INTELLECTUAL DISABILITY AND DEMENTIA PRACTICE

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Tucson, Arizona

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TODAY'S SPEAKERS

Matthew P. Janicki, Ph.D. is the co-chair of the US National Task Group on Intellectual Disabilities and Dementia Practices, as well as a research associate professor in the Department of Disability and Human Development at the University of Illinois Chicago. Formerly, he was director for aging and special populations for the New York State Office for People with Developmental Disabilities. Currently, he is leading a study of specialized group homes designed for dementia related care of adult with intellectual disabilities.

Seth M. Keller, MD is a neurologist and the co-President of the National Task Group in Intellectual Disabilities and Dementia Practices. He maintains a neurology practice in New Jersey and is a past-President of the American Academy of Developmental Medicine and Dentistry as well as the founding chair of the Intellectual Disabilities Interest Group within the American Academy of Neurology.

Thomas J. Buckley, Ed.D. serves as a consultant to several provider organizations, functions as an expert witness on disability discrimination suits, and serves on the Board of CARF. He was instrumental in aiding several organizations with setting up dementia-capable services for individuals with intellectual disabilities and is the author of numerous useful guides and products focusing on dementia care planning.

INTRODUCTION & OBJECTIVES

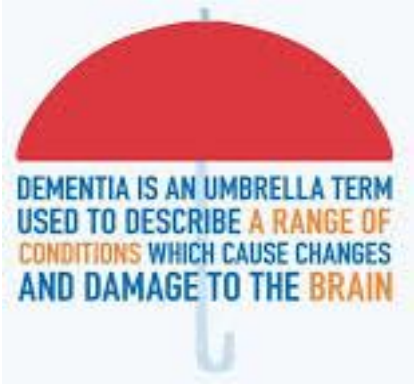
QUICK OVERVIEW OF DEMENTIA AND WHAT DOES IT MEAN

CONTENT

- Basics – Dementia and Intellectual Disabilities
- Federal and National Perspectives
- Dementia Processes
- Dementia Services and Supports
- Medical and Health Factors
- Individualized Care and Meeting Standards

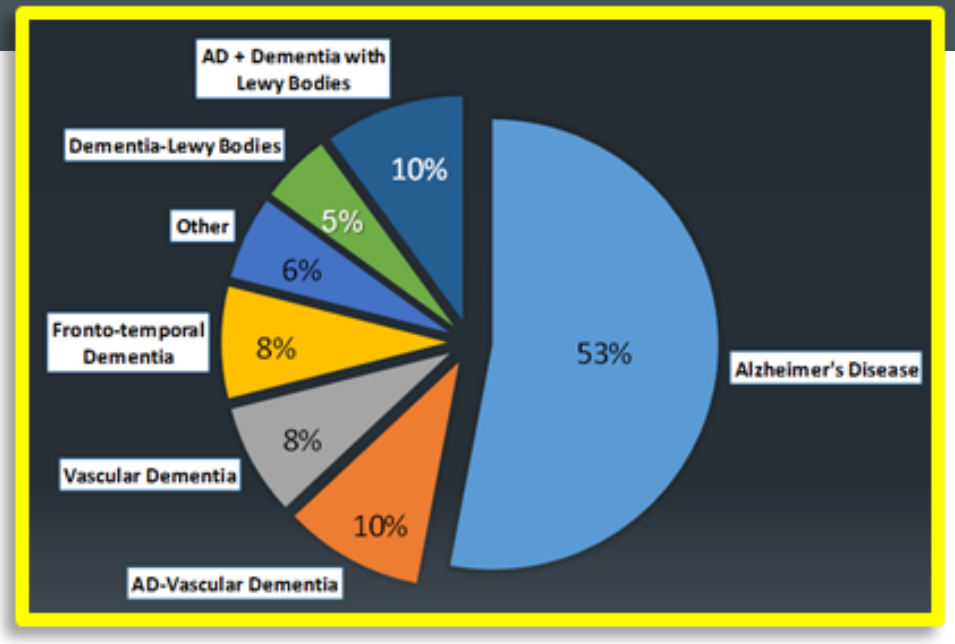
ALZHEIMER'S VS. DEMENTIA

Alzheimer's disease is the name of a neuropathology – or brain disease – that leads to general dysfunction



Dementia is the behavioral expression of the brain disease – usually via memory loss and behavioral dysfunction

… losses occur in memory, language skills, orientation, ADLs [activities of daily living], and changes in personality and global functioning



Causes of dementia among aging adults

- Alzheimer's disease
- Stroke and related vascular accidents
- Neurological diseases (Parkinson's)
- Idiopathic changes in the brain (Lewy, FTD)

INTELLECTUAL DISABILITY (ID)

Characterized by

- Below normative intellectual functioning, due to cognitive impairment (organic or functional) present since birth or infancy
- *Not a mental illness or psychiatric impairment*
- Varies in degree and co-impairment
- Compensated by training, education, remediation, habilitation, supports for life activities

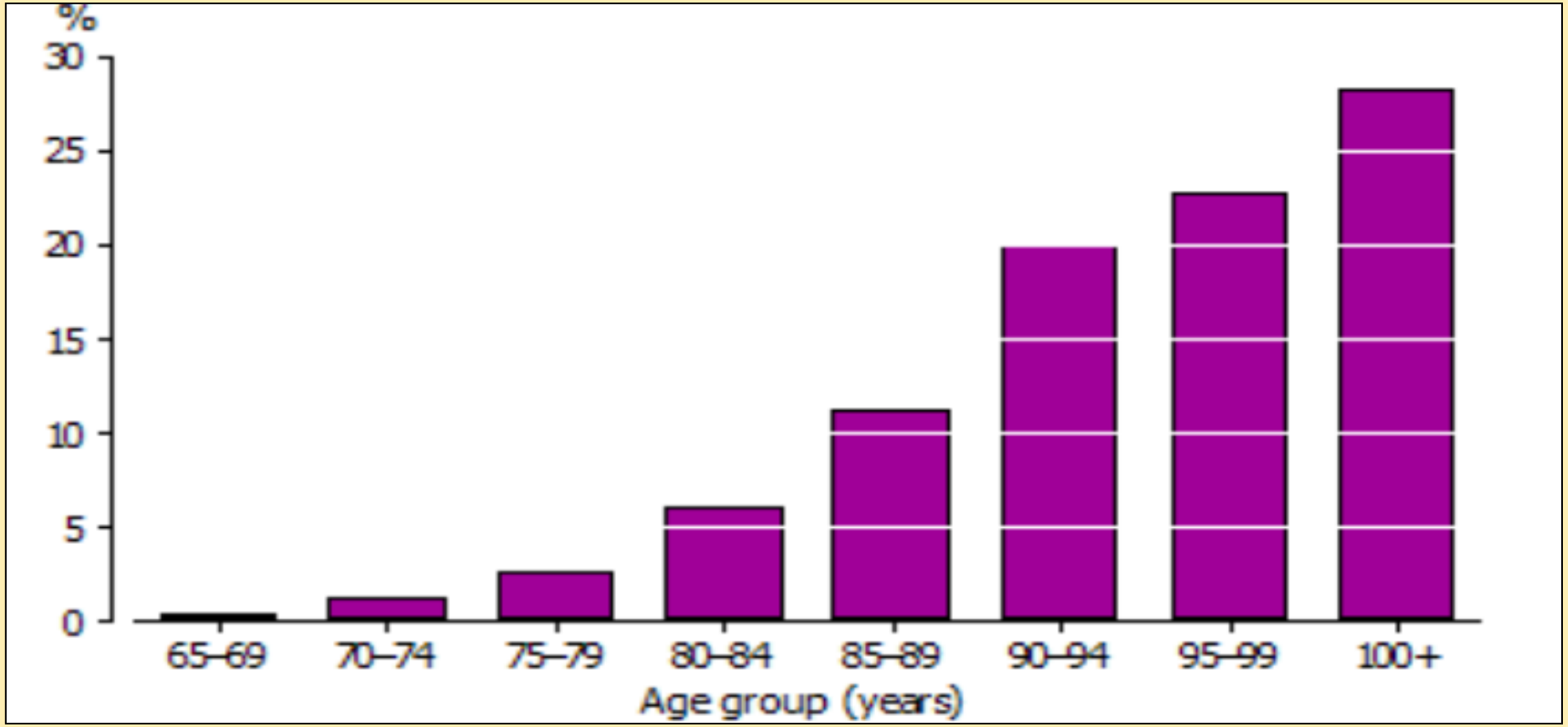
Down syndrome is a chromosomal abnormality present at birth (#21) associated with ID – In adults, age 40+, occurs in 10-12% of agency clientele

Developmental disabilities

- Neuro-developmental conditions leading to impairments in physical, learning, language, or behavior areas
- Originate in the developmental period, may impact day-to-day functioning, and usually last throughout a person's lifetime
- Intellectual disability is one of the developmental disabilities

https://www.cdc.gov/ncbddd/developmentaldisabilities/facts.html

PROPORTION OF AGE GROUPS IDENTIFIED AS HAVING DEMENTIA OR ALZHEIMER'S DISEASE



Source: Australian Bureau of Statistics Survey of Disability, Ageing, & Carers (2009); http://www.abs.gov.au/AUSSTATS/abs@.nsf/Lookup/4102.0Main+Features50Dec+2012

THE ‘NAPA’ & NTG

NATIONAL ALZHEIMER’S PROJECT ACT

THE 'NAPA'

The **National Alzheimer's Project Act** required the creation of a national strategic plan to address the rapidly escalating Alzheimer's disease crisis and calls for coordination of Alzheimer's disease research and caregiver support efforts by the federal government

- **National Alzheimer's Project Act** *(became law in early 2011)*
  - Requires DHHS to submit an annual Alzheimer's plan to Congress – from 2012 to 2025
- Administered by federal **Department on Health Human Services** (DHHS)
- **Advisory Council on Alzheimer's Research, Care, and Services**
  - Council composed of Presidential appointees and federal agency staff
  - Creates the **National Plan to Address Alzheimer's Disease** with annual updates

National Plan called for -- among other things….

☑ Issuance of practice guidelines for care and supports and expanded public education

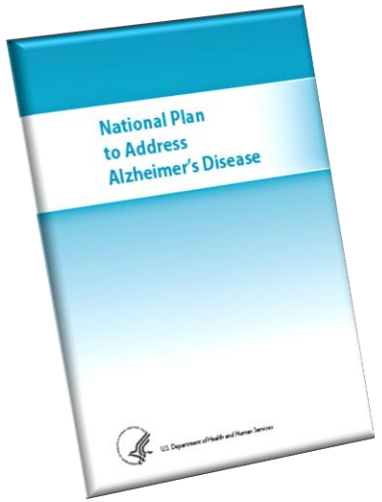
☑ Promotion of assessment tool for detection of cognitive impairment as part of the annual wellness visit

☑ Enhanced supports for caregivers

☑ Expanded research

☑ Special population focus - I/DD

First released on May 15, 2012
Will be updated annually until 2025!



IMPLICATIONS OF NAPA FOR PROVIDERS?

