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Psychological Society  
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Division of  
Clinical Psychology  
Faculty for People with  
Intellectual Disabilities



## Dementia and People with Intellectual Disabilities

*Guidance on the assessment, diagnosis, interventions and support of people with intellectual disabilities who develop dementia*

April 2015

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This publication has been produced by the Faculty for People with Intellectual Disabilities of the British Psychological Society Division of Clinical Psychology and the Intellectual Disabilities Faculty of the Royal College of Psychiatrists, and represents the views and expert contributions of the members of those faculties only.

Printed and published by the British Psychological Society.

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REP77 4/2015

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# Membership of the working group

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## Acknowledgements

The working group acknowledges the help of the following people in providing their views, advice and contributions: Members of the DCP Faculty for People with Intellectual Disabilities and the Royal College of Psychiatrists.

## Foreword

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It is a great pleasure to be asked to provide a foreword for this excellent guidance on dementia and people with intellectual disabilities. The awareness of dementia has never been higher and the emphasis on providing support to people following a diagnosis is most important. We know that dementia attracts stigma and that is of at least equal if not more relevance in people with intellectual disabilities. Professionals and the public are generally more aware of the increased risk of developing dementia in people with intellectual disabilities.

The guidance is comprehensive across the pathway from diagnosis, post diagnostic support including assessment, through management and end of life care. Its strength is in its comprehensive nature, including, in the same breath, those practical and clinical suggestions which are so important in the day-to-day care of people as well as the evidence base for these actions. The multi-professional nature of the guidance is so important.

One of the things which has been achieved in dementia in the general population, particularly older people, is to convey a message that dementia is a challenge for everybody. Information and guidance has been key to this and the information provided in this guide will go a long way to reassuring people that issues regarding cognitive impairment and dementia in people with intellectual disabilities are not, in all cases and as a matter of course, the purview of the expert and the specialist.

Everyone knows that, thanks to advances in medical care and the social environment, people with intellectual disabilities are now living longer, a great success and a tribute to society, and that brings with it some challenges in terms of the need for more services for people with cognitive impairment and intellectual disabilities.

This guidance has been created carefully by the recognised authorities in the arena and has been woven together expertly. It is accessible and yet comprehensive, it is practical but not simplistic but most importantly it whets the appetite of the reader to learn more. Although concentrating on initiatives in England, an international audience could read it with benefit.

I certainly learnt more about dementia in people with intellectual disabilities in reading this than I have ever before.

**Professor Alistair Burns**

*National Clinical Director of Dementia*

*NHS England*

## Purpose of this guidance

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This report is a revision to the original joint British Psychological Society and the Royal College of Psychiatrists (2009) guidance on dementia and people with intellectual disabilities. It has been written by a joint working group of the DCP Intellectual Disabilities Faculty of the British Psychological Society and the Royal College of Psychiatrists.

The main purpose of the guidance is to enable those working in clinical and social care services to improve the quality of life of people with intellectual disabilities who develop dementia, by providing guidance to inform assessment, diagnosis, interventions and support. The guidance is aimed at clinicians in intellectual disabilities and older peoples' mental health services and services for younger people with dementia. The decision about which services provide which part of the care pathway is a local decision to be taken by commissioners and providers, but ensuring that all elements of this guidance are considered and in place for people with intellectual disabilities and dementia.

Since the original guidance was published there has been a far greater awareness about dementia in the general population and a proliferation of strategies and standards documents. However, dementia and people with intellectual disabilities has still received minimal focus. The current guidance has been updated using both the current research literature and the experience of senior clinicians working in the field. Some areas of the guidance have only needed minor changes, whilst others have had a more extensive re-write. Three completely new sections have been added.

As before, our main focus has been to highlight the key factors that we consider are the elements of an excellent service, and to help those working in services evaluate how they might help the increasing numbers of people with intellectual disabilities who are developing dementia given improvements in life-expectancy.

It is with these principles in mind that this report has been produced and has the following aims:

- To bring together available and relevant evidence-based practice with a consensus of clinical opinion and experience.
- To provide a framework for good practice and for the development of multi-agency care pathways.
- To promote effective and timely assessment, diagnosis, and interventions for people with intellectual disabilities suspected or confirmed as having dementia and to ensure quality support to them and their staff and other carers.
- To provide guidance for service providers, developers and commissioners.
- To provide a set of standards of good practice against which service provision can be benchmarked and audited.
- To provide a quality outcome measure to use to evaluate what the person with intellectual disabilities and dementia is experiencing in their care.
- To promote the development of comprehensive and effective local services and to reduce the number of individuals who are failed by current service provision.

- To provide a framework for training health and social care professionals and paid support staff and carers.
- To guide the future development of services.

The report was produced through the combined work of members of the working group and drew on the published evidence base and from the working group members' extensive clinical experience in this area. Although this report has been undertaken primarily by psychologists and psychiatrists, we recognise that people with dementia must have effective multi-agency and multi-disciplinary services. We believe that this report will therefore be relevant to anyone who has an interest in dementia and people with intellectual disabilities, including health and social care professionals, families, paid staff, advocates, service providers and commissioners.

We still cannot be prescriptive within the document, particularly about the choice of assessment tools, because the evidence is not available to support particular instruments. Decisions will still need to be made locally depending on local resources and configurations. However, the report is intended to highlight the specific issues that people with intellectual disabilities and dementia present, and to ensure that local services are timely, effective and ensure that the person continues to have a high quality, safe and person-centred lifestyle as the dementia progresses.

This is a rapidly developing area with new knowledge and practice developing all the time. Research with people with intellectual disabilities and dementia is increasing, and will add to the evidence base.

Finally, the report hopes to complement other publications and guidance in this area and to provide a way forward for supporting people with intellectual disabilities who develop dementia.

## Section 1 – Context

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In March 2012 the Prime Minister, David Cameron, set out what he called ‘the Dementia Challenge’ (DH, 2012a). Recognising the urgent need for change, he set targets for improvements by 2015 in three broad areas: 1) health and care, 2) the establishment of dementia friendly communities, and 3) improving dementia research. This document has been revised with these challenges in mind, paying particular attention to how they apply to people with intellectual disabilities. It sets out standards of clinical practice in the areas of assessment, diagnosis and interventions for people with intellectual disabilities who develop dementia with an emphasis on how people with intellectual disabilities who develop dementia can be best supported in a manner that maintains their dignity and quality of life.

Although this document primarily references the context of dementia in England and the other UK nations, publications from other nations are equally as relevant and have the same underlying principles and practices outlined in them, for example Australia’s *National Framework for Action on Dementia* (Australian Government, 2006) and the Netherlands’ *National Dementia Plan* (Ministry of Health, 2004).

### 1.1 Dementia publications

Interest in the field of dementia in the general population has further increased since the publication of the first edition of this guidance in 2009. At that time the National Dementia Strategy for England had just been published (*Living well with dementia: A National dementia strategy*; DH, 2009). The strategy has three key steps: improved awareness and understanding of dementia and removal of the stigma that surrounds it; early diagnosis and intervention; and improving the quality of care for people with dementia by developing a range of services for people with dementia and their carers which fully meets their changing needs over time. The strategy had 17 objectives to be met in a five-year plan; the objectives included improving awareness and diagnosis, better access to care and advice, the needs of carers, the commissioning of health and social care services, better end of life care, workforce planning, and research. Similar strategies have been published in Scotland (*Scotland’s National Dementia Strategy 2013–2016*; Scottish Government, 2013), Wales (*National Dementia Vision for Wales*; Welsh Assembly Government, 2011) and Northern Ireland (*Improving Dementia Services in Northern Ireland – A Regional Strategy*; DHSSPSNI, 2011).

The National Institute for Health and Care Excellence (NICE) and the Social Care Institute for Excellence (SCIE) published a joint clinical guideline on the management of dementia in 2006 (NICE, 2006). Key recommendations included: integrated working across all agencies; provision of memory assessment services as a point of referral for diagnosis of dementia; assessment, support and treatment (where needed) for carers; assessment and treatment of non-cognitive symptoms and behaviour that challenges; dementia care training for all staff working with older people; and improvement of care for people with dementia in general hospitals. Since then NICE has published quality standards and audit tools (NICE, 2010, 2013) covering in detail principles of care, risk



factors and prevention, early identification, diagnosis and assessment, promoting choice, providing support, integrating and co-ordinating care and service provision, and promoting independence and maintaining function and also guidance on interventions, living arrangements, hospital and palliative care. These are all relevant to people with intellectual disabilities, given the risk of dementia at a younger age in people with intellectual disabilities (recognised in the NICE guidance), particularly affecting people with Down's syndrome, with the added complexity of the potential difficulty recognising the possibility of dementia and arriving at an accurate diagnosis given pre-existing cognitive impairments.

The NICE guidance and the quality standards are an evidence-based and informed synthesis of research, previous reports and policies. As with the earlier Department of Health and the Care Services Improvement Partnership (CSIP) guidance *Everybody's Business: Integrated Mental Health Services For Older Adults: A Service Development Guide* (DH/CSIP, 2005), the NICE guidance recognises that effective services will only be achieved through joint working and efficient partnerships. The earlier National Audit Office Report (NAO, 2007) had concluded that overall, services were not currently delivering value for money to taxpayers or people with dementia and their families; that too few people were being diagnosed or being diagnosed early enough, and that early intervention was needed to improve quality of life; and, finally, that services in the community, care homes and at the end of life are not delivering consistently or cost-effectively against the objective of supporting people to live independently as long as possible in the place of their choosing. The NAO advocated a 'spend to save' approach, with upfront investment in services for early diagnosis and intervention and improved specialist services, community services and in general hospitals, resulting in long-term cost savings from prevention of transition into care homes and decreased length of hospital stay. The Commission for Social Care Inspection had also reported on people's experiences of living in a care home in their report *See Me, Not Just the Dementia* (CSCI, 2008) confirming the importance of a positive communication style with people with more advanced dementia. These are all very much echoed in the National Dementia Strategy and in the Prime Minister's challenge, suggesting that these and other previous reports are now having an impact. This is a very positive development.

The above are examples of dementia-specific policies and guidance; however, there is also a broader context which is relevant to this updated guidance. First, is the serious problem of the potential for neglect and abuse within health and social care settings. The reports of the Mid Staffordshire NHS Foundation Trust Public Enquiry (Francis, 2013) and from the Department of Health on the independent hospital, *Winterbourne View – Transforming Care: A National Response to Winterbourne View Hospital* (DH, 2012), have highlighted the particular vulnerabilities of older people with dementia and people with intellectual disabilities in institutional hospital settings. Earlier concerns about the abuse of people who were vulnerable in the community had led in England and Wales to the publication in 2008 of *No Secrets* (DH, 2008a) and to all areas of the country establishing safeguarding policies. Abuse of people with intellectual disabilities and those with dementia is not uncommon, whether in hospitals, family homes or residential care. The challenge is to prevent it, as far as possible, and to detect it and respond quickly where it is identified as having taken place.

Secondly, the move towards individual health and social care budgets that aim to give choice and control to people in need of health and social care services including people with intellectual disabilities and people with dementia has become a reality. For people with disabilities this had its origins in the 1996 Community Care (Direct Payments) Act but in reality only in the last few years has it become the funding model of choice with the intention of extending it to other areas of need and to the funding of health care.

Thirdly, there have been changes in the law and in international conventions that have implications for the way in which care is provided and in which choices are made as to the services people receive.

Mental Capacity issues are laid out in legislation in many nations, e.g. Adults with Incapacity Act (Scotland) 2000 and the Mental Capacity Act (England and Wales) 2005. The Mental Capacity Act 2005 is well established in England and Wales although a recent post-legislative review by the House of Lords has raised concerns about its implementation. However, the amendment to the Act that introduced the Deprivation of Liberty Safeguards (DoLS) was severely criticised by the House of Lords Committee and also has been subject to a recent ruling from the Supreme Court that has implications for the support of people with intellectual disabilities and also those with dementia whether or not they have intellectual disabilities. This ruling has broadened the definition of what is meant by ‘deprivation’ and it is likely that many more people with intellectual disabilities, and particularly those who have developed dementia, will be considered to be deprived of their liberties and in need of the safeguards. In 2008 the UN Convention on the Rights of People with Disabilities (UN, 2008) also challenged concepts of legal capacity arguing that everyone must be presumed to have ‘legal capacity’. The Equality Act has also been enacted, requiring authorities to make ‘reasonable adjustments’ when seeking to meet the needs of people with disabilities. These various legal developments when taken together have very significant implications for the group of people that are the focus of this guidance.

## **1.2 Assumptions made within the guidance document**

Within this document, a number of assumptions are made throughout. These include the need for effective partnership working between all agencies involved in the care of people with intellectual disabilities and dementia as highlighted in reports cited in the previous section. The working party recognises that each area will be configured to meet local need, but effective care can only be provided when there is good partnership working within health services – between intellectual disabilities and older people’s services, and across statutory, private and voluntary agencies.

Regardless of how each service is configured, the working party has assumed that certain principles and ways of working are already integral to the delivery of services for people with intellectual disabilities, and that these will also be available to people with intellectual disabilities who develop dementia. It has been assumed that services already deliver care in line with *Valuing People* (DH, 2001) ensuring that everyone who wants one has a Person Centred Plan and a Health Action Plan, as well as an individualised care plan. It has also been assumed that services will be delivered in line with both their relevant mental capacity legislation, the Human Rights Act and relevant National Standards for dementia care (e.g. NICE, 2006, 2010, 2013).

## Section 2 – Epidemiology

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### 2.1 Older population with intellectual disabilities

There have been significant improvements in the mean life expectancy of people with intellectual disabilities from an estimated 18.5 years in the 1930s to 66 years in the 1990s (Braddock, 1999). The life expectancy of people with mild intellectual disabilities now approaches that in the general population of a similar socio-economic status, but the life expectancy of people with more severe levels of intellectual disabilities remains reduced compared with the general population.

Given these improvements, the overall population with intellectual disabilities is steadily increasing and it has been predicted that the proportion of people with intellectual disabilities over 65 years of age will have doubled by 2020, with over a third of all people with intellectual disabilities being over 50 years of age by that time (Janicki & Dalton, 2000; McConkey et al., 2006). This is also true for the population of people with Down's syndrome. The birth prevalence of Down's syndrome in England and Wales has remained relatively stable despite antenatal screening (which has been offset by an increase in Down's syndrome resulting from younger maternal age), while mean life expectancy has increased to 58 years (Wu & Morris, 2013). This has resulted in a growing population of older adults with Down's syndrome. It is because of these factors that consideration needs to be given to age-related illnesses that most commonly occur in later life, such as dementia.

Estimating prevalence rates of dementia can be problematic because of a number of methodological issues. These include diagnostic difficulties associated with dementia in a population which has pre-existing cognitive and functional impairments and the complexity of establishing accurate population samples of people with intellectual disabilities. However, there is evidence from several studies that people with intellectual disabilities have an increased risk of developing dementia compared to that observed in the general population. In particular, those with trisomy 21 resulting in Down's syndrome have an earlier age-related risk of developing dementia of the Alzheimer type. There is very limited research investigating the specific risk of dementia and other age-related disorders in those with other specific syndromes, some of which are associated with a reduced life-expectancy, or with autism.

### 2.2 Prevalence rates of dementia among older people with intellectual disabilities (excluding people with Down's syndrome)

Several studies have investigated rates of clinical dementia among people with intellectual disabilities living within the community and used established or modified criteria based on systems for the diagnosis of dementia. Moss and Patel (1995) reported that 12 per cent of a group of people with intellectual disabilities over age 50 years had dementia. Cooper (1997) found that the rate of dementia increased as expected in a population-based study with just over 20 per cent of those over the age of 65 years meeting criteria for dementia. Strydom et al. (2007) reported findings from a two-stage population-based survey of adults

with intellectual disabilities (without Down's syndrome) across several London boroughs. They found that prevalence rates varied depending on the diagnostic criteria used, with DSM-IV criteria resulting in the highest rate and ICD-10 the lowest. Rates for dementia, of whatever cause, using DSM-IV criteria were 13.1 per cent in those 60 years and over and 18.3 per cent in those 65 years or over. This compares to prevalence rates in the general population of 1 per cent for 60–65-year-olds to 13 per cent for 80–85-year-olds and 32 per cent for 90–95-year-olds (Hofman et al., 1991). See Figure 1 below for a comparison of dementia rates between individuals with Down's syndrome, intellectual disabilities and the general population.

Alzheimer's disease was found to be the most common type of dementia, and had a prevalence of 8.6 per cent in those aged 60 and older, three times greater than comparable general older adult population rates, but cases of Lewy body dementia, vascular dementia, and fronto-temporal dementia were also identified based on clinical observations while rates varied according to diagnostic criteria used – DSM-IV criteria were more inclusive than ICD10 (Strydom et al., 2007). Prevalence rates increased with age though shifted towards younger ages compared to the general population but did not differ significantly between mild, moderate and severe intellectual disabilities groups (Strydom et al., 2009).

Overall, the incidence rate for dementia in those aged 60 and older was estimated to be 54.6/1000 person years with the highest incidence rate in the age group 70–74 (Strydom et al., 2013).

### **2.3 Prevalence and incidence rates of dementia among people with Down's syndrome**

The association between Down's syndrome and the risk of 'precipitated senility' was first reported by Fraser and Mitchell in 1876, with Struwe (1929) describing the significant Alzheimer-like neuropathological changes in the brains of people with Down's syndrome, and almost all older adults with Down's syndrome were found to have the neuropathological hallmarks of Alzheimer's disease at post-mortem (Mann, 1988) which has since also been demonstrated with in-vivo amyloid PET imaging studies (Landt et al., 2011; Hartley et al., 2014).

Studies have reported rates of dementia meeting the necessary criteria that start at a few percent from 30 to 39 years of age, increasing to 10–25 per cent in the 40 to 49-year-old group and to 20 per cent and 50 per cent in the 50 to 59-year-old group and between 30 per cent and 75 per cent aged 60 years or older (Hewitt et al., 1985; Wisniewski et al., 1985; Lai & Williams, 1989; Holland et al., 1998). Between 50 and 60 years of age the prevalence of dementia doubles with each five-year interval (Coppus et al., 2006). Studies have varied in their findings beyond the age of 60: some studies have found that prevalence rates continue to increase, with most individuals eventually diagnosed with dementia (Visser et al., 1997; Tyrrell et al., 2001) whilst others described a decrease in prevalence in the older age group due to the increased mortality associated with dementia (Coppus et al., 2006). Incidence increased steadily with increasing age and did not decline after age 60, from 2.5 per 100 person years in those aged <50 to 13.31 per 100 person years in those aged 60 and older (Coppus et al., 2006).

Case reports, cross-sectional and longitudinal studies have all confirmed an increase in the

prevalence rates of clinically diagnosed dementia with increasing age that starts when people with Down's syndrome are in their 30s and steadily increases in prevalence into the 60s. These rates are not as great as neuropathological studies initially suggested would be the case and, whilst the precise rates differ between studies, it is clear that not all people with Down's syndrome present with the pattern of memory loss and functional decline characteristic of dementia in later life. Nevertheless, using cumulative incidence rates it has been calculated that nearly 70 per cent of older adults with Down's syndrome are likely to develop dementia symptoms should they all live to age 70 (Zigman et al., 2002).

Recent analysis by the ADSID collaboration (Strydom 2014) using data from assessment clinics across England ( $N = 338$  cases with Down's syndrome and dementia) established that the majority of individuals with Down's syndrome who are diagnosed with dementia presented in their 50's (interquartile range 50.9 – 59.3 years), with a mean age at diagnosis of approximately 55 years (SD 6.5). A quarter was diagnosed before the age of 50, and a quarter after age 60. Survival varied considerably, but did not appear to be much shorter than the general population with a mean survival time of 4 years following diagnosis.

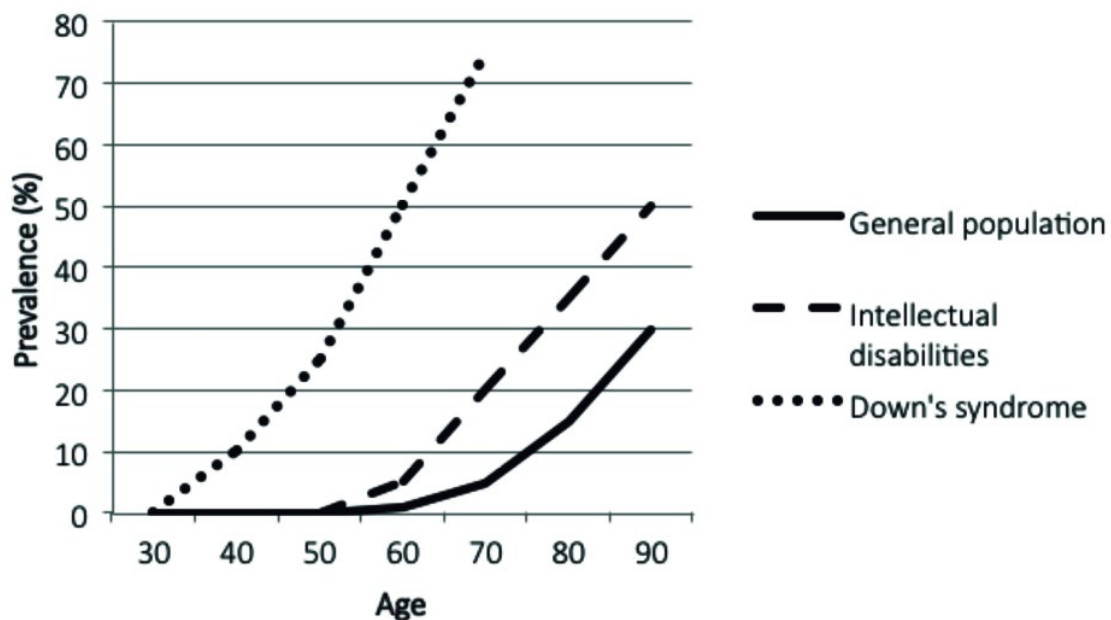


Figure 1: Comparison of dementia prevalence rates by age

Figure 1 summarises the age-related prevalence rates of dementia in people with Down's syndrome, those with intellectual disabilities without Down's syndrome, and in the general population. The exact rates have to be considered with caution but the trend represented in this figure is now increasingly accepted. The early presentation and course of dementia is now well established for people with Down's syndrome. For those with intellectual disabilities but without Down's syndrome, age-related prevalence rates are brought forward to a small degree compared to the general population but not to the same extent as for people with Down's syndrome. This latter group would appear to have a uniquely early risk for developing dementia, almost invariably of the Alzheimer's type. For the former group the full range of causes of dementia is observed.

Amyloid is the protein that, in an insoluble form, forms the neural plaques characteristic of Alzheimer's disease. The gene coding for the Amyloid Precursor Protein (APP) is

located on chromosome 21, and this is likely to account for the increase in risk of Alzheimer's disease in people with Down's syndrome. Children with Down's syndrome have been found at post mortem to have evidence of diffuse cerebral amyloid deposition and in adult life plaques and tangles characteristic of Alzheimer's disease are found. The assumption therefore is that this slow deposition of amyloid in the brain leads to a cascade of adverse neural events over time and ultimately to the full pathology of Alzheimer's disease. Further evidence implicating the extra copy of the APP gene in Alzheimer's disease in Down's syndrome was reported in a rare case of partial trisomy 21 without triplication of the APP gene. Neuropathological changes associated with Alzheimer's disease did not occur; neither did clinical dementia develop despite the advanced age of the individual (Prasher et al., 1998). However, whilst the brain pathology characteristic of Alzheimer's disease would seem to be near universal in later life, it is clear that not all of the older people with Down's syndrome with full trisomy 21 develop the clinical features of dementia. The reason for this remains unclear.

### Key points

- People with intellectual disabilities have a higher risk of developing dementia compared to the general population, with a significantly increased risk for people with Down's syndrome and at a much earlier age.
- Life expectancy of people with Down's syndrome has increased significantly and the number of older people with Down's syndrome has been increasing.
- The incidence and prevalence of Down's syndrome is relatively stable.

## Section 3 – Baseline assessment and monitoring

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### 3.1 Reactive, baseline assessment and prospective monitoring

A core feature of dementia is a decline from a baseline level of the person's functioning. Establishing pre-morbid skills, abilities and personality can be challenging in the intellectual disability population due to variance in cognitive functioning and abilities, frequent poor record keeping from childhood and the possible lack of consistent involvement of family or staff throughout the person's lifespan.

Signs of early dementia can be subtle and require careful observation to identify concerns in a timely way. Families and staff carers can often be so close to the person that they become less able to recognise minor changes in functioning through adapting to the person's needs. Similarly, for some people there is an absence of people who can comprehensively describe and evidence the person's baseline of functioning, the role of baseline cognitive assessment becomes apparent.

Services need to consider what type of service they offer to people with intellectual disabilities who may develop dementia. Services will need to do the following:

- Provide reactive screening. This relates to the assessment of functioning and the development of a formulation exploring the reasons for observed deterioration in any adult with intellectual disabilities after concerns have been raised. Information relating to dementia care pathways and the roles of the multidisciplinary team can be found in Section 6.
- Establish a baseline for every adult with Down's syndrome whilst they are healthy – ideally at age 30.

Services may also consider undertaking prospective screening for dementia for adults with Down's syndrome conducted at intervals from the age of 30 onwards.

Good practice guidance from the Foundation for People with Learning Disabilities (Turk et al., 2001) recommended that every service for people with intellectual disabilities should set up a register of adults with Down's syndrome, conduct a baseline assessment of cognitive and adaptive functioning by the age of 30 years (being mindful of likely continued brain development throughout the second decade of life). Despite this, the availability of assessment and treatment across the UK remains inequitable. Most areas now offer reactive assessment for those with signs of deterioration, and a number of services now offer baselines and prospective screening to adults with Down's syndrome such as that described by Hobson et al., 2012, Cairns et al., 2010; Jervis & Prinsloo, 2007; McBrien et al., 2005.

Tools used for dementia assessment are described in section 6.

### 3.2 Reactive monitoring

Reactive monitoring is the most common service provided by intellectual disabilities services. However, the reliability and efficiency of reactive screening can be greatly enhanced if a baseline assessment is available, as data can be compared in a timely way.

Reactive assessment means conducting a thorough assessment of cognitive and social functioning after concerns about deterioration have been raised. This relies on carers noticing relevant signs of change and making a referral to the GP or intellectual disability service. This does not always occur in a timely manner. Due to high turnover of staff in many intellectual disabilities residential settings, staff may not be aware of changes in a person's presentation and diagnostic overshadowing can occur. To enhance this brief dementia checklists can be used by residential staff or at annual reviews (e.g. Whitwham et al., 2011), and staff awareness training can be provided for services supporting people with intellectual disabilities.

### **3.3 The importance of baseline assessment**

There is no definitive 'test' for dementia. Its presence is a matter of eliciting a clinical history suggesting dementia and establishing evidence of change in function from a known baseline and excluding other diagnoses that may mimic dementia. In the mainstream population, it is more straightforward to gauge pre-morbid functioning from self-report or employment history than it is in the population of people with intellectual disabilities where self-report is limited and few paid carers are in possession of a full history.

Unless a baseline is established when the person is healthy, it is difficult to know whether there has been a deterioration later in life. By the time an individual is referred with concerns, considerable deterioration may have already occurred and an accurate account of pre-morbid functioning may be difficult to construct. Longitudinal data is then needed to establish decline from a baseline as it is not possible to compare results of an assessment with an 'average' result for the person with Down's syndrome, intellectual disabilities or general population.

Whilst it may be possible to establish a diagnosis of dementia from a one-off assessment when there is good historic data from which to compare, or the clinical picture is extremely clear, there is a risk of false negative or positive diagnoses.

Prompt diagnosis ensures that attention can be paid in a timely way to necessary changes to a care package, medication, preparing family carers and support staff for the inevitable changes and challenges that dementia will bring.

#### **When should baseline assessment occur?**

There is currently no clear evidence base as to the best age at which this should occur; however, setting a baseline when the client is healthy and functioning at their best will clearly be most helpful at re-screen should concerns arise. It has been suggested that an assessment by the age of 30 years would be helpful (Turk et al., 2001; McBrien, 2009). Ongoing neurodevelopment in late adolescence and early adulthood should be considered so as not to set baselines at too early an age when the brain is still developing. Carr (2000) demonstrated stability in intellectual ability and daily living skills for her cohort of people with Down's syndrome during the age period 21–30 years. This suggests that a baseline conducted by the age of 30 would helpfully capture people prior to any cognitive decline.

Ideally, a baseline assessment at age 30 years would capture an individual's functioning once the brain is thought to be relatively fully developed, and before the potential onset of



dementia. A copy of the baseline assessment should be given to the person and their carers to keep in the person's health action plan for future reference.

Functional limitations of electronic health record systems may require the development of additional databases to capture relevant clinical information about clients with Down's syndrome, or people on dementia care pathways. It is important to be aware of relevant data protection issues and organisational protocols (such as registering your database with the Trust's information governance department) and be mindful of the potential need to gain consent/assent from service users and/or their carers to have personal information recorded in such a way.

Some services seek simultaneous consent for inclusion on dementia databases alongside permissions to contact clients regarding possible research projects that may be of interest in the future.

### **3.4 Prospective monitoring**

Prospective monitoring entails checking for early signs of dementia by repeating the baseline assessment at regular intervals. This necessitates, as do baseline assessments, having a register of all adults with Down's syndrome and additionally a method of recalling people for a re-assessment. Jethwa and Cassidy (2010) and O'Caomh et al. (2013) suggest an accurate and extensive record of baseline skills levels in people with intellectual disabilities is crucial and regular comparison with baseline is key to early diagnosis.

The frequency of prospective monitoring for dementia should be matched to the rising risk with age. For example, the baseline assessment should take place at 30 years; then every two years for those in their 40s; and annually for those aged 50 and over. Assessment is non-invasive and is usually enjoyable for the participant (see Section 6).

#### **Additional benefits of regular assessment**

One argument in favour of prospective screening concerns the known health risks for all people with Down's syndrome. Many treatable illnesses can produce symptoms of cognitive decline independent of dementia. The evidence is that all too often, such disorders remain undetected in intellectual disabilities populations (Watchman, 2014). This applies particularly to people with Down's syndrome who are prone to certain health problems (Coppus, 2014). Many carers and indeed some GPs are not aware, for example, of the need for routine thyroid function tests for adults with Down's syndrome. There are useful factsheets, available free of charge on the internet from the Down's Syndrome Association, that can be used to raise awareness of this issue.

A review of six months of routine prospective screens carried out in one intellectual disability service showed that of 33 prospective assessments of apparently healthy adults with Down's syndrome, 12 (36 per cent) revealed concerns that had not previously been identified by carers. These included dementia-like symptoms (memory and behavioural change), physical or mental health concerns that could be immediately treated, and action required by social services, including one requiring the instigation of the safeguarding protocol (Major & McBrien, 2011). It is hoped that improved access to regular health screening (e.g. annual GP health checks) will help to minimise the prevalence of undetected physical and mental health concerns.

### Prospective assessments – should we or shouldn't we?

The evidence does not currently indicate a need for prospective screening for people with intellectual disabilities without Down's syndrome. Screening for people with Down's syndrome needs to be justified in terms of likely gain, the demands placed on the service, and there needs to be consideration of the issue of informed consent. Some services may decide against it for reasons of intrusion, commissioning intentions and/or resource limitations. Apparent changes in scores on cognitive assessments do not in themselves indicate dementia but do require further investigation.

## 3.5 Ongoing Monitoring

Regardless of whether a service offers prospective monitoring, it is important that a programme of regular monitoring and reassessment is established once the person is suspected of having dementia. This will enable the progression of the disease to be described, suitable medical, psychosocial and environmental interventions to be put in place, and the quality outcome for the person to be measured. See Sections 6, 12 and 18.

### Key points

- It is recommended to assess every adult with Down's syndrome by the age of 30 to establish a baseline against which to compare future suspected changes in functioning.
- It is worth considering screening all adults with Down's syndrome over 40 regularly because of the increased risk of dementia and the prevalence of undetected but treatable illnesses. This should link to the person's health action plan.
- Once the person has been suspected of having dementia, a programme of regular monitoring and reassessment needs to be established.

## Section 4 – Possible reasons for apparent decline in functioning in adults with intellectual disability

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Changes in functional ability with or without behaviour change are often the initial presentation of dementia in adults with intellectual disabilities. It is important to remember, however, that some changes may be part of the normal ageing process. There are a number of other reasons an individual with intellectual disabilities may show a cognitive decline. The list below, whilst not exhaustive, describes the most common reasons for change in ability. It is important to recognise that two or more conditions can co-exist.

- 1. Dementia:** The typical presentation of dementia is one of gradual loss of skills along with change in personality and cognitive decline. Dementia is a diagnosis of exclusion therefore, it is important to consider other conditions that may cause loss of skills and cognitive decline, especially as many of these are treatable.
- 2. Physical problems** include such conditions as hypothyroidism, anaemia, uncontrolled epilepsy and chronic infections. Electrolyte abnormalities, hypo- or hyper-glycaemia, nutritional deficiencies particularly vitamin B12 or folate could also cause functional decline in adults with intellectual disabilities. A thorough physical examination and relevant clinical tests are required at the time of initial assessment. These may need to be repeated from time to time as necessary.
- 3. Sensory impairments:** People with intellectual disabilities in general and those with Down's syndrome in their middle/old age specifically are likely to develop hearing and visual impairments. Visual impairment could be due to development of cataract or conditions such as keratoconus in individuals with Down's syndrome. Some 60–80 per cent of people with Down's syndrome will have hearing problems at some point in their lives. Conductive hearing deficit caused by earwax and the narrow acoustic canal is frequently seen in people with Down's syndrome.
- 4. Mental health problems:** The most common differential diagnosis is depressive illness (McBrien, 2003) but other conditions such as the exacerbation of an existing psychotic disorder can mimic the presentation of dementia. Severe anxiety can also cause an apparent decline in functioning.
- 5. Sleep problems:** Obstructive sleep apnoea or other sleep disorders can cause day time drowsiness, mental slowing as well as confusion, and are particularly common in people with Down's syndrome. Day time drowsiness and slowing could be interpreted as dementia if the sleep problem is not identified.
- 6. Iatrogenic (medication related) causes:** Medications with anticholinergic side effects can cause cognitive impairments in elderly people and people with intellectual disabilities. Use of high dose psychotropic/anti-epileptic medications and multiple medications can contribute to cognitive impairment as well. Some of the common medications that could cause a dementia-like presentation are listed in Table 1.

**Table 1:** Some common medications that could cause a dementia-like presentation  
(Diagnosis and treatment guideline, Mayo Foundation for Medical Education and Research, (Moran et al., 2013))

Medication class	Examples	Side effects that contribute to cognitive decline
Antipsychotic medications	Chlorpromazine, Olanzapine Clozapine	Sedation, mental slowing, effect of anti-cholinergic properties affecting cognition
Anti-epileptic medications	Phenobarbitone, Phenytoin, Sodium Valproate	Sedation and mental slowing
Antidepressants	Clomipramine	Same as above
Benzodiazepines, particularly long acting preparations	Clonazepam Temazepam Diazepam	Sedation, confusion, mental slowing
Older generation antihistamines	Diphenhydramine Hydroxyzine Promethazine	Sedation
Pain medications	Meperidine Propoxyphene	Confusion, dizziness, tiredness Meperidine can cause seizures

**7. Impact of life events:** People with intellectual disabilities in their middle age can face a number of life events such as loss of a parent or long-term carer, moving away from home or loss of day activities. In some individuals, the impact of life events may lead to a regressive state with apparent loss of skills. Changes in routine such as new structure to day opportunities or changes in support staff can cause profound reactions in an individual with intellectual disabilities leading to functional decline and a dementia-like presentation.

