

Dementia Evaluation and Care in Aging Adults with IDD; Remembering Bill



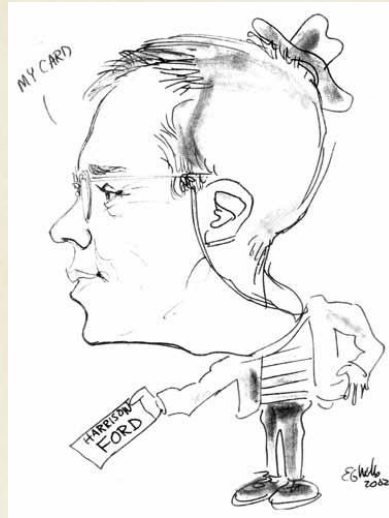
Seth M. Keller, MD

Matthew P. Janicki, PhD



Bill “Harrison Ford” Hogan






- Bill developed a number of medical problems including ear infections, hearing loss, osteoporosis and ultimately he moved into a group home
- Bill's mom "Jo" died unexpectedly in 1997
- 1998 Bill fell and broke his hip leading eventually to a hip replacement, he used a cane after that time
- A serious spine infection and bowel prolapse also complicated his life
- Bill began to show signs of cognitive decline in 2003

Concerns Cited by Families

- Incidents raising parental/carer concerns:
 - Falling
 - difficulty eating
 - no longer talking
 - increased aggression
 - short-term memory loss
 - throwing self on the floor
 - decline in general abilities
 - undressing inappropriately
 - difficulty getting out of bed
 - increased conflict with peers
 - becoming disinterested in activities
 - medical problems (e.g., seizures, incontinence)
 - other problems (such as 'trying to make guests leave house')

Recent studies have shown that carer concerns may be as good as clinical assessment in identifying dementia...

Source: Jamieson-Craig et al. (2010). Reliance on carer reports of early symptoms of dementia among adults with intellectual disabilities. *Journal of Policy & Practice in Intellectual Disabilities*, 7(1), 34–41.

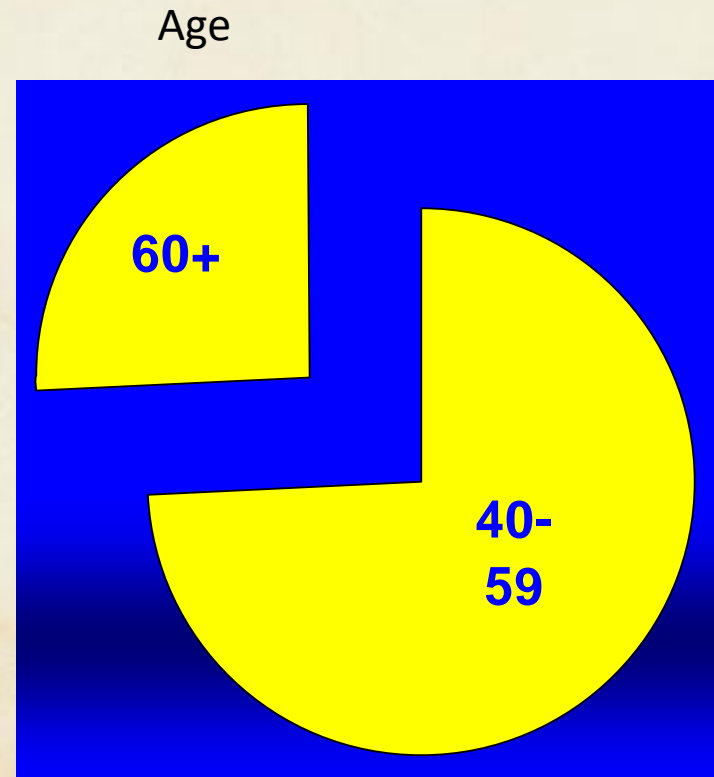


What is the research telling us about age-associated decline?

