# Decline in Adults with an Intellectual and Developmental Disability; Concerns for Dementia


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ntg
National Task Group on Intellectual Disabilities and Dementia Practices

Project ECHO®
University of Washington
Seattle, WA

UW Medicine
MEMORY & BRAIN WELLNESS CENTER

AADMD

# Case

- 45 yo female with DS, with PMHX of hypothyroidism and depression
- Last 6 months been forgetful at home and work
- Lives with father, and sister has been very supportive
- On Synthroid, TSH wnl
- On Zoloft
- Concerns for early onset AD

# 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

ADA
Americans with Disabilities Act

Special Olympics


CONVENTION on the RIGHTS of PERSONS with DISABILITIES







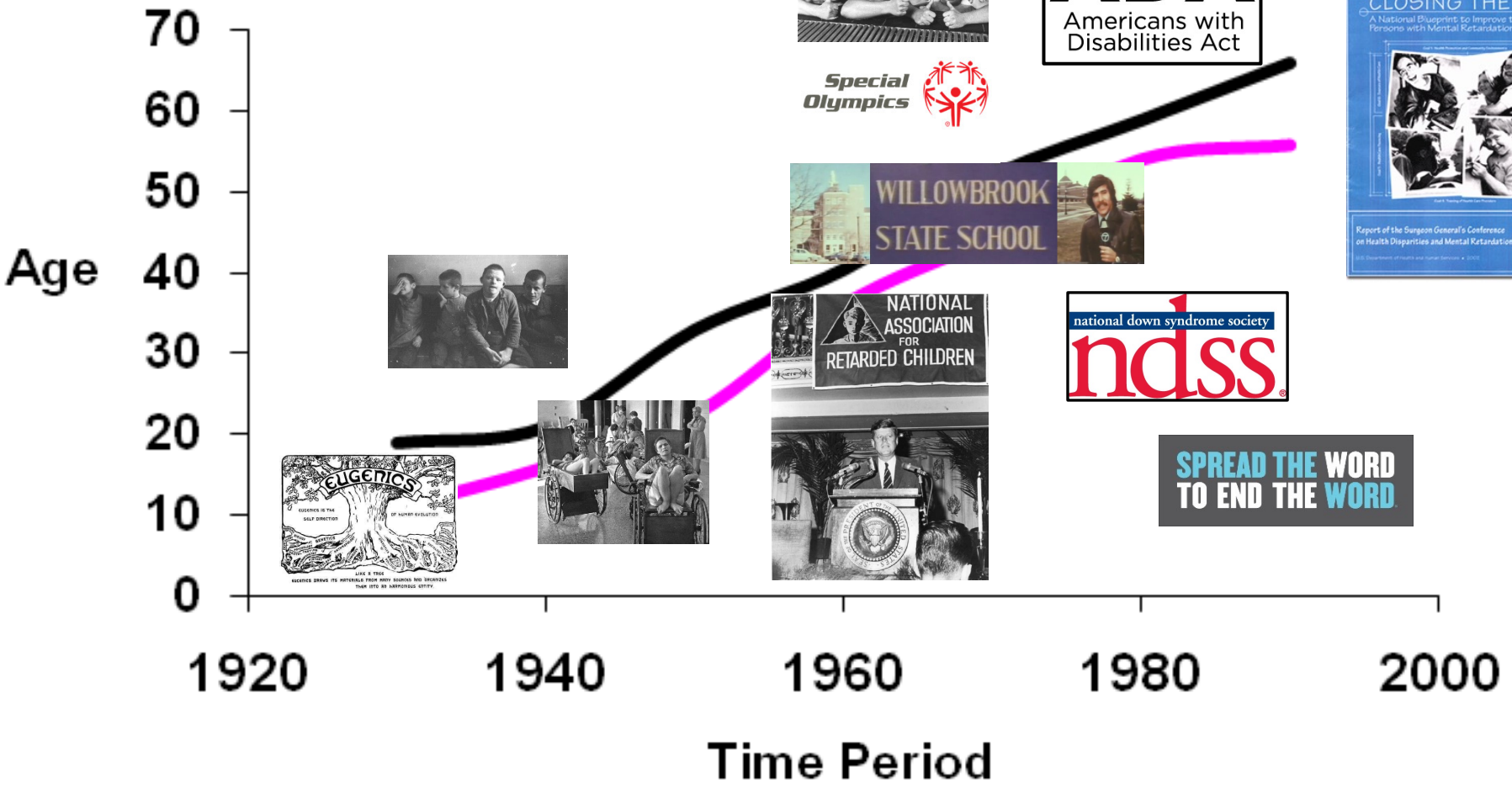


national down syndrome society
ndss






SPREAD THE WORD TO END THE WORD

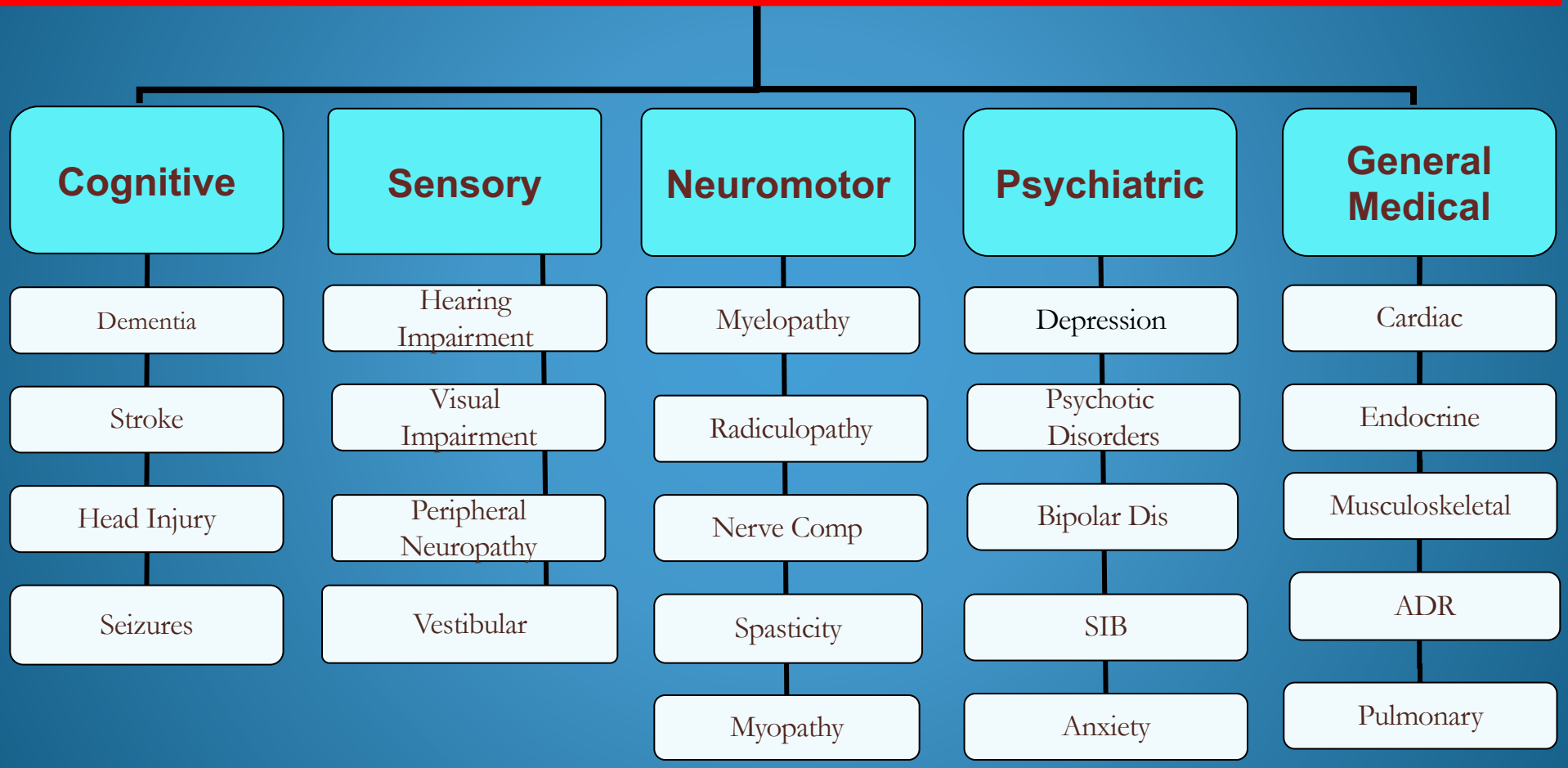



Source: Carter & Jancar, 1983, Janicki, Dalton, Henderson & Davidson, 1999

# 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

**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**



# DD Specific Aging and Health Complications

## Down Syndrome

- Sleep disturbances, depression, sensory loss
- Obesity
- Thyroid dysfunction, B12/folate deficiency
