Evaluation of Cognitive Decline in a Person with Intellectual and Developmental Disabilities

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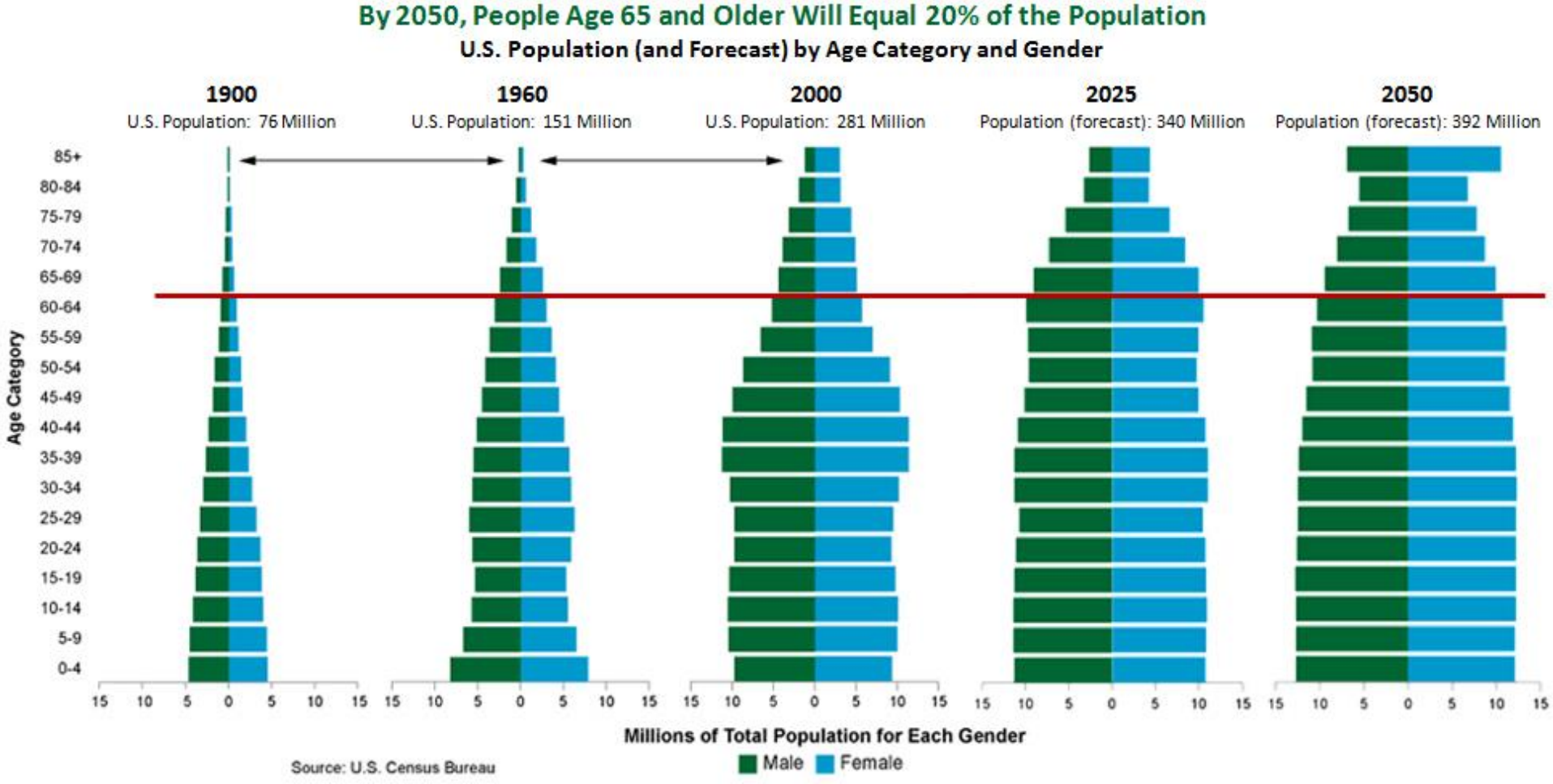
ntg
National Task Group on Intellectual Disabilities and Dementia Practices

UL SCHOOL OF MEDICINE

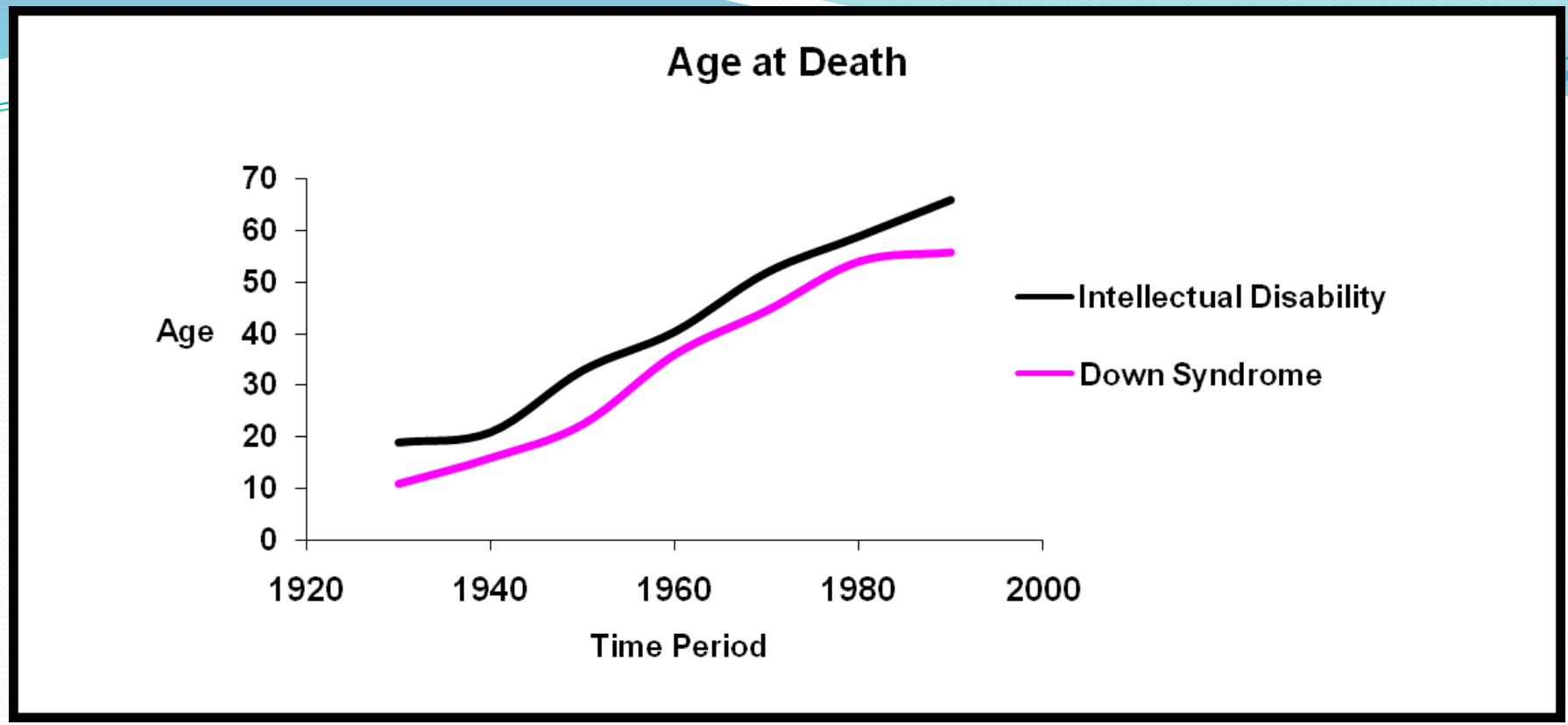
American Academy of Developmental Medicine and Dentistry

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



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

- Currently estimated life expectancy of a 1-year-old child with DS is between 43 and 55 years

Curr Gerontol Geriatr Res. 2012; 2012: 412-536.

