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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ntg
National Task Group
on Intellectual Disabilities
and Dementia Practices

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

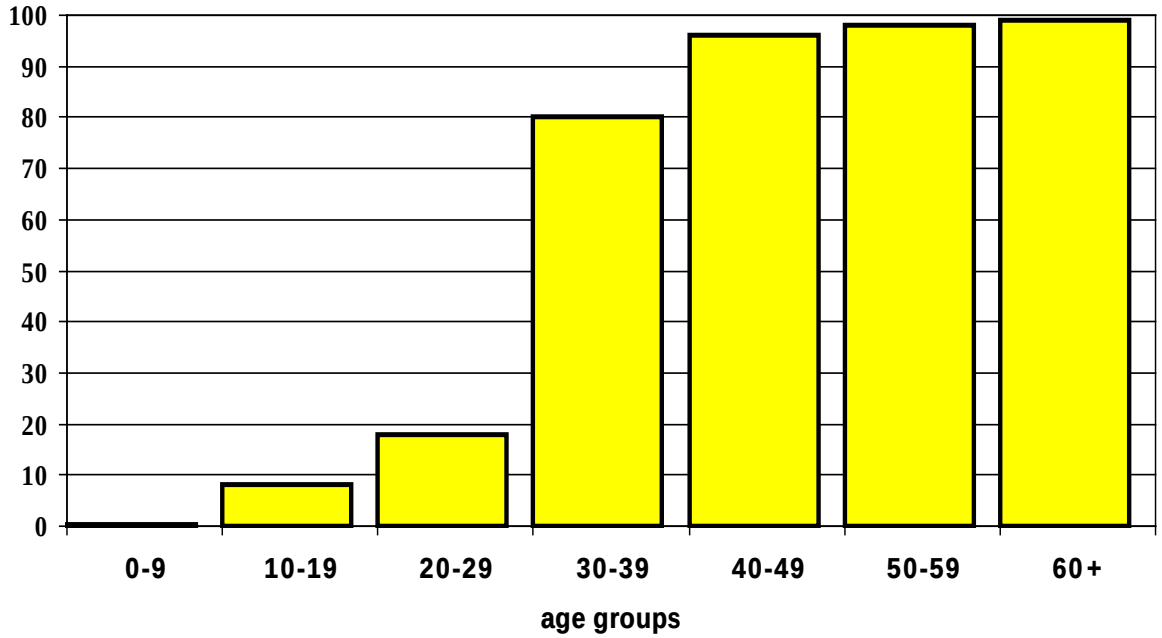
Unknowns…

- 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:	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.

Percent persons with Down syndrome showing evidence of neurofibrillary tangles (NFT) and senile plaques (SP) at autopsy



Source: Mann (1993) – [based on 39 published studies n=434]

Issues that arise with respect to dementia and Down syndrome

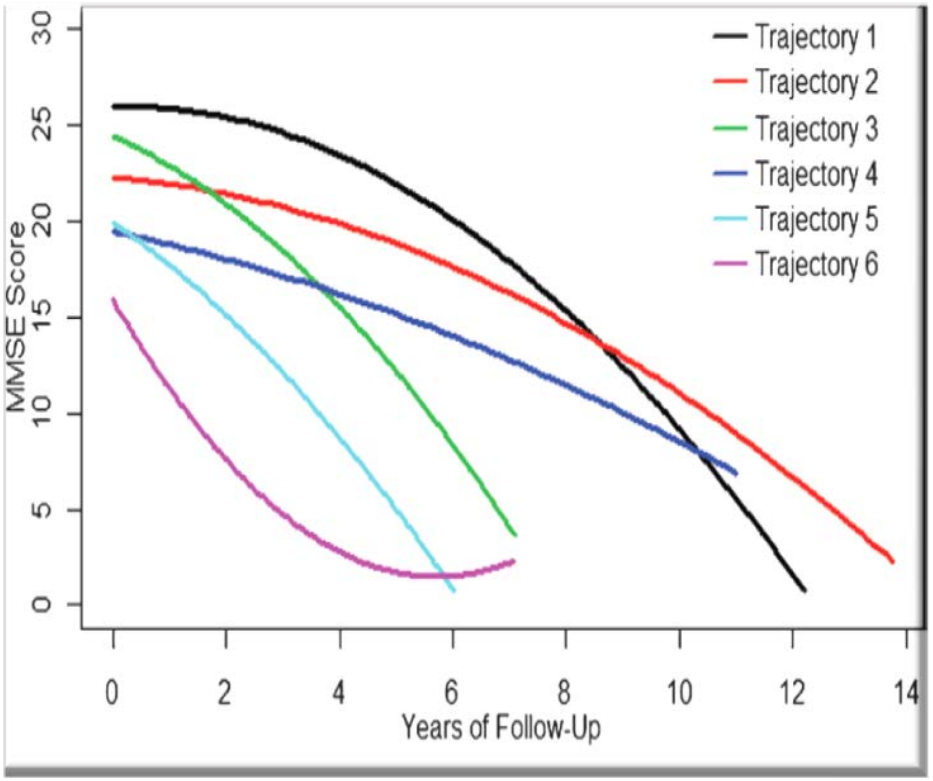
- ✓ Much higher prevalence of neuropathology indicative of AD in most adults w/Down syndrome (DS)
- ✓ Generally dementia of the Alzheimer's type is prevalent in adults with DS
- ✓ Average onset age in early 50s for DS (late 60s for others)
- ✓ Most DAT diagnosed within 3 years of "onset" in adults w/DS
- ✓ 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

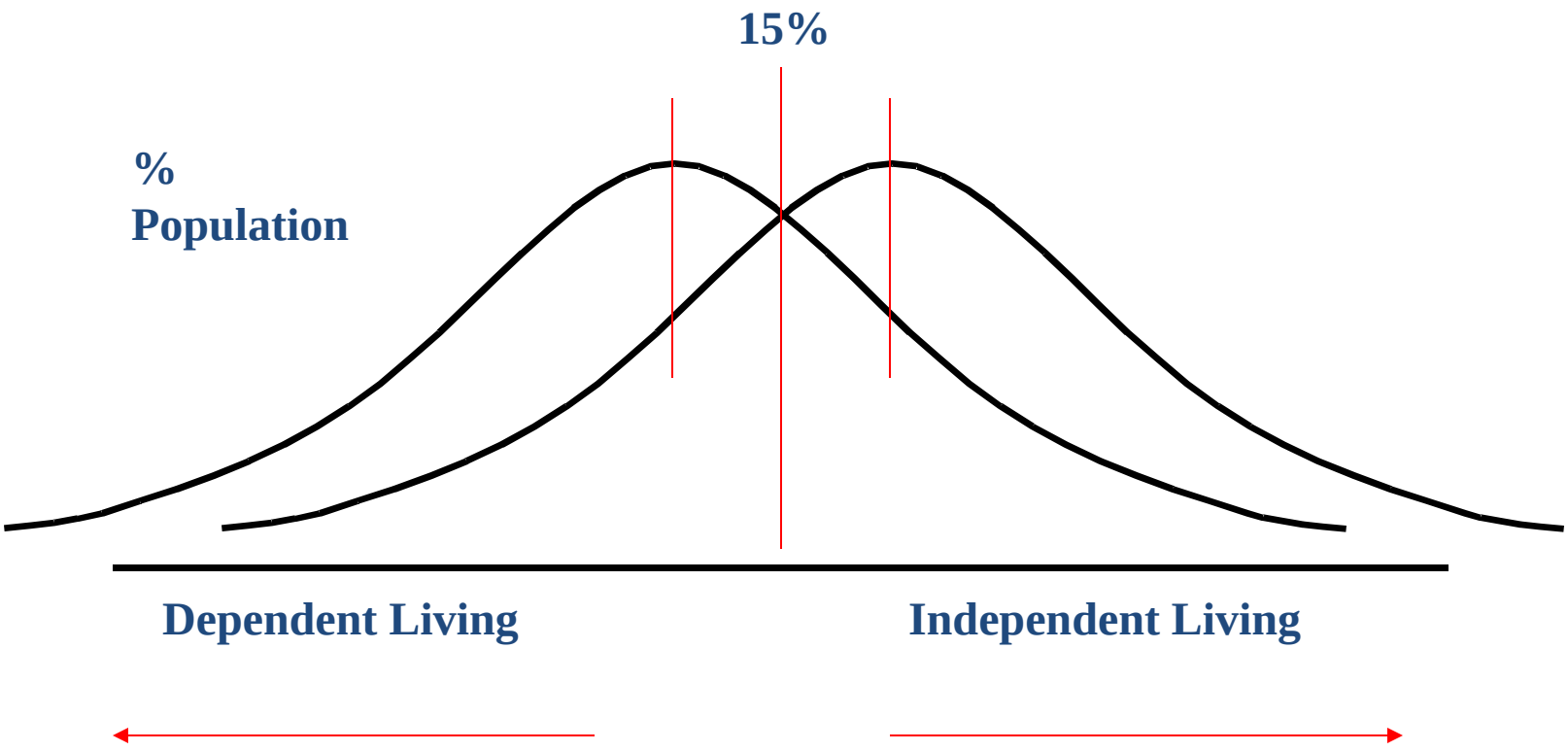
- Degree of retention of function
- Expected trajectory of progressive dysfunction
- Duration (remaining life years)
- Type of dementia
- Health status
- Environmental accommodations



