

Dementia in Intellectual and Developmental Disabilities: Recognition and Management

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**2019 Memory Loss
Conference: Synchronizing
Science & Support**

**Memorial Center for
Learning and Innovation**


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SIU MEDICINE
FORWARD. FOR YOU.

DISCLOSURE



- **PI, Co-I on Industry Funded Studies:**
 - **ASPIRE II, Janssen**
 - **OBSERVEMDD0001, Janssen**
 - **RECOVER, LivaNova**
 - **OASIS, Alkermes**
 - **MYCITE, Otsuka**
 - **Member of American Neuropsychiatric Association**
 - **Former Medical Director at a State Operated Intermediate Care Facility for Developmental Disabilities**
 - **Adult Relative with Developmental Disability**
- 

WHAT IS INTELLECTUAL DISABILITY?



“Intellectual disability is a disability characterized by significant limitations in both intellectual functioning and in adaptive behavior, which covers many everyday social and practical skills. This disability originates before the age of 18.”

AAIDD, 2019

<http://wilderdom.com/intelligence/IQWhatScoresMean.html>



WHAT IS DEMENTIA?



- Acquired, after 18-22 years
- Greater than 50 causes
 - Alzheimer's Disease
 - Vascular Disease
 - "Mixed"
 - Frontotemporal
 - Lewy Body Disease
 - And more than 50 others...
- Static vs Progressive
- Declining Multiple domains of Cognition and Functionality



PEDIATRIC NEURODEGENERATIVE DISORDERS WHICH CAN PRESENT AS DEMENTIA IN ADULTS

- Neuronal intranuclear inclusion disease
- Alexander's Disease
- Lafora's Disease
- Kuf's Disease
- Cerebrotendinous xanthomatosis
- PLO-SL
 - Polycystic Lipomembranous Osteodysplasia with Sclerosing Leukoencephalopathy
- Adrenoleukodystrophy
- Gangliosidosis 1-type III
- Gangliosidosis 2
- Gaucher's type 1
- Niemann-Pick II-C
- Mucopolysaccharidosis III-B
- Mitochondrial disorders (MERRF, MELAS)
- Metachromatic Leukodystrophy
- Wilson's Disease
- Fabry's Disease
- Krabbe's Disease

“REVERSIBLE (TREATABLE) DEMENTIAS”



- B-12 Deficiency
 - Folate Deficiency
 - Hypothyroidism
-
- (“Not Very Reversible Dementias”)



EDUCATIONAL OBJECTIVES



- Describe trends in lifespan and dementia risk in people with intellectual disabilities
- Depict the barriers to proper recognition and treatment
- List several of the tools which can be used for detection





Agenda	Time
Introduction: Life Expectancy in People with Intellectual and Developmental Disabilities and the Emerging Geriatric Landscape	10
Cost of Dementia	5
Recognition, Evaluation, and Diagnosis	10
Caregiver Burden and Workforce Needs	10
Evolving Evidence-Based Treatments	5
Possible Solutions and Pathways	5
Discussion	



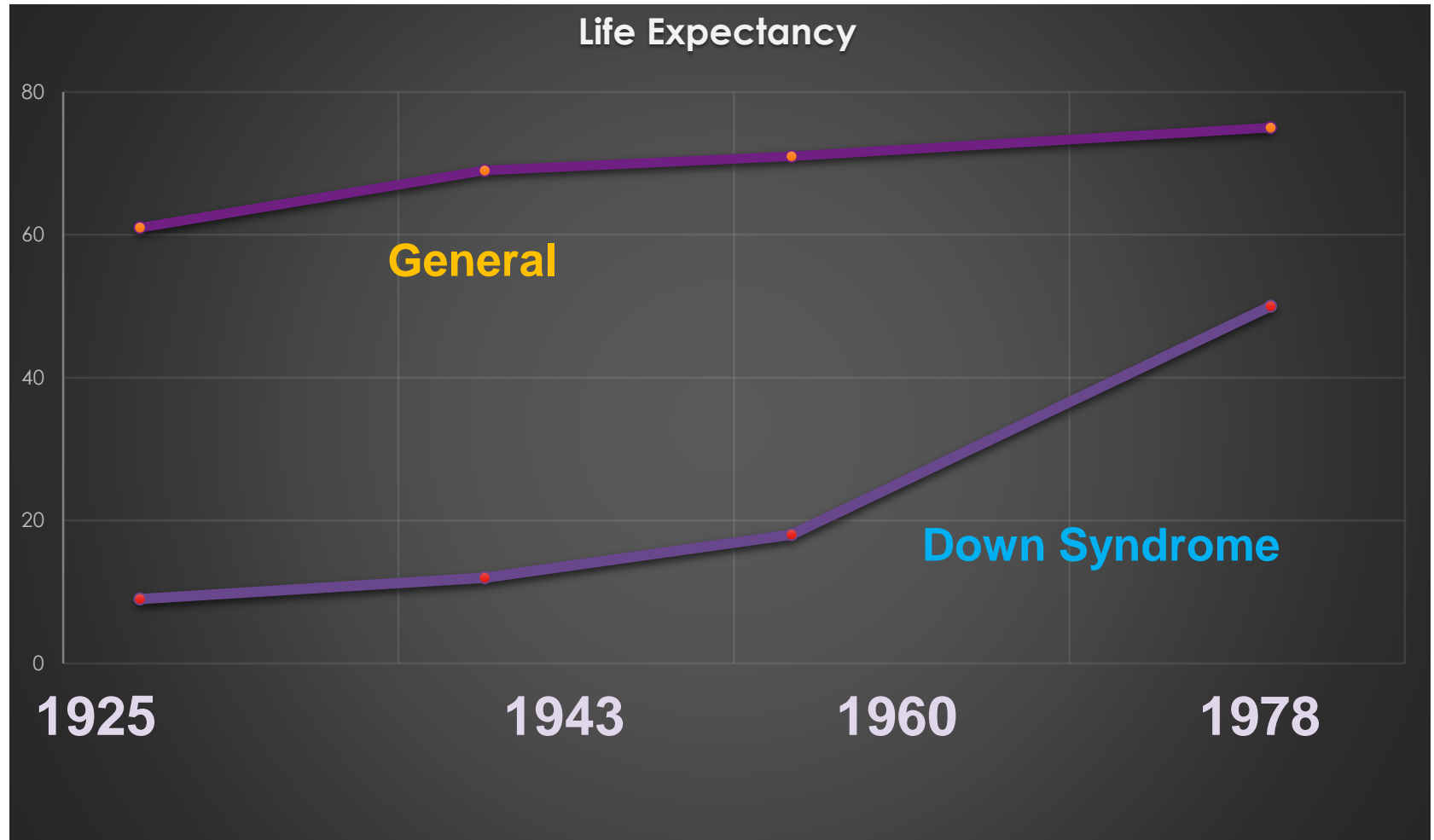


“Uncle Phil”

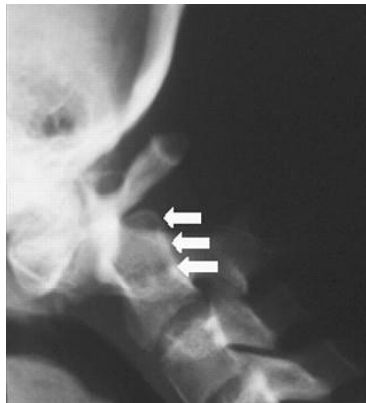
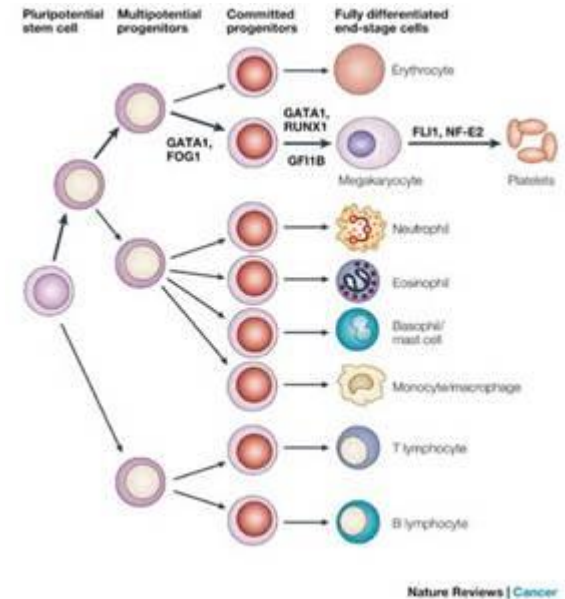
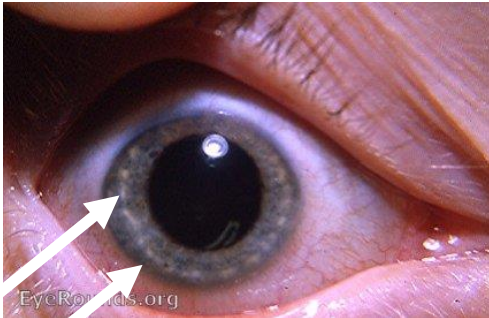