- 25% of persons with Down syndrome are still alive at 65 years

Rubin & Crocker,2006; Yang Rasmussen & Friedman, 2002

Expected Physical Changes of Aging

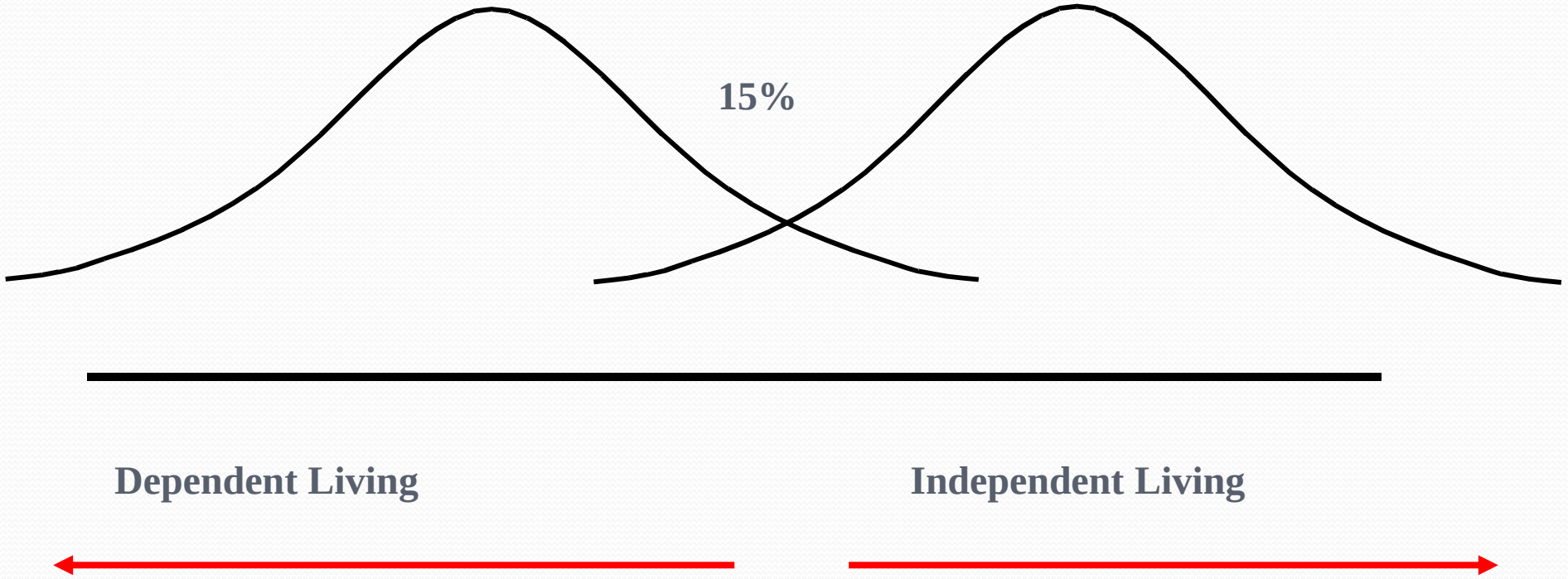
- **Osteopenia/Osteoporosis** - normal aging-related bone loss
- **Sarcopenia** - progressive loss of muscle mass
- **Presbyopia:** the lens of the eye becomes stiffer and less flexible – affecting the ability to focus on close objects (accommodation)
- **Presbycusis** – aging related change in the ability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

Does aging always bring decline in function?

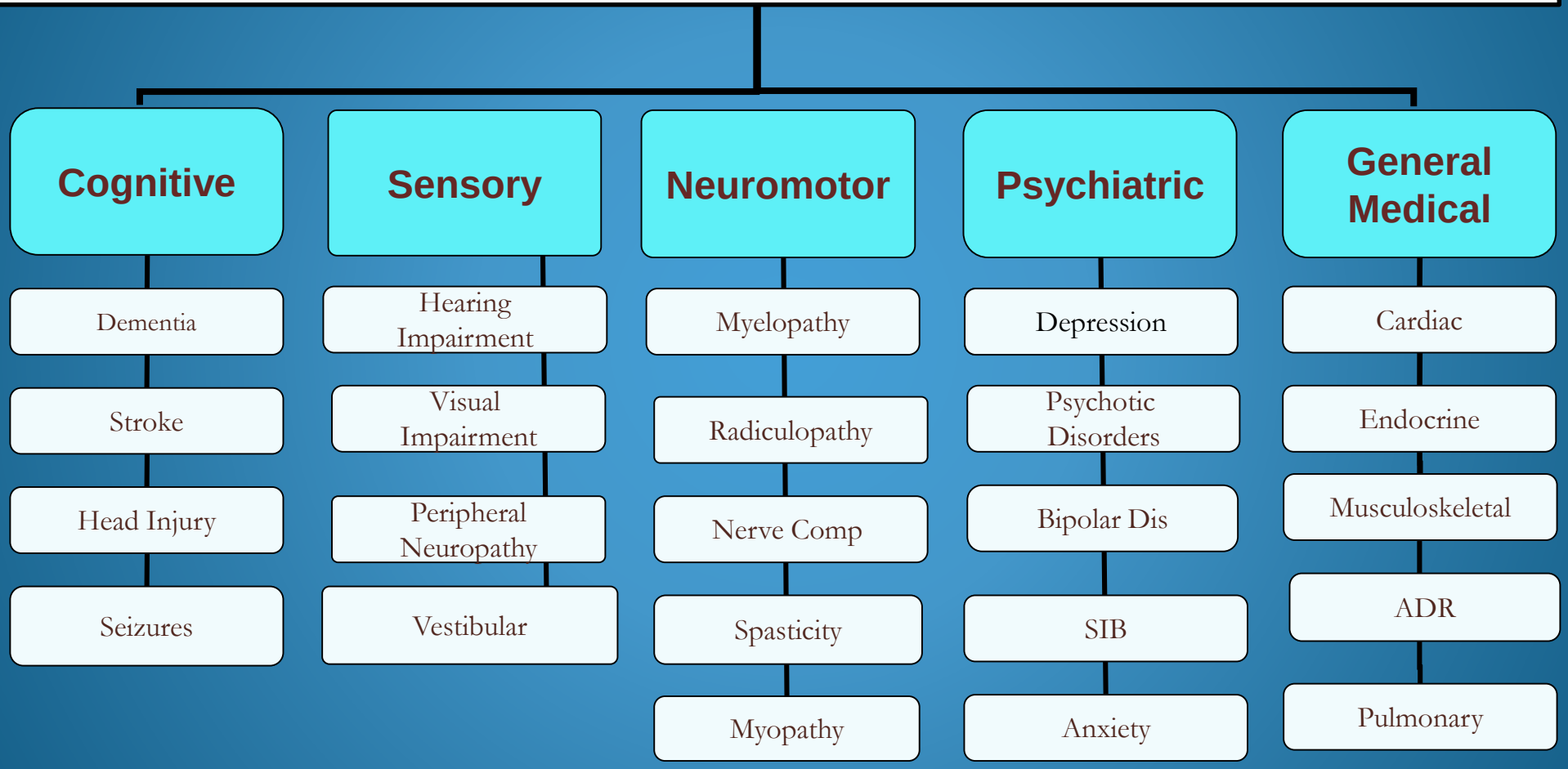
What change is Normal or Not??

