HEALTHCARE CONCERNS FOR ADULTS AGING WITH INTELLECTUAL DISABILITIES: FOCUS ON COGNITIVE IMPAIRMENT AND DEMENTIA

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- Past President AADMD
- Co-President NTG

- June 8, 2021

ntg
National Task Group on Intellectual Disabilities and Dementia Practices

CATHOLIC CHARITIES HAWAI'I
CIRCLE OF CARE FOR DEMENTIA

Sponsored by

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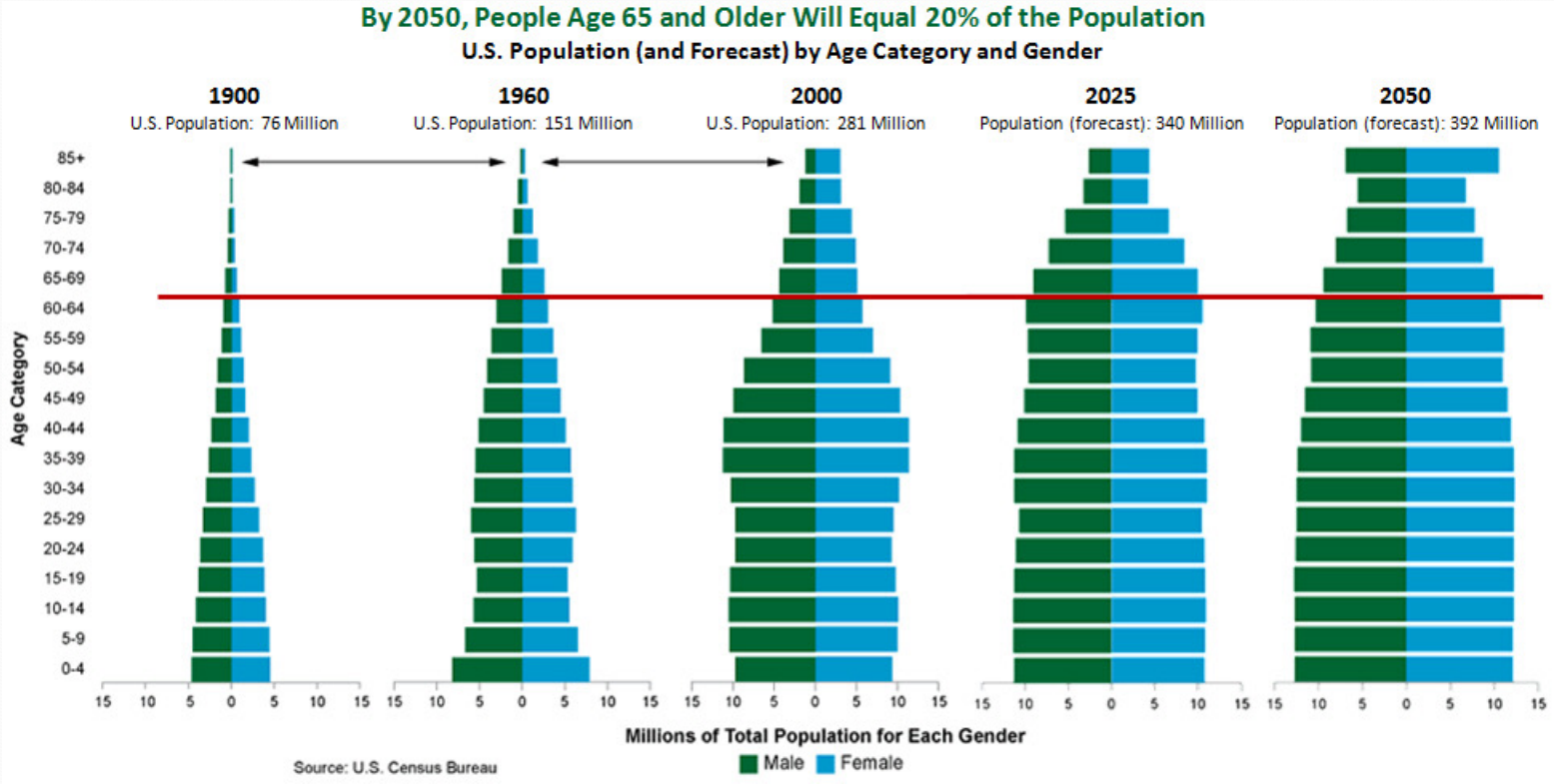
Outline

- Introduction
- When Change Occurs
- Making the Diagnosis of dementia
- Treatment
- Following the course of disease
- Supporting the caregiver
- Next Steps