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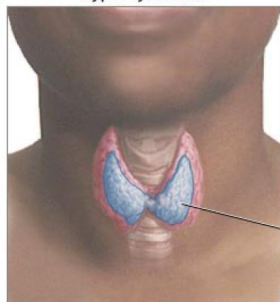
Introduction: Life Expectancy and Geriatric Medicine



Features of Down Syndrome

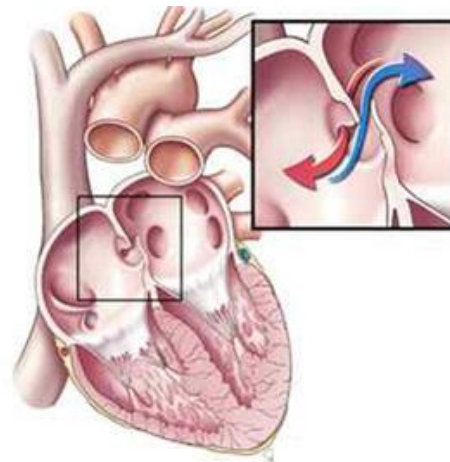


Hypothyroidism



In hypothyroidism, levels of thyroid hormone are low. The thyroid gland can be small or large (goiter), depending on the cause of the disorder

Atrophied thyroid



“DIVERSITY OF SYNDROME-SPECIFIC AGING ISSUES”



- **Example: Down’s Syndrome Accelerated Aging**
 - cataracts
 - hearing loss
 - hypothyroidism
 - osteoporosis
 - epilepsy
 - sleep apnea
 - genetically elevated risk of developing Alzheimer disease



OTHER SYNDROME RELATED AGING



- **Cerebral Palsy**
 - loss of mobility
 - Osteoporosis
 - chronic fatigue
 - chronic pain
- **Williams Syndrome**
- **Prader-Willi Syndrome**
- **Fragile X Syndrome**
- **Other rare syndromes ??????**



GENERAL POPULATION BASED PROJECTIONS

figure 4:

Projected Numbers of People Age 65 and Over in the U.S. Population with Alzheimer's Disease Using the U.S. Census Bureau Estimates of Population Growth*

Numbers
(in millions)

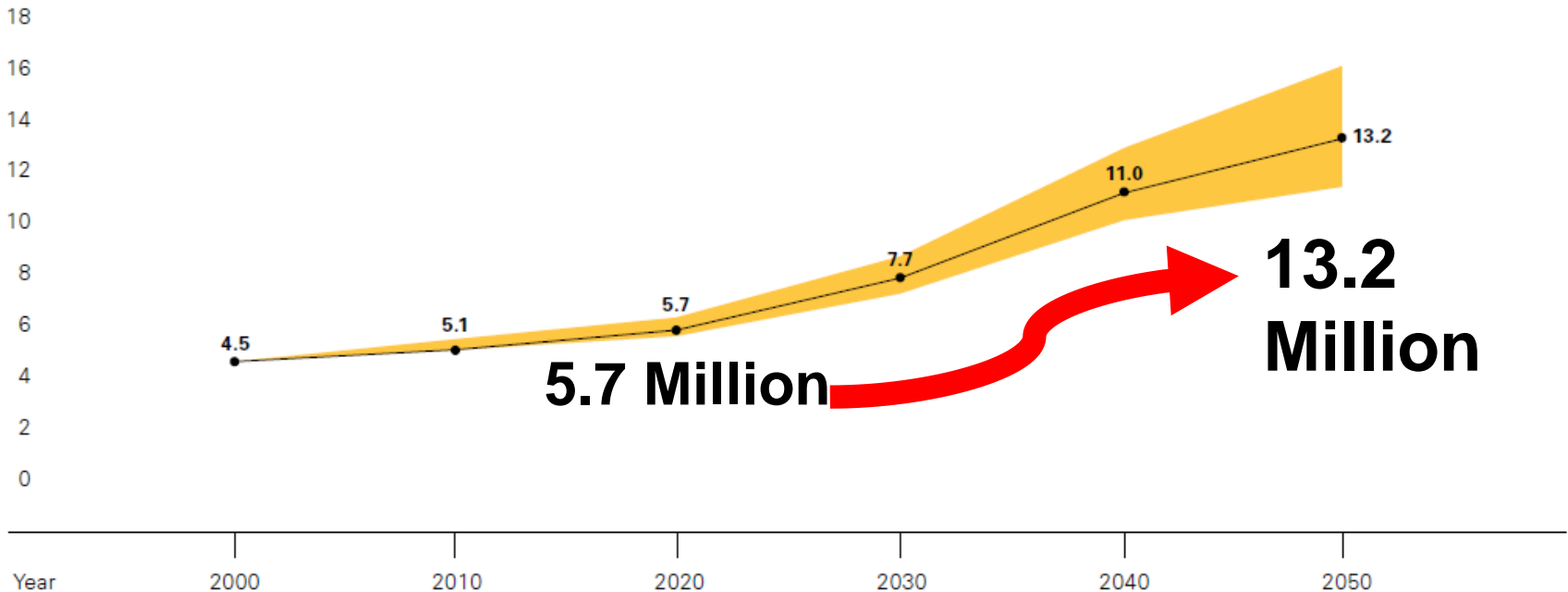
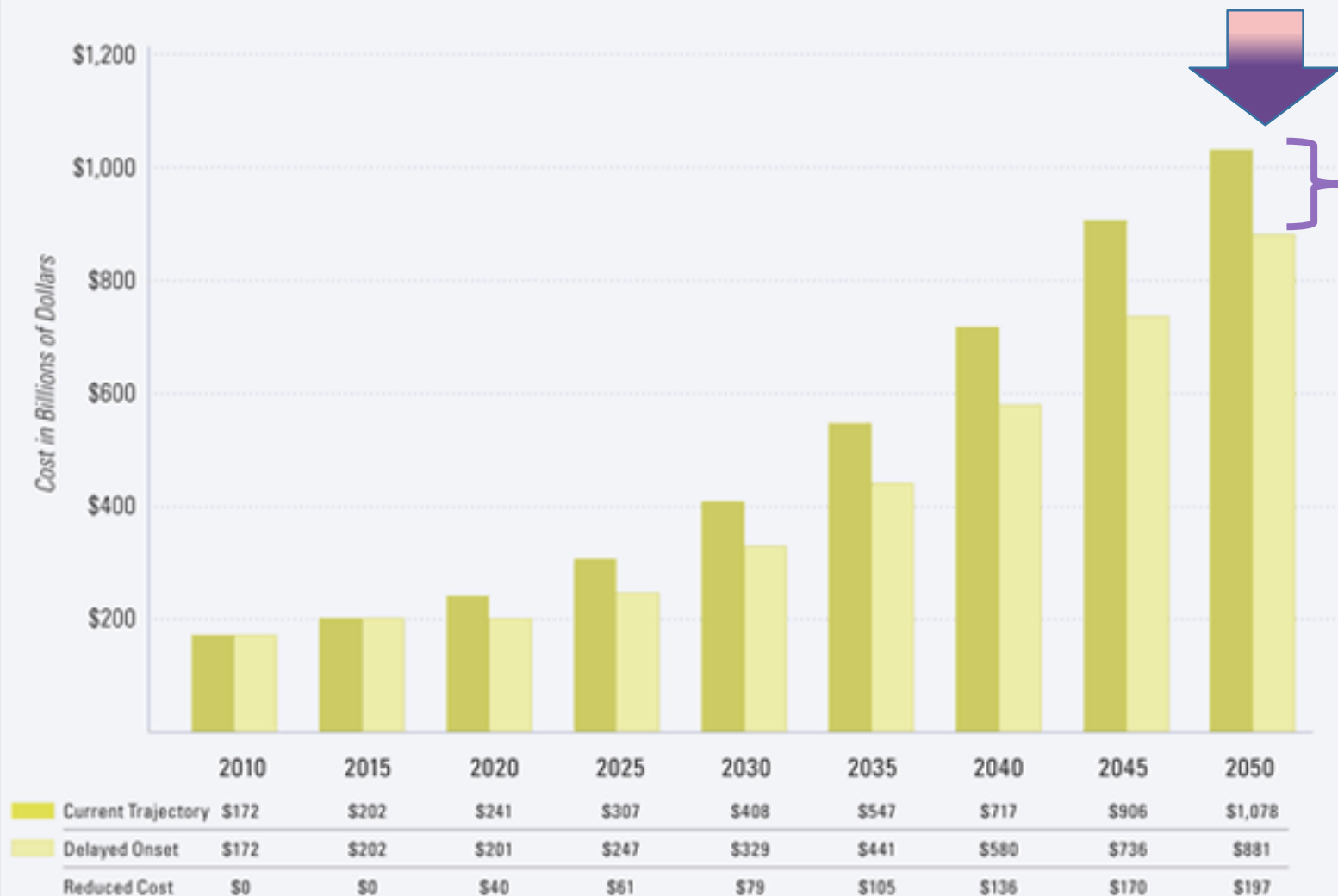