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

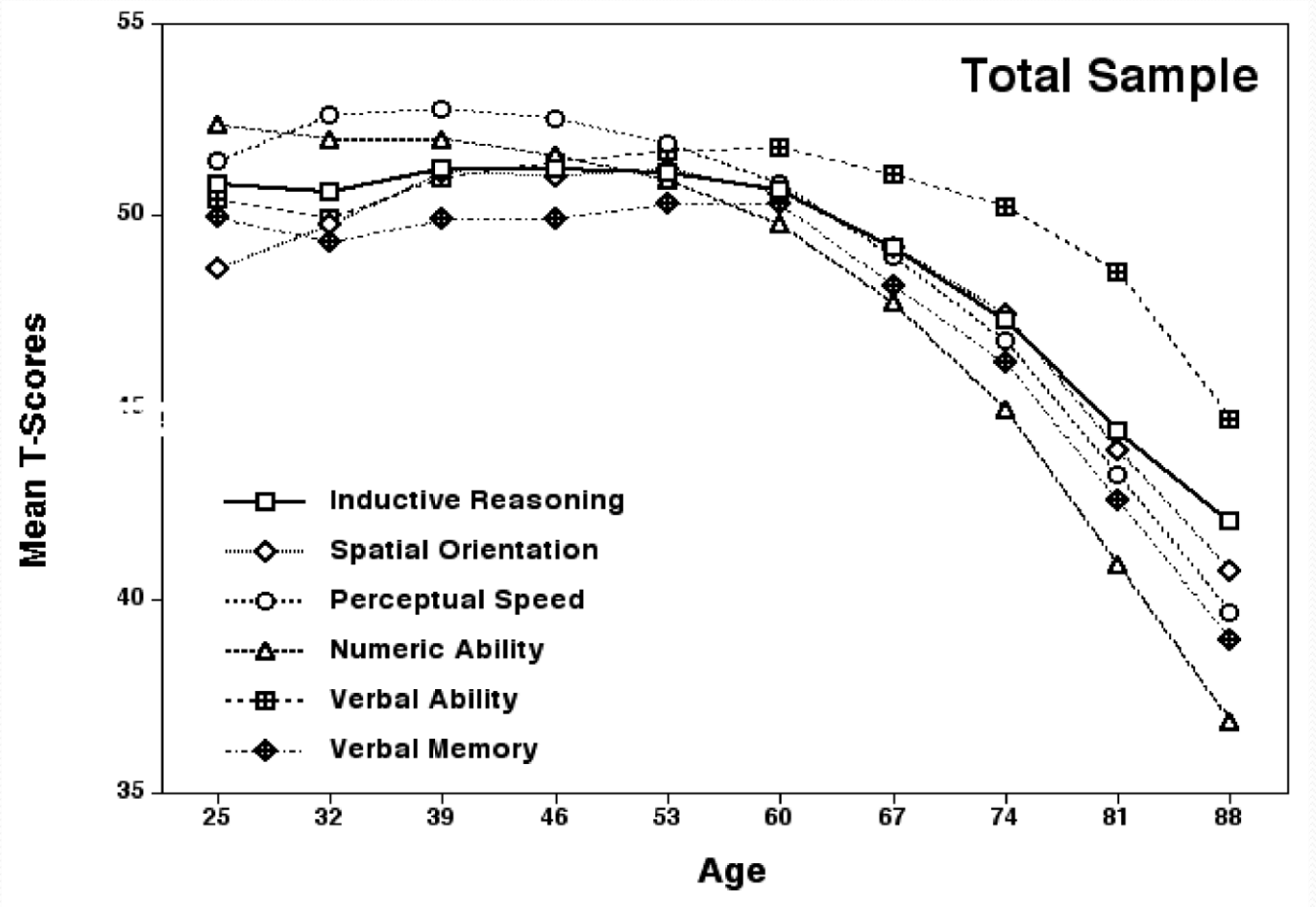
Cerebral Palsy

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

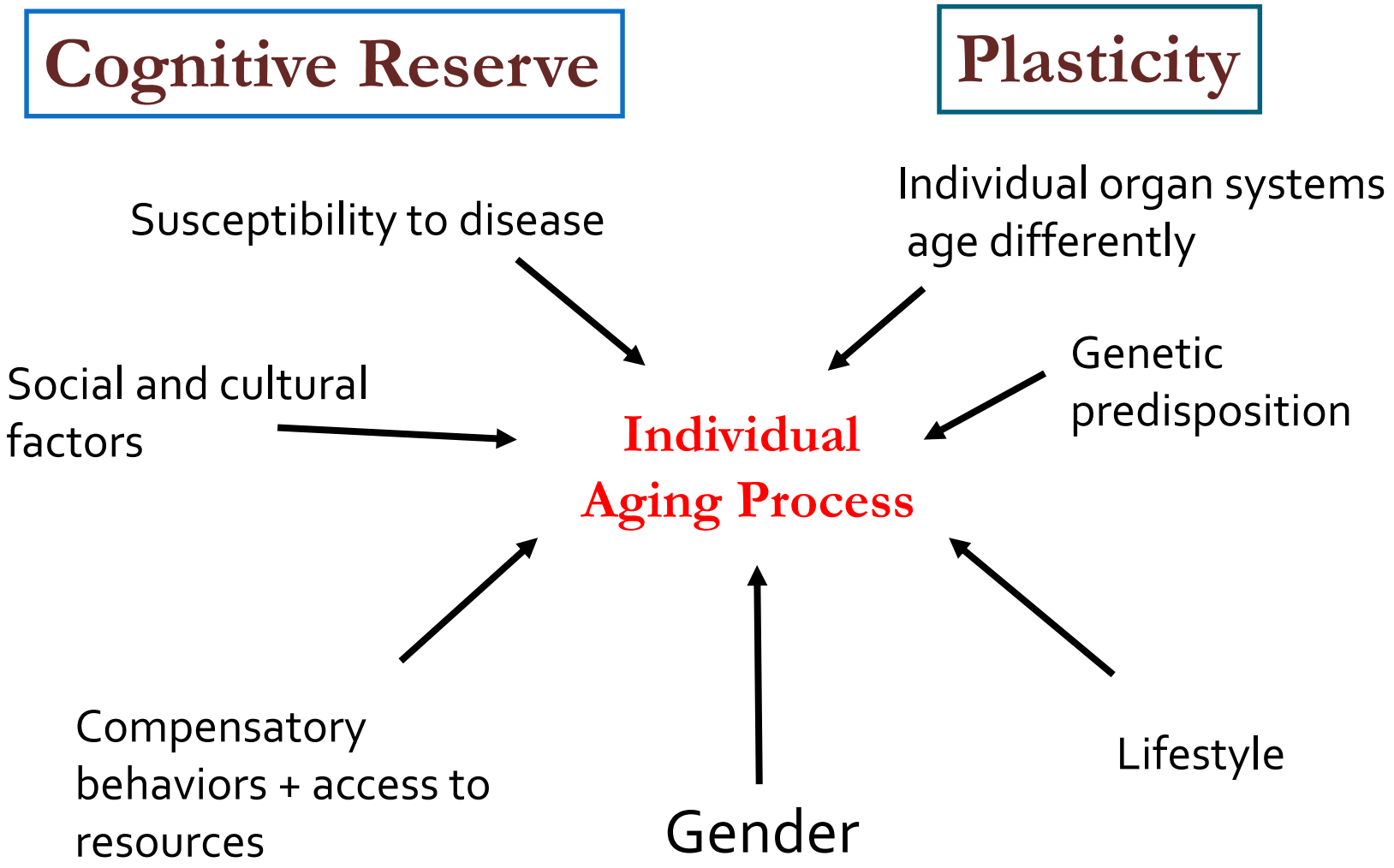
Autism

- 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

Longitudinal Age Changes



Schaie, K. W. (2005). Developmental influences on adult intelligence: The Seattle Longitudinal Study. New York: Oxford University Press



Diversity of the Aging Process

Does having a IDD predispose to getting dementia?