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

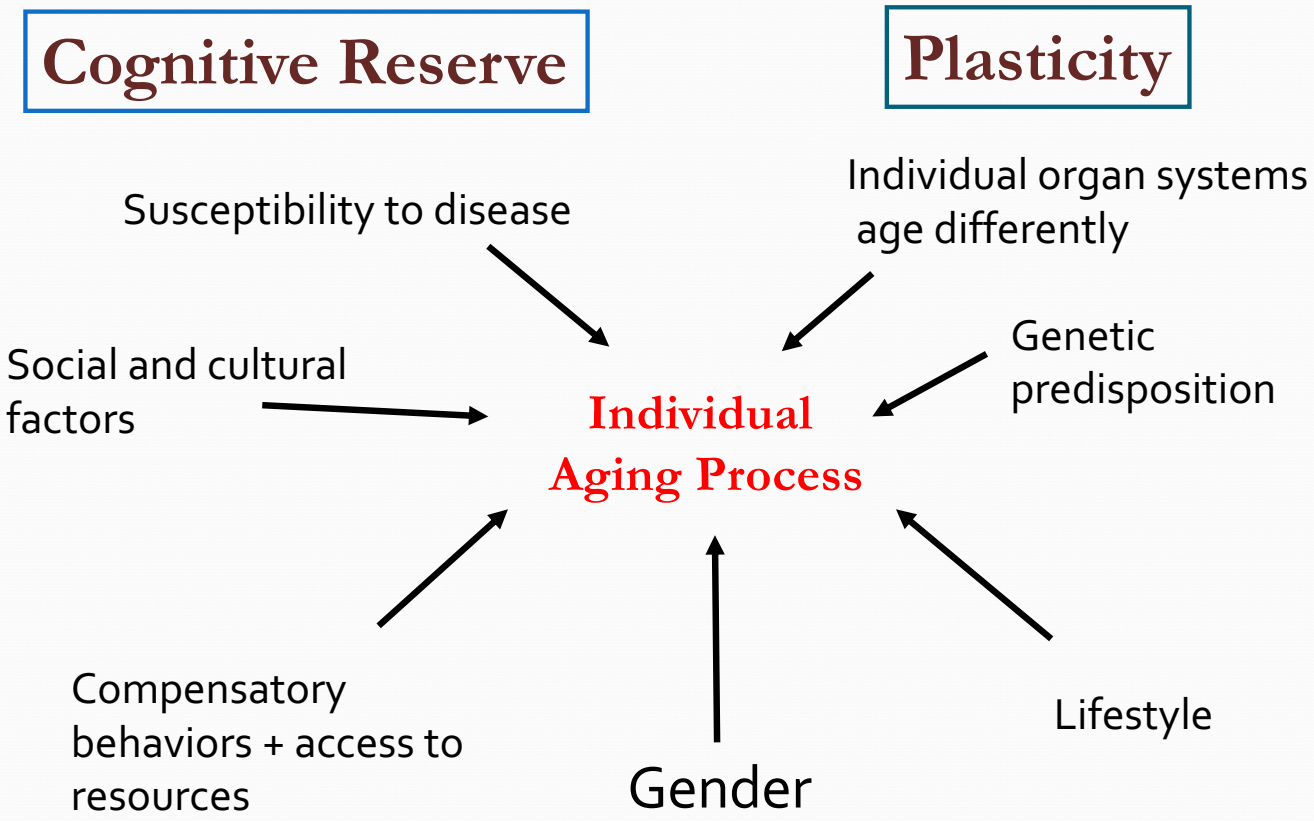
Source: Carter & Jancar, 1983, Janicki, Dalton, Henderson & Davidson, 1999

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

Expected Physical Changes of Aging

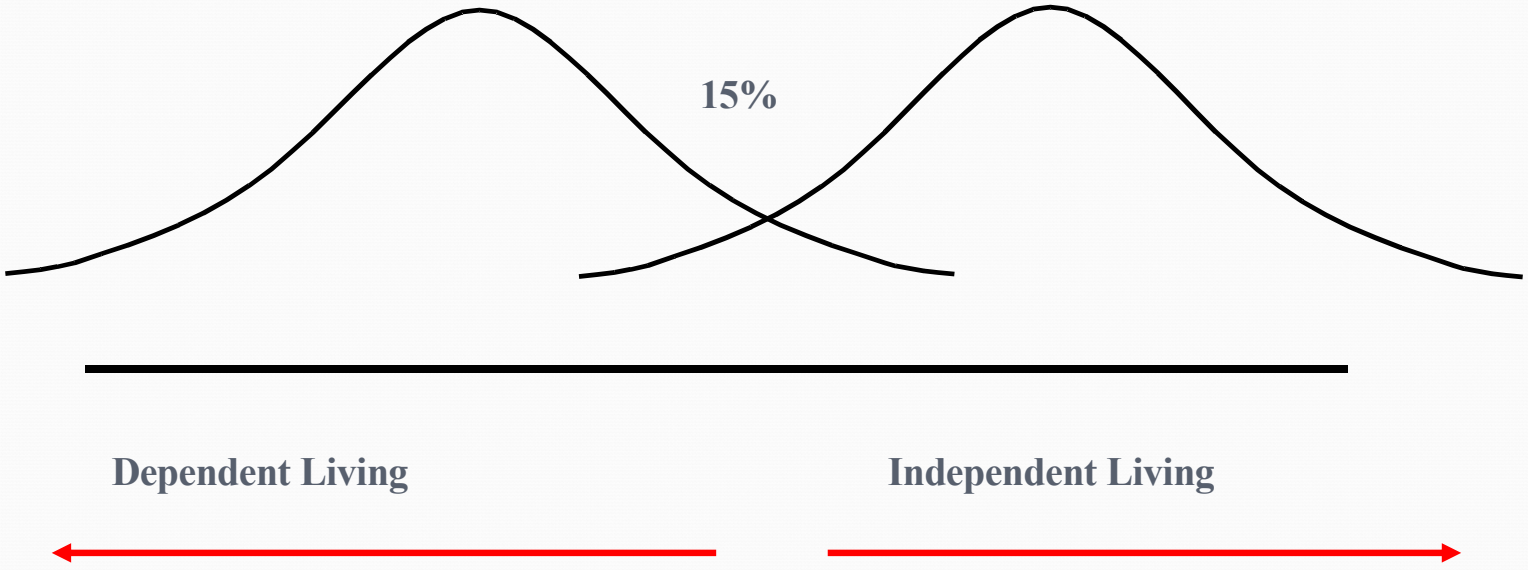
- **Osteopenia/Osteoporosis** - normal aging-related bone loss
- **Sarcopenia** - progressive loss of muscle mass
- **Presbyopia:** the lens of the eye becomes stiffer and less flexible – affecting the ability to focus on close objects (accommodation)
- **Presbycusis** – aging related change in the ability 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



