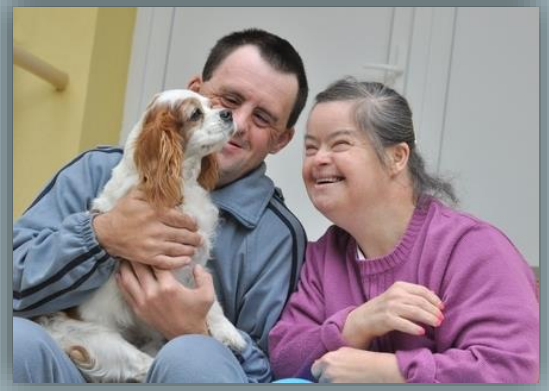
AGING IN ADULTS WITH INTELLECTUAL AND DEVELOPMENTAL DISABILITIES; CONCERNS AND HOPE



Seth M. Keller, MD

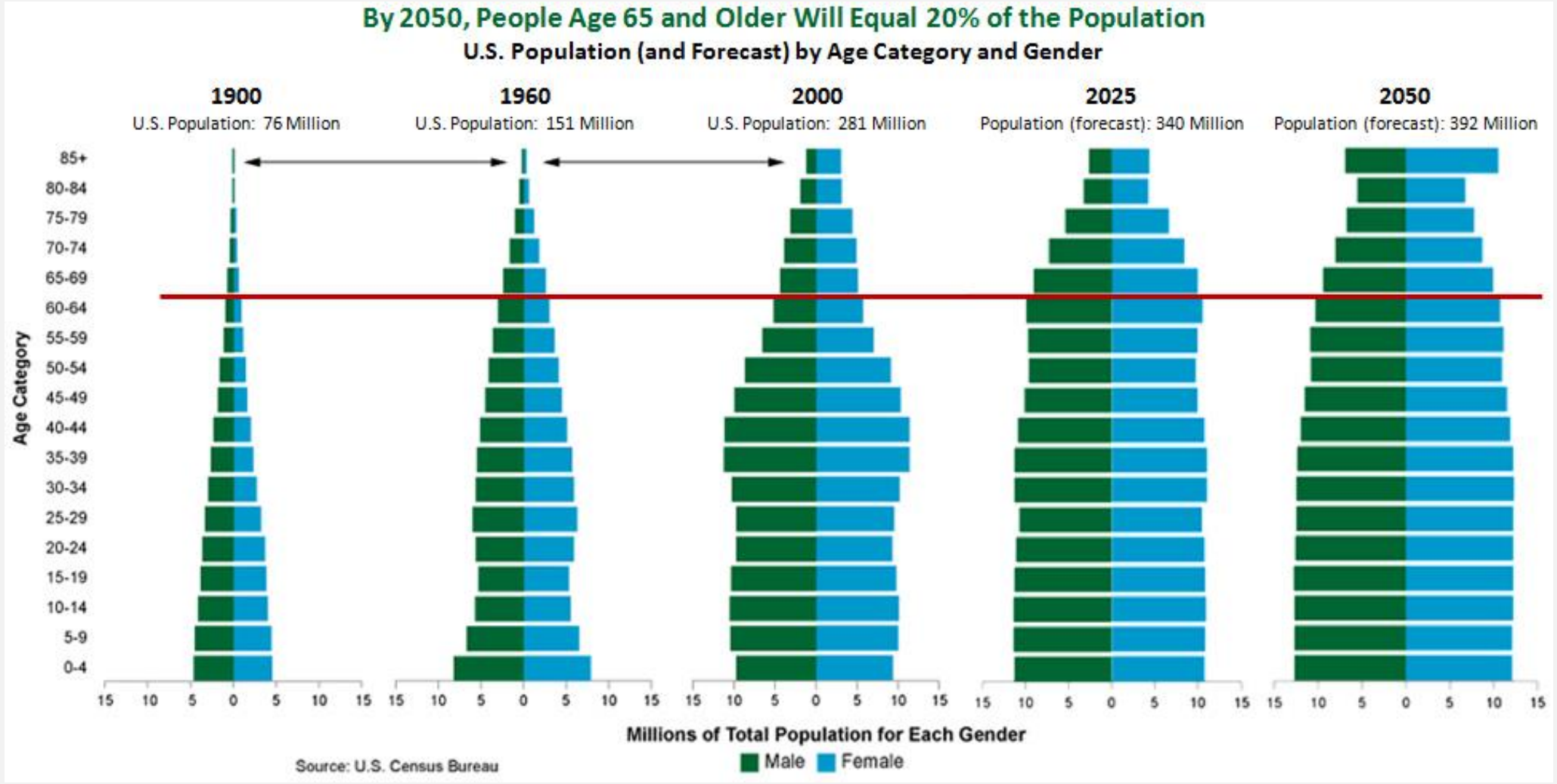
April 11, 2019

AADMD

Down Syndrome Alliance of the Midlands

ntg
National Task Group on Intellectual Disabilities and Dementia Practices

Changing US Population Demographics



AGING AND INTELLECTUAL AND DEVELOPMENTAL DISABILITIES

- In 2002, an estimated 641,000 adults with IDD were older than 60.
- In 2002 about 75% of all older adults with IDD were in the 40-60 year old age range.
- The number of adults with IDD age 60 years and older is projected to nearly double from 641,860 in 2000 to 1.2 million by 2030 due to increasing life expectancy and the aging of the baby boomer generation

