Healthcare for Adults with I/DD and Dementia

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American Academy of Developmental Medicine and Dentistry
Co-Chair NTG
Health Care

- Health Care Advocacy for Family/Agency Communication for Adults with I/DD
- Health Care Assessment, Diagnosis, Treatment, Long term care
Tips for Health Care Advocacy

- Be aware of myths and stereotypes about aging in persons with I/DD
- Know the individual; who they are and how they’ve been focusing on specific ADL’s
- 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 parent and sibling issues
- Understand and create support structure; aging and I/DD
- Determine expectations and goals
Prepare for Appointments

- Gather pertinent information
  - Know past pertinent medical history
    - Always compare the person to who she or he has been throughout a lifetime, not to others of similar chronological age
  - Different shifts and perspectives of team/family
  - Symptoms as specific as possible, time of day, what behaviors, patterns, who is present
  - Prioritize symptoms
  - Share the information with advocate who will be going
The National Task Group on Intellectual Disabilities and Dementia Practices Consensus Recommendations for the Evaluation and Management of Dementia in Adults with Intellectual Disabilities


Lifespan Prevention

▪ Develop activities for healthy movement and weight bearing early in life
▪ Mentor good practices such as hydration, moderate diet, and exercise (You are the role model)
▪ Make the exercises fun and part of everyday life
▪ What you do will influence others around you

Rehabilitation Research and Training Center (RRTC) on Aging with Developmental Disabilities: Lifespan Health and Function, UIC at Chicago
http://www.rrtcadd.org/
“We can no longer overlook the fear and concern shared by aging Americans and their adult children. Additional work is needed to ensure that the nation is informed about cognition and its impact on families and communities, and updated over time as scientific discoveries emerge.”

Marilyn Albert, PhD
Johns Hopkins Alzheimer's Disease Research Center

“Unless public health engages our partners to act quickly and strategically, issues of aging—falls, mobility, cognitive health—will consume our healthcare system.”

Toni Miles, MD, PhD
Institute of Gerontology, University of Georgia

“We must capitalize on state efforts to coordinate public health chronic disease programs and include cognitive health in that larger picture.”

Sharon Moffatt, R.N, BSN, MSN
Association of State and Territorial Health Officials

The Healthy Brain Initiative
The Public Health Road Map for State and National Partnerships, 2013–2018
Caregiving

Care for people with dementia, such as Alzheimer’s disease, is often provided in the home by family members or friends. These caregivers typically have a longer and harder caregiving journey than caregivers for people with other conditions. While they provide a tremendous service to the person with dementia and to society, they are at greater risk for developing health problems.

Family Partners
The vast majority of caregivers for people with dementia, such as Alzheimer’s disease, are family members in home settings.

Risks of Caregiving
Caregivers of people with Alzheimer’s disease are at greater risk for anxiety, depression, and poorer quality of life compared to caregivers of people with other chronic conditions.
CDC’s Healthy Brain Research Network

▪ Outlines how state and local public health agencies and their partners can promote cognitive functioning, address cognitive impairment for individuals living in the community, and help meet the needs of care partners

▪ Identifies priority actions of the Road Map in the four traditional domains of public health:
  ▪ Monitor and evaluate
  ▪ Educate and empower the nation
  ▪ Develop policy and mobilize partnerships
  ▪ Ensure a competent workforce
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
Functional Decline

Cognitive
- Dementia
- Stroke
- Head Injury
- Seizures

Sensory
- Hearing Impairment
- Visual Impairment
- Peripheral Neuropathy
- Vestibular

Neuromotor
- Myelopathy
- Radiculopathy
- Nerve Comp
- Spasticity

Psychiatric
- Depression
- Psychotic Disorders
- Bipolar Dis
- SIB
- Anxiety

General Medical
- Cardiac
- Endocrine
- Musculoskeletal
- ADR
- Pulmonary
What Should be Ruled Out?