**8. Abuse:** Current or recent physical, emotional or sexual abuse in people with intellectual disabilities may result in loss of skills and regression and the development or exacerbation of behaviour problems that might superficially mimic dementia.

**9. Impact of poor environment:** An unsuitable environment associated with a lack of stimulation, isolation and lack of social opportunities for positive interaction can lead to loss of skills. In the event that this is also associated with changes in support structure, where people do not know the individual well, these changes may be attributed to dementia.

**10. Acute organic brain syndrome:** This may co-exist with dementia or be part of the differential diagnosis. Someone with uncontrolled epilepsy, for example, could present with confusional state that may mimic dementia. People with dementia may deteriorate rapidly and develop an acute confusional state when they have an acute physical health problem such as a urinary tract or respiratory infection.

### Key points

- The common differential diagnoses for individuals with Down's syndrome presenting with loss of skills are depressive illness, sensory impairments (hearing or visual), hypothyroidism, obstructive sleep apnoea and dementia.
- Sometimes conditions can co-exist.
- Do not forget about the following causes of apparent functional decline as these are often missed:
  - iatrogenic causes of cognitive impairment particularly when the individual is taking multiple medications;
  - impact of the environment particularly in relation to occupational deprivation and under stimulation; and
  - impact of abuse on the individuals with ID.

## Section 5 – Clinical presentation of dementia

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### 5.1 Criteria for dementia

Within the general population, diagnostic criteria have been developed in order to improve the accuracy of the clinical diagnosis of dementia. These include the ICD-10 and DSM-5 criteria. These are summarised below.

#### ICD-10 (WHO, 1992) criteria (summary)

1. Evidence of decline in memory, most evident in the learning of new information. The impairment applies to both verbal and non-verbal material and is sufficient to interfere with everyday function.
2. A decline in other cognitive abilities and daily living skills, characterised by deterioration in judgment and thinking such as planning and organising, and in the general processing of information, to a degree leading to impaired functioning in daily living. These include:
  - language comprehension and expression;
  - perception;
  - praxis;
  - executive function;
  - usual daytime activities; and
  - use of household utensils and equipment.
3. Absence of clouding of consciousness/delirium.
4. Decline in emotional control, motivation or social behaviour in at least one of the following:
  - emotional lability;
  - irritability;
  - apathy; and
  - coarsening of social behaviour.
5. The duration in changes in memory must be longer than six months.

#### DSM-5 criteria (American Psychiatric Association, 2013)

The DSM-5 has renamed dementia as ‘neurocognitive disorders’ and recognises both minor and major neurocognitive disorder.

Major neurocognitive disorder (dementia) is defined as:

1. Evidence of a significant decline from a previous level of performance in one or more cognitive domain (such as attention, executive function, learning and memory, perceptual motor or social cognition) which is based on:
  - a. report of significant decline in cognitive function by the individual themselves, a knowledgeable informant, or a clinician;
  - b. a substantial impairment in cognitive performance is documented by clinical assessment, preferably standardised neuropsychological testing.

2. The cognitive deficits interfere with independence in everyday activities (which is defined as at a minimum requiring assistance with complex instrumental activities of daily living such as paying bills).
3. The deficits do not occur exclusively in the context of a delirium.
4. The deficits are not better explained by another mental disorder (e.g. major depressive disorder or schizophrenia).

Subtypes should be specified (e.g. Alzheimer's disease).

It is not yet known how the DSM–5 criteria for dementia will perform in this population, but in the general population, DSM–IV criteria have been found to be more inclusive compared to ICD10 criteria. This was also the case in people with intellectual disabilities (Strydom et al., 2007). ICD10 and DSM–IV dementia criteria both showed substantial reliability and satisfactory validity in older adults with intellectual disabilities without Down's syndrome, although caution is advised when using these criteria in people with more severe intellectual disabilities and/or co-morbid sensory disabilities and predictive validity was slightly worse than in the general population (Strydom et al., 2013). ICD10 criteria were more likely to accord with clinician diagnoses than DSM–IV criteria in people with Down's syndrome who have been diagnosed clinically with dementia, possibly due to the inclusion of behavioural symptoms in the ICD10. However, both sets of criteria underdiagnosed dementia compared to clinician judgement, suggesting that experienced clinicians used their clinical knowledge of the slightly atypical dementia presentation in Down's syndrome to diagnose the disorder at an earlier stage than would be possible if they applied the diagnostic criteria (Sheehan et al., 2014).

## 5.2 Course of the disease

Dementia is at present untreatable and has a progressive course. In the general population each of the different types of dementia has characteristic clinical features. The presentation and course of dementia has been studied in people with Down's syndrome but less so in those with intellectual disabilities. However, some general observations can be made.

### People with Down's syndrome

The most common form of dementia in people with Down's syndrome is Alzheimer's disease. It is known that the brain pathology of Alzheimer's disease is almost universally found in later life in people with Down's syndrome. Vascular disease, and therefore the risk of dementia of a vascular origin, is rare.

Dementia in people with Down's syndrome may present atypically with changes in behaviour and/or personality that can precede the full clinical picture of dementia by some years.

A number of studies have reported that behavioural changes, rather than functional memory decline, are the early signs of developing dementia and Ball et al. (2006) have proposed that this may be explained by limited reserve capacity of the frontal lobes of people with Down's syndrome. Longitudinal neuropsychological studies have also found that some discrete cognitive abilities, as measured using established neuropsychological tests, show evidence of increasing impairment over time starting in the early and pre-

clinical stages of dementia. Thus memory and orientation are affected early, with praxis, language and visuo-spatial skills becoming impaired later in the progression of the disorder (see Ball et al., 2006 for review).

The behavioural problems associated with dementia may take the form of behavioral excesses such as irritability, aggression or self-injury and then behavioral deficits which would include slowness, apathy loss of interest and lessened social engagement (Oliver et al., 2011).

The middle and later course of Alzheimer's disease in people with Down's syndrome and for those with other causes for their intellectual disabilities are comparable in characteristics to those experienced by people in a similar stage of dementia but without pre-existing intellectual disabilities, though in individuals with Down's syndrome neurological symptoms are common, and include the onset of seizures for the first time in that person's life. Late stage symptoms are characterised by the individual's lack of response to the environment, loss of mobility, loss of communication skills, incontinence, seizures and may include Parkinsonian features (Visser et al., 1997; Strydom et al., 2010).

The progression of dementia in people with Down's syndrome with dementia has been reported to be more rapid than in the general population. However, it may be that diagnosis occurs later in this population (Bush & Beail, 2004).

People with intellectual disabilities are at high risk for additional co-morbid illness that might impact on both the way that dementia presents and its progression over time. Unexpected deterioration or changes in presentation or course should be investigated in case there are treatable additional physical or psychiatric co-morbid illnesses.

### **Atypical presentations in people with Down's syndrome**

There have been anecdotal reports of people with Down's syndrome in their teens or early adult life who deteriorate, often following a life event, and either never or, only after many months or years, recover. There have been limited reports in the literature (e.g. Worley et al., 2014) but the characteristics of the decline may superficially resemble that of dementia or depressive illness but it neither seems to progress (as would be expected with dementia) or resolve (as would be expected with depressive illness).

The clinical picture is dominated by the development of a general slowness in mental and/or physical activity, apparent loss of interest in previous activities, and a level of functioning that is below that previously observed. In some people the presentation may include autistic features. At present it is unclear how such problems should be best conceptualized. If depressive illness is a possible factor, a trial of anti-depressant medication may be indicated, with careful monitoring of outcomes. Other conditions such as Hashimoto's encephalopathy catatonia and neurological diseases need to be excluded (Brodthmann, 2009, Jap & Ghaziuddin, 2011). Regular review of neuropsychological function is helpful in order to check that there is no progressive disorder such as dementia. Therapeutically the approach taken is primarily a rehabilitative one with attempts to help the person progressively back to their previous state of function.

### **People with intellectual disabilities without Down's syndrome**

For those people with intellectual disabilities not due to Down's syndrome, the picture may be variable. There is more likely to be a range of pathologies resulting in dementia, as is



the case in the general population, and for the same reason differences in presentation and the course of the dementia.

Experience from the study of older people with Down's syndrome suggests that the clinical presentation may be affected by an interaction between how brain development is affected due to having Down's syndrome and the developing pathology of Alzheimer's disease. The same is likely to be true among people with intellectual disabilities not due to Down's syndrome as there will be a great variation across this population in terms of level of abilities. The following are likely to be useful guiding principles with respect to the presentation and course of dementia in those with intellectual disabilities not due to Down's syndrome.

- The presentation and course of dementia among people with mild intellectual disabilities is likely to be similar to that which is observed in the general population.
- The presentation and course of dementia in people with more severe intellectual disabilities may initially be atypical and present with changes in behaviour and for this reason dementia may not be suspected. As the illness progresses careful questioning of staff and carers may identify evidence of developing memory and functional impairments or neurological symptoms such as incontinence and dysphagia.
- The age of onset of dementia in this group of people may be a few years earlier than is commonly found in the general population but not as early as in people with Down's syndrome (Strydom et al., 2007).
- As with those in the general population it is important to investigate the likely cause of dementia as the full range of causes for dementia are likely to be found in this group of people and this may have important therapeutic and management implications.
- There is evidence that the onset of epilepsy in later life for the first time may be a marker for developing dementia in people with Down's syndrome. Whether this is the case for other people with intellectual disabilities is uncertain but onset of epilepsy in later life for the first time should always be investigated and increasing difficulty controlling pre-existing epilepsy may be an indication for considering the possibility of developing dementia.

### Key points

- The course of dementia in people with Down's syndrome has been well studied, and may be atypical with early development of behavioural or personality change, though memory problems are also prominent.
- New onset of epilepsy or worsening of existing epilepsy in an older person with Down's syndrome should always raise the possibility of Alzheimer's disease.
- Diagnostic criteria are reliable in the intellectual disability population, but dementia is more difficult to diagnose in those with severe disabilities or comorbid problems and may require sequential assessment. Clinicians should also keep in mind the slightly atypical presentation of dementia in people with Down's syndrome.
- A small number of young people with Down's syndrome seem to present with decline in their teens or early twenties, often with no clear aetiology.
- People with intellectual disabilities without Down's syndrome who develop dementia may have the same range of pathologies as the general population.

## Section 6 – Assessment

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There is great variability of functioning within the population of people with intellectual disabilities. Comparison with ‘peer-related’ norms is not possible as it is with mainstream dementia assessments. Assessment of decline needs to be personalised to each individual, with their own unique ‘baseline of functioning’ being the comparison when concerns arise. Holistic assessment should also encompass wider systemic issues (e.g. Care Quality Commission (CQC) concerns, Safeguarding Adult investigations). These issues can be identified through thorough informant and carer interview, direct observation as well as gathering knowledge from members of the multi-disciplinary team.

NICE Quality Standard 1, *Dementia* (NICE, 2010) states: ‘People with suspected dementia are referred to a memory assessment service specialising in the diagnosis and initial management of dementia.’ The process of assessment essentially has three stages that follow NICE guidance for dementia (NICE, 2006). For people with intellectual disabilities, this usually occurs within the context of the intellectual disability service rather than in mainstream memory services due to the specialist skills and expertise in assessing people with learning disabilities (Barrett & Burns, 2014).

Assessment should include a file review and systematic history-taking from the person and multiple informants across settings and services, who have known the individual for a significant period of time.

Assessment should include physical and mental state examinations, cognitive assessments and other investigations to enable the evaluation of present functioning and the identification of other possible causes of decline. The medical investigations should be guided by the clinical picture but invariably include investigations of a person’s basic physical state and specific tests, such as measures of thyroid function and B12, or specialist assessment of hearing and/or vision. Where the clinical picture is unusual, the diagnosis in doubt, or there are features that prompt concern (e.g. focal findings on neurological exam, a recent fall/head injury, rapid or sudden decline) a CT or MRI brain scan may be indicated (refer to Section 4 for detail regarding differential diagnosis and below for further details regarding scans).

The diagnostic process leads to a formulation that brings together all of the information from the various interviews, assessments and investigations and finally determines the likely cause of the observed clinical changes and sets them in the context of the individual. This forms the basis for making a possible diagnosis and developing an individualised care plan.

### 6.1 Assessment process

The section outlines in more detail the key factors to be considered during the assessment process.

#### 6.1.1 History and information gathering

- Nature of the presenting problem(s), origin, rate/pattern of progress (sudden or gradual), presence of seizures and other associated conditions, impact on the person’s overall functioning and personality.

- History of significant physical and medical history including past and present medical conditions, e.g. diabetes, hypertension, thyroid or cerebrovascular disease, B12 deficiency.
- History of or current presence of psychiatric symptoms such as depression, anxiety or other mental health problems.
- Record developmental history and best level of historic functioning. Ascertain if there are any previous neuropsychological test data on record and compare data with previous assessment results. Record historic daily living skills, interests/hobbies/skills and details of personality.
- Family history: dementia or other mental health and medical conditions (particularly in first-degree relatives).
- Assess for psychosocial issues, changes or life events. These include house moves, health decline/death of loved ones, change of caregivers, changes to, or retirement from work/day services.
- Information gathering should be undertaken through a combination of informant interview (preferably with a family member, when relevant and appropriate) or an informant who has known the person well for a period of six months at least) and directly from the person where possible.

### 6.1.2 Mental state examination

Observation of level of alertness, orientation to time, place and person, any evidence of alterations in consciousness, psychomotor activity, mood, thoughts, evidence of any abnormal mental beliefs or experiences, and perceptual abnormalities. Evaluation of memory and other cognitive functions via formal assessment (see below).

### 6.1.3 Physical examination

Where possible a complete physical examination should be undertaken. This should occur in the context of primary care, although support may be provided by specialist services.

The key issues are:

- Cardiovascular system – focal deficits, evidence of cardiovascular accident etc.
- Detailed neurological examination (focal deficits, gait abnormalities, speech abnormalities, etc.).
- Endocrine system: signs of hypothyroidism.
- Careful recording of historic/newly added/current medications (with particular attention to those that are psychoactive, antiepileptic, sedating or anticholinergic).

Complete physical examination may be difficult in a significant proportion of individuals. The physical health issues could be addressed in these individuals by a combination of:

- brief physical checks, e.g. blood pressure, pulse;
- observations for any evidence of physical health issues;
- observational tools (e.g. pain questionnaires); and
- information from carers, e.g. The *OK Health Checks* (Matthews, 2006), *Health Action Plans* (DH, 2001) or bespoke nursing assessments developed to meet the needs of local intellectual disability services can be used to structure this process.

#### 6.1.4 Physical investigations

Recommended routine investigations are:

- full blood count;
- urea & electrolytes;
- blood sugar;
- thyroid function tests;
- liver function tests;
- B12 and Folate level;
- lipid profile; and
- sensory screening – vision and hearing.

Optional tests are:

- Electro encephalograph (EEG) – if there is evidence of the occurrence of seizures.
- Neuro-imaging: this may not be practical as a routine investigation for the assessment of dementia but may be of value when vascular dementia or other brain lesions are suspected (see section on ‘Neuroimaging’ below).
- Electro Cardiograph (ECG) – particularly if anti-dementia medication is to be used or there is evidence of cardiovascular problems.

#### 6.1.5 Environmental assessment (see also Section 11)

- Quality of the person’s physical environment.
- Staffing levels (day and night).
- The mix of people with intellectual disabilities in the residential and day care settings.
- Quality and quantity of day activities.
- Staff characteristics: attitudes and competence, including consistency of approach.
- Scrutiny/review of historic/current support package.

#### Key points

- Carrying out investigations may be difficult in some individuals with intellectual disabilities. In this situation, clinicians have to make a decision using information from the history, physical assessment and direct observation as to whether investigations are essential or not. There may be scope to work with the person to improve the chances of success in engaging with physical assessments (e.g. systematic desensitisation for blood tests, brain scans, eye-tests etc.).
- Mental Capacity Act (2005) and its code of practice guidelines must be followed where investigation is considered and the person is unable to consent and cooperate.

*A minimum reactive dementia assessment should include/consider:*

- Physical health check.
- Longer-term and recent life events.
- Semi-structured interview exploring negative change in relation to functional skills, memory, behavior, orientation, mood and consider potential alternatives for these changes.
- Mental state examination (may utilise a depression or anxiety measure).
- Objective assessment of cognitive and adaptive skills and abilities.

## 6.2 Information to support the assessment process

Assessment tools for dementia in the general population are not appropriate for people with intellectual disabilities. For example, widely-used tools such as the Mini-Mental State Examination (MMSE) (Folstein & Folstein, 2001) or the Addenbrookes Cognitive Examination – 3rd Edition (Hodges, 2012) assume the pre-morbid level of functioning to have been within the average range.

There are a wide variety of tools utilised in the UK to assess dementia in people with intellectual disabilities. A systematic review (Zeilinger et al., 2013) found some measures currently used in the UK were neither designed for the assessment of dementia, nor for people with intellectual disabilities. There is currently no agreed battery of assessments with which to assess dementia in this population and there is often great variation in screening/assessment methods. There continues to be a lack of research data to secure agreement in order for there to be better uniformity across services and for future data to be pooled and compared. Lack of research data and the nature of the intellectual disability population requires each individual to be viewed independently in terms of their own functioning. Cognitive and informant-based assessments serve as a unique baseline to that individual.

- Colleagues in the US (National Task Group on Intellectual Disabilities and Dementia Practices (NTG), Moran et al., 2013) have recently developed practice guidelines that helpfully describe the key stages of a dementia evaluation in this population and has published an Early Detection Screening Tool (NTG-Early Detection Screen for Dementia).
- Always look for evidence from previous assessments on file that might indicate pre-morbid functioning. Where possible, the same test should then be repeated for comparison.
- The tests used should include (where possible) direct assessment with the person and questionnaire/interview-based assessments with well-informed carers. Information should be collected from both carers at home and from carers within the person's day services to ensure concerns are not situation-specific.
- For some individuals with profound intellectual disabilities, pre-morbid cognitive ability may be so poor that changes may not be detected by any available standardised testing. Carer reports have to take precedence.
- Whatever battery of assessments is chosen, it should be used longitudinally within the service to enable comparison of performance over time for that individual.

## 6.3 Direct testing with the person with Down's syndrome/ intellectual disabilities

It is well recognised that the early changes associated with dementia in people with Down's syndrome can often relate to behaviour or personality rather than memory functioning (Oliver et al., 2011; Adams & Oliver, 2010; Ball et al., 2006b, 2008, 2010). These studies suggest that there is often compromised function associated with the frontal lobes early in the course of the disease. For this reason, assessments that tap into executive functioning are important to consider, especially in terms of establishing baselines when a person is still healthy and well.

Assessments should cover as a minimum (although may be dependent on intellectual functioning and ability to engage in direct assessment):

- a validated instrument for the cognitive assessment of dementia in people with intellectual disabilities (see ‘Neuropsychological Assessments and Informant Questionnaires’ sections below for examples);
- prospective, short- and long-term memory (visual and verbal);
- executive functioning;
- orientation;
- language (expressive and comprehension); and
- recording of evidence of new learning.

Other assessment to consider:

- Mood state.
- Examples of reading/writing/mathematic skills can support a helpful baseline to ascertain changes to these skills over time. Examples/references to these skills can often be found in historic files.
- Direct questioning of known previous skills or knowledge personal to the individual (e.g. order of potting balls in snooker, knowledge about a favourite entertainer.)

This is not a complete list and the assessor need to be responsive to assessing specific issues in greater detail if required (e.g. praxis, attention, perception). The tools listed below are those most commonly used in services in the UK. Many services have developed their own ‘assessment battery’ combining a variety of psychometric measures and assessments that tap into the areas of functioning described above. Specific recommendations cannot be made until there is more research evidence comparing their efficacy.

### 6.3.1 Direct neuropsychological assessment

**Neuropsychological Assessment of Dementia in Adults with Intellectual Disabilities (NAID)** (Crayton et al., 1998). The NAID is a battery of very simple tests covering memory, orientation, language and praxis. There is no manual, rather the instructions are in Crayton et al. (1998) and data in Adams and Oliver (2006). This battery takes about 45 minutes to administer. The majority of people with Down’s syndrome can attempt most of it. It is said by the authors to be in use in over 30 intellectual disabilities services in the UK.

**CAMCOG-DS** is the neuropsychological assessment part of the CAMDEX-DS (Ball et al., 2006). It is a concise neuropsychological test battery which is based on the CAMCOG, which was designed to meet the need to assess all the cognitive deficits specified in criteria for dementia, i.e. memory impairment, aphasia, apraxia, agnosia and disturbance in thinking (executive function). The CAMCOG-DS includes assessments of orientation, language, memory, attention, praxis, abstract thinking and perception, giving individual subscale scores as well as a total score.

**Severe Impairment Battery (SIB)** (Saxton et al., 1993). The SIB is designed to assess cognitive abilities at the lower end of the intellectual range in the general population (age range 51–91). There are 40 items and administration is said to take about 20 minutes. It is composed of very simple one-step commands which are presented in conjunction with gestural cues (e.g. ‘what’s your name?’, ‘please write your name here’, ‘what do you call the thing you drink coffee from?’). The SIB is divided into scoreable subscales, each sampling

within the range expected of the severely-impaired individual. The six major subscales are attention, orientation, language, memory, visuospatial ability and construction. There are also brief evaluations of praxis and the person's ability to respond appropriately when his/her name is called (orienting to name). In addition, there is an assessment of social interaction skills. It yields scores out of 100, to assess mild to moderate dementia. There is no cut-off for 'normal' as the test should only be used with people known to be severely impaired.

**Test for Severe Impairment** (Albert & Cohen, 1992). This is a 24-item test covering eight domains which was designed for people with severe cognitive dysfunction, but not specifically intellectual disabilities. This test may not be sensitive to change over time, and only includes a few memory items.

### **Tests of executive functioning**

Whilst there have been some recent studies exploring the measurement of executive functioning in people with intellectual disabilities (e.g. Ball et al., 2009) there is currently no clear agreement as to which tests should be adopted for the assessment of dementia.

Individual assessments such as tests of verbal/category fluency, response inhibition tests such as the Cats and Dogs Test or scramble boxes (see Ball et al., 2008 for descriptions), and set/rule switching tasks such as card sorting tests (Weigl sorting; Dimensional Change Card Sort test) are sometimes used as a part of wider bespoke test batteries to establish functioning in these areas. This is helpful when setting a baseline of functioning that may be used for future comparison.

Executive test batteries developed for the non-intellectual disabilities population (such as the Delis-Kaplan Executive Function System – D-KEFS (Delis et al., 2001) or the Behavioural Assessment of the Dysexecutive Syndrome – BADS (Wilson et al., 1996) tend to be too difficult for many people with intellectual disabilities, although some sub-tests (e.g. key search) may be used successfully in people with mild intellectual disabilities.

The BADS-ID (Webb & Dodd, 2014) is under development and may assist in filling the current gap in relation to suitable tests of executive functioning in people with intellectual disabilities.

The Measure of Everyday Planning (MEP) is a tool under development (Webb et al., 2014) and is a flexible tool designed to help identify the issues underlying difficulties that adults with intellectual disabilities may have with independently initiating, planning and carrying out everyday activities. It explores subtle factors that can impair the performance of individuals who, superficially, appear to have the ability to carry out tasks but, in reality, struggle with them.

The informant/carer versions of the DEX (found in the BADS, Wilson et al., 1996) and the Behaviour Rating Inventory of Executive Function – Adult Version – BRIEF-A (Roth et al., 2000) may be useful in ascertaining areas of deficit in relation to executive functioning. These can be used as baseline and repeated at intervals, or when concerns are raised to monitor changes in symptoms common to those with executive dysfunction.

## 6.4 Informant questionnaires

These should aim to cover those areas of function that are known to deteriorate with the development of dementia including: short and long term memory, general mental functioning, dyspraxia and dysphasia, daily living skills and personality and behaviour.

**Dementia Questionnaire for People with Learning Disabilities – DLD** – formerly known as the DMR (Evenhuis et al., 2007). The DMR and its recent successor, the DLD, is widely used to longitudinally assess the development of dementia in adults with intellectual disabilities in the UK and Europe. It is a screening tool for the early detection of dementia in adults with intellectual disabilities, completed by carers, consisting of 50 items. There are eight sub-scales: short term memory, long term memory, orientation (making up Sum of Cognitive Scores), speech, practical skills, mood, activity and interest and behavioural disturbance (making up Sum of Social Scores). Evenhuis (1992) reported that the DMR had sensitivity of up to 100 per cent in identifying dementia and suggested change scores and cut-off scores that might be indicative of dementia. Prasher (1997) conducted an independent evaluation of the DMR on 100 adults with Down’s syndrome in the UK and, finding poor specificity, suggested modifications to the cut-off scores. A prospective 14-year longitudinal study (McCarron et al., 2014) stated the DMR/DLD to be the most sensitive tool in their battery for tracking change in symptoms over time.

**The Dementia Scale for Down Syndrome – DSDS** (Gedye, 1995). This is designed for use with people with Down’s syndrome but may also be useful for people with intellectual disabilities generally according to NICE (2006). It gives a measure of early, middle and late stages of dementia and includes the time course of the deterioration and a differential diagnosis scale. The psychometric property of the DSDS has never been published in a peer-reviewed journal. Its administration is restricted to clinical and other qualified psychologists and trained psychometricians.

**Dementia Screening Questionnaire for Individuals with Intellectual Disabilities – DSQUIID** (Deb et al., 2007). The DSQUIID is designed to be ‘a user-friendly observer-rated dementia screening questionnaire with strong psychometric properties for adults with intellectual disabilities’, according to the authors. It comprises 43 questions in three sections. The scoring system overcomes the floor effect found in some other assessments.

**Adaptive Behaviour Dementia Questionnaire – ABDQ** (Prasher et al., 2004). This is a 15-item questionnaire derived from the AAMD Adaptive Behavior Scale (Nihira et al., 1974) which is used to detect change in adaptive behaviour. It has been developed to screen specifically for dementia in Alzheimer’s disease in people with Down’s syndrome. It sets out to collect information on how the person compares now to their previous normal level of social functioning. It gives criteria for the presence of Alzheimer’s disease and a rating of severity, but the threshold scores may require revision as these do not appear to be accurate in clinical practice.



## 6.5 Measures of psychological issues

### 6.5.1 Measures of mental health

**The PAS-ADD Checklist** (Moss, 2002a) is a 25-item questionnaire designed for use primarily by care staff and families to help them decide whether further assessment of an individual's mental health may be helpful. The scoring system includes threshold scores which, if exceeded, indicate the presence of a potential psychiatric problem, which may then be more fully assessed using the Mini PAS-ADD. The PAS-ADD Checklist produces three scores, relating to affective or neurotic disorder; possible organic condition (including dementia); and psychotic disorder.

**The Mini-PAS-ADD Interview** (Moss, 2002b) is designed to provide highly reliable information on psychiatric symptoms, usually by informant interview. The assessment produces scores relating to seven diagnostic categories: Depression; Anxiety; Expansive mood; Obsessive compulsive disorder; Psychosis; Unspecified disorder (mostly dementia and other organic problems in our field-trial sample); and Autistic spectrum disorder. Threshold scores are provided for each of the above seven diagnostic areas. If the person reaches or exceeds a threshold, the implication is that they probably warrant a diagnosis. However, a strong emphasis is placed on clinical interpretation of the results.

### 6.5.2 Measures of depression

**The Glasgow Depression Scale** (client version) – GDS-LD (Cuthill et al., 2003). A 20-item questionnaire designed for use with people with intellectual disabilities. It has a three point Likert-type response scale ('never', 'sometimes', 'a lot'). It has a suggested cut-off score for suspected depressive illness.

**Glasgow Depression Scale** (Carer Supplement) – GDS-CS (Cuthill et al., 2003). This is a 16-item questionnaire about depressive symptoms, completed by a carer. It is designed to be applicable to people with intellectual disabilities. No cut-off scores are suggested by the authors, so its usefulness depends on repeat completion, although one-off use can highlight obvious areas of concern.

### 6.5.3 Measure of anxiety

**Glasgow Anxiety Scale** (Mindham & Espie 2003) is a 27-item tool to help discriminate anxious from non-anxious clients with mild intellectual disabilities. It is reported to have good test-retest reliability and internal consistency.

### 6.5.4 Carer burden

**Caregiver Activity Survey-Intellectual Disability** – CAS-ID (McCarron et al., 2002) provides a measure of carer burden that can be helpful as a measure of increasing care needs and therefore in determining level of social care required. Carers record the amount of time needed to care for a person with intellectual disabilities in a 24-hour period, across eight domains such as nursing care, behaviour, supervision and personal hygiene. Care staff require careful instructions to complete it reliably.

### 6.5.5 Life events scales

These are useful to gauge whether the presenting symptoms may be attributable to an adverse life event. There are many different ones available or they may be designed by the assessor to cover typical life events occurring in the past two years.

## 6.6 Assessments of adaptive functioning

### 6.6.1 Assessments of daily living skills through direct observation of the person

If repeated at intervals, these can shed additional light on changes in skills. The Assessment of Motor Process Skills (AMPS, Fisher, 2006) is utilised in some services, although its delivery is limited to occupational therapists who have received specialist training, and it can be hard to generalize the findings.

### 6.6.2 Assessments of daily living skills completed with a carer

If repeated at intervals, these can shed additional light on changes in skills. There are many to choose from including:

- **AAMD Adapted Behaviour Scales, 1974 revision** – ABS (Nihira et al., 1974), which is a standardised measure of daily living skills and maladaptive behaviour. It can be useful for assessing those with profound intellectual disabilities, who cannot undertake direct assessments, but may be cumbersome to complete.
- **Hampshire Social Services – Staff Support Levels Assessment** (Hampshire Social Services, 1987) – a 32 item assessment which has behavioural anchors and therefore reports excellent test–retest validity and inter-rater reliability.
- **Vineland Adaptive Behaviour Scales: Second Edition** (Sparrow et al., 2007) provides analysis of a broad range of skills and behaviour. These include communication, daily living skills, socialisation, motor skills and maladaptive behaviour. Vineland Adaptive Behaviour Scales: The Second Edition is available in both long and short form, providing either a summary assessment of adaptive behaviours or a more detailed assessment. Semi-structured interview and questionnaire formats make it easier to assess those who have difficulty performing in test situations. It covers 0–90 years. It is an American tool and hence some of the items will be unfamiliar to UK users, and like the ABS and ABAS-II may be cumbersome to complete.
- **Adaptive Behaviour Assessment System-II** – ABAS-II (Harrison & Oakland, 2003) provides a comprehensive norm-referenced assessment of the adaptive skills of individuals aged from birth to 89 years. The clinician can use the ABAS–II to diagnose and classify disabilities and disorders; identify an individual’s strengths and limitations; and to document and monitor the individual’s performance over time. It provides standard scores that are directly comparable to those found in the Wechsler IQ tests. It too is an American publication.

In looking at changes in adaptive functioning, it is important to assess not only the ability of the person to complete each task, but to probe carefully whether there is a qualitative change in performance of each task.

## 6.7 Who conducts assessments and how are they organised?

A dementia care pathway is helpful to guide the multi-disciplinary team through the sequence of events from referral, through screening and diagnosis, to treatment/intervention and end of life care. It helps to ensure a co-ordinated approach and effective multi-disciplinary working. Typically, a community nurse may conduct the health screen as the first stage of assessment, in partnership with the GP and/or intellectual disabilities psychiatrist for necessary additional tests or investigations.

The assessments of memory, mood and behaviour are often conducted by clinical or other qualified psychologists but in some cases, may be undertaken by other multi-disciplinary team members. The need to refer for specialised psychological assessment for people with intellectual disabilities was identified in Department of Health guidelines for GPs in 2014 (Barrett & Burns, 2014). There is a need to ensure that non-psychologists undertaking such assessments are suitably trained in undertaking psychometric assessment that are well supervised in the formulation and interpretation of the data. This is commonly provided by clinical or other qualified psychologists.

AMPS assessments, if used, require special training (usually the preserve of occupational therapists). The role of the intellectual disabilities psychiatrist or clinical or other qualified psychologist is crucial to making the differential diagnosis once all the assessment data have been collected.

Some services have a dedicated or virtual team using an agreed battery of assessments (e.g. Cairns et al., 2010; Jervis & Prinsloo, 2007; McBrien et al., 2005). This may be known as a memory clinic or dementia assessment/screening programme. The team should at the minimum include a psychiatrist in intellectual disabilities, clinical or other qualified psychologist and community nurse.

### **6.7.1 Where to see people/observations**

Careful attention needs to be paid to where, when and how to assess individuals. A holistic assessment should include meeting the person being assessed and their carers in their normal living and daytime environments. Cognitive assessments may be more effectively offered in a healthcare or similar setting if this can provide consistency for repeat assessment and provide standardised testing conditions that are free of distractions and give an appropriate context. However, this would need to be balanced with potential client anxiety and a better picture of functioning which can be achieved at the person's home or other place preferred by them.

### **6.7.2 Explaining assessments to people with intellectual disabilities**

It is important that the assessment process is discussed with the person with intellectual disabilities, and that their consent to participate is obtained. People will need to have different communication methods used that are tailored to the individual, including clear verbal communication and the use of picture booklets to explain the assessment process. Explanations should be related to people's prior understanding of the issues, and couched in a way as not to cause the person anxiety, with an emphasis on helping people to understand the process and the support available. Assessment of the person's capacity may need to be undertaken, and where the person does not have capacity, the decision re assessment needs to be made using a best interest process.

### **6.7.3 Conditions for reassessment**

Repeat cognitive assessments need to be rigorously administered and interpreted to take account of normal fluctuations in the assessed person's performance (e.g. tiredness, mood, good day/bad day), specific changes (e.g. hearing aids, glasses, medication effects), testing environment changes (place, layout, distractions) and tester effects (skills, experience, relationship with the person with intellectual disabilities, testing style, administration and scoring anomalies). Ideally the same tester should use the same tests in the same

environment using strict criteria for similar administration/prompts, and where possible seeking information from the same informant on each occasion, although this may not be possible in practice. The standard error of measurement and normal ageing deterioration must also be considered when considering results showing some evidence of cognitive deterioration.

### Key points

- Multi-disciplinary assessment is important.
- Assessments should include direct assessment of the person together with preferably, multiple-informant based questionnaire/assessments.
- Staff undertaking psychometric assessment as a part of the assessment process need to be suitably trained in psychometrics and receive appropriate supervision.
- Assessment for other co-morbid conditions is essential.
- Consideration should be given to practical issues in assessment (e.g. location).
- Consider test re-test issues (e.g. different informants/testers).

## 6.8 Neuro-imaging

### 6.8.1 Indications

The most consistent structural change in the early stage of Alzheimer's disease is the atrophy of the medial temporal lobe.

People with Down's syndrome have medial temporal lobe atrophy even without dementia. However, normative values have not yet been established, so neuro-imaging currently has limited value in the early diagnosis of Alzheimer's disease in people with Down's syndrome. Its value is mainly to rule out structural lesions other than atrophy (e.g. space occupying lesions). It should therefore be used only when the clinical picture suggests the possibility of such lesions.