- Physical changes in old age occur with predictability
- Decline occurs in expected patterns
- Adults with motor, neurological and other significant co-conditions impacted much more adversely
- Alzheimer's disease & other dementias occur at norm except for certain conditions (e.g., Down syndrome)

Proportion of older population of adults w/intellectual disabilities

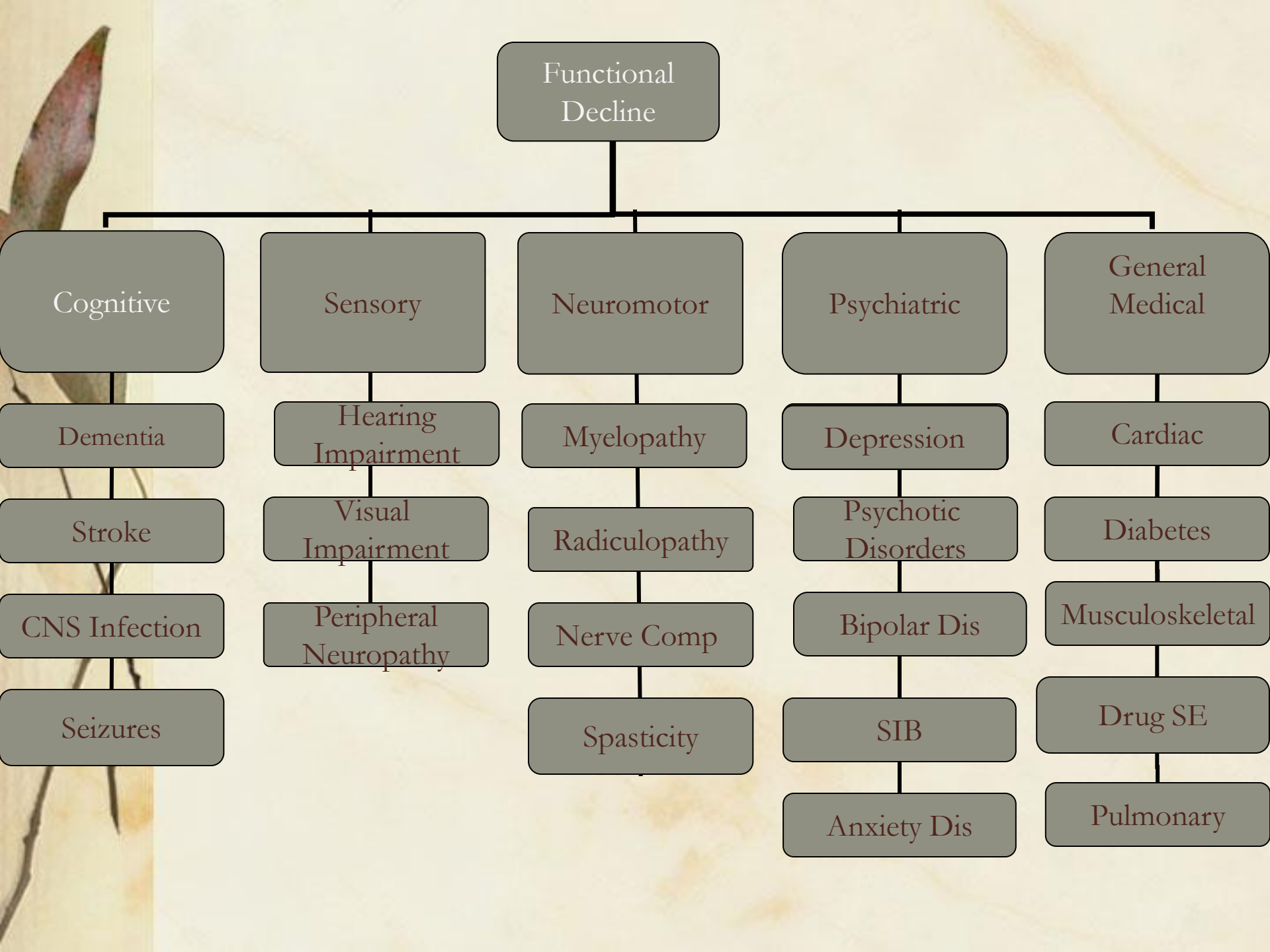
- Currently about 75% of all older adults with intellectual disabilities are in the 40-to-60 age group
- Expectations are that the 60+ age group will increase threefold over the next 20 years
- Estimated 9,000 current DD with Dementia in US
- Estimated to grow to 30,000 by 2025






Functional Decline Definition

- A process in which a person is unable to perform at the same level of activity as previously performed
 - Cognitive
 - Physical
- This is usually how persons with intellectual disabilities present when medically/psychiatrically compromised