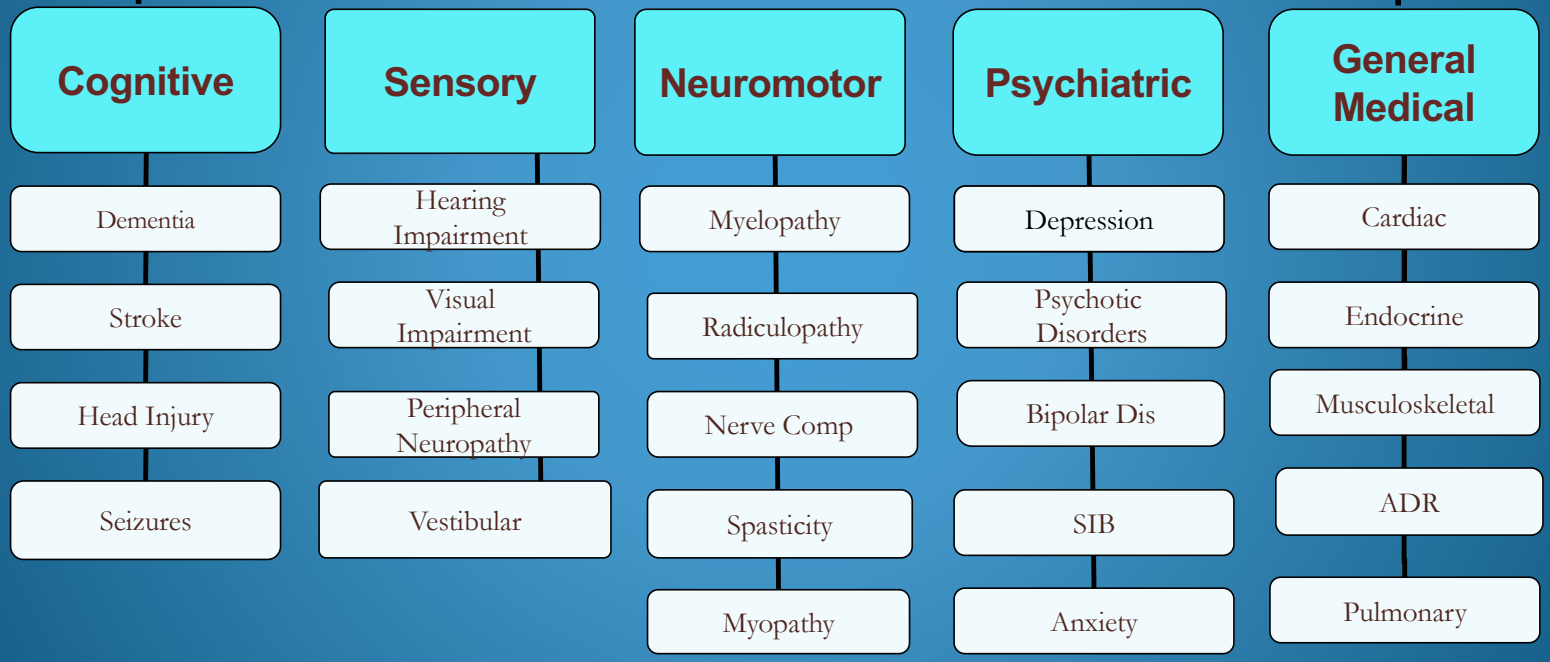
Diversity of the Aging Process

Aging and Decline Affects QOL

Small Change in Cognitive Capability could have profound impact on Independence



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

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!!

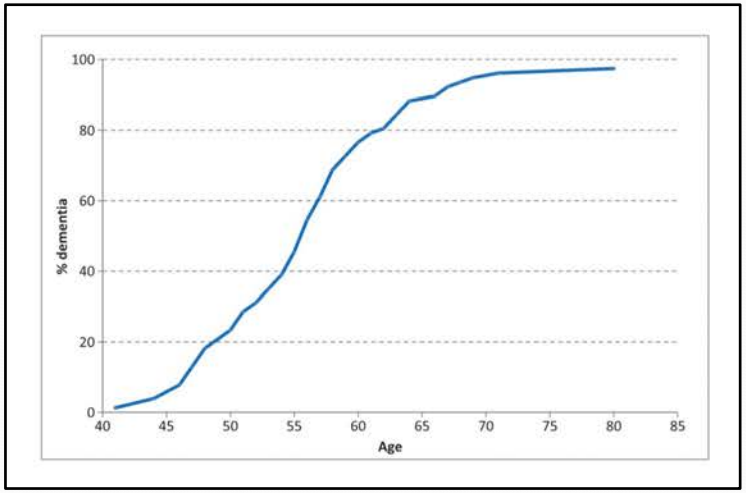
Cognitive Changes with Aging

- 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

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

Early Assessment

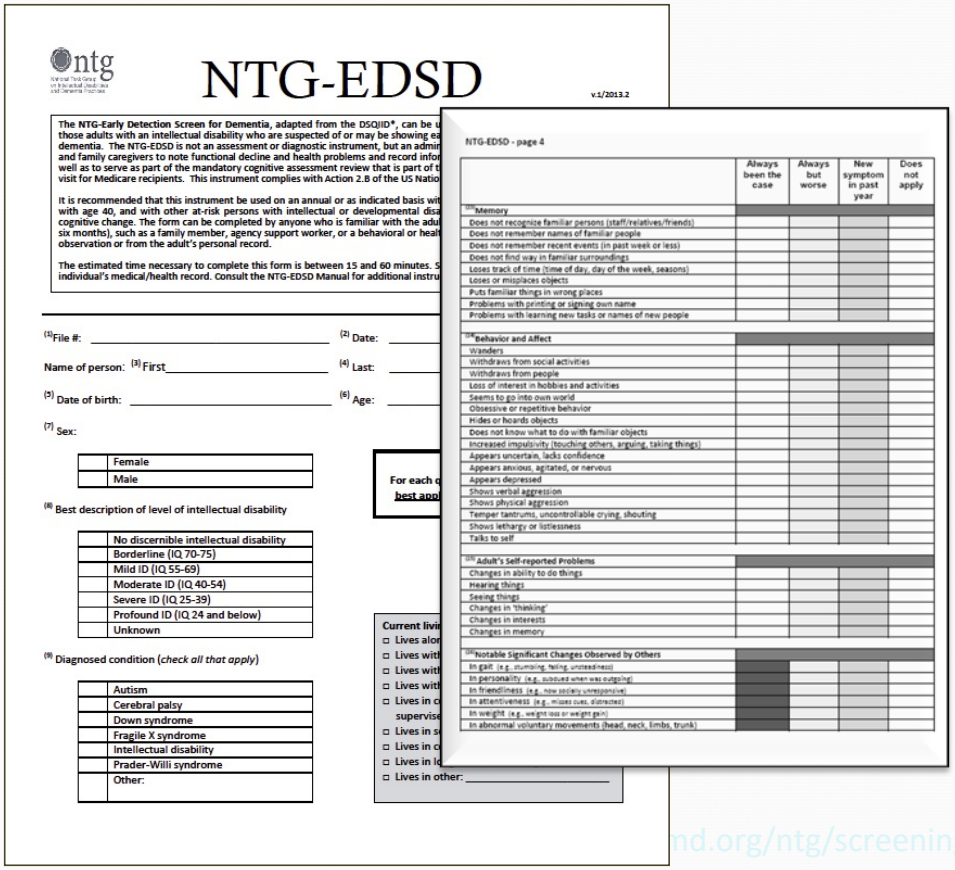
History is the most important part of assessment and it must include:

- Onset and progression of symptoms
- Medical history and medication
- Psychiatric symptoms: focussing on memory, speech, and mood
- Personal history including habits (Life Story)
- Family history
- Recent life changes
- Caregivers account
- NTG-EDSD

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



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NTG-EDSD

v.1/2013.2

The NTG-Early Detection Screen for Dementia, adapted from the DSQIID*, can be u
those adults with an intellectual disability who are suspected of or may be showing ea
dementia. The NTG-EDSD is not an assessment or diagnostic instrument, but an admin
and family caregivers to note functional decline and health problems and record infor
well as to serve as part of the mandatory cognitive assessment review that is part of t
visit for Medicare recipients. This instrument complies with Action 2.B of the US Natio

It is recommended that this instrument be used on an annual or as indicated basis wit
with age 40, and with other at-risk persons with intellectual or developmental disa
cognitive change. The form can be completed by anyone who is familiar with the adu
six months), such as a family member, agency support worker, or a behavioral or healt
observation or from the adult's personal record.

The estimated time necessary to complete this form is between 15 and 60 minutes. S
individual's medical/health record. Consult the NTG-EDSD Manual for additional instru

(1) File #: _____ (2) Date: _____

Name of person: (3) First_____ (4) Last: _____

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

(7) Sex:

	Female
	Male

(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:

For each q
best appl

Current livi
- Lives alon
- Lives with
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- Lives with
- Lives in c supervise
- Lives in s
- Lives in c
- Lives in Ic
- Lives in 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, failing, 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)				