- Sleep Apnea
- Gait dysfunction
- Seizure Disorder
- Early onset Alzheimer's Disease

## Cerebral Palsy

- Chronic Pain
- Dysphagia, aspiration, Esophageal strictures, gastritis
- Dental caries, erosion
- Motor dysfunction, inc spasticity and spinal cord dysfunction
- Osteoporosis
- Worsening bladder/bowel dysfunction

## Autism

- Lifespan outcomes with Autism are unpredictable: some improve, some plateau, some lose skills
- Restrictive behaviors such as ritualistic, compulsive or self injurious behaviors tend to become more infrequent with age
- Seizures, accidental deaths (drowning, suffocation), earlier death from heart disease, aspiration pneumonia

# When Change Occurs

- Normative or a sign of disease
- Life Stories
- Health Co-morbidities
- Psychosocial changes
- Dementia/Alzheimer's disease
- Biases/stereotypes/Diagnostic Over Shadowing

# Cognitive Changes with Aging in Adults with Down Syndrome

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

- Regression
- Medical
- Psychological
- Normal aging
- Mild cognitive impairment
- Early-onset Alzheimer's dementia; 60% by age 60!!

# 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**
- Depression = ***chronic unless treated, poor quality , I "don't know", "I just can't" responses, no pleasure***