Dementia

- Widespread loss of mental skills and abilities so that capability to care for oneself is lost
- Losses occur in memory, language skills, orientation, ADLs, and changes evident in personality and global functioning
- Syndrome which refers to progressive decline in intellectual functioning severe enough to interfere with person's normal daily activities and social relationships
- Key – loss from previous level of function
- Involves cognitive decline from any organic cause affecting the brain – such as head injury, brain disease, stroke, anoxia, AIDS – that occurs at any stage of life beyond childhood
- Causes in old age include – Alzheimer's disease (AD), stroke and related vascular accidents (VAD), neurological diseases (Parkinson's), others



DSM-IV DIAGNOSTIC CRITERIA FOR Alzheimer's Disease

- Development of cognitive deficits manifested by both
 - impaired memory
 - aphasia, apraxia, agnosia, disturbed executive function
- Significantly impaired social, occupational function
- Gradual onset, continuing decline
- Not due to CNS or other physical conditions (e.g., Parkinson's, delirium)
- Not due to an Axis I disorder (e.g., schizophrenia)



Dementia vs. Intellectual Disability

- Intellectual disabilities involve undeveloped or underdeveloped mental or intellectual skills and abilities
- Dementia is a widespread loss of mental or intellectual skills and abilities
 - Dementia related losses occur in memory, language skills, orientation, ADLS [activities of daily living], and changes evident in personality and global functioning
- Key – behavioral presentation may be similar, but it's the loss from previous level of function that differentiates the two

Prevalence of Dementia in Adults with intellectual disabilities

▶ Dementia found in adults with ID

- 3% > 40 years of age and older
- 6% > 60 years of age and older
- 12% > 80 years of age and older

▶ Dementia found in adults with DS

- 25% > 40 years of age and older
- 66% > 60 years of age and older

▶ Except for DS, the 60+ data correspond with the data reported for the population in general 60+

*Dementia is found
In 5-6% of adults
age 60 and older
in the general
population*

Sources: ¹Janicki, M.P. & Dalton, A.J. (2000). Prevalence of dementia and impact on intellectual disability services. *Mental Retardation*, 38, 277-289; ²Zigman, W.B., Schupf, N., Devenny, D., et al. (2004). Incidence and prevalence of dementia in elderly adults with MR without DS. *AJMR*, 109, 126-141.



Comments on dementia and ID

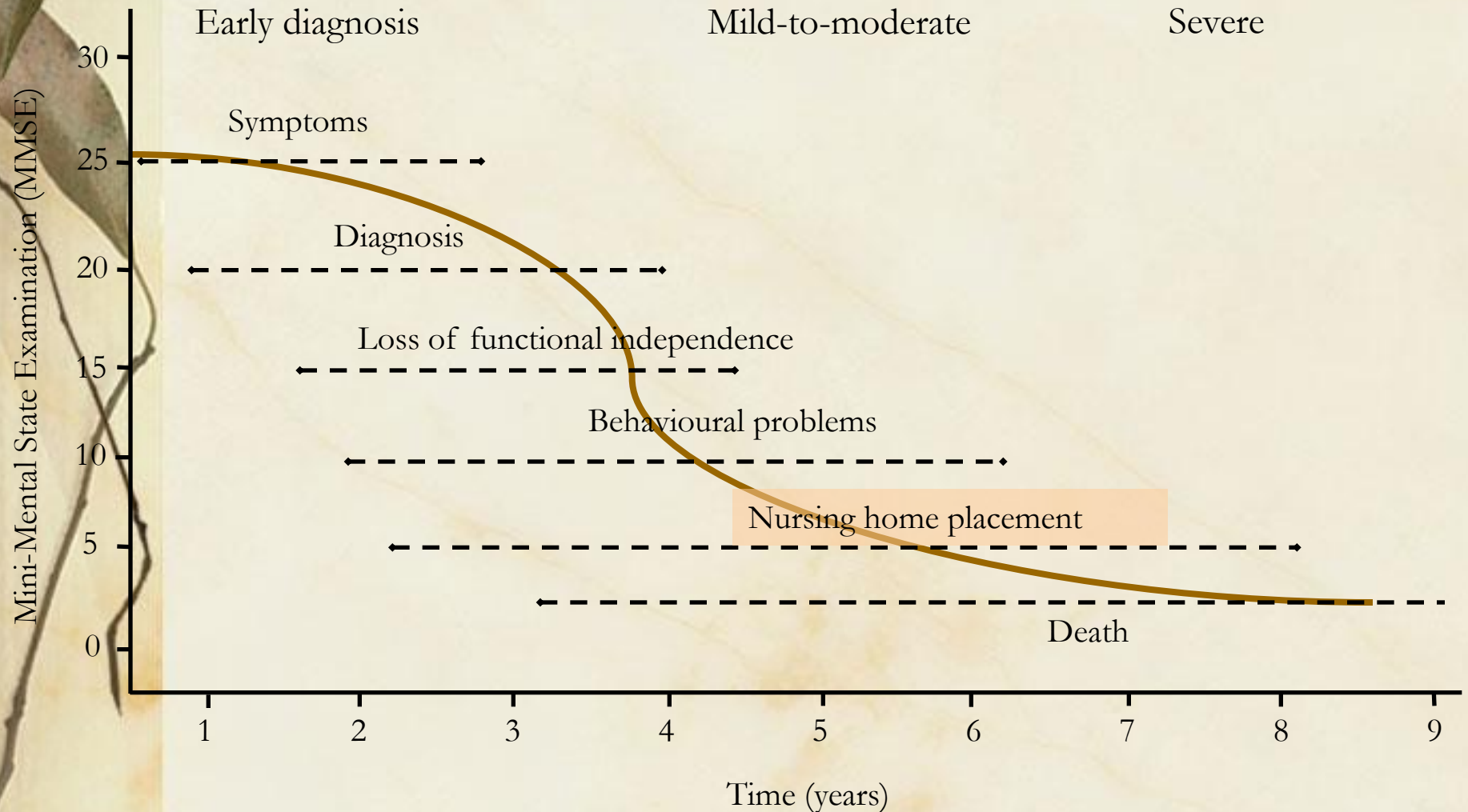
- Population of older adults with ID is growing as is the number with dementia
- Down syndrome poses a high risk for dementia of AD type
- Other-ID adults at same risk as general population
- Assessment is based on comparing the person to him or herself
- Need for awareness of warning signs and raising “index of suspicion”
- Assessment and evaluation needs to be individual-specific
- Be alert that onset and duration may vary when Down syndrome is present
- Community care is similar to other people affected by dementia

