

ALZHEIMER'S DISEASE IN ADULTS WITH DOWN SYNDROME

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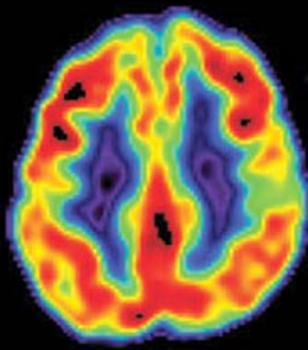
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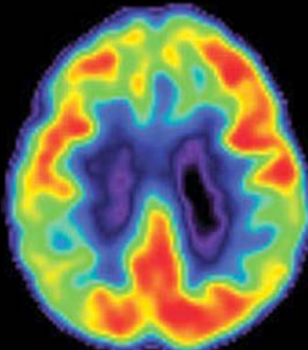
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ALZHEIMER'S DISEASE

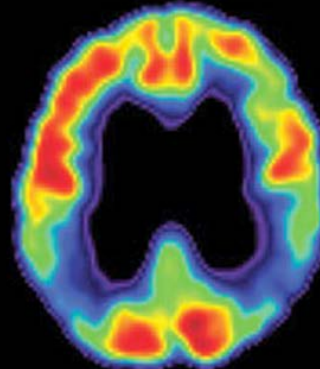
- Most common type of dementia
- Progressive deterioration of cognitive functioning that ultimately prevents performance of everyday activities



Normal



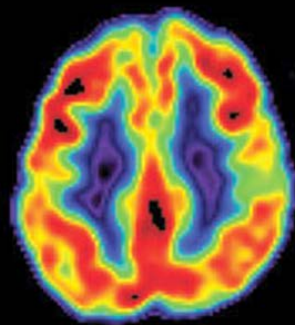
Mild cognitive
impairment



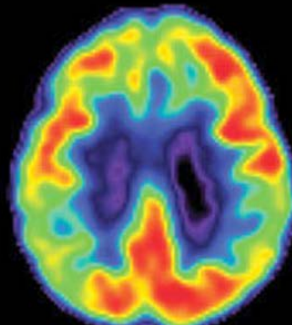
Alzheimer's
disease

ALZHEIMER'S DISEASE

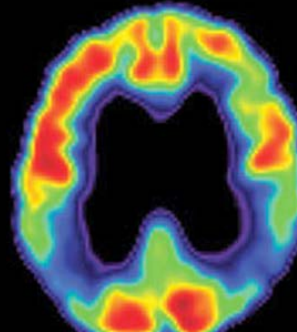
- 5.3 million Americans; 110,000 in Wisconsin
- Prevalence will increase; ~ 10,000 Baby Boomers turn 65 per day
- 7th leading cause of death