LIFE EXPECTANCY

Carter & Jancar, 1983, Janicki, Dalton, Henderson, & Davidson, 1999

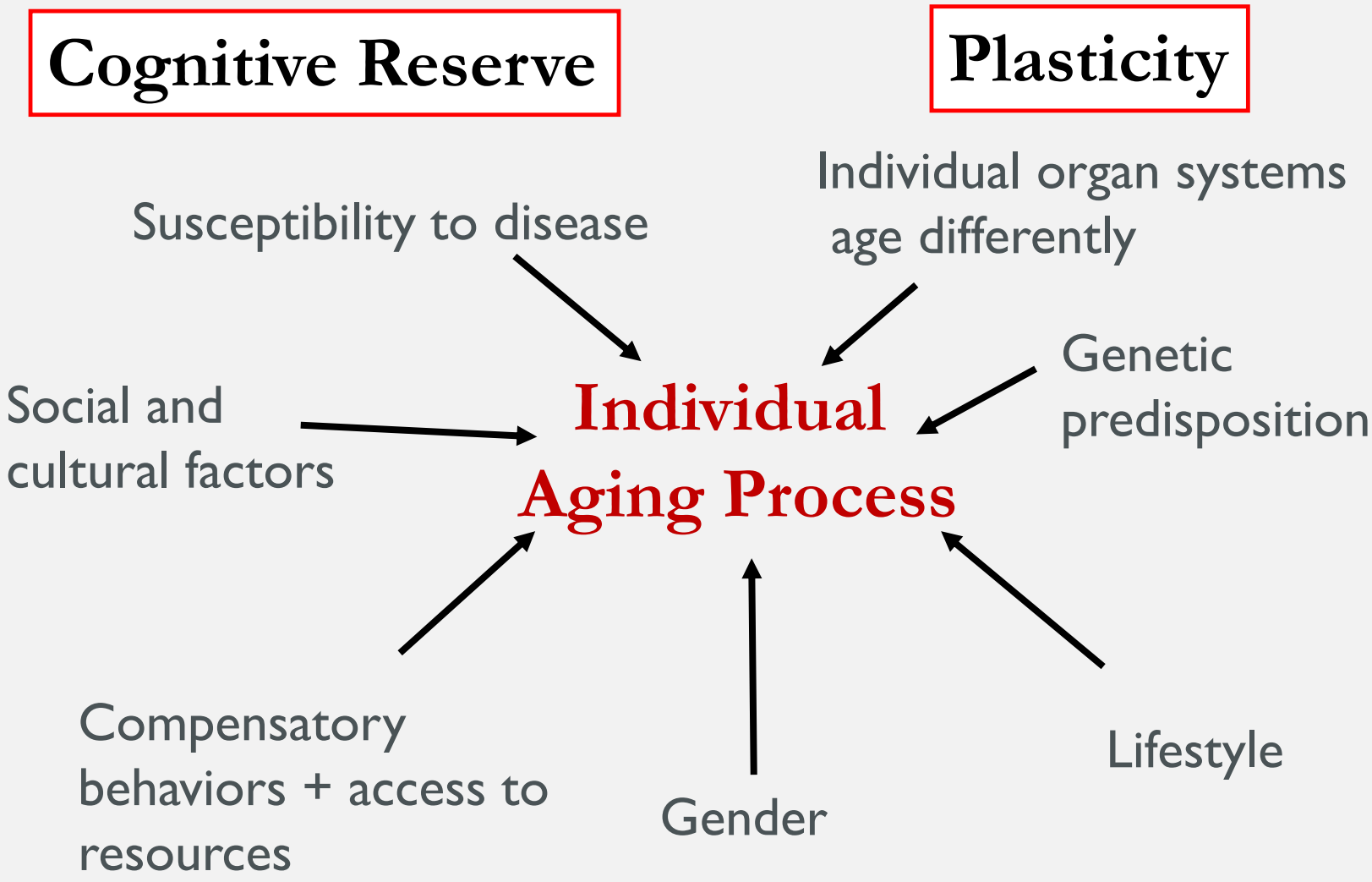
EXPECTED PHYSICAL CHANGES OF AGING

- **Osteopenia/Osteoporosis** - normal aging-related bone loss
- **Sarcopenia** - progressive loss of muscle mass
- **Presbyopia:** the lens of the eye becomes stiffer and less flexible – affecting the ability to focus on close objects (accommodation)
- **Presbycusis** – aging related change in the ability to detect higher pitches – more noticeable in those age 50+
- **Gustation** (i.e. the sense of taste) decrements become more noticeable beyond 60+
- **Olfaction** (i.e. the sense of smell), decrements become more noticeable after age 70+
- **Somatosensory System -** Reduction in sensitivity to pain, touch, temperature, proprioception
- **Vestibular** – Reduction in balance and coordination
- **Cognitive** – Reduction in short term memory loss, attention, and, retrieval
- **Homeostenosis** – narrowing of reserve capacity

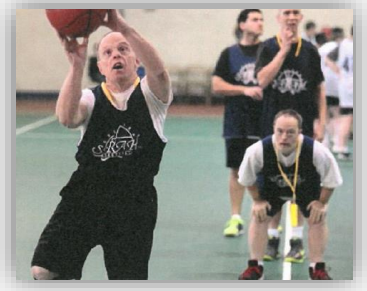
AGE RELATED HEALTH COMPLICATIONS

- Seizures
- Osteoporosis
- Falls and fractures
- Behavioral challenges
- Visual and hearing deficits
- Dementia
- Gait dysfunction
- Cardiopulmonary disease
- Strokes
- Cancer
- Spinal disease
- Liver and Kidney disease
- GI disturbances
- Changes in medication metabolism

Diversity of the Aging Process



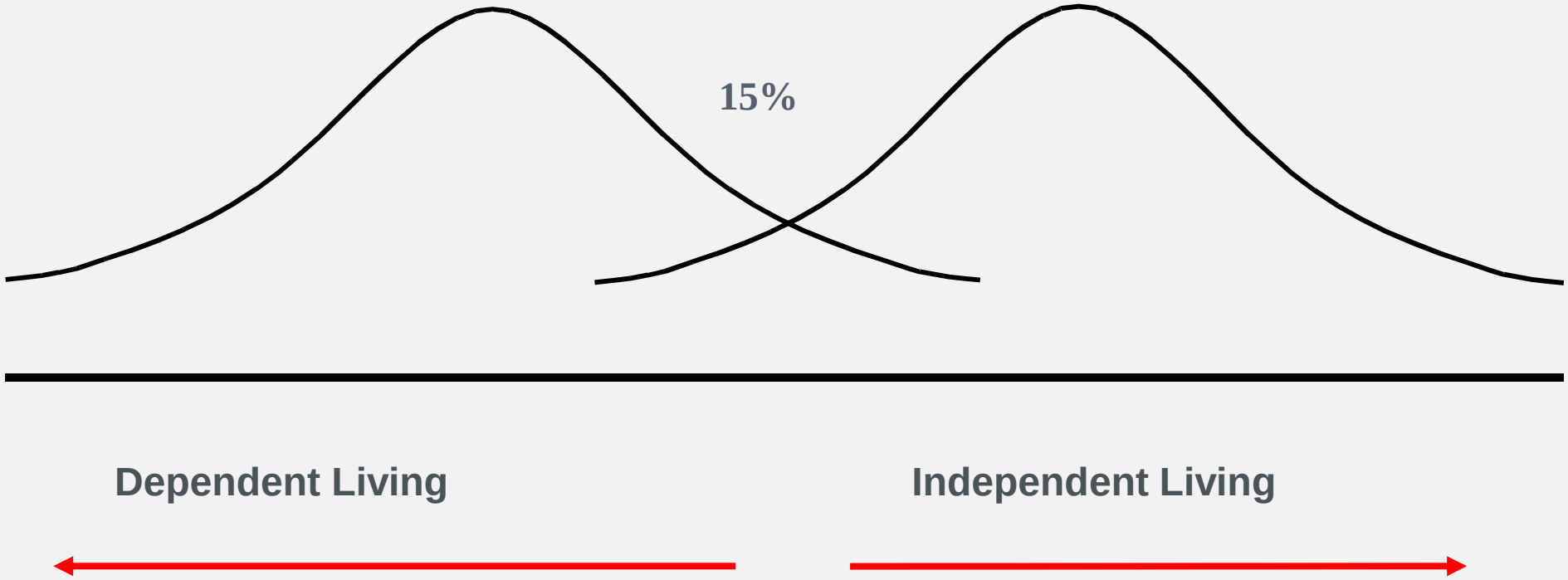
Aging Persons with Intellectual and Developmental Disabilities (IDD)



- Individuals with IDD are living longer and some experience age-related functional and/or cognitive decline
- Normal aging vs pathologic aging
- Syndrome specific aging concerns
- Change in interests
- Aging support networks; siblings and parents
- Younger support networks not adapted to seniors
- Direct support staff/agencies not trained in recognizing the changes of aging nor trained in most age related conditions including dementia care and support
- Participation in competitive physical activity-based sports may become more difficult as one gets older
- Adults with IDD often drop out of Special Olympics as they get older
- Aging and health promotion is not routinely a part of most programs

