor, B.A.
Determinants of aggression, and adaptive and maladaptive behavior in older people with Down's syndrome with and without dementia.
Journal of Intellectual Disability Research, 1999, 43(5), 393-399.
Abstract: In a cross-sectional study of aggression, and adaptive and maladaptive behavior in 128 subjects with Down's syndrome (DS), 29 of whom had dementia, the current authors found that the presence of dementia was not predictive of aggression or maladaptive behavior. However, the level of adaptive behavior was shown to be lower in subjects with dementia, and in those with lower levels of cognitive functioning, as measured on a rating instrument, the Test for Severe Impairment. Although the presence of aggressive behaviors is not higher in subjects with dementia and DS on cross-sectional review, it remains to be seen whether aggression will increase in individual cases with the onset or progression of dementia. The decline in adaptive behavior shown in the present study confirms the findings of previous studies and indicates a direction for service development for persons with the dual diagnosis of dementia and DS.

Cosgrove, M.P., Tyrrell, J., McCarron, M., Gill, M., & Lawlor, B.A.
Age at onset of dementia and age of menopause in women with Down's syndrome.
Journal of Intellectual Disability Research, 1999, 43(6), 461-465.
Abstract: Menstrual status and the age of menopause were investigated in 143 Irish females with Down's syndrome (DS). The average age of menopause in 42 subjects (44.7 years) was younger than in the general population. The age at onset of dementia correlated with the age of menopause. This finding may be a manifestation of accelerated ageing in DS or point to oestrogen deficiency being an independent risk factor for the development of Alzheimer's

Courtenay, K., Jokinen, N.S., & Strydom, A.
Caregiving and adults with intellectual disabilities affected by dementia
Journal of Policy and Practice in Intellectual Disabilities, 2010, 7(1), 26-33. https://doi.org/10.1111/j.1741-1130.2010.00244.x
Abstract: Authors conducted a systematic review of the available Dutch, English, and German language literature for the period 1997–2008 on the current knowledge on social-psychological and pharmacological caregiving with respect to older adults with intellectual disabilities (ID) affected by dementia. Authors note that caregiving occurs on a personal level between the person and their carer and organizational and interorganizational supports have an impact on the quality of care provided. However, the lack of robust evidence to meet the needs of adults with ID affected by dementia means that service organizations often have to extrapolate from the evidence base of dementia care practices in the general population. The review showed that concerns over staff burden, behavioral interventions, and staff training, and applications of models of care were emerging, but were not systematically studied. Authors noted that pharmacological agents and nonpharmacological, psychosocial techniques were being used to assist carers manage behavior, but the evidence base of both nonpharmacological and pharmacological interventions that can help people with ID and dementia and their carers is insufficient because of the absence of systematic and robust studies. The authors note a need for an international research agenda that begins to address gaps in knowledge. With more adults projected to be affected by dementia, a robust evidence-based body of literature on dementia care in people with ID can help with planning for and providing quality dementia-capable services.

Cox, S.
Home solutions: Housing & support for people with dementia
London: The Housing Associations Charitable Trust [78 Quaker Street, London, England E1 6SW; e/m: hact@hact.org.uk] (1998)
112 pp.
Abstract: Publication details some 10 case studies of housing options and accommodations for persons affected by dementia (and applicable to adults with intellectual disabilities). Models covered include: support in a person's own home, support in a shared home, specialist dementia support with communal facilities, and different types and levels of support on one site. Sections also deal with housing and support solutions for people with dementia from ethnic minority communities and the repair, remodeling, adaptation and renovation of ordinary housing. Case models contain full descriptions of settings and accommodations.

Coyle, C.E., Kramer, J., & Mutchler, J.E.
Aging together: Sibling carers of adults with intellectual and developmental disabiliities
Journal of Policy and Practice in Intellectual Disabilities, 2014, 11(4), 302-312; doi: 10.1111/jppi.12094
Abstract: Family care provision is the norm for adults with intellectual and developmental disabilities (I/DD), even as they and their support networks grow older. As families age together, the role of primary carer frequently transitions from the parent to a sibling, as aging parents die or become too frail to provide continued support. This paper explores the transition in care from the perspective of a sibling who has replaced parents as the primary carer for an individual aging with I/DD. Data are drawn from semi-structured, in-depth interviews with a sample of adults over age 40, living in the United States, and caring for a sibling with I/DD(n = 15). Data were analyzed using a constant comparative qualitative approach. Results reveal themes impacting the adjustment to the role of primary carer, the extent to which aging transformed the content of care needs, the importance of planning, and the availability of supplementary support. First, we found that the aging process permeated the careproviding role, requiring ongoing modi?cation of that role as a result of the aging of the adult with I/DD and their family support system. A second key theme is that planning shapes the adjustment to the carer role.A third key theme is that the support systems surrounding the sibling dyad contribute to successful adjustment to care providing.Findings from this study underscore the need to develop long-term

services and supports as well as educational resources that accommodate this population of carers as they age together with their sibling with I/DD.

Cutler, N.R., Heston, L.L., Davies, P., Haxby, J.V., & Schapiro, M.B.
NIH Conference. Alzheimer's disease and Down's syndrome: new insights.
Annuals of Internal Medicine, 1985,103(4), 566-578.
Abstract: Neuropathologic and neurochemical studies of older adults with Down's syndrome and those with Alzheimer's disease reveal striking similarities. Genetic studies indicate that near relatives of patients with Alzheimer's disease are at increased risk of developing Alzheimer's disease, and the risk appears to be age specific. These families with familial Alzheimer's disease have also been found to have a high incidence of Down's syndrome. Neurochemical data suggest that a cholinergic deficiency must be present for dementia to develop, and serial assessments of brain metabolic function with positron emission tomography in Alzheimer's disease have shown that the parietal lobe has reductions in metabolic function before the onset of neuropsychologic deficits in this brain region. Neuropsychologic testing indicates that patients with Down's syndrome over 35 years old have poorer cognitive skills than do younger patients. Brain metabolic function is excessively reduced in the demented adults with Down's syndrome.

Dalton, A.J., Fedor, B.L., Patti, P.J., Tsiouris, J.A., & Mehta, P.D.
TheMultidimensional Observation Scale for Elderly Subjects (MOSES): studies in adults with intellectual disability
Journal of Intellectual & Developmental Disability, 2002, 27(4), 310-324.
https://doi.org/10.1080/1366825021000029348
Abstract: This report describes the results of five studies aimed at evaluating the usefulness, reliability, and validity of the Multidimensional Observation Scale for Elderly Subjects (MOSES) in the assessment of change in ageing persons with intellectual disability. Three hundred and thirty-six individuals with an average age of 49.8 years, including an equal number of men and women, were participants in one or more of the five studies. There were 220 participants with Down syndrome, 81 persons without Down syndrome with intellectual disability, and 35 persons from the general ageing population who were clinically diagnosed with Alzheimer's disease using NINCD/ADRDA criteria. Persons with Down syndrome 40 years of age and older could be distinguished from their younger peers with Down syndrome by statistically significant poorer scores on the MOSES, with those 50 years of age and older showing the worst scores. Comparisons of adults with intellectual disability diagnosed with dementia of the Alzheimer type (DAT), using DSM-IV criteria, with or without Down syndrome, as well as comparisons of patients with clinically diagnosed depression, provided evidence that subtests of the MOSES were sensitive to DAT but less so to depression. These clinical groups also showed significantly poorer scores on the MOSES when compared with those of the normative sample. It was concluded that the MOSES is a behavioural observation scale that can provide useful information in clinical settings as well as in research.

Dalton, A.J., Mehta, P.D., Fedor, B.L. & Patti, P.J.
Cognitive changes in memory precede those in praxis in aging persons with Down syndrome.
*Journal of Intellectual & Developmental Disability,*1999, *24*(2), 169-187.
Abstract: Experimental tests of cognitive functions were developed and standardised to detect the onset and progression of the early stage of Alzheimer disease in persons with Down syndrome. The aim was to determine whether or not there was a specific sequence of cognitive changes over a 3-year period for the test measures. When compared with a young group (17-39years of age at the start), an old group of persons with Down syndrome (40-58years of age at the start) showed small but statistically significant changes over time suggestive of "pre-clinical signs" of dementia. When the data were sorted into 4 subgroups on the basis of age, a more detailed analysis revealed that the subgroup that was 50 years of age and older at the start showed changes in scores which were of a magnitude more clearly indicative of early dementia on the test measures. Deterioration in learning/ memory functions began at a mean age of 54.2 years, followed later by deterioration in movement-related functions (praxis) at a mean age of 56.9 years. Deterioration in scores on an informant-based behavior rating scale (MOSES) occurred at an intermediate age of 55.0 years. The results provide preliminary support for the hypothesis that persons with Down syndrome who are 50 years of age and older may develop a specific sequence of functional changes during the early stage of dementia. They also illustrate ways in which small sample norms can be effectively used to increase the practical usefulness of tests intended to evaluate dementia in persons with intellectual disabilities.

Das J.P., Davis B., Alexander J., Rauno K.P. & Naglieri J.A.
Cognitive decline due to aging among persons with Down's syndrome.
Research in Developmental Disabilities, 1995, Nov-Dec, 16(6): 461-4788 doi: 10.1016/0891-4222(95)00030-5.
Abstract: This study examined decline in cognitive functions in individuals with Down syndrome (DS) over the age of 40 in comparison to participants of the same age and comparable mental handicap without Down syndrome (NonDS). Both DS (n = 32) and NonDS (n = 31) samples were divided into "younger" (40-49 years) and "older" (50-62) groups. Cognitive processes were examined by tests of general intellectual functioning (Dementia Rating Scale, Peabody Picture Vocabulary Test-Revised, and the Matrix Analogies Test-Expanded form), as well as planning, attention, simultaneous, and successive processing tests taken from Das-Naglieri Cognitive Assessment System. The older individuals with Down syndrome performed more poorly than those in the other three groups. The differences were particularly evident in tasks requiring planning and attention. Authors note the possibility of using these tests as indicators of the early signs of Alzheimer's disease.

 Davis, D.R.
A parent's perspective
In M.P. Janicki & A.J. Dalton (Eds.), Dementia, aging, and intellectual disabilities. pp. 42-50
Philadelphia: Brunner-Mazel (1999)
Abstract: Book chapter that provides an account of the experiences of a family with an adult son with Down syndrome who eventually succumbs to dementia of the Alzheimer's type. Includes a discussion of the difficult early years of the son's life and the challenges the family faced as he aged. It also examines the family's problems in recognizing that their son was experiencing the onset of dementia and his gradual decline until his death at age 46.

Day, K., Carreon, D., & Stump, C.
The therapeutic design of environments for people with dementia: A review of the empirical research
Gerontologist, 2000, 40, 397-416
Abstract: Design of the physical environment is increasingly recognized as an important aid in caring for people with dementia. This article reviews the empirical research on design and dementia, including research concerning facility planning (relocation, respite and day care, special care units, group size), research on environmental attributes (noninstitutional character, sensory stimulation, lighting, safety), studies concerning building organization (orientation, outdoor space), and research on specific rooms and activity spaces (bathrooms, toilet rooms, dining rooms, kitchens, and resident rooms). The analysis reveals major themes in research and characterizes strengths and shortcomings in methodology, theoretical conceptualization, and application of findings.

Davies, M., McGllade, A., & Bickerstaff, D.
A needs assessment of people in the Eastern Health and Social Services Board (Northern Ireland) with intellectual disability and dementia
Journal of Learning Disabilities, 2002, 6, 23-33.
Abstract: Article details a study undertaken by the Eastern Health and Social Services Board (Northern Ireland) which aimed to identify the number of people with intellectual disability within this area who were diagnosed with or were thought to have dementia. The objectives of the study were to collate demographic details and to profile the needs of this group. Key workers were asked to provide this information and were invited to comment on gaps in existing service provision and on future needs. A number findings emerged: diagnostic services were patchy; people with dementia were living in a range of residential settings; carers wished to care for their clients for as long as practically possible, but they required extra resources and training to do so; and some individuals with an intellectual disability were excluded from elderly services. A report was compiled incorporating 12 recommendations.

Davys, D., & Haigh, C.
Older parents of people who have a learning disability: Perceptions of future accommodation needs
British Journal of Learning Disabilities, 2007, 36, 66–72.
doi:10.1111/j.1468-3156.2007.00447.x
Abstract: The aim of this qualitative study was to provide an insight into the perceptions of older parents of adults with intellectual diability on the future

accommodation needs of their adult children. Semi-structured interviews were used to seek parental awareness of residential options available, concerns in relation to future accommodation and the preferred accommodation options for their offspring. Four couples who shared the family home with an adult who has an intellectual disabilty took part in the study and data were analyzed using a step-by-step form of content analysis as described by Burnard *[Nurse Educ Today 11 (1991), 461*]. Emergent themes from transcripts were then organized into main categories. The results of this study suggest that older parents are dissatisfied with both statutory and private services, that they have concerns for their other children, and their own aging. Being a parent to a person who has an intellectual disability is seen to be a difficult task, were worried about what the adult's brothers and sisters may have to do in the future, but also wanted to provide support at home for as long a possible. Of the parents who participated in this study, three couples wanted to maintain their adult child at home for as long as possible and the parents who were actively seeking accommodation outside the family home expected to be involved in all aspects of their daughter's care for the long-term future.

de Franca Bram, J.M., Talib, L.L., Giroud Joaquim, H.P., Carvalho, C.L., Gattaz, W.G., & Forlenza, O.V.
Alzheimer's disease-related biomarkers in aging adults with Down syndrome: systematic review
Current Psychiatry Research and Reviews, 2019, 15(1), 49-57. DOI: 10.2174/1573400515666190122152855
Abstract: Down syndrome (DS) is associated with a high prevalence of cognitive impairment and dementia in middle age and older adults. Given the presence of common neuropathological findings and similar pathogenic mechanisms, dementia in DS is regarded as a form of genetically determined, early-onset AD. The clinical characterization of cognitive decline in persons with DS is a difficult task, due to the presence intellectual disability and pre-existing cognitive impairment. Subtle changes that occur at early stages of the dementing process may not be perceived clinically, given that most cognitive screening tests are not sensitive enough to detect them. Therefore, biological markers will provide support to the diagnosis of DS-related cognitive impairment and dementia, particularly at early stages of this process. To perform a systematic review of the literature on AD-related biomarkers in DS. We searched PubMed, Web of Science and Cochrane Library for scientific papers published between 2008 and 2018 using as primary mesh terms 'Down', 'Alzheimer', 'biomarker'. 79 studies were retrieved, and 39 were considered eligible for inclusion in the systematic review: 14 post-mortem studies, 10 neuroimaging, 4 addressing cerebrospinal fluid biomarkers, and 11 on peripheral markers. There is consistent growth in the number of publication in this field over the past years. Studies in DS-related dementia tend to incorporate many of the diagnostic technologies that have been more extensively studied and validated in AD. In many instances, the study of CNS and peripheral biomarkers reinforces the presence of AD pathology in DS.

de Sola, S., de la Torre, R., Sánchez-Benavides, G., Benejam, B. , Cuenca-Royo, A., del Hoyo, L., Rodríguez, J., Catuara-Solarz, S., Sanchez-Gutierrez, J., Dueñas-Espin, I., Hernandez, G., Peña-Casanova, J., Langohr, K., Videla, S., Blehaut, H., Farre,M,. Dierssen, M. & The TESDADStudy Group
A new cognitive evaluation battery for Down syndrome and its relevance for clinical trials.
Frontiers in Psychology, 2015, 6, 708. doi:10.3389/fpsyg.2015.00708.
Abstract: The recent prospect of pharmaceutical interventions for cognitive impairment of Down syndrome (DS) has boosted a number of clinical trials in this population. However, running the trials has raised some methodological challenges and questioned the prevailing methodology used to evaluate cognitive functioning of DS individuals. This is usually achieved by comparing DS individuals to matched healthy controls of the same mental age. We propose a new tool, the TESDAD Battery that uses comparison with age-matched typically developed adults. This is an advantageous method for probing the clinical efficacy of DS therapies, allowing the interpretation and prediction of functional outcomes in clinical trials. In our DS population the TESDAD battery permitted a quantitative assessment of cognitive defects, which indicated language dysfunction and deficits in executive function, as the most important contributors toother cognitive and adaptive behavior outcomes as predictors of functional change in DS. Concretely, auditory comprehension and functional academics showed the highest potential as end-point measures of therapeutic intervention for clinical trials: the former as a cognitive key target for therapeutic intervention, and the latter as a primary functional outcome measure of clinical efficacy. Our results also emphasize the need to explore the modulating effects of IQ, sex, and age on cognitive enhancing treatments. Noticeably, women performed significantly better than men of the same age and IQ in most cognitive tests, with the most consistent differences occurring in memory and executive functioning and negative trends rarely emerged on quality of life linked to the effect of age after adjusting for IQ and sex. In sum, the TESDAD battery is a useful neurocognitive tool for probing the clinical efficacy of experimental therapies in interventional studies in the DS population suggesting that age-matched controls are advantageous for determining normalization of DS.

De Vreese, L.P., Gomiero, T., Uberti M., De Bastiani, E., Weger, E., Mantesso, U., & Marangoni, A.
Functional abilities and cognitive decline in adult and aging intellectual disabilities. Psychometric validation of an Italian version of the Alzheimer's Functional Assessment Tool (AFAST): analysis of its clinical significance with linear statistics and artificial neural networks.
Journal of Intellectual Disability Research, 2015, Apr; 59(4), 370-84. doi: 10.1111/jir.12113.
Abstract: A psychometric validation of an Italian version of the Alzheimer's Functional Assessment Tool scale (AFAST-I), designed for informant-based assessment of the degree of impairment and of assistance required in seven basic daily activities in adult/elderly people with intellectual disabilities (ID) and (suspected) dementia; and a pilot analysis of its clinical significance with traditional statistical procedures and with an artificial neural network. AFAST-I was administered to the professional caregivers of 61 adults/seniors with ID with a mean age (± SD) of 53.4 (± 7.7) years (36% with Down syndrome). Internal consistency (Cronbach's a coefficient), inter/intra-rater reliabilities (intra-class coefficients, ICC) and concurrent, convergent and discriminant validity (Pearson's r coefficients) were computed. Clinical significance was probed by analysing the relationships among AFAST-I scores and the Sum of Cognitive Scores (SCS) and the Sum of Social Scores (SOS) of the Dementia Questionnaire for Persons with Intellectual Disabilities (DMR-I) after standardization of their raw scores in equivalent scores (ES). An adaptive artificial system (AutoContractive Maps, AutoCM) was applied to all the variables recorded in the study sample, aimed at uncovering which variable occupies a central position and supports the entire network made up of the remaining variables interconnected among themselves with different weights. AFAST-I shows a high level of internal homogeneity with a Cronbach's a coefficient of 0.92. Inter-rater and intra-rater reliabilities were also excellent with ICC correlations of 0.96 and 0.93, respectively. The results of the analyses of the different AFAST-I validities all go in the expected direction: concurrent validity (r=-0.87 with ADL); convergent validity (r=0.63 with SCS; r=0.61 with SOS); discriminant validity (r=0.21 with the frequency of occurrence of dementia-related Behavioral Excesses of the Assessment for Adults with Developmental Disabilities, AADS-I). In our sample age and gender do not correlate with the scale and comparing the distribution of the AFAST-I and DMR-SCS and DMR-SOS expressed as ES, it appears that memory disorders and temporal and spatial disorientation (SCS) precede the loss of functional abilities, whereas changes in social behaviour (SOS) are less specific in detecting cognitive deterioration sufficient to provoke functional disability and vice versa. The results of AutoCM analysis reveal that the hub (core) of the entire network is represented by the functional domain 'personal/oral hygiene' in the entire study sample and 'use of toilet' in a subgroup of subjects who obtained an ES equal to 0 at DMR-SCS. These results confirm the reliability and validity of AFAST-I and emphasise the complexity of the relationship among functional status, cognitive functioning and behaviour also in adults/seniors with ID.

De Vreese, L. P., Mantesso, U., De Bastiani, E., Weger, E., Marangoni, A.C., & Gomiero, T.
Procedures on cognition and behavior in older adults with intellectual disabilities: A 3-year follow-up study.
Journal of Policy and Practice in Intellectual Disabilities, 2012, 9(2), 92-102.
Abstract: Dementia appears at a higher rate among some adults with intellectual disabilities (ID) and this potentially poses a greater risk of nursing home admission. Yet, to date, there is no evidence on the efficacy of general dementia-derived environment-, personnel-, and patient-oriented intervention strategies in delaying onset of dementia or in slowing down its rate of progression in this population. To investigate the feasibility and efficacy of a multicomponent nonpharmacological approach, the authors studied a sample of 14 adults with worsening cognition and everyday functioning who were no longer manageable by their family or staff in day centers or, and who were relocated in a model

special care unit (SCU) designed to proactively accommodate the needs of people with ID and dementia. Baseline level and rate of decline across a 3-year period were assessed by means of the Dementia Questionnaire for Persons with Intellectual Disabilities and compared to two control groups not in dementia-capable programs matched for age, sex, and severity of ID. After 3 years, the authors found some improvement in cognition and stabilization in everyday functioning and behaviors in the SCU residents and a worsening in the control groups. The authors noted that enrollment in a dementia-capable program facilitated daily practice of residents' residual skills and abilities, enhancing their memory and verbal communication, that the prosthetic environment contributed to activity maintenance and appropriate intellectual challenges, and that the greater participation on an individual level added to the skill maintenance. Although the interpretation of these positive findings is not straightforward, they confirm the validity of this "in-place progression" model and provide a platform for continuing progress in person-centered services and care for aging persons with ID.

De Vreese, L.P. Mantesso, U., de Bastiani, E., Marangoni, A., & Gomiero, T.
Psychometric evaluation of the Italian version of the AADS questionnaire: A caregiver-rated tool for the assessment of behavioral deficits and excesses in persons with intellectual disabilities and dementia
International Psychogeriatrics, 2011, 23, 1124-1132.
Abstract: The aim of this study was to verify the reliability and validity of the Italian version of the Assessment for Adults with Developmental Disabilities (AADS-I), the only available measure specifically designed to assess the frequency, management difficulties and impact on the quality of life (QoL) of positive and negative non-cognitive symptoms in persons with intellectual disabilities (ID) and dementia. AADS-I was administered to professional carers of 63 aging ID individuals. We computed the internal consistency separately of the frequency, management difficulty and effect on the QoL subscales of Behavioral Excesses and Behavioral Deficits and their inter-rater and test-retest reliabilities. Homogeneity of AADS-I was found to range from good to excellent: Cronbach's a coefficients were 0.77, 0.83 and 0.82, respectively for frequency, management difficulty and effect on the QoL of Behavioral Excesses, and 0.82, 0.76 and 0.79 of Behavioral Deficits. Intraclass correlation coefficients (ICC) between two independent carers were 0.67, 0.79 and 0.73 and 0.67, 0.67 and 0.67 for frequency, management difficulty and effect on the QoL of Behavioral Excesses and Deficits, respectively. Corresponding ICC for test-retest reliability were 0.80, 0.75, 0.78 and 0.70, 0.81, 0.81. Age, sex and typology of ID did not correlate with the AADS-I subscale scores, whereas the severity of ID related only with the frequency subscale of Behavioral Deficits. This subscale also correlated with the Dementia Questionnaire for Persons with Intellectual Disabilities. Behavioral deficits are more frequent in subjects with dementia. These results confirm the reliability and validity of the Italian version of AADS.

De Vreese, L.P., Uberti, M., Mantesso, U., De Bastiani, E., Weger, E., Marangoni, A.C., Weiner, M.F. & Gomiero, T.
Measuring quality of life in intellectually disabled persons with dementia with the Italian version of the quality of life in late-stage dementia (QUALID) scale.
Journal of Alzheimer's Disease and Parkinsonism, 2012, 2:104e.
doi:10.4172/2161-0460.1000104
Abstract: The aim of this study is to verify a cross-cultural adaptation of an Italian version of the Quality of Life in Late-Stage Dementia (QUALID) scale in a sample of aging people with intellectual disabilities (ID). The QUALID was translated according to standardized procedures. Internal consistency was analyzed using Cronbach's alpha. A Principal Component Analysis verified its multidimensionality. Inter-rater and test-retest reliabilities were also assessed using the Intraclass Correlation Coefficient (ICC). Convergent validity was probed by Spearman's correlations among the QUALID score and the six sub-scores of the Assessment for Adults with Development Disabilities (AADS), a proxy-based questionnaire rating behavioral excesses and deficits commonly found in people with intellectual disabilities and dementia. Clinical validity was assessed by comparing QUALID scores obtained by subjects with and without dementia using the Mann-Whitney U test. A total of 40 adults/older people with ID at five ID-specific centers in the province of Trento and Cremona participated in the study. Findings show optimal levels of internal consistency (a = 0.80) and confirm the factors identified in the Spanish validation study (symptoms of discomfort, positive social interaction and depression). The scale has high inter-rater (ICC = 0.95) and good test-retest reliabilities (ICC = 0.89). The total QUALID score correlates significantly with the AADS sub-scores for behavioral excesses, but does not differ between individuals with and without dementia, though two out of the three identified factor scores are significantly higher in the dementia subgroup. The authors conclude that the Italian version of the QUALID is a reliable and valid instrument for estimating quality of life in aging adults with ID and dementia.

Deb, S., Hare, M.A., Prior, L., & Bhaumik, S.
Dementia screening questionnaire for individuals with intellectual disabilities.
British Journal of Psychiatry, 2007, 190, 440-444.
doi:10.1192/bjp.bp.106.024984.
Abstract: Many adults with Down syndrome develop Alzheimer's dementia relatively early in their lives, but accurate clinical diagnosis remains difficult. The authors set out too develop a user-friendly observer-rated dementia screening questionnaire with strong psychometric properties for adults with intellectual disabilities. They used qualitative methods to gather information from carers of people with Down syndrome about the symptoms of dementia. This provided the items for the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) which was then tested for its psychometric properties. The DSQIID was administered to carers of 193 adults with Down syndrome, 117 of whom were examined by clinicians who confirmed a diagnosis of dementia for 49 according to modified ICD-10 criteria. They established that a total score of 20 provides maximum sensitivity (0.92) and optimum specificity (0.97) for screening. The DSQIID has sound internal consistency ($\propto$=0.91) for all its 53 items, and good test-retest and interrater reliability. The authors established a good construct validity by dividing the questionnaire items into four factors. The authors conclude that the DSQIID is valid, reliable and user-friendly observer-rated questionnaire for screening for dementia among adults with Down syndrome.

Deb, S., Hare, M. & Prior, L.
Symptoms of dementia among adults with Down's syndrome: a qualitative study.
Journal of Intellectual Disability Research, 2007, 51, 726-739.
DOI:10.1111/j.1365-2788.2007.00956.x
Abstract: Dementia is common among adults with Down's syndrome (DS); yet the diagnosis of dementia, particularly in its early stage, can be difficult in this population. One possible reason for this may be the different clinical manifestation of dementia among people with intellectual disabilities. The aim of this study was to map out the carers' perspective of symptoms of dementia among adults with DS in order to inform the development of an informant-rated screening questionnaire. Unconstrained information from carers of people with DS and dementia regarding the symptoms, particularly the early symptoms of dementia, was gathered using a qualitative methodology. Carers of 24 adults with DS and dementia were interviewed. The interviews were recorded and fully transcribed. The transcripts were then analyzed using qualitative software. There appeared to be many similarities in the clinical presentation of dementia in adults with DS and the non-intellectually disabled general population. Like in the non-intellectually disabled general population, forgetfulness especially, impairment of recent memory combined with a relatively intact distant memory and confusion were common, and presented early in dementia among adults with DS. However, many 'frontal lobe'-related symptoms that are usually manifested later in the process of dementia among the general population were common at an early stage of dementia among adults with DS. A general slowness including slowness in activities and speech, other language problems, loss of interest in activities, social withdrawal, balance problems, sleep problems, loss of pre-existing skills along with the emergence of emotional and behavior problems were common among adults with DS in our study. This study highlighted the similarities in the clinical presentation of dementia among the general population and people with DS with a particular emphasis on the earlier appearance of symptoms associated with the frontal lobe dysfunction among adults with DS.

Deb, S.S. , Strydom, A., Hithersay, T., Gomiero, T., De Vreese,. L.P., Janicki, M.P., Jokinen, N.S., Service, K.
Dementia in People with Intellectual Disabilities. In: Bertelli, M.O., Deb, S., Munir, K., Hassiotis, A., Salvador-Carulla, L. (eds) Textbook of Psychiatry for Intellectual Disability and Autism Spectrum Disorder., 2022.
Springer, Cham. https://doi.org/10.1007/978-3-319-95720-3_28
Abstract: With the increasing life expectancy in the last five decades, people with intellectual disabilities (ID) are exposed to the risk of age-related neurodegenerative disorders including dementia. The prevalence of dementia is increased in people with ID compared with that in the general population. People with Down syndrome (DS) are at even a higher risk of developing Alzheimer's dementia (AD) compared with non-DS people with ID. However, there are

difficulties in making an early and accurate diagnosis of dementia in individuals with ID and the screening instruments such as the Mini Mental Status Examination that are used in the general population often show floor effects when used for individuals with ID because of their pre-existing cognitive impairment, the level of which varies depending on the severity of ID. Both informant-rated and direct neuropsychological tests have been used for the case detection of dementia for individuals with ID. However, direct neuropsychological tests cannot be used for those who have severe ID and their validity could still be questionable in a number of cases of mild to moderate ID. There are many similarities and some differences in the clinical manifestation of dementia in individuals with ID and the non-ID general population. Impaired recent memory and confusion in the context of relatively intact distant memory are likely to be the early symptoms in individuals with ID who have a mild ID, whereas loss of skills and change in behaviour are likely to be the early features for those with more severe ID. Many symptoms, including features of `frontal lobe dysfunction' that tend to appear late in dementia in the general population, may appear early in individuals with ID and DS. Ideally, individuals with ID should be screened for signs of dementia from before the age of 30/35. A multi-disciplinary approach should be taken for diagnosis of dementia in individuals with ID using a combination of informant-rated scales and neuropsychological tests in a longitudinal fashion over time. Important differential diagnoses include hypothyroidism, depression, and sensory impairment. Assessment should include physical, psychological, and social aspects including appropriate examinations and investigations. The evidence-base for the pharmacological management of dementia in ID and DS is poor, which does not allow to draw any definitive conclusion about their efficacy in this population. Therefore, the same pharmacological regime that is used in the non-ID population along with non-pharmacological interventions should be considered for people with ID.

Dekker, A.D., Coppus, A.M.W., Strydom, A., Vermeiren, Y., Grefelman, S., Eleveld, J., Beugelsdijkm G., ... De Deyn, P.P.
Behavioural and psychological symptoms of dementia in Down syndrome: European development of the novel BPSD-DS evaluation scale
European Neuropsychopharmacology, 2016 (Oct.), 26, S641 [Presentation at ECNP Congress 2016, Vienna, Austria] DOI: 10.1016/S0924-977X(16)31739-4
Abstract: Core to the dementia are behavioral and psychological symptoms of dementia (BPSD), an umbrella term for various behavioural changes associated with the presence of dementia, such as apathy, anxiety, disinhibition, irritability and psychosis. BPSD are associated with increased suffering, a reduced quality of life, earlier institutionalization and accelerated cognitive decline for patients, and increased caregiver burden. Although accurate recognition of BPSD may increase acceptance and understanding, enable adaptive caregiving and, importantly, allow for therapeutic interventions, BPSD have not been comprehensively evaluated in the Down syndrome (DS) population. BPSD are extensively assessed in AD patients in the general population using focussed behavioural scales, such as the Neuropsychiatric Inventory. However, not a single behavioural assessment scale has been adapted for the DS population, thus not considering the DS-specific circumstances. The authors sought to develop an adapted evaluation scale for comprehensive assessment of BPSD in DS, taking pre-existing behaviour and limitations associated with intellectual disability into account. A multidisciplinary collaboration of major DS expertise centers in Europe has been established to develop the novel BPSD-DS evaluation scale. Development consisted of a series of phases, starting with a thorough evaluation of the rather limited knowledge on BPSD in DS and extensive expert feedback rounds to identify relevant behavioural items. A structured, informant interview has been adopted, since people with DS often have difficulty to verbalize their emotions and feelings. Using defined scoring definitions for frequency, severity and caregiver burden at two periods of time, the scales enables identification of specific changes over time, i.e. disentangling behavioural alterations from characteristic behaviour that has always been typical for an individual. Multiple optimization rounds and a pilot study yielded the final version that is tested until summer 2016 in a multi-center European validation phase including approx. 250 DS individuals with and without dementia. The BPSD-DS evaluation scale exclusively focuses on behaviour, including 83 items in twelve clusters: anxiety & nervousness, sleep disturbances, irritability, obstinacy, agitation & stereotypical behaviour, aggression, apathy & aspontaneity, depression, delusions, hallucinations, disinhibition & sexual behaviour, and eating and drinking behaviour. Scale development, the final version, as well as the first results from the validation phase will be presented. Current clinical experiences and preliminary results for interrater reliability (>.90) are promising. BPSD have been largely neglected in DS. The novel BPSD-DS evaluation scale is the first comprehensive tool for its systematic assessment in the DS population, and serves a three-fold purpose in the future: (1) to monitor behavioural changes in daily care, (2) to identify early BPSD in DS in longitudinal research studies, and (3) to assess behavioural outcome measures in clinical trials for dementia in DS.

Dekker, A.D., Sacco, S., Carfi, A., Benejam, B., Vermeiren, Y., Beugelsdijk, G., Schippers, M., Hassefras, L,, Eleveld, J,, Grefelman, S,, Fopma, R,, Bomer-Veenboer, M,, Boti, M,, Oosterling, G.D.E, Scholten, E., Tollenaere, M., Checkley, L., Strydom, A., Van Goethem, G., Onder, G., Blesa, R., Zu Eulenburg, C., Coppus, A.M.W., Rebillat, A.S., Fortea, J., De Deyn, P.P.
The Behavioral and Psychological Symptoms of Dementia in Down Syndrome (BPSD-DS) Scale: Comprehensive assessment of psychopathology in Down syndrome
Journal of Alzheimers Disease, 2018, 63(2), 797-819. doi: 10.3233/JAD-170920.
Abstract: People with Down syndrome (DS) are prone to develop Alzheimer's disease (AD). Behavioral and psychological symptoms of dementia (BPSD) are core features, but have not been comprehensively evaluated in DS. In a European multidisciplinary study, the novel Behavioral and Psychological Symptoms of Dementia in Down Syndrome (BPSD-DS) scale was developed to identify frequency and severity of behavioral changes taking account of life-long characteristic behavior. 83 behavioral items in 12 clinically defined sections were evaluated. The central aim was to identify items that change in relation to the dementia status, and thus may differentiate between diagnostic groups. Structured interviews were conducted with informants of persons with DS without dementia (DS, n = 149), with questionable dementia (DS+Q, n = 65), and with diagnosed dementia (DS+AD, n = 67). First exploratory data suggest promising interrater, test-retest, and internal consistency reliability measures. Concerning item relevance, group comparisons revealed pronounced increases in frequency and severity in items of anxiety, sleep disturbances, agitation & stereotypical behavior, aggression, apathy, depressive symptoms, and eating/drinking behavior. The proportion of individuals presenting an increase was highest in DS+AD, intermediate in DS+Q, and lowest in DS. Interestingly, among DS+Q individuals, a substantial proportion already presented increased anxiety, sleep disturbances, apathy, and depressive symptoms, suggesting that these changes occur early in the course of AD. Future efforts should optimize the scale based on current results and clinical experiences, and further study applicability, reliability, and validity. Future application of the scale in daily care may aid caregivers to understand changes, and contribute to timely interventions and adaptation of caregiving.

Dekker, A.D., Strydom, A., Coppus, A,M,W., Nizetic, D., Vermeiren,, Y., Naude, P.J.W., Van Dam, D., Potier, M-C., Fortea, J., De Deyn, P.P.
Behavioural and psychological symptoms of dementia in Down syndrome: Early indicators of clinical Alzheimer's disease?
Cortex, 2015, 75, 36-61. doi: 10.1016/j.cortex.2015.07.032. Epub 2015 Aug 13.
Abstract: Behavioral and psychological symptoms of dementia (BPSD) are a core symptom of dementia and are associated with earlier institutionalization and accelerated cognitive decline for adults with Down syndrome (DS) and increased caregiver burden. Despite the extremely high risk for DS individuals to develop dementia due to Alzheimer's disease (AD), BPSD have not been comprehensively assessed in the DS population. Due to the great variety of DS cohorts, diagnostic methodologies, sub-optimal scales, covariates and outcome measures, it is questionable whether BPSD have always been accurately assessed. However, accurate recognition of BPSD may increase awareness and understanding of these behavioral aberrations, thus enabling adaptive caregiving and, importantly, allowing for therapeutic interventions. Particular BPSD can be observed (long) before the clinical dementia diagnosis and could therefore serve as early indicators of those at risk, and provide a new, non-invasive way to monitor, or at least give an indication of, the complex progression to dementia in DS. This review found that various BPSD appear to be altered in demented DS individuals, but study results have not always been consistent. From childhood to adulthood, externalizing behavior likely decreases and internalizing behavior increases. Frontal lobe symptoms have been suggested as early signs of AD in DS. Disinhibition and apathy, as well as executive dysfunction, seem to be omnipresent in the prodromal phase, whereas reports are inconsistent for depression. Regarding activity disturbances, studies indicated decreasing hyperactivity levels towards adulthood. Excessive activity in demented DS individuals should be a fairly easy observable sign, however, general slowness has been reported and apathy itself might cause reduced activity. Agitation appears to be more prevalent in demented than in non-demented DS individuals,

but reports on aggression are inconsistent, though aggression seems to be reduced in the overall DS population. Sleep disturbances are markedly present in both demented and non-demented DS individuals. Although sleep disorders may not yet differentiate between those with and without AD, they are important to consider as such sleep disorders may aggravate cognitive decline and BPSD.

Dekker, A.D., Wissing, M.B.G., Ulgiati, A.M., Bijl, B., van Gool, G., Groen, M.R., Grootendorst, E.S., van der Wal, I.A., Hobbelen, J.S.M., De Deyn, P.P., & Waninge, A.
Dementia in people with severe or profound intellectual (and multiple) disabilities: Focus group research into relevance, symptoms and training needs. *Journal of Applied Research in Intellectual Disability*, 2021, Jul 2. doi: 10.1111/jar.12912. Online ahead of print.
Abstract: Differentiating dementia from baseline level of functioning is difficult among people with severe/profound intellectual (and multiple) disabilities. Moreover, studies on observable dementia symptoms are scarce. This study examined (a) the relevance of dementia diagnosis, (b) observable symptoms and (c) training/information needs. Four explorative focus groups were held with care professionals and family members who have experience with people with severe/profound intellectual (and multiple) disabilities (≥40 years) and decline/dementia. Thematic analysis showed that participants wanted to know about a dementia diagnosis for a better understanding and to be able to make informed choices (question 1). Using a categorization matrix, cognitive and behavioural changes were shown to be most prominent (question 2). Participants indicated that they needed enhanced training, more knowledge development and translation, and supportive organizational choices/policies (question 3). Timely identifying/diagnosing dementia allows for a timely response to changing needs. This requires a better understanding of symptoms.

Dekker, A.D., Ulgiati, A.M., Groen, H., Boxelaar, V.A., Sacco, S., Falquero, S., Carfi, A.., di Paola, A., Benejam, B., Valldeneu, S., Fopma, R., Oosterik, M,. Hermelink, M., Beugelsdijk, G., Schippers, M., Henstra, H., Scholten-Kuiper, M., Willink-Vos, J., de Ruiter, L., Willems, L., Loonstra-de Jong, A., Coppus, A.M.W., Tollenaere, M., Fortea, J., Onder, G.,Rebillat, A-S., Van Dam, D., De Deyn, P.P.
The Behavioral and Psychological Symptoms of Dementia in Down Syndrome Scale (BPSD-DS II): Optimization and Further Validation.
Journal of Alzheimer's Disease, 2021, 82(3), 1371-1371, 2021
Abstract: [Corrections: In Figure 8, p. 1519, the second graph (item 10.3) reads: "10.3 making inappropriate comments". `Making inappropriate comments' is the description belonging to item 10.2 of the BPSD-DS II. Instead, the second graph (item 10.3) should read: "10.3 loss of decorum".]

Dekker, A.D., Ulgiati, A.M., Groen, H., Boxelaar, V.A., Sacco, S., Falquero, S., Carfi, A.., di Paola, A., Benejam, B., Valldeneu, S., Fopma, R., Oosterik, M,. Hermelink, M., Beugelsdijk, G., Schippers, M., Henstra, H., Scholten-Kuiper, M., Willink-Vos, J., de Ruiter, L., Willems, L., Loonstra-de Jong, A., Coppus, A.M.W., Tollenaere, M., Fortea, J., Onder, G.,Rebillat, A-S., Van Dam, D., De Deyn, P.P.
'The Behavioral and Psychological Symptoms of Dementia in Down Syndrome Scale (BPSD-DS II): Optimization and further validation'.
Journal of Alzheimer's Disease, 2021, 81(4), 1505-1527. DOI 10.3233/JAD-201427
Abstract: People with Down syndrome (DS) are at high risk to develop Alzheimer's disease dementia (AD). Behavioral and psychological symptoms of dementia (BPSD) are common and may also serve as early signals for dementia. However, comprehensive evaluation scales for BPSD, adapted to DS, are lacking. Therefore, we previously developed the BPSD-DS scale to identify behavioral changes between the last six months and pre-existing life-long characteristic behavior. To optimize and further study the scale (discriminative ability and reliability) in a large representative DS study population. Optimization was based on item irrelevance and clinical experiences obtained in the initial study. Using the shortened and refined BPSD-DS II, informant interviews were conducted to evaluate 524 individuals with DS grouped according to dementia status: no dementia (DS, N = 292), questionable dementia (DS + Q, N = 119), and clinically diagnosed dementia (DS + AD, N = 113). Comparing item change scores between groups revealed prominent changes in frequency and severity for anxious, sleep-related, irritable, restless/stereotypic, apathetic, depressive, and eating/drinking behavior. For most items, the proportion of individuals displaying an increased frequency was highest in DS + AD, intermediate in DS + Q, and lowest in DS. For various items within sections about anxious, sleep-related, irritable, apathetic, and depressive behaviors, the proportion of individuals showing an increased frequency was already substantial in DS + Q, suggesting that these changes may serve as early signals of AD in DS. Reliability data were promising. The optimized scale yields largely similar results as obtained with the initial version. Systematically evaluating BPSD in DS may increase understanding of changes among caregivers and (timely) adaptation of care/treatment.

Dennehy, H., Allen AP, McGlinchey E, Buttery N, García-Domínguez L, Chansler R, Corr C, Dunne P, Kennelly S, Daly L, McCallion P, McCarron M.
A scoping review of post-diagnostic dementia supports for people with intellectual disability.
Aging & Mental Health, 2022 Oct 11:1-10. doi: 10.1080/13607863.2022.2130171. Epub ahead of print. PMID: 36218056.
Abstract: People with intellectual disability, particularly people with Down syndrome, are at an increased risk for early-onset dementia, in comparison to people without an intellectual disability. The aim of this review was to scope the current landscape of post-diagnostic dementia supports for people with intellectual disability.Method: A systematic search of five electronic databases (CINAHL, Medline, PsycArticles, PsycInfo and Web of Science) was conducted for this scoping review. Results were screened independently by two reviewers, with a third reviewer for arbitration where necessary. Forty-two studies met the inclusion criteria, and relevant information was extracted. The articles included focused on the experiences of people with intellectual disability and dementia, as well as the role of carers, family members and staff. Key themes included ageing in place, environmental supports for people with intellectual disability and dementia, dementia-specific interventions and therapies, as well as the feasibility of these interventions. Besides the studies that focussed on these themes, other studies focussed on staff training and family supports. This review highlights the importance of implementing timely and appropriate post-diagnostic supports for people living with intellectual disability and dementia. More controlled trials are required on post-diagnostic dementia supports for people with intellectual disability.

Devenny, D.A., Krinsky-McHale, S.J., Sersen, G., & Silverman, W.P.
Sequence of cognitive decline in dementia in adults with Down's syndrome.
Journal of Intellectual Disability Research 2000, 44(6), 654-665. doi:10.1046/j.1365-2788.2000.00305.x
Abstract: Because of lifelong intellectual deficits, it is difficult to determine the earliest signs and characteristics of age-associated decline and dementia among adults with Down syndrome. In a longitudinal study in which all participants were healthy at the time of their entry into the study, the present authors compared the amount of decline on the subtests of the WISC-R to determine the sequence of cognitive decline associated with varying stages of dementia. Twenty-two individuals with varying degrees of cognitive decline were compared to 44 adults with DS who have remained healthy. All participants functioned in the mild or moderate range of intellectual disability at initial testing. On each subtest of the WISC-R, the amount of change experienced by the healthy participants over the study period was compared to the amount of change found for each of the groups with decline. Out of the individuals who showed declines, 10 adults with DS were classified as having 'questionable' decline based on the presence of memory impairment, and five and seven adults with DS were classified as in the 'early stage' and 'middle stage' of DAT, respectively, based on the presence of memory impairment, score on the Dementia Scale for Down Syndrome and a physician's diagnosis. It was found that participants who were identified as 'questionable', in addition to the memory loss that determined their classification, also showed significant declines on the Block Design and Coding subtests. The five adults in the early stage of dementia showed declines on these subtests, and in addition, on the Object Assembly, Picture Completion, Arithmetic and Comprehension subtests. The seven adults in the middle stage of dementia showed declines on these subtests, plus declines on Information, Vocabulary and Digit Span subtests. The Picture Arrangement and Similarities subtests were not useful in distinguishing between the groups because of baseline floor effects for a substantial proportion of participants. The present longitudinal study showed a sequence of cognitive decline associated with DAT, beginning with a possible 'pre-clinical' stage, and progressing through the early and middle stages. This approach begins to define the sequence of declining cognitive capacities that contributes to the observed functional deterioration caused by Alzheimer's disease and that is likely to reflect the involvement of cortical areas as the disease progresses.

Dick, M.B., Doran, E., & Phalen, M., & Lott,I.
Cognitive profiles on the Severe Impairment Battery are similar in Alzheimer disease and Down syndrome with dementia
Alzheimer Disease and Associated Disorders, 2015, Dec 22 [Epub ahead of print]
Abstract: Previous research has revealed similarities in the neuropathology, clinical presentation, and risk factors between persons with Alzheimer disease from the general population (GP-AD) and those with Down syndrome (DS-AD). Less is known, however, about the extent of similarities and differences in the cognitive profiles of these 2 populations. Fifty-one moderate to severely demented GP-AD and 59 DS-AD individuals participated in this study which compared the cognitive profiles of these 2 populations on the Severe Impairment Battery (SIB), controlling for sex as well as level of functional ability using a modified version of the Bristol Activities of Daily Living Scale. Overall, the neuropsychological profiles of the higher-functioning individuals within the DS-AD and advanced GP-AD groups, as represented by mean difference scores on the SIB as a whole and across the 9 separate cognitive domains, were very similar to one another after adjusting for sex and functional impairment. To our knowledge, this is the first study to directly compare the cognitive profiles of these 2 populations on the SIB. Findings suggest that the underlying dementia in GP-AD and DS-AD may have corresponding and parallel effects on cognition.

Dillane, I., & Dood, O.
Nursing people with intellectual disability and dementia experiencing pain: An integrative review.
Journal of Clinical Nursing, 2019, 10.1111/jocn.14834, 28(13-14), 2472-2485
Abstract: To explore the current evidence of nurses caring for people with intellectual disability and dementia who experience pain. People with intellectual disability are aging and are experiencing age-related health conditions including dementia and conditions associated with pain, but at an earlier age. Addressing the needs of people with intellectual disability who develop dementia is a new challenge for nurses. The authors undertook an integrative literature review and a systematic search of databases: CINAHL, MEDLINE, PsycINFO, Cochrane, EMBASE, Academic Search Complete, Scopus and Web of Science between 27 October 2017-7 November 2017. Hand searching and review of secondary references were also undertaken. Quality appraisal (Crowe Critical Appraisal Tool), thematic data analysis (Braun and Clarke, Qualitative Research in Psychology, 3, 2006, 77) and reporting using the PRISMA guidelines. Seven papers met the inclusion criteria, and three themes emerged from this review: nurses knowledge of ageing, dementia and pain; recognising pain in people with intellectual disability and dementia; and the role of nurse education. People with intellectual disability and dementia have difficulty communicating their pain experience compounded by pre-existing communication difficulties. A pain experience can present similar to behavioral and psychological symptoms of dementia, and diagnostic overshadowing often occurs whereby a pain need is misinterpreted as behavioral and psychological symptoms resulting in inappropriate treatment. Nurses need greater knowledge about the presence of pain and potential causes in people with intellectual disability and dementia, and education can be effective in addressing this knowledge deficit. Pain assessment tools for people with intellectual disability and dementia need to include behavioural elements, and baseline assessments are required to identify changes in presentation. Nurses need to recognize and respond to pain based on the evidence in order to deliver quality care.

Dillenburger, K., & McKerr,L.
'How long are we able to go on?' Issues faced by older family caregivers of adults with disabilities
British Journal of Learning Disabilities, 2011 March, 39(1), 29-38. https://doi.org/10.1111/j.1468-3156.2010.00613.x
Abstract: Research-informed policy and practice is needed for older caregivers of adult sons/daughters with disabilities. These caregivers are often under tremendous stress because of failing health, financial pressures, bereavement and worry about the future of their sons/daughters. Twenty-nine older parents/caregivers of 27 adults with intellectual and/or developmental disabilities were interviewed to explore their views and experiences regarding long-term care and service arrangements, health and psychological needs and 'future planning'. Findings show a severe lack of support, respite care and future planning which causes high stress levels for caregivers. Policy makers and researchers working in this field need to take into consideration the needs of older caregivers when making future plans for adults with disabilities.

Dodd, K.
Commentary on "Estimating the number of people with Down's syndrome in Scotland and the cohort at elevated risk of early onset dementia"
Tizard Learning Disability Review, 2017, 22(3), 172-176. 10.1108/TLDR-04-2017-0019
Abstract: The purpose of this paper is to consider the implications for people with Down's syndrome and their families of identifying those people who are at risk of developing dementia from the research study "Estimating the number of people with Down's syndrome in Scotland and the cohort at elevated risk of early onset dementia". The commentary is based on a review of the associated literature. Estimating the numbers is important but has serious implications for people who have an elevated risk and their families. Preparation and ongoing support and planning are vital to ensure that quality of life is maintained as dementia is identified and progresses.

Dodd, K.
Supporting people with Down's syndrome and dementia
Tizard Learning Review, 2003, 8(4), 14-18
Abstract: Brief review of literature and concepts dealing with the prevalence of dementia among people with Down syndrome in England, ethical issues in assessment and diagnosis, the value of early diagnosis, and an explication of service options and management strategies. Review concludes with a prognosis for services in the future.

Dodd, K., Watchman, K., Janicki, M.P., Coppus, A., Gaertner, C., Fortea, J., Santos, F. H., Keller, S.M., & Strydom. A.
Consensus Statement of the International Summit on Intellectual Disability and Dementia related to post-diagnostic support
Aging & Mental Health, 2018, 22(11), 1406-1415. DOI: 10.1080/13607863.2017.1373065
Abstract: Post diagnostic support (PDS) has varied definitions within mainstream dementia services and different health and social care organizations, encompassing a range of supports that are offered to adults once diagnosed with dementia until death. An international summit on intellectual disability and dementia held in Glasgow, Scotland in 2016 identified how PDS applies to adults with an intellectual disability and dementia. The Summit proposed a model that encompassed seven focal areas: post-diagnostic counseling; psychological and medical surveillance; periodic reviews and adjustments to the dementia care plan; early identification of behavior and psychological symptoms; reviews of care practices and supports for advanced dementia and end of life; supports to carers/ support staff; and evaluation of quality of life. It also explored current practices in providing PDS in intellectual disability services. The Summit concluded that although there is limited research evidence for pharmacological or non-pharmacological interventions for people with intellectual disability and dementia, viable resources and guidelines describe practical approaches drawn from clinical practice. Post diagnostic support is essential, and the model components in place for the general population, and proposed here for use within the intellectual disability field, need to be individualized and adapted to the person's needs as dementia progresses. Recommendations for future research include examining the prevalence and nature of behavioral and psychological symptoms (BPSD) in adults with an intellectual disability who develop dementia, the effectiveness of different non-pharmacological interventions, the interaction between pharmacological and non-pharmacological interventions, and the utility of different models of support.

Donaldson S.
Work stress and people with Down syndrome and dementia.
Down's Syndrome, Research and Practice, 2002, 8(2), 74-78.
Abstract: Author assessed how staff ratings of challenging behavior for people with Down syndrome and dementia affected the self-reported well-being of care staff. Data were collected from 60 care staff in 5 day centers in a large city in England. The data were collected by use of a questionnaire. There was no significant difference between those who cared for individuals with Down syndrome and dementia and those caring for service users with other non-specified learning disabilities without dementia, regarding their self-reported well-being. Self-reported well-being did correlate with staff rating of challenging behavior in both those who cared for people with Down syndrome and dementia and those who did not care for such service users, with well-being declining as perceived challenging behavior increased. The findings indicate that challenging behavior prevention and reduction may be of benefit to both service users and care staff well-being.

Dunne, P., Reilly, E., Judge, R., Lowe, F. & McCarron, M.
Giving meaning to life - the role of digital life Stories in supporting people with intellectual disability and dementia
Journal of Intellectual Disability Research, 2019, 63(8), 643.
Abstract: Over a four-year period individuals with an intellectual disability (ID) in a large service provider in the Republic of Ireland were supported to create their personalized digital life story using selected multi media apps. An easy read survey was distributed to 380 people with ID to gauge their readiness and interest in engaging in digital life story. A bespoke training course was developed to support the introduction of digital life story activities. The use of the personalized digital life stories by the individuals and their support staff, family and social network was captured through recorded observations and individual use was tracked through a developed audit tool. Over the four-year period eighty people with an ID commenced their digital life story. Case studies within this cohort showed a variety of use which varied depending on degree of ID and stage of dementia. In all cases life stories were instrumental to enhanced communication and social interactions. Key factors in uptake and sustainability were staff training, iPad clubs and champions across the organization Implications: Digital life stories were key in supporting meaningful interactions across the continuum of dementia both in day to day interactions with family, staff, volunteers, and peers and as a tool for social engagement through digital life story clubs.

Eady, N., Sheehan, R., Rantell, K., Sinai, A., Bernal, J., Bohnen, I., Courtney, K., Dodd, K., Gazizova, D., Hassiotis, A., ... Strydom, A.
Impact of cholinesterase inhibitors or memantine on survival in adults with Down syndrome and demenita: Clincal cohort study.
British Journal of Psychiatry, 2018 Mar, 212(3), 155-160.
doi:10.1192/bjp.2017.21
Abstract: There is little evidence to guide pharmacological treatment in adults with Down syndrome and Alzheimer's disease. Authors investigated the effect of cholinesterase inhibitors or memantine on survival and function in adults with Down syndrome and Alzheimer's disease. This was a naturalistic longitudinal follow-up of a clinical cohort of 310 people with Down syndrome diagnosed with Alzheimer's disease collected from specialist community services in England. Median survival time (5.59 years, 95% CI 4.67-6.67) for those on medication (n = 145, mainly cholinesterase inhibitors) was significantly greater than for those not prescribed medication (n = 165) (3.45 years, 95% CI 2.91-4.13, log-rank test P<0.001). Sequential assessments demonstrated an early effect in maintaining cognitive function. Cholinesterase inhibitors appear to offer benefit for people with Down syndrome and Alzheimer's disease that is comparable with sporadic Alzheimer's disease; a trial to test the effect of earlier treatment (prodromal Alzheimer's disease) in Down syndrome may be indicated.

Ebbing, K.; von Gunten, A.; Guinchat, V.; Georgescu, D.; Bersier, T.; Moad, D.; Verloo, H.
Barriers Facing Direct Support Professionals When Supporting Older Adults Presenting with Intellectual Disabilities and Unusual Dementia-Related Behavior: A Multi-Site, Multi-Methods Study.
Disabilities, 2022, 2(4), 662-680. https://doi.org/10.3390/disabilities2040047
Abstract: Increased life expectancy among people with intellectual disabilities (ID) raises the risk of their diagnosis being superimposed by behavioral and psychological symptoms of dementia (BPSD). The difficulties facing direct support professionals dealing with this is an emerging, under-investigated issue. The study investigates direct support professionals' perceptions and experiences of their daily support for aging people with ID presenting with superimposed BPSD. Twenty-four direct support professionals from long-term care facilities responded to clinical vignettes and attended focus groups conducted to investigate perceptions and lived experiences of the barriers and struggles they faced. Direct support professionals' reactions to vignettes revealed their difficulties recognizing BPSD superimposed on the known challenging behaviors of people with ID. Focus groups highlighted daily struggles with BPSD, the lack of knowledge about detecting and dealing with them, and associated somatic and psychopathological diseases of aging. Improved knowledge transfer about good practices for person-centered support to aging people with ID presenting with BPSD is strongly recommended.

Eisner, D.A.
Down's syndrome and aging: Is senile dementia inevitable?
Psychological Reports 1983, 52(1), 119-124.
https://doi.org/10.2466/pr0.1983.52.1.119
Abstract: Numerous studies have reported that in elderly Down's Syndrome individuals there is a high preponderance of senile dementia. An examination of these investigations shows that, while there is accelerated neurological aging, there is not a high incidence of behavioral or overt senile dementia. Changes in cognitive functioning for Down's Syndrome persons are similar to those found in non-Down's retarded populations.

Elliott-King, J., Shaw, S., Bandelow, S., Devshi, R., Kassam, S., & Hogervorst, E.
A critical literature review of the effectiveness of various instruments in the diagnosis of dementia in adults with intellectual disabilities
Alzheimers and Dementia (Amst), 2016,Jun30, 4(1), 126-148. doi: 10.1016/j.dadm.2016.06.002. eCollection 2016.
Abstract: Currently, there is no consensus on dementia diagnostics in adults with intellectual disabilities (ID). There are three types of assessments available: direct cognitive tests, test batteries, and informant reports. A systematic literature search was conducted in four databases yielding 9840 records. Relevant studies were identified and selected using predefined inclusion and exclusion criteria and then coded and classified according to assessment type. This was completed by two independent researchers, with a third consulted when discrepancies arose. The review collates diagnostic instruments and presents strengths and weaknesses. Overall 47 studies met the search criteria, and 43 instruments were extracted from the selected studies. Of which, 10 instruments were classified as test batteries, 23 were classified as direct cognitive tests, and the remaining 10 were informant reports. This review can recommend that cognitive test batteries can offer the most practical and efficient method for dementia diagnosis in individuals with ID.

Engdahl, J.M.K.
Alzheimer's disease & Down syndrome: A practical guide for caregivers.
36 pp.
Bozeman, Montana: Author [723 South 13th Street, Bozeman, MT 59715] (1995)
Abstract: Training manual developed to provide primary information about care practices for parents and other primary carers of adults with Down syndrome affected by Alzheimer's disease. Covers, in brief format, recognizing signs and symptoms, diagnostic advice, care management practice (communication, dealing with problem behaviors, helping with activities of daily living, promoting alternative activities) and help for carers.

Englund, A., Jonsson, B., Zander, C.S., Gustafsson, J,. & Annerén, G.
Changes in mortality and causes of death in the Swedish Down syndrome population
American Journal of Medical Genetics [Part A], 2013 Apr, 161A(4), 642-649, doi: 10.1002/ajmg.a.35706.
Abstract: During the past few decades age at death for individuals with Down syndrome (DS) has increased dramatically. The birth frequency of infants with DS has long been constant in Sweden. Thus, the prevalence of DS in the population is increasing. The aim of the present study was to analyze mortality and causes of death in individuals with DS during the period 1969-2003. All individuals with DS that died between 1969 and 2003 in Sweden, and all individuals born with DS in Sweden between 1974 and 2003 were included. Data were obtained from the Swedish Medical Birth Register, the Swedish Birth Defects Register, and the National Cause of Death Register. Median age at death has increased by 1.8 years per year. The main cause of death was pneumonia. Death from congenital heart defects decreased. Death from atherosclerosis was rare but more frequent than reported previously. Dementia was not reported in any subjects with DS before 40 years of age, but was a main or contributing cause of death in 30% of the older subjects. Except for childhood leukemia, cancer as a cause of death was rare in all age groups. Mortality in DS, particularly infant mortality, has decreased markedly during the past decades. Median age at death is increasing and is now almost 60 years. Death from cancer is rare in DS, but death from dementia is common.

ENIDA
Face to face: Respectful coping with dementia in older people with intellectual disability
52 minutes
Working Group on Coping with Dementia in Older People with Intellectual Disability, European Network on Intellectual Disability and Ageing [ENIDA - c/o Patricia Noonan Walsh, Ph.D., Director, Centre for the Study of Developmental Disabilities, University College Dublin, Belfield, Dublin 4, IRELAND – e-mail:

patricia.walsh@ucd.ie] (2000)
Abstract: A 52-minute video with an accompanying information booklet, which uses a number of case vignettes from France, Belgium and the Netherlands to illustrate the various symptoms and stages of dementia among older people with intellectual disability. Examples of practices to promote "respectful coping" with dementia, death and dying on the part of direct support professionals and clinicians are presented. Devised for staff training and development, Face to Face may be viewed in short segments. A version with English subtitles and English booklet is available in formats suitable for Europe and for North America. Developed with funding and support from: ENIDA, Fondation de France, the European Union, and University College Dublin, Ireland.

Ericksson, M., & Sundin, M.
Developing early detection of dementia with people with intellectual and developmental disabilities
Poster presented at the 27th Annual Conference of Alzheimer Europe, Berlin, Germany, October 3, 2017. (PO3.26)
Abstract: In Finland there is a lack of a unified approach to the early detection of dementia for people with intellectual and developmental disabilities (IDD). The aim of this presentation is to describe a currently underway collaboration project, which responds to this challenge. In our opinion, there are two essential elements in early detection of dementia with people with IDD: 1) an overall description of the psychosocial functioning of the person and 2) a screening method for dementia, which developed for persons with IDD. These two components are shortly outlined. Psychosocial functioning: Early detection of neurocognitive disorders in people with IDD is multidisciplinary teamwork. The Finnish Association of Intellectual and Developmental Disabilities (FAIDD) has published two methods for this purpose (the Toimi and the Psyto), and we recommend using these methods in the assessment of dementia in people with IDD as well. For example, it is important to distinguish the symptoms of dementia from other possible psychological disorders (like mood disorders and psychotic symptoms). Translation of the National Task Group Early Detection Screen for Dementia (NTG-EDSD): Commonly used assessment methods (like the Cerad and the Mini-Mental) may not be applicable for people with IDD. In our project, we decided to translate the NTG-EDSD into Finnish. The EDSD is already available online in many other languages (www.aadmd.org/ntg/screening). As the authors write, the NTG-EDSD is not an assessment or diagnostic instrument, but an administrative screen that can be used by people who know the client well. The Finnish version of the NTG-EDSD is being introduced into practice in 2017. FAIDD trains staff and other people working with people with IDD in using the screen within the perspective of psychosocial functioning.

Esbensen, A.J.
Health conditions associated with aging and end of life of adults with Down syndrome.
International Review of Research in Mental Retardation, 2010, 39c, 107-126. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3010180/
Abstract: Expectations for the life course of individuals with Down syndrome (DS) have changed, with life expectancy estimates increasing from 12 in 1949 to nearly 60 years of age today. Along with this longer life expectancy comes a larger population of adults with DS who display premature age-related changes in their health. There is thus a need to provide specialized health care to this aging population of adults with DS who are at high risk for some conditions and at lower risk for others. This review focuses on the rates and contributing factors to medical conditions that are common in adults with DS or that show changes with age. The review of medical conditions includes the increased risk for skin and hair changes, early onset menopause, visual and hearing impairments, adult onset seizure disorder, thyroid dysfunction, diabetes, obesity, sleep apnea and musculoskeletal problems. The different pattern of conditions associated with the mortality of adults with DS is also reviewed.

Esbensen, A.J., Boshkoff Johnson, E., Amaral, J.L., Tan, C.M., & Macks, R.
Differentiating aging among adults with Down syndrome and comorbid psychopathology.
American Journal on Intellectual and Developmental Disabilities, 2016, 121 (1), 13-24.
Abstract: Differences were examined between three groups of adults with Down syndrome in their behavioral presentation, social life/activities, health, and support needs. We compared those with comorbid dementia, with comorbid psychopathology, and with no comorbid conditions. Adults with comorbid dementia were more likely to be older, have lower functional abilities, have worse health and more health conditions, and need more support in self-care. Adults with comorbid psychopathology were more likely to exhibit more behavior problems and to be living at home with their families. Adults with no comorbidities were most likely to be involved in community employment. Differences in behavioral presentation can help facilitate clinical diagnoses in aging in Down syndrome, and implications for differential diagnosis and service supports are discussed.

Esbensen, A.J., Mailick, M.R., & Silverman, W.
Long-term Impact of parental well-being on adult outcomes and dementia status in individuals with Down syndrome.
American Journal on Intellectual and Developmental Disabilities, 2013, 118(4), 294-309.
Abstract: Parental characteristics were significant predictors of health, functional abilities, and behavior problems in adults with Down syndrome (n = 75) over a 22-year time span, controlling for initial levels and earlier changes in these outcomes. Lower levels of behavior problems were predicted by improvements in maternal depressive symptoms. Higher levels of functional abilities were predicted by prior measures of and improvements in maternal depressive symptoms. Better health was predicted by prior measures of maternal depressive symptoms, paternal positive psychological well-being, relationship quality between fathers and their adult children, and improvements in maternal positive psychological well-being. Dementia status was also predicted by parental characteristics. The study suggests the importance of the family context for healthy aging in adults with Down syndrome.

Esralew, L., Janicki, M.P., & Keller, S.M.
National Task Group Early Detection Screen for Dementia (NTG-EDSD)
Chapter in V. Prasher (ed.), *Neuropsychological Assessments of Dementia in Down Syndrome and Intellectual Disabilities*. Pp. 197-213. 2018. Doi:10.1007/978-3-319-61720-6
Abstract: Early identification of signs and symptoms of cognitive and functional decline associated with dementia is an important first step in managing the course of dementia and providing quality care. Studies show that persons with intellectual disabilities are as susceptible to the causes of dementia as are other adults, with the relative distribution of etiologies mirroring those of other adults. The challenge, as noted by the World Health Organization, is to identity early those adults susceptible and affected so that assessment and diagnostic work-ups can be undertaken. A screening tool can be used to substantiate changes in adaptive skills, behavior and cognition. With early detection, assessment and diagnosis can be carried out to determine whether cognitive changes are the result of a neuropathological process related to disease or trauma to the brain, or attributable to other causes, which may be treatable and reversible. The National Task Group Early Detection Screen for Dementia (NTG-EDSD) is an informant-based rating tool for use with adults with intellectual and developmental disabilities who are suspected of experiencing changes in thinking, behavior, and adaptive skills suggestive of mild cognitive impairment or dementia. This chapter offers background on the NTG-EDSD and describes benefits that may accrue from using a dementia screening instrument.

Evans, E., Bhardwaj, A., Brodaty, H., Sachdev, P., Draper, B., & Trollor, J.N.
Dementia in people with intellectual disability: insights and challenges in epidemiological research with an at-risk population
International Review of Psychiatry. 2013, 25(6), 755-763. doi: 10.3109/09540261.2013.866938.
Abstract: The population with intellectual disability (ID) is aging, but age-related health concerns such as dementia have received little research attention thus far. We review evidence regarding the prevalence and incidence of dementia in people with ID, and discuss some possible explanations for an increased risk, such as shared genetic risk factors, co-morbid physical and mental disorders, lifestyle factors, trauma, and lowered brain reserve. We discuss practical and theoretical challenges facing researchers in this field, before highlighting the implications of findings to date for future research and clinical care. Research on dementia in this at-risk population has the potential to help us understand dementia in general and to improve services for this group of vulnerable individuals.