MRI scans have a number of advantages over CT scans. However the sensitivity of CT scans has improved considerably and the procedure is shorter and less complex than for MRI scans.

### 6.8.2 Informing and preparing

Neuro-imaging requires explicit informed consent. Detailed information regarding the rationale and the procedure should be given to the patient in an appropriate user-friendly format. Assessment of capacity will be needed to identify whether the individual is able to provide valid consent for the procedure. Where the person does not have capacity best interest principles will need to be applied.

Anxiety about the procedure can be allayed by a visit to the radiology unit and familiarization with the procedure, and occasionally sedation may be required.

### 6.8.3 Process

Although, with adequate preparation, some people with intellectual disabilities may be able to go through the procedure without any other interventions, others may benefit from the use of one dose of oral anxiolytic medication such as lorazepam or diazepam an hour beforehand. Some clinicians prefer to use buccal midazolam, which provides rapid and short-term sedation and therefore may be given immediately before the procedure. Some individuals may find it difficult to have a scan in spite of all these measures.

Generally, the MRI procedure is longer and more anxiety provoking. In these situations, discussion with the radiologist may be helpful in deciding if CT can be used as an alternative. New generation CT scans are much more user friendly and less anxiety provoking.

If the individual clearly needs neuro-imaging but is unable to co-operate in spite of all these measures, it can be undertaken under general anaesthesia. However, risks and benefits should be evaluated and thorough discussion held with the carers and other professionals to arrive at a best interests decision, if the person lacks the capacity to consent to this specific investigation. If the person has the capacity to consent to the scan it is for him/her to decide whether or not to have the scan, after being given clear information on the procedure, risks and benefits.

### 6.8.4 Sharing the findings

It is important to share the findings of the scan with the person with intellectual disabilities, carers and other professionals. This should include the nature of the findings as well as implications for the management. This discussion should be clearly documented.

#### Key points

- Neuro-imaging (CT/MRI) is not an essential investigation for the diagnosis of dementia in Down's syndrome.
- It may be of value where other brain lesions/vascular dementia are suspected.
- New generation CT scanners are as sensitive as MRI scanners and may be more acceptable for people with intellectual disabilities.
- Amyloid PET scans may become more available in the near future, but its value in the diagnosis of dementia in individuals with Down's syndrome who all have amyloid deposits in their brains remain uncertain.

## Section 7 – Establishing the diagnosis and breaking the news

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### 7.1 Background to an effective and person-centred approach.

The increased risk of developing dementia at an earlier age is well-established for people with intellectual disabilities, and particularly for people with Down's syndrome (see Section 2 – Epidemiology). This knowledge raises the ethical dilemma of how this information should be sensitively communicated to the individual and to families and carers. It can be helpful to begin conversations during the transition process, building from people's current understanding of the aetiology of their intellectual disabilities. Ongoing conversations can explore people's awareness of health risks that may develop over their lifetime, together with specific risks associated with different syndromes.

The awareness of difference is personally challenging and it is likely that this process will be painful and it will be necessary to give ongoing support.

There is much work to be done in this area as currently many individuals with intellectual disability have not been informed about the nature of their intellectual disability and any associated health risks. The individual and their network of support need to be given the opportunity to understand how age-related risks can be monitored and identified at the earliest possible stage by regular health screening and the promotion of healthy ageing (see Section 3 Baseline and Monitoring). They also need to be informed of what assessment and interventions, including adaptations, accommodation, support and care, are available if difficulties associated with dementia do develop (see Section 11 – Environments and Section 12 – Interventions).

### 7.2 Establishing the diagnosis of dementia

The first step in establishing the diagnosis is to bring the assessment information from different sources together (see Section 6 – Assessment). It is important for the multidisciplinary group to combine the assessments of the psychiatrist, psychologist, occupational therapist, community nurse, speech and language therapist and other relevant professionals with the results of the investigations and physical examination that the GP or psychiatry team may have carried out. The clinical or other qualified psychologist and psychiatrist will be the key disciplines involved in reviewing the outcome of multidisciplinary assessment and then arriving at a diagnosis, with support from the multidisciplinary team.

#### 7.2.1 Diagnostic process

For a significant proportion of individuals, it may not be possible to reach a clear diagnosis at an early stage. It may only be possible to have a range of differential diagnoses with a true picture emerging over a period of time (see Section 4 – Possible Reasons for Decline). The diagnostic evaluation may need to be repeated regularly, e.g. at six monthly intervals. It is worthwhile remembering that for some individuals at this stage the diagnosis of dementia may be clearly ruled out and the presenting problems formulated in a different way and appropriate interventions offered.

Assessment information may need to be referred to regularly or may not need to be referred to again for several years, therefore it is important to ensure copies are securely held where they may be found in future. Some services copy their assessment reports to the GP, the residential home or family carer as well as to the person's NHS file held within the local service for people with intellectual disabilities. Accessible reports should be considered for the person with intellectual disabilities.

### **7.3 Sharing the information with the person with intellectual disabilities**

Consistent with national guidance concerning sharing information about diagnosis of serious illness with the patient, all UK dementia strategies state that everyone should be told their diagnosis and this has been conceptualised as a human right.

A person-centred approach that does not further isolate, depersonalise and marginalise the person means steps should be taken to share the information with the person with intellectual disabilities as early as possible, and at all stages, so that they can be supported to understand and cope with their changing experiences (Watchman, 2012; Tuffrey-Wijne & Watchman, 2014).

This will also help them be involved as much as possible in decisions about their support and care and medical treatments, including concerning establishing their views and preferences about future care before their illness progresses.

Where health professionals from services other than specialist teams for people with intellectual disabilities are in direct communication with the person concerning their diagnosis and illness, they may need support and guidance concerning the communication needs of the person with intellectual disabilities, as generic breaking-bad-news models are not likely to meet their needs.

However, the process of sharing the diagnosis also needs to be sensitive to the expressed wishes of the individual concerning knowledge of their physical health and illness and, as for the general population, it is likely to be painful for the person to hear this information and accept their diagnosis.

However, it must be undertaken as good practice irrespective of the mental capacity of the person concerned so that the person is sensitively given opportunities to know about the health changes they are experiencing and a psychological approach, with additional direct psychological interventions when necessary, should be used to give them emotional support and lessen anxiety and distress.

A wide range of personal factors, including culture, personal history and current relationships, also need to be taken into account when deciding how to share the diagnosis and to facilitate an understanding of the illness and experiences.

The model of best practice concerning breaking the news to people with intellectual disabilities emphasises the need to communicate information over time and based on 'chunks' appropriate to the person's current framework of knowledge and lived experience and their awareness of the future.

All those who know the individual can contribute to a current understanding of the

person's information needs and preferences and how material can be presented to maximise understanding and retention. This process may involve additional input from a speech and language therapist to make the information as accessible as possible, including when the person no longer communicates verbally. As the illness progresses the person's framework of knowledge will change and is likely to be related to an earlier period of their life.

Practice guidelines, developed within the multidisciplinary team, concerning how to approach communicating with the person about their dementia and with suggestions about how to personalise this, can be helpful.

In order for the person to have adequate support it will generally be appropriate to share the diagnosis with family, friends, carers and support workers around the time of telling the person themselves.

This is an ongoing process and all involved with the person, whether multidisciplinary professionals, carers in different contexts, and family members, need to feel confident to give accurate and accessible information to the person and to be supported to give this information repeatedly and helpfully in the context of changing information needs. The psychologist and other members of the multidisciplinary team are an important resource that needs to be available to support those in day to day contact with the person about their continuing communications concerning the diagnosis and the person's changing experiences, whilst still maintaining a positive approach to support. The use of a 'communication passport' that incorporates the communication needs linked to the progression of dementia can be very helpful.

Booklets such as *The Journey of Life* and *About Dementia* (Dodd et al., 2005 a, & c), and *What is Dementia?* (Kerr & Innes, 2000) are useful resources for helping the person with intellectual disabilities understand their condition and experience, although it is clear they should always be introduced with sensitivity and care.

## **7.4 Sharing the information with family and carers**

This is also a complex process as it will involve giving information on the diagnosis, including likely course and prognosis, and many family members and carers and support workers will understandably find it painful and difficult to be given this information.

In some cases families and carers will have had no prior knowledge of the increased risk of people with intellectual disabilities developing dementia and they will require both emotional support, psychoeducation and, where relevant, training. Families can find it devastating to learn that their loved one is developing an impairment additional to their intellectual disability. However, it is very important to help family and carers understand how dementia is affecting the person with a pre-existing intellectual disabilities and to maintain an awareness of both diagnoses so that they can understand and cope with the changing situation.

Families may need a lot of support to accept that the person with intellectual disabilities needs to be told their diagnosis and that not knowing is likely to be linked to increased confusion and distress. Once again a helpful approach needs to be informed by an awareness of how family and carer factors, such as culture, religion and personal history,



might influence their attitude and understanding of the needs of the person with intellectual disabilities and dementia. On occasion family members may need focused psychological interventions concerning their acceptance of the diagnosis.

A partnership approach needs to be adopted with the person, their family and friends, and their carers. A palliative approach also needs to be adopted from the point of diagnosis and effort should always be made to keep families and carers informed and involved at all stages (see Section 15 – Palliative Care and End of Life). Family and carers are understandably anxious about future care and they need help in planning and thinking ahead including concerning specific supports and accommodation. Their understanding of the support and management plans and the rationale behind them can be increased by their involvement in structured meetings such as those using the Quality Outcome Measure for Individuals with Dementia (see Section 18 – Outcomes).

Families and carers will need support and information to prepare for end of life care at the later stage of illness and they need to be sensitively informed about this early on to minimise their sense of isolation and anxiety about the future.

There are information booklets to assist carers to understand dementia, for example, *Keep Talking about Dementia* (Watchman, 2006) and *Down's Syndrome and Alzheimer's Disease: A guide for Parents and Carers* (Holland, 2013). Useful information can also be found on the Alzheimer's Society website.

## **7.5 Sharing the information with friends and peers**

Those involved with the person with intellectual disabilities who is affected by dementia also need to inform the friends and peers of the person, who may themselves have intellectual disabilities, about the diagnosis and help them understand what this means. Friends and peers benefit greatly, both personally and in their capacity to support the person with dementia, from the guided use of specific resources and psychological work or group work aimed at helping them understand what is happening to the person.

There are resources to help friends and peers with intellectual disabilities, e.g. *About My Friend* (Dodd et al., 2005b), *Ann has Dementia* (Hollins et al., 2012) and *Easy Read Factsheets* (BILD, 2014).

## **7.6 Multidisciplinary discussion and care coordination**

In the first post-diagnostic multidisciplinary meeting, which wherever possible should include social work practitioners, it is important to agree on the process of care planning at this early stage and identify an appropriate professional to be the care coordinator. This should form part of the Care Pathway and lead into the development of a new Health Action Plan (HAP) and a reviewed person-centred support plan (see Section 10 – Philosophy of Care). The HAP and support plan will need to be reviewed regularly in the context of the person's changing condition and anticipated needs. The Quality Outcome Measure for Individuals with Dementia is a useful tool to use in this process and can involve family, paid carers and professionals in partnership.

## 7.7 Management

Irrespective of the degree of certainty attached to the diagnosis, interventions focused on the needs of the individual and management of any risks should be thoroughly discussed in the first care planning meeting. All interventions and management plans need to have the person as the centre of focus and this will therefore require a holistic approach that takes into account the person's expressed views and a wide range of personal factors, including profile of abilities and communication needs, cultural background, life experiences, significant relationships with family, carers and peers, and preferred activities, as well as working to minimise known risks.

Decisions about specific interventions and treatments that are likely to be effective in supporting the person to live well, maintain independence and minimise disabilities, need to be discussed and reviewed in regular multidisciplinary meetings. The frequency of such meetings will vary depending on the current needs of the person, and are likely to range from monthly to six monthly.

Please refer to other relevant sections of guidance for details of evidenced-based interventions and treatment currently available.

As the person's physical care needs intensify and they approach the later stage of dementia, the team of professionals around the person needs to consider NHS Continuing Care Funding. In addition there will be an increasing need for palliative care and end of life planning (see Section 15 – Palliative Care and End of Life).

### Key points

- People with intellectual disabilities, their families and carers need to be given opportunities to understand the nature of the intellectual disability and information about any associated health risks from an early point in their life and particularly from transition to adulthood onwards.
- People with intellectual disabilities need to be told about their diagnosis of dementia and given ongoing opportunities to understand their diagnosis and their experience of dementia.
- Family members and carers need to be informed about the diagnosis and involved as much as possible in support and management plans and, as appropriate, be given opportunities for education and training.
- The person's peers and friends are also important people to involve in giving information about the diagnosis and this will both help them cope and help them support the person affected by dementia.
- People with intellectual disabilities and their families and carers may need psychological interventions to enable them to feel emotionally supported and to begin to understand the diagnosis.

## Section 8 – Additional health co-morbidities associated with dementia

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As we get older we become more susceptible to additional health challenges, and this is no different for people diagnosed with dementia. However, there are some health conditions that are specifically associated with advancing dementia and these should be taken into consideration.

### 8.1 Epilepsy

#### 8.1.1 Prevalence

Over 80 per cent of people with Down's syndrome and dementia develop seizures (Menendez, 2005). There are two peaks for the development of epilepsy in people with Down's syndrome, one in adolescence and one in later life. Older people with Down's syndrome (over 45 years) are more likely to have seizures than younger people, and the development of epilepsy in later life should raise the possibility of Alzheimer's disease. A younger age of onset of dementia is associated with a higher risk of developing seizures (Menendez, 2005).

#### 8.1.2 The nature of seizures

The most common seizures in people with Down's syndrome and dementia are myoclonic and tonic-clonic types although the whole range of generalized and partial seizures may be seen.

- In the general population, people with Alzheimer's disease generally develop seizures at a late stage of dementia but in people with Down's syndrome seizures can be a presenting symptom. Seizures are generally thought to occur earlier in the course of the illness in people with intellectual disabilities than is found in the general population.
- A proportion of people with Down's syndrome who develop dementia will have a previous history of epilepsy, and there may be a change in seizure frequency, pattern or severity with the onset of dementia.
- Myoclonic seizures tend to occur more frequently and may initially present as mild jerks, although the intensity and frequency can vary considerably. As the dementia progresses, they can become both more severe and more frequent. Tonic clonic seizures are more obvious and similar precautions, as in people without dementia, should be taken.

#### 8.1.3 Investigations and diagnosis

The diagnosis of epilepsy is clinical, and is usually made following two witnessed, unprovoked seizures. Where people with a progressive condition such as dementia, which is associated with the development of epilepsy, have a seizure, many clinicians will initiate treatment after the first seizure rather than waiting to confirm a second seizure (particularly where the person has had seizures before or where the person has had a tonic clonic seizure).

Investigation guidance for the diagnosis of epilepsy suggests a range of investigations including EEG, CT/MRI scans, ECG and blood tests. Whilst a full physical examination and blood tests should always be considered, neuroradiological investigations and EEG's should be undertaken if there is a suspicion of a space occupying lesion or some other potentially treatable cause of dementia.

#### 8.1.4 Risks

Seizures are associated with additional physical health problems ranging in severity from physical injury to death. The risks increase with poorly controlled seizures and polypharmacy. It is important to establish a monitoring system so that staff and carers can take responsive action when needed. Staff and carers should be encouraged to maintain regular seizure charts to record the nature, frequency, intensity and duration of seizures, and to complete appropriate risk assessment. Issues relating to both the seizures and associated treatments should be addressed, including eating and drinking guidelines, management of falls and personal care.

#### 8.1.5 Management

- a) **Drug management** – seizures in people with dementia generally respond to a single antiepileptic drug. Drugs with a broad spectrum of action are normally used as the first line treatment. It is important to note, when choosing a drug, the potential for further impairment of cognitive function because of the sedative effect of the antiepileptic drug. For this reason, newer, less sedating drugs are usually chosen. Where the presenting seizures are myoclonic, Levetiracetam or Sodium Valproate are first line choices (remember that myoclonic seizures can be worsened by administering Carbamazepine, Gabapentin, Pregabalin and perhaps Lamotrigine). Where the presenting seizures are tonic-clonic in nature, Lamotrigine, Levetiracetam and Sodium Valproate are first line choices. The following good practice principles should be borne in mind:
  - i. Promote the use of a single medication whenever possible.
  - ii. The treatment goal should be a healthy balance between quality of life and seizure control.
  - iii. Unacceptable side effects should not be present and the prescriber should take immediate action if side effects are reported. Careful monitoring of the side effects by staff and carers is the key and it is important for clinicians to explain what to look for in relation to side effects.
  - iv. The care plan should cover how to minimize seizure related risks especially in relation to falls and injuries.
  - v. Avoid Phenytoin as this causes significant cognitive slowing and can debilitate an individual with limited cognitive reserve.
- b) **Use of rescue medication** – Midazolam is currently the treatment of choice for prolonged or clusters of seizures. This can be administered via buccal or intranasal routes. The standard doses of both are 10mg for adults weighing more than 50kg. Training for staff and carers in the use of rescue medication is essential. The use of rescue medication should be documented in the person's care plan.

- c) **Side effects** – The main side effects of Sodium Valproate are weight gain, gastro-intestinal problems, and a negative impact on cognitive function. The main side effects of Levetiracetam are cognitive slowing, (particularly if the dose is escalated rapidly) and gastro-intestinal. Some clinicians report behavioural change, this is almost certainly related to side effects and rapid dose escalation. With Lamotrigine, the concerns are for skin rash and gastro-intestinal side effects. With diminishing cognitive reserve, drugs that effect cognitive function have a much more dramatic effect meaning that, for example, sedation can be a significant problem.
- d) Do's and don'ts
- Do not use rescue medication unless there is a clear indication and this is documented in the care plan.
  - Do train carers and provide appropriate information.
  - Remember that quality of life has a higher priority than total seizure freedom.
  - Remember that the presence of active epilepsy should not limit community participation.
  - Do monitor seizures and the side effects of anti-epileptic drugs.
  - Remember that both seizures and drug side effects can affect the person's ability to eat, drink and function effectively.

### Key points

- The occurrence of seizures for the first time is very common in people with Down's syndrome who have developed dementia.
- The use of a single antiepileptic medication should be encouraged.
- Careful monitoring is required of seizure patterns and side effects of medication.

## 8.2 Pain

Poor recognition and treatment of pain in people with intellectual disabilities and dementia is common. Many issues contribute to the low level of pain recognition. These include:

- Staff attitudes towards, and experience of, 'behaviour that challenges'.
- Diagnostic over-shadowing.
- Problems with communication.
- Beliefs about pain thresholds.
- The impact of past treatment on willingness to complain of pain.
- The use of temporary agency/bank staff who are less likely to observe a change in a person's level of distress.

Sometimes staff are not sufficiently aware that people with intellectual disabilities who are getting older will experience painful conditions, such as arthritis, that can be associated with older age. Difficulties are sometimes ascribed to the dementia process rather than there being a consideration of whether people are in pain.

Research and practice both indicate that there is inadequate training about dementia and people with intellectual disabilities of staff, at all levels and from all professional backgrounds. In addition, little attention is paid to the recognition and management of pain in this group.

There is also little use of pain assessment and recognition tools. There is a range of effective pain/distress tools available for staff and carers to use to identify pain or distress in people with intellectual disabilities and dementia. Tools to consider include the Abbey Pain Scale (Abbey et al., 2004) and the Disability Distress Assessment Tool (DisDAT) (Regnard et al., 2007).

### Key points

- Pain recognition and management for people with intellectual disabilities and dementia is often very poor.
- Diagnostic overshadowing is a frequent occurrence, and staff are often unaware of the range of painful health conditions that may present with increasing age.
- There are tools available to help staff and carers identify pain in people with intellectual disabilities.

## 8.3 Sleep disorders

People with intellectual disabilities have a high prevalence of sleep difficulties which worsen with age and with associated conditions such as dementia. The sleep disturbances in dementia typically include a reversal of the sleep-wake cycle (sleeping during day time and wandering at night time) and a reduction in the slow wave sleep; which may be due to the loss of cholinergic nerve cells.

Clinicians should exclude the following treatable conditions or situations before considering biological attributes (loss of cholinergic neurons):

- Co-morbid psychological problems including depression, anxiety, fear and nightmares.
- Alcohol/substance misuse.
- Physical health problems, e.g. pain, epilepsy, sleep apnoea, heart-failure, respiratory disorders, nocturnal enuresis.
- Side-effects of medication, e.g. stimulant drugs.
- Poor sleep hygiene – uncomfortable bed, noisy household, poor light and temperature adjustment, late evening coffee and recent change in the environment.

People with intellectual disabilities and dementia should have a routine assessment of sleep hygiene. Associated factors can be easily overlooked, especially in people with intellectual disabilities who may have poor communication skills. The assessment and the sleep history from the carer should include the following:

- The person's sleep pattern.
- Information on sleep hygiene, bedroom and bedtime routine.
- Onset, duration and nature of the problem (e.g. difficulty in going to sleep, frequent awakening, early morning insomnia, motor activities, snoring during sleep and evidence of daytime sleepiness).
- Effect of insomnia on the person and others including family and/or carers.
- Past sleep difficulties and previous treatments.
- Psychiatric and medical diagnoses (including epilepsy).
- Current medications.
- Family history of sleep problems.
- Risks associated with sleep difficulties, e.g. wandering behaviours or falls.

Accurate information is required to make an informed decision about the nature of an individual's sleep pattern. This can be difficult if there are no informant reports available. The use of assistive technology or waking night staff has helped greatly in trying to provide some of the answers, but is not a panacea. Some effort may be required to get adequate sleep data; however, accurate information is required to inform the appropriate management strategies. Consideration should also be given to the use of, for example, the Epworth Sleepiness scale (Johns, 1991) or related scale used by local sleep apnoea clinics, in the event of daytime sleepiness in order to establish whether onward referral to a sleep clinic is warranted.

### 8.3.1 Management

The approach to managing sleep disorders is, first and foremost, to ensure the problem is appropriately understood and that there are adequate checks and balances in place to ensure that interventions can be monitored, in order to understand the outcome of each intervention. The least restrictive options should be used first.

- Treat any associated physical and/or psychological problems.
- Use non pharmacological strategies (sleep hygiene).
  - Encourage daily activities and if possible exercise.
  - Avoid day time napping.
  - Reduce caffeine and alcohol intake before bed time.
  - Eliminate factors that impede sleep (watching TV into the early hours).
  - Use the bed just for sleeping.
  - Set and maintain a regular routine of rising and retiring at the same time everyday.
  - Ensure sleeping environment is conducive to sleep.

If the above approaches do not produce any significant benefits and the risks continue, a pharmacological approach may be considered along with non-pharmacological approaches.

The pharmacological approaches include the use of:

- Melatonin – recently licensed under the name of Circadin in patients over the age of 55 (there is an age related decrease in endogenous melatonin secretion which contributes towards age related insomnia).
  - Z-drugs (Zopiclone, Zolpidem and Zaleplon).
- Benzodiazepines (Temazepam, Loprazolam or Lormetazepam).

The choice of medication should depend on the individual patient's needs and should be based on NICE Guidance CG 76 (Clinical Update 2009):

- Melatonin can be used for long-term insomnia in those over 55 (with or without dementia), licensed preparation is long-acting Circadin. Monitor results, if improvement after three weeks, treatment can be extended to 10 weeks in the first instance.
- Use short acting benzodiazepines (Temazepam, Loprazolam or Lormetazepam), but be careful about the long-term effects of such drugs.
- Consider use of z-drugs (Zopiclone, Zolpidem and Zaleplon) as an alternative.
- There are no differences in the efficacy of z-drugs and if one of them is not effective the others should not be used.

- Switch from one z-drug to another only if there is an adverse effect directly related to that particular drug.
- Use the minimum effective dose.
- Use medication on an ‘as and when necessary’ basis (every second or third night if required) rather than on a regular basis.
- Do not continue hypnotics for longer than four weeks.
- Discuss the discontinuation with the person and carer, and taper/stop it very gradually.
- Warn the person and carer of withdrawal symptoms and rebound insomnia.
- Advise on adverse interactions with alcohol and other sedative agents.
- Do not use in hepatic failure, chronic respiratory diseases and people who have a history of substance misuse.
- Be mindful of the side effects including day time sedation, falls and sun downing (confusional state in the evening).

### Key points

- Sleep difficulties are commonly experienced in people with intellectual disabilities as they get older and/or develop dementia.
- Assessment should include ruling out co-morbid mental health problems, substance misuse, physical health problems and poor sleep hygiene.
- The management of sleep difficulties in dementia should be based on non-pharmacological approaches including good practice of sleep hygiene.
- Medication should only be used if other approaches have failed or risks are significant.
- NICE recommendations should be adhered to if pharmacological approaches are used.

## 8.4 Gastrointestinal disorders

Gastrointestinal disorders are common in people with intellectual disabilities. Gastro-oesophageal reflux disease (GORD), constipation, dysphagia and nutritional issues are all found in people with advancing dementia and should be monitored. Disorders of the gastrointestinal tract can cause changes in behaviour due to pain or food refusal and so should be included in any analysis.

A swallowing assessment as part of the baseline investigation of dementia would be best practise. This is due to the fact that dysphagia (swallowing difficulties) and apraxia (purposeful movement difficulties) are both associated with feeding difficulties in people with dementia, but might not be the presenting feature.

Swallowing difficulty (with resulting aspiration) is a major risk for the development of aspiration pneumonia. Often, secretions are aspirated during sleep; however, a normal cough reflex and normal immune mechanisms protect the airways from repeated infections. As dementia progresses, however, this simple aspiration will extend to aspiration of food and drinks during eating making the chance of developing pneumonia much more likely.

Choking during eating usually starts with liquids because the swallowing of liquids requires the best coordination of the muscles involved. Choking can sometimes be prevented by



switching from thin liquids to thick liquids, e.g. from milk to yogurt. This should be part of the assessment completed by a qualified speech and language therapist and or dietitian (see Section 14 – Eating and Drinking).

Apraxia results in inability to use utensils but people with dementia may be still able to feed themselves finger food. With the progression of dementia, individuals may ultimately be unable to feed themselves or drink without assistance. However, adequate nutrition can be provided by hand feeding using a modified diet that is adapted to the ability of the individual paying attention to their individual chew and to their swallowing difficulties. Hand feeding can often be provided until the time when all physiological processes start to shut down.

At the point where food refusal starts to become an issue and where there are concerns about malnutrition, considerations turn to the use of nasogastric tube feeding or the insertion of percutaneous endoscopic gastrostomy (PEG) tube. The Cochrane review (Sampson et al., 2009) concluded that tube feeding does not prevent or improve pressure ulcers, does not reduce the risk of infections, does not improve functional status or comfort of the patient, does not decrease aspiration pneumonia (and could increase it) and does not prevent the consequences of malnutrition. Tube feeding should therefore be carefully considered and appropriately indicated. The most appropriate use of this treatment is where it is likely to be short term to allow an individual to recover from an acute illness.

#### Key points

- All people who have been diagnosed with dementia warrant a dysphagia assessment and should have eating and drinking guidelines in place.
- Eating and drinking guidelines should include the opportunity for ‘finger foods’ in order to allow the individual to maintain their ability to feed themselves.
- Nasogastric and PEG feeding are not associated with significantly improved outcomes and should be carefully considered.

## 8.5 Infections

People with dementia commonly succumb to a number of infections including, most commonly, urinary tract infection (UTI), respiratory tract infection, skin infection, gastrointestinal infection and eye infection.

These infections are almost an inevitable consequence of advanced dementia; the reason includes reduced immune responses. The risk of development of urinary tract infections is increased by incontinence (especially in women) and by urinary retention (in men). Swallowing difficulties with aspiration increase the risk of developing respiratory infections and reduced physical activity increases the risk of urinary and respiratory infections, deep vein thrombosis and infected pressure ulcers.

Communication difficulties in people with intellectual disabilities and dementia further complicate the diagnosis of illness as they are even less likely to report cough, rash, gastrointestinal symptoms, and joint pain than people with intellectual disabilities alone. Functional impairment is also an important factor because dependence in feeding and oral care is a significant factor in the development of aspiration pneumonia.

Ultrasound can be useful in the detection and future management of residual urine. This is usually the remit of the continence teams who are well equipped to manage these situations. The avoidance of internal urinary catheters, where possible, is an important prevention strategy for UTIs because the bladder is colonised with bacteria soon after the insertion of an indwelling catheter. Prophylactic antibiotics can seem like a tempting solution; however, this can lead to the development of antibiotic-resistant bacteria.

Strategies to prevent aspiration pneumonia include oral hygiene, avoidance of smoking and endotracheal intubation, and potentiation of the cough reflex. Bad dental hygiene is a risk factor for development of pneumonia. Oral care has been shown to decrease the incidence of pneumonia (and death) and the number of febrile days.

### Key points

- People with dementia have a high rate of infection and so any change in presentation should include an assessment for an infection.
- Infections and the causes for these need to be addressed and, where possible, changes should be made to prevent recurrence.
- Where possible, steps should be taken to prevent infections rather than to treat each as they arise.

## Section 9 – Conceptual understanding of the dementia process

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Conceptual understanding of the psychological and social consequences of dementia is essential for senior staff to guide timely decision-making about interventions and approaches to individuals/service developments. The social model (NICE/SCIE, 2006) proposes that whilst people with dementia have an impairment, they may be further disabled by the way they are treated by or excluded from society. The advantages of this framework are that carers and staff will understand:

- that dementia is not the fault of the individual;
- that the focus is on the remaining skills rather than losses;
- that the individual can be fully understood (their history, likes/dislikes, etc.);
- the influence of an enabling or supportive environment;
- the key value of appropriate communication; and
- opportunities for stress-free and failure-free activities.

This means that the responsibility to continue to reach out to people with dementia lies with people who do not have dementia. Carers and staff need to change their approach to 'go with' the person and their continuing changes. Brawley (1997) concluded that '90 per cent of the catastrophic behaviours in dementia are induced by carers or the environment'.

### 9.1 Models of dementia

Downs et al. (2006) show how different models, e.g. as a neurological condition; as a neuro-psychiatric condition; as a normal part of ageing; and seeing dementia from a person-centred perspective; can be used to understand dementia.

One approach to understanding dementia and its psychological consequences has been put forward by Buijssen (2005). He proposes two laws of dementia, and asserts that by understanding them, and their consequences, we have a framework to understand and respond to people appropriately.

#### Law 1: Law of disturbed encoding

In this law, the person is no longer able to transfer information successfully from their short-term memory and store it in their long-term memory. This means that the person is unlikely to remember things that have just happened to them.

The main consequence of disturbed encoding is that the person is unable to form any new memories for the things they experience or for things they are told. This means that:

- They experience disorientation in an unfamiliar environment.
- They experience disorientation in time.
- The person asks the same questions repeatedly.
- The person quickly loses track of conversations.
- The person is less able to learn anything new.

- The person easily loses things.
- The person is unable to recall people whom they have recently met.
- Appointments are quickly forgotten.
- The person experiences anxiety and stress.

### Law 2: Law of roll-back memory

Long-term memory contains all the memories that have been acquired starting with the most recent memories, and working back toward childhood memories. When dementia develops, the person will be less able to form any new memories after this time. At first their long-term memories will remain intact, but as dementia progresses, long-term memories will also begin to deteriorate and eventually disappear altogether. Deterioration of memory will begin with the most recent memories and will progress until only memories of early childhood remain, hence memory can be said to be ‘rolling back’.

The consequences of roll-back memory are:

- Loss of daily skills such as using kitchen appliances.
- Memory loss for events, beginning with the most recent, e.g. last holiday.
- Decreased social skills and increased inappropriate behaviour.
- Decreased vocabulary and inability to find words.
- Disorientation towards people, e.g. inability to recognise family and relatives.
- The person may begin to have ‘flashbacks’ and see people from their past.
- Self-care skills will begin to deteriorate.
- Changes in personality.
- The person believes that they are younger and that time has actually ‘rolled back’.

## 9.2 Understanding the process of change

It is important that staff and family carers understand that dementia is a progressive disease, and that the skills, abilities and needs of the person will be constantly changing.

Alzheimer’s disease in people with Down’s syndrome can be thought about in three stages: early stage, middle stage and late stage. The person’s cognitive ability will deteriorate across these three stages and their level of dependence will increase.

It is important to remember that the person may appear to move into the next stage, often because of an illness such as pneumonia or a urinary tract infection (UTI), and may return to the previous stage once the illness has been treated, although the recovery can take time. Therefore when there is a sudden decline, a physical health cause should always be considered and appropriate investigations undertaken.

### Early Stage

At this stage, the person is showing signs of decline from their usual level of functioning in the following areas:

- Subtle changes in behaviour and mood.
- Performance at day placements deteriorate.
- Memory problems, particularly for recent events.
- Ability to learn new information is affected.
- Language and word finding problems.

- Decline in social, community and daily living skills.
- Disorientation.
- Difficulties with steps, stairs and kerbs due to depth perception problems.

### Middle Stage

- Memory loss become more pronounced and the individual may forget personal information or the names of familiar people.
- Language problems become more evident.
- Confusion and disorientation around time, place and may have problems finding their way around familiar environments.
- Difficulties with and then loss of self care skills.
- More severe changes in personality and social behaviour, e.g. mood changes, inactivity or apathy, behavioural disturbances such as wandering, sleep problems, agitation, hallucinations and delusions.
- Problems with eating and drinking.
- Disturbed sleep patterns.
- Increasing incontinence.

### Late Stage

- Myoclonus and epilepsy.
- Incontinence (bladder and bowels).
- Loss of eating/drinking skills.
- Problems with walking and balance, individuals become chair- or bed-bound.
- Problems with recognising people.
- Often require 24-hour care.
- Will become bedridden and inactive.
- Greater risk of infections, particularly pneumonia.

Deterioration of people with intellectual disabilities and dementia is not predictable. People will deteriorate at different rates and patterns. Some deteriorate steadily, others less predictably, and this can change at any time. Furthermore, people will often not be clearly in one stage and will show signs of more than one stage.

Frequently people offering care and support on a daily basis to a person with dementia adapt to the changes in the person as they occur. It is often when the person is assisted to have a holiday and is out of their familiar environment that staff and carers realise the extent of the change. Similarly, if staff are new to caring for the person, they might not know the person's history and not understand the abilities of the person before they were diagnosed with dementia.

### Case Study

John has Down's syndrome and Alzheimer's disease. He resides in a supported living home with a staff team that have known him for many years and have learnt to support him effectively. The staff team has been supported through 'whole staff team' training and regular reviews of John's care needs.

The psychologist was contacted as the psychiatrist raised concerns that the staff team was beginning to struggle with understanding John's needs, and that he was losing weight. The psychologist offered a consultation session to the manager and senior support workers.

As a result of the consultation, it became apparent that staff who regularly supported John had not recognised that John's dementia had progressed. This meant that the style of support and prompts that they used were no longer helpful for John and not appropriate for his current needs. Once this was clarified, the team was able to reflect on John's changing needs and put in place a more appropriate care plan to support him effectively, especially with regard to his eating and drinking. This led to John gaining weight.

### Key points

- The social model gives a conceptual model for staff and carers to understand dementia.
- Understanding what happens as dementia progresses and its consequences gives a framework to understand and respond appropriately.
- Think about what the person is actually experiencing, and use that to inform the care and support that is required.

