People with Intellectual or Developmental Disabilities and Dementia

2013 NIH/ACL Alzheimer’s Webinar Series
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WELCOME

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Dementia in Adults with Intellectual and Developmental Disabilities

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Dementia in adults with I/DD

• Understanding dementia
• ‘Early onset’ dementia in Down syndrome
• Challenges to diagnosis and care
• Assessments
• Care guidelines
• Tips for advocates
• The “Team” and needed services
Understanding 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
- Effects of dementia will be progressive and eventually lead to death
- Early interventions can aid in adapting to changes and prolonging lucid periods

**Unknowns...**

- Who will be affected?
- How pronounced will be early changes?
- How dramatic will be the changes in function?
- How long will person live after diagnosis?
- What other diseases or medical conditions may be co-incident?
- What particular dementia-related behaviors will be more evident?
Percentage of people with Down syndrome who develop dementia at different ages:

<table>
<thead>
<tr>
<th>Age Group</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>30’s</td>
<td>2%</td>
</tr>
<tr>
<td>40’s</td>
<td>10-15%</td>
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<tr>
<td>50’s</td>
<td>33%</td>
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<tr>
<td>60’s</td>
<td>50-70%</td>
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Source: Mann (1993) – [based on 39 published studies n=434]
Issues that arise with respect to dementia and Down syndrome

✔ Much higher prevalence of neuropathology indicative of AD in most adults with Down syndrome (DS)

✔ Generally dementia of the Alzheimer’s type is prevalent in adults with DS

✔ Average onset age in early 50s for DS (late 60s for others)

✔ Most DAT diagnosed within 3 years of “onset” in adults w/DS

✔ More initial personality change in DS (rather than memory loss)

✔ Late onset seizures found in large number of adults w/DS

✔ Duration generally is from 2 to 7 years

✔ Aggressive forms of AD can lead to death <2 years of onset in adults w/DS
Issues that arise with respect to dementia and Down syndrome

- Older adults with Down are at high risk of Alzheimer’s disease
- Not every adult will show signs of dementia as he or she ages
- Age-associate decline may be due to aging and not dementia
- Institute baseline for (‘personal best’) functioning at age ~40
- Useful to know the signs of MCI and dementia and keep track of capabilities after age 40
- Early detection screening useful to identify possible progression into MCI or dementia
- Early referral for assessment or diagnosis if signs present is advised
Critical factors

- Degree of retention of function
- Expected trajectory of progressive dysfunction
- Duration (remaining life years)
- Type of dementia
- Health status
- Environmental accommodations

Varying trajectories have implications for continual assessment and adaptations to care management

Source: Figure 1 from Wilkowsz et al., (2009). Trajectories of cognitive decline in Alzheimer’s disease. *International Psychogeriatrics*, 21, 1-10
Small change in cognitive capability could have profound impact on independence
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 and diagnostic overshadowing
- Environmental influences may be more important in I/DD
- Aging parents and siblings
- Lack of research, education, and training
Assessing the “problem”

• Knowing that a change in function is a concern; when is a change in function part of normal aging or not?
• Diagnostic overshadowing
• Benefits of early diagnosis
• Documenting a change in function has occurred from a prior established baseline (NTG-EDSD)
• Diagnostic Assessment tools
• Change of care perspective
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

Use: to provide information to physician or diagnostician on function and to begin the conversation leading to possible assessment/diagnosis

http://aadmd.org/ntg/screening
Informant-report and objective measures for clinical assessment of dementia in people with intellectual disabilities

- Adaptive Behaviour Dementia Questionnaire (ABDQ)
- Assessment for Adults with Developmental Disabilities (AADS)
- Dementia Questionnaire for People with Learning Disabilities (DLD)
- Dementia Scale for Down Syndrome (DSDS)
- Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID)
- Prudhoe Cognitive Function Test
- Test for Severe Impairment
Important Care Perspectives

• Change of care focus
  • Going from making gains to that of maintaining function and dealing with eventual loss and decline