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

Source: Figure 1 from Wilkosz et al., (2009). Trajectories of cognitive decline in Alzheimer's disease. *International Psychogeriatrics*, 28,1-10

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”

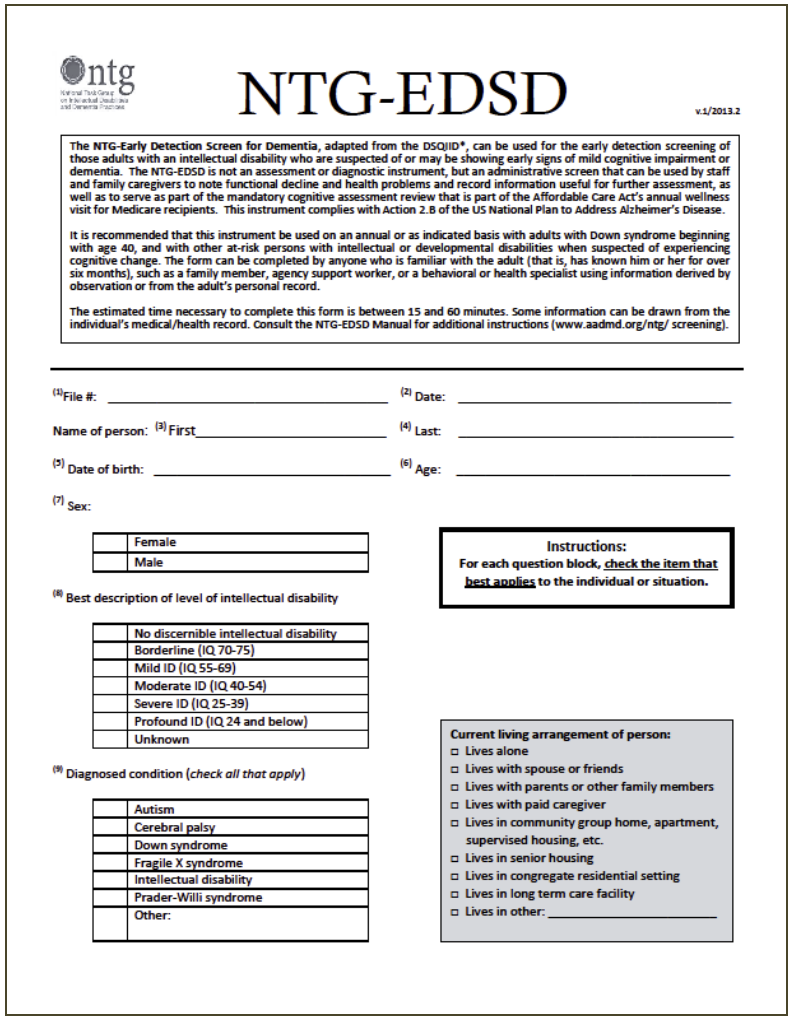
- Knowing that a change in function is a concern; when is a change in function part of normal aging or not?
- Diagnostic overshadowing
- Benefits of early diagnosis
- Documenting a change in function has occurred from a prior established baseline (NTG-EDSD)
- Diagnostic Assessment tools
- Change of care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



ntg

NTG-EDSD

v.1/2013.2

The NTG-Early Detection Screen for Dementia, adapted from the DSQIID*, can be used for the early detection screening of those adults with an intellectual disability who are suspected of or may be showing early signs of mild cognitive impairment or dementia. The NTG-EDSD is not an assessment or diagnostic instrument, but an administrative screen that can be used by staff and family caregivers to note functional decline and health problems and record information useful for further assessment, as well as to serve as part of the mandatory cognitive assessment review that is part of the Affordable Care Act's annual wellness visit for Medicare recipients. This instrument complies with Action 2.B of the US National Plan to Address Alzheimer's Disease.

It is recommended that this instrument be used on an annual or as indicated basis with adults with Down syndrome beginning with age 40, and with other at-risk persons with intellectual or developmental disabilities when suspected of experiencing cognitive change. The form can be completed by anyone who is familiar with the adult (that is, has known him or her for over six months), such as a family member, agency support worker, or a behavioral or health specialist using information derived by observation or from the adult's personal record.

The estimated time necessary to complete this form is between 15 and 60 minutes. Some information can be drawn from the individual's medical/health record. Consult the NTG-EDSD Manual for additional instructions (www.aadmd.org/ntg/ screening).

(1) File #: _____ (2) Date: _____

Name of person: (3) First_____ (4) Last: _____

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

(7) Sex:

	Female
	Male

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

(8) Best description of level of intellectual disability

	No discernible intellectual disability
	Borderline (IQ 70-75)
	Mild ID (IQ 55-69)
	Moderate ID (IQ 40-54)
	Severe ID (IQ 25-39)
	Profound ID (IQ 24 and below)
	Unknown

(9) Diagnosed condition (*check all that apply*)

	Autism
	Cerebral palsy
	Down syndrome
	Fragile X syndrome
	Intellectual disability
	Prader-Willi syndrome
	Other:

Current living arrangement of person:
- ☐ Lives alone
- ☐ Lives with spouse or friends
- ☐ Lives with parents or other family members
- ☐ Lives with paid caregiver
- ☐ Lives in community group home, apartment, supervised housing, etc.
- ☐ Lives in senior housing
- ☐ Lives in congregate residential setting
- ☐ Lives in long term care facility
- ☐ Lives in other: _____

