Nurse Practitioner Education in Developmental Disabilities Webinar Series

Neurologic Complications in Adults with I/DD

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Outline

- Epilepsy
- Movement Disorders
- Gait Dysfunction
- Spasticity
- Dementia
Epilepsy in I/DD
STATISTICS

- 3% of the population has I/DD
- 10-20% of all I/DD individuals have epilepsy
- 50% of individuals with I/DD & CP have epilepsy (More common in tetra or hemiplegic than dystonic or diplegic)
- 21% of I/DD with IQ > 50 have epilepsy
- 50% of I/DD with IQ < 50 have epilepsy
- 40% of individuals living in large residential facilities have epilepsy

Difficulties with Seizure Care

- Refractory epileptic syndromes
- Multiple seizure types
- Frequent status epilepticus and clusters
- Lifelong AED use
- Polypharmacy (epilepsy and medical)
- Side-effects are frequent but hard to detect
- Side-effect tolerance; status quo
- Co-occurrence with challenging behaviors including Autism
- Challenges in obtaining data and communication
- Transitioning of care
- Staff knowledge/training
- Acute seizure care
Issues in Developing Optimal Plans for Seizure Care

- All paroxysmal events are not seizures
- All seizures are not dangerous
- Not all seizures are refractory
- Multiple drugs are usually not necessary
- Side-effects are very important
- Seizures may not be lifelong
- Data collection and communication
  - Seizuretracker.com
  - Expectations from patient/families/staff/providers
Differential Diagnosis of Seizures

- Syncope
- Behavior
- Toxicity
- Pseudoseizures
- Panic attacks
- Hypoglycemia
- Vertigo
# Epilepsy Treatments

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Age</th>
<th>Indication</th>
<th>Efficacy</th>
<th>Side Effects</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>AEDs</strong></td>
<td>Children</td>
<td>Specific AEDs for specific seizure types</td>
<td>64% sz freedom&lt;sup&gt;1&lt;/sup&gt;</td>
<td>Vary by AED, typically CNS- and endocrine-related</td>
</tr>
<tr>
<td></td>
<td>Adults</td>
<td>All seizure types</td>
<td>54% pts &gt;50% sz reduction at 3 months&lt;sup&gt;2&lt;/sup&gt;</td>
<td>Lipid disorders, ketoacidosis</td>
</tr>
<tr>
<td><strong>Ketogenic Diet</strong></td>
<td>Primarily children</td>
<td>All seizure types</td>
<td>70% in select patients sz freedom&lt;sup&gt;3&lt;/sup&gt;</td>
<td>Cognitive effects, surgery-related risks</td>
</tr>
<tr>
<td><strong>Epilepsy Surgery</strong></td>
<td>Children</td>
<td>Pharmacoresistant or localization-related epilepsy</td>
<td>43% of pts &gt;50% sz reduction at 3 years&lt;sup&gt;4&lt;/sup&gt;</td>
<td>Voice alteration, cough, pharyngitis, dyspnea</td>
</tr>
<tr>
<td></td>
<td>Adults</td>
<td>Pharmacoresistant epilepsy, partial seizures</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>VNS Therapy</strong></td>
<td>12 and older</td>
<td>Pharmacoresistant epilepsy, partial seizures</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Antiepileptic Drug Options

Partial
- Simple
- Complex
- Secondarily Generalized

Generalized
- Tonic
- Tonic-clonic
- Myoclonic
- Atonic
- Infantile Spasms
- Absence

Pregabalin, Phenytoin, Carbamazepine, Phenobarbital, Gabapentin, Tiagabine, Oxcarbazepine, Lacosamide

ACTH
- Vigabatrin
- Topiramate
- Zonisimide

Ethosuximide

Valproate, Lamotrigine, Topiramate, Zonisimide
- Levetiracetam, Felbamate, Rufinamide, Clobazam
Treatment/Evaluation Sequence for Pharmacoresistant Epilepsy

1st Monotherapy AED Trial

2nd Monotherapy AED Trial

3rd Monotherapy/Polytherapy AED Trial

Strongly consider videoEEG Monitoring

Psychogenic, migraine, syncope, sleep disorders, movement disorder’s, etc.