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

**TABLE 1. Common Medication Classes Associated With Possible Worsening of Cognitive Function in Patients With Dementia**

<table>
<thead>
<tr>
<th>Medication class</th>
<th>Examples</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Antihistamines, especially first generation</td>
<td>Diphenhydramine</td>
<td>Anticholinergic adverse effects, urine retention, confusion, sedation</td>
</tr>
<tr>
<td></td>
<td>Hydroxyzine</td>
<td></td>
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<tr>
<td></td>
<td>Promethazine</td>
<td></td>
</tr>
<tr>
<td>Bladder agents</td>
<td>Oxybutynin</td>
<td>Anticholinergic adverse effects, urine retention, confusion, sedation</td>
</tr>
<tr>
<td></td>
<td>Tolterodine</td>
<td></td>
</tr>
<tr>
<td>Certain pain medications</td>
<td>Meperidine</td>
<td>Meperidine: increased risk of seizures with renal impairment</td>
</tr>
<tr>
<td></td>
<td>Propoxyphene</td>
<td></td>
</tr>
<tr>
<td>Tricyclic antidepressants</td>
<td>Amitriptyline</td>
<td>Risks and benefits of this medication should be guided by a psychiatrist with familiarity with patients with I/DD</td>
</tr>
<tr>
<td></td>
<td>Clomipramine</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Doxepin</td>
<td></td>
</tr>
<tr>
<td>Certain antipsychotics</td>
<td>Chlorpromazine</td>
<td>Atypicals have been associated with increased mortality when used to treat behavioral problems in elderly patients with dementia, but no such studies have been conducted in Down syndrome or I/DD in general</td>
</tr>
<tr>
<td></td>
<td>Clozapine</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Pimozide</td>
<td></td>
</tr>
<tr>
<td>Long-acting benzodiazepines</td>
<td>Clonazepam</td>
<td>Very sedating; caution for gait impairment, dizziness</td>
</tr>
<tr>
<td></td>
<td>Temazepam</td>
<td>If a benzodiazepine is required for anxiety, consider short-acting agents (appropriately dosed): alprazolam, lorazepam</td>
</tr>
<tr>
<td></td>
<td>Diazepam</td>
<td></td>
</tr>
</tbody>
</table>

I/DD = intellectual and developmental disabilities.

Adults with Down Syndrome: Specialty Clinic Perspectives

Chicoine, B., McGuire, D., Rubin, S.

Diagnosed Disorders for 148 Adults Who Presented with a Decline in Function

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Frequency</th>
<th>Percent of Diagnosed Disorders (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mood</td>
<td>76</td>
<td>31</td>
</tr>
<tr>
<td>Anxiety</td>
<td>31</td>
<td>13</td>
</tr>
<tr>
<td>Obsessive-Compulsive</td>
<td>29</td>
<td>12</td>
</tr>
<tr>
<td>Behavior</td>
<td>23</td>
<td>9</td>
</tr>
<tr>
<td>Hypothyroid</td>
<td>22</td>
<td>9</td>
</tr>
<tr>
<td>Adjustment</td>
<td>12</td>
<td>5</td>
</tr>
<tr>
<td>Alzheimer's</td>
<td>11</td>
<td>4</td>
</tr>
<tr>
<td>B12 Deficiency</td>
<td>7</td>
<td>3</td>
</tr>
<tr>
<td>Menopause</td>
<td>7</td>
<td>3</td>
</tr>
<tr>
<td>Attention Deficit / Hyperactive</td>
<td>6</td>
<td>2</td>
</tr>
<tr>
<td>Gastrointestinal or Urinary</td>
<td>6</td>
<td>2</td>
</tr>
<tr>
<td>Sensory Impairment</td>
<td>6</td>
<td>2</td>
</tr>
<tr>
<td>Psychotic</td>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td>Other Medical Conditions*</td>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td>Cardiac Conditions</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td><strong>TOTAL</strong></td>
<td><strong>247</strong></td>
<td><strong>100</strong></td>
</tr>
</tbody>
</table>
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
Diagnosis of I/DD and Dementia