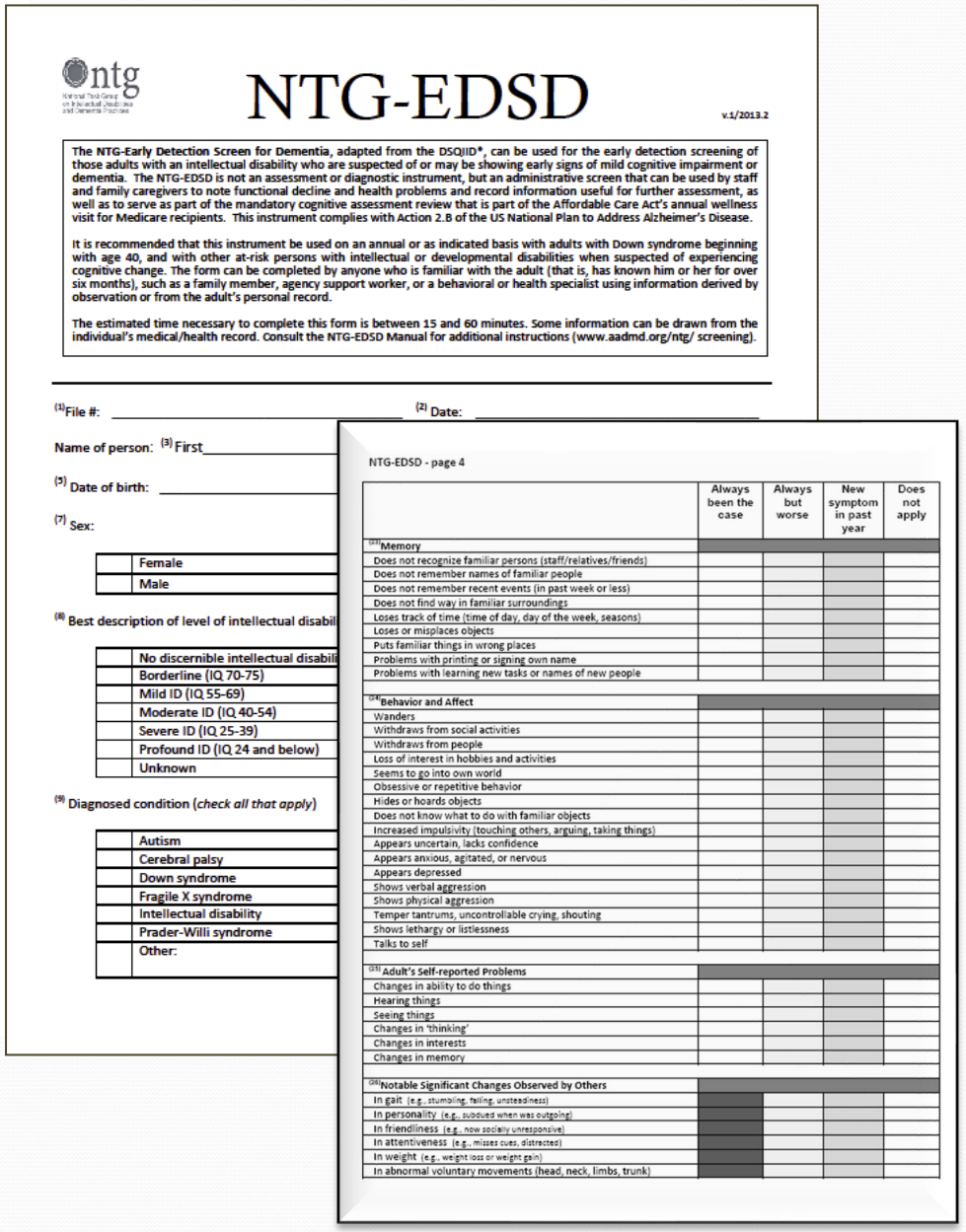
  *can look like agitation & confusion*

# Making the Diagnosis of Dementia

- Having enough information; Life Story
- Differential diagnosis
  - Hypothyroidism, B12/folate, Sleep Apnea, Depression/Adjustment
  - ADR's, Neuromuscular
- Neuropsychological Assessment
- Imaging
- Biomarkers

## 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



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# 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.B 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_____

(5) Date of birth: _____

(7) Sex:

| | |
|---|---|
| | Female |
| | Male |

(8) Best description of level of intellectual disabil

| | |
|---|---|
| | No discernible intellectual disabili |
| | 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: |

NTG-EDSD - page 4

| | Always been the case | Always but worse | New symptom in past year | Does not apply |
|---|---|---|---|---|
| (10) 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 | | | | |
| (11) 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 | | | | |
| (12) Adult's Self-reported Problems | | | | |
| Changes in ability to do things | | | | |
| Hearing things | | | | |
| Seeing things | | | | |
| Changes in 'thinking' | | | | |
| Changes in interests | | | | |
| Changes in memory | | | | |
| (13) 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) | | | | |

NTG-EDSD (the-ntg.org)

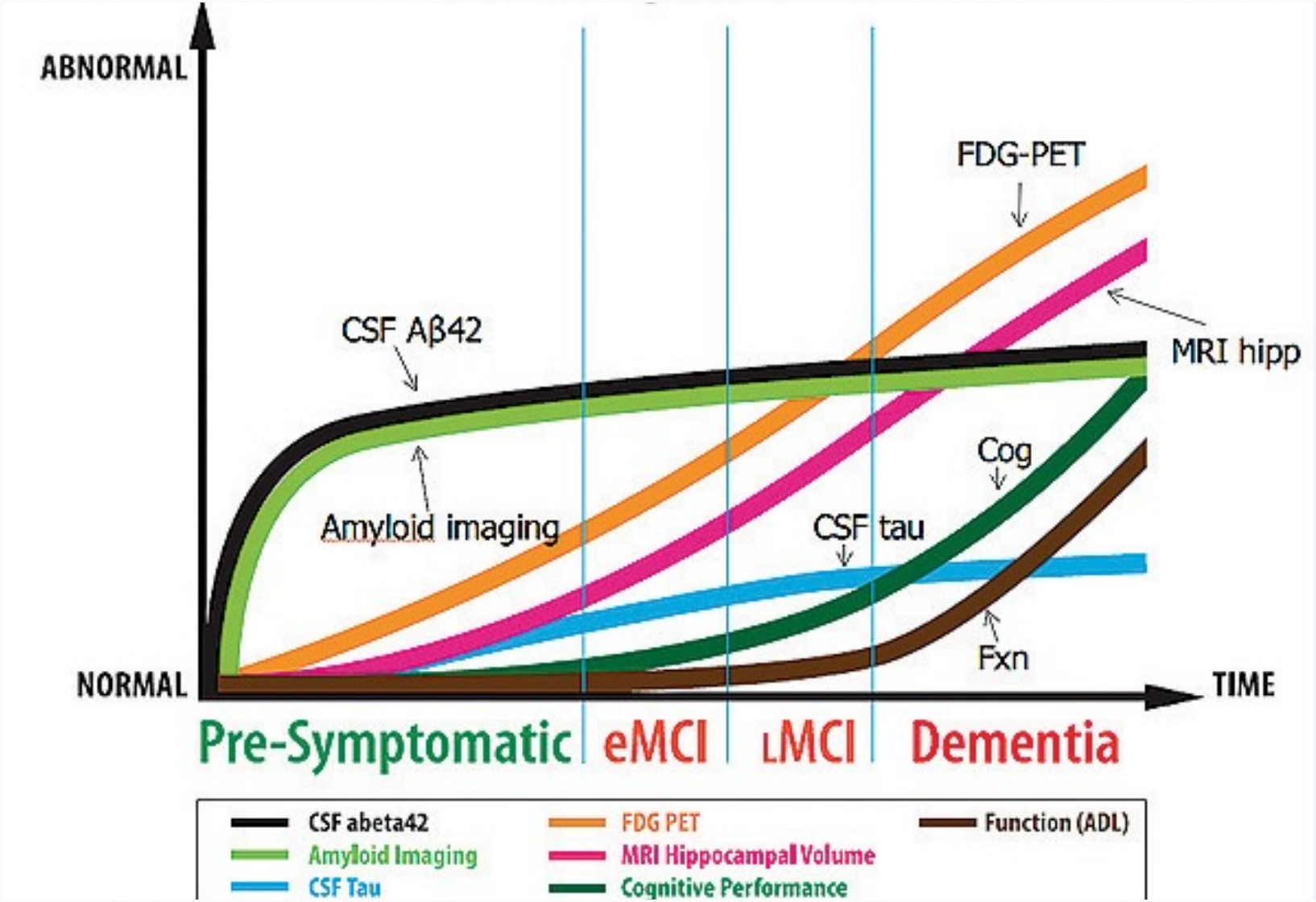
# Neurocognitive Assessments

**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)
- 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)
- Fuld Object Memory Evaluation (1990)