- IDD at disadvantage due to developmental immaturity of brain architecture
- Down syndrome and early onset Alzheimer's disease
- Social restrictions
- Less access to health care
- Less health promotion activities
- Usage of polypharmacy
- History of mental health comorbidities
- Usage of neuroleptics
- History of falls and head injuries
- History of SIB

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, 9-10 years from first symptoms
- Infrequently rapid decline can occur
- Sensory impairments (vision: 92.8%; hearing: 62.3%) 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.

McCarron et al., (2013). A prospective 14-year longitudinal follow-up of dementia in persons with Down syndrome. Journal of Intellectual Disability Research

Percentage of people with Down syndrome who develop dementia at different ages: Age percentage with clinical signs of dementia	
30's	2%
40's	10-15%
50's	33%
60's	50-70%

Source: Neil, M. (2007). Alzheimer's dementia: What you need to know, what you need to do. Understanding intellectual disability and health. Accessed from http://www.intellectualdisability.info/mental-health/alzheimers-dementia-what-you-need-to-know-what-you-need-to-do.

What Could It Be?

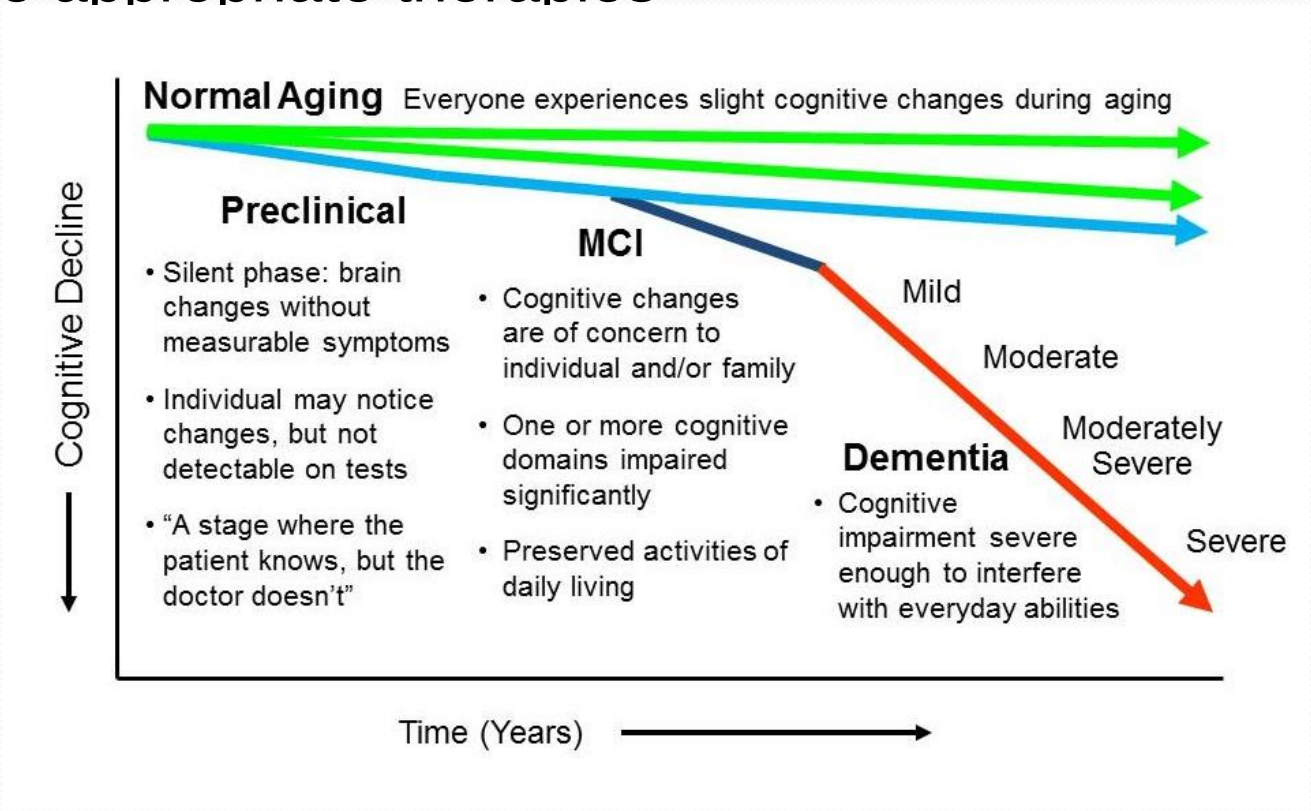
- Another medical condition
- Medication side-effect
- Hearing loss or vision loss
- Depression
- Acute illness
- Severe but unrecognized pain
- Normal aging
- Dementia

Importance of accurate diagnosis

- Many people in the early stages prefer to know
- Patients and caregivers need accurate and timely information in order to plan ahead
- Access to support and counselling
- Associated welfare benefits
- Legal advice and driving
- Availability of drug treatments
- May find reversible causes

Importance of accurate diagnosis Cont.’

- Anticipate course of the disease and its complications
- Provide appropriate therapies



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

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

Assessment

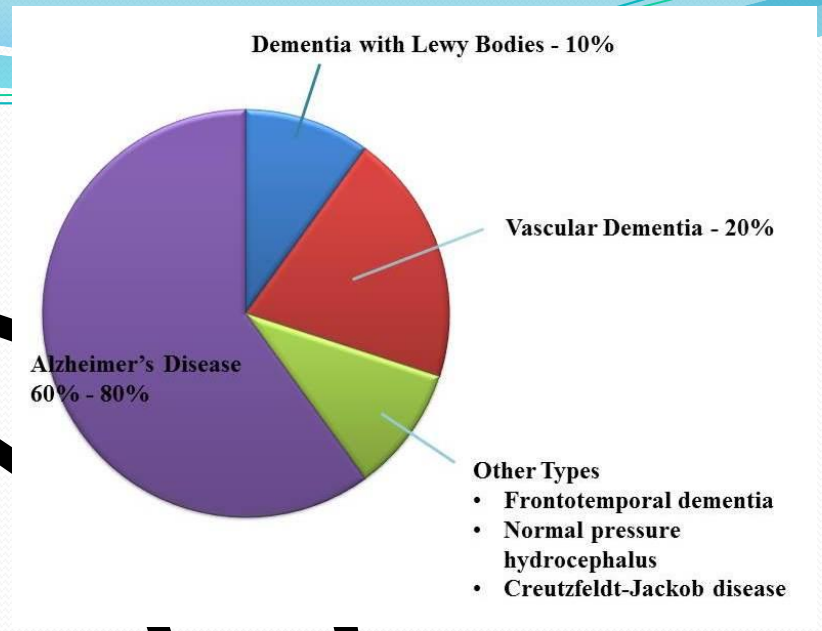
History is the most important part of assessment and it must include:

- 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
- Caregivers account

Normal Changes vs. Alzheimer's

Normal	Early Alzheimer's Disease
Can't find your keys	Routinely place important items in odd places, such as keys in the fridge, wallet in the dishwasher
Search for casual names and words	Forget names of family members and common objects, or substitute words with inappropriate ones
Briefly forget conversation details	Frequently forget entire conversations
Feel the cold more	Dress regardless of the weather, wear several skirts on a warm day, or shorts in a snow storm
Can't find a recipe	Can't follow recipe directions
Forget to record a check	Can no longer manage checkbook, balance figures, solve problems, or think abstractly
Cancel a date with friends	Withdraw from usual interests and activities, sit in front of the TV for hours, sleep far more than usual
Make an occasional wrong turn	Get lost in familiar places, don't remember how you got there or how to get home
Feel occasionally sad	Experience rapid mood swings, from tears to rage, for no discernible reason

www.helpguide.org/alzheimers



DEMENTIA

Alzheimer's Disease

- **Early - Young Onset**
- **Normal Onset**

Vascular Dementias
(Multi-infarct)

Dementia with Lewy Bodies

Fronto-Temporal Lobe Dementias

Other Dementias
- Genetic syndromes
- Metabolic pxs
- ETOH related
- Drugs/toxin exposure
- White matter diseases
- Mass effects
- Depression(?) or Other Mental conditions
- Infections – BBB cross
- Parkinson's
- NPH
- Multiple Sclerosis

How to differentiate between types of Dementia's?