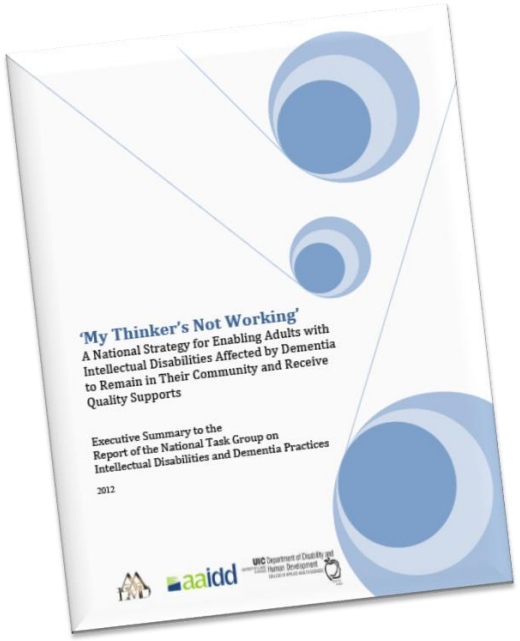
- **Tie-in to State Alzheimer's Plans' objectives**
  - *https://aspe.hhs.gov/pdf-document/national-plan-address-alzheimers-disease*
- **GWEPs* – enhancing the capacity of the workforce (working in dementia-related areas)**
  - *http://bhw.hrsa.gov/grants/geriatricsalliedhealth/index.html*
- **Potential implications of CMS' Setting Rule – Dementia housing**
  - *https://www.medicaid.gov/medicaid/hcbs/index.html*
- **CDC's Healthy Brain Initiative**
  - *http://www.cdc.gov/aging/healthybrain/index.htm*
- **Alzheimer's Disease Program Initiative – Annual funding call-out**
  - *http://www.aoa.acl.gov/AoA_Programs/HPW/Alz_Grants/*
  - ID-oriented grant projects funded in various states

*Geriatrics Workforce Enhancement Program

'My Thinker's Not Working'

National Task Group on Intellectual Disabilities and Dementia Practices

The **National Task Group** is a not-for-profit corporation charged to advocate, educate, provide technical assistance and program protocols, and guide public policy. Its members are composed of provider agency personnel, clinicians, academics, government officials, family members, and others.



Mission…

- ✓ To define best practices that can be used by agencies in delivering supports and services to adults with intellectual disabilities affected the various dementias
- ✓ To identify a workable national a 'first-instance' early detection / screening instrument
- ✓ To produce educational materials of use to families, people with ID, and providers of services
- ✓ To further public policy with respect to dementia as it affects adults with intellectual disabilities

The NTG is associated with the National Down Syndrome Society, is part of the LEAD Coalition in Washington, and has connections with university aging programs and community organizations.

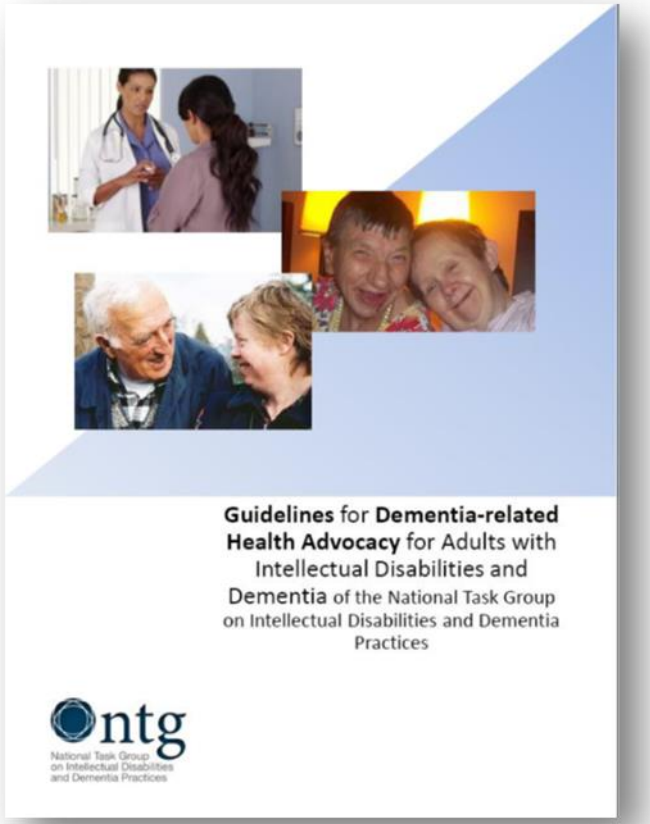
www.the-ntg.org

THE FUNCTIONS OF THE 'NTG'

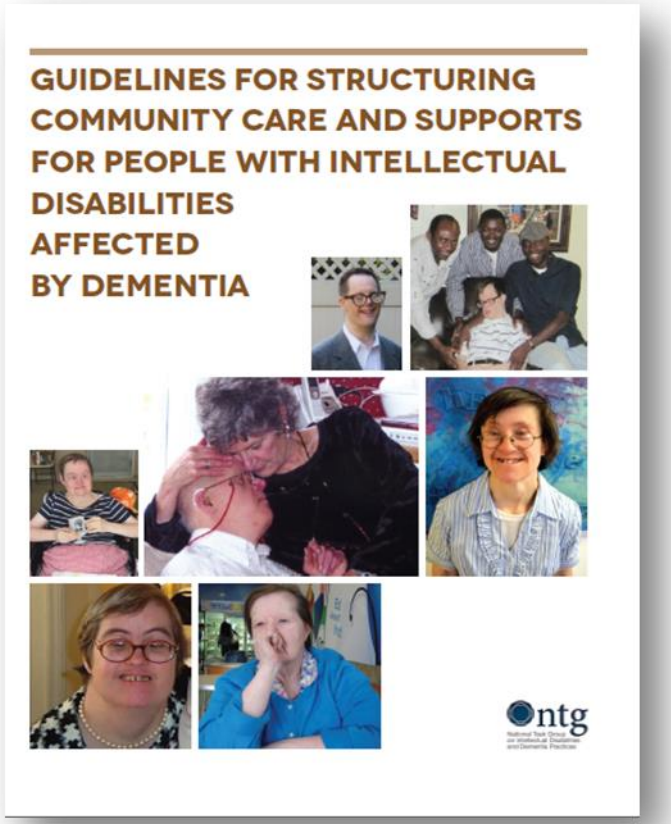
- Advocacy
- Education & training
- Family aids
- Policy
- Information dissemination
- Diagnostics and assessment
- National and international connections

www.the-ntg.org

NTG GUIDELINES



Health advocacy



Community living & supports

Diagnostics and medical care

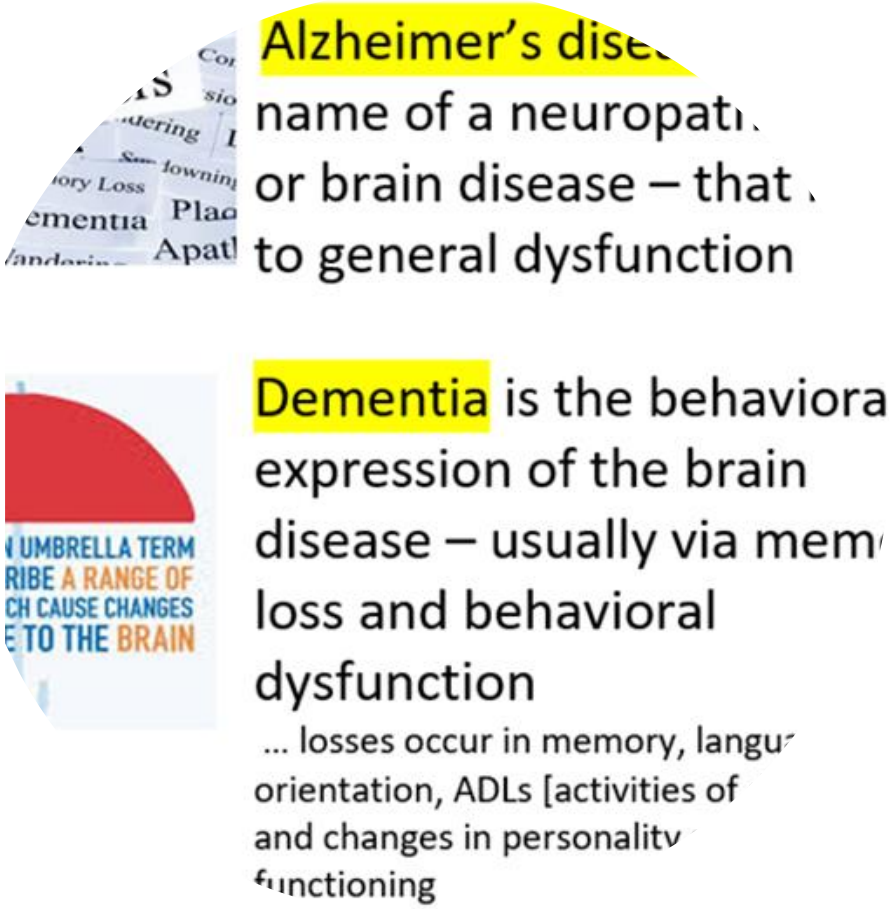
KEY FEATURES AND ISSUES

DEMENTIA PROCESSES AND INTELLECTUAL DISABILITY

WHY SOMETHING TO THINK ABOUT?

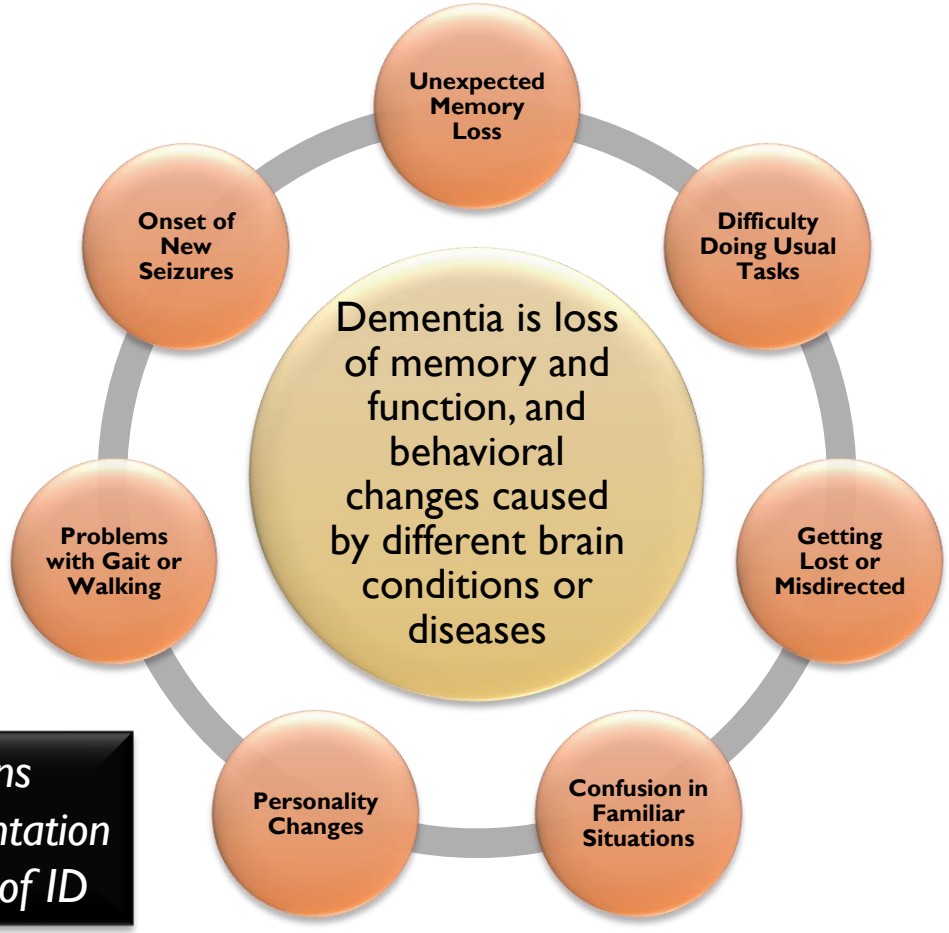
- **Dementia is the result of a brain disease or injury**, such as Alzheimer's disease, Lewy body disease, or a brain injury or trauma
- With progression an adult with dementia is **increasingly less able to take care** of him or herself … and requires supervision and someone to help him or her with necessities
- Main **dementia care options** for most agencies are to support the person in place (whether at home or in their residential accommodation), refer to a long-term care facility, or admit to a specialty dementia-capable group home
- Dealing with dementia calls upon agencies to make some **critical decisions** about dementia care and developing support resources