Evenhuis H.M.
The natural history of dementia in Down's syndrome.
Archives of Neurology, 1990, Mar;47(3):263-7.

Abstract: In a prospective longitudinal study with death as the end point in 17 middle-aged patients with Down's syndrome, dementia was clinically diagnosed in 15 patients, by means of careful observations in daily circumstances. Autopsies were performed in 10 cases: 8 demented patients and 2 nondemented patients. Neuropathologically, Alzheimer-type abnormalities were demonstrated in 9 patients, both demented and nondemented, and combined Alzheimer-type abnormalities with infarctions were demonstrated in 1 patient. In the 14 demented patients who did not show evidence of cerebrovascular or systemic vascular disease, dementia had an early onset and was rapidly progressive (mean age at onset, 51.3 years in the moderately retarded patients and 52.6 years in the severely retarded patients; mean duration of symptoms, respectively, 4.9 and 5.2 years). Cognitive and behavioral decline corresponded to symptoms of dementia of the Alzheimer's type in patients without Down's syndrome, but often were not recognized early. In the present group of patients, there was a remarkably high incidence of gait and speech deterioration. Also, the incidence of epileptic seizures and myoclonus was about eightfold, as compared with dementia of the Alzheimer's type in patients without Down's syndrome.

Evenhuis, H.M.
The Dementia Questionnaire for People with Learning Disabilities.
Chapter in V. Prasher (ed.), *Neuropsychological Assessments of Dementia in Down Syndrome and Intellectual Disabilities*. Pp. 43-56. 2018.
10.1007/978-3-319-61720-6
Abstract: To facilitate the diagnosis of dementia in persons with intellectual disabilities (ID), based on observations of caregivers, since 1980 the Dementie Vragenlijst voor Zwakzinnigen (DVZ) has been developed by Heleen Evenhuis, ID physician, and Margeen Kengen and Harry Eurlings, behavioral therapists, all working in De Bruggen center for people with ID, Zwammerdam, the Netherlands. The Dementia Questionnaire for People with Learning Disabilities (DLD) is an English translation of this instrument. Formally known as the Dementia Questionnaire for Mentally Retarded Persons (DMR). After many years of distribution through De Bruggen, its publication has now been taken over by Harcourt Test Publishers]. In this chapter, the author reviews the development of the DMR (DLD) along with its clinical applications.

Evenhuis, H.M., Hermans, H., Hilgenkamp, T.I.M., Bastiaanse, L.P., & Echteld, M.A.
Frailty and disability in older adults with intellectual disabilities: results from the healthy ageing and intellectual disability study
Journal of the American Geriatrics Society, 2012, May, 60(5), 934-938.
doi: 10.1111/j.1532-5415.2012.03925.x.
Abstract: To obtain first insight into prevalence and correlates of frailty in older people with intellectual disability (ID). This was a population-based cross-sectional study in persons using formal ID services. Three Dutch care provider services, with 848 individuals with borderline to profound ID aged 50 and older participated in the Healthy Ageing and Intellectual Disability (HA-ID) Study. All participants underwent an extensive health examination. Frailty was diagnosed according to Cardiovascular Health Study criteria. Associations between frailty and participant characteristics were investigated using multivariate logistic regression analysis. Prevalence of frailty was 11% at age 50 to 64 and 18% at age 65 and older. Age, Down syndrome, dementia, motor disability, and severe ID were significantly associated with frailty, but only motor disability had a unique association with frailty. In a regression model with these variables, 25% of the variance of frailty was explained. At age 50 to 64, prevalence of frailty is as high as in the general population aged 65 and older (7-9%), with a further increase after the age of 65. Motor disability only partially explains frailty. Future studies should address health outcomes, causes, and prevention of frailty in this population.

Fahey-McCarthy, E., McCarron, M., Connaire, K., & McCallion, P.
Developing an education intervention for staff supporting persons with an intellectual disability and advanced dementia.
Journal of Policy and Practice in Intellectual Disabilities, 2009, 6(4), 267-275.
https://doi.org/10.1111/j.1741-1130.2009.00231.x
Abstract: Generally, staff working in settings that provide care for adults with intellectual disabilities (ID) have not received specific education with respect to extended care for terminal illnesses or late-stage dementia. Equally, staff working in specialist palliative care often are not familiar with the unique issues of supporting persons with intellectual disabilities affected by dementia. To understand care concerns with respect to supporting persons with ID and advanced dementia, and to develop, deliver, and evaluate an educational intervention with staff in ID settings and specialist palliative care services, 14 focus group interviews were conducted with staff across six ID services and one specialist palliative care provider in the greater Dublin (Ireland) area. Qualitative descriptive analysis resulted in the emergence of key themes and formed the development of an educational intervention. Pre- and posttest questionnaires assessed responses to a pilot delivery of the educational intervention. Formal feedback from staff indicated that the educational intervention was highly valued and addressed key training concerns. They agreed that the training supported "aging in place," and the preparation for a "good death" including support for staff, peers, and family in their grief and bereavement. An educational intervention in the form of a trainer manual was produced to support cross-service system in-service training on issues of addressing advanced dementia in persons with ID.

Fazio, S., Pace, D., Kallmyer, B., & Pike, J.
Alzheimer's Association towards Guidelines for Dementia Care Practice: Recommendations with emphasis on high-quality, person-centered care in long-term and community-based care settings.
Alzheimer's & Dementia, 2018, 14(4), 520-521.
https://doi.org/10.1016/j.jalz.2018.03.001
Abstract: Speaks to the Dementia Care Practice Recommendations that were developed to better define quality care across all care settings and throughout the disease course. Notes that they are intended for professional care providers who work with individuals living with dementia and their families in residential and community based care settings.

Fazio, S., Pace, D., Kallmyer, B., & Maslow, K., & Zimmerman, S.
Alzheimer's Association dementia care practice recommendations.
Gerontologist, 2018, Jan 18, 58(suppl1), S1-S9.
doi: 10.1093/geront/gnx182.
Abstract: The Alzheimer's Association 'Dementia Care Practice Recommendations' outline guidance on quality care practices based on a comprehensive review of current evidence, best practice, and expert opinion. The 'Dementia Care Practice Recommendations' were developed to better define quality care across all care settings, and throughout the disease course. They are intended for professional care providers who work with individuals living with dementia and their families in residential and community-based care settings. With the fundamentals of person-centered care as the foundation, the 'Dementia Care Practice Recommendations' posit goals for quality dementia care in the following areas: (a) person-centered care, (b) detection and diagnosis, (c) assessment and care planning, (d) medical management, (e) Information, education, and support, (f) ongoing care for behavioral and psychological symptoms of dementia, and support for activities of daily living, (g) staffing, (h) supportive and therapeutic environments, and (I) transitions and coordination of services.

Firth, N.C., Startin, C.M., Hithersay, R., Hamburg, S., Wijeratne, P.A., Mok, K.Y., Hardy, J.. Alexander, D.C., LonDownS Consortium, & Strydom, A.
Aging related cognitive ch anges associated with Alzheimer's diseaese in Down syndrome.
Annals of Clinical and Translational Neurology, 2018, May 20, 5(6), 741-751. doi: 10.1002/acn3.571. eCollection 2018 Jun.
Abstract: Individuals with Down syndrome (DS) have an extremely high genetic risk for Alzheimer's disease (AD), however, the course of cognitive decline associated with progression to dementia is ill-defined. Data-driven methods can estimate long-term trends from cross-sectional data while adjusting for variability in baseline ability, which complicates dementia assessment in those with DS. We applied an event-based model to cognitive test data and informant-rated questionnaire data from 283 adults with DS (the largest study of cognitive functioning in DS to date) to estimate the sequence of cognitive decline and individuals' disease stage. Decline in tests of memory, sustained attention/motor coordination, and verbal fluency occurred early, demonstrating that AD in DS follows a similar pattern of change to other forms of AD. Later decline was found for informant measures. Using the resulting staging model, we showed that adults with a clinical diagnosis of dementia and those with APOE 3:4 or 4:4 genotype were significantly more likely to be staged later, suggesting that the model is valid. Our results identify tests of memory and sustained attention may be particularly useful measures to track decline in the preclinical/prodromal stages of AD in DS whereas informant-measures may be useful in later stages (i.e. during conversion into dementia, or postdiagnosis). These results have implications for

the selection of outcome measures of treatment trials to delay or prevent cognitive decline due to AD in DS. As clinical diagnoses are generally made late into AD progression, early assessment is essential.

Folin, M., Baiguera, S., Conconi, M.T., Pati, T., Grandi, C., Parnigotto, P.P., & Nussdorfer, G.G.
The impact of risk factors of Alzheimer's disease in the Down syndrome
International Journal of Molecular Medicine, 2003, Feb,11(2), 267-270.
https://pubmed.ncbi.nlm.nih.gov/12525890/
Abstract: Down syndrome (DS) patients, after the fourth decade of life, display some neuropathological features of the Alzheimer's disease (AD). Several hypotheses suggested that apoE4 protein, an AD risk factor, might promote amyloid formation by stabilizing an aggregated conformation of the beta amyloid protein (Abeta). This peptide is the major proteinaceous component of the senile plaques either in AD or DS, and it is a proteolytic product of the amyloid precursor protein (APP). Both brain and platelets express three APP transcripts of the apparent molecular weight of 106, 110 and 130 kDa. In DS the Abeta deposits may ensue, at least in part, from the overexpression of the Abeta precursor gene located on chromosome 21. Aims of the present study were to evaluate the frequency of apoE4 isoform in DS population, and to ascertain whether the ratio between the 130 and the 106-110 kDa platelet APP isoforms is lower in DS, as seems to occur in AD patients. ApoE4 frequency was significantly lower in DS when compared to AD patients. E4 allele frequency of older DS patients was about half that of younger ones. The 130 to 106-110 kDa APP isoform ratio was similar in young DS and control subjects, and markedly lower in AD patients. Our results indicate that: i) in DS patients the early, selective accumulation of Abeta peptides is independent of the ApoE genotype, but the allele epsilon4 predisposes to various causes of premature death; and ii) platelet APP isoform abnormalities, which can be observed in AD patients, do not occur in young DS patients, suggesting a different processing of APP platelets in DS with respect to AD.

Fromage, B., & Anglade, P.
[Le vieillissement des sujets trisomiques] [The aging of adults with Down syndrome] *Article in French*
L'Encéphale, 2002, May-June, 28(3 Pt 1), 212-216.
https://pubmed.ncbi.nlm.nih.gov/12091781/
Abstract: The normal aging of adults with Down syndrome is comparable to that observed in individuals who have an equivalent cognitive deficit. However it is earlier and is related to the former intellectual level and life story of the person. Before 50 years, there is no significant reduction of memory. After this age short-term memory, the speed of information processing and selective attention weaken. These changes are similar to those in older adults with intellectual disability, giving the impression of early ageing in adults with Down syndrome. In terms of autonomy in everyday life, it is possible to establish an average evolutionary profile. From 60 years old, deterioration is estimated at 45% of the score obtained at 40 years, affecting in particular the skills necessary for the carrying out daily tasks (washing, dressing, feeding without assistance.). We have little knowledge of the psychiatric evolution of this people because older handicapped people are a new group in society. In the three fields of cognition, autonomy and mental health, the ageing of adults with Down syndrome is very sensitive to their environment. Pathological aging of adults with Down syndrome is associated with the dementia syndrome that, with varying degrees, combines disorders of the cognitive functions and behavior, modifying the personality. The clinical diagnosis of dementia is difficult to establish in adults with Down syndrome and opinions diverge, also it is important to comply with three rules: 1) to establish an individual base line and to observe, with the help of regular evaluations, a clear change in performance; this must be confirmed by similar modifications in daily conducts; 2) the decline depends not only on the resources of the subject, but also on the demands made by environment; 3) lasting deterioration of capacities must be clearly greater than that observable in normal ageing to signify dementia. As a function of actual age, adults with Down syndrome show early signs of ageing compared to the general population. One notes the presence of pathological anatomic lesions from 36 years old, which are similar to those observable among adults diagnosed with Alzheimer disease. However it seems that about 20% of adults with Down syndrome do not show clinical signs of dementia 20 years later. The diagnosis, which is delicate to establish, requires an ecological process consistent over time, underlining the influence of the context and the human environment on aging amond adiults with Down syndrome.

Fonseca, L.M., Padilla, C., Jones, E., Neale, N., Haddad, G.G., Mattar, G.P., Barros, E., Clare, I.C.H., Busatto, G.F., Bottino, C.M.C., Hoexter, M.Q., Holland, A.J., & Zaman, S.
Amnestic and non-amnestic symptoms of dementia: An international study of Alzheimer's disease in people with Down's syndrome
International Journal of Geriatric Psychiatry, 2020, Jun, 35(6), 650-661. doi: 10.1002/gps.5283. Epub 2020 Mar 15. PMID: 32100307
Abstract: The presence of age-related neuropathology characteristic of Alzheimer's disease (AD) in people with Down syndrome (DS) is well-established. However, the early symptoms of dementia may be atypical and appear related to dysfunction of prefrontal circuitry. To characterize the initial informant reported age-related neuropsychiatric symptoms of dementia in people with DS, and their relationship to AD and frontal lobe function. Non-amnestic informant reported symptoms (disinhibition, apathy, and executive dysfunction) and amnestic symptoms from the CAMDEX-DS informant interview were analyzed in a cross-sectional cohort of 162 participants with DS over 30 years of age, divided into three groups: stable cognition, prodromal dementia, and AD. To investigate age-related symptoms prior to evidence of prodromal dementia we stratified the stable cognition group by age. Amnestic and non-amnestic symptoms were present before evidence of informant-reported cognitive decline. In those who received the diagnosis of AD, symptoms tended to be more marked. Memory impairments were more marked in the prodromal dementia than the stable cognition group (OR = 35.07; P < .001), as was executive dysfunction (OR = 7.16; P < .001). Disinhibition was greater in the AD than in the prodromal dementia group (OR = 3.54; P = .04). Apathy was more pronounced in the AD than in the stable cognition group (OR = 34.18; P < .001). Premorbid amnestic and non-amnestic symptoms as reported by informants increase with the progression to AD. For the formal diagnosis of AD in DS this progression of symptoms needs to be taken into account. An understanding of the unique clinical presentation of DS in AD should inform treatment options.

Forbat, L., & Service, K.P.
Who cares? Contextual layers in end-of-life care for people with intellectual disability and dementia.
Dementia, 2005, 4(3), 413-431.
Abstract: The complexity of the relationship between intellectual disability (ID) and dementia is increasingly acknowledged. In order to operationalize a route towards person-centered care, we introduce the hierarchy model (Pearce, 1999) as a tool to focus the attention of policy and practice on all aspects of caregiving. This tool, which is taken from the family therapy literature, enables practitioners to examine the broad systems that impact on the delivery and receipt of care. In this article, we focus on its utility in scrutinizing end-of-life and later stages of dementia by illustrating its use with three key areas in dementia care. These three areas provide some of the most challenging situations at the end stages, because of the possible treatment options, they are: nutrition, medical interventions, and the location of care provision. This model enables a focused approach to understanding how meaning is created within social interaction. The article draws out implications for practice and policy and has applications for practice internationally.

Fortea, J., Zaman,S,H., Hartley, S., Rafii, M.S., & Head,H, Maria Carmona-Iragui, M.
Alzheimer's disease associated with Down syndrome: a genetic form of dementia
The Lancet Neurology, 2021 (Nov. 01), 20(11), 930-942.
DOI:https://doi.org/10.1016/S1474-4422(21)00245-3
Abstract: Adults with Down syndrome develop the neuropathological hallmarks of Alzheimer's disease and are at very high risk of developing early-onset dementia, which is now the leading cause of death in this population. Diagnosis of dementia remains a clinical challenge because of the lack of validated diagnostic criteria in this population, and because symptoms are overshadowed by the intellectual disability associated with Down syndrome. In people with Down syndrome, fluid and imaging biomarkers have shown good diagnostic performances and a strikingly similar temporality of changes with respect to sporadic and autosomal dominant Alzheimer's disease. Most importantly, there are no treatments to prevent Alzheimer's disease, even though adults with Down syndrome could be an optimal population in whom to conduct Alzheimer's disease prevention trials. Unprecedented research activity in Down syndrome is rapidly changing this bleak scenario that will translate into disease-modifying therapies that could benefit other populations.

Fortea J, Vilaplana E, Carmona-Iragui M, Benejam B, Videla L, Barroeta I,

Fernández S, Altuna M, Pegueroles J, Montal V, Valldeneu S, Giménez S, González-Ortiz S, Muñoz L, Estellés T, Illán-Gala I, Belbin O, Camacho V, Wilson LR, Annus T, Osorio RS, Videla S, Lehmann S, Holland AJ, Alcolea D, Clarimón J, Zaman SH, Blesa R, Lleó A.
Clinical and biomarker changes of Alzheimer's disease in adults with Down syndrome: a cross-sectional study.
Lancet. 2020 Jun 27;395(10242):1988-1997. doi: 10.1016/S0140-6736(20)30689-9
Abstract: Alzheimer's disease and its complications are the leading cause of death in adults with Down syndrome. Studies have assessed Alzheimer's disease in individuals with Down syndrome, but the natural history of biomarker changes in Down syndrome has not been established. We characterised the order and timing of changes in biomarkers of Alzheimer's disease in a population of adults with Down syndrome. We did a dual-centre cross-sectional study of adults with Down syndrome recruited through a population-based health plan in Barcelona (Spain) and through services for people with intellectual disabilities in Cambridge (UK). Cognitive impairment in participants with Down syndrome was classified with the Cambridge Cognitive Examination for Older Adults with Down Syndrome (CAMCOG-DS). Only participants with mild or moderate disability were included who had at least one of the following Alzheimer's disease measures: apolipoprotein E allele carrier status; plasma concentrations of amyloid ß peptides 1-42 and 1-40 and their ratio (Aß1-42/1-40), total tau protein, and neurofilament light chain (NFL); tau phosphorylated at threonine 181 (p-tau), and NFL in cerebrospinal fluid (CSF); and one or more of PET with 18F-fluorodeoxyglucose, PET with amyloid tracers, and MRI. Cognitively healthy euploid controls aged up to 75 years who had no biomarker abnormalities were recruited from the Sant Pau Initiative on Neurodegeneration. We used a first-order locally estimated scatterplot smoothing curve to determine the order and age at onset of the biomarker changes, and the lowest ages at the divergence with 95% CIs are also reported where appropriate. Between Feb 1, 2013, and June 28, 2019 (Barcelona), and between June 1, 2009, and Dec 31, 2014 (Cambridge), we included 388 participants with Down syndrome (257 [66%] asymptomatic, 48 [12%] with prodromal Alzheimer's disease, and 83 [21%] with Alzheimer's disease dementia) and 242 euploid controls. CSF Aß1-42/1-40 and plasma NFL values changed in individuals with Down syndrome as early as the third decade of life, and amyloid PET uptake changed in the fourth decade. 18F-fluorodeoxyglucose PET and CSF p-tau changes occurred later in the fourth decade of life, followed by hippocampal atrophy and changes in cognition in the fifth decade of life. Prodromal Alzheimer's disease was diagnosed at a median age of 50·2 years (IQR 47·5-54·1), and Alzheimer's disease dementia at 53·7 years (49·5-57·2). Symptomatic Alzheimer's disease prevalence increased with age in individuals with Down syndrome, reaching 90-100% in the seventh decade of life.

Interpretation: Alzheimer's disease in individuals with Down syndrome has a long preclinical phase in which biomarkers follow a predictable order of changes over more than two decades. The similarities with sporadic and autosomal dominant Alzheimer's disease and the prevalence of Down syndrome make this population a suitable target for Alzheimer's disease preventive treatments.

Foundation for People with Learning Disabilities
Down's syndrome and dementia - Briefing for Commissioners
London: The Foundation for People with Learning Disabilities [c/o Mental Health Foundation, 20/21 Cornwall Terrace, London, England NW1 4QL; e/m mhf@mhf.org.uk; www.learningdisabilities.org.uk] (February 2001)
8 pp.
Abstract: Backgrounder document, written for funders of services in the United Kingdom, outlines the epidemiology of dementia and Down's syndrome and identifies key support services necessary as part of a package of local services to be established for persons affected by dementia and intellectual disabilities (ID). While titled for dementia and Down's syndrome applicable for all persons with ID. Written in brief style, covers main issues and funding considerations and serves as an excellent planning tool for establishing services. Also covers basic clinical diagnostic information and basis for care management decision making. Routes the reader to associated organizations for further information

Fray, M.T.
Caring for Kathleen: A sister's story about Down's syndrome and dementia.
Kidderminster, United Kingdom: British Institute of Learning Disabilities [BILD, Wolverhampton Road, Kidderminster, Worcestershire, UK DY10 3PP – www.bild.demon.co.uk] (2000)
44 pp.
Abstract: Biographical monograph on the aging and eventual decline and death of a woman with Down syndrome as told by her sister. Provides many insights in service barriers and successes, while also providing a vivid case example of how Alzheimer's disease affects a family carer of a person with an intellectual disability.

Fredericksen, J. & Fabbre, V.
Down syndrome and Alzheimer's disease: Issues and implications for social work.
Journal of Gerontological Social Work, 2018, 61(1), 4-10. DOI: 10.1080/01634372.2017.1393480.
Owing to recent medical advancements, people with Down Syndrome (DS) are now able to live considerably longer lives and thus experience a variety of complex issues as they age. Alzheimer's Disease (AD) frequently occurs in older adults who have DS, but few practice guidelines exist to inform social work practice with older adults who have this dual diagnosis. This commentary will highlight the connection between these two conditions within a neurobiological framework and discuss implications for practice based on the available literature on this intersection of ability status, cognitive status, and age.

Frederiksen, K..S., Nielsen, T.R., Winblad, B., Schmidt, R., Kramberger, M.G., Jones, R.W., Hort, J., Grimmer, T., Georges, J., Frölichm L., et al.
European Academy of Neurology/European Alzheimer's Disease Consortium position statement on diagnostic disclosure, biomarker counseling, and management of patients with mild cognitive impairment
European Journal of Neurology, 2021, 28(7), 2147-2155. doi: 10.1111/ene.14668.
Abstract: Careful counseling through the diagnostic process and adequate postdiagnostic support in patients with mild cognitive impairment (MCI) is important. Previous studies have indicated heterogeneity in practice and the need for guidance for clinicians. A joint European Academy of Neurology/European Alzheimer's Disease Consortium panel of dementia specialists was appointed. Through online meetings and emails, positions were developed regarding disclosing a syndrome diagnosis of MCI, pre- and postbiomarker sampling counseling, and postdiagnostic support. Prior to diagnostic evaluation, motives and wishes of the patient should be sought. Diagnostic disclosure should be carried out by a dementia specialist taking the ethical principles of "the right to know" versus "the wish not to know" into account. Disclosure should be accompanied by written information and a follow-up plan. It should be made clear that MCI is not dementia. Prebiomarker counseling should always be carried out if biomarker sampling is considered and postbiomarker counseling if sampling is carried out. A dementia specialist knowledgeable about biomarkers should inform about pros and cons, including alternatives, to enable an autonomous and informed decision. Postbiomarker counseling will depend in part on the results of biomarkers. Follow-up should be considered for all patients with MCI and include brain-healthy advice and possibly treatment for specific underlying causes. Advice on advance directives may be relevant. Guidance to clinicians on various aspects of the diagnostic process in patients with MCI is presented here as position statements. Further studies are needed to enable more evidence-based and standardized recommendations in the future.

Fromage, B., & Anglade, P.
Avancée en âge du sujet atteint d'une trisomie 21 [The aging of Down's syndrome subjects].
Encephale, 2002, May-Jun, 28(3 Pt 1), 212-216.
Abstract: The normal ageing of adults with Down syndrome is comparable to that observed in individuals who have an equivalent cognitive deficit. However it is earlier and is related to the former intellectual level and life story of the person. Before 50 years, there is no significant reduction of memory. After this age short-term memory, the speed of information processing and selective attention weaken. These changes are similar to those in older adults with an intellectual disabiliy, giving the impression of early ageing in Down syndrome subjects. In terms of autonomy in everyday life, it is possible to establish an average evolutionary profile. From 60 years old, deterioration is estimated at 45% of the score obtained at 40 years, affecting in particular the skills necessary for the carrying out daily tasks (washing, dressing, feeding without assistance.). We have little knowledge of the psychiatric evolution of this people because older handicapped people are a new group in society. In the three fields of cognition, autonomy and mental health, the ageing of Down syndrome subjects is very sensitive to their environment. Pathological aging of adults ith Down syndrome is associated with the dementia syndrome that, with varying degrees, combines

disorders of the cognitive functions and behavior, modifying their personality. The clinical diagnosis of dementia is difficult to establish in subjects with Down syndrome and opinions diverge, also it is important to comply with three rules: 1) to establish an individual base line and to observe, with the help of regular evaluations, a clear change in performance; this must be confirmed by similar modifications in daily conducts; 2) the decline depends not only on the resources of the subject, but also on the demands made by environment; 3) lasting deterioration of capacities must be clearly greater than that observable in normal ageing to signify dementia. As a function of actual age, adults with Down syndrome show early signs of aging compared to the general population. One notes the presence of pathological anatomic lesions from 36 years old, which are similar to those observable among patients with Alzheimer's disease. However it seems that about 20% of adults with Down syndrome do not show clinical signs of dementia 20 years later. The diagnosis, which is delicate to establish, requires an ecological process consistent over time, underlining the influence of the context and the human environment on the ageing of adults with Down syndrome.

Funaki, Y., Kaneko, F., & Okamura, H.
Study of factors associated with changes in quality of life of demented elderly persons in group homes
Scandinavian Journal of Occupational Therapy, 2005, 12(1), 4-9. doi:10.1080/11038120510031725.
Abstract: The purpose of the present study was to attempt to identify changes in quality of life (QOL) and factors associated with them shortly after demented elderly residents entered a group home. The subjects were 25 demented elderly persons who had entered a group home within the previous 3 months. Their QOL and factors associated with it were evaluated on two occasions, at baseline and 3 months later. An objective scale for dementia, the Quality of Life Questionnaire for Dementia (QOL-D), was used to evaluate their QOL. The results showed a significant change between the QOL-D scores at baseline and 3 months later, and changes in housekeeping item scores were extracted as factors associated with changes in QOL-D. These findings suggest that the QOL score rises soon after entering a group home, and that the acquisition of roles within the group home may influence the increase in QOL.

Furness, K.A., Loverseed, A., Lippold, T., & Dodd, K.
The views of people who care for adults with Down's syndrome and dementia: A service evaluation.
British Journal of Learning Disabilities, 40(4), 318-327.
https://doi.org/10.1111/j.1468-3156.2011.00714.x
Abstract: It is well established that people with Down's syndrome are more likely to develop dementia than other people and that onset of dementia is likely to occur earlier at an earlier age. The article reports on a specialist service for people with Down's syndrome and dementia. The service has offered dementia screening and assessment to people with Down's syndrome for over 10 years and has also developed to offer support and training for carers. Semi-structured interviews were conducted with family carers, relatives and staff about the impact on them of caring for someone with Down's syndrome and how the dementia service supports them in this role. The resulting data were analyzed using interpretative phenomenological analysis. The responses provide rich insights into the areas of 'knowledge and information', 'coping and support' and 'concerns about the future'. Interviewees also identified services they wanted for the future. As a result of this evaluation, a number of changes have been proposed and begun to be implemented within the service. The results have important implications for other health, social care, and voluntary organizations.

Ghazirad, M., Hewitt, O. & Walden, S.
What outcome measures are most useful in measuring the effectiveness of anti-dementia medication in people with intellectual disabilities and dementia?
Advances in Mental Health and Intellectual Disabilities, 2022, 16(2), 87-101.
https://doi.org/10.1108/AMHID-10-2021-0038
Abstract: The use of anti-dementia medication in people with intellectual disabilities has been controversial and requires additional research to assess the efficacy of such medications. An essential part of this treatment (both in terms of research and clinical practice) is having robust outcome measures to assess the efficacy of these medications for individuals. Currently there is no consensus in the UK regarding which outcome measures, in conjunction with clinical judgement, are effective in informing clinicians' decision-making regarding anti-dementia medication management and this paper aims to present useful outcome measures. A comprehensive literature search was conducted to identify relevant outcome measures. Outcome measures focused on aspects of patients' presentation such as cognition, activities of daily living, neuropsychiatric presentation or the impact of their presentation (either on themselves, or on others). These outcome measures were critically appraised to ascertain their suitability in informing clinician's decisions regarding management of anti-dementia medication. The focus of this appraisal was on good quality measures that are practical and accessible and can be easily used within clinical NHS services. This paper provides advice for clinicians on using appropriate outcome measures, depending on patients' presentations and the symptoms of dementia being targeted, that can be used alongside their clinical assessment to enhance their anti-dementia medication management. Two case studies are presented to illustrate the use of such outcome measures. The case for using a range of assessments that are both broad in focus, and those specifically selected to measure the areas of functioning targeted by the anti-dementia medication, is presented.

Ghezzo, A., Salvioli, S., Solimando, M.C., Palmieri, A., Chiostergi, C., Scurti,M., Lomartire,L., Bedetti, F., Cocchi, G., Follo, D., Pipitone, E,, Rovatti,P., Zamberletti, J., Gomiero, T., Castellani, G., & Franceschi, C.
Age-related changes of adaptive and neuropsychological features in persons with Down syndrome.
PLoSONE. 2014, 9(11): e113111. doi:10.1371/journal.pone.0113111
Abstract: Down syndrome (DS) is characterised by premature aging and an acceleratedecline of cognitive functions in the vast majority of cases. As the life expectancy of DS persons is rapidly increasing, this decline is becoming a dramatic healthproblem. The aim of this study was to thoroughly evaluate a group of 67 non-demented persons with DS of different ages (11 to 66 years), from neuropsychological, neuropsychiatric and psychomotor point of view in order to evaluate in a cross-sectional study the age-related adaptive and neuropsychological features, and to possibly identify early signs predictive of cognitive decline. The main finding of this study is that both neuropsychological functions and adaptive skills are lower in adult DS persons over 40 years old, compared to younger ones. In particular, language and short memory skills, frontal lobe functions, visuo-spatial abilities and adaptive behavior appear to be the more affected domains. A growing deficit in verbal comprehension, along with social isolation, loss of interest and greater fatigue in daily tasks, are the main features found in older, non demented DS persons evaluated in our study. It is proposed that these signs can be alarm bells for incipient dementia, and that neuro-cognitive rehabilitation and psycho-pharmacological interventions must start as soon as the fourth decade (or even earlier) in DS persons, i.e. at an age where interventions can have the greatest efficacy.

Gholipour., T, Mitchell, S., Sarkis, R.A., & Chemali, Z.
The clinical and neurobehavioral course of Down syndrome and dementia with or without new-onset epilepsy.
Epilepsy & Behavior, 2017 Mar; 68, 11-16. doi: 10.1016/j.yebeh.2016.12.014. Epub 2017 Feb 10.
Abstract: Adult patients with Down syndrome (DS) are at higher risk of developing Alzheimer-type dementia and epilepsy. The relationship between developing dementia and the risk of developing seizures in DS is poorly characterized to date. In addition, treatment response and medication tolerability have not been rigorously studied. We identified 220 patients with a diagnosis of DS and dementia. Those without a history of developing seizures (DD) were compared to patients with new-onset seizures (DD+S) after the age of 35. Electronic records were reviewed for demographics, seizure characteristics, cognitive status, and psychiatric comorbidities. Of the patients included for analysis, twenty-six out of 60 patients had new-onset seizures or developed seizures during the follow-up period (the DD+S group) with a median onset of 2.0years after the dementia diagnosis. Generalized tonic-clonic seizures were the most common seizure type (61.5% of DD+S). Sixteen (61.5%) patients were reported to have myoclonus. Levetiracetam was the most commonly used initial medication, with the majority (73%) of patients treated achieving partial or complete seizure control. The DD+S patients tended to have a similar burden of new-onset neuropsychiatric symptoms compared to the DD group. New-onset epilepsy seems to occur early in the course of dementia in DS patients. Patients generally respond to treatment. A great burden of neuropsychiatric symptoms is seen. Future studies need to explore the relationship between ß-amyloid accumulation and epileptiform activity and attend to the care and needs of DS patients with dementia and seizures.

Giménez, S., Tapia, I.E., Fortea, J., Levedowski, D., Osorio, R., Hendrix, J., & Hillerstrom, H.

Caregiver knowledge of obstructive sleep apnoea in Down syndrome.
Journal of Intellectual Disability Research, 2023 Jan;67(1), 77-88. doi: 10.1111/jir.12990. Epub 2022 Nov 22. PMID: 36416001.
Abstract: Down syndrome (DS) population has a very high prevalence of obstructive sleep apnoea (OSA), but this remains underdiagnosed. Hence, we aimed to evaluate caregiver's knowledge of OSA and related sociodemographic factors that could contribute to OSA screening patterns in this population. An online survey though the LuMind IDSC Foundation focused on OSA diagnosis, treatments and the number of sleep studies performed. Data were compared between subjects born before and after the American Academy of Pediatrics (AAP) recommendations for OSA screening. Of the caregivers, 724 (parents 96.3%), responded to the survey. The median [interquartile (IQR)] age of the subjects with DS was 12 [20;7] years. The majority (84.3%) had sleep apnoea diagnosis, and half of them were initially referred for a sleep study due to disturbed sleep symptoms. Only 58.7% of the responders were aware of the AAP recommendations. This was linked to higher socioeconomic and/or educational level and to an earlier OSA diagnosis. The median (IQR) age of OSA diagnosis was lowered after the AAP guidelines publication compared with before its publication (3 [4;2] years vs. 10 [18;5] years, P < 0.000). Adenotonsillectomy (81.9%) and continuous positive airway pressure (61.5%) were the most commonly prescribed treatments. Few had discussed other new therapies such as hypoglossal nerve stimulation (16.0%). Only 16.0% of the subjects repeated the sleep study to monitor OSA with ageing, and 30.2% had to wait more than 4 years between studies. This study reinforces the need to improve OSA knowledge of caregivers and clinicians of individuals with DS to promote an earlier diagnosis and optimal treatment of OSA in this population.

Gitlin, L.N., and Corcoran, M.
Making homes safer: environmental adaptations for people with dementia
Alzheimer's Care Quarterly, 2000, 1(1), 50-58
Abstract: Evaluating the safety of the home environment is an important component of clinical care for persons with dementia. This article discusses safety concerns for persons with dementia living at home alone or with family members, specific modifications to the physical environment to address these issues, and guiding principles for implementing environmental changes. A wide range of environmental strategies can be introduced to maximize home safety. Different adaptations may need to be implemented with progressive memory loss thus necessitating periodic reevaluation of the home.

Glasson, E.J., Dye. D.E., & Bittles, A.H.
The triple challenges associated with age-related comorbidities in Down syndrome.
Journal of Intellectual Disability Research, 2014, Apr; 58(4), 393-398. doi:10.1111/jir.12026. Epub 2013 Mar 19. PMID: 23510031
Abstract: Major increases in the survival of people with Down syndrome during the last two generations have resulted in extended periods of adulthood requiring specialist care, which in turn necessitates greater understanding of the nature, timing and impact of comorbidities associated with the disorder. The prevalence of five comorbidities reported as common in adults with Down syndrome, visual impairment, hearing impairment, epilepsy, thyroid disorders and dementia was assessed by decade of life. From early adulthood, people with Down syndrome are at enhanced risk of developing new comorbidities and they may present with multiple conditions. Three specific challenges are identified and discussed: are comorbidities detected in a timely manner, is the clinical progress of the disorder adequately understood, and who is responsible for the provision of care? Further detailed investigations into the development and treatment of comorbidities across the lifespan are needed for a successful longitudinal approach to healthcare in people with Down syndrome. Authors note that iImplementation of this approach will better inform healthcare providers to ensure continuity of care with advancing age.

Gomiero, T., Bertelli, M., Deb, S., Weger, E., Marangoni, A., De Bastiani, E., Mantesso, U., & De Vreese, L.P.
A multicentre Italian validation study in aging adults with Down syndrome and other forms of intellectual disabilities: Dementia Screening Questionnaire for Individuals with Intellectual Disabilities.
Current Alzheimer Research, 2017, 14(7), 709-721. doi:10.2174/1567205014666170117094757.
Abstract: The USA National Task Group (NTG) guidelines advocate the use of an adapted version of Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) for dementia screening of individuals with Down syndrome (DS) and with other forms of ID (non-DS). In order to meet these guidelines, this study verifies the psychometric properties of an Italian version of the original DSQIID in a population composed of adults aged 40 years and over with DS and non-DS ID. Internal consistency, inter-rater and intra-rater reliabilities, structural validity, convergent validity and known group differences of DSQIID-I were assessed with 200 individuals with ID (mean of 55.2 years; range: 40-80 years) recruited from 15 different centers in Italy. Diagnosis of dementia was done according to IASSID diagnostic criteria and its degree of clinical certainty was defined according to Silverman et al.'s classification (2004). Cronbach's alpha for the DSQIID-I was 0.94. The ICCs for inter-rater and test-retest reliability were both 0.89. A Principal Component analysis revealed three domains, namely memory and confusion- related items, motor and functional disabilities, depression and apathy, which explained almost 40% of the overall variance. The total DSQIID-I score correlated significantly with DMR and differed significantly among those individuals (n = 34) with cognitive decline from those without (n = 166). Age, gender and severity of ID were unrelated to the DSQIID-I. The present study confirms the cross-cultural value of DSQIID which was proved to be a psychometrically valid and user-friendly observer- rated scale for dementia screening in adults with both DS and non-DS ID.

Goodman, C., Evans, C., Wilcock, J., Froggatt, K., Drennan, V., Sampson, E., Blanchard, M, Bissett, M., & Iliffe, S.
End of life care for community dwelling older people with dementia: an integrated review.
International Journal of Geriatric Psychiatry, 2010, Apr, 25(4), 329-337. doi:10.1002/gps.2343.
Abstract: [Non-ID population study] Authors reviewed the evidence for end-of-life care for community dwelling older people with dementia (including those resident in care homes). An integrated review synthesised the qualitative and quantitative evidence on end-of-life care for community dwelling older people with dementia. English language studies that focused on prognostic indicators for end-of-life care, assessment, support/relief, respite and educational interventions for community dwelling older people with dementia were included. A user representative group informed decisions on the breadth of literature used. Each study selected was screened independently by two reviewers using a standardised check list. Sixty eight papers were included. Only 17% (12) exclusively concerned living and dying with dementia at home. Six studies included direct evidence from people with dementia. The studies grouped into four broad categories: Dementia care towards the end of life, palliative symptom management for people with dementia, predicting the approach of death for people with dementia and decision-making. The majority of studies were descriptive. The few studies that developed dementia specific tools to guide end of life care and outcome measures specific to improve comfort and communication, demonstrated what could be achieved, and how much more needs to be done. Research on end-of-life care for people with dementia has yet to develop interventions that address the particular challenges that dying with dementia poses. There is a need for investigation of interventions and outcome measures for providing end-of-life care in the settings where the majority of this population live and die.

Greenwood, N., Pound, C., & Brearley, S.
'What happens when I can no longer care?' Informal carers' concerns about facing their own illness or death: a qualitative focus group study
BMJ Open, 2019 Sep 3;9(8):e030590. doi: 10.1136/bmjopen-2019-030590.
Abstract: Older informal carers play an increasingly important role in supporting others with long-term health conditions. This study aimed to explore in depth the perspectives of older carers (70+ years) supporting others with a variety of conditions and disabilities focusing on their thoughts and experiences about when they are unable to continue caring. The design was qualitative with four focus groups and the setting was Greater London in England. Participants were 28 older carers (70+ years) recruited from the voluntary sector participated in this study. Most were women and many were spouses caring for partners with age-related conditions such as dementia, arthritis and visual impairment. Nearly a third were parents of adult children with severe physical or cognitive disabilities. Findings were that the thematic analysis identified two main aspects for carers when contemplating the future-when they are unable to care in the short term or long term if they die or can no longer manage. Themes included the following: the impact of age, health conditions and relationships on future planning; anxiety about future care; carers' ambivalence and challenges in broaching the subject; interventions that might help older carers talk about and plan for the future of those they care for. Authors note that services need to be open to talking about this difficult topic. Our findings suggest that frank discussions about when older carers cannot care and having plans in place, whether these are financial or

address other practical issues, makes it easier for all concerned. However, this issue is not easily broached and its timing and ways to access this support must be carefully and individually gauged. Future research with more diverse demographic groups is needed to improve understanding of these carers' perspectives. Research is also needed to develop interventions to support older carers to talk about and plan for the future.

Hahn, H.E., & Cadogan, M.P.
Development and Evaluation of a staff training program on palliative care for persons with intellectual and developmental disabilities
Journal of Policy and Practice in Intellectual Disabilities, 2011, Mar, 8(1), 42–52
Abstract: Persons with intellectual and developmental disabilities (I/DD) face barriers and disparities at end of life. Among thesebarriers are limited educational opportunities and a paucity of targeted training materials on palliative care for staff who providetheir day-to-day care. This paper reports on a three-phase project undertaken to develop, implement, and evaluate a palliative carecurriculum and educational program that is responsive to the unique learning needs of staff providing services and supports forindividuals with I/DD living in long-term care settings. Participants' ratings of their levels of preparation and con?dence to providepalliative care improved from pretraining to posttraining. Posttraining use of materials and practice changes in palliative careoccurred.When training is developed in partnership with the staff who will use these training resources,it has the potential to sustainits use and to alter the care practices to address the palliative care needs of persons with I/DD.

Hammond, B., & Beneditti, P.
Perspectives of a care provider
In M.P. Janicki & A.J. Dalton (Eds.), Dementia, Aging, and Intellectual Disabilities.
pp. 32-41
Philadelphia: Brunner-Mazel (1999)
Abstract: Book chapter that provides a descriptive chronology of a middle-aged woman with Down syndrome who, once diagnosed with Alzheimer disease, follows a classic course of decline and eventual debilitation and death. Staff of her residence chronicled the progression of her dementia and provide some insights into the care management practices used in providing for her care. The authors place the course of her disease in perspective and offer comments on the stresses and strains on agency resources. Suggestions are offered for agencies facing similar challenge in providing day to day care for adults with dementia.

Handen, B.L.
The search for biomarkers of Alzheimer's disease in Down syndrome
American Journal of Intellectual and Developmental Disabilities, 2020,125(2), 97-99. doi: 10.1352/1944-7558-125.2.97.
Abstract: Adults with Down syndrome are at high risk for Alzheimer's disease (AD), with most individuals developing clinical dementia by their late 60s. This increased risk for AD has been attributed, at least in part, to triplication and overexpression of the gene for amyloid precursor protein (APP) on chromosome 21, leading to elevated levels of amyloid ß peptides. This article offers a brief overview of our current knowledge of AD in the DS population. In addition, the NIA/NICHD-funded, multicenter longitudinal study of biomarkers of AD in adults with DS is explored.The Alzheimer's Biomarkers Consortium–Down Syndrome (ABC-DS) is a longitudinal study of Alzheimer Disease biomarkers in adults with Down syndrome supported by federal grants from the National Institute on Aging (NIA) and the Eunice Kennedy Shriver National Institute of Child Health and Human Development (NICHD). The primary goal of ABC-DS is to understand the factors that moderate the relationship between Aß, neurodegeneration and dementia in DS and biomarkers for those factors that could be critically important in the design of effective therapeutic trials for AD, not only in DS, but in the general population as well.

Harp, J.P., Koehl, L.M., Van Pelt, K.I., Hom, C.I., Doran, E., Hea\d, E., Lott, I.T., & Schmitt, F.A.
Cognitive and behavioral domains that reliably differentiate
Normal aging and dementia in Down syndrome
Brain Science, 2021 Aug 25, 11(9), 1128.
https://doi.org/10.3390/brainsci11091128
Abstract: : Primary care integration of Down syndrome (DS)-specific dementia screening is strongly advised. The current study employed principal components analysis (PCA) and classification and regression tree (CART) analyses to identify an abbreviated battery for dementia classification. Scale- and subscale-level scores from 141 participants (no dementia n = 68; probable Alzheimer's disease n = 73), for the Severe Impairment Battery (SIB), Dementia Scale for People with Learning Disabilities (DLD), and Vineland Adaptive Behavior Scales—Second Edition (Vineland-II) were analyzed. Two principle components (PC1, PC2) were identified with the odds of a probable dementia diagnosis increasing 2.54 times per PC1 unit increase and by 3.73 times per PC2 unit increase. CART analysis identified that the DLD sum of cognitive scores (SCS < 35 raw) and Vineland-II community subdomain (< 36 raw) scores best classified dementia. No significant difference in the PCA versus CART area under the curve (AUC) was noted (D(65.196) = -0.57683; p = 0.57; PCA AUC = 0.87; CART AUC = 0.91). The PCA sensitivity was 80% and specificity was 70%; CART was 100% and specificity was 81%. These results support an abbreviated dementia screening battery to identify at-risk individuals with DS in primary care settings to guide specialized diagnostic referral

Hassiotis, A., Strydom, A., Allen, K., & Walker, Z.
A memory clinic for older people with intellectual disabilities
Aging & Mental Health, 2003, 7(6), 418-423
Abstract: Cognitive decline in older people with intellectual disabilities (ID) is often under-recognized. Following the publication of the National Service Framework for Older People and the white paper Valuing People, older people with intellectual disabilities of all aetiologies should have access to a systematic assessment of their cognitive function in order to detect decline in cognition and adaptive skills and implement appropriate treatments as early as possible. The development of a memory clinic for older people with ID is described, including instruments used and characteristics of attendees. Such projects are in line with current UK government policies and can contribute to the improvement of standards of care and support research in this vulnerable group of people.

Hartley, S.L., Handen, B.L., Devenny, D., Tudorascu, D. Piro-Gambetti, B., Zammit, M.B., Laymon, C.V., Klunk, W.E., Zaman,S., Cohen, A., & Christian, B.T.
Cognitive indicators of transition to preclinical and prodromal stages of Alzheimer's disease in Down syndrome.
Alzheimer's & Dementia: Diagnosis, Assessment & Disease Monitoring, 2020; 12: 1-10 e12096. https://doi.org/10.1002/dad2.12096

Abstract: There is a critical need to identify measures of cognitive functioning sensitive to early Alzheimer's disease (AD) pathophysiology in Down syndrome to advance clinical trial research in this at-risk population. The objective of the study was to longitudinally track performance on cognitive measures in relation to neocortical and striatal amyloid beta (Aß) in non-demented Down syndrome. The study included 118 non-demented adults with Down syndrome who participated in two to five points of data collection, spanning 1.5 to 8 years. Episodic memory, visual attention and executive functioning, and motor planning and coordination were assessed. Aß was measured via [C-11] Pittsburgh Compound-B (PiB) PET. PiB was associated with level and rate of decline in cognitive performance in episodic memory, visual attention, executive functioning, and visuospatial ability in models controlling for chronological age. The Cued Recall Test emerged as a promising indicator of transition from preclinical to prodromal AD.

Hatzidimitriadou, E., & Alisoun Milne, A.
Planning ahead: Meeting the needs of older people with intellectual disabilities in the United Kingdom
Dementia, 2005, 4(3), 341-359. https://doi.org/10.1177/1471301205055027
Abstract: Despite the acknowledged increase in the number of older people with intellectual disabilities (ID) in the UK, the age-related health and social care needs of this population have yet to be fully understood and addressed. Although there is some evidence of positive development, the current picture of service provision is characterized by fragmentation and limited choice of resources and specialist care. Policy aims are variably met and inconsistently applied. Research suggests that service planning is often incoherent, that many older people with ID and their carers receive poor quality non-specialist care and that staff are inadequately trained to manage the often multiple and complex needs of this user group. There is a considerable co-joined service development and research challenge in this emerging field. If older people with ID and their carers are to receive quality provision, a coherent and well-funded service planning system is required which is underpinned by articulated agency partnerships, informed by good practice developments in the fields of ID, gerontology and dementia care, and linked to evidence about effective models of care and services. The

incorporation of the perspectives of users and carers in the planning process is an essential pre-requisite as is a commitment to the development of effective support across the life course of all individuals with ID.

He, P., Chen, G., Wang, Z., Guo., C., Li, N., Yun, C., & Zheng, X.
Adults with intellectual disabilities in China: comorbid psychiatric disorder and its association with health service utilisation
Journal of Intellectual Disability Research, 2018 Feb, 62(2), 16-114. doi: 10.1111/jir.12451. Epub 2017 Nov 26.
Abstract: Adults with intellectual disabilities (ID) often have multiple comorbidities. Psychiatric disorders in this population have been poorly studied in developing countries. We aimed to investigate the prevalence of psychiatric disorders in adults with ID and whether comorbid psychiatric disorders were associated with health service utilization. We obtained data from the Second National Sample Survey on Disability, conducted in 31 provinces of China and selected a subsample of 13 631 adults aged 18 years and above with ID. ID were defined by intelligence quotient score under 70, deficits in two or more adaptive behaviours and age of onset under 18 years. Psychiatric disorders were identified according to the International Statistical Classification of Diseases, Tenth Revision. Logistic regressions were used for data analyses. The prevalence of psychiatric disorders in adults with ID was 16.7%. The most prevalent type of psychiatric disorder was **dementia.** Older adults, females, being minorities, urban residents, being literate, low-income groups and having severe ID, were associated with elevated risk of psychiatric disorder among adults with ID. Compared with individuals without psychiatric disorders, those with comorbid psychiatric disorders were more likely to use medical service and less likely to use rehabilitation service. The prevalence of psychiatric disorder in adults with ID was strikingly higher than that in the general population. Health service utilisation among Chinese adults with ID remained a big challenge. There is a possibility of diagnostic overshadowing by local clinicians, which may have resulted in overdiagnosis of dementia and underdiagnosis of common mental disorders. This study informs further investigations regarding common mental disorders among people with ID and has implications for public health strategies and health policies to meet health service need for this population.

Head, E., Lott, I.T., Wilcock, D.M., & Lemere, C.A.
Aging in Down syndrome and the development of Alzheimer's disease neuropathology
Current Alzheimer Research, 2016, 13(1),18-29. doi:10.2174/1567205012666151020114607.
Abstract: Chromosome 21, triplicated in Down Syndrome, contains several genes that are thought to play a critical role in the development of AD neuropathology. The overexpression of the gene for the amyloid precursor protein (APP), on chromosome 21, leads to early onset beta-amyloid (Aß) plaques in DS. In addition to Aß accumulation, middle-aged people with DS develop neurofibrillary tangles, cerebrovascular pathology, white matter pathology, oxidative damage, neuroinflammation and neuron loss. There is also evidence of potential compensatory responses in DS that benefit the brain and delay the onset of dementia after there is sufficient neuropathology for a diagnosis of AD. This review describes some of the existing literature and also highlights gaps in our knowledge regarding AD neuropathology in DS. It will be critical in the future to develop networked brain banks with standardized collection procedures to fully characterize the regional and temporal pathological events associated with aging in DS. As more information is acquired regarding AD evolution in DS, there will be opportunities to develop interventions that are age-appropriate to delay AD in DS

Head, E., Powell, D., Gold, B.T., Schmitt, F.A..
Alzheimer's disease in Down syndrome
European Journal of Neurodegenerative Diseases, 2012, 1(3): 353–364.
Abstact: A key challenge to adults with Down syndrome (DS) as they age is an increased risk for cognitive decline, dementia, and Alzheimer disease (AD). In DS persons ranging from 40-49 years of age, 5.7-55% may be clinically demented and between 50-59 years, dementia prevalence ranges from 4-55% (reviewed in [1]). Despite the wide ranges reported for dementia prevalence, a consistent feature of aging in DS is the progressive accumulation of AD brain pathologies. By the age of 40 years, virtually all have sufficient senile plaques and neurofibrillary tangles for a neuropathological diagnosis of AD [2]. Thus, there is dissociation between the age of onset of AD neuropathology (40 years) and increasing signs of clinical dementia. We discuss the hypothesis that frontal impairments are a critical factor affecting cognitive function and are associated with white matter (WM) and AD neuropathology. While these may be an early sign of conversion to dementia, we also review several other clinical comorbidities that may also contribute to dementia onset.

 **Hellen, C.R.**
Alzheimer's disease - activity-focused care (2nd Ed.)
Boston: Butterworth-Heinemann (1998)
436 pp.
Abstract: A 13-chapter text that provide voluminous information on developing and provision of activities for persons affected by Alzheimer's disease and related dementias - with application to persons with intellectual disabilities. Written from a practitioner viewpoint, it is designed to promote an individual's cognitive, physical and psychosocial well-being. It includes forms and profiles for use by program personnel, presents a holistic intervention program, features content on refocusing activities for physically combative or violent situations. Contains chapters on communication, daily living care activities, aiding at mealtimes, facilitating physical wellness (mobility and exercise), addressing dementia induced behaviors, creating meaningful activities for daily life, and aiding in terminal care, among others.

Heller, T., Scott, H.M., & Janicki, M.P.
Caregiving, intellectual disability, and dementia: Report of the Summit Workgroup on Caregiving and Intellectual and Developmental Disabilities
Alzheimers & Dementia - Translational Reserach & Clinical Interventions,. 2018, 4, 272-282.
Published online 2018 Jul 10. doi: 10.1016/j.trci.2018.06.002
Abstract: A specially commissioned working group produced a report on caregiving, intellectual and developmental disabilities (IDDs), and dementia for the National Institutes of Health-located National Research Summit on Care, Services, and Supports for Persons with Dementia and Their Caregivers. Experts in caregiving, dementia, and IDDs examined the current state of research, policy, and practice related to caregiving and supports; identified the similarities and dissimilarities between IDD-related care and services and the general population affected by dementia; and considered how these findings might contribute to the conversation on developing a dementia care research and services development agenda. Five major areas related to programs and caregiving were assessed: (1) challenges of dementia; (2) family caregiving interventions; (3) supportive care settings; (4) effects of diversity; and (5) bridging service networks of aging and disability. Recommendations included increasing supports for caregivers of adults with IDDs and dementia; increasing research on community living settings and including caregivers of persons with IDDs in dementia research; acknowledging cultural values and practice diversity in caregiving; increasing screening for dementia and raising awareness; and leveraging integration of aging and disability networks.

Herron, D.L., & Priest, H.M.
Support workers' knowledge about dementia: A vignette study.
Advances in Mental Health and Intellectual Disabilities, 2013, 7(1), 27-39. https://doi.org/10.1108/20441281311294675
Abstract: It is widely acknowledged that people with intellectual disabilities are highly likely to experience mental health problems, but that support workers' knowledge and skill in this area is sometimes lacking. There is little research explicitly exploring knowledge about the mental health of older people with intellectual disabilities and the purpose of this paper is to attempt to fill this gap. In total, 14 support workers completed a questionnaire in which three vignettes presented progressively worsening indicators of dementia in an older person with intellectual disabilities. Participants explained what they thought was happening and what action they would take. Data were analysed using Braun and Clarke's framework. Few participants had undertaken any mental health training, and only one in relation to older people. They were generally poor at judging early and intermediate indicators of dementia, but were able to identify more overt later signs. However, they believed these advanced indicators to be the onset of dementia. Nonetheless, they would generally take appropriate action, such as observation and referral. Abuse was often considered as a causal factor. The most significant implication is the need for training in the mental health needs of older people and in particular, the general and specific indicators and expected trajectory of dementia in this population.

Herron, D.L., Priest, H.M., & Read, S.
Supporting people with an intellectual disability and dementia: A constructivist grounded theory study exploring care providers' views and experiences in the UK.

Journal of Applied Research in Intellectual Disabilities, 2020, 33(6), 1405-1417. doi: 10.1111/jar.12768
Abstract: There is a need to better understand the experiences and support needs of paid and family carers of people with an intellectual disability and dementia, and the role of Intellectual Disability Dementia Care Pathways (IDDCPs). This study explored the experiences of carers, and IDDCPs and other support structures within those experiences. A constructivist grounded theory methodology was implemented. Data were obtained through 23 semi-structured interviews with two family carers, eight paid carers and eight healthcare professionals. The study's theory produced five interrelated categories: Impact of Dementia, Challenging the Diagnosis Process, Continuum of Support, Continuity and Continuum of Understanding. Findings have demonstrated the importance of planning and supporting carers' holistic needs; the role of an IDDCP in the post-diagnostic support (or lack of it) for carers; and the importance of a timely diagnosis of dementia. An important implication of these findings is the need for local services to develop inclusive specialized IDDCPs. Having IDDCP healthcare professionals, who have expertise in both intellectual disability and dementia, allows support, advice and information to be tailored to both the person's intellectual disability and dementia. This may also help to address some of the challenges people with an intellectual disability and dementia, and their carers, experience when encountering generic services without the appropriate expertise in intellectual disability care. Knowledge and understanding of both intellectual disability and dementia are essential to initiating the diagnosis process and providing person-centred support. Carers need both a theoretical understanding of dementia and how to address dementia-related changes through dementia training courses, and hands on experience where they could apply this training. Based on their work, the authors recommend the (1) need for local health services to develop inclusive specialized IDDCPs, (2) development of a comprehensive, accessible training package, informed by these findings and the concept of person-centred care, (3) need for organizations and services to address the reactive culture sometimes seen, and implement procedures for effective dementia care planning, and (4) need to ensure a reliable, timely diagnosis and early dementia care planning, through reactive assessments, proactive baselining and screening, and associated guidance.

Heston, L.L.
Down's syndrome and Alzheimer's dementia: defining an association
Psychiatric developments,1984, Winter, 2(4), 287-294.
https://pubmed.ncbi.nlm.nih.gov/6241313/
Abstract: The typical neuropathological features of Alzheimer's disease, plaques and tangles, appear in virtually all patients with Down's Syndrome after the age of 40. Clinically, changes in cognitive performance and behavior appear to correlate with these neuropathological changes, although a satisfactory operational definition of dementia in a context of mental retardation is not available. It is unknown whether the cholinergic losses in the nucleus basalis, which are a feature of early onset Alzheimer's disease, also occur late in Down's syndrome. Two family studies have supported a greater than expected incidence of Down's cases among relatives of probands dying with Alzheimer-type dementia, but the association is not strong. It is noteworthy that in both studies, phenotypically normal carriers of the rare 15/21 translocation had severe early onset dementia, although this translocation is responsible for less than 0.4 per cent of Down's cases. An increased incidence of dementia among carriers of the more common 14/21 translocation has not been reported. In any case, it is proposed that a gene product originating from the long arm of chromosome 21 (21q) is necessary for Alzheimer-type pathology, since a segregating gene could not be responsible for the 100 per cent incidence of these changes among 21q trisomics.

Higgins, L., & Mansell, J.
Quality of life in group homes and older persons' homes.
British Journal of Learning Disabilities, 2009, 37, 207–212
Abstract: Older people with intellectual disabilities sometimes live in older people's homes rather than homes for people with intellectual disabilities. Little is known about their quality of life in these homes. A non-equivalent comparison group design was used to compare the quality of life of 59 people in three groups; older people without an intellectual disability living in older people's homes (n = 20), older people with an intellectual disability living in older people's homes (n = 19) and older people with an intellectual disability living in intellectual disability homes (n = 20). Data were collected on participant characteristics, adaptive behavior and three aspects of quality of life; community involvement, participation in domestic living and choice making. The three groups were comparable in terms of gender, ethnicity and additional impairments but the older people without an intellectual disability were older and had more adaptive skills than the other groups. Older people with an intellectual disability experienced better quality of life outcomes in terms of participation in meaningful activity and community access when they lived in intellectual disability homes compared with older people's homes. It was not possible to achieve reliability on the measure of choice-making. This study provides some evidence to suggest that older people with an intellectual disability may be best served in intellectual disability homes rather than older people homes and that it is an area of research which needs further exploration.

Hithersay, R., Hamburg, S., Knight, B., & Strydom, A.
Cognitive decline and dementia in Down syndrome.
Current Opinion in Psychiatry, 2017, Mar; 30(2), 102-107.
doi:10.1097/YCO.0000000000000307.
Abstract: Alzheimer's disease is most likely universal in older individuals with Down syndrome, due to having three copies of the amyloid precursor protein gene, resulting in amyloid-beta plaque deposition. Down syndrome is an important population in which to consider clinical trials of treatments to prevent or delay the development of dementia. However, assessment of subtler cognitive changes is challenging due to the presence of intellectual disability.
Recent research confirmed that older adults with Down syndrome often present with cognitive decline: more than 80% may experience dementia by age 65 years. Efforts have been made to improve and validate neuropsychological assessment and to describe the relationship with comorbidities such as epilepsy and haemorrhagic stroke. There have also been advances in biomarkers such as neuroimaging using amyloid PET. Clinical trials of treatments, particularly in the presymptomatic phase of Alzheimer's disease, are important to consider in individuals with Down syndrome given their high dementia burden, and may also serve as proof of concept for other forms of Alzheimer's disease. However, further work is required to improve outcome measures and better understand the biomarkers of progression of disorder and their relationship with symptom development during the presymptomatic period.

Hithersay, R,, Startin, C.M., Hamburg, S., Mok, K.Y., Hardy, J., Fisher, E.M.C., Tybulewicz, V.L.J., Nizetic, D., & Strydom, A.
Association of dementia with mortality among adults with Down syndrome older than 35 years
JAMA Neurology, 2019, Feb 1, 76(2), 152-160.
doi:10.1001/jamaneurol.2018.3616.
Abstract: This work quantifies the fatal burden of dementia associated with Alzheimer disease in individuals with Down syndrome (DS). To explore the association of dementia associated with Alzheimer disease with mortality and examine factors associated with dementia in adults with DS. Prospective longitudinal study in a community setting in England. Data collection began March 29, 2012. Cases were censored on December 13, 2017. The potential sample consisted of all adults 36 years and older from the London Down Syndrome Consortium cohort with 2 data times and dementia status recorded (N = 300); 6 withdrew from study, 28 were lost to follow-up, and 55 had a single data collection point at time of analysis. The final sample consisted of 211 participants, with 503.92 person-years' follow-up. Dementia status, age, sex, APOE genotype, level of intellectual disability, health variables, and living situation. Crude mortality rates, time to death, and time to dementia diagnosis with proportional hazards of predictors. Of the 211 participants, 96 were women (45.5%) and 66 (31.3%) had a clinical dementia diagnosis. Twenty-seven participants (11 female; mean age at death, 56.74 years) died during the study period. Seventy percent had dementia. Crude mortality rates for individuals with dementia (1191.85 deaths per 10 000 person-years; 95% CI, 1168.49-1215.21) were 5 times higher than for those without (232.22 deaths per 10 000 person-years; 95% CI, 227.67-236.77). For those with dementia, APOE e4 carriers had a 7-fold increased risk of death (hazard ratio [HR], 6.91; 95% CI, 1.756-27.195). For those without dementia, epilepsy with onset after age 36 years was associated with mortality (HR, 9.66; 95% CI, 1.59-58.56). APOE e4 carriers (HR, 4.91; 95% CI, 2.53-9.56), adults with early-onset epilepsy (HR, 3.61; 95% CI, 1.12-11.60), multiple health comorbidities (HR, 1.956; 95% CI, 1.087-3.519), and those living with family (HR, 2.14; 95% CI, 1.08-4.20) received significantly earlier dementia diagnoses. Dementia was associated with mortality in 70% of older adults with DS. APOE e4 carriers and/or people with multiple comorbid health conditions were at increased risk of dementia and death, highlighting the need for good health care. For those who died without a dementia diagnosis, late-onset epilepsy was the only significant factor associated with death, raising questions about potentially undiagnosed dementia cases in

this group

Hobson, B., Webb, D., Sprague, L., Grizzell, M., Hawkins, C., & Benbow, S.M.
Establishing a database for proactive screening of adults with Down's syndrome: when services work together
Advances in Mental Health and Intellectual Disabilities, 6(2), 99-105. https://doi.org/10.1108/20441281211208464
Abstract: This paper describes a service improvement project with two aims: to identify and screen all adults with Down syndrome aged over 30 years in a defined locality using a standardised instrument to establish functional baselines; and to set up a database to facilitate early diagnosis of dementia in this population. An assistant psychologist used a standardised instrument to screen participants who were identified through contact with health, social, and third sector, and housing services. Eligible people were identified and screened using an informant-based measure. Three groups were identified: group 1 showed no significant change; group 2 showed significant change but no signs of dementia; and group 3 showed significant change plus signs of dementia. People with suspected dementia were referred on for further investigation/ assessment and supportive services. Terminology is important in engaging families in a screening project, as is the opportunity to provide information. A proactive screening project can be established by employing working partnerships between intellectual disability and older adult services to aid diagnosis. Adults with Down syndrome aged over 30 years in a defined locality can be identified through contact with health, social, and third sector, and housing services. Those identified can be screened using a standardised instrument and a database of screening results established in order to establish baselines against which future re-screening can be conducted. Partnership working between older adult mental health services and intellectual disability services can improve the diagnostic service to adults with Down syndrome.

Hoekman, J., & Maaskant, M.A.
Comparison of instruments for the diagnosis of dementia in individuals with intellectual disability
Journal of Intellectual & Developmental Disability, 2002, 27(4), 296-309. https://doi.org/10.1080/1366825021000029339
Abstract: The authors describe the agreement among the results (dementia/no dementia) of three instruments used for the potential diagnosis of dementia in persons with intellectual disability. The instruments are: the Dementia Questionnaire for Mentally Retarded Persons (DMR), the Checklist with Symptoms of Dementia (CLD) and the Delayed Match-to-Sample Test (DMTS). The results were compared with the expert opinion of a physician/educational specialist/psychologist. The participants were 329 adults affiliated with centres for people with intellectual disability in The Netherlands. It was found that the agreement among the three instruments was low (kappa < 0.5). The agreement between the expert opinion and the results of the tests was also found to be low. It was concluded that the instruments do not mutually agree upon which of the adults can be regarded as dementing or not dementing and they also provide inconsistent agreement with the expert opinion when dementia is present. It was further concluded that it is not advisable to use a single instrument when attempting to diagnose dementia in people with intellectual disability.

Holingue, C., Wise, E., Caoili, A., Klein, A., Kalb, L.G., & Beasley, J.
Screening for Dementia among Adults with Intellectual Disability: Outcomes from a Pilot Study
Journal of Mental Health Research in Intellectual Disabilities, 2022, 15(1), 20-36. https://doi.org/10.1080/19315864.2021.1965270
Abstract: Screening for dementia among individuals with ID is important to identify individuals in need of care and support. The objective of this pilot study was to identify obstacles associated with screening and assessment of dementia among older adults with ID in a crisis-prone population. The NTG's Early Detection Screen for Dementia (EDSD) was administered to eligible enrollees ages 50 years and older within the START (Systemic, Therapeutic, Assessment, Resources, and Treatment) program. Focus groups were carried out to understand the barriers to screening and diagnosis of dementia. Of the 95 eligible enrollees, 63 participants had dementia screening tools completed. Obstacles identified through focus groups included difficulty differentiating changes from baseline function, competing priorities in this crisis-prone population, lack of access to providers, and an unclear understanding of the benefit or purpose of screening among some caregivers. START coordinators noted that the EDSD provided a helpful way to collect information and document changes in the enrollee's functioning. The EDSD may be helpful for capturing potential dementia-associated changes over time in crisis-prone adults with ID, though obstacles remain to the access of further evaluation for dementia.