Normal



Mild cognitive
impairment



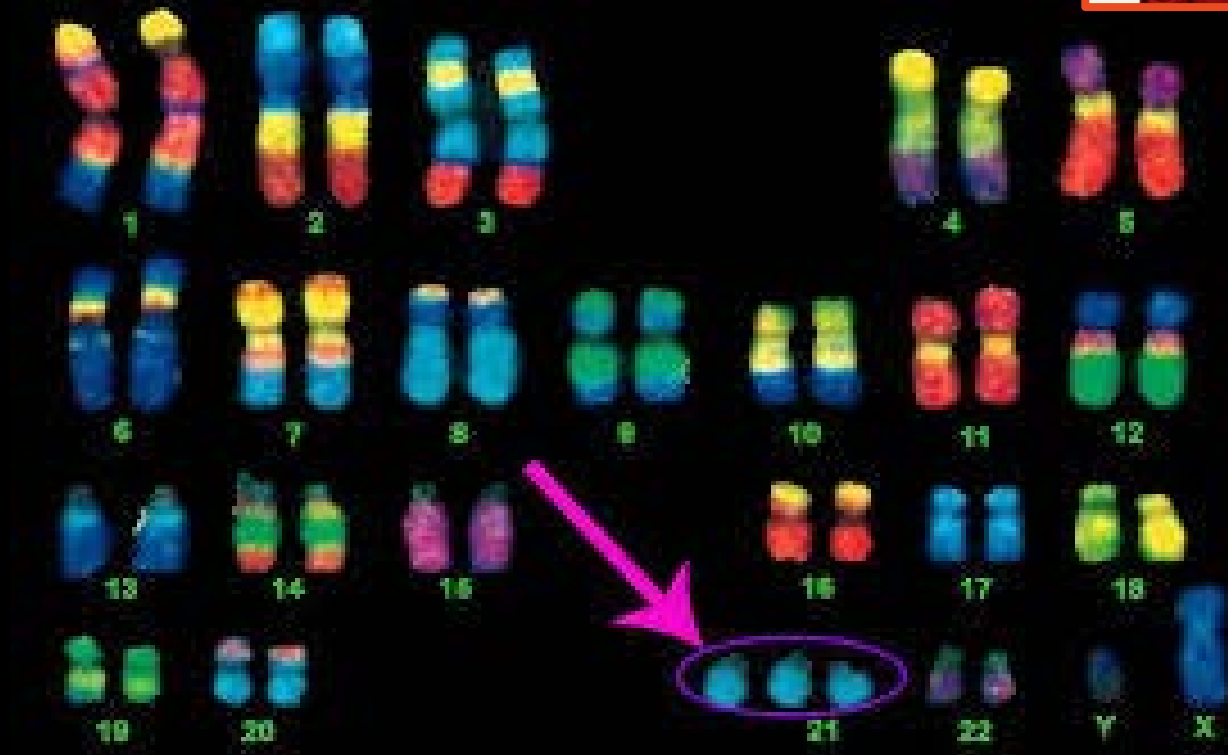
Alzheimer's
disease

ALZHEIMER'S DISEASE

- No cure
- Treat symptoms; temporarily slow the progression of disease
- Critical need to find ways to treat the disease, delay onset, and prevent



DOWN SYNDROME



Reprinted from Shaw, 2013

DOWN SYNDROME



- 1 in 800 live births worldwide; ~255,000 children in US
- Intellectual disability
- Impairments in language, motor, and cognitive skills
- Facial appearance - flat face, short neck, slanting eyes, etc.
- Physical features - low muscle tone, loose joints
- Health conditions - problems with thyroid, heart, intestines, hearing loss

DOWN SYNDROME AND ALZHEIMER'S DISEASE

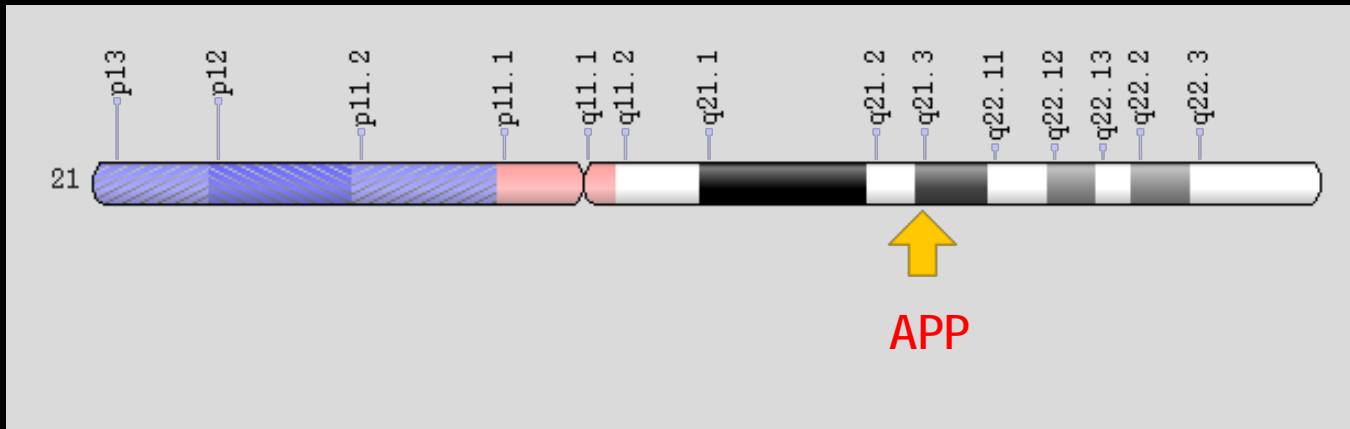


General population: Rare before age 50; 5-10% of adults aged 65+ yrs; 15-30% of those aged 80+ yrs

Down syndrome: 9% of adults in 40; 33% of adults in 50s; 50% of adults in 60s+ yrs

WHY THE HEIGHTENED RISK?