*Originally named the Dementia Questionnaire for Mentally Retarded

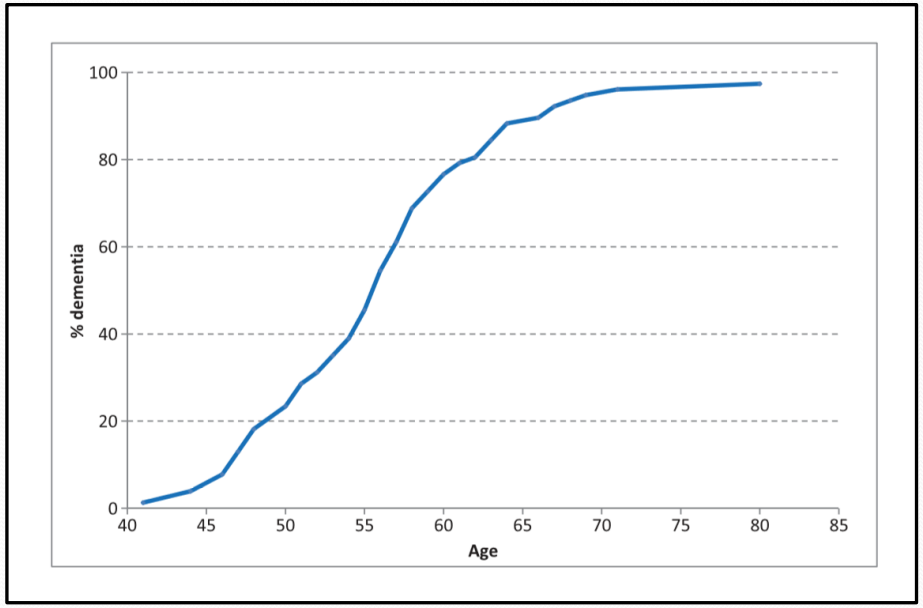
# Alzheimer's Disease Biomarkers




**Report of the task force on designing clinical trials in early (predementia) AD**
P.S. Aisen, S. Andrieu, C. Sampaio, M. Carrillo, Z.S. Khachaturian, et al, Neurology Jan 2011, 76 (3) 280-286