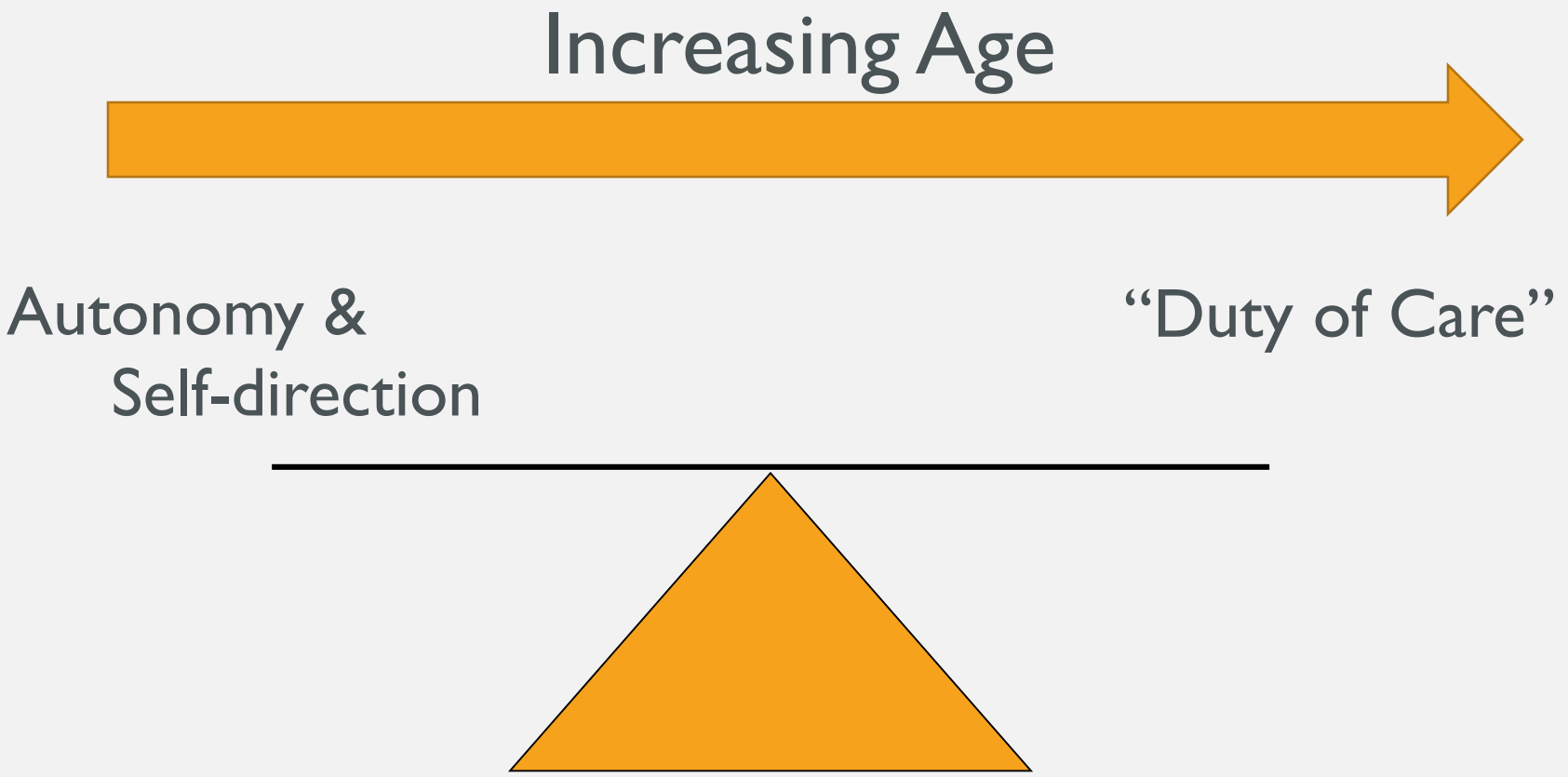
AGING AND DECLINE AFFECTS QOL

Small Change in Cognitive Capability could have profound impact on Independence

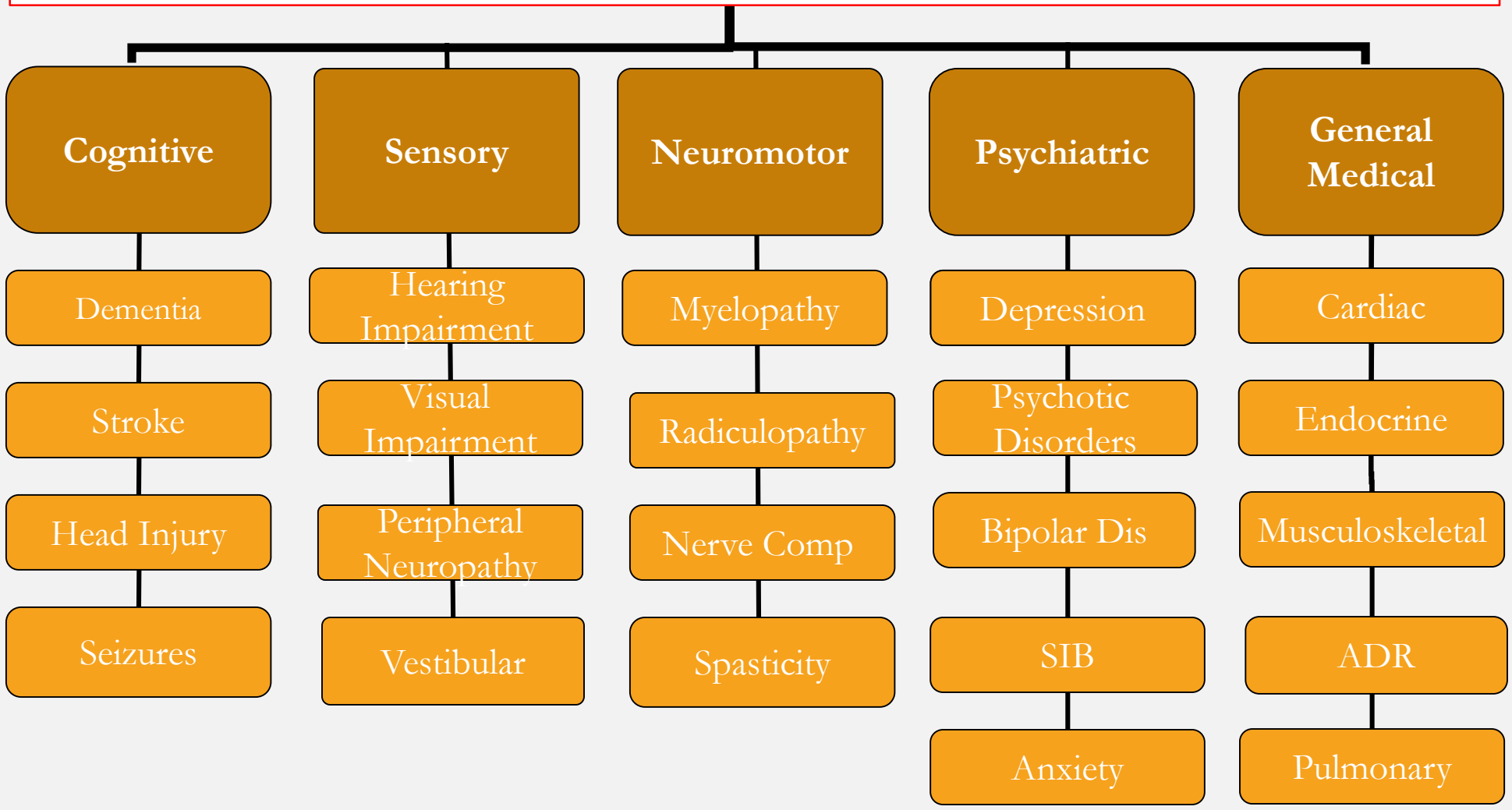


SUPPORTING THROUGHOUT THE LIFESPAN

- A balancing act of guiding philosophies



Functional decline is the decrement in physical and/or cognitive functioning and occurs when a person is unable to engage in activities of daily living



COGNITIVE CHANGES WITH AGING

- Normal changes = **more forgetful & slower to learn**
- MCI – Mild Cognitive Impairment =
  - Immediate recall, word finding, or complex problem solving problems (½ of these folks will develop dementia in 5 yrs)
- Dementia = **Chronic thinking problems in > 2 areas**
- Delirium =**Rapid changes in thinking & alertness**

  (seek medical help immediately)
- Depression **= *chronic unless treated, poor quality , I "don't know", "I just can't" responses, no pleasure***

  can look like agitation & confusion

Cognitive Changes with Aging
In those with Down Syndrome

Who I Am: My Stories, My Memory, My Life History

- Regression
- Medical
- Psychological
- Normal Aging
- Mild Cognitive Impairment
- Dementia (Alzheimer's)

ADULTS WITH DOWN SYNDROME: SPECIALTY CLINIC PERSPECTIVES

CHICOINE, B., MCGUIRE, D., RUBIN, S.