- Chromosome 21 codes for the amyloid- β precursor protein (APP) gene



- Accumulation of amyloid- β plaques in brain plays key role in development of Alzheimer's disease

NEUROPATHOLOGY IN ALZHEIMER'S DISEASE

Healthy



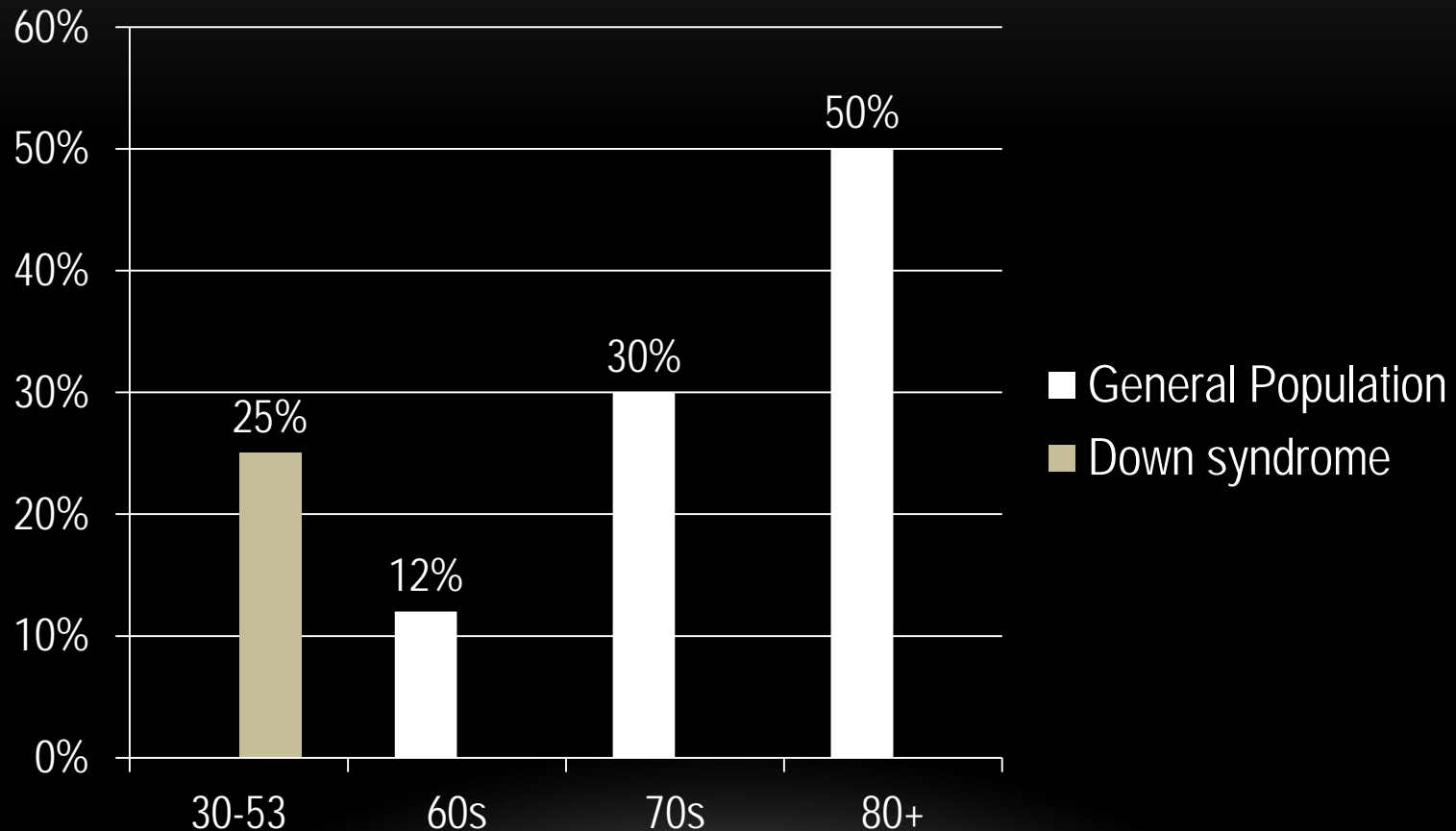
Alzheimer's Disease



Neurofibrillary
tangles

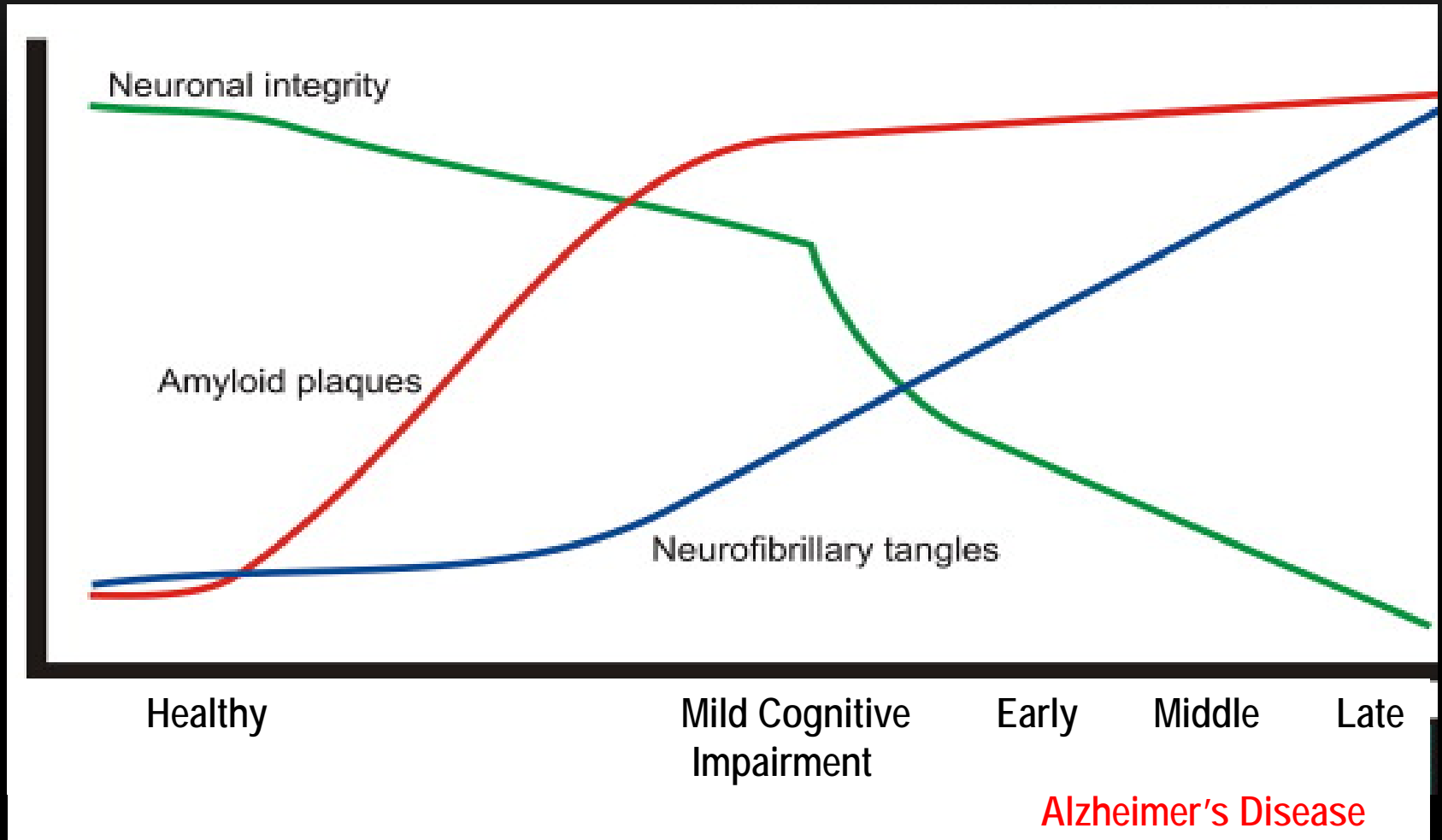