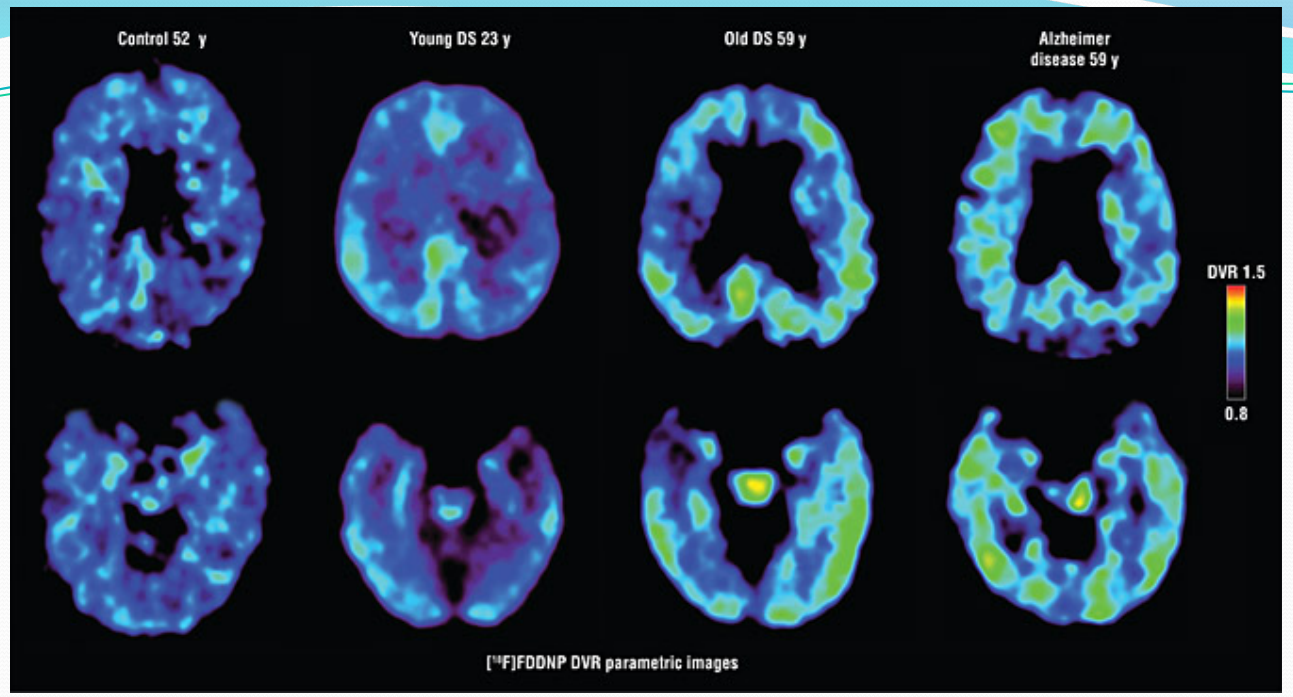
# Alzheimer's Disease in Down Syndrome

- 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**

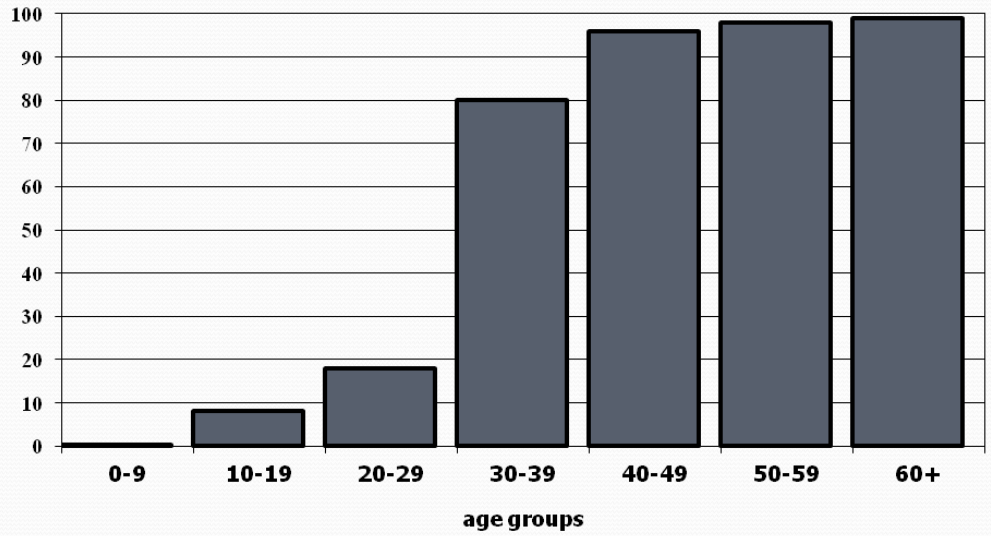



## Representative Amyloid Scans in DS and AD

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



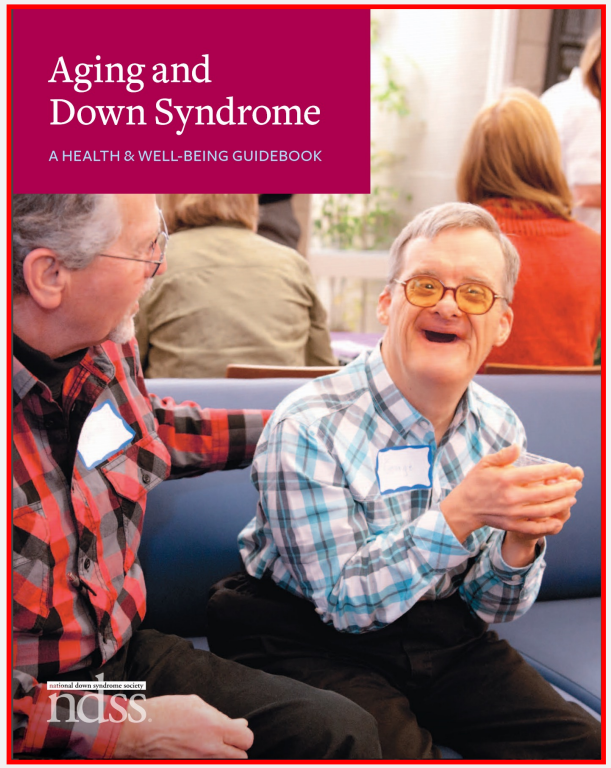
# Caring

- "Caring" with empathy and sensitivity
- Cognitive therapeutics
- Behavioral care and supports
- Health and wellness
- Concomitant health complications; seizures, sleep apnea, falls, etc.
- Anticipating progression and its various needs
- Palliative and end stage supports
- Research opportunities
- Financial and legal supports
- Transition of care