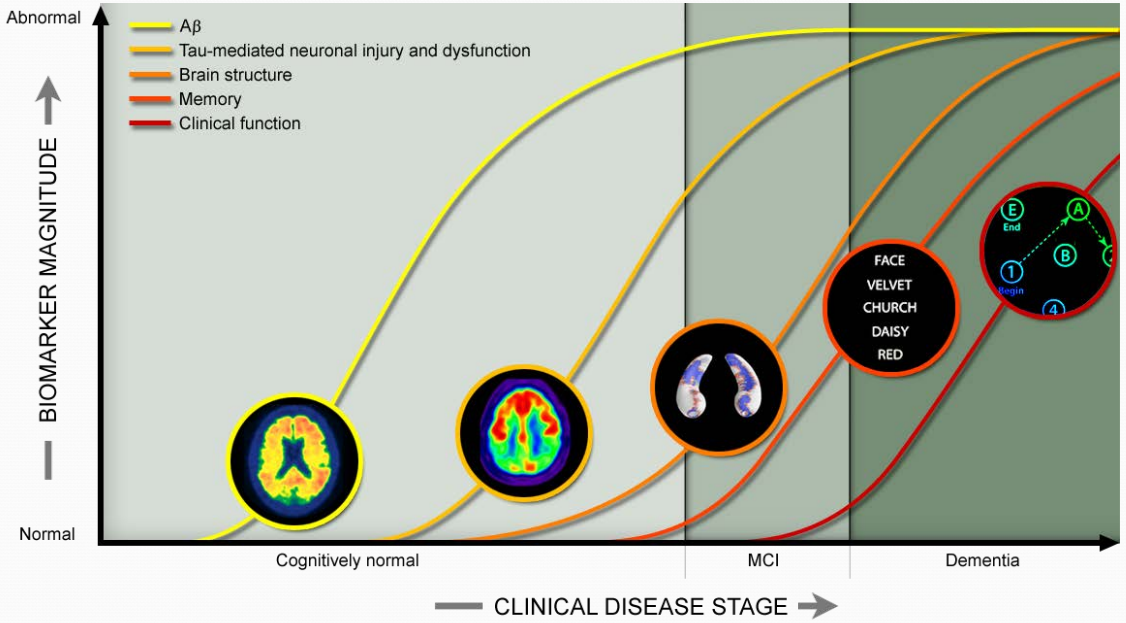
nd.org/ntg/screening

Making the Diagnosis of Dementia

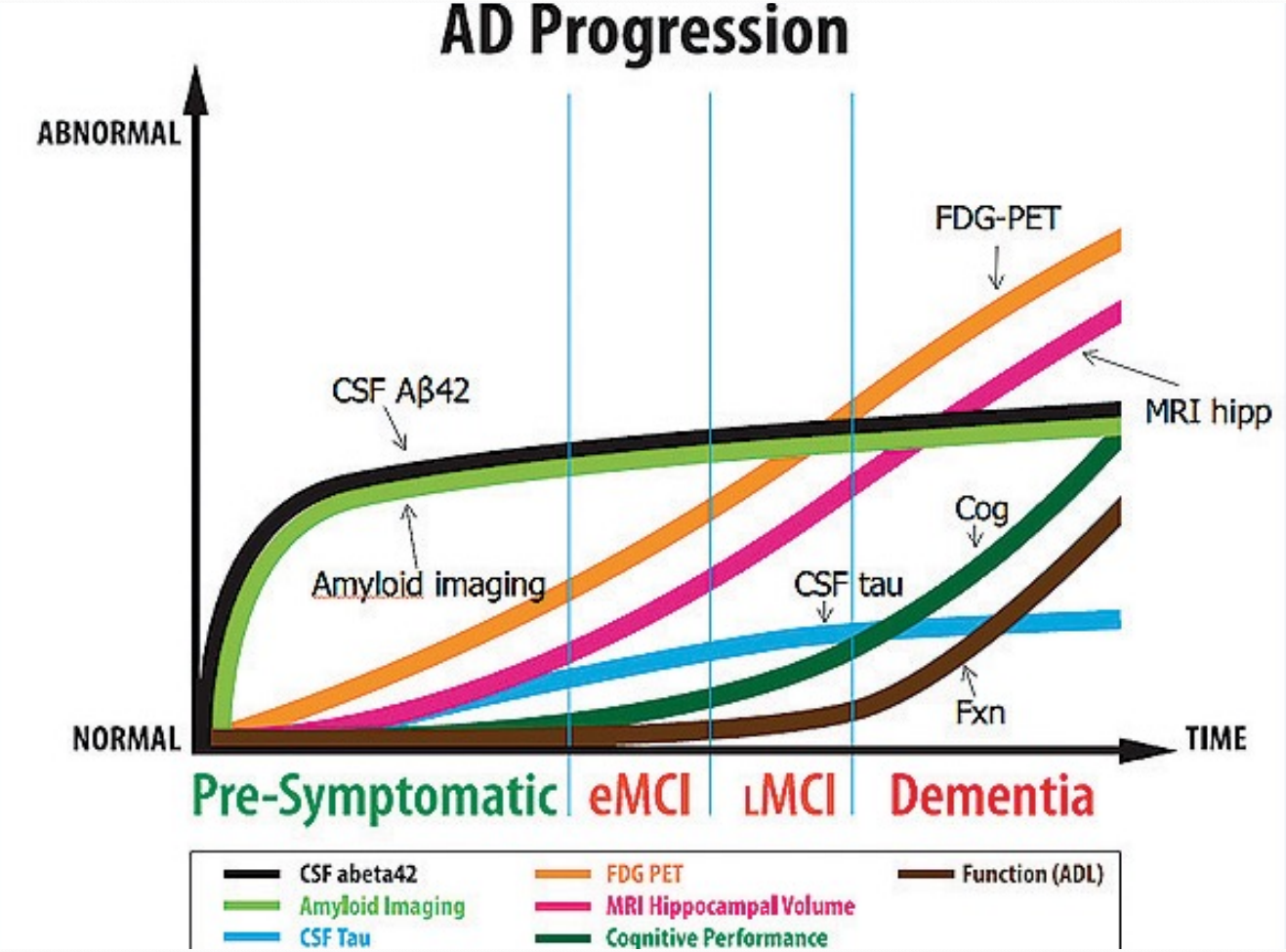
- Having enough information
- Differential diagnosis
  - Hypothyroidism, B12/folate, Sleep Apnea, Depression/Adjustment
  - ADR's, Neuromuscular
- Neuropsychological Assessment
- Imaging
- Biomarkers

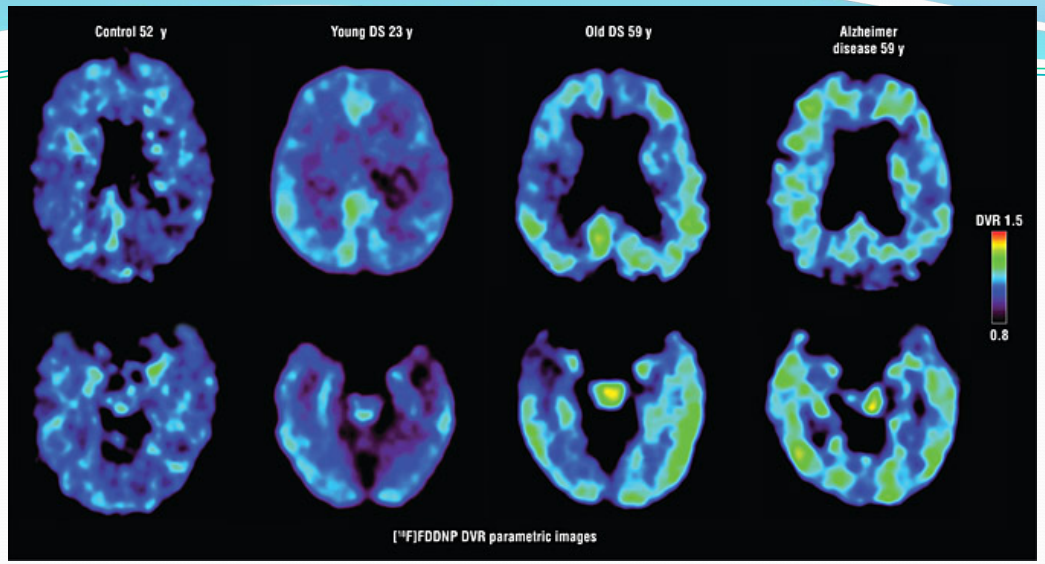
Diagnostic Accuracy

- History of current difficulties
- NTG-EDSD
- Neuropsychological testing
- Physical exam
- Family and social history
- Blood testing
- EEG
- Brain Imaging
- Biomarkers