THINGS TO KNOW ABOUT DEMENTIA



- **Dementia an umbrella term** for a range of changes in behavior and function affecting aging adults and usually linked to brain disease (e.g., Alzheimer's) or injury (e.g., stroke)
  - Alzheimer's is **a disease of the brain** – dementia describes the resulting behavior
  - Most adults with Down syndrome (DS) are at high **risk of Alzheimer's disease** and consequently dementia; same risk as general population for adults with other ID
  - **Average age of 'onset'** in Down syndrome is about **53** and +60s/-70s for ID; Alzheimer's begins some 20 years before 'onset'
  - **Changes in memory** often signal dementia in ID; changes in personality often signal dementia in DS
  - After diagnosis **progressive decline in DS** can last for from 1 to 7+ years; up to 20 years in other ID
  - Care after the early stage can become more challenging as memory, self-care, communication, and walking become more difficult… eventually leads to **advanced dementia**

DEMENTIA-RELATED FUNCTIONAL CHANGE



These problems must be notable and usually occur in a cluster

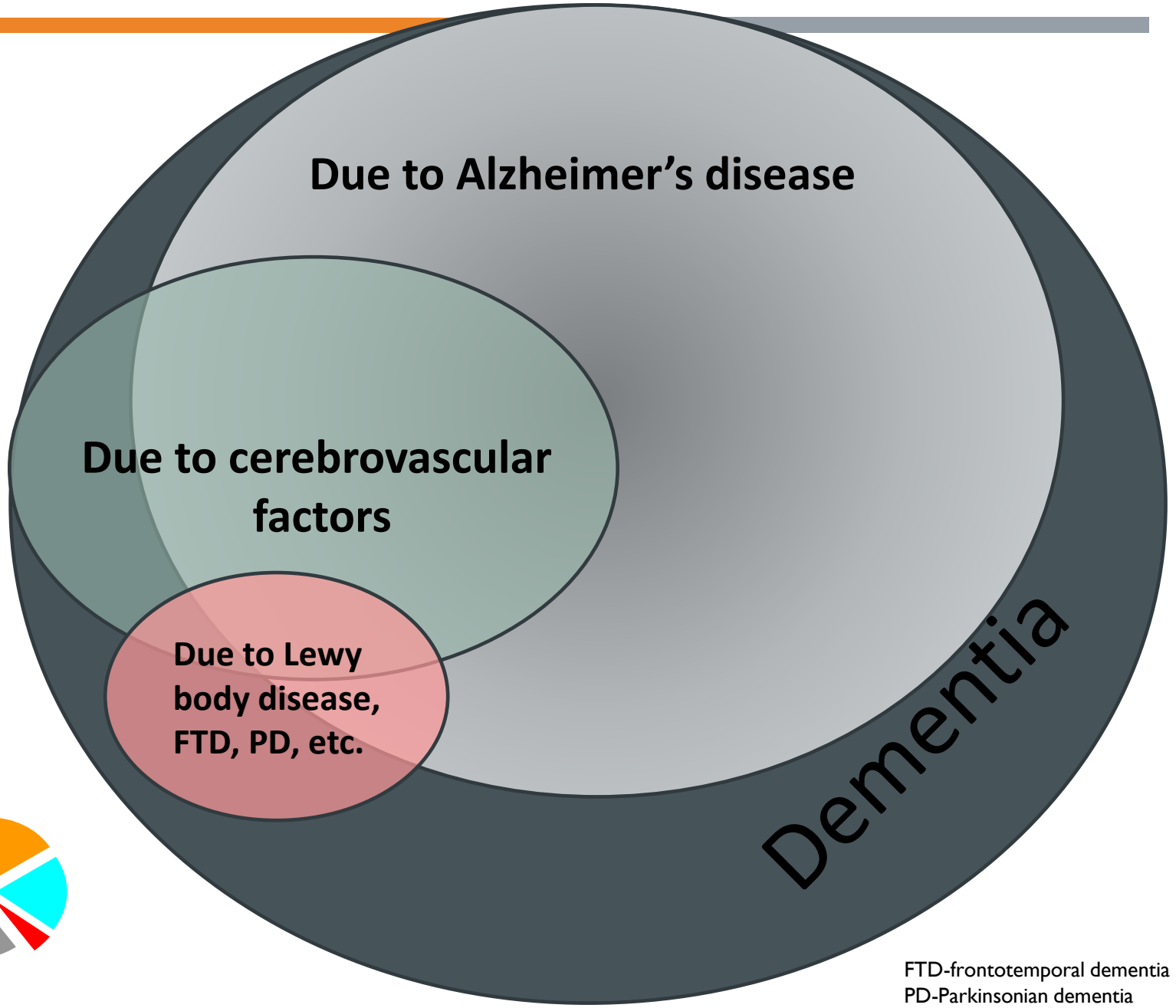
In ID, similar signs but of varying presentation depending on level of ID

Alzheimer's disease pathology often co-occurs with other pathologies, particularly cerebrovascular pathology

The effects of these different pathologies are additive, and may at least interact

At later ages, mixed dementia is very common, and additional pathologies remain to be identified

Pie chart illustrations can be misleading…



Source: Blacker, D. (2020) [graphic from Weuve, J.]

FTD-frontotemporal dementia
PD-Parkinsonian dementia

KEY WARNING SIGNS OF DEMENTIA IN DOWN SYNDROME

- Adults with Down syndrome are at **high risk** for Alzheimer's disease and dementia
- Researchers are finding that the first signs of Alzheimer's disease, some new changes to brain cells (the 'plaques and tangles'), occur some **20 years before** behavioral changes will be noticed
- Researchers have also found that adults with Down syndrome **show early symptoms** in a different way…
  - Noticeable first are changes in **personality** and in **general decision making**, then in memory (in contrast memory is usually affected first in other people)

- Abrupt **onset of seizures** when there had been none in the past
- **Incontinence** when an individual has always been toileting appropriately
- **Sleep/wake cycle** changes or disruptions
- Loss of sociability – a noticeable **change in personality**

KEY DIFFERENCES IN ADULTS WITH INTELLECTUAL DISABILITY

Some adults have early onset and shorter duration

- Younger-age (or early) onset is found in adults with Down syndrome and head injury
- Most adults with Down syndrome survive less than 7 years after the onset of dementia

Some differences in symptom presentation

- Most early symptoms are the same, except in Down syndrome where there are more notable early personality changes

Assessments are conducted differently

- Standard tests used with typical adults with dementia are not useful – With adults with intellectual disability need to use comparisons of the same individual over time

Differentiating factors - ID and dementia

Rate of occurrence[1,3,5,8,14]	➢ Age-cohort percent for adults with ID is same as in general population (~5-6% over 60) ➢ Much higher prevalence (60% >age 60) and neuropathology indicative of AD in most adults with Down syndrome (DS)
Dementia type[2,9]	➢ Generally, dementia of the Alzheimer's type is prevalent in DS ➢ Similar range of dementias found in other ID as in other people
Risk[15]	➢ DS and head trauma are significant risk factors in ID
Onset and duration[1,2,3,10]	➢ Average onset age in early 50s for DS – late 60s for others ➢ Most DAT diagnosed within 3 years of "onset" in adults with DS
Behavioral changes[2,3,6,11,12,13]	➢ In DS - early change in personality more evident ➢ In other ID - initial memory loss more evident ➢ Notable changes in behavior - apathy, sleep disturbance, agitation, incontinence, uncooperativeness, irritability, aggressiveness
Neurological signs[1,2,4,7,16,17,18,19]	➢ Late onset seizures in 24%-53% of adults w/DS ➢ Late onset seizures in DS indicator of life expectancy of less than 2 years, probable death within 3 years, and death almost invariably within 5 years of onset ➢ Seizures more common at end-stage (84.0%) versus at mid-stage Alzheimer's disease (39.4%)
Prognosis[2, 17]	➢ Aggressive AD can lead to death <2 years of onset in DS ➢ 2-7+ years mean duration in DS; probable death within 3 years, and death usually within 5 years of onset ➢ Same duration expected among other ID as in other people