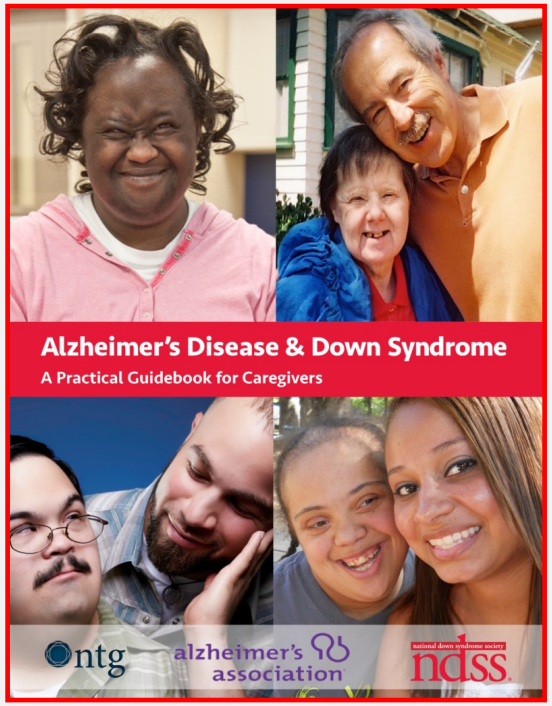
# IMPACT ON FAMILIES AND CAREGIVERS

- 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

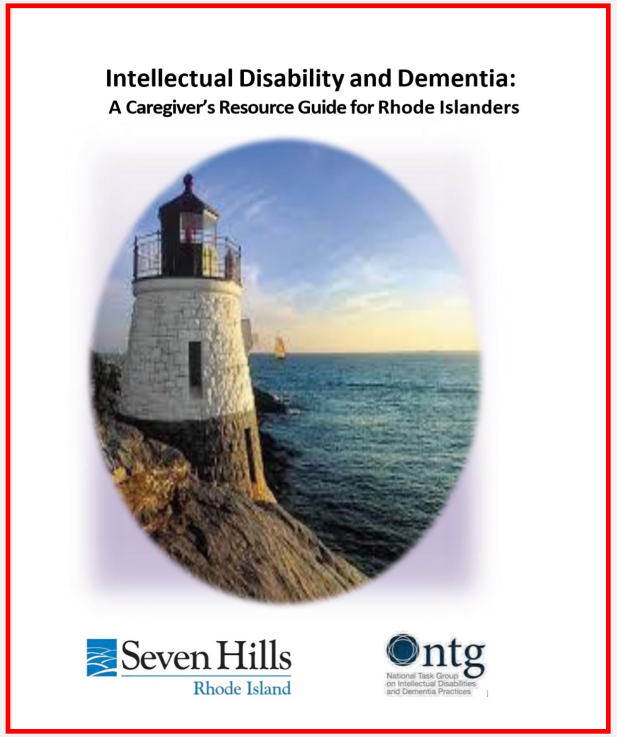
# PERSON/FAMILY CENTERED RESOURCES



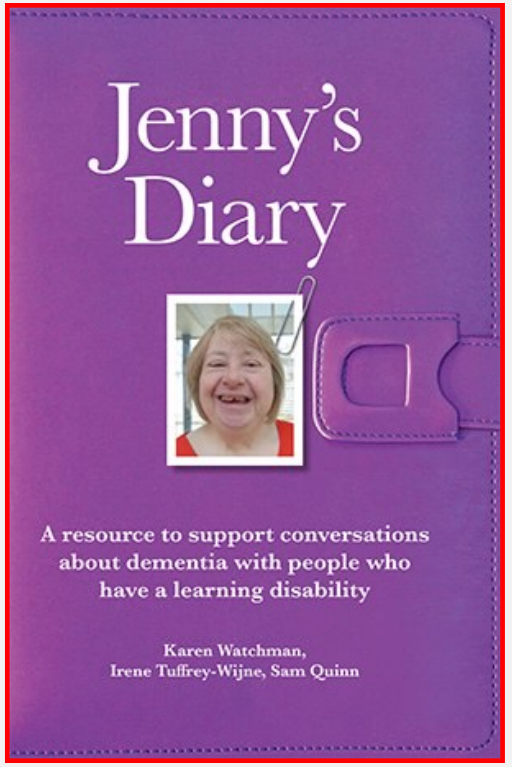

http://www.ndss.org/wp-content/uploads/2017/11/Aging-and-Down-Syndrome.pdf




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

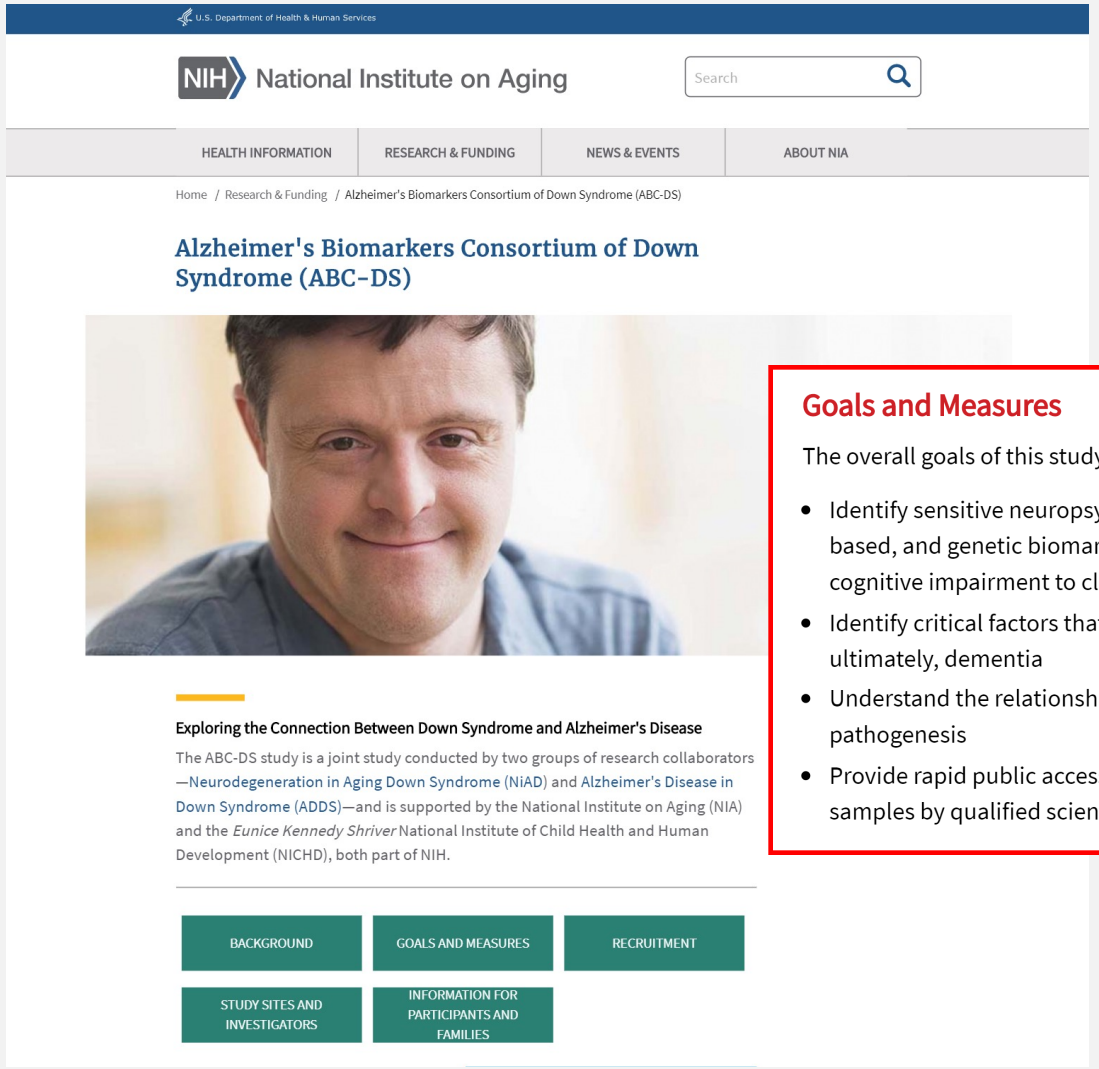



http://www.sevenhills.org/uploads/SHRI-IDD-ADRD-Resource-Guide.pdf




www.learningdisabilityanddementia.org/jennys-diary.html

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




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

## 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<br>William Klunk, M.D., Ph.D., Co-PI<br>Cathy Wolfe, Study Coordinator |
| University of Wisconsin Madison, WI | Brad Christian, Ph.D., Co-PI<br>Renee Makuch, Study Coordinator |
| Barrow Neurological Institute Phoenix, AZ | Marwan Sabbagh, M.D., Site PI<br>Sandy Quintanilla, Study Coordinator |
| University of Cambridge Cambridge, UK | Shahid Zaman, M.D., Ph.D., Site PI<br>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<br>Deborah Pang, Study Coordinator |
| Kennedy Krieger Institute/Johns Hopkins Medical Center Baltimore, MD | Wayne Silverman, Ph.D., Co-PI |
| University of California, Irvine Irvine, CA | Ira Lott, M.D., Co-PI<br>Eric Doran, Study Coordinator<br>Alicia Hernandez, Study Coordinator |
| Harvard/Massachusetts General Hospital Boston, MA | Florence Lai, M.D., Site PI<br>Diana Rosas, M.D., Site PI<br>Nusrat Jahan, Study Coordinator<br>Courtney Jordan, Study Coordinator |
| The New York State Institute for Basic Research in Developmental Disabilities Staten Island, NY | Sharon Krinsky-McHale, Ph.D., Site PI<br>Deborah Pang, Study Coordinator |
| University of North Texas Health Science Center Fort Worth, TX | Sid O'Bryant, Ph.D., Site PI |