Alzheimer's Disease Biomarkers



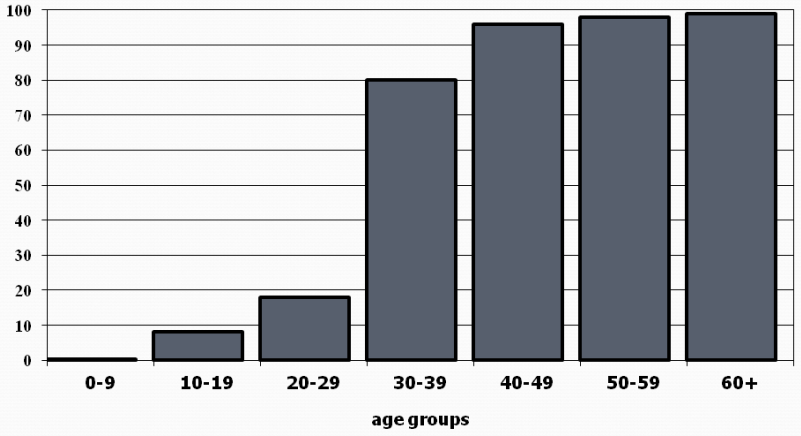


Representative Amyloid Scans in DS and AD

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

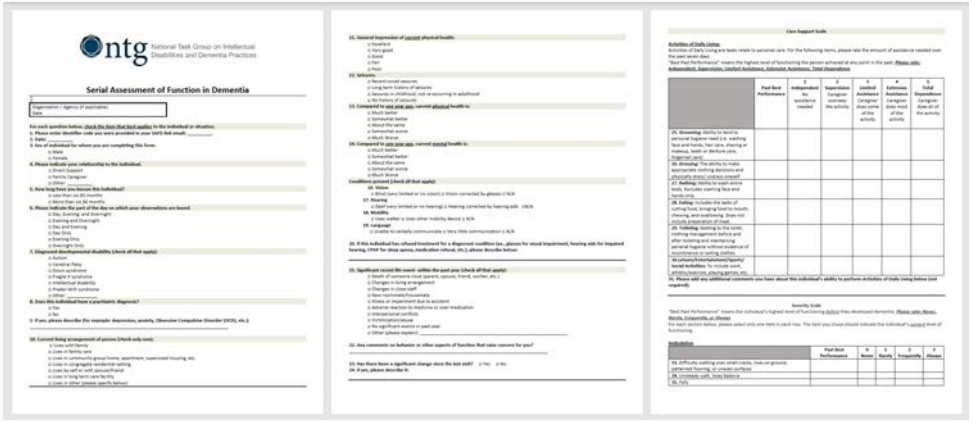


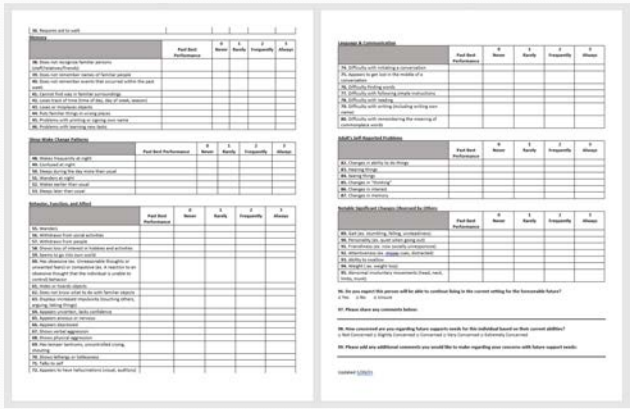
Following Course of Disease

- To determine stage, prognosis, anticipatory guidance, QOL
- Determine levels of supports
- Determine efficacy of therapies
- Determine need for palliative and hospice care

Serial Assessment of Change

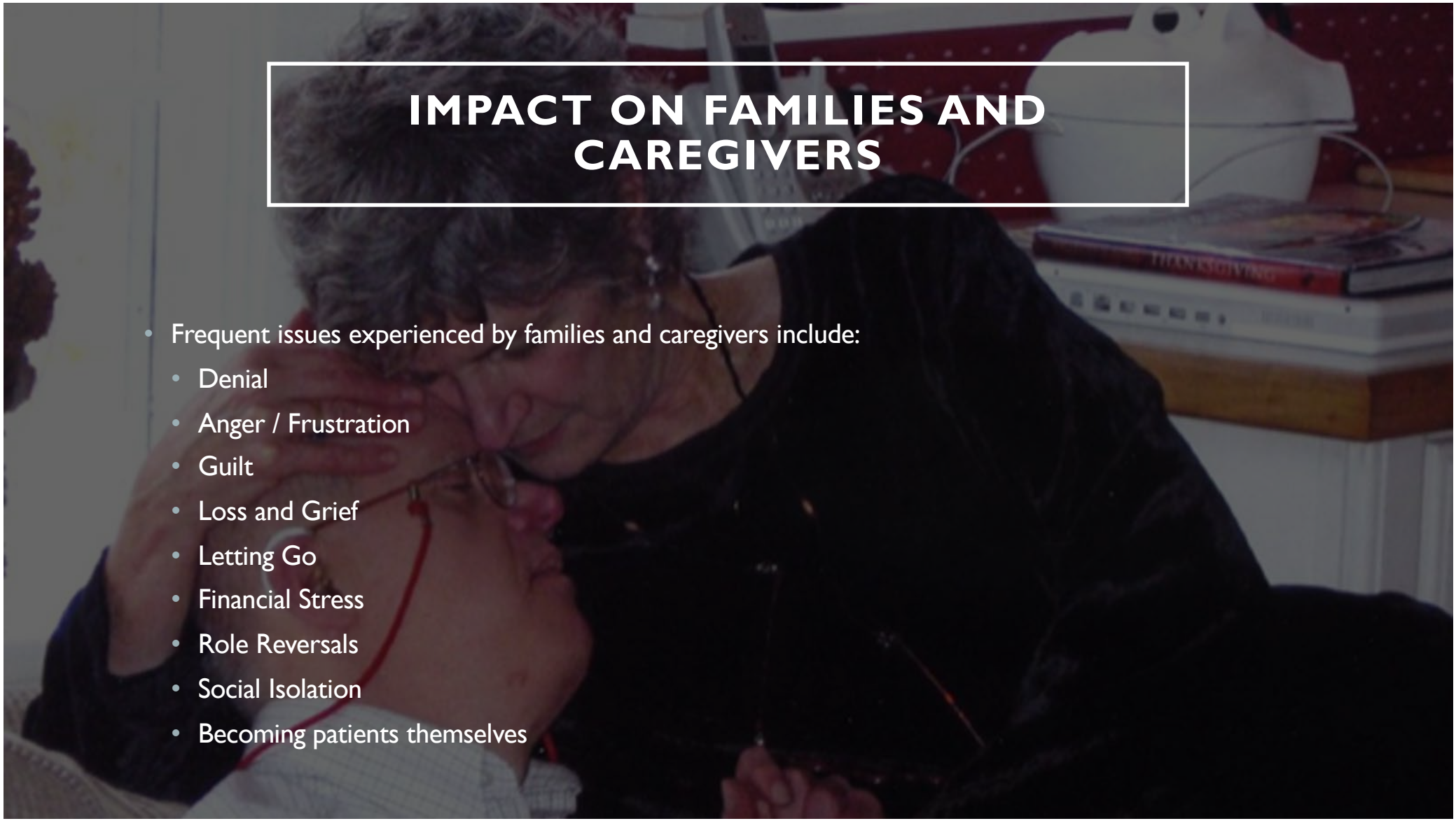
- Serial Assessment of Function in Dementia (SAFD)
- Based on the NTG-EDSD
- Informant Based
- Likert scale
- Care Support Scale
- Severity Scale
- Caregiver support concerns
- May be able to show evidence of serial changes in function due to disease and therapies
- Validity study on going, Jefferson Univ Dept Occ. Therapy. WITH Foundation support