## Section 10 – Philosophy of care

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### 10.1 Individualised philosophy of care

There are a number of models of person-centred dementia care in existence within the general dementia literature, including Person-Centred Care (Kitwood, 1997); VIPs (Brooker, 2006); and Feelings Matter Most (Sheard, 2008). Sheard (2013) has recently introduced the concept of ‘mattering’ which he says ‘brings together the core elements of; emotional care, skills, quality of life and environment with the culture of a shared relaxed community, thereby evidencing that individuals matter’.

*The State of Care* report from the Care Quality Commission (2014) showed that those services that maintain people’s dignity and treat them with respect all have a number of things in common: they recognise the individuality of each person in their care, help the person to retain their sense of identity and self-worth; take time to listen to what people say; are alert to people’s emotional needs as much as their physical needs and give the person control over their care and the environment around them.

Excellence in dementia care requires staff and family carers to understand and know the person, understand dementia and its consequences for the person, and consequently to be able to think ahead and predict ‘stressors’. They need to adapt their approach as much as possible to ensure that the person with dementia has a stress-free, failure-free but individualised care that is consistent but without time pressures. This needs to be incorporated into the person’s person-centred plan and care plan.

### 10.2 Life Story work

This can only be achieved by having a thorough knowledge and understanding of the person and their history. This can be aided by the use of Life Story work (Gibson, 1994). The individual themselves should be encouraged to participate in the making of the life story as much as possible.

- Where possible the individual should have the final say as to what is included in the book/box. People should only put things in their life story book that they are comfortable with – even if it means leaving out huge chunks of their life.
- A Life Story does not have to stick to one particular format – it should be whatever the person wants it to be. It can include sensory items that people can feel or smell which have a significance to the person.
- Life story books/boxes should not finish. Staff, family and the individual can continually add to them.
- Life Story work can be a great way to increase an individual’s self-esteem because the life story is all about them and what they like, etc. (without having to mention a diagnosis or any other labels they may have.)

- If including photographs of people it is important they are put into some sort of context, for example, what their name is, how they are known to the person, date taken, event, why it is important to the person. That way, anyone can pick the book up and be able to use it in a meaningful way with the individual. This is absolutely vital so that if someone loses the ability to remember, the staff still have the cues.
- If the person does not have many pictures of their life/growing up, an alternative would be to use pictures from magazines, books and the internet of anything that the individual remembers or just likes.

Staff and family carers need to be clear about what they are trying to achieve. This is not the time for learning new skills, achieving goals or facing change. They need to consider the person's happiness, comfort and security. The focus of care should change from goal orientated to emphasis on enabling quality of life. Staff and carers need to remember that people compensate for their deterioration in functioning by making greater use of remaining abilities (e.g. earlier memories). This may mean that the person finds comfort in activities and objects from their childhood. Roll-back memory may also mean that the person is not oriented to the present day. Care needs to be taken not to challenge the person's beliefs repeatedly as this will add to their stress. This approach fits with the values and model of Positive Behaviour Support (Gore et al., 2013) that is central to services for people with intellectual disabilities.

Care includes not only the social aspects of the person's life, but an increasing awareness of the physical consequences of getting older and additionally having dementia. Care needs to be taken to ensure that diagnostic overshadowing does not occur, where all changes are attributed to the dementia. Research (e.g. Kerr et al., 2006) indicates that pain recognition and management is extremely inadequate in this group of people. All people should have comprehensive health checks and a Health Action Plan, updated annually at minimum. Further information on health co-morbidities associated with dementia can be found in Section 8.



## Dementia Consultation

Mary, a lady with Down's syndrome, was diagnosed with mid stage dementia. The home manager was worried about how the staff team would manage the diagnosis and also change their working practices. She was concerned that they would feel de-skilled.

A systemic staff consultation was held, exploring the meaning and beliefs that support staff had about dementia, and how support needs changed with the diagnosis. They highlighted the change in emphasis from valuing independence and choice to maintaining skills and reducing anxiety. Different perspectives of 'good support' were considered, for example: the organisation, intellectual disability team, client, client's family, home manager, and support team. They discussed the how organisational values were different to what they now felt might be best for Mary.

Personal stories of dementia, death, what might make a 'good death,' were explored and the confusion of being a professional in someone's life and expectations around this, yet knowing someone and being sad about the changes in the person they supported. We talked about space that can be made for all of this. The home manager noticed changes in the way staff communicated with each other about their emotions as well as the practical issues. She noticed that staff seemed more confident and creative in changing their practice to suit Mary. For example, the staff team asked for training in intensive interaction so they would be better able to communicate with Mary as she could no longer speak. Mary was able to stay at home until she died.

## Key points

- Staff and carers need to ensure that people with intellectual disabilities and dementia have stress-free, failure free and consistent care.
- This approach leads to a more supportive environment and lower levels of behaviour and distress.
- Services need to review regularly the support given to people with intellectual disabilities and dementia, particularly the amount of staff support provided both during the day and through the night.

## Section 11 – Environments

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This section of the guidance focuses on how carers and services can create capable environments that enhance the quality of life for people with intellectual disabilities and dementia.

### 11.1 Where should people with intellectual disabilities and dementia live?

Janicki and Dalton (1998) proposed a potential pathway that someone might follow after a diagnosis of dementia. This we have adapted to reflect the current position of the Care Services Improvement Partnership and the Department of Health, with the options given in order of preference for people with intellectual disabilities and dementia.

#### Preferred option:

**‘Dying in place’** where the person can stay where they are currently living with appropriate supports adapted and provided. This means that the person stays with what is familiar in their long-term memory.

#### Compromise option:

**‘Moving to more specialist intellectual disability provision’** where the person has had to move from their current home, but moves into provision supported by intellectual disabilities services.

#### Least preferred option:

**‘Referral out of intellectual disability services’** where the person will be moved to services for older people, either residential or nursing.

By staying where they are, the person will stay with familiar people (family, peers, familiar carers) and in an environment that they know. Although their needs will change as the dementia progresses, every effort should be made to maintain their home life. This may necessitate environmental changes and adaptations to support the person, increases in staffing levels and careful thinking about the supports required.

In some instances it is not possible to maintain the person in their existing home:

- It may be that the design of the building is inappropriate and cannot be changed.
- The person may be being looked after by older family carers who may not be able to continue caring as the needs of the person increase, or their own health changes.
- Risk assessments should be completed regularly to ensure that the environment remains safe for both the person and their carers.
- Staff may reach a ‘tipping point’, where there are insurmountable problems with the placement (often the quality of life for the other residents or staff issues).
- Funding may be an issue.

If the person has to move then the most appropriate provision may be a specialised intellectual disability service which is able to meet the person’s increasing health needs and provide palliative care (Thompson & Wright, 2001). Although some moves may be of

benefit to the person, the majority of the evidence suggests that moving increases the rate of decline and can precipitate deterioration in health and behaviour and lead to an earlier than expected death (Wilkinson et al., 2004). Multiple moves must always be avoided. The use of an advocate or an Independent Mental Capacity Advocate (if the person has no family) may assist in decision-making, particularly where the person is lacking capacity (Mental Capacity Act, 2005). Relationships with significant others e.g. families, staff, friends and advocates must be considered and maintained.

If the person has to move, it is vital that other opportunities for continuity, for example, day activities, leisure and social opportunities which are in the person's long-term memory, are maintained. Maintaining social networks will also involve working with the person's peers to help them understand dementia and the changes in the person. This is discussed further in Section 17. There is a range of materials to assist with this (Dodd et al., 2005a, b & c).

## 11.2 What are the features of a capable environment?

There is a wealth of literature within general dementia care, and a smaller amount within the intellectual disabilities arena about the importance of the environment in enabling the person with dementia (Watchman et al., 2010; Kerr, 1997, 2007; Dodd et al., 2009; Dodd et al., 2006). Most environments where people with intellectual disabilities live are not 'dementia-enabled'.

According to Marshall (1998) the environmental design should not impede an individual but should have positive and beneficial effects for both the person and the staff. The environment should:

- compensate for the disability;
- maximise independence;
- enhance self-esteem and confidence;
- demonstrate care for staff;
- be orientating and understandable;
- reinforce personal identity;
- welcome relatives and the local community; and
- allow control of stimuli.

Adaptations will need to be made to ensure that the environment does not add more stress. Many of the adaptations required are not too expensive but can have a very positive effect on the quality of life for the person with dementia.

Environments need to be:

- Calm.
- Predictable and make sense.
- Familiar.
- Suitably stimulating.
- Safe and risk assessed.

### Calm and stress free

Environments can easily become stressful for someone with dementia. To avoid this and keep the environment calm and allow the person to focus better, many aspects of the environment can be adjusted.

## Noise

Noise is one factor which needs to be controlled, although not eliminated. People with dementia may have impaired hearing and in addition because of their cognitive impairments, do not know which sounds to attend to or not to attend to. For example it would be difficult for them to know whether they need to attend to the radio playing in the background or the staff talking to them. This impairment can be very distressing and is why noise levels should be controlled where possible. Two types of noise need to be considered – noise from the outside and noise from within the building.

- Noise from outside the building may cause the person to be distracted, or prevents them hearing conversation. Double glazing and lined curtains can help to reduce the level of external noise.
- Within the building, noise levels from competing sources need to be reviewed. Noise can be from TVs, radios and music systems, conversation or people calling out, telephones ringing.
- Staff need to consider the level of noise appropriate for each activity, person, and time of day.
- Music can be very calming but must be used selectively, and not be a constant background noise. Selecting music that is in the person's long term-memory can be beneficial, giving the person something that they can relate to.
- Distractions from other people – either staff, other residents or visitors – can also cause problems, especially if they are at times when you want the person to concentrate on a task, activity or interaction. Thinking about possible distractions and taking preventative measures can help, as can simple environmental measures such as moving the position of the telephone.

## Predictable and making sense

It is important to have an environment that is both predictable for the person and makes sense to them, otherwise the result will be disorientation and confusion. To avoid this, it is important to first recognise two important visual changes within individuals with dementia. One is that their depth perception is lost and, secondly, that they see the red end of the colour spectrum better than the blue end, and as a result colours such as red, orange and yellow are more easily seen.

- With regards to the loss of depth perception, one of the obvious consequences of this is that stairs become impossible to use as the depth of each step is hard to judge. As a result they should be avoided.
- Another consequence of this difficulty is that a change in colour can be perceived as a change in level. For example the point at which one carpet changes colour to another in a different room may be perceived by a person with dementia as a step, and cause them to be hesitant in doorways, or unwilling to enter a room. Staff may not understand what is causing the person's distress and may wrongly ascribe it to the person being difficult, stubborn or challenging.
- Furthermore, dark areas on light flooring can look like holes so the person with dementia will be inclined to avoid them by walking around them. Lighting which is not too bright or too dark is recommended to prevent shadow effects on the flooring which can look like holes.

- In addition to this, bathrooms and kitchens typically can be problematic as they usually have shiny floors which can look like pools of water to someone with dementia. It is recommended to have matt flooring which is the same colour as the flooring in the room that leads to it.
- With regards to the change in colour perception, it is recommended that red, orange and yellow are used to make certain objects or rooms more obvious, for example, painting a toilet door red, having red toilet seats, and using contrasting colours for crockery and table mats.
- With regard to colours for decoration and furniture, the most important issue is to ensure colour contrasts – so that the walls are a different colour from the flooring, which is different again from the furniture. This enables the person with dementia to be more able to distinguish items in their environment and helps to avoid accidents and incidents.
- As people get older, they need their environment to have more light. It is important that the *tone* of colours makes the environment look lighter and brighter, rather than having colours that make the environments seem darker and ‘suck out the light’.

These issues can cause the person to get very distressed and can also increase the possibility of falls. Risk assessments for the environment need to be reviewed regularly to keep the person safe.

- Aside from the issues mentioned, memory impairments mean individuals cannot find their way around familiar settings. This can be frightening for them so simple changes such as adding good signage to doors (e.g. personalised acrylic boxes with personal/significant items of the person within them) at the right level for older people can make a difference.
- Nolan et al. (2002) showed that placing a portrait such as a photograph and personal memorabilia in a display case outside the room of each person with dementia increased room finding by 45 per cent.
- Changes such as using rooms for a single clear function wherever possible; using the same room consistently for an activity (e.g. always using one particular bathroom for washing).
- Using picture rather than verbal cues is vital for people with dementia, e.g. picture timetables, pictures to indicate where things are, signage, picture menus, etc.
- Camouflaging doors that people do not need to use by painting them the same colour as the rest of the wall or using curtains to make the top half of a fire door look like a window; all can reduce confusion dramatically.
- Memory loss means mirrors can become scary to look as they do not recognise themselves. This is because they see themselves at a younger age, so mirrors may need to be covered up (e.g. at night).

### Familiar

Due to roll back memory (See Section 9 – Conceptual Understanding of the Dementia Process) people will not recognise things that were not around at the age they are ‘experiencing’ now. For example, someone who is 80 years old but whose reality is now the 1950s will have problems recognising and understanding how to use mixer taps, futuristic looking kettles, lamps, chairs, clocks, etc.

- It is important to use traditional style objects that will be recognisable to the person from their own past. For example, clocks with hands, traditional style kettles, cookers, lamps, chairs, clocks and curtains rather than blinds, etc. Basically, it is best to avoid modern futuristic styles.
- Aside from this issue, other adaptations that could be made to make the environment more familiar is to use small-scale, domestic, homely furnishings and to use objects/pictures for orientation (eg. picture of a toilet on the toilet door). Cohen and Weisman (1990) stated that familiar objects, activities, and spaces can trigger personal associations and even encourage social interactions and meaningful activity.

### Suitably stimulating

Although the environment needs to be calm, it is still important that the environment is sufficiently stimulating. There must be a happy balance between over stimulation and under stimulation. The study by Morgan and Stewart (1999) supported this notion and showed a curvilinear relationship between environmental demands and negative behaviour in people with dementia. Thus negative outcomes were associated with both under stimulation and over stimulation.

- To achieve this suitably stimulating environment, appropriate levels of noise for that person, activity and time of day are important.
- In addition clear views of the outside world and small quiet areas are important as they allow individuals suitable stimulation and allow peaceful and calming times.

### Safe

Safety is of paramount importance and there are a number of issues which need to be considered with people with dementia in a home.

- One behaviour which people with dementia may exhibit is wandering, and although this can lead to some safety problems, it should not be totally discouraged. As a result it is important all exits are monitored and/or alarmed and that fire exits are camouflaged so they are not subject to misuse.
- In addition it is important that steps, stairs and uneven flooring are all considered within the home due to the depth perception problems experienced in early and middle stages of dementia.
- It is important the garden is secure and safe for wandering, with safe plants and even surfaces. Ideally a home and garden should be designed with wandering in mind, thus a circular design is optimal for a home, allowing clients to wander around and return to where they started. This notion of a circular arrangement would also be applicable to the garden in terms of a pathway.
- In addition to this, the temperature of water in the kitchen, bathrooms and toilets need to be controlled and all possible hazardous objects, such as knives, sharp corners, hot kettles, need to be stored away safely.

Assistive technology, e.g. bed alarms, epilepsy monitors, are a useful adjunct to the environment for people with intellectual disabilities and dementia, but should not be used as a replacement for sufficient and competent support.

### Key points

- Wherever possible, people with intellectual disabilities and dementia should remain in familiar environments.
- If the person has to move, then this should be within an intellectual disabilities service.
- Environments can be adapted to make them dementia-friendly and enable the person with dementia to make sense of where they are living.
- It is important that environments provide stimulation for the person without putting them under any stress.

## Section 12 – Meeting changing needs/interventions

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### 12.1 Overview of approaches to intervention

It is important that the diagnostic process, which can be lengthy, does not preclude the implementation of interventions. Regardless of the eventual outcome of the diagnostic process, support is required to address current areas of need. Interventions need to be tailored to the individual, particularly the person's existing health, disability and social circumstances. There needs to be an emphasis on enhancing the psychological well-being of the individual, and minimizing the impact of changes being felt by the person's carers and/or peers.

Most of the interventions likely to be required are ones that are not specific to dementia, and likely to be beneficial regardless of the eventual outcome of the diagnostic process. The interventions will be familiar as core to the work of intellectual disability professionals with emphasis on a person centered approach with interdisciplinary and multiagency working. For example; the provision of accurate and timely information; formulation; interventions to maximise and maintain independence, skills and health; and at the same time promoting safety comfort, and dignity. Reducing excessive demands and simplifying routines are key, while minimising avoidable changes, especially any exclusion from appropriate services. It is notable that where someone is in a totally unsuitable environment for their needs (e.g. isolated or bullied) and may be depressed, then a change can be beneficial and aid the diagnostic process. Furthermore, awareness needs to be maintained regarding the person's changing needs and interventions reviewed at regular intervals.

The use of Person Centred Planning (DH, 2001) and the Care Programme Approach (DH, 2008) provide the same overall framework for individualised planning and care coordination as for anyone else with intellectual disabilities and changing or complex needs. All care planning, of course, has to be in the context of the individual and their family's religious/cultural context. Consent, capacity and risk management issues need to be considered at all stages, alongside the need for advocacy, advance directives, living wills and other mechanisms to facilitate best interest or complex decision making, according to changing legislative frameworks and ethical practice. Furthermore the Quality Outcome Measure for Individuals with Dementia (Dodd & Bush, 2013) provides guidance in providing quality person centered care for people with Intellectual disabilities and dementia (see Appendix 2).

### 12.2 Meeting changing needs

As the dementia progresses, the emphasis of care changes from enabling the person to maintain their skills with support to increasingly taking on tasks for the person with dignity and respect.

At the early stage of dementia, this will involve reminding the person of the day, time, place; simplifying routines and reducing choices; introducing memory aids such as diaries, timetables and objects of reference; simplifying communication, and using additional cues and prompts.



As the dementia progresses, care changes to trying to preserve abilities for as long as possible, using techniques of reminiscence, identifying favourite activities and strengths, and finding failure-free activities. At this stage, health monitoring becomes essential. Attention to weight, adequate nutrition and hydration, physical health including epilepsy continence, pain and mobility are all vital.

Towards the end of the person’s life, swallowing and dysphagia, skin and pressure sore care, moving and handling all become paramount. Even at the end stage of dementia it is important that the person has positive interactions throughout their day (Sharp, 2007). The tasks of daily living often become the activities of the day, and should be pleasurable and enjoyable for the person. Appropriate touch and verbal interaction are essential. There is further information regarding specific interventions for different stages of dementia within Watchman’s (2014) book.

**Table 2:** Suggested activities for each stage of dementia. (Source: Kalsy–Lillico et al., (2012) reproduced in Watchman (2014).

Early Stage	Early–Middle stage	Middle Stage	End Stage
Board games – card games such as snap are good, as are large sized dominoes	Music – play something that resonates, is liked by the person and has a good bass and beat	Movement and exercise – can be done standing or sitting	Smiling and laughing – don’t underestimate this as an activity
Ball games – throwing soft balls to each other, standing or sitting	Dance – chair dancing is good, swaying and rocking in time to music	Multi-sensory environments – use lights, sounds, smells, touch, Snoezlen	Singing – humming along to popular tunes, radio jingles or TV adverts
Discussion – about people, places and things	Art and ‘pottery’ – working with dough, clay, plasticine or sand	Massage – hand and feet spa treatments	Stroking – positive touch of people and objects that have different textures
Relaxation – progressive relaxation, massage or aromatherapy activities	Movement – guided walks, progressive relaxation	One-step cooking tasks, such as mixing items, peeling food	Gentle rocking – can relax and establish physical contact
Arts and crafts – painting, coloring in, making bean bags, poster	Drama	One-step gardening tasks, such as watering plants, digging pots	Holding – as above
End-product activities – anything where there is an immediate end results such as flower arranging, drawing, cooking, baking	Reminiscence – using familiar items, mementoes, touch, taste, smells, sounds, pictures or phots that reminds people of times gone by	One-step daily living tasks, such as plumping up cushions	Cuddling – as above
Use visual planners and cues to structures activities/day	Storytelling – talking about old friends, stores about special times, memories or what’s on TV	Walking: along routes that are circular, with focus points	
	Spiritual or religious activity	Stacking and folding – clothes, papers and magazines	
		Soft toys – touch can help anxious feelings	
		Baths, bubbles, balloons – remind people of fun	

## 12.3 Understanding behaviour in people with intellectual disabilities and dementia

By implementing the philosophy of care outlined earlier, for example, Buijssen (2005) psychosocial model of dementia, many of the difficulties that can occur in dementia can be prevented or minimised. However, there will often be times when people present with behaviours that staff or family find difficult to understand and respond to.

Positive behaviour support approaches (PBS) have become established as a preferred approach when working with people with intellectual disabilities who present behaviours that can be perceived as challenging. This is evidenced in *Positive and Proactive Care: reducing the need for restrictive interventions* (DH, 2014), British Psychological Society guidelines (BPS, 2004) and the joint guidelines of the Royal College of Psychiatrists, the BPS and the Royal College of Speech and Language Therapists (RCPsych et al., 2007). PBS emphasises person-centred values, aims to enhance community presence, increase personal skills and competence and ensure dignity and respect is maintained for the person being supported. When a diagnosis of dementia is suspected a shift in value from increasing skills to maintaining skills can be helpful in terms of reducing stress of the individual.

In all cases where there is behaviour that challenges, the standards of assessment of the behaviour and subsequent intervention should occur as outlined in *Challenging behaviour: a unified approach* (RCPsych et al., 2007) This includes ensuring that any intervention addresses the person, the environment and the interaction between the two. As the report says:

*'A comprehensive assessment should address: a functional assessment of behaviour, underlying medical and organic factors, psychological/psychiatric factors. Detailed functional assessment and diagnosis are both integral features of an assessment of challenging behaviour and should lead to a clear formulation of the presenting problem. Interventions should be delivered in a person-centred context and a framework of positive behavioural support. They should include proactive and reactive strategies. Interventions described include psychotherapy, communication, positive programming, physical and/or medical and psychopharmacological.'* (p10).

Additional aspects, related to the dementia, may need additional consideration in the assessment of the behaviour of concern:

- The problem behaviour may be transitory to the current stage of the person's dementia and not need an intervention. Similarly, it may be possible to withdraw interventions if/when the person's skills and behaviour change again.
- The situation should be viewed through the eyes of the person with dementia – i.e. their current reality. Continual correction by staff/carers of a false reality (e.g. person with dementia asking when a dead parent will visit) will not reduce their immediate confusion and distress. Emphasis should be on validation of feelings rather than the 'truth' of the situation.
- The behaviour should be viewed as an attempt by the person to communicate or to make sense of a bewildering environment (e.g. GP practice waiting room is confused with an airport lounge).

- The behaviour may be an exacerbation or return of previous behaviours. If the person had difficult behaviours/personality traits previously, these may return/reoccur with roll back memory.
- The behaviour may be caused by a return to a long-term memory that is now inappropriate, e.g. childhood urinating outdoors in the countryside whilst on long walks.
- There may be an underlying neurological change, e.g. taste bud changes leading to food fads and a liking for stronger flavours, refusing baths resulting from problems with depth perception and/or stepping into the bath.
- Simple and practical solutions may work e.g. a net to catch items thrown out of the window.
- Simple environmental alterations may alter the behaviour e.g. removing a mirror.

There is a good description of many of the most common problem behaviours seen in dementia in Dodd et al. (2009) *Down's syndrome and dementia resource pack*. Throughout the progress of the dementia, any changes should be clearly documented. There are tools available to facilitate this, e.g. *Down's syndrome and dementia workbook* (Dodd et al., 2006), and care mapping approaches (Brooker & Surr, 2005).

## 12.4 Psychological interventions

Within the literature emphasis has been placed on assessment and diagnosis. However, psychological interventions described in the general dementia literature can be adapted for use with people with intellectual disabilities. Interventions need to consider the multiple influences on behaviour (behavioural, systemic and biomedical) in order to support the individual and carers to understand, cope and manage behaviour and emotional distress (Kalsy-Lillico, 2014). This approach places responsibility on the system to change and to both communicate effectively with the individual whilst enhancing the person's current capabilities. Kalsy-Lillico et al. (2012) describe psychological interventions in relation to four orientations: behavior, emotion, cognitive and stimulation. Table 3 presents a framework for organising psychological interventions and practices into these four broad groups. We would also recommend the use of systemic approaches for working with families and staff teams. Intensive interaction also appears useful in enabling staff teams to communicate with a person at their current level of ability.

**Table 3:** A framework for psychological interventions. Source Kalsy-Lillico et al., 2012 quoted in Watchman, 2014.

<b>Behaviour-orientated</b>	A full functional analysis of the behavior in question will enable a systemic understanding of the behavior as a form of communication. The best practice principles that should be considered when supporting ageing adult with intellectual disabilities and dementia include simplifying multistep activities/skills, matching the level of demand on the individual with that of their current capacities, employing a range of prompts to facilitate communication and to modify the environment insofar as possible to compensate for deficits and capitalise on the individual's strengths. For carers, recommended practice also encourages the adoption of a proactive approach to identify potential stressors (or triggers) that can lead to distressed behaviours and moderate change as necessary.
<b>Emotion-orientated</b>	The underlying principles for such interventions are to reduce distress, validate a sense of self, enhance emotional wellbeing and support coping strategies. Psychodynamic approaches appear helpful for understanding intrapsychic concerns, cognitive/ behavioural techniques assist individuals in the early stage to build coping strategies and reduce distress, reminiscence and life review approaches provide individuals in the mild to moderate stage of dementia with interpersonal connections.
<b>Cognition-orientated</b>	The aim of these techniques is to compensate for cognitive deficits by utilising behavioural approaches to focus on specific cognitive and behavioural impairments and help to optimise remaining abilities. These techniques include skills training.
<b>Stimulation-orientated</b>	These treatments include recreational activities (such as crafts, games, pets) and art therapies (music, dance, art) to provide stimulation and enrichment that will engage the individual's available cognitive and emotional resources. Approaches such as life work that include life stores, valuables and memories pictures/photographs/objects are powerful ways of relating to the individual with dementia in a person-centered way. Reminiscence work is also important as a process of recalling experiences and events memorable for the individual by using different mediums such as verbal, visual, musical, tactile and smell. Anecdotally, in using reminiscence with groups of aging adults with intellectual disabilities, Kalsy-Lillico has found that its associative process is that one memory leads to another so that one person's shared recollection usually sparks off associated recollections or 'memoires' in others, has had positive effects on engagement and communication as reminiscence makes connections between a person's past, present and future.

## 12.5 Other interventions

There are also many useful non-dementia-specific interventions available. All depend on competent staff assessing and intervening as appropriate at the level that is required (i.e. individual/ family/service). The main interventions are outlined below in Table 4.

**Table 4:** Summary of useful interventions to improve the care of people with dementia

<b>Person-Centred Approaches to Support</b>	<ul style="list-style-type: none"> <li>• Person Centered plan.</li> <li>• Health Care Plan.</li> <li>• Communication Passport.</li> <li>• Life Story Book.</li> <li>• Advanced Directives &amp; end of life planning.</li> </ul>
<b>Physical Health</b>	<ul style="list-style-type: none"> <li>• Health checks leading to a Health Action Plan and any treatment. Tools available e.g. OK Health Check (Matthews, 2006) End of Life support - Gold Standard Framework (Thomas &amp; DH, 2005).</li> <li>• Management of weight.</li> <li>• Management of pain.</li> <li>• Management of sleep.</li> <li>• Management of epilepsy.</li> <li>• Management of medication.</li> </ul>
<b>Mobility</b>	<ul style="list-style-type: none"> <li>• Strategies to maintain mobility, promote exercise, address posture especially regarding respiratory function, correct gait and reduce the risk of falls. Consider equipment needed and promote safe manual handling.</li> <li>• Control of pain and discomfort.</li> <li>• Pressure area care.</li> <li>• Treatment of any difficulties of motor function, adaptation and teaching of skills to include compensatory techniques.</li> <li>• Equipment (eg. hoists, profiling beds, etc.).</li> </ul>
<b>Eating/Drinking</b>	<ul style="list-style-type: none"> <li>• Eating and drinking, strategies for maintenance of adequate oral intake in a safe manner. Swallowing assessments, eating programme with dietetic advice and advice regarding posture.</li> <li>• Diet to reduce risk of constipation.</li> <li>• Dysphagia management.</li> </ul>
<b>Continence</b>	<ul style="list-style-type: none"> <li>• Aids/adaptations.</li> <li>• Help to maintain continence.</li> </ul>
<b>Communication</b>	<ul style="list-style-type: none"> <li>• Strategies to improve communication.</li> <li>• Use of a 'communication passport' i.e. information on how best the person receives information and expresses themselves.</li> <li>• Use of objects of reference, pictures.</li> <li>• Environmental signposting/signs and symbols.</li> <li>• Intensive interaction.</li> </ul>

Table 4 continued

<b>Environment</b>	<ul style="list-style-type: none"> <li>• The person lives and spends time in environments that are familiar to them.</li> <li>• Depth perception problems are minimized for example flooring colour is consistent.</li> <li>• Necessary aids/adaptations to help find their way around and meet their needs e.g. red toilets doors, toilets seats, good signage, handrails, chairs at a good height.</li> </ul>
<b>Self Help Skills</b>	<ul style="list-style-type: none"> <li>• Modeling / guiding / prompting / pacing.</li> <li>• Adding supports to maintain self-care and domestic skills.</li> <li>• Adaptation of the environment to aid daily living.</li> <li>• Assistive technology.</li> </ul>
<b>Occupation/Activity</b>	<ul style="list-style-type: none"> <li>• Sensory stimulation (including Snoezelen).</li> <li>• Aromatherapy.</li> <li>• Music and art therapy.</li> <li>• Support for people to engage in activities.</li> <li>• Other failure free activities e.g. looking at magazines, carers sitting with the person describing what is happening.</li> <li>• Music activities.</li> </ul>
<b>Orientation/Confusion</b>	<ul style="list-style-type: none"> <li>• The person is able to understand their daily routine through the use of appropriate cues and aid for example, daily picture timetable, picture menus and staff rotas.</li> <li>• Reminiscence.</li> <li>• Life story work.</li> </ul>
<b>Mental Health and Behaviour</b>	<ul style="list-style-type: none"> <li>• Anxiety Management.</li> <li>• Intervention for Depression.</li> <li>• Relaxation techniques.</li> <li>• Promotion of positive behaviour and feelings of self esteem.</li> <li>• Intensive Interaction.</li> <li>• Observations to gain an understanding of behaviours.</li> <li>• Functional analysis and setting up of programmes for challenging behaviours utilising a Positive Behaviour Support Approach.</li> <li>• Setting up of crisis intervention plans.</li> </ul>
<b>Work with families and carers</b>	<ul style="list-style-type: none"> <li>• Education about dementia and how to care in a psychological way.</li> <li>• Training for staff team.</li> <li>• Support offered to family and carers, through systemic consultation opportunities for formulation and problem solving.</li> </ul>

### Key points

- It is important to utilise the skills of the multi-disciplinary team to support the individual with intellectual disabilities and dementia and their network of support.
- A range of interventions should be available to meet the person's need as their dementia progresses.
- Other agencies as well as those in the statutory sector can help to support the person and their carers.

## Section 13 – Medication

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### 13.1 Anti-dementia medications in people with intellectual disabilities and dementia

The degeneration of nerve cells in the brain leads to a reduction in neurotransmitters (a group of chemicals which have an important role in the transmission of signals between nerve cells). The neurotransmitter Acetylcholine is particularly affected in Alzheimer's disease. Acetylcholine plays a key role in the way the brain processes and consolidates information.

All the anti-dementia medication, with the exception of Memantine, increase the level of Acetylcholine available for transmission of nerve signals by delaying its breakdown. This is achieved by inhibiting an enzyme – Acetylcholinesterase – which is responsible for Acetylcholine breakdown and normally helps to maintain the balance of chemicals in the brain. Another enzyme involved in the breakdown is Butyl Cholinesterase and one of the anti-dementia drugs (Rivastigmine) inhibits this enzyme in addition to its effect on Acetyl Cholinesterase. It is important to note that none of the anti-dementia drugs developed to date are disease modifying; rather, they treat symptoms, but are unlikely to have a significant impact upon the pathological processes and are therefore not considered to prolong life.

- Acetylcholinesterase inhibitors are used mainly in Alzheimer's type dementia, but also have demonstrable improvements in other dementia types, particularly Lewy Body dementia
- NICE guidance – CG 42 (2012) recommends the use of Acetylcholinesterase inhibitors for mild to moderate Alzheimer's dementia to delay the progress of the illness. Memantine is to be used in those individuals with moderate dementia or where the Acetylcholinesterase inhibitors have proven unsuccessful (due to side effects or unsuitability). NICE has however restated that the difficulty in staging dementia in people with intellectual disabilities should not disadvantage them, thus introducing some flexibility for prescribers.
- These medications can also be used in the management of behaviour/psychological problems in people with Alzheimer's disease and Lewy Body dementia where psychological/environmental measures alone are not successful.
- Although there is no conclusive evidence that these drugs are effective in slowing cognitive decline in people with intellectual disabilities, the available evidence suggests that they may improve the quality of life both for the person and their carers.

Table 5: Anti-dementia medications and reviews of their effectiveness

Method of Action	Drug	Evidence
Acetyl & Butyl Cholinesterase Inhibitor	Rivastigmine	<p>Prasher et al. (2005): In a non-randomised trial (17 treatment, 13 controls), people who were treated with Rivastigmine had less decline over 24 weeks in global functioning and adaptive behaviours but no statistical difference.</p> <p>Prasher et al. (2013): In a non-randomised trial (27 treatment, 13 controls) both oral and transdermal Rivastigmine treatment was associated with significantly less decline in both cognitive and global functioning over a six-month period.</p>
Acetylcholinesterase inhibitor	Donepezil	<p>Prasher et al. (2002): Double blind placebo controlled trial of Donepezil, showed that the improvement at 24 weeks was statistically non-significant. The sample size of the study was too small to explore the efficacy in the subgroups of mild to moderate disease.</p> <p>Lott et al. (2002) in their open label study on Donepezil found that treatment resulted in significant improvement in scores on the Down's Syndrome Dementia Scale (Gedye, 1995). However, there were methodological drawbacks.</p> <p>Prasher et al. (2003), in their open label study on Donepezil treatment for people with Down's syndrome, found that treatment with the anti-dementia drug was associated with initial improvement in global functioning and adaptive behaviours. Follow-up at 104 weeks found that, whilst there was deterioration in both treatment and control groups, it was significantly less in the treatment group.</p> <p>Kishnani et al. (2010) published a trial of Donepezil in children and adolescents with Down's syndrome to improve cognitive functioning. They finally concluded that Donepezil does not significantly improve memory and other cognitive functions after a large (<math>N = 129</math>) randomised placebo controlled trial in 10–17-year-olds. These studies are notable in that they demonstrated that Donepezil is well tolerated, even in children with intellectual disabilities.</p>
	Galantamine	No studies in people with Down's syndrome.



Method of Action	Drug	Evidence
NMDA antagonist	Memantine	Hanney et al. (2012): Randomised, double-blind, placebo-controlled trial of Memantine for dementia in adults older than 40 years with Down's syndrome (MEADOWS); the authors included older adults with DS who did not have clear signs of dementia but argued that due to the high risk for AD in this population and the potential neuroprotective effect of memantine such an approach was justifiable. This was a well-designed study that included 88 patients actively in treatment and 85 patients receiving placebo and demonstrated no difference between the active and control group and concluded that Memantine was not beneficial for people with Down's syndrome and cognitive decline.
Antioxidants	900IU alpha-tocopherol, 200 mg ascorbic acid & 600 mg alpha-lipoic acid	Lott et al. (2011): Randomised placebo controlled trial of antioxidants in adults with DS and dementia ( <i>N</i> = 53). Antioxidant supplementation was found to be safe, though ineffective.