# Potential Aging and IDD Partners

Families

DD Councils

IDD Advocacy groups

Community Aging

Cultural/Spiritual Advocacy

Alzheimer's Association

AADMD

Community Support Organizations

NTG

Healthcare Professionals

Sports and Fitness

Pharma

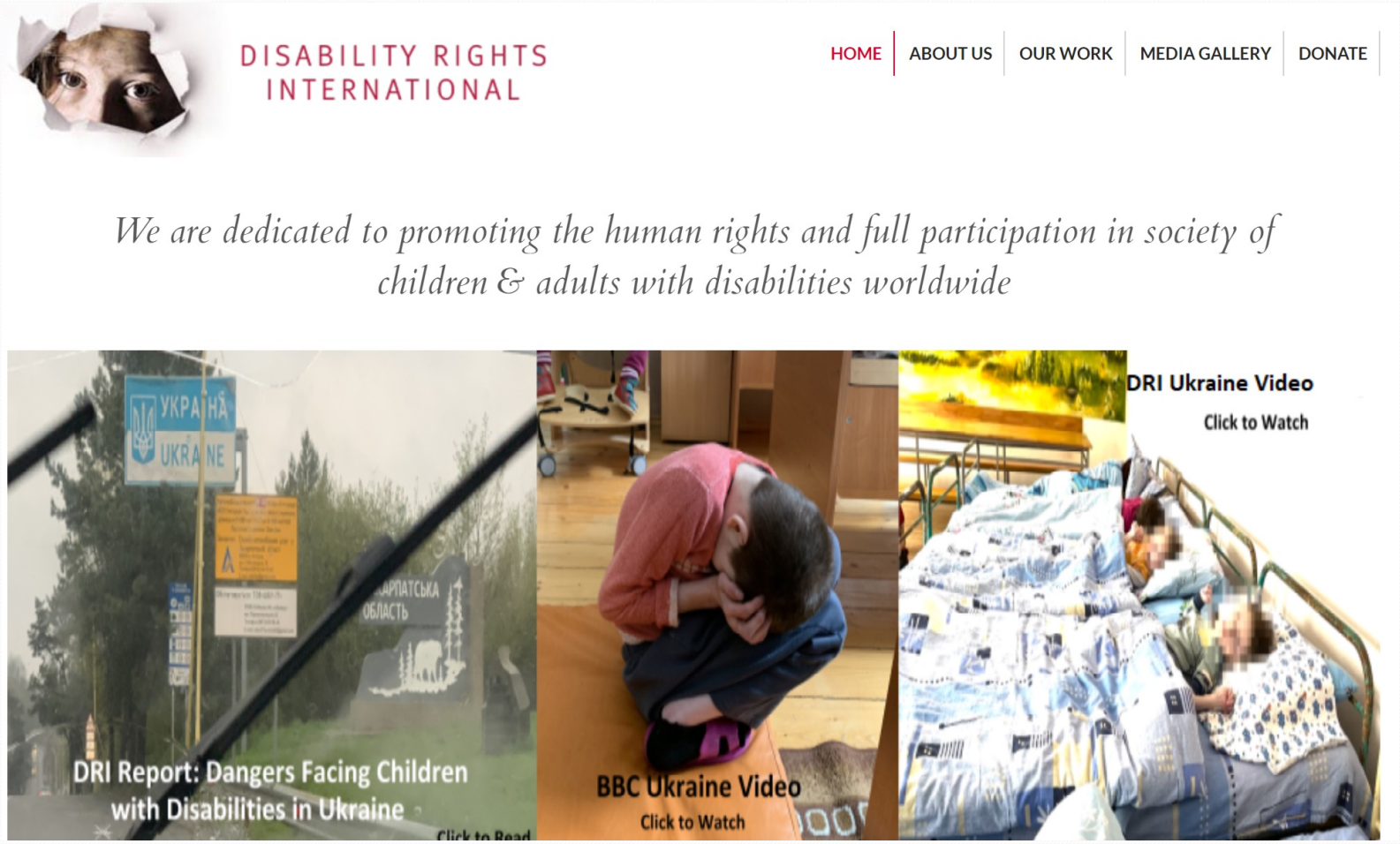
UCEDD/LEND

Health Insurance

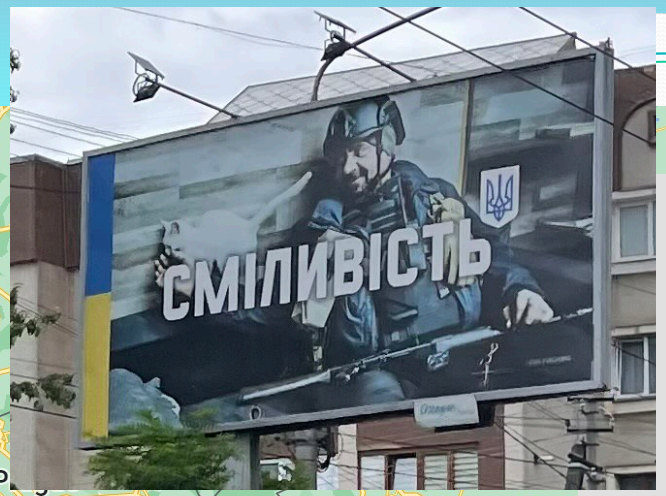
# Back to the Case; more information

- His mother died 1 year ago
- Sister has been taking more of an active role in her life
- What should next steps be for assessment and care??
  - Ask a lot more questions
  - Don’t jump to conclusions