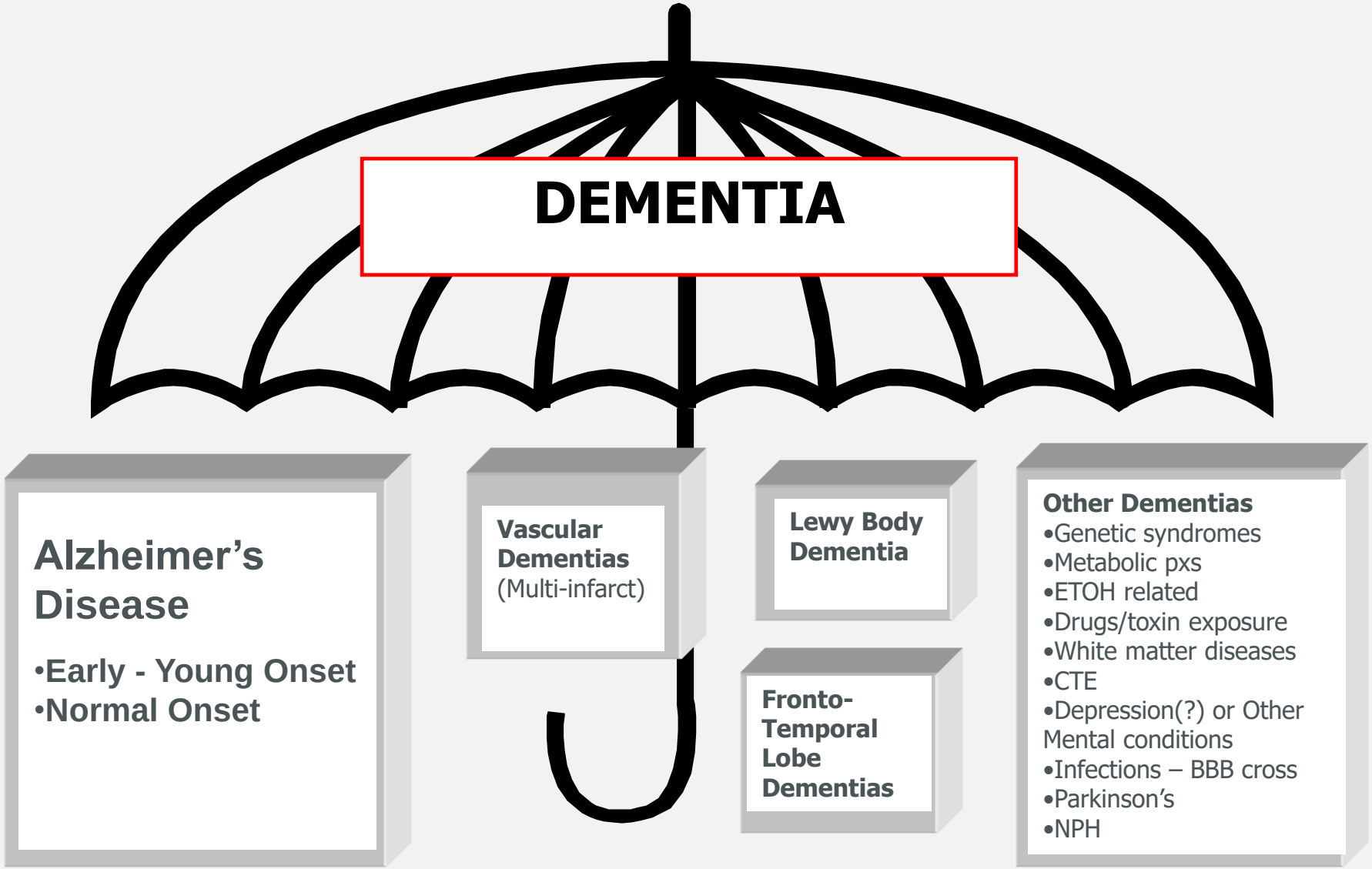
Dementia, Aging and Intellectual Disabilities: A Handbook
ed. by Janicki and Dalton (Taylor and Francis, 1999)

Diagnosed Disorders for 148 Adults Who Presented with a Decline in Function		
Disorder	**Frequency**	**Percent of Diagnosed Disorders (%)**
Mood	76	31
Anxiety	31	13
Obsessive-Compulsive	29	12
Behavior	23	9
Hypothyroid	22	9
Adjustment	12	5
Alzheimer's	11	4
B12 Deficiency	7	3
Menopause	7	3
Attention Deficit / Hyperactive	6	2
Gastrointestinal or Urinary	6	2
Sensory Impairment	6	2
Psychotic	4	2
Other Medical Conditions*	4	2
Cardiac Conditions	3	1
TOTAL	247	100

THE DIAGNOSIS OF DEMENTIA

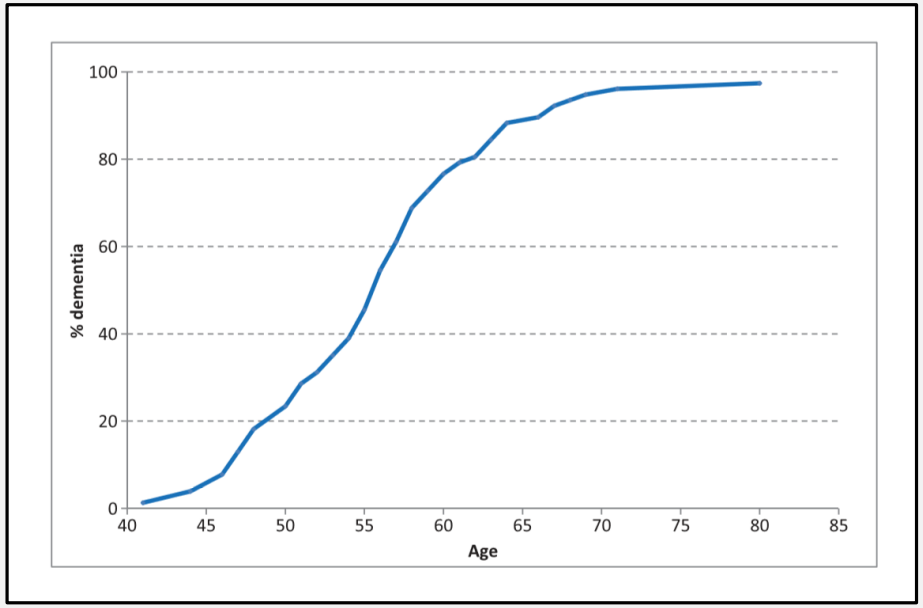
- An acquired syndrome consisting of a decline in memory and other realms of cognitive functioning
- At least one of the following deficits
  - Language difficulties (aphasia)
  - Difficulty with common tasks (apraxia)
  - Unable to identify common objects (agnosia)
  - Disturbance in executive functioning
    - Planning, judgment, decision making

Source: *Diagnostic and Statistical Manual of Mental Disorders.* DSM-IV


DEMENTIA
Alzheimer's Disease
•Early - Young Onset
•Normal Onset
Vascular Dementias
(Multi-infarct)
Lewy Body Dementia
Fronto-Temporal Lobe Dementias
Other Dementias
•Genetic syndromes
•Metabolic pxs
•ETOH related
•Drugs/toxin exposure
•White matter diseases
•CTE
•Depression(?) or Other Mental conditions
•Infections – BBB cross
•Parkinson's
•NPH

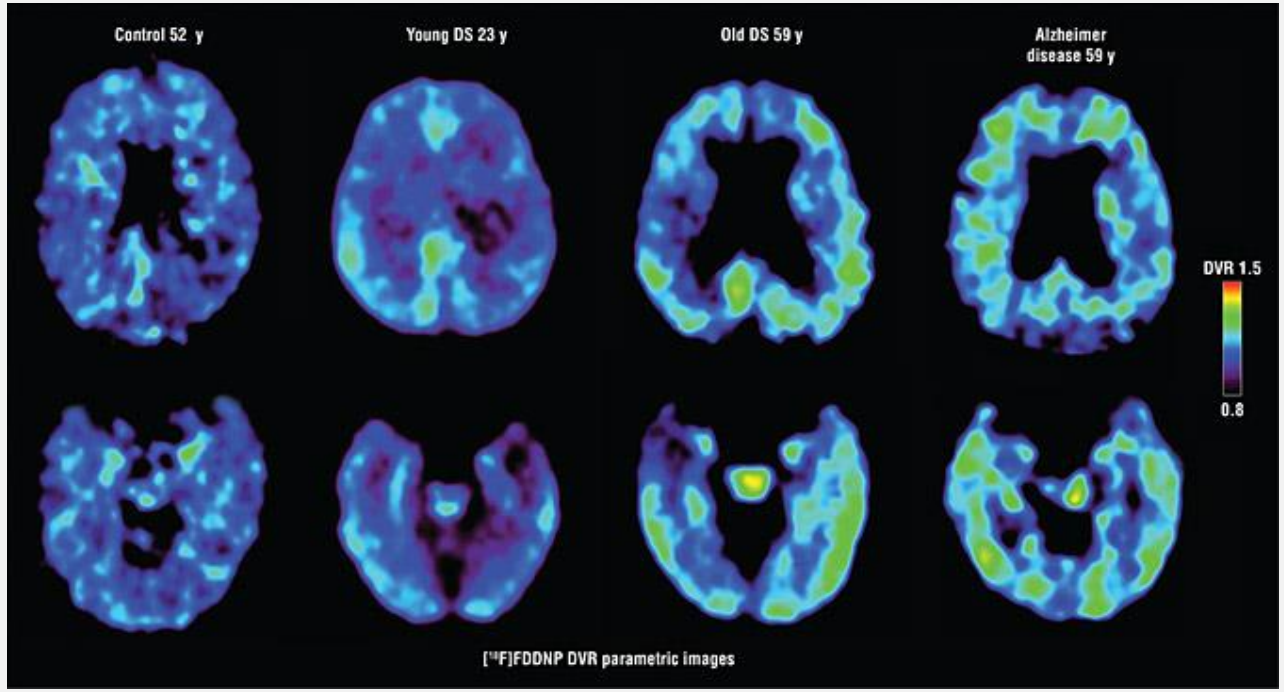
ALZHEIMER'S DISEASE IN DOWN SYNDROME

- Women with Down's syndrome are more at risk of developing Alzheimer's disease than men in the 40 to 65 age group
- People with Down's syndrome who develop Alzheimer's disease live, on average, 4-10 years from first symptoms; median 7 years
- Rapid decline can occur
- Sensory impairments (vision: 93.3%; hearing: 61.3%) were evident in adults with dementia
- Late onset seizures were evident in 73.9%; with epilepsy dx at mean age of 55.4, and interval of about ½ year following dx of dementia.