• Supporting family caregivers
  • Recognize the challenges faced by the aging caregiver
  • Support sibling and parent care providers
Guidelines for Structuring Community Care and Supports for People With Intellectual Disabilities Affected by Dementia

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Nancy Jokinen
Matthew P. Janicki
Seth M. Keller
Philip McCallion
Lawrence T. Force and the National Task Group on Intellectual Disabilities and Dementia Practices

http://aadmd.org/NTG
## Some Key Features of the Guidelines

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<th>Section</th>
<th>Features</th>
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<td><strong>Background</strong></td>
<td>NTG initiative, underlying principles, dementia as it affects people with ID</td>
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<td><strong>Staging Model</strong></td>
<td>Early recognition of symptoms → late stages</td>
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<td>Expected changes in behaviour &amp; function</td>
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<td>Recommended actions</td>
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<tr>
<td><strong>Early Detection</strong></td>
<td>Use of an early detection tool</td>
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<td>Assessment &amp; diagnosis period</td>
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<td><strong>Program / Support Options</strong></td>
<td>Critical concerns in varying circumstances</td>
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<td>Non pharmacological approaches</td>
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<td><strong>Auxiliary Issues</strong></td>
<td>Abuse, financial, managing choice &amp; liability, medications and nutrition</td>
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<tr>
<td><strong>Collaboration</strong></td>
<td>Intellectual disability, aging, Alzheimer’s, health</td>
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<tr>
<td></td>
<td>Policies, programs, services</td>
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</table>
Tips for Health Care Advocacy

• Be aware of myths and stereotypes about aging in persons with I/DD
• Know the individuals; who they are and how they’ve been focusing on specific ADLs
• Never assume it is the result of normal aging! Diagnostic overshadowing
• Know the possible side effects and interactions for medications used by the individual
• Differential diagnosis
• Be prepared for visit
Tips for Health Care Advocacy

• Be empowered
• Form alliances and partnerships with health care team
• Appreciate aging parents’ issues
• Understand and create support structure; aging and I/DD
• Determine expectations and goals
What kinds of services are needed for people with ID and dementia?

• Supports for continued living with families when available and appropriate
• Engaging activities in community settings
• Health reviews and surveillance by clinicians who understand ID, aging, and neuropathologies
• Appropriate screening and assessments for aging-related conditions
• Health maintenance – nutrition and regular physical exercise
• Supports for ‘dementia-capable’ care in community care settings that can change as the disease progresses; including education and training
Team Approach to Care

- Improving outcomes
- Respect for need and opinions of team members
- Helps anticipate and prepare for decline
- Who is the team??
Aging adults with ID...

- are a vulnerable population and may need special help when dementia symptoms arise
- may have significant co-morbidities - from a lifetime of challenges
- often need specialized housing and care settings to preclude being institutionalized as they age
- could be residing with older parents who themselves are declining and who may need additional help
- may be difficult to assess due to lifelong cognitive impairments or inabilities to self-report
- can benefit from partnership arrangements between aging network and I/DD providers
Help for DD and dementia

On-line
- [http://aadmd.org/ntg](http://aadmd.org/ntg)
  - For publications, screening tools, and other resources on dementia and I/DD
  - For information on Down syndrome and dementia
- [http://www.ndss.org](http://www.ndss.org)
  - Look for their new booklet: *Aging and Down Syndrome: A Health & Well-being Guidebook*

State, regional, local
- Contact
  - Your state developmental disabilities agency
  - Your state aging agency
  - Your area agency on aging
  - Your state chapter of The Arc
  - Your state developmental disabilities planning council
  - Your state or local chapter of the Alzheimer’s Association
  - Your state’s Caregiver Support Program
  - Local dementia assessment clinics or centers
The National Task Group is largely supported by the American Academy of Developmental Medicine and Dentistry and the University of Illinois at Chicago’s RRTC on Aging and Developmental Disabilities – Lifespan Health & Function