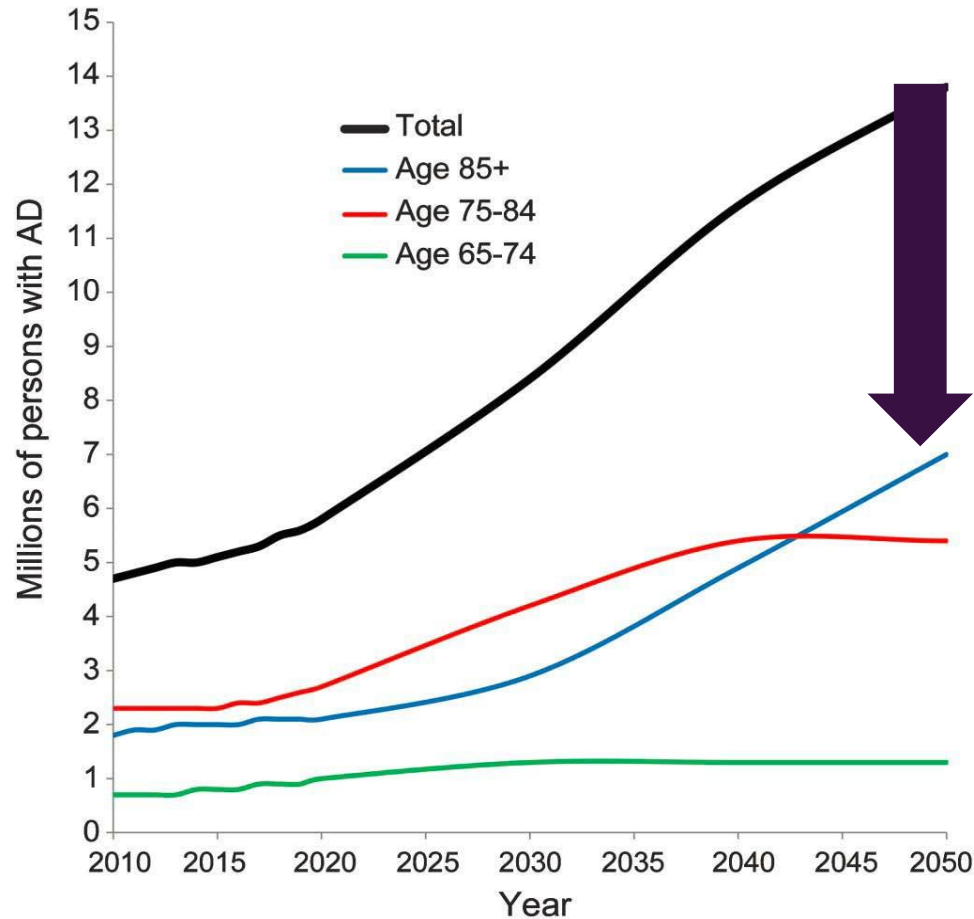
Figure 12:

Impact of a Slowed Progression on Costs, Americans Age 65 and Older with Alzheimer's Disease, 2010–2050



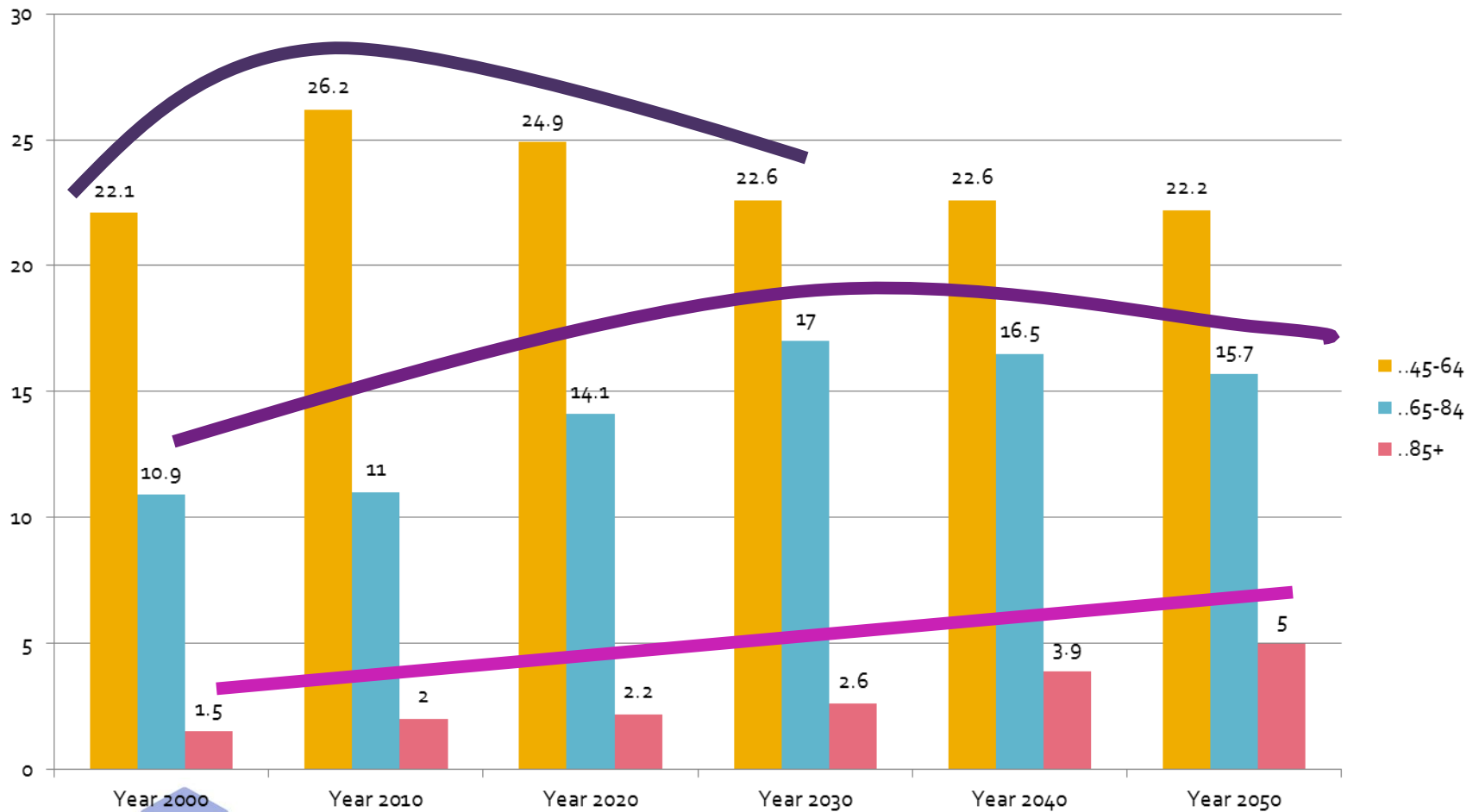
ALZHEIMER'S DISEASE PREVALENCE

Estimated number of people with Alzheimer's disease (AD) in the United States in 2010 and projections through 2050



Hebert, L. E., Weuve, J., Scherr, P. A., & Evans, D. A. (2013). Alzheimer disease in the United States (2010–2050) estimated using the 2010 Census. *Neurology*, 80(19), 1778–1783.