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

Journal of Policy and Practice in Intellectual Disabilities

Volume 10, Issue 1, pages 1–24, March 2013

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Lawrence T. Force and the
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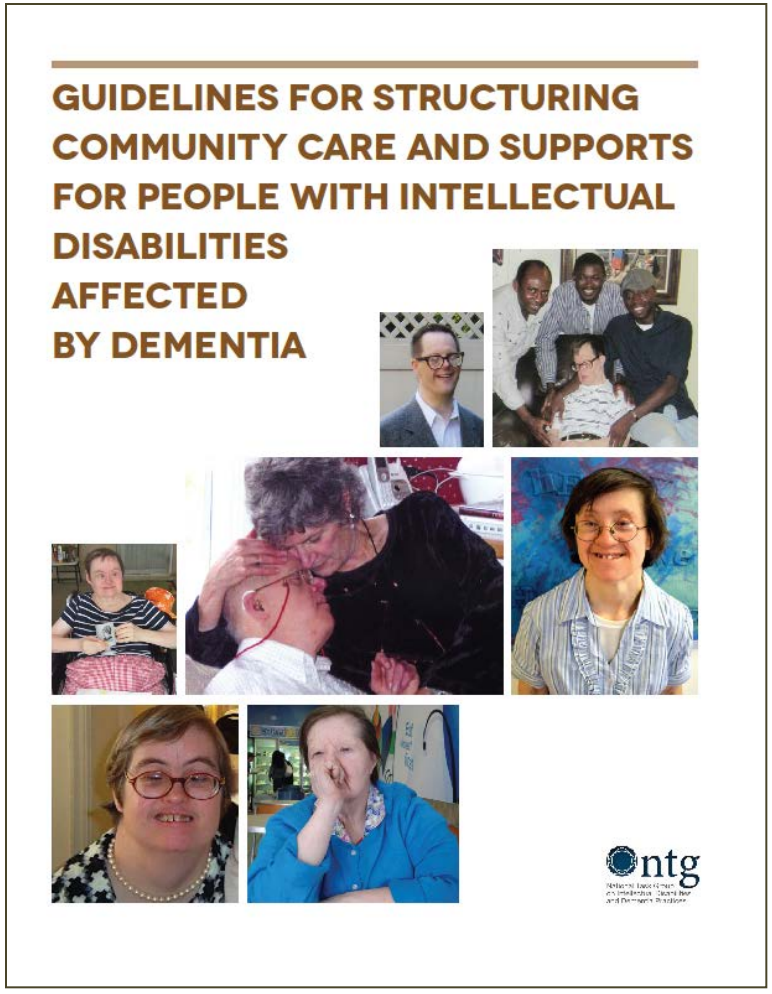


http://aadmd.org/NTG

Some **Key Features** of the Guidelines

Background	• NTG initiative, underlying principles, dementia as it affects people with ID
Staging Model	• Early recognition of symptoms → late stages • Expected changes in behaviour & function • Recommended actions
Early Detection	• Use of an early detection tool • Assessment & diagnosis period
Program / Support Options	• Critical concerns in varying circumstances • Non pharmacological approaches
Auxiliary Issues	• Abuse, financial, managing choice & liability, medications and nutrition
Collaboration	• Intellectual disability, aging, Alzheimer's, health • Policies, programs, services

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

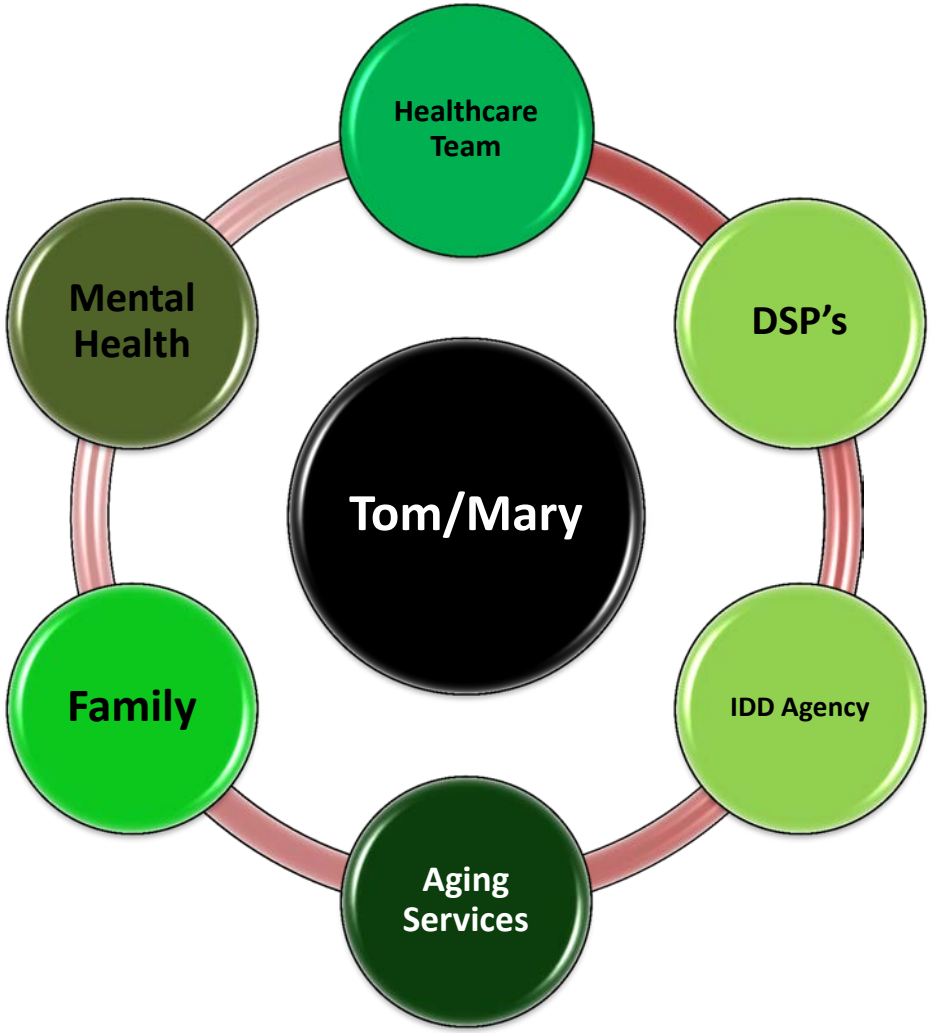
- Be empowered
- Form alliances and partnerships with health care team
- Appreciate aging parents' issues
- Understand and create support structure; aging and I/DD
- Determine expectations and goals

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
  - For publications, screening tools, and other resources on dementia and I/DD
- http://www.alz.org/dementia/down-syndrome-alzheimers-symptoms.asp
  - For information on Down syndrome and dementia
- 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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Visit us at – http://aadmd.org/ntg

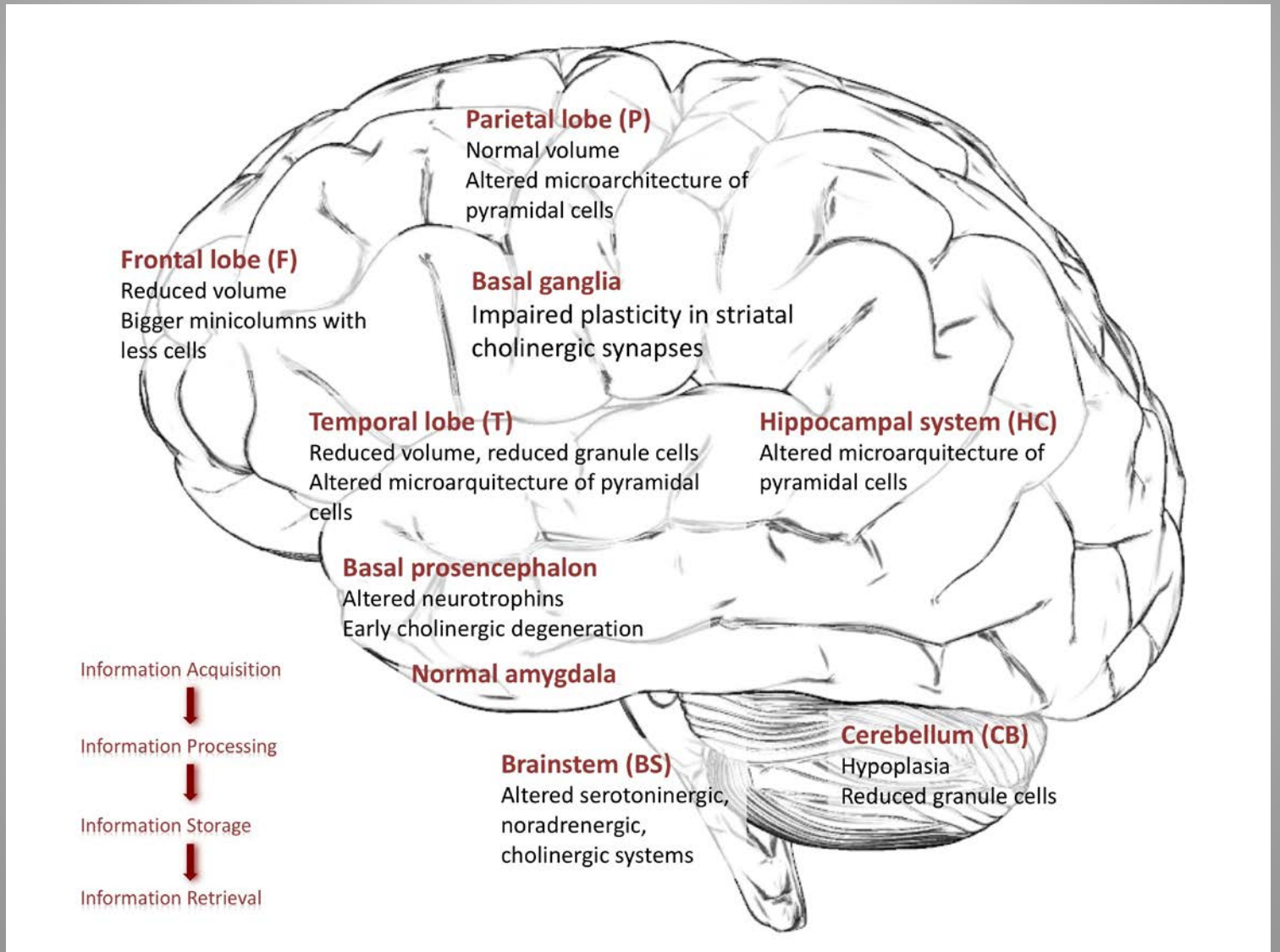
Overview of Research on Down Syndrome and Aging: Opportunities and Challenges