Challenges to diagnosis and care

- Individuals with IDD may not be able to report signs and symptoms
- Subtle changes may not be observed
- Commonly used dementia assessment tools are not relevant for people with IDD
- Difficulty of measuring change from previous level of functioning
- Conditions associated with IDD maybe mistaken for symptoms of dementia - Diagnostic overshadowing
- Aging parents and siblings
- Lack of research, education, and training



IMPACT ON FAMILIES AND CAREGIVERS

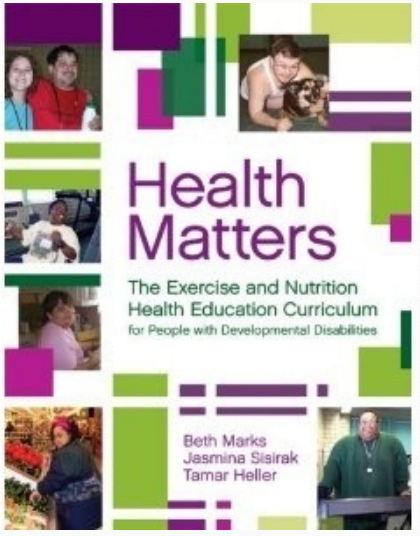
- Frequent issues experienced by families and caregivers include:
  - Denial
  - Anger / Frustration
  - Guilt
  - Loss and Grief
  - Letting Go
  - Financial Stress
  - Role Reversals
  - Social Isolation
  - Becoming patients themselves