Holland, A.J.
Ageing and its consequences for people with Down's syndrome
Fact Sheet Series - Learning about intellectual disabilities and health
Accessed 24 August 2004 at
http:www.intellectualdisability.info/lifestages/ds_ageing.htm
Down Syndrome Association (UK) and the Department of Mental Health & Learning Disability at St. George's Hospital Medical School, University of London. 9 pp.
Abstract: Fact sheet outlines the evidence which suggests that ageing and t he problems of old age are particularly relevant to people with Down syndrome as some of these age-related problems develop earlier in life than would normally be the case. Topics covered include: aging and the brain, aging and dementia, behavioral features of dementia in people with Down syndrome, apparent decline in later life - cases to consider, difficulties in detecting dementia in people with intellectual disabilities, differential diagnosis - which conditions mimic dementia, common causes of decline in later life in people with Down syndrome, genetic mechanisms, treatment, supporting the individual, and the future.

Holland, A.J., Karlinsky, H. & Berg, J.M.
Alzheimer's disease in persons with Down syndrome: Diagnostic and management considerations
In J.M. Berg, H. Karlinsky, A.J. Holland (Eds.), Alzheimer's Disease, Down Syndrome, and Their Relationship.
pp. 96-114
Oxford: Oxford University Press (1993)
Abstract: Book chapter that examines the implications of Alzheimer's disease for adults with Down syndrome, including assessment and diagnosis and specialty service provision. Authors note that assigning a tenable diagnosis of Alzheimer disease requires careful and comprehensive data assembly, including medical history, clinical examination, neuropsychological assessment and laboratory investigations. Once the diagnosis is established, effective ongoing management should focus on supporting not only the affected individual (including advocacy for his or her rights) but also the family and professional carers. During the course of the illness various medical, psychiatric and psychological interventions can be helpful as can changes in the environment. A wide range of services for persons with Down syndrome who develop Alzheimer's disease makes it possible for affected individuals, despite deterioration, to remain in the family home or in community residential settings. Authors proffer some general suggestions for services and adaptations.

Holland, A.J., Hon, J., Huppert, F.A., & Stevens, F.
Incidence and course of dementia in people with Down's syndrome: findings from a population-based study.
Journal of Intellectual Disability Research, 2000, 44(2), 138-146. DOI: 10.1046/j.1365-2788.2000.00263.x
Abstract: The prevalence rate of Alzheimer's disease (AD) in people with Down's syndrome (DS) increases significantly with age. However, the nature of the early clinical presentation, course and incidence rates of dementia are uncertain. The aims of the present study were to investigate the characteristics of age-related clinical changes and incidence rates for dementia in a population-based sample of people with DS aged 30 years and older at the age of risk for dementia. A modified version of the Cambridge Examination for Mental Disorders of the Elderly informant interview was used to determine the extent and nature of changes in memory, personality, general mental functioning and daily living skill 18 months after a similar assessment. At the time of the first assessment, the initial changes reported were predominately in behavior and personality. At the second assessment, overall estimated incidence rates for frontal-like dementia were high (0.24), mainly in the younger groups, with incidence rates of AD, meeting both ICD-10 and DSM-IV criteria, of 0.04 predominately in the older groups. The present authors have hypothesized that the observed personality changes and the high estimated incidence rates of frontal-like dementia in the younger groups may indicate that functions served by the frontal lobes are the first to be compromised with the progressive development of Alzheimer-like neuropathology in people with DS.

Holst, G., Johansson, M., & Ahlstrom, G.
Signs in people with intellectual disabilities: Interviews with managers and staff

on the identification process of dementia.
Healthcare (Basel)), 2018, 6(3), 103. https:// doi.org/10.3390/healthcare6030103
Abstract: The life expectancy of people with intellectual disabilities (ID) has steadily increased, which has been accompanied by an increased risk of dementia. Staff and managers are key resources for safety diagnosis since they deliver information about people with ID behavior every day. The aim of the present study was to explore the identification process employed by staff and managers to detect signs of suspected dementia in people with an ID within intellectual disability services (ID-services). Twenty managers and 24 staff within an ID-service were interviewed and qualitative latent content analysis was applied. A model consisting of three themes on three levels of resources for the identification process of signs of suspected dementia emerged from the analysis. On the first level was the time and continuity in the care relationship, which is crucial for identifying and responding to changes in cognitive ability that indicate dementia. On the second level, the staff identify deficiencies in their own knowledge, seek support from colleagues and managers within their workplace and, on the third level, outside their workplace. Staff and managers expressed a need for early and continuous guidance and education from specialists in dementia and primary healthcare. This finding indicates an urgent need for intervention research and digital support for staff in dementia care. Only when these resources are used will it be possible for staff to identify, and react to, dementia symptoms in a safe and knowledge-based way, initiate examination, design well-adapted care, and, if necessary, consider moving the person to adapted housing. A close collaboration with specialists within dementia care may contribute to a good working environment for staff with skilled guidance and support, continuous education, and the initiation of intervention research concerning the best accommodation form and digital support for people with an ID and staff.

Hom, C.
Neuropsychological subtypes of incident mild cognitive impairment in Down syndrome.
AAIC 2020, Poster presentation, July 29, 2020. *Alzheimer's & Dementia*, 2020, 16 (S6), https://doi.org/10.1002/alz.043299
Abstract: Past attempts to characterize the earliest cognitive changes as individuals with Down Syndrome (DS) transition from cognitively stable to mild cognitive impairment (MCI) have been equivocal (Garcia-Alba et al., 2019; Lautarescu et al., 2017). Difficulties identifying MCI in this population are complicated by variability in pre-morbid cognitive abilities, the use of neuropsychological tests that were created for the neurotypical population, and participants scoring at floor on the baseline assessment (Krinsky-McHale and Silverman, 2013). We examined data from 151 individuals with Down Syndrome (M age=50.25, SD age=6.94). Their pre-morbid level of intellectual impairment ranged from mild to severe. All participants received comprehensive evaluations. Following data collection, the clinical status of each participant was rated at consensus review that considered performance on a core neuropsychological test battery and the clinical data for each participant. Data from the non-demented and MCI groups are examined: Cognitive Stable (N=107, 70.9%) and MCI-DS (N=44, 29.1%). The full battery consists of 27 subtests that were hypothesized a priori to measure five cognitive domains: language, memory, executive function, visuospatial reasoning, and motor coordination. Factor analysis revealed 7 principal components that maximally discriminated between test scores in older adults with DS who have not reached clinical AD status: (1) general intelligence (2) sensorimotor, (3) memory, (4) language comprehension and expression, (5) executive function/speed, (6) attention/language expression, and (7) visuomotor. Cluster analysis for the MCI group produced 3 distinct groups: (1) dysexecutive (n=4), (2) dysnomic/visuospatial impaired (n=28), and (3) amnestic/motor impaired (n=12). Author concludes that the neuropsychological battery assesses 7 distinct cognitive functions in older adults with DS. It can also capture cognitive decline, as we were able to empirically identify three distinct neuropsychological subtypes of MCI: amnestic/visuomotor impaired, dysexecutive, and dysnomic. These subtypes are generally consistent with those that have been found within the neurotypical population (Edmonds et al., 2015; Dick et al., 2016), strengthening the evidence that AD has a similar course in the DS population and late onset AD.

Horvath, S,. Garagnani, P., Bacalini, M.G., Pirazzini, C., Salvioli, S., Davide, G.,Di Blasio, A.M., Giuliani, C.,Tung, S., Vinters, H.V., & Franceschi, C.
Accelerated epigenetic aging in Down syndrome
Aging Cell, 2015, 1–5, eprint. Doi: 10.1111/acel.12325
Abstract: Down syndrome (DS) entails an increased risk of many chronic diseases that are typically associated with older age. The clinical manifestations of accelerated aging suggest that trisomy 21 increases the biological age of tissues, but molecular evidence for this hypothesis has been sparse. Here, we utilize a quantitative molecular marker of aging (known as the epigenetic clock) to demonstrate that trisomy 21 significantly increases the age of blood and brain tissue (on average by 6.6 years, $P = 7.0\ 3\ 10^{-14}$).

Humphreys, L., Bigby, C., & Iacono, T.
Dimensions of group home culture as predictors of quality of life outcomes
Journal of Applied Research in Intellectual Disabilities, 2020 Nov, 33(6), 1284-1295. doi: 10.1111/jar.12748. Epub 2020 May 27.
Abstract: Research has shown that there is variability in quality of life (QOL) outcomes for people with intellectual disabilities who live in group homes. The aim was to examine dimensions of group home culture as predictors of QOL outcomes. The Group Home Culture Scale (GHCS) was used to measure staff perceptions of culture in 23 group homes. QOL data were available from 98 people with intellectual disabilities. Multilevel modelling was used to examine the associations between the GHCS subscales and four QOL-dependent variables. Of the GHCS subscales, Effective Team Leadership and Alignment of Staff with Organizational Values significantly predicted residents' engagement in activities. Supporting Well-Being significantly predicted residents' community involvement. None of the GHCS subscales significantly predicted domestic participation and choice making. The findings suggest that strategies to improve Effective Team Leadership and Supporting Well-Being dimensions of culture may contribute to enhancing certain QOL outcomes.

Huxley, A., Van-Schaik, P., & Witts, P.
A comparison of challenging behavior in an adult group with Down's syndrome and dementia compared with an adult Down's syndrome group without dementia.
British Journal of Learning Disabilities, 2005, 33(4), 188-193.
Abstract: This study investigated the frequency and severity of challenging behavior in adults with Down's syndrome with and without signs of dementia. Care staff were interviewed using the Aberrant Behavior Checklist-Community version (M.G. Aman & N.N. Singh, Slosson, East Aurora, NY, 1994), to investigate the frequency and severity of challenging behavior. Individuals' 'dementia status' was assessed by using the Dementia Scale for Down's Syndrome (Gedye Research and Consulting, Vancouver, 1995). The results showed that the dementia group displayed more frequent and severe forms of challenging behavior than the nondementia group. The difference in reported levels of challenging behavior of both groups with the general learning disabilities population was not considered to be clinically significant and levels fell predominantly within the 'normal range'. The findings of this study suggest that frequent and severe forms of challenging behavior in adults with Down's syndrome is more likely to be a behavioral symptom associated with the onset of a dementing illness and not due to normal aging alone.

Iacono, T., Bigby, C., Carling-Jenkins, R., & Torr, J.
Taking each day as it comes: Staff experiences of supporting people with Down syndrome and Alzheimer's disease in group homes,
Journal of Intellectual Disability Research, 2013; 58(6). DOI:10.1111/jir.12048.
Abstract: Disability staff are being increasingly required to support adults with Down syndrome who develop Alzheimer's disease. They have little understanding of the nature of care required, and may lack input from aged care and dementia services, which lack knowledge of intellectual disability. The aim of this study was to report on the experiences of disability staff in group homes supporting residents with Down syndrome and Alzheimer's disease in relation to their under understanding of what was happening to these residents, their responses to them, and how they felt about their support role. Disability support staff for nine adults with Down syndrome who had a diagnosis of Alzheimer's disease were interviewed twice, over intervals of 6-9 months. Interviews were transcribed and analyzed for themes. Authors foiund that three key themes emerged - (I) struggling to understand change, (ii) taking each day as it comes, and (iii) he's got a disability and that's our job. Staff had only limited understanding of how Alzheimer's disease impacted the adults with Down syndrome, their responses to changes were ad hoc, and they used strategies on a trial and error basis. They were committed to providing care, but at the risk of rejecting input and support. The need for collaboration across disability, and aged and disability care was evident to facilitate aging-in-place and planned care transitions.

Ilacqua, A., Benedict, J., Shoben, A., Skotkp, B.G., Mathews, T. Benson, B., & Allain, D.C.

Alzheimer's disease development in adults with Down syndrome: Caregivers' perspectives
American Journal of Medical Genetics, 2020, 182(1), 104-114
Abstract: Research about Alzheimer's disease (AD) in individuals with Down syndrome (DS) has predominantly focused on the underlying genetics and neuropathology. Few studies have addressed how AD risk impacts caregivers of adults with DS. This study aimed to explore the perceived impact of AD development in adults with DS on caregivers by assessing caregiver knowledge, concerns, effect on personal life, and resource utilization via a 40-question (maximum) online survey. Survey distribution by four DS organizations and two DS clinics resulted in 89 caregiver respondents. Only 28 caregivers correctly answered all three AD knowledge questions. Caregivers gave an average AD concern rating of 5.30 (moderately concerned) and an average impact of possible diagnosis rating of 6.28 (very strong impact), which had a significant negative correlation with the age of the adult with DS (p = .009). Only 33% of caregivers reported utilization of resources to gain more information about the AD and DS association, with low levels of perceived usefulness. Our data reveal caregivers' misconceptions about AD development in DS, underutilization of available resources, and substantial concerns and perceived impacts surrounding a possible AD diagnosis. This study lays the foundation for how the medical community can better serve caregivers of aging adults with DS.

Innes, A., McCabe, L., & Watchman, K.
Caring for older people with an intellectual disability
Maturitas, 2012 Aug;72(4):286-95. doi: 10.1016/j.maturitas.2012.05.008.
Abstract: This review critically evaluates the available research literature on aging among people with an intellectual disability. 42 papers meeting the review inclusion criteria are presented under three themes: studies with a service user perspective (13), studies of carers of older people with ID (14) and studies of service provision for older people with ID (15). User view specific findings relate to concerns about accommodation; experiences of services; and perceptions of aging; with a common underlying finding from all user focused themes that of unmet need. Carer specific findings relate to fear of the future; experiences of older carers; and planning for the future. Services themes reflect the debate over specialist or generalist services as people age; accommodation; retirement from day services; and staff training. Overall this review reveals a lack of robust research evidence concerning the lives of older people with ID and a need for more research that directly engages with older people with ID and their carers.

Iulita, M.F., Garzón Chavez, D., Klitgaard Christensen, M., Valle Tamayo, N., Plana-Ripoll, O., Rasmussen, S.A., Roqué Figuls, M., Alcolea, D., Videla, L., Barroeta, I., Benejam, B., Altuna, M., Padilla, C., Pegueroles, J., Fernandez, S., Belbin, O., Carmona-Iragui, M., Blesa, R., Lleó, A., Bejanin, A., & Fortea, J.
Association of Alzheimer disease with life expectancy in people with Down syndrome.
JAMA Netw Open. 2022 May 2;5(5):e2212910. doi: 10.1001/jamanetworkopen.2022.12910. PMID: 35604690; PMCID: PMC9127560.
Abstract: People with Down syndrome have a high risk of developing Alzheimer disease dementia. However, penetrance and age at onset are considered variable, and the association of this disease with life expectancy remains unclear because of underreporting in death certificates. The authors assessed whether the variability in symptom onset of Alzheimer disease in Down syndrome is similar to autosomal dominant Alzheimer disease and to assess its association with mortality. This study combines a meta-analysis with the assessment of mortality data from US death certificates (n = 77 347 case records with an International Classification of Diseases code for Down syndrome between 1968 to 2019; 37 900 [49%] female) and from a longitudinal cohort study (n = 889 individuals; 46% female; 3.2 [2.1] years of follow-up) from the Down Alzheimer Barcelona Neuroimaging Initiative (DABNI). A meta-analysis was conducted to investigate the age at onset, age at death, and duration of Alzheimer disease dementia in Down syndrome. PubMed/Medline, Embase, Web of Science, and CINAHL were searched for research reports, and OpenGray was used for gray literature. Studies with data about the age at onset or diagnosis, age at death, and disease duration were included. Pooled estimates with corresponding 95% CIs were calculated using random-effects meta-analysis. The variability in disease onset was compared with that of autosomal dominant Alzheimer disease. Based on these estimates, a hypothetical distribution of age at death was constructed, assuming fully penetrant Alzheimer disease. These results were compared with real-world mortality data. The authors found that in this meta-analysis, the estimate of age at onset was 53.8 years (95% CI, 53.1-54.5 years; n = 2695); the estimate of age at death, 58.4 years (95% CI, 57.2-59.7 years; n = 324); and the estimate of disease duration, 4.6 years (95% CI, 3.7-5.5 years; n = 226). Coefficients of variation and 95% prediction intervals of age at onset were comparable with those reported in autosomal dominant Alzheimer disease. US mortality data revealed an increase in life expectancy in Down syndrome (median [IQR], 1 [0.3-16] years in 1968 to 57 [49-61] years in 2019), but with clear ceiling effects in the highest percentiles of age at death in the last decades (90th percentile: 1990, age 63 years; 2019, age 65 years). The mortality data matched the limits projected by a distribution assuming fully penetrant Alzheimer disease in up to 80% of deaths (corresponding to the highest percentiles). This contrasts with dementia mentioned in 30% of death certificates but agrees with the mortality data in DABNI (78.9%). Important racial disparities persisted in 2019, being more pronounced in the lower percentiles (10th percentile: Black individuals, 1 year; White individuals, 30 years) than in the higher percentiles (90th percentile: Black individuals, 64 years; White individuals, 66 years). These findings suggest that the mortality data and the consistent age at onset were compatible with fully penetrant Alzheimer disease. Lifespan in persons with Down syndrome will not increase until disease-modifying treatments for Alzheimer disease are available

Jacobs, J., Schwartz, A., McDougle, C.J., & Skotko, B.G.
Rapid clinical deterioration in an individual with Down syndrome
American Journal of Medical Genetics, Part A 9999A:1-4 DOI 10.1002/ajmg.a.37674
Abstract: A small percentage of adolescents and young adults with Down syndrome experience a rapid and unexplained deterioration in cognitive, adaptive, and behavioral functioning. Currently, there is no standardized work-up available to evaluate these patients or treat them. Their decline typically involves intellectual deterioration, a loss of skills of daily living, and prominent behavioral changes. Certain cases follow significant life events such as completion of secondary school with friends who proceed on to college or employment beyond the individual with DS. Others develop this condition seemingly unprovoked. Increased attention in the medical community to clinical deterioration in adolescents and young adults with Down syndrome could provide a framework for improved diagnosis, evaluation, and treatment. This report presents a young adult male with Down syndrome who experienced severe and unexplained clinical deterioration, highlighting specific challenges in the systematic evaluation and treatment of these patients.

Jacobs, P., Watchman, K., Wilkinson, H., Hoyle, L., & McGenily, L.
Experiences of people with intellectual disability and dementia: A systematic review.
Journal of Applied Research in Intellectual Diabilities, 2022 Dec 23. doi: 10.1111/jar.13063. Epub ahead of print. PMID: 36562340.
Abstract: Dementia disproportionately affects people with intellectual disability. Most qualitative studies explore their experiences by utilising proxy-reports. A smaller number of studies illustrate the possibility of exploring perspectives directly from people with intellectual disability and dementia. This systematic review synthesized findings from existing studies (n = 8) that involve people with intellectual disability and dementia as participants to understand their experiences of dementia. Searches were conducted using CINAHL, PsychInfo and Social Services Abstracts. Findings include descriptions of changes in individual functioning, a narrowing of social worlds and of how people made sense of the changes despite often having no knowledge of their dementia diagnosis. Additionally, discussion focuses on how people's experiences are shaped by their environments. People's subjective experiences and views on how dementia affected their lives were more ambiguous compared to contextual data and the descriptive portrayal of how people's lives had changed. Thus, this review highlights the importance of researchers needing consider how to facilitate conversations with people about their experiences and the ethics involved in conducting qualitative research with people with intellectual disability and dementia, particularly how respond to participants not knowing about their dementia diagnosis. Spending time with participants over a longer period, getting to know how people communicate and the use of visual aids or everyday items were examples of approaches that supported research involvement. The review recognized the complexities of speaking to people with intellectual disability about dementia, challenges views that people with intellectual and dementia cannot be involved in research and makes recommendations to support inclusion in future studies.

Jamieson-Craig, R., Scior, K., Chan, T., Fenton, C., & Strydom, A.
Reliance on carer reports of early symptoms of dementia among adults with

intellectual disabilities
Journal of Policy and Practice in Intellectual Disabilities, 2010, 7(1), 34 - 41. https://doi.org/10.1111/j.1741-1130.2010.00245.x
Abstract: As clinicians often rely on carer reports to identify adults with intellectual disabilities (ID) with early signs of dementia, this study focused on carer-reported symptoms to ascertain whether carer reports of decline in everyday function would be a more effective screening method to detect possible cases of dementia than reports of memory decline in older adults with ID. Subjects were 154 participants who were reassessed along with their carers two to three years after baseline. A questionnaire for carer-reported change in everyday function and the Dementia Questionnaire for Persons with Mental Retardation (DMR) were used to assess carer views of everyday function and memory. The diagnosis of dementia was confirmed by two psychiatrists working independently. Participants who developed dementia displayed both everyday function and memory decline. Overall, decline in everyday function appeared to be the best indicator of new dementia cases. Retrospective carer report of change in everyday function was as good as, if not better than, prospective ratings to identify dementia; however, in those with mild ID, memory change was a better indicator of dementia, while in those with more severe ID, decline in everyday function was a better indicator. Decline in everyday function (whether prospective change from baseline or reported retrospectively by carers) appears to be a better screening method for dementia than memory decline, particularly for participants with moderate/severe ID.

Janicki, M.P.
Health status and comorbidities of adults with dementia and ID-implications for screening and healthcare
Innovation in Aging, 2017 July, 1(suppl_1), 712. https://doi.org/10.1093/geroni/igx004.2554
Abstract: Declining health status and comorbidities are often markers for associated mild cognitive impairment or dementia. A group of community-dwelling adults with intellectual disability diagnosed with dementia (along with controls) have been tracked longitudinally with respect to their health and function. With time, the dementia-capable group home residents with dementia are showing significantly varied health status and comorbidities as well as marked behavioral changes. By tracking the health and function longitudinally, outcome information can pinpoint markers that are associated with premorbid dementia and can help health providers maintain surveillance over select functions and health conditions of those adults already affected. Screening instruments, incorporating these markers, can more precisely be used to identify at-risk adults for ADRD and aid providers design remediation programs earlier.

Janicki, M.P.
Dementia capable group homes for adults with intellectual disability: development implications
Innovation in Aging, 2018 Nov, 2(suppl_1), 532-533. https://doi.org/10.1093/geroni/igy023.1967
Abstract: Community housing for adults with intellectual disability (ID) and dementia (AD) is becoming more prevalent. An opportunistic longitudinal study (2011-2018) of group homes (GHs) for such adults examined a number of resident and administrative factors. Study followed 2 cohorts of adults with ID, 15 w/dementia and 15 matched controls (CO), over 7 years (8 time intervals). Initial cohort of 15 AD (Xage=59.1; NDS=5) resided in 3 (5-bed-each) dementia GHs. COs resided in general GHs/apartments. Over 7 years, 8 AD died, and 7 replacements (Xage=58.7; NDS=2) were added to the study. Instruments captured subject characteristics, behavior/health/function, staffing, time spent on caregiving, and administrative factors. Findings noted differences in health and function factors between the ADs and COs. Deaths occurred at age-norms. Comparative co-morbidities showed AD residents had significantly more classic cognitive and physical health issues associated with dementia and physical debilitation. An ebb and flow was observed of residents affected by dementia when an agency has multiple dementia GHs, as well as variations in staffing patterns and periods of intensity of care during the day. Over time, the 3 dementia GHs, by inter-home transfers and selective new admissions, have trended toward stage/level specific care settings. Findings can help with planning long-term use of such GHs and can providing dementia capable care for adults with ID in a community-based specialized setting. With dementia affecting an increasing number of adults w/ID (due to aging) more attention needs to be given to functional GHs providing in-community long-term mid-stage and advanced dementia capable care.

Janicki, M.P.
Quality outcomes in group home dementia care for adults with intellectual disabilities.
Journal of Intellectual Disability Research, 2011, 55(8), 763-776. [doi: 10.1111/j.1365-2788.2011.01424.x].
Abstract: Dementia, as a public health challenge, is a phenomenon vexing many care organizations providing specialized residential and family supports for older adults with intellectual disabilities. With increasing survivorship to ages when risk is greatest, expectations are that many more adults in service will present with cognitive decline and diagnosed dementia as they grow older. As persons with dementia present with new needs, there is often a call for a reorientation of services. With respect to residential supports, agencies may need to adapt current methods of care, with particular attention to providing care in small group homes. However, dementia-related care also must be quality care and applicable standards need to be met. The author reviewed relevant policy and practice organizational guidelines and applied research literature addressing components of care and service provision that were critical to quality care and that were consistent with professional practice. Examined were the nuances and contributing factors of quality dementia care and it was proposed that quality of care criteria need to be universally applicable and serve as a framework for adapting extant residential environments and make them 'dementia-capable'. It is proposed that efforts to evaluate dementia-related care provision with respect to quality need to consider quality of care provision components such as (1) clinically relevant early and periodic assessment; (2) functional modifications in the living setting; (3) constructive staff education and functionality for stage-adapted care; and (4) flexible long-term services provision that recognizes and plans for progression of decline and loss of function.

Janicki, M.P.
On-going activities of the National Task Group on Intellectual Disabilities and Dementia Practices
Gerontologist, 2018, 56(Suppl_3), 573.
Abstract: The National Task Group on Intellectual Disabilities and Dementia Practices (NTG), organized in 2011, has been actively involved in stimulating development of services for people with intellectual disabilities (ID) affected by dementia. The NTG has created several sets of practice guidelines, a screening and early detection instrument for use by families and agencies, web-based informational materials,and a national curriculum on ID and dementia, and has undertaken the provision of workforce development workshops across the US on dementia and ID. The NTG works to compliment the activities being undertaken under the National Plan to Address Alzheimer's Disease and consults with various national organizations focusing on dementia and lifelong disabilities. The goal of the NTG is to continue to affect change and improve the quality of community dementia care provision corresponding with National Plan updates.

Janicki, M.P.
Small group homes as "dementia-capable" settings for people with intellectual disabilities and early stage dementia.
Alzheimer's & Dementia, 3(3S), Part_3, First published: 01 July 2007. https://doi.org/10.1016/j.jalz.2007.04.369
Abstract: Localities are beginning to feel the impact of the growing number of older adults with lifelong intellectual disabilities (ID) who are also affected by dementia. Many local organizations are attempting to adapt their support and residential services to help this group be served more effectively within the community. Yet, questions have been raised as to the models that may most reasonably be used and how to address the discordance between traditional ID service and "dementia-capable" services. Investigated was how localities and organizations have adapted to the onset of dementia and address early stage care demands and determine practices that are effective in promoting "dementia-capable" care. Several studies were conducted to determine how government entities and local providers are adapting services to identify models prevalent in the provider sector, and specifically to identify staff training needs, physical and environmental adaptations, and differential time spent by staff in providing dementia care. Data showed that most US states are not prepared to address growing onset of dementia in select parts of the ID population, that responses to early stage service needs have been mostly handled by local entities and service organizations, that most have not developed extensive training programs for staff and are experimenting with best practice methods to deliver care - primarily via small group homes - and that dementia care takes up a disproportionate amount of staff time in small care settings. To address early stage dementia related services in the most effective manner, a concerted effort needs to be in place to aid local service entities adapt services to

dementia-related presentations among ID clientele, set up coordinated training for staff, secure funds for adapting group homes for community "dementia-capable" care, and construction of clinical support services and augmentation of family support services for parents and other kin carers.

Janicki, M..P.
Community-based housing and NPI-care practices for adults with intellectual disability and dementia
AAIC 2020 Conference, Poster presentation, July 30. 2020. *Alzheimer's & Dementia*, 16(S8), First published: 07 December 2020.
https://doi.org/10.1002/alz.047061
Abstract: Aging persons with intellectual disability (ID) represent a vulnerable population with respect to cumulative neuropathological conditions, including dementia. Adults with Down syndrome (DS), a subset, have a recognized high risk for Alzheimer's disease. With dementia present, how to provide post-diagnostic supports is challenging. Dementia care group homes (GHs) along with NPIs are emerging as a mode for providing out-of-home community supports. Data from a longitudinal study provide insights on what care organizations need to consider when organizing specialty group home care. The study, begun in 2011, followed three co-located homes providing NPIs to 15 adults with dementia. Findings revealed trajectories of changes over time, housing need/function level patterning, and health status outcomes. Key findings noted 3 age-of-admission clusters (X=50.5; ?=57.1; X=66.8); overall mortality (Xage-death=65.4; ID=69.3; DS=56.3) – half of original entrants died within 7 years; age at entry (X= 59.1); years from entry to death (X= 5.4 yrs); LOS (X=49.4 months/4.12 yrs); morbidities (number of co-morbidities decreased among survivors). In same period, 8/15 deaths in GHs vs 3/15 deaths in Controls. NPI-related practices included day program activities (adults in mid- to later stages were engaged in regular off-site day activities that agency provided; adults with advanced dementia remained in homes), staffing patterns differed based on level of care – more staff assigned to homes with residents with advanced dementia, and staff training included dementia capable communications, engagement, and managing daily routines. Trends showed adults with Down syndrome were admitted to homes earlier but had more life-years in the GHs than older adults admitted at later age but who succumbed earlier to disease complications. Dementia care GHs should expect varied trajectories of decline; mortality linked to complexity of pre-existing conditions and progression of dementia; changes in the focus of care needs over time (including advanced dementia and end-of-life care). Dementia care GHs can enable provision of in-community group housing and quality care in accord with stage-defined functional changes and needs if structured in a planful way (factoring in dementia-stage, dementia type, mortality expectations, health status, patterns of care needs, dementia-related behaviors, aging-related issues, and probable trajectories of decline of the residents).

Janicki,M.P., Dalton, A.J., McCallion, P., Davies Baxley, D., & Zendell, A.
Group home care for adults with intellectual disabilities and Alzheimer's disease
Dementia, 2005, 4, 361-385. https://doi.org/10.1177/1471301205055028
Abstract: The growing numbers of individuals with intellectual disabilities affected by Alzheimer disease and related dementias has raised new challenges for community care providers. This paper examines means of providing community group home-based care in a sample of care providers in five different countries. The aim is to identify trends that have emerged. Two samples of group homes for adults with intellectual disabilities affected by dementia were studied to determine: (1) what are the physical characteristics of the homes; (2) what physical environmental adaptations have been made in response to behavioral deterioration expressed by residents with dementia, and (3) what are the demands on staff resulting from dementia care. The first sample of group homes in five countries provided comparative international data on home designs, staffing, costs, and residents. The second sample, drawn from homes in the USA and the UK, provided data on the impact of dementia. Findings revealed staffing and design of homes varied but generally abided by general practices of dementia care; homes relied on existing resources to manage changes posed by dementia care; programmatic and environmental adaptations were implemented to address progression of dementia; and residents with dementia presented more demands on staff time with respect to hygiene maintenance and behavior management when compared to other residents not affected by dementia.

Janicki, M. P., Heller, T., Seltzer, G., & Hogg, J.
Practice guidelines for the clinical assessment and care management of Alzheimer's disease and other dementias among adults with intellectual disability
Journal of Intellectual Disability Research, 1996, 40, 374-382
Abstract: The AAMR/IASSID practice guidelines, developed by an international workgroup, provide guidance for stage-related care management of Alzheimer's disease, and suggestions for the training and education of carers, peers, clinicians, and program staff. The guidelines suggest a three step intervention activity process, that includes: (1) recognizing changes, (2) conducting assessments and evaluations, and (3) instituting medical and care management. They provide guidance for public policies that reflect a commitment for aggressive care of people with Alzheimer's disease and intellectual disability, and avoidance of institutionalization solely because of a diagnosis of dementia. [This report is available also on www.aamr.org at the following URL: http://161.58.153.187/Bookstore/Downloadables/index.shtml]

 **Janicki, M.P., McCallion, P., & Dalton, A.J.**
Supporting people with dementia in community settings.
In M.P. Janicki & A.F. Ansello (Eds.), Community Supports for Aging Adults with Lifelong Disabilities.
pp. 387-413
Baltimore, Maryland: Paul H. Brookes Publishing (2000)
Abstract: Due to the "greying" of the nation's population, dementia associated with Alzheimer's disease and other causes, has become another challenge for providers of services to adults with intellectual disabilities. In this book chapter, the authors explore the factors, policies, and support structures that can help agencies provide continued "aging-in-place" dementia-capable care, develop "in-place progression" dementia specific programs, or chose alternative care settings. It also explores some features of dementia-related behaviors that may need to be taken into account in program design and makes suggestions for staff training and planning for dementia programs.

Janicki, M.P., McCallion, P., & Dalton, A.J.
Dementia-related care decision-making in group homes for persons with intellectual disabilities
Journal of Gerontological Social Work, 2002, 38(½), 179-196.
https://doi.org/10.1300/J083v38n01_04
Abstract: The number of age-associated pathologies is increasing, with the increase in the number of elderly persons. One such age-associated condition, Alzheimer's disease and related dementias, affects a significant number of adults with intellectual disability (ID), in particular those with Down syndrome. Many affected adults live in small community group homes or with their families. How to provide sound and responsive community care is becoming a challenge for agencies faced with an increasing number of affected adults. This study reports the outcome of a survey of group homes serving adults with ID and dementia, explores the onset, duration and effects of dementia and their impact on planning for community care of adults with ID. It also examines emerging community care models that provide for "dementia capable" supports and services. Two models, "aging in place," and "in place progression" are examined with regard to care practices and critical agency decision making. An approach, the ECEPS model, for responding to dementia is offered.

Janicki, M.P. & Dalton A.J.
Care management, diagnostic and epidemiologic considerations in adults with intellectual disabilities and Alzheimer disease
British Journal of Developmental Disabilities, 1996, 42(Supplement), s84
Abstract: Review of the process and outcome of the Invitational International Colloquium on Alzheimer Disease among Persons with intellectual Disabilities held in Minneapolis, Minnesota (USA) and the subsequent development of a set of international practice guidelines and reports on the assessment, epidemiology, and care management of adults with intellectual disabilities affected by dementia.

 **Janicki, M.P., & Dalton, A.J.**
Dementia in developmental disabilities
In N. Bouras (Ed.), Psychiatric and Behavioral Disorders in Developmental Disabilities and Mental Retardation (1999)
pp. 121-153
Cambridge: Cambridge University Press
Abstract: This book chapter provides a brief overview of the current status of knowledge about dementia and its relationship to intellectual disability, touching on current developments in the evaluation of possible comorbid psychiatric, medical and age-associated conditions. The clinical presentation of dementia is examined as well as relevant contemporary issues related to diagnosis,

assessment, and care management. Lastly, questions of dementia policy and suggestions for training programs on dementia and intellectual disability are addressed.

Janicki, M.P., & Dalton, A.J.
Dementia and public policy considerations
In M.P. Janicki & A.J. Dalton (eds.), Dementia, Aging, and Intellectual Disabilities (1999)
pp. 388-414
Philadelphia: Brunner-Mazel
Abstract: This book chapter examines a number of the major public policy considerations related to the aging of adults with intellectual disabilities who evidence change due to dementia. Specifically addressed is the changing structure of at-risk adult populations with intellectual disabilities in service systems, the programmatic and policy issues raised by providers attempting to cope with these changes, needs for further training, education and dissemination of information on aging, and lastly, the challenges and policy imperatives to be confronted with the new millennium.

Janicki, M.P., & Dalton, A.J.
Dementia, aging, and intellectual disabilities: A handbook
488pp.
Philadelphia: Brunner-Mazel [http://www.taylorand francis.com] (1999)
Abstract: 21 chapter text on dementia issues and intellectual disabilities. Six parts: Introduction, Biomedical considerations, Assessment considerations, Clinical considerations, Program considerations, and Education and policy considerations. Text provides most up-to-date information available about Alzheimer's disease and related dementias as they affect persons with mental disabilities. Text examines biology and physiology of dementia, neurological and medical complications associated with dementia, best practices to meet the needs of aging persons with intellectual disabilities, policy issues raised by the growing number of older adults with ID, and case studies of affected individuals. Contains glossary of terms, and appendices with AAMR/IASSID practice guidelines for dementia diagnosis and care management in adults with intellectual disabilities, as well as Newroth & Newroth guidelines for coping with Alzheimer's disease in persons with Down syndrome.

Janicki, M.P., & Dalton, A.J.
Prevalence of dementia and impact on intellectual disability services
Mental Retardation, 2000, 38, 277-289.
Abstract: A statewide survey, conducted to ascertain the administrative prevalence of dementia in adults with an intellectual disability, found a prevalence of about 3% of the adult service population over the age of 40 years (a rate of 28/1000), 6.1% of the population over the age of 60 years, and 12.1% of the population over the age of 80 years (or rates of 68.7/1000 and 121.3/1000, respectively). The rate of dementia was consistent with that for adults in the general population, except for those adults with Down syndrome (who made up a third of the overall group) who had a much higher rate: 22.1% among adults age 40 and older and 56.4% among adults age 60 and older. Onset was observed to occur in the mid-60s (early 50s for Down syndrome). Alzheimer-type dementia was the most frequent diagnosis. Late-onset seizures were reported in about 12% of the cases. With the occurrence of dementia expected to rise proportionately with the increase of longevity among adults with an intellectual disability, it is clear that care systems will have to raise the "index of suspicion" among staff and families, adapt to become "dementia capable," and improve their diagnostic and technical resources, as well as their community-based care management supports.

Janicki, M.P., & Dalton, A.J.
Alzheimer disease in a select population of older adults with mental retardation
The Irish Journal of Psychology, 1993, 14(1), 38-47
doi:10.1080/03033910.1993.10557913
Abstract: Twenty New York State Office of Mental Retardation and Developmental Disabilities district offices were surveyed to determine the prevalence of older adults with mental retardation who also had suspected or diagnosed Alzheimer disease (AD). The survey identified 123 individuals in this category, about 1% of an overall case roll of 10,878 people of 40 years of age and older for the 17 reporting offices. Of these 123 individuals, 64% were people with Down syndrome. Typically, a person with suspected Alzheimer disease was identified through staff observation or report of changes in behaviour leading to suspicion of Alzheimer disease. Information on training needs indicated that a workshop on AD was a primary need. Training in various content areas was indicated, including confirming suspicions, diagnosis, programming, and individual program planning.

Janicki, M.P., McCallion. P., Splaine, M., Santos, F.H., Keller, S.M., & Watchman, K.
Consensus statement of the international summit on intellectual disability and dementia related to nomenclature
Intellectual and Developmental Disabilities, 2017, 55(5), 338–346. DOI: 10.1352/1934-9556-55.5.338.
Abstract: A working group of the 2016 International Summit on Intellectual Disability and Dementia was charged to examine the terminology used to define and report on dementia in publications related to intellectual disability (ID). A review of related publications showed mixed uses of terms associated with dementia or causative diseases. Like general applications, language related to dementia in ID field often lacked precision and could lead to a misunderstanding of the condition(s) under discussion. Most articles related to ID and dementia reporting clinical or medical research generally provided a definition of dementia or related terms; social care articles tended toward term use without definition. Toward terminology standardization within studies/ reports on dementia and ID, the Summit recommended (a) gaining familiarity with dementia-related diagnostic, condition-specific, and social care terms (as identified in the working group's report), (b) creating a guidance document on accurately defining and presenting information about individuals or groups referenced, and (c) that in reports on neuropathologies or cognitive decline or impairment, definitions are used and data include subjects' ages, sex, level of ID, residential situation, basis for dementia diagnosis, presence of Down syndrome (or other risk conditions), years from diagnosis, and if available, scores on objective measures of changing function.

Janicki, M.P. Zendell, A., & DeHaven. K.
Coping with dementia and older families of adults with Down syndrome.
Dementia, 2010, 9(3), 391-407.
Abstract: The authors studied a group of older carers of aging adults with Down syndrome (DS) to ascertain what effects such caregiving may have on them given the presence or possibility of age-associated decline or dementia. The study also examined the comparative levels of care provided, key signs noted when decline was beginning, the subjective burden experienced, and what were the key associated health factors when carers faced a changed level of care. The authors found that this group was made up of long-term, committed carers who had decided early to look after their relative with DS over their lifetime. When faced with the onset and ongoing progression of dementia, their commitment was still evident as evidenced by adopting physical accommodations and finding ways to continue to provide care at home, while also seeking help from outside sources. Most saw a family or group home environment as the place of choice for their relative with DS when they decided they could no longer offer care. The study did not ascertain any burn-out or significant health related problems associated with their continued caregiving save for their concerns about day-to-day strain and what will happen in the future.

Janicki, M.P., & McCallion, P.
A group home cluster model for providing community-based dementia care.
Paper presented at the 21st annual conference of Alzheimer Europe, Warsaw, Poland. (2011, October).
C:/Users/Janicki%20Associates/Downloads/P3.4%20Janicki%20(2).pdf
Abstract: Paper reports on a study undertaken of an innovation group home program operated by a provider organization serving older adults with intellectual disabilities. The provider built three co-located group homes for five adults within a neighborhood setting. Each of the adults resident at the homes have some degree of diagnosed dementia. The adults were both males and females, all were age 50+, and some had Down syndrome. The homes are staffed by paid staff working 24/7. The residents were studied for health co-morbidities, program activities, and degrees of impairment and compared with a matched group of adults without dementia. The study examined administrative and programmatic factors related to the operation of the homes, as well as shifts in characteristics related to their intellectual disability and the effects of dementia

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Jaycock, S., Persaud, M. & Johnson, R.
The effectiveness of dementia care mapping in intellectual disability residential services: A follow-up study.
Journal of Intellectual Disabilities, 2006, 10(4), 365-375.

Abstract: The authors present a follow-up to exploratory work published in the Journal of Intellectual Disabilities in 2001. This article describes a study that aimed to assess the effectiveness of dementia care mapping in supporting practice improvement in intellectual disability residential services. An average of 9 hours of observational data were collected using dementia care mapping in relation to 14 adults with severe or profound intellectual disabilities (but who not have dementia). Sixteen interviews were also undertaken with staff over a 4 month period.The findings provided a detailed picture of the activities and interactions between the participants involved in the study and raised some issues about 'organizational culture' when developing person-centered approaches. These data have helped strengthen the case that care mapping has the potential to be a useful addition to the existing repertoire of tools to support effective practice improvement and person-centered planning.

Jenkins, R., Davies, R., Sardi, I., Llewellyn, P., Northway, R., O'Connor, C., Trudgeon, C. & Keeling, D.
Adults with learning disabilities presenting with dementia [Final Report - May 2009]
University of Glamorgan (Faculty of Health, Sport, and Science), 2008. 78pp. https://www.choiceforum.org/docs/pdem.pdf
Abstract: As life expectancy improves, increasing numbers of people with intellectual (learning) disabilities are affected by dementia. Supporting someone with a dual diagnosis of intellectual disability and dementia presents unique challenges to both those responsible for delivering services and carers. In response to these challenges Gwent Healthcare NHS Trust developed a Dementia Care Pathway in 2005. The aims were: (1) to ensure early and appropriate diagnosis, (2) to provide a co-ordinated approach to assessment and intervention, (3) to develop intervention plans that will support both client and carers, (4) to provide a process for monitoring the person over time, and (5) to support, carers, clients, and professionals via the proviso of information and training. The study demonstrated that the stated aims of the Dementia Care Pathway are being met. Stage one results found that the training provided to paid support staff was received enthusiastically and had a positive impact on knowledge, confidence, and competence levels in relation to working iwth clients who have, or may develop, dementia. In most cases the knowledge was well maintained at six month follow-up. There is some evidence, however, that although staff who attended the training reported it to be 'interesting' and 'very relevant, but they still doubted its usefulness in practical day-to-day situations when caring for their clients. In order for the potential of the pathway to be further maximized attention should be paid to increasing the level of information given to carers, widening contributions to pathway review meetings, and educating carers on the diverse range of interventions possible through the pathway.

Jervis, N., & Prinsloo, L.
How we developed a multidisciplinary screening project for people with Down's syndrome given the increased prevalence of early onset dementia
British Journal of Learning Disabilities, 2008, 36 (1), 13–21.
Abstract: Much research has identified an increased prevalence of dementia in adults with Down syndrome when compared with the general population. Neuropathological changes associated with Alzheimer's dementia in the brain have been found in most people with Down syndrome who die over the age of 35 years. Given the limitations of many assessments for dementia in relation to people with Down syndrome for a single completion, it has been recommended that all people with Down syndrome are assessed at least once in early adulthood in order that they have their own baseline which can be compared with in the future if changes in skills and abilities occur. The authors have had many requests from other services enquiring about their project and how a similar initiative could be set up. Therefore, this article focuses on the way the Manchester Learning Disability Partnership approached screening 135 adults with Down syndrome and details the assessments used, practical considerations, what has been learned and future service implications.

Johannsen, P., & Mai, J.
Alzheimer-type demens og Downs syndrom [Alzheimer-type dementia and Down syndrome] *Article in Danish*
Ugeskrirft for Laeger, 1995 Feb 20, 157(8), 1021-1024.
Abstract: Dementia is seen earlier and more often in patients with Down syndrome (DS) than in the general population. Patients with DS develop neuropathological changes similar to the changes seen in dementia of Alzheimer's type (DAT). The literature concerning DAT in DS is reviewed. The clinical course and differential diagnoses are discussed. Before the diagnosis of DAT is made, treatable causes of dementia should be excluded.

Johannsen, P., Christensen, J.E.J., & Mai, J.
The prevalence of dementia in Down syndrome
Dementia, 1996, 7(4), 221-225.
Abstract: The authors assess the prevalence of clinical dementia in three age groups of persons with Down syndrome in the county of Aarhus, Denmark. Group 1 was composed of 14-16 year olds (n=13), group 2 was composed of 23-29 year olds (n=34), and group 3 was composed of 50-60 year olds (n=25). Of the 85 subjects, 72 (85%) participated. Carers were interviewed and a neurological examination was performed. An EEG was recorded in 50 of the Ss. Definite clinical dementia was defined as a acquired and progressive decline in 4 or more out of 17 items that are considered to indicate dementia in people with Down syndrome. Possible dementia was considered when 1-3 items were affected. Six adults (24%) in group 3 had definite clinical dementia and 6 adults in group 3 and 2 (6%) in group 2 had possible dementia. Authors note that this was the first Danish population-based study of the prevalence of dementia in people with Down syndrome.

Johansson, P.E., & Terenius, O.
Development of an instrument for early detection of dementia in people with Down syndrome
Journal of Intellectual & Developmental Disabilities, 2002, 27(4), 325-345. https://doi.org/10.1080/1366825021000029357
Abstract: The successful detection of early signs of dementia in people with Down syndrome could form a basis for useful early support and for drug treatment. This report describes the development and preliminary application of an interview and test instrument for the assessment of dementia among people with intellectual disability, as well as a framework for diagnosis that combines the findings of an interview and a test with the diagnostic criteria of ICD-10, DSM-IV and NINCDS-ADRDA. From among the number of tests and interview questions developed, those showing the most significant differences between participants in three groups of differing levels of intellectual disability and estimated dementia were kept. Reported are the assumptions for the items used, descriptions of the process and items used, and the associations of test items with predicting the presence of dementia. The authors conclude that a protocol combining testing and interview has promise and potential for detecting early signs of dementia in this population and could prove feasible for use in practice.

Johansson, M., Holst,G., & G Ahlström
Signs in people with intellectual disabilities: Interviews with managers and staff on the identification process of dementia
Journal of Intellectual Disability Research, 2019, 63(8), 649.
Abstract: An increasing number of people with intellectual disability (ID) are reaching older ages and an increased risk of dementia diseases. Staff and managers give support in daily living and can deliver information about residents' changes in behavior. The aim of this Swedish study was to explore the identification process employed by staff and managers to detect signs of suspected dementia in people with ID within intellectual disability services (ID-services). Twenty managers and 24 staff within ID-service were interviewed and qualitative latent content analysis was applied. A model consisting of three themes on three levels of resources for the identification process of signs of suspected dementia emerged from the analysis. On the first level was the time and continuity in the care relationship, which is crucial for identifying and responding to changes in cognitive ability that indicate dementia. On the second level, the staff identifies deficiencies in their own knowledge, seek support from colleagues and managers within their workplace and, on the third level, outside their workplace. Staff and managers expressed needs for guidance and education from specialists in dementia and primary healthcare. This finding indicates an urgent need for intervention research and digital support for staff in dementia care.

Johnson, N., Fahey, C., Chicoine, B., Chong, G., & Gitelman, D.
Effects of donepezil on cognitive functioning in Down syndrome
American Journal on Mental Retardation, 2003,108(6), 367-372
Abstract: This study to determined whether donepezil, an acetylcholinesterase inhibitor, would improve cognitive functioning in 19 subjects with Down syndrome and no dementia. They were assigned to either a donepezil or placebo group. Cognitive functioning and caregiver ratings were measured at baseline, 4 weeks, and 12 weeks. With the exception of one area (language), no improvement was noted in any of the cognitive subtests, behavioral scores, or caregiver ratings. Subjects in the donepezil group showed an improvement in language scores compared to subjects in the placebo group. The results suggest that donepezil may improve language performance in subjects with Down syndrome and no

dementia, but further studies need to be done on a larger group to confirm this result.

Jokinen, N.S., Janicki,M.P., Hogan,M., & Force, L.T.
The middle years and beyond: Transitions and families of adults with Down syndrome
Journal on Developmental Disabilities, 2012, 18(2), 59-69.
Abstract: Normally expected transitions connect the various periods of life. Often these transitions are prompted by life events that require adaptation to a changed circumstance and may challenge both individual and family quality of life. Such transitions may be *planful* (proactive) or *demand* (reactive). Little, however, has been written about the nature of such transitions and how they specifically affect older-aged families of adults with Down syndrome. Such families are often predominate lifelong carers of adults with Down syndrome. Drawing on research and experience, the authors examined three transition points from a family perspective. Each of these points of change requires that people adapt and may lead to various outcomes, including at times outcomes that are unexpected, stressful, and challenging. The three points of transition examined include moving away from the parental home, changes occurring within a residential service (e.g., staff changes, relocations), and the reactions to the onset and course of dementia. Vignettes and quotes illustrate the complexities of these transitions and show that, even with planful management, often such transitions can go awry and produce unpredictable outcomes.

Jokinen, N.
The content of available practice literature in dementia and intellectual disability.
Dementia: The International Journal of Social Research and Practice, 2005, 4(3), 327-339.
Abstract: Adults with intellectual disability are living to ages seen within the general population and they, too, are at risk of developing dementia. This review was to conducted to identify the nature and content of the literature related to adults with intellectual disability and dementia and bring together guidelines for services and staff providing care. The preponderance of work between 1995 and 2004 focuses on the biomedical, diagnosis and assessment aspects of the disease. Although guidelines exist, there is a lack of published literature on the efficacy of practice strategies to guide the provision of daily care. Future research is discussed that could support continued community living and high quality of life during all stages of the disease.

Jokinen, N., Service, K., Marsack-Topolewski, C., & Janicki, M.P.
Support-staging model for caregivers of adults with intellectual disability affected by dementia
AAIC2020,Poster presentation, July 30, 2020. *Alzheimer's & Dementia,* 16(S8), First published: 07 December 2020. https://doi.org/10.1002/alz.047274
Abstract: Adults with intellectual disability (ID) and dementia are a sub-population of persons who are often un- or underserved. Most adults with ID are integrated within the general community (living autonomously, or in apartments/group residences); but significant numbers also reside with their families, particularly adults with Down syndrome. Family help/counseling approaches, such as the New York University-Caregiver Intervention (NYUCI), might benefit from a support-staging model assessment focus on what specific aid a family requires to meet their needs. Patterns of such needs have been identified that can help with providing dementia-capable care. Objective needs include: (a) information on signs and symptoms, (b) diagnostic advice, (c) understanding behavioral changes and managing dementia-related behaviors, (d) adapting homes, (e) determining daily routines most conducive to calming, (f) planning for the future, (g) finding and navigating resources, and (h) responding to end-of-life needs. Subjective needs include: (a) being informed at time of diagnosis and throughout the course of dementia, (b) coping with a profound sense of loss from knowing the diagnosis, (c) fearing the future [including financial concerns], (d) formulating long-term plans, (e) accessing community-based coordinated care, (f) facing difficulties from the medical community, (g) feeling overwhelmed by caregiving demands, (h) feeling a sense of isolation and abandonment, and (i) facing end-of-life issues. A working group emanating from the 2016 Glasgow Summit on Intellectual Disability and Dementia organized a schema encapsulating these concerns into a support-staging model. The schema suggested four fluid stages: Diagnostic (seeking cause of changes in function, abilities, personality), Explorative (exploring dementia capable interventions), Adaptive (coping with and managing the symptoms/changes), and Closure (resolving / relief from responsibilities). Using this schema, a process (utilizing the NYUCI) is underway to operationalize a support-staging assessment instrument which would enable counseling staff to frame the state of a family's concerns, build relationships through this knowledge of the caregiver and provide tailored services to them. The outcome will enable systematic coding and organizing both objective and subjective data so that specific interventions and counseling can be adapted to meet both intermittent and continuous caregiver needs.

Jokinen, N., Janicki, M.P., Keller, S.M., McCallion, P., Force,L.T., & National Task Group on Intellectual Disabilities and Dementia Practices
Guidelines for structuring community care and supports for people with intellectual disabilities affected by dementia
Journal of Policy and Practice in Intellectual Disabilities, 2013, 10(1), 1–24.
Abstract: To assist families and organizations in their planning for extended care that accompanies the diagnosis of dementia, the National Task Group on Intellectual Disabilities and Dementia Practices (NTG) in the United States adopted a set of practice guidelines covering the period from when suspicions are aroused to when care ends with eventual death. These guidelines are drawn from the research literature as well as clinical experiences and demonstrated best practices. The guidelines delineate what actions should be undertaken and are presented in a manner that reflects the progressive nature of prevalent dementias. To enable the development of the most appropriate and useful services and care management for adults with intellectual disabilities affected by dementia, the NTG adopted the staging model generally accepted for practice among generic dementia services. The staging model follows the flow from a prediagnosis stage when early recognition of symptoms associated with cognitive decline are recognized through to early, mid, and late stages of dementia, and characterizes the expected changes in behavior and function. In keeping with the National Plan to Address Alzheimer's Disease recommendations for earlier and more widespread efforts to detect possible symptoms, the guidelines cite the application of the NTG-Early Detection Screen for Dementia as a first step in documenting early signs of cognitive and functional changes among people with intellectual disabilities. The guidelines also provide information on nonpharmacological options for providing community care for persons affected by dementia as well as commentary on abuse, financial, managing choice and liability, medication, and nutritional issues.

Jokinen, N., Gomiero, T., Watchman, K., Janicki, M.P. Hogan, M., Larsen, F., Service, K., & Crowe, J .
Perspectives on family caregiving of people aging with intellectual disability affected by dementia: Commentary from the International Summit on Intellectual Disability and Dementia
Journal of Gerontological Social Work, 61(4), 411-431. DOI: 10.1080/01634372.2018.1454563
Abstract: This article, an output of the 2016 International Summit on Intellectual Disability and Dementia, examines familial caregiving situations within the context of a support-staging model for adults with intellectual disability (ID) affected by dementia. Seven narratives offer context to this support-staging model to interpret situations experienced by caregivers. The multidimensional model has two fundamental aspects: identifying the role and nature of caregiving as either primary (direct) or secondary (supportive); and defining how caregiving is influenced by stage of dementia. We propose staging can affect caregiving via different expressions: (1) the "diagnostic phase," (2) the "explorative phase," (3) the "adaptive phase," and (4) the "closure phase." The international narratives illustrate direct and indirect caregiving with commonality being extent of caregiver involvement and attention to the needs of an adult with ID. We conclude that the model is the first to empirically formalize the variability of caregiving within families of people with ID that is distinct from other caregiving groups, and that many of these caregivers have idiosyncratic needs. A support-staging model that recognizes the changing roles and demands of carers of people with ID and dementia can be useful in constructing research, defining family-based support services, and setting public policy.

Kåhlin, I., Kjellberg, A., & Hagberg, J-E.
Ageing in people with intellectual disability as it is understood by group home staff
Journal of Intellectual & Developmental Disability, 2016. 41(1),1-10. https://doi.org/10.3109/13668250.2015.1094038
Abstract: The number of older residents in group homes for people with intellectual disability (ID) is increasing. This interview study was focused on how group home staff address issues of ageing and being old among people with ID. Twelve members of staff at 4 different group homes in Sweden were interviewed. Findings revealed old age as something unarticulated in the group home. Group home staff felt unprepared to meet age-related changes in residents. The study

also revealed that group home staff had a one-tracked way of describing the process of ageing among people with ID, which was seemingly rooted in a medical paradigm of disability. Based on this study's findings, we suggest that there is a need to raise issues and give guidance related to ageing and ID in disability policy documents to support the development of a formal culture that addresses old age and ID in disability services.

Kalsy, S., McQuillan, S., Adams, D., Basra, T., Konstantinidi, E., Broquard, M., Peters, S., Lloyd, V., & Oliver, C.
A proactive psychological strategy for determining the presence of dementia in adults with Down syndrome: Preliminary description of service use and evaluation
Journal of Policy and Practice in Intellectual Disabilities, 2005, 2(2), 116-125. https://doi.org/10.1111/j.1741-1130.2005.00025.x
Abstract: The authors describe and assess the experience of providing proactive screening for dementia in older adults with intellectual disabilities (ID) through a dedicated clinical psychology service within the National Health Service in England. Subjects were the first 18 participants who were referred to the clinical service or were identified as showing early signs of probable dementia in a proactive screening strategy. The screening process involved combining neuropsychological, behavioral, and health data with information from a clinical assessment of the presenting problem in a case series approach. The process of psychological assessment and formulation is illustrated together with an outline of the psychological interventions employed for early-, mid-, and late-stage dementia. An appraisal of the service strategy showed that a dedicated psychology service for dementia assessment can be effective when offering a defined and workable psychological response to the increasing presentation of dementia-associated behaviors among people with ID. Ancillary services included supporting carers in contributing to the assessment and intervention process so as to ensure appropriately responsive and respectful care management for the person with ID and dementia. The authors recommend that a multimodal stage model of intervention founded on direct performance and informant-based assessments (within a framework of differential diagnosis) be employed in supporting people with ID and dementia.

Kalsy, S., McQuillan, S., Oliver. C., Hall, S.
Manual for the "Assessment for Adults with Developmental Disabilities" (A.A.D.S.) Questionnaire
School of Psychology, University of Birmingham, Edgbaston, Birmingham B15 2TT (2000).
Scales designed to assess behaviors associated with dementia and levels of caregiving. American version is available for download from www.uic.edu/orgs/rrtcamr/dementia.

Kalsy, S., Heath, R., Adams, D., & Oliver, C.
Effects of training on controllability attributions of behavioural excesses and deficits shown by adults with Down syndrome and dementia.
Journal of Applied Research in Intellectual Disabilities, 2007, 20(1), 64 -68.
Abstract: Whereas there is a knowledge base on staff attributions of challenging behavior, there has been little research on the effects of training, type of behavior and biological context on staff attributions of controllability in the context of people with intellectual disabilities and dementia. A mixed design was used to investigate the effects of three factors on care staff attributions of the controllability of challenging behavior. Pre- and post-training measures were administered to participants (n = 97) attending training on ageing, dementia and people with intellectual disabilities. Authors found no significant effects of diagnosis or type of behavior on attributions were found. There was a significant increase in knowledge after training ($P < 0.001$) and training was found to significantly decrease the attribution of controllability ($P < 0.001$). Conclusion was that the results suggest that training that focuses on aspects of change relevant to behavior can favorably influence care staff's knowledge and attributions of controllability within the context of people with Down syndrome and dementia.

Keater, D.B., Phelen, M.J., Taylor, L., Doran, E., Krinsky-McHale, S., Price, J., Ballard, E.E., Kreisl, W.C., Hom, C., Nguyen, D., Pulsifer, M., Lai, F., Rosas, D.H., Brickman, A.M., Schupf, N., Yassa, M.A., Silverman, W., & Lott, I.T.
Down syndrome: Distribution of brain amyloid in mild cognitive impairment
Alzheiimer's Dementia: Diagnosis, Assessment & Disease Monitoriing, 2020, 12(1), e12013. https://doi.org/10.1002/dad2.12013
Abstract: Down syndrome (DS) is associated with a higher risk of dementia. We hypothesize that amyloid beta (Aß) in specific brain regions differentiates mild cognitive impairment in DS (MCI-DS) and test these hypotheses using cross-sectional and longitudinal data. 18F-AV-45 (florbetapir) positron emission tomography (PET) data were collected to analyze amyloid burden in 58 participants clinically classified as cognitively stable (CS) or MCI-DS and 12 longitudinal CS participants. The study confirmed our hypotheses of increased amyloid in inferior parietal, lateral occipital, and superior frontal regions as the main effects differentiating MCI-DS from the CS groups. The largest annualized amyloid increases in longitudinal CS data were in the rostral middle frontal, superior frontal, superior/middle temporal, and posterior cingulate cortices. Authors note that this study helps us to understand amyloid in the MCI-DS transitional state between cognitively stable aging and frank dementia in DS. The spatial distribution of Aß may be a reliable indicator of MCI-DS in DS.

📖 **Keller, S.M., Janicki, M.P., & Esralew, L.**
Dementia: screening, evaluation, diagnosis and management
In: Rubin I.L., Merrick J., Greydanus D.E., Patel D.R. (eds) *Health Care for People with Intellectual and Developmental Disabilities across the Lifespan. Springer*, Cham. https://doi.org/10.1007/978-3-319-18096-0_116.
Abstract: The emergent acknowledgment of the increase rate of dementia among adults with intellectual and developmental disability (IDD) has led to the recognition of a lifespan approach to securing and providing services with relevant supports. Consideration of what is most needed in older age has taken on more prominence. Service organizations are responding to the emergence of age-associated neuropathologies as coincident to lifelong conditions and are attempting to adapt their services for continued community care until death. The historical focus on aging among adults with IDD and the recent focus on dementia has heightened awareness of the latter age needs of adults with IDD - and in particular those at-risk of or already affected by dementia. The positive outcome of these public health initiatives is that more dementia-capable services are being developed, technologies are improving, and there is an increased interest in maintaining quality of life through to the end-of-life, irrespective of the nature and complexity of conditions prevalent in older age.

Kerins, G., Petrovic, K., Bruder, M.B., & Gruman, C.,
Medical conditions and medication use in adults with Down syndrome: A descriptive analysis.
Down Syndrome Research and Practice, 2008, 12(2), 141-147. [http://www.down-syndrome.org/reports/2009/reports-2009.pdf]
Abstract: Authors the presence of medical conditions and medication use within a sample of adults with Down syndrome. The author employed a retrospective chart review using a sample of 141 adults with Down syndrome and age range of 30 to 65 years. They identified 23 categories of commonly occurring medical conditions and 24 categories of medications used by adults with Down syndrome. From their work, the authors concluded that approximately 75% of older adults with Down syndrome in their sample experienced memory loss and dementia. Hypothyroidism, seizures, and skin problems also occurred commonly. The prevalence of cancer (i.e., solid tumors) and hypertension was extremely low. Older adults with Down syndrome used anticonvulsants more often than younger adults with Down syndrome. The use of multivitamins and medications such as pain relievers, prophylactic antibiotics, and topical ointments was common.

📖 **Kerr, D.**
Down's syndrome and dementia
76 pp.
Birmingham, UK: Venture Press (1997)
Abstract: Text providing a comprehensive review of issues and practices relative to adults with Down syndrome affected by Alzheimer's disease. Covered are a range of topics related to care management, including assessment of need, communication, creating a therapeutic environment, how to maintain skills, and dealing with challenging behaviors. Also covered are specific interventions and supporting carers.

Kerr, D., Cunningham, C., & Wilkinson, H.
Learning disability and dementia Are we prepared?
Journal of Dementia Care, 2006, May/June, 17-19
Abstract: None provided

Kerr, D., Cunningham, C. & Wilkinson, H.
Responding to the pain experiences of people with a learning difficulty and dementia

Joseph Rowntree Foundation, 2006. (The Homestead, 40 Water End, York YO30 6WP). https://www.choiceforum.org/docs/jrfpainfindings.pdf
Abstract: The report explores knowledge and practice in relation to pain recognition and management amongst direct support staff, members of community learning [intellectual] disability teams and general practitioners. It also examines the understanding and experiences of pain amongst people with a learning difficulty [intellectual disability] and dementia. It identifies the dilemmas and obstacles to effective pain management, and reports on examples of good practice. The report found that the pain experiences and management of people with a learning difficulty [intellectual disabiltiy] who have dementia mirrored findings in relation to people in the general population. It did, however, identify extra and compounding issues in relation to people with a learning difficulty [intellectual disability]. The authors proffer recommendations for practitioners and service providers.

Kirk, L.J., Hick, R., & Laraway, A.
Assessing dementia in people with learning disabilities: The relationship between two screening measures.
Journal of Intellectual Disabilities, 2006, 10(4), 357-364.
Abstract: As life expectancy increases for people with intellectual disabilities, the impact of dementia on people with intellectual disabilities and their families, carers and services is becoming more apparent. Psychological services for intellectual disabilities are receiving an increasing number of referrals requesting dementia assessment. Health and social care services are adapting to the diverse needs of an ageing population with intellectual disabilities. The authors describe a study investigating the relationship between two assessments for dementia in people with intellectual disabilities. Carers of people with intellectual disabilities over the age of 50 (or 40 if the individual had Down syndrome) completed the Dementia Questionnaire for Mentally Retarded People (DMR) and the Adaptive Behavior Scale–Residential and Community (ABS). Overall, the two questionnaire measures showed significant relationships. However, results suggested that both assessments have clinical value in informing individual needs and aiding diagnosis. The authors discuss the Implications for both clinical and social care services.

Kirwan, R., Sheerin, F., McGlinchey, E., McCallion, P., & McCarron, M.
Functional loss in older adults with intellectual disabilities and dementia.
Learning Disability Practice, 2022, 25(5), ??-?? doi: 10.7748/ldp.2022.e2184
Abstract: Diagnosing dementia in people with intellectual disabilities can be challenging due to pre-existing cognitive impairment. In this population, functional loss could be an early indicator of dementia, but the relationship between functional loss and dementia is not well understood. This means there is a risk of delayed diagnosis and therefore a delay in care planning. To identify and compare the prevalence and age distribution of dementia in people with intellectual disabilities, differentiating between those with Down's syndrome and those with an intellectual disability not attributable to Down's syndrome, and to identify and compare functional loss in people with intellectual disabilities who have developed dementia, differentiating between those with Down's syndrome and those with an intellectual disability not attributable to Down's syndrome. This was a secondary analysis of data from waves 1 and 3 of the Intellectual Disability Supplement to the Irish Longitudinal Study on Ageing (IDS-TILDA), a nationally representative study in Ireland of older adults with intellectual disabilities as they age. Functional loss was determined by participants' level of difficulty with activities of daily living (ADLs) and instrumental activities of daily living (IADLs). The prevalence of dementia was higher among participants with Down's syndrome than among those with an intellectual disability not attributable to Down's syndrome, and the age distribution of dementia differed markedly between the two groups. Both groups experienced similar patterns of difficulty with ADLs and IADLs. IADLs posed more difficulty than ADLs, and among ADLs self-care activities posed the most difficulty. It is important to continue to investigate functional loss in older adults with intellectual disability who have developed dementia, as this should lead to earlier assessment and care planning.