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

Outline

- Mouse models
- Cognitive functioning
- Clinical trials
- Telemedicine

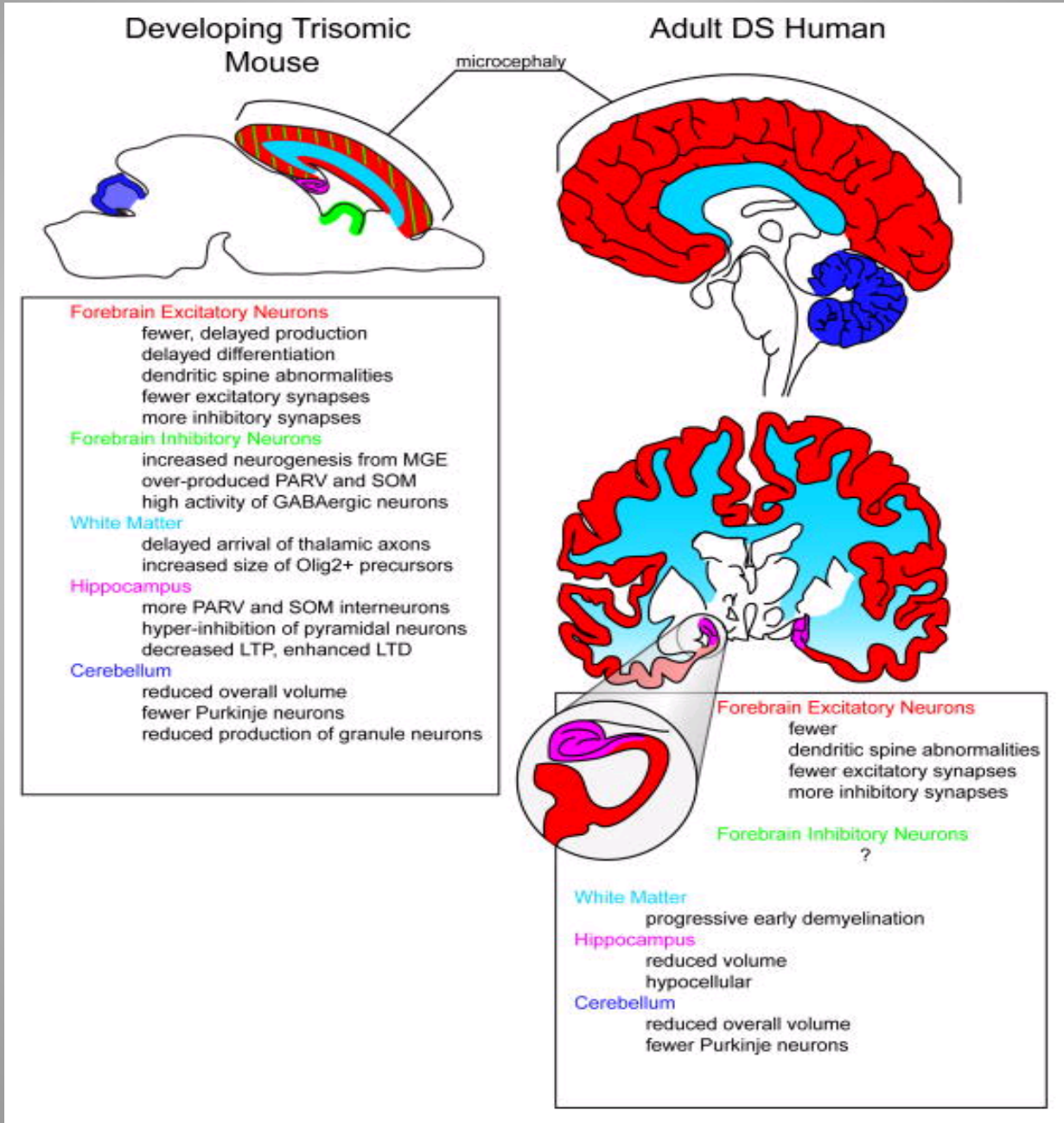
Neuropathological Topography (Lott and Diersessen, 2010)



Mouse Models for Down Syndrome

- Overexpression of same genes
- Can study individual genes
- Can experimentally manipulate model
- Offers a platform for translational research