- At present there is no known cure for Alzheimer's dementia. Use of anti-dementia drugs at best may improve global functioning or reduce the rate of decline significantly. The natural progression of the disease may be delayed for a period by the medication but ultimately will continue. Eventually tolerance to the medication may decrease with an increased susceptibility to side effects. At this point, medication will need to be discontinued. It is therefore imperative to make it clear to carers and service users at the outset that the medication may be withdrawn at some point in the future.
- There is anecdotal evidence to suggest that people with Down's syndrome respond well to lower doses of the medication. It is, therefore, feasible to maintain treatment at the lowest effective dose. If symptoms re-emerge at a later stage, the dose can be increased.
- The effect/side effects of the medication should be monitored closely. This can be done using a number of different tools, but as general principle should include measures of cognitive, social and adaptive skills. Medication should be used for as long as it is effective (i.e. stabilizes or delays the progression of symptoms).
- There are a number of case studies, audits and anecdotal evidence that suggests people with Down's syndrome tolerate Acetylcholinesterase medication well and that it is perhaps more effective at managing the symptoms of dementia in people with Down's syndrome than it is in the general population.

- There is a theory that myoclonic epilepsy is mediated via cholinergic pathways. Myoclonic seizures are commonly associated with the epilepsy that occurs in people with Down's syndrome and Alzheimer's dementia. Although the Acetylcholinesterase inhibitors do not increase cholinergic pathways per se, they do improve the efficacy of neurotransmission. It is thought that this may result in reduced myoclonic seizures in those people taking this medication. There have been no published trials and there is no firm evidence to support this at present, but it is important to monitor the onset and progression of neurological symptoms such as myoclonic epilepsy as part of clinical follow-up following initiation of treatment.
- There are no longitudinal studies that describe the progress of dementia in people with Down's syndrome. Although post mortem studies detail that neurofibrillary tangles and neurofibrillary plaques are found in people with Down's syndrome over the age of 40, there is still a significant distribution in age of diagnosis of dementia. This may in part account for the significant variation in life expectancy following the diagnosis.

## **13.2 Psychotropic and other medications in people with intellectual disabilities and dementia.**

Use of psychotropic medications to control behaviour problems in dementia should only be considered as a last resort. The vast majority of people with intellectual disabilities and dementia with behaviour problems can be managed with environmental and other psychosocial approaches which are detailed in this guidance in Sections 11 and 12. Every effort should be made to carry out a thorough assessment to determine the reasons for such behaviours and take the necessary remedial actions (BPS, 2004; RCPsych et al., 2007).

In spite of the use of environmental and psychological interventions, there may be a small group of individuals with dementia who may benefit from use of psychotropic medications. The following are examples where a use of a low dose of medication may be considered:

- The presence of delusions and hallucinations causing significant distress may warrant the possible use of antipsychotic medications.
- People with depressive symptoms may benefit from an antidepressant treatment.
- In addition, there will be a small number of people with behaviour problems where clinicians would consider psychotropic medications to minimize the risk to the individuals themselves or other people.
- In these situations medications are considered as an option due to one of the following reasons:
  - the behaviour is continuing with significant risk to self or other people in spite of the use of the psychosocial / environmental approaches;
  - or
  - a short-term management where it is not practically possible to achieve the risk reduction with any other measures immediately.

The NICE guideline CG 42 on dementia (NICE, 2012b) states that 'people with Alzheimer's disease, vascular dementia, mixed dementias or Dementia with Lewy Bodies with severe non-cognitive symptoms (psychosis and/or agitated behaviour causing significant distress) may be offered treatment with an anti-psychotic drug after the following conditions have been met'. The conditions listed are:

1. There should be a full discussion with the person with dementia and/or carers about the possible benefits and risks of treatment. In particular, cerebro-vascular risk factors should be assessed and the possible increased risk of stroke/transient ischaemic attack and possible adverse effects on cognition discussed.
2. Changes in cognition should be assessed and recorded at regular intervals. Alternative medication should be considered if necessary.
3. Target symptoms should be identified, quantified and documented.
4. Changes in target symptoms should be assessed and recorded at regular intervals.
5. The effect of comorbid conditions, such as physical illness, pain and depression, should be considered.
6. The choice of antipsychotic should be made after an individual risk–benefit analysis.
7. The dose should be low initially and then titrated upwards.
8. Treatment should be time limited and regularly reviewed (every three months or according to clinical need).

For people with Lewy Body dementia, healthcare professionals should monitor carefully for the emergence of severe untoward reactions, particularly neuroleptic sensitivity reactions (which manifest as the development or worsening of severe extrapyramidal features after treatment in the accepted dose range or acute and severe physical deterioration following prescription of antipsychotic drugs for which there is no other apparent cause).

It is important to follow good practice principles (Deb et al., 2006, Tyrer et al., 2008) in treating people with intellectual disabilities and dementia:

- For people with intellectual disabilities and dementia, any such treatments should be considered on a best interest basis if the person is unable to provide informed consent.
- People with intellectual disabilities and dementia may be unusually sensitive to certain types of psychotropic medications due to reduced drug metabolism, reduced drug clearance and reduced plasma protein binding.
- Some underlying conditions such as Lewy Body dementia may make the person particularly sensitive to the use of even small doses of a neuroleptic, e.g. haloperidol. For these reasons it is important to start medications at a lower dose, titrate slowly and review frequently (with a minimum of three-monthly reviews).
- Every effort should be made to keep the dose at the lowest level at which the drug is effective.
- Side effects may not always be reported by the service users due to impaired communication in many, and it is therefore essential for the prescribing clinician to make both the users and carers aware of what to look for in terms of side effects.

### **Use of antidepressants:**

To treat symptoms of depression selective serotonin reuptake inhibitors (SSRIs) are preferred but attention is needed to the risk of developing low sodium levels. Sertraline has been found to be effective in the treatment of depression in people with dementia in a randomized control trial (Lyketsos et al., 2003). There is emerging evidence that SSRIs may be used for treatment of agitation in dementia (Nyth & Gottfries, 1990; Pollock et al., 2002,. Many clinicians prefer trazodone for which there is some evidence available for its effectiveness (Sultzer et al., 2002).

### Use of mood stabilisers:

Drugs such as Carbamazepine or Valproate may be considered if there is evidence of rapid cycling mood disorder or significant mood fluctuations.

#### Key points

- Acetylcholinesterase inhibitors are used mainly in Alzheimer's type dementia. NICE guidance (2013) recommends their use in mild to moderate dementia to delay the progress of the illness. NICE has however restated that the difficulty in staging dementia in people with intellectual disabilities should not disadvantage them, thus introducing some flexibility for prescribers.
- Psychotropic medications have only a limited role in the management of neuropsychiatric symptoms in people with intellectual disabilities and dementia and should only be considered if other environmental/psychosocial approaches have produced only very limited or no benefit and the risk from the symptoms is assessed as high.
- Antidepressant medications are useful in the management of depressive symptoms in people with dementia and intellectual disabilities.
- Caution should be exercised in the use of antipsychotic medication in the context of the evidence of a high risk for cerebrovascular events and mortality.
- Where psychotropic medications are used, target symptoms should be clearly recorded; risks and benefits discussed with the person and/ or carers and the minimum effective dose should be used for the shortest length of time.

## Section 14 – Safe eating and drinking

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Eating and drinking are basic human functions. It is vital that the person with dementia is assisted to make eating and drinking as enjoyable and stress free as possible. Each person will have their own beliefs about food. Eating is often a social experience as well as a means of gaining nutrition. Individuals have different reactions to different foods that come from their experience, culture and taste, and these need to be taken into account in understanding the person with dementia.

As the dementia progresses, safe eating and drinking becomes more of an issue. Staff need to be flexible and person-centred, remembering that the person will have good days and bad days. This may mean that carers and staff will need to completely rethink how to approach eating and drinking for the person in order to ensure that it is a safe, enjoyable and nutritious activity.

### 14.1 What makes people want to eat and drink?

Each person will respond to a large number of cues that make them want to eat and drink. These include: time of the day, smell of food, rattle of crockery and cutlery, the table being set, the colour of the food, how the food is presented and the taste of the food. Similarly there are a number of reasons that make people less inclined to eat or drink. These include when the smell is off-putting, the place is noisy or dirty, the food doesn't look nice, the person doesn't like the people they are sitting with, they don't feel well or are too tired. A person may also not wish to eat as they do not recognise that the feeling coming from within is one of hunger, as the connection between the sensation in their stomach and the thought that they need to eat is lost.

It is therefore essential that the chances of the person enjoying their food and successfully eating and drinking are maximised by paying attention to a number of factors. These include:

- The opportunity to be involved in preparing what they eat all but vanishes. Preparing what you eat can improve levels of wellness and positively affects a person's ability to chew and swallow food. It also impacts on quality of life, not just from a nutritional perspective, but also in terms of wellbeing and it is a great social opportunity for people.
- Promoting people to do things for themselves can have a significant impact on improving a person's appetite. All stages of meal/snack preparation will stimulate saliva production and activate the centres in the brain involved in swallowing.
- People should be involved in every aspect of planning and preparing a meal, from creating a shopping list, selecting the produce off the shelves, to the final serving of the meal. This will often mean thinking creatively and it may not always be about preparing a main meal, but may be providing the opportunity to do part of the task which focuses on people's remaining abilities.

- Eating in a dining room environment: a communal environment may contribute favourably to food consumption. Set up an environment that allows the individual to wander to eat finger foods along their path. Keep auditory distractions to a minimum (noise from television, radio, and people moving in and out of the dining room).
- Posture and positioning of the person is important to safe eating.
- Understanding why people need to eat and drink and ways of encouraging eating are important, e.g. looking at what people are eating and when – including safe foods, presentation, finger foods, amount of food, and reacting promptly to eating and drinking difficulties.

## 14.2 Eating and drinking difficulties

As the dementia progresses, eating and drinking difficulties become more apparent. The types of difficulty can be characterised as follows:

- **Difficulty chewing, grinding or moving the food in the mouth:** this means that food does not break down and form a bolus and therefore cannot be propelled to the back of the mouth efficiently. Some people (particularly those with Down's syndrome) may have a backwards/forwards motion of the tongue which is different from the more full range of movement that most other people have. This more restricted movement is also frequently observed in people who have been unable to eat a normal diet requiring them to develop good chewing patterns e.g. People who have cerebral palsy or other physical disabilities. It can also be observed in people in the advanced stages of dementia. For people with dementia this may not be a motor problem to chew, but being able to remember to chew and difficulty with the sensation in the mouth triggering the idea they need to chew.
- **General muscle weakness or stiffness in the face and neck area** may mean that the person experiences difficulty controlling liquids in the mouth. Liquids and soft foods may dribble out of the sides or front of the mouth or disappear down the throat in an uncontrolled fashion, possibly entering the unprotected airway.
- **Changes in sensation** in the mouth whether reduced or heightened can drastically affect the eating or drinking process for the person. Changes in sensation may affect someone's tolerance of extreme temperatures or their enjoyment of certain flavours. People with dementia often respond to stronger tasting foods.
- **The swallow reflex sequence** can become uncoordinated. The swallow may be triggered too early or too late. This means that the bolus or liquid will pass down the throat at a point when the airway is not properly protected. This can lead to inhalation into the larynx or aspiration onwards towards the lungs. The mouth or the throat may become full and overflow. A swallow may be triggered, but some of the residue may be inhaled into the larynx or windpipe. It may be possible to hear this if the person's voice sounds 'wet' or more gruff than usual.
- **Saliva:** We produce a large amount of saliva which we normally swallow without even realising. Someone with a swallowing problem may appear to produce too much saliva, which then escapes from the mouth leading to drooling. Conversely, some people produce too little saliva, which can produce a dry mouth. A dry mouth makes it much harder to swallow and bacteria and organisms may begin to build up, making the person much more prone to infections such as thrush.

Once eating and drinking difficulties have been suspected, it is important that an immediate referral is made to their local speech and language therapy department. Carers and staff need to be aware of the key signs to watch for: coughing, choking, a red face, and watery eyes or loss of breath during eating or drinking.

Working with carers and staff needs to be ongoing to ensure that issues that help to minimise difficulties are recognised and acted on. These issues include:

- Being aware (by keeping a food and drink diary) of what textures and liquids seem to cause problems and which do not. Staff and carers need to avoid those that cause problems and make sure that everybody knows which they are.
- Checking that dentures fit properly. It is important to minimise the number of teeth removed as this will cause gum shrinkage and gum muscle weakness and dentures will then be harder to keep in place.
- Correctly positioning the body and particularly the head may greatly assist people to eat and drink safely. It is important that the person is referred to the speech and language therapist who will assess and advise on this together with multi-disciplinary team members.
- Remembering that even if the mouth itself feels and looks empty, there may be a build up of residue in the throat. It is advisable to encourage people to take their time, to make additional swallows, in order to clear as much of the residue down the food pipe into the stomach as possible. If left this residue may silently drip down into the airway. This is known as ‘silent aspiration’. Food and liquids silently aspirated onto the lungs may cause chest infections (aspiration pneumonia).
- A consistent approach is key. All persons who are going to assist an individual to eat and drink should ensure that they have consulted and understood any eating and drinking guidelines that are in place before they begin.
- Treat each meal, snack or drink as a new episode as the difficulties will fluctuate and people need to be aware of how the person is ‘in the moment’.
- Timed snacks can improve hydration and nutrition as well as medication compliance.
- Successful feeding in dementia may take up to one hour depending on the severity of the disease. Remember that people can become fatigued and may need a break before feeding is restarted.
- Interruptions during mealtimes distracts people with dementia, resulting in reduced nutrition and calorific intake.

Where staff or carers have concerns they need to keep a record of episodes of coughing, throat clearing, ‘wet’ voice quality, choking and also chest infections and consult a speech and language therapist.

Saliva can also be an issue for people with dementia. If people with muscle weakness are experiencing loss of saliva, etc. from the mouth then staff and carers need to gently dab away the moisture – never wipe it, as the action of wiping across the lips has the effect of stimulating more saliva, as there is a salivary gland in the chin area. Where too little saliva is being produced, the person should be offered frequent small drinks to keep the mouth hydrated and care taken with a regular oral hygiene routine. Wiping the lips in this instance may be beneficial as it stimulates the sub-lingual salivary gland to produce saliva but this needs to be discussed with a speech and language therapist or other relevant specialist.

### Key points

- Eating and drinking are basic human functions.
- It is vital that the person with dementia is assisted to make eating and drinking as enjoyable and stress free as possible.
- All eating and drinking difficulties must be taken extremely seriously and an urgent referral made to a speech and language therapist.



## Section 15 – Palliative care and end of life issues

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Dementia is a progressive and, at present, terminal illness. Its course, whilst varying in detail from person to person, is by definition associated with the progressive loss of skills and the ability to communicate, ending in a final stage where the person becomes completely dependent on others for their every need. As the illness progresses, so it becomes harder for others to ascertain the wishes of the person suffering from dementia and this is likely to be particularly the case when the person has pre-existing intellectual disabilities. There is much evidence that historically, and currently, people with intellectual disabilities experience barriers in accessing quality health care for serious illness and underuse palliative care services.

For these reasons preparing for the end of life and taking a palliative care approach is of importance from the time of establishing a diagnosis onwards. Forward planning concerning these issues needs to be embedded in the person-centred plan. (See Section 7 – Breaking the News). The generally fairly long course of dementia allows time for the person him/herself and for others to prepare and to ascertain how they wish to be supported and to plan for how their health and well-being will be maintained and for how they will be supported at the end of life and when dying.

### 15.1 End of Life strategy

End of life issues have been addressed significantly over the last five years in all areas of Britain. Strategy documents in England, Scotland, Northern Ireland, Ireland and Wales are consistent in the information, guidance and imperatives they give concerning the approach to palliative and end of life care and they all stress that this should focus on the person rather than the disease and aim to ensure quality of life for those living with an advanced non-curative condition.

For people with intellectual disabilities there is now a strong emphasis on involving them in their end of life care as for all other aspects of their life. This is made explicit in *Improving the Health and Well-being of People with Intellectual Disabilities: An evidence-based commissioning guide for clinical commissioning groups* (Learning Disabilities Observatory et al., revised 2013) which states that people with intellectual disabilities should be able to have the same end-of-life care planning and access the same palliative care services as everyone else, as this contributes to effective and coordinated care and a good death. *The Route to Success in End of Life Care – Achieving quality for people with intellectual disabilities* is a practical guide to improving end of life care for people with intellectual disabilities, within the NHS National End of Life Care Programme (2011). The NICE *Quality Standard for End of Life Care* (2012a) makes it clear that it is now an agreed standard that people with intellectual disabilities should receive the same palliative and end of life care as the rest of the population.

In England the Department of Health *End of Life Strategy* (DH, 2008b) set out the key areas for action to ensure that people have excellence in end of life care. This included developing strong links with specialised end of life services, e.g. hospices, palliative care services, and admiral nurses.

NICE *Quality Standard for End of Life Care* (2012a) covers all settings and services in which care is provided by health and social care staff to all adults approaching the end of life and also covers support for the families and carers.

## 15.2 Addressing end of life issues

The approach advocated by both the DH (2008b) and NICE (2012a) is a holistic one which addresses psychological and physical needs, social and practical needs and spiritual and religious needs. It also gives guidance on access to specialist palliative care, care of the body after death and support with bereavement. For all areas it specifies cultural sensitivity and dignity and explicitly states that a person should be offered spiritual and/or religious support appropriate to their needs and preferences.

Health and social care professionals are required to act to identify people approaching the end of life in a timely way and to give them and their family and carers opportunity to discuss, develop and review a personalised care plan for current and future support and treatment. The treatment plan is for symptom management and for current and anticipated physical and specific psychological needs. The care plan needs to outline personalised support for their social, practical and emotional needs, appropriate to their preferences, and should aim to maximise independence and social participation for as long as possible. There is a focus on essential services being available and accessible at all times to those approaching the end of life who need them.

This includes people approaching the end of life receiving specialist palliative care if their usual care team are unable to relieve their symptoms adequately. It is emphasised that people should be offered this care in a timely way at any time of day or night.

The NICE Quality Standard was updated in late 2013 following the review of the Liverpool Care Pathway and the subsequent announcement of the phasing out of this pathway. There is now a new approach and new priorities to looking after people who are dying, launched by the Leadership Alliance for the Care of Dying People (LACDP). These priorities state that decisions about care and treatment are to be made in accordance with the needs and wishes of the person and should be reviewed and revised regularly, communication between staff and the person who is dying should be sensitive, the dying person is to be involved in decisions about their care, people important to the dying person are to be listened to and their needs respected, and that care should be tailored to the individual and delivered with compassion.

It is clear that good practice in dementia care for people with intellectual disabilities requires working with the individual with dementia, their families and carers from an early stage concerning palliative and end of life care and that this means being aware of and taking into consideration a range of factors in a person's life including their personal history and preferences, their family and other relationships and their cultural and religious background with particular reference to ideas and beliefs about serious illness, dying and death.

Good practice guidance also states that people in significant relationships to the person who has died need to be supported in their loss and bereavement and peers and co-residents with intellectual disabilities need to be included in interventions that give support after the person's death.

As new problems arise their occurrence should not be a surprise and should have been anticipated. In the early stages of dementia the issues listed below should be considered as these will all become relevant towards the end of the illness or at the person's death. The individual with intellectual disabilities and dementia should always be involved whatever their capacity. If they lack capacity or their capacity diminishes then it will be necessary for the appropriate professionals to make Best Interest decisions within the context of the Mental Capacity Act (2005). For complex and serious decisions or where there are conflicts concerning treatment, for instance between the family and the medical team, it will be necessary to involve an Independent Mental Capacity Advocate (IMCA) and the Court of Protection.

Early consideration needs to be given to:

- An understanding of where, how and by whom the person would like to be supported towards the end of their life and whether he/she would like to appoint someone to make healthcare decisions on his/her behalf when he/she no longer has the capacity to do so.
- Future management of financial affairs such as the appointment of a lasting power of attorney for property and affairs or guardianship or appointee arrangements
- The making of a will.
- An understanding of the person's preference with respect to the use of or withholding of more invasive treatments in the advanced stages of dementia when he/she will no longer have the capacity to consent. These may include the use of artificial nutrition and hydration, ventilation, and the treatment of infections. If, at an earlier stage of the illness, the person has the capacity to do so they may be supported to make an advanced statement with respect to these matters. As noted, the Liverpool Care Pathway is no longer in place and the approach and priorities outlined by the LACDP (LACDP, 2014), which emphasises that the person who is dying and the people important to the person must always be involved in decisions about care and treatment, should be followed
- Funeral arrangements, again in discussion with the person and the people important to them.

### **15.3 Palliative care and the role of the palliative care team**

It is important to develop working partnerships and to have close collaboration between different health services and a mutual understanding of philosophies of care including professionals in intellectual disabilities services understanding the role and timing of palliative care in terms of access to specialist support and appropriate symptom management. This needs to include considerations about getting support in the home so that unnecessary hospital admissions can be avoided. The local intellectual disabilities hospital liaison nurse can be very important in promoting joint working. It is also helpful to have a clinician within intellectual disabilities services focusing on palliative and end of life care who can promote this collaboration and lead training within their services about end stage dementia and the role of palliative care.

The nature of dementia makes it difficult to predict whether a person is reaching the end of their life, and this can lead to difficulties in terms of access to palliative care services.

The aims of this approach are to support quality of life, to help the person die with dignity and without pain and in a place of their choosing and to provide support to relatives and friends to help them prepare for the death.

The following are areas that may require particular attention towards the end of the person's life:

**Eating and drinking:** People should be supported and encouraged to eat and drink by mouth for as long as possible, with specialist advice being obtained concerning any feeding and swallowing difficulties from multidisciplinary team professionals particularly speech and language therapists, occupational therapists and physiotherapists (concerning seating and posture) and also from dieticians. Weight loss is common in late-stage dementia. Nutritional support, including artificial (tube) feeding should be considered if dysphagia is thought to be a transient phenomenon. NICE guidance (2013) does not recommend that such artificial means are used in those with severe dementia for whom dysphagia or disinclination to eat is a manifestation of dementia severity. As specific ethical and legal principles apply with respect to withholding or withdrawing nutritional support expert advice should be sought if there is any lack of clarity or disagreement in this area. Further information can be found in Section 14.

**Resuscitation:** It is generally considered that cardiopulmonary resuscitation is unlikely to succeed in cases of cardiopulmonary arrest in people with end stage dementia. For those people who have retained their capacity to make a decision on this matter, it is for them to decide whether or not to agree to resuscitation if it were needed. For those who now lack the capacity to make such decisions, any wishes expressed in a valid and applicable advanced decision to refuse treatment must be respected if the circumstances are applicable. The policies and procedures set out in the Mental Capacity Act 2005 (or similar legislation in other jurisdictions) should be followed.

**Pain relief:** Unexplained changes in behaviour or evidence of distress in a person with dementia may be indicative of underlying pain. The possible cause for pain needs to be investigated and necessary treatments undertaken and both pharmacological and non-pharmacological approaches to pain relief considered.

**Posture:** Management of posture is important in order to optimise remaining abilities and prevent the development and/or progression of secondary complications such as pain, fatigue, muscle shortening, joint deformity, respiratory complications and pressure ulcers. There needs to be assessment of postural management equipment needs such as standing aids (where appropriate), specialist seating provision and sleep systems. Good postural management can also facilitate safe eating and drinking and improve respiratory function.

**Resources:** Accessible booklets are available to help people with intellectual disabilities plan their end of life care, e.g. *When I Die, Sunderland People First* in association with the PCPLD Network (Palliative Care for People with Learning Disabilities). The network is a voluntary organisation made up of people with intellectual disabilities and palliative care professionals concerned by their experience that people with intellectual disabilities who were facing a life-limiting illness did not seem to access the same services or receive the same quality of service as the rest of the population. This group also offers other useful resources

*Making a Will*, an accessible leaflet for people with intellectual disabilities concerning writing a will, is available from Mencap (2014).

### Key points

- Preparation for palliative and end of life care should be ongoing as the dementia progresses from diagnosis onwards and embedded in personalised plans for care and support at every stage.
- People with intellectual disabilities must be able to have the same end-of-life care planning and access the same palliative care services as everyone else.
- All care should be provided in accordance with the provisions of the Mental Capacity Act. Care includes medical care, physical care and psychological and emotional care of the person, and psychological and emotional care of family, friends and peers, and carers and staff, and this needs to be delivered with cultural sensitivity and where appropriate, religious support.
- Partnership working and close collaboration between professionals in intellectual disability and other health services, particularly palliative care, is very important in terms of ensuring appropriate access and timing to specialist support and appropriate symptom management. A mutual understanding of philosophies of care in these services needs to be developed.

## **Section 16 – Capable commissioning for people with intellectual disabilities and dementia**

Commissioners of health and social care services are now far more aware of the increase in the prevalence of dementia in the general population, but it is still unclear about their knowledge and expertise in relation to commissioning services for people with intellectual disabilities and dementia, even though guidance was first distributed to commissioners in 2001 (Turk et al., 2001). As is the case post-Winterbourne, there is a need for local services and local competent teams who have expertise in working with people with intellectual disabilities who develop dementia.

Commissioners of health services need to be clear about the care pathway for the assessment, diagnosis, interventions and support for people with intellectual disabilities who develop dementia from primary care, through to appropriate secondary care services, and on to palliative care services. In practice, good quality care will involve active partnership-working between intellectual disability services, older people's services, primary and secondary health care, palliative care and social care. These areas should develop an integrated dementia strategy for the care of people with intellectual disabilities and dementia. This should involve the development of an integrated care pathway involving all relevant agencies. Specialist health professionals e.g. speech and language therapists, physiotherapists, occupational therapists, dietitians, community intellectual disabilities nurses are all essential partners in providing excellence in care.

Commissioners of social care need to recognise that dementia is by definition a deteriorating condition, and that peoples' needs will increase over time, and therefore increased and timely funding will be needed to ensure safe practice. This will inevitably involve the funding of waking night staff as the dementia progresses. Best practice is that people with intellectual disabilities should receive their residential care within the intellectual disabilities arena rather than in generic services for people with dementia, as the quality of the services gives people the best opportunities for a good quality of life for both their intellectual disabilities and their dementia. There will need to be efficient processes and understanding in place to ensure that Continuing Healthcare Assessments are undertaken promptly and funding agreed as needed.

Staff involved in assessment, diagnosis, interventions and support need to be trained in dementia care and be able to offer both holistic and specialist assessments and a range of interventions aimed at meeting the needs of people with intellectual disabilities and dementia. Care managers have an essential role to play in ensuring that services are actively monitored to ensure that they are responsive to the changing needs of the person. People with intellectual disabilities and dementia should have access to regular reviews (monthly–six monthly depending on the rate of deterioration) by an identified care manager.

End of life care needs to be planned in advance, using the same principles and services available to the general population. Good partnership-working with palliative care services is essential, both to support the person and the carers (see Section 15).

## 16.1 What are the elements of an excellent service?

Commissioners will want to ensure that there is:

- Demographics are known, including having a database of all adults with intellectual disabilities which includes identification of people with Down's syndrome and those in out-of-area placements.
- A multi-agency dementia strategy.
- A multi-agency care pathway for assessment, diagnosis, interventions and support of people with intellectual disabilities who develop dementia.
- A multi-disciplinary approach to assessment and diagnosis and support.
- Prompt access to assessment and diagnostic services including baseline assessment for people with Down's syndrome by the age of 30.
- Person-centred dementia care.
- Effective care management and review system.
- Prompt access to the full range of medical, psychological, therapeutic and social interventions.
- All living and day service environments are dementia friendly.
- The person is supported to remain in their familiar home with additional supports provided in a timely manner.
- Support is available to family carers and service providers.
- There is a capable trained workforce able to deliver excellence in dementia care.
- End of life care follows the requirements of the National End of Life Strategy.

## 16.2 What should the commissioners expect as outcomes of an excellent service?

- Increase in prompt differential diagnosis of the person's difficulties.
- Increase in other conditions being treated promptly.
- Increase in accurate diagnosis of dementia.
- Reduction in behavioural difficulties.
- Increase in quality of life indicators for the person.
- Reduction in moves to other placements.
- Reduction in the need for emergency one-to-one cover.
- Reduction in out of area placements.
- Increased carer support and satisfaction.
- Reduction in staff stress.

## 16.3 Potential risks if services are not available/not effective

- Increased costs of one-to-one, new in area or out of area placements.
- More complaints.
- Potential safeguarding issues.
- Increase in behaviours leading to abuse or harm to self and others.
- Carer breakdown.

### Key points

- Each area should develop a dementia strategy and integrated care pathway to support the provision of high quality assessment, intervention and care for people with intellectual disabilities who develop dementia.

## Section 17 – Capable support

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Excellence in dementia care for people with intellectual disabilities and dementia is underpinned by the knowledge and skills of the people who support them, and their ability to continuously adapt to the person's changing needs.

### 17.1 Family carers

Family carers need specific emotional and practical support. Many family carers find the diagnosis of dementia traumatic, as it may bring back emotions surrounding the birth, life expectancy and disability of the person. Furthermore, many families will also have prior experiences of family and friends being diagnosed with dementia and thus will have ideas about the journey ahead. Where the family are the main carers, they must be offered a comprehensive Carer's Assessment. Many carers, particularly the parents of people with Down's syndrome, may themselves be at risk of developing dementia or other age related conditions. Often diagnosis of dementia comes at a time when family carers are themselves reaching a time when they are requiring more support due to their own ageing.

Services need to be sensitive to the needs and beliefs of families, and to see things from their perspective. Some carers believe that it is their duty to care and may find it very difficult to accept support and help into their own home, or try to cope even when the person has needs that are greater than they can cope with. Carers need to have prompt access to appropriate information about supports and resources available, including short breaks (both within and away from the home), individualised budgets and direct payments, and aids and adaptations including assistive technology. They need to be involved in assessments and review meetings, even when their family member does not live with them.

Appropriate use of the National Framework for Continuing Health Care (DH, 2012b) should be made to ensure that funding of health and social care is fair and transparent.

Staff need to be very sensitive to the small number of carers who cannot cope with seeing their family member deteriorating, and may opt out of being involved. Life Story work is one positive way of enabling family carers to maintain a relationship or to stay involved in the person's care. Carers often need a great deal of support to prepare for the eventual death of the person they are supporting. Advanced planning can help carers to be involved in sharing future wishes, and to talk through issues and plan for the last years/months of life.

### 17.2 Paid care staff characteristics

Staffing numbers need to be appropriate to ensure the person is safe and that staff are able to meet the person's changing needs. In practice, this means that staffing levels will need to increase as the dementia progresses. By mid-stage dementia, people usually require waking night staff to ensure safety, and often an increase in staffing levels to manage self care and to respond to the distress that people with dementia often exhibit.

Excellent dementia care can only be provided by a consistent staff group. Agency staff,



unless very well known to the person, should always be avoided as this can add to the person's stress level. Staff will need to have access to regular training and resources to meet the varied demands of caring for people with intellectual disabilities and dementia. There are a range of resources available including the *Resource Pack for Carers of Adults with Down's Syndrome and Dementia* (Dodd et al., 2009), the *Down's Syndrome and Dementia Workbook for Staff* (Dodd et al., 2006), *Supporting Derek* (Watchman et al., 2010) and a range of DVDs from the Down's Syndrome Association. Dementia training may also be offered by local Community Intellectual Disabilities Teams. In our experience, the best care is provided by staff who are flexible in their approach, deal with changing situations with compassion and humour, cope with the person's declining abilities and can reach out to the person.

Staff need support to cope with the deterioration in the person with intellectual disabilities and dementia and to prepare for the eventual death of the person they are supporting. Research has indicated that caring for people at late stage of dementia raises specific issues related to their readiness to respond to end of life needs; fear of swallowing difficulties; and environmental concerns and ageing in place. See Section 14 for more details.

Staff are the key component in ensuring that people who live in any form of supported living or residential provision can 'live well' with dementia. The timing of training is important. It is important that a service is 'dementia-ready' before their residents begin to develop symptoms of dementia. Staff need to have a thorough understanding of the person, of dementia and the consequences of having dementia, and then how to adapt their care as the dementia progresses. This can only be achieved by having a clear framework to underpin the training and support provided to services. Evidence suggests that where staff had received relevant and targeted training that was practice-based and person-centred, they displayed an appreciable difference in confidence, quality of care and support and they also reported reduced stress levels.

Evaluation of training models used with staff who are supporting people with intellectual disabilities with other comorbid conditions suggest that the best outcomes occur when there is interactive training which involves the development of care plans; follow-up consultation and support for implementing care plans; and where there are changes in how the organisation understands and responds to people with dementia.

The importance of developing a shared vision on which to build practice is now well-recognised as the pre-requisite of good care. Without this solid foundation, values, expectations and approaches are likely to differ greatly amongst staff. This will ultimately generate conflict and frustration and will in turn place unnecessary demands on the already confused person with intellectual disabilities and dementia. Sheard (2013) recently introduced the concept of 'mattering' which 'brings together the core elements of emotional care, skills, quality of life and environment with the culture of a shared relaxed community, thereby evidencing that individuals matter'. He emphasises that developing the emotional competency in staff is the only foundation on which progress in dementia care will be sustained and on which dementia care training will finally deliver effectiveness (see Sections 9 and 10).

### 17.3 Delivery of training to staff

Dementia awareness training is often delivered to groups of staff from a range of services, often with junior staff attending. The only advantage of this model is that it allows staff to meet with colleagues from other settings and to learn from each other. However, this approach rarely results in change in practice once the staff member returns to their care setting. The person who attended the training has to try to ‘sell’ their knowledge and skills to the staff team, often with little commitment from management to implement changes. Even when this person is designated the ‘dementia champion’, this rarely results in the needed change in staff knowledge across the whole staff team (Dodd, 2014).

Experience in training staff teams indicates that improving dementia care for people with intellectual disabilities and dementia requires a whole-system approach within staff teams.

Staff need to understand what is dementia, its specific links to people with intellectual disabilities, and the signs of symptoms that they need to recognise both at pre-diagnosis and at each stage of the disease. However, this needs to be underpinned by a model of dementia that helps them to really understand what is happening to the person who they are supporting who is developing dementia, and to be able to put themselves in that person’s shoes (see Section 9).

### 17.4 Training content

Training should use a variety of mediums, e.g. didactic; group work; use of DVDs; discussions; role plays; case studies; homework tasks. Training can be supplemented by the use of specific resources e.g. the *Dementia Workbook for Staff* (Dodd et al., 2006), *Resource Pack for Carers of Adults with Down’s Syndrome and Dementia* (Dodd et al., 2009), *Supporting Derek* (Watchman et al 2010), *Happy Eating* (Dodd, 2012).

Table 6 suggests types of training, support and outcomes that should be achieved for each stage of dementia (Dodd 2014). Each stage builds on the information and supports from previous stages and knowledge and ideas can be re-iterated and developed with the staff team. For further details of the Outcomes required, see the chapter on outcomes later in this guidance.

### 17.5 Working with other residents

The effect on other people with intellectual disabilities of seeing a peer deteriorate is rarely considered. Helping peers to understand dementia can reduce their distress and help them to be more considerate and understanding of the changes occurring with the person with dementia, in some cases helping to prevent placements breaking down.