Some Background on Dementia & ID

- ▶ Rate of occurrence^{1, 3, 5, 8}
 - Age-cohort percent is same as in general population for adults with ID
 - Much higher prevalence & neuropathology indicative of AD in most adults with Down syndrome (DS)
 - ▶ *Adults w/DS make up about 10-12% of adult ID population*
- ▶ Dementia type^{2,9}
 - Type of dementia differs between DS and other types of intellectual disability
 - ▶ Generally dementia of the Alzheimer's type is prevalent in DS
 - ▶ Range of dementias found in other people with ID
 - Problem of 'diagnostic overshadowing'
- ▶ Onset, prevalence, and duration^{1, 2, 3,6,10}
 - Average onset age in early 50s for DS – late 60s for others
 - Most DAT diagnosed within 3 years of "onset" in adults w/DS
 - 26-75% of DS > 60 yo
 - 4-19% Non-DS IDD >75 yo
- ▶ Behavioral changes^{2, 3, 6}
 - More personality change in DS
 - More initial memory loss in other ID
- ▶ Neurological signs^{1, 2, 4, 7}
 - Late onset seizures found in 12%-84% of adults w/DS
- ▶ Prognosis²
 - Aggressive forms of AD can lead to death <2 years of onset in DS
 - ▶ 2-7 years mean duration in DS
 - Same longevity expected for other ID

Sources: ¹Janicki, M.P. & Dalton, A.J. (2000). Prevalence of dementia and impact on intellectual disability services. *Mental Retardation*, 38, 277-289; ²Janicki, M.P., & Dalton, A.J. (1999). *Dementia, Aging, and Intellectual Disabilities: A Handbook*. Philadelphia: Brunner-Mazel; ³Bush, A., & Beail, N. (2004). Risk factors for dementia and Down syndrome. *AJMR*, 109, 83-97 ⁴Menendez M. (2005). Down syndrome, Alzheimer's disease and seizures. *Brain Development*, 27(4), 246-252. ⁵Zigman, W.B., Schupf, N., Devenny, D., et al. (2004). Incidence and prevalence of dementia in elderly adults with MR without DS. *AJMR*, 109, 126-141. ⁶Ball, S.L., Holland, A.J., Hon, J., Huppert, F.A., Treppner, P., & Watson, P.C. Personality and behavior changes mark the early stages of Alzheimer's disease in adults with Down's syndrome: findings from a prospective population-based study. *International Journal of Geriatric Psychiatry*, 2006, [Jun 26]. ⁷Crespel, A., Gonzalez, V., Coubes, P., & Gelisse, P. (2007). Senile myoclonic epilepsy of Genton: Two cases in Down syndrome with dementia and late onset epilepsy. *Epilepsy Research*, 77, 165-168. ⁸Evenhuis, H. (1997). The natural history of dementia in ageing people with intellectual disabilities. *JIDR*, 41, 92-96. ⁹Strydom, A., Livingston, G., King, M., & Hassiotis, A. (2007). Prevalence of dementia in ID using different diagnostic criteria. *Br. J. Psychiatry*, 191, 150-157. ¹⁰Margallo-Lana et al., (2007). Fifteen year follow-up of 92 hospitalized adults with DS. *JIDR*, 51, 463-477.