Sources: [1]Janicki, M.P. & Dalton, A.J. (2000). Prevalence of dementia and impact on intellectual disability services. Mental Retardation,38, 277-289. [2]Janicki, M.P., & Dalton, A.J. (1999). Dementia, Aging, and Intellectual Disabilities: A Handbook. Philadelphia: Brunner-Mazel; [3]Bush, A., & Beail, N. (2004). Risk factors for dementia and Down syndrome. AJMR, 109, 83-97. [4] Menendez M. (2005). Down syndrome, Alzheimer's disease and seizures. Brain Development, 27(4), 246-252. [5]Zigman, W.B., Schupf, N., Devenny, D., et al. (2004). Incidence and prevalence of dementia in elderly adults with MR without DS, AJMR, 109, 126-141. [6]Ball, S.L., Holland, A.J., Hon, J., Huppert, F.A., Treppner, P., & Watson, P.C. Personality and behavior changes mark the early stages of Alzheimer's disease in adults with Down's syndrome: findings from a prospective population-based study. International Journal of Geriatric Psychiatry, 2006, [Jun 26]. [7]Crespel, A., Gonzalez, V., Coubes, P., & Gelisse, P. (2007). Senile myoclonic epilepsy of Genton: Two cases in Down syndrome with dementia and late onset epilepsy. Epilepsy Research, 77,165-168. [8]Evenhuis, H. (1997). The natural history of dementia in ageing people with intellectual disabilities. JIDR. 41(1), 92-96. [9]Strydom, A., Livingston, G., King, M., & Hassiotis, A. (2007).Prevalence of dementia in ID using different diagnostic criteria. Br. J. Psychiatry, 191, 150-157. [10] Margallo-Lana et al., (2007). Fifteen year follow-up of 92 hospitalized adults with DS. JIDR, 51, 463-477. [11]Gianpietro, N. (2013). Research in dementia in Down syndrome. Presentation at the SR Congresso Internazionale sulla Sindrome di Down, Roma, Italy, November 9, 2013. [12]Deb S. , Hare M., & Prior L. (2007). Symptoms of dementia among adults with Down's syndrome: a qualitative study. Journal of intellectual disability Research , 51(9), 726-739. [13]Cooper, S-A. (1997). A population-based health survey of maladaptive behaviours associated with dementia in elderly people with learning disabilities. Journal of Intellectual Disability Research, 41(6), 481-487. [14] Torr, J.,& Davis, R. (2007). Ageing and mental health problems in people with intellectual disability. Current Opinion in Psychiatry, 20(5), 467-471. [15]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 Clinic Proceedings, 88(8), 831-840. [16] Esbenson, A.J., (2010). Health conditions associated with aging and end of life of adults with Down syndrome. Int Rev Res Ment Retard. 2010 ; 39(C): 107–126. [17] Prasher, V.P. & Corbett, J.A> (1993). Onset ofseizures as a poor indicator of longevity in people with down syndrome and dementia. International Journal of Geriatric Psychiatry, 8(11), 923–927. [18] Robertson, Hatton, Emerson, & Baines. (2015). Prevalence of epilepsy among people with intellectual disabilities: A systematic review. Seizure, 29, 46–62. [19] McCarron M, Gill M, McCallion P, Begley C. (2005). Health co-morbidities in ageing persons with Down syndrome and Alzheimer's dementia. J Intellect Disabil Res., 49, 560–566.
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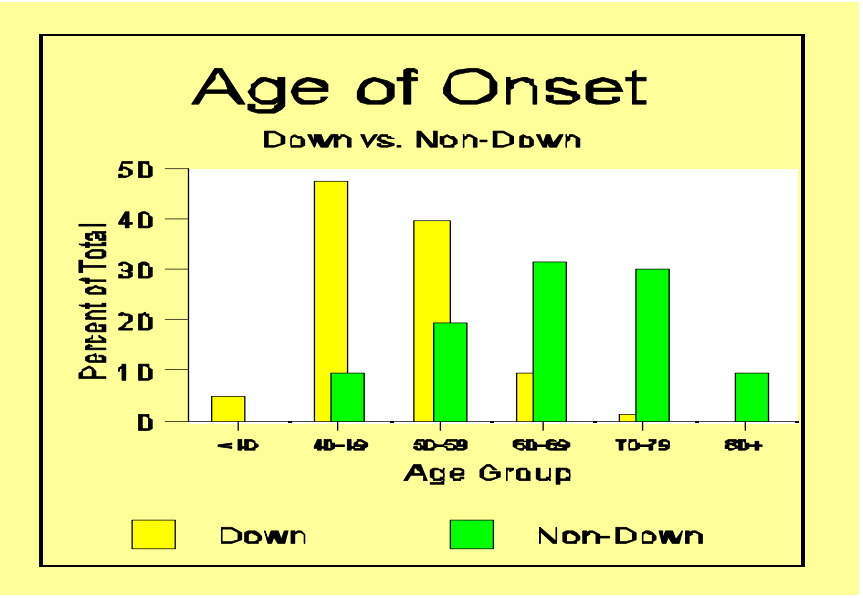
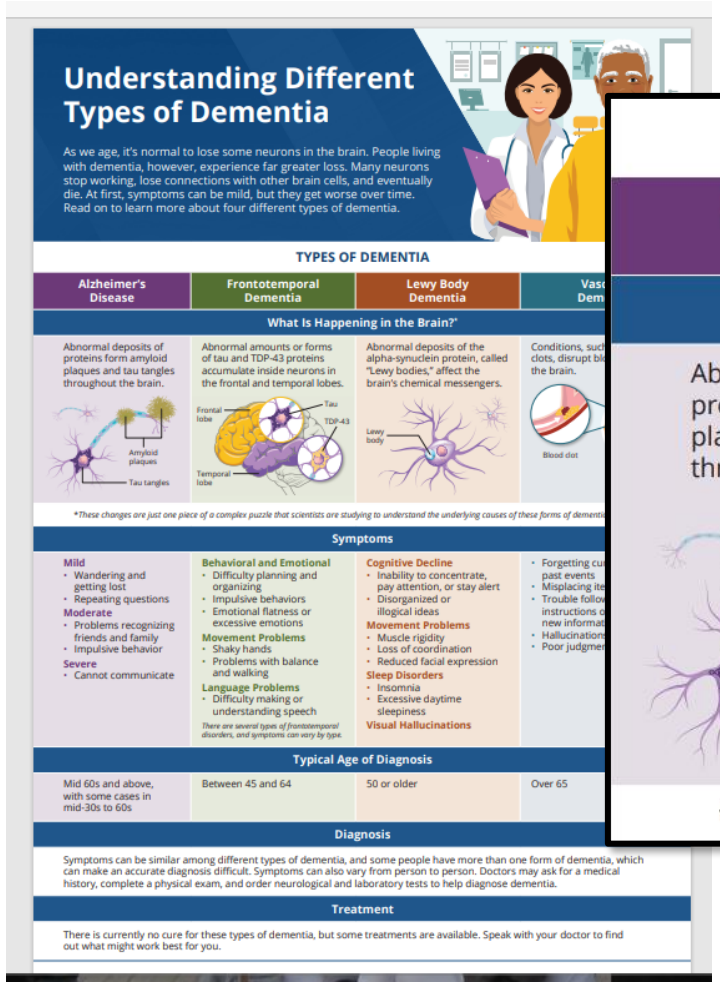
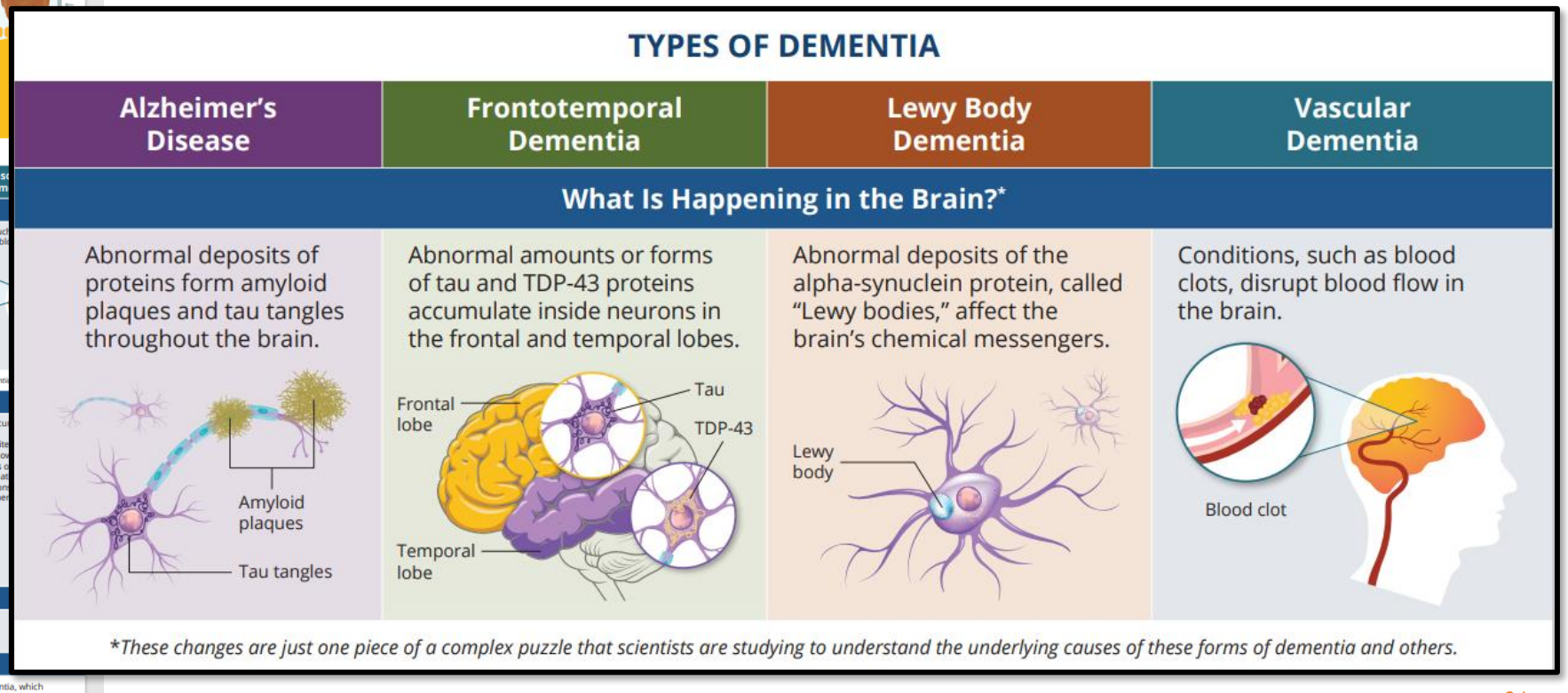
WHY IS RECOGNITION OF 'ONSET' IMPORTANT?

- **Knowing expected onset gives a 'head's-up' for initiating surveillance**
  - Look for changes
  - Introduce periodic screening
  - Alert staff to be watchful
  - Provides for an 'index of suspicion'
- **Helps us to begin to reformulate services and care practices**
  - Creating safer environments
  - Introducing cues for movement and way-finding
  - Engaging in planning ahead for eventualities
  - Setting goals for terms of service – adapting personal program plans