Resources now exist to help explain dementia to adults with intellectual disabilities. Dodd et al., (2005 a,b & c) published three booklets, *About Dementia*, *About My Friend and The Journey of Life*, to support this aspect of work. These booklets explain dementia in terms of the lifecycle. They can be used individually or in a group setting, as long as supported by confident staff/carers. Both Lyngaard and Alexander (2004) and Dodd (2008) have reported on successful short courses for people with intellectual disabilities living with someone with dementia.

**Table 6:** Suggested training, support and outcome for each stage of dementia

Stage of dementia	Suggested content of formal training	Suggested on-going support	Outcome required
Prediagnosis:	<ul style="list-style-type: none"> <li>• Signs and symptoms of dementia</li> <li>• The diagnostic process and how to make a referral</li> <li>• Life Story work</li> <li>• Person-centred care plans including health action plan, communication passport, end of life plans</li> </ul>	<ul style="list-style-type: none"> <li>• Understanding the diagnostic process</li> <li>• Working with the person and their family / friends to develop their life story work</li> <li>• Support to develop end of life plans whilst the person has capacity</li> </ul>	<ul style="list-style-type: none"> <li>• Person is supported through the diagnostic process</li> <li>• Other health issues are identified and treated</li> <li>• Social issues are identified and resolved</li> <li>• Plans are in place</li> </ul>
Early stage:	<ul style="list-style-type: none"> <li>• Model of dementia (Buijssen)</li> <li>• Philosophy of care</li> <li>• Physical environments</li> <li>• Importance of picture cues</li> <li>• Medication</li> </ul>	<ul style="list-style-type: none"> <li>• Helping staff to accept the diagnosis and the changes</li> <li>• Grief and loss</li> <li>• Implementing the philosophy of care</li> <li>• Importance of consistency of approach</li> </ul>	<ul style="list-style-type: none"> <li>• Person is supported to maintain their current lifestyle with additional supports and prompts</li> </ul>
Mid stage:	<ul style="list-style-type: none"> <li>• Communication</li> <li>• Supporting peers of the person with dementia</li> <li>• Failure-free activities</li> <li>• Maintaining health and additional health issues e.g. epilepsy, mobility, continence</li> <li>• Pain recognition and management</li> <li>• Reminiscence</li> </ul>	<ul style="list-style-type: none"> <li>• Understanding the meaning of behaviours and exploring solutions</li> <li>• Avoidance of confrontation</li> <li>• Understanding and coping with agitation and distress</li> </ul>	<ul style="list-style-type: none"> <li>• Person is supported to live as full a life as possible focussing on preferred activities and reminiscence activities and without unnecessary changes</li> </ul>
Late and end stage:	<ul style="list-style-type: none"> <li>• Safe manual handling</li> <li>• Safe eating and drinking</li> <li>• Skin and pressure care</li> <li>• Mobility, falls management, posture and positioning</li> <li>• Meeting spiritual needs</li> </ul>	<ul style="list-style-type: none"> <li>• Support for end of life care for both the staff and their support of the person's peers and family</li> <li>• Support in getting appropriate aids in a timely manner e.g. specialist wheelchair, seating, profiling bed, hoist, bathing aids</li> </ul>	<ul style="list-style-type: none"> <li>• Person receives care that allows them to continue to experience activities and support that is familiar to them</li> <li>• Person is supported in all daily living needs in a dignified and safe manner</li> <li>• Person experiences end of life care that results in a 'good death' in their preferred place.</li> </ul>

## 17.6 Ethnicity Issues

Ethnic background has been shown to affect the nature of care giving practices, carer stress and help seeking in people with dementia and their carers (Ilfie & Manthorpe, 2004). In people with intellectual disabilities in general there are significant differences in the nature of care giving, carer stress and carers' psychological and physical burden (McGrother et al., 2002, Devapriam et al., 2008). Contrary to the popular view, very few enjoy the support of extended family networks (Hatton et al., 2002). Material disadvantage, lack of informal networks and high need for services are some of the key issues related to people with intellectual disabilities from an ethnic minority background (Hatton et al., 1998).

However, it is not clear how these processes are affected when an individual with intellectual disabilities from an ethnic minority background develops dementia. Clinical experience suggests that the lack of availability of culturally appropriate respite and home care provision cause difficulty. Lack of awareness of services and language barriers may prevent people from ethnic minority communities from receiving appropriate levels of support. This can result in significant distress for both the person with intellectual disabilities and their carers.

This highlights the need for more cohesive action by health, social care and voluntary sector services together to improve access to and the experience of services by people with intellectual disabilities and dementia from black and minority ethnic (BME) communities. The plan of action should include the following:

- A clear strategy for staff training on diversity.
- Proactive measures to develop partnerships with community and voluntary organisations.
- Facilitation of active participation by people with intellectual disabilities and carers from BME background in the stakeholder discussions about the development of culturally appropriate services for people with intellectual disabilities and dementia.
- Direct payments and individualised budgets may provide an opportunity for increasing the range of culturally appropriate services available.
- Further research is required to understand the views of people with intellectual disabilities with dementia from BME communities and their carers about culturally appropriate way of meeting their needs.

### Key points

- Staff are key to ensuring that people with intellectual disabilities and dementia can 'live well' with the disease.
- Training must include developing a shared vision on which to build practice – this can only be achieved by training the whole staff team together.
- Training is not a 'one-off' activity, but should be delivered on an ongoing basis as the person's dementia progresses to ensure that staff are capable of meeting the person's changing needs and achieving the best outcomes for the person.
- Capable dementia services need to be aware of, and meet the needs of, the person with dementia, those of staff and carers and other people with intellectual disabilities who live with the person, including those from BME communities.

## Section 18 – Outcomes

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There are a range of quality standard documents which have been produced by different nations to define the expected standards of care for people with dementia.

The National Institute for Health and Care Excellence (NICE) has produced two quality standards (QS1, NICE, 2010; QS30, NICE, 2013) which define what constitutes a high standard of care for people with dementia for people in England and Wales.

NICE dementia quality standard QS1 (NICE, 2010) covers care provided by health and social care staff in direct contact with people with dementia in hospital, community, home-based, group care, residential or specialist care settings. This quality standard requires that dementia services should be commissioned from and coordinated across all relevant agencies encompassing the whole dementia care pathway. It states that an integrated approach to provision of services is fundamental to the delivery of high quality care to people with dementia. According to NICE (2010), this quality standard provides clinicians, managers and service users with a description of what a high-quality dementia service should look like. It describes markers of high-quality, cost effective care that, when delivered collectively, should contribute to improving the effectiveness, safety, experience and care for adults with dementia

NICE dementia quality standard QS30 (NICE, 2013) covers supporting people to live well with dementia. This set of standards applies to all social care settings and services working with and caring for people with dementia.

Unfortunately both these quality standards have been written as inputs rather than outcomes, and the measures for assessing compliance with these quality standards are purely quantitative in nature and that desired levels of achievement should be defined locally.

### 18.1 Measuring outcomes for services for people with intellectual disabilities and dementia

In our original guidance on dementia (BPS, 2009) a self-assessment checklist with 15 standards was developed that can be used to evaluate the provision of dementia care across health, social care and voluntary agencies in a geographical area. Many areas have used this as a basis of developing their local dementia strategy for people with intellectual disabilities and to benchmark their services and develop an action plan (e.g. Surrey Learning Disabilities & Dementia Strategy, 2011).

The self-assessment checklist took a similar approach to that in the ‘Green Light toolkit’ (Cole & Gregory, 2004) and *Challenging Behaviour: a unified approach* (Royal College of Psychiatrists et al., 2007). The checklist reflected the content of the report, and translates the guidance into ‘Standards you should see if the recommendations are being met’. However, it differs from previous quality standards in that each standard is written as an outcome rather than an input. The self assessment checklist has been updated and can be found in Appendix 1. Each standard should be rated using a red–amber–green system, with clear descriptors given for each of these for every standard.

## 18.2 Measuring outcomes for people with intellectual disabilities and dementia

Outcome measurement for people with intellectual disabilities and dementia is still in its infancy. Clinicians and professionals working with people with intellectual disabilities are often clear about what works, but this has not resulted in a substantial evidence base.

Specific areas have been looked at for groups of individuals, e.g. effectiveness of staff training, type of living establishment, use of personalised technology.

Within the general population of people with dementia, a range of measures have been developed and evaluated, but many instruments are not sensitive enough to detect change. However, it is clear from the literature that current measures see dementia as a ‘stable’ disease rather than one which is progressive. It seems vital that any measure is sensitive to the progression of the disease and not just for people with early or mid stage dementia.

One of the key components of excellence in dementia care is the ability of the system around the person with dementia to continuously adapt their understanding, care and resources as the person’s dementia progresses. Staff who work in homes and day services for people with intellectual disabilities usually receive training as people are identified as having dementia. This allows them to become proficient at meeting the needs of people at the newly diagnosed/early stage. However, as the dementia progresses they are less able to adapt what they do in line with the person’s changing needs. This often leads to staff feeling that they cannot care for the person with changing needs and that they are always ‘lagging behind’. In turn, this can lead to poorer outcomes for the person with mid or late stage dementia.

Outcome measurement needs to relate to the stages of dementia and how the care for the person is adapted as the disease progresses. It also needs to focus on outcomes for the person and not for processes.

The *Quality Outcome Measure for Individuals with Dementia* (Dodd & Bush, 2013; Dodd et al., 2015) has been developed from the BPS /RCPsych guidance (BPS, 2009). The measure has 17 domains. Each domain has a description of the required quality outcome for each of the three main stages of dementia – suspected/early, mid and late stage. The measure can be found in Appendix 3.

The QOMID should be completed by the professional in discussion with the relevant people for the particular stage of dementia that the person has. Wherever possible, and depending on ability, the person with dementia should be asked how they would rate their experience in each domain. Additional information for the professional to make an inclusive judgement may come from family, support staff, advocates, care managers or anyone else involved with the person and their support, often at a care review meeting.

In supporting the person with dementia, the aim is for them to have high quality outcomes throughout the progression of their dementia. As dementia is a progressive condition, it is vital to ensure that the person’s changing needs are recognised and met. This may mean that scores may fluctuate during the course of the dementia as support ‘catches up’ with the person’s changing needs.

The QOMID is also designed to help everyone involved supporting the person to work with

the person and their carers to both prevent deterioration in quality outcome and to forward plan effective care. For each domain that is scored at less than the maximum, the support team is asked to specify what needs to be put in place to improve the person's quality outcome for that domain. These actions can then be included in the person's support plan. In addition, by looking at the descriptions for the next stage of dementia, the professional can begin to help the person and their supporters to think about what needs to be put in place to maintain the person's quality outcome.

### Case Study

Brian was diagnosed with dementia. The clinical psychologist and occupational therapist met with the staff team and care manager. The QOMID was used to evaluate his quality outcome. The results were used to create a dementia care plan based on the interventions described in this guidance. This provided a clear overview of who was doing what in relation to Brian's support needs, e.g. the occupational therapist to source further equipment, the training that was needed, and the need for the community incontinence service to be involved.

Regular reviews ensured that everyone was aware of any changes in Brian. It was hoped that this would reduce the need for crisis intervention. The home manager reported the reviews as helpful as they often did not notice deterioration until they had space to reflect, as they were with the person daily. It also meant better quality care for Brian as all those involved in his care were accountable for certain outcomes. The home manager felt that they were better supported by the community intellectual disabilities team whereas previously they felt that they had been left to struggle until a crisis occurred.

### Key points

- Each area should use the Self-Assessment Checklist to measure the Outcome of their services for people with intellectual disabilities and dementia, and to assist in the development of a local strategy document.
- Individual quality outcomes for each person with intellectual disabilities and dementia can be measured using the Quality Outcome Measure for Individuals with Dementia (QOMID).

## Section 19 – Future directions and research

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### 19.1 Policy context

Alzheimer's disease and dementia is increasingly becoming a key research priority for many countries, and the aim of the international community is to ensure that it has the same prominence as cancer research. In England, Earl Howe announced in March 2013 the intention of the Government to increase research and innovation in health and social care emphasising the investment in health research through the National Institute for Health Research (NIHR) and the Medical Research Council (MRC). This policy document lists the Government initiatives including the research networks, the NIHR research advisory service and the recently established 15 Academic Health Science Networks for England. Scotland, Wales and Northern Ireland are having their own initiatives. Within this context, the development of new treatments for dementia, and trials to assess such treatments, have a high priority.

Clinical trials are not just about pharmaceutical developments but also about psychological treatments and other approaches, the aim of which might be to maintain function and the dignity of those affected by dementia. This section has therefore been added to the revised guidance largely because we recognise that new treatments for dementia are being, and will be, developed and it is imperative that people with intellectual disabilities, in general, and people with Down's syndrome, specifically, have the opportunity to partake in trials and subsequently benefit from the treatments that are found to be safe and effective.

### 19.2 Advances in understanding

Advances in the understanding of dementia in people with intellectual disabilities, in general, and people with Down's syndrome, in particular has become more possible with improvements in diagnosis and in the means of tracking the cognitive and functional changes associated with dementia over time. Research in the UK and internationally has clearly established that people with Down's syndrome have a high risk for developing the clinical features of dementia from their 30's with a peak incidence in their early 50's. Similarly, people with intellectual disabilities (not due to Down's syndrome) have a slightly earlier age-related prevalence profile of dementia than the typically developing population.

For people with Down's syndrome the focus of research has been on the role of the amyloid precursor protein (APP) gene on chromosome 21 (therefore inherited in triplicate in people with Down's syndrome) leading to the 'amyloid cascade hypothesis' for dementia of the Alzheimer's type in this population. New structural (MRI) and ligand-based (PET) neuroimaging studies provide the means for investigating this relationship and large scale genetic association studies enable the influence of variations at other genetic loci on the course of dementia to be studied.

Particularly in people with Down's syndrome, the longer term objective of research is the development of a preventative treatment. These proposed new treatments will need to be



tested in formal double blind placebo controlled trials. At present treatment developments aimed at preventing dementia in people with Down's syndrome are focused on the modification of beta amyloid production in the brain. For people with intellectual disabilities not due to Down's syndrome advances in treatment are likely to emerge from research in the typically developing population, however, these may need more formal testing (particularly with respect to side effect profiles) in the intellectual disabilities population.

Outside of pharmaceutical developments, which may still be some years away, there has been a lack of research involving people with intellectual disabilities in the environmental and psychological strategies used in the general population to optimise function and maintain dignity and quality of life, such as cognitive stimulation or the Enhancing the Healing Environment (EHE) initiative. Research, whether of a pharmaceutical agent or of some support strategy requires the identification and involvement of people with intellectual disabilities and those who support them.

### **19.3 Clinical trials**

For treatments to be developed that might prevent or treat dementia it is necessary to engage people with dementia in such research and specifically in clinical trials. This is particularly the case for people with Down's syndrome where the risk of developing Alzheimer's disease relatively early in life is high and treatment aimed at prevention is therefore a priority. Such research requires collaboration between people with dementia and their families, clinicians, social care providers and clinical academics and basic scientists. Research of this type has particular challenges if the right balance is to be achieved between enabling research that will lead to new treatments, on the one hand, and, on the other, ensuring the people with intellectual disabilities and dementia are not exposed to excessively intrusive research or to exploitation. It will be clinicians and support workers who are at the forefront when it comes to recruitment to such trials.

### **19.4 The role of clinicians in research**

The following points highlight how clinicians can work in partnership to facilitate such research.

To enable research a major priority is to improve the recruitment of patients into research through building on the NHS Constitution pledge to inform patients about opportunities to participate in research and engaging the NHS in the implementation of RAFT (the Recruitment and Feasibility Tool – a database for patients to register their interest in dementia research). Clinicians and those who support people with intellectual disabilities are the gateway to recruitment and the attitude taken to research by these two groups of people has a powerful impact on whether the potential participant is willing to meet those doing the research. Ultimately it must be for the person with intellectual disabilities to decide or, where he/she lacks the capacity to consent, the protocols and safeguards in the appropriate European and national legislations then apply (e.g. European Clinical Trials Regulation 536 (2014), Mental Capacity Act (2005), Adults with Incapacity [Scotland] Act (2000).

With the advent of electronic health records searches are now possible according to particular diagnostic categories. The identification of potential participants for research has therefore become more feasible. Specialist services for adults with intellectual disabilities should ensure that all people with intellectual disabilities seen and who have received a diagnosis of dementia are identifiable by the service and at the time of diagnosis those concerned and those supporting them are informed about the importance of research, including any trials that are taking place. Specialist services have a responsibility for the identification of people with intellectual disabilities and dementia and to be willing to approach those meeting the necessary criteria for inclusion in a study on behalf of the research group undertaking the study.

In clinical practice different clinicians and services may have their own approaches but for research it is usually necessary to have more formal and time-consuming assessments so that findings can be compared across studies – see section 6 for examples of diagnostic instruments and cognitive and functional assessments. Whilst it is through additional research funding that these more extensive assessments can be undertaken than is possible in clinical practice the use of agreed diagnostic assessments and of standard cognitive assessments would enhance recruitment on a larger scale for studies of, for example, risk and protective factors or for treatment trials. Clinicians working in local specialist services should establish diagnostic and assessment protocols that are agreed and in general use.

## **19.5 Infrastructure and funding**

In England, the NIHR has established a Clinical Research Network (CRN) with regional hubs that support research recruitment and ensure that clinicians and patients from all parts of the country are able to participate in and benefit from research. In particular, the Mental Health Research Network (MHRN) and the Dementia and Neurocognitive Diseases Research Network (DeNDRON) should be able to support clinical trials in adults with Down's syndrome or intellectual disabilities who has dementia. Similar networks have been established in Wales (Clinical Research Collaboration Cymru – CRC Cymru) and in Scotland (the Scottish Clinical Research Network). The Scottish Dementia network (SDCRN) has been very supportive of dementia research in the intellectual disabilities population. Intellectual Disabilities Services clinicians who are interested in research in this population could also join research groups associated with their professional organisations, such as the Dementia in Intellectual Disabilities special interest group (DID-SIG) associated with the Intellectual Disability Faculty of the Royal College of Psychiatrists.

There are no specific funding streams for dementia research in intellectual disabilities, and it may be seen as a 'niche' area for mainstream funders. Funders may therefore benefit from being made aware of the importance of research in this area.

## 19.6 Recommendations

1. Staff in services seeing adults with intellectual disabilities and, specifically adults with Down's syndrome, where the diagnosis of dementia is being considered, should ensure that when a diagnosis of dementia is made the person concerned and those who support them are made aware of research projects being undertaken and permission requested to pass on their details to any approved and relevant research project.
2. Services have the means to retrospectively identify any person with intellectual disabilities diagnosed as having dementia and specifically people with Down's syndrome either in the age at risk for dementia or with a diagnosis of dementia and be willing to be a point of contact with them if approved and appropriate research is being undertaken that is looking for potential participants.
3. In specialist memory clinics and in services for adults with intellectual disabilities the use of standardised diagnostic and neuropsychological assessments for the diagnosis and monitoring of dementia, as it affects people with intellectual disabilities, is encouraged. This will ensure that comparison can be made across services and over time thereby providing consistent and reliable data on prevalence and incidence of dementia in this population and also enabling recruitment into future trials of any new intervention or treatment.

## References

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- Abbey, J., Piller, N., Bellis de, A., Esterman, A., Parker, D., Giles, L. & Lowcay, B. (2004). The Abbey pain scale: a 1-minute numerical indicator for people with end stage dementia. *International Journal of Palliative Nursing*, 10, 6–14.
- Adams, D. & Oliver, C. (2010). The relationship between acquired impairments of executive function and behavior change in adults with Down syndrome. *Journal of Intellectual Disability Research*, 54(5), 393–405.
- Albert, M. & Cohen, C. (1992). The Test for Severe Impairment: an instrument for the assessment of people with severe cognitive dysfunction. *Journal of the American Geriatric Society*, 40, 449–53.
- American Psychiatric Association (2013). *Diagnostic and statistical manual of mental disorders* (5th edn.). Arlington, VA: American Psychiatric Publishing.
- Australian Government (2006). *The National Framework for Action on Dementia 2006–2010*. Retrieved from [www.alz.co.uk/sites/default/files/australia\\_plan.pdf](http://www.alz.co.uk/sites/default/files/australia_plan.pdf).
- Ball, S.L., Holland, A.J., Huppert, F.A., Treppner, P., Watson, P. & Hon, J. (2006). 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*, 21, 661–673.
- Ball, S.L., Holland, A.J., Huppert, F.A., Treppner, P. & Dodd, K. (2006). *CAMDEX-DS: The Cambridge examination for mental disorders of older people with down's syndrome and others with intellectual disabilities*. Cambridge: Cambridge University Press.
- Ball, S.L., Holland, A., Treppner, P., Watson, P.C. & Huppert, F.A. (2008). 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*, 47(1), 1–29.
- Ball, S., Holland, A.J., Watson, P.C., & Huppert, F.A. (2010). Theoretical exploration of the neural bases of behavioural disinhibition, apathy and executive dysfunction in preclinical Alzheimer's disease in people with Down's syndrome: potential involvement of multiple frontal-subcortical neuronal circuits. *Journal of Intellectual Disability Research*, 54(4), 320–336.
- Barrett, E. & Burns, A. (2014). *Dementia revealed. What primary care needs to know. A primer for general practice*. Prepared in partnership by NHS England and Hardwick CCG with the support of the Department of Health and Royal College of General Practitioners.
- Brawley, E.C. (1997). *Designing for Alzheimer's disease*. Chichester: John Wiley.
- Braddock, D. (1999). Aging and developmental disabilities: demographic and policy issues affecting American families. *Mental Retardation*, 37, 155–161.
- British Institute of Learning Disabilities (2014). *Easyread facts sheets on dementia*. Kidderminster: BILD

- British Psychological Society (2004). *Challenging behaviours: Psychological interventions for severely challenging behaviours shown by people with learning disabilities*. Leicester: British Psychological Society.
- British Psychological Society & Royal College of Psychiatrists (2009). *Dementia and people with learning disabilities: Guidance on the assessment, diagnosis, treatment and support of people with learning disabilities who develop dementia*. Leicester: British Psychological Society.
- Brodthmann, A. (2009). Hashimoto encephalopathy and Down syndrome. *Archives of Neurology*, 66(5), 663–666.
- Brooker, D. & Surr, C.A. (2005). *Dementia care mapping: Principles and practice*. Bradford: University of Bradford.
- Brooker, D. (2006). *Person centred dementia care: Making services better*. London: Jessica Kingsley Publishers.
- Buijssen, H. (2005). *The simplicity of dementia*. London: Jessica Kingsley Publishers.
- Bush, A. & Beail, N. (2004). Risk factors for dementia in people with Down syndrome: Issues in assessment and diagnosis. *American Journal on Mental Retardation*, 109, 83–97.
- Cairns, V., Lamb, I. & Smith, E. (2010). 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*, 39(3), 198–208.
- Care Quality Commission (2014). *The state of care report*. Retrieved from [www.cqc.org.uk/content/state-care-201314](http://www.cqc.org.uk/content/state-care-201314).
- Carr, J. (2000). Intellectual and daily living skills of 30-year-olds with Down's syndrome: continuation of a longitudinal study. *Journal of Applied Research in Intellectual Disabilities*, 13, 1–16.
- Cohen, U. & Weisman, G.D. (1990). Environmental design to maximize autonomy for older adults with cognitive impairments. *Generations*, 14, 75–78.
- Cole, A. & Gregory, M. (2004). *Green light for mental health: a service improvement toolkit*. London: Foundation for People with Learning Disabilities.
- Commission for Social Care Inspection (2008). *See me, Not the dementia: understanding people's experiences of living in a care home*. London: CSCI.
- Cooper, S.A. (1997). Epidemiology of psychiatric disorders in elderly compared with younger adults with learning disabilities. *British Journal of Psychiatry*, 170, 375–380.
- Coppus, A., Evenhuis, H., Verberne, G.-J., Visser, F., Van Gool, P., Eikelenboom, P. & Van Duijin, C. (2006). Dementia and mortality in persons with Down's syndrome. *Journal of Intellectual Disability Research*, 50, 768–777.
- Coppus, A.M.W. (2014). The outpatient clinic for adults with Down syndrome; a model to diagnose dementia. In K. Watchman (Ed.) *Intellectual disability and dementia: research into practice*. London: Jessica Kingsley Publishers.

- Crayton, L., Oliver, O., Holland, A., Bradbury, J. & Hall, S. (1998). The neuropsychological assessment of age-related cognitive deficits in adults with Down's syndrome. *Journal of Applied Research in Intellectual Disabilities*, 11, 255–272.
- Cuthill, F.M., Espie, C.A. & Cooper, S.A. (2003). Development and psychometric properties of the Glasgow Depression Scale for people with a learning disability. *British Journal of Psychiatry*, 182, 347–353.
- Deb S., Clarke D. & Unwin G. (2006). *Using medication to manage behaviour problems among adults with a learning disability: Quick reference guide*. University of Birmingham, MENCAP, Royal College of Psychiatrists.
- Deb, S., Hare, M., Prior, L. & Bhaumik, S. (2007). The dementia screening questionnaire for individuals with intellectual disabilities. *British Journal of Psychiatry*, 190, 440–444.
- Delis, D.C., Kaplan, E. & Kramer, J.H. (2001). *Delis-Kaplan Executive Function System*. Oxford: Pearson.
- Department of Health (2001). *Valuing people. A new strategy for learning disability for the 21st Century*. London: TSO (The Stationery Office).
- Department of Health (2008a). *No secrets*. London: TSO (The Stationery Office).
- Department of Health (2008b). *End of life strategy. Promoting high quality care for all adults at the end of life*. London: TSO (The Stationery Office).
- Department of Health (2008c). *Refocusing the care programme approach. Policy and positive practice guidance*. London: TSO (The Stationery Office).
- Department of Health (2009). *Living well with dementia: A national dementia strategy*. London: TSO (The Stationery Office).
- Department of Health (2012a). *The dementia challenge: Fighting back against dementia*. <http://dementiachallenge.dh.gov.uk/about-the-challenge/>
- Department of Health (2012b). *Winterbourne View – Transforming care: A national response to Winterbourne View Hospital*. London: TSO (The Stationery Office).
- Department of Health (2012c). *The National Framework for NHS Continuing Healthcare and NHS-Funded Nursing Care – Revised*. TSO (The Stationery Office).
- Department of Health (2014). *Positive and proactive care: Reducing the need for restrictive interventions*. Retrieved from [www.official-documents.gov.uk](http://www.official-documents.gov.uk).
- Department of Health/Care Services Improvement Partnership (2005). *Everybody's business: Integrated mental health services for older adults: a service development guide*. London: TSO (The Stationery Office).
- Department of Health, Social Services and Public Safety, Northern Ireland (2011). *Improving dementia services in Northern Ireland – A regional strategy*. Retrieved from [www.dhsspsni.gov.uk/improving-dementia-services-in-northern-ireland-a-regional-strategy-november-2011.pdf](http://www.dhsspsni.gov.uk/improving-dementia-services-in-northern-ireland-a-regional-strategy-november-2011.pdf).
- Devapriam, J., Raju, B., Gangadharan, S. & Bhaumik, S. (2008). A comparative study of stress and unmet needs in carers of South Asian and white adults with intellectual disability. *Ethnicity and Inequalities in Health and Social Care*, 1, 35–43.

- Dodd, K. (2008). Transition to old age – what can we do to aid the process? *Advances in Mental Health and Learning Disabilities*, 2, 7–11.
- Dodd, K. (Ed). on behalf of the multidisciplinary team (2012). *Happy eating: Making eating and drinking safe an enjoyable for people with dementia*. Brighton: Pavilion Publishing.
- Dodd, K. (2014). Staff knowledge and training. In K. Watchman (Ed.) *Intellectual disability and dementia: Research into practice*. Jessica Kingsley Publishers. London.
- Dodd, K. & Bush, A. (2013). *Quality Outcome Measure for Individuals with Dementia (QOMID)*. Accessible from [http://dcp-ld.bps.org.uk/dcp-ld/useful-links-and-info/useful-links-and-info\\_home.cfm](http://dcp-ld.bps.org.uk/dcp-ld/useful-links-and-info/useful-links-and-info_home.cfm).
- Dodd, K., Bush, A. & Livesey, A. (2015). *Developing and piloting the QOMID – Quality Outcome Measure for Individuals with Intellectual Disabilities and Dementia*. *Advances in Mental Health and Intellectual Disabilities*. Accepted for publication.
- Dodd, K., Turk, V. & Christmas, M. (2009). *Resource pack for carers of adults with Down's syndrome and dementia (2nd edn)*. Birmingham: BILD Publications.
- Dodd, K., Kerr, D. & Fern, S. (2006). *Down's syndrome and dementia workbook for staff*. London: Down's Syndrome Association.
- Dodd, K., Turk, V. & Christmas, M. (2005a). *The journey of life: How people change from babies to older people*. Birmingham: BILD Publications.
- Dodd, K., Turk, V. & Christmas, M. (2005b). *About my friend: For friends of people with Down's syndrome and dementia*. Birmingham: BILD Publications.
- Dodd, K., Turk, V. & Christmas, M. (2005c). *About dementia – for people with learning disabilities*. Birmingham: BILD Publications.
- Downs, M., Small, N. & Froggatt, K. (2006). Explanatory models of dementia: links to end-of-life care. *International Journal of Palliative Nursing*, 12, 209–213.
- European Union (2014). *European clinical trials regulation*. Retrieved from [http://ec.europa.eu/health/files/eudralex/vol-1/reg\\_2014\\_536/reg\\_2014\\_536\\_en.pdf](http://ec.europa.eu/health/files/eudralex/vol-1/reg_2014_536/reg_2014_536_en.pdf)
- Evenhuis, H.M. (1992). Evaluation of a screening instrument for dementia in ageing mentally retarded persons. *Journal of Intellectual Disability Research*, 36, 337–47.
- Evenhuis, H.M., Kengen, M.M.F. & Eurlings, H.A.L. (2007). *Dementia questionnaire for people with learning disabilities (DLD). UK adaptation*. Antonio, TX: Harcourt Assessment.
- Fisher, A.G. (2006). *Assessment of motor and process skills. Vol. 2: User manual (6th edn.)*. Fort Collins, CO: Three Star Press.
- Folstein, M. & Folstein, S. (2001). *The mini mental state examination (MMSE): a guide for people with dementia and their carers*. Lutz, FL: Psychological Assessment Resources.
- Francis, R. (2013). *Report of the Mid Staffordshire NHS Foundation Trust Public Inquiry*. London. TSO (The Stationery Office).
- Fraser, J. & Mitchell, A. (1876). Kalmuc idiocy: report of a case with autopsy with notes on 62 cases. *Journal of Mental Science*, 22, 161.

- Gedye, A. (1995). *Dementia scale for Down syndrome – Manual*. Vancouver, BC: Gedye Research & Consulting.
- Gibson, F. (1994). *Reminiscence and recall: A guide to good practice*. London: Age Concern.
- Gore, N.J., McGill, P., Toogood, S., Allen, D., Hughes, J.C., Baker, P., Hastings, R., Noone, S.J. & Denne, L.D. (2013). Definition and scope for positive behavioural support. *International Journal of Positive Behavioural Support*, 3(2), 14–23.
- Hampshire Social Services (1987). *Hampshire Social Services – Staff Support Levels Assessment*. Winchester: Hampshire Social Services.
- Hanney, M., Prasher, V., Williams, N., Jones, E. L., Aarsland, D., Corbett, A. & Ballard, C. (2012). Memantine for dementia in adults older than 40 years with Down’s syndrome (MEADOWS): a randomised, double-blind, placebo-controlled trial. *The Lancet*, 379(9815), 528–536.
- Harrison, P.L. & Oakland, T. (2003). *Adaptive Behaviour Assessment System (2nd edn.) (ABAS-II)*. New York: The Psychological Corporation.
- Hartley, S.L., Handen, B.L., Devenny, D.A., Hardison, R., Mihaila, I., Price, J.C. & Christian, B.T. (2014). Cognitive functioning in relation to brain amyloid- $\beta$  in healthy adults with Down syndrome. *Brain*, 137(9), 2556–2563.
- Hatton, C., Akram, Y., Shah, R., Robertson, J. & Emerson, E. (2002). *Supporting South Asian families with a child with severe disabilities: A report to the Department of Health*. Lancaster: Institute for Health Research, Lancaster University.
- Hatton, C., Azmi, S., Caine, A. & Emerson, E. (1998). Informal carers of adolescents and adults with learning difficulties from the South Asian communities: Family circumstances, service support and carer stress. *British Journal of Social Work*, 28, 821–837.
- Hewitt, K.E., Carter, G. & Jancer, J. (1985). Ageing in Down’s syndrome. *British Journal of Psychiatry*, 147, 58–62.
- Hobson, B., Webb, D., Sprague, L., Grizzell, M., Hawkins, C. & Benbow, S.M. (2012). 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.
- Hodges, J. (2012). *Addenbrooke’s Cognitive Examination-III (ACE-III)*. Retrieved from [www.neura.edu.au/frontier/research/test-downloads](http://www.neura.edu.au/frontier/research/test-downloads).
- Hofman, A., Rocca, W.A.M., Brayne, C., Breteler, M., Clarke, M. & Cooper, B. (1991). The prevalence of dementia in Europe: a collaborative study of 1980–1990 findings. *International Journal of Epidemiology*, 20, 736–48.
- Holland, A.J. (2013). *Down’s syndrome and Alzheimer’s disease: A guide for parents and carers*. London: Down’s Syndrome Association.
- Holland, A.J., Hon, J., Huppert, F.A., Stevens, F. & Watson, P. (1998). Population-based study of the prevalence and presentation of dementia in adults with Down’s syndrome. *British Journal of Psychiatry*, 172, 493–8.