Knox, K., Stanley, J., Hendrix, J.A., Hillerstrom, H., Dunn, T., Achenbach, J., Chicoine, B.A., Lai, F., Lott, I., Stanojevic, S., Howlett, S.E., & Rockwood, K.
Development of a symptom menu to facilitate Goal Attainment Scaling in adults with Down syndrome-associated Alzheimer's disease: a qualitative study to identify meaningful symptoms.
Journal of Patient Reported Outcomes, 2021 Jan 11, 5(1), 5. doi: 10.1186/s41687-020-00278-7.
Abstract: : As life expectancy of people with Down syndrome (DS) increases, so does the risk of Alzheimer's disease (AD). Identifying symptoms and tracking disease progression is especially challenging whenever levels of function vary before the onset of dementia. Goal Attainment Scaling (GAS), an individualized patient-reported outcome, can aid in monitoring disease progression and treatment effectiveness in adults with DS. Here, with clinical input, a validated dementia symptom menu was revised to facilitate GAS in adults living with Down Syndrome-associated Alzheimer's disease (DS-AD). Four clinicians with expertise in DS-AD and ten caregivers of adults living with DS-AD participated in semi-structured interviews to review the menu. Each participant reviewed 9-15 goal areas to assess their clarity and comprehensiveness. Responses were systematically and independently coded by two researchers as 'clear', 'modify', 'remove' or 'new'. Caregivers were encouraged to suggest additional items and recommend changes to clarify items. Median caregiver age was 65 years (range 54-77). Most were female (9/10) with 15 years of education (10/10). Adults with DS-AD had a median age of 58 years (range 52-61) and either a formal diagnosis (6/10) or clinical suspicion (4/10) of dementia. The initial symptom menu consisted of 67 symptoms each with 2-12 descriptors (589 total). The clinicians' adaptation yielded 58 symptoms each with 4-17 descriptors (580 total). Of these 580 descriptors, caregivers identified 37 (6%) as unclear; these were reworded, and one goal area (4 descriptors) was removed. A further 47 descriptors and one goal area were added to include caregiver-identified concepts. The final menu contained 58 goal areas, each with 7-17 descriptors (623 total). A comprehensive symptom menu for adults living with DS-AD was developed to facilitate GAS. Incorporating expert clinician opinion and input from caregivers of adults with DS-AD identified meaningful items that incorporate patient/caregiver perspectives.

Kobayashi, S., Yamamoto-Mitani, N., Nagata, S., & Murashima, S
End-of-life care for older adults with dementia living in group homes in Japan
Japan Journal of Nursing Science, 2008, 5(1), 31-40. doi: 10.1111/j.1742-7924.2008.00097.x.
Abstract: The aim of this study was to elucidate the components of end-of-life care provided to older adults with dementia who live in group homes (GHs) in Japan. The number of GHs in Japan is rapidly increasing. Although GHs were originally not established to care for elderly people with advanced-stage dementia, many residents remain in the GH even after their stage of dementia advances; thus, end-of-life care is required. Interviews were conducted with seven GH administrators on their experience in providing end-of-life care to their residents. The constant comparative approach was used for data collection and analysis. Four themes emerged as essential components of end-of-life care in the GH setting: (i) maintaining a familiar lifestyle; (ii) minimizing physical and mental discomfort; (iii) proactively utilizing desirable medical care; and (iv) collaborating with family members. The combination of the four components seems to be a unique characteristic of end-of-life care in GHs in Japan. These findings may be used to establish a framework for end-of-life care at GHs.

Koehl, L.H., Harp, J., Van Pelt, K.L., Head, E., & Schmitt, F.A.
Longitudinal assessment of dementia measures in Down syndrome.
Alzheimer's & Dementia (Amst), 2020, Nov 14, 12(1), e12075. doi: 10.1002/dad2.12075. eCollection 2020.
Abstract: Early detection of dementia symptoms is critical in Down syndrome (DS) but complicated by clinical assessment barriers. The current study aimed to characterize cognitive and behavioral impairment using longitudinal trajectories comparing several measures of cognitive and behavioral functioning. Measures included global cognitive status (Severe Impairment Battery [SIB]), motor praxis (Brief Praxis Test [BPT]), and clinical dementia informant ratings (Dementia Questionnaire for People with Learning Disabilities [DLD]). One-year reliability was assessed using a two-way mixed effect, consistency, single measurement intraclass correlation among non-demented participants. Longitudinal assessment of SIB, BPT, and DLD was completed using linear mixed effect models. One-year reliability (n = 52; 21 male) was moderate for DLD (0.69 to 0.75) and good for SIB (0.87) and BPT (0.80). Longitudinal analysis (n = 72) revealed significant age by diagnosis interactions for SIB ($F(2, 115.02) = 6.06$, $P = .003$), BPT ($F(2, 85.59) = 4.56$, $P = .013$), and DLD ($F(2, 103.56) = 4.48$, $P = .014$). SIB progression (PR) had a faster decline in performance versus no-dementia (ND) ($t(159) = -2.87$; $P = .013$). Dementia had a faster decline in BPT performance versus ND ($t(112) = -2.46$; $P = .041$). PR showed quickly progressing scores compared to ND ($t(128) = -2.86$; $P = .014$). Current measures demonstrated moderate to good reliability. Longitudinal analysis revealed that SIB, BPT, and DLD changed with age depending on diagnostic progression; no change rates were dependent on baseline cognition, indicating usefulness across

a variety of severity levels in DS.

Koenig, B.R.
Aged and dementia care issues for people with an intellectual disability: Best practices (vol. 2).
80 pp.
Brighton, South Australia: MINDA, Inc. (1995)
Abstract: Text covering a range of useful topics related to service provision for dementia among persons with intellectual disabilities. Highly detailed chapters cover health issues, physical decline, behavioral changes, and social aspects. Specific remedial information is provided on communication issues and adapting the environment. A chapter also addresses counseling strategies, examining a diverse range of approaches.

Kozma, C.
Down syndrome and dementia.
Topics in Geriatric Rehabilitation, 2008, 24(1), 41–53. doi: 10.1097/01.TGR.0000311405.01555.3b
Abstract: Down syndrome (DS) is one of the most common genetic conditions with an estimated incidence of 1 in 750 in the general population. It results from an extra chromosome 21 with the total chromosome count being 47 instead of the normal 46. The classic features of DS include hypotonia, atypical facial characteristics, an increased incidence of major and minor anomalies, vision and hearing deficits, other health problems, and intellectual disabilities. People with DS are living longer and experiencing premature aging, specifically Alzheimer disease (AD). The incidence of AD among adults with DS varies significantly according to studies averaging between 11% to 22% for people aged 40 to 49 years, 24.9% for people aged 50 to 59 years, and 25.6% to 77% for people older than 60 years. All studies indicate an early onset of AD as well as an exponential increase in prevalence with age. Furthermore, senile plaques and neurofibrillary tangles, the neuropathological characteristics of AD, are seen in the brain of all people with DS. Annual screening for AD should become part of routine medical practice of older adults with DS, because an early diagnosis is important for comprehensive care.

Krinsky-McHale SJ, & Silverman W.
Dementia and mild cognitive impairment in adults with intellectual disability: issues of diagnosis.
Developmental Disabilities Research Reviews, 2013,18(1).31-42. doi: 10.1002/ddrr.1126.
Abstract: Individuals with intellectual disability (ID) are now living longer with the majority of individuals reaching middle and even "old age." As a consequence of this extended longevity they are vulnerable to the same age-associated health problems as elderly adults in the general population without ID. This includes dementia, a general term referring to a variety of diseases and conditions causing substantial loss of cognitive ability and functional declines; adults with Down syndrome are at especially high risk. A great deal of recent effort has focused on the very earliest detectable indicators of decline (and even prodromal stages of dementia-causing diseases). A condition called mild cognitive impairment (MCI) has been conceptually defined as a decline in functioning that is more severe than expected with typical brain aging but not severe enough to meet criteria for a diagnosis of dementia. Consensus criteria for both dementia and MCI have been developed for typically developing adults but are of limited applicability for adults with ID, given their pre-existing cognitive impairments. Early diagnosis will continue to be of growing importance, both to support symptomatic treatment and to prevent irreversible neuropathology when interventions are developed to slow or halt the progression of underlying disease. While the intellectual and developmental disabilities field has for some time recognized the need to develop best-practices for the diagnosis of MCI and dementia, there remains a pressing need for empirically based assessment methods and classification criteria.

Krinsky-McHale, S.J., & Silverman. W.
Dementia and mild cognitive impairment in adults with intellectual disability: Issues of diagnosis
Developmental Disabilities Research Reviews, 2013, 18, 31- 42. https://doi.org/10.1002/ddrr.1126
Abstract: Individuals with intellectual disability (ID) are now living longer with the majority of individuals reaching middle and even "old age." As a consequence of this extended longevity they are vulnerable to the same age-associated health problems as elderly adults in the general population without ID. This includes dementia, a general term referring to a variety of diseases and conditions causing substantial loss of cognitive ability and functional declines; adults with Down syndrome are at especially high risk. A great deal of recent effort has focused on the very earliest detectable indicators of decline (and even prodromal stages of dementia-causing diseases). A condition called mild cognitive impairment (MCI) has been conceptually defined as a decline in functioning that is more severe than expected with typical brain aging but not severe enough to meet criteria for a diagnosis of dementia. Consensus criteria for both dementia and MCI have been developed for typically developing adults but are of limited applicability for adults with ID, given their pre-existing cognitive impairments. Early diagnosis will continue to be of growing importance, both to support symptomatic treatment and to prevent irreversible neuropathology when interventions are developed to slow or halt the progression of underlying disease. While the intellectual and developmental disabilities field has for some time recognized the need to develop best-practices for the diagnosis of MCI and dementia, there remains a pressing need for empirically based assessment methods and classification criteria.

Krinsky-McHale, S.J., Zigman, W.B., Lee, J.H., Schupf, N., Pang, D., Listwan, T., Kovacs, C., & Silverman. W.
Promising outcome measures of early Alzheimer's dementia in adult with Down syndrome
Alzheimers Dement (Amst). 2020; 12(1): e12044.
Published online 2020 Jul 5. doi: 10.1002/dad2.12044
Abstract: Adults with Down syndrome (DS) are at high risk for developing Alzheimer's disease (AD) and its associated dementia, warranting the development of strategies to improve early detection when prevention is possible. Using a broad battery of neuropsychological assessments, informant interviews, and clinical record review, we evaluated the psychometrics of measures in a large sample of 561 adults with DS. We tracked longitudinal stability or decline in functioning in a subsample of 269 participants over a period of 3 years, all initially without indications of clinically significant aging-related decline. Results identified an array of objective measures that demonstrated sensitivity in distinguishing individuals with incident "mild cognitive impairment" (MCI-DS) as well as subsequent declines occurring with incident dementia. Several instruments showed clear promise for use as outcome measures for future clinical trials and for informing diagnosis of individuals suspected of experiencing early signs and symptoms of a progressive dementia process.

Kruse, B., Müller, S,V., & Sappok, T.
Demenz bei Menschen mit geistiger Behinderung [Dementia in people with intellectual disabilities]
*NeuroTransmitte*r, 2019, 30(3), 36-45. doi: 10.1007/s15016-019-6594-y,
Abstract :People with a mental disability are getting older and age-typical diseases are more common in them, including dementia. The symptoms and the diagnostic process, however, differ significantly in comparison to people without intellectual disabilities. However, recognizing the diagnosis is essential for adequate treatment and support.

Kuske, B., Wolfe, C., Gövert, U., & Müller, S.V.
Early detection of dementia in people with an intellectual disability – A German pilot study
Journal of Applied Research in Intellectual Diabilities, 2017, Dec, 30(S1), 49-57. https://doi.org/10.1111/jar.12347
Abstract: This study investigated the application of a newly developed neuropsychological assessment, the Wolfenbütteler Dementia Test for Individuals with Intellectual Disabilities (WDTIM) in combination with the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID). The instruments were evaluated in a prospective 2-year follow-up study. A total of 102 people with an intellectual disability were assessed at 6-month intervals. Data were analysed using qualitative and statistical analyses. Four groups of individuals emerged from the analysis: (1) confirmed suspicion, (2) no suspicion, (3) questionable suspicion and (4) early suspicion. Significant differences were found between groups 1 and 2. The WDTIM could be administered to 90%–100% of all participants exhibiting mild-to-moderate intellectual disability and to 50% with severe intellectual disability. The WDTIM was shown to have good applicability to people with mild-to-moderate intellectual disability and to be appropriate for detecting cognitive changes. Using the two instruments in combination achieved greater accuracy in reinforcing a dementia suspicion than did using the DSQIID alone.

Lane, A.M., Reed, M.B., & Hawranik, P.
Aging individuals with Down syndrome and dementia as teachers: Learnings from

staff in a developmental disability program in long-term care, Journal of *Gerontological Nursing,* 2019, 45(5), 17-22. Doi: 10.3928/00989134-20190328-02
Abstract: Older adults with Down syndrome (DS) and dementia are an emerging sub-population. With much longer life spans than decades ago, issues have arisen as to where these aging adults will live and how nurses and other staff in facilities can provide effective care to these individuals. This article presents a research study that examined the learnings of nurses and staff members working within a western Canadian program for older adults with DS and dementia. These learnings include: the importance of learning from each other; importance of collaboration; how individuals with developmental disabilities communicate; older adults with DS and dementia differ from older adults with dementia; and residents' impact on staff.

Lautarescu, B.A., Holland, A.J., & Zaman, S.H.
The early presentation of dementia in people with Down syndrome: A systemic review of longitudinal studies
Neuropsychology Review, 2017, Mar, 27(1), 31-45. doi: 10.1007/s11065-017-9341-9. Epub 2017 Mar 13. PMID: 28289920
Abstract: Adults with Down syndrome (DS) are at a very high risk of developing early onset Alzheimer's disease (AD) due to trisomy of chromosome 21. AD is preceded by a prolonged prodromal "pre-clinical" phase presenting with clinical features that do not fulfil the diagnostic criteria for AD. It is important to clinically characterize this prodromal stage to help early detection of the disease as neuropathology of AD is almost universal by the fifth decade in DS. There is a lack of knowledge of the trajectory of decline associated with the onset of dementia in this population and early signs may be overlooked or misdiagnosed, negatively affecting the quality of life of those affected and the use of early pharmacological or psychosocial interventions. The objective of this systematic review was to evaluate the published literature on longitudinal data in order to identify the cognitive and behavioral changes occurring during the prodromal and early stages of AD in this population. Fifteen peer-reviewed articles met the inclusion criteria, including a total number of 831 participants, with the duration between baseline and follow up varying from 1 year to 47 years. Results suggest that, compared to the general population for which short-term (episodic) memory loss is the most common indicator associated with the onset of AD, in people with DS, executive dysfunction and Behavioural and Psychological Symptoms of Dementia (BPSD) are commonly observed during pre-clinical and early stages and may precede memory loss. The review highlights the importance of using a broad spectrum of assessments in the context of heterogeneity of symptoms. Theoretical and practical implications are discussed, as well as the need for further research.

Lazenby, T.
The Impact of aging on eating, drinking, and swallowing function in people with Down's syndrome
Dysphagia, 2008, 23, 88–97. https://doi.org/10.1007/s00455-007-9096-1
Abstract: Many people with Down's syndrome (DS) experience eating, drinking, and swallowing (EDS) difficulties, which can potentially lead to life-threatening conditions such as malnutrition, dehydration, and aspiration pneumonia. As the life expectancy of people with DS continues to improve, there is an increasing need to examine how the aging process may further affect these conditions. Published research studies have yet to address this issue; therefore, this article draws on the literature in three associated areas in order to consider the dysphagic problems that might develop in aging people with DS. The areas examined are EDS development in children and adolescents with DS, EDS changes associated with aging, and EDS changes associated with **dementia** of the Alzheimer's type (DAT) because this condition is prevalent in older adults with DS. This article concludes that unlike in the general population, the aging process is likely to cause dysphagic difficulties in people with DS as they get older. Therefore, it is suggested that longitudinal studies are needed to examine the specific aspects of EDS function that may be affected by aging and concomitant conditions in DS.

Lao, P.J., Gutierrez, J., Keator, D., Rizvi, B., Banerjee, A., Igwe, K.C., Laing, K.K., Sathishkumar, M., Moni, F., Andrews, H., Krinsky-McHale, S., Head, E., Lee, J.H., Lai, F., Yassa, M.A., Rosas, H.D., Silverman, W., Lott, I.T., Schupf, N., & Brickman, A.M.
Alzheimer-related cerebrovascular disease in Down syndrome
Annals of Neurology, 2020, Dec, 88(6), 1165-1177. doi: 10.1002/ana.25905. Epub 2020 Oct 9.
Abstract: Adults with Down syndrome (DS) develop Alzheimer disease (AD) pathology by their 5th decade. Compared with the general population, traditional vascular risks in adults with DS are rare, allowing examination of cerebrovascular disease in this population and insight into its role in AD without the confound of vascular risk factors. We examined in vivo magnetic resonance imaging (MRI)-based biomarkers of cerebrovascular pathology in adults with DS, and determined their cross-sectional relationship with age, beta-amyloid pathology, and mild cognitive impairment or clinical AD diagnostic status. Participants from the Biomarkers of Alzheimer's Disease in Down Syndrome study (n = 138, 50 ± 7 years, 39% women) with MRI data and a subset (n = 90) with amyloid positron emission tomography (PET) were included. We derived MRI-based biomarkers of cerebrovascular pathology, including white matter hyperintensities (WMH), infarcts, cerebral microbleeds, and enlarged perivascular spaces (PVS), as well as PET-based biomarkers of amyloid burden. Participants were characterized as cognitively stable (CS), mild cognitive impairment-DS (MCI-DS), possible AD dementia, or definite AD dementia based on in-depth assessments of cognition, function, and health status. There were detectable WMH, enlarged PVS, infarcts, and microbleeds as early as the 5th decade of life. There was a monotonic increase in WMH volume, enlarged PVS, and presence of infarcts across diagnostic groups (CS < MCI-DS < possible AD dementia < definite AD dementia). Higher amyloid burden was associated with a higher likelihood of an infarct. The findings highlight the prevalence of cerebrovascular disease in adults with DS and add to a growing body of evidence that implicates cerebrovascular disease as a core feature of AD and not simply a comorbidity.

Lao, P., Zimmerman, M. E., Hartley, S. L., Gutierrez, J., Keator, D., Igwe, K. C., ... & Brickman, A. M.
Obstructive sleep apnea, cerebrovascular disease, and amyloid in older adults with Down syndrome across the Alzheimer's continuum.
Sleep Advances, 2022 May 5, 3(1):zpac013. doi: 10.1093/sleepadvances/zpac013.
Abstract: We determined the extent to which obstructive sleep apnea (OSA) is associated with increased cerebrovascular disease and amyloid burden, and the relation of the two processes across clinical Alzheimer's disease (AD) diagnostic groups in adults with Down syndrome (DS). Adults with DS from the Biomarkers of Alzheimer's Disease in Down Syndrome (ADDS) study were included given available research MRI (n = 116; 50 ± 8 years; 42% women) and amyloid PET scans (n = 71; 50 ± 7 years; 39% women) at the time of analysis. Participants were characterized as cognitively stable (CS; 64%), with mild cognitive impairment-DS (MCI-DS; 23%), with possible AD dementia (5%), or with definite AD dementia (8%). OSA was determined via medical records and interviews. Models tested the effect of OSA on MRI-derived cerebrovascular biomarkers and PET-derived amyloid burden, and the moderating effect of OSA and AD diagnosis on biomarkers. OSA was reported in 39% of participants, which did not differ by clinical AD diagnostic group. OSA was not associated with cerebrovascular biomarkers but was associated with greater cortical amyloid burden. White matter hyperintensity (WMH) volume (primarily in the parietal lobe), enlarged perivascular spaces, and cortical and striatal amyloid burden were greater across clinical AD diagnostic groups (CS<MCI-DS<possible AD<definite AD). OSA increased the differences in WMH volumes across clinical AD diagnostic groups, primarily in the frontal and temporal lobes. Adults with DS and OSA had greater amyloid burden and greater cerebrovascular disease with AD. Importantly, OSA may be a modifiable risk factor that can be targeted for intervention in this population at risk for AD.

Lautarescu, B.A., Holland, A.J., & Zaman, S.H.
The early presentation of dementia in people with Down syndrome: a systematic review of longitudinal studies
Neuropsychology Review, 2017, Mar, 27(1), 31-45. doi: 10.1007/s11065-017-9341-9. Epub 2017 Mar 13.
Abstract: Adults with Down syndrome (DS) are at a very high risk of developing early onset Alzheimer's disease (AD) due to trisomy of chromosome 21. AD is preceded by a prolonged prodromal "pre-clinical" phase presenting with clinical features that do not fulfil the diagnostic criteria for AD. It is important to clinically characterise this prodromal stage to help early detection of the disease as neuropathology of AD is almost universal by the fifth decade in DS. There is a lack of knowledge of the trajectory of decline associated with the onset of dementia in this population and early signs may be overlooked or misdiagnosed, negatively affecting the quality of life of those affected and the use of early pharmacological or psychosocial interventions. The objective of this systematic review is to evaluate the published literature on longitudinal data in order to identify the cognitive and behavioural changes occurring during the prodromal and early stages of AD in this population. Fifteen peer-reviewed articles met the

inclusion criteria, including a total number of 831 participants, with the duration between baseline and follow up varying from 1 year to 47 years. Results suggest that, compared to the general population for which short-term (episodic) memory loss is the most common indicator associated with the onset of AD, in people with DS, executive dysfunction and Behavioural and Psychological Symptoms of Dementia (BPSD) are commonly observed during pre-clinical and early stages and may precede memory loss. The review highlights the importance of using a broad spectrum of assessments in the context of heterogeneity of symptoms. Theoretical and practical implications are discussed, as well as the need for further research.

Lessov-Schlaggar, C.N., Del Rosario, O.L., Morris, J.C., Ances, B.M., Schlaggar, B.L., & Constantino, J.N.
Adaptation of the Clinical Dementia Rating Scale for adults with Down syndrome
Journal of Neurodevelopmental Disorders, 2019, Dec 16, 11(1), 39. doi:10.1186/s11689-019-9300-2.
Abstract: Adults with Down syndrome (DS) are at increased risk for Alzheimer disease dementia, and there is a pressing need for the development of assessment instruments that differentiate chronic cognitive impairment, acute neuropsychiatric symptomatology, and dementia in this population of patients. We adapted a widely used instrument, the Clinical Dementia Rating (CDR) Scale, which is a component of the Uniform Data Set used by all federally funded Alzheimer Disease Centers for use in adults with DS, and tested the instrument among 34 DS patients recruited from the community. The participants were assessed using two versions of the modified CDR-a caregiver questionnaire and an in-person interview involving both the caregiver and the DS adult. Assessment also included the Dementia Scale for Down Syndrome (DSDS) and the Raven's Progressive Matrices to estimate IQ. Both modified questionnaire and interview instruments captured a range of cognitive impairments, a majority of which were found to be chronic when accounting for premorbid function. Two individuals in the sample were strongly suspected to have early dementia, both of whom had elevated scores on the modified CDR instruments. Among individuals rated as having no dementia based on the DSDS, about half showed subthreshold impairments on the modified CDR instruments; there was substantial agreement between caregiver questionnaire screening and in-person interview of caregivers and DS adults. The modified questionnaire and interview instruments capture a range of impairment in DS adults, including subthreshold symptomatology, and the instruments provide complementary information relevant to the ascertainment of dementia in DS. Decline was seen across all cognitive domains and was generally positively related to age and negatively related to IQ. Most importantly, adjusting instrument scores for chronic, premorbid impairment drastically shifted the distribution toward lower (no impairment) scores

Li, R. S. Y., Kwok, H.W.M., Deb, S., Chui, E.M.C., Chan, L.K., & Leung, D.P.K.
Validation of the Chinese version of the dementia screening questionnaire for individuals with intellectual disabilities (DSQIID-CV).
Journal of Intellectual Disability Research, 2015, Apr, 59(4), 385-95. doi: 10.1111/jir.12173.
Abstract: An increasing number of people with intellectual disabilities (ID) are at risk of developing age-related disorders such as dementia because of a dramatic increase in life expectancy in this population in the recent years. There is no validated dementia screening instrument for Chinese people with ID. The Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) was reported to be a valid, user-friendly, easy-to-use observer-rated instrument. It was developed in the UK and has good psychometric properties. Validation of a Chinese version of the DSQIID will facilitate its application among the Chinese population. The DSQIID was translated into the Chinese version (DSQIID-CV). By purposive sampling, service users with ID aged 40 years or over were recruited through two large centres serving adults with ID in Hong Kong. Carers who had taken care of the participants continuously for the past 6 months were invited to complete the DSQIID-CV. All participants were examined by qualified psychiatrists to determine the presence or absence of dementia.. Two hundred people with ID whose age ranged between 40 and 73 years (mean 51 years, SD=7.34 years) were recruited to the study. A clinical diagnosis of dementia was established in 13 participants. An overall total score of 22 as a screening cut-off provided the optimum levels of specificity (0.995) and sensitivity (0.923). The DSQIID-CV showed good internal consistency (alpha=0.945) for all its 53 items, and excellent test-retest reliability (0.978, n=46) and inter-rater reliability (1.000, n=47). Exploratory factor analysis resulted in a four-factor solution explaining 45% of the total variance. The DSQIID-CV is shown to have robust psychometric properties. It is the first valid and reliable dementia screening instrument for Chinese adults with ID.

Lin, J-D., Lin, L-P., Hsia, Y-C., Hsu, S-W., Wu, C-L., & Chu, C.M.
A national survey of caregivers' perspective of early symptoms of dementia among adults with an intellectual disability based on the DSQIID scale
Research in Autism Spectrum Disorders, 2014, 8(3), 275-280.
Abstract: As life expectancy increases for persons with an intellectual disability, concerns have been raised that there will be an increased demand for health or social services, particularly to address the challenges posed by the problems of dementia. To plan services for people with an intellectual disability who might experience the consequences of aging, an important first step is to obtain epidemiological data on the prevalence of dementia in this vulnerable population. This study aimed to investigate the dementia prevalence rate and its associated demographical factors in adults with an intellectual disability in Taiwan. A national survey was conducted to recruit 460 community residents of at least 45 years of age with an intellectual disability. The Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) was administered to caregivers to determine the symptoms of dementia in adults with an intellectual disability. The results indicated that 16.5% of the adults with an intellectual disability might have dementia conditions (DSQIID score ? 20). After controlling for other factors in a multiple logistic regression analysis, the older adults with intellectual disability (?55 vs. 45–54, OR = 2.594, 95% CI = 1.438–4.679) and those individuals with a comorbid diagnosis of mental illness or neurological disease (with vs. without, OR = 2.826, 95% CI = 1.593–5.012) had a higher risk of dementia than their counterparts. This study suggests that further longitudinal studies are needed to examine the specific aspects of the functions of living and morbidity that might be affected by aging and concomitant conditions in adults with an intellectual disability.

Lin, L.P., Hsu, S.W., Hsia, Y.C., Wu, C.L., Chu, C., & Lin, J.D.
Association of early-onset dementia with activities of daily living (ADL) in middle-aged adults with intellectual disabilities: the caregiver's perspective.
Research in Developmental Disabilities, 2014, 35(3), 626-631. doi: 10.1016/j.ridd.2013.12.015. Epub 2014 Jan 24.
Abstract: Few studies have investigated in detail which factors influence activities of daily living (ADL) in adults with intellectual disabilities (ID) comorbid with/without dementia conditions. The objective of the present study was to describe the relation between early onset dementia conditions and progressive loss of ADL capabilities and to examine the influence of dementia conditions and other possible factors toward ADL scores in adults with ID. This study was part of the "Healthy Aging Initiatives for Persons with an Intellectual Disability in Taiwan: A Social Ecological Approach" project. We analyzed data from 459 adults aged 45 years or older with an ID regarding their early onset symptoms of dementia and their ADL profile based on the perspective of the primary caregivers. Results show that a significant negative correlation was found between dementia score and ADL score in a Pearson's correlation test ($r=-0.28$, $p<0.001$). The multiple linear regression model reported that factors of male gender (ß=4.187, $p<0.05$), marital status (ß=4.79, $p<0.05$), education level (primary: ß=5.544, $p<0.05$; junior high or more: ß=8.147, $p<0.01$), Down's syndrome (ß=-9.290, $p<0.05$), severe or profound disability level (ß=-6.725, $p<0.05$; ß=-15.773, $p<0.001$), comorbid condition (ß=-4.853, $p<0.05$) and dementia conditions (ß=-9.245, $p<0.001$) were variables that were able to significantly predict the ADL score ($R(2)=0.241$) after controlling for age. Disability level and comorbidity can explain 10% of the ADL score variation, whereas dementia conditions can only explain 3% of the ADL score variation in the study. The present study highlights that future studies should scrutinize in detail the reasons for the low explanatory power of dementia for ADL, particularly in examining the appropriateness of the measurement scales for dementia and ADL in aging adults with ID.

Llewellyn, P.
The needs of people with learning disabilities who develop dementia: A literature review
Dementia (London), 2011, 10(2), 235-247.
https://doi.org/10.1177/1471301211403457
Abstract: People with learning disabilities are living longer and are increasingly developing age related conditions including dementia. If this occurs, their medical and social needs pose many challenges for services. A literature review was undertaken of articles published between 1996— 2006. Data was collected relating to the needs of people with learning disabilities and dementia, their carers and their peers. The primary medical need is for timely and accurate diagnosis. There is a multitude of diagnostic tools and advice is available as to

which are most suitable for different client groups. The needs of carers are intertwined with those of people with learning disabilities and dementia and meeting their needs for education, training and increased staff numbers, has proved beneficial. Although multiple services will be responsible for the needs of this client group, there is a consensus that learning disability services should be at the heart of service provision.

Lloyd, V. , Kalsy, S., & Gatherer, A.
The subjective experience of individuals with Down syndrome living with dementia
Dementia, 2007, 6(1), 63-88.
Abstract: An increasing number of studies have begun to explore the subjective experience of individuals with dementia. However, despite the increased prevalence of dementia in individuals with Down syndrome, no such published research has been undertaken within this population. The aim of this study was to explore the perspectives and subjective experiences of six individuals with Down syndrome and dementia. Semi-structured interview accounts were analyzed using Interpretative Phenomenological Analysis, in order to gain a level of understanding concerning the impact of dementia upon respondents' lives and sense of self. Five main themes emerged: (1) Self-image, (2) The Relational Self, (3) Making Sense of Decline,(4) Coping Strategies and (5) Emotional Experience. Whilst the process of adjusting to dementia appeared comparable to the general population, the content of this was influenced by multiple levels of context specific to having a concomitant intellectual disability.

Lloyd, V., Kalsy, S., & Gatherer, A.
Impact of dementia upon residential care for individuals with Down syndrome
Journal of Policy and Practice in Intellectual Disabilities, 2008, 5(1), 33-38.
Abstract Despite the increased prevalence of dementia in individuals with Down syndrome, relatively little is known about its impact upon care provision. Carers may be familiar with the demands of assisting a person with Down syndrome, but generally have little knowledge about the course or impact of dementia. This dissonance may lead to stress, which can have a detrimental effect on the carer and the quality of care for the recipient. In this exploratory study, the authors examined the objective and subjective impact of dementia upon paraprofessional paid carers of individuals with Down syndrome working in residential settings. The study used the Caregiver Activities Scale—Intellectual Disabilities (CAS-ID), the Caregiver Difficulties Scale—Intellectual Disabilities (CDS-ID), and the Maslach Burnout Inventory (MBI). Responses given for these measures by paraprofessional carers of individuals with Down syndrome and dementia (n = 9) were compared with responses from those caring for recipients with Down syndrome and no additional cognitive decline (n = 11). No significant differences were found in the responses from these sets of carers on measures of objective (CAS-ID) or subjective burden (CDS-ID). However, the MBI revealed that carers of individuals with Down syndrome and dementia reported significantly increased levels of emotional exhaustion. Findings suggested that, while even when there is little difference in the level of caregiving tasks or the subjective difficulties of caregiving, the onset of dementia in individuals with Down syndrome resulted in increased emotional exhaustion for carers. Additional factors not considered within this study, such as challenging behavior, may also be pertinent to carer burden.

Lott, J.D.
The rate of decline of social skills across dementing and non-dementing individuals with intellectual disabilities: A longitudinal study.
Dissertation Abstracts International: Section B: The Sciences and Engineering, 2007, 67(8-B), 2007, 4715.
The author sought to establish rate of decline of adaptive skills in a population of individuals with intellectual disability (ID) with dementia compared to similar adults with ID and without dementia, as well as examining the variability of positive and negative social behaviors across diagnostic classes. The participants were matched for age, sex, Down syndrome, and level of ID. The control group was screened for the presence of dementia with the Early Signs of Dementia Checklist. Rate of decline within groups was assessed by the Vineland Adaptive Behavior Scales and changes in positive and negative behaviors were measured by the Matson Evaluation of Social Skills for the Severely Retarded. (MESSIER) Prior to a diagnosis of dementia groups were equivalent. No significant differences were found for adaptive behaviors. Visual analysis of plotted means supports predicted decline in skills for both groups. Significant differences were found across time for positive social skills. Significant correlations were observed between the VABS and the MESSIER Positive domains. The findings provide support for the diagnostic utility of the MESSIER with dementia. However, no support was observed for different variances of negative behaviors across diagnostic groups. This would suggest that the measure of negative behaviors is not supported as a diagnostic tool at this time.

Lott, I.T., & Head, E.
Dementia in Down syndrome: unique insights for Alzheimer disease research
Nat Rev Neurol 15, 135–147 (2019). https://doi.org/10.1038/s41582-018-0132-6
Abstract: Virtually all adults with Down syndrome (DS) show the neuropathological changes of Alzheimer disease (AD) by the age of 40 years. This association is partially due to overexpression of amyloid precursor protein, encoded by APP, as a result of the location of this gene on chromosome 21. Amyloid-ß accumulates in the brain across the lifespan of people with DS, which provides a unique opportunity to understand the temporal progression of AD and the epigenetic factors that contribute to the age of dementia onset. This age dependency in the development of AD in DS can inform research into the presentation of AD in the general population, in whom a longitudinal perspective of the disease is not often available. Comparison of the risk profiles, biomarker profiles and genetic profiles of adults with DS with those of individuals with AD in the general population can help to determine common and distinct pathways as well as mechanisms underlying increased risk of dementia. This Review evaluates the similarities and differences between the pathological cascades and genetics underpinning DS and AD with the aim of providing a platform for common exploration of these disorders.

Lott, I.Y., Doran, E., Nguyen, V.Q., Tournay, A., Movsesyan, N., & Gillenc, D.L.
Down syndrome and dementia: Seizures and cognitive decline
Journal of Alzheimer's Disease, 2012, 29(1),177-185.
doi:10.3233/JAD-2012-111613
Abstract: The objective of this study was to determine the association of seizures and cognitive decline in adults with Down syndrome (DS) and Alzheimer's-type dementia. A retrospective data analysis was carried out following a controlled study of antioxidant supplementation for dementia in DS. Observations were made at baseline and every 6 months for 2 years. Seizure history was obtained from study records. The primary outcome measures comprised the performance-based Severe Impairment Battery (SIB) and Brief Praxis Test (BPT). Secondary outcome measures comprised the informant-based Dementia Questionnaire for Mentally Retarded Persons and Vineland Adaptive Behavior Scales. Because a large proportion of patients with seizures had such severe cognitive decline as to become untestable on the performance measures, time to "first inability to test" was measured. Adjustments were made for the potentially confounding co-variates of age, gender, APOE44 status, baseline cognitive impairment, years since dementia onset at baseline, and treatment assignment. The estimated odds ratio for the time to "first inability to test" on the SIB comparing those with seizures to those without is 11.02 (95% CI: 1.59, 76.27), a ratio that is significantly different from 1 (p = 0.015). Similarly, we estimated an odds ratio of 9.02 (95% CI: 1.90, 42.85) on the BPT, a ratio also significantly different than 1 (p = 0.006). Results from a secondary analysis of the informant measures showed significant decline related to seizures. We conclude that there is a strong association of seizures with cognitive decline in demented individuals with DS. Prospective studies exploring this relationship in DS are indicated.

Lott, I.T., & Lai, F.
Dementia in Down's syndrome: observations from a neurology clinic.
Applied Research in Mental Retardation, 1982, 3, 233–239. doi: 10.1016/0270-3092(82)90017-0.
Abstract: Clinical manifestations of dementia were reviewed in 15 Down's syndrome (DS) patients referred to a neurological clinic over a 24-month period for mental deterioration. The ages ranged from 32-64 years. One hundred percent showed personality changes and loss of independent daily living skills, the presenting symptoms in two-thirds of the cases. Other manifestations included seizures (53%), gait deterioration (73%), sphincteric incontinence (40%), and pathological release reflexes (67%). All 7 patients with CT-scans showed moderate or severe central and peripheral cortical atrophy. Detailed clinical information is presented for two patients, one of whom showed a temporary remission with imipramine. A characteristic dementia syndrome appears to be present in a subpopulation of aging DA patients with radiographic findings of Alzheimer's disease.

Lynggard, H., & Alexander, N.
'Why are my friends changing?' Explaining dementia to people with learning

disabilities
British Journal of Learning Disabilities, 2004, 32(1), 30-34.
Abstract: Many publications seek to explain the causes and effects of dementia to the general population and there is evidence of the benefit of supporting carers and of establishing support groups. However, there is much less published material aimed at people with intellectual disabilities, and little focus on the specific needs of people who share their homes and lives with other people with learning disabilities who develop dementia. This article, based on group work, describes residents who had expressed bewilderment at the gradual changes they were witnessing in two of their housemates with dementia with whom they had shared a home and friendships over many years. Employing a wide range of visual aids, equipment, role plays and exercises, we sought to make the explanation of dementia as accessible and concrete as possible. The group also provided a forum for the residents to talk about the effects of living with others who develop dementia. Evaluation showed how a relatively short intervention can result in positive changes for both the people with learning disabilities who develop dementia and their peers.

Lyons, V., Oliver, E., Knifton, & Molesworth, S.
Role of Admiral Nurses in supporting people with learning disabilities and dementia
Learning Disability Practice. 2022, 25(5), ??-?? doi: 10.7748/ldp.2022.e2180
Abstract: The average age of people with learning disabilities is increasing, meaning that the number of people with learning disabilities and dementia is also rising. The care trajectory for people with learning disabilities and dementia is complex, starting with challenges in obtaining an appropriate diagnosis through to receiving appropriate and high-quality end of life care. The charity Dementia UK recognises the issues that families experience when someone in their family has a learning disability and dementia, and has developed a model of care in which Admiral Nurses, who are specialist dementia nurses, work in learning disability services. This article explores the role of the Admiral Nurse in learning disability services and examines the areas in which these specialist nurses provide tailored support. The article also outlines the expected outcomes of the service provided by these nurses.

MacDonald, S., & Summers, S.J.
Psychosocial interventions for people with intellectual disabilities and dementia: A systematic review
Journal of Applied Research in Intellectual Disabilities, 27 February 2020 https://doi.org/10.1111/jar.12722
Abstract: People with intellectual disability experience a higher prevalence of dementia, at an earlier age, than the general population. The aim of this review was to establish the psychological interventions and outcomes for individuals with intellectual disability and dementia. A search of eight electronic databases and reference lists of all included articles was conducted using PRISMA guidelines. Data were synthesized using an integrative method. Initial searching produced 2,331 papers. Twenty-one studies met the inclusion criteria. Interventions were deductively categorized into behavioural, systemic and therapeutic. All studies reported positive findings for individuals and for the systems which support them, but limited by methodological issues and neglect of the direct experience and impact on individuals themselves. The lack of a synthesis of psychosocial interventions within clinical practice, and the associated evidence, has invariably led to a lack of knowledge in practice. This is clearly evidenced by the omission of psychosocial interventions within established intellectual disability and dementia care pathways. The findings are discussed in relation to the wider literature and evidence base. Future research should aim to adopt methodologically robust designs that are inclusive of the individual experience of people with intellectual disability. The authors also posit that this review will provide essential knowledge for enacting policies that all individuals diagnosed with dementia and their carers have access to meaningful post-diagnostic care, including social and psychological care and support.

Manji, S.W.L.U.
Aging with dementia and an intellectual disability: A case study of supported empowerment in a community living home.
Dissertation Abstracts International Section A: Humanities and Social Sciences, 2009, 70(1-A), 352.
Abstract: Case study explored the qualitative experience of 4 adults with intellectual disability (ID) and dementia residing in a specializing dementia support group home. Participant observation, daily living log notes, and interviews with family/friend carers, direct-care staff, and administrators were used to obtain data. The three study questions were: (I) how the onset of dementia in people with ID changes their needs, what adjustments have to be made in the support practices, and what service barriers and successes are experienced; (ii) how adults with ID and dementia experience living in a home specializing in dementia support and how stakeholders perceive this model of support; and (iii) what are the ways policymakers can better respond to the changing needs of people with ID and dementia. Two social processes were identified: 'marginalization' and 'supported empowerment'. Marginalization depicted how dementia affected adults with ID as they incurred multiple losses in ability, home, and community. Despite losses, the adults maintained their 'selfhood' with good health support, decision-making, self-agency, and autonomy as the home provided an individualized transition process, consistent and person-centered support, and elevated empathy to facilitate freedom of choice. Supported empowerment was found as an empowering social model with micro-practices that harnessed elements of empowerment necessary to support people with dual disabilities. Seven policy considerations that prevent premature placement in nursing homes, enable aging in place, and maintain a participatory life in community were recommended.

Mansell, J., Beadle-Brown, J., Whelton, B., Beckett, C., & Hutchinson, A.
Effect of service structure and organization on staff care practices in small community homes for people with intellectual disabilities
Journal of Applied Research in Intellectual Disabilities, 2008, Sep, 21(5), 398-413. https://doi.org/10.1111/j.1468-3148.2007.00410.x
Abstract: An important question in community living is what factors influence the extent to which staff provide 'active support'. Engagement, care practices and a range of staff and organizational characteristics were studied in 72 residential homes serving 359 adults with intellectual disabilities. Managers in 36 settings were trained in person-centred active support (PCAS). A group comparison design and multivariate analysis was used to investigate the relationship between variables. The PCAS group showed more active support, assistance, other contact from staff and engagement in meaningful activity but no difference in choice-making or assessment of participation in activities of daily living. The PCAS group had more staff with a professional qualification, were more likely to think that challenging behaviour was caused by lack of stimulation, had attitudes more in line with a policy of community care, rated most care tasks as less difficult, and were more organized to deliver active support. The comparison group were more likely to think that challenging behaviour was learned negative behaviour, showed more teamwork and were more satisfied. Multivariate analysis identified a range of staff and organizational variables associated with engagement and active support. The results suggest that some variables which have not hitherto been studied in relation with active support are associated with it. Professional qualification, knowledge and experience appear to be important as do some staff attitudes, clear management guidance, more frequent supervision and team meetings, training and support for staff to help residents engage in meaningful activity.

Margallo-Lana M.L., Moore, P.B., Kay, D.W., Perry, R.H., Reid, B.E., Berney, T.P., Tyrer, S.P.
Fifteen-year follow-up of 92 hospitalized adults with Down's syndrome: incidence of cognitive decline, its relationship to age and neuropathology
Journal of Intellectual Disability Research, 2007, 51(6), 463-477. doi: 10.1111/j.1365-2788.2006.00902.x.
Abstract: The clinical and neuropathological features associated with dementia in Down's syndrome (DS) are not well established. To examine clinico-pathological correlations and the incidence of cognitive decline in a cohort of adults with DS. A total of 92 hospitalized persons with DS were followed up from 1985 to December 2000. At outset, 87 participants were dementia-free, with a median age of 38 years. Assessments included the Prudhoe Cognitive Function Test (PCFT) and the Adaptive Behavior Scale (ABS), to measure cognitive and behavioral deterioration. Dementia was diagnosed from case records and caregivers' reports. Eighteen (21%) patients developed dementia during follow-up, with a median age of onset 55.5 years (range 45-74). The PCFT demonstrated cognitive decline among those with a less severe intellectual disability (mild and moderate) but not among the profoundly disabled people (severe and profound). Clinical dementia was associated with neuropathological features of Alzheimer's disease, and correlated with neocortical neurofibrillary tangle densities. At the age of 60 years and above, a little more than 50% of patients still alive had clinical evidence of dementia. Authors concluded that clinical dementia associated with measurable cognitive and functional decline is frequent in people with DS after middle age, and can be readily diagnosed among less severely intellectually disabled persons using measures of cognitive function such as the PCFT and behavioral scales

such as the ABS. In the more profoundly disabled people, the diagnosis of dementia is facilitated by the use of behavioral and neurological criteria. In this study, the largest prospective DS series including neuropathology on deceased patients, the density of neurofibrillary tangles related more closely to the dementia of DS than senile plaques. In people with DS surviving to middle and old age, the development of dementia of Alzheimer type is frequent but not inevitable, and some people with DS reach old age without clinical features of dementia.

Markar, T.N., Cruz, R., Yeoh, H., & Elliott, M.
A pilot project on a specialist memory clinic for people with learning disabilities
British Journal of Developmental Disabilities, 2006, 52(102), 37-46.
https://doi.org/10.1179/096979506799103640
Abstract: The aim of this pilot project was to evaluate the usefulness of establishing aspecially designed " Memory Clinic" for the assessment and diagnosis of dementia in people with learning disabilities. This pilot memory clinic was set up by re-organization of existing resources, especially in terms of professional time. The core team members were a psychiatrist, psychologist and a community nurse from the specialist learning disability team with a special interest in dementia and its treatment. Over a period of 8 months a total of 12 assessments were carried out. Seven females and 4 males were assessed, the age range being 41 years – 83 years; one being a repeat assessment. In 3 individuals a definitive diagnosis of dementia was made. Six of these service users alsohad associated Down's syndrome. In the Downs syndrome group 2 had a definitive diagnosis of dementia and for1 the diagnosis was inconclusive at the initial assessment. Authors note that the professionals involved in the assessment process need to be specially trained in dealing with the issues related to identifying and interpreting the significance of changes in function, behavior, and personality in adults with learning disabilities

Marler, R., & Cunningham, C.
Down's Syndrome and Alzheimer's Disease: A Guide for Carers.
39 pp.
London: Down's Syndrome Association [155 Mitcham Road, London, UK SW17 9PG] (1994).
Abstract: This booklet for community carers and agency staff covers some of the fundamentals concerning adults with Down syndrome and Alzheimer's disease, including information on obtaining diagnoses, approaches to care management, and securing services in the UK. Contains some vignettes and a small glossary and references.

Marsack-Topolewski, C.N. & Sameul, P.S.
Caregivers' perceptions of family quality of life of individuals with developmental disabilities comorbid with dementia: A pilot study.
Journal for ReAttach Therapy and Developmental Diversities, 2020 Dec 25; 3(2), 56-70. https://doi.org/10.26407/2020jrtdd.1.38
Abstract: Although individuals with intellectual/ developmental disabilities (I/DD) are living longer than in the past, they also are exposed to age-related changes in health and well-being. They are prone to acquire dementia that often manifests earlier and more frequently than in the general population. However, there is sparse knowledge on the daily challenges that affect the quality of life of the individuals with I/DD and comorbid dementia and their family caregivers. This pilot study examined strengths and challenges of individuals with dual diagnoses of I/DD and dementia using the family quality of life (FQOL) framework. Cross-sectional data was gathered from a convenience sample of family caregivers using a web-based electronic survey. This study aims to identify the common and differential elements of the DLD (SLI) and LD through a quantitative and qualitative analysis. The variables of interest in this study were the levels of importance and satisfaction attributed to the nine FQOL domains, and overall FQOL. The mean level of importance was higher than the associated ratings of satisfaction in eight of the nine domains, with an overall importance mean of 4.15 and satisfaction mean of 3.28. Analysis of the open-ended comments indicated that the negative impact of social isolation, compound caregiving, and dynamically changing caregiving needs on overall FQOL was balanced by participants' values and beliefs. A statistical analysis (Student's t test) was conducted in order to compare the children in LD and DLD groups. The data obtained from this analysis along with LSA indicate that the language skills differ between the two groups in the following aspects: lexical, pragmatic, semantic, syntactic, morphological and phonological. Significant differences ($p < .05$) occur for the start of speech therapy age, phonological disorder, passive vocabulary and language psychological age. Results for active vocabulary did not indicate a statistical difference between LD and DLD children. The discrepancies in the FQOL domains pertaining to formal and informal services and social supports elucidate a need to empower families with high caregiving needs through research, practice and policy. Providers should be cognizant of the needs of individuals with I/DD and dementia comorbid, as well as the needs of their family caregivers.

Mascarenhas Fonseca, L., Prado Mattar, G., Guerra Haddad, G., Burduli, E., McPherson, S.M., Maria de Figueiredo Ferreira Guilhoto, L., Busatto Filho, G., Sanches Yassuda, M., Bottino, C.M.C., Queiroz Hoexter, M. & Chaytor, N.S.
Prevalence of neuropsychiatric symptoms related to dementia in individuals with Down syndrome
AAIC2020, Poster presentation, July 29, 2020. *Alzheimer's & Dementia,* 16(S6), First published: 07 December 2020. https://doi.org/10.1002/alz.047603
Abstract: Neuropsychiatric symptoms (NPS) are significant manifestations of dementia, with important consequences for patients and caregivers. Despite the established genetic link between Down syndrome (DS) and Alzheimer's disease (AD), studies investigating NPS in individuals with DS and dementia are scarce. The Neuropsychiatric Inventory (NPI) was developed to identify symptoms of dementia and while it is widely used for the assessment of individuals with dementia in the general population, we have found no studies using the NPI in those with DS. The aim of this study is to characterize NPS in a sample with DS with heterogeneous cognitive profiles using the NPI Participants (N=92) with DS, =30 years of age were assessed with the Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities (CAMDEX-DS). They were classified by a psychiatrist into three categories: AD, prodromal dementia, and stable cognition. Another psychiatrist blinded to the CAMDEX-DS dementia diagnosis, evaluated the participants using the NPI. Chi-square tests were used to check for significant differences in frequency of symptoms. Thirteen participants (14.1%) had AD, 17 (18.4%) were classified as prodromal dementia and 62 (67.5%) were in the stable cognition group. Prevalence of delusion, depression, anxiety, disinhibition, irritability, appetite abnormalities and total NPI did not differ between groups. Anxiety and irritability were common across all groups (~50% of the total), while euphoria was not present in any participant. Hallucination, agitation, apathy, aberrant motor behaviour and night-time behaviour disturbance showed significant difference among the groups ($p<0.05$), with higher prevalence in the group with AD. Authors note that the results indicate that individuals with DS and AD have some of the symptoms that are characteristically present among the general population with AD. Future studies are needed to understand if AD in DS is associated with a similar pattern of NPS observed among people with AD in the general population or may follow a specific NPS pattern. The high frequency of some NPS in individuals with DS and stable cognition should be considered in the diagnostic process in order to reduce the odds of generating a false positive.

Mascarenhas Fonseca, L., Prado Mattar, G., Guerra Haddad, G., Burduli, E., McPherson, S.M., de Figueiredo Ferreira Guilhoto, L.M., Sanches Yassuda, M. Filho Busatto, G., Machado de Campos Bottino, C., Queiroz Hoexter, M., & Sage Chaytor, N.
Neuropsychiatric symptoms of Alzheimer's disease in Down Syndrome and Its impact on caregiver distress
Journal of Alzheimer's Disease, 2021, 81(1), 137-154.doi: 10.3233/JAD-201009.
Abstract: Neuropsychiatric symptoms (NPS) are non-cognitive manifestations common to dementia and other medical conditions, with important consequences for the patient, caregivers, and society. Studies investigating NPS in individuals with Down syndrome (DS) and dementia are scarce. Characterize NPS and caregiver distress among adults with DS using the Neuropsychiatric Inventory (NPI). We evaluated 92 individuals with DS (≥30 years of age), divided by clinical diagnosis: stable cognition, prodromal dementia, and AD. Diagnosis was determined by a psychiatrist using the Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities (CAMDEX-DS). NPS and caregiver distress were evaluated by an independent psychiatrist using the NPI, and participants underwent a neuropsychological assessment with Cambridge Cognitive Examination (CAMCOG-DS). Symptom severity differed between-groups for delusion, agitation, apathy, aberrant motor behavior, nighttime behavior disturbance, and total NPI scores, with NPS total score being found to be a predictor of AD in comparison to stable cognition (OR for one-point increase in the NPI = 1.342, $p = 0.012$). Agitation, apathy, nighttime behavior disturbances, and total NPI were associated with CAMCOG-DS, and 62% of caregivers of individuals with AD reported severe distress related to NPS. Caregiver distress was most impacted

by symptoms of apathy followed by nighttime behavior, appetite/eating abnormalities, anxiety, irritability, disinhibition, and depression (R2 = 0.627, F(15,76) = 8.510, p < 0.001). NPS are frequent and severe in individuals with DS and AD, contributing to caregiver distress. NPS in DS must be considered of critical relevance demanding management and treatment. Further studies are warranted to understand the biological underpinnings of such symptoms.

Mascarehas Fonseca, L., Padilla, C., Jones, E., Neale, N., Haddad, G.G., Mattar, G.P., Barros, E., Clare, I.C.H., Busatto, G.F., Bottino, C.M.C., Hoexter, M.Q., Holland, A.J., & Zaman, S.
Amnestic and non-amnestic symptoms of dementia: An international study of Alzheimer's disease in people with Down's syndrome
International Journal of Geriatric Psychiatry, 2020 Jun, 35(6), 650-661.
doi: 10.1002/gps.5283. Epub 2020 Mar 15.
Abstract: The presence of age-related neuropathology characteristic of Alzheimer's disease (AD) in people with Down syndrome (DS) is well-established. However, the early symptoms of dementia may be atypical and appear related to dysfunction of prefrontal circuitry. The authors sought to characterize the initial informant reported age-related neuropsychiatric symptoms of dementia in people with DS, and their relationship to AD and frontal lobe function. Non-amnestic informant reported symptoms (disinhibition, apathy, and executive dysfunction) and amnestic symptoms from the CAMDEX-DS informant interview were analyzed in a cross-sectional cohort of 162 participants with DS over 30 years of age, divided into three groups: stable cognition, prodromal dementia, and AD. To investigate age-related symptoms prior to evidence of prodromal dementia we stratified the stable cognition group by age. Amnestic and non-amnestic symptoms were present before evidence of informant-reported cognitive decline. In those who received the diagnosis of AD, symptoms tended to be more marked. Memory impairments were more marked in the prodromal dementia than the stable cognition group (OR = 35.07; P < .001), as was executive dysfunction (OR = 7.16; P < .001). Disinhibition was greater in the AD than in the prodromal dementia group (OR = 3.54; P = .04). Apathy was more pronounced in the AD than in the stable cognition group (OR = 34.18; P < .001). Premorbid amnestic and non-amnestic symptoms as reported by informants increase with the progression to AD. For the formal diagnosis of AD in DS this progression of symptoms needs to be taken into account. An understanding of the unique clinical presentation of DS in AD should inform treatment options.

Mascarenhas Fonseca, L., Rufino Navatta, A.C., Bottino, C.M.C., & Miotto, E.C.
Cognitive rehabilitation of dementia in adults with Down syndrome: A review of non-pharmacological interventions
Dementia and Geriatric Cognitive Disorders Extra, 2015, Sep-Dec, 5(3), 330–340.
Published online 2015 Sep 18. doi: 10.1159/000438858
Abstract: There is a close genetic relationship between Alzheimer's disease (AD) and Down syndrome (DS), AD being the most severe mental disorder affecting aging individuals with DS. The objective of the present study was to evaluate the efficacy of cognitive rehabilitation interventions in DS patients with AD by means of a critical literature review. Because AD is progressive and irreversible, treatment is aimed at delaying and reducing the cognitive and functional decline in order to preserve or improve quality of life. The effects that pharmacological treatments and cognitive interventions have on elderly individuals with AD are well documented. Recent clinical trials have investigated the use of pharmacological treatment in DS patients with AD, generating preliminary results that have been unfavorable. There is a clear lack of studies addressing the efficacy of cognitive rehabilitation interventions in DS patients with AD, and there is an urgent need for studies providing evidence to inform decisions regarding the appropriate choice of treatment strategies.

Mattheys, K., Boustead, I., Doyle, A., & Watchman, K.
'Life through a lens': understanding the impact of dementia, a participatory action project led by people with intellectual disability
Journal of Intellectual Disability Research, 2019, 63(8), 650.
Abstract: Co-researchers with an intellectual disability are part of a team looking at the effects of non-drug interventions with people who have dementia, including people with Down syndrome. Photovoice is a method of data collection and analysis combining photography with social action supporting the inclusion of people typically excluded from research. Three co-researchers with intellectual disabitly working on the 'Life through a Lens' research project attended training in photovoice methodology and use of the camera, followed by a series of practice exercises. Each engaged in participant observation to understand the impact of non-drug interventions on peers with an intellectual disabitly and dementia. Photographs were then taken that represented their feelings about the intervention followed by a group discussion with wider research team. Two of the co-researchers believed that their peers benefitted from the non-drug interventions. For example, after observing changes to the home environment of one participant, the co-researcher discussed the relaxing and calming effect this created and how it helped her to be safer at home; his photography reflected the security he observed. Such photography can help with reflecting perceptions of the effect of non–drug interventions in dementia care, offering greater authority to co-researchers with intellectual disability.

May, H.L., Fletcher, C., Alvarez, N., Zuis, J., & Cavallari, S.G.
Alzheimer's disease and Down syndrome: A manual of care
Wrentham, Mass.: Alzheimer's Committee of Wrentham Developmental Center (1996)
89 pp.
Abstract: A 9-chapter staff training manual covering the basic issues related to the occurrence of Alzheimer's disease in adults with Down syndrome. Chapters include an introduction, Alzheimer's disease and Down syndrome, assessment, family and guardian considerations, early Alzheimer's disease, mid-stage Alzheimer's disease, feeding and nutrition concerns, and understanding difficult behaviors. Appendix contains a "Level of Capacity Scale," and table outlining implications and treatment suggestions for persons with intellectual disabilities affected by dementia.

Margallo-Lana, M.L., Moore, P.B., Kay, D.W., Perry, R.H., Reid, B.E., Berney, T.P. & Tyrer, S.P.
Fifteen-year follow-up of 92 hospitalized adults with Down's syndrome: incidence of cognitive decline, its relationship to age and neuropathology
Journal of Intellectual Disability Research, 2007 Jun;51(Pt. 6):463-477.
doi: 10.1111/j.1365-2788.2006.00902.x.
Abstract: The clinical and neuropathological features associated with dementia in Down's syndrome (DS) are not well established. Aims To examine clinico-pathological correlations and the incidence of cognitive decline in a cohort of adults with DS. A total of 92 hospitalized persons with DS were followed up from 1985 to December 2000. At outset, 87 participants were dementia-free, with a median age of 38 years. Assessments included the Prudhoe Cognitive Function Test (PCFT) and the Adaptive Behavior Scale (ABS), to measure cognitive and behavioural deterioration. Dementia was diagnosed from case records and caregivers' reports. Eighteen (21%) patients developed dementia during follow-up, with a median age of onset 55.5 years (range 45-74). The PCFT demonstrated cognitive decline among those with a less severe intellectual disability (mild and moderate) but not among the profoundly disabled people (severe and profound). Clinical dementia was associated with neuropathological features of Alzheimer's disease, and correlated with neocortical neurofibrillary tangle densities. At the age of 60 years and above, a little more than 50% of patients still alive had clinical evidence of dementia. Clinical dementia associated with measurable cognitive and functional decline is frequent in people with DS after middle age, and can be readily diagnosed among less severely intellectually disabled persons using measures of cognitive function such as the PCFT and behavioural scales such as the ABS. In the more profoundly disabled people, the diagnosis of dementia is facilitated by the use of behavioural and neurological criteria. In this study, the largest prospective DS series including neuropathology on deceased patients, the density of neurofibrillary tangles related more closely to the dementia of DS than senile plaques. In people with DS surviving to middle and old age, the development of dementia of Alzheimer type is frequent but not inevitable, and some people with DS reach old age without clinical features of dementia.

McBrien, J., Whitwham, S., Olverman, K., & Masters, S.
Screening adults with Down's syndrome for early signs of Alzheimer's disease.
Tizard Learning Disability Review, 2005, 10(4), 23-32.
Abstract: Given the now well-recognized risk of Alzheimer's Disease (AD) for adults with Down's Syndrome (DS) as they reach middle age, services for people with learning disability (LD) need to meet this new challenge. Good practice guidance from the Foundation for People with Learning Disabilities recommended that every service for people with learning disability should set up a register of adults with DS, conduct a baseline assessment of cognitive and adaptive functioning before the age of 30 years, develop specialist skills in this area, offer training to other professionals, front-line staff and carers, and seek high-quality

co-ordination between agencies. This article reports the progress of one LD service in meeting these challenges, highlighting the successes and difficulties that may guide other teams considering such a development.

McCallion, P.
Maintaining communication
In M.P. Janicki & A.J. Dalton (Eds.), Dementia, Aging, and Intellectual Disabilities
pp. 261-277
Philadelphia: Brunner-Mazel (1999)
Abstract: This book chapter is based on the premise that progression of dementia among persons with intellectual disabilities appears to be similar to that in the general population. Therefore, it explores how existing service models and programs may be adapted for the population with intellectual disabilities. A five part program, Maintaining Communication and Independence (MCI), is proposed which adapts an existing program for persons with dementia to better meet the needs of persons with intellectual disabilities. The five parts to MCI are: (1) strengths identification and deficit assessment, (2) environ-mental modification, (3) good communication, (4) memory aids, and (5) taking care of the carer.

McCallion, P., & Janicki, M.P.
Intellectual disabilities and dementia (Computer-based Course)
2 CD-Rom set
Center for Excellence in Aging Services, School of Social Welfare, Richardson 208, University at Albany, Albany, New York 12222 (2002)
Abstract: 2 disk set - usable on Windows 9.X/2000 on 233 MHZ Pentium or faster with audio/video playback. Instructional course on aging, intellectual disabilities and dementia. Contains digital video version of "Dementia and People with Intellectual Disabilities– What Can We Do?"

McCallion, P., McCarron, M., & Force, L.T
A measure of subjective burden for dementia care: the Caregiving Difficulty Scale – Intellectual Disability
Journal of Intellectual Disability Research, 2005, 49(5), 365-371.https://doi.org/10.1111/j.1365-2788.2005.00670.x
Abstract: It has been suggested in the literature on family caregiving for persons with Alzheimer's dementia (AD) that levels of objective and subjective burden among carers often predict institutionalization of the persons with AD. There is a paucity of measures to assess whether perceived burden among formal caregivers may also predict movement to more restrictive settings for persons with intellectual disabilities (ID) and AD. This study focused upon the development of a measure of subjective burden, The Caregiving Difficulty Scale – Intellectual Disability (CDS-ID) as a first step in addressing this measurement deficit. An existing caregiver subjective burden scale, the Caregiving Hassles Scale (CHS) was adapted for use with 203 staff caregivers of persons with ID and AD. Preliminary testing of existing CHS items and proposed new items was carried out in two countries, Ireland and the USA. Confirmatory factor analysis with the existing items and exploratory factor analysis with existing and proposed new items for the scale was used to establish the content and test the psychometric properties of a revised scale, the CDS-ID. On the existing CHS items, staff carers appeared to experience greater subjective burden than has been reported for family caregivers. However, the psychometric properties of the CHS found with this population were poor. Factor analysis produced a revised scale, the CDS-ID with three subscales with Cronbach alphas ranging from 0.75 to 0.93 and 38 items overall. This new scale when used with objective burden and other scales offers an opportunity to more systematically measure the difficulties staff experience when caring for persons with ID who present with symptoms of AD.

McCallion, P., Nickle, T., & McCarron, M.
A comparison of reports of caregiver burden between foster family care providers and staff caregivers in other settings: A pilot study
Dementia (London), 2005, 4(3), 401-412
https://doi.org/10.1177/1471301205055034
Abstract: There has been increasing concern about the impact of dementia symptoms on the lives and on the care being provided for persons with intellectual disability (ID) in out-of-home settings. One such setting that has received little attention is foster family care homes. These settings in the USA replicate family living and while some supports and resources are provided, they are not designed to meet intensive care needs. As a preliminary step in understanding family experiences and to expand the range of interest in Alzheimer's disease (AD) in persons with ID beyond traditional out-of-home settings, a pilot study was initiated that included aging persons with ID and symptoms of AD who were living in foster family care settings in two regions of New York State as well as more traditional out-of-home care subjects. Comparisons of matched samples on subjective and objective burden measures suggest that there are few differences in experiences. The limitations of these findings are considered and recommendations made for future, related research.

McCarron, M.
Some issues in caring for people with the dual disability of Down's syndrome and Alzheimer's dementia
Journal of Learning Disabilities for Nursing, Health and Social Care, 1999, 3(3), 123-129. https://doi.org/10.1177/174462959900300302
Abstract: Virtually all individuals with Down's syndrome over the age of 35 years have neurological changes characteristic of Alzheimer's disease. It has become increasingly recognized that people with Down's syndrome and dementia have very special needs, and those who care for them require specialist knowledge and skills. This paper aims to explore some important issues in caring for persons with this dual disability. It commences with a brief outline on the prevalence of dementia in this population. Diagnostic issues and the clinical presentation of dementia in persons with Down's syndrome are reviewed. In an attempt to help staff respond to the opportunities and challenges they encounter, issues discussed, include: promoting well-being, developing a shared vision on which to build practice, mealtimes -- a therapeutic event, reality orientation and validation therapy, communication, activity and entertainment.

McCarron, M., Gill, M., Lawlor, B., & Begley, C.
Time spent caregiving for persons with the dual disability of Down's syndrome and Alzheimer's dementia: Preliminary findings
Journal of Learning Disabilities, 2002, 6(3), 263-279.
https://doi.org/10.1177/1469004702006003036
Abstract: Persons with Down's syndrome (DS) are at increased risk of Alzheimer's type dementia (AD) compared with the general population. Little attention has been paid to the current and future impact of AD on caregivers and clients in residential and community settings. This study sought to test if the Caregiver Activity Survey-Intellectual Disability (CAS-ID) would be useful in measuring time spent by professional caregivers aiding persons with DS and AD. Preliminary findings suggest that staff caregiving time increases significantly when a person with DS experiences symptoms of dementia. No significant differences were reported in time spent caregiving for subjects at mid-stage versus end-stage dementia; however, the nature and tasks of caregiving change as dementia progresses. This study supports the utility of the CAS-ID in measuring time spent caregiving for persons with AD and DS. Care providers must plan appropriate models of health and social care to effectively address these needs.

McCarron, M., Gill, M., Lawlor, B., & Beagly, C.
A pilot study of the reliability and validity of the Caregiver Activity Survey – Intellectual Disability (CAS-ID)
Journal of Intellectual Disability Research, 2002, 46, 605-612. doi: 10.1046/j.1365-2788.2002.00437.x.
Abstract: Authors undertook to amend the Caregiver Activity Survey (Davis et al., 1997) and apply it for use with caregivers of persons with intellectual disabilities. Under this study, the CAS-ID was tested with 30 adults and convergent validity was assessed by comparing the CAS-ID with other measures of cognitive and functional impairment of adults with intellectual disabilities. Final version of the CAS-ID contains 8 items: dressing, bathing/showering, grooming, toileting, eating and drinking, housekeeping, nursing care-related activities, and supervision/behavior management. Authors content that the CAS-ID has the potential for identifying and measuring care and resource requirements for people experiencing decline associated with dementia.

McCarron, M., Gill, M., McCallion, P., & Begley, C.
Health co-morbidities in ageing persons with Down syndrome and Alzheimer's dementia.
Journal of Intellectual Disability Research, 2005, 49(7), 560-566. doi:10.1111/j.1365-2788.2005.00704.x.
Abstract: Consideration of the relationship between physical and mental health co-morbidities in ageing persons with Down syndrome (DS) and Alzheimer's dementia (AD) is of clinical importance both from a care and resource perspective. To investigate and measure health co-morbidities in ageing persons

with Down syndrome with and without AD. Recorded physical and mental health needs were ascertained for 124 persons with DS >35 years through a systematic and detailed search of individual medical and nursing case records. Differences in persons with and without AD were investigated, by stage of dementia and by level of intellectual disability (ID). A summed score for health co-morbidities was created and compared using r-tests. Persons with AD had significantly higher co-morbidity scores than persons without AD ® = -8.992, d.f. = 121, P<0.0001). There was also a significant difference in summed co-morbidity scores for persons at end-stage vs. persons at midstage AD (t = -6.429, d.f. = 56, P < 0.0001). No differences were found by level of ID. Increasing health co-morbidities in persons with DS and AD have important implications for care and resources. Appropriate environmental supports combined with competent skilled staff are crucial and will have an important impact on the quality of life for this increasingly at risk population.