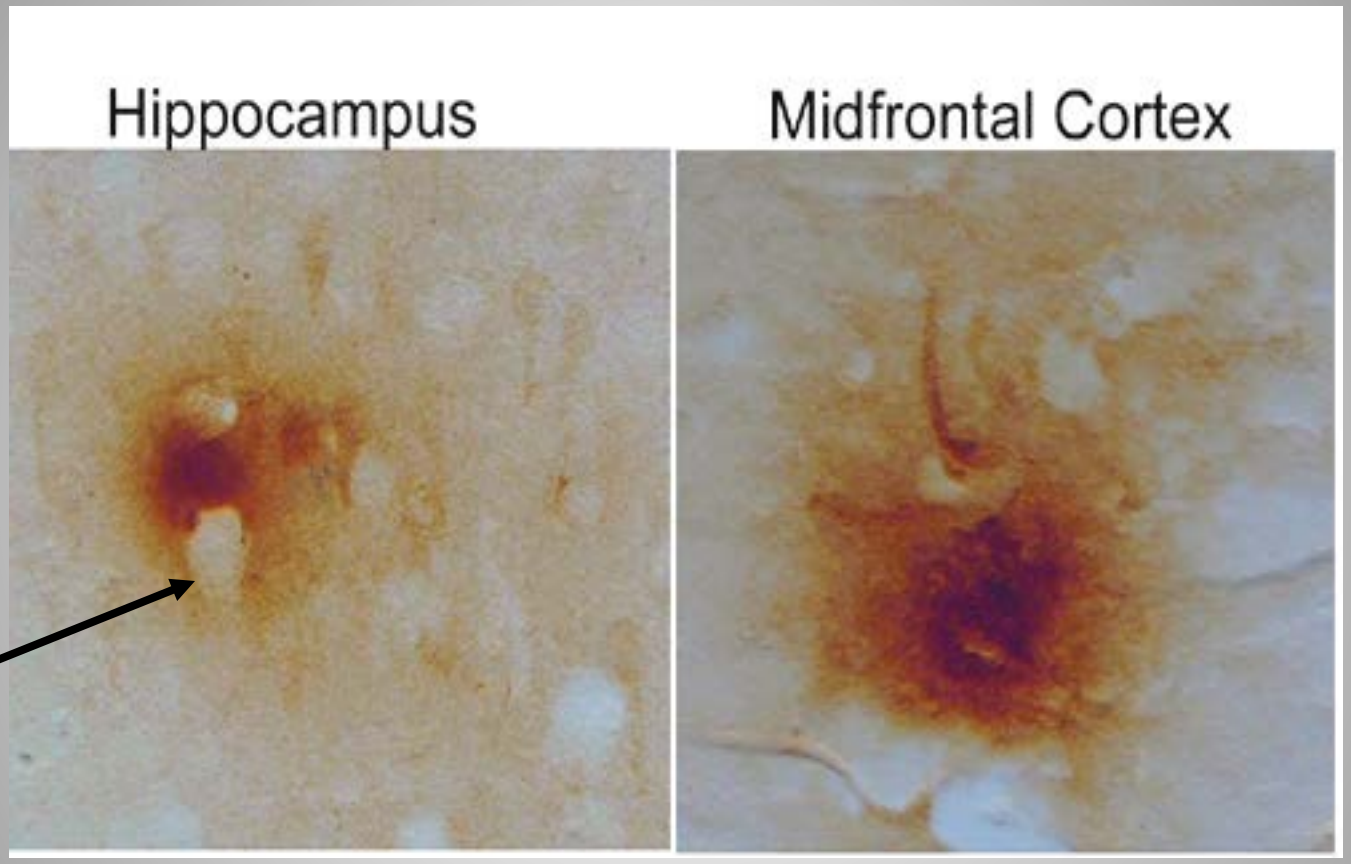
Haydar and Reeves, 2012



Intellectual disability in DS (Dierssen review, 2012)

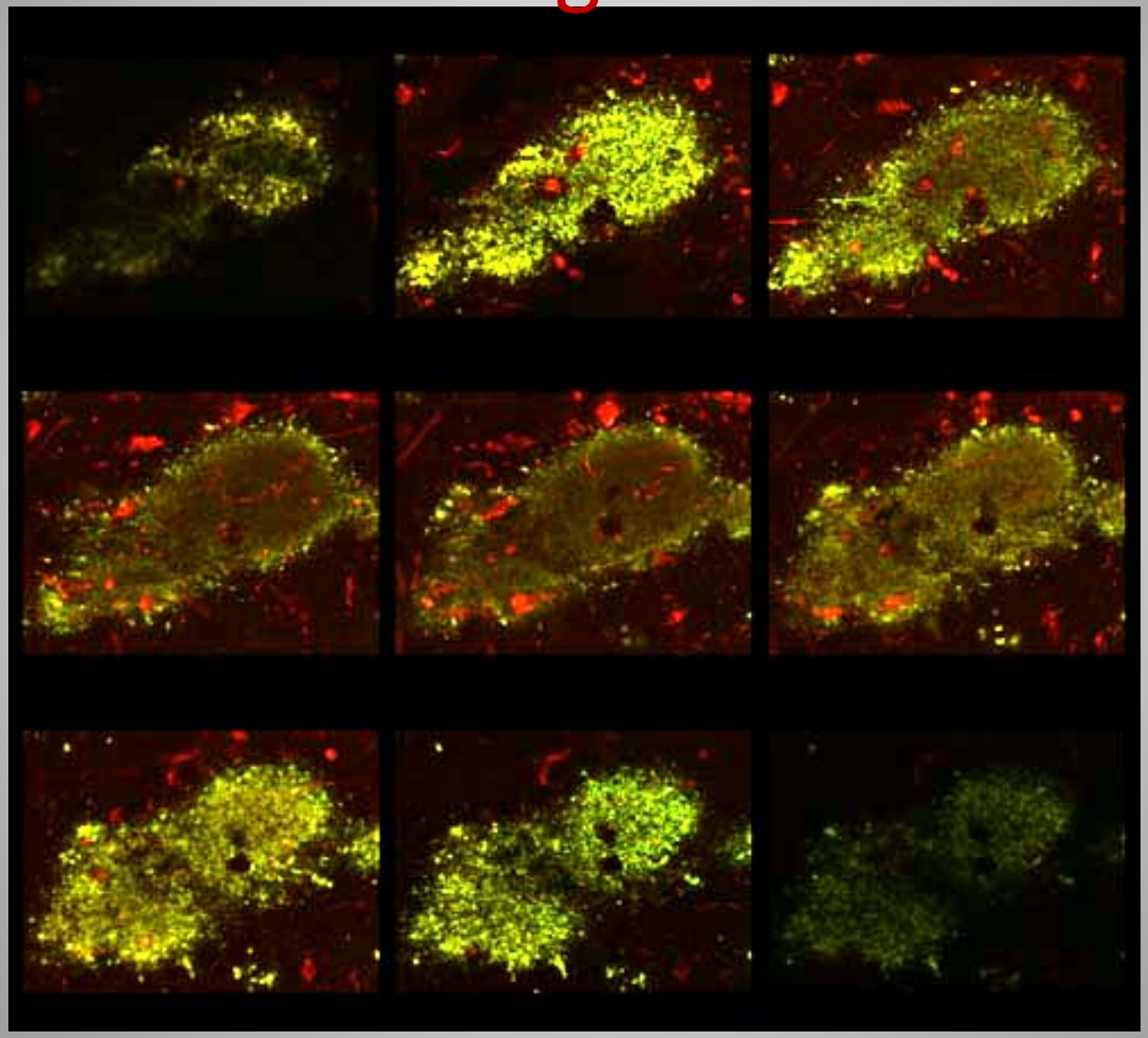
- Disruption in keeping incoming information on line, mental computation, and storage
- Uneven working memory
- Visual short term memory worse than visual-spatial memory
- May impair downfield cognitive performance in language, vocabulary and problem solving

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)

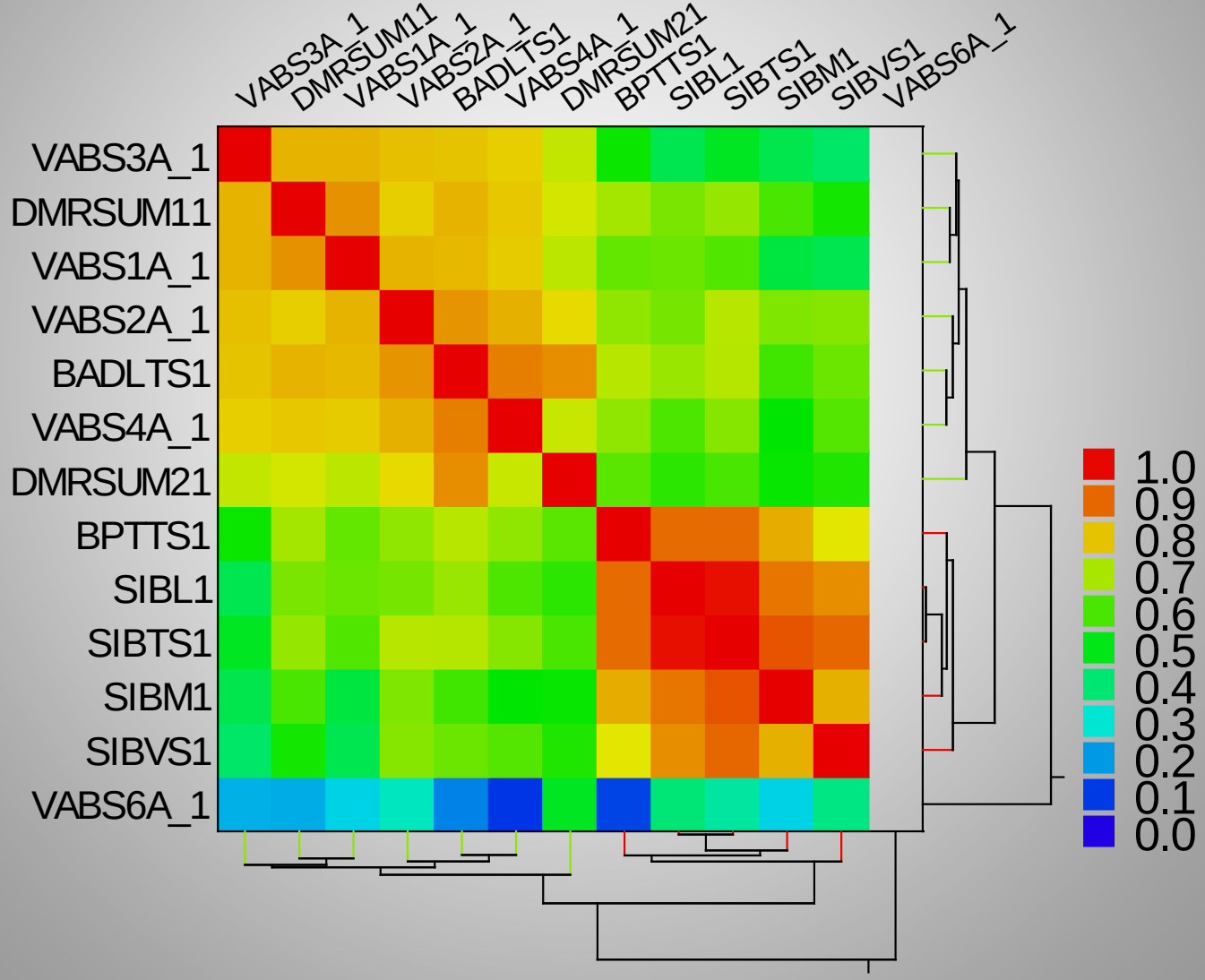
- CAMDEX informant measures show impaired frontal functioning in preclinical AD in DS