Natural history of Alzheimer's Disease



What Should be Ruled Out?

- Thyroid abnormality
- Depression
- Vision & hearing loss
- Medication reactions
- Urinary tract disturbance
- Gastro-intestinal disturbance
- Nutritional deficiency
- Vitamin deficiency
- Head trauma
- Brain tumor
- Folic acid abnormalities in people taking anti-convulsants





Ideal Test

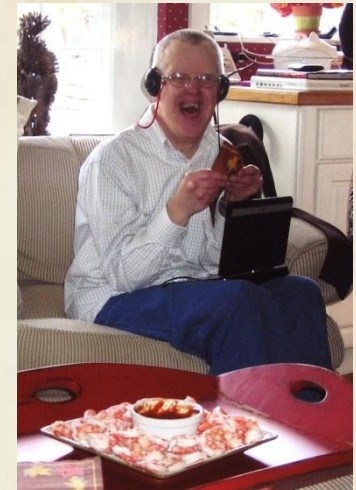


- Sensitive
- Specific
- Easy to Administer by staff/family
- Reproducible
- For various levels of ID/Verbal skills
- For different types of IDD causes/syndromes
- Differentiate cognitive decline from other causes of decline





Bill



- Diagnosed with AD January 2006, had difficulties establishing baseline cognitive functioning
- Mood dysfunction requiring *Zoloft*
- *Namenda* started then stopped due to dizziness
- *Aricept* started April 2008
- Worsening behavior, Thyroid levels markedly elevated despite taking *Synthroid* for years



What Makes Diagnosis Difficult?

- Individuals with DD may not be able to report signs and symptoms
- Subtle changes may not be observed
- Most dementia assessment tools are not relevant for people with DD
- Difficulty of measuring change from previous level of functioning
- Conditions associated with DD may mistaken for signs of dementia
- Challenges of communication between care givers and healthcare providers

National Task Group on Intellectual Disabilities and Dementia Practices



Co-Chairs
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The recently enacted National Alzheimer's Project Act (NAPA) will lead to the development of a coherent and coordinated national strategy on dealing with Alzheimer's disease in the United States. To complement this federal initiative and to address the myriad requests for more specific information and practice models for providing quality care for people with intellectual disabilities affected by dementia, the American Association on Intellectual Disabilities (AAIDD), the American Academy of Developmental Medicine and Dentistry (AADMD), along with the Rehabilitation Research and Training Center on Aging and Developmental Disabilities-Lifespan Health and Function at the University of Illinois at Chicago, have created the National Task Group on Intellectual Disabilities and Dementia Practices (NTG).

It is the intent of the National Task Group to feed into the NAPA effort and ensure that the concerns and needs of people with intellectual disabilities and their families, when affected by dementia, are considered as part of this national strategy.

The overall goal of the NTG is to review and update the technological and clinical practices used by agencies in delivering supports and services to adults with ID affected by the various dementias and produce a document which will

- (1) suggest a workable screening instrument that will help substantiate suspicions of dementia-related decline (Group S)**
- (2) produce a new set of practice guidelines for post-determination health care and supports (Group H)**
- (3) examine and recommend models of community-based support and long term care of persons with ID affected by dementia (Group C)**

For more information, access: www.aadmd.org/ntg



NTG Task Group Screen (S)