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Co-chairs

Visit us at – http://aadmd.org/ntg
Overview of Research on Down Syndrome and Aging: Opportunities and Challenges

Ira T. Lott, MD
Professor Emeritus
University of California, Irvine
Outline

• Mouse models
• Cognitive functioning
  • Clinical trials
• Telemedicine
Neuropathological Topography (Lott and Diersessen, 2010)
Mouse Models for Down Syndrome

• Overexpression of same genes

• Can study individual genes

• Can experimentally manipulate model

• Offers a platform for translational research
Haydar and Reeves, 2012

Developing Trisomic Mouse

- Forebrain Excitatory Neurons
  - fewer, delayed production
  - delayed differentiation
  - dendritic spine abnormalities
  - fewer excitatory synapses
  - more inhibitory synapses

- Forebrain Inhibitory Neurons
  - increased neurogenesis from MGE
  - over-produced PARV and SOM
  - high activity of GABAergic neurons

- White Matter
  - delayed arrival of thalamic axons
  - increased size of Olig2+ precursors

- Hippocampus
  - more PARV and SOM interneurons
  - hyper-inhibition of pyramidal neurons
  - decreased LTP, enhanced LTD

- Cerebellum
  - reduced overall volume
  - fewer Purkinje neurons
  - reduced production of granule neurons

Adult DS Human

- Forebrain Excitatory Neurons
  - fewer
  - dendritic spine abnormalities
  - fewer excitatory synapses
  - more inhibitory synapses

- Forebrain Inhibitory Neurons
  - ?

- White Matter
  - progressive early demyelination

- Hippocampus
  - reduced volume
  - hypocellular

- Cerebellum
  - reduced overall volume
  - fewer Purkinje neurons
Intellectual disability in DS (Dierssen review, 2012)

• Disruption in keeping incoming information on line, mental computation, and storage
  • Uneven working memory
  • Visual short term memory worse than visual-spatial memory
  • May impair downfield cognitive performance in language, vocabulary and problem solving
Early Onset Aβ in Down syndrome

4 months old – anti-Aβ1-16 immunostaining in free-floating formic acid pretreated 50 µm thick formalin-fixed sections.
Diffuse plaques are associated with neurodegeneration.
Effect of synaptic disruption on executive functioning in DS

- Executive function refers to cognitive operations that regulate other processes such as attention, planning, working memory

- Adolescents with DS show impairments in task assessment, set shifting, and working memory reflective of executive dysfunction (Lafranchi 2010)

- Executive dysfunction becomes more marked with age and dementia changes (Ball, 2008; Adams 2010)
Emotional Changes in Early Dementia

• Apathy, indifference, pragnosia associated with abnormal neurological findings and atrophy on brain MRI (Nelson et al 2001)

• CAMDEX informant measures show impaired frontal functioning in preclinical AD in DS

• Measures of cognition, receptive language, behavior and executive functioning implicate frontal lobes in early dementia in DS
Examples

• Arizona test battery (Edgin et al, 2010)

• Broad social and cognitive function measures (Zigman et al 2008)

• Working group on assessment of aging and dementia in DS (Ayward and Burt, 2000)
Factor analysis of neuropsychological tests and domain correlations
Beta-amyloid Deposition in Dogs: Comparison with Human Brain

Oxidative damage is a key feature of the aged canine brain
Mitochondria and Oxidative Stress in Down syndrome (Coskun et al)

- Control region mutations seen in brain and peripheral tissues from individuals with DS, DS+AD, and AD in the general population
Research participation flowchart

1. Invited for screening\n   \n   - Did not attend screening\n     \n   - Enrolled/Screened\n     \n   - Eligible\n     \n   - Randomized\n     \n   - Allocated to antioxidant\n     \n   - Initiated allocated intervention\n     \n   - Evaluated - Year 1\n     \n   - Evaluated - Year 2\n     \n   - Discontinued\n     \n   - Allocated to placebo\n     \n   - Initiated allocated intervention\n     \n   - Evaluated - Year 1\n     \n   - Evaluated - Year 2\n     \n   - Discontinued