- Cognitive symptoms
- Age of onset
- Reversible/Irreversible
- Male/female ratio
- Rate of progression
- Comorbidities
- Behavioral symptoms
- Treatment options
- Risk in IDD
- Genetics
- Cortical/Subcortical
- Imaging
- Neurodegeneration
- Pathology
- Biomarkers

Diagnosis of I/DD and Dementia

- Suspicion that pathologic decline in cognitive function is occurring
- Use of early warning screening and EDSD
- Neurocognitive assessments
- Avoid Diagnostic Overshadowing
- Workup and rule out/rule in accurate diagnosis
- Empiric diagnosis; Possible, Probable, Definite
- Usage of Biomarkers
- Autopsy proven

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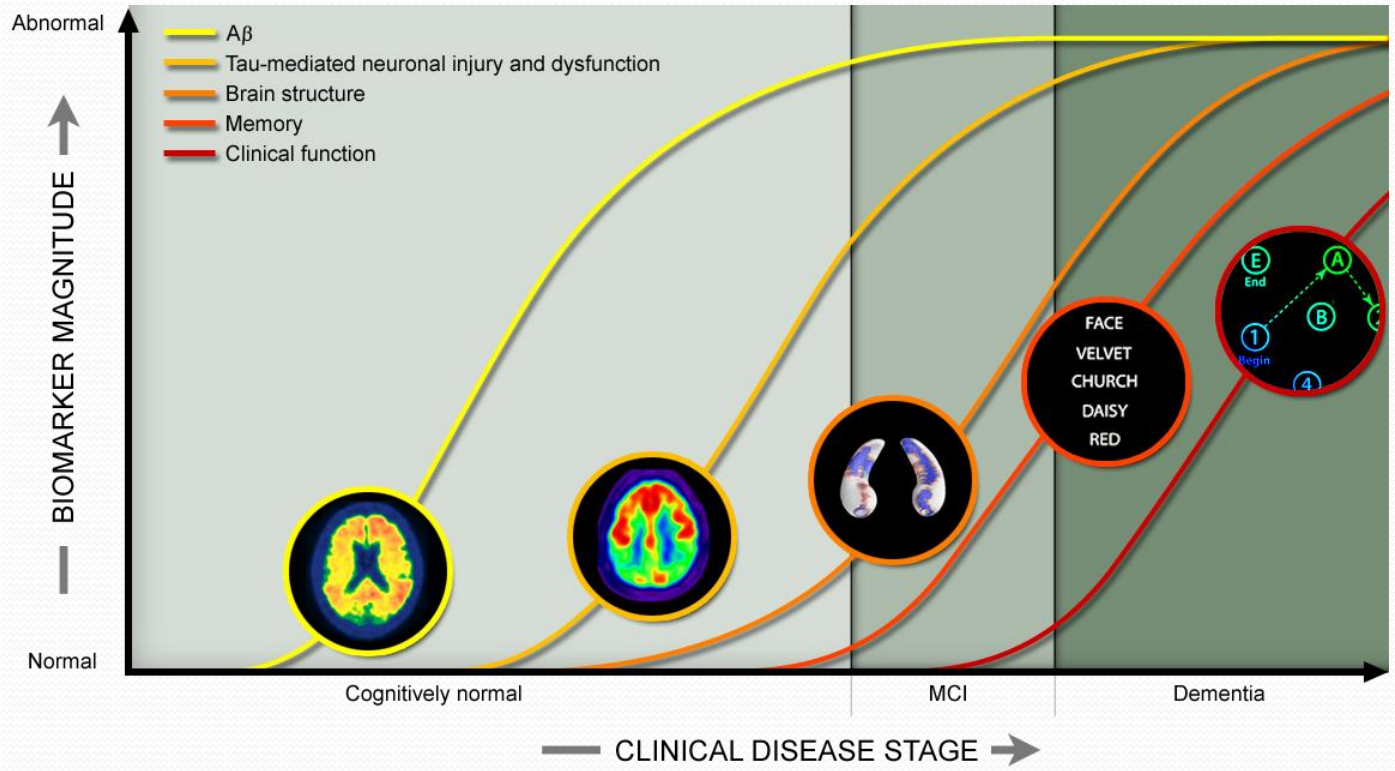
Diagnostic Accuracy

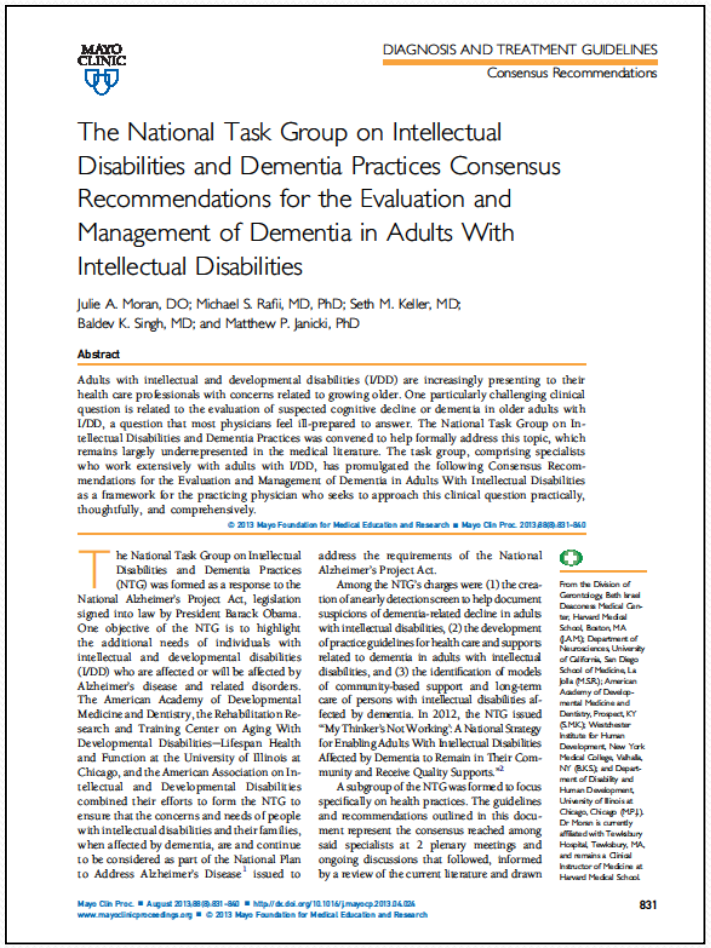
- History of current difficulties
- NTG-EDSD/Neuropsychological testing
- Physical exam
- Family and social history
- Blood testing
- EEG
- Brain Imaging
- Biomarkers



Challenges to diagnosis and care

- Individuals with I/DD 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 I/DD
- Difficulty of measuring change from previous level of functioning
- Conditions associated with I/DD maybe mistaken for symptoms of dementia - Diagnostic overshadowing
- Aging parents and siblings
- Lack of research, education, and training

The NTG Consensus Recommendations for the Health Advocacy, Evaluation and Management of Dementia in Adults with Intellectual Disabilities



MAYO CLINIC

DIAGNOSIS AND TREATMENT GUIDELINES
Consensus Recommendations

The National Task Group on Intellectual Disabilities and Dementia Practices Consensus Recommendations for the Evaluation and Management of Dementia in Adults With Intellectual Disabilities

Julie A. Moran, DO; Michael S. Rafii, MD, PhD; Seth M. Keller, MD; Baldev K. Singh, MD; and Matthew P. Janicki, PhD

Abstract

Adults with intellectual and developmental disabilities (I/DD) are increasingly presenting to their health care professionals with concerns related to growing older. One particularly challenging clinical question is related to the evaluation of suspected cognitive decline or dementia in older adults with I/DD, a question that most physicians feel ill-prepared to answer. The National Task Group on Intellectual Disabilities and Dementia Practices was convened to help formally address this topic, which remains largely underrepresented in the medical literature. The task group, comprising specialists who work extensively with adults with I/DD, has promulgated the following Consensus Recommendations for the Evaluation and Management of Dementia in Adults With Intellectual Disabilities as a framework for the practicing physician who seeks to approach this clinical question practically, thoughtfully, and comprehensively.