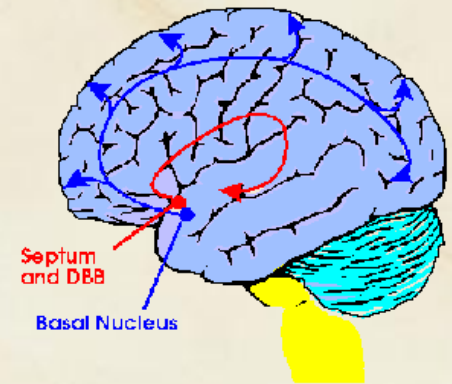
- The group will develop a protocol for a dementia-behavior sensitive screen which should be standardized and valid for the detection of cognitive decline in adults with ID who are aging.
- The screen should be relatively **brief** and contain items most sensitive for **detecting functional and cognitive change indicative of early dementia** and which can be **administered** by and interpreted by a variety of levels of staff (including **trained DSP's, nurses, and primary care docs**).
- The items chosen should be **sensitive to progressive changes** and useful for staff in making a recommendation, when viewed over time, for further diagnostics.

Dementia and Goals of Care

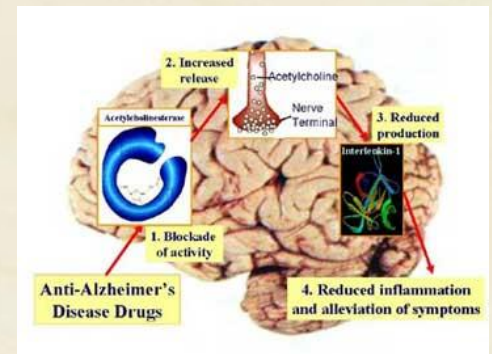
- Prolonging life
- Enhancing Q of L
- Prevent functional decline
- Slow progression
- Decrease psychiatric/behavioral problems
- Fall reduction program
- Pharmacologic and behavioral interventions
- Palliative Care
- End of Life Care



Treatment Options



- Cholinesterase Inhibitors; Aricept, Exelon, Razadyne
 - Heller, J. American Journal of Medical Genetics, Oct. 15, 2004; vol 130: pp 324-326
 - Lott IT *et al.* Arch Neurol. 2002;59:1133-1136
 - Kishnani PS *et al.* (1999) Lancet 353: 1064
- NMDA (N-methyl-D- aspartate) receptor antagonist; Namenda
- Behavioral Modification/Psychotropics
- Research
 - Anticholinergics
 - Nicotine
 - Homocysteine
 - Huperzine A
 - NSAIDS



Response to medication

Donepezil in Adults with Down Syndrome With and Without Dementia					
Author	Journal	Year	No subjects	Study Type	Results
Kishnani, P.S., et al.,	Lancet	1999	4	Case Reports No dementia	Improvement
Heller, J. H. et al	AJ Medical Genetics	2003	6	Case Reports No dementia	Improvement Language
Johnson, N. et al	AJMR	2003	19	RCT No dementia	Improvement Language
Prasher, V.P., et al.,	Intl J Ger Psych.	2002	27	RCT Alzheimer's Disease	Non significant improvement
Lott, I.T., et al.,	Archives Neurology	2002	15	Case Control Alzheimer's Disease	Significant Improvement
Prasher, V.P., et al	Intl J Ger Psych.	2003	25	Open label extension Alzheimer's Disease	Significant improvement
Kondoh, T. Et al	Annals Pharmacotherapy	2005	2	Case Reports Alzheimer's Disease	Improvement



Behavioral Changes in Persons with Alzheimer's Disease

- Nearly all persons with AD exhibit behavioral changes
- Diverse behavioral symptoms occur
- Multiple symptoms occur simultaneously
- Behavioral changes become more frequent with disease progression
- Behaviors are recurrent after onset

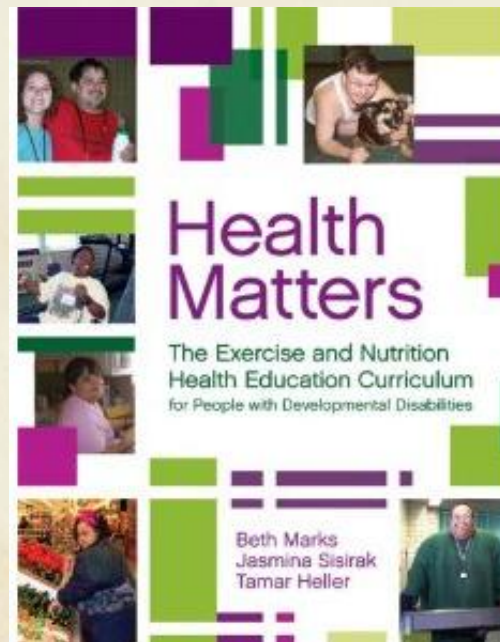


Nonpharmacological Strategies

Also Remember the four S's

- Maximize Safety and Limit the Risks
- Promote Structure and Consistency
- Enhance Serenity and Limit Confusing Stimuli
- Nurture Sanity and Supports for All Caregivers