McCarron, M., & Lawlor, B.A.
Responding to the challenge of ageing and dementia in intellectual disability in Ireland
Aging and Mental Health, 2003, 7(6), 413-417.
doi:10.1080/1360786031000159465 5.
Abstract: The intellectual disability (ID) population in Ireland is ageing and the number of older persons with the dual disability of ID and dementia is increasing. In spite of these demographic trends, as in other countries adequate policy and service provision for this population are lacking. This paper draws upon data available on the population with ID and dementia, reviews both generic and ID specific literature, considers the policy context and argues for a specific model of service provision. A service model is proposed for the development of multidisciplinary specialist teams within ID, delivered through mobile regional ID dementia clinics.

McCarron, M., Gill, M., Mccallion, P., Begley, C.
Alzheimer's dementia in persons with Down's syndrome: predicting time spent on day-to-day caregiving.
Dementia, 2005, 4(4), 521-538. https://doi.org/10.1177/1471301205058305
Abstract: The aim of this study was to investigate the amount of time formal caregivers spend addressing activities of day-to-day care activities for persons with Down's syndrome (DS) with and without Alzheimer's dementia (AD). Caregivers completed for 63 persons with DS and AD, and 61 persons with DS without AD, the Caregiving Activity Survey-Intellectual Disability (CAS-ID). Data was also gathered on co-morbid conditions. Regression analysis was used to understand predictors of increased time spent on day-to-day caregiving. Significant differences were found in average time spent in day-to-day caregiving for persons with and without AD. Mid-stage and end-stage AD, and co-morbid conditions were all found to predict increased time spent caregiving. Nature and tasks of day-to-day caregiving appeared to change as AD progressed. The study concluded that staff time to address day-to-day caregiving needs appeared to increase with onset of AD and did so most dramatically for persons with moderate intellectual disability. Equally, while the tasks for staff were different, time demands in caring for persons at both mid-and end-stage AD appeared similar.

McCarron, M., McCallion, P., Fahey-McCarthy, E., Connaire, K., & Dunn-Lane, J.
Supporting persons with Down syndrome and advanced dementia: Challenges and care concerns
Dementia, 2010, 9, 285-298. https://doi.org/10.1177/1471301209354025
Abstract: To understand staff perceptions of critical issues in caring for persons with intellectual disability (ID) and advanced dementia. There has been growing interest in addressing resource, training, and service redesign issues including an increase in collaborative practices in response to the growing incidence of dementia among persons with ID. Most recently this has included consideration of the specific issues in advanced dementia. Thirteen focus group interviews were held involving staff in six ID services and one specialist palliative care provider in Ireland. A qualitative descriptive approach was taken to analysis. Staff identified three key themes: (1) readiness to respond to end of life needs, (2) the fear of swallowing difficulties, and (3) environmental concerns and ageing in place. Four underlying issues that emerged in this study offer clues to solutions: (a) differences in staff preparation associated with settings, (b) lack of understanding and lack of collaboration with palliative care services, © uncertainties about the ability to transfer existing palliative care models to persons with ID and dementia and (d) the need to develop training on end stage dementia and related care approaches

McCarron, M. McCallion, P., Fahey-McCarthy, E., & Connaire, K.
Staff perceptions of essential prerequisites underpinning end-of-life care for persons with intellectual disability and advanced dementia.
Journal of Policy and Practice in Intellectual Disabilities, 2010, 7(2), 143–152. https://doi.org/10.1111/j.1741-1130.2010.00257.x
Abstract To better address palliative care and end-of-life issues for persons with intellectual disability (ID) and dementia, work was undertaken to understand the perspectives of agency staff in both the ID services and specialist palliative care fields. A qualitative descriptive design composed of 13 focus group interviews involved 50 participants drawn from six ID service providers and seven participants from one specialist palliative care service. Analysis was an iterative process; codes were identified and through thematic analysis, collapsed into two core themes: building upon services' history and personal caring—offering quality and sensitive care, and supporting comfort and optimal death in persons with ID and advanced dementia. Challenges were raised for service systems in the areas of aging in place, person-centered care, and interservice collaboration. Authors recommend both more practice relationship based and collaborative approaches to care and a stronger evidence-based research program on the timing and the efficacy of palliative care for persons with ID and dementia.

McCarron, M., McCallion, P., Fahey-McCarthy, E., & Connaire, K.
The role and timing of palliative care in supporting persons with intellectual disability and advanced dementia.
Journal of Applied Research in Intellectual Disabilities, 2011, 24, 189–198. https://doi.org/10.1111/j.1468-3148.2010.00592.x
Abstract: To better describe the role and timing of palliative care in supporting persons with intellectual disabilities and advanced dementia (AD). Specialist palliative care providers have focused mostly on people with cancers. Working with persons with intellectual disabilities and AD offers opportunities to expand such palliative care to other populations and disease conditions and to better understand the timing and role of palliative care delivery. Thirteen focus group interviews were held involving staff in six intellectual disability services and one specialist palliative care provider in Ireland. A qualitative descriptive approach was taken to analysis. Specialist palliative care staff recognized that person-centered care delivered in intellectual disability services was consistent with palliative approaches, but staff in intellectual disability services did not consider advanced dementia care as 'palliative care'. Both groups were unsure about the role of palliative care at early stage of dementia but appreciated specialist palliative care contributions in addressing pain and symptom management challenges. Successful extension of palliative care principles, philosophy and services to persons with intellectual disabilities and AD will require in-depth understanding of prevailing care philosophies and agreement regarding timing and the unique contributions of specialist palliative care services.

McCarron, M., McCallion, P., Reilly, E., & Mulryan, N.
Responding to the challenges of service developmental to address dementia needs for people with an intellectual disability and caregivers
Watchman, K. (ed.), 2014, *Intellectual Disability and Dementia: Research into Practice*, Jessica Kingsley Publishers, London, pp. 241-70.
Abstract: Book chapter addressing the association between intellectual disability and dementia; the experiences of dementia in people with intellectual disabilities; and service planning. This book chapter pertains to the latter. It acknowledges the challenges faced by traditional intellectual disability services and explains the approach adopted by one such service in ROI to supporting and accommodating people with an intellectual disability who develop dementia. It exemplifies the move to an in place progression model in traditional intellectual disability services, whereby services are adapted so that people with an intellectual disability can be supported and accommodated as they progress through dementia.

McCarron, M., & Riley, E.
Supporting persons with intellectual disability and dementia: Quality dementia care standards - A guide to practice
39 pp.
Dublin, Ireland: Trinity College Dublin (2010)
Source: http://www.docservice.ie/includes/documents/Dementia%20Publication%202011.pdf
Abstract: Document contains a series of six standards covering a range of areas concerned with care affecting adults with intellectual disabilities affected by dementia. Drawn from standards affecting the general population, this document

groups together focal areas under six main categories reflecting person-centered dementia care. The standards consist of statements, indicators, and criteria for assessing evidence. The standards cover (1) appropriately trained staff and service development, (2) memory assessment services, (3) health and personal care, (4) communication and behavior, (5) promoting well-being and social connectedness, and (6) supporting persons with advanced dementia.

McCarron, M., Reilly, E., & Dunne, P.
Achieving quality environments for person centred dementia care
45 pp.
Dublin, Ireland: Daughters of Charity Service
Abstract: Provides an overview of principles and practices designed to enable the operation of small group homes, including covering the planning process, design of private and public spaces, as well as therapeutic uses. Illustrated by two Daughters of Charity homes established for dementia specific care for people with ID. One home offers care for people with moderate dementia and includes 4 permanent beds and 2 respite beds for people both living with their families in the community and community group homes. Home also has a 6 bed step-down palliative care unit for people with ID in the later stages of dementia. These purpose built facilities were designed to be responsive to the changing needs of persons across the continuum of dementia. The home-like environments support people with dementia and staff to participate and complete tasks together, as well as informal impromptu unplanned activities. The homes are designed so that each resident has his or her own bedroom, with numerous communal areas including sitting rooms and garden areas.

McGuire, B. E.; Whyte, N. , & Hardardottir, D.
Alzheimer's disease in Down Syndrome and intellectual disability: A review.
The Irish Journal of Psychology, 2006, 27(3-4), 114-129.
https://doi.org/10.1080/03033910.2006.10446235
The authors review the literature on Alzheimer's disease (AD) in persons with general intellectual disabilities and those with Down syndrome. It focuses on the prevalence, clinical manifestations, diagnosis and management of AD in these populations. The literature indicates that people with Down syndrome have a greatly increased risk of dementia from their early 40s, while people with general intellectual disabilities have similar rates of AD to the general population. Taking into account the life expectancy of people with intellectual disabilities and those with Down syndrome, guidelines are provided for estimating the proportion of service users in a population that are at risk of developing dementia. The difficulties around diagnosis are reviewed and a particular emphasis is placed on the range of psychometric measures that may contribute to assessment and diagnosis. The management of service users who develop dementia is also reviewed and the implications for service providers are highlighted.

McKenzie, K., Harte, C., Patrick, S., Matheson, E., & Murray, G.C.
The assessment of behavioural decline in adults with Down's syndrome
Journal of Learning Disabilities, 2002, 6, 175-184.
https://doi.org/10.1177/146900470200600206
Abstract: Article reports study the examined two methods of using the Vineland Adaptive Behavioral Scales (VABS) to measure behavioral change in adults with Down syndrome who were surmised to be at-risk of Alzheimer's disease. The first approach used the VABS within a semi-structured interview and all areas of behavioral change identified by staff were noted. The second approach used the basal rule of the VABS as indicated in the Scales' manual. Comparison of the two approaches indicated that using the second approach highlighted significant decline in scores (for adults meeting the criteria for "probable Alzheimer's disease) on a number of domains between baseline and 12-24 months. One limitation of this approach that was noted was that this scoring method appeared to miss more subtle changes on behavior, which may be indicative of early Alzheimer's disease – which were picked up by the first approach. Authors recommend flexibility in using the VABS for assessment purposes and caution researchers to be explicit in reporting how the VABS was used in studies assessing dementia.

McKenzie, K., Metcalfe, D., Michie, A., & Murray, G.
Service provision in Scotland for people with an intellectual disability who have, or who are at risk of developing, dementia.
Dementia (London), 2020, 19(3), 736-749. doi: 10.1177/1471301218785795.
Abstract: This research aimed to identify current national provision by health services in Scotland in relation to proactive screening and reactive assessment for people with an intellectual disability in Scotland who have, or are at risk of developing, dementia. Staff from 12 intellectual disability services, representing the 11 health board areas in Scotland, completed an online questionnaire which asked about proactive screening and reactive assessment for people with intellectual disability who had, or were at risk of developing, dementia as well as suggested areas for improvement. All of the areas provided services for people with intellectual disability who have, or are at risk of developing, dementia, but differed as to whether this was reactive, proactive or both. Nine services offered intervention following diagnosis. The most common elements used across both proactive screening and reactive assessment were conducting a health check, using a general dementia questionnaire designed for people with an intellectual disability and direct assessment with the person. Clinical psychology and community learning disability nurses were the professions most likely to be involved routinely in both proactive screening and reactive assessments. The psychometric properties of the most commonly used assessments of cognitive and behavioral functioning were mixed. The areas of improvement suggested by practitioners mainly related to ways of improving existing pathways. This research represents the first step in providing an overview of service provision in Scotland. There was some inconsistency in relation to the general and specific components which were involved in proactive screening and reactive assessment. Implications for service provision are discussed.

McKenzie, K., Metcalfe, D., & Murray, G.
A review of measures used in the screening, assessment and diagnosis of dementia in people with an intellectual disability.
Journal of Applied Research in Intellectual Disabilities, 2018, 31(5), 725-742. doi: 10.1111/jar.12441.
Abstract: The increasing number of individuals with an intellectual disability who are at risk of developing dementia highlights the need to use measures with strong psychometric properties as part of the screening, assessment and diagnostic process. Searches were made of clinical and good practice guidelines and English language journal articles sourced from Proquest, Web of Science and Scopus databases (up to July 2017) for tools which were designed or adapted for the purpose of helping to diagnose dementia in people with intellectual disability. Based on a detailed review of 81 articles and guidelines, the present authors identified 22 relevant tools (12 cognitive, 10 behaviour). These were reviewed in terms of their psychometric properties. A number of tools were found to be available for use with people with intellectual disability; however, few were specifically standardized for this purpose which also had comprehensive information about reliability and validity.

McLaughlin, K., & Jones, D.
'It's all changed: 'carers' experiences of caring for adults who have Down's syndrome and dementia.
British Journal of Learning Disabilities, 2011, 39(1), 57-63.
https://doi.org/10.1111/j.1468-3156.2010.00618.x
Abstract: A qualitative interview study was undertaken to determine the information and support needs of carers of adults who have Down's syndrome and dementia. The data were analysed thematically. People who care for someone with Down's syndrome and dementia were asked about what it was like being a carer. Four of the carers were brothers or sisters of the person who had dementia, and two were paid carers not family members. All carers said that they wanted more information about some of the health and social problems that come with having dementia. Carers said that they wanted to know more information and meet other carers and other people who have dementia. Carers' information and support needs were seen to change at pre-diagnosis, diagnosis, and post-diagnosis. Helping carers to manage the changing nature of the adult with dementia is seen to be an essential part of the health professional's role.

McQuillan, S., Kalsy, S., Oyebode, J., Millichap, D., Oliver, C., & Hall, S.
Adults with Down's syndrome and Alzheimer's disease
Tizard Learning Review, 2003, 8(4), 4-13
Abstract: Adults with Down's syndrome are at risk of developing Alzheimer's disease in later life. This paper gives an overview of the current research in the area and discusses the implications it raises for individuals, carers, and service providers. Information on the link between Down's syndrome and Alzheimer's disease and prevalence rates are given. The clinical symptoms of Alzheimer' disease and a stage model documenting the progression of the disease are presented. Attention is drawn to the problems inherent in assessing and diagnosing Alzheimer's disease in a person with a pre-existing intellectual disability. Also discussed are the management of Alzheimer's disease, a focus on care management practices, and recommendations for service provision (including guidelines for supporting individuals which include maintaining skills, adapting a person-centered approach, implementing psychosocial interventions,

and multi-disciplinary care management. Recommendations for the future include increasing education and awareness, implementing screening services, improving assessment methods, and developing appropriate services.

McVicker, R.W., Shanks, O.E., & McClelland, R.J.
Prevalence and associated features of epilepsy in adults with Down's syndrome
British Journal of Psychiatry, 1994, Apr;164(4), 528-532. doi:10.1192/bjp.164.4.528.
Abstract: The aim of this study was to establish the prevalence of epilepsy in persons with Down's syndrome aged 19 years and over. A total of 191 adults with Down's syndrome were identified, giving a prevalence of 0.76/1000 (95% CI 0.75 to 0.77). Of these, 18 had epilepsy, giving a prevalence of 9.4% (95% CI 5.3% to 13.5%). The prevalence of epilepsy increased with age, reaching 46% in those over 50. The neurophysiological (EEG) findings of the epilepsy group were compared with those of a control group of Down's syndrome adults without epilepsy. Paroxysmal abnormalities consistent with a diagnosis of epilepsy were found in 80% of the epilepsy group, compared with only 13% of controls (P < 0.001). Epilepsy of late onset was associated with diffuse EEG abnormalities and clinical evidence of dementia. The age distribution and EEG findings suggest two independent processes in the causation of epilepsy: late-onset epilepsy associated with clinical evidence of dementia, and early-onset epilepsy in the absence of dementia.

Menéndez M.
Down syndrome, Alzheimer's disease and seizures.
Brain Development, 2005, 27(4), 246-252. doi:10.1016/j.braindev.2004.07.008.
Abstract: Neuropathologically, Alzheimer-type abnormalities are demonstrated in patients with Down syndrome (DS), both demented and nondemented and more than a half of patients with DS above 50 years develop Alzheimer's disease (AD). The apolipoprotein E epsilon4 allele, oestrogen deficiency, high levels of Abeta1-42 peptide, elevated expression of BACE2, and valine polymorphism of prion protein gene are associated with earlier onset of dementia in DS individuals. Advanced AD alone may be an important risk factor for new-onset seizures in older adults and age above 60 years is a recognized risk factor for poor outcome from convulsive and nonconvulsive status epilepticus. DS patients aged over 45 years are significantly more likely to develop Alzheimer's disease than those less than 45 years and up to 84% demented individuals with DS develop seizures. Late-onset epilepsy in DS is associated with AD, while early-onset epilepsy is associated with an absence of dementia. In AD patients with a younger age of dementia onset are particularly susceptible to seizures. DS adults with epilepsy score significantly higher overall on the adaptive behaviour profile. Language function declined significantly more rapidly in AD patients with seizures and there is a good correlation between the severity of EEG abnormalities and cognitive impairment whereas in DS slowing of the dominant occipital rhythm is related to AD and the frequency of the dominant occipital activity decreases at the onset of cognitive deterioration.

Millichap, D., Oliver, C., McQuillan, S., Kalsy, S., Lloyd, V., & Hall, S.
Descriptive functional analysis of behavioral excesses shown by adults with Down syndrome and dementia.
International Journal of Geriatric Psychiatry, 2003, 18, 844-854. doi: 10.1002/gps.930
Abstract: The study examined the hypothesis that a functional relationship exists between social environmental events and behavioral excesses in individuals with Down syndrome and dementia. A case-series design was employed (n = 4) using an direct observation-based descriptive functional assessment procedure. Observations were conducted in the natural environments of four participants over periods ranging from 11 to 15.4 hours. Data were collected on non-verbal and verbal behavioral excesses, appropriate engagement and verbal interaction with others. Social environmental events observed including both staff and peer behavior. Analysis of co-occurrence for behavioral excesses and social environmental events indicated significant relationships for some behaviors consistent with operant reinforcement processes. Sequential analysis showed that changes in the probability of social contact occurred in the period directly preceding and following verbal behaviors. Results support the hypothesis that, consistent with literature for older adults with dementia in the general population, some behavioral excesses were functional in nature and not randomly occurring events. No relationship was found between appropriate engagement and staff.

Mircher, C., Cieuta-Walti, C., Marey, I., Rebillat, A-S., Cretu, L., Milenko, E., Conte, M., Sturtz, F., Rethore, M-O, & Ravel, A.
Acute regression in young people with Down syndrome.
Brain Science, 2017, Jun, 7(6), 57. Published online 2017 May 27. doi: 10.3390/brainsci7060057
Abstract: Adolescents and young adults with Down syndrome (DS) can present a rapid regression with loss of independence and daily skills. Causes of regression are unknown and treatment is most of the time symptomatic. We did a retrospective cohort study of regression cases: patients were born between 1959 and 2000, and were followed from 1984 to now. We found 30 DS patients aged 11 to 30 years old with history of regression. Regression occurred regardless of the cognitive level (severe, moderate, or mild intellectual disability (ID)). Patients presented psychiatric symptoms (catatonia, depression, delusions, stereotypies, etc.), partial or total loss of independence in activities of daily living (dressing, toilet, meals, and continence), language impairment (silence, whispered voice, etc.), and loss of academic skills. All patients experienced severe emotional stress prior to regression, which may be considered the trigger. Partial or total recovery was observed for about 50% of them. In our cohort, girls were more frequently affected than boys (64%). Neurobiological hypotheses are discussed as well as preventative and therapeutic approaches.

Mohan, M., Bennet, C., & Carpenter, P.K.
Rivastigmine for dementia in people with Down syndrome
Cochrane Systemic Review - Intervention, 2009, 1.
https://doi.org//10.1002/14651858.CD007658
Abstract: Alzheimer's dementia (AD) is the most common form of dementia in people with Down Syndrome (DS). Acetylcholine is a chemical found in the brain that has an important role in memory, attention, reason and language. Rivastigmine is a “pseudo-irreversible” inhibitor of acetylcholinesterase, which is thought to maintain levels of acetylcholine. Rivastigmine can improve cognitive function and slow the decline of AD in the general population over time. It is important to note that people with DS tend to present with AD at a much younger age than the normal population as well as having subtle differences in physiology (e.g. metabolism and heart rate) and may therefore have different requirements from the general population. The authors sought to determine the effectiveness and safety of rivastigmine for people with DS who develop AD by using the following search methods, CENTRAL, MEDLINE, EMBASE, CINAHL, PsycINFO, BIOSIS, SCI, SSCI and the NRR, up to October 2008. They also contacted the manufacturers of rivastigmine as well as experts in the field, to ask about reports of unpublished or ongoing trials. Selection criteria included randomised controlled trials of participants with DS and AD in which treatment with rivastigmine was administered compared with a placebo group. Authors found that no study was identified which met inclusion criteria for this review and concluded that as there are no included trials, recommendations cannot be made about rivastigmine for AD in DS. Well-designed, adequately powered studies are required.

Molnar, F.J., Benjamin, S., Hawkins, S.A., Briscoe, M., & Ehsan, S.
One size does not fit all: Choosing practical cognitive screening tools for your practice.
JAGS (Journal of the American Geriatrics Society), 2020, 68(10), 2207-2213. DOI: 10.1111/jgs.1671
Abstract: Every year, millions of patients worldwide undergo cognitive testing. Unfortunately, new barriers to the use of free open access cognitive screening tools have arisen over time, making accessibility of tools unstable. This article is in follow-up to an editorial discussing alternative cognitive screening tools for those who cannot afford the costs of the Mini-Mental State Examination and Montreal Cognitive Assessment (see www.dementiascreen.ca). The current article outlines an emerging disruptive“free-to-fee”cycle where free open access cognitive screening tools are integrated into clinical practice and guidelines, where fees are then levied for the use of the tools, resulting in clinicians moving on to other tools. This article provides recommendations on means to break this cycle, including the development of tool kits of valid cognitive screening tools that authors have contracted not to charge for (i.e., have agreed to keep free open access).The PRACTICAL.1 Criteria (PRACTIcing Clinician Accessibility and Logistical Criteria Version 1) are introduced to help clinicians select from validated cognitive screening tools, considering barriers and facilitators, such as whether the cognitive screening tools are easy to score and free of cost. It is suggested that future systematic reviews embed the PRACTICAL.1 criteria, or refined future versions, as part of the standard of review. Methodological issues, the need for open access training to insure proper use of cognitive screening tools, and the need to anticipate growing ethnolinguistic diversity by developing tools that are less sensitive to educational, cultural, and linguistic bias are discussed in this opinion piece.

Moran, J.A., Rafii, M.S., Keller, S.M., Singh, B.K., Janicki, M.P.
The National Task Group on Intellectual Disabilities and Dementia Practices consensus recommendations for the evaluation and management of dementia in adults with intellectual disabilities.
Mayo Clinic Proceedings, 2013 Aug;88(8):831-840. doi: 10.1016/j.mayocp.2013.04.024. Epub 2013 Jul 10.
Abstract: Adults with intellectual and developmental disabilities (I/DD) are increasingly presenting to their health care professionals with concerns related to growing older. One particularly challenging clinical question is related to the evaluation of suspected cognitive decline or dementia in older adults with I/DD, a question that most physicians feel ill-prepared to answer. The National Task Group on Intellectual Disabilities and Dementia Practices was convened to help formally address this topic, which remains largely under-represented in the medical literature. The task group, comprising specialists who work extensively with adults with I/DD, has promulgated the following Consensus Recommendations for the Evaluation and Management of Dementia in Adults With Intellectual Disabilities as a framework for the practicing physician who seeks to approach this clinical question practically, thoughtfully, and comprehensively.

Moriconi, C., Schlamb, C., & Harrison, B.
Down syndrome and dementia: Guide to identification, screening, and managment.
Journal for Nurse Practitioners, 2015, 11(8), 812-818.
https://www.npjournal.org/article/S1555-4155(15)00602-9/pdf
Abstract: Down syndrome (DS) is an intellectual disability due to the genetic disorder trisomy21. Many individuals with DS are living into middle and older adulthood,experiencing chronic health problems, and are at risk for dementia. This article describes the primary care management of adults with DS, the relationship between DS and Alzheimer's dementia, and screening protocols for primary care. [Extract follows] The family of the person with DS and AD needs a team to navigate the changes that are inevitable during the course of this disease. An open discussion with both the NP and social worker will assist most families in providing the best quality of life for aging adults with DS. Maintaining social contact with family and friends may be difficult for aging DS adults, but is vital for well-being. Expectations for self-care and new learning must be readjusted and give way to an emphasis on a positive approach to the adult with DS. Families and caregivers need to proactively create a safe and calming home environment to promote quality of life.As AD progresses, assessment of nonverbal communication is essential in the care of adults with DS, but it may be challenging. Communication may be based on nonverbal cues and gestures. Additional response time is needed to allow for verbal or nonverbal communication of needs. Utilizing communication boards and pictures of common persons or objects is beneficial in this population. Safe relationships grounded in familiarity and trust are paramount in providing a secure environment for adults with DS and AD.

Moss, S., Lambe, L., & Hogg, J.
Physical and mental health
Ageing Matters - Pathways for Older People with Learning Disabilities: Manager's Reader.
pp. 41-60
Kidderminster: British Institute of Learning Disabilities [Wolverhampton Road, Kidderminster, Worcestershire DY10 3PP United Kingdom] (1998)
Abstract: This unit, one of six that is used for training staff, covers briefly some of the key issues related to physical and mental health, and touches on dementia. Although not specifically developed for care management of adults with dementia, the text, in total, can be a useful resource for staff working in care settings when one or more of the adults in the setting are affected by dementia.

Moss, S., & Patel, P.
Dementia in older people with intellectual disability: symptoms of physical and mental illness, and levels of adaptive behavior.
Journal of Intellectual Disability Research, 1997, 41(1), 60-69. doi: 10.1111/j.1365-2788.1997.tb00677.x.
Abstract: Detailed data on health and functional ability of 101 people with intellectual disability over 50 years of age are presented. Using a combination of informant interviewing, observation and measurement of cognitive change over a 3-year period, 12 of these individuals were identified as suffering from dementia. Their data are compared to those of the non-dementia sufferers. The people suffering from dementia had a greater number of chronic physical health problems and chronic disability resulting from physical health problems. Their capacity for self-directed activity was lower. The subjects had a reduced capacity to enjoy things, and were more irritable and more prone to violence. However, the outlook is somewhat different from a strategic perspective. The population of people with intellectual disability shows considerable epidemiological changes across the lifespan because of the effects of differential survival. The interaction of these factors tends to mask the impact of dementia-related skill loss in this population

Mullins, D., Daly, E., Simmons, A., Beacher, F., Foy, C.M.L., Lovestone, S.,, Hallahan, B., Murphy, K.C., & Murphy, D. G.
Dementia in Down's syndrome: an MRI comparison with Alzheimer's disease in the general population.
Journal of Neurodevelopmental Disorders 2013, 5, 19-00. doi:10.1186/1866-1955-5-19.
Abstract: Down's syndrome (DS) is the most common genetic cause of intellectual disability. People with DS are at an increased risk of Alzheimer's disease (AD) compared to the general population. Neuroimaging studies of AD have focused on medial temporal structures; however, to our knowledge, no *in vivo* case–control study exists comparing the anatomy of dementia in DS to people with AD in the general population. We therefore compared the *in vivo* brain anatomy of people with DS and dementia (DS+) to those with AD in the general population. Using MRI in 192 adults, we compared the volume of whole brain matter, lateral ventricles, temporal lobes and hippocampus in DS subjects with and without dementia (DS+, DS-), to each other and to three non-DS groups. These included one group of individuals with AD and two groups of controls (each age-matched for their respective DS and general population AD cohorts). AD and DS+ subjects showed significant reductions in the volume of the whole brain, hippocampus and temporal lobes and a significant elevation in the volume of the lateral ventricle, compared to their non-demented counterparts. People with DS+ had a smaller reduction in temporal lobe volume compared to individuals with AD. DS+ and AD subjects have a significant reduction in volume of the same brain regions. We found preliminary evidence that DS individuals may be more sensitive to tissue loss than others and have less 'cognitive reserve'

Nagdee, M.
Dementia in intellectual disability: a review of diagnostic challenges.
African Journal of Psychiatry (Johannesburg), 2011, 14, 194-199. doi: 10.4314/ajpsy.v14i3.1.
Abstract: The evaluation of dementia in individuals with intellectual disability, which will guide subsequent intervention, care and management depends on the systematic review of a number of factors: (1) the individual historical context, obtained from multiple sources, (2) evaluation of the pre-existing cognitive, behavioral, psychiatric, medical and adaptive skill profile, (3) the constellation, and pattern of evolution, of presenting signs and symptoms, (4) results of focused investigations, and (5) refinement of the differential diagnosis. In patients with ID, standard clinical methods need to be supplemented by careful, longitudinal behavioral observations, and individually tailored assessment techniques. Co-morbidity, multiple biological, psychological and socio-environmental factors, and complex interactions among events, are the reality for many ageing people with ID. Determining the various influences is often a formidable clinical task, but should be systematically carried out using medical, cognitive, behavioral, neuropsychiatric and psycho-social frameworks.

National Task Group on Intellectual Disabilities and Dementia Practices.
My thinker's not working': A national strategy for enabling adults with intellectual disabilities affected by dementia to remain in their community and receive quality supports.
42pp.
National Task Group on Intellectual Disabilities and Dementia Practices [www.aadmd.org/ntg]. (2012).
Abstract: 'My Thinker's Not Working' is the short title for the 42-page summative report issued by the National Task Group on Intellectual Disabilities and Dementia Practices, a planning and advocacy group organized to produce a national plan on dementia and intellectual disabilities. The report offers 20 recommendations for the improvement of services nationally and locally and suggests that its findings and recommendations be considered and integrated into the reports and plans being developed by the federal Advisory Council on Alzheimer's Research, Care, and Services -- under the National Alzheimer's Project Act. The document reviews the main issue facing adults with intellectual disabilities as they age when they are affected by dementia, as well as their

families and provider organizations. The document is composed of 7 sections (Charge and Purpose, The Population, Challenges Facing the Population, Community Services, Education and Training, Financing, and Possible Solutions) and the National Dementia and Intellectual Disabilities Action Plan.

NAMHI
Alzheimer's Dementia in persons with intellectual disabilities: Some common questions and concerns
NAMHI, 5 Fitzwilliam Place, Dublin 2, Ireland
Abstract: 28 page booklet with 18 sections/question areas outlining basic information about Alzheimer's disease and people with ID, diagnostic resources, and service to help cope with the course of the disease. Developed by Dr. Mary McCarron of Trinity College Dublin.

Nelson, L., Lott, I., Touchette, P., Satz, P., & D'Elia, L.D.
Detection of Alzheimer disease in individuals with Down syndrome
American Journal of Mental Retardation, 1995 May;99(6):616-622.
Abstract:A comprehensive baseline of emotional functioning was established for adults with Down syndrome. Five emotional factors were studied using groups of (a) adults with Down syndrome (n = 30), (b) clinical control subjects with dementia of the Alzheimer type (n = 18), and (c) elderly control subjects without intellectual disability (n = 25). Results of planned statistical comparisons showed indifference, pragnosia, and inappropriateness as primary emotional factors separating Down syndrome and Alzheimer disease groups from elderly control subjects without intellectual disabiity. Indifference was also shown to covary with cognitive mental state, whereby increased levels of indifference were associated with decreased levels of cognitive functioning. There is the possibility of noncognitive variables signaling dementia of the Alzheimer type in individuals with Down syndrome.

Nelson L.D., Orme, D., Osann, K., & Lott, I.T.
Neurological changes and emotional functioning in adults with Down Syndrome.
Journal of Intellectual Disability Research, 2001, 45, 450-456. doi: 10.1046/j.1365-2788.2001.00379.x.
Abstract: Study examined emotional changes in adults with Down Syndrome (DS) over time and to determine whether changes in these psychological variables were associated with brain atrophy on MRI scan and the presence of pathological reflexes on the neurological examination. Participants were 26 adults with DS and their caregivers. Caregivers completed a measure of emotional functioning about individuals with DS at two different time points (1 year apart). Levels of cognitive functioning were measured and neurological and MRI examinations were performed on all subjects at initial testing. Significant group effect separated those with and without pathological findings on MRI and neurological exam across three different scales: depression, indifference, and pragmatic language functioning. Problems of poor pragmatic language functioning appeared later in the course of suspected Alzheimer's disease (AD), as demonstrated by a significant group effect at time 2, but not at initial testing. In these subjects, the primary emotional change was a decline in social discourse (e.g. conversational style, literal understanding, verbal expression in social contexts). These emotional levels were stable over time, regardless of degree of cognitive decline. Specific emotional changes occur during the course of AD which were associated with abnormal findings from MRI and from neurological examination. These results, along with abnormalities in brain imaging and the presence of pathological reflexes, suggested that frontal lobe dysfunction is likely to be an early manifestation of Alzheimer's Disease in Down Syndrome.

New York State Developmental Disabilities Planning Council
When people with developmental disabilities age
18 minutes
New York State Developmental Disabilities Planning Council [155 Washington Avenue, Albany, New York 12222] (1992).
Abstract: A 18-minute video outlining the major physical and social change issues affecting adults with intellectual and developmental disabilities as they age, including a brief mention of Alzheimer's disease and Down syndrome. Available in VHS and CD-Rom format.

New York State Developmental Disabilities Planning Council
Dementia and people with intellectual disabilities – What can we do?
23 minutes
New York State Developmental Disabilities Planning Council [155 Washington Avenue, Albany, New York 12222] (2001).
Abstract: An instructional video which covers the basics of how dementia affects adults with intellectual disabilities, and provides information on diagnostics and suggestions on providing supports and services in community care settings. Produced by the University at Albany, this video can serve as primer on dementia and intellectual disabilities and provides information on basic design and service issues. Available in VHS and CD-Rom format.

Newroth, S., & Newroth, A.
Coping with Alzheimer disease: a growing concern.
28 pp.
Downsview: Ontario: National Institute on Mental Retardation (Kinsmen NIMR Building, York University Campus, 4700 Keele Street, Ontario, Canada, M3J IP3) (1981)
Abstract: Monograph describing one residential program's experience in caring for persons with Down syndrome who developed Alzheimer's disease; includes a chart of observations and guidelines for care. The guidelines are reproduced as an appendix in Janicki & Dalton (1999).

Ng, N., Flygare Wallen, E., & Ahlstrom, G.
Mortality patterns and risk among older men and women with intellectual disability: a Swedish national retrospective cohort study
BMC Geriatrics, 2017, 17(1), 269. doi: 10.1186/s12877-017-0665-3.
Abstract: Sweden has closed all institutions and imposed legislation to ensure service and support for individuals with intellectual disability (ID). Understanding mortality among older individuals with ID is essential to inform development of health promotion and disease control strategies. We investigated patterns and risk of mortality among older adults with ID in Sweden. This retrospective cohort study compared older adults aged 55 years and older with ID with a control population. Participants were followed during 2002-2015 or death, and censored if they moved out of Sweden. Individuals with ID were identified from two national registers: one covering all specialist health-care visits (out-patient visits and hospitalization) and the other covering people accessing social/support services. Individuals with ID (n = 15,289) were matched with a control population by sex, birth year, and year of first hospitalisation/out-patient visit/access to LSS services. Cause-of-death data were recorded using International Classification of Diseases, Tenth Revision. Cox proportional hazards regression were conducted to assess if overall and cause-specific mortality rate among individuals with ID was higher than in the Swedish population. The overall mortality rate among individuals with ID was 2483 per 100,000 people compared with 810 in the control population. Among those who died, more individuals with ID were younger than 75 years and unmarried. Leading causes of death among individuals with ID were circulatory diseases (34%), respiratory diseases (17%) and neoplasms (15%). Leading causes of death in a sub-sample with Down syndrome (DS) were respiratory diseases (37%), circulatory diseases (26%) and mental/behavioural disorders (11%). Epilepsy and pneumonitis were more common among individuals with ID than controls. **Alzheimer's disease was common in the control population and individuals with DS, but not among those with ID** when DS was excluded. Individuals with ID had a higher overall mortality risk (hazard ratio [HR] 4.1, 95% confidence interval [CI] 4.0-4.3) and respiratory disease death risk (HR 12.5, 95% CI 10.9-14.2) than controls. Older adults with ID in Sweden carry a higher mortality risk compared with the general population, mainly attributable to respiratory, nervous and circulatory diseases. Care for this group, particularly during the terminal stage of illness, needs **to be tailored based on understanding of their main health problem.**

Nieuwenhuis-Mark, R.E.
Diagnosing Alzheimer's dementia in Down syndrome: Problems and possible solutions.
Research in Developmental Disabilities, 2009, 30(5), 827-838. doi:10.1016/j.ridd.2009.01.010.
Abstract: It is widely accepted that people with Down syndrome are more likely than the general population to develop Alzheimer's dementia as they age. However, the diagnosis can be problematic in this population for a number of reasons. These include: the large intra-individual variability in cognitive functioning, the different diagnostic and methodological procedures used in the field and the difficulty in obtaining baseline levels of cognitive functioning in this population with which to assess cognitive and behavioral change. Recent researchers have begun to suggest ways around these difficulties. This review explores these recent developments and provides recommendations which may aid clinicians in their attempts to diagnose Alzheimer's dementia in the early

stages in the Down syndrome population.

Noelker, E.A. & Somple, L.C.
Adults with Down syndrome and Alzheimer's
In K.A. Roberto (Ed.), The Elderly Caregiver: Caring for Adults with Developmental Disabilities.
pp. 81-92
Newbury Park: SAGE Publications (1993)
Abstract: Book chapter providing a brief summary of significant assessment and care issues affecting adults with Down syndrome who have Alzheimer's disease. Noted are the needs for education of carers and families, as well as specialty care provision and community services.

Northway, R., Holland-Hart, D., & Jenkins, R.
Meeting the health needs of older people with intellectual disabilities: exploring the experiences of residential social care staff.
Health and Social Care in the Community, 2017 May, 25(3), 923-931. doi: 10.1111/hsc.12380. Epub 2016 Aug 31.
Abstract: Older people with intellectual disabilities often experience high levels of health needs and multiple morbidities but they may be supported by residential care staff with little or no previous experience of identifying and meeting health needs. Little is known regarding how they undertake this health-related role and this exploratory study seeks to address this gap. A purposive sample of 14 managers of supported living accommodation in Wales were interviewed in 2014 to determine their experiences of supporting tenants in relation to age-related health needs. The semi-structured interviews were transcribed and thematically analysed. Three of the emerging themes are reported in this paper: meeting health needs, the consequences of ageing and relationships. Findings indicate that residential care staff support older people with intellectual disabilities with complex and multiple health needs: they monitor health status, support access to healthcare, provide additional support arising from changing health needs and advocate for tenants in the context of healthcare. However, their role is often not understood by healthcare professionals. The importance of staff having a long-term relationship with those they support was identified as being important to identifying any health-related changes. The need to develop effective relationships with healthcare staff was also noted. It is concluded that there is a need for better understanding among health staff of the role of residential social care workers and for further research regarding health-related communication.

O'Bryant, S.E., Zhang, F., Silverman, W., Lee, J.H., Krinsky-McHale, S.J., Pang, D., Hall, J., & Schupf, N.
Proteomic profiles of incident mild cognitive impairment and Alzheimer's disease among adults with Down syndrome
Alzheimer's & Dementia: Diagnosis, Assessment & Disease Monitoring, 12, 1, https://doi.org/10.1002/dad2.12033
Abstract: We sought to determine if proteomic profiles could predict risk for incident mild cognitive impairment (MCI) and Alzheimer's disease (AD) among adults with Down syndrome (DS). In a cohort of 398 adults with DS, a total of n = 186 participants were determined to be non-demented and without MCI or AD at baseline and throughout follow-up; n = 103 had incident MCI and n = 81 had incident AD. Proteomics were conducted on banked plasma samples from a previously generated algorithm. The proteomic profile was highly accurate in predicting incident MCI (area under the curve [AUC] = 0.92) and incident AD (AUC = 0.88). For MCI risk, the support vector machine (SVM)-based high/low cut-point yielded an adjusted hazard ratio (HR) = 6.46 (P < .001). For AD risk, the SVM-based high/low cut-point score yielded an adjusted HR = 8.4 (P < .001). on
The current results provide support for our blood-based proteomic profile for predicting risk for MCI and AD among adults with DS.

O'Caoimh, R., Clune, Y., & Molloy, D.W.
Screening for Alzheimer's disease in Downs syndrome
Journal of Alzheimers Disease & Parkinsonism, 2013, S7; 001. http://dx.doi.org/10.4172/2161-0460.S7-001
Abstract: Down syndrome (DS), is associated with an increased incidence of Alzheimer's disease (AD). Although pathological changes are ubiquitous by 60 years of age, prevalence rates are lower. The diagnosis of AD in persons with DS is challenging, complicated by atypical presentations, baseline intellectual disability and normal age associated cognitive decline. Effective screening is limited by a paucity of diagnostic criteria, cognitive screening instruments and screening programs. Both observer-rated questionnaires and direct neuropsychological testing are suggested to screen for cognitive impairment, each with different strengths and weaknesses. This paper reviews commonly used screening instruments and explores the unique challenges of screening for AD in persons with DS. It concludes that single, one-dimensional screening tools and opportunistic evaluations are insufficient for detecting dementia in this population. These should be replaced by batteries of tests, incorporating informant questionnaires, direct neuropsychological testing, assessment of activities of daily living and behaviors, measured at baseline and reassessed at intervals. Developing these strategies into organized screening programs should improve diagnostic efficiency and management.

O'Dwyer, M., Finnerty, S., Henman, M., Carroll, R., McCallion, P., & McCarron, M.
Prevalence and treatment of dementia in older adults with intellectual disability in Ireland
Journal of Intellectual Disability Research, 2019, 63(8), 645.
Abstract: High rates of dementia have been reported among older adults with intellectual disability (ID), particularly those with Down Syndrome. As the use of dementia drugs in this patient group lacks an evidence base, their rates of use are of interest. Incidence and prevalence rates were determined using a combined dementia variable for three waves of the IDS-TILDA study, a nationally representative study of older adults with ID in Ireland. Incidence of dementia was defined as participants newly reporting a diagnosis and/or newly receiving dementia drug(s) at each wave. Prevalence of dementia was defined those who had reported a diagnosis at a previous wave and/or received dementia drug(s) at a previous wave. Drugs for dementia were included as a proxy for dementia diagnosis, in those with no diagnosis. Dementia incidence remained similar across Waves: 5.0% at Wave 1, 4.3% at Wave 3. Prevalence increased, 5% at Wave 1, to 9.6% by Wave 3. Those receiving receiving dementia drug(s) decreased, from 54.1% of those with dementia at Wave 1 to 28.8% at Wave 3. Three dementia drugs were reported: donepezil, memantine and rivastigmine. It was found that use of drugs for dementia decreased, despite an increased incidence. Further research into efficacy of use of a drugs is needed.

O'Leary, L., Cooper, S-A., & Hughes-McCormack, L.
Early death and causes of death of people with intellectual disabilities: A systematic review
Journal of Applied Research in Intellectual Disabilities, 2018, 31(3), 325-342. https://doi.org/10.1111/jar.12417
Abstract: Death of people with intellectual disabilities is considered to be earlier than for the general population. Databases were searched for key words on intellectual disabilities and death. Strict inclusion/exclusion criteria were used. Information was extracted from selected papers, tabulated and synthesized. Prospero registration number: CRD42015020161. Of 19,111 retrieved articles, 27 met criteria. Death was earlier by 20 years. It has improved in recent decades; however, the same inequality gap with the general population remains. More severe intellectual disabilities, and/or additional comorbidities rendered it shortest. Standardized mortality rates showed a greater inequality for women than men. Respiratory disease and circulatory diseases (with greater congenital and lesser ischemic disease compared with the general population) were the main causes of death. Cancer was less common, and cancer profile differed from the general population. Some deaths are potentially avoidable. All research is from high-income countries, and cause of death is surprisingly little investigated. Authors concluded that improved health care, including anticipatory care such as health checks, and initiatives addressing most relevant lifestyle behaviors and health risks are indicated.

Oliver, C,, Adams, D,, Holland, A,J,, Brown, S,S,G,, Ball, S,, Dodd, K,, & Carr, J.
Acquired mild cognitive impairment in adults with Down syndrome: Age-related prevalence derived from single point assessment data normed by degree of intellectual disability.
International Journal of Geriatric Psychiatry, 2021 Dec 24, 37(2), 10.1002/gps.5674. doi: 10.1002/gps.5674. Epub ahead of print.
Abstract: Individuals with Down syndrome (DS) are at significant risk for early onset Alzheimer's disease (AD), likely due to the triplication of genes on chromosome 21 that facilitate AD neuropathology. To aid the effective early diagnosis of dementia in DS, we demonstrate the strategy of using single point assessment of cognitive performance with scoring normed for degree of intellectual disability to generate age related prevalence data for acquired mild

cognitive impairment (AMCI). Four hundred and twelve adults with DS were assessed using the Neuropsychological Assessment of dementia in adults with Intellectual Disability. Normative data, banded by degree of intellectual disability, allowed identification of AMCI by atypical deviation from expected performance. AMCI was evident in approximately 20% of adults with DS aged 40 and under, 40% aged 41-50 and 45% aged 51 and over. Relative risk increased significantly in those aged 46 and over. Analysis of prevalence by 5-year age bands revealed two peaks for higher prevalence of AMCI. Psychometric data indicate single point assessment of AMCI is possible for the majority of adults with DS. Two peaks for age-related prevalence of AMCI suggest the risk for onset of AD conferred by trisomy of chromosome 21 is moderated by another factor, possibly ApoE status.

Oliver, C., & Holland, A.J.
Down's syndrome and Alzheimer's disease: a review.
Psychological Medicine, 1986, 16(2), 307-322. doi: 10.1017/s0033291700009120.
Abstract: Neuropathological change found in nearly all individuals with Down syndrome over the age of 35 years closely resembles that of Alzheimer's disease. The extent to which dementia occurs as a result of this change is unclear, and the studies which have investigated presumed cognitive deficits are reviewed. The theories put forward to explain the association between these two disorders and their possible significance to the understanding of the aetiology of Alzheimer's disease are discussed.

Oliver, C., Crayton, L., Holland, A., & Hall, S.
Cognitive deterioration in adults with Down syndrome: effects on the individual, caregivers, and service use
American Journal on Mental Retardation, 2000, 103, 455-465
Abstract: Individuals with Down syndrome (N = 49) who had participated in serial neuropsychological assessments were assigned to one of three groups comparable in level of premorbid intellectual disability: (1) those showing cognitive deterioration, (2) those comparable in age but not showing cognitive deterioration and (3) those not showing cognitive deterioration but younger. Those experiencing cognitive deterioration were less likely to receive day services, had more impoverished life experiences, and required more support compared to groups without cognitive deterioration. When age was controlled for, cognitive deterioration was significantly positively associated with carer difficulties and service use and negatively associated with life experiences for the individual. Results suggest a potential role for carer difficulties in influencing life experiences of adults with Down syndrome showing cognitive decline.

Oliver, C., Kalsy, S., McQuillan, S., & Hall, S.
Behavioural excesses and deficits associated with dementia in adults who have Down syndrome.
Journal of Applied Research in Intellectual Disabilities, 2011, 24, 208–216. https://doi.org/10.1111/j.1468-3148.2010.00604.x
Abstract: Informant-based assessment of behavioral change and difference in dementia in Down syndrome can aid diagnosis and inform service delivery. To date few studies have examined the impact of different types of behavioral change. The Assessment for Adults with Developmental Disabilities (AADS), developed for this study, assesses behavioral excesses (11 items) and deficits (17 items) associated with dementia. Inter-informant reliability, internal consistency and concurrent validity were evaluated and found to be robust. A comparison of the AADS subscale scores for three groups (n = 12) of adults with Down syndrome demonstrated more frequent deficits and excesses and greater management difficulty and effects on the individual in a dementia group than age comparable and younger groups. The AADS is a promising dementia specific measure for people with intellectual disability. Further research should evaluate change as dementia progresses and the nature of management difficulty and effects on the individual.

Olsen, R.V., Ehrenkrantz, E., & Hutchings, B.L.
Creating the movement-access continuum in home environments for dementia care
Topics in Geriatric Rehabilitation, 1996, 12(2),1-8.
DOI:10.1097/00013614-199612000-00003
Abstract: Since the majority of people with Alzheimer's disease receive some care at home, the environment of that home must be safe and supportive. In-depth interviews of 90 "seasoned" caregivers identified tactics for creating these settings through home modifications and technology. A successful modification strategy follows a three-stage movement-access continuum that responds to the disease course -- assistance, restriction with compensation, and wheelchair accessibility. Approaching home modifications along this continuum encourages independence and movement when appropriate while providing safety and control. With a sensitive and ongoing modification strategy, the home environment can become an asset rather than a liability for caregiving.

Olsen, R.V., Ehrenkrantz, E., & Hutchings, B.
Creating supportive environments for people with dementia and their caregivers through home modifications
Technology and Disability, 1993, 2(4), 47-57
Abstract: Article examines what caregivers did to enhance or modify their homes when a spouse or other family member had dementia. Authors address controlling access (using locking techniques, blocking access with gates and partial doors, and the like, as examining modifications to kitchens, bathrooms, and furniture. Data showed that many built ramps, double railings, hand grips, as well as extending landings for ease of wheelchair use, reducing riser heights, removing steps, and installing electric chair lifts. Home owners also reconfigured space and rooms. Authors conclude that home owners modified spaces to increase access and independence in some life areas and to limit or curtail access in others. Article is a good source of information for how the process and outcome of families tackle home modifications

📖 **Olsen, R.V., Ehrenkrantz, E., & Hutchings, B.**Homes that help: Advice from caregivers for creating a supportive home (Alzheimer's and Related Dementias)
77 pp.
Newark, New Jersey: New Jersey Institute of Technology [Architecture and Building Science Research Group, School of Architecture, NJIofT, University Heights, Newark, New Jersey 07102-1982] (1993)
Abstract: Manual that details examples of how to adapt a home for persons affected by dementia, covering care management techniques, physical adaptations, and personal monitoring strategies.

Owens, D., Dawson, J. C., & Losin, S.
Alzheimer's disease in Down's syndrome.
American Journal of Mental Deficiency, 1971, 75, 606–612.
Abstract: Although neuropathologists describe Alzheimer's changes in the brains of all victims of Down's syndrome over 35 yr. of age, only 3 cases of clinical dementia in such individuals are described in the literature. In order to establish clinical correlates of Alzheimer's disease, psychiatric and neurologic findings obtained from a middle-aged group were compared to those of Down's syndrome patients in their early 20s. The older group exhibited significantly greater incidence of abnormality in (a) object identification, (b) snout reflex, (c) Babinski sign, and (d) palmomental sign. Both groups displayed mild hypertonia rather than hypotonia, and face-hand test was abnormal in 75% of Ss tested. While dementia is uncommon, subtle neurological changes reflect neuropathological findings present in aging sufferers of Down's syndrome.

Paiva, A.F., Nolan, A., Thumser, C., & Santos, F.H.
Screening of cognitive changes in adults with intellectual disabilities: A systematic review
Brain Sciences, 2020, 10(11), 848; https://doi.org/10.3390/brainsci10110848
Abstract: Screening and assessment of cognitive changes in adults with Intellectual Disabilities (ID), mainly Down Syndrome (DS), is crucial to offer appropriate services to their needs. Authors present a systematic review of the existing instruments assessing dementia, aiming to support researchers and clinicians' best practice. Searches were carried out in the databases Web of Science; PubMed; PsycINFO in March 2019 and updated in October 2020. Studies were selected and examined if they: (1) focused on assessing age-related cognitive changes in persons with ID; (2) included adults and/or older adults; (3) included scales and batteries for cognitive assessment. Forty-eight cross-sectional studies and twenty-seven longitudinal studies were selected representing a total sample of 6451 participants (4650 DS and 1801 with other ID). In those studies, we found 39 scales, questionnaires, and inventories, and 13 batteries for assessing cognitive and behavioural changes in adults with DS and other ID. It was noted that the most used instrument completed by an informant or carer was the Dementia Questionnaire for Learning Disabilities (DLD), and its previous versions. The authors explore the strengths and limitations of the instruments and outline recommendations for future use.

Pape, S.

Dementia diagnostic criteria in persons with IDD.
Journal of Intellectual Disability Research, 2019, 63(8), 641.
Abstract: Dementia is a common clinical presentation among older adults with IDD, particularly those with Down syndrome. The presentation of dementia may differ compared with typical Alzheimer's disease, and criteria thus require validation in IDD populations. Data from memory assessments in individuals with Down syndrome were presented to expert raters who rated the case as dementia or no dementia using ICD-10, DSM-IV-TR and DSM-5 criteria and their own clinical judgement. Estimates were then made of the concurrent validity and reliability of clinicians' diagnoses of dementia against these manualised diagnoses. Validity of clinical diagnoses were explored by establishing the stability of diagnoses over time. Similar data from previous studies in other individuals with intellectual disabilities were compared. It was found that clinical diagnoses of dementia in Down syndrome were valid and reliable and could be used as the standard against which new criteria such as the DSM-5 are measured. Criteria had good inter-rater reliability but concurrent validity varied. Author cautions that clinicans should consider the reliability and validity of dementia diagnostic criteria when applying these in clinical settings.

Pape, S.E., Baksh, R.A., Startin, C., Hamburg, S., Hithersay, R., & Strydon, A.
The association between physical activity and CAMDEX-DS changes prior to the onset of Alzheimer's disease in Down syndrome
Journal of Clinical Medicine, 2021, Apr 27, 10(9), 1882. doi: 10.3390/jcm10091882.
Abstract: People with Down syndrome are at ultra-high risk of developing Alzheimer's dementia. At present, there are no preventative or curative treatments. Evidence from sporadic Alzheimer's disease literature suggests that lifestyle factors including physical activity may help maintain cognitive and functional skills and reduce dementia risk. Our study aimed to explore the association between regular exercise undertaken by participants with Down syndrome and changes in dementia-related domains of cognition and function. This was to consider whether physical activity may be a protective measure to delay cognitive decline and dementia in Down syndrome. Demographic, lifestyle, and health information was collected at baseline and at a two year follow up from 214 adults with Down syndrome without dementia, who also underwent assessment using the Cambridge Examination for Mental Disorders of Older People with Down Syndrome and Others with Intellectual Disabilities (CAMDEX-DS) and genetic analysis. Logistic regression models were used to examine the potential associations between decline in CAMDEX-DS domains and exercise whilst controlling for key variables. At baseline, engaging in moderate intensity exercise was associated with a 47% reduced risk of everyday skills decline and engaging in high intensity exercise was associated with a 62% reduced risk of decline in personality and behaviour. At follow-up, high levels of exercise were associated with an 87% reduced risk of decline in personality and behaviour. Moderate intensity exercise at baseline was associated with a 62% reduction in risk of decline during the follow-up period in memory and orientation. Based on our data it appears that regular moderate and high intensity exercise could reduce the risk of clinically detectable decline in a Down syndrome population with possible long-term benefits. People with Down syndrome may engage in less physical activity than their peers, and barriers remain which can prevent people with Down syndrome engaging in exercise. Our work highlights how important it is that people with Down syndrome are supported to be physically active, and to promote exercise as part of a healthy ageing plan. Clinical trials in this area would be justified to determine if engaging in exercise can lead to realistic improvements in maintaining functioning and delaying dementia onset in Down syndrome and to help develop guidance in this area.

Paton, J., Johnston, K., Katona, C., & Livingston, G
What causes problems in Alzheimer's disease: Attributions by caregivers. A qualitative study
International Journal of Geriatric Psychiatry, 2004, Jun, 19(6), 527-532. doi:10.1002/gps.1118.
Abstract: Authors soiught to gain insight into caregivers' understanding of the causes of behaviours they find problematic in people with Alzheimer's disease in order to inform the development of educational strategies. A qualitative, semi-structured interview was used. Participants were 205 caregivers for a person with Alzheimer's disease, all of whom were aware of the diagnosis and who had been recruited as part of a larger longitudinal study. Participants were from inner-city and suburban London/semi-rural Essex. The main outcome measures were caregivers' understanding of: the cause of problematic behaviour; the ability of the person with dementia to control this behaviour; the prognosis of the illness. Most carers attribute the cognitive, behavioural and psychological symptoms of dementia to causes other than dementia; many believe that the person with dementia has control over their behaviour and substantial numbers believe the person with dementia will return to normal. This study suggests that providing facts about the illness to caregivers is not enough, as caregivers may not understand that the symptoms they observe are related to the diagnosis. Education by clinicians should focus on the understanding of caregivers and in particular explore the caregivers' attributions of the symptoms which are present in the person for whom they care.

Patti, P., Amble, K. & Flory, M.
Placement, relocation and end of life issues in aging adults with and without Down's syndrome: A retrospective study.
Journal of Intellectual Disability Research, 2010, 54(6), 538-546. doi: 10.1111/j.1365-2788.2010.01279.x.
Abstract: Aging adults with Down's syndrome (DS) experience more relocations and other life events than adults with intellectual disabilities aged 50 and older without DS. Age-related functional decline and the higher incidence of dementia were implicated as the contributing factors that led to relocation and nursing home placement. A retrospective study of adults with intellectual disabilities who were born prior to the year 1946 was conducted to analyze the number of relocations experienced over a 5- and 10-year period. The cohort consisted of 140 individuals (61 with DS between ages 50–71 years, and 79 without DS between ages 57–89 years) who had been referred to a diagnostic and research clinic. Analyses revealed the number of relocations over a 5- and 10-year period were significantly greater in the DS group. Placement in a nursing home for end of life care was significantly higher in the DS group whereas the majority (90%) in the non-DS group remained in a group home setting. Mortality was significantly earlier in the DS group with the mean age at death to be 61.4 years compared with 73.2 years in the non-DS group. The authors concluded that the present results suggest that aging adults with DS encounter more relocations, and ar e more likely to have their final placement for end of life care in a nursing home. In contrast, the adults without DS were subjected to less relocation and remained in the same group home setting.

Pary, R.J.
Differential diagnosis of functional decline in Down's syndrome
Habilitative Mental Healthcare Newsletter, 1992, 11(6), 37-41.
https://www.worldcat.org/title/habilitative-mental-healthcare-newsletter/oclc/21277851
Abstract: (non provided - extract from article) Breif overview of examining adults with Down syndrome (DS) for functional decline. Provides brief summary of Alzheimer's disease and the prevalence and occurrence of AD in persons with DS. Provides two tables, Table 1: Disorders which are common in DS and Miscellaneous cerebral conditions with no apparent predilections for individuals with DS; Table 2: Work-up for functional decline in DS. Discusses complicating factors such as thyroid disease, major depression, sensory impairment, infection, and other conditions. Provides brief description of factors comprising a diagnostic workup. References (n-49) provide historical glimpse into extant topical literature prior to 1992.

Perera, B., Kamieniarz, L., Iftikhar, M. & Solomou, S.
Screening and diagnosing dementia in people with Down's syndrome: implications of using the DLD questionnaire
Advances in Mental Health and Intellectual Disabilities, 2022, 16(4), 239-248. https://doi.org/10.1108/AMHID-04-2022-0015
Abstract: The Dementia Questionnaire for People with Learning Disabilities (DLD) is one of the main screening and monitoring tools for dementia in people with Down's syndrome (DS). As part of a quality improvement project to improve the care for people with DS and dementia in an intellectual disability service, the authors studied the screening and monitoring process by retrospectively investigating the use of DLD and exploring clinicians' experience of using it. DLDs completed in the service was retrospectively assessed. Changes in DLD scores were matched against people who received a clinical diagnosis of dementia. Data were analysed to estimate sensitivity, specificity and predictive values of DLD. A questionnaire was used to assess clinicians' experience. Data for 20 service users was collected. DLD cognitive scores showed 80% sensitivity and 60% specificity for the diagnosis of dementia, with a positive predictive value of 40% and negative predictive value of 90%. Staff found DLD to be easy to perform but time consuming. This led to the preparation of a decision tool for appropriateness of performing a DLD. **The results show that a negative DLD helps to exclude dementia where there is concern over cognitive decline,**

but a positive result is not specific enough to suggest the possibility of dementia. This shows that DLD may have limitations if used as a screening tool alone but could be used for the monitoring of the disease trajectory of those with a confirmed diagnosis as well as to establish a baseline DLD when a person is screened for dementia first.

Persaud, M., & Jaycock, S.
Evaluating care delivery: the application of dementia care mapping in learning disability residential services
Journal of Learning Disabilities, 2001, 5(4), 345-352.
https://doi.org/10.1177/146900470100500
Abstract: Measurement and evaluation in intellectual disability services is still in its infancy. This report explores how good practice in relation to quality of care initiatives in dementia care transpose into intellectual disability settings. The authors applied dementia care mapping (DCM) to evaluate its effectiveness and efficiency in generic intellectual disability settings. Results showed that the application of the method to be partially successful. The data produced compared favorably in quality, quantity and detail with those collected in dementia care areas. Analysis of data revealed great potential for the method; however, result indices and coding frameworks need to be modified and adapted in future studies. No subject had dementia.

Peterson, M.E., & O'Bryant, S.E.
Blood-base biomarkers for Down syndrome and Alzheimer's disease: A systematic review
Developmental Neurobiology, 2019, 79(7), 699-710.
https://doi.org/10.1002/dneu.22714
Abstract: Down syndrome (DS) occurs due to triplication of chromosome 21. Individuals with DS face an elevated risk for development of Alzheimer's disease (AD) due to increased amyloid beta (Aß) resulting from the over-expression of the amyloid precursor protein found on chromosome 21. Diagnosis of AD among individuals with DS poses particular challenges resulting in an increased focus on alternative diagnostic methods such as blood-based biomarkers. The aim of this review was to evaluate the current state of the literature of blood-based biomarkers found in individuals with DS and particularly among those also diagnosed with AD or in prodromal stages (mild cognitive impairment [MCI]). A systematic review was conducted utilizing a comprehensive search strategy. Twenty-four references were identified, of those, 22 fulfilled inclusion criteria were selected for further analysis with restriction to only plasma-based biomarkers. Studies found Aß to be consistently higher among individuals with DS; however, the link between Aß peptides (Aß1-42 and Aß1-40) and AD among DS was inconsistent. Inflammatory-based proteins were more reliably found to be elevated leading to preliminary work focused on an algorithmic approach with predominantly inflammatory-based proteins to detect AD and MCI as well as predict risk of incidence among DS. Separate work has also shown remarkable diagnostic accuracy with the use of a single protein (NfL) as compared to combined proteomic profiles. This review serves to outline the current state of the literature and highlights the potential plasma-based biomarkers for use in detecting AD and MCI among this at-risk population.

Petronis, A.
Alzheimer's disease and down syndrome: from meiosis to dementia
Experimental Neurology, 1999, Aug, 158(2), 403-413.
doi:10.1006/exnr.1999.7128.
Abstract: Several molecular and clinical similarities have been detected in Alzheimer's disease (AD) and Down syndrome (DS). The most remarkable feature is abnormal accumulation of beta-amyloid in the brains of both individuals affected with AD and aging DS patients followed by dementia. In addition, AD patients exhibit dermatoglyphic patterns similar to those in DS, and late maternal age is a risk factor in both diseases. AD and DS could be related genetically because AD families exhibit a higher rate of DS cases and vice versa. Although numerous discoveries have been made in the elucidation of the etiopathogenic factors in AD and DS, little progress has been achieved in understanding the origin of the common features of the two diseases. This article reviews clinical and molecular similarities in DS and AD and also chromosome 21 studies in both diseases. A new hypothesis explaining the association between AD and DS is suggested, and this hypothesis is based on the poorly understood molecular phenomenon of aberrant meiotic recombination. Aberration in meiotic recombination has been consistently detected in chromosomal diseases including trisomy 21 and sex chromosomes. There are no studies dedicated to meiotic recombination in genetic diseases; however, evidence for disturbed recombination has been documented in several neurological diseases such as Huntington's disease, myotonic dystrophy, and fragile X syndrome. Interestingly, the rate of trisomic XXY children born to mothers transmitting fragile X mutation is higher than expected. This finding suggests that AD could be associated with DS in a similar way to which fragile X syndrome is related to trisomy of sex chromosomes. Based on analogy with fragile X syndrome, it can be predicted that AD should demonstrate aberrant meiotic recombination in chromosome 21, most likely in the region D21S1/S11-D21S16 which is linked to early onset familial AD. Based on the same rationale, different patterns of meiotic recombination in the nondisjunct chromosome 21 within DS patients grouped according to the concomitant disease are predicted.

Prasher, V.
End-stage dementia in adults with Down syndrome
International Journal of Geriatric Psychiatry, 1995, 10(12), 1067-1069.
https://doi.org/10.1002/gps.930101213
Abstract: End-stage dementia in adults with Down syndrome has not been fully investigated. Available information, 6 months prior to death, for 20 adults with Down syndrome who had died with Alzheimer's disease was reviewed. A terminal stage of severe intellectual deterioration, marked personality and mood changes, loss of sphincter control, seizure activity, immobility with hypertonia and complete loss of self-care skills was found. These findings have important clinical and service implications.

Prasher, V.P.
Review of donepezil, rivastigmine, galantamine and memantine for the treatment of dementia in Alzheimer's disease in adults with Down syndrome: implications for the intellectual disability population
International Journal of Geriatric Psychiatry, 2004, 19, 509 - 515. doi: 10.1002/gps.1077.
Abstract: The management of dementia in Alzheimer's disease has dramatically changed since the development of anti-dementia drugs. However, there is limited information available regarding the bio-medical aspects of the differing drugs; particularly relating to adults with intellectual disability. Indeed the information available for the intellectual disabled population is limited to adults with Down syndrome. This review highlights the important pharmacological and clinical aspects of donepezil, rivastigmine, galantamine and memantine and supports the view that such drugs play an important part in the management of dementia in adults with intellectual disability. Future clinical and research issues are discussed.

Prasher, V.P., & Corbett, J.A.
Onset of seizures as a poor indicator of longevity in people with down syndrome and dementia
International Journal of Geriatric Psychiatry,1993, 8(11), 923-927.
https://doi.org/10.1002/gps.930081106
Abstract: An association between Down syndrome and dementia of Alzheimer type is well established. This study demonstrates that late onset seizures in people with Down syndrome are a strong indicator of a dementing process. Further, late onset seizures in people with Down syndrome may be used as a prognostic indicator, indicating life expectancy of less than a further 2 years, probable death within 3 years and death almost invariably within 5 years of onset.

Prasher, V., Farooq, A. & Holder, R.
The Adaptive Behavior Dementia Questionnaire (ABDQ): screening questionnaire for dementia in Alzheimer's disease in adults with Down syndrome
Research in Developmental Disabilities, 2004, 25(4), 385-397. doi: 10.1016/j.ridd.2003.12.002.
Abstract: The diagnosis of dementia in Alzheimer's disease remains at times problematic in adults with intellectual disability. The analysis of 5-year consecutive data developed a researched-based clinical screening tool for dementia in Alzheimer's disease in adults with Down syndrome. The Adaptive Behavior Dementia Questionnaire (ABDQ) is a 15-item questionnaire, which is used to detect change in adaptive behavior. The scale has good reliability and validity, with an overall accuracy of 92%. It is one of the first clinical tools designed specifically to screen for dementia in Alzheimer's disease in adults with Down syndrome.

Prasher, V.P.,& **Filer, A.**
Behavioural disturbance in people with Down's syndrome and dementia.
Journal of Intellectual Disabilities Research, 1995, 39(5), 432-436. doi: 10.1111/j.1365-2788.1995.tb00547.x.

Abstract: Behavioral disturbance associated with dementia in people with Down syndrome has not been fully researched. This study investigated such problems in subjects with Down syndrome and dementia and controls with Down syndrome but free of dementia. Changes in mood, difficulty with communication, gait deterioration, loss of self-care skills, sleep disturbance, day-time wandering and urinary incontinence were found to be associated with dementia. Problems giving the greatest cause for concern to carers were restlessness, loss of communication skills, urinary incontinence and wandering. Care provision specifically focused on management of behavioral disturbance in individuals who develop dementia is recommended.