Amyloid- β
plaques

PERCENTAGE OF HEALTHY ADULTS WITH HIGH AMYLOID-B PLAQUES



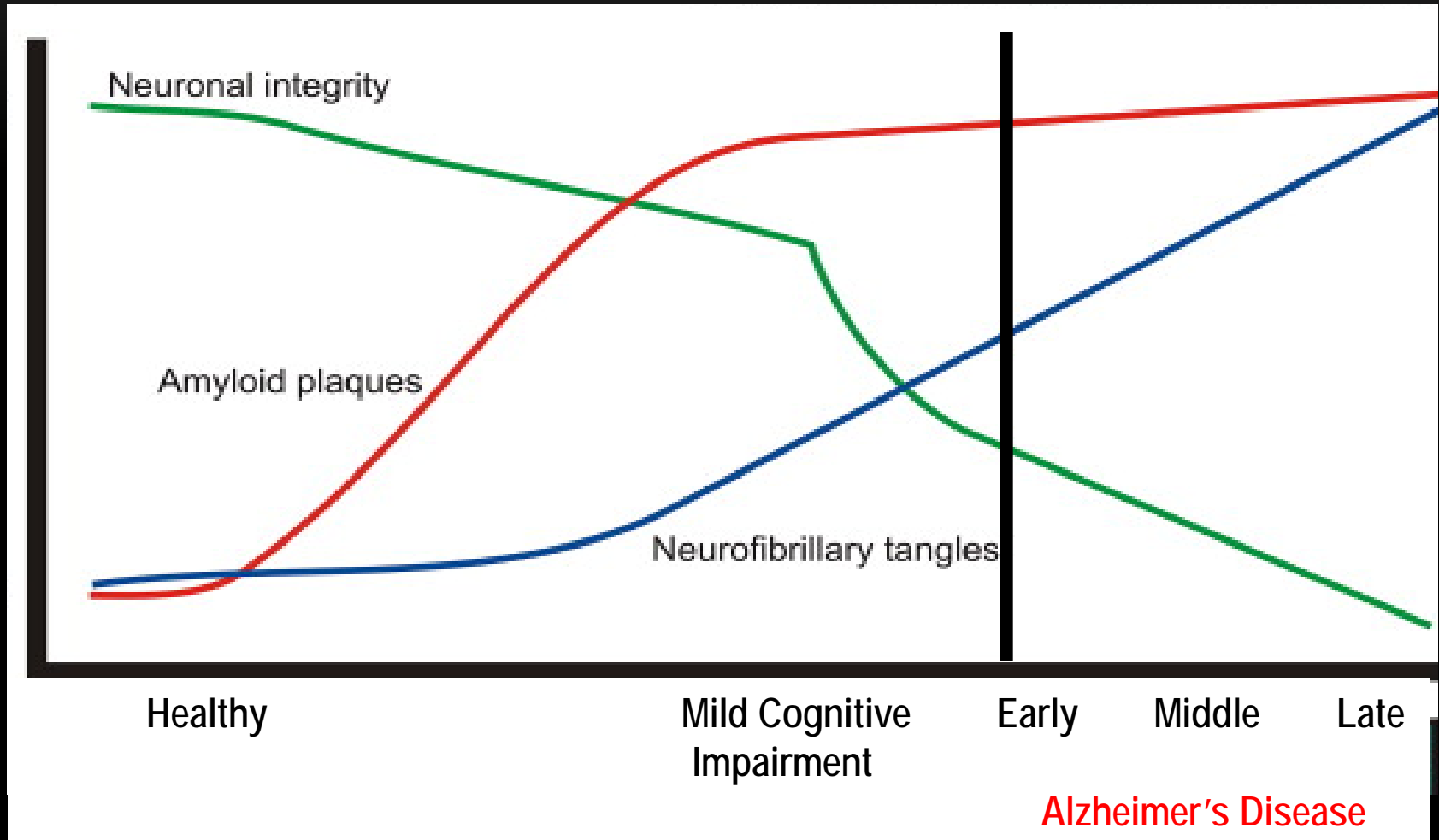
General population data: Rowe & Villemagne, 2011
Down syndrome data: Hartley et al., 2014

PROGRESSION OF ALZHEIMER'S DISEASE



Reprinted from Tarawneh & Holtzman, 2010

PROGRESSION OF ALZHEIMER'S DISEASE



Reprinted from Tarawneh & Holtzman, 2010

NEURODEGENERATION IN AGING DOWN SYNDROME (NIAD STUDY)

- Track early brain changes associated with Alzheimer's disease in adults with Down syndrome
- How does Alzheimer's disease develop? When could we intervene? Why do symptoms progress faster in some individuals than others? Can we come up with accurate early screeners?