- Hollins, S., Blackman, N. & Ely, R., (2012). *Ann has dementia*. London: Books Beyond Words.
- Iliffe, S. & Manthorpe, J. (2004). The debate on ethnicity and dementia: from category fallacy to person-centred care. *Aging and Mental Health*, 8, 283–292.
- Janicki, M. & Dalton, A. (Eds.) (1998). *Dementia, aging and intellectual disabilities*. Philadelphia, PA: Brunner/Mazel.
- Janicki, M.P. & Dalton, A.J. (2000). Prevalence of dementia and impact on intellectual disability services. *Mental Retardation*, 38, 276–88.
- Jap, S.N. & Ghaziuddin, N. (2011). Catatonia among adolescents with Down syndrome: a review and 2 case reports. *Journal of ECT*, 27(4), 334–337.
- Jervis, N. & Prinsloo, L. (2007). 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*, 36(1), 13–21.
- Jethwa, H. & Cassidy, G. (2010). Difficulties of dealing with dementia in individuals with intellectual disabilities: the healthcare perspective. *Advances in Mental Health and Intellectual Disabilities*, 4(4), 48–52.
- Johns, M.W. (1991). A new method for measuring daytime sleepiness: The Epworth Sleepiness Scale. *Sleep*, 14, 540–5.
- Kalsy-Lillico, S. (2014). Living life with dementia: Enhancing psychological wellbeing. In K. Watchman (Ed.) *Intellectual disability and dementia: Research into practice*. London: Jessica Kingsley Publishers.
- Kalsy-Lillico, S, Adams, D. & Oliver, C. (2012). Older adults with intellectual disabilities: Issues in ageing and dementia. In E. Emerson, C. Hatton, K. Dickson, R. Gone, A. Caine & J. Bromley (Eds.) *Clinical psychology and people with intellectual disabilities (2nd edn.)*. Chichester: Wiley.
- Kerr, D., Cunningham, C. & Wilkinson, H. (2006). *Responding to the pain needs of people with a learning disability*. York: York Publishing Services.
- Kerr, D. (1997). *Down's syndrome and dementia: a practitioner's guide*. Birmingham: Venture Press.
- Kerr, D. (2007). *Understanding learning disability and dementia: Developing effective interventions*. London: Jessica Kingsley Publishers.
- Kerr, D. & Innes, M. (2000). *What is dementia? A booklet for adults with a learning disability*. Edinburgh: Scottish Down's Syndrome Association.
- Kishnani, P.S., Heller, J.H., Spiridigliozzi, G.A., Lott, I., Escobar, L., Richardson, S., Zhang, R. & McRae, T. (2010). Donepezil for treatment of cognitive dysfunction in children with Down syndrome aged 10–17. *American Journal of Medical Genetics Part A*, 152(12), 3028–3035.
- Kitwood, T (1997). *Dementia reconsidered: The person comes first*. Milton Keynes: Open University Press

- Lai, F. & Williams, R.S. (1989). A prospective study of Alzheimer disease in Down syndrome. *Archives of Neurology*, 46, 849–53.
- Landt, J., D’Abrera, J.C., Holland, A.J., Aigbirhio, F.I., Fryer, T.D., Canales, R., Hong, Y.T., Mendon, D.K., Baron, J.C. & Zaman, S.H. (2011). Using positron emission tomography and Carbon 11–Labeled Pittsburgh compound B to image brain fibrillar  $\beta$ -Amyloid in adults with Down syndrome: Safety, acceptability, and feasibility. *Archives of Neurology*, 68(7), 890–896.
- Leadership Alliance for the Care of Dying People (2014). *One chance to get it right: Improving people’s experience of care in the last few days and hours of life*. London: LACDP.
- Learning Disabilities Observatory, RCGP, Public Health England. (2013). *Improving the health and wellbeing of people with learning disabilities: An evidence-based commissioning guide for clinical commissioning groups (CCGs)*. Retrieved 24 February 2015 from <http://www.improvinghealthandlives.org.uk/publications>.
- Lott, I.T., Osann, K., Doran, E. & Nelson, L. (2002). Down syndrome and Alzheimer disease: response to donepezil. *Archives of Neurology*, 59, 1133–6.
- Lott, I.T., Doran, E., Nguyen, V.Q., Tournay, A., Head, E. & Gillen, D.L. (2011). Down syndrome and dementia: a randomized, controlled trial of antioxidant supplementation. *American Journal of Medical Genetics Part A*, 155(8), 1939–1948.
- Lyketsos, C.G., DelCampo, L., Steinberg, M., Miles, Q., Steele, C.D., Munro, C., Baker, A.S., Sheppard, J.E., Frangakis, C., Brandt, J. & Rabins, P.V. (2003). Treating depression in Alzheimer disease: efficacy and safety of sertraline therapy, and the benefits of depression reduction: the DIADS. *Archives of General Psychiatry*, 60, 737–746.
- Lyngaard, H. & Alexander, N. (2004). ‘Why are my friends changing?’ Explaining dementia to people with learning disabilities. *British Journal of Learning Disabilities*, 32, 30–34.
- Major, B. & McBrien, J. (2011). The value of prospective screening for dementia in adults with Down’s syndrome. *Clinical Psychology Forum*, 224, 44–48.
- Mann, D. (1988). The pathological association between Down syndrome and Alzheimer disease. *Mechanisms of ageing and development*, 43(2), 99–136.
- Marshall, M. (1998). How it helps to see dementia as a disability. *Journal of Dementia Care*, 6(1), 15–17.
- Matthews, D.R. (2006). *The ‘OK’ health check*. Preston: Fairfield Publications.
- McBrien, J.A. (2003). Assessment and diagnosis of depression in adults with intellectual disabilities: a review. *Journal of Intellectual Disability Research*, 47, 1–13.
- McBrien, J. (2009). Screening adults with Down’s syndrome for early signs of dementia. *Journal of Integrated Care*, 17(3), 3–7
- McBrien, J.A., Whitwham, S., Olverman, K. & Masters, S. (2005). Screening adults with Down’s Syndrome for early signs of Alzheimer’s disease. *Tizard Learning Disability Review*, 10, 23–32.

- McCarron, M., Gill, M., Lawlor, B. & Begley, C. (2002). A pilot study of the reliability and validity of the Caregiver Activity Survey – Intellectual Disability (CAS-ID). *Journal of Intellectual Disability Research*, 46, 605–13.
- McCarron, M., McCallion, P., Reilly, E. & Mulryan, N. (2014). A prospective 14-year longitudinal follow-up of dementia in persons with Down syndrome. *Journal of Intellectual Disability Research*, 58(1), 61–70.
- McConkey, R., Mulvany, F. & Barron, S. (2006). Adult persons with intellectual disabilities on the island of Ireland. *Journal of Intellectual Disability Research*, 50, 227–236.
- McGrother, C.W., Bhaumik, S., Thorp, C.F., Watson, J.M. & Taub, N.A. (2002). Prevalence, morbidity and service need among South Asian and white adults with intellectual disability in Leicestershire, UK. *Journal of Intellectual Disability Research*, 46, 299–309.
- Mencap (2014). *Making a will*. Accessed at <https://www.mencap.org.uk/our-services/wills-and-trusts/guide-wills-and-trusts>
- Menendez, M. (2005). Down syndrome, Alzheimer’s disease and seizures. *Brain and Development*, 27(4). 246–252.
- Mindham, J. & Espie, C.A. (2003). Glasgow Anxiety Scale for people with an intellectual disability (GAS-ID): development and psychometric properties of a new measure for use with people with mild intellectual disability. *Journal of Intellectual Disability Research*, 47(1), 22–30.
- Ministry of Health (2004). *National Dementia Plan*. Netherlands.
- Moran, J.A., Rafii, M.S., Keller, S.M., Singh, B.K. & Janicki, M.P. (2013). 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 Clin Proceedings*, 88(8), 831–840.
- Morgan, D.G. & Stewart, N.J. (1999). The physical environment of special care units: Needs of residents with dementia from the perspective of staff and family. *Qualitative Health Research*, 9(1), 105–118.
- Moss, S. (2002a). *The PAS-ADD checklist*. Brighton: Pavilion Publishing.
- Moss, S. (2002b). *The Mini-PAS-ADD interview*. Brighton: Pavilion Publishing.
- Moss, S. & Patel, P. (1995). Psychiatric symptoms associated with dementia in older people with learning disability. *British Journal of Psychiatry*, 163, 471–80.
- National Audit Office (2007). *Improving services and support for people with dementia*. London: TSO.
- National Institute of Health and Care Excellence (2006). *Dementia: Supporting people with dementia and their carers in health and social care. Clinical guidelines, CG42*. Retrieved from [www.nice.org.uk/CG42](http://www.nice.org.uk/CG42).
- National Institute of Health and Care Excellence (2009). *Medicines adherence: Involving patients in decisions about prescribed medicines and supporting adherence. Clinical guidelines CG 76*. Retrieved from [www.nice.org.uk/guidance/cg76](http://www.nice.org.uk/guidance/cg76).

- National Institute for Health and Clinical Excellence (2010). *Dementia Quality Standard 1*. Accessed at <http://www.nice.org.uk/guidance/qs1/chapter/list-of-statements>
- National Institute for Health and Clinical Excellence (2012a). *Quality Standard for End of Life Care. Quality Standard 13*. Accessed at <http://www.nice.org.uk/guidance/QS13>
- National Institute for Health and Clinical Excellence (2012b). *Dementia: Supporting people with dementia and their carers in health and social care. Clinical guideline CG42*. Accessed at <http://www.nice.org.uk/guidance/CG42>
- National Institute for Health and Clinical Excellence (2013). *QS30 Supporting People to Live well with dementia*. Retrieved from <https://www.nice.org.uk/guidance/qs30>
- NHS National End of Life Care Programme (2011). *The route to success in end of life care – Achieving quality for people with intellectual disabilities*. Retrieved from [www.endoflifecareforadults.nhs.uk](http://www.endoflifecareforadults.nhs.uk)
- Nihira, K., Foster, R., Shellhaas, M. & Leyland, H.. (1974). *AAMD Adaptive Behaviour Scale 1974 Revision*. Washington, DC: American Association on Mental Deficiency.
- Nolan, B., Mathews, R.M., Truesdell-Todd, G. & VanDorp, A. (2002). Evaluation of the effect of orientation cues on wayfinding in persons with dementia. *Alzheimer's Care Quarterly*, 3, 46–49.
- Nyth, A. L. & Gottfries, C G. (1990). The clinical efficacy of citalopram in treatment of emotional disturbances in dementia disorders in Nordic multicentric study. *British Journal of Psychiatry*, 157, 894–901.
- O’Caoimh, R., Clune, Y. & Molloy, D.W. (2013). Screening for Alzheimer’s disease in Downs syndrome. *Journal of Alzheimers Disease and Parkinsonism*, <http://dx.doi.org/10.4172/2161-0460.s7-001>
- Oliver, C., Kalsy, S., McQuillan, S. & Hall, S. (2011). Behavioural excesses and deficits associated with dementia in adults who have Down syndrome. *Journal of Applied Research in Intellectual Disabilities*, 24(3), 208–216.
- Pollock, B.G., Mulsant, B.H., Rosen J, Sweet, R.A., Mazumdar, S., Bharucha, A., Marin, R., Jacob, N.J., Huber, K.A., Kastango, K.B. & Chew, M.L. (2002). Comparison of citalopram, perphenazine, and placebo for the acute treatment of psychosis and behavioral disturbances in hospitalized, demented patients. *American Journal of Psychiatry*. 159, 460–465.
- Prasher, V.P. (1997). Dementia questionnaire for persons with mental retardation (DMR). Modified criteria for adults with Down syndrome. *Journal of Applied Research in Intellectual Disabilities*, 10, 54–60.
- Prasher, V.P., Farrer, M.J., Kessling, A.M., Fisher, E.M.C., West, R.J., Barber, P.C. & Butler, A.C. (1998). Molecular mapping of Alzheimer-type dementia in Down’s syndrome. *Annals of Neurology*, 43, 380–383.
- Prasher, V.P., Huxley, A. & Haque, M.S. (2002). A 24-week, double-blind, placebo-controlled trial of donepezil in patients with Down syndrome and Alzheimer’s disease – Pilot study. *International Journal of Geriatric Psychiatry*, 17, 270–8.

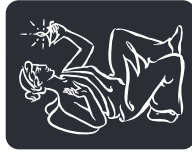
- Prasher, V.P., Adams, C. & Holder, R. (2003). Long-term safety and efficacy of donepezil in the treatment of dementia in Alzheimer's disease in adults with Down syndrome: open-label study. *International Journal of Geriatric Psychiatry*, 8, 549–51.
- Prasher, V.P., Holder, R. & Asim, F. (2004). The Adaptive Behaviour Dementia Questionnaire (ABDQ): Screening questionnaire for dementia in Alzheimer's disease in adults with Down syndrome. *Research in Developmental Disabilities*, 25, 385–97.
- Prasher, V.P., Fung, N. & Adams, C. (2005). Rivastigmine in the treatment of dementia in Alzheimer's disease in adults with Down syndrome. *International Journal of Geriatric Psychiatry*, 20, 496–7.
- Prasher, V.P., Sachdeva, N., Adams, C. & Haque, M.S. (2013). Rivastigmine transdermal patches in the treatment of dementia in Alzheimer's disease in adults with Down syndrome – pilot study. *International Journal of Geriatric Psychiatry*, 28(2), 219–220.
- Regnard, C., Reynolds, J., Watson, B., Matthews, D., Gibson, L. & Clarke, C. (2007). Understanding distress in people with severe communication difficulties: developing and assessing the Disability Distress Assessment Tool (DisDAT). *Journal of Intellectual Disability Research*, 51, 277–292.
- Roth, R.M., Isquith, P.K. & Gioia, G.A. (2000). Behaviour Rating Inventory of Executive Function - Adult version (BRIEF-A). Hogrefe.
- Royal College of Psychiatrists, British Psychological Society & Royal College of Speech and Language Therapists (2007). *Challenging behaviour: A unified approach*. London: Royal College of Psychiatrists.
- Sampson, E.L., Candy, B. & Jones, L. (2009). Enteral tube feeding for older people with advanced dementia. *Cochrane Database of Systematic Reviews*, 2. art. no.: CD007209. DOI: 10.1002/14651858.CD007209.pub2.
- Saxton, J., McGonigle, K.L., Swihart, A.A. & Boller, F. (1993). *The severe impairment battery*. Oxford: Pearson.
- Scottish Government (2013). *Scotland's National Dementia Strategy 2013–2016*. Retrieved from <http://www.scotland.gov.uk/Resource/0042/00423472.pdf>.
- Sharp, S. (2007). *Home from home*. London: Alzheimer's Society.
- Sheard, D.M. (2008). *Feelings matter most*. Brighton: Dementia Care Matters.
- Sheard, D.M. (2013). The feeling of 'mattering': the positioning of emotions in dementia care. *Journal of Dementia Care*, 21, 23–27.
- Sheehan, R., Sinai, A., Bass, N., Blatchford, P., Bohnen, I., Bonell, S. & Strydom, A. (2014). Dementia diagnostic criteria in Down syndrome. *International Journal of Geriatric Psychiatry*. doi: 10.1002/gps.4228.
- Sparrow, S.S., Cicchetti, D.V. & Balla, D.A. (2007). *Vineland-II: Vineland Adaptive Behavior Scales (2nd edn.)*. Livonia, MN: Pearson.
- Struwe, F. (1929). Histopathologische Untersuchungen über Entstehung und Wesen der senilen plaques. *Zeitschrift für die gesamte Neurologie und Psychiatrie*, 122, 291–307.
- Strydom, A. (2014). *Report of the ADSID research group*. Personal communication.

- Strydom, A., Livingston, G., King, M. & Hassiotis, A. (2007). Prevalence of dementia in intellectual ability using different diagnostic criteria. *British Journal of Psychiatry*, 191, 150–157.
- Strydom, A., Lee, L.A., Jokinen, N., Shooshtari, S., Rayka, V., Tor, J., Tsiouris, J.A., Courtenay, K., Bass, N., Sinnema, M., Maaskant, M.A. (2009). *Report on the state of science on dementia in people with intellectual disabilities*. IASSID Special Interest Research Group on Ageing and Intellectual Disabilities. Retrieved 24 February 2015 from <https://www.iassidd.org/uploads/legacy/images/documents/Aging/state%20of%20the%20science%20dementia%202009.pdf>.
- Strydom, A., Shooshtari, S., Lee, L., Raykar, V., Torr, J., Tsiouris, J. & Maaskant, M. (2010). Dementia in older adults with intellectual disabilities—epidemiology, presentation, and diagnosis. *Journal of Policy and Practice in Intellectual Disabilities*, 7(2), 96–110.
- Strydom, A., Chan, T., Fenton, C., Jamieson-Craig, R., Livingston, G. & Hassiotis, A. (2013). Validity of criteria for dementia in older people with intellectual disability. *American Journal of Geriatric Psychiatry*, 21(3), 279–288.
- Sultzer, D.L., Gray, K.F., Gunay, I., Wheatley, M.V. & Mahler, M.E. (2002). Does behavioral improvement with Haloperidol or Trazodone treatment depend on psychosis or mood symptoms in patients with dementia? *Journal of American Geriatric Society*, 49, 1294–1300.
- Surrey County Council (2011). *Surrey Learning Disabilities & Dementia Strategy*. [www.surreycc.gov.uk](http://www.surreycc.gov.uk)
- Thomas, K. & Department of Health (2005). *Gold standards framework*. London: Department of Health.
- Thompson, D. & Wright, S. (2001). *Misplaced and forgotten: People with learning disabilities in residential services for older people*. London: Foundation for People with Learning Disabilities.
- Tuffrey-Wijne, I. & Watchman, K. (2014). Sharing the diagnosis of dementia: Breaking bad news to people with an intellectual disability. In K. Watchman (Ed.) *Intellectual disability and dementia: Research into practice*, chapter 10. London: Jessica Kingsley Publishers
- Turk, V., Dodd, K. & Christmas, M. (2001). *Down's syndrome and dementia: Briefing for Commissioners*. London: Foundation for People with Learning Disabilities.
- Tyrrell, J., Cosgrave, M., McCarron, M., McPherson, J., Calvert, J., Kelly, A., McLaughlin, M., Gill, M. & Lawlor, B.A. (2001). Dementia in people with Down's syndrome. *International Journal of Geriatric Psychiatry*, 16(12), 1168–1174.
- Tyrer, P., Oliver-Africano, P.C., Ahmed, Z., Bouras, N., Cooray, S., Deb, S., Murphy, D., Hare, M., Meade, M., Reece, B., Kramo, K., Bhaumik, S., Harley, D., Regan A., Thomas, D., Rao, B., North, B., Eliahoo, J., Karatela, S., Soni, A. & Crawford, M. (2008). Risperidone, haloperidol, and placebo in the treatment of aggressive challenging behaviour in patients with intellectual disability: a randomised controlled trial. *The Lancet*, 371, 57–63.

- United Nations (2008). *The United Nations convention on the rights of people with disabilities*. Retrieved from [www.un.org/disabilities/convention/conventionfull.shtml](http://www.un.org/disabilities/convention/conventionfull.shtml).
- Visser, F.E., Aldenkamp, A.P., Van Huffelen, A.C. & Kuilman, M. (1997). Prospective study of the prevalence of Alzheimer-type dementia in institutionalized individuals with Down syndrome. *American Journal on Mental Retardation*, 101(4), 412.
- Watchman, K. (2006). *Keep talking about dementia: Information for siblings and professionals about Down's syndrome and dementia*. Edinburgh: Scottish Down's Syndrome Association.
- Watchman, K., Kerr, D. & Wilkinson, D. (2010). *Supporting Derek*. York: Joseph Rowntree Foundation.
- Watchman, K. (2012). People with a learning disability and dementia: reducing marginalization. *Journal of Dementia Care*, 20(5), 35–39.
- Watchman, K. (2014). Towards understanding individual experiences of people ageing with an intellectual disability and dementia In K. Watchman (Ed.) *Intellectual disability and dementia: Research into practice*. London: Jessica Kingsley Publishers.
- Webb, Z. & Dodd, K. (2014). *BADS-ID*. Presentation at DCP ID Faculty Advancing Practice Conference, Llandudno. 27 March 2014.
- Webb, Z., Dodd, K., Joseph, T., Marlow, E. & Thomas, H. (2014). *Measure of everyday planning*. Surrey and Borders Partnership NHS Trust.
- Welsh Assembly Government (2011). *National dementia vision for Wales*. Retrieved from <http://wales.gov.uk/docs/dhss/publications/110302dementiaen.pdf>.
- Whitwham, S., McBrien, J. & Broom, W. (2011). 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*, 39(1), 17–21.
- Wilkinson, H., Kerr, D., Cunningham, C. & Rae, C. (2004). *'Home for good?' Models of good practice for supporting people with a learning disability and dementia*. Brighton: Pavilion.
- Wilson, B., Emslie, H., Evans, J., Alderman, N. & Bridges, P. (1996). *Behavioural assessment of the dysexecutive syndrome*. Oxford: Pearson.
- Wisniewski, K.E., Wisniewski, H.M. & Wen, G.Y. (1985). Occurrence of neuropathological changes and dementia of Alzheimer's disease in Down syndrome. *Annals of Neurology*, 17, 278–82.
- World Health Organisation (1992). *International classification of diseases (ICD). 10*. Geneva: World Health Organisation.
- Worley, G., Crissman, B.G., Cadogan, E., Milleson, C., Adkins, D.W. & Kishnani, P.S. (2014). Down syndrome disintegrative disorder new-onset autistic regression, dementia, and insomnia in older children and adolescents with Down syndrome. *Journal of Child Neurology*, 0883073814554654.
- Wu, J. & Morris, J.K. (2013). The population prevalence of Down's syndrome in England and Wales in 2011. *European Journal of Human Genetics*, 21(9), 1016–1019.

## Appendix 1

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The British  
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**The British Psychological Society**

**The Royal College of Psychiatrists**

### **Dementia and People with Intellectual Disabilities:**

**Guidance on the assessment, diagnosis, treatment and support of  
people with intellectual disabilities who develop dementia**

### **Good Practice Standards – Self Assessment checklist**



This framework is designed to be used by members of Partnership Boards or their equivalent, and senior professionals (clinicians, social workers, managers, commissioners and those responsible for inspection and review) who have responsibility within a defined area or population for the provision of services to people with intellectual disabilities who develop or at risk of developing dementia. It is a self-assessment checklist that should be used to establish the extent to which local processes and practices reflect 'best practice' as described in more detail elsewhere in this document.

The current provision and practices that are followed by services should be reviewed against the standards that are described. Users, advocates and carers should also be involved in the review process. The standards apply to people living in family homes, health and social care provision (within the statutory and independent sectors). They also apply when out of area placements have been purchased by the relevant authorities.

The checklist should be:

- Reviewed in a multi-disciplinary / multi-agency setting, with the aim of achieving a consensus view about how local services compare against the Good Practice Standards.
- The team that carries out the review of standards should ensure that they have the appropriate membership to achieve a broad view of services.
- The team should include: commissioners, practitioners from the Community Intellectual Disabilities Team, practitioners from the relevant Older Adults Mental Health Service, Social Services care managers, residential providers, day service providers, inspection teams, people with intellectual disabilities and carers.
- The reviewing team should refer to the relevant section in the guidance document to clarify the detail required to show compliance with the standard.
- Evidence for compliance with each standard should be demonstrated.
- A joint action plan should be developed to address any areas of need, or to build on current good practice.
- The action plan should be reviewed regularly to update on progress.

1. Legal framework and guidance				
Standard	Green Amber Red	Action required	By whom	By when
<p>1. People who develop, or are at risk of developing dementia have access to assessments and interventions that are delivered within current legislation and national guidance.</p> <p>Refer to Section 1.</p>				
<p><b>Green:</b> People who develop, or are at risk of developing dementia have services delivered within current legal and best practice requirements. These will include:</p> <ul style="list-style-type: none"> <li>• Clear health and social care pathways that incorporate assessment of capacity, in line with the Mental Capacity Act.</li> <li>• The presence of a local advisory group with clear terms of reference, to consider issues of best interest for people who lack capacity.</li> <li>• Local care pathways are in place to ensure compliance with national clinical guidelines on dementia and their National Dementia Strategy.</li> <li>• All people with intellectual disabilities who develop (or are at risk of developing) dementia have a person-centred plan.</li> <li>• Annual Health Checks and Health action plans are offered to all people with intellectual disabilities who are at risk of developing dementia.</li> <li>• People with intellectual disabilities who are at risk of developing dementia have the opportunity to plan their future care and have considered advance decisions and end of life planning as part of their care package.</li> <li>• There is a published local Safeguarding Adults policy that people with intellectual disabilities, clinicians and carers can access.</li> <li>• Where people have 'out of area' placements purchased for them, the purchasing authority regularly checks that the provider also achieves these standards.</li> </ul>				
<p><b>Amber:</b> There are a few elements that require action in order to meet national guidance and to comply with current legislation.</p>				
<p><b>Red:</b> There are significant gaps between national guidance/ legislation and local practice.</p>				

2. Population				
Standard	Green Amber Red	Action required	By whom	By when
2. Each area has a register / database of people with intellectual disabilities, which identifies people with Down's Syndrome that can be used to plan and deliver effective services. Refer to Section 2.				
<p><b>Green:</b> There is a regularly updated register / database of all adults with intellectual disabilities known to both health and social care in the catchment area. This includes the identification of people with Down's syndrome, and people who are the responsibility of the area but have services purchased for them in other areas.</p> <p><b>Amber:</b> There is only a partial database, or it is not regularly updated, or it does not identify people with Down's syndrome, or it only identifies people with Down's syndrome, or excludes people who are placed by the authority in out if area accommodation.</p> <p><b>Red:</b> There is no register / database or it is not regularly updated.</p>				

4. Care Pathway			
Standard	Green Amber Red	Action required	By whom By when
<p>4. People who develop, or are at risk of developing dementia have assessment, diagnosis, interventions and support delivered according to an agreed multi-agency care pathway which includes access to other services beyond the specialist intellectual disability areas as applicable (e.g. Older People's Mental Health Services, neurology, advocacy). Refer to Section 16.</p>			
<p><b>Green:</b> There is a multi-agency care pathway for assessment, diagnosis, interventions and support of people who develop, or are at risk of developing dementia, which has been agreed by the Partnership Board or its equivalent.</p>			
<p><b>Amber:</b> There is a single service care pathway, or only a partial care pathway or it is not fully agreed.</p>			
<p><b>Red:</b> No care pathway has been agreed.</p>			

5. Multidisciplinary approach to assessment, diagnosis and support				
Standard	Green Amber Red	Action required	By whom	By when
<p>5. People who develop, or are at risk of developing dementia are offered assessments, diagnosis and specialist support from the full range of dedicated and skilled professionals within intellectual disabilities services. They have easy access to specialist assessments within the general health services (e.g. neurology)</p> <p>Refer to Section 16.</p>				
<p><b>Green:</b> There is a full range of suitably trained professionals within the intellectual disability specialist services that fully matches the care pathway. They have easy access to specialist input from mainstream services such as neurology.</p>				
<p><b>Amber:</b> The workforce only partially matches the care pathway.</p>				
<p><b>Red:</b> There are significant gaps in staffing to deliver the care pathway.</p>				

6. Assessment and diagnosis			
Standard	Green Amber Red	Action required	By whom By when
6. People who develop, or are at risk of developing dementia have easy access to comprehensive assessment and diagnostic services according to an agreed protocol.  Refer to Sections 3 – 7.			
<p><b>Green:</b> Within each area, people who develop, or are at risk of develop dementia have easy access to comprehensive assessment and diagnostic services where:</p> <ul style="list-style-type: none"> <li>• Assessments are undertaken according to an agreed assessment protocol that includes decisions on whether the service offers baselines, reactive screening and / or prospective screening.</li> <li>• Assessments follow an agreed assessment battery including both direct assessment of the person with intellectual disabilities and informant assessment.</li> <li>• Assessments include physical, psychological, social and environmental factors.</li> <li>• There is a timely response to referrals.</li> <li>• Diagnosis is multi-disciplinary and based on established diagnostic criteria.</li> <li>• There is a written formulation.</li> <li>• There is a protocol for how to share the diagnosis of dementia both with people with intellectual disabilities and carers.</li> <li>• Other illnesses / conditions are identified and treated promptly.</li> </ul>			
<b>Amber:</b> There are a few gaps that require action in order to achieve a comprehensive assessment and diagnosis service.			
<b>Red:</b> There are significant gaps between the best practice standards for assessment and diagnosis, and the current local procedures.			

7. Person Centred Dementia Care				
Standard	Green Amber Red	Action required	By whom	By when
7. People who develop, or are at risk of developing dementia have their care provided according to person centred principles and this is individualised to meet the needs of the person.  Refer to Section 10.				
<p><b>Green:</b> People who develop, or are at risk of developing dementia have:</p> <ul style="list-style-type: none"> <li>• A person centred plan that has been drawn up with the person to include future needs.</li> <li>• Dementia care provided according to an agreed philosophy of care that places the person at the centre of the process.</li> <li>• Care provided following the principles of the Mental Capacity Act and least restrictive practices.</li> <li>• Care able to meet their changing needs without risk or delay.</li> <li>• Care provided that promotes social inclusion.</li> </ul>				
<p><b>Amber:</b> Most people will receive care that meets this standard, but there are some gaps for a few people.</p>				
<p><b>Red:</b> Fewer than half of the people who have dementia or are at risk of developing dementia receive care that meets this standard.</p>				

8. Care management and review			
Standard	Green Amber Red	Action required	By whom By when
8. People who develop, or are at risk of developing dementia have their care purchased, monitored and reviewed by an effective care management system, whether in the geographical area or in an out of area placement. Refer to Section 16.			
<p><b>Green:</b> There is a clear inter-agency care co-ordination system that ensures that all people with intellectual disabilities and dementia have:</p> <ul style="list-style-type: none"> <li>• A named care manager.</li> <li>• A written multi-disciplinary care plan incorporating the person's person centred plan.</li> <li>• Regular care reviews.</li> <li>• A health action plan that identifies pain signals and management.</li> <li>• Proactive risk assessment and management plans in place.</li> <li>• Access to flexible funding to meet changing needs without delay.</li> <li>• Outcomes monitored regarding their quality of life.</li> <li>• Staffing levels increased as required including the provision of waking night staff.</li> <li>• Care that recognises that people with intellectual disabilities and dementia should not be moved unless absolutely necessary.</li> <li>• No multiple moves.</li> </ul>			
<p><b>Amber:</b> Most elements of a comprehensive care management system are in place but there are a few gaps.</p>			
<p><b>Red:</b> There are significant gaps in the system.</p>			



9. Interventions			
Standard	Green Amber Red	Action required	By whom By when
<p>9. People who develop dementia have prompt access to the full range of medical, psychological, therapeutic and social interventions as required.</p> <p>Refer to Sections 12 &amp; 13.</p>			
<p><b>Green:</b> People who develop dementia have, as required, prompt access to staff who deliver:</p> <ul style="list-style-type: none"> <li>• Medications in line with NICE guidance, according to a locally agreed protocol.</li> <li>• Strategies, equipment and aids to maintain mobility, promote exercise, address posture, correct gait and reduce the risk of falls.</li> <li>• Specialist pressure area care.</li> <li>• Strategies for maintenance of adequate oral intake in a safe manner. This includes swallowing assessment, eating programme with dietetic advice along with advice regarding posture, and diet to reduce risk of constipation.</li> <li>• Aids / adaptations to help to maintain continence.</li> <li>• Strategies to aid communication including communication passports, objects of reference, pictures, signposting, signs and symbols.</li> <li>• Additional supports to maintain self care and domestic skills and adaptation of the environment to aid daily living.</li> <li>• Support for people to engage in failure free activities including sensory stimulation, aromatherapy, and other therapeutic activities.</li> <li>• Reminiscence, reality orientation, life story books, validation therapy.</li> <li>• Dementia care mapping.</li> <li>• Positive Behaviour Support including promotion of positive behaviour and feelings of self esteem, anxiety management, functional analysis and setting up of programmes for challenging behaviours, setting up of crisis intervention plans</li> </ul>			
<p><b>Amber:</b> Most elements of a comprehensive intervention service are in place, but there are a few gaps.</p>			
<p><b>Red:</b> There are significant gaps in the range of services that are required.</p>			

10. Dementia friendly environments			
Standard	Green Amber Red	Action required	By whom By when
<p>10. People who develop, or are at risk of developing dementia, have accommodation and day and leisure activities which are dementia friendly and are commissioned to meet their changing needs.</p> <p>Refer to Sections 11.</p>			
<p><b>Green:</b> Most people in the area, who develop or are at risk of developing dementia, have accommodation and day and leisure activities which are dementia friendly and can meet their changing needs. Any accommodation and / or day and leisure activity will be appropriate in terms of:</p> <ul style="list-style-type: none"> <li>• Have been identified as being 'dementia friendly'.</li> <li>• Have ensured that environments are safe, suitably stimulating, make sense and are predictable.</li> <li>• Have ensured appropriate use of colour in homes and services.</li> <li>• Have ensured appropriate furnishings are provided.</li> <li>• Provide appropriate aids and adaptations in a timely manner.</li> <li>• Provide appropriate wheelchairs, special beds and special seating in a timely manner.</li> <li>• Use appropriate assistive technology.</li> </ul>			
<p><b>Amber:</b> Some people in the area will have access to accommodation and activities that meet these quality standards.</p>			
<p><b>Red:</b> Few people in the area have access to accommodation and activities that meet these quality standards.</p>			

11. 'Dying in place'				
Standard	Green Amber Red	Action required	By whom	By when
11. People who develop dementia are supported to 'die in place', with additional supports provided in a timely manner.  Refer to Section 11.				
<p><b>Green:</b> There are processes in place to ensure that people who develop dementia are supported in ways that help to maintain them in their own home with:</p> <ul style="list-style-type: none"> <li>• Good advance planning</li> <li>• Additional staff as required including waking night staff</li> <li>• Environmental adaptations as necessary</li> <li>• A system for rapid availability of funding to meet changing needs</li> <li>• Good end of life care</li> </ul> <p>If the person needs to move, then the new service should be:</p> <ul style="list-style-type: none"> <li>• Close to the person's existing home</li> <li>• Within intellectual disabilities services</li> <li>• Allow links to existing day and leisure opportunities to be maintained</li> <li>• A final move</li> </ul> <p><b>Amber:</b> Most people will receive care that meets this standard, but there are some gaps for a few people, such that they are moved out of the intellectual disability services</p> <p><b>Red:</b> Fewer than half of the people who have dementia receive care that meets this standard, or people experience significant delays in providing necessary supports, or people are placed in nursing homes.</p>				

12. Choices and rights of people with intellectual disabilities and dementia			
Standard	Green Amber Red	Action required	By whom By when
12. The wishes, choices and rights of the person with intellectual disabilities who develops, or is at risk of developing dementia are respected, and this is evidenced in the care provided. Refer to Section 10.			
<p><b>Green:</b> Within the identified area, people with intellectual disabilities who develop, or are at risk of developing dementia all have:</p> <ul style="list-style-type: none"> <li>• A regularly updated Person Centred Plan.</li> <li>• A regularly updated Health Action Plan.</li> <li>• An individualised care plan that is developed with all services that the person utilises, and is coordinated across services.</li> <li>• Accessible reports about their care.</li> <li>• Accessible information about dementia.</li> <li>• Support for their peers with regard to dementia.</li> <li>• Appropriate advocacy to meet their needs.</li> </ul>			
<p><b>Amber:</b> There are some gaps in services so that most, but not all people have access to all these provisions.</p>			
<p><b>Red:</b> There are significant gaps, such that few people have access to all these provisions.</p>			

13. Support to family carers				
Standard	Green Amber Red	Action required	By whom	By when
13. Family carers are assisted to understand and support their relative who has dementia. Their needs as carers are met.  Refer to Section 17.				
<p><b>Green:</b> Within the identified area, family carers are assisted to understand and support their relative who has dementia:</p> <ul style="list-style-type: none"> <li>• All carers of a person with intellectual disabilities and dementia are offered a carer's assessment in line with Local Authority guidance.</li> <li>• Respite care / short breaks are available to those with dementia who are living with a family member. The respite provision is able to provide 'dementia friendly' accommodation and services.</li> <li>• Family carers are offered accessible information including: dementia in people with intellectual disability/Down's Syndrome, thyroid disorder, depression, pain awareness, etc, as relevant to the individual person.</li> <li>• Family carers are provided with support from the local Intellectual Disabilities Community Team as required to manage, for example, challenging behaviour, depression, epilepsy, eating and drinking, aids and adaptations.</li> <li>• Family carers are offered training sessions to increase awareness of dementia and its management in the home setting.</li> <li>• Family carers are consulted about the development of services to people with intellectual disabilities in their area.</li> </ul>				
<b>Amber:</b> Carers have access to most of these services and there are only a few gaps in such provision.				
<b>Red:</b> There are significant gaps in the provision for carers.				