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/

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

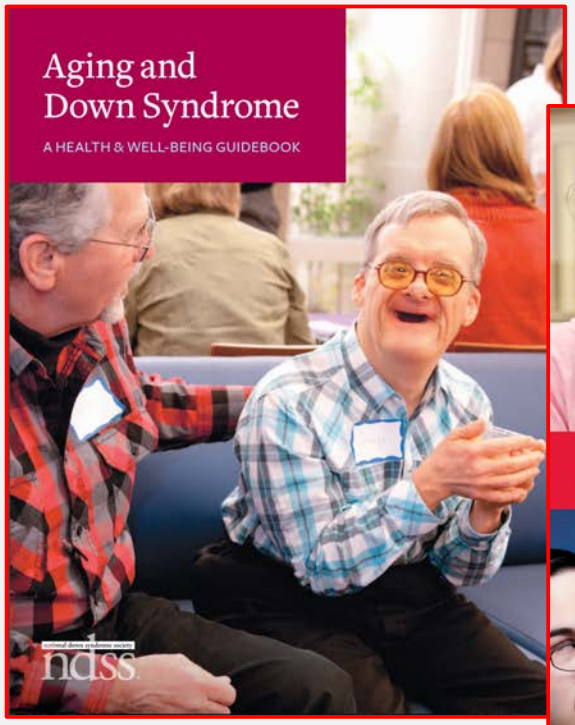
CATHOLIC CHARITIES HAWAIʻI
CIRCLE OF CARE FOR DEMENTIA

ADRC HAWAIʻI
AGING AND DISABILITY RESOURCE CENTER

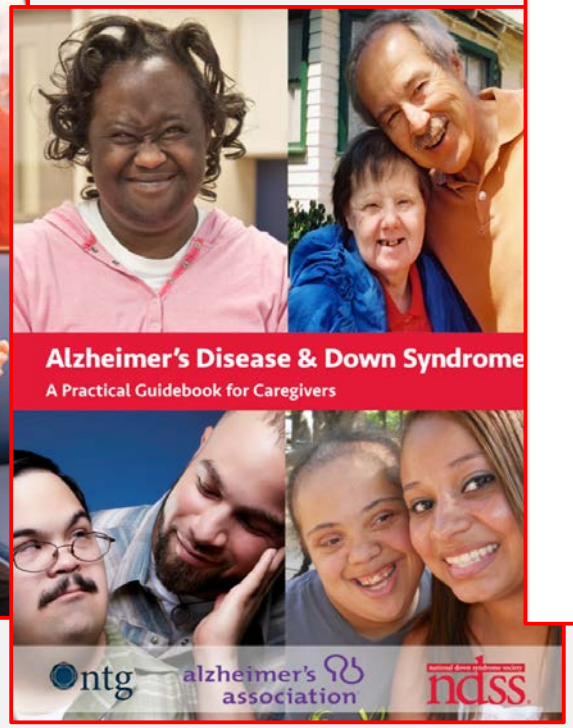
Special Olympics Hawaii

alzheimer's association

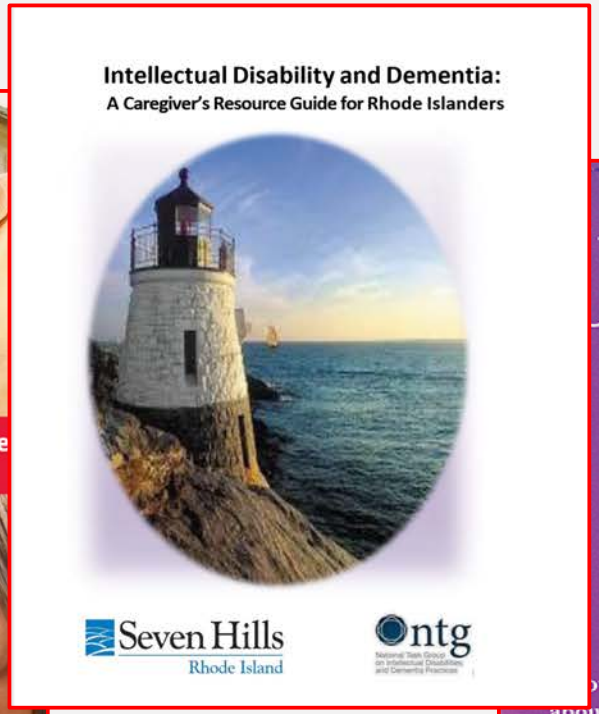
PERSON/FAMILY CENTERED RESOURCES



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

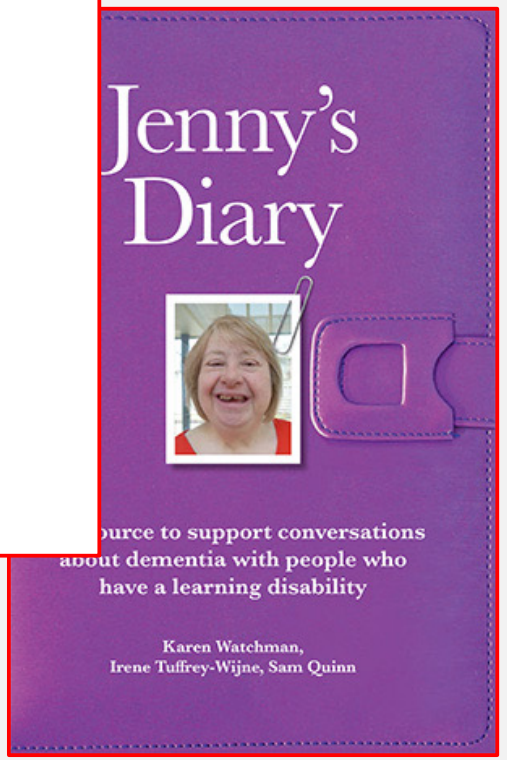


http://www.ndss.org/wp-content/uploads/2017/11/NDSS_Guidebook_FINAL.pdf

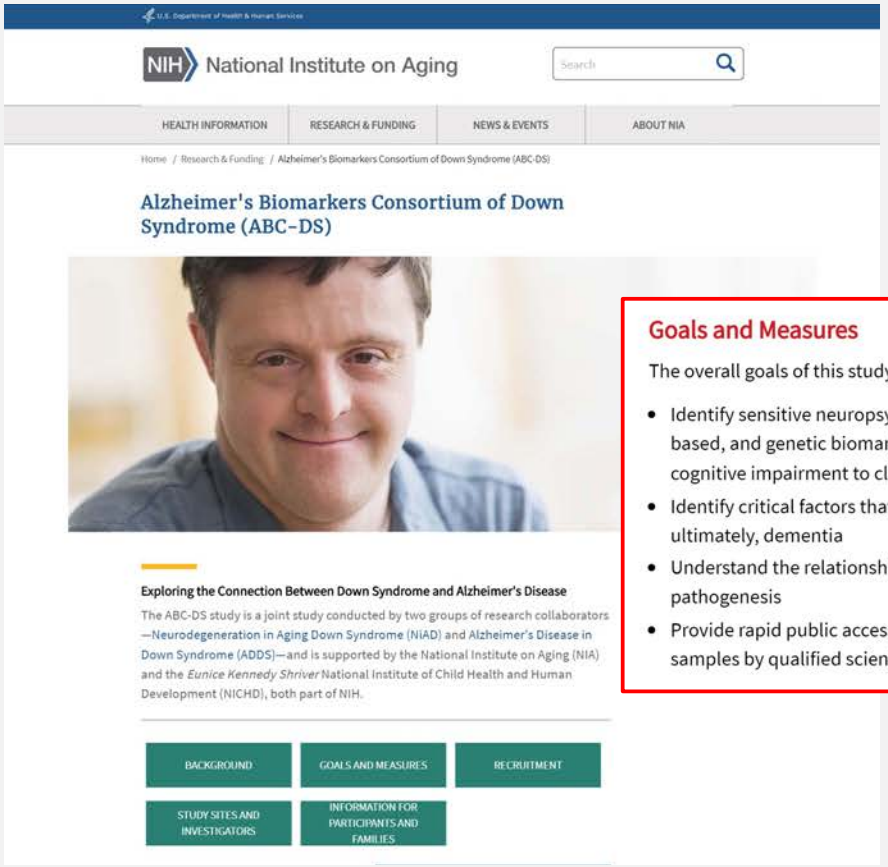


http://www.sevenhills.org/uploads/SHRI-IDD-ADRD-Resource-Guide.pdf

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.

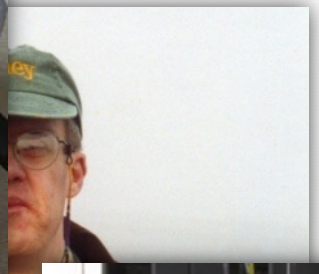
Neurodegeneration in Aging Down Syndrome (NiAD)

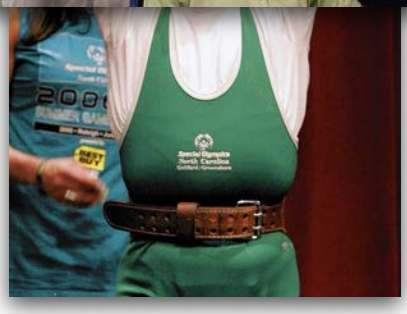
Site	Investigator & Study Coordinator
University of Pittsburgh (Coordinating Center), Pittsburgh, PA	Ben Handen, Ph.D., Co-PI William Klunk, M.D., Ph.D., Co-PI Cathy Wolfe, Study Coordinator
University of Wisconsin Madison, WI	Brad Christian, Ph.D., Co-PI Renee Makuch, Study Coordinator
Barrow Neurological 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
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 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
The New York State Institute for Basic Research in Developmental Disabilities Staten Island, NY	Sharon Krinsky-McHale, Ph.D., Site PI Deborah Pang, Study Coordinator
University of North Texas Health Science Center Fort Worth, TX	Sid O'Bryant, Ph.D., Site PI

Who do you see?





Where do individuals and their supports now go to get assessment, diagnosis and long term care?

What is the best way to document early changes in function?

How can the care of those with dementia be improved?

How can a team approach to care be created and sustained? Who are the team members?


Thank You!!
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Intellectual Disabilities and Dementia (the-ntg.org)