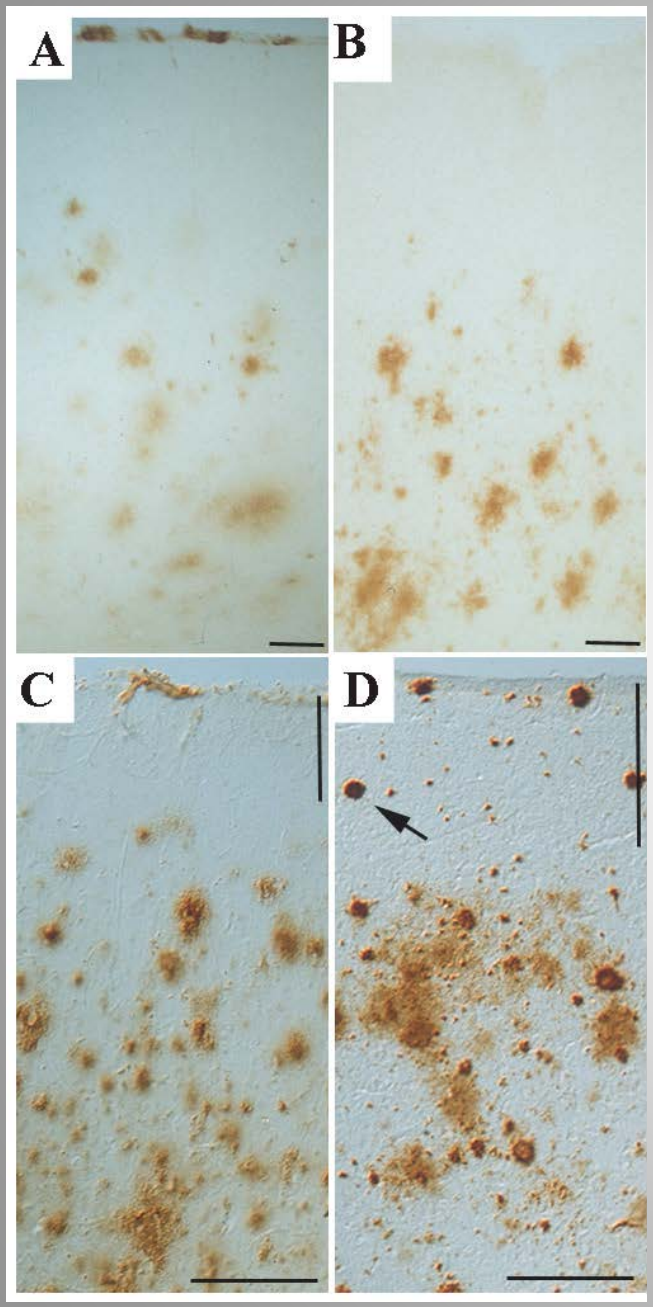
- Measures of cognition, receptive language, behavior and executive functioning implicate frontal lobes in early dementia in DS

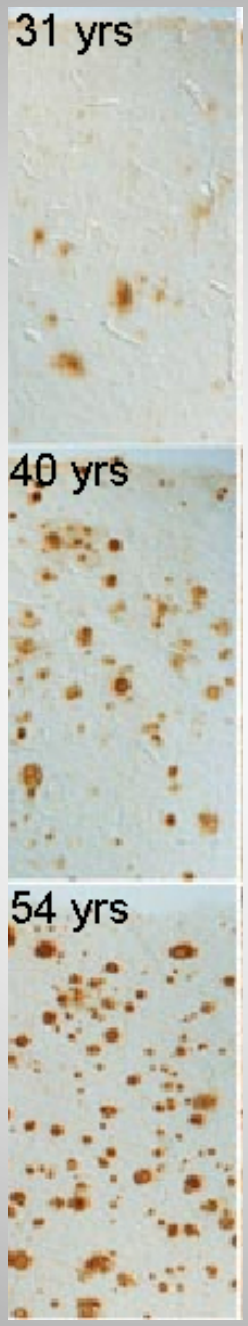
Examples

- 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







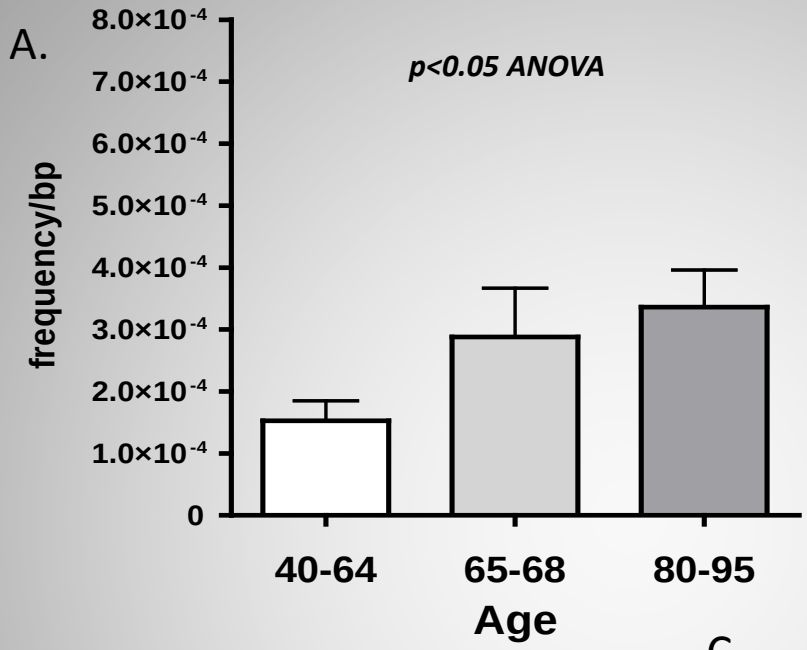
Beta-amyloid Deposition in Dogs: Comparison with Human Brain