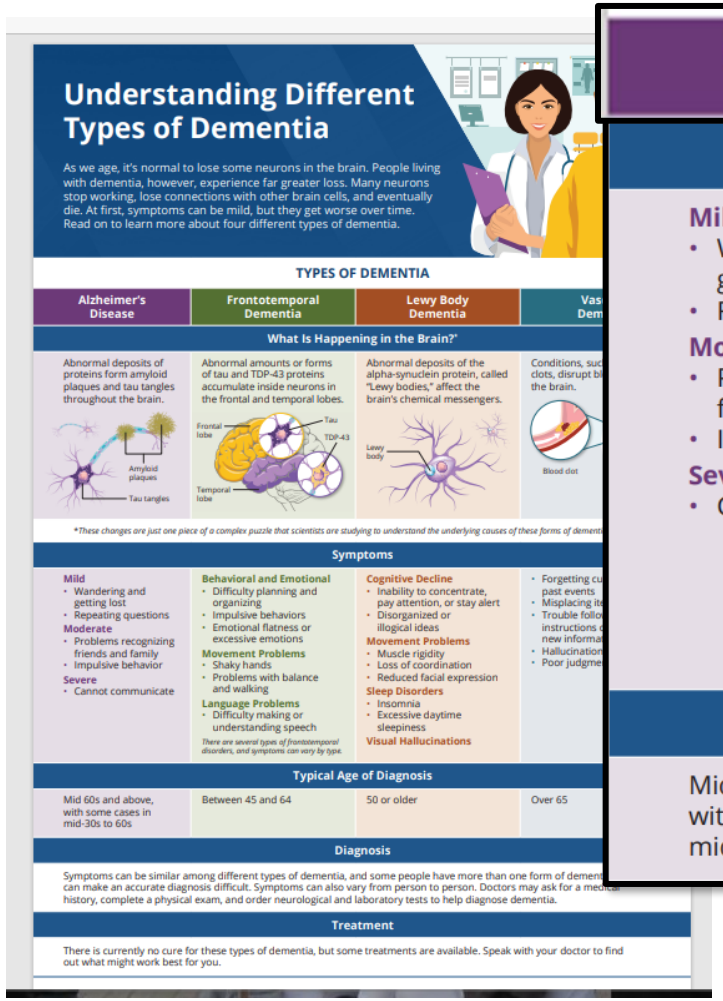


TYPES OF DEMENTIA





SYMPTOMS BY TYPE OF DEMENTIA



Understanding Different Types of Dementia

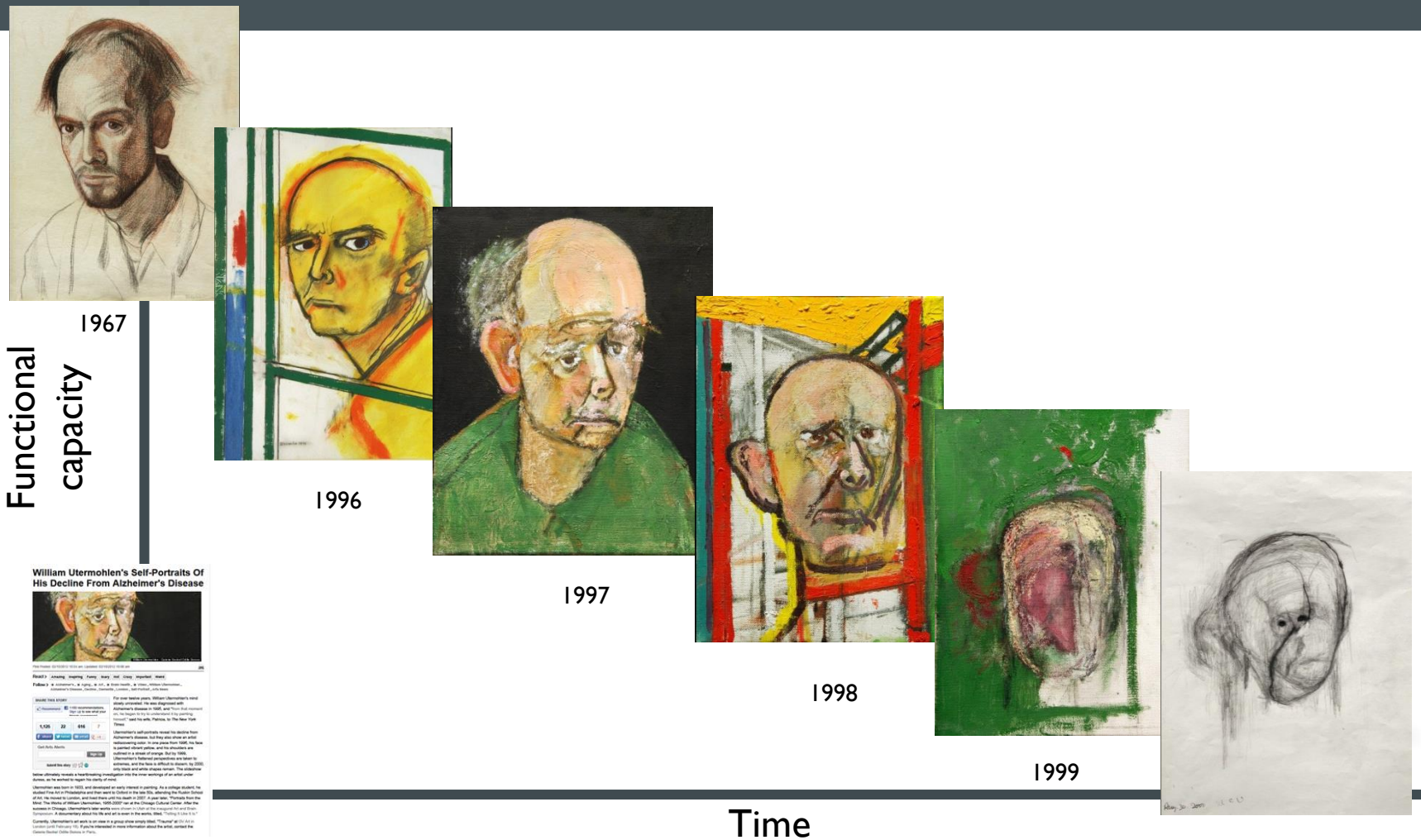
As we age, it's normal to lose some neurons in the brain. People living with dementia, however, experience far greater loss. Many neurons stop working, lose connections with other brain cells, and eventually die. At first, symptoms can be mild, but they get worse over time. Read on to learn more about four different types of dementia.

TYPES OF DEMENTIA

Alzheimer's Disease	Frontotemporal Dementia	Lewy Body Dementia	Vas Dem
What Is Happening in the Brain?*			
Abnormal deposits of proteins form amyloid plaques and tau tangles throughout the brain.	Abnormal amounts or forms of tau and TDP-43 proteins accumulate inside neurons in the frontal and temporal lobes.	Abnormal deposits of the alpha-synuclein protein, called "Lewy bodies," affect the brain's chemical messengers.	Conditions, suc clots, disrupt bl the brain.
Amyloid plaques; Tau tangles	Frontal lobe; Tau; TDP-43; Temporal lobe	Lewy body	Blood clot
*These changes are just one piece of a complex puzzle that scientists are studying to understand the underlying causes of these forms of dement			
Symptoms			
Mild • Wandering and getting lost • Repeating questions **Moderate** • Problems recognizing friends and family • Impulsive behavior **Severe** • Cannot communicate	**Behavioral and Emotional** • Difficulty planning and organizing • Impulsive behaviors • Emotional flatness or excessive emotions **Movement Problems** • Shaky hands • Problems with balance and walking **Language Problems** • Difficulty making or understanding speech *There are several types of frontotemporal disorders, and symptoms can vary by type.*	**Cognitive Decline** • Inability to concentrate, pay attention, or stay alert • Disorganized or illogical ideas **Movement Problems** • Muscle rigidity • Loss of coordination • Reduced facial expression **Sleep Disorders** • Insomnia • Excessive daytime sleepiness **Visual Hallucinations**	• Forgetting cu past events • Misplacing it • Trouble follo instructions new informa • Hallucination • Poor judgme
Typical Age of Diagnosis			
Mid 60s and above, with some cases in mid-30s to 60s	Between 45 and 64	50 or older	Over 65
Diagnosis			
Symptoms can be similar among different types of dementia, and some people have more than one form of dement can make an accurate diagnosis difficult. Symptoms can also vary from person to person. Doctors may ask for a medica history, complete a physical exam, and order neurological and laboratory tests to help diagnose dementia.			
Treatment			
There is currently no cure for these types of dementia, but some treatments are available. Speak with your doctor to find out what might work best for you.			

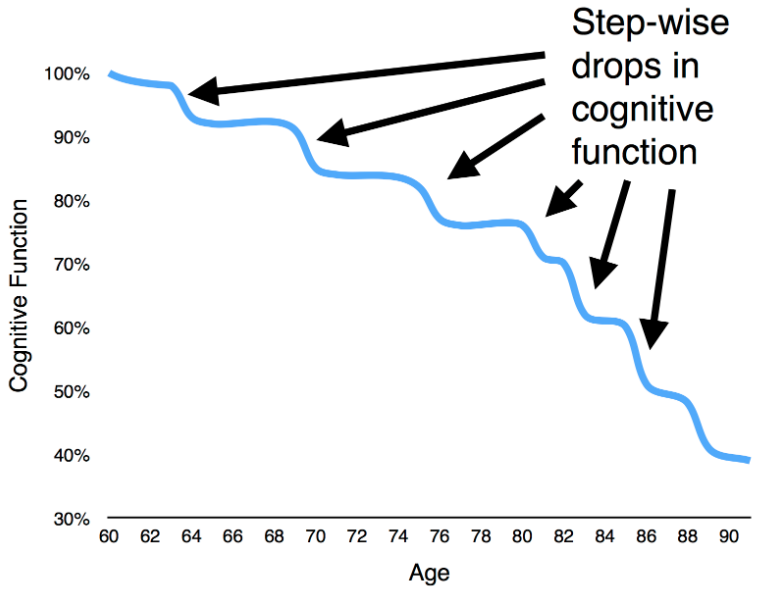
Alzheimer's Disease	Frontotemporal Dementia	Lewy Body Dementia	Vascular Dementia
Symptoms			
Mild • Wandering and getting lost • Repeating questions **Moderate** • Problems recognizing friends and family • Impulsive behavior **Severe** • Cannot communicate	**Behavioral and Emotional** • Difficulty planning and organizing • Impulsive behaviors • Emotional flatness or excessive emotions **Movement Problems** • Shaky hands • Problems with balance and walking **Language Problems** • Difficulty making or understanding speech *There are several types of frontotemporal disorders, and symptoms can vary by type.*	**Cognitive Decline** • Inability to concentrate, pay attention, or stay alert • Disorganized or illogical ideas **Movement Problems** • Muscle rigidity • Loss of coordination • Reduced facial expression **Sleep Disorders** • Insomnia • Excessive daytime sleepiness **Visual Hallucinations**	• Forgetting current or past events • Misplacing items • Trouble following instructions or learning new information • Hallucinations or delusions • Poor judgment
Typical Age of Diagnosis			
Mid 60s and above, with some cases in mid-30s to 60s	Between 45 and 64	50 or older	Over 65

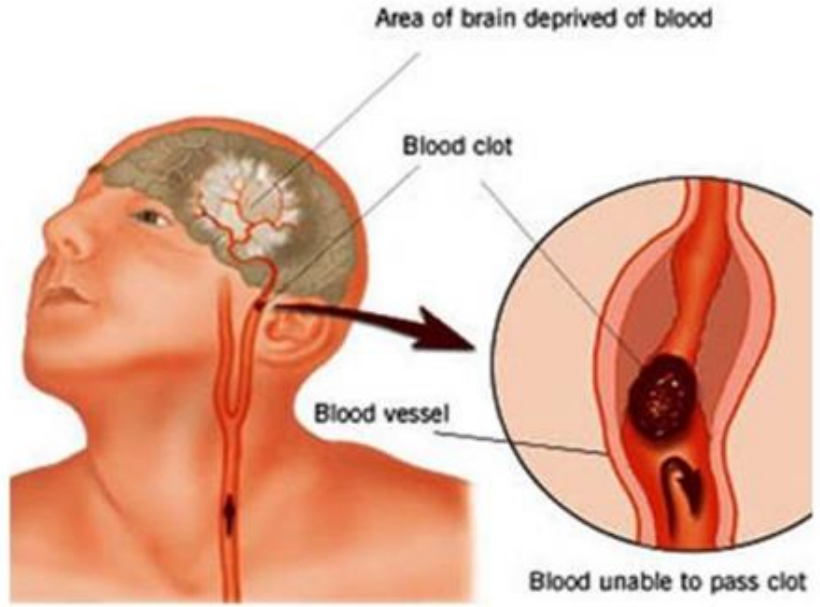
PROGRESSIVE COGNITIVE DETERIORATION DUE TO ALZHEIMER'S DISEASE



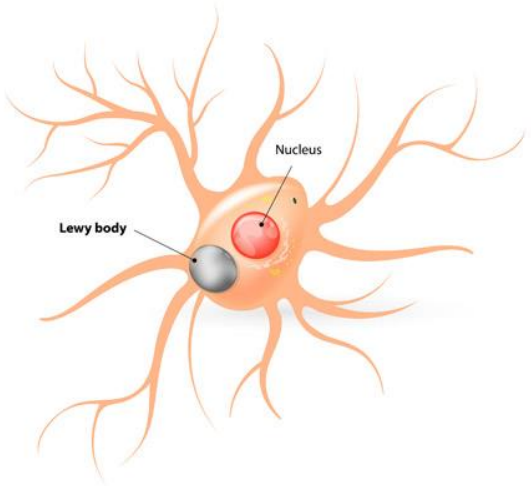
Source: http://www.huffingtonpost.com/2012/02/09/william-utermohlens-self-portraits_n_1265712.html