PROJECTED AGE PROGRESSION IN INTELLECTUAL DISABILITIES POPULATION

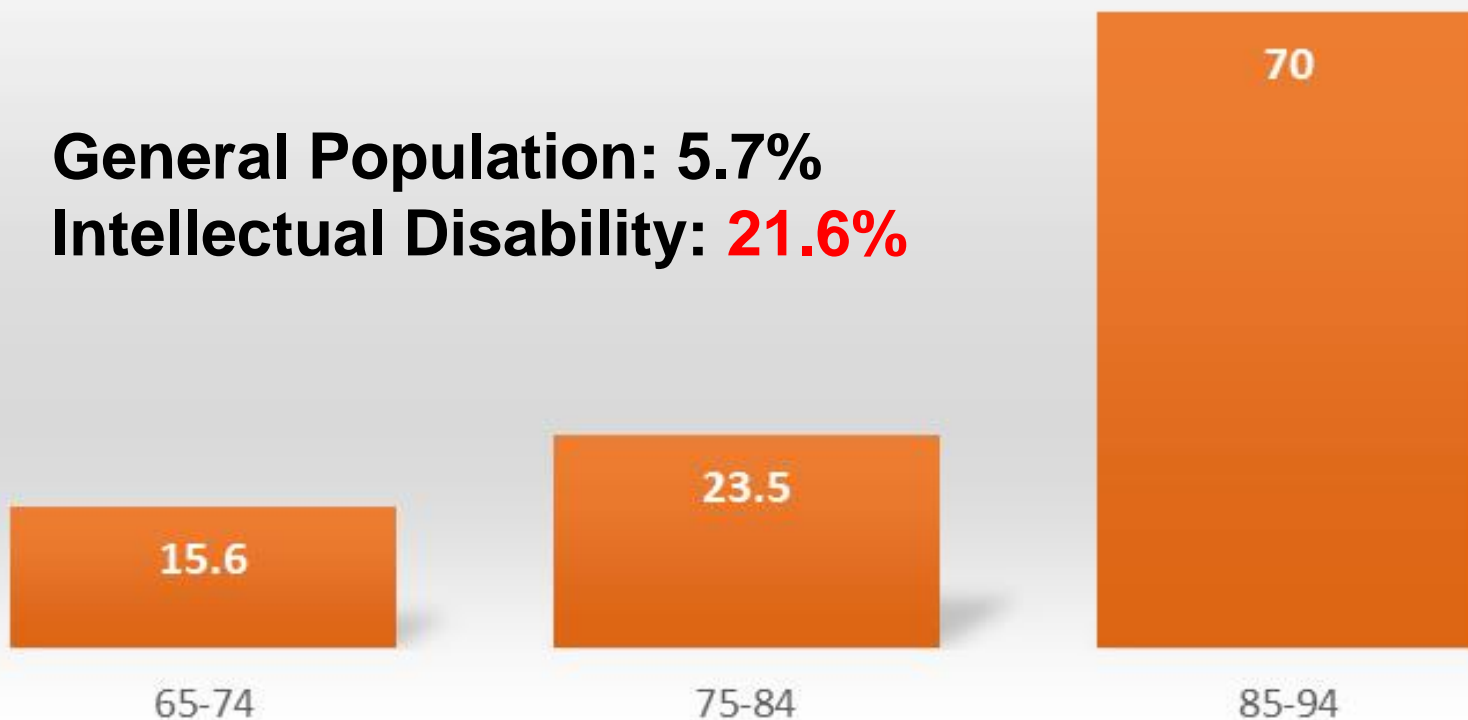


U.S. Census Bureau 2004

DEMENTIA IN PEOPLE WITH INTELLECTUAL DISABILITIES

Percent with Dementia

General Population: 5.7%
Intellectual Disability: 21.6%





- Greater number of people with intellectual disability:
- Living in the community
- Needing psychiatric and behavioral services
- Living into the older age range



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Causes of Dementia and Co- morbidity

THE IMPACT OF ALZHEIMER'S AND RELATED DEMENTIAS

Patient

More than 5 million affected
Increasing with aging
population
70% live at home

More than 70% of patients
with Alzheimer's disease
live at home, and almost
75% of home care is
provided by family and
friends

Family

Loss of productivity of
caregivers
Painful process
Significant incidence of
depression and medical illness

46% to 59% of caregivers
reported depression,
according to various
studies

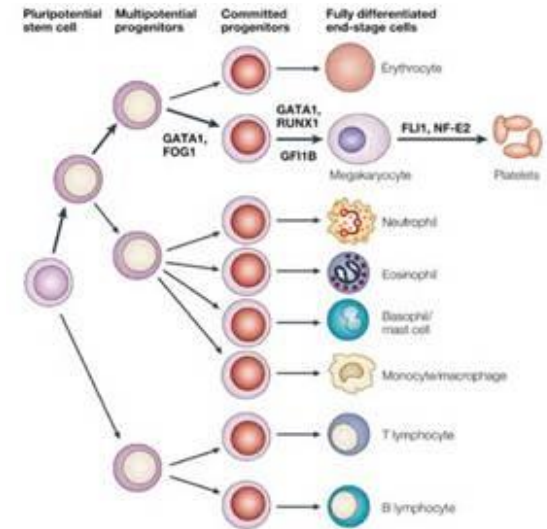
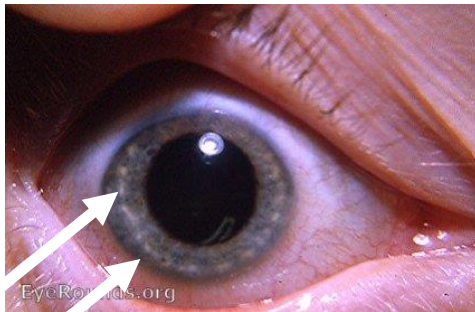
Society