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

Charts


A.
p<0.05 ANOVA
frequency/bp
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7.0×10-4
6.0×10-4
5.0×10-4
4.0×10-4
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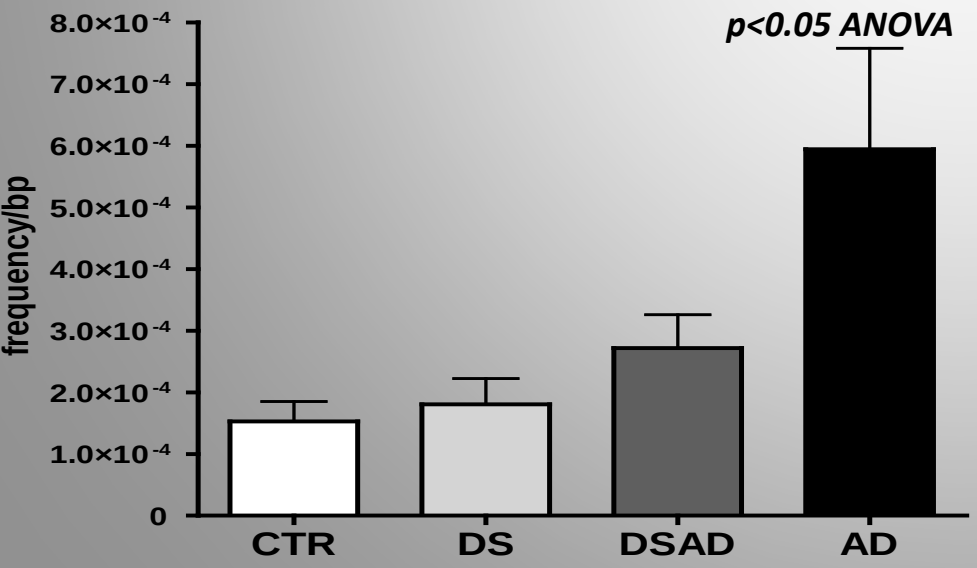
1.0×10-4
0
40-64
65-68
80-95
Age


B.
p<0.05 ANOVA
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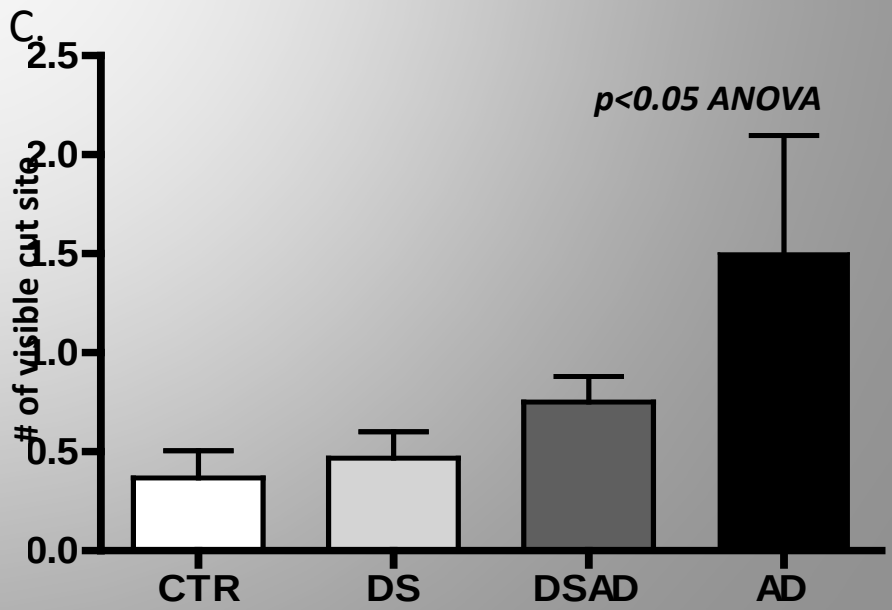
DS
DSAD
AD


C.
p<0.05 ANOVA
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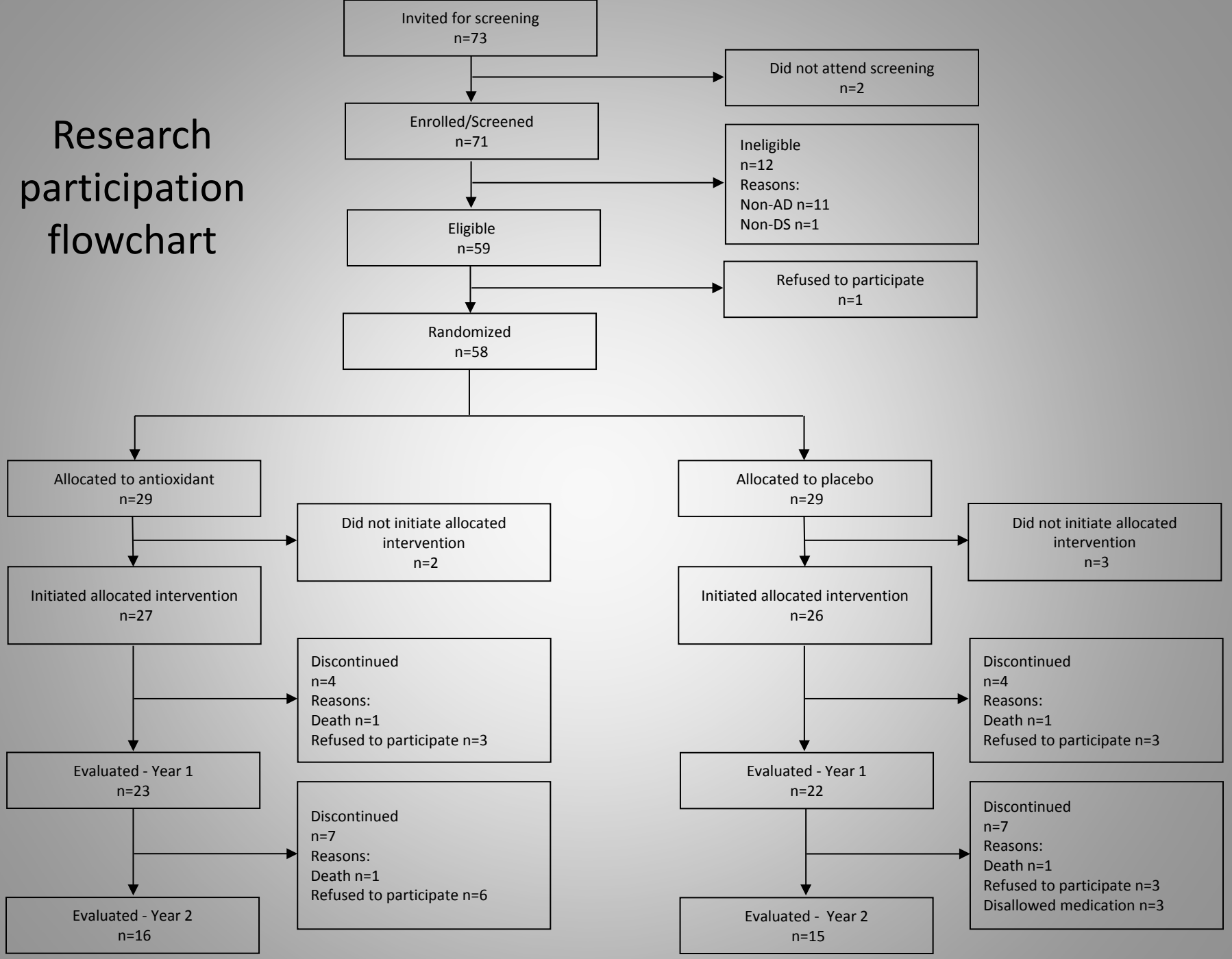
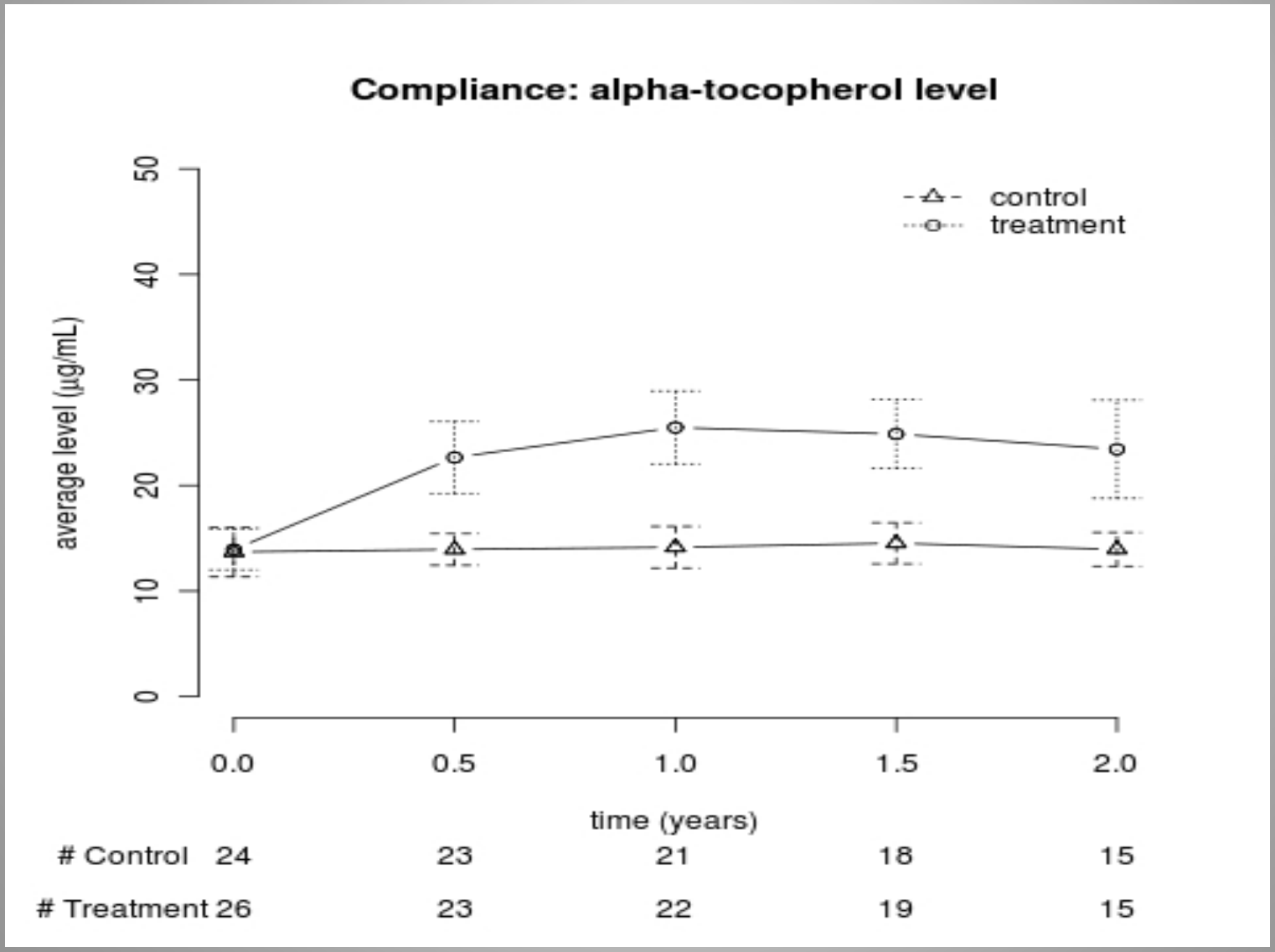
2.0
1.5
1.0
0.5
0.0
CTR
DS
DSAD
AD