Prasher, V.P. & Krishnan, V.H.R.
Age of onset and duration of dementia in people with Down syndrome: Integration of 98 reported cases in the literature
International Journal of Geriatric Psychiatry, 1993 Nov, 8(11), 915-922 https://doi.org/10.1002/gps.930081105
Abstract: The published case reports of 98 people with Down Syndrome were studied with respect to the age of onset and duration of clinically diagnosed dementia. The incidence of dementia was unimodal, increasing rapidly from 40 years to a peak of 30% incidence in the fifth decade of life, followed by a further rapid decline. Females with Down syndrome had an earlier onset. Duration of dementia decreased with increasing age of onset for both males and females.

Prasher, V.P., Janicki, M.P., Jozsval, E., Berg, J.M., Lovering, J.S., Rashid, A., Fung, W.L.A., & Percy, M.
Alzheimer's disease and dementia: Implications for people with Down syndrome and other intellectual and developmental disabilities.
Chapter 49 (pp. 709-733) in Wehmeyer, M., Brown, I., Percy, M., Shogren, K.A., Fung, W.L.A., (Eds.), *A Comprehensive Guide to Intellectual and Developmental Disabilities*. Baltimore, MD: Paul Brookes Publishing.
Abstract: Chapter provides comprehensive information in its first three sections on Alzheimer's disease and dementias and how these disorders affect people with Down syndrome and other intellectual disabilities, including assessment and early detection of dementia. The fourth section covers the multidisciplinary management of dementia as occuring in people with intellectual disabilities. Subsections cover pharmacological approaches and nonpharmacological interventions, as well as environmental considerations for safe living, and services and resources needed by people with dementia and their caregivers.

Prasher, V.P., Mahmood, H., & Mitra, M.
Challenges faced in managing dementia in Alzheimer's disease in patients with Down syndrome.
Degenerative Neurological and Neuromuscular Disease, 2016, 6, 85-94. doi: 10.2147/DNND.S91754. eCollection 2016.
Abstract: Dementia in Alzheimer's disease (DAD) is more common in adults with Down syndrome (DS), with characteristically an earlier onset. The treatment of DAD is not too dissimilar in the general population and in people with intellectual disabilities. However, the underlying intellectual disability can make the management of DAD more challenging in older adults with DS. This literature review aimed to look at the management of DAD in people with DS. The management of dementia is holistic. This includes treating reversible factors, aiming to slow the cognitive decline, psychological therapies, ensuring that the environment is appropriate, and use of psychotropic medication when necessary to manage behavioral problems, psychotic symptoms, depressive symptoms, and sleep difficulty. Antidementia medications have a role to play but remain limited. The management of DAD in the DS population can be at times challenging, but good clinical practice should involve accurate diagnosis of dementia, treating any reversible additional factors, consideration of psychological and behavioral management, use of antidementia medication, and a multidisciplinary team approach.

Prasher, V.P., Metseagharun, T., & Haque, S.
Weight loss in adults with Down syndrome and with dementia in Alzheimer's disease.
Research in Developmental Disabilities, 2004, Jan-Feb, 25(1), 1-7. doi: 10.1016/j.ridd.2003.04.005.
Abstract: An association between weight loss and Alzheimer's disease has been established in the general population but little information is available regarding this association in people with intellectual disabilities. A 4-year longitudinal study of adults with Down syndrome with and without Alzheimer's disease was undertaken. Age-associated weight loss was seen in virtually all older adults with Down syndrome. A significant association between weight loss and Alzheimer's disease was found for older adults with Down syndrome. This study highlights important research and clinical issues regarding weight loss and nutrition in Down syndrome adults with dementia.

Prasher, V.P., Sachdeva, N., & Tarrant, N.
Diagnosing dementia in adults with Down's syndrome. 2015, *Neurodegenerative Disease Management*, 5(3), 249-256.
Abstract: Individuals with Down's syndrome (DS) are living longer and many will survive into their fifth or sixth decade of life. Among the DS population, the prevalence of dementia in Alzheimer's disease increases from 9.4% in age group 30-39 years to 54.5% age group 60-69 years. The psychopathology of dementia in Alzheimer's disease is similar to that seen in the general population although differences are apparent due to the underlying intellectual disability in DS and on the reliance on collateral information from informants. The diagnostic workup follows accepted practice although neuropsychological tests and neuroimaging will only be adjuncts to the clinical assessment; such investigations have limited diagnostic value. Presently, research is focused on identifying genetic and biological measures of Alzheimer's disease in DS.

Prasher, V.P., Percy, M,, Janicki, M.P., Jozsvai, E., Fung, W.L.A., & Brown, I.
Implications of dementia for adults with developmental disabilities.
Chapter (pp. 699-721) in Brown, I. & Percy, M. (2020). *Developmental Disabilities in Ontario* (4th Ed.), Toronto, Ontario, Canada: Delphi Graphic Communications. https://oadd.org/publications/textbook/
Abstract: Chapter provides an introduction to the topic of dementia in persons with developmental disabilities. Dementia, as a worldwide public health concern, is increasing in prevalence markedly because the world's population if living longer and aging in greater numbers. Chapter covers the physiology of dementia, options for services, mechanisms for multidisciplinary management, and advances in advocacy, dementia prevention, and dementia research.

Proveda, B., & Broxholme, S.
Assessments for dementia in people with learning disabilities: Evaluation of a dementia battery developed for people with mild to moderate learning disabilities
Learning Disability Practice, 2016, 19(1), 31-40. doi.org/10.7748/ldp.19.1.31.s23
Abstract: An intellectual disabilities' dementia battery was developed to assess cognitive abilities in individuals referred to the intellectual disabilities service because of concerns of possible dementia. The present study aimed to establish concurrent validity with previously validated measures of cognitive ability and its clinical effectiveness in detecting dementia in this population. Fifty-five individuals aged 29 and over (range: 29 to 71), received a baseline and a follow-up assessment using the dementia battery between 2000 and 2010. Differences in performance between individuals allocated to 'probable', 'unsure' and 'no' dementia groupings were investigated at domain and subtest level, as well as overall performance. Results on the battery were compared with clinically relevant measures of dementia also included in the local assessment protocol. Significant differences in overall performance were found between the 'probable' and 'no' dementia groups as well as cognitive domain-specific differences. No differences were found at subtest level. Good concurrent validity was found between the battery and comparable measures of change within the dementia assessment protocol, namely the VABS, DMR and BPVS II. The intellectual disabilities' dementia battery appears to be a good measure, which can be used longitudinally, to detect change in individuals and help establish a diagnosis of dementia. It is also comparable with other measures of change incorporated in the dementia assessment protocol. Subtests included in the language domain appear to be the most relevant at detecting significant changes between baseline and follow up. Future studies should attempt to standardize this measure and establish cut-off scores.

PTAC [PASRR Technical Assistance Center]
How does a categorical determination for dementia and intellectual disability affect the PASRR process?
PTAC, April 10, 2018;
https://www.pasrrassist.org/resources/How-does-a-categorical-determination-for-dementia-and-intellectual-disability-affect-the-PASRR-process%3F
Abstract: 42 CFR 483.130(h) provides that the State intellectual disability authority may make categorical determinations that individuals with dementia, which exists in combination with an intellectual disability (ID) or a related condition, do not need specialized services. A categorical determination for dementia and ID can be applied at the Level I screening, but the categorical must

be determined at the Level II evaluation phase of PASRR. Beyond the specialized services determination, there is no basis for ending a Level II evaluation for an individual with an intellectual disability diagnosis, as the evaluation still determines if nursing facility services are needed. Under 42 CFR § 483.128(j), findings must be issued in the form of an abbreviated written evaluative report which— (1) Identifies the name and professional title of the person applying the categorical determination and the data on which the application was made; (2) Explains the categorical determination(s) that has (have) been made and, if only one of the two required determinations can be made categorically, describes the nature of any further screening which is required; (3) Identifies, to the extent possible, based on the available data, NF services, including any mental health or specialized psychiatric rehabilitative services, that may be needed; and (4) Includes the bases for the report's conclusions. A categorical determination for dementia and intellectual disability also requires the issuing of a written determination notice per the following 483.130(k)(l) requirements: (k) Notice of determination. The State mental health or intellectual disability authority must notify in writing the following entities of a determination made under this subpart: 1. The evaluated individual and legal representative; 2. The admitting or retaining NF; 3. The individual or resident's attending physician; and 4. The discharging hospital, unless the individual is exempt from preadmission screening as provided at §483.106(b)(2). (l) Contents of notice. Each notice of the determination made by the State mental health or intellectual disability authority must include: 1. Whether a NF level of service is needed; 2. Whether specialized services are needed; 3. The placement options that are available to the individual consistent with these determinations; and 4. The rights of the individual to appeal the determination

Puri, B.K., Ho, K.W., & Singh, I.
Age of seizure onset in adults with Down's syndrome.
International Journal of Clinical Practice, 2001, 55(7), 442-444.
https://pubmed.ncbi.nlm.nih.gov/11594252/
Abstract: In a cohort of 68 adults (35 males and 33 females) with Down's syndrome aged 29-83 years, a history of seizures was found in 26.5%. The overall mean age of onset of seizures was 37 years, males (22 years) being significantly younger than females (51 years). The age of onset was bimodally distributed, with the first peak occurring in the first two decades, and a late-onset peak occurring in the fifth and sixth decades. A strong association between Alzheimer's disease and seizures was confirmed. Of those with a history of seizures, those aged over 45 years were significantly more likely to develop Alzheimer's disease than those younger than 45. It is suggested that late-onset epilepsy in Down's syndrome is associated with Alzheimer's disease, while early-onset epilepsy is associated with an absence of dementia.

Pulsifer, M.B., Evans, C.L., Hom, C., Krinsky-McHale, S.J., Silverman, W., Lai, F., Lott, I., Schupf, N., Wen, J ., Rosas, H.D.
Language skills as a predictor of cognitive decline in adults with Down syndrome
Alzheimers Dement (Amst), 2020, Aug 25, 12(1), e12080.
doi: 10.1002/dad2.12080. eCollection 2020.
Abstract: Adults with Down syndrome (DS) are at high risk for early onset Alzheimer's disease (AD), characterized by a progressive decline in multiple cognitive domains including language, which can impact social interactions, behavior, and quality of life. This cross-sectional study examined the relationship between language skills and dementia. A total of 168 adults with DS (mean age = 51.4 years) received neuropsychological assessments, including Vineland Communication Domain, McCarthy Verbal Fluency, and Boston Naming Test, and were categorized in one of three clinical groups: cognitively stable (CS, 57.8%); mild cognitive impairment (MCI-DS, 22.6%); and probable/definite dementia (AD-DS, 19.6%). Logistic regression was used to determine how well language measures predict group status. Vineland Communication, particularly receptive language, was a significant predictor of MCI-DS. Semantic verbal fluency was the strongest predictor of AD-DS. Assessment of language skills can aid in the identification of dementia in adults with DS. Clinically, indications of emerging language problems should warrant further evaluation and monitoring.

Rafii, M.S.
Tau PET imaging for staging of Alzheimer's disease in Down syndrome
Developmental Neurobiology, 2019, 79(7), 711- 715.
https://doi.org/10.1002/dneu.22658
Abstract: Alzheimer's disease (AD) pathology and early-onset dementia develop almost universally in Down syndrome (DS). AD is defined neuropathologically by the presence of extracellular plaques of aggregated amyloid ß protein and intracellular neurofibrillary tangles (NFTs) of aggregated hyperphosphorylated tau protein. The development of radiolabeled positron emission tomography (PET) ligands for amyloid plaques and tau tangles enables the longitudinal assessment of the spatial pattern of their accumulation in relation to symptomatology. Recent work indicates that amyloid pathology develops 15–20 years before neurodegeneration and symptom onset in the sporadic and autosomal dominant forms of AD, while tau pathology correlates more closely with symptomatic stages evidenced by cognitive decline and dementia. Recent work on AD biomarkers in DS illustrates similarities between DS and sporadic AD. It may soon be possible to apply recently developed staging classifications to DS to obtain a more nuanced understanding of the development AD in DS and to provide more accurate diagnosis and prognosis in the clinic.

Rafii, M., & Santoro, S.L.
Prevalencd and severity of Alzheimer disease in individuals with dementia.
JAMA Neurology, 2019, 76, 142-143.
Abstract: The median life expectancy for a child with Down syndrome (DS) born in the 1950s was less than 10 years of age, with congenital heart defects being the main cause of death. With advances in medical care and improvements in the overall health of individuals with DS, life expectancy has increased dramatically; for children with DS born in 2010, median life expectancy is estimated to be 65 years. However, along with this longer lifespan comes the prospect of a considerable increase in the risk of developing dementia associated with Alzheimer disease (AD), with a prevalence of nearly 80% for those with DS who are older than 65 years.

Rafii, M.S., Ances, B.M., Schupf, N., Krinsky-McHale, S.J., Mapstone, M., Silverman, W., Lott, I....O'Bryant, S.
The AT(N) framework for Alzheimer's disease in adults with Down syndrome.
Alzheimer's & Dementia (Amst), 2020,Oct 27,12(1),e12062. doi: 10.1002/dad2.12062. eCollection 2020.
Abstract: The National Institute on Aging in conjunction with the Alzheimer's Association (NIA-AA) recently proposed a biological framework for defining the Alzheimer's disease (AD) continuum. This new framework is based upon the key AD biomarkers (amyloid, tau, neurodegeneration, AT[N]) instead of clinical symptoms and represents the latest understanding that the pathological processes underlying AD begin decades before the manifestation of symptoms. By using these same biomarkers, individuals with Down syndrome (DS), who are genetically predisposed to developing AD, can also be placed more precisely along the AD continuum. The A/T(N) framework is therefore thought to provide an objective manner by which to select and enrich samples for clinical trials. This new framework is highly flexible and allows the addition of newly confirmed AD biomarkers into the existing AT(N) groups. As biomarkers for other pathological processes are validated, they can also be added to the AT(N) classification scheme, which will allow for better characterization and staging of AD in DS. These biological classifications can then be merged with clinical staging for an examination of factors that impact the biological and clinical progression of the disease. Here, we leverage previously published guidelines for the AT(N) framework to generate such a plan for AD among adults with DS.

Rafii, M.S., Wishnek, H., Brewer, J.B., Donohue, M.C., Ness, S., Mobley, W.C., Aisen, P.S., & Rissman, R.A.
The Down syndrome biomarker initiative (DSBI) pilot: Proof of concept for deep phenotyping of Alzheimer's disease biomarkers in Down syndrome
Frontiers in Behavioral Neuroscience, 2015, Sep 14, 9, 239.
doi: 10.3389/fnbeh.2015.00239. eCollection 2015.
Abstract: To gain further knowledge on the preclinical phase of Alzheimer's disease (AD), we sought to characterize cognitive performance, neuroimaging and plasma-based AD biomarkers in a cohort of non-demented adults with down syndrome (DS). The goal of the down syndrome biomarker Initiative (DSBI) pilot is to test feasibility of this approach for future multicenter studies. We enrolled 12 non-demented participants with DS between the ages of 30-60 years old. Participants underwent extensive cognitive testing, volumetric MRI, amyloid positron emission tomography (PET; 18F-florbetapir), fluorodeoxyglucose (FDG) PET (18F-fluorodeoxyglucose) and retinal amyloid imaging. In addition, plasma beta-amyloid (Aß) species were measured and Apolipoprotein E (ApoE) genotyping was performed. Results from our multimodal analysis suggest greater hippocampal atrophy with amyloid load. Additionally, we identified an inverse relationship between amyloid load and regional glucose metabolism. Cognitive and functional measures did not correlate with amyloid load in DS but did correlate with regional FDG PET measures. Biomarkers of AD can be readily studied in adults with DS as in other preclinical AD populations. Importantly, all

subjects in this feasibility study were able to complete all test procedures. The data indicate that a large, multicenter longitudinal study is feasible to better understand the trajectories of AD biomarkers in this enriched population.

Raj,S., Stanley, M., Mackintosh,S., & Fryer, C.
Scope of occupational therapy practice for adults with both Down syndrome and dementia: A cross-sectional survey
Australian Ocupational Therapy Journal, 2022, 67(3), 218-228. https://doi.org/10.1111/1440-1630.12645
Abstract: Dementia in adults with Down syndrome causes a progressive decline in daily occupations impacting both persons with Down syndrome and their informal caregivers. This study aimed to explore the scope of occupational therapy practice for adults with both Down syndrome and dementia and their informal caregivers living in their homes. A survey was conducted with occupational therapists having clinical experience in providing interventions for adults with Down syndrome. A web-based survey was developed to explore occupational therapy practice for this group of people with Down syndrome and their informal caregivers. Responses to closed-ended questions were analysed descriptively, and inductive content analysis was used for open-ended questions. Forty-three occupational therapists from Australia, Canada, United Kingdom and the United States of America participated in the survey. Two-thirds were from the United Kingdom, most of whom were employed in the public sector and had at least 10 years of clinical experience. Over 90% of respondents received one or more referrals in a typical month for adults with Down syndrome having dementia, 68% of which were for a decline in activities of daily living. Home environment and activities of daily living were frequently assessed areas, and the commonest interventions were compensatory strategies and environmental modifications. Only half the respondents provided interventions for informal caregivers. Risk and safety and manual handling were frequently addressed domains for informal caregivers. Collaboration and developing clinical expertise were the two key perceived enablers for providing effective occupational therapy services. Fragmentation of services and a lack of client-centred care were the common perceived barriers. Occupational therapists often address decline in activities of daily living for individuals with both Down syndrome and dementia. To support participation in meaningful occupations for these people and support the needs of their informal caregivers, it is essential that services are offered in a collaborative approach.

Raj, S.E., Mackintosh,S., Kernot,J., Fryer, C., & Stanley, M.
Development and feasibility testing of an evidence-based occupational therapy program for adults with both Down syndrome and dementia
Journal of Policy and Practice in Intellectual Disabilities, 2022, First published: 28 June 2022 https://doi.org/10.1111/jppi.12435
Abstract: This paper describes the development of a home-based occupational therapy intervention program for people with Down syndrome who experience early on-set dementia causing a decline in their performance skills and increasing care dependency on their informal caregivers. A six-step methodological process adapted from the Medical Research Council framework for developing and evaluating complex interventions was formulated to develop an evidence-based occupational therapy program for people with both Down syndrome and dementia and their informal caregivers. The first two steps gathered evidence through systematic reviews of the literature and determined the scope of current occupational therapy practice. The gathered evidence was synthesised in step three to develop a client-centred occupational therapy intervention program for persons with both Down syndrome and dementia and their informal caregivers. In steps four and five, opinions were sought from occupational therapists working in this area of practice on the content of the developed program and its feasibility within the Australian disability services context. The final testing step can be conducted in the future using a single-case experimental design study. It is important to use rigorous frameworks and gather comprehensive evidence using multiple methods to develop interventions for small heterogeneous populations. The developed occupational therapy program for persons with both Down syndrome and dementia and their informal caregivers appears feasible to be implemented within the Australian disability services; however, funding limitations imposes barriers for its implementation in clinical practice.

Rebillat, A-S., Hiance-Delahaye, A., Falquero, S., Radice, G., & Sacco, S.
The French translation of the dementia screening questionnaire for individuals with intellectual disabilities is a sensitive tool for screening for dementia in people with Down Syndrome.
Research in Developmental Disabilities, 2021 Nov; 118, 104068. doi: 10.1016/j.ridd.2021.104068. Epub 2021 Aug 28.
Abstract: People with Down Syndrome (DS) are at an increased risk of developing Alzheimer's Disease (AD) relatively early in life. The dementia screening questionnaire for individuals with intellectual disabilities (DSQIID) has been developed for people with intellectual disabilities and was shown to have high discriminative power to distinguish between people with and without dementia. The objective of this study was to verify if the French version of the DSQIID (DSQIID-F) had a good diagnostic specificity and to determine the optimal cut-off for screening people with DS for dementia. This was a single-centre, retrospective, medical chart review study in people with DS aged =40 years. Demographics, level of intellectual disability, DSQIID-F data and clinical assessment of dementia were extracted from medical records. Sensitivity and specificity for different DSQIID-F cut-offs were calculated to determine the optimal cut-off. Some 151 people with DS were included with a median age of 51 years. The optimal DSQIID-F cut-off was 19, sensitivity was 0.940 (95 % CI: 0.830; 0.985) and specificity was 0.941 (95 % CI: 0.873; 0.975). Results were comparable to those for the English DSQIID (cut-off: 20; sensitivity: 0.92; specificity: 0.97). However, the psychometric qualities of the DSQIID-F, used for clinical follow-up, have not been verified. The DSQIID-F has good discriminative power and represents a useful tool to screen people with DS for dementia

Reppermund, S., & Trollor, J.N.
Successful ageing for people with an intellectual disability
Current Opinion in Psychiatry, 2016, March, 29(2), 149-154. doi: 10.1097/YCO.0000000000000228
Abstract: Successful ageing has not yet been defined in people with an intellectual disability. The purpose of this review is to discuss and define successful ageing in the context of intellectual disability and to propose strategies to improve health and wellbeing for this population. People with an intellectual disability experience higher rates of diabetes, hypertension, obesity and cardiovascular disease, and higher rates of mental disorders than people without an intellectual disability. People with an intellectual disability engage in more passive leisure activities because many active leisure activities require the participation of or assistance by others. Health promotion programmes tailored to people with an intellectual disability consisting of exercise and health education can result in more positive attitudes toward exercise and improvements in psychosocial outcomes. With modifications for people with an intellectual disability, the concept of successful ageing can be used as a template for development of strategies to improve health and wellbeing for people with an intellectual disability as they age. Targeted programmes focused on health promotion and prevention of age-related morbidities is required. There is a need for policies addressing positive ageing, including social participation and maximizing community participation. Appropriate and ongoing education for people with an intellectual disability and their carers on healthy living in areas of physical, social, and cognitive activity, nutrition and avoidance of risk factors is essential.

Reid, A. H., & Aungle, P. G.
Dementia in ageing mental defectives: A clinical psychiatric study. Journal of *Mental Deficiency Research,* 1974, 18, 15–23. Doi: 10.1111/j.1365-2788.1974.tb01214.x
Abstract: Review of literature on dementia and Down syndrome to date [no published abstract]

Robertson, J., Hatton, C., Emerson, E., Baines, S.
Prevalence of epilepsy among people with intellectual disabilities: A systematic review.
Seizure, 2015, 29, 46-62. doi: 10.1016/j.seizure.2015.03.016.
Abstract: Epilepsy is more common in people with intellectual disabilities than in the general population. However, reported prevalence rates vary widely between studies. This systematic review aimed to provide a summary of prevalence studies and estimates of prevalence based on meta-analyses. Studies were identified via electronic searches using Medline, Cinahl and PsycINFO and cross-citations. Information extracted from studies was tabulated. Prevalence rate estimates were pooled using random effects meta-analyses and subgroup analyses were conducted. A total of 48 studies were included in the tabulation and 46 studies were included in meta-analyses. In general samples of people with intellectual disabilities, the pooled estimate from 38 studies was 22.2% (95% CI 19.6-25.1). Prevalence increased with increasing level of intellectual disability. For samples of people with Down syndrome, the pooled estimate from data in 13 studies was 12.4% (95% CI 9.1-16.7), decreasing to 10.3% (95% CI 8.4-12.6) following removal of two studies focusing on older people. *Prevalence increased*

with age in people with Down syndrome and was particularly prevalent in those with Alzheimer's/dementia. Epilepsy is highly prevalent in people with intellectual disabilities. Services must be equipped with the skills and information needed to manage this condition.

Robinson, A., Spencer, B., & White, L.
Understanding difficult behaviors: Some suggestions for coping with Alzheimer's disease and related illnesses
80 pp.
Geriatric Education Center of Michigan (Alzheimer's Education Program, Eastern Michigan University, P.O. Box 981337, Ypsilanti, MI 48198-1337; www.emich.edu/public/alzheimers) (1999 rev.)
Abstract: Manual format publication providing detailed information on addressing difficult behaviors and understanding their causes and environmental relationships. Specific detailed sections on angry, agitated behavior; hallucinations and paranoia; incontinence; problems with bathing, dressing, eating, sleeping and wandering; repetitive actions, screaming and verbal noises, and wanting to go home. Appendix contains selected readings, and audio-visual materials. Does not specifically focus on intellectual disabilities, but is good generic resource.

Ross, W.T., & Olsen, M.
Care of the adult patient with Down syndrome
Southern Medical Journal, 2014, 107(11), 715-721.
doi:10.14423/SMJ.0000000000000193.
Abstract: Individuals with Down syndrome have an increased risk for many conditions, including cardiovascular disease, cancer, infections, and osteoporosis, and endocrine, neurological, orthopedic, auditory, and ophthalmic disorders. They also are at increased risk for abuse and human rights violations and receive fewer screenings and interventions than the population without Down syndrome. In this literature review, the most common health conditions associated with Down syndrome are examined, along with the topics of sexual abuse, menstrual hygiene, contraception, and human rights. Clinical guidelines for this population are summarized in an effort to assist practicing physicians in improving their provision of health care to the adult patient with Down syndrome.

Rösner, P., Berger, J., Tarasova, D., Birkner, J., Kaiser, H., Diefenbacher, A., & Sappok, T.
Assessment of dementia in a clinical sample of persons with intellectual disability
Journal of Applied Research in Intellectual Disability, 2021, X(x), 1-12.
doi:10.1111/jar.12913
Abstract: Assessment of age-associated disorders has become increasingly important. In a clinical setting, people with intellectual disability with and without dementia were assessed retrospectively using the Neuropsychological Test Battery (NTB) and the Dementia Questionnaire for People with Learning Disabilities (DLD) at two different times to analyse neuropsychological changes and diagnostic validity. One group (n = 44) was assessed with both instruments, while the DLD was applied in 71 patients. In the NTB (n = 44), only patients with dementia (n = 26) showed a decline in the NTB total score and three subscales. Receiver operating characteristic analysis revealed a diagnostic sensitivity of .67, a specificity of .81, and an area under the curve (AUC) of .767. In the DLD group (n = 71), only those with dementia displayed a decrease in the cognitive and social scale; diagnostic sensitivity and specificity values were low (.61/.63) and the AUC was .704. Neuropsy-chological assessment was sensitive to detect cognitive changes over time. Sensitivity values of both instruments suggest a reassessment at a later time point

Rowe, M.
Will general practitioners be adequately prepared to meet the complexities of enhanced dementia screening for people with learning disabilities and Down syndrome: key considerations.
British Journal of Learning Disabilities, 2014, 44(1), 43-48.
https://doi.org/10.1111/bld.12108
Abstract: This article provides a timely response in regard to the UK's Department of Health's current initiative to financially reward general practitioners (GPs) to prioritise and undertake dementia screening for people with learning disabilities over the age of 50 years and for people with Down syndrome over the age of 40 years. Whilst GPs are becoming increasingly aware of their responsibility to care for the complex needs of people with learning disabilities, the implementation of dementia screening poses a multitude of challenges. Research has continued to suggest how difficult it is to detect the early and often ambiguous signs of dementia for someone who has pre-existing cognitive impairments and may present with atypical symptomology. And it continues to be a difficult process even for those who specialise within this area. However, GPs who choose to opt into this financially incentivised scheme will now be offering dementia screening. This article outlines the main GP aims within the dementia screening process and the difficulties that may be encountered, with specific focus upon (1) offering a dementia screen, (2) obtaining consent, (3) undertaking screening within the most appropriate setting, (4) choosing a dementia screening tool and developing a baseline, and (5) detecting early dementia signs.

Rubenstein, E., Hartley, S., & Bishop, L.
Epidemiology of dementia and Alzheimer disease in individuals with Down syndrome
JAMA Neurology, 2020, 77(2), 262-264. doi:10.1001/jamaneurol.2019.3666
Abstract: We describe prevalence and incidence of dementia and AD in DS in a full Medicaid population of adults with DS in Wisconsin from 2008 through 2018. We assessed Medicaid claims for adults (=21 years) who ever had 2 DS claims over their lifetime (based on International Classification of Diseases, Ninth Revision and Tenth Revision codes) on 2 separate days during Medicaid enrollment. Dementia claims were extracted from codes for any dementia (with AD as a subset) from the Centers for Medicare & Medicaid Services Chronic Conditions Data Warehouse. We required 3 or more years of Medicaid enrollment for adults with DS to ensure validity of dementia claims, therefore, beneficiaries entered the cohort at any point between 2008 and 2015. We categorized age at first and last claims (<40 years, 40-54 years, and =55 years) to account for confounding by age. A total of 2,968 individuals were included, of whom 1,507 (50.8%) were male. The median (interquartile range) age at first claim was 39 (25-48) years. In the category of individuals aged 55 years or older, 490 of 938 had dementia claims (52.2%), 307 of 938 had AD claims (32.7%), and dementia incidence was 102 (95% CI, 87-119) cases per 1000 person-years. Among individuals aged 40 to 54 years, 190 of 1013 had dementia claims (18.8%), and dementia incidence was 49 (95% CI, 44-53) cases per 1000 person-years. The probability of an incident dementia claim was 40% (95% CI, 41%-47%) over 11 years of enrollment for adults with DS who were aged 40-54 years at cohort entry and 67% (95% CI, 60%-74%) for those 55 years and older at cohort entry (Figure). There were no sex differences for dementia among individuals younger than 40 years (prevalence ratio, 1.07 [95% CI, 0.63-1.81]) or among those 55 years and older (prevalence ratio, 0.94 [95% CI, 0.69-1.29]). Dementia prevalence was higher in female individuals than male individuals aged 40 to 54 years (prevalence ratio, 1.23 [95% CI, 1.02-1.50]). Findings from a statewide health system confirm that both dementia and AD in individuals with DS present in claims data at rates similar to those ascertained from clinical samples. The hypothesized causative mechanism and similar eligibility requirements between state Medicaid programs for people with DS likely mean that other state Medicaid systems experience high incidence and prevalence of dementia and AD in individuals with DS. Dementia and AD prevalence and incidence in Medicaid beneficiaries with DS highlight the need to identify prodromal presentations and develop dementia services and supports for adults with DS as they age and continue to rely on Medicaid and Medicaid-funded assisted living or skilled nursing facilities.

Ryan, A., Taggart, L., Truesdale-Kennedy, M. & Slevin, E.
Issues in caregiving for older people with intellectual disabilities and their ageing family carers: a review and commentary
International Journal of Older People Nursing, 2014, Sep, 9(3), 217-226.
doi: 10.1111/opn.12021.
Abstract: In keeping with worldwide demographic changes and an ageing population, people with intellectual disabilities are living longer and all the evidence suggest that this trend will continue. This 'new' population of older people and their carers will pose challenges for health and social care providers. This paper presents a review of the literature on key issues influencing caregiving for older people with intellectual disabilities and their ageing family carers. The review was undertaken using a framework adapted from the NHS Centre for Reviews and Dissemination. Papers were identified through the use of databases including CINAHL, Science Direct, PsychoInfo, Blackwell Synergy, the Cochrane Library and MEDLINE. The key themes which emerged from the literature and which consequently form the basis of this review include: ageing family carers, future planning and support services. In the context of family caregiving, older people with intellectual disabilities represent a unique group insofar as they are unlikely to be married and therefore have no spouse or dependents to care for

them in later life. As a result, parents (usually mothers) have to continue caring for their son or daughter with an intellectual disability as they both grow older, often resulting in a mutually dependent relationship. The caregiving situation is further complicated by poor emergency and future planning and by a lack of appropriate services for this group of individuals. In light of the emergence of a 'new' population of older people with intellectual disabilities, there is an urgent need to develop services and support structures which will enable these individuals and their ageing carers to 'age in place' and when this is no longer possible, to have appropriate alternatives that recognise the duality of their needs as older people and as people with intellectual disabilities.

Ryan, C., MacHale, R., & Hickey, E.
"Forgetting familiar faces": Staff perceptions of dementia in people with intellectual disabiliities
British Journal of Learning Disabilities, 2018, 46(3), 155-162. [On-line version of 29 May 2018]; https://doi.org/10.1111/bld.12233
Abstract: Living with dementia is challenging, but poses unique difficulties for adults with an intellectual disability. The demands of dementia are also challenging for family, carers, and friends. The authors explored the impact of dementia on direct care staff using a focus group methodology. Thematic analysis was used to investigate the staff narratives. There were four key themes that emerged: (a) the difficulty of recognizing symptoms of dementia in people with intellectual disability, (b) the process of diagnosis, (c) the challenge of dementia for the person, (d) the emotional impact of dementia for other people. The authors concluded that the themes identified a number of important potential targets for supporting staff and peers when dementia is present in an adult witih an intellectual disability.

Ryan, J., & Carey, E..
Developing person-centred planning in dementia care.
Learning Disability Practice. 12(5), 24-28. doi:10.7748/ldp2009.06.12.5.24.c7064
Abstract: This article provides an overview of the current literature on person-centered planning, health action planning and associated concepts such as facilitation and advocacy. It explores how flexible approaches to person-centered planning and health action planning were introduced and adapted to meet the individual daily changing needs of Anna, an older person with Down syndrome and dementia.

Ryan, K., Guerin, S., Dodd, P., & McEnvoy, J.
End-of-life care for people with intellectual disabilities: Paid carer perspectives
Journal of Applied Research in Intellectual Disability, 2011, 24(3), 199-207. https://doi.org/10.1111/j.1468-3148.2010.00605.x
Abstract: Little is known of paid carers' perspectives when caring for people with intellectual disabilities at the end-of-life. Sixty four individuals from intellectual disability services took part in 12 focus groups. Interviews were analysed using framework analysis. Participants wanted to provide palliative care and felt the experience enriched practice. However, they were inadequately prepared to meet need and this often led to staff stress. A number of issues appeared to heighten stress: situations when end-of-life care decision making was challenging, when staff felt 'pushed out' by relatives and when staff did not have sufficient support or time to provide care or mourn the loss of service users. The study describes issues which contribute to the development of staff stress when providing palliative care and draws attention to areas where strategies should be developed in order to improve the quality of care provided to people with intellectual disabilities.

Salehi, A., Ashford, J.W., & Mufson, E.J.
The link between Alzheimer's disease and Down syndrome. A historical perspective
Current Alzheimer Research, 2016, 13(1), 2–6.
https://doi.org/10.2174/1567205012999151021102914
Abstract: Approximately 40-80% of persons with Down syndrome (DS) develop Alzheimer's disease (AD)-like dementia by the fifth to sixth decade of life, a much younger age than is typically seen in sporadic AD. The onset of dementia symptoms in DS parallels the development of classic brain neuropathological lesions (i.e., amyloid plaques) similar to that evident in AD. Both disorders appears to have a similar genetic linkage, which is supported by the triplication of the gene that codes for amyloid beta (A4) precursor protein (APP) in persons with DS and an extra copy of the APP gene causes familial AD in persons without DS. Despite an overlap in the genetics of these disorders, the clinical presentation of dementia differs between persons with DS and AD. Whereas `forgetfulness' is a typical symptom of early-phase dementia for persons in the general population, behavioral problems and personality changes are early signs of dementia for persons with DS. There are indications that amyloid burden begins in the frontal cortex before spreading to other brain regions in those with DS-AD, something that is not always the case in sporadic AD suggesting pathological variances between these disorders. These differences beg the question: Are the genetic and neuropathological commonalities found in DS- and AD-related dementia an associated similarity or do these disorders share a common pathogenesis? To address this query, we briefly review the clinical, histopathological, and genetic research supporting a putative link between dementia in DS and AD.

Salem, L.C., & Jørgensen, K.
Demens hos personer med Downs syndrom [Dementia in people with Down syndrome]
Ugeskrift for Læger, 2014, Jun 23, 176(26), V04120217
https://pubmed.ncbi.nlm.nih.gov/25294572/
Abstract: In developed countries the population of elderly people with Down syndrome expands resulting in an increasing incidence of age-related diseases, including dementia. The assessment of dementia in individuals with intellectual disability is often complicated due to large intra-individual variability in cognitive functioning prior to dementia and to lack of standardised measures to detect dementia. Structured observations of symptoms of dementia and assessment techniques tailored for people with intellectual disability are increasingly needed.

Santoro, S.L., Campbell, A., Balasubramanian, A., Haugen, K., Schafer, K., & Mobley, W.
Specialty clinics for adults with Down syndrome: A clinic survey
American Journal of Medical Genetics A, 2021, Mar 17. DOI: 10.1002/ajmg.a.62169
Abstract: Specialty centers improve care for patients with Down syndrome. The cohort of adults with Down syndrome is increasing, but the capacity for specialty centers to meet their medical care needs is unknown. Electronic survey of staff of specialty clinics for adults with Down syndrome was conducted. Review of online clinic list ings, and calculation of the number of adults with Down syndrome were performed. Analysis identified the percent of adults with Down syndrome who could have their medical care needs met in a current specialty clinic. Fourteen specialty clinics report providing care for 4038 adults with Down syndrome. Respondents reported gaps in care including: limitations of existing clinics, need for additional clinics, and knowl edgeable health professionals in Down syndrome. Survey-respondent clinic capacity
would meet needs of 3% of adults with Down syndrome. Twenty-five clinics for adults with Down syndrome were listed online with capacity to care for 6517 adults with Down syndrome meeting the needs of 5% of the population. Additional clinic capacity is needed to meet the needs of adults with Down syndrome. Survey of exis ting clinics provides guidance to create additional clinics, including: must-have team members, current sources of clinic financial support, and gaps in current clinical care.

Santos, F.H., Watchman, K., Janicki, M.P. and the Summit on Intellectual Disability and Dementia.
Highlights from the International Summit on Intellectual Disability and Dementia Implications for Brazil
Dementia & Neuropsychologia, 2018, 12(4), 329-336.
doi:10.1590/1980-57642018dn12-040001
Abstract: In October of 2016, an interdisciplinary group representing North and South American and European countries met in Glasgow, Scotland, to scrutinize universal issues regarding adults with intellectual disability (ID) affected by dementia and to produce recommendations and guidelines for public policy, practice, and further research. The aim of this paper is to apprise relevant outcomes of the Summit targeting Brazilian researchers, clinicians, and nongovernmental organizations in the field of ageing and dementia that are committed to developing the Brazilian national dementia plan. Three core themes were covered by the Summit: i) human rights and personal resources, ii) personalized services and caregiver support, and iii) advocacy and public impact. The exploration of the themes highlighted variations across countries, and revealed consensual views on matters such as international networks, guidance for practices, and advocacy on behalf of both people with ID affected by dementia, and their families. The authors outline the challenges Brazil must confront regarding ageing and dementia and proffer recommendations to address the needs of adults with ID affected by dementia within this scenario; both of which would help in developing the Brazilian national dementia plan.

Sauna-Aho, O., Bjelogrlic-Laakso, N., Siren, A., & Arvio, M.
Signs indicating dementia in Down, Williams and Fragile X syndromes.
Molecular Genetics and Genomic Medicine, 2018, 6(5), 855-860. https://doi.org/10.1002/mgg3.430
Abstract: Intellectual disability (ID) and dementia reflect disturbed cortical function during and after developmental age, respectively. Due to the wide heterogeneity of ID population the decline in cognitive and adaptive skills may be different in distinct genetic subgroups. Using the British Present Psychiatric State-learning Disabilities assessment (PPS-LD) questionnaire the dementia signs were screened in 62, 22 and 44 individuals (> 35 year of age) with Down (DS, OMIM number 190685), Williams (WS, OMIM number, 194050), and Fragile X syndrome (FXS, OMIM number 309550), respectively. The median age of those with FXS (59 years) was higher than of those with DS (50 years) and WS (53 years). Most study participants with DS (80%) and FXS (89%) were or had been moderately or severely intellectually disabled while most participants with WS (73%) were or had been mildly or moderately disabled at adolescent age. The adolescent (premorbid) level of ID did not correlate with the dementia score. The median scores were 11/27, 1/27, and 0/27 in DS, WS, and FXS subgroups, respectively. Dementia that was confirmed by brain imaging, manifested as Alzheimer disease and as moya-moya disease associated vascular dementia in DS and as vascular dementia in WS. This survey suggests that the risk of dementia varies depending on the cause of ID and that the severity of ID in adolescence does not predict the development of dementia at a later age. Consequently, the ID and dementia should be understood as separate clinical entities that need to be taken into account in the health management of intellectually disabled people. This is important for the arrangement of appropriate and timely interventions, which can be expected to delay the need for institutionalization.

Schaap, F.D., Dijkstra, G.J., Finnema, E.J., & Reijneveld, S..A.
The first use of dementia care mapping in the care for older people with intellectual disability: a process analysis according to the RE-AIM framework
Aging & Mental Health, 22(7), 912-919. DOI: 10.1080/13607863.2017.1401582
Abstract: The aging of the population with intellectual disability (ID), with associated conseqences as dementia, creates a need for evidence-based methods to support staff. Dementia Care Mapping (DCM) is perceived to be valuable in dementia care and promising in ID-care. The aim of this study was to evaluate the process of the first use of DCM in ID-care. DCM was used among older people with ID and care-staff in 12 group homes of six organisations. We obtained data on the first use of DCM in ID-care via focus-group discussions and face-to-face interviews with: care-staff (N = 24), managers (N = 10), behavioural specialists (N = 7), DCM-ID mappers (N = 12), and DCM-trainers (N = 2). We used the RE-AIM framework for a thematic process-analysis. All available staff (94%) participated in DCM (reach). Regarding its efficacy, staff considered DCM valuable; it provided them new knowledge and skills. Participants intended to adopt DCM, by continuing and expanding its use in their organisations. DCM was implemented as intended, and strictly monitored and supported by DCM-trainers. As for maintenance, DCM was further tailored to ID-care and a version for individual ID-care settings was developed, both as standards for international use. To sustain the use of DCM in ID-care, a multidisciplinary, interorganisational learning network was established. DCM tailored to ID-care proved to be an appropriate and valuable method to support staff in their work with aging clients, and it allows for further implementation. This is a first step to obtain an evidence-based method in ID-care for older clients.

Schaap, F.D., Dijkstra, G.J., Stewart, R.E., Finnema, E.J., & Reijneveld,J S..A.
Effects of Dementia Care Mapping on well-being and quality of life of older people with intellectual disability: A quasi-experimental study
Journal of Applied Research in Intellectual Disabilities, 2019, July, 32(4), 849-860. https://doi.org/10.1111/jar.12576
Abstract: The ageing of people with intellectual disability, accompanied with consequences like dementia, challenges intellectual disability-care staff and creates a need for supporting methods, with Dementia Care Mapping (DCM) as a promising possibility. This study examined the effect of DCM on the quality of life of older people with intellectual disability. We performed a quasi-experimental study in 23 group homes for older people with intellectual disability in the Netherlands, comparing DCM (n = 113) with care-as-usual (CAU; n = 111). Using three measures, we assessed the staff-reported quality of life of older people with intellectual disability. DCM achieved no significantly better or worse quality of life than CAU. Effect sizes varied from 0.01 to -0.22. Adjustments for covariates and restriction of analyses to people with dementia yielded similar results. The finding that DCM does not increase quality of life of older people with intellectual disability contradicts previous findings and deserves further study.

Schaap, F.D., Finnema, E.J., Dijkstra, G.J., & Reijneveld, M.
What can we learn from dementia care in the care of older people with intellectual disability?
Journal of Intellectual Disability Reserach, 2019, 63(8), 645-646.
Abstract: The ageing of people with intellectual disability (ID) increases rates of dementia, starting earlier and are more prevalent than in the general population. ID-care staff call for methods, knowledge, and skills to support their older residents. Person-centred methods derived from dementia care can fill this gap, but are often used unsystematically, and not adapted to ID-care. Moreover, their effectiveness in ID-care is not yet clear. One person-centred method adapted to ID-care, is Dementia Care Mapping (DCM). The aim of this study is to examine the experiences of care staff with DCM. We assessed this after two applications of DCM in twelve group homes for older people with ID, with a qualitative study (N = 24) and a quantitative study on care-staff (N = 136). Our study showed that DCM provided better understanding of the behaviour of their residents with and without dementia, more reflection and awareness of their own professional behaviour, and new knowledge and (dementia-care) skills. Furthermore, relating the needs and interpretation of the behaviour of residents to the theory of person-centred care provided care-staff a rationale and significance in daily care. Finally, DCM led to more team coordination of care. Authors concluded that evidence from dementia care can improve the quality of care for older people with ID, if adequately embedded in ID-care.

Schäper, S. & Graumann, S.
Alter(n) als wertvolle Lebensphase erleben [Aging and quality of life: challenges and opportunities for people with intellectual disabilities]
Zeitschrift für Gerontologie und Geriatrie, 2021, Oct, 45(7), 630-636. doi:10.1007/s00391-012-0388-1.
Abstract: In the coming years, a growing number of people with an intellectual disability will reach retirement age. In line with the change of paradigms, the leading ideas of participation, inclusion and self-determination have become the principles of the ideological and conceptual framework in social services for people with disabilities. However, in many places convincing concepts and arrangements of support for elderly people with intellectual disabilities are lacking, particularly beyond institutionalized concepts. The research project "Lebensqualität inklusiv(e)" (quality of life included) tries to bridge this gap. On the base of an estimation of the demographic development for this group of people, models of best practice have been documented and evaluated focusing on living conditions and the special requirements for elderly people with intellectual disabilities in order to gather ideas for the development of arrangements of support. The results show that an interdisciplinary cooperation is indispensable

Schlamb, C.D., & Moriconi, C.D.
Betsy: A case study of a client with Down's syndrome and dementia
Advancing Care Excellence for Seniors. 8pp.
https://digitalcommons.wcupa.edu/cgi/viewcontent.cgi?article=1000&context=nurs_facpub
Abstract: This case study is about an aging woman experiencing Down syndrome (DS) and dementia. People with Down syndrome are living longer than ever before. Since the 1980s their life expectancy has doubled and many now live into their 60s, most likely because of advances in medical treatment and improved living conditions. Adults with DS and dementia typically experience several residential relocations during their lifetime and these may be traumatic events for these individuals. This study explores the complex needs of aging clients with intellectual disabilities. Target students for this teaching strategy have completed medical-surgical or geriatric nursing.

Schupf, N., Kapell, D., Lee, J.H., Ottman, R. & Mayeux, R.
Increased risk of Alzheimer's disease in mothers of adults with Down's syndrome
Lancet,1994, Aug 6, 344(8919), 353-356. doi: 10.1016/s0140-6736(94)91398-6.
Abstract: Most adults with Down's syndrome (DS) develop neuropathology characteristic of Alzheimer's disease (AD) by the age of 40. Most of the non-dysjunction events in DS are of maternal origin. We postulated therefore that a shared genetic susceptibility to DS and AD would be associated with an increased frequency of AD among mothers, but not fathers, of individuals with DS. We further hypothesised that the shared susceptibility could involve an accelerated ageing process, leading to the birth of a child with DS to a relatively

young mother and to an increased risk of dementia in the mother and her relatives. Families of 96 adults with DS and of 80 adults with other forms of mental retardation were ascertained through the New York State Developmental Disabilities services network. A semi-structured interview was used to obtain information on the presence or absence of non-stroke-related dementia and other disorders in parents. There was an increase in risk of dementia among mothers of DS probands compared with control mothers (risk ratio 2.6 [95% CI 0.9-7.3]). The risk of dementia among mothers who were 35 or younger when their DS children were born was 5 times that of control mothers (4.9 [1.6-15.4]). There was no increase in risk of dementia among mothers who were older (> 35 years) at the proband's birth (0.8 [0.2-3.4]). There was no difference in risk of dementia between fathers of DS cases and fathers of controls (1.2 [0.4-3.9]) and no discernible influence of age on this risk. Familial aggregation of dementia among mothers of adults with DS supports the hypothesis of a shared genetic susceptibility to DS and AD.

Schupf, N., Kapell, D., Nightingale, B, Rodriguez, A., Tycko, B., & Mayeux, R.
Earlier onset of Alzheimer's disease in men with Down syndrome.
Neurology, 1998, 50(4), 991-995. https://doi.org/10.1212/WNL.50.4.991
Abstract: Virtually all individuals with Down syndrome (DS) have neuropathologic changes characteristic of Alzheimer's disease (AD) beginning at 40 years of age. Few studies have examined factors that influence age at onset of AD in DS. We investigated whether sex differences in age at onset and risk of AD among adults with DS are similar to those observed in the general population and whether the effect of sex on risk of AD is modified by apolipoprotein E(*APOE*) genotype. A community-based sample of 111 adults with cytogenetically confirmed DS (34 to 71 years of age) was ascertained through the New York State Developmental Disabilities system. A semi-structured interview with caregivers and review of medical records was used to ascertain the presence or absence of AD. *APOE* genotyping was carried out without knowledge of the subject's medical history or clinical diagnosis. Both male gender and the presence of an *APOE* ?4 allele were associated with an earlier onset of AD. Compared with women, men with DS were three times as likely to develop AD. Compared with those with the *APOE* 3/3 genotype, adults with DS with the 3/4 or 4/4 genotypes were four times as likely to develop AD. No individual with an *APOE* ?2 allele developed AD. No evidence of interaction of sex and *APOE* genotype was found in risk of AD. The higher risk of AD in men may be related to differences in hormonal function between men and women with DS that are distinct from those in the general population.

Schupf, N., Winsten, S., Patel, B., Pang, D., Ferin, M., Zigman, W.B., Silverman, W., & Mayeux R.
Bioavailable estradiol and age at onset of Alzheimer's disease in postmenopausal women with Down syndrome.
Neuroscience Letter, 2006, Oct 9, 406(3), 298-302.
Abstract: Several lines of evidence suggest that loss of estrogen after menopause may play a role in the cognitive declines associated with Alzheimer's disease (AD). Women with Down syndrome (DS) experience early onset of both menopause and AD. This timing provides a model to examine the influence of endogenous estrogen deficiency on risk of AD. We hypothesized that low serum levels of bioavailable estradiol (E2) would be associated with increased risk of AD. One hundred and nineteen postmenopausal women with DS, 42-59 years of age, were ascertained through the New York State developmental disability service system and followed at 18-month intervals. Information from cognitive assessments, caregiver interviews, medical record review and neurological examination was used to establish the diagnosis of dementia. Women with DS who developed AD had lower levels of bioavailable E2, lower levels of total estradiol, higher levels of sex-hormone binding globulin, and lower levels of dehydroepiandrosterone sulfate at baseline than women who remained dementia free over the course of follow-up. Women who had low levels of bioavailable E2 at baseline were four times as likely to develop AD (HR=4.1, 95% CI: 1.2-13.9) and developed AD, on average, 3 years earlier, than those with high levels of bioavailable E2, after adjustment for age, level of mental retardation, ethnicity, body mass index, history of hypothyroidism or depression and the presence of the apolipoprotein varepsilon4 allele. Our findings support the hypothesis that reductions in estrogen following menopause can contribute to the cascade of pathological processes leading to AD.

Schupf, N., Pang, D., Patel, B.N., Silverman, W., Schubert, R., Lai, F., Kline, J.K., Stern, Y., Ferin, M., Tycko, B., & Mayeux, R.
Onset of dementia is associated with age at menopause in women with Down's syndrome.
Annuals of Neurology, 2003, 54(4), 433-438. https://doi.org/10.1002/ana.10677
Abstract: Women with Down's syndrome experience early onset of both menopause and Alzheimer's disease. This timing provides an opportunity to examine the influence of endogenous estrogen deficiency, indicated by age at menopause, on risk of Alzheimer's disease. A community-based sample of 163 postmenopausal women with Down's syndrome, 40 to 60 years of age, was ascertained through the New York State Developmental Disability service system. Information from cognitive assessments, medical record review, neurological evaluation, and caregiver interviews was used to establish ages for onset of menopause and dementia. We used survival and multivariate regression analyses to determine the relation of age at menopause to age at onset of Alzheimer's disease, adjusting for age, level of mental retardation, body mass index, and history of hypothyroidism or depression. Women with early onset of menopause (46 years or younger) had earlier onset and increased risk of Alzheimer's disease (AD) compared with women with onset of menopause after 46 years (rate ratio, 2.7; 95% confidence interval [CI], 1.2-5.9). Demented women had higher mean serum sex hormone binding globulin levels than nondemented women (86.4 vs 56.6 nmol/L, p = 0.02), but similar levels of total estradiol, suggesting that bioavailable estradiol, rather than total estradiol, is associated with dementia. Our findings support the hypothesis that reductions in estrogens after menopause contribute to the cascade of pathological processes leading to AD.

Schweber, M.S.
Alzheimer's disease and Down syndrome
Progress in Clinical and Biological Research, 1989, 317, 247-267.
https://pubmed.ncbi.nlm.nih.gov/2532369/
Abstract: This report contains a summary of an extensive survey of autopsy data for persons with intellectual disability. Among adults with Down syndrome (DS), the brain neuropathology of AD was universal in those age 37 and over; claimed exceptions were indefensible. The behavioral evaluations of the DS adults, however, could be classified into three divisions: 1. "quiescent" (neither seizures nor dementia, 2). "partial" (seizures but no dementia), and 3). "active" (dementia +/- seizures). Thus, it is reasonable to argue that all persons with DS develop AD itself upon aging. However, DS cannot be used uncritically as an AD model since no increased incidence of active AD was found in DS with aging beyond the critical threshold age (mid-30's). Improved accurate quantification of Southern blots produced 100% accuracy in decoding blind samples of DS and non-DS samples. Using this system, DNA levels similar to those of DS have been demonstrated for all categories of AD at a small subsection of chromosome 21 near to, or within the DS DNA location on chromosome 21. Increased amounts of a complete, structural gene sequence were not found (or expected). The results provide evidence for a unitary hypothesis for DS and all forms of AD.

Scottish Down's Syndrome Association
What is dementia? - A booklet about dementia for adults who have a learning disability.
14pp
Edinburgh: Scottish Down's Syndrome Association [158-160 Balgreen Road, Edinburgh, Scotland EH11 3AU; e/m: info@sdsa.org.uk; www.sdsa.org.uk] [n.d.] [Source: http://www.rrtcadd.org/TA/Dementia_Care/Resources/Info.html]
Abstract: Written for the Scottish Down's Syndrome Association by Diana Kerr and Mo Innes this A4 size booklet is designed to explain dementia and its nuances to persons with intellectual disabilities (termed "learning disabilities in Scotland). Using drawings and easy language this booklet covers many of the symptoms and behaviors classically associated with Alzheimer's disease.

Service, K.P.
Considerations in care for individuals with intellectual disability with advanced dementia
Journal of Gerontological Social Work, 2002, 38, 213-224.
Abstract: A number of physical, psychosocial, or ethical issues related to the care of the individual with advanced dementia are reviewed and related to individuals with intellectual disabilities. The sources used include the published literature and illustrations drawn from personal observations. The author notes that through anticipation and early planning, advanced directives and service planning (which looks to adaptation of services and other care management interventions), can effectively impact care at the end. Areas that need to be addressed include technical information, including a review of and, as appropriate, adaptation of general advanced dementia resources, relief, rest, support, reassurance, receipt

of on-going information, participation in planning, a sense of humor, and appreciation. Also noted, are the differences experienced because of the presence of paid staff as carers and residence outside of the family home. It is concluded that, although the goals of quality care is the same for all people with advanced dementia, the process by which to reach these goals often needs further consideration and adaptation for people with intellectual disabilities.

Service, K..P., & Clifford, C.J.
What do I really need? Assessment of caregiver supports for people with intellectual and developmental disability and dementia.
AAIC 2020 Conference (Amsterdam, NL - virtual), Poster presentation, July 30. 2020. Alzheimer's & Dementia, 16(S7), First published: 07 December 2020. https://doi.org/10.1002/alz.047106
Abstract: The increased needs of people with an intellectual and developmental disability (IDD) and a dementia related disorder can strain caregivers and existing community support systems. The project team, comprised of IDD and Aging experts and funded, in part, through a federal grant, conducted a needs assessment on the awareness and use of typical community-based resources such as senior centers. The assessment consisted of both telephone interviews and home visits with group home, shared living, and family caregivers. Nurse Practitioner (NP) conducted a total of 95 interviews with 54 site visits, and the evaluator completed 40 interviews with caregivers of people with IDD and dementia diagnosis. Analysis included both qualitative and quantitative data. Caregivers were asked about the following: functional and health status of person with IDD since dementia diagnosis, receipt of dementia specific caregiving training, care confidence levels, perceived barriers to care, and access to community-based aging resources. Caregiver's most frequent concerns included lack of suitable day programming, planning for the person's future, and caregiver burnout and stress. 78% reported feeling confident providing care currently and 68% were confident about providing care in the future. Most caregivers are aware of local community resources such as senior centers, Alzheimer's Association Counseling and an on-line training series on aging, but rarely used the resources. Authors note that caregivers generally relied on support from provider agencies indicating a need for increased collaboration across the IDD and Aging human service support systems. Trainings, delivered to both aging advocates and caregivers of people with IDD, and designed to improve communication and collaboration focused on dementia capable care, state systems, and available community resources. In addition, a series of web-based resources were developed with a focus on IDD and dementia. Results of the assessment will continue to guide resource and training development to improve collaboration and support the relationship between the Aging and IDD communities.

📖 **Service, K.P., Lavoie, D. Herlihy, J.E.**
Coping with losses, death and grieving
In M.P. Janicki & A.J. Dalton (eds.), Dementia, Aging, and Intellectual Disabilities.
pp. 330-351
Philadelphia: Brunner-Mazel (1999)
Abstract: This book chapter uses a composite case to demonstrate strategies to address the issues related to losses and death for people with intellectual disability and the diagnosis of dementia and for their families and staff. Dealing with the diagnosis and the changes are explained in the framework of the stages of death and dying as developed by Kubler-Ross. The responses to the losses of dementia which are manifested by affected individuals and members of their personal networks are reflective of a number of factors. The dilemma related to personal value systems, professional roles, and philosophies of care is explored in the context of ethical concerns. The impact of program considerations such as rules, regulations, policies, and economics is examined. Bereavement work for peers and housemates can be further developed for carers, family, and staff. Recommendations for research and interventions for public policy are given.

Service, K., Watchman, K., Hogan, M., Janicki, M.P., Cadovius, N., & Beránková, A.
Dying well with an intellectual disability and dementia
Journal of Dementia Care, 2017, 25(4), 25-31.
https://www.researchgate.net/publication/318723617_Dying_well_with_an_intellectual_disability_and_dementia
Abstract: An international summit on intellectual disability and dementia identified three areas where the added complexity of advanced dementia warrants particular attention around end-of-life services in people with an intellectual disability. The three areas were: (a) ascertainment of advanced stage of dementia, (b) place of care, and (c) active support. The authors discuss each of these three issues and note the particular challenges that arise when someone with dementia also has an intellectual disability. The summit proffered a series of recommendations that included ongoing exchange of experiences and skills across professions, development of tools and scales that facilitate understanding of the progression of dementia, and more equitable access to palliative care and hospice services with increased and timely referral

Sheehan, R., Ali, A., & Hassiotis, A.
Dementia in intellectual disability
Current Opinion in Psychiatry, 2014, March, 27(2), 143-148
Abstract: Dementia is emerging as a significant condition in the population with intellectual disability. This review is aimed at clinicians working in the field. We revisit what is known on the subject and expand on this with results from recent research. The emphasis of this review is on the clinical research rather than laboratory or molecular research. Research has encompassed all aspects of dementia in intellectual disability, from epidemiology, assessment and diagnosis, through to management. There remains a lack of evidence concerning both pharmacological and nonpharmacological treatment of dementia in people with intellectual disability. Recent research has tended to focus on dementia in Down syndrome. More research is necessary in order to translate improvements in the understanding of the neuropathology of intellectual disability and dementia into effective treatments. There is also a need to investigate the optimum environment in which to provide holistic care for individuals affected.

Sheehan, R., Sinai, A., Bass, N., Blatchford, P., Bohnen, I,, Bonell, S., Courtenay, K., Hassiotis, A., Markar, T., McCarthy, J., Mukherji, K., Naeem, A,. Paschos, D., Perez-Achiaga, N., Sharma, V., Thomas, D., Walker, Z., Strydom, A.
Dementia diagnostic criteria in Down syndrome.
International Journal of Geriatric Psychiatry, 2015, 30(8), 857-863. doi: 10.1002/gps.4228.
Abstract: Dementia is a common clinical presentation among older adults with Down syndrome. The presentation of dementia in Down syndrome differs compared with typical Alzheimer's disease. The performance of manualised dementia criteria in the International Classification of Diseases (ICD)-10 and Diagnostic and Statistical Manual of Mental Disorders-IV-Text Revision (DSM-IV-TR) is uncertain in this population. We aimed to determine the concurrent validity and reliability of clinicians' diagnoses of dementia against ICD-10 and DSM-IV-TR diagnoses. Validity of clinical diagnoses were also explored by establishing the stability of diagnoses over time. The authors used clinical data from memory assessments of 85 people with Down syndrome, of whom 64 (75.3%) had a diagnosis of dementia. The cases of dementia were presented to expert raters who rated the case as dementia or no dementia using ICD-10 and DSM-IV-TR criteria and their own clinical judgement. The authors found that clinician's judgement corresponded best with clinically diagnosed cases of dementia, identifying 84.4% cases of clinically diagnosed dementia at the time of diagnosis. ICD-10 criteria identified 70.3% cases, and DSM-IV-TR criteria identified 56.3% cases at the time of clinically diagnosed dementia. Over time, the proportion of cases meeting ICD-10 or DSM-IV-TR diagnoses increased, suggesting that experienced clinicians used their clinical knowledge of dementia presentation in Down syndrome to diagnose the disorder at an earlier stage than would have been possible had they relied on the classic description contained in the diagnostic systems. The authors concluded that clinical diagnosis of dementia in Down syndrome is valid and reliable and can be used as the standard against which new criteria such as the DSM-5 are measured.

Sheehan. R., Sinai, A., Bass, N., Blatchford, P., Bohnen, I,, Bonell, S., Courtenay, K., Hassiotis, A., Markar, T., McCarthy, J., Mukherji, K., Naeem, A,. Paschos, D., Perez-Achiaga, N., Sharma, V., Thomas, D., Walker, Z., Strydom, A.
Dementia diagnostic criteria in Down syndrome.
International Journal of Geriatric Psychiatry, 2014, Aug, 30(8), 857-863. doi: 10.1002/gps.4228. Epub 2014 Nov 3.
Abstract: Dementia is a common clinical presentation among older adults with Down syndrome. The presentation of dementia in Down syndrome differs compared with typical Alzheimer's disease. The performance of manualised dementia criteria in the International Classification of Diseases (ICD)-10 and Diagnostic and Statistical Manual of Mental Disorders-IV-Text Revision (DSM-IV-TR) is uncertain in this population. We aimed to determine the concurrent validity and reliability of clinicians' diagnoses of dementia against ICD-10 and DSM-IV-

TR diagnoses. Validity of clinical diagnoses were also explored by establishing the stability of diagnoses over time. The authors used clinical data from memory assessments of 85 people with Down syndrome, of whom 64 (75.3%) had a diagnosis of dementia. The cases of dementia were presented to expert raters who rated the case as dementia or no dementia using ICD-10 and DSM-IV-TR criteria and their own clinical judgement. The authors found that clinician's judgement corresponded best with clinically diagnosed cases of dementia, identifying 84.4% cases of clinically diagnosed dementia at the time of diagnosis. ICD-10 criteria identified 70.3% cases, and DSM-IV-TR criteria identified 56.3% cases at the time of clinically diagnosed dementia. Over time, the proportion of cases meeting ICD-10 or DSM-IV-TR diagnoses increased, suggesting that experienced clinicians used their clinical knowledge of dementia presentation in Down syndrome to diagnose the disorder at an earlier stage than would have been possible had they relied on the classic description contained in the diagnostic systems. The authors concluded that clinical diagnosis of dementia in Down syndrome is valid and reliable and can be used as the standard against which new criteria such as the DSM-5 are measured.

Sheth, A.J.
Intellectual disability and dementia: perspectives on environmental influences.
Quality in Ageing and Older Adults, 2019, 20(4), 179-189.
https://doi.org/10.1108/QAOA-11-2018-0060.
Abstract: The purpose of this paper was to improve understanding of environmental influences on participation in routine and familiar activities for people with intellectual disability and dementia from first-person and caregiver perspectives. The methodology involved four adults with intellectual disability and dementia participating in 2 nominal group technique sessions and 12 family and staff caregivers participating in 5 standard focus groups. Transcripts were analyzed utilizing thematic analysis centering the findings from nominal group technique sessions and an ecological systems lens. The findings revealed that participants with intellectual disability and dementia identified six important themes: activity access, caregiver assistance, social interactions, responsibilities, privacy, and health and wellness. Their perspectives focused primarily at an immediate environment level, while caregiver input added additional understandings from broader ecological systems levels. This study provides a beginning point to establishing a framework for creating supports and addressing barriers to participation for adults with intellectual disability and dementia based on direct input from potential service consumers and their caregivers. People with intellectual disabilities and dementia provide valuable insights into their experiences through engagement in accessible research.

Sheth, A.J., Kramer, J.M., Magasi, S., Heller, T., Nishida, A., & Hammel, J.
"It's not the same without you:" Exploring the experience and perception of transition for people with intellectual disabilities and dementia
British Journal of Learning Disabilities, 2021 (Sep), 49(3), 365-372.
https://doi.org/10.1111/bld.12412
Abstract: For people with intellectual disabilities and dementia, transitions are likely to become increasingly common as they age. While transitions experienced by people with intellectual disabilities in young adulthood are frequently studied, less is known about transitions in older adults, including residential, vocational and leisure changes. This article aims to explore the experiences of transition from the perspectives of people with intellectual disabilities and dementia, including the impact on their daily lives. Three women with intellectual disabilities and dementia living in residential settings participated in participant observations and informal interviews across a variety of environments and activities. Field notes and interview transcripts underwent a thematic analysis focusing on transitions. Participants experienced the impact of transitions in their residential placements, day programming, leisure activities and relationships. Themes related to their experiences of transitions included making sense of transitions, utilising peer care networks for support and tackling the looming threat of loss and transition. Peer care networks and friendships are crucial in supporting people during and after transitions. Receiving effective supports to maintain relationships, roles and activities, even in seemingly minor ways, is an important right for people with intellectual disabilities and dementia, particularly as care needs increase.

Shirai, Y., Bishop, K., & Kushner, M.
National dementia capable care training: A model implementation and evaluation
Intellectual and Developmental Disabilities, 2021, 59 (5), 422–435.
https://doi.org/10.1352/1934-9556-59.5.422
Abstract: With a growing need for specialized training for direct caregivers and support staff of persons with intellectual and developmental disabilities (IDD) affected by dementia, the National Task Group on Developmental Disabilities and Dementia Practices (NTG) developed a comprehensive evidence-informed Dementia Capable Care Training (DCCT). To overcome the challenge of the training length and cost, and to extend its dissemination, the Sonoran Center developed a shorter version of the NTG-DCCT while retaining its core components, and implemented it in seven cities in the U. S. Southwest (N = 368). The pre- and post-training evaluation (n =260) demonstrated that the short version of the NTG-DCCT is effective in significantly improving participants' knowledge and/or confidence in dementia capable care. The follow-up semi-structured interviews of participants (n = 7) provide some insights.

Shooshtari, S., Stoesz, B.M., Udell, L., Fenez, L., Dik, N., Burchill, C., Sachs, E., Menec, V.
Aging with intellectual and developmental disabilities and dementia in Manitoba
Advances in Mental Health and Intellectual Disabilities, 11(4), 134-144.
https://doi.org/10.1108/AMHID-03-2017-0007
Abstract: Information on the risk of dementia in aging persons with intellectual and/or developmental disability(IDD) in Manitoba, Canada is lacking. The purpose of this paper is to estimate dementia prevalence in adults with IDD. Anonymized population-level health and non-health administrative data (1979-2012) contained in the Population Health Research Data Repository of the Manitoba Centre for Health Policy (MCHP) were linked to identify adults with IDD, and estimate the prevalence of dementia based on the presence of ICD codes. Prevalence of dementia was estimated for persons aged 18-55 years and 55+ years, and was reported by sex, type of residence, region of residence, neighbourhood income quintiles, and IDD diagnostic category.Of the 8,655 adults with IDD identified, 8.1 per cent had an indication of dementia in their medical records; an estimate three times greater than that found for those without IDD (2.6 per cent). More than 17 per cent of Manitobans with IDD aged 55+ years had an indication of dementia, which was nearly twice the rate reported previously. Of those with IDD and dementia, 34.7 per cent lived in long-term care facilities. Health and social support services are typically available to individuals with dementia aged 65+ years; thus, younger adults with IDD and dementia may not be eligible for those supports. To promote equity in health and access to care, dementia screening and increased supports for aging individuals with IDD are recommended.