2. \n   \n   - Ineligible\n     \n   - Refused to participate

3. \n   \n   - Non-AD\n     \n   - Non-DS
Alpha-tocopherol Level in Plasma shows compliance with regimen
Considerations

• Future clinical trials need to address health disparities in DS and AD

• Telemedicine is the use of health care technology when distance separates the doctor and patient
Telemedicine
TM program UCI 2001-PRESENT

- 2100 clinics
- 8400 consults
- California Center for Connected Health
- California Telehealth Network
## Telemedicine Screening for Dementia in DS

<table>
<thead>
<tr>
<th>Description</th>
<th>Count</th>
</tr>
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<tbody>
<tr>
<td>Subjects identified</td>
<td>358</td>
</tr>
<tr>
<td>Subjects screened</td>
<td>334</td>
</tr>
<tr>
<td>Subjects screened positive</td>
<td>97</td>
</tr>
<tr>
<td>Subjects declined to participate</td>
<td>24</td>
</tr>
<tr>
<td>Subjects examined</td>
<td>37</td>
</tr>
<tr>
<td>Subjects diagnosed with dementia</td>
<td>8</td>
</tr>
<tr>
<td>Subjects placed on medication therapy</td>
<td>8</td>
</tr>
<tr>
<td>Subjects diagnosed with a pseudodementia</td>
<td>4</td>
</tr>
</tbody>
</table>
UCI Down Syndrome Team

• Eric Doran, MS-Program Manager

• Nina Movsesyan, PhD-Research Coordinator
  
  • Anne Tournay, MD-Neurologist
  
  • Mindora Totoui, MD, PhD-Neurologist

• Pinar Coskun, PhD-Mitochondria Studies
  
  • David Walsh, PhD-Psychologist

• Supported in part by HD25912, HD065160, AG16572; State of California
Federal Resources

Andrew Morris, MPH
Administration on Intellectual and Developmental Disabilities
Administration for Community Living
E-mail: andrew.morris@acl.hhs.gov
Phone: 202-690-5985
AIDD Resources

- AIDD programs
  - University Centers for Excellence on Developmental Disabilities
    • Have dementia related grants and research
  - Protection & Advocacy
    • Legal and advocacy services for people with disabilities
  - Developmental Disability Councils
    • Policy and advocacy resources
  - Projects of National Significance
ACL and Resources

• Alzheimer’s Disease Supportive Services Program
  – Supports efforts to expand the availability of community-level supportive services for persons with Alzheimer’s and their caregivers and improve the responsiveness of the home and community-based care system to persons with dementia. Includes translation of evidence-based interventions into effective supportive service programs at the community level.
ACL and Resources

• National Alzheimer’s Call Center
  – National information and counseling service for persons with Alzheimer’s disease, their family members, and unpaid caregivers. Available in 56 states and territories, 24 hours a day, 7 days a week, 365 days a year, the Call Center provides expert advice, care consultation, information, and referrals nationwide, at the national and local levels.
National Alzheimer’s Contact Center

Call 800-272-3900
Staffed 24/7
Information also available on-line, via email, message boards, etc.
ACL and Resources

• National Family Caregiver Support Program
  – Funds a range of supports that assist family and informal caregivers to care for their loved ones at home for as long as possible. The program supports five services: information to caregivers about available services; assistance to caregivers in gaining access to the services; individual counseling, organization of support groups, and caregiver training; respite care; and supplemental services.
  – Go to http://eldercare.gov to find program specifics for local areas
ACL and Resources

• Lifespan Respite Care Program
  – Support, expands, and streamlines the delivery of planned and emergency respite services while also providing for the recruitment and training of respite workers and caregiver training and empowerment.
Alzheimer's is...