University of Pittsburgh



University of Cambridge, UK



Waisman Center, University of Wisconsin-Madison

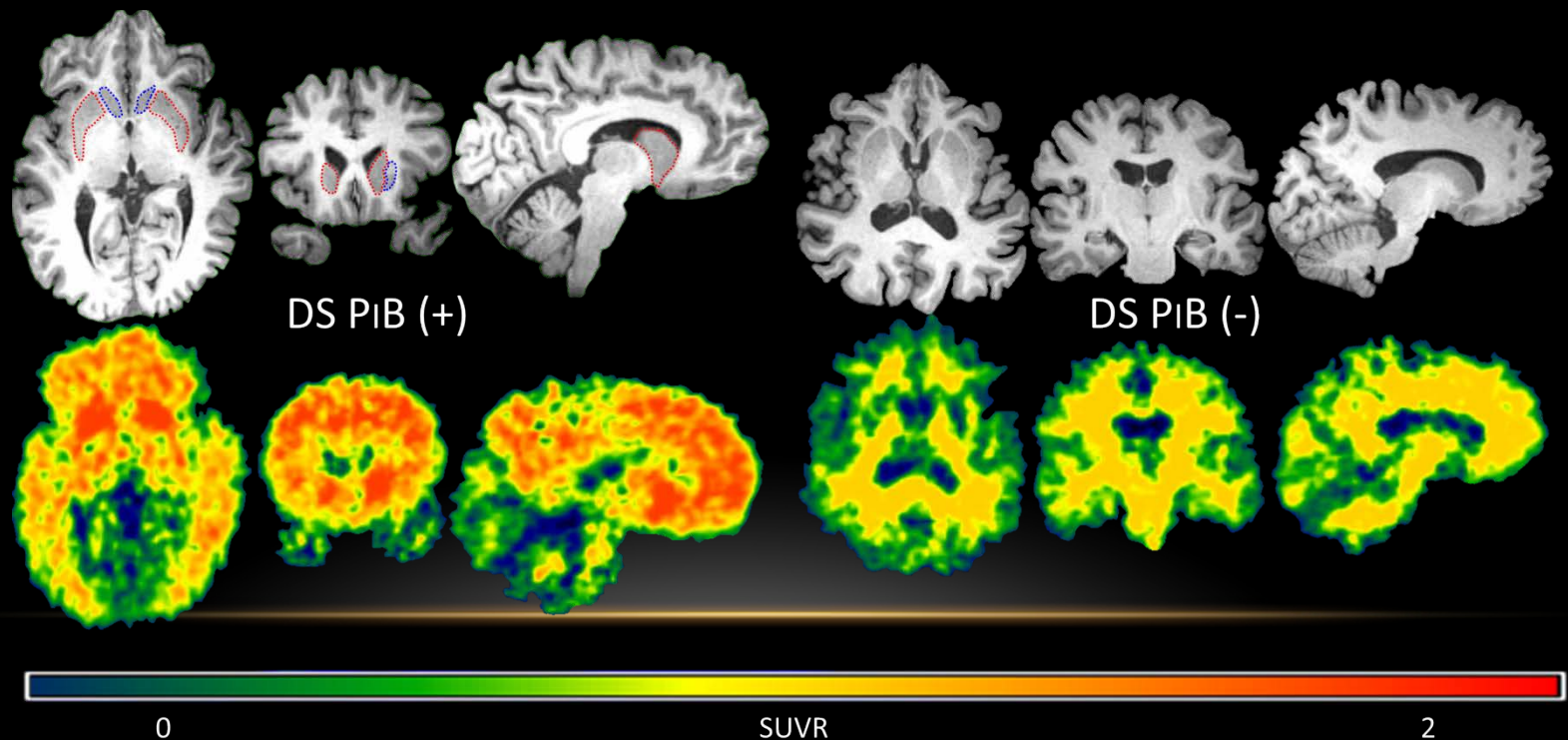
BRAIN IMAGING

- Magnetic resonance imaging (MRI)
- Positron emission tomography (PET)



AMYLOID-B

- Tissue ratios calculated for cortical regions-of-interest (ROI) and normalized to cerebellum (SUVR)
- PiB+ = above the cutoff in cortical areas of the brain



NEUROPSYCHOLOGICAL TESTS




SCREENING AND DIAGNOSIS ALZHEIMER'S DISEASE IN DOWN SYNDROME



SCREENING INTERVIEWS

- National Task Group Early Detection Screen for Dementia (NTG-EDSD)
- Dementia Scale for Down Syndrome (DSDS)



National Task Group
on Intellectual Disabilities
and Dementia Practices

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 _____ (4) Last: _____

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

(7) Sex:

<input type="checkbox"/>	Female
<input type="checkbox"/>	Male

(8) Best description of level of intellectual disability

<input type="checkbox"/>	No discernible intellectual disability
<input type="checkbox"/>	Borderline (IQ 70-75)
<input type="checkbox"/>	Mild ID (IQ 55-69)
<input type="checkbox"/>	Moderate ID (IQ 40-54)
<input type="checkbox"/>	Severe ID (IQ 25-39)
<input type="checkbox"/>	Profound ID (IQ 24 and below)
<input type="checkbox"/>	Unknown

(9) Diagnosed condition (check all that apply)

<input type="checkbox"/>	Autism
<input type="checkbox"/>	Cerebral palsy
<input type="checkbox"/>	Down syndrome
<input type="checkbox"/>	Fragile X syndrome
<input type="checkbox"/>	Intellectual disability
<input type="checkbox"/>	Prader-Willi syndrome
<input type="checkbox"/>	Other: _____

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

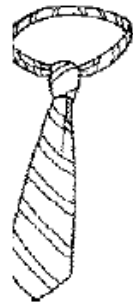
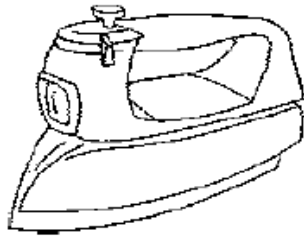
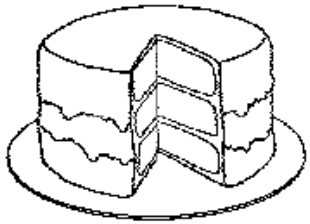
Current living arrangement of person:

- ☐ 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: _____

DIRECT ASSESSMENTS

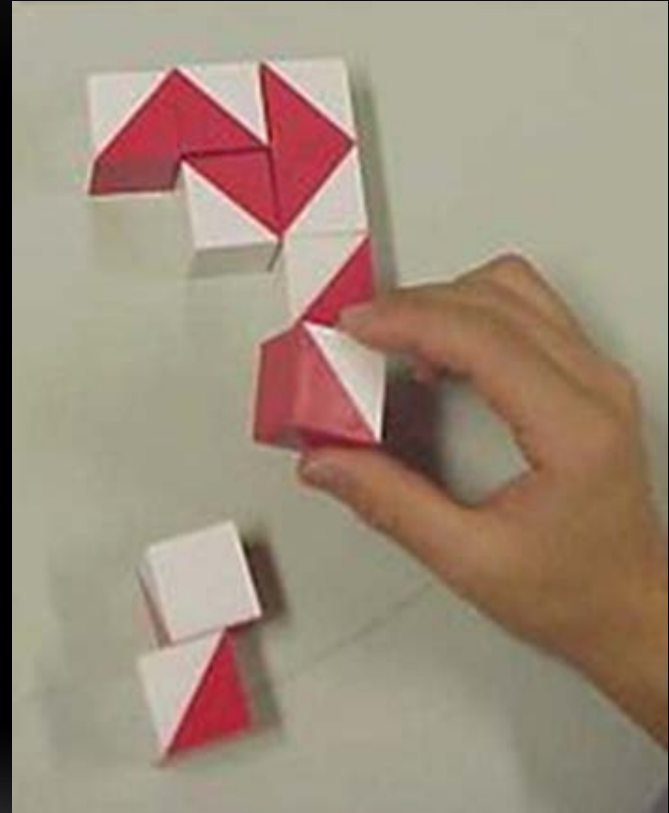
Area	Measures
Verbal Learning/Memory	Cued Recall Test, Wechsler Memory Scale, 4 th edition Story Recall Logical Memory I and II subtests
Visual Memory	Rivermead Behavioral Memory Test for Children Visual Memory subtests
Attention/Processing Speed	WISC-Revised Digits Forward, Corsi Block Tapping Forward, NEPSY Visual Attention subtest
Executive Functioning/Working Memory	Stroop Dog and Cat Task, WISC-IV Digit Span Backwards, Corsi Block Tapping Backward
Visuospatial Construction	Developmental Test of Visual-Motor Integration, 5 th edition, Purdue Pegboard, WISC-IV Block Design and Haxby Extension
Language	NEPSY 2 nd edition Word Generation Semantic Fluency subtest, Expressive-One Word Picture Vocabulary Test, Peabody Picture Vocabulary Test, 4 th edition