Epilepsy Surgery/VNS Therapy/Neuropace Evaluation

Polytherapy AED Trials
Resective Surgery
Stimulator Therapy

Seizure control at what cost?

- Toxicity
- Cognitive
- Physiologic
- Behavioral
- Financial considerations

**Psychiatric adverse events during levetiracetam therapy**
M. Mula, MR. Trimble, et al
Neurology. 2003 Sep 9;61(5):704-6

**Topiramate and Psychiatric Adverse Events in Patients with Epilepsy**
M. Mula, MR. Trimble, et al
Behavioral Outcomes with Eliminating Sedating Agents

- Improved alertness and interaction
- Improved maladaptive behavior
- Reduced psychotropic medication usage

Coulter DL. *AM J Ment Retard.* 1988;93:320-327
Acute Seizure Care

1. Cushion head, remove glasses.
2. Loosen tight clothing.
3. Turn on side and keep airway clear.
4. Note the time a seizure starts and the length of time it lasts.
5. Don’t put anything in mouth.
6. Don’t hold down.
7. As seizure ends… offer help.

- Recognition of the event and appreciate when significant
  - Clusters, Status Epilepticus, Seizure types
- First aid
- Usage of VNS magnet
- Diastat acudial
- 9-1-1
- First responder care
- ED and hospital care
- Document details of the event
Sudden Unexpected Death in Epilepsy (SUDEP)

May be the cause of death when:
- A healthy person with epilepsy dies suddenly without drowning or trauma
- The person may or may not have had a seizure before death
- No other reason for death is found upon exam after death
  - Person was not using illegal drugs (example: cocaine)
  - Person did not have a heart attack

Risk of SUDEP increases when:
- Seizures are not well controlled (treatment resistant epilepsy)
- Treatment resistant epilepsy = failure of 2 rounds of appropriate and tolerated seizure medication
  - Treatment resistant epilepsy is common in patients with autism
- A patient suffers from generalized tonic-clonic seizures
- Seizures happen at night when the person is sleeping

Some common theories causing SUDEP include:
- Heart arrhythmias
- Breathing trouble
- Brain shutdown

1 out of 1,000 patients with epilepsy die unexpectedly each year

In those with uncontrolled epilepsy, risk increases to 1 out of every 150 people
Movement Disorders in I/DD
Movement Disorders Classification

- Hyperkinetic vs Akinetic
- By type of movement; dyskinesia, myoclonic, tremor, dystonia, chorea
- Age of onset
- Acquired vs genetic
- Behavior vs organic
Extrapyramidal Effects

- Tardive Dyskinesia
- Akathisias
- Parkinson’s
- Dystonic Rx

0.5%-56% TD in long term usage of Neuroleptics

Management

- Stop offending agent
- Switch agent
- Reduce dosage
- Add Benztropine, Diphenhydramine
- L-Dopa Therapy
Gait Dysfunction and Spasticity in I/DD
Normal vs Pathologic Changes of Gait in Adults with IDD

- What is baseline and why did past dysfunction occur?
- What normal aging changes are expected?
- How to discern pathologic changes
- What are the risks and complications of altered gait and spasticity?
Abnormal Gait and IDD

- Pain
- Impaired Joint Mobility (arthritis, contractures)
- Muscle weakness (Spina bifida, low tone)
- Spasticity (stroke, cord lesion, Cerebral Palsy)
- Sensory/balance deficit (neuropathy, stroke, vision, vestibular)
- Impaired central processing (dementia, stroke, delirium, drugs)
- Cognitive Impairment
- Syndrome specific (Down syndrome, FASD, Fragile X)
Consequences of Gait Dysfunction

- Falls
  - Injury, fracture, CHI, hospitalizations
- Pain
- Osteoporosis
- Risks to skin integrity, cardiopulmonary system
- DVT’s/PE’s
- ADL’s
- QOL/Independence
- Impact upon care team
### Upper Motor Neuron Syndrome

A group of symptoms that may be caused by damage or injury to motor neuron pathways or brain regions that control movement\(^2\).\(^3\)

<table>
<thead>
<tr>
<th>Characterization</th>
<th>Positive Symptoms(^4)</th>
<th>Negative Symptoms(^4)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Examples</td>
<td>Muscle overactivity</td>
<td>Muscle underactivity</td>
</tr>
<tr>
<td></td>
<td>Spasticity, clonus, flexor/extensor spasm, hyperreflexia, dystonia, and rigidity</td>
<td>Decreased dexterity, weakness, paralysis, fatigability, and slowness of movement</td>
</tr>
</tbody>
</table>