Percentage of people with Down syndrome who develop dementia at different ages:	
30's	2%
40's	10-15%
50's	20-50%
60's	60-90%

McCarron et al., (2017). A prospective 20-year longitudinal follow-up of dementia in persons with Down syndrome Journal of Intellectual Disability Research Sep;61(9):843-852

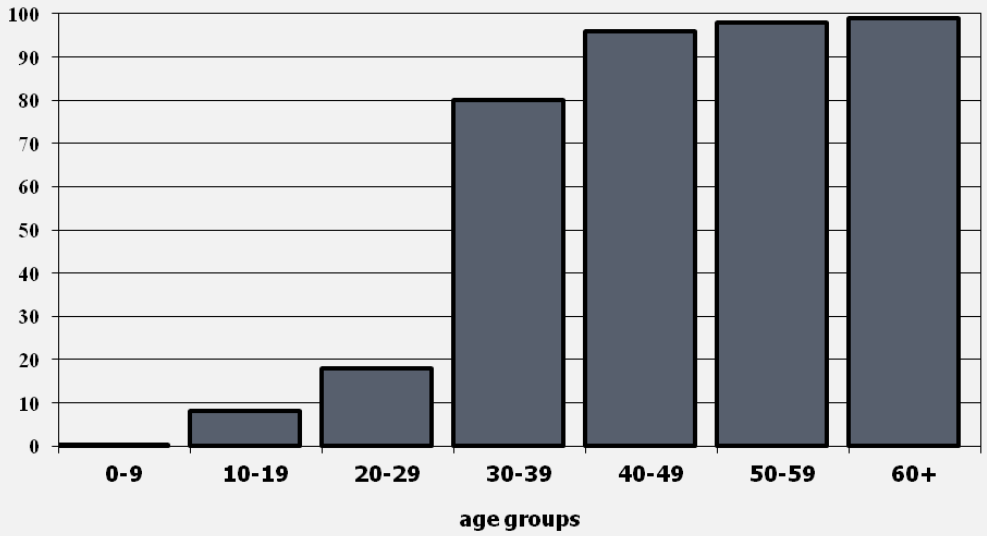


Representative Amyloid Scans in DS and AD

Nelson, L. D. et al. Arch Neurol 2011;68:768-774.

Percent persons with Down syndrome showing evidence of neurofibrillary tangles (NFT) and senile plaques (SP) at autopsy

Mann, D.M.A. (1993). Association between Alzheimer disease and Down syndrome: Neuropathological observations. In J.M. Berg, H. Karlinsky, & A.J. Holland (Eds.),Alzheimer disease and Down syndrome and their relationship (pp. 71-92). Oxford University Press

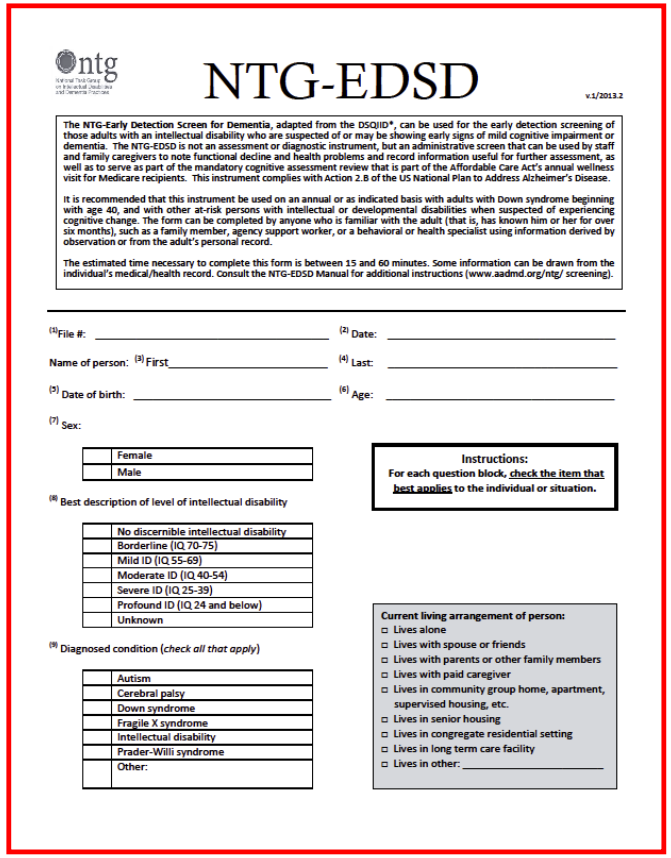


EARLY DETECTION/SCREENING

'NTG-Early Detection Screen for Dementia' (NTG-EDSD)

- Usable by support staff and caregivers to note presence of key behaviors associated with dementia
- Picks up on health status, ADLs, behavior and function, memory, self-reported problems
- Available in multiple languages

Use: to provide information to physician or diagnostician on function and to begin the conversation leading to possible assessment/diagnosis



ntg

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.

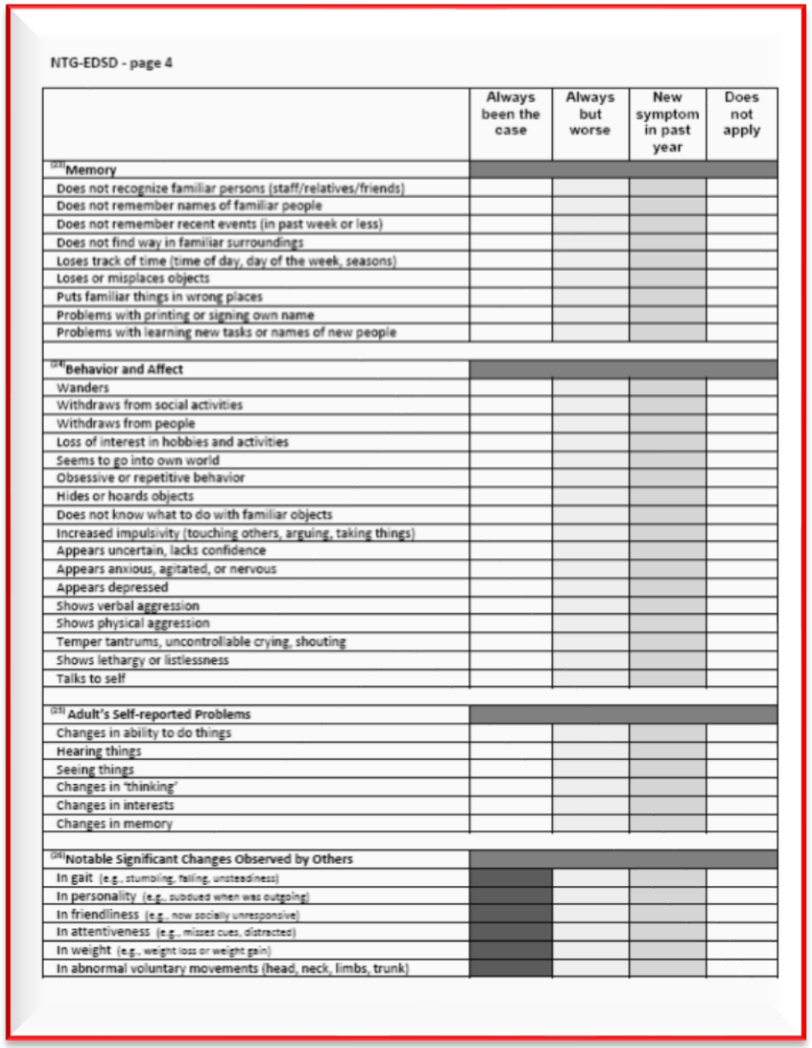
(8) Best description of level of intellectual disability