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

- Maintaining QOL
- Prolonging life
- Prevent functional decline
- Slow progression
- Decrease psychiatric/behavioral problems
- Fall reduction program
- Reduce hospitalization
- Watch for signs of abuse and neglect
- Cholinesterase Inhibition and Memantine
- Pharmacologic and behavioral interventions
- Palliative Care
- End of Life Care
Impact of Coexisting Medical Conditions

Cognitive Impairment

Prevalence of coexisting conditions in PWD

2.4 conditions/pt
HTN 82%
DM 39%
CAD 21%
CHF 14%
Stroke 10%

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
Behavioral and Psychological Symptoms of Dementia (BPSD)

- Symptoms of disturbed perception, thought content, mood or behaviour that frequently occur in patients with dementia

(Finkel & Burns, 1999)
Dementia-related behavior

Complex interaction between:
- Cognitive deterioration
- Physical health
- Emotional health/Psychiatric issues
- Life story, Personality Style
- Environmental issues (Physical and Social)
Dementia-Related Behaviors

- 90% of people with dementia will have at least one symptom
  - Depression—40%
  - Delusions—63%
  - Hallucinations—4-41%
  - Aggression—31-42%
  - Apathy
Other Symptoms of BPSD

- Sleep disturbance (day/night reversal)
- Hoarding
- Shadowing
- Disinhibition (stripping)
- Sexually inappropriate behaviour
- Sundowning
- Wandering
Implications of Dementia-related Behavior

- Associated with worse prognosis
- More rapid cognitive decline
- Increased caregiver burden
- Leads to earlier admission to institutional care
- Increased healthcare cost
Common Triggers

- **Physical**
  - Acute illness/infection, medications, pain, poor vision, hearing, poor sleep

- **Cognitive**
  - Inability to understand, express oneself, lack of insight, misinterpretation of environment, difficult to problem solve

- **Emotional**
  - Fear, anxiety, depression, frustration, apathy, boredom

- **Environmental**
  - Changes in caregiver, confrontational approach, tasks that exceed abilities, change in routine, over/understimulation, lack of visual cues
Nonpharmacological Strategies

Also Remember the four S’s

▪ Maximize safety and Limit the Risks
▪ Promote Structure and Consistency
▪ Enhance serenity and Limit Confusing Stimuli
▪ Nuture Sanity and Supports for All Caregivers
Do not:

▪ Argue – it will make the situation worse
▪ Tell the person what they can’t do – tell them what they can do
▪ Talk down to the person as if they are a young child
▪ Ask a lot of questions
▪ Talk about a person with dementia as if they are not present even if you think they cannot understand you
BPSD Typically Not Amenable To Pharmacologic Management

- Undressing
- Repetitive Activity
- Vocally disruptive behavior
- Eating inedibles
- Tugging at restraints

- Pushing wheelchair bound co-residents
- Hiding & hoarding
- Inappropriate Voiding
- Poor Social Skills
- Wandering

C. Ward Hillside Centre IHA
When should we consider pharmacologic treatment of BPSD?

- Behavior is dangerous, distressing, disturbing, damaging to social relationships and persistent

AND

- Has not responded to comprehensive non-pharmacologic treatment plan. Including removal of possibly offending drugs

OR

- Requires emergency treatment to allow proper investigation of underlying problems

OR

- Clear medication-responsive psychiatric comorbidity identifiable

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Questions to be Answered in Evaluating Medication Use

- What is the target problem being treated?
- Is the drug necessary?
- Are nonpharmacologic therapies available?
- Is this the lowest practical dose?
- Does this drug have adverse effects that are more likely to occur in an older patient?
- By what criteria, and at what time, will the effects of therapy be assessed?
- Who are we treating the individual or us??