MEMORY





VISUOSPATIAL ORGANIZATION

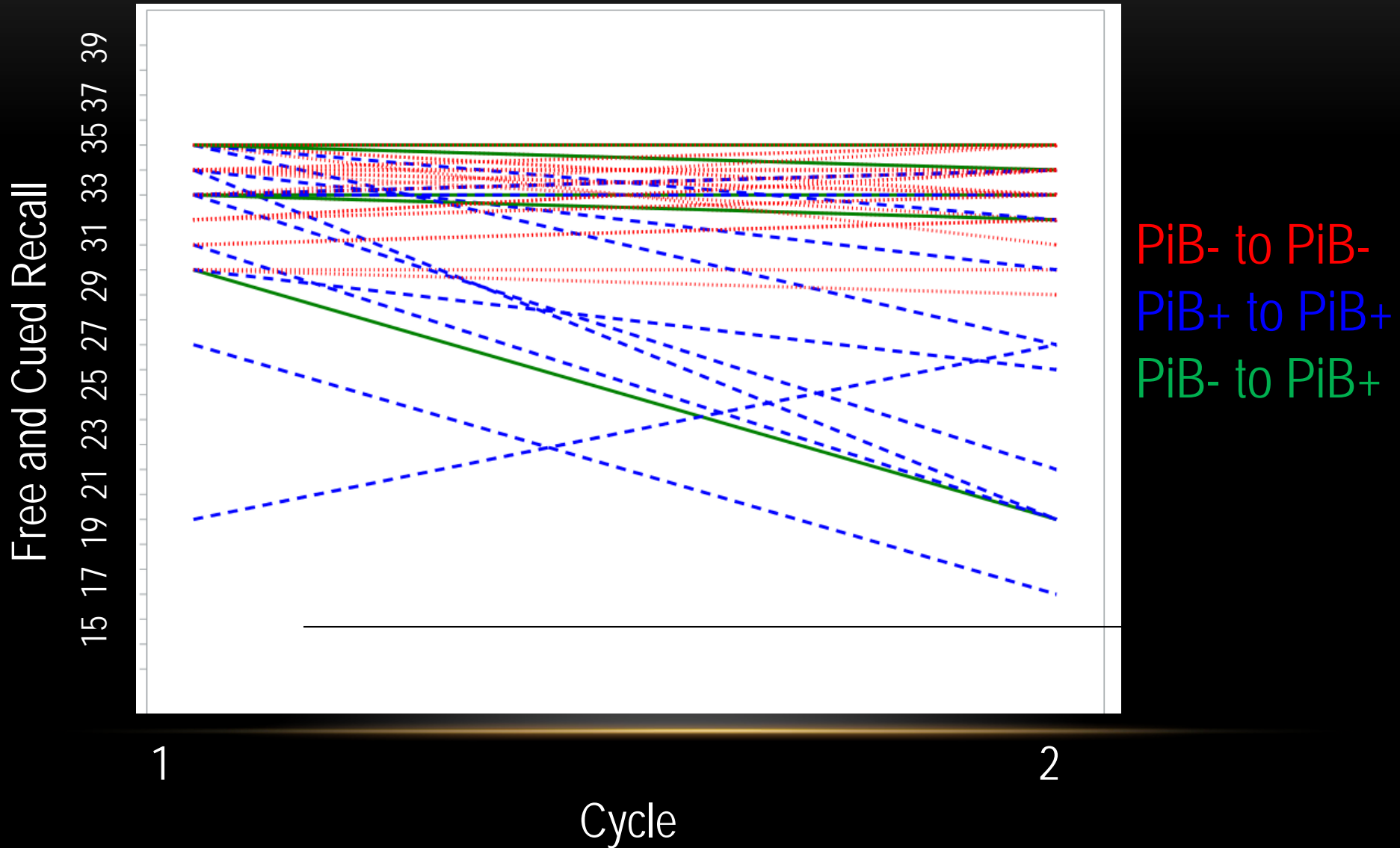


EXECUTIVE FUNCTIONING



BLUE	RED	YELLOW	ORANGE
GREEN	BLUE	PURPLE	RED
PURPLE	YELLOW	RED	BLUE
ORANGE	BLUE	YELLOW	RED
RED	GREEN	ORANGE	BLUE
PURPLE	YELLOW	BLUE	ORANGE

CHANGE CYCLE 1 TO CYCLE 2 BY PIB GROUP



BASELINE AND CONTEXT

- **Baseline assessment** by age 35 years
 - cognitive abilities, memory, motor functioning, everyday living skills, and social and behavioral functioning
- **Consider medical conditions**
 - Vision loss/impairment, hearing loss, hypothyroidism, sleep apnea, celiac disease
- **Consider life transitions**
 - Transfer of care, death of parents, work or staff transitions

COMMUNICATION TIPS FOR PROFESSIONALS AND CAREGIVERS



COMMUNICATING

- **Body language** – your mood affects their mood
- **Positive non-verbal communication** – comfort, care, and demonstration
- **Gain attention** - sit in front of them and at same level
- **Simple and clear** - break down activities into a set of simple (one-step) instructions; speak clearly and at a natural rate of speech
- **Avoid open-ended questions or conversations require recent memory** – may add confusion and agitation
- **Distract and redirect** – go for a walk, change the mood

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Patrick Lao

Michael Bergnic

Christy Matan

Chester Mathis

Tami Hogg

Regina Hardison

Ansel Hillmer

Rameshwari

Tumuluru

Dhanabalan Murali

Annie Cohen

Julie Price

Darlynne Devenny

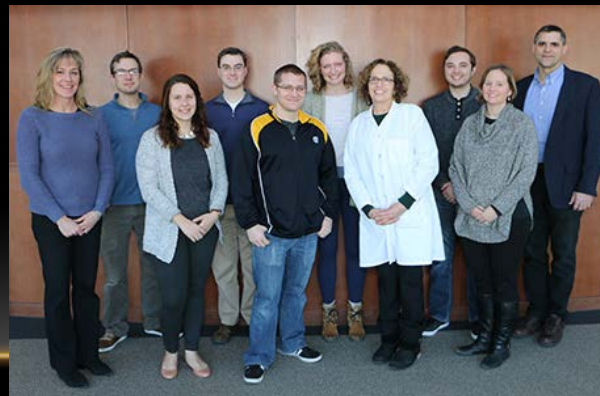
Peter Bulova



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