	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: _____



NTG-EDSD - page 4

	Always been the case	Always but worse	New symptom in past year	Does not apply
(23) Memory				
Does not recognize familiar persons (staff/relatives/friends)				
Does not remember names of familiar people				
Does not remember recent events (in past week or less)				
Does not find way in familiar surroundings				
Loses track of time (time of day, day of the week, seasons)				
Loses or misplaces objects				
Puts familiar things in wrong places				
Problems with printing or signing own name				
Problems with learning new tasks or names of new people				
(24) Behavior and Affect				
Wanders				
Withdraws from social activities				
Withdraws from people				
Loss of interest in hobbies and activities				
Seems to go into own world				
Obsessive or repetitive behavior				
Hides or hoards objects				
Does not know what to do with familiar objects				
Increased impulsivity (touching others, arguing, taking things)				
Appears uncertain, lacks confidence				
Appears anxious, agitated, or nervous				
Appears depressed				
Shows verbal aggression				
Shows physical aggression				
Temper tantrums, uncontrollable crying, shouting				
Shows lethargy or listlessness				
Talks to self				
(25) Adult's Self-reported Problems				
Changes in ability to do things				
Hearing things				
Seeing things				
Changes in 'thinking'				
Changes in interests				
Changes in memory				
(26) Notable Significant Changes Observed by Others				
In gait (e.g., stumbling, falling, unsteadiness)				
In personality (e.g., subdued when was outgoing)				
In friendliness (e.g., now socially unresponsive)				
In attentiveness (e.g., misses cues, distracted)				
In weight (e.g., weight loss or weight gain)				
In abnormal voluntary movements (head, neck, limbs, trunk)				

http://aadmd.org/ntg/screening

NEUROCOGNITIVE ASSESSMENTS

INFORMANT-REPORT AND OBJECTIVE MEASURES FOR CLINICAL ASSESSMENT OF DEMENTIA IN PEOPLE WITH INTELLECTUAL DISABILITIES

- Adaptive Behaviour Dementia Questionnaire (ABDQ), Prasher et al. (2004)
- Assessment for Adults with Developmental Disabilities (AADS), Kalsy et al. (2000); Oliver et al. (2011)
- Dementia Questionnaire for People with Learning Disabilities (DLD)*, Evenhuis (1992); Evenhuis (1996); Eurlings, Evenhuis & Kengen (2006); Evenhuis et al. (2007)

  *Originally named the Dementia Questionnaire for Mentally Retarded

- Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID), Deb et al. (2007)
- Prudhoe Cognitive Function Test (shorter versions), Kay et al. (2003)
- Test for Severe Impairment (Modified), Albert & Cohen (1992)
- Dementia Scale for Down Syndrome (DSDS), Gedye (1995)

REALISTIC GOALS OF DEMENTIA TREATMENT

- Attenuate cognitive and functional decline
- Prevent / decrease behavioral and psychiatric symptoms
- Delay nursing home placement
- Lengthen period of self-sufficiency
- Reduce caregiver burden/support families
- Palliative Care
- End of Life Care
- Determining and measuring outcomes

BEHAVIORAL AND PSYCHOLOGICAL SYMPTOMS OF DEMENTIA (BPSD)

- Depression—40%
- Delusions—63%
- Hallucinations—4-41%
- Aggression—31-42%
- Apathy
- Pseudobulbar Affect

- Sleep disturbance (day/night reversal)
- Hoarding
- Shadowing
- Disinhibition (stripping)
- Sexually inappropriate behavior
- Sundowning
- Wandering

- Associated with worse prognosis
- More rapid cognitive decline
- Increased caregiver burden
- Leads to earlier admission to institutional care
- Increased healthcare costs

COMMON TRIGGERS

- Physical
  - Acute illness/infection, medications, pain, poor vision, hearing, poor sleep
- Cognitive
  - Inability to understand, express oneself, lack of insight, misinterpretation of environment, difficult to problem solve
- Emotional
  - Fear, anxiety, depression, frustration, apathy, boredom
- Environmental
  - Changes in caregiver, confrontational approach, tasks that exceed abilities, change in routine, over/understimulation, lack of visual cues

NONPHARMACOLOGICAL APPROACHES

- Familiar environment—avoid frequent moves
- Soft lighting
- Calm colors
- Places to walk
- Access to outdoor spaces
- Home-like environment
- Low stimuli—minimize background noise
- Time out space
- Reminiscing
- Individualized Care Planning
- Careful analysis of care interactions
- Meaningful activity
- Art/Music Therapy
- Exercise/Movement
- Snoezelen (multisensory stimulation program)
- Aromatherapy
- Yoga

QUESTIONS TO BE ANSWERED IN EVALUATING MEDICATION USE

- What is the target problem being treated?
- Is the drug necessary?
- Are nonpharmacologic therapies available?
- Is this the lowest practical dose?
- Does this drug have adverse effects that are more likely to occur in an older patient?
- By what criteria, and at what time, will the effects of therapy be assessed?
- Safety of the medication

Drug use in the nursing home
Avorn J, Gurwitz JH. Ann Intern Med. 1995 Aug 1;123(3):195-204

MANAGING BPSD: PHARMACOLOGIC INTERVENTIONS

Drug class	Chemical name	Dosage range (mg)	Side effects of class
Antipsychotics	Aripiprazole* Haloperidol Risperidone* Quetiapine* Olanzapine*	2.5-15 0.5-5 0.25-2 25-200 2.5-15	Sedation, EPS, NMS, metabolic syndrome, QTc prolongations, increased risk of CVE and mortality
Antidepressants	Fluoxetine Citalopram Paroxetine Sertraline Trazadone	10-80 10-60 10-50 25-200 25-200	Anxiety, headaches, sedation, GI symptoms, sexual dysfunction
Mood stabilizers	Carbamazepine Divalproex sodium Oxcarbazepine	100-400 250-1000 300-600	Sedation, gait and balance issues, falls, liver dysfunction, hyperammonemia, thrombocytopenia

Adapted from Tampi et al. *Clin Geriatr*. 2011;19:31-32.

PBA: Dextromethorphan/Quiidine (Nuedexta) 20/10 mg
Hepatotoxicity, QTc prolongation, thrombocytopenia

COGNITIVE ENHANCERS

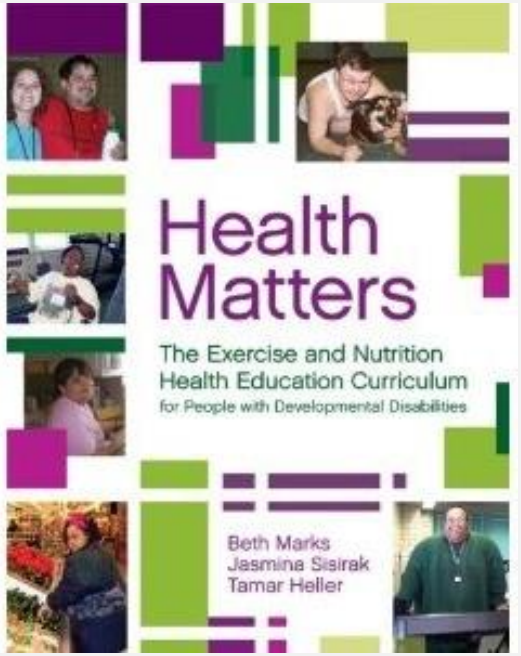
- Cholinesterase Inhibitors; Aricept, Exelon, Razadyne
  - **Eady, N., et al. The British Journal of Psychiatry (2018) 212, 155–160**
    - **Heller, J. American Journal of Medical Genetics, Oct. 15, 2004; vol 130: pp 324-326**
    - ***Lott IT et al.* Arch Neurol. 2002;59:1133-1136**
    - **Kishnani PS *et al.* (1999*)* Lancet 353: 1064**
- NMDA (N-methyl-D- aspartate) receptor antagonist; Namenda
  - **Hanney, Prasher, The Lancet, Volume 379, Issue 9815, 528 - 536, 11 February 2012**
- Herbal Supplements/Vitamins; Vit E, Gingo Biloba
  - **Sano, M *et al*. (1997) A controlled trial of selegiline, alpha-tocopherol, or both as treatment for Alzheimer's disease. NEJM 336: 1216-22**

Possible preventive strategies against dementia

- **Promoting healthy lifestyles**
  - non-smoking
  - moderate alcohol intake
  - physical activity