Shultz, J.M., Aman, M.G., & Rojahn, J..
Psychometric evaluation of a measure of cognitive decline in elderly people with mental retardation.
Research in Developmental Disabilities, 1998, 19, 63-71.
Abstract: Forty elderly persons with mental retardation were assessed by their care providers on a modified version of the Short Informant Questionnaire on Cognitive Decline in The Elderly (IQCODE) an instrument designed to quantify cognitive decline in elderly people in the general population. They were also assessed for IQ, aberrant behavior, and current mental status; test-retest and interrater reliability were evaluated as well. Internal consistency, as assessed by coefficient alpha, was moderately high (alpha = .86). Test-retest reliability was mediocre and interrater reliability levels did not reach statistical significance. The Short IQCODE was not correlated with a variety of demographic features or with behavior ratings, showing evidence of divergent validity. However, the Short IQCODE was only weakly (nonsignificantly) correlated with a measure of current mental status, which challenges its concurrent validity. The Short IQCODE probably needs to be modified further for satisfactory psychometric performance in people with mental retardation. However, some features of this study may have resulted in suboptimal estimates of the Short IQCODE's psychometric characteristics.

Sigal, M.J,, & Levine, N.
Down's syndrome and Alzheimer's disease.
Journal of the Canadian Dental Association, 1993, 59(10), 823-5, 829.
https://pubmed.ncbi.nlm.nih.gov/8221282/
Abstract: Individuals with Down's syndrome (DS) who live to be 40 years of age will demonstrate neuropathological changes that are consistent with Alzheimer's disease (AD). Due to modern medical intervention, we are now observing an aging DS population. Middle-aged Down's syndrome adults are actually considered to be "very old," and it is not uncommon to observe a progressive loss of cognitive function and a decline in the ability to perform daily tasks consistent with that seen in Alzheimer's disease. At this stage, the DS individual will not be able to perform daily preventive dental care and may be unable to cooperate for professional dental care. Clinicians who care for DS adults must be aware of this problem when preparing their dental treatment plans, which must emphasize

preventive care prior to the onset of dementia and the maintenance of that program during their patients' cognitive decline. In the latter stages of AD, it may be necessary to extract all the remaining teeth due to the inability of the individual or care giver to provide adequate oral hygiene to prevent dental caries or periodontal disease.

Simard, M., & van Reekum, R.
Dementia with Lewy bodies in Down's syndrome.
International Journal of Geriatric Psychiatry, 2001, Mar;16(3), 311-20.
Abstract: The association between Down's syndrome (DS) and Alzheimer's disease is well established. This paper presents a review of the literature, suggesting a possible association between DS and the more recently recognized dementia with Lewy bodies (DLB). Patients with DLB frequently present with changes in affect and behavior, and in particular with psychotic symptoms. The literature suggests a possible role for atypical neuroleptics in the management of psychosis in DLB.

Sinai, A., Mokrysz, C., Bernal, J., Bohnen, I., Bonell, S., Courtney, K., Dodd, K., ... Strydom, A.
Predictors of age of diagnosis and survival of Alzheimer's disease in Down syndrome.
Journal of Alzheimer's Disease, 2018, 61(2), 717-728.
doi: 10.3233/JAD-170624.
Abstract: People with Down syndrome (DS) are an ultra-high risk population for Alzheimer's disease (AD). Understanding the factors associated with age of onset and survival in this population could highlight factors associated with modulation of the amyloid cascade. This study aimed to establish the typical age at diagnosis and survival associated with AD in DS and the risk factors associated with these. Data was obtained from the Aging with Down Syndrome and Intellectual Disabilities (ADSID) research database, consisting of data extracted from clinical records of patients seen by Community Intellectual Disability Services (CIDS) in England. Survival times when considering different risk factors were calculated. **The mean age of diagnosis was 55.80 years**, *SD 6.29. Median survival time after diagnosis was 3.78 years, and median age at death was approximately 60 years.* Survival time was associated with age of diagnosis, severity of intellectual disability, living status, anti-dementia medication status, and history of epilepsy. Age at diagnosis and treatment status remained predictive of survival time following adjustment. This study provides the best estimate of survival in dementia within the DS population to date, and is in keeping with previous estimates from smaller studies in the DS population. This study provides important estimates and insights into possible predictors of survival and age of diagnosis of AD in adults with DS, which will inform selection of participants for treatment trials in the future.

Silverman, W., Krinsky-McHale, S.J., Lai, F., Rosas, H.D., Hom, C., Doran, E., Pulsifer, M., Lott. I., Schupf, N, and Alzheimer's Disease in Down Syndrome (ADDS) Consortium
Evaluation of the National Task Group-Early Detection Screen for Dementia: Sensitivity to 'mild cognitive impairment' in adults with Down syndrome
Journal of Applied Research in Intellectual Disabilities, 2020, Dec 13.
doi:10.1111/jar.12849.
Abstract: The accuracy of the National Task Group-Early Detection Screen for Dementia (NTG-EDSD) was evaluated in a sample of 185 adults with Down syndrome (DS), emphasizing 'mild cognitive impairment (MCI-DS)'. Knowledgeable informants were interviewed with the NTG-EDSD, and findings were compared to an independent dementia status rating based on consensus review of detailed assessments of cognition, functional abilities and health status (including physician examination). Results indicated that sections of the NTG-EDSD were sensitive to MCI-DS, with one or more concerns within the 'Memory' or 'Language and Communication' domains being most informative. The NTG-EDSD is a useful tool for evaluating dementia status, including MCI-DS. However, estimates of sensitivity and specificity, even for detecting frank dementia, indicated that NTG-EDSD findings need to be supplemented by additional sources of relevant information to achieve an acceptable level of diagnostic/screening accuracy.

Silverman, W., Schupf, N., Zigman, W., Devenny, D., Miezejeski, C., Schubert, R., & Ryan, R.
Dementia in adults with mental retardation: Assessment at a single point in time.
American Journal of Mental Retardation, 2004, 109, 111-125.
https://doi.org/10.1352/0895-8017(2004)109<111:DIAWMR>2.0.CO;2
Abstract: Dementia status of 273 adults with mental retardation was rated based upon two extensive evaluations conducted 18 months apart. Overall, 184 individuals did not have dementia, 33 had possible or definite dementia, and 66 had findings suggesting uncertain or questionable status. These ratings were compared to binary classifications (dementia vs. no dementia) generated from the Dementia Questionnaire for Persons With Mental Retardation (Evenhuis, 1995) and the IBR Mental Status Examination (Wisniewski & Hill, 1985). When performance was referenced to IQs (established earlier in adulthood), quantitative criteria effectively distinguished between individuals with and without dementia based upon assessment at a single point in time. Findings suggest that procedures of this type could soon contribute to more accurate and rapid diagnoses of dementia.

Smith, D., & Chicoine, B.
Difficulties of diagnosing and managing dementia in people with Down syndrome
British Journal of Psychiatry, 2018, Nov, 213(5), 668-669.
doi:10.1192/bjp.2018.199.
Abstract: In our experience as the directors of Down syndrome clinics for adults, the big issue is how the diagnosis of dementia Is made. Clinicians tend to easily apply the diagnosis or Alzheimer's dementia without looking at all of the potential causes of pseudodementia in this population. They often assume that loss of ability is due to dementia because of a study published in 1985 that showed plaques and tangles in the brain tissue of all people with Down syndrome over the age of 35. Wisniewskl & Rabe subsequently wrote that there was a discrepancy between neuropathology and the occurrence of dementia in people with Down syndrome. Just as the population of typically developed older adults the diagnosis of Alzheimer's dementia in people with Down syndrome should be made only after evaluation for causes of pseudodementia.

Social Care Institute for Excellence
Learning disabilities and dementia
SCIE, Isosceles Head Office, One High Street, Egham TW20 9HJ, (2020).
https://www.scie.org.uk/dementia/living-with-dementia/learning-disabilities/
Abstract: Webpage informational matter on dementia and intellectual disability, recognising early changes, diagnostic and assessment issues, environmental design, and health care.

Soliman, A, & Hawkins, D.
The link between Down's syndrome and Alzheimer's disease: 1.
British Journal of Nursing, 1998, Jul 9-22, 7(13), 779-784.
Abstract: This article, the first of two parts, considers the link between Down's syndrome and Alzheimer's disease and how this link has been a significant factor with regards to research into the aetiology of Alzheimer's disease. It describes some of the suggested causes of Alzheimer's disease in people with Down's syndrome. The diagnosis, signs and symptoms of Alzheimer's disease are briefly discussed. The second article concludes with the implications of Alzheimer's disease in people with Down's syndrome for family careers, services and nurses.

Soliman, A., & Hawkins, D.
The link between Down's syndrome and Alzheimer's disease: 2.
British Journal of Nursing, 1998, Jul 23-Aug 12, 7(14), 847-850.
Abstract: In this article, the second of two parts, the needs of family and professional carers of people with Down's syndrome and Alzheimer's disease are examined. Substantial numbers of people with Down's syndrome survive to the age of 50 and beyond and so work still needs to be done on finding solutions to the problems faced by this client group and its carers. As well as the difficulties faced by any family carer of a person with dementia, those caring for someone with Down's syndrome and Alzheimer's disease may also have to deal with additional worries and problems. Consideration is given to service provision and the implications for nursing. A case study will illustrate some of the points made.

Starkey, H., Bevns, S., & Bonell, S.
The role of prospective screening in the diagnosis of dementia in people with Down's syndrome
Advances in Mental Health and Intellectual Disabilities, 2014, 8(5), 283-291.
https://doi.org/10.1108/AMHID-12-2013-0067
Abstract: People with Down's syndrome are at increased risk of developing early onset Alzheimer's disease. It has been recommended that all adults with Down's syndrome receive baseline neuropsychological testing for dementia. In certain areas prospective screening of people with Down's syndrome takes place to ensure the early diagnosis of the condition. However, little has been published on the value of this type of screening. The purpose of this paper is to report on a prospective screening programme and asks whether the programme is effective

in identifying dementia-related changes in people with Down's syndrome and whether the current screening intervals are appropriate. All adults with Down's syndrome in Plymouth (UK) are identified and offered a comprehensive test battery at baseline at the age of 20 and then have testing biennially from 40 to 50 and annually after 50. All individuals diagnosed with dementia between 2001 and 2013 were identified and their case notes examined. The symptoms at the time of diagnosis were identified and whether these symptoms had been identified through the screening programme or by other routes were recorded. Prevalence data and age at diagnosis were also recorded. In total, 26 people were diagnosed with dementia during the study period. Of these, the diagnosis of dementia followed concerns being identified during the routine screening programme in 54 per cent of cases. In the younger age group (age 40-49) 63 per cent of people were identified through the screening programme. At the time of diagnosis a mean of 5.5 areas of concern were in evidence. This paper adds to the growing evidence base around the value of prospective dementia screening in people with Down's syndrome. It is also one of a few studies exploring the frequency of screening. Additionally, it adds further data about prevalence of dementia in people with Down's syndrome.

Startin, C.M., Hamburg, S., Hithersay, R., Al-Janabi, T., Mok, K.Y., Hardy, J. LonDownS Consortiu, & Strydom, A.
Cognitive markers of preclinical and prodromal Alzheimer's disease in Down syndrome.
Alzheimer's & Dementia, 2019, Feb, 15(2), 245-257.
doi: 10.1016/j.jalz.2018.08.009. Epub 2018 Nov 28.
Abstract: Down syndrome (DS) is associated with an almost universal development of Alzheimer's disease. Individuals with DS are therefore an important population for randomized controlled trials to prevent or delay cognitive decline, though it is essential to understand the time course of early cognitive changes. We conducted the largest cognitive study to date with 312 adults with DS to assess age-related and Alzheimer's disease-related cognitive changes during progression from preclinical to prodromal dementia, and prodromal to clinical dementia. Changes in memory and attention measures were most sensitive to early decline. Resulting sample size calculations for randomized controlled trials to detect significant treatment effects to delay decline were modest. Our findings address uncertainties around the development of randomized controlled trials to delay cognitive decline in DS. Such trials are essential to reduce the high burden of dementia in people with DS and could serve as proof-of-principle trials for some drug targets.

Startin, C.M., Lowe, B., Hamburg, S., Hithersay, R., Strydom, A., & LonDowns Consortium.
Validating the Cognitive Scale for Down Syndrome (CS-DS) to detect longitudinal cognitive decline in adults with Down syndrome.
Frontiers in Psychiatry, 2019, Apr 16,10, 158. doi: 10.3389/fpsyt.2019.00158. eCollection 2019.
Abstract: Down syndrome (DS) is associated with intellectual disability and an ultra-high risk of developing dementia. Informant ratings are invaluable to assess abilities and related changes in adults with DS, particularly for those with more severe intellectual disabilities and/or cognitive decline. We previously developed the informant rated Cognitive Scale for Down Syndrome (CS-DS) to measure everyday cognitive abilities across memory, executive function, and language domains in adults with DS, finding CS-DS scores are a valid measure of general abilities, and are significantly lower for those with noticeable cognitive decline compared to those without decline. To further test the validity of the CS-DS in detecting changes associated with cognitive decline we collected longitudinal data across two time points, approximately 1.5-2 years apart, for 48 adults with DS aged 36 years and over. CS-DS total scores (78.83 ± 23.85 vs. 73.83 ± 25.35, p = 0.042) and executive function scores (46.40 ± 13.59 vs. 43.54 ± 13.60, p = 0.048) significantly decreased between the two time points, with scores in the memory domain trending towards a significant decrease (22.19 ± 8.03 vs. 20.81 ± 8.63, p = 0.064). Adults with noticeable cognitive decline at follow-up showed a trend to significantly greater change in total scores (7.81 ± 16.41 vs. 3.59 ± 16.79, p = 0.067) and significantly greater change in executive function scores (5.13 ± 9.22 vs. 1.72 ± 9.97, p = 0.028) compared to those without decline. Change in total scores showed significant correlations with change in scores from other informant measures of everyday adaptive abilities and symptoms associated with dementia, and participant assessment of general cognitive abilities (all p < 0.005), while change in memory scores (R 2 = 0.28, p = 0.001) better predicted change in participant cognitive assessment scores than change in executive function (R 2 = 0.15, p = 0.016) or language (R 2 = 0.15, p = 0.018) scores. These results suggest informants may better detect changes in the executive function domain, while change in informant rated memory scores best predicts change in assessed cognitive ability. Alternatively, memory domain scores may be sensitive to changes across both early and late cognitive decline, whereas executive function domain scores are more sensitive to changes associated with later noticeable cognitive decline. Our results provide further support for the validity of the CS-DS to assess everyday cognitive abilities and to detect associated longitudinal changes in individuals with DS.

Startin, C.M., Rodger, R., Fodor-Wynne, L., Hamburg, S., & Strydom, A.
Developing an informant questionnaire for cognitive abilities in Down syndrome: The Cognitive Scale for Down Syndrome (CS-DS)
PLOS ONE, Published: May 6, 2016
https://doi.org/10.1371/journal.pone.0154596
Abstract: Down syndrome (DS) is the most common genetic cause of intellectual disability (ID). Abilities relating to executive function, memory and language are particularly affected in DS, although there is a large variability across individuals. People with DS also show an increased risk of developing dementia. While assessment batteries have been developed for adults with DS to assess cognitive abilities, these batteries may not be suitable for those with more severe IDs, dementia, or visual / hearing difficulties. Here we report the development of an informant rated questionnaire, the Cognitive Scale for Down Syndrome (CS-DS), which focuses on everyday abilities relating to executive function, memory and language, and is suitable for assessing these abilities in all adults with DS regardless of cognitive ability. Complete questionnaires were collected about 128 individuals with DS. After final question selection we found high internal consistency scores across the total questionnaire and within the executive function, memory and language domains. CS-DS scores showed a wide range, with minimal floor and ceiling effects. We found high interrater (n = 55) and test retest (n = 36) intraclass correlations. CS-DS scores were significantly lower in those aged 41+ with significant cognitive decline compared to those without decline. Across all adults without cognitive decline, CS-DS scores correlated significantly to measures of general abilities. Exploratory factor analysis suggested five factors within the scale, relating to memory, self-regulation / inhibition, self-direction / initiation, communication, and focussing attention. The CS-DS therefore shows good interrater and test retest reliability, and appears to be a valid and suitable informant rating tool for assessing everyday cognitive abilities in a wide range of individuals with DS. Such a questionnaire may be a useful outcome measure for intervention studies to assess improvements to cognition, in addition to detecting dementia-related cognitive decline. The CS-DS may also be a useful tool for other populations with ID.

Stephens, M.M., Herge, E., & Wright, C.
Down Syndrome and Dementia: A Patient and Care-Giver Centered Approach.
Delaware Journal of Public Health, 2021, Sep 27. 7(4), 128-130. doi: 10.32481/djph.2021.09.016.
https://djph.org/wp-content/uploads/2021/09/djph-74-016.pdf
Abstract: Reviewed are the common characteristics and co-morbidities of individuals with Down syndrome (DS), he epidemiology of AD and clinical presentation of AD in this cohort, and discussed are practical considerations for diagnosis and treatment as well as strategies to maximize support for caregivers. Consistent with the National Plan to Address Alzheimer's Disease, we will focus on the importance of the early detection of AD and what it means to patients, caregivers, and the healthcare system. The most commonly used screening tool for evaluating the early indicators of dementia in DS is the National Task Group - Early Detection Screen for Dementia (NTG-EDSD). The NTG-EDSD scoring scale helps clinicians compare patients with DS to their own baseline as the questions are scored always been the case, always but worse, new symptom in the past year, or does not apply. The framework of analysis reflects transitions from "clinically stable" to concern for MCI-DS to suspicious for AD. Pharmacotherapy and non-pharmacologic approaches to treat co-existing depression and anxiety, and improve quality of life are important considerations in the care of patients with AD and DS. Caring for and supporting patients with DS and MCI-DS and AD is best managed from an interprofessional team approach to achieve the best outcomes. Social workers, case managers, nurses and occupational therapists can evaluate the needs of the patient and caregiver and provide recommendations to support patient participation in activities and routines which can improve patient overall health and well-being as well as reduce unwanted behaviors. The goal of the primary care physician is to recognize and manage the medical and socioeconomic disparity that exists for patients with DS, especially as they age.

Struckmeyer, L.R., & Pickens, N.
Home modifications for people with Alzheimer's disease: A scoping review
American Journal of Occupational Therapy, 2015, 70(1), 1-9. DOI:10.5014/ajot.2015.016089Corpus ID: 31243965.
Abstract: The purpose of this review was twofold: (1) to gain insight into what is known from the literature about home modifications for people with Alzheimer's disease (AD) and (2) to identify gaps in the literature that could lead to opportunities for research. A systematic scoping review of peer-reviewed articles published from 1994 through 2014 explored home modifications and AD. Seventeen articles met the inclusion criteria. The three major findings pertain to (1) the caregiver role and caregiver training, (2) a client-centered collaborative approach to assessment and intervention, and (3) modifications for safety and function. Home modifications involved the physical and social environments as well as cognitive strategies at the task level. Opportunities exist for the development of assessment procedures, the exploration of home modifications in the later stages of AD, and the study of home modification needs of people with dementia who live alone.

Strydom, A.
Clinical trials to prevent or delay Alzheimer's disease in individuals with Down syndrome
Journal of Intellectual Disability Research, 2019, 63(8), 640-641. https://onlinelibrary.wiley.com/doi/abs/10.1111/jir.12651
Abstract: Adults with Down syndrome (DS) have neuropathological features identical to individuals with sporadic Alzheimer's disease (AD) and this discovery played an important role in the identification of the amyloid precursor protein gene on chromosome 21. Individuals with DS have a lifetime risk for dementia in excess of 90% and DS is now acknowledged to be the most common genetic cause of AD, but this group is often excluded from AD medication trials. This review will discuss primary and secondary prevention trials for AD in DS and the potential barriers and solutions to such trials. It will include a brief overview of the epidemiology, diagnosis and outcome measurement issues pertinent to prevention trials, as well as important ageing-related co-morbidities that need to be considered in the design of such trials. Ddescribed is the work of the Europe-wide Horizon21 Consortium and other efforts to establish DS clinical trials networks, as well as to consider the methodological issues for trials to prevent or delay AD in DS. It was noted that individuals with DS could benefit from treatments to prevent or delay AD. Improved knowledge of pathogenic processes and their clinical consequences in DS will hopefully lead to new clinical trials.

Strydom, A., & Hassiotis, A.
Diagnostic instruments for dementia in older people with intellectual disability in clinical practice
Aging & Mental Health, 2003, 7(6), 431-437. doi: 10.1080/13607860310001594682.
Abstract: There is a need for simple and reliable screening instruments for dementia in the intellectual disability (ID) population that can also be used to follow their progress, particularly if they are being treated with anti-dementia drugs. Commonly used tests for the general population such as the Mini Mental State Examination (MMSE) are not appropriate for many people with ID. This paper is a literature review of alternative instruments that have been used in research or recommended by experts since 1991 and have the potential to be used as screening instruments. Two types of tests have been identified: those administered to informants, and those that rely on direct assessment of the individual. The most promising informant rated screening tool in most adults with ID including Down syndrome (DS) diagnosis is the Dementia Questionnaire for Persons with Mental Retardation (DMR). However, sensitivity in single assessments is variable and cut-off scores need further optimization. In those with DS, the Dementia Scale for Down Syndrome (DSDS) has good specificity but mediocre sensitivity. The Test for Severe Impairment and Severe Impairment Battery are two direct assessment tools that show promise as screening instruments, but need further evaluation.

Strydom, A., Al-Janabi, T., Houston, M., & Ridley, J.
Best practice in caring for adults with dementia and learning disabilities.
Nursing Standard (Royal College of Nursing UK) , 2016, Oct, 31(6), 42-51. doi: 10.7748/ns.2016.e10524.
Abstract: People with intellectual [learning] disabilities, particularly Down's syndrome, are at increased risk of dementia. At present, services and care tailored to people with both dementia and a intellectual disability are unsatisfactory. This article reviews the literature specific to dementia in people with learning disabilities, including: comprehensive screening, diagnosis, management, environmental considerations, end of life care and training issues for nursing staff. Recommendations for best practice and service improvement are made to improve the quality of life for individuals with dementia and learning disabilities, pre and post-diagnosis.

Strydom, A., & Hassiotis, A., **Livingston, G., & King, M.**
Prevalence of dementia in older adults with intellectual disability without Down syndrome
Journal of Applied Research in Intellectual Disabilities, 2006, 19, 253.
Abstract: The aim of this study was to determine the prevalence of dementia in older adults with intellectual disability(ID) without Down syndrome. The authors identified the total population of adults with ID aged 60+ in the five London boroughs served through local social services registers, ID teams and residential services providers and then screened the Ss with a simple object memory task, information about functional status, and the Dementia Questionnaire for Persons with Mental Retardation (DMR). Screen positives on the DMR, or those with unexplained functional decline or memory deficits underwent detailed examination. Full assessment of cognitive and physical function was undertaken and additional information was collected from informants and medical records. All information was summarized to determine dementia status with IDC-10, DSM-IV, and DC-LD criteria. The authors identified 264 adults with ID and 222 (84%) participated in the study. One in four screened positive. The prevalence rate for ICD-10 or DSM-IV was 12%. Prevalence differed between those with mild and severe ID, and between diagnostic criteria. The authors concluded that dementia is common in older adults with ID without DS, but prevalence in severe ID deviated from prediction and the use of diagnostic criteria needs to be reviewed.

Strydom, A., Hassiotis, A., & Livingston, G.
Mental health and social care needs of older people with intellectual disabilities
Journal of Applied Research in Intellectual Disabilities, 2005, 18(3), 229-235. DOI:10.1111/J.1468-3148.2005.00221.X
Abstract: Older people with intellectual disabilities (ID) are a growing population but their age-related needs are rarely considered and community services are still geared towards the younger age group. We aimed to examine the mental health and social care needs of this new service user group. We identified all adults with ID without Down syndrome (DS) aged 65+ living in the London boroughs of Camden and Islington. The Psychiatric Assessment Schedule for Adults with a Developmental Disability (PASADD) checklist was used to detect psychiatric disorder, the Vineland Behavior Scale (maladaptive domain) for problem behaviors and the Dementia Questionnaire for Persons with Mental Retardation (DMR) to screen for dementia. Carers reported health problems and disability. Needs were measured with the Camberwell Assessment of Need for adults with Intellectual Disabilities (CANDID-S). A total of 23 older people with ID (13 had mild ID and nine more severe ID) and their carers participated in the survey. In which, 74% had one or more psychiatric symptoms; 30% were previously known with a diagnosis of mental illness. One-third of the older people screened positive for dementia (range: 17-44%, depending on sensitivity of DMR scores used). Three quarters of the group had physical health problems, 74% had poor sight, 22% had hearing loss and 30% had mobility problems. Carers rated unmet needs for accommodation (22%), day activities, and eyesight and hearing. The people with ID rated unmet needs to be social relationships (44%), information and physical health. Authors concluded that older people with ID without DS have considerable prevalence of health problems and psychiatric disorders, including symptoms of functional decline and dementia. Such symptoms are often not recognized and further research into their needs is a priority.

Strydom, A., Livingston, G., King, M., & Hassiotis. A.
Prevalence of dementia in intellectual disability using different diagnostic criteria.
British Journal of Psychiatry, 2007, 191(2), 150-157. doi: 10.1016/j.ridd.2013.02.021. Epub 2013 Apr 9.
Abstract: Diagnosis of dementia is complex in adults with intellectual disability owing to their pre-existing deficits and different presentation. To describe the clinical features and prevalence of dementia and its subtypes, and to compare the concurrent validity of dementia criteria in older adults with intellectual disability. The Becoming Older with Learning Disability (BOLD) memory study is a two-stage epidemiological survey of adults with intellectual disability without Down syndrome aged 60 years and older, with comprehensive assessment of people who screen positive. Dementia was diagnosed according to ICD–10, DSM–IV and DC–LD criteria. The DSM–IV dementia criteria were more inclusive. Diagnosis using ICD–10 excluded people with even moderate dementia. Clinical subtypes of dementia can be recognized in adults with

intellectual disability. Alzheimer's dementia was the most common, with a prevalence of 8.6% (95% CI 5.2–13.0), almost three times greater than expected. Dementia is common in older adults with intellectual disability, but prevalence differs according to the diagnostic criteria used. This has implications for clinical practice.

Strydom, A., Hassiotis, A., King, M., Livingston, G.
The relationship of dementia prevalence in older adults with intellectual disability (ID) to age and severity of ID.
Psychological Medicine, 2008, 15, 1-9. doi: 10.1017/S0033291708003334.
Abstract: Previous research has shown that adults with intellectual disability (ID) may be more at risk of developing dementia in old age than expected. However, the effect of age and ID severity on dementia prevalence rates has never been reported. We investigated the predictions that older adults with ID should have high prevalence rates of dementia that differ between ID severity groups and that the age-associated risk should be shifted to a younger age relative to the general population. A two-staged epidemiological survey of 281 adults with ID without Down syndrome (DS) aged 60 years; participants who screened positive with a memory task, informant-reported change in function or with the Dementia Questionnaire for Persons with Mental Retardation (DMR) underwent a detailed assessment. Diagnoses were made by psychiatrists according to international criteria. Prevalence rates were compared with UK prevalence and European consensus rates using standardized morbidity ratios (SMRs). Dementia was more common in this population (prevalence of 18.3%, SMR 2.77 in those aged 65 years). Prevalence rates did not differ between mild, moderate and severe ID groups. Age was a strong risk factor and was not influenced by sex or ID severity. As predicted, SMRs were higher for younger age groups compared to older age groups, indicating a relative shift in age-associated risk. Criteria-defined dementia is 2-3 times more common in the ID population, with a shift in risk to younger age groups compared to the general population.

Strydom, A., Shooshtari, S., Lee, L., Raykar, V., Torr, J., Tsiouris, J., Jokinen, N., Courtenay, K., Bass, N., Sinnema, M., & Maaskant, M.
Dementia in older adults with intellectual disabilities—epidemiology, presentation, and diagnosis
Journal of Policy and Practice in Intellectual Disabilities, 2010, 7(2), 96-110. https://doi.org/10.1111/j.1741-1130.2010.00253.x
Abstract: As life expectancy of people with intellectual disabilities (ID) extends into older age, dementia is an increasing cause of morbidity and mortality. To update and summarize current knowledge on dementia in older adults with ID, the authors conducted a comprehensive review of the published literature from 1997–2008 with a specific focus on: (1) epidemiology of dementia in ID in general as well as in specific genetic syndromes; (2) presentation; and (3) diagnostic criteria for dementia. The review drew upon a combination of searches in electronic databases Medline, EMBASE, and PsycINFO for original research papers in English, Dutch, or German. The authors report that varied methodologies and inherent challenges in diagnosis yield a wide range of reported prevalence rates of dementia. Rates of dementia in the population with intellectual disability not because of Down syndrome (DS) are comparable with or higher than the general population. Alzheimer's disease onset in DS appears earlier and the prevalence increases from under 10% in the 40s to more than 30% in the 50s, with varying prevalence reported for those 60 and older. Incidence rates increase with age. Few studies of dementia in other genetic syndromes were identified. Presentation differs in the ID population compared with the general population; those with DS present with prominent behavioral changes believed to be because of frontal lobe deficits. Authors recommend large-scale collaborative studies of high quality to further knowledge on the epidemiology and clinical presentation of dementia in this population.

Strydom, A., Chan, T., King, M., Hassiotis, A., & Livingston, G.
Incidence of dementia in older adults with intellectual disabilities
Research in Developmental Disabilities, 2013, 34(6), 1881-1885. doi:10.1016/j.ridd.2013.02.021. Epub 2013 Apr 9.
Abstract: Dementia may be more common in older adults with intellectual disability (ID) than in the general population. The increased risk for Alzheimer's disease in people with Down syndrome (DS) is well established, but much less is known about dementia in adults with ID who do not have DS. We estimated incidence rates from a longitudinal study of dementia in older adults with ID without DS and compared them to general population rates. 222 participants with ID without DS aged 60 years and older were followed up an average of 2.9 years later to identify those who had declined in functional or cognitive abilities. Those who screened positive had a comprehensive assessment for dementia, diagnosed using ICD 10 and DSM IV criteria. 134 participants who did not have dementia at initial assessment were alive and interviewed at follow up; 21 (15.7%) were diagnosed with dementia. Overall incidence rate for those aged = 60 was 54.6/1000 person years (95% CI 34.1-82.3). The highest incidence rate (97.8/1000 person years) was in the age group 70-74. Standardised incidence ratio for those aged = 65 was 4.98 (95% CI 1.62-11.67). Incidence of dementia in older people with intellectual disabilities are up to five times higher than older adults in the general population. Screening may be useful in this population given the high incident rates, particularly as more effective treatments become available. Studies to explore the underlying aetiological factors for dementia associated with intellectual disability could help to identify novel protective and risk factors.

Strydom, A., Coppus, A., Blesa, R., Danek, A., Fortea, J., Hardy, J., Levin, J., Nuebling G., Rebillat A.S., Ritchie, C., van Duijn, C., Zaman, S., & Zetterberg, H.
Alzheimer's disease in Down syndrome: An overlooked population for prevention trials.
Alzheimers Dementia (N Y), 2018, 13(4), 703-713. doi: 10.1016/j.trci.2018.10.006.
Abstract: The discovery that adults with Down syndrome (DS) have neuropathological features identical to individuals with sporadic Alzheimer's disease (AD) played a key role in the identification of the amyloid precursor protein gene on chromosome 21 and resulted in the amyloid cascade hypothesis. Individuals with DS have a lifetime risk for dementia in excess of 90%, and DS is now acknowledged to be a genetic form of AD similar to rare autosomal-dominant causes. Just as DS put the spotlight on amyloid precursor protein mutations, it is also likely to inform us of the impact of manipulating the amyloid pathway on treatment outcomes in AD. Ironically, however, individuals with DS are usually excluded from AD trials. This review will discuss primary and secondary prevention trials for AD in DS and the potential barriers and solutions to such trials and describe the Europe-wide Horizon21 Consortium to establish a DS-AD prevention clinical trials network.

Stuart, C.
Estimating the number of people with Down's syndrome in Scotland and the cohort at elevated risk of early onset dementia.
Tizard Learning Disability Review, 2017, 22(3), 164-171 https://doi.org/10.1108/TLDR-11-2016-0041
Abstract: The purpose of this study was to ascertain the size of the population of adults with Down syndrome in Scotland and provide a basis for estimating their number and age ranges with dementia. Data were requested from all general practitioners (GP) in Scotland on people with an identified READ code denoting Down syndrome. A statistical weighting model was then applied to account for non-response bias. Findings were that there were 3,261 adults with Down estimated by the application of a statistical weighting model. Of these, 1,118 (34%) were aged between 40 and 59. This age group includes those adults with the highest incidence of early onset dementia. It was not possible to apply a benchmark to the percentage of observed data so as to determine the accuracy of the estimates. Adults with Down have an elevated risk of developing dementia significantly earlier than the general population and require specific age appropriate supports and services to meet their needs both pre- and post-diagnosis. The reality of this is currently not fully realized in either standard practice or national policy concerning the issue.

Sung, H., Hawkins, B.A., Eklund, S.J., Kim, K.A., Foose, A., May, M.E., Rogers, N.B.
Depression and dementia in aging adults with Down syndrome: a case study approach.
Mental Retardation, 1997, 35(1), 27-38. https://doi: 10.1352/0047-6765(1997)035<0027:DADIAA>2.0.CO;2
Abstract: Patterns of symptoms associated with depression and dementia were examined in 3 aging adults with Down syndrome. A case study approach (Yin, 1994) was employed to identify and link these symptoms. Results of the case analyses provide further insight into distinguishing between depression and dementia in older persons with Down syndrome.

Svanelov, E.
An observation study of power practices and participation in group homes for people with intellectual disability.
Disability and Society, 2020, 35(9), 1419-1440.

https://doi.org/10.1080/09687599.2019.1691978
Abstract: This study explored how participation constitutes and is constituted by practices of power in group homes for people with intellectual disability. The study used disciplinary power as theoretical perspective and was based on 50 h of observation in two group homes with a total of 15 residents. The analysis identifies practices of power and their relationship to individual agency and participation. The results show that institutional structures construct practices of power that define codes of conduct for the group home residents and their possibility for participation. This study offers implications for the daily lives of residents in group homes for people with intellectual disability.

te Boekhorst, S., Depla, M.F.I.A., de Lange, J., Pot, A.M. & Eefsting, J.A.
The effects of group living homes for older people with dementia: A comparison with traditional nursing home care.
International Journal of Geriatric Psychiatry, 2009, 24(9), 970-978. doi:10.1002/gps.2205.
Abstract: [Non-ID population] The aim of this study was to investigate the effects of group living homes on quality of life and functioning of people with dementia. The study had a quasi-experimental design with a baseline measurement on admission and an effect measurement six months later. Participants were 67 residents in 19 group living homes and 97 residents in seven traditional nursing homes. DQOL and QUALIDEM measured quality of life, functional status was examined with MMSE, IDDD, RMBPC, NPI-Q and RISE from RAI. Use of psychotropic drugs and physical restraints was also assessed. Linear and logistic regression analyses analyzed the data. After adjustment for differences in baseline characteristics, residents of group living homes needed less help with ADL and were more socially engaged. There were no differences in behavioral problems or cognitive status. Also after adjusting, two of the 12 quality of life subscales differed between the groups. Residents of group living homes had more sense of aesthetics and had more to do. While there were no differences in prescription of psychotropic drugs, residents of group living homes had less physical restraints. Group living homes had some beneficial effects on its residents, but traditional nursing homes performed well as well. Possible study limitations included the baseline differences between the study groups and the use of different informants on T0 and T1. Future nursing home care may very well be a combination of the best group living care and traditional nursing home care have to offer.

te Boekhorst, S., Willemse, B., Depla, F.I.A., Eefsting, J.A., & Pot, A.M.
Working in group living homes for older people with dementia: The effects on job satisfaction and burnpout and the role of job characteristics
International Psychogeriatrics, 2008, 20(5), 927-940. doi:10.1017/S1041610208007291. Epub 2008 May 19.
Abstract: [Non-ID population] Group living homes are a fast-growing form of nursing home care for older people with dementia. This study seeks to determine the differences in job characteristics of nursing staff in group living homes and their influence on well-being. We examined the Job Demand Control Support (JDCS) model in relation to 183 professional caregivers in group living homes and 197 professional caregivers in traditional nursing homes. Multilevel linear regression analysis was used to study the mediator effect of the three job characteristics of the JDCS-model (demands, control and social support) on job satisfaction and three components of burnout (emotional exhaustion, depersonalization and decreased personal accomplishment). Demands were lower in group living homes, while control and social support from co-workers were higher in this setting. Likewise, job satisfaction was higher and burnout was lower in group living homes. Analysis of the mediator effects showed that job satisfaction was fully mediated by all three psychosocial job characteristics, as was emotional exhaustion. Depersonalization was also fully mediated, but only by control and social support. Decreased personal accomplishment was partially mediated, again only by job characteristics, control and support. This study indicates that working in a group living home instead of a traditional nursing home has a beneficial effect on the well-being of nursing staff, largely because of a positive difference in psychosocial job characteristics.

Taggert, L., Truesdale-Kennedy, M., Ryan, A., McConkey, R.
Examining the support needs of ageing family carers in developing future plans for a relative with an intellectual disability
Journal of Intellectual Disabilities, 2012 Sep;16(3):217-234. doi: 10.1177/1744629512456465.
Abstract: Planning for the future care of adults with an intellectual disability after the main family carer ceases their care, continues to be a sensitive and difficult time posing challenges for service providers internationally. Limited research has been undertaken on this topic because until recently, people with intellectual disability usually pre-deceased their parents. This study examined ageing carers' preferences for future care and the support systems required to make such future plans. The study was conducted in one region of the United Kingdom with a high proportion of family carers. A mixed methods design was employed. In Stage 1, a structured questionnaire was used to collate information on the health, caregiving demands and future planning preferences of 112 parent and sibling carers; aged 60-94 years. In Stage 2, 19 in-depth semistructured interviews were undertaken with a sample of carers to explore a range of issues around future planning. Over half of the carers were lone carers, mainly female, with many reporting a wide range of health problems. A third of these carers reported that their caregiving resulted in high levels of anxiety. The main preference of the carers was for the person to remain in the family home, with either the family and/or paid staff to support them. A minority of parent carers preferred the person to move into the home of a sibling, although some favored the person moving to a residential facility with other people with intellectual disabilities. The majority of carers did not want their relative to move into an older people's residential/nursing facility. In the qualitative data, four main themes were identified around future planning: unremitting apprehension, the extent of planning, obstacles encountered and solutions for future planning. Avoidance, lack of guidance and a lack of appropriate residential provision were cited as obstacles to making future plans compounded by the emotional upset experienced by carers in thinking about the future. Findings of this study clearly identify the emotional, informational and practical supports required by these ageing family carers. These findings have national and international relevance in influencing how governments and service providers support parent and sibling carers to proactively plan for the future, and in the development of both in-home and out-of-home options when a family carer can no longer provide care. This is more urgent than ever given the growing numbers of older persons with intellectual disabilities in future decades.

Takenoshita, S., Kuwano, R., Inoue, T., Kurozumi, T., Terada, S., Yamada, N., & **Suemitsu, S.**
Prevalence of dementia in people with intellectual disabilities without Down syndrome in Japan
Journal of Applied Research in Intellectual Disabilities, 2021, 34(5), 1305. doi: 10.1002/gps.5258.
Abstract:There are few large-scale studies of the prevalence of dementia in people with intellectual disabilitywho do not have Down syndrome, although Down syndrome is well known to be related with Alzheimer's disease. We investigated 1831 adults with intellectual disabilitybut without Down syndrome who were residents of a facility for people with intellectual disabilityin Japan. The caregivers answered a questionnaire, and physicians directly examined the participants suspected of cognitive decline. Of the 1831 patients, 118 were diagnosed with dementia and 51 with mild cognitive impairment (MCI). The prevalence of dementia was 5.6% for the 55 to 64 age group, 13.2% for the 65 to 74 age group, 22.2% for the 75 to 84 age group, and 42.3% for the 85 to 94 age group. The prevalence of MCI was 2.1% for the 55 to 64 age group, 7.4% for the 65 to 74 age group, and 11.5% for the 85 to 94 age group. Sixty-five patients (55.1%) had not been diagnosed with dementia before the survey. People with intellectual disability without Down syndrome may develop dementia at an earlier age than those without intellectual disability. Our nationwide survey suggested there are many undiagnosed dementia patients in the community of people with intellectual disability.

Takenoshita, S., Terada, S., Kuwano, R., Inoue, T., Cyoju, A., Suemitsu, S., & Yamada, N.

Prevalence of dementia in people with intellectual disabilities: Cross-sectional study
International Journal of Geriatric Psychiatry, 2020 (April), 35(4), 314-422. https://doi.org/10.1002/gps.5258
Abstract: There are only a few studies of the prevalence of dementia in people with intellectual disability (ID) without Down syndrome (DS), and there is a large difference in the prevalences between reported studies. Moreover, the prevalence of mild cognitive impairment (MCI) in ID has not been reported. We aimed to evaluate the prevalence of dementia in adults of all ages and the prevalence of MCI in people with ID. Furthermore, we tried to clarify the differences depending on the various diagnostic criteria. The survey included 493 adults with ID at 28 facilities in Japan. The caregivers answered a questionnaire, and physicians directly examined the participants who were suspected of cognitive decline. Dementia and MCI were diagnosed according to ICD-10, DC-LD, and DSM-5 criteria. The prevalence of dementia was 0.8% for the 45 to 54 years old group, 3.5% for the 55 to 64 years old group, and 13.9% for the 65 to 74 years old group in people with ID without DS. The prevalence of MCI was 3.1% for patients 45 to 54, 3.5% for patients 55 to 64, and 2.8% for patients 65 to 74 with ID without DS. DSM-5 was the most inclusive in diagnosing dementia and MCI in people with ID. People with ID without DS may develop dementia and MCI at an earlier age and higher rate than the general population. Among the diagnostic criteria, DSM-5 was the most useful for diagnosing their cognitive impairment.

Takenoshita, S., Terada, S., Kuwano, R., Inoue, T., Kurozumi, T., Choju, A., Suemitsu, S., & Yamada, N.
Validation of the Japanese version of the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities.
Journal of Intellectual Disability Research, 2020 Dec; 64(12), 970-979. doi: 10.1111/jir.12788. Epub 2020 Oct 5.
Abstract: Dementia in people with intellectual disabilities (IDs) is difficult to detect because of preexisting cognitive deficits. An effective screening method is required. The Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) was developed as an observer rating tool to screen dementia in people with ID. The aim of this study was to verify the screening accuracy of the DSQIID for Japanese people with ID. Four-hundred ninety-three subjects with ID participated in this study. Caregivers who had observed the participants for more than 2 years scored the Japanese version of the DSQIID (DSQIID-J) of the participants. Three doctors examined participants directly and diagnosed dementia using the Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition criteria. To identify the key screening items that predict dementia, the specificities of a single and pairs of items with 100% sensitivity were evaluated relative to the dementia diagnosis. Of 493 participants, 34 were people with Down syndrome (DS), and 459 were people without DS. Seventeen participants were diagnosed with dementia. The suitable cut-off score of the DSQIID-J was 10/11 (sensitivity 100% and specificity 96.8%) for screening dementia. The inter-rater reliability, test-retest reliability and internal consistency of the DSQIID-J were excellent. Regarding key items, there was no single item with 100% sensitivity, and the best two-item combination was the pair of 'Cannot dress without help' and 'Walks slower' (sensitivity 100% and specificity 93.5%). We identified several important question items of the DSQIID-J related to the diagnosis of dementia in people with ID. The DSQIID-J is a useful screening tool for dementia in adults with ID.

Temple, V., & Konstantareas, M.M.
A comparison of the behavioural and emotional characteristics of Alzheimer's dementia in individuals with and without Down syndrome.
Canadian Journal of Aging, 2005, 24(2), 179-190
Abstract: The behavioral and emotional changes associated with Alzheimer's disease (AD) are compared for individuals with Down syndrome and AD and individuals with AD from the general population (AD-only). The primary caregivers of 30 people with Down syndrome and AD and 30 people with AD-only completed the BEHAVE-AD and the Apathy subscale of the CERAD. As well, behavioral observations at adult day programs were undertaken with selected participants (n=26). The Down syndrome group experienced fewer delusions and had lower total scores on the BEHAVE-AD, indicating fewer problem behaviors overall. Day program observations suggested that the AD-only group were more likely to be sedentary and observe the activities of others, while the Down syndrome group were more physically active. Improving our understanding of the similarities and differences between these two groups may help facilitate the integration of individuals with Down syndrome into adult day programs for the general population.

Temple, V., Jozsvai, E., Konstantareas, M.M., & Hewitt, T.A.
Alzheimer dementia in Down's syndrome: the relevance of cognitive ability.
Journal of Intellectual Disability Research, 2001, 45(1), 47-55. doi:10.1046/j.1365-2788.2001.00299.x.
Abstract: More years of education have been found to be associated with a lower rate of Alzheimer disease (AD) in individuals without intellectual disability. It has been proposed that education reflects greater 'synaptic reserve' and that greater synaptic reserve may defer the development of AD. The present study compared individuals with Down's syndrome (DS) who were found to have symptoms of dementia with those who remained symptom-free to determine if the two groups differed in their level of education, employment, recreational activities, years in an institution or overall level of cognitive functioning. Thirty-five adults with DS aged between 29 and 67 years were assessed. The participants were recruited from a community health facility and included individuals with a wide range of ability levels. Neuropsychological testing, caregiver report and the Dementia Scale for Down Syndrome (Gedye 1995) were used to identify decline in participants over periods of 6 months to 3 years. After the effect of age was statistically removed, multiple regression analyses revealed that level of cognitive functioning was significantly associated with decline such that a higher level of cognitive functioning predicted less decline. None of the environmental variables (i.e. educational level, years in an institution and employment) were directly associated with decline; however, a post hoc regression using level of cognitive functioning as the outcome variable revealed that level of cognitive functioning itself was associated with these environmental variables. A higher level of cognitive functioning was associated with fewer cases of dementia in individuals with DS, and level of cognitive functioning appears to be associated with environmental factors such as level of education, years in an institution and employment. The present findings suggest that environmental interventions aimed at improving level of cognitive functioning may also be useful in deferring the onset of dementia.

Thalen, M., Volkers, K.M., van Oorsouw, W.M.W.J.,& Embregts, P.J.C.M.
Psychosocial interventions for older people with intellectual disabilities and the role of support staff: A systematic review.
Journal of Applied Research in Intellectual Disabilities, 2022 Mar;35(2):312-337. doi: 10.1111/jar.12953. Epub 2021 Nov 15.
Abstract: The life expectancy of people with intellectual disabilities has increased. The implications of ageing have resulted in changes in their support needs and challenges to support staff. Access to evidence-based strategies for support staff providing care to elderly with intellectual disabilities remains scarce. A systematic review was conducted to provide an overview of available psychosocial interventions. Four databases were searched and assessed according to the PRISMA guidelines. A narrative, integrative method of analysis was conducted to synthesize quantitative and qualitative data. The 36 studies included in the review reported on interventions aimed at either identifying and meeting the needs or perceptions of older individuals or at improving their behavior and skills. Furthermore, the role of support staff in the implementation of interventions was either active, assisting or undefined. This overview of studies could contribute to the existing body of knowledge and help to optimize psychosocial support for a growing population.

Thase, M.E., Tigner, R., Smeltzer, D.J., & Liss. L
Age-related neuropsychological deficits in Down's syndrome
Biological Psychiatry, 1984 Apr; 19(4), 571-585. https://pubmed.ncbi.nlm.nih.gov/6234031/

Abstract: Down's syndrome (DS) has been suggested as a high-risk condition for Dementia Alzheimer's type (DAT). In the present study, neuropsychological variables were assessed in 165 DS subjects and 163 matched controls with intellectual disability. Overall, DS subjects had lower scores for orientation, digit span, visual memory, object-naming, and general knowledge, as well as more "released" reflexes. Impairments were most evident in DS subjects greater than 50 years old. These findings provide further support for an association between aging and DAT in Down's syndrome. Methodological issues and areas for future research are discussed.

The Arc
Developmental disabilities and Alzheimer disease: what you should know.
43 pp.
Silver Spring, Maryland: The Arc of the United States [1010 Wayne Avenue, Suite 650, Silver Spring, MD 20910 – www.TheArc.org] (1995)
Abstract: A booklet covering some of the fundamentals concerning adults with intellectual disabilities and Alzheimer's disease including what is Alzheimer's disease, its course and outcome, diagnostic suggestions, care considerations, and how to obtain assistance. Contains resource list and glossary.

Thompson, D.J., Ryrie, I., & Wright, S.
People with intellectual disabilities living in generic residential services for older people in the UK
Journal of Applied Research in Intellectual Disabilities, 2004, 17(2), 101-108. https://doi.org/10.1111/j.1360-2322.2004.00187.x
Abstract: As part of a UK program of work focusing on older people with ID, the circumstances of those who reside in generic services for older people were investigated. Some 215 people with ID were identified living in 150 homes. They were significantly younger than other residents and were placed in these homes more because of organizational change or the aging/death of family carers, rather than due to their own needs. Of the residents, 24 adults had Down syndrome, 8 of whom were noted to have dementia. Of the 215, 45 had dementia. Average age of people with DS upon entry was 60 and those remaining at the homes was about 65.

Tom, S.E., Hubbard, R.A., Crane, P.K., Haneuse, S.J., Bowen, J., McCormick, W.C., McCurry, S., & Larson, E.B.
Characterization of dementia and Alzheimer's disease in an older population: updated incidence and life expectancy with and without dementia
American Journal of Public Health, 2015 Feb,105(2), 408-413. doi: 10.2105/AJPH.2014.301935.
Abstract: Authors estimated dementia incidence rates, life expectancies with and without dementia, and percentage of total life expectancy without dementia. Authors studied 3605 members of Group Health (Seattle, WA) aged 65 years or older who did not have dementia at enrollment to the Adult Changes in Thought study between 1994 and 2008. We estimated **incidence rates of Alzheimer's disease and dementia**, as well as **life expectancies** with and without dementia, defined as the average number of years one is expected to live with and without dementia, and percentage of total life expectancy without dementia. Dementia incidence increased through ages 85 to 89 years (74.2 cases per 1000 person-years) and 90 years or older (105 cases per 1000 person-years). Life expectancy without dementia and percentage of total life expectancy without dementia decreased with age. Life expectancy with dementia was longer in women and people with at least a college degree. Percentage of total life expectancy without dementia was greater in younger age groups, men, and those with more education. Efforts to delay onset of dementia, if successful, would likely benefit older adults of all ages.

Torr, J., & Davis, R.
Ageing and mental health problems in people with intellectual disability
Current Opinions in Psychiatry, 2007, 20(5), 467-471. doi: 10.1097/YCO.0b013e328278520d.
Abstract: Increasing numbers of people with intellectual disability are now living well into old age. This paper will review the recent literature pertaining to the mental health of older people with intellectual disability. Overall, the prevalence of mental health problems is high in adults of all ages with intellectual disability. A major epidemiological study did not report sufficient detail to examine the effect of ageing on specific disorders or the differential effects of ageing and early mortality in people with Down's syndrome. At least a third of people with Down's syndrome can expect to develop Alzheimer's disease in middle age whilst for other people with intellectual disability, Alzheimer's disease is probably no more common than in the general population. Diagnosis and management of dementia is complicated by the high rates of comorbid physical and mental health problems. Overall, mental health problems in older people with intellectual disability are similar to younger people with intellectual disability, however there are more cases of dementia and physical health problems. Further research is needed to improve our understanding of the effects of ageing on the mental health and care needs of older people with intellectual disability.

Torr, J., Strydom, A., Patti, P. & Jokinen N.
Aging in Down syndrome: Morbidity and mortality.
Journal of Policy and Practice in Intellectual Disabilities, 2010, *7*(1), 70-81. https://doi.org/10.1111/j.1741-1130.2010.00249.x
Abstract: The life expectancy of adults with Down syndrome has increased dramatically over the last 30 years, leading to increasing numbers of adults with Down syndrome now living into middle and old age. Early-onset dementia of the Alzheimer type is highly prevalent in adults with Down syndrome in the sixth decade, and this has overshadowed other important conditions related to aging among adults with Down syndrome. The authors' aim was to update and summarize current knowledge on these conditions, and examine causes of morbidity and mortality in older people with Down syndrome by conducting a systematic review of the published literature for the period: 1993–2008. They reviewed English-language literature drawn from searches in the electronic databases Medline, CINAHL, and PsycINFO, as well as supplementary historical papers. The authors conclude that functional decline in older adults with Down syndrome cannot be assumed to be due only to dementia of the Alzheimer type (which is not inevitable in all adults with Down syndrome). Functional decline may be the result from a range of disorders, especially sensory and musculoskeletal impairments. Given the high rates of early-onset age-related disorders among adults with Down syndrome, programmatic screening, monitoring, and preventive interventions are required to limit secondary disabilities and premature mortality. With respect to assessment and treatment, in the absence of specialist disability physicians, geriatricians have a role to play.

Tsang, W., Oliver, D., & Triantafyllopoulou, P.
Quality of life measurement tools for people with dementia and intellectual disabilities: A systematic review.
Journal of Applied Research in Intellectual Disabilities, 2023 Jan, 36(1), 28-38. doi: 10.1111/jar.13050.
Abstract: Adults with intellectual disabilities are an at-risk group of developing dementia. In the absence of a cure for dementia, emphasis on treatment is the promotion of Quality of life (QoL). The aim of this review is to identify and describe QoL tools for people with intellectual disabilities and dementia. A systematic review was carried out using 10 databases and papers from up to March year 2021. Two instruments were identified and examined. The QoL in late-stage dementia, which showed evidence of good levels of internal consistency, intra-rater reliability, test-retest reliability, and convergent validity. The Dementia Quality of Life - proxy was also used; however, its psychometric properties have yet to be studied within the intellectual disabilities population. Due to the degenerative nature of dementia involving cognitive impairment such as memory and language difficulties, there can be significant barriers in capturing the subjective experience of QoL in a person with dementia. Therefore, it may not be possible to capture the patient's perceived QoL from self-report measures. Interpretation of concepts can be problematic, especially when life situation of the respondent is vastly different to that of a researcher. Also, the conceptualisation of QoL can vary in different stages of dementia in the general population, such that in the early stages of dementia enjoyment of daily activities is a relevant domain for QoL, but may no longer seem to be applicable in severe or advanced

dementia. It is recommended instruments should be developed and psychometrically tested specifically for adults with intellectual disabilities and dementia to help inform policy makers, measure outcomes of interventions and personal outcomes.

Tse, M.M., Kwan, R.Y, & Lau, J.L.
Ageing in individuals with intellectual disability: issues and concerns in Hong Kong
Hong Kong Medical Journal, 2018, Feb, 24(1), 68-72.
doi: 10.12809/hkmj166302. Epub 2018 Jan 12.
Abstract: The increasing longevity of people with intellectual disability is testimony to the positive developments occurring in medical intervention. Nonetheless, early-onset age-related issues and concerns cause deterioration of their overall wellbeing. This paper aimed to explore the issues and concerns about individuals with intellectual disability as they age. Articles that discussed people older than 30 years with an intellectual disability and those that identified ageing health issues and concerns were included. Only studies reported in English from 1996 to 2016 were included. We searched PubMed, Google Scholar, and Science Direct using the terms 'intellectual disability', 'ageing', 'cognitive impairment', 'health', and 'screening'. Apart from the early onset of age-related health problems, **dementia** is more likely to develop by the age of 40 years in individuals with intellectual disability. Geriatric services to people with intellectual disability, however, are only available for those aged 60 years and older. Cognitive instruments used for the general population are not suitable for people with intellectual disability because of floor effects. In Hong Kong, the Chinese version of the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities is the only validated instrument for people with intellectual disability. The use of appropriate measurement tools to monitor the progression of age-related conditions in individuals with intellectual disability is of great value. Longitudinal assessment of cognition and function in people with intellectual disability is vital to enable early detection of significant deterioration. This allows for therapeutic intervention before substantial damage to the brain occurs such as dementia that hastens cognitive and functional decline.

Tsiouris, J.A., & Patti, P.J.
Drug treatment of depression associated with dementia or presented as "pseudodementia" in older adults with Down syndrome.
Journal of Applied Research in Developmental Disabilities, 1997, *10* (4), 312-322, https://doi.org/10.1111/j.1468-3148.1997.tb00026.x
Abstract: The response to antidepressant drugs, mainly the selective serotonin reuptake inhibitors (SSRIs), was evaluated in adults with intellectual disability (ID) and Down syndrome (DS) who presented with depression and decline in activities of daily living (ADL) skills. Among other patients with ID referred to a specialised clinic for diagnostic work-up, 37 adults with DS over the age of 40 and a mean age of 51.4 years were evaluated and 34 were followed-up. Depression associated with dementia was diagnosed in 16 cases, and depression presented as functional decline 'pseudodementia' was found in 4 cases. Recommendations for treatment with antidepressants were followed in 10 cases with a marked improvement in functioning compared to a rapid decline in 10 cases where treatment was refused. Treatment with the SSRI antidepressant drugs resulted in improved quality of life, differentiated 'pseudodementia' from dementia, and possibly delayed the dementing process in adults with DS and presentation of depression associated with dementia.

Tsiouris, J.A., Patti, P.J., & Flory, M.J.
Effects of antidepressants on longevity and dementia onset among adults with Down syndrome: a retrospective study
Journal of Clinical Psychiatry, 2014, 75(7), 731-737.
doi:10.4088/JCP.13m08562.
Abstract: To investigate the effects of antidepressants on longevity, age at dementia onset, and survival after onset among adults with Down syndrome, controlling for late-onset seizures, trisomy 21 mosaicism, and cholinesterase inhibitor use. The charts of 357 adults with Down syndrome (mean age at first visit = 46.3 years, SD = 9.0) evaluated in a metropolitan diagnostic and research clinic between 1990 and 2008 were reviewed. Seventeen patients had trisomy 21 mosaicism; 155 patients were diagnosed with depressive disorders using DSM-III-R and IV criteria, 78 of whom received antidepressants for over 90 days. Of 160 patients who developed dementia, **the estimated mean age at onset was 52.8 years**. Fifty-six patients (demented and nondemented) had late-onset seizures. Longevity and age at estimated onset among those receiving and not receiving antidepressants were compared. Cox proportional hazards models examined risks for dementia onset and death. The mean age at dementia onset among those receiving antidepressants before onset was 53.75 years versus 52.44 years among others. Proportional hazards models showed a significant delay of onset among those taking antidepressants (hazard ratio = 0.69; 95% CI, 0.48–0.98; P = .038). Mean age at death or at end of study for those receiving antidepressants was 54.71 years; among others, it was 52.60 years (hazard ratio = 0.63; 95% CI, 0.42–0.94; P = .024). Among the 35 adults with late-onset seizures and dementia who died, mean survival after seizure onset was 4.23 years. The findings in this retrospective study revealed that antidepressant use was associated with delayed dementia onset and increased longevity in adults with Down syndrome; mean survival after late-onset seizures was longer than previously reported. Further studies, however, are needed to confirm these associations, optimally in a clinical trial to confirm causality.

Tsiouris, J.A., Patti, P.J., Tipu, O. & Raguthu, S..
Adverse effects of phenytoin given for late-onset seizures in adults with Down syndrome.
Neurology, 2002, Sept 10, 59(5), 779-780. doi: 10.1212/wnl.59.5.779.
Summary (no abstract provided): A brief report that indicates the adverse effects of therapeutic levels of phenytoin and the improvement observed when phenytoin was replaced with other antiepileptics in 17 adults with DS, Alzheimer disease (AD) and late-onset seizures (LOS). The reported deterioration in the patients' condition was found to be due to the adverse effects from phenytoin and not to AD. It was suggested that practitioners avoid prescribing phenytoin to treat LOS in persons with DS and AD. If phenytoin is already prescribed, it should be replaced with another anticonvulsive agent.

Tsou, A.Y., Bulova, P., Capone, G., Chicoine, B., Gelaro, B., Odell Harville, T., Martin, B.A., McGuire, D.E., McKelvey, K.D. Peterson, M., Tyler, C., Wells, M., Whitten, M.S. & the Global Down Syndrome Foundation Medical Care Guidelines for Adults with Down Syndrome Workgroup
Medical care of adults with Down syndrome: A clinical guideline
JAMA, 2020, 324(15), 1543-1556. doi:10.1001/jama.2020.17024
Abstract: Down syndrome is the most common chromosomal condition, and average life expectancy has increased substantially, from 25 years in 1983 to 60 years in 2020. Despite the unique clinical comorbidities among adults with Down syndrome, there are no clinical guidelines for the care of these patients.
To develop an evidence-based clinical practice guideline for adults with Down syndrome. The Global Down Syndrome Foundation Medical Care Guidelines for Adults with Down Syndrome Workgroup (n=13) developed 10 Population/ Intervention/ Comparison/Outcome (PICO) questions for adults with Down syndrome addressing multiple clinical areas including mental health (2 questions), dementia, screening or treatment of diabetes, cardiovascular disease, obesity, osteoporosis, atlantoaxial instability, thyroid disease, and celiac disease. These questions guided the literature search in MEDLINE, EMBASE, PubMed, PsychINFO, Cochrane Library, and the TRIP Database, searched from January 1, 2000, to February 26, 2018, with an updated search through August 6, 2020. Using the GRADE (Grading of Recommendations, Assessment, Development, and Evaluation) methodology and the Evidence-to-Decision framework, in January 2019, the 13-member Workgroup and 16 additional clinical and scientific experts, nurses, patient representatives, and a methodologist developed clinical recommendations. A statement of good practice was made when there was a high level of certainty that the recommendation would do more good than harm, but there was little direct evidence. From 11295 literature citations associated with 10 PICO questions, 20 relevant studies were identified. An updated search identified 2 additional studies, for a total of 22 included studies (3 systematic reviews, 19 primary studies), which were reviewed and synthesized. Based on

this analysis, 14 recommendations and 4 statements of good practice were developed. Overall, the evidence base was limited. Only 1 strong recommendation was formulated: screening for Alzheimer-type dementia starting at age 40 years. Four recommendations (managing risk factors for cardiovascular disease and stroke prevention, screening for obesity, and evaluation for secondary causes of osteoporosis) agreed with existing guidance for individuals without Down syndrome. Two recommendations for diabetes screening recommend earlier initiation of screening and at shorter intervals given the high prevalence and earlier onset in adults with Down syndrome. These evidence-based clinical guidelines provide recommendations to support primary care of adults with Down syndrome. The lack of high-quality evidence limits the strength of the recommendations and highlights the need for additional research.

Tuffrey-Wijne I.
How to Break Bad News to People with Intellectual Disabilities: A Guide for Carers and Professionals.
Jessica Kingsley Publishers, London (2012).
Abstract: How to tell bad news to people with dementia? First assess the person's current framework of knowledge, then give the new chunks of information one by one, then finally check and reassess the person's knowledge. This might be useful for bereaved people with dementia to fill the gap.

Tuffrey-Wijne, I., & Watchman, K.
Breaking bad news to people with learning disabilities and dementia
Learning Disability Practice, 2015 Sept, 18(7), 16-23.
DOI:10.7748/ldp.18.7.16.e1672
Abstract: People with learning disabilities are now enjoying a longer life expectancy than ever before as a result of enhanced medical and social interventions and improved quality of life. Some, particularly individuals with Down's syndrome, are susceptible to dementia at a significantly younger age than the average age of onset in the rest of the population. Currently, there is limited guidance on how to talk to people with learning disabilities about dementia and, until such information is shared, individuals cannot be positioned as an authority on their own condition. The new model presented here suggests a way of supporting staff and families to have enabling conversations about dementia that center on the person's current situation, level of understanding and capacity.

Tuffrey-Wijne, I., & Watchman, K.
Sharing the diagnosis of dementia: Breaking bad news to people with intellectual disabilities.
In: Watchman, Karen, (ed.) Intellectual disability and dementia: research into practice. London : Jessica Kingsley Publishers. pp. 184-203. ISBN 9781849054225
Abstract: Not available - see Tuffrey-Wijne & Watchman 2015

Turky, A., Felce, D., Jones, G., & Kerr, M.
A prospective case control study of psychiatric disorders in adults with epilepsy and intellectual disability
Epilepsia, 2011 July, 52(7), 1223-1230.
https://doi.org/10.1111/j.1528-1167.2011.03044.x
Abstract: No study to date has prospectively investigated the impact of epilepsy on psychiatric disorders among adults with an intellectual disability (ID). This study aimed to determine prospectively the influence of epilepsy on the development of psychiatric disorders in adults with ID. Psychiatric symptoms were measured prospectively over a 1-year period among 45 adults with ID and active epilepsy and 45 adults with ID without epilepsy, matched on level of ID. The 1-year incidence rate (IR) of commonly occurring Axis 1 psychiatric disorders was compared with and without controlling for possible confounding factors. Total psychiatric symptom scores over the period were compared between the two groups using repeated-measures analysis of covariance. Adults with epilepsy and ID had a more than seven times increased risk for developing psychiatric disorders, particularly depression and unspecified disorders of presumed organic origin, including dementia, over a 1-year period compared to those with ID only. Comparison of the psychiatric scores showed the epilepsy group to have significantly higher unspecified disorder and depression symptom scores. The findings point to an increased risk of depression and unspecified disorders, including dementia, among adults with ID and epilepsy. Further exploration of the nature and treatment of these unspecified disorders may help the care of people with epilepsy and ID.

Tyler, C.V., & Shank, J.C.
Dementia and Down syndrome
The Journal of Family Practice, 1996, 42(6), 619-621.
https://cdn.mdedge.com/files/s3fs-public/jfp-archived-issues/1996-volume_42-43/JFP_1996-06_v42_i6_dementia-and-down-syndrome.pdf
Abstract: Case report of a 43-year old woman with Down syndrome and progressive decline over three years that was attributed to dementia of the Alzheimer's type. Authors describe the medical conditions evident during decline, whilst living with her family. Identifies typical features associated with decline for persons with Down syndrome and defines areas for concern during examinations by physicians.

Tyrrell, J., Cosgrave, M., McCarron, M., McPherson, J., Calvert, J., Kelly, A., McLaughlin, M., Gill, M.,& Lawlor, B.A.
Dementia in people with Down's syndrome.
International Journal of Geriatric Psychiatry, 2001, Dec, 16(12), 1168-1174.
Abstract: To determine the prevalence of dementia in an Irish sample of people with Down's syndrome (DS) and to examine associated clinical characteristics of dementia in this group. Some 285 people with DS (Age 35-74 years, mean age +/- SD 46.5 +/- 8.2 years) were included in this cross-sectional study. The diagnosis of dementia was made using modified DSMIV criteria. Cognitive tests used were the Down's Syndrome Mental Status Examination (DSMSE), Test for Severe Impairment (TSI) and adaptive function was measured by the Daily Living Skills Questionnaire (DLSQ). The overall prevalence of dementia was 13.3%. The presence of dementia was associated with epilepsy, myoclonus, and head injury. The demented DS group were significantly older (n = 38, mean age 54.7 years SD +/- 7.5) than the non-demented (n = 246, mean age 45.6, SD +/- 7.3). The TSI and DLSQ had a satisfactory spread of scores without 'floor' or 'ceiling' effects in people with moderate and severe learning disability. Median scores in demented versus the non-demented groups were significantly different for each measure of function. Authors conclude that dementia had a prevalence of 13.3% and occurred at a mean age of 54.7 years. The combination of DLSQ score, age and presence of epilepsy were found to predict presence of dementia.