STEP-WISE PROGRESSION OF VASCULAR DEMENTIA



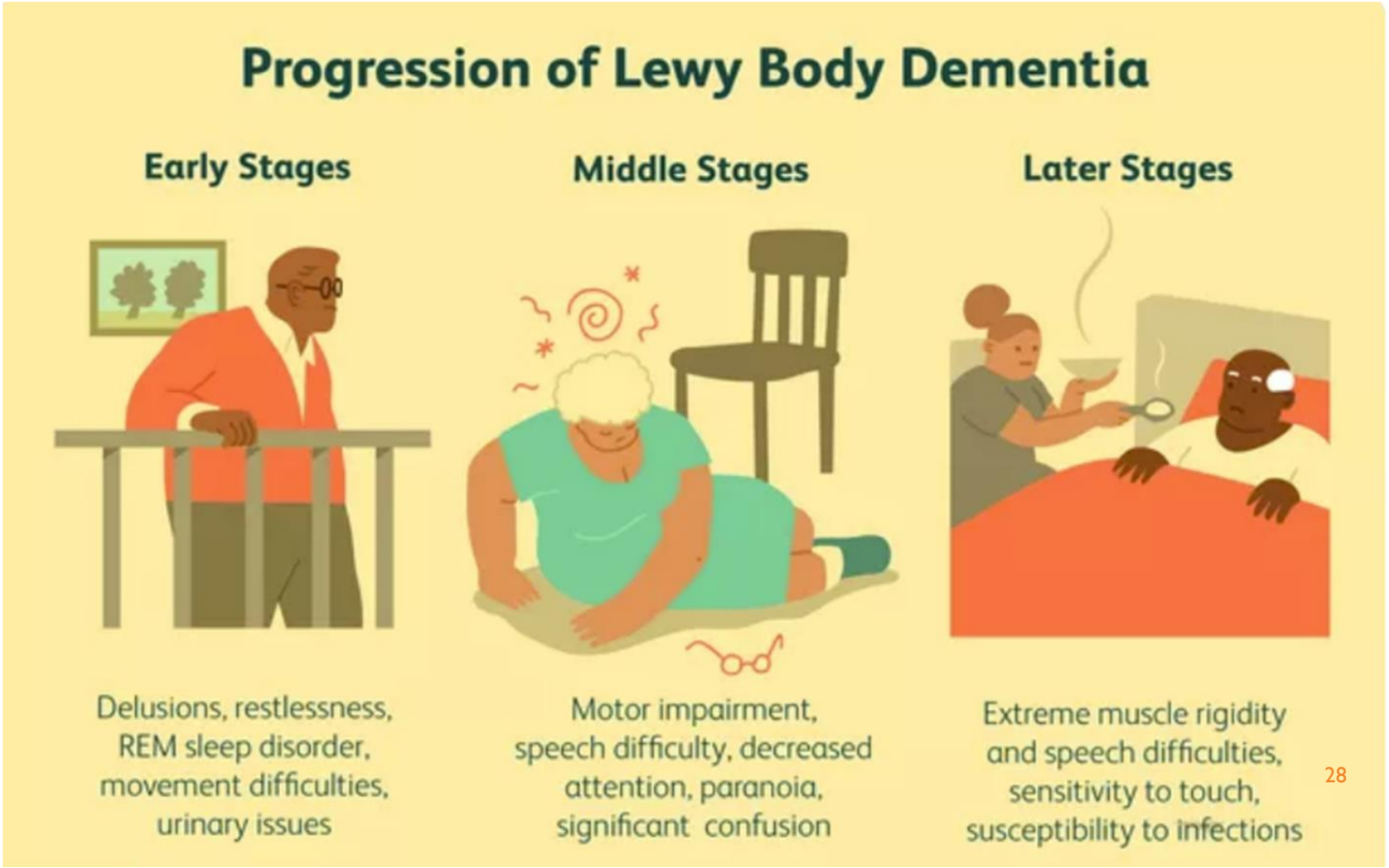


LEWY BODY DEMENTIA



Men more at risk

Caused by build-up of Lewy Body proteins in the brain



FRONTO-TEMPORAL DEMENTIA

speech

personality

Signs and symptoms of frontotemporal dementia

- Changes in personality. The person may become quiet and withdrawn, or even unusually cheerful and outgoing
- Emotional changes, such as seeming cold and distant

- Difficulty understanding what is considered appropriate behavior, or seemingly having 'no filter'
- Change in language, such as words coming out in the wrong order
- Becoming easily distracted or confused, they may feel easily overwhelmed when trying to process information from their senses

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- Difficulty with movement
- Repetitive behavior, such as obsessive cleaning, collecting or exercising

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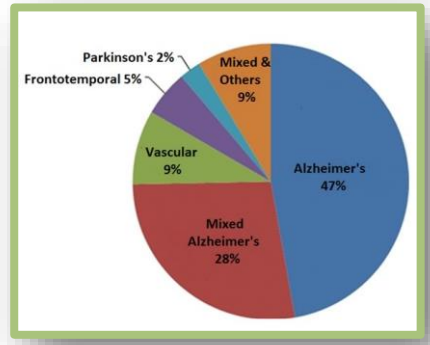
- Not looking after themselves
- Binge eating. *The person will recognize that they are full but not stop eating, because their brain doesn't tell them to.*

TYPE OF DEMENTIA CAN INFLUENCE CARE PLANNING

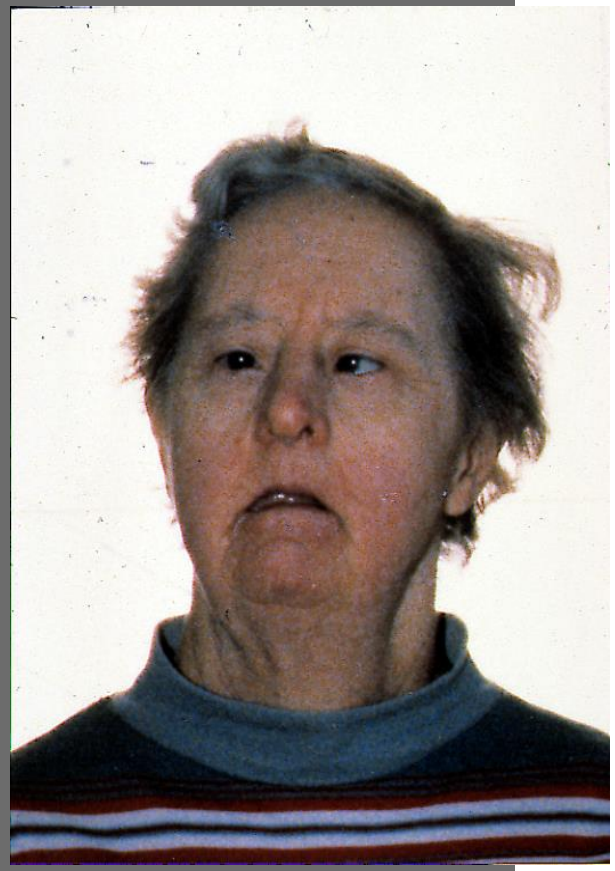
- Most persons with Down syndrome will have dementia of the Alzheimer's type – caused by Alzheimer's disease
- Persons with ID may have a variety of dementias (in norm with the general population)

- Why is it important (or useful) to know type?
  - To determine 'course of treatment' and expectations of staging and rate of decline
  - To help with determining best ways to handle 'challenging behaviors'
  - To help with organizing staffing patterns and clinical supports



RUBY

Ruby at age 62



Courtesy: A.J. Dalton (2000)

Sign or Symptom	Age
Early	
Impaired memory function	54.7
Impaired learning abilities	56.7
Hearing loss	57.0
Disorientation	58.0
Hypothyroidism	59.0
Middle	
Personality changes	60.5
Deterioration of ADL skills	63.0
Abnormal reflexes	64.5
Late	
Hallucinations	64.5
Seizures	65.0
Incontinence	65.4
Has to be fed	65.4
Apathy	65.4
Complete care required	65.4
Death	65.5

Ruby spent most of her life in a large congregate care institution … back in the 80s.

Contemporary practices would have offered her a different life and opportunities… specially when dealing with her decline and eventual succumbing to dementia.

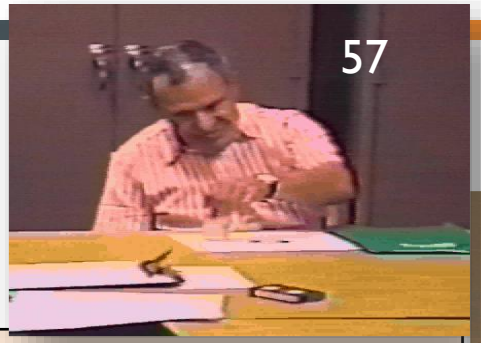
Ruby's decline illustrates a typical progression of stage associated losses of function, onset of comorbidities, and aging

TERMINOLOGY

- Mild cognitive impairment (MCI)
- Early onset dementia
- Early-stage dementia
- Mid-stage dementia
- Late-stage or advanced dementia

Is it "Possible, Probable, vs. Definite dementia"?

General stage durations for Alzheimer's dementia in typical adults



EARLY or MILD STAGE

2 to 4 years or longer

MID- or MODERATE STAGE

2 to 10 years

LATE or SEVERE STAGE

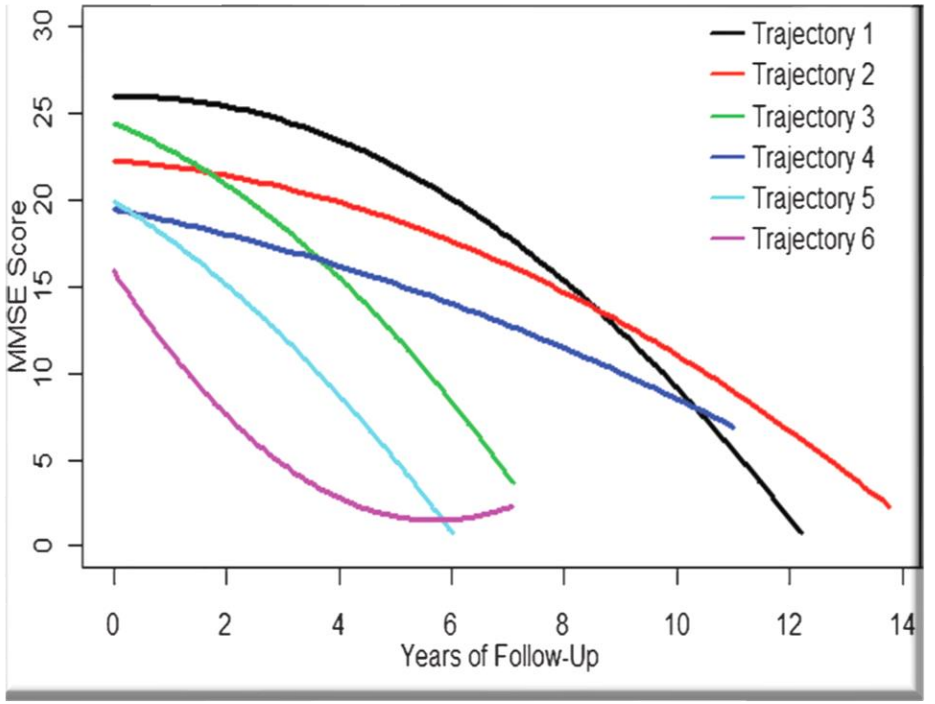
1 to 3 years of longer

For adults with Down syndrome there is compressed staging of Alzheimer's dementia