- **Decreasing vascular burden**
  - hypertension
  - heart failure
  - diabetes
  - stroke

- **Increasing brain reserve**



Rehabilitation Research and Training Center (RRTC) on Aging with Developmental Disabilities: Lifespan Health and Function, UIC at Chicago http://www.rrtcadd.org/

CHANGE IN FOCUS OF SUPPORTS PROVIDED

- Maintaining skills
- Stabilizing the environment
- Minimizing choices
- Giving reassurance
- Personal care
- Assessing and meetings medical needs
- Meaningful activities

PROGRESSION OF DISEASE; ANTICIPATORY GUIDANCE

- Cognitive Skills will decline
- Support needs will increase
- Increase risks of falls, injuries
- Swallowing dysfunction, clots, pneumonia, bladder infections
- Seizures
- Watch for signs of abuse and neglect
- Watch for signs of caregiver burn out
- End of Life care; Palliative and Hospice

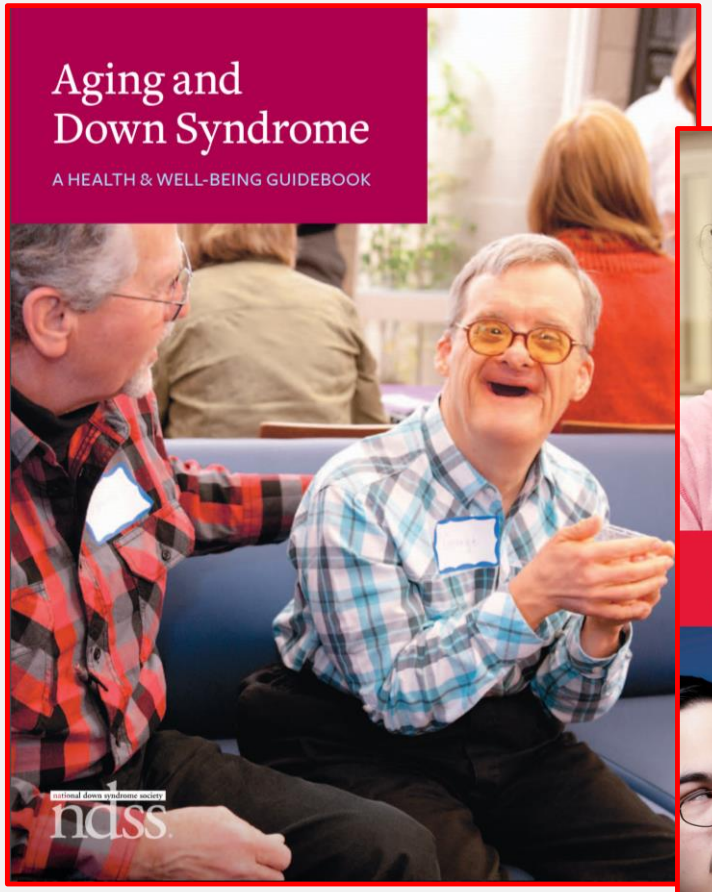
IMPACT ON FAMILIES AND CAREGIVERS

- Frequent issues experienced by families and caregivers include:
  - Denial
  - Anger / Frustration
  - Guilt
  - Loss and Grief
  - Letting Go
  - Financial Stress
  - Role Reversals
  - Social Isolation
  - Becoming patients themselves

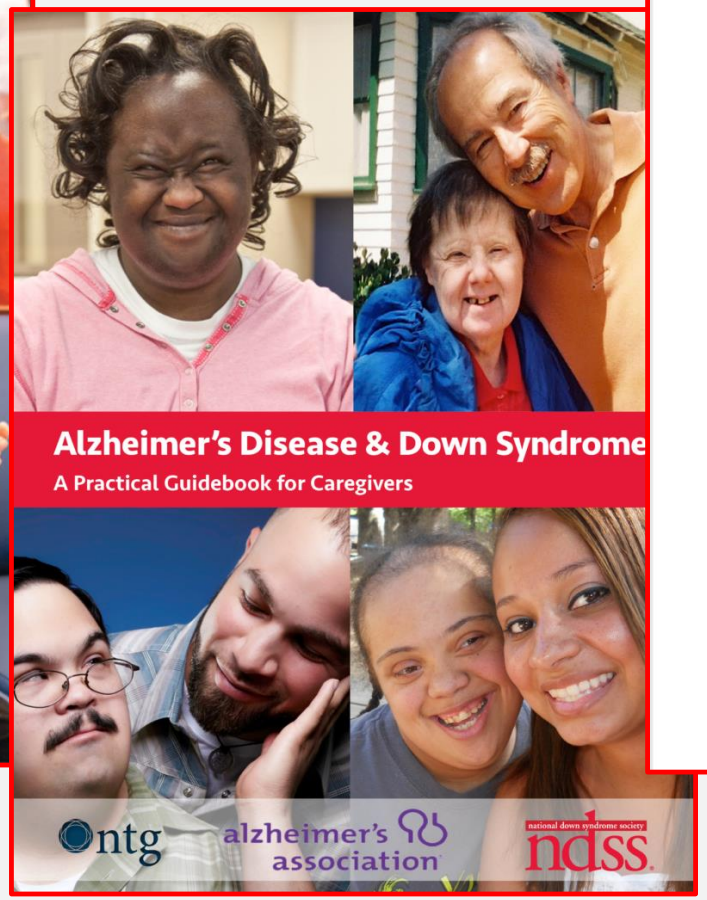
COMMUNITY, STATE AND NATIONAL SUPPORTS

- Community support provider agencies
  - Private
  - Public – state/local government entities
- Area Agencies on Aging (AAA)
  - Aging and Disability Resource Centers (ADRC)
- State and local Alzheimer's Association chapters
  - As well as other local dementia care groups
- State and local Protection and Advocacy Networks
- AADMD-NTG
- Special Olympics
- Faith-based organizations