\$100 Billion / year

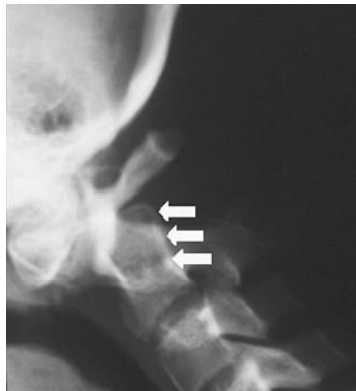
Down Syndrome - Faces from the Internet



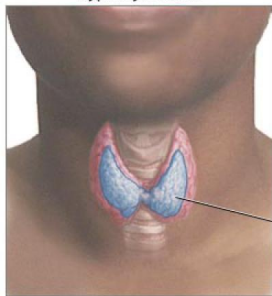
Features of Down Syndrome



Nature Reviews | Cancer

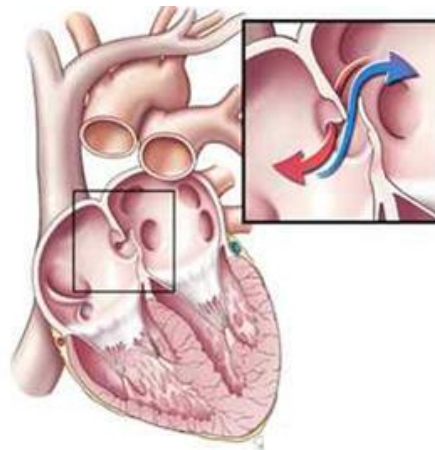


Hypothyroidism

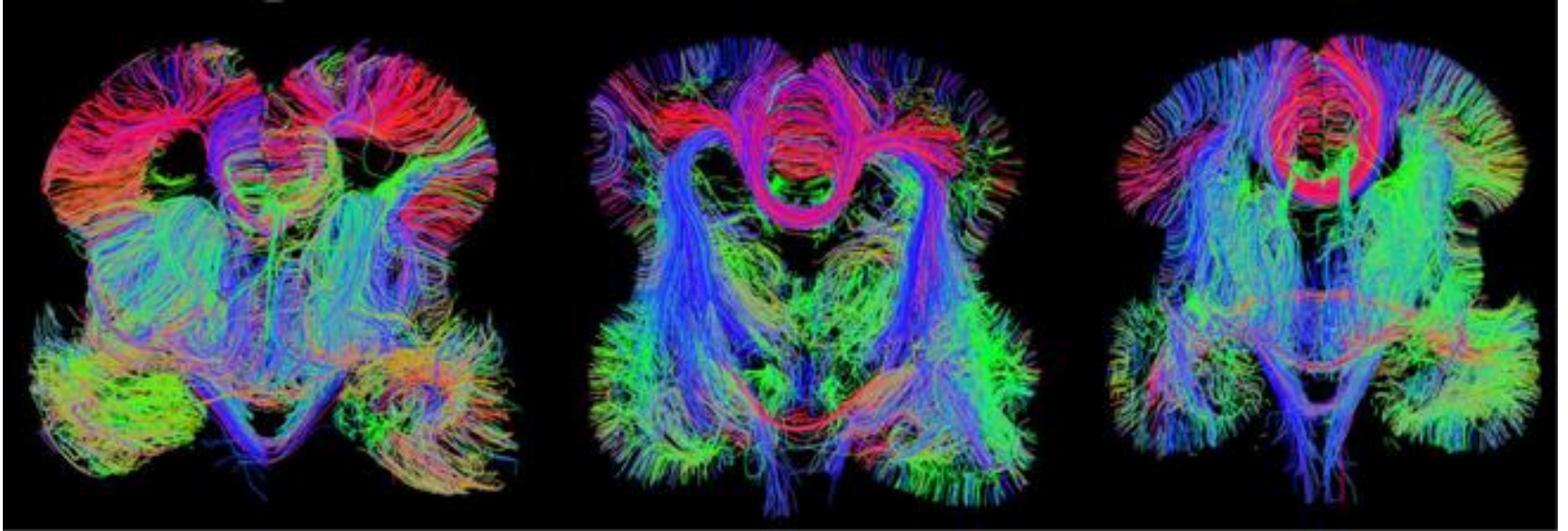


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Atrophied thyroid



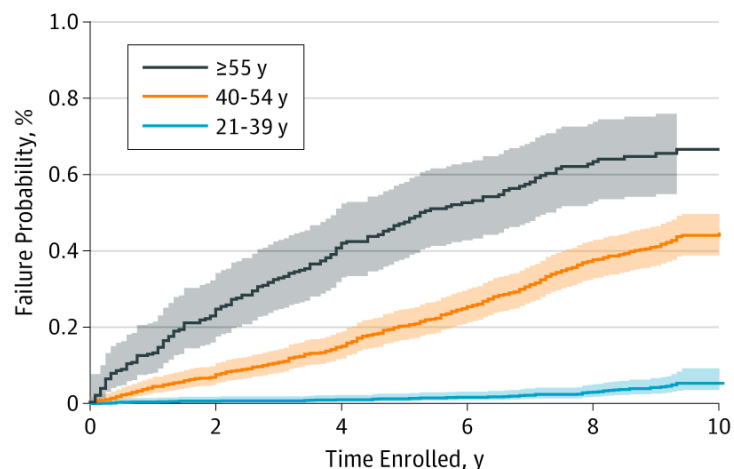
Why Down Syndrome?



From: **Epidemiology of Dementia and Alzheimer Disease in Individuals With Down Syndrome**

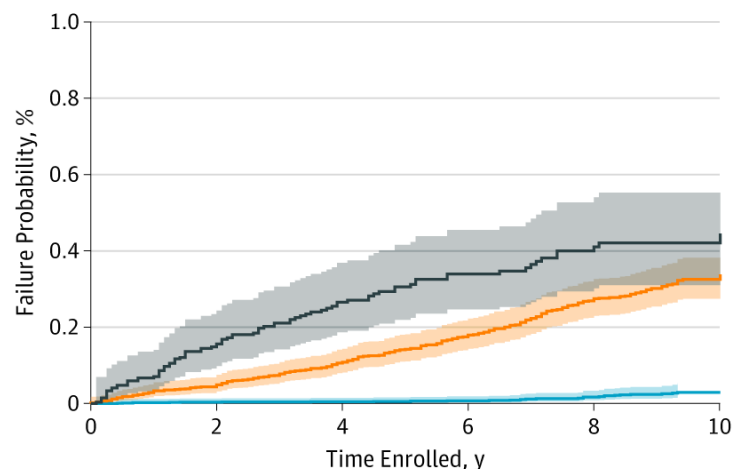
JAMA Neurol. Published online October 28, 2019. doi:10.1001/jamaneurol.2019.3666

A Any dementia



No. at risk											
21-39 y	1540	1534	1530	1456	1373	1274	1174	1042	910	660	224
40-54 y	1072	1029	999	931	854	761	691	615	523	393	153
≥55 y	279	243	215	176	139	109	91	76	57	46	24