14. Capable workforce				
Standard	Green Amber Red	Action required	By whom	By when
<p>14. People who develop, or are at risk of developing dementia, and their families, receive support, advice and care from a capable workforce that is appropriately skilled.</p> <p>Refer to Section 17.</p>				
<p><b>Green:</b> Within the identified area:</p> <ul style="list-style-type: none"> <li>Members of the Intellectual Disabilities Community Team are up to date in their knowledge and skills through continuing professional development opportunities with regard to: the risks of dementia, its early symptoms and progress and the methods of managing it. As a consequence, they can provide evidence-based assessment and advice to individuals and care teams.</li> <li>A rolling programme of dementia training is provided to the care staff via whole team training in the identified area throughout the year. A log is kept of attendance. Homes caring for someone with dementia and intellectual disabilities are specifically targeted.</li> <li>Intellectual Disabilities Community Team staff provide training and support to care staff teams in residential, respite and day activity settings concerning the management of any individual with dementia, as required.</li> <li>There is a workforce development plan that addresses the issues of dementia care.</li> </ul>				
<p><b>Amber:</b> There are some gaps in the workforce development, with some staff not being suitably trained.</p>				
<p><b>Red:</b> There are significant gaps in the workforce development, with many staff not having the necessary skills to support people who have dementia.</p>				

15. End of Life Care				
Standard	Green Amber Red	Action required	By whom	By when
15. People with intellectual disabilities and dementia have End of Life care delivered in line with the national strategy. Refer to Section 15.				
<p><b>Green:</b> Within the identified area, End of Life care includes all the following elements:</p> <ul style="list-style-type: none"> <li>• The person with intellectual disabilities and dementia has been involved as fully as possible in decisions and preferences with regard to end of life care.</li> <li>• Carers of the person with intellectual disabilities and dementia have been involved in end of life discussions.</li> <li>• Mental capacity assessment has taken place with regard to specific end of life decisions.</li> <li>• Where possible, the person with intellectual disabilities has been given the opportunity to make Advance decisions about their care.</li> <li>• Where the person does not have capacity, decisions are made following Best Interests guidance.</li> <li>• Palliative care services are able to meet the needs of people with intellectual disabilities and dementia.</li> </ul> <p><b>Amber:</b> There are a few gaps in the provision of a comprehensive end of life care for people with intellectual disabilities, but most of the elements are available.</p> <p><b>Red:</b> There are significant gaps in provision, such that few people receive comprehensive end of life care.</p>				

16. Quality Outcomes				
Standard	Green Amber Red	Action required	By whom	By when
16. The quality outcomes for people with learning disabilities and dementia are measured regularly to ensure that there is evidence that people are receiving excellent person centred dementia care.  Refer to Section 18.				
<p><b>Green:</b> Within the identified area,</p> <ul style="list-style-type: none"> <li>• The person with intellectual disabilities and dementia has had their quality outcome measured at least at each stage of dementia</li> <li>• The person with intellectual disabilities and dementia and their staff and family carers have been involved in assessing the person's quality outcome.</li> <li>• The quality outcome measure has been used to identify where improvements in care can be made</li> <li>• The improvements have been put in place within the agreed timescales.</li> <li>• Further reviews have taken place to ensure the quality outcome is maintained especially with the progression across the stages of dementia.</li> </ul> <p><b>Amber:</b> There are some gaps in the measurement of the person's quality outcome and the timely improvements needed</p> <p><b>Red:</b> There are significant gaps in the measurement of the person's quality outcome and the timely improvements needed</p>				



## Appendix 2

### QUALITY OUTCOME MEASURE FOR INDIVIDUALS WITH DEMENTIA (QOMID)

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#### INSTRUCTION BOOKLET

##### What is the QOMID?

The Quality Outcome Measure for Individuals with Dementia (QOMID) has been designed to measure the quality outcomes for any person with dementia. It is easy to use, and in initial analysis, has good internal consistency as a measure of quality outcome.

The QOMID consists of 17 domains which explore the key areas that ensure that the person with dementia is experiencing a good quality experience.

The QOMID is staged for the three main stages of dementia – suspected/ early; mid and late stage. Although the domains are the same for each stage, the description of quality outcomes may change across the stages to reflect the different requirements as dementia progresses.

The QOMID must be used in its entirety, and should not be amended.

The QOMID is available free of charge from: <http://www.bps.org.uk/networks-and-communities/member-microsite/dcp-faculty-people-intellectual-disabilities/quality-outcome-measures-individuals-deme>

**We would like to collect further data on its use so that we can do further analysis on its usefulness and statistical properties. Each time you complete it, please could you upload anonymised data to: <https://www.surveymonkey.com/s/K6LB3ZD>**

##### When to use the QOMID

The QOMID should be used sequentially to rate the quality outcome of the person as their dementia progresses. The QOMID should be used as part of a regular review of the person's care to ensure that the person maintains good quality experience through the course of their dementia.

### **Who should complete the QOMID?**

- The QOMID should be completed by the professional in discussion with relevant people for each stage of dementia.
- It is particularly effective when completed as part of a review of the person's care.
- Wherever possible, and depending on ability, the person with dementia should be asked how they would rate their experience in each domain.
- Additional information for the professional to make an inclusive judgement may come from family, support staff, advocates, care managers or anyone else involved with the person and their support.
- The person completing the QOMID should record the evidence for their decision for each domain.

### **How to use the QOMID**

1. **Staging of dementia:** The professional completing the QOMID needs to use a combination of results from formal assessments together with their professional judgement to decide which stage of dementia the person is currently experiencing.

	<b>Suspected / Early stage</b>	<b>Mid stage</b>	<b>Late Stage</b>
<u>Early stage dementia</u> is usually characterised by gradual minor changes in the person's abilities or behaviour, especially loss of short term memory. The person may also start to become anxious and agitated. They may experience distress over their failure to manage tasks, and will need reassurance and emotional support. People who fall into Care Cluster 18 (Cognitive Impairment- Low Need) may fall into this group.			
<u>Mid stage dementia</u> is characterised by the changes becoming more marked. The person will need more support to help them manage their day-to-day living. They may need frequent reminders or help to eat, wash, dress and use the toilet. They are likely to become increasingly forgetful - particularly of names - and may sometimes repeat the same question or phrase over and over because of the decline in their short-term memory. They will also experience loss of long-term memories resulting in them failing to recognise people or confuse them with others, and for older memories to become more vivid. Some people at this stage become very easily upset, angry or aggressive - perhaps because they are feeling frustrated - or they may lose their confidence and become very clingy. People who fall into Care Clusters 19 (Cognitive Impairment- Moderate Need) or Care Cluster 20 (Cognitive Impairment or Dementia Complicated- High Need) may fall into this group.			

Late stage dementia is when the person with dementia will need even more help, and will gradually become totally dependent on others for nursing care. Loss of memory may become very pronounced, with the person unable to recognise familiar objects or surroundings or even those closest to them, although there may be sudden flashes of recognition. The person may also become increasingly frail. They may start to shuffle or walk unsteadily, eventually becoming confined to bed or a wheelchair. People who fall into or Care Cluster 21 (Cognitive Impairment or Dementia- High Physical or Engagement) may fall into this group.

Use the column for that stage of dementia and rate each domain using the following rating scale:

<b>1</b>	<b>2</b>	<b>3</b>	<b>4</b>
<b>This is rarely achieved for this person</b>	<b>This is sometimes achieved for this person</b>	<b>This is mostly achieved for this person</b>	<b>This is completely and consistently achieved for this person</b>

For each domain, enter the rating at this current time onto the **Scoring Sheet**. All domains should be completed. If the domain is rated less than 4, specify what needs to happen to improve the person's quality outcome in that area of their life as an action in the **Scoring Sheet**.

**Expected scoring:** The aim, in supporting the person with dementia, is for them to have an excellent quality outcome throughout the progression of their dementia.

As dementia is a progressive condition, it is vital to ensure that the person's changing needs are recognised and met. This means that as the person moves into each stage of dementia, the quality outcome score for each domain may start at 2 or 3, but as people work together to improve the person's quality outcome, the scores should reach the maximum of 4 in each domain.

Scores may fluctuate during the course of the dementia as support 'catches up' with the person's changing needs. Scores should be entered onto the **Scoring sheet**, putting the evidence for the rating.

**A score of 60 – 68 indicates that the person has an excellent quality outcome.**

**A score of 51 – 59 indicates that the person has a good quality outcome.**

**A score of 43 – 50 indicates that the person has an adequate quality outcome.**

**A score of 34 – 42 indicates that the person has a poor quality outcome.**

**A score of 33 or less indicates that the person has an unacceptable quality outcome.**

**3. Forward planning:** The QOMID is designed to help the support team and the professionals to work with the person to both prevent deterioration in quality and to forward plan effective care.

For each domain that is scored at less than 4, the support team is asked to specify what needs to be put in place to improve the person's quality outcome for that domain. These actions should be entered into the **Scoring Sheet** and can then be included in the person's support plan.

In addition, by looking at the descriptions for the next stage of dementia, the professional can begin to help the person and their supporters to think about what needs to be put in place to maintain their quality experience.

For further information please contact: Karen Dodd – [DrKaren.dodd@sabp.nhs.uk](mailto:DrKaren.dodd@sabp.nhs.uk)

**QUALITY OUTCOME MEASURE FOR INDIVIDUALS WITH DEMENTIA (QOMID)**

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**DOMAIN REFERENCE BOOKLET**

AREA	SUSPECTED / EARLY STAGE DEMENTIA	MID STAGE DEMENTIA	LATE STAGE DEMENTIA
<b>1. PERSON CENTRED APPROACHES TO SUPPORT</b>	<p>The person experiences support which is underpinned by planning based on:</p> <ul style="list-style-type: none"> <li>• the person's own wishes,</li> <li>• their capacity (maximising their decision making wherever possible),</li> <li>• their needs and history as shown in their individualised support plan which includes                             <ul style="list-style-type: none"> <li>○ their person centred plan,</li> <li>○ health care plan,</li> <li>○ communication passport,</li> <li>○ life story book,</li> <li>○ advanced directives, and</li> <li>○ end of life planning.</li> </ul> </li> </ul>	<p>The person experiences support which is underpinned by planning based on:</p> <ul style="list-style-type: none"> <li>• the person's own wishes,</li> <li>• their capacity (maximising their decision making wherever possible),</li> <li>• their needs and history as shown in their individualised support plan which includes                             <ul style="list-style-type: none"> <li>○ their person centred plan,</li> <li>○ health care plan,</li> <li>○ communication passport,</li> <li>○ life story book,</li> <li>○ advanced directives, and</li> <li>○ end of life planning.</li> </ul> </li> </ul>	<p>The person experiences support which is underpinned by planning based on:</p> <ul style="list-style-type: none"> <li>• the person's own wishes,</li> <li>• their capacity (maximising their decision making wherever possible),</li> <li>• their needs and history as shown in their individualised support plan which includes                             <ul style="list-style-type: none"> <li>○ their person centred plan,</li> <li>○ health care plan,</li> <li>○ communication passport,</li> <li>○ life story book,</li> <li>○ advanced directives, and</li> <li>○ end of life planning.</li> </ul> </li> </ul>
<b>2. POSITIVE RISK TAKING</b>	<p>The person is supported to take appropriate risks that enhance their opportunities to live an independent, fulfilled life.</p>	<p>The person is supported to take appropriate risks that enhance their opportunities to live an independent, fulfilled life.</p>	<p>The person is supported by people who take positive action to ensure that the person still has a range of fulfilling life experiences.</p>

AREA	SUSPECTED / EARLY STAGE DEMENTIA	MID STAGE DEMENTIA	LATE STAGE DEMENTIA
<b>3. RESPECT FOR HUMAN RIGHTS</b>	<p>The person's human rights are fully respected by ensuring that there is full compliance with:</p> <ul style="list-style-type: none"> <li>• Prescribed medication is in line with NICE guidelines</li> <li>• Mental Capacity Act- choice and decision making. The person is supported to make as many decisions for themselves as is possible</li> <li>• Best Interest Decisions that are made on behalf of the person are fully documented</li> <li>• Absence of inappropriate restrictions</li> </ul> <p>If the person tends to wander, this is managed through respectful and positive approaches that do not impact on their human rights</p>	<p>The person's human rights are fully respected by ensuring that there is full compliance with:</p> <ul style="list-style-type: none"> <li>• Prescribed medication is in line with NICE guidelines</li> <li>• Mental Capacity Act- choice and decision making. The person is supported to make as many decisions for themselves as is possible</li> <li>• Best Interest Decisions that are made on behalf of the person are fully documented</li> <li>• Absence of inappropriate restrictions</li> </ul> <p>If there is a need to deprive somebody of their liberty, the appropriate Deprivation of Liberty Safeguards are in place.</p>	<p>The person's human rights are fully respected by ensuring that there is full compliance with:</p> <ul style="list-style-type: none"> <li>• Prescribed medication is in line with NICE guidelines</li> <li>• Mental Capacity Act- choice and decision making. The person is supported to make as many decisions for themselves as is possible</li> <li>• Best Interest Decisions that are made on behalf of the person are fully documented</li> <li>• Absence of inappropriate restrictions</li> </ul> <p>If there is a need to deprive somebody of their liberty, the appropriate Deprivation of Liberty Safeguards are in place.</p>
<b>4. CONSISTENCY OF APPROACH</b>	<p>The person experiences consistency of approach in all settings e.g.</p> <ul style="list-style-type: none"> <li>• They are supported by familiar people</li> <li>• Family /Staff fully understand the content of their support plan</li> <li>• New staff are properly introduced to the person before they start working with them</li> <li>• The use of unfamiliar or agency staff is minimised and staff are well briefed about the person before they start to support them.</li> </ul>	<p>The person experiences consistency of approach in all settings e.g.</p> <ul style="list-style-type: none"> <li>• They are supported by familiar people</li> <li>• Family /Staff fully understand the content of their support plan</li> <li>• New staff are properly introduced to the person before they start working with them</li> <li>• The use of unfamiliar or agency staff is minimised and staff are well briefed about the person before they start to support them.</li> <li>• They are not moved unnecessarily because of funding issues (e.g. need for waking night staff)</li> </ul>	<p>The person experiences consistency of approach in all settings e.g.</p> <ul style="list-style-type: none"> <li>• They are supported by familiar people</li> <li>• Family /Staff fully understand the content of their support plan</li> <li>• New staff are properly introduced to the person before they start working with them</li> <li>• The use of unfamiliar or agency staff is minimised and staff are well briefed about the person before they start to support them.</li> <li>• They are not moved unnecessarily because of funding issues (e.g. need for waking night staff)</li> </ul>

AREA	SUSPECTED / EARLY STAGE DEMENTIA	MID STAGE DEMENTIA	LATE STAGE DEMENTIA
<b>5. INTERACTION WITH OTHERS</b>	The person experiences calm and constructive interaction with family, staff and friends, who adapt the amount of language used and use symbols and pictures as required to ensure the person experiences positive interactions.	The person experiences calm and constructive interaction with family, staff and friends, with no confrontation; no time pressures; and validation of roll back memories. The person experiences positive interactions and is always approached from the front to prevent surprise and panic.	The person experiences calm and constructive interaction with family, staff and friends, with protected 1:1 time each waking hour to ensure that the person experiences positive interactions.
<b>6. EMOTIONAL REASSURANCE TO COPE WITH THE CHANGES</b>	The person receives explanations about their dementia and reassurance about the effects of the disease as appropriate to their wishes and level of ability.	The person is reassured about the changes they are experiencing through both verbal and non verbal interaction.	The person is reassured about their condition by the way people interact both verbally and through appropriate touch.
<b>7. ORIENTATION</b>	The person is oriented to time and place through approaches that are appropriate to their level of ability. Their support plan describes routines that are likely to be important to the person as the dementia progresses. There is evidence that the team has made plans to ensure that any future changes that are envisaged for the person are properly considered and take account of possible effect on the person's orientation	The person is able to understand their daily routine through the use of appropriate cues and aids e.g. daily picture timetable, picture menus, picture staff rotas. There is evidence that the team has made plans to ensure that any future changes that are envisaged for the person are properly considered and take account of possible effect on the person's orientation	The person feels safe in having a consistent and familiar routine.
<b>8. DAILY LIVING</b>	The person is able to complete personal care and daily living activities as much as they are able, but without pressure. The person's abilities and additional assistance required to help maintain independence are recognised, and the person is supported appropriately e.g. having increased prompting.	The person is able to complete parts of personal care and daily living tasks that they can do and are assisted as necessary so they do not fail. Their support plan details the additional assistance required to help maintain as much independence as possible in a failure free manner.	The person experiences care that is dignified and respectful of them as a person for all their personal care and daily living activities.

<b>AREA</b>	<b>SUSPECTED / EARLY STAGE DEMENTIA</b>	<b>MID STAGE DEMENTIA</b>	<b>LATE STAGE DEMENTIA</b>
<b>9. CARRYING OUT PREFERRED ACTIVITIES</b>	The person continues to access and enjoy activities which build upon their lifelong interests and preferences and are appropriate to their level of ability and dementia. Activities are adapted to meet their changing needs.	The person continues to access and enjoy activities which build upon their lifelong interests and preferences and are appropriate to their level of ability and dementia. Activities are adapted to take account of their attention span and memory and ensure that the person is not stressed or experiences failure.	The person continues to access and enjoy activities appropriate to their level of ability and dementia. The person has opportunities to interact with people / objects which give them enjoyment and in ways that take full account of their preferences and attention span.
<b>10. FLEXIBILITY OF SUPPORT</b>	The person continues to attend familiar social, leisure, work, respite and recreational activities in their local community, with adjustments made as appropriate to meet their needs.	The person continues to enjoy familiar social, leisure, work, respite and recreational activities in their local community through flexible supports e.g. short days, flexible transport, 1:1 support following the person.	The person continues to access and enjoy the community as much as their dementia allows and as agreed in their support plan.
<b>11. ENVIRONMENT</b>	The person lives and spends their time in environments that are familiar to them and can find their way around easily with depth perception problems minimised. e.g. flooring colour is consistent.	The person lives and spends their time in environments that are familiar to them and help them find their way around and meet their needs, and minimises risks of falls. e.g. red toilet doors, red toilet seats, colour contrasts, good signage, handrails, chairs at right height.	The person lives and spends their time in environments that are familiar to them and have all the necessary adaptations to meet their needs. e.g. hoists, adapted bath / shower, special bed, appropriate wheelchair and armchair, changing facilities.



AREA	SUSPECTED / EARLY STAGE DEMENTIA	MID STAGE DEMENTIA	LATE STAGE DEMENTIA
12. BEHAVIOUR	<p>Behavioural issues are minimised by ensuring that the person experiences support that:</p> <ul style="list-style-type: none"> <li>• Understands the context of their behaviour</li> <li>• responds with compassion and avoids confrontation.</li> </ul> <p>If the person needs support from services because of their behaviour this is underpinned by:</p> <ul style="list-style-type: none"> <li>• a comprehensive assessment of the person, their care and the environment,</li> <li>• a formulation that enables carers or staff to understand the likely reasons for the behaviour,</li> <li>• a proactive support plan that includes triggers to be avoided,</li> <li>• reactive strategies that are non-restrictive and the effectiveness of the approach is reviewed regularly.</li> </ul>	<p>Behavioural issues are minimised by ensuring that the person experiences support that:</p> <ul style="list-style-type: none"> <li>• Understands the context of their behaviour</li> <li>• responds with compassion and avoids confrontation.</li> </ul> <p>If the person needs support from services because of their behaviour this is underpinned by:</p> <ul style="list-style-type: none"> <li>• a comprehensive assessment of the person, their care and the environment,</li> <li>• a formulation that enables carers or staff to understand the likely reasons for the behaviour,</li> <li>• a proactive support plan that includes triggers to be avoided,</li> <li>• reactive strategies that are non-restrictive and the effectiveness of the approach is reviewed regularly.</li> </ul>	<p>Behavioural issues are minimised by ensuring that the person experiences support that:</p> <ul style="list-style-type: none"> <li>• Understands the context of their behaviour</li> <li>• responds with compassion and avoids confrontation.</li> </ul> <p>If the person needs support from services because of their behaviour this is underpinned by:</p> <ul style="list-style-type: none"> <li>• a comprehensive assessment of the person, their care and the environment,</li> <li>• a formulation that enables carers or staff to understand the likely reasons for the behaviour,</li> <li>• a proactive support plan that includes triggers to be avoided,</li> <li>• reactive strategies that are non-restrictive and the effectiveness of the approach is reviewed regularly.</li> </ul>

AREA	SUSPECTED / EARLY STAGE DEMENTIA	MID STAGE DEMENTIA	LATE STAGE DEMENTIA
<p><b>13. HEALTH</b></p>	<p>The person's physical and mental health needs are met promptly and appropriately including attention to:</p> <ul style="list-style-type: none"> <li>• Pain recognition and management</li> <li>• Thyroid function</li> <li>• Vision</li> <li>• Hearing</li> <li>• Blood pressure</li> <li>• Diabetes</li> <li>• Mental wellbeing</li> </ul> <p>Medication is prescribed appropriately and reviewed regularly.</p> <p>The person experiences care with regard to Vitamin D in line with DH guidance.</p>	<p>The person's physical and mental health needs are met promptly and appropriately including attention to:</p> <ul style="list-style-type: none"> <li>• Pain recognition and management</li> <li>• Thyroid function</li> <li>• Vision</li> <li>• Hearing</li> <li>• Blood pressure</li> <li>• Diabetes</li> <li>• Mental wellbeing</li> </ul> <p>Medication is prescribed appropriately and reviewed regularly.</p> <p>The person experiences care with regard to Vitamin D in line with DH guidance.</p>	<p>The person's physical and mental health needs are met promptly and appropriately including attention to:</p> <ul style="list-style-type: none"> <li>• Pain recognition and management</li> <li>• Thyroid function</li> <li>• Vision</li> <li>• Hearing</li> <li>• Blood pressure</li> <li>• Diabetes</li> <li>• Mental wellbeing</li> </ul> <p>Medication is prescribed appropriately and reviewed regularly.</p> <p>The person has:</p> <ul style="list-style-type: none"> <li>• No pressure sores</li> <li>• No aspiration</li> <li>• No urinary tract infections</li> </ul> <p>The person experiences care with regard to Vitamin D in line with DH guidance.</p>
<p><b>14. SUPPORT FROM WELL COORDINATED AGENCIES</b></p>	<p>The person's needs are met by people from providers in primary care, secondary care, social services and voluntary sector who have a good understanding of the needs of people with dementia, and who work well together with the person and their families.</p>	<p>The person's needs are met by people from providers in primary care, secondary care, social services and voluntary sector who have a good understanding of the needs of people with dementia, and who work well together with the person and their families. Where necessary good links are made with neurology services re management of epilepsy.</p>	<p>The person's needs are met by people from providers in primary care, secondary care, social services and voluntary sector who have a good understanding of the needs of people with dementia, and who work well together with the person and their families. Good links are made with local palliative care services.</p>

AREA	SUSPECTED / EARLY STAGE DEMENTIA	MID STAGE DEMENTIA	LATE STAGE DEMENTIA
<b>15. NUTRITION</b>	The person enjoys a good and appetising diet and adequate hydration. The person maintains an appropriate weight which is monitored through regular weight checks.	The person enjoys a good and appetising diet and adequate hydration as appropriate to their needs over each 24 hour period. The person maintains an appropriate weight which is monitored through regular weight checks. Any swallowing difficulties are identified and support plans take these into full account	The person enjoys a good and appetising diet and adequate hydration as appropriate to their needs over each 24 hour period which also prevents dysphagia and aspiration. There is a full assessment of all eating and swallowing problems by an appropriate clinician. Any needs are well documented, a support plan is in place and staff are trained to deliver it safely. The person maintains an appropriate weight which is monitored through regular weight checks.
<b>16. MOBILITY</b>	The person maintains good mobility. They access regular exercise that is appropriate to their needs and interests.	The person is able to mobilise safely and has appropriate aids and adaptations in place. They access regular exercise that is appropriate to their needs and interests. Risk assessments are in place to prevent falls.	The person is supported to be moved appropriately. They access regular exercise that is appropriate to their needs and interests. Risk assessments are in place to prevent falls.
<b>17. CONTINENCE</b>	The person maintains their baseline level of continence.	The person maintains their baseline level of continence through environmental changes e.g. clear signage for toilets; regular prompting to use the toilet; and attention to relevant health issues where possible. Continence products are only used when the person needs them.	The person experiences dignified management of incontinence through the use of appropriate aids and continence products.

**QUALITY OUTCOME MEASURE FOR INDIVIDUALS  
WITH LEARNING DISABILITIES AND DEMENTIA (QOMID)**

Karen Dodd and Alick Bush (2013) ©

**SCORING SHEET**

We would like to collect further data on its use so that we can do further analysis on its usefulness and statistical properties. Each time you complete it, please could you upload anonymised data to: <https://www.surveymonkey.com/s/K6LB3ZD>

<b>PERSON'S NAME:</b>	<b>DOB:</b>
<b>ADDRESS:</b>	<b>DATE OF COMPLETION:</b>
<b>NAME OF PERSON COMPLETING:</b>	<b>DESIGNATION:</b>
<b>PEOPLE PRESENT:</b>	
<b>Current stage of dementia (please circle)</b>	<b>LATE STAGE DEMENTIA</b>
<b>SUSPECTED / EARLY STAGE DEMENTIA</b>	<b>MID STAGE DEMENTIA</b>

DOMAIN	SCORE	EVIDENCE FOR RATING	ACTION NEEDED TO IMPROVE QUALITY OUTCOME
1. Person Centred Approaches to Support			
2. Positive Risk Taking			
3. Respect for Human Rights			
4. Consistency of approach			
5. Interaction with others			

1	2	3	4
This is rarely achieved for this person	This is sometimes achieved for this person	This is mostly achieved for this person	This is completely and consistently achieved for this person

DOMAIN	SCORE	EVIDENCE FOR RATING	ACTION NEEDED TO IMPROVE QUALITY OUTCOME
6. Emotional reassurance to cope with the changes			
7. Orientation			
8. Daily Living			
9. Carrying out preferred activities			
10. Flexibility of support			

1	2	3	4
This is rarely achieved for this person	This is sometimes achieved for this person	This is mostly achieved for this person	This is completely and consistently achieved for this person

DOMAIN	SCORE	EVIDENCE FOR RATING	ACTION NEEDED TO IMPROVE QUALITY OUTCOME
11. Environment			
12. Behaviour			
13. Health			
14. Support from well co-ordinated agencies			
15. Nutrition			

1	2	3	4
This is rarely achieved for this person	This is sometimes achieved for this person	This is mostly achieved for this person	This is completely and consistently achieved for this person

DOMAIN	SCORE	EVIDENCE FOR RATING	ACTION NEEDED TO IMPROVE QUALITY OUTCOME
16. Mobility			
17. Continence			
Total score (Max = 68):			

1	2	3	4
This is rarely achieved for this person	This is sometimes achieved for this person	This is mostly achieved for this person	This is completely and consistently achieved for this person

**EXPLANATION OF TOTAL SCORE:**

- A score of 60 – 68 indicates that the person has an excellent quality outcome.
- A score of 51 – 59 indicates that the person has a good quality outcome.
- A score of 43 – 50 indicates that the person has an adequate quality outcome.
- A score of 34 – 42 indicates that the person has a poor quality outcome.
- A score of 33 or less indicates that the person has an unacceptable quality outcome.



# Appendix 3 - Leaflet for Commissioners of Services for People with Intellectual Disabilities and dementia

## 1. The Specific Issues

The life-expectancy of people with mild intellectual disabilities now approaches that in the general population of a similar socio-economic status, but the life-expectancy of people with more severe levels of intellectual disabilities remains reduced compared to the general population. Given these improvements, the overall population with intellectual disabilities is steadily increasing and it has been predicted that the proportion of people with intellectual disabilities over 65 years of age will have doubled by 2020, with over a third of all people with intellectual disabilities being over 50 years of age by that time. This is also true for the population of people with Down's syndrome.

There is an increase in the prevalence rates of clinically diagnosed dementia with increasing age that starts when people with Down's syndrome are in their 30s and steadily increases in prevalence into the 60s. It has been calculated that nearly 70% of older adults with Down's syndrome are likely to develop dementia symptoms should they all live to age 70.

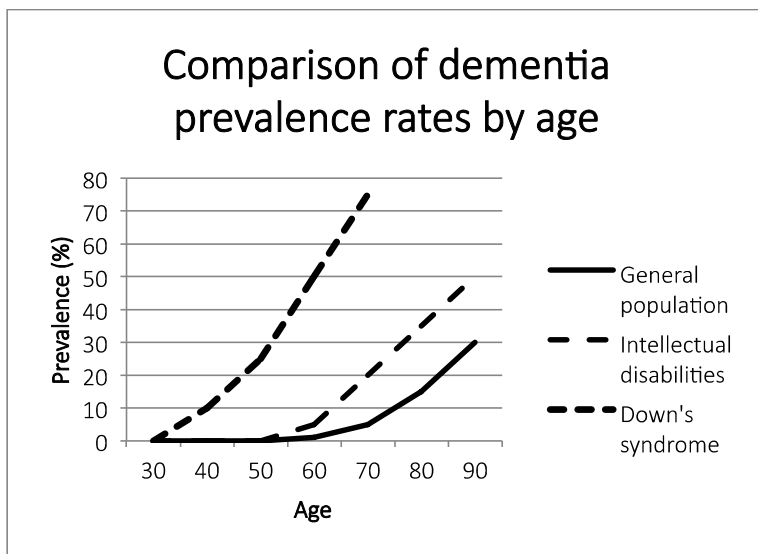


Figure 1 summarises the age-related prevalence rates of dementia in people with Down's syndrome, those with intellectual disabilities without Down's syndrome, and in the general population. The exact rates have to be considered with caution but the trend represented in this figure is now increasingly accepted. The early presentation and course of dementia is now well established for people with Down's syndrome. For those with intellectual disabilities but without Down's syndrome, age-related prevalence rates are brought forward to a small degree compared to the general population but not to the same extent as for people with Down's syndrome. This latter group would appear to have a uniquely early risk for developing dementia, almost invariably of the Alzheimer's-type. For the former group the full range of causes of dementia is observed.

## **2. What should happen**

Commissioners of health services need to be clear about the care pathway for the assessment, diagnosis, interventions and support for people with intellectual disabilities who develop dementia from primary care, through to appropriate secondary care services, and on to palliative care services. In practice, good quality care will involve active partnership-working between intellectual disability services, older people's services, primary and secondary health care, palliative care and social care. These areas should develop an integrated dementia strategy for the care of people with intellectual disabilities and dementia. This should involve the development of an integrated care pathway involving all relevant agencies. Specialist health professionals e.g. speech and language therapists, physiotherapists, occupational therapists, dietitians, community intellectual disabilities nurses are all essential partners in providing excellence in care.

Commissioners of social care need to recognise that dementia is by definition a deteriorating condition, and that peoples' needs will increase over time, and therefore increased and timely funding will be needed to ensure safe practice. This will inevitably involve the funding of waking night staff as the dementia progresses. Best practice is that people with intellectual disabilities should receive their residential care within the intellectual disabilities arena rather than in generic services for people with dementia, as the quality of the services gives people the best opportunities for a good quality of life for both their intellectual disabilities and their dementia. There will need to be efficient processes and understanding in place to ensure that Continuing Healthcare Assessments are undertaken promptly and funding agreed as needed.

Staff involved in assessment, diagnosis, interventions and support need to be trained in dementia care and be able to offer both holistic and specialist assessments and a range of interventions aimed at meeting the needs of people with intellectual disabilities and dementia. Care managers have an essential role to play in ensuring that services are actively monitored to ensure that they are responsive to the changing needs of the person. People with intellectual disabilities and dementia should have access to regular reviews (monthly - six monthly depending on the rate of deterioration) by an identified care manager. End of Life care needs to be planned in advance, using the same principles and services available to the general population. Good partnership-working with palliative care services is essential, both to support the person and the carers.

### **3.0 What are the elements of an excellent service?**

Commissioners will want to ensure that there is:

- Demographics are known including having a database of all adults with intellectual disabilities which includes identification of people with Down's syndrome and those in out of area placements.
- A multi-agency dementia strategy.
- A multi-agency care pathway for assessment, diagnosis, interventions and support of people with intellectual disabilities who develop dementia.
- A multi-disciplinary approach to assessment and diagnosis and support.
- Prompt access to assessment and diagnostic services including baseline assessment for people with Down's syndrome by the age of 30.
- Person-centred dementia care.
- Effective care management and review system.
- Prompt access to the full range of medical, psychological, therapeutic and social interventions.
- All living and day service environments are dementia friendly.

- The person is supported to remain in their familiar home with additional supports provided in a timely manner.
- Support is available to family carers and service providers.
- There is a capable workforce able to deliver excellence in dementia care.
- End of Life care follows the requirements of the National End of Life Strategy.

#### **4.0 What should the commissioners expect as outcomes of an excellent service?**

- Increase in prompt differential diagnosis of the person's difficulties.
- Increase in other conditions being treated promptly.
- Increase in accurate diagnosis of dementia.
- Reduction in behavioural difficulties.
- Increase in quality of life indicators for the person.
- Reduction in moves to other placements.
- Reduction in the need for emergency one-to-one cover, as a result of planned increases in support as the dementia progresses to enable people to continue to access preferred activities and for personal care.
- Reduction in out of area placements.
- Increased carer support and satisfaction.
- Reduction in staff stress.

#### **5.0 Potential risks if services are not available/not effective**

- Increased costs of one-to-one, new in area or out of area placements.
- More complaints.
- Potential safeguarding issues.
- Increase in behaviours leading to abuse or harm to self and others.
- Carer breakdown.

#### **6.0 Further Information**

##### a) Dementia Organisations

Alzheimer's Society  
 Devon House  
 58 St Katharine's Way  
 London E1W 1LB                      Tel: 0300 222 11 22  
 www.alzheimers.co.uk

##### b) Learning Disability Organisations

Down's Syndrome Association  
 Langdon Down Centre  
 2a Langdon Park  
 Teddington  
 Middlesex  
 TW11 9PS                              Tel: 0333 1212 300  
 www.downs-syndrome.org.uk

Scottish Down's Syndrome Association  
 158/160 Balgreen Road  
 Edinburgh EH11 3AU              Tel: 0131 313 4225  
 www.dsscotland.org.uk

MENCAP

123 Golden Lane  
London EC1Y 0RT  
www.mencap.org.uk

Tel: 020 7454 0454

BILD

Birmingham Research Park  
97 Vincent Drive  
Edgbaston  
Birmingham  
B15 2SQ  
www.bild.org.uk

Tel: 0121 415 6960

Publications

Supporting Derek ( Watchman et al., 2010)

<http://www.jrf.org.uk/publications/supporting-derek>

Down's Syndrome and dementia- a resource for carers and support staff  
(Dodd et al., 2009)

<http://www.bild.org.uk/our-services/books/bestsellers/downs-syndrome-and-dementia/>

Down's Syndrome and dementia workbook for staff. (Dodd et al 2006)

<http://www.downs-syndrome.org.uk/shop/publications/medical-and-health-for-professionals/1091-downs-syndrome-and-dementia-workbook.html>

About Dementia

The journey of life

About my friend

(Dodd et al., 2005)

<http://www.bild.org.uk/our-services/books/health-and-well-being/about-dementia/>

The Simplicity of Dementia. (Buijssen 2005) Jessica Kingsley Publishers.  
London.

Understanding Learning Disability and Dementia: Developing Effective Interventions.  
(Kerr 2007) Jessica Kingsley Publishers. London

Printed and published by the British Psychological Society.

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