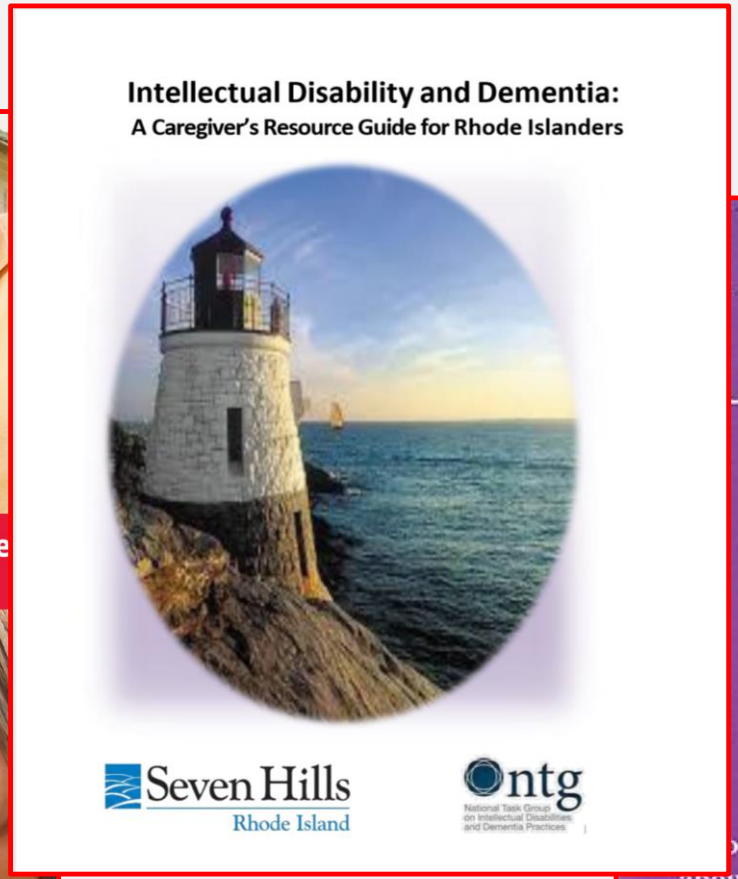
PERSON/FAMILY CENTERED RESOURCES



http://www.ndss.org/wp-content/uploads/2017/11/Aging-and-Down-Syndrome.pdf

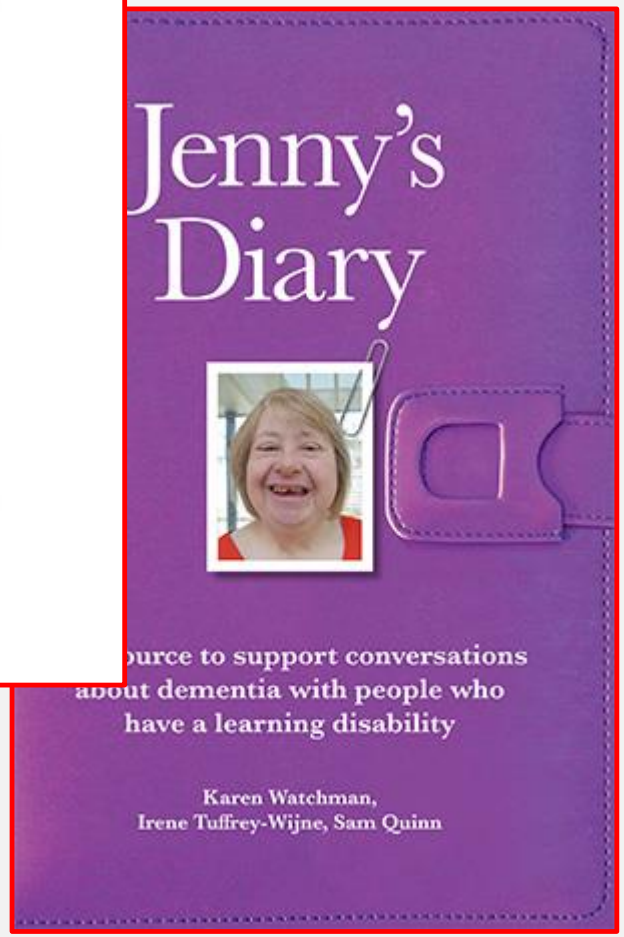


http://www.ndss.org/wp-content/uploads/2017/11/NDSS_Guidebook_FINAL.pdf

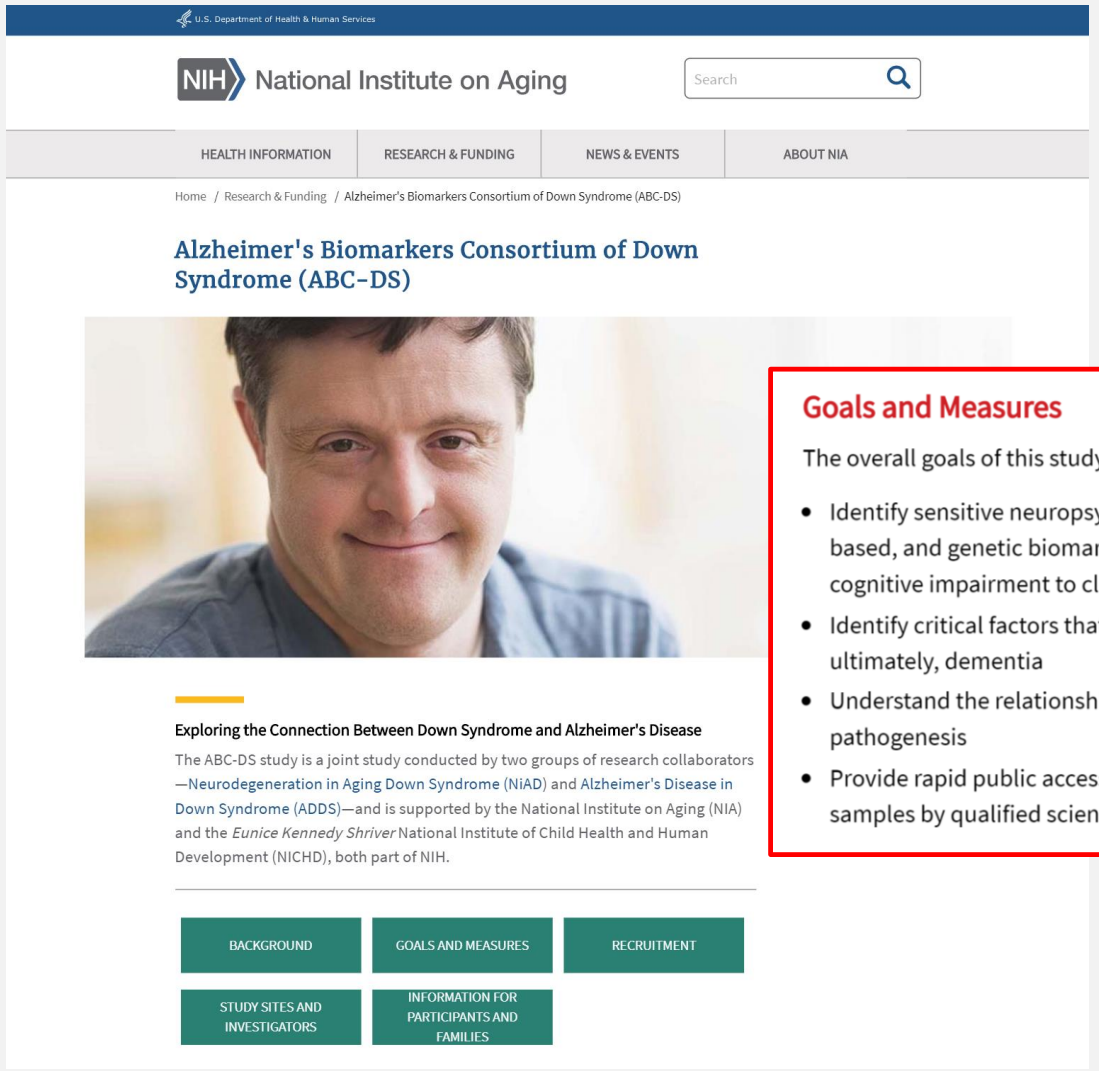


http://www.sevenhills.org/uploads/SHRI-IDD-ADRD-Resource-Guide.pdf

www.learningdisabilityanddementia.org/jennys-diary.html



ALZHEIMER'S BIOMARKERS CONSORTIUM OF DOWN SYNDROME (ABC-DS)



https://www.nia.nih.gov/research/abc-ds

Get More Information

Scientific Contacts for ABC-DS

NIA
Laurie Ryan PhD, ryanl@mail.nih.gov

NICHD
Melissa Parisi
PhD, parisima@mail.nih.gov

Goals and Measures

The overall goals of this study are to:

- Identify sensitive neuropsychological measures of cognitive decline, imaging, blood-based, and genetic biomarkers associated with transition from normal aging to mild cognitive impairment to clinical dementia in adults with DS
- Identify critical factors that link cerebral Aβ deposition to neurodegeneration and, ultimately, dementia
- Understand the relationships between biomarkers and pathways implicated in AD pathogenesis
- Provide rapid public access to all data, without embargo, and access to the biological samples by qualified scientific investigators

Recruitment

The NiAD sites will recruit 180 adults with DS (10% with dementia) and 40 sibling controls, age 25 years and older. The ADDS sites will recruit 225-300 adults with DS, 40 years and older.

Neurodegeneration in Aging Down Syndrome (NiAD)

Site	Investigator & Study Coordinator
University of Pittsburgh (Coordinating Center), Pittsburgh, PA	Ben Handen, Ph.D., Co-PI William Klunk, M.D., Ph.D., Co-PI Cathy Wolfe, Study Coordinator
University of Wisconsin Madison, WI	Brad Christian, Ph.D., Co-PI Renee Makuch, Study Coordinator
Barrow Neurological Institute Phoenix, AZ	Marwan Sabbagh, M.D., Site PI Sandy Quintanilla, Study Coordinator
University of Cambridge Cambridge, UK	Shahid Zaman, M.D., Ph.D., Site PI Concepcion Padilla, Study Coordinator

Alzheimer's Disease in Down Syndrome (ADDS)

Site	Investigator & Study Coordinator
Columbia University (Coordinating Center) New York, NY	Nicole Schupf, Ph.D., Co-PI Deborah Pang, Study Coordinator
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University of California, Irvine Irvine, CA	Ira Lott, M.D., Co-PI Eric Doran, Study Coordinator Alicia Hernandez, Study Coordinator
Harvard/Massachusetts General Hospital Boston, MA	Florence Lai, M.D., Site PI Diana Rosas, M.D., Site PI Nusrat Jahan, Study Coordinator Courtney Jordan, Study Coordinator
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WHO DO YOU SEE?

Thank You!!

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