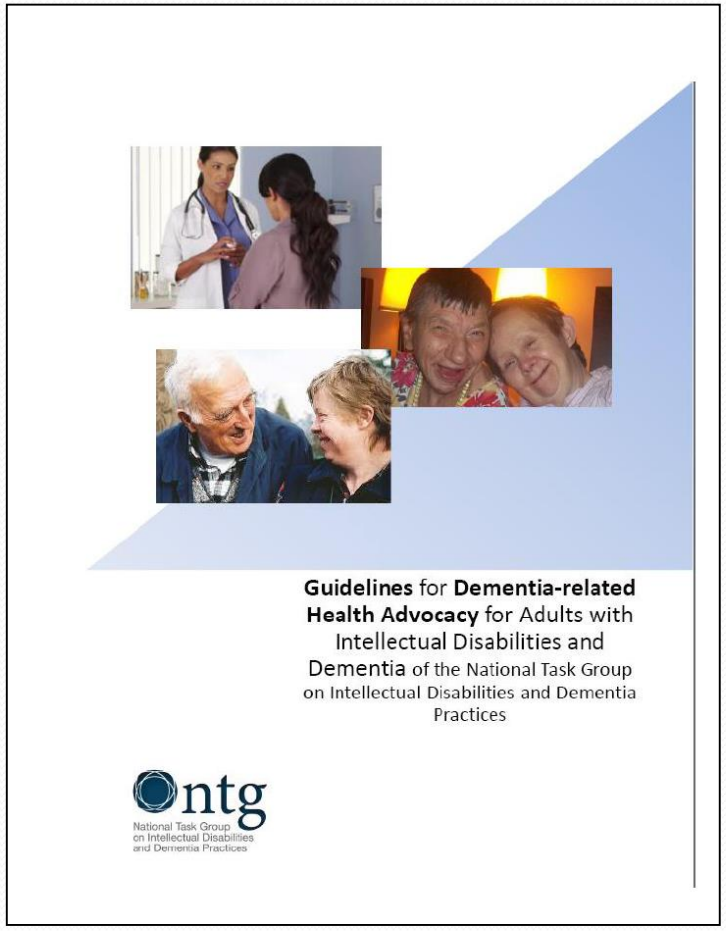
The National Task Group on Intellectual Disabilities and Dementia Practices (NTG) was formed as a response to the National Alzheimer's Project Act, legislation signed into law by President Barack Obama. One objective of the NTG is to highlight the additional needs of individuals with intellectual and developmental disabilities (I/DD) who are affected or will be affected by Alzheimer's disease and related disorders. The American Academy of Developmental Medicine and Dentistry, the Rehabilitation Research and Training Center on Aging With Developmental Disabilities—Lifespan Health and Function at the University of Illinois at Chicago, and the American Association on Intellectual and Developmental Disabilities combined their efforts to form the NTG to ensure that the concerns and needs of people with intellectual disabilities and their families, when affected by dementia, are and continue to be considered as part of the National Plan to Address Alzheimer's Disease[1] issued to address the requirements of the National Alzheimer's Project Act.

Among the NTG's charges were (1) the creation of an early detection screen to help document suspicions of dementia-related decline in adults with intellectual disabilities, (2) the development of practice guidelines for health care and supports related to dementia in adults with intellectual disabilities, and (3) the identification of models of community-based support and long-term care of persons with intellectual disabilities affected by dementia. In 2012, the NTG issued "My Thinker's Not Working": A National Strategy for Enabling Adults With Intellectual Disabilities Affected by Dementia to Remain in Their Community and Receive Quality Supports."[2]

A subgroup of the NTG was formed to focus specifically on health practices. The guidelines and recommendations outlined in this document represent the consensus reached among said specialists at 2 plenary meetings and ongoing discussions that followed, informed by a review of the current literature and drawn

From the Division of Gerontology, Beth Israel Deaconess Medical Center, Harvard Medical School, Boston, MA (JAM); Department of Neurosciences, University of California, San Diego School of Medicine, La Jolla (MSR); American Academy of Developmental Medicine and Dentistry, Prospect, KY (SMK); Westchester Institute for Human Development, New York Medical College, Valhalla, NY (BKS); and Department of Disability and Human Development, University of Illinois at Chicago, Chicago (MPJ). Dr Moran is currently affiliated with Tewksbury Hospital, Tewksbury, MA, and remains a Clinical Instructor of Medicine at Harvard Medical School.

Moran JA, et al Mayo Clin Proc 2013; 88(8): 831-840.



Guidelines for **Dementia-related Health Advocacy** for Adults with Intellectual Disabilities and Dementia of the National Task Group on Intellectual Disabilities and Dementia Practices

ntg
National Task Group on Intellectual Disabilities and Dementia Practices

Bishop KM, et al Intellect Dev Disabil. 2015 Feb;53(1):2-29.

Outcome Assessment of Care: Are Therapeutic Interventions Effective?

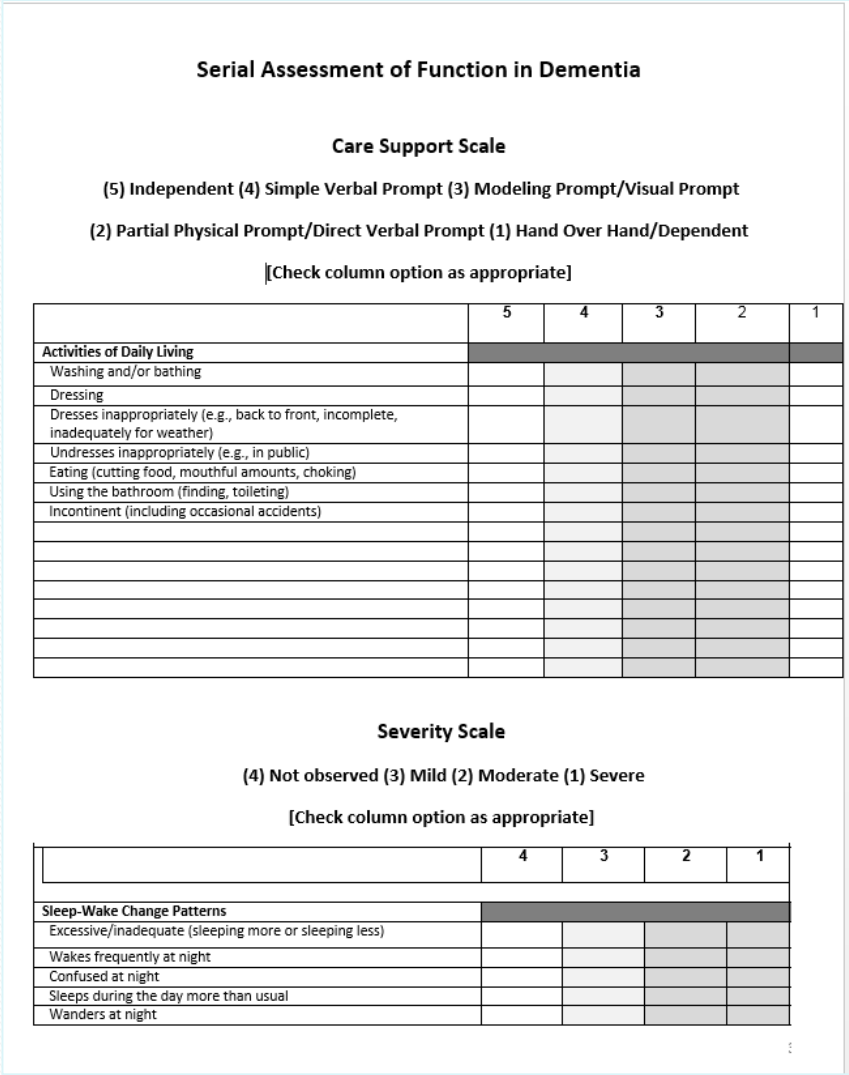
- Therapies can be positive, ineffective, or detrimental
- The degree and impact of the treatment needs to be known
- Clarity of expectations need to be discussed
- Communication of objective outcome assessments need to be defined
- Care tied to reimbursements