Early Stage	Middle Stage	Late Stage
Confusion and memory loss	Difficulties with ADLs ["activities of daily living"]	Loss of speech
Disorientation in space	Anxiety, paranoia, agitation and other compromising behaviors	Loss of appetite, weight loss
Problems with routine tasks	Sleep difficulties	Loss of bladder and bowel control
Changes in personality and judgment	Sleep difficulties	Loss of mobility
	Difficulty recognizing familiar people	Total dependence on others
		~Death

CRITICAL FACTORS

- Degree of retention of function
- Expected trajectory of progressive dysfunction
- Duration (remaining life years)
- Type of dementia
- Health status
- Environmental accommodations



Varying trajectories have implications for continual assessment and adaptations to care management

Source: Figure 1 from Wilkosz et al., (2009). Trajectories of cognitive decline in Alzheimer's disease. *International Psychogeriatrics*, 28,1-10

IMPLICATIONS OF TRAJECTORIES AND DURATION

- Knowing something about variations in trajectories
  - Anchors around potential duration of 'stay' at same level of functioning
  - Provides ideas about potential changes and their nature
  - Creates a schedule for timing changes in service orientation – planning care, evaluating patterns of care, and organizing staffing and environmental modification
  - Provides an empirical basis for expectations of co-morbidities
  - Gives staff information about anticipating changes
  - Helps with introducing ameliorative interventions or aids for day-to-day functioning
  - Long-term planning for care financing (budgeting for shifts in staff and housing)

FEATURES RELATED TO DEMENTIA

- Older adults with Down syndrome are at high risk of Alzheimer's disease
- Not every adult will show signs of dementia as he or she ages
- Age-associate decline may be due to aging and not dementia
- Institute baseline for ('personal best') functioning at age ~40 for Down and at ~60 for other ID
- Useful to know the signs of MCI* and dementia and keep track of capabilities after age 40
- Early detection screening useful to identify possible progression into MCI or dementia
- Early referral for assessment or diagnosis if signs present is advised (to rule out alternative bases for physical or cognitive changes)

*mild cognitive impairment

WHAT TO DO WHEN DEMENTIA IS SUSPECTED - ID?

Start with an administrative screen

- Capture visuals on functioning (preferably 'personal best')
  - *digital recording of behavior*
  - Screening instrument
- Observe if screen provides 'hits' on 'warning signs'

Refer for clinical assessment and diagnosis

- Clinicians reapply ID-specific measures to look for longitudinal patterns
- If evidence points to dementia-like symptoms, refers for full diagnostic evaluation (for differential dx)

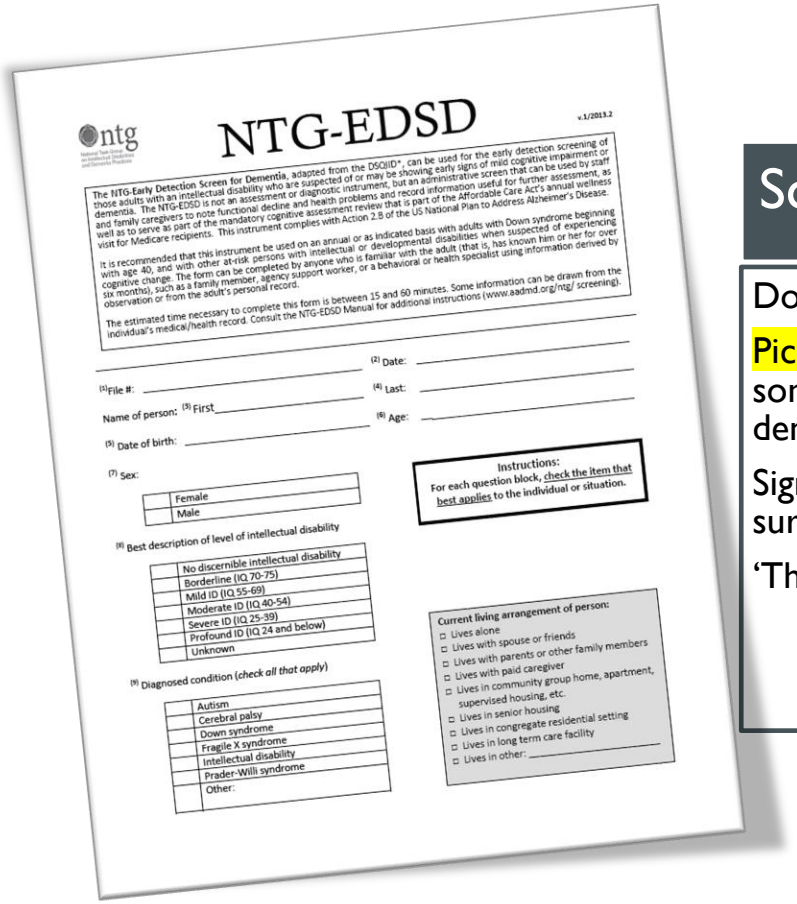
Post-diagnosis support

- Post-dx – decide on value of pharmacological tx
- Implement non-pharmacological strategies
- Support through stage changes

Who does assessments?

www.the-ntg.org

SCREENING-ASSESSMENT-DIAGNOSIS PROCESS



NTG-EDSD

v.1/2013.2

The NTG-Early Detection Screen for Dementia, adapted from the DSQIID*, can be used for the early detection screening of those adults with an intellectual disability who are suspected of or may be showing early signs of mild cognitive impairment or dementia. The NTG-EDSD is not an assessment or diagnostic instrument, but an administrative screen that can be used by staff and family caregivers to note functional decline and health problems and record information useful for further assessment, as well as to serve as part of the mandatory cognitive assessment review that is part of the Affordable Care Act's annual wellness visit for Medicare recipients. This instrument complies with Action 2.8 of the US National Plan to Address Alzheimer's Disease.

It is recommended that this instrument be used on an annual or as indicated basis with adults with Down syndrome beginning with age 40, and with other at-risk persons with intellectual or developmental disabilities when suspected of experiencing cognitive change. The form can be completed by anyone who is familiar with the adult (that is, has known him or her for over six months), such as a family member, agency support worker, or a behavioral or health specialist using information derived by observation or from the adult's personal record.

The estimated time necessary to complete this form is between 15 and 60 minutes. Some information can be drawn from the individual's medical/health record. Consult the NTG-EDSD Manual for additional instructions (www.aadmd.org/ntg/ screening).

(1) File #: (2) Date:

Name of person: (3) First (4) Last:

(5) Date of birth: (6) Age:

(7) Sex:

	Female
	Male

Instructions:
For each question block, check the item that best applies to the individual or situation.

(8) Best description of level of intellectual disability

	No discernible intellectual disability
	Borderline (IQ 70-75)
	Mild ID (IQ 55-69)
	Moderate ID (IQ 40-54)
	Severe ID (IQ 25-39)
	Profound ID (IQ 24 and below)
	Unknown

(9) Diagnosed condition (*check all that apply*)

	Autism
	Cerebral palsy
	Down syndrome
	Fragile X syndrome
	Intellectual disability
	Prader-Willi syndrome
	Other:

Current living arrangement of person:
- ☐ Lives alone
- ☐ Lives with spouse or friends
- ☐ Lives with parents or other family members
- ☐ Lives with paid caregiver
- ☐ Lives in community group home, apartment, supervised housing, etc.
- ☐ Lives in senior housing
- ☐ Lives in congregate residential setting
- ☐ Lives in long term care facility
- ☐ Lives in other: _____

www.the-ntg.org

Screening

Done by carers or staff

Picks up key problem areas – some associated with MCI or dementia – confirms suspicions

Signals areas for further surveillance by carers or staff

'The starting point'

Assessment

Form used by carers or staff to begin discussion about concerns with clinician

Clinician uses noted items for more in-depth assessment and tracking

Suspicions can be confirmed or revisited periodically

Neurological scans can be ordered

Diagnosis

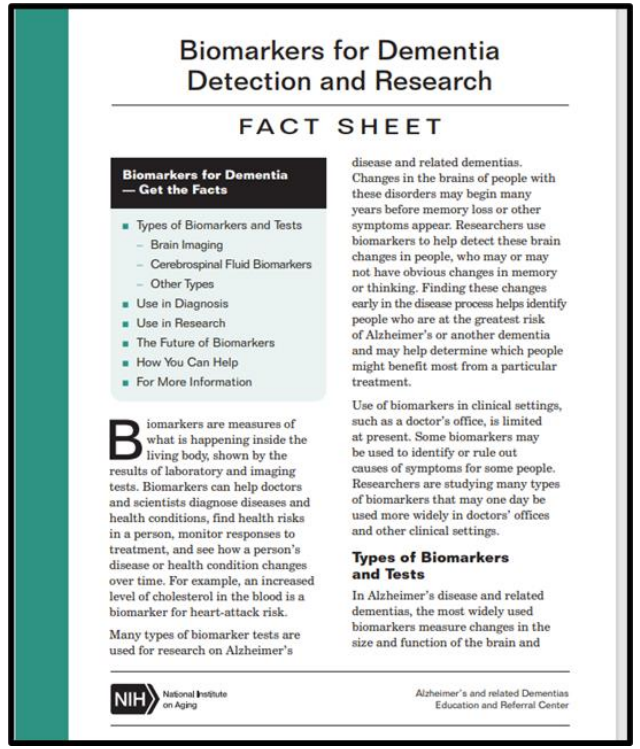
Clinic or clinicians ascertain whether dementia dx is viable

Eventual review of symptoms and application of range of general dementia ascertainment measures

Designation of possible or probable dementia and potentially clinical etiology

WHAT IS THE FUTURE IN DIAGNOSTICS

- The National Institute on Health has funded numerous studies of **biomarkers**, both within the general population and of adults with Down syndrome
- Biomarkers provide detailed **measures of abnormal changes in the brain**, which can aid in early detection of possible disease in people with very mild or unusual symptoms
- People with Alzheimer's disease and related dementias progress at different rates, and **biomarkers may help predict and monitor their progression**
- Given the difficulty of using traditional office-methods of diagnosing dementia in adults with intellectual disability, **biomarkers will offer a means of confirming a clinician's suspicions** or diagnostic outcomes



Biomarkers for Dementia Detection and Research

FACT SHEET

Biomarkers for Dementia — Get the Facts

- Types of Biomarkers and Tests
  - Brain Imaging
  - Cerebrospinal Fluid Biomarkers
  - Other Types
- Use in Diagnosis
- Use in Research
- The Future of Biomarkers
- How You Can Help
- For More Information

Biomarkers are measures of what is happening inside the living body, shown by the results of laboratory and imaging tests. Biomarkers can help doctors and scientists diagnose diseases and health conditions, find health risks in a person, monitor responses to treatment, and see how a person's disease or health condition changes over time. For example, an increased level of cholesterol in the blood is a biomarker for heart-attack risk.

Many types of biomarker tests are used for research on Alzheimer's disease and related dementias. Changes in the brains of people with these disorders may begin many years before memory loss or other symptoms appear. Researchers use biomarkers to help detect these brain changes in people, who may or may not have obvious changes in memory or thinking. Finding these changes early in the disease process helps identify people who are at the greatest risk of Alzheimer's or another dementia and may help determine which people might benefit most from a particular treatment.

Use of biomarkers in clinical settings, such as a doctor's office, is limited at present. Some biomarkers may be used to identify or rule out causes of symptoms for some people. Researchers are studying many types of biomarkers that may one day be used more widely in doctors' offices and other clinical settings.