Exercise and Activities as Brain Food



Rehabilitation Research and Training Center (RRTC) on Aging with
Developmental Disabilities: Lifespan Health and Function, UIC at Chicago



What Makes Evaluation and Treatment Difficult?

- Assessment and diagnostic tools not uniform
- Communication between support staff and healthcare providers inadequate
- Access to and availability of Caring Providers
- Data collection of decline inconsistent
- Treatment responses inadequately assessed
- Preventive Care inconstantly provided
- Toxicity and polypharmacy poorly assessed
- Pharmacologic and non-pharmacologic behavior management inadequately assessed
- Transition care problematic
- Palliative care and End of Life care inadequate



NTG Task Group Health (H)

- The group will focus on a standardized plan for a process to follow if decline is suspected, usage of the **screening tools**, and implementation of **outcome measures** to assess treatment outcomes when medical therapies are employed.
- The effort should lead to the definition of a set of practice guidelines that could be adopted for clinical practice and by regulatory agencies for defining **best practice in following through on suspicions and post-confirmation of presence of dementia**.
- The Group will focus on applications of a **multidisciplinary** approach and eventual adoption of a person centered care plan for persons with ID affected.

Bill



- May 2009 -Walking problems progressed including falls, he went from using a cane to a walker to a wheel chair
- Sleep problems increased, *Trazadone* was added
- Seizures began, *Keppra* was begun, but it increased agitation. Switched to *Lamictal*. Myoclonic jerks began; *Klonopin* added temporarily.
- Behavioral outbursts and agitation increased, appetite and weight deteriorated
- Bill moved into another group home in July 2009 to help with his increasingly difficult behavior and worsening health issues
- November 2009- *Tramadol* was prescribed for apparent pain and oxygen at night



Preparing for dementia

Knowns...

- People with ID have same rate of dementia as general population
- Some people with ID have higher rates (e.g., Down syndrome, head injury)
- Some % of any adult client pool will be affected

Unknowns...

- Who will be affected
- How long will person live after dx
- What other diseases or medical conditions may be co-incident
- What particular dementia-related behaviors will be more evident



Dementia & families

- Persons with DS & ID who live with parents – and develop dementia – pose severe challenges for parents
 - Behavior will deteriorate
 - Person may remain ambulatory
 - Physical needs will become more prominent
- Aging parents may less capable of continuing to provide care at home
- Situation may lead to crisis at home



Edinburgh Principles for people with DD and dementia

- Promote utmost QOL: base services on person-centered approach
- Individual strengths, capabilities, skills and wishes should be overriding consideration in decision-making
- Involve individual and family in all phases of assessment, service planning and provision
- Ensure appropriate diagnostic, assessment and intervention services available

Edinburgh Principles

- Plan and provide supports and services that optimize remaining in chosen home and community
- Ensure same access to services and supports as those in general population affected by dementia
- Ensure generic, cooperative and proactive strategic planning

Dementia care planning

	Early stage	Mid stage	Late stage
Administrative	Plan forward for more focused care with progression; lock in funding to cover extended care	Set up specialty care programs or support teams for in-home care; augment staffing	Adjustments to staff and physical features of home
Clinical	Early screening and diagnostics; medical oversight	Periodic reassessments and follow-up; check for coincident conditions	Nursing care evaluations; medical oversight
Programmatic	Introduce memory aids; watch for changes in function; reduce options	Closer supervision; specialized care; wandering controls; memory exercise; move to specialty homes	Careful attention to avoid dehydration and malnutrition; avoid 'bed sores'; introduce hospice care


Training/ staff education

- Aging
- Diseases and conditions of older age
- Medications
- Dementias
 - Alzheimer's, vascular, fronto-temporal, Lewy body, etc.
- Care management
- Communication & activities
- Environmental adaptations





E EARLY SCREENING AND DIAGNOSTICS
C CLINICAL SUPPORTS
E ENVIRONMENTAL MODIFICATIONS
P PROGRAM ADAPTATIONS
S SPECIALIZED CARE



Dementia care Options

- *Institutional care* – long term care facilities, nursing homes, old age homes, dementia special care units
- *Neighborhood group care* – generic group homes, specialized ID group homes
 - “in-place progression” model
 - “aging-in-place” model
- *Family care* – living with family, other relatives, or other family members or carers



What Makes Supportive Care Difficult?