Assessing the Course and Magnitude of the Disease

- Helps determine therapeutic effect
- Helps to better determine the magnitude severity of symptoms of dementia
- Helps determine levels of support
- Determining level of function/ADL component tied to cognitive skills.
- The level and degree of support will increase as cognitive skills decline
- Maybe able to help with Anticipatory Guidance and disease progression and planning

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
- May be able to show evidence of serial changes in function due to disease and therapies
- Developing as a on-line tool



Serial Assessment of Function in Dementia

Care Support Scale

(5) Independent (4) Simple Verbal Prompt (3) Modeling Prompt/Visual Prompt

(2) Partial Physical Prompt/Direct Verbal Prompt (1) Hand Over Hand/Dependent

[Check column option as appropriate]

	5	4	3	2	1
Activities of Daily Living					
Washing and/or bathing					
Dressing					
Dresses inappropriately (e.g., back to front, incomplete, inadequately for weather)					
Undresses inappropriately (e.g., in public)					
Eating (cutting food, mouthful amounts, choking)					
Using the bathroom (finding, toileting)					
Incontinent (including occasional accidents)					

Severity Scale

(4) Not observed (3) Mild (2) Moderate (1) Severe

[Check column option as appropriate]

	4	3	2	1
Sleep-Wake Change Patterns				
Excessive/inadequate (sleeping more or sleeping less)				
Wakes frequently at night				
Confused at night				
Sleeps during the day more than usual				
Wanders at night				

Can cognitive decline and dementia be halted or slowed as we age?

Health Promotion and Successful Aging

Increasing Complex Cognitive Activity Later in Life *might:*

- Increase sense of purpose, motivation, and hope
- Decrease depression
- Improve level of socialization
- Offer additional outlets for emotional expression
- Decrease stress and improve coping
- Improve use of compensatory strategies
- Increase depth of processing
- Increase level of physical activity
- Increase engagement with good “role models”

Finnish Geriatric Intervention Study to Prevent Cognitive Impairment and Disability

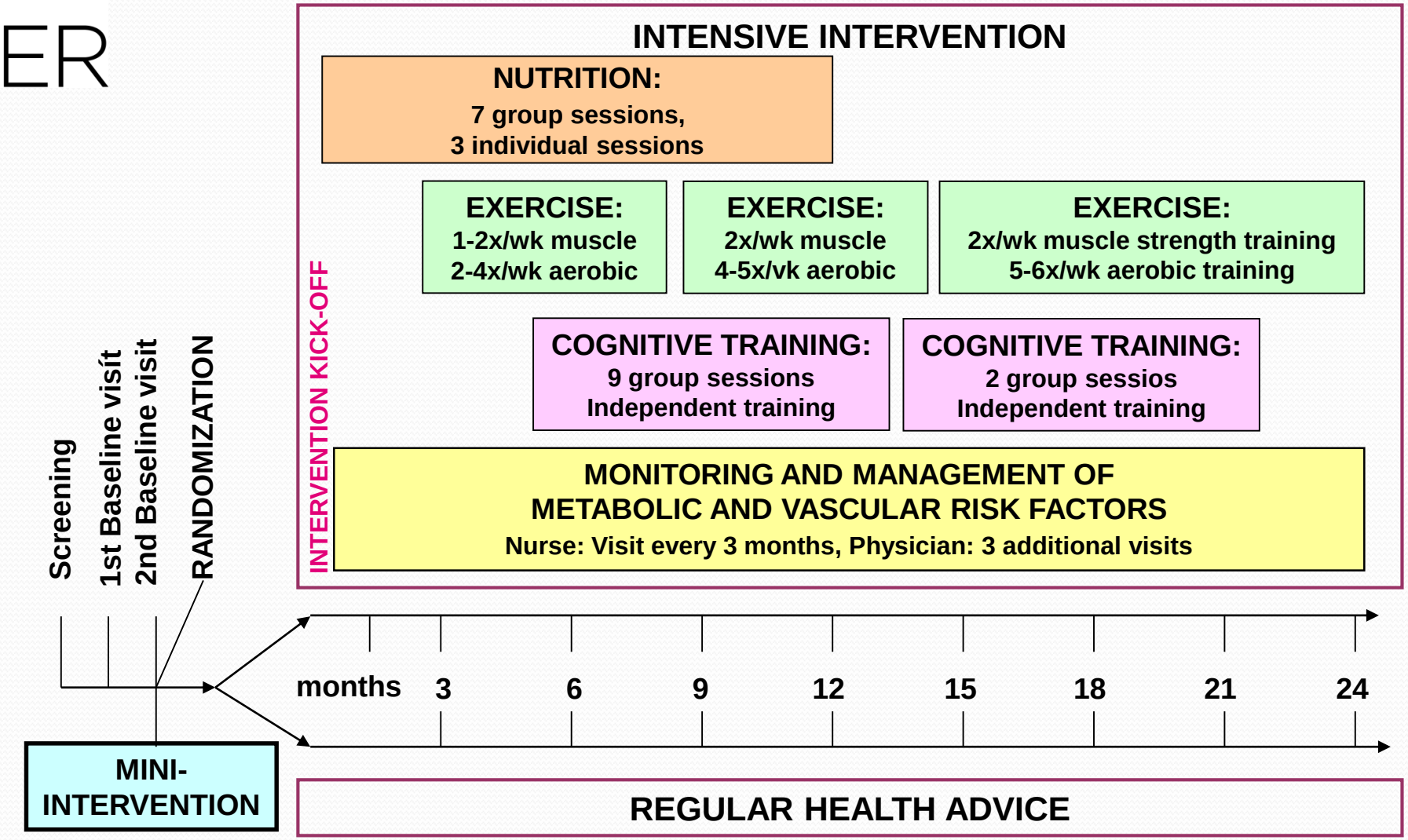
Aims to reduce cognitive impairment in an at risk population through a 2-year multi-domain life-style intervention including:

- Nutritional guidance
- Physical activity
- Cognitive and social activities
- Monitoring and management of metabolic and vascular risk factors: hypertension, dyslipidemia, obesity, impaired glucose tolerance