Udell, L.
Supports in small group home settings
In M.P. Janicki & A.J. Dalton (Eds.), Dementia, Aging, and Intellectual Disabilities.
pp. 316-329
Philadelphia: Brunner-Mazel (1999)
Abstract: This book chapter covers what organizations that provide residential supports to adults with an intellectual disability need to consider in terms of planning and implementing program changes. Covered are areas that examine the nature of dementia and its possible impact on service provision, Its particular focus is on how agencies that decide to support people with dementia in small group home settings can accommodate their organizational and operational structure and offers insight ion the perspectives and questions that agencies need to consider. Suggestions are offered on how to address some of the difficulties that organizations will encounter.

University of Maryland School of Medicine
Hi Buddy... The developmentally delayed individual with Alzheimer disease
19 minutes
VideoPress, the University of Maryland School of Medicine [100 North Greene Street, Suite 300, Baltimore, Maryland USA (1 800 328 7450; fax: 1 410 706 8471; www.videopress.org)]

Abstract: Video on the subject of Alzheimer's disease and adults with developmental disabilities.

University of Stirling
Building networks - Conference on learning disabilities and dementia
58 pp.
Dementia Services Development Centre, Department of Applied Social Science, Faculty of Human Sciences, University of Stirling, Stirling, Scotland FH9 4LA (2000).
Abstract: Proceedings of conference on community dementia care and people with intellectual disabilities held in Dunblane, Scotland (November 11, 1999). The report summarizes the main points made by the numerous speakers at the conference. The conference highlighted the need for wider awareness among managers and service personnel of the need for (and for resources and developing expertise on) training staff in residential and home support services on responding to the needs of people with intellectual disabilities who have dementia. The 16 papers range from the theoretical to the practical.

Urv, T.K., Zigman, W.B., & Silverman,W.
Psychiatric symptoms in adults with down syndrome and Alzheimer's disease.
American Journal on Intellectual and Developmental Disabilities, 2010, 115(4), 265-276.
Abstract: Changes in psychiatric symptoms related to specific stages of dementia were investigated in 224 adults 45 years of age or older with Down syndrome. Findings indicate that psychiatric symptoms are a prevalent feature of dementia in the population with Down syndrome and that clinical presentation is qualitatively similar to that seen in Alzheimer's disease within the general population. Psychiatric symptoms related to Alzheimer's disease vary by the type of behavior and stage of dementia, but do not seem to be influenced by sex or level of premorbid intellectual impairment. Some psychiatric symptoms may be early indicators of Alzheimer's disease and may appear prior to substantial changes in daily functioning. Improvements in understanding the progression of dementia in individuals with Down syndrome may lead to improved diagnosis and treatment.

van Hoof, J., & Kort, H.
Supportive living environments: A first concept of a dwelling designed for older adults with dementia.
Dementia, 2009, 8, 293 - 316. doi:10.1177/1471301209103276
Abstract: The vast majority of older adults want to remain living independently at home, with or without a sufficient amount of professional home care, even when overall health is starting to decline. The ageing of society and the increase in the number of very old elders goes together with an increase in the number of people with dementia. About two thirds of the diagnosed people in the Netherlands live at home. Dementia has severe implications to the quality of daily life, in particular to independent functioning. This sets extra demands to living environments. Older adults with dementia and their partners ask for living environments that support independence, compensate for declining vitality, and lower the burden of family care. For this purpose, a first concept of a design for a dementia dwelling is presented in this paper, which incorporates modifications in terms of architecture, interior design, the indoor environment, and technological solutions. These design features were derived from literature search and focus group sessions. Current design guidelines are frequently based on practical experience only, and therefore, more systematic field research should be carried out to find evidence for the various design modifications. Also, it needs to be studied how the design features of the dementia dwelling can be incorporated into the existing housing stock.

van Hoof, J., Kort, H.S.M., van Waarde, H., & Blom, M.M.
Environmental interventions and the design of homes for older adults with dementia: An overview.
American Journal of Alzheimer's Disease & Other Dementias, 2010, 25(3), 202-232. doi:10.1177/1533317509358885
Abstract: In Western societies, the vast majority of people with dementia live at home and wish to remain doing so for as long as possible. Aging in place can be facilitated through a variety of environmental interventions, including home modifications. This article provides an overview of existing design principles and design goals, and environmental interventions implemented at home, based on literature study and additional focus group sessions. There is a multitude of design principles, design goals, and environmental interventions available to assist with activities of daily living and functions, although few systematic studies have been conducted on the efficacy of these goals and interventions. The own home seems to be a largely ignored territory in research and government policies, which implies that many problems concerning aging in place and environmental interventions for dementia are not adequately dealt with.

van Hoof, J., Kort, H.H., Duijnstee, M.S., Rutten, P.P., & Hensen, J.J.
The indoor environment and the integrated design of homes for older people with dementia.
Building and Environment, 2010, 45, 1244-1261. doi: 10.1016/J.BUILDENV.2009.11.008
Abstract: There are currently about 6 million - mainly older - people with dementia in the European Union. With ageing, a number of sensory changes occur. Dementia syndrome exacerbates the effects of these sensory changes and alters perception of stimuli. People with dementia have an altered sensitivity for indoor environmental conditions, which can induce problematic behavior with burdensome symptoms to both the person with dementia and the family carer. This paper, based on literature review, provides an overview of the indoor environmental parameters, as well as the integrated design and implementation of relevant building systems. The overview is presented in relation to the intrinsic ageing of senses, the responses of older people with dementia and the impact on other relevant stakeholders through the combined use of the International Classification of Functioning, Disability and Health, and the Model of Integrated Building Design. Results are presented as indicators of the basic value, functional value and economic value, as well as a synthesis of building-related solutions. Results can help designers and building services engineers to create optimal environmental conditions inside the living environments for people with dementia, and can be used to raise awareness among health care professionals about of the influence of the indoor environment on behavior of the person with dementia.

Vaughan, R.M., McGee, C., Guerin, S., Tyrrell, J., & Dodd, P.
The challenges of diagnosis and treatment of dementia in Down's syndrome
Irish Journal of Psychological Medicine, 2016, 33(3), 151-158
Abstract: This study analysed retrospective data on twenty adults with Down's syndrome (DS), who are clients of a specialist service in Dublin. The aim was to compare the practice of this service on diagnosis and treatment of dementia, with the consensus recommendations of the Royal College of Psychiatry, British Psychological Society and National Institute for Health and Care Excellence. Further aims were to establish average time to make a diagnosis and commence pharmacotherapy, and to describe tolerability to acetylcholinesterase inhibitors. It was found that screening for dementia did not take place before the age of 30yrs with the mean age for first assessment being 48yrs and average age at diagnosis being 51yrs. Average length of time from first identified symptoms to diagnosis was 1.3yrs. Of those diagnosed, 83% were prescribed acetylcholinesterase inhibitors but the authors were concerned at the continued use of the drug even when there appeared to be no benefit to the person. They found that a wide range of assessments were in use and that longitudinal assessment and follow up were not practiced. The authors recommend the streamlining of assessment tools and repeating assessment on a longitudinal basis.

Verbeek, H., van Rossum, E., Zwakhalen, S.M., Kempen, G.I., & Hamers, J.P.
Small, homelike care environments for older people with dementia: a literature review.
International Psychogeriatrics, 2009, 21(2), 252-264. doi: 10.1017/S104161020800820X.
Abstract: There is large cross-national variation in the characteristics of small, domestic-style care settings which emphasize normalized living. However, a

systematic overview of existing types is lacking. This study provides an international comparison of the care concepts which have adopted a homelike philosophy in a small-scale context. Insight into their characteristics is vital for theory, planning and implementation of such dementia care settings. A literature search was performed using various electronic databases, including PubMed, Medline, CINAHL and PsycINFO. In addition, "gray" literature was identified on the internet. Concepts were analyzed according to five main characteristics: physical setting, number of residents, residents' characteristics, domestic characteristics and care concept. 75 papers were included covering 11 different concept types in various countries. Similarities among concepts reflected a focus on meaningful activities centered around the daily household. Staff have integrated tasks and are part of the household, and archetypical home-style features, such as kitchens, are incorporated in the buildings. Differences among concepts were found mainly in the physical settings, numbers of residents and residents' characteristics. Some concepts have become regular dementia care settings, while others are smaller initiatives. The care concepts are implemented in various ways with a changing staff role. However, many aspects of these small, homelike facilities remain unclear. Future research is needed, focusing on residents' characteristics, family, staff and costs.

Verlinden, V.J.A., van der Geest, J.N., de Bruijn, R.F.A.G., Hofman, A., Koudstaal, P.J., & Ikram, M.A.
Trajectories of decline in cognition and daily functionng in preclinical dementia
Alzheimer's & Dementia, 2016, Feb, 12(2), 144-153.
doi: 10.1016/j.jalz.2015.08.001. Epub 2015 Sep 9.
Abstract: [*Note: this article refers to adults in the general population]* Although preclinical dementia is characterized by decline in cognition and daily functioning, little is known on their temporal sequence. We investigated trajectories of cognition and daily functioning in preclinical dementia, during 18 years of follow-up. In 856 dementia cases and 1712 controls, we repetitively assessed cognition and daily functioning with memory complaints, mini-mental state examination (MMSE), instrumental activities of daily living (IADL), and basic activities of daily living (BADL). Dementia cases first reported memory complaints 16 years before diagnosis, followed by decline in MMSE, IADL, and finally BADL. Vascular dementia related to earlier decline in daily functioning but later in cognition, compared with Alzheimer's disease. Higher education related to larger preclinical cognitive decline, whereas apolipoprotein E (APOE) e4 carriers declined less in daily functioning. These results emphasize the long hierarchical preclinical trajectory of functional decline in dementia. Furthermore, they show that various pathologic, environmental, and genetic factors may influence these trajectories of decline.

Videla, L., Benejam, B., Pegueroles, J., Carmona-Iragui, M., Padilla, C., Fernández, S., Barroeta, I., Altuna, M., Valldeneu, S., Garzón, D., Ribas, L., Montal, V., Arranz Martínez, J., Rozalem Aranha, M., Alcolea, D., Bejanin, A., Iulita, M.F., Videla Cés, S., Blesa, R., Lleó, A., & Fortea, J.
Longitudinal clinical and cognitive changes along the Alzheimer disease continuum in Down syndrome.
JAMA Netw Open. 2022 Aug 1;5(8):e2225573. doi: 10.1001/jamanetworkopen.2022.25573. PMID: 35930282; PMCID: PMC9356319.
Abstract: Alzheimer disease (AD) is the main medical problem in adults with Down syndrome (DS). However, the associations of age, intellectual disability (ID), and clinical status with progression and longitudinal cognitive decline have not been established. To examine clinical progression along the AD continuum and its related cognitive decline and to explore the presence of practice effects and floor effects with repeated assessments, authors used a single-center cohort study of adults (aged >18 years) with DS with different ID levels and at least 6 months of follow-up between November 2012 and December 2021. The data are from a population-based health plan designed to screen for AD in adults with DS in Catalonia, Spain. Individuals were classified as being asymptomatic, having prodromal AD, or having AD dementia. The main outcome was clinical change along the AD continuum. Cognitive decline was measured by the Cambridge Cognitive Examination for Older Adults With Down Syndrome and the modified Cued Recall Test. Authors assessed a total of 632 adults with DS (mean [SD] age, 42.6 [11.4] years; 292 women [46.2%]) with 2847 evaluations (mean [SD] follow-up, 28.8 [18.7] months). At baseline, there were 436 asymptomatic individuals, 69 patients with prodromal AD, and 127 with AD dementia. After 5 years of follow-up, 17.1% (95% CI, 12.5%-21.5%) of asymptomatic individuals progressed to symptomatic AD in an age-dependent manner (0.6% [95% CI, 0%-1.8%] for age <40 years; 21.1% [95% CI, 8.0%-32.5%] for age 40-44 years; 41.4% [95% CI, 23.1%-55.3%] for age 45-49 years; 57.5% [95% CI, 38.2%-70.8%] for age ?50 years; P < .001), and 94.1% (95% CI, 84.6%-98.0%) of patients with prodromal AD progressed to dementia with no age dependency. Cognitive decline in the older individuals was most common among those who progressed to symptomatic AD and symptomatic individuals themselves. Importantly, individuals with mild and moderate ID had no differences in longitudinal cognitive decline despite having different performance at baseline. This study also found practice and floor effects, which obscured the assessment of longitudinal cognitive decline. This study found an association between the development of symptomatic AD and a high risk of progressive cognitive decline among patients with DS. These results support the need for population health plans to screen for AD-related cognitive decline from the fourth decade of life and provide important longitudinal data to inform clinical trials in adults with DS to prevent AD.

Visootsak, J., & Sherman, S.
Neuropsychiatric and behavioral aspects of trisomy 21
Current Psychiatry Reports, 2007, 9(2), 135-140.
Abstract: Down syndrome (DS), or trisomy 21, is the most common identifiable genetic cause of mental retardation. The syndrome is unique with respect to its cognitive, behavioral, and psychiatric profiles. The well-known cheerful and friendly demeanor often creates a personality stereotype, with parents and observers commenting on the positive attributes. Despite these strengths, approximately 20% to 40% of children with DS have recognized behavioral problems. Such problems persist through adulthood, with a decrease in externalizing symptoms of aggressiveness and attention problems and the emergence of internalizing symptoms of depression and loneliness. In adulthood, the presence of early-onset dementia of the Alzheimer type and cognitive decline may pose a challenge in recognizing these internalizing symptoms. Understanding the age-related changes in cognitive functioning and behavioral profiles in individuals with DS provides insight into clinical and treatment implications.

Visser, F.E., Aldenkamp, A.P., van Huggelen, A.C., Kuilman, M., Overweg, J., & van Wijk, J.
Prospective study of the prevalence of Alzheimer-type dementia in institutionalized individuals with Down syndrome
*American Journal on Mental Retardation,*1997, Jan, 101(4), 400-412.
https://pubmed.ncbi.nlm.nih.gov/9017086/
Abstract: Institutionalized patients with Down syndrome (n = 307) were monitored for 5 to 10 years prospectively to determine prevalence of Alzheimer-type dementia. Clinical signs, cognitive functioning, and EEGs were assessed. When possible, postmortem neuropathological examinations were conducted. Progressive mental and physical deterioration was found for 56 of the residents. **Mean age at onset of dementia was 56 years**. Prevalence increased from 11% between ages 40 and 49 to 77% between 60 and 69. All patients 70 and over had dementia. Neuropathological findings were consistent with clinical diagnosis. Use of a dementia checklist, cognitive skills inventory, and EEG reliably detected Alzheimer-type dementia at an early stage.

Walaszek, A., Schroeder, M., Krainer, J., Pritchett, G., Wilcenski, M., Endicott, S. , Albrecht, T., Carlsson, C.M., & Mahoney, J.
Effectively training professional caregivers to screen and refer persons with dementia and intellectual/developmental disability
AAIC 2020 Conference, Poster presentation, July 30. 2020. *Alzheimer's & Dementia*, 16(S8), First published: 07 December 2020.
https://doi.org/10.1002/alz.037966

Abstract: By age 40, almost all people with Down syndrome, the most common cause of intellectual/developmental disability (I/DD), have neuropathological changes consistent with Alzheimer's disease; by age 60, about half have dementia. Detecting dementia in persons with I/DD can be challenging because baseline cognitive impairment can be severe and because persons with I/DD may have difficulty reporting symptoms. The National Task Group Early Detection Screen for Dementia (NTG-EDSD) was developed to aid detection of cognitive impairment in adults with I/DD. We implemented an educational curriculum to increase the ability of professional caregivers to screen for dementia in persons with I/DD using the NTG-EDSD. In November 2018 to April 2019, we held five training sessions for professional caregivers of persons with I/DD, partnering with various managed care organizations (MCO), aging and disability resource centers, adult day programs, and adult family homes. We assessed knowledge and attitudes at baseline, immediately after training, and one week, one month and six months after training. Participants (N=154) included direct care workers, case managers, healthcare providers, and other social services staff. Participants reported a marked increase in confidence in their ability to detect changes associated with mild cognitive impairment or dementia ($p<0.001$), decline in activities of daily living ($p=0.02$), and changes in behavior and affect ($p<0.001$). Satisfaction with the training was very high, and 94.0% of participants agreed or strongly agreed they could use the NTG-EDSD tool with their clients. Following the training, one MCO we partnered with, serving 62 of 72 counties in Wisconsin, made the NTG-EDSD a standard part of the assessment of adults with Down syndrome starting at age 40. Authors note that a wide variety of social services and healthcare professionals can be effectively trained to screen for dementia in persons with I/DD using a standardized screening tool, the NTG-EDSD. Satisfaction with the training was high, and use of the NTG-EDSD was thought to be feasible. This educational intervention led to change in practice at a systems level within an MCO. Next steps could include assessing impact of such training on the quality of life and healthcare outcomes of persons with I/DD.

Walaszek, A., Albrecht, T., LeCaire, T., Sayavedra, N., Schroeder, M., Krainer, J., Prichett, G., Wilcenski, M., Endicott, S., Russmann, S., Carlsson, C.M., & Mahoney, J. Training professional caregivers to screen for report of cognitive changes in persons with intellectual disability.
Alzheimer's & Dementia: Translational Research & Clinical Interventions, 2022 Aug, 8(1), 1-10, e12345 https://doi.org/10.1002/trc2.12345
Abstract: By age 60, 60% of adults with Down syndrome (DS) have dementia. Detecting dementia in persons with intellectual disability (ID) can be challenging because their underlying cognitive impairment can confound presentation of dementia symptoms and because adults with ID may have difficulty reporting symptoms. The National Task Group Early Detection Screen for Dementia (NTG-EDSD) was developed to aid detection of report of cognitive impairment in adults with ID. We implemented an educational curriculum using the NTG-EDSD and evaluated the impact of the intervention on professional caregivers' self-assessed capacity to identify persons with ID and dementia. We held five in-person training sessions for professional caregivers of persons with ID, partnering with various managed care organizations and social services agencies. We assessed knowledge and attitudes at baseline; immediately after training; and 1 week, 1 month, and 6 months after training. A total of 154 direct care workers, case managers, health-care providers, and other social services staff attended the trainings. Satisfaction with the NTG-EDSD training was high; 94% of attendees agreed or strongly agreed that they could use the NTG-EDSD with their clients. After training, attendees reported a marked increase in confidence in their ability to track various health circumstances and detect functional decline in their clients, although some gains were not sustained over time. As a result of the training, one managed care organization made the NTG-EDSD a standard part of its assessment of adults with DS starting at age 40. Social services and health-care professionals can learn to document signs of cognitive decline in adults with ID using the NTG-EDSD. Attendees were highly satisfied with the training, experienced an increase in confidence in their care of persons with ID, and found the NTG- EDSD feasible to use. Because not all gains were sustained over time, booster trainings may be necessary.

Walker, B., MacBryer, S., Jones, A., & Law, J.
Interinformant agreement of the dementia questionnaire for people with learning disabilities
British Journal of Learning Disabilities, 2015, Sept, 43(3), 227-233..
https://doi.org/10.1111/bld.12102
Abstract: Because of difficulties with neuropsychological assessments for dementia in people with intellectual disabilities, professionals in clinical practice have relied heavily on carer interviews, one of the most widely used being the Dementia Questionnaire for People with Learning Disabilities (DLD – Evenhuis et al. 2006 Dementia questionnaire for people with intellectual disabilities manual (second edition). Amsterdam, Netherlands, Harcourt Test Publishers). Because dementia is indicated by the magnitude of changes in scores between longitudinal assessments, interinformant agreement is paramount. We carried out the DLD interview independently with two carers for each of 26 people with Down syndrome. Only 15% of pairs of carers achieved 'good' agreement. Levels of agreement varied widely across the DLD subscales and individual questions. Interinformant agreement was better for less able people with Down syndrome than for more able individuals.

📖 **Walker, C.A., & Walker, A.**
Uncertain Futures: people with learning difficulties and their ageing family carers
54 pp.
Brighton, UK: Pavilion Publishing (1998)
Abstract: This monograph provides an overview of research, policy and practice relating to service responses to adults with learning difficulties living at home with older family carers in the UK. The authors' premise is that as life expectancy increases, a growing proportion of people with learning difficulties continues to live with family members, most frequently parents, whose caring role is being extended into their own advanced old age. Highlighted are some of the issues raised by service users, carers and service providers, including care for someone with diminishing abilities. The text argues that there is urgent need for the paid service sector to work with families to provide the necessary support and planning to take the uncertainty out of the future.

Wallace, E., Harp, J., Van Pelt,K.I., Koehl, L., Caban-Holt, .A.M,., Anderson-Mooney, A.J., Robertson, W., Lightner, D., Jicha, G.A., Head, E., & A Schmitt, F.A.
Validity of the Severe Impairment Battery, Brief Praxis Test, and Dementia Questionnaire for Persons with Intellectual Disabilities in differentiating dementia status in individuals with Down syndrome
AAIC2020, Poster presentation, July 29, 2020. *Alzheimer's & Dementia*, 16 (S6), First published: 07 December 2020. https://doi.org/10.1002/alz.044227
Abstract: Individuals with Down syndrome (DS) are at high risk for dementia, specifically Alzheimer's disease (AD). However, many measures regularly used for the detection of AD in the general population are not suitable for individuals with DS. Some measures, including the Severe Impairment Battery (SIB), Brief Praxis Test (BPT), and Dementia Questionnaire for Persons with Intellectual Disabilities (DMR), have been used in clinical trials and other research with this population. Validity research is limited, however, particularly regarding identification of predementia symptoms in the DS population. The current project presents baseline cross-sectional SIB, BPT, and DMR performance in order to characterize their ability to discriminate normal cognition, possible AD, and probable AD in DS. Baseline SIB, BPT, and DMR performances from 117 individuals were analyzed as part of a large longitudinal cohort of aging individuals with DS. Receiver operating characteristic (ROC) curves were calculated to investigate accuracy in differentiating levels of dementia status. In comparing no/possible AD vs. probable AD, the SIB and BPT exhibited fair discrimination ability (AUC = .78 and .79, respectively). In comparing no/possible AD vs. probable AD, the DMR exhibited good discrimination ability (AUC = .89), with qualitatively similar performance of the DMR-Cognitive and DMR-Social subscales (AUC = .89 and .83, respectively). In comparing no AD vs. possible AD, the SIB and BPT failed to differentiate these groups (AUC = .53 and .55, respectively), whereas the DMR exhibited good differentiation (AUC = .80). Au thors note that the results suggest that the SIB, BPT, and DMR are able to

discriminate between levels of dementia status in individuals with DS, supporting their continued use in the clinical assessment of dementia in DS. Specifically, the DMR, based on informant ratings of social and cognitive behaviors of daily living, outperformed the SIB and BPT, tests of cognitive performance, in discriminating no/possible AD vs. probable AD as well as no AD vs. possible AD. Such findings suggest that the DMR is better equipped to identify symptoms of overt dementia as well as predementia in this population. Findings reinforce the importance of including informant behavior ratings in assessment of this population.

Walsh, D.M., Doran, E., Silverman, W., Tournay, A., Movsesyan, N., & Lott, I.T.
Rapid assessment of cognitive function in down syndrome across intellectual level and dementia status
Journal of Intellectual Disability Research, 2015, Nov, 59(11), 1071-1079. doi:10.1111/jir.12200. Epub 2015 May 29.
Abstract: Adults with Down syndrome (DS) are at risk of developing dementia and cognitive assessment is a fundamental part of the diagnostic process. Previously, we developed a Rapid Assessment for Developmental Disabilities (RADD), a brief, broadly focused direct test of cognition. In the current report, we assess whether the RADD is sensitive to dementia in DS and the degree to which it compares with other cognitive measures of dementia in this population. In a sample of 114 individuals with DS, with dementia diagnosed in 62%, the RADD was compared with the Dementia Questionnaire for Mentally Retarded Persons (DMR), the Bristol Activities of Daily Living Scale, Severe Impairment Battery (SIB), and the Brief Praxis Test (BPT). The RADD showed predicted effects across intellectual disability (ID) levels and dementia status ($p < 0.001$). Six-month test-retest reliability for the subset of individuals without dementia was high ($r(41) = 0.95$, $p < 0.001$). Criterion-referenced validity was demonstrated by correlations between RADD scores and ID levels based upon prior intelligence testing and clinical diagnoses ($rs (114) = 0.67$, $p = 0.001$) and with other measures of cognitive skills, such as the BPT, SIB, and DMR-Sum of Cognitive scores (range 0.84 through 0.92). Using receiver operating characteristic curves for groups varying in pre-morbid severity of ID, the RADD exhibited high sensitivity (0.87) and specificity (0.81) in discriminating among individuals with and without dementia, although sensitivity was somewhat lower (0.73) for the subsample of dementia cases diagnosed no more than 2 years prior to their RADD assessment. Taken together, findings indicated that the RADD, a relatively brief, easy-to-administer test for cognitive function assessment across ID levels and dementia status, would be a useful component of cognitive assessments for adults with DS, including assessments explicitly focused on dementia.

Waninge, A., Wissing, M., Hobbelen, H., Fokkens, A., Dekker, A., & De Deyn, P.
Dementia in people with severe/profound intellectual disabilities
Journal of Applied Research in Intellectual Disabilities, 2021, 34(5), 1214-1215. https://doi.org/10.1111/jar.12917
Abstract: In people with severe or profound intellectual disabilities, it is difficult to diagnose dementia. As timely identification and diagnosis of dementia allows for a timely response to changing client wishes and needs, this study examined symptoms, and diagnosis of dementia in practice. Family members and professionals were invited to fill out a survey about symptoms and diagnosis of dementia in people with severe or profound intellectual disabilities. Results of the survey were further explored within semi-structured interviews with professionals having experience with signaling and diagnosing dementia in these people. Symptoms found in the survey and transcripts of the interviews were qualitatively analyzed, using thematic analyses based on a developed symptom-matrix. The survey was filled out completely by 14 family members and 90 professionals with different backgrounds. Results showed that behavioral changes were recognized more frequently than cognitive decline. Compared to those without dementia, epilepsy and motor decline were more present in case of dementia. Fifteen interviews (until saturation) with professionals provided an in-depth view into the symptoms, and how to identify them, again stressing behavioral alterations and to a lesser extent cognitive symptoms.

Ward, S., Opie, J., O'Connor, D.W.
Family carer's responses to behavioural and psychological symptoms of dementia
International Journal of Geriatric Psychiatry, 2003, Nov, 18(11), 1007-1012. doi: 10.1002/gps.1005.
Abstract: The author sought to describe the responses of family carers to the behavioural and psychological symptoms associated with dementia. Thirty family carers of people with dementia were identified in a survey of mental disorder in general practice. Another 20 were referred by local aged mental health services. Carers were interviewed using the Manchester and Oxford University Scale for the Psychopathological Assessment of Dementia (MOUSEPAD) which rates behavioural and psychological disturbances. Carers' customary responses to current symptoms were recorded verbatim and categorised using a structured typology. Symptom frequency increased in line with dementia severity. Disturbances were generally well tolerated. Most were ignored where possible, except for wandering from home. Other common responses included avoiding triggers, providing reassurance, reality orientation, diversion, and collusion with false beliefs. Restrictive or punitive responses were uncommon. Few carers articulated clear strategies to deal with behavioural and psychological symptoms. For most, tolerance proved more effective and less distressing than arguments and reprimands. Carers' responses are likely to be influenced by social and cultural factors and may differ in other settings.

Wark, S., Hussain, R., & Parmenter, T.
Down syndrome and dementia: Is depression a confounder for accurate diagnosis and treatment?
Journal of Intellectual Disabilities, 2014, 18(4), 305-314. doi: 10.1177/1744629514552152.
Abstract: The past century has seen a dramatic improvement in the life expectancy of people with Down syndrome. However, research has shown that individuals with Down syndrome now have an increased likelihood of early onset dementia. They are more likely than their mainstream peers to experience other significant co-morbidities including mental health issues such as depression. This case study reports a phenomenon in which three individuals with Down syndrome and dementia are described as experiencing a rebound in their functioning after a clear and sustained period of decline. It is hypothesized that this phenomenon is not actually a reversal of the expected dementia trajectory but is an undiagnosed depression exaggerating the true level of functional decline associated with the dementia. The proactive identification and treatment of depressive symptoms may therefore increase the quality of life of some people with Down syndrome and dementia.

Watchman, K., Kerr, D., & Wilkinson, H.
Supporting Derek: A new resource for staff working with people who have a learning difficulty and dementia.
58 pp.
York, United Kingdom: Joseph Rountree Foundation (2010)
Access: http://www.jrf.org.uk/publications/supporting-derek
Abstract: This resource pack published by the Joseph Rowntree Foundation in partnership with the University of Edinburgh, is aimed at staff supporting people with intellectual disability who develop dementia. Its focus in on helping care staff and training officers from intellectual disability and dementia care settings, as well as community, housing and health care staff. The pack is composed of 10 topic area (chapters), including basics on dementia, understanding behavior, development care environments, pain, communication, meaningful activities, friends with dementia, nutrition and hydration, night-time care, and palliative care. The pack includes a DVD and training materials which cover many of the key issues related to diagnosing and responding to dementia in people with intellectual disabilities. A short drama included on the DVD (acted by people with an intellectual disability) provides an insight into the reality of dementia and how it

might feel to the individual affected.

Watchman, K.
Critical issues for service planners and providers of care for people with Down's syndrome and dementia.
British Journal of Learning Disabilities, 2003, 31(2), 81-84.
https://doi.org/10.1046/j.1468-3156.2003.00228.x
Abstract: This discussion paper raises critical issues that need to be addressed along with suggestions as to how they may be met with. Author notes that the role of service planners and providers of care is one that cannot be understated while considering the future needs of people with Down's syndrome and dementia. Discussed are appropriateness of accommodations, care management, diagnosis, and training.

Watchman, K.
Why wait for dementia?
Journal of Learning Disabilities, 2003, 7, 221-230.
https://doi.org/10.1177/14690047030073
Abstract: Adults with Down syndrome living in supported accommodation, who develop dementia, may also experience other preventable difficulties caused by the environment in which they live. This can result in their enforced move to a different accommodation. Yet it is known that it is beneficial for people with intellectual disabilities and dementia to remain in familiar surroundings for as long as possible. This article puts forward a new set of guidelines suggesting the modification of the living environment of adults with Down syndrome before they develop dementia. The guidelines are discussed along with possible barriers to their implementation.

Watchman, K.
Practitioner-raised issues and end-of-life care for adults with Down syndrome and dementia
Journal of Policy and Practice in Intellectual Disabilities, 2005, 2(2), 156-162.
https://doi.org/10.1111/j.1741-1130.2005.00026.x
Abstract: The author interviewed a small group of practitioners working in intellectual disability and palliative care settings about their perceptions of a number of end-of-life issues related to people with Down syndrome who were affected by dementia. The study, which took place in Scotland, identifed a number of issues and perceptions expressed by the subjects as well as gaps in services and practice. Key among the findings were the need for people with Down syndrome to be more involved in planning for their own end-of-life care; a lack of communication between those persons working in palliative care and intellectual disability settings; identifcation of a “care culture clash;” deficits in training programs for staff involving dying, death, and bereavement; and that end-of-life care for people with Down syndrome and dementia is a neglected area of research. The author highlights the lack of uniform practice when working with people with Down syndrome in the end stages of dementia and provides some recommendations for further discourse and research.

Watchman, K.
Changes in accommodation experienced by people with Down syndrome and dementia in the first five years after diagnosis
Journal of Policy and Practice in Intellectual Disabilities, 2008, 5(1), 65-68.
Abstract: Research that has tracked living situation changes is lacking for people with Down syndrome post-diagnosis of dementia. Extant studies have not considered reasons for a move, the stage at which it happened, and how involved in the decision the person with Down syndrome was. To study this, a postal questionnaire was used with 35 carers of persons with Down syndrome who had been diagnosed with dementia during the previous five years. Results showed that there are fewer accommodation changes in the early stages of dementia among people with Down syndrome than have previously been suspected and that confusion exists over the interpretation of existing care models. Findings also revealed that adults with Down syndrome were often denied the opportunity to take part in discussions about their future accommodations and there was a lack of forward planning on the part of carers.

Watchman, K.
People with a learning disability and dementia: reducing marginalisation
Journal of Dementia Care [Research Review], 2012, 20(5), 34- 39.
http://www.learningdisabilityanddementia.org/uploads/1/1/5/8/11581920/journal_of_dementia_care.pdf
Abstract: The awareness that people with a learning disability, particularly Down’s syndrome, are at risk of dementia at a younger age brings an associated need for clarity over service planning and delivery. In order to record changes and developments in approaches, research literature documents the changing history of people with a learning disability and, separately, people with dementia. We not have the same knowledge about the most appropriate ways of supporting individuals who have both a learning disability and dementia. People will already experience social exclusion due to society’s interpretation of their learning disability and this review identifies from the literature factors that have contributed to the further marginalization of this group. The review highlights the need for accurate data and statistics, an individualized approach to sharing information about the diagnosis, general and specialist training, an increased use of adapting methods of communication as dementia progresses and a consistent staff approach across care settings.

📖 **Watchman, K**.
Intellectual Disability and Dementia: Research into Practice.
336 pp.
London/Philadelphia: Jessica Kingsley Publishers (2014).
Abstract: In 16 chapters, this edited text offers a balanced appraisal of the evidence base on people with intellectual disabilities who develop dementia. It includes a range of resources, and is split into three sections that address the following: (1) The association between intellectual disabilities and dementia: what do we know? (2) Experiences of dementia in people with intellectual disabilities: how do we know?, and (3) Service planning: what are we going to do? Section one explores issues such as defining and diagnosing dementia in people with intellectual disabilities, prevalence and incidence and treatment options. The authors explain the differing theories about why people with Down’s syndrome are more likely to experience dementia, which provides a useful foundation for discussions about the use of medication. Section two explores the perspectives of people with learning disabilities and their families and the experiences of families via case studies. This section also explores some checklists for use with family members to help plan for the future. Section three focuses on service planning by describing a framework that can be used by practitioners for discussing diagnosis and prognosis of dementia. This section also considers the issues related to ageing in place and dementia-specific services and suggests that training is important for staff supporting those with learning disabilities and dementia.

Watchman, K., Janicki, M.P., and the members of the International Summit on Intellectual Disability and Dementia
The intersection of intellectual disability and dementia: Report of the international summit on intellectual disability and dementia
Gerontologist, 2019, 59(3), 411-419. doi: 10.1093/geront/gnx160.
Abstract: An International Summit on Intellectual Disability and Dementia, held in Glasgow, Scotland (October 13-14, 2016) drew individuals and representatives of numerous international and national organizations and universities with a stake in issues affecting adults with intellectual disability (ID) affected by dementia. A discussion-based consensus process was used to examine and produce a series of topical reports examining three main conceptual areas: (1) human rights and personal resources (applications of the Convention for Rights of People with Disabilities and human rights to societal inclusion, and perspectives of persons with ID), (2) individualized services and clinical supports (advancing and advanced dementia, post-diagnostic supports, community supports and services, dementia-capable care practice, and end-of-life care practices), and (3) advocacy, public impact, family caregiver issues (nomenclature/ terminology, inclusion of persons with ID in national plans, and family caregiver issues). Outcomes included recommendations incorporated into a series of publications and topical summary bulletins designed to be international resources, practice

guidelines, and the impetus for planning and advocacy with, and on behalf of, people with ID affected by dementia, as well as their families. The general themes of the conceptual areas are discussed and the main recommendations are associated with three primary concerns.

Watchman, K., Janicki, M.P., Splaine, M., Larsen, F.K., Gomiero, T., & Lucchino, R.
International summit consensus statement: Intellectual disability inclusion in national dementia plans.
American Journal of Alzheimer's Disease & Other Dementias, 2017, Jun, 32(4), 230-237 https://doi.org/10.1177/1533317517704082
Abstract: The World Health Organization (WHO) has called for the development and adoption of national plans or strategies to guide public policy and set goals for services, supports, and research related to dementia. It called for distinct populations to be included within national plans, including adults with intellectual disability (ID). Inclusion of this group is important as having Down's syndrome is a significant risk factor for early-onset dementia. Adults with other ID may have specific needs for dementia-related care that, if unmet, can lead to diminished quality of old age. An International Summit on Intellectual Disability and Dementia, held in Scotland, reviewed the inclusion of ID in national plans and recommended that inclusion goes beyond just description and relevance of ID. Reviews of national plans and reports on dementia show minimal consideration of ID and the challenges that carers face. The Summit recommended that persons with ID, as well as family carers, should be included in consultation processes, and greater advocacy is required from national organizations on behalf of families, with need for an infrastructure in health and social care that supports quality care for dementia.

Watchman, K., Janicki, M.P., Udell, L., Hogan, M., Quinn, S., Beránková, A.
Consensus statement of the International Summit on Intellectual Disability and Dementia on valuing the perspectives of persons with intellectual disability.
Journal of Intellectual Disabilities,2019 Jun;23(2):266-280. doi: 10.1177/1744629517751817. Epub 2018 Jan 17.
Abstract: The International Summit on Intellectual Disability and Dementia covered a range of issues related to dementia and intellectual disability, including the dearth of personal reflections of persons with intellectual disability affected by dementia. This article reflects on this deficiency and explores some of the personal perspectives gleaned from the literature, from the Summit attendees and from the experiences of persons with intellectual disability recorded or scribed in advance of the two-day Summit meeting. Systemic recommendations included reinforcing the value of the involvement of persons with intellectual disability in (a) research alongside removing barriers to inclusion posed by institutional/ethics review boards, (b) planning groups that establish supports for dementia and (c) peer support. Practice recommendations included (a) valuing personal perspectives in decision-making, (b) enabling peer-to-peer support models.

Watchman, K., Mattheys, K., & McKernon, M.
Effects of the implementation of non-drug Interventions on behaviour and psychosocial symptoms of dementia in people who have an Intellectual disability
Journal of Intellectual Disability Research, 2019, 63(8), 647.
Abstract: This three-year study investigates if non-drug interventions result in positive changes in behaviour associated with dementia in people with intellectual disability. People with intellectual disability are involved as advisors (n = 1) and co-researchers (n = 4) in both cycles. Cycle 1 (concluded) included 7 participants with intellectual disability in the early stage of dementia (4 with Down syndrome) and 12 support staff. Cycle 2 (ongoing) includes participants who have a more profound intellectual disability, and/or are experiencing advanced dementia. In both cycles, a goal-setting tool firstly helped to identify individualised non-drug interventions. In Cycle 1, a pre- and post-behaviour change tool (NPI-Q), was completed alongside semi-structured interviews, a bespoke tool to measure 'in the moment' changes, intervention diaries, and photovoice. Cycle 1 interventions included reminiscence, life story, music playlists, cookery, aromatherapy, environmental design change, exercise and cognitive games. Of 239 separate intervention over a 6-month period in Cycle 1, 193 resulted in positive behaviour change with 75% of goals being achieved or exceeded. The study offers insight into the support of people with intellectual disability and dementia. Use of non-drug supports in response to distress has led to cultural change within participating organizations with less reliance on medication as a first response.

Watchman, K., McKernon, M., Boustead, L., & Doyle, A.
Intellectual disability and dementia: Understanding the effectiveness of psychosocial interventions
Journal of Applied Research in Intellectual Disabilities, 2021, 34(5),1273. https://onlinelibrary.wiley.com/doi/epdf/10.1111/jar.12763
Abstract: The study aim was to identify effectiveness of psychosocial interventions with people who have an intellectual disability and dementia. This mixed-method participatory action study used goal-setting theory with 16 participants with intellectual disability and dementia, and 22 social care staff across 11 sites. Five core researchers with intellectual disability were part of an inclusive research team collecting data using existing and bespoke tools, including photovoice. Psychosocial interventions included: music playlists, reminiscence, animal therapy, robotic animals, and design changes. Analysis used descriptive and inferential statistics and framework analysis. Found that 74% of individual goals met or exceeded expectations with reduction in some "as required" medication. Qualitative findings include themes of enabling care and interventions as tools for practice. Photovoice provided insight into previously unreported fears about dementia. This poster combines an easy-read and pictorial summary of the study. Recommendations are made to maximise wellbeing and ensure the perspectives of people with dementia are heard: medication review, design changes to the home of a person with dementia, staff training, and talking about dementia more with people who have an intellectual disability. Individualized psychosocial interventions have potential to reduce distress or agitation in persons with intellectual disability and dementia, and to increase quality of life.

Warner, M.L.
The complete guide to Alzheimer's-proofing your home.
470 pp.
West Lafayette, Indiana: Purdue University Press (1998)
Abstract: General text on adapting homes and living environments for persons with dementia; applicable to home and other residential situations for adults with intellectual disabilities and dementia.

Webber, R., Bowers, B., & Bigby, C
Confidence of group home staff in supporting the health needs of older residents with intellectual disability
Journal of Intellectual and Developmental Disabilities, 2016, 41(2), 107-114. doi.org/10.3109/13668250.2015.1130218
Abstract: Increased life expectancy for people with intellectual disability is accompanied by increased age-related health concerns. People ageing with intellectual disability experience more health conditions and are relocated to aged care earlier than their age peers. Group home staff were surveyed about their (a) training and confidence in 11 health conditions and 7 health procedures, and (b) attitude to relocating residents with health needs to aged care. Staff training in each of 10 health conditions and 7 health procedures was positively associated with increased confidence in supporting residents with those health issues. Higher staff confidence in caring for residents with 9 conditions and requiring 4 procedures was negatively associated with a likelihood of recommending that a person with those health needs should relocate to aged care. Targeted training of staff in age-related health issues may contribute to better health care and delay residents relocating to aged care.

Webber, R., Bowers, B. & McKenzie-Green, B.
Staff responses to age-related health changes in people with an intellectual disability in group homes.
Disability and Society, 2010, 25(6), 657-671.

https://doi.org/10.1080/09687599.2010.505736
Abstract: The purpose of this study was to explore how supervisors in group homes caring for people with intellectual disability responded to the development of age-related health changes in their residents. Ten group home supervisors working in the disability sector were interviewed once. Data were analyzed using Dimensional Analysis. The study identified several factors related to whether a resident could stay 'at home' or would need to be moved to residential aged care (nursing home) including: nature and extent of group home resources, group home staff comfort with residents' health changes, staff skill at navigating the intersection between the disability and ageing sectors, and the supervisor's philosophy of care. The ability of older people with an intellectual disability to 'age in place' is affected by staff knowledge about and comfort with age-related illnesses, staff skills at navigating formal services, staffing flexibility, and the philosophy of group home supervisors. Despite the growing international concern for the rights of people with disability, particularly in relation to decision making, questions about the older person's choice of residence and participation in decision making about what was best for them, were almost nonexistent. Rather, decisions were made based on what was considered to be in 'the best interest.

Whitehouse, R., Chamberlain, P., & Tunna, K.
Dementia in people with learning disability: a preliminary study into care staff knowledge and attributions
British Journal of Learning Disabilities, 2000, 28(4) 148- 153.
https://doi.org/10.1046/j.1468-3156.2000.00057.x
Abstract: This paper describes the findings of a pilot study funded by the NHS Executive Primary and Community Care Research Initiative Small Projects Scheme that investigated the knowledge and attributions of dementia held by care staff who work with older adults with learning disability. Meetings took place with 21 members of care staff identified as “keyworkers” to older adults with learning disability living in residential houses provided by Solihull Healthcare NHS Trust, Solihull, UK. The results suggest that staff have knowledge of ageing at a similar level to that of college students. Forgetfulness was the sign that they would most expect to see if they thought someone was suffering from dementia. When a change in behavior was attributed to dementia, it was most likely to be viewed as 'stable, uncontrollable' with staff feeling pessimistic about being able to change the behavior.

Whittick, J.E.
Dementia and mental handicap: attitudes, emotional distress and caregiving
British Journal of Medical Psychology, 1989, 62, 181-189. doi: 10.1111/j.2044-8341.1989.tb02825.x.
Abstract: Against the current climate of hospital closure programs and community care, attitudes to caregiving were examined in three groups of carers, namely mothers caring for a mentally handicapped child, mothers caring for a mentally handicapped adult and daughters caring for a parent with dementia. An 'attitude questionnaire' was developed by the author and administered, postally, to the three groups. Daughters were found to be more likely than the mothers to see their caring role in a negative way and were more inclined to favor institutional care. Possible reasons for this are discussed. The relationship between attitudes and emotional distress (as measured by the GHQ-30) were also examined for the sample as a whole. Negative and pro-institutional attitudes towards the caregiving situation were associated with elevated levels of emotional distress. Implications at both a local and a national level for all those involved with carers are discussed in the light of these findings.

Whitwham, S., McBrien, J., & Broom, W.
Should we refer for a dementia assessment? A checklist to help know when to be concerned about dementia in adults with Down syndrome and other intellectual disabilities.
British Journal of Learning Disabilities, 2011, 39(1), 17-21.
https://doi.org/10.1111/j.1468-3156.2009.00606.x
Abstract: The aim of this research was to develop a simple screening checklist to help carers and professionals know when to make a referral for a dementia assessment. A checklist was completed for all new referrals to a dementia service for people with intellectual disabilities. The obtained scores were compared to the diagnostic outcome of a comprehensive dementia assessment. The data (n = 159) indicate a higher score on the checklist correlates significantly with a subsequent diagnosis of dementia. Cut-off scores are explored. The checklist appears to be a useful tool to prompt referrals for a full dementia assessment. By helping the referrer to know when to be concerned about dementia, it may reduce the number of people referred late or not at all.

Wiener, J.M., & Pazzaglia, F.
Ageing- and dementia-friendly design: theory and evidence from cognitive psychology, neuropsychology and environmental psychology can contribute to design guidelines that minimise spatial disorientation.
Cognitive Processing, 2021, 22, 715-730 (2021).
https://doi.org/10.1007/s10339-021-01031-8
Abstract: Many older people, both with and without dementia, eventually move from their familiar home environments into unfamiliar surroundings, such as sheltered housing or care homes. Age-related declines in wayfinding skills can make it difficult to learn to navigate in these new, unfamiliar environments. To facilitate the transition to their new accommodation, it is therefore important to develop retirement complexes and care homes specifically designed to reduce the wayfinding difficulties of older people and those with Alzheimer's disease (AD). Residential complexes that are designed to support spatial orientation and that compensate for impaired navigation abilities would make it easier for people with dementia to adapt to their new living environment. This would improve the independence, quality of life and well-being of residents, and reduce the caregivers' workload. Based on these premises, this opinion paper considers how evidence from cognitive psychology, neuropsychology and environmental psychology can contribute to ageing- and dementia-friendly design with a view to minimising spatial disorientation. After an introduction of the cognitive mechanisms and processes involved in spatial navigation, and the changes that occur in typical and atypical ageing, research from the field of environmental psychology is considered, highlighting design factors likely to facilitate (or impair) indoor wayfinding in complex buildings. Finally, psychological theories and design knowledge are combined to suggest ageing- and dementia-friendly design guidelines that aim to minimize spatial disorientation by focusing on residual navigation skills.

Wiese, M., Stancliffe, R.J., Dew, A., Balandin, S., & Howarth, G.
What is talked about? Community living staff experiences of talking with older people with intellectual disability about dying and death
Journal of Intellectual Disability Research, 2014 Jul, 58(7), 679-690. doi: 10.1111/jir.12065
Abstract: This study explored what community living staff talked about and did with people with intellectual disability (ID) to assistthem to understand dying and death. Guided by grounded theory methodology,focus groups and one-to-one interviews were conducted with 22 staff who had talked about any topic relating to dying and death with their clients. There was little evidence that staff talked with, or did things with clients to assist understanding of the end of life, both prior to and after a death. Prior to death staff assisted clients in a limited way to understand about determining wishes in preparation for death, and what dying looks like by observance of its passage. Following a death staff offered limited assistance to clients to understand the immutability of death, and how thedead can be honored with ritual, and remembered.
The findings have implications for why people with ID have only partial understanding of the end of life, the staff skills required to support clients’ understanding, and when conversations about the end of life should occur

Wilkinson, H., Janicki, M.P., & Edinburgh Working Group on Dementia Care Practices (EWGDCP).
The Edinburgh Principles with accompanying guidelines and recommendations.
Journal of Intellectual Disability Research, 2002, 46, 279-284. doi: 10.1046/j.1365-2788.2002.00393.x.

Abstract: A panel of experts attending a 3-day meeting held in Edinburgh, UK, in February 2001 was charged with producing a set of principles outlining the rights and needs of people with intellectual disability (ID) and dementia, and defining service practices which would enhance the supports available to them. The Edinburgh Principles, seven statements identifying a foundation for the design and support of services to people with ID affected by dementia, and their carers, were the outcome of this meeting. The accompanying guidelines and recommendations document provides an elaboration of the key points associated with the Principles and is structured toward a four-point approach: (1) adopting a workable philosophy of care; (2) adapting practices at the point of service delivery; (3) working out the coordination of diverse systems; and (4) promoting relevant research. It is expected that the Principles will be adopted by service organizations world-wide, and that the accompanying document will provide a useful and detailed baseline from which further discussions, research efforts and practice development can progress.

Wilkinson, H., Kerr, D., & Rae, C.
People with a learning disability: their concerns about dementia
Journal of Dementia Care, 2003, 11(1), 27-29.
Abstract: With people with a learning disability live longer, more of them are developing dementia. In planning the services they need, an important first step is to ask them what they think. Authors report information from surveying a group of older adults with intellectual disabilities.

Wilkinson, H., Kerr, D., & Cunningham, C.
Equipping staff to support people with an intellectual disability and dementia in care home settings.
Dementia -The International Journal of Social Research and Practice, 2005, 4(3), 387-400. https://doi.org/10.1177/147130120505
Abstract: The knowledge, experiences and skills of direct care staff working in care home settings are essential in ensuring a good quality of life and care for a person with an intellectual disability (ID) who develops dementia. Drawing on the findings of a wider study, the issues of training, support and the wider needs of staff when trying to support a resident who develops dementia are explored, specifically as relating to the role played by staff and the need to determine their experiences and related training needs. Following an introduction to the policy and practice context for working with people with an ID and dementia, and a brief description of the research method, the authors discuss the attitudes and practices of staff; supportive changes at an organizational level; and the knowledge and training needs of staff and specific gaps in knowledge. The authors argue that, within the policy and practice context of aiming to support residents to 'age in place', support for staff is a crucial aspect of ensuring that such an approach is effective and provides a coordinated approach to planning, resourcing and support.

Wisniewski, K., Howe, J., Williams, D. G., & Wisniewski, H. M.
Precocious aging and dementia in patients with Down's syndrome.
Biological Psychiatry, 1978,13, 619–627. PMID: 153156
Abstract: Studied 50 unselected institutionalized patients with Down's syndrome to determine the clinical course of precocious aging and mental and neurological deterioration. Significant differences were established in neurological and psychiatric abnormalities and mental deterioration in patients below and above age 35, indicating progressive changes in the CNS. Demonstrated were higher incidence of recent memory loss, impairment of short-term visual retention, frontal release signs, hypertonia, hyperreflexia, long-tract signs, and psychiatric problems. Also noted was the presence of external features of precocious aging. Down's syndrome appears to be a human chromosomal abnormality in which genetically determined biochemical defects leading to precocious aging and dementia can be studied.

Wisniewski, K.E., Wisniewski, H.M., & Wen, G.Y.
Occurrence of neuropathological changes and dementia of Alzheimer's disease in Down's syndrome.
Annals of Neurology,1985, Mar;17(3), 278-282. doi: 10.1002/ana.410170310.
Abstract: One hundred brains of patients with Downs syndrome (DS) who died in institutions for chronic care were examined for clinicopathological correlation of Alzheimer's disease. Fifty-one were below and 49 were above age 30 years at death. Tissues from the right, prefrontal, and hippocampal cortices were processed for microscopy using H&E and Bodian-periodic acid-Schiff impregnation. Morphometric evaluations of plaques and tangles were carried out. Plaques or plaques and tangles were found in the brains of 56 patients with DS, 7 below age 30 and 49 above that age. A history of dementia was evident in the medical records of 15 of these patients; of these only 2 were below the age of 30. The brains of the patients with DS who also had clinical dementia had more than twenty plaques or plaques and tangles per 1.5 X 10(6) micron 2 of cortex. The numbers of plaques and tangles found in the brains of the patients with DS above the age of 30 greatly increased with age but varied from brain to brain. These observations suggest a correlation among dementia, the density of plaques and tangles, and age. All 100 brains studied showed early arrest of brain growth and brain atrophy, a condition that may have been due to prenatal arrest of neurogenesis mainly in the granular cell layers, prenatal and postnatal arrest of synaptogenesis, and early aging. Plaques and tangles developed twenty to thirty years earlier and dementia was clinically detected at least three times more frequently (20 to 30%) in DS than it is known to occur in the non-DS population.

Wissing, M. B. G., Ulgiati, A. M., Hobbelen, J. S. M., De Deyn, P. P., Waninge, A., & Dekker, A. D.
The neglected puzzle of dementia in people with severe/profound intellectual disabilities: A systematic literature review of observable symptoms.
Journal of Applied Research in Intellectual Disabilities, 2022, 35, 24-45. https://doi.org/10.1111/jar.12920
Abstract: Dementia is increasingly prevalent in people with severe/profound intellectual disabilities. However, early detection and diagnosis of dementia is complex in this population. This study aimed to identify observable dementia symptoms in adults with severe/profound intellectual disabilities in available literature. A systematic literature search was conducted in PubMed, PsycINFO and Web of Science with an exhaustive search string using a combination of search terms for severe/profound intellectual disabilities and dementia/ageing. Eleven studies met inclusion criteria. Despite the few small-sized studies, a range of dementia symptoms were identified, subdivided in cognitive decline (e.g., memory loss, for getfulness, deterioration in speech, losses of social skills), decline in activities of daily living (e.g., self-cares skills, everyday functioning/ skills), BPSD (e.g., apathy, aggression, irritability, altered eating/ drinking behaviour) as well as neurologic and other physical symptoms (e.g., incontinence, (late-onset) epilepsy, hypotonia, gait deterioration). Cognitive decline, behavioral and psychological alterations, decline in activities of daily living as well as neurological and physical changes were found. Only a very limited number of studies reported symptoms ascribed to dementia in adults with severe/profound intellectual disabilities. Given the complexity of signaling and diagnosing dementia, dedicated studies are required to unravel the natural history of dementia in this population.

Wissing, M.B.G., Dijkstra, R., van der Wal, I.A., Grootendorst, E.S., Hobbelen, J.S.M., van der Putten, A.A.J., De Deyn, P.P., Waninge, A., Dekker, A.D.
Dementia in people with severe/profound intellectual (and multiple) disabilities: applicability of items in dementia screening instruments for people with intellectual disabilities
Journal of Mental Health Research in Intellectual Disabilities, 2022, 15(4), 322-363. https://doi.org/10.1080/19315864.2022.2111737
Abstract: Diagnosing dementia in people with severe/profound intellectual (and multiple) disabilities (SPI(M)D) is complex. Whereas existing dementia screening instruments as a whole are unsuitable for this population, a number of individual items may apply. Therefore, this study aimed to identify applicable items in existing dementia screening instruments. Informant interviews about 40 people with SPI(M)D were conducted to identify applicable items in the Dementia Scale for Down Syndrome, Behavioral and Psychological Symptoms of Dementia in

Down Syndrome II scale, Dementia Questionnaire for persons with Mental Retardation and Social competence Rating scale for people with Intellectual Disabilities. Among 193 items, 101 items were found applicable, categorized in β5 domains: behavioral and psychological functioning (60 items), cognitive functioning (25), motor functioning (6), activities of daily living (5) and medical comorbidities (5). Identifying applicable items for people with SPI(M)D is an essential step in developing a dedicated dementia screening instrument for this population.

Wissing, M.B.G., Fokkens, A.S., Dijkstra, R., Hobbelen, J.S. M., van der Putten, A.A.J., De Deyn, P.P., Waninge, A., & Dekker, A.D.
Dementia in people with severe/profound intellectual (and multiple) disabilities: practice-based observations of symptoms
Journal of Mental Health Research in Intellectual Disabilities, 2022, 15(4), 364-393. https://doi.org/10.1080/19315864.2022.2061092
Abstract: Observable dementia symptoms are hardly studied in people with severe/profound intellectual (and multiple) disabilities (SPI(M)D). Insight in symptomatology is needed for timely signaling/diagnosis. This study aimed to identify practice-based observations of dementia symptoms in this population. Care professionals and family members were invited to complete a survey about symptoms. Quantitatively analyzed survey data were further deepened through semi-structured interviews with care professionals having vast experience in signaling/diagnosing dementia in this population. Symptoms were categorized using a symptom matrix. Survey respondents and interviewees frequently observed a decline in activities of daily living (ADL) functioning and behavioral and psychological changes, like increased irritability, anxiety, apathy and decreased eating/drinking behavior. Cognitive symptoms were particularly recognized in persons with verbal communication and/or walking skills. To lesser extent motor changes and medical comorbidities were reported. Increased insight in dementia symptoms contributes to developing a dedicated screening instrument for dementia in people with SPI(M)D.

Woods, R.T., Moniz-Cook, E., Lliffe, S., Campion, P., Vernooij-Dassen, M., Zanetti, O., & Franco, M.
Dementia: Issues in early recognition and intervention in primary care.
Journal of the Royal Society of Medicine, 2003, 96, 320-324.
Abstract: Generic article about the need for quality and accurate screening and assessment of adults suspected of showing signs of Alzheimer's disease and the need for psychosocial interventions and family carer supports. Authors note need for better training of medical practitioners who may be screening for dementia, indicating that there is a need for timely detection and diagnosis that will prevent crises, facilitate adjustment and provide access to treatments and supports.

Zaman S, Fortea J.
The crucial history of Down syndrome.
Lancet Neurol. 2022 Mar;21(3):222. doi: 10.1016/S1474-4422(22)00047-3. PMID: 35182507.
Full article: In 1866, John Langdon Down was the first clinician to provide a detailed account of Down syndrome. A decade later, Fraser and Mitchell highlighted the early onset of senility in this population. The observations of neuropathological features of Alzheimer's disease in Down syndrome were published in 1929 by Struwe. In 1948, Jervis noted, "[with the] exception of the age of onset, the clinical and pathological manifestations are those of senile dementia". In 1984, using samples taken from the brain of people with Down syndrome, Glenner and Wong biochemically characterized amyloid β (Aβ), predicted that the gene for Alzheimer's disease would be on chromosome 21, and proposed a pathogenic role for Aβ. Using the Aβ peptide sequence, the locus of the APP gene was discovered on chromosome 21. Genetic linkage studies later identified the first mutations in APP, PSEN1, and PSEN2. Furthermore, Down syndrome studies have shown the importance of endosomal misprocessing of APP in Alzheimer's disease. Down syndrome is now regarded as a genetically determined form of Alzheimer's disease, similar to autosomal dominant Alzheimer's disease. Clinical and biomarker changes show a parallel natural history in these two forms. These genetic types of Alzheimer's disease provide the strongest evidence supporting the amyloid cascade hypothesis and give researchers a unique opportunity to study pathophysiology and its temporality, and to undertake preventive trials.

Zeilinger, E.L., Gärtner, C, Janicki, M.P., Esralew, L., Weber, G.
Practical applications of the NTG-EDSD for screening adults with intellectual disability for dementia: A German-language version feasibility study.
Journal of Intellectual and Developmental Disability, 2016, 41(1), 42-49. https://doi.org/10.3109/13668250.2015.1113238
Abstract: Authors evaluated the feasibility of using the German-language version of a recently developed screening tool for dementia for persons with intellectual disability (ID): the National Task Group – Early Detection Screen for Dementia (NTG-EDSD). Some 221 paid carers of ageing persons with ID were asked to use the NTG-EDSD and report back on its utility and on 4 feasibility dimensions, and to provide detailed feedback on aspects deemed critical or missing. All feasibility dimensions were rated good to very good, and 80% of respondents found the NTG-EDSD useful or very useful for the early detection of dementia. This highlights a high acceptability of this instrument by the main target group. The positive feasibility evaluation of the NTG-EDSD indicates the usability and adequacy of this instrument for application of early detection of dementia in persons with ID.

Zeilinger, E.L., Komenda, S., Zrnic, I., Franken, F., & Woditschka, K.
Informant-based assessment instruments for dementia and their measurement properties in persons with intellectual disability: systematic review protocol
BMJ Open, 2020, 10(12), 1136. https://bmjopen.bmj.com/content/10/12/e040920
Abstract: Persons with intellectual disability (ID) are at a higher risk of developing dementia than persons without ID, with an expected earlier onset. Assessment methods for the general population cannot be applied for persons with ID due to their pre-existing intellectual and functional impairments. As there is no agreed-upon measure to assess dementia in persons with ID, multiple instruments for this purpose have been developed and adapted in the past decades. This review aimed to identify all available informant-based instruments for the assessment of dementia in persons with ID, to evaluate and compare them according to their measurement properties, and to provide a recommendation for the most suitable instruments. Additionally, an overview of the amount and quality of research on these instruments was provided.

Zeilinger, E.L., Stiehl, K.A.M., & Weber, G.
A systematic review on assessment instruments for dementia in persons with intellectual disabilities.
Research in Developmental Disabilities, 2013, 34, 3962–3977. doi:10.1016/j.ridd.2013.08.013
Abstract: This work describes an extensive systematic literature review on assessment instruments for dementia in persons with intellectual disability (ID). Existing instruments for the detection of dementia in persons with ID were collected and described systematically. This allows a direct and quick overview of available tools. Additionally, it contributes to the availability and usability of information about these instruments, thus enhancing further developments in this field. A systematic literature search in five databases (CINAHL, PsycInfo, PubMed, Scopus, and Web of Science) was conducted. In order to include gray literature an invisible college approach was used. Relevant studies were identified and selected using defined inclusion and exclusion criteria. After the selection process all instruments were coded and classified. It was determined which concepts they assess, whether they were especially developed or adapted for persons with ID, and whether they were designed to assess dementia. The selection of relevant papers, as well as the coding of instruments was done independently by two researchers. In total, 97 records met the search criteria. Out of these, 114 different instruments were extracted. There were 79 instruments to be completed by the person with ID, and 35 informant-based instruments. Additionally, four test batteries were found. Some of these instruments were neither designed for the assessment of dementia, nor for persons with ID. There are a variety of different tools used for the assessment of

dementia in ID. Nevertheless, an agreed-upon approach or instrument is missing. Establishing this would improve the quality of assessment in clinical practice, and benefit research. Data collected would become comparable and combinable, and allow research to have more informative value.

Zigman, W.B
Atypical aging in Down syndrome
Developmental Disabilities Research Review, 2013, 18(1), 51-67. doi:10.1002/ddrr.1128.
Abstract: At present, there may be over 210,000 people with Down syndrome (DS) over the age of 55 in the United States (US) who have significant needs for augmented services due to circumstances related to ordinary and/or pathological aging. From 1979 through 2003, the birth prevalence of DS rose from 9.0 to 11.8 (31.1%) per 10,000 live births in 10 representative US regions. This increase, largely due to women conceiving after age 35, portends an ever-growing population of people with DS who may be subject to pathogenic aging. Whereas Trisomy 21 is one of the most widespread genetic causes of intellectual disability (ID), it still is one of the least understood of all genetic ID syndromes. While longevity in people with DS has improved appreciably in as modest a period as 30 years, age-specific risk for mortality still is considerably increased compared both with other people with ID or with the typically developing population. The penetrance of the phenotype is widely distributed, even though a consistent genotype is assumed in 95% of the cases. Some, but not all body systems, exhibit signs of premature or accelerated aging. This may be due to both genetic and epigenetic inheritance. We now know that the long-term outcome for people with DS is not as ominous as once contemplated; a number of people with DS are living into their late 60s and 70s with few if any major signs of pathogenic aging. Alzheimer's disease (AD), a devastating disease that robs a person of their memory, abilities and personality, is particularly common in elder adults with DS, but is not a certainty as originally thought, some 20% to 30% of elder adults with DS might never show any, or at most mild signs of AD. DS has been called a mature well-understood syndrome, not in need of further research or science funding. We are only beginning to understand how epigenetics affects the phenotype and it may be feasible in the future to alter the phenotype through epigenetic interventions. This chapter is divided into two sections. The first section will review typical and atypical aging patterns in somatic issues in elder adults with DS; the second section will review the multifaceted relationship between AD and DS.

Zigman, W.B,, & Lott. I.T.
Alzheimer's disease in Down syndrome: Neurobiology and risk.
Mental Retardation and Developmental Disabilities Research Reviews, 2007, 13, 237–246. https://doi.org/10.1002/mrdd.20163
Abstract: Down syndrome (DS) is characterized by increased mortality rates, both during early and later stages of life, and age-specific mortality risk remains higher in adults with DS compared with the overall population of people with mental retardation and with typically developing populations. Causes of increased mortality rates early in life are primarily due to the increased incidence of congenital heart disease and leukemia, while causes of higher mortality rates later in life may be due to a number of factors, two of which are an increased risk for Alzheimer's disease (AD) and an apparent tendency toward premature aging. In this article, we describe the increase in lifespan for people with DS that has occurred over the past 100 years, as well as advances in the understanding of the occurrence of AD in adults with DS. Aspects of the neurobiology of AD, including the role of amyloid, oxidative stress, Cu/ZN dismutase (SOD-1), as well as advances in neuroimaging are presented. The function of risk factors in the observed heterogeneity in the expression of AD dementia in adults with DS, as well as the need for sensitive and specific biomarkers of the clinical and pathological progressing of AD in adults with DS is considered.

Zigman, W.B., .Devenny, D.A., Krinsky-McHale, S.J., Jenkins, E.C., Urv, T.K., Wegiel, J., Schupf, N., & Silverman, W.
Alzheimer's disease in adults with Down syndrome.
International Review of Research in Mental Retardation, 2008, 36, 103-145.
Abstract: Down syndrome is associated with increased mortality rates due to congenital cardiac defects and leukemia early in life, and with Alzheimer's disease and a tendency toward premature aging later in life. Alzheimer's disease was once considered an inexorable result of growing old with Down syndrome, but recent data indicate that risk does not reach 100%. Although some individuals exhibit signs and symptoms of Alzheimer's disease in their 40s, other individuals have reached the age of 70 without developing dementia. This chapter presents a wealth of data from a longstanding longitudinal study with the overall objective of understanding and recounting the mechanisms responsible for these substantial individual differences.

Zigman, W.B., Schupf, N., Devenny, D., Miezejeski, C., Ryan, R., Urv, T.K., Schubert, R., & Silverman, W.
Incidence and prevalence of dementia in elderly adults with mental retardation without Down syndrome.
American Journal on Mental Retardation, 2004, 109, 126–141
Abstract: Rates of dementia in adults with mental retardation without Down syndrome were equivalent to or lower than would be expected compared to general population rates, whereas prevalence rates of other chronic health concerns varied as a function of condition. Given that individual differences in vulnerability to Alzheimer's disease have been hypothesized to be due to variation in cognitive reserve, adults with mental retardation, who have long-standing intellectual and cognitive impairments, should be at increased risk. This suggests that factors determining intelligence may have little or no direct relationship to risk for dementia and that dementia risk for individuals with mental retardation will be comparable to that of adults without mental retardation unless predisposing risk factors for dementia are also present.

Denotes videocassette
Denotes book or chapter in book
Denotes CD-ROM
Denotes report
Denotes booklet or agency issued manual
Denotes website

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