- Staff education and training
- Lack of a appropriate dementia specific environment
- Inability to be proactive and flexible with dementia progression and complications
- Inadequate communication amongst support team
- Lack of a palliative care and end of life program
- Inadequate funding stream to support needs of the individual
- Lack of federal, state, and county dementia policies



What can be done at agency level

- Collect ‘personal best’ data
 - video, charting, biographical data
- Institute screening
 - Formal screening instruments
 - Informal assessment of behavior
- Refer for formal diagnostics
 - Local expert ‘memory’ & Alzheimer’s clinics
- Continue re-assessments over time
 - Monitor change
 - Confirm preliminary diagnoses
- Consult with diagnosticians



NTG Task Group Community Supports (C)

- The group will examine models of informal and formal support and care that can offer long term care in community settings, including continued aging in family homes, specialized support in 'dementia capable' group homes, and aging in place supports during early stage care.
- The group will define specific **supports that agencies and interest organizations may adopt** to anticipate signals of decline or symptoms of early stage dementia, adaptations to physical conditions and designs of the living settings, adaptations of day supports and services, and tracking of trajectories of decline... also, to be defined will be **staffing needs** and patterns, as well as training topics.
- The group will also examine and comment on **adaptations for long-term care that are stage specific.**

National planning – strategic plans

- In USA... *National Task Group on Intellectual Disabilities and Dementia Practice*
- Aims

- To identify a workable screening instrument applicable in all USA
- To define practice guidelines for medical and allied health community
- To define ‘best practices’ for community care of adults with ID and dementia
- Tie NTG into National Alzheimer’s Project Act (NAPA)

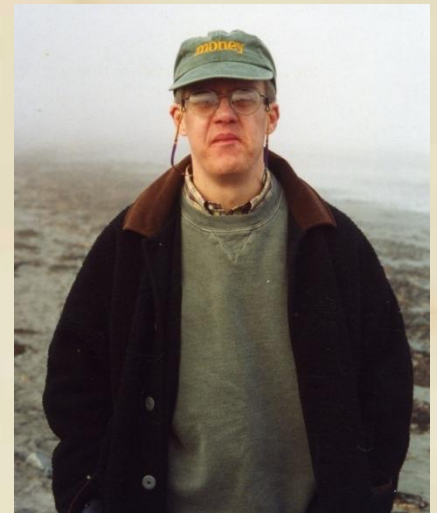
The screenshot shows the website for the National Task Group on Intellectual Disabilities and Dementia Practices. The main heading is "National Task Group on Intellectual Disabilities and Dementia Practices". Below the heading, there is a video player with a play button. To the left of the video, there are navigation links: "D LINKS", "Overview", "Group Description", "Group Sign-up", "Meeting Minutes", "Members", "Supporting Organizations", and "Documents". Below these links, there is a section titled "DMD Information" with sub-sections: "AAIDD Board Members", "About AAIDD", "Board Meeting Minutes", "Scientific Tools", "Find a Doctor", "Membership Benefits", and "National Task Group on ID and Dementia". To the right of the video, there is a section titled "Find us on Facebook" with the logo for "The American Academy of Developmental Medicine and Dentistry". Below the Facebook link, there is a small image of a person and some text. At the bottom of the page, there is a section titled "Meeting Opportunities" with sub-sections: "Understand and Use Change Goals", "Special Olympics", "ADDA", "AAIDD Conference", "Annual AAIDD Conference", "The National Leadership Consortium on ID", and "The National Task Group on Intellectual Disabilities and Dementia Practices".

<http://www.aadmd.org/NTG>



Bill Hogan

4/30/60 - 2/25/10



Mark Twain was once quoted as saying, "Let us live so that when we come to die, even the undertaker will be sorry."

Bill's was a life well lived. We bid him adieu and can almost hear him signing off with his favorite, "Too-da-doo." Toodle loo to you, too, Bill. See you on the other side.

Dedicated to Bill, and all of the past, current, and future sons, daughters, brothers and sisters affected



Thank You!

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