Types of Biomarkers and Tests

In Alzheimer's disease and related dementias, the most widely used biomarkers measure changes in the size and function of the brain and

NIH National Institute on Aging

Alzheimer's and related Dementias Education and Referral Center

New FDA approved Alzheimer's therapeutics

https://order.nia.nih.gov/sites/default/files/2020-07/biomarkers-dementia-research-508.pdf

EFFECTS OF DEMENTIA

SOME INFORMATION FOR SUPPORT SERVICES

UNDERSTANDING DEMENTIA

Knowns…

- People with ID have same rate of dementia as general population
- Some people with ID have higher rates (e.g., Down syndrome, head injury)
- Some % of any adult client pool will be affected
- Early interventions can aid in adapting to changes and prolonging lucid periods
- Effects of dementia will be progressive and eventually lead to death

Unknowns…

- Who will be affected?
- How pronounced will be early changes?
- How dramatic will be the changes in function?
- How long will person live after diagnosis?
- What other diseases or medical conditions may be co-incident?
- Which particular dementia-related behaviors will be more evident?

EXPECTATION OF CHANGE AND FACTORS IN ID AND DEMENTIA UNDERLYING HOUSING AND CARE PRACTICES

Expectations of change

- Cognitive skills will decline
- Support needs will increase
- Increase risks of falls, injuries
- Swallowing dysfunction, clots, pneumonia, bladder infections, nutritional deficiencies, seizures

Care factors

- Watch for signs of abuse and neglect (including self-neglect)
- Watch for signs of caregiver burn-out and stress at home … affected on adult's behavior
- Watch for advanced dementia and needs for end-of-life care (palliative care and hospice)

ID associated issues that extenuate these factors:

- Co-incident conditions that may affect gait, sensory faculties, and cognition
- Co-morbidities or diseases that may affect physiological functions
- Previously identified 'mental health' issue
- Late-onset seizures
- Precocious (early) aging effects
- Expressive language difficulties
- Nutritional deficiencies & diet inadequacies
- Presence of polypharmacy

WHAT ARE NEEDED SUPPORTS?

- Help for caregivers and the person
- Advanced planning for alternative care
- Diagnostic and intervention assistance
- Support groups for caregivers (family or staff)
- Dementia capable community housing (group homes)
- Respite for caregivers
- Health care and social supports

"… *people have, on average, six years of living independently once mild cognitive impairment starts.*"*

Dementia is a condition that lessens an adult's ability to be left alone – thus, living without supervision is not an option *as the condition progresses*

*https://www.bbc.com/news/health-63749586

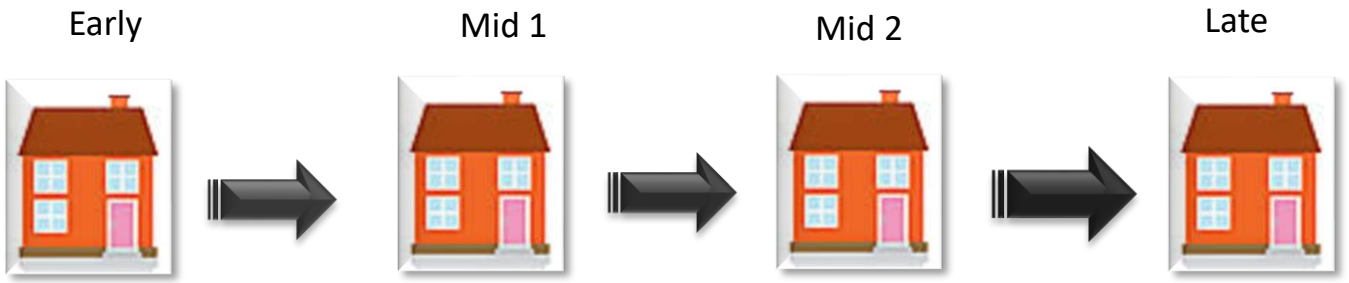
OPTIONS FOR DEMENTIA CARE SETTINGS

Staying	**Staying at home** • Continued care by family members until eventual advanced dementia and end-of-life • *Considerations*: home adaptation, close supervision for safety and avoiding self-harm or neglect 24/7, possible wheelchair use, palliative and/or hospice aid	**Agency focus** Outreach and community supports (HCBS) Helping support family caregivers
Leaving	**Leaving home** • Admission to a nursing facility after non-ambulatory care is necessary • *Consideration*: SNF capability & understanding of DS? • Looking for an agency run specialty dementia care group home • Other options – perhaps memory care centers, assisted living programs?	**Agency Focus** Securing housing with dementia specialty care Clinical team supports Training for staff

PREVALENT MODELS OF GROUP HOME-BASED DEMENTIA CARE

AGING-IN-PLACE

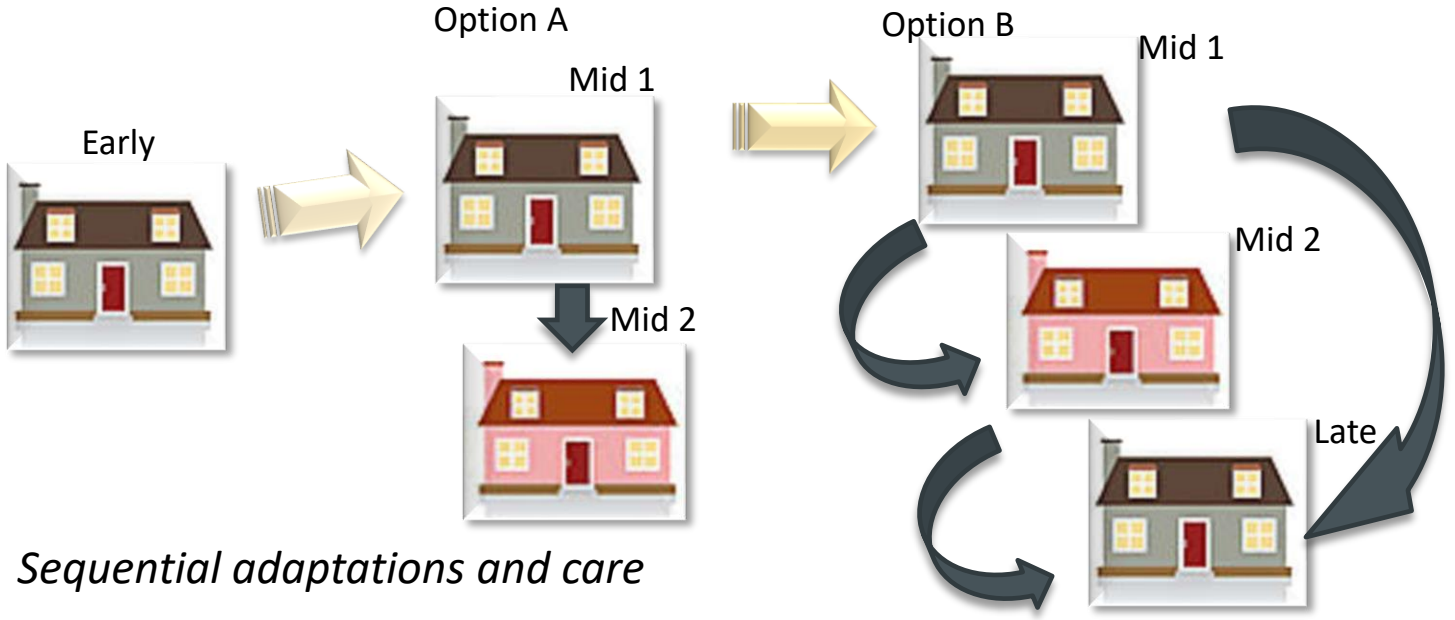
- *single care home and stable stay*



Linear adaptations and care

IN-PLACE-PROGRESSION

- *multiple care homes & movement with progression*



Sequential adaptations and care

Mid = mid-level

Source: JANICKI (2010)

WHAT'S IMPORTANT TO KNOW

TRAINING

- The difference between normal aging changes and pathological aging changes
- Early signs of functional change associated with dementia
- Types of dementia and their main characteristics, what will be the behavioral/functional changes, and their duration
- When is it best to refer for assessment and to whom
- What options exist for early dementia-related supports
- What options exist (or need to be put in place) for long-term dementia capable care/supports

Who should receive training and resource materials?

- **Direct support staff**
- **Clinicians**
- **Program managers**
- **Agency Admin personnel**

EARLY STAGE

Engage	Engage the individual and their family, and/or other carers or guardians in advance care **planning** (and prepare advance directives) consistent with state or other requirements.
Identify and plan	Identify and plan to remediate the **environmental challenges** to help maintain community living
Establish	Establish a **daily regime** that provides for purposeful engagement based on individual needs and preferences, yet is organized so as not to cause anxiety and confusion
Provide	Provide ongoing **clinical supports** to address behavioral and psychological symptoms associated with dementia
Redesign	Redesign **day activities and programs** so that participation in valued activities and opportunities for interaction with others continues and respite for families and other caregivers is possible

MID STAGE

- Provide increased assistance with **personal care and hygiene** when needed
- Secure appropriate residential supports and consider housing options to **accommodate increasing losses** in independent functioning
- Continue surveillance and periodic **assessments** to determine extent of change and progressive dysfunction as well as the possible development of comorbid conditions
- Monitor any **medications** being taken to prevent ADRs
- Enhance **training of staff and family** as well as consultation to carers around coping with behaviors and adapting routines
- Institute planning for **long-term services** and supports
- Ensure **protections are in place** to preclude abuse or harm in both formal and informal settings.

(Source: Jokinen et al., 2013)

LATE / END STAGE

- Reorganize care management toward **nonambulatory care**
- Reassign staff to activities more structured around nursing and **personal care** including the support of family carers who wish to maintain the person at home
- Obtain support from **palliative care** or hospice specialists
- Institute procedures to maintain **dignity, comfort**, and **address pain** and symptom management
- Organize **end-of-life supports** and post-death arrangements

DEMENTIA CARE PLANS

Formal dementia care plans, organized with support team, family, and the person, enable continual supports and assistance to maintain maximum function and enhance quality of life… and ease pressures on caregivers

- Focus on knowledge of needs of individual and support system
- Core elements
  - Assessment and re-assessment of dementia and it impact on health and function
  - Daily living supports, including housing
  - Health reviews for long-standing conditions and any emerging co-incident conditions
  - Dietary and nutrition
  - Mobility and physical functions
  - Medications
  - Special needs
  - Aid to caregivers

WHAT CAN YOU DO?

- Improve understanding of aging and dementia
- Be alert to risk and early signs decline
- Adapt living environments to minimize risk
- Help with futures planning (health and social care)
- Aid families who are carers
- Enhance staff skills – training with respect to dementia
- Quality checks on services
- Provide stage-related services
- Plan for future growth of aging segment of population

KEY RESOURCE WWW.THE-NTG.ORG

- Matthew Janicki

  mjanicki@uic.edu

- www.the-ngt.org

MEDICAL AND HEALTH FACTORS IN ID AND DEMENTIA

SETH M. KELLER, MD
NEUROLOGY ASSOCIATES
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THOUGHTS ON SERVICES AND INDIVIDUAL CARE

THOMAS BUCKLEY, ED.D.
CARF INTERNATIONAL BOARD OF TRUSTEES
TUCSON, ARIZONA

DISCUSSION AND Q&A