B Alzheimer disease

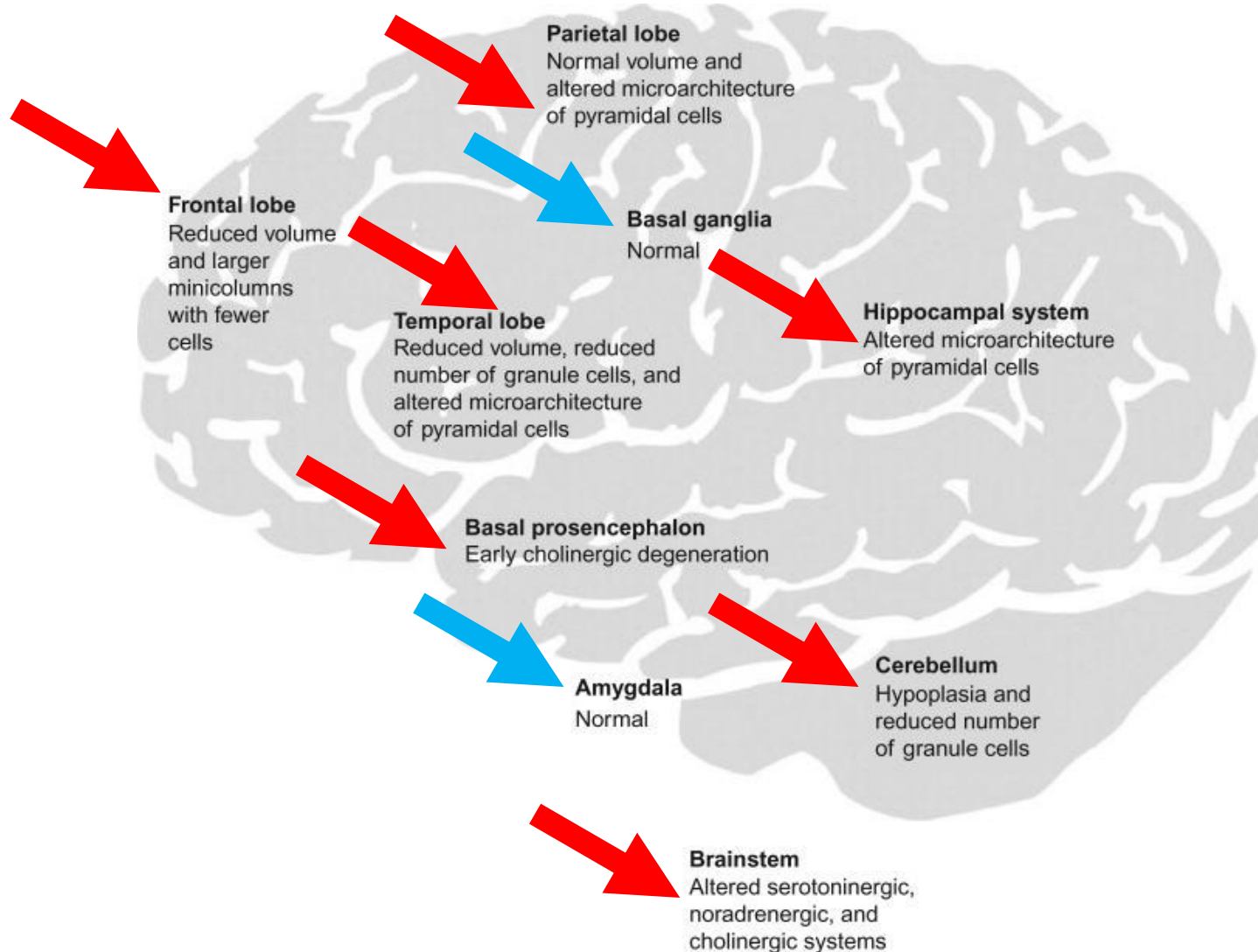


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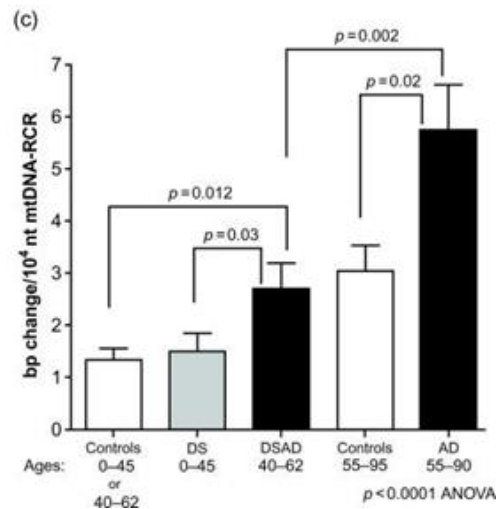
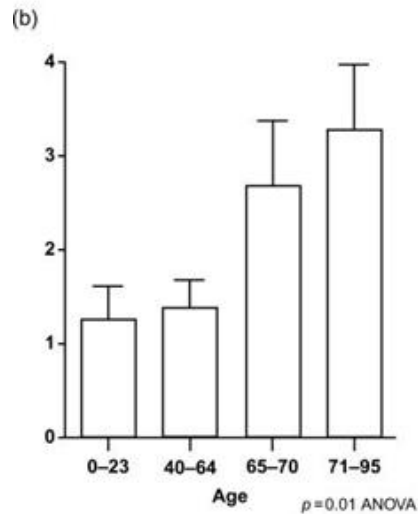
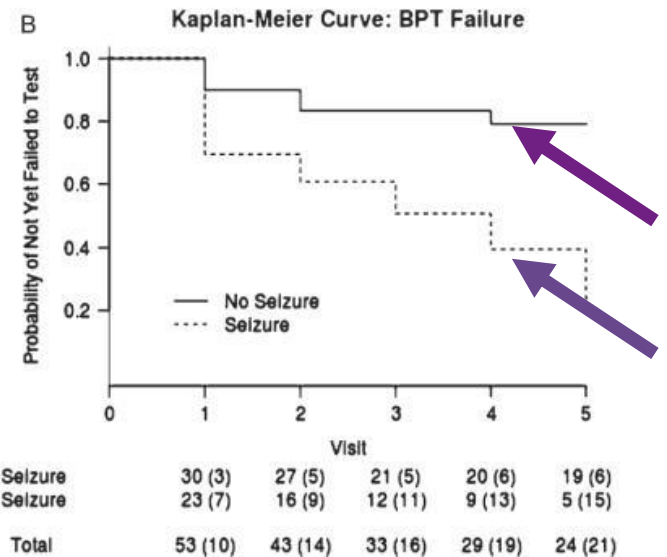
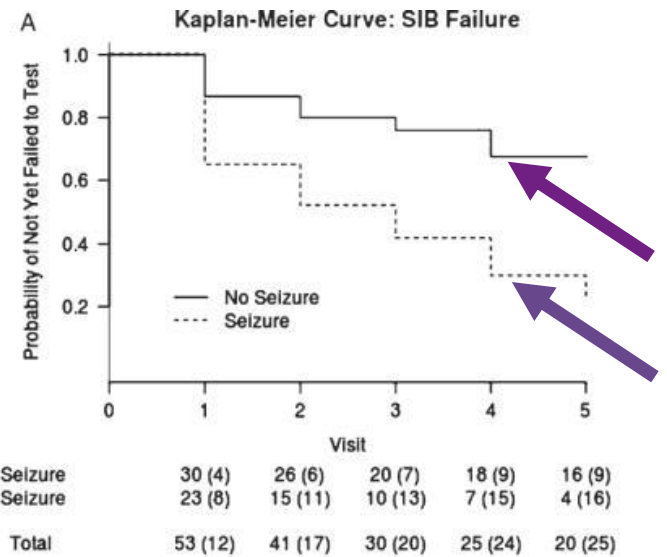
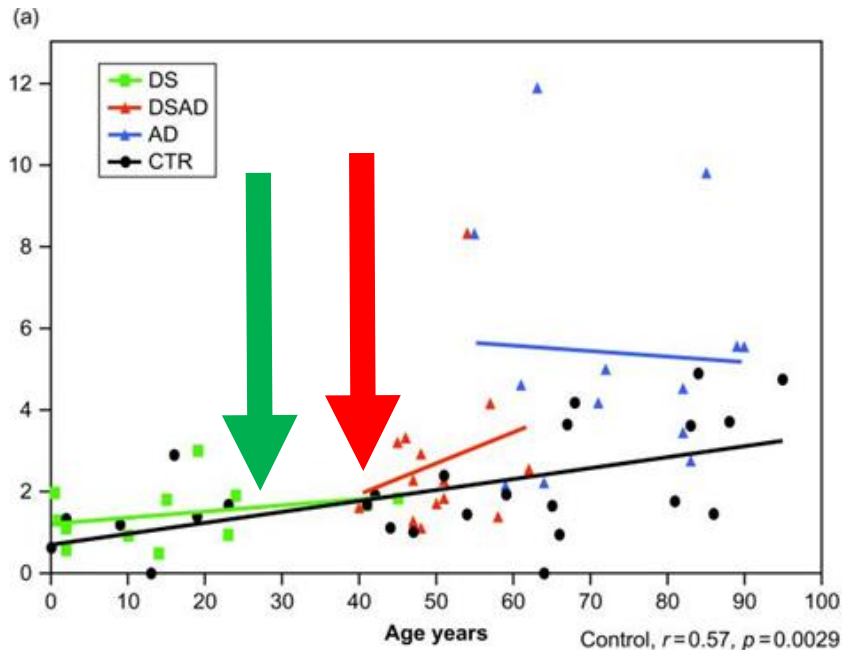
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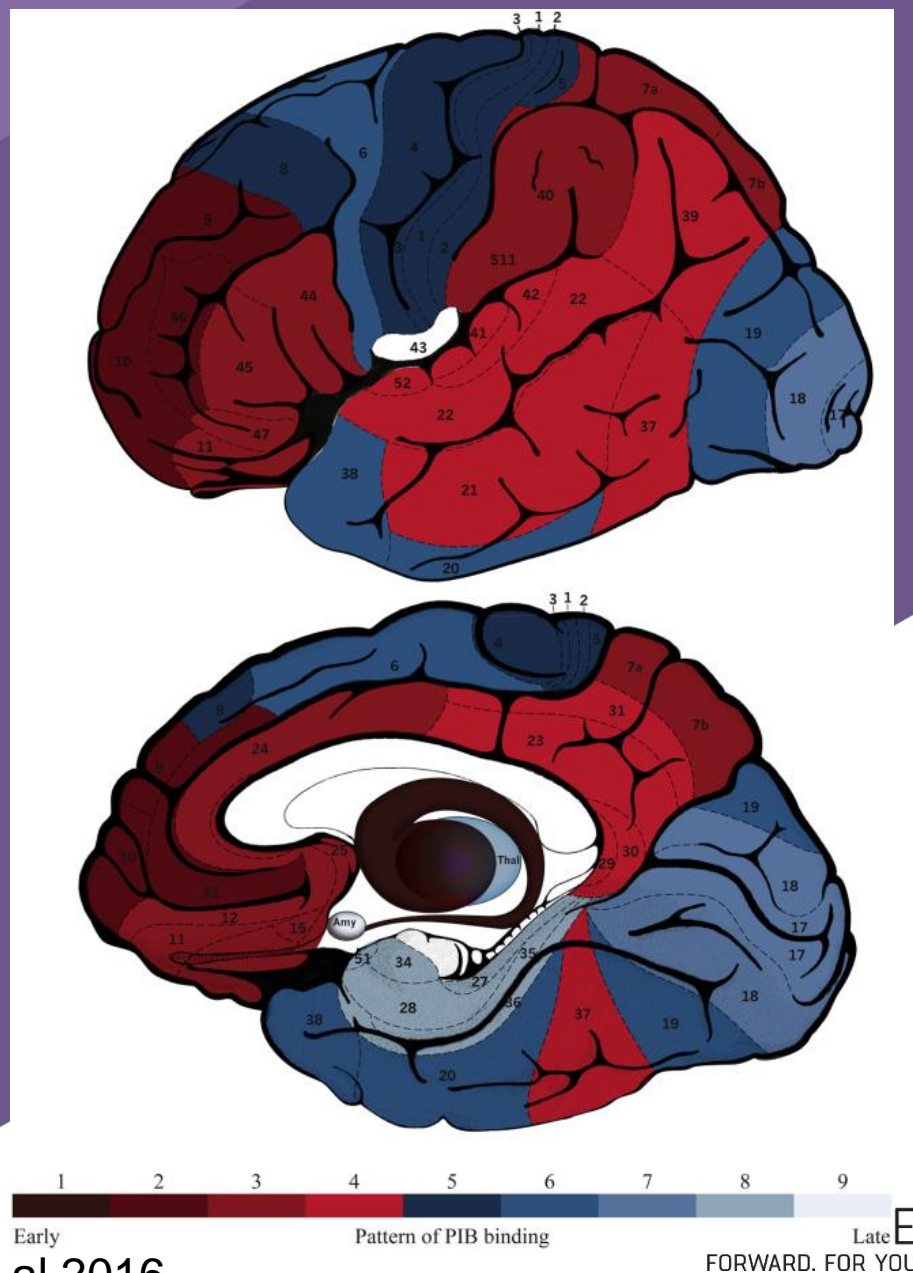
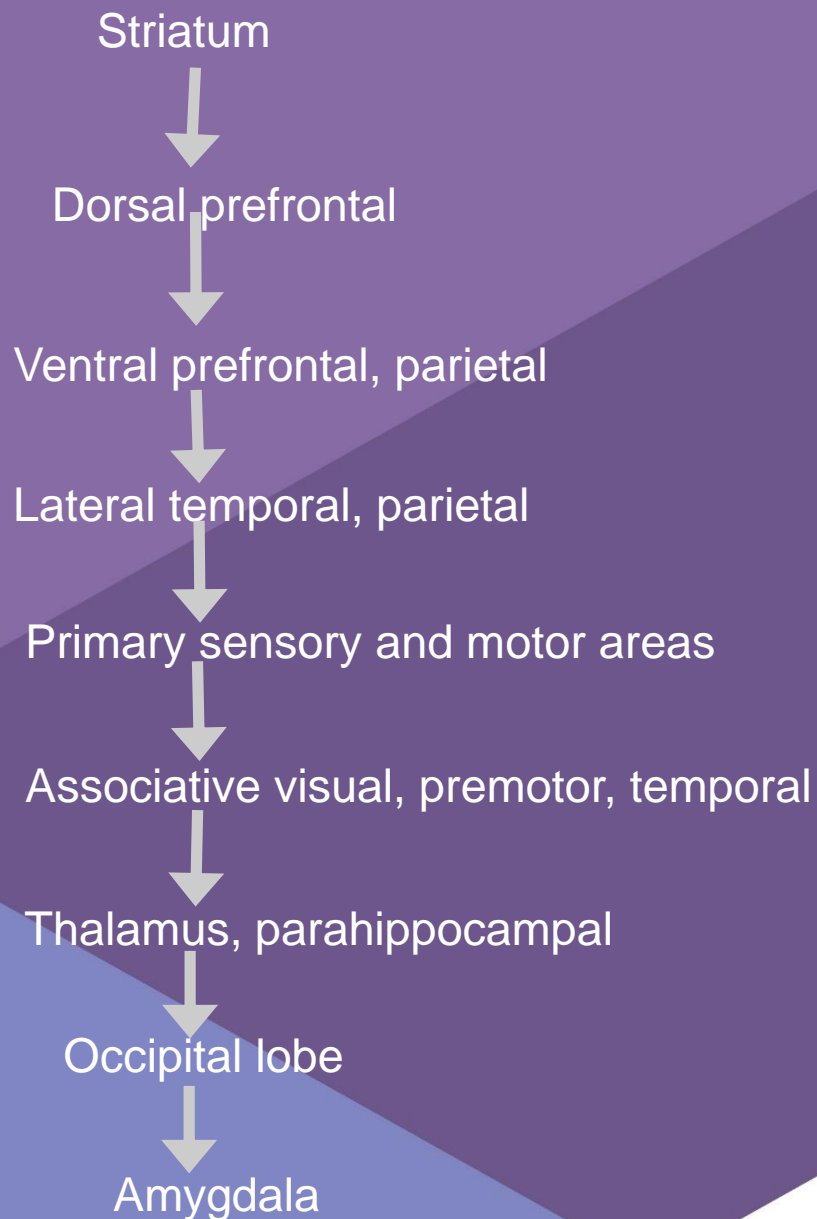
Cumulative Incidence Functions for Incident Dementia Claim Among Medicaid Enrollees With Down Syndrome in Wisconsin, 2008 Through 2018 Members enrolled in Medicaid for at least 3 years were included; a washout period of 1 year without a dementia claim was included.

The Brain in Down Syndrome



Age and Dementia





Annus et al 2016

DOWN SYNDROME

In people with Down syndrome, changes in overall function, personality, and behavior may be more common early signs of Alzheimer's than memory loss and forgetfulness.