The most common form of dementia causes problems with memory, behavior, and thinking that worsen over time, eventually leading to death. There is no cure. Over 6 million people in the United States have the disease. Alzheimer’s is not a normal part of aging.

Alzheimer's and Down Syndrome

Alzheimer's disease occurs three to five times more often among people with Down syndrome than the general population. People with Down syndrome are also more likely to develop Alzheimer's disease at a younger age than other adults.

As with all adults, advancing age also increases the chances that a person with Down syndrome will develop Alzheimer's disease. Estimates vary, but it is reasonable to conclude that 25 percent or more of people with Down syndrome who are older than 35 show clinical signs and symptoms of Alzheimer’s-type dementia.

However, it is important to note that not everyone with Down syndrome develops Alzheimer's symptoms.

Important Links

The NIH Alzheimer's Disease Education and Referral Center
Information about symptoms at each stage and how the disease progresses

The Memory Loss Tapes

Alzheimer’s Association
What is Dementia?

Down Syndrome and Alzheimer's Disease

Dementia and Intellectual Disabilities (PDF)
ACL Alzheimer’s Page

http://www.aoa.gov/AoARoot/AoA_Programs/H
PW/Alz_Grants/index.aspx, includes the following:

• Information on ADSSP

• Links to prior webinars on various dementia-related topics
NIA’s Alzheimer’s Disease Education and Referral (ADEAR) Center
1-800-438-4380
Mon-Fri, 8:30 am-5:00 pm Eastern Time
adear@nia.nih.gov
• Focus on research-based information
• Referral to government and organization resources
NIA-funded Alzheimer’s Disease Centers (ADCs):

- ADCs conduct research to improve diagnosis and care and test treatments
- Help with obtaining diagnosis and medical management
- Opportunities to participate in research

Alzheimer’s Disease Research Centers

The National Institute on Aging funds Alzheimer’s Disease Centers (ADCs) at major medical institutions across the U.S. Researchers at these Centers are working to translate research advances into improved diagnosis and care for Alzheimer’s disease (AD) patients while, at the same time, focusing on the program’s long-term goal—finding a way to cure and possibly prevent AD.

Areas of investigation range from the basic mechanisms of AD to managing the symptoms and helping families cope with the effects of the disease. Center staff conduct basic, clinical, and behavioral research and train scientists and health care providers who are new to AD research.

Although each center has its own unique area of emphasis, a common goal of the ADCs is to enhance research on AD by providing a network for sharing new ideas as well as research results. Collaborative studies draw upon the expertise of scientists from many different disciplines.

For patients and families affected by AD, the ADCs offer:

- Diagnostic and medical management (costs may vary—centers may accept Medicare, Medicaid, and private insurance).
- Information about the disease, services, and resources.
- Opportunities for volunteers to participate in drug trials, support groups, clinical research projects, and other special programs for volunteers and their families.

Some ADCs have satellite facilities which offer diagnostic and treatment services and research opportunities in underserved, rural, and minority communities.

National NIA-funded AD resources are listed at the end of the directory.

For more information, contact any of the centers in the directory below. The directory is also available in PDF format (2.2MB).

Find an Alzheimer’s Disease Center (ADC): Click on a city name in the map below or browse the list of ADCs by state.

http://www.nia.nih.gov/alzheimers/alzheimers-disease-research-centers#adcs
NIA ADEAR Search for Alzheimer’s Clinical Trials

Find trials by:

- Location
- Eligibility criteria
- Drugs tested
- Featured trials
- ADEAR phone support

1-800-438-4380

Sign up to receive e-alerts, RSS when new trials posted/updated
Research Resources

Web Address:  
http://www.alz.org/trialmatch  
Phone:  1.800.272.3900
Questions?

Registration for Webinars 3 & 4 Now Open at: https://aoa-events.webex.com

Slides, audio and transcript for 2013 webinar series will be available under Resources and Useful Links at: http://www.aoa.gov/AoARoot/AoA_Programs/HPW/Alz_Grants/index.aspx