(Avorn et al. 1995)
Medications Specifically for Behavioral Psychological and Symptoms in Dementia (BPSD)

• Clear indication, potential benefits and risks
  • FDA Black Box Warning for Antipsychotics in usage in patients with dementia. Studies have shown an increased rate of mortality secondary to vascular complications including strokes and cardiac events\(^1\)

• Identify target symptoms

• Expected time to response

• Risks associated with and without Rx

• Appropriate dose range

• Monitoring for side effects and response

• When to consider dose reduction, discontinuation.

Medications Specifically for Alzheimer’s Symptoms: Behavioral Psychological and Symptoms in Dementia (BPSD)

<table>
<thead>
<tr>
<th>Target Symptoms</th>
<th>Medication</th>
</tr>
</thead>
<tbody>
<tr>
<td>Delusions</td>
<td>Atypical Antipsychotics:</td>
</tr>
<tr>
<td>Hallucination</td>
<td>• risperidone</td>
</tr>
<tr>
<td>Aggression</td>
<td>• olanzapine</td>
</tr>
<tr>
<td>“Agitation”</td>
<td>• quetiapine</td>
</tr>
<tr>
<td>Sadness</td>
<td>Antidepressants</td>
</tr>
<tr>
<td>Irritability</td>
<td>• citalopram</td>
</tr>
<tr>
<td>Anxiety</td>
<td>• sertraline</td>
</tr>
<tr>
<td>Insomnia</td>
<td>• venlafaxine</td>
</tr>
<tr>
<td></td>
<td>• mirtazapine</td>
</tr>
<tr>
<td></td>
<td>• trazodone</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Target symptoms</th>
<th>Medication</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mood swings</td>
<td>Mood stabilizers:</td>
</tr>
<tr>
<td>Euphoria</td>
<td>• valproic acid</td>
</tr>
<tr>
<td>Impulsivity</td>
<td>• carbamazepine</td>
</tr>
<tr>
<td>Agitation</td>
<td>Cholinesterase Inhibitors.</td>
</tr>
<tr>
<td>Apathy</td>
<td>Memantine</td>
</tr>
<tr>
<td>Irritability</td>
<td></td>
</tr>
<tr>
<td>Anxiety (short term</td>
<td>Anxiolytics:</td>
</tr>
<tr>
<td>use in predictable</td>
<td>• lorazepam</td>
</tr>
<tr>
<td>situations)</td>
<td>• oxazepam</td>
</tr>
</tbody>
</table>

Safety and Antipsychotics

- Over-sedation
- Postural Hypotension
- Impaired cognition
- Falls
- Weight gain
- Hyperglycemia
- QTc prolongation
- Extra-pyramidal symptoms (EPS)
- Tardive Dyskinesia
- Cerebrovascular events
- Mortality
Cognitive Enhancers

- **Cholinesterase Inhibitors; Aricept, Exelon, Razadyne**
  - *Lott IT et al.* Arch Neurol. 2002;59:1133-1136

- **NMDA (N-methyl-D-aspartate) receptor antagonist; Namenda**

- **Herbal Supplements/Vitamins**
  - Ginkgo Biloba

- **Research**
  - Anticholinergics
  - Nicotine
  - Homocysteine
  - Huperzine A
  - NSAIDS
  - Beta Amyloid antagonists
  - Vaccination trials
## Response to dementia medication in DS