- Reduced interest in being sociable, conversing, or expressing thoughts
- Decreased enthusiasm for usual activities
- Decline in ability to pay attention
- Sadness, fearfulness, or anxiety
- Irritability, uncooperativeness or aggression
- Restlessness or sleep disturbances
- Seizures that begin in adulthood
- Changes in coordination and walking
- Increased noisiness or excitability
- Researchers don't yet know why early Alzheimer's symptom patterns may tend to differ among those with and without Down syndrome

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Recognition of Dementia in People with Intellectual Disabilities

DIAGNOSING DEMENTIA

- **Detailed patient history**
- **How and when symptoms appear**
- **Family history**
- **Assessment of emotional status and living environment**
- **Physical and Neurological exam and laboratory tests**
- **Neuropsychological testing**
- **Memory, language skills assessment**
- **Mathematic, drawing and other brain function tests**
- **X-ray, CT, or MRI to rule out other diagnosis**





BARRIERS TO ROUTINE APPROACH IN DIAGNOSING DEMENTIA

- **Misidentification by the family and caregivers of early signs of Alzheimer's disease (AD) as normal aging process**
- **Social skills often maintained in early AD**
- **Denial and lack of insight by patient**
- **Lack of routine or definitive screening tools**
- **Diagnostic “Overshadowing”**

DOWN SYNDROME

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BARRIERS TO DIAGNOSIS

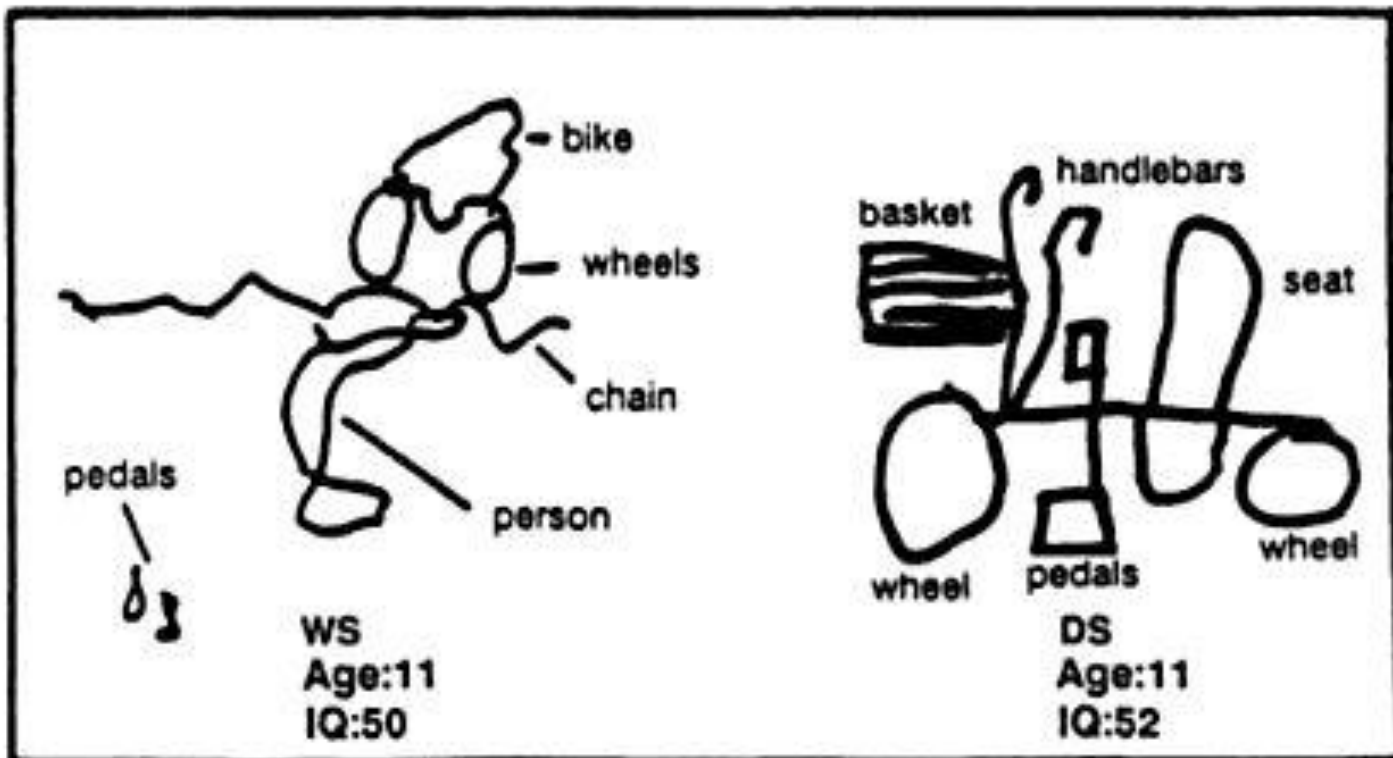
- **In addition to the barriers to the routine approach:**
 - **Detailed patient history**
 - May not be present; may be subjective
 - **How and when symptoms appear**
 - Historical perspective not apparent because of staff turnover
 - **Family history**
 - Many families aren't closely involved
 - **Assessment of emotional status and living environment**
 - Systems issues

BARRIERS TO DIAGNOSIS IN PEOPLE WITH IDD

- **Physical exam and laboratory tests**
 - Lack of understanding of best practices in working with populations; diagnostic overshadowing
- **Neuropsychological testing**
 - Success rate differs
- **Memory, language skills assessment**
 - Often missing; differing opinions
- **Mathematic, drawing and other brain function tests**
 - Aimed at neuro-typical
- **X-ray, CT, or MRI to rule out other diagnosis**
 - Compliance is hard

Drawing of a Bicycle

Age and IQ Matched WS and DS Subjects



TREATMENT



- What is the MMSE in IDD?
- Nootropic agents
 - Donepezil
 - Rivastigmine
 - Memantine
 - Galantamine
- Other syndrome specific agents?
- Early Intervention
- Healthy lifestyle
- Role of Behavioral Treatment, Psychotropics



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Caregiver Burden and Workforce Needs

Workforce – Direct Service Professionals

- The national average annual turnover rate of DSPs is **45%**
- The field needs **574,200 new DSPs** every year
- National cost of replacing DSPs estimated to be:
- **\$2,338,716,600 (2.4 Billion)** in 2015
- **9% of all** available DSP positions go unfilled
- Estimated time required to care for clients with ID and dementia is **3-4 times** that for ID alone

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Possible Solutions and Pathways

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).

**Yearly: 40 years on, people with Down
Syndrome, 50 years on for other people
15-60 minutes to complete**

EVALUATION TOOLS



- National Task Group Early Detection Screen for Dementia (NTG-EDSD)
 - Down – yearly at 40 and above
 - Idiopathic IDD – yearly at 50 and above
 - All - whenever suspicion of cognitive change
- Manual and tool available at:
- www.aadmd.org/ntg/screening



RESOURCES

- National Task Group on Intellectual Disabilities and Dementia Practices -- <https://aadmd.org/NTG>
- National Alzheimer's and Dementia Resource Center -- <https://nadrc.acl.gov/>
- National Down Syndrome Society -- www.ndss.org
- National Down Syndrome Congress -- www.ndsccenter.org
- Centers for Disease Control and Prevention. Facts about Down syndrome. Updated March 3, 2016. www.cdc.gov/ncbddd/birthdefects/downsyndrome.html

1. improving professional identity and recognition;
2. teaching business and organizational leaders skills to improve their ability to recruit, select and retain employees;
3. the use of self-directed services that permit individuals and families to recruit, select and retain their own DSPs;
4. using worker cooperative and independent provider models;
5. using competency-based training models that lead to credentialing or certification of staff and yield wage increases; and
6. using technology-enhanced supports.

TRAINING



- **Recognizing signs of decline**
- **Palliative care**
- **Behavioral interventions**
- **Recognizing pain**
- **Mealtimes support**
- **Challenging behavior**
- **Coping strategies (for staff)**
- **Collaborative Teamwork**



HEALTH ADVOCACY



- **The role of leadership, nursing, and social work**
 - **Promote health literacy among families and staff**
 - **Train caregivers on the signs of decline**
 - **Provide guidelines on what to report and to whom regarding observed decline in functioning**
 - **Allow for and encourage for discussions with health care professionals**
 - **Encourage systematic assessment and guidance about next steps**



- **Encourage open and honest communication between health care professionals and all members of the individual's support system with one another to develop the appropriate delivery of services and allow for future planning**



HEALTH ADVOCACY-PERSON CENTERED

- **Exercise**
- **Proper Diet**
- **Social activity**
- **Music therapy**
- **Pet therapy**
- **Spiritual therapy**
- **Structured day activity**
- **Caregiver support**



SUMMARY

- Alzheimer's and other Dementias are challenging to recognize and diagnose
- Dementia is increasing in prevalence and represents a major challenge to the IDD field in terms of:
 - Cost
 - Workforce
 - Diagnosis
 - Treatment
- Possible solutions
 - Adoption of Routine Screening
 - Realignment of workforce initiatives
 - Collaborative care
 - Research

Thanks!

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**Next Steps,
Questions and Answers**