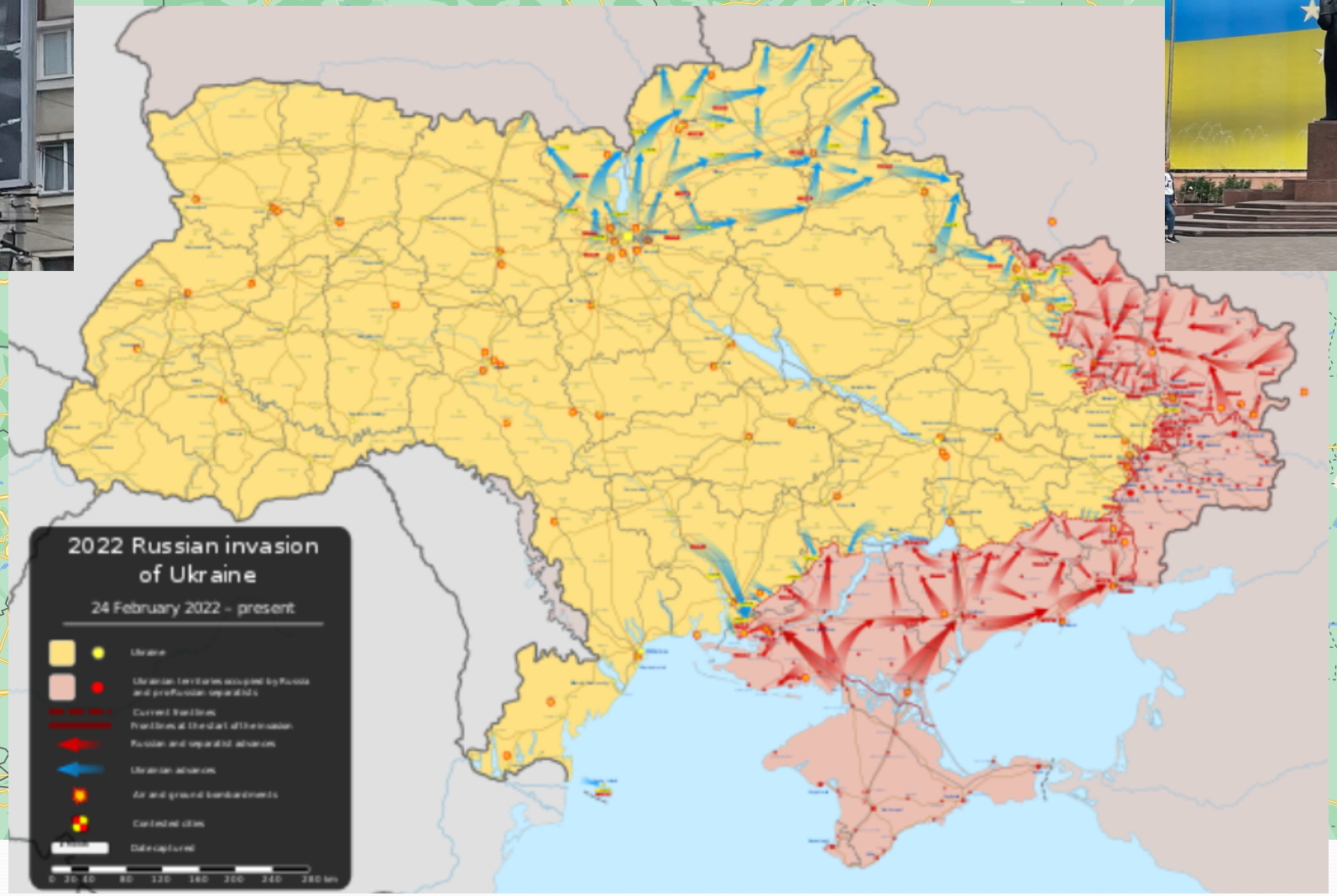
# My Mission to Ukraine; June 3-12, 2022




Disability Rights International - Disability Rights International promotes the human rights and full community integration of persons with disabilities worldwide (driadvocacy.org)

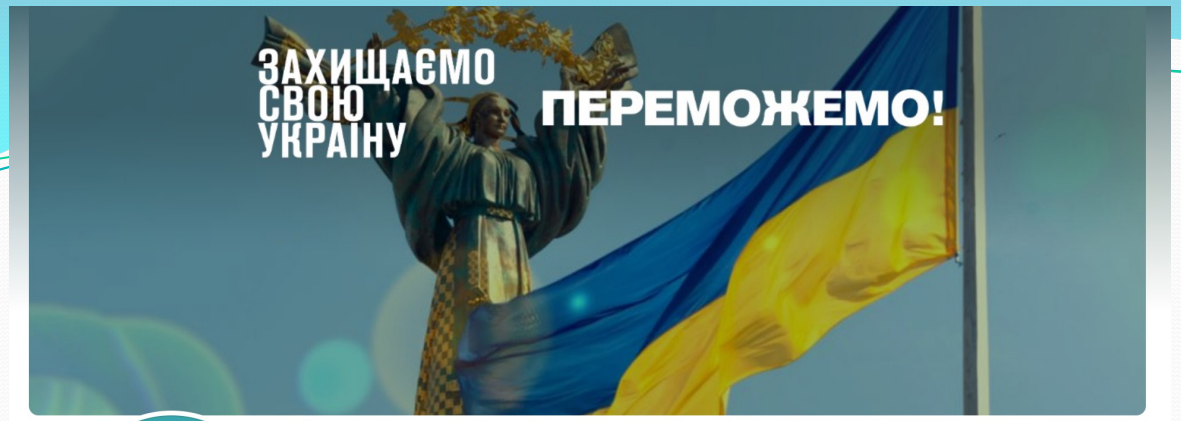







Belarus
Berlin
Czechia
Slovakia
Vienna

[How To Help Disabled Ukrainian Children Left Behind During War (today.com)]




**Чернівецька обласна військова адміністрація**

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# Thank You!!

Intellectual Disabilities and Dementia (the-ntg.org)

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