Research participation flowchart



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

Telemedicine Screening for Dementia in DS

Subjects identified	358
Subjects screened	334
Subjects screened positive	97
Subjects declined to participate	24
Subjects examined	37
Subjects diagnosed with dementia	8
Subjects placed on medication therapy	8
Subjects diagnosed with a pseudodementia	4

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

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Administration on Intellectual and Developmental Disabilities
Administration for Community Living
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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

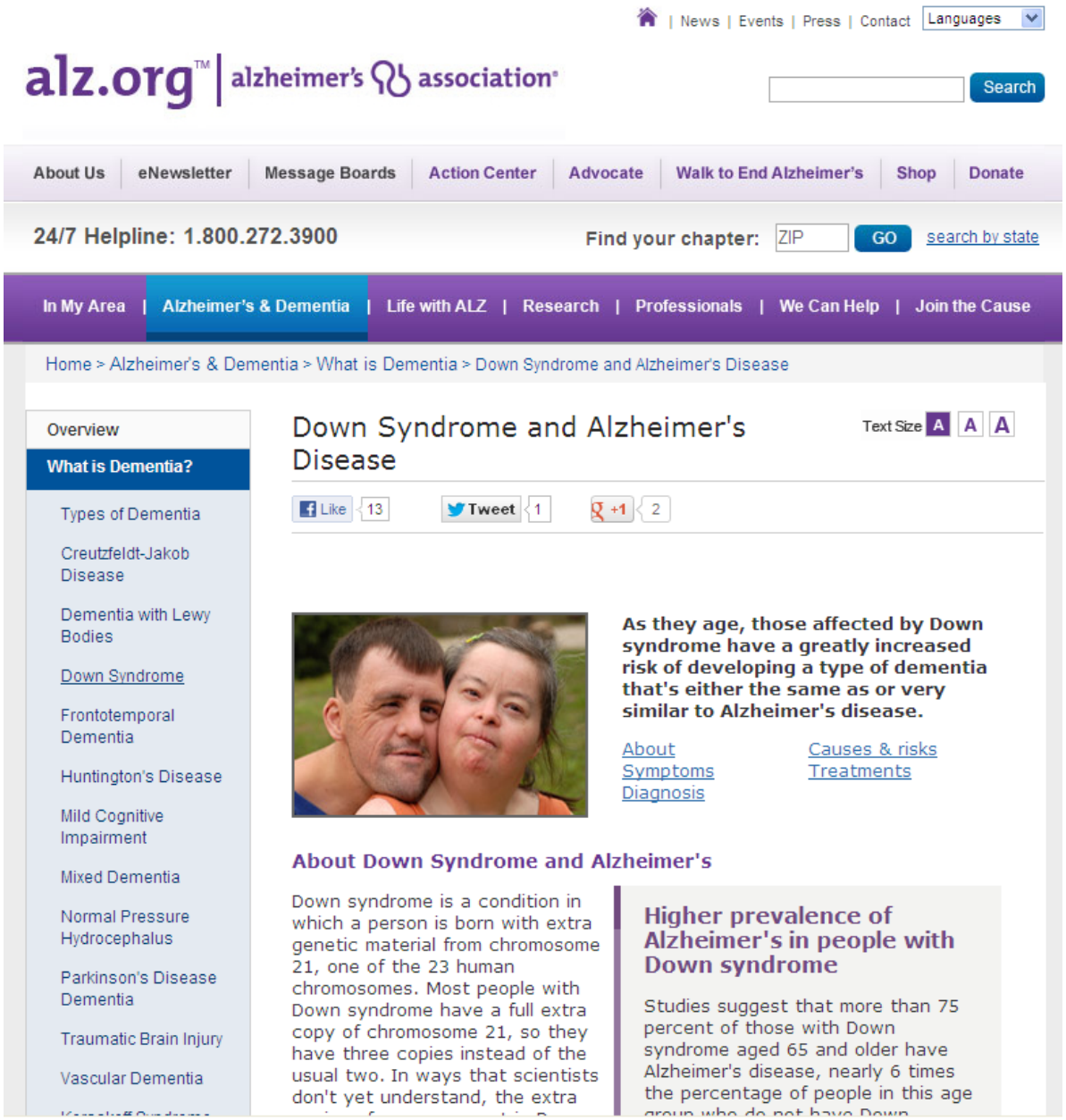
ACL and Resources

- 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.

http://alzheimers.gov/

http://alzheimers.gov/down_syndrome.html

ACL Alzheimer's Page

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

- Information on ADSSP
- Links to prior webinars on various dementia-related topics

http://www.nia.nih.gov/alzheimers

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

http://www.nia.nih.gov/alzheimers/alzheimers-disease-research-centers#adcs

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:

- Diagnosis 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 (253K).

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

News & Highlights

Publications

Research Centers

Clinical Trials

AD Library

Questions? Contact ADEAR

Call to talk to an Information Specialist:
1-800-438-4380 (toll-free)
Mon-Fri, 8:30 am-5:00 pm Eastern Time

or send an email to: adear@nia.nih.gov

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

http://www.nia.nih.gov/alzheimers/clinical-trials

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