### Donepezil in Adults with Down Syndrome With and Without Dementia

<table>
<thead>
<tr>
<th>Author</th>
<th>Journal</th>
<th>Year</th>
<th>No subjects</th>
<th>Study Type</th>
<th>Results</th>
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</thead>
<tbody>
<tr>
<td>Kishnani, P.S., et al.</td>
<td>Lancet</td>
<td>1999</td>
<td>4</td>
<td>Case Reports</td>
<td>Improvement</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>No dementia</td>
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<tr>
<td>Heller, J. H. et al</td>
<td>AJ Medical Genetics</td>
<td>2003</td>
<td>6</td>
<td>Case Reports</td>
<td>Improvement</td>
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<tr>
<td></td>
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<td></td>
<td></td>
<td>No dementia</td>
<td>Language</td>
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<tr>
<td>Johnson, N. et al</td>
<td>AJMR</td>
<td>2003</td>
<td>19</td>
<td>RCT</td>
<td>Improvement</td>
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<tr>
<td></td>
<td></td>
<td></td>
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<td>No dementia</td>
<td>Language</td>
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<tr>
<td>Prasher, V.P., et al.,</td>
<td>Intl J Ger Psych.</td>
<td>2002</td>
<td>27</td>
<td>RCT</td>
<td>Non significant improvement</td>
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<td></td>
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<td></td>
<td></td>
<td>Alzheimer’s Disease</td>
<td></td>
</tr>
<tr>
<td>Lott, I.T., et al.,</td>
<td>Archives Neurology</td>
<td>2002</td>
<td>15</td>
<td>Case Control</td>
<td>Significant</td>
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<td>Improvement</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Alzheimer’s Disease</td>
<td>improvement</td>
</tr>
<tr>
<td>Kondoh, T. et al</td>
<td>Annals Pharmacotherapy</td>
<td>2005</td>
<td>2</td>
<td>Case Reports</td>
<td>Improvement</td>
</tr>
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<td></td>
<td></td>
<td></td>
<td></td>
<td>Alzheimer’s Disease</td>
<td></td>
</tr>
</tbody>
</table>

- NMDA (N-methyl-D-aspartate) receptor antagonist; Memantine DB, PC 88 received drug/85 Placebo
  No Improvement

Cognitive Enhancers and BPSD

- Some evidence that they decrease some symptoms of BPSD
- Improvement seen in hallucinations, apathy, anxiety, and aberrant motor behavior
- Improvement may take weeks to months to occur
- Question of whether they may prevent BPSD from emerging
- Limited value in acute situation
- Consider discontinuation with emergence of agitation and decline in function.
Targets for Future Therapies

- Aβ
  - β-secretase inhibitors
  - γ-secretase inhibitors
  - Monoclonal antibodies
- Tau protein
- Inflammation
- Insulin resistance
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
Serial Assessment of Change

- Serial Assessment of Function in Dementia
- 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
Assessing Cognitive Decline

- Helps determine therapeutic effect
- 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
Progression of Disease; Anticipatory Guidance

- Cognitive Skills will decline
- Support needs will increase
- Increase risks of falls, injuries
- Swallowing dysfunction, clots, pneumonia, bladder infections
- Seizures
- Watch for signs of abuse and neglect
- Watch for signs of caregiver burn out
- End of life decisions
Palliative and End of Life Care

▪ The realization that Alzheimer’s disease progresses with increasing risks of health complications impacting ones QOL/ADL’s

▪ Respecting ones wishes for level of care and quality of life

▪ Defining, anticipating, and preparing for end of life

▪ Supporting families, friends, and supports
Clinical Supports

- Usage of trained professionals, i.e. physicians, nurses, psychologists, social workers, and others trained in Dementia assessment and intervention
- Can aid in developing programs or care management plans
- Help to both staff and individuals with dementia including in-service training
Team Approach to Care

- Improving outcomes
- Respect for need and opinions of team members
- Who is the team??
Multidisciplinary Care

Healthcare Team

Mental Health

Family

Direct Support/ DD Agency

Aging Agencies
The Future of Alzheimer’s Disease

▪ Earlier recognition
  ▪ Dependent on reliable biomarkers

▪ New medications
  ▪ Current medications only address symptoms
  ▪ New medications in development
    ▪ Disease-modifying therapy
    ▪ Combination disease-modifying and symptomatic therapy

▪ Prevention

▪ Improved support network