FINGER

Kivipelto et al., Alzheimer & Dementia 2013

FINGER



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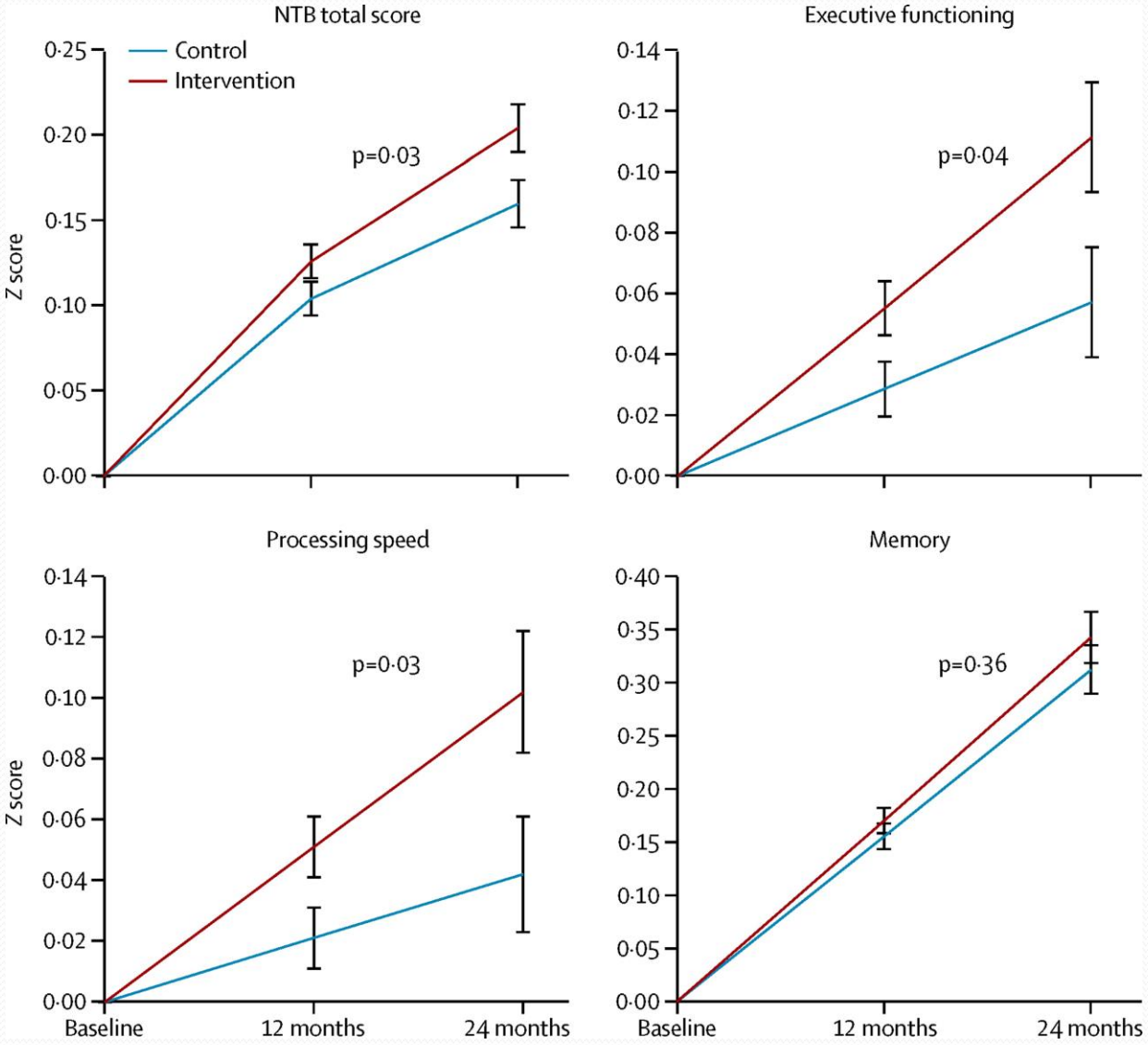

FINGER

Articles

A 2 year multidomain intervention of diet, exercise, cognitive training, and vascular risk monitoring versus control to prevent cognitive decline in at-risk elderly people (FINGER): a randomised controlled trial

Tiia Ngandu, Jenni Lehtisalo, Alina Solomon, Esko Levälahti, Satu Ahtiluoto, Riitta Antikainen, Lars Bäckman, Tuomo Hänninen, Antti Jula, Tiina Laatikainen, Jaana Lindström, Francesca Mangialasche, Teemu Paajanen, Satu Pajala, Markku Peltonen, Rainer Rauramaa, Anna Stigsdotter-Neely, Timo Strandberg, Jaakko Tuomilehto, Hilkka Soininen, Miia Kivipelto

Summary

Background Modifiable vascular and lifestyle-related risk factors have been associated with dementia risk in observational studies. In the Finnish Geriatric Intervention Study to Prevent Cognitive Impairment and Disability (FINGER), a proof-of-concept randomised controlled trial, we aimed to assess a multidomain approach to prevent cognitive decline in at-risk elderly people from the general population.

Methods In a double-blind randomised controlled trial we enrolled individuals aged 60–77 years recruited from previous national surveys. Inclusion criteria were CAIDE (Cardiovascular Risk Factors, Aging and Dementia) Dementia Risk Score of at least 6 points and cognition at mean level or slightly lower than expected for age. We randomly assigned participants in a 1:1 ratio to a 2 year multidomain intervention (diet, exercise, cognitive training, vascular risk monitoring), or a control group (general health advice). Computer-generated allocation was done in blocks of four (two individuals randomly allocated to each group) at each site. Group allocation was not actively disclosed to participants and outcome assessors were masked to group allocation. The primary outcome was change in cognition as measured through comprehensive neuropsychological test battery (NTB) Z score. Analysis was by modified intention to treat (all participants with at least one post-baseline observation). This trial is registered at ClinicalTrials.gov, number NCT01041989.

Findings Between Sept 7, 2009, and Nov 24, 2011, we screened 2654 individuals and randomly assigned 1260 to the intervention group (n=631) or control group (n=629). 591 (94%) participants in the intervention group and 599 (95%) in the control group had at least one post-baseline assessment and were included in the modified intention-to-treat analysis. Estimated mean change in NTB total Z score at 2 years was 0·20 (SE 0·02, SD 0·51) in the intervention group and 0·16 (0·01, 0·51) in the control group. Between-group difference in the change of NTB total score per year was 0·022 (95% CI 0·002–0·042, p=0·030). 153 (12%) individuals dropped out overall. Adverse events occurred in 46 (7%) participants in the intervention group compared with six (1%) participants in the control group; the most common adverse event was musculoskeletal pain (32 [5%] individuals for intervention *vs* no individuals for control).

Interpretation Findings from this large, long-term, randomised controlled trial suggest that a multidomain intervention could improve or maintain cognitive functioning in at-risk elderly people from the general population.

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Chronic Disease Prevention Unit (T Ngandu PhD, J Lehtisalo MSc, E Levälahti MSc, S Ahtiluoto MD, Prof A Jula PhD, Prof T Laatikainen PhD, J Lindström PhD, Prof M Peltonen PhD, Prof J Tuomilehto PhD, Prof M Kivipelto PhD) and Welfare and Health Promotion Unit (S Pajala PhD), National Institute for Health and Welfare, Helsinki, Finland; Karolinska Institutet Center for Alzheimer Research, Stockholm, Sweden (T Ngandu, A Solomon PhD, Prof M Kivipelto); Institute of Clinical Medicine/Neurology (A Solomon, Prof H Soininen PhD, Prof M Kivipelto) and Institute of Public Health and Clinical Nutrition (Prof T Laatikainen), University of Eastern Finland, Kuopio, Finland; Aging Research Center, Karolinska Institutet-Stockholm University, Stockholm, Sweden (A Solomon, Prof L Bäckman PhD, F Mangialasche PhD, Prof M Kivipelto); Institute of

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Where do we go from here?

Thank You!!

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http://aadmd.org/ntg