Treatment Goals

Major Classes of Treatment Goals with Examples of Each \(^1\), \(^2\)

**Technical Objectives**
- Increase range of motion
- Reduce tone
- Reduce spasm

**Functional Objectives**
- Improve activities of daily living (e.g., dressing, hygiene)
- Reduce pain
- Enhance ease of care
- Improve limb position
- Improve gait

**Preventive Objectives**
- Prevent contracture
- Prevent skin maceration
- Prevent skin ulcers

\(^1\) Gormley ME, Jr., O'Brien CF, Yablon SA. A clinical overview of treatment decisions in the management of spasticity. *Muscle Nerve Suppl* 1997; 6:S14-20

Traditional Step-Ladder Approach to Management of Spasticity

- Neurosurgical procedures
- Orthopedic procedures
- Neurolysis
- Oral medications
- Rehabilitation Therapy
- Remove noxious stimuli
Spasticity Treatment Team

- Rehabilitative Therapy
  - Physiatry
  - Physical Therapy
  - Occupational Therapy
- Neurologist
- Primary Care Provider
- Nursing
- Direct Care Staff
- Orthopaedic Surgeon
- Neurosurgeon
- Anesthesiologist
- Family
Dementia in I/DD
Functional Decline

- A process in which a person is unable to perform at the same level of activity as previously performed
  - Cognitive
  - Physical
- What is normative aging and what is pathologic?
- Functional decline has an impact upon ones ADL's, QOL, and needs for supports
Domains of Cognition and Dementia

- Memory
  - Short and long term
- Attention
- Executive function
- Language
- Visuospacial
- Praxis

- Progressive decline in cognition and function with evolution of symptoms over time
Classification of Dementias

**Potentially Reversible**
- Drug Toxicity
- Metabolic Disturbance
- Normal Pressure Hydrocephalus
- Mass Lesion (Tumor, Chronic Subdural)
- Infectious Process (Meningitis, Syphilis)
- Collagen-Vascular Disease (SLE, Sarcoid)
- Endocrine Disorder (Thyroid, Parathyroid)
- Nutritional Disease (B12, thiamine, folate)
- Mood dysfunction
- Sleep dysfunction

**Irreversible**
- Alzheimer’s Disease
- Frontotemporal Dementia
- Parkinson’s Dementia
- Lewy body Disease
- Primary Progressive Aphasia
- Huntington's Chorea
- Kufs Disease
- Multi-infarct Dementia
- Jacob-Cruzefeldt Disease
- Head injuries
- HIV Dementia
- Multiple Sclerosis
Alzheimer’s Disease in Down Syndrome

- Women with Down’s syndrome are more at risk of developing Alzheimer’s disease than men in the 40 to 65 age group.
- People with Down’s syndrome who develop Alzheimer’s disease live, on average, 9-10 years from first symptoms.
- Infrequently rapid decline can occur.
- Late on-set seizures.
- From diagnosis to death is on average 8.2 years.

Percentage of people with Down syndrome who develop dementia at different ages:

<table>
<thead>
<tr>
<th>Age</th>
<th>Percentage</th>
</tr>
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<tbody>
<tr>
<td>30’s</td>
<td>2%</td>
</tr>
<tr>
<td>40’s</td>
<td>10-15%</td>
</tr>
<tr>
<td>50’s</td>
<td>33%</td>
</tr>
<tr>
<td>60’s</td>
<td>50-70%</td>
</tr>
</tbody>
</table>

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

Plaque of Amyloid Beta-Protein. Visible as a black globular mass when stained. The plaque is surrounded by abnormal neurites and degenerating neurons.
Natural history of Alzheimer’s Disease

Updated model integrating Alzheimer's disease immunohistology and biomarkers. The threshold for biomarker detection of pathophysiological changes is denoted by the black horizontal line.

## Adults with Down Syndrome: Specialty Clinic Perspectives

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

<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
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
- Available in multiple languages

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
The NTG’s recommended nine-step approach for assessing health and function.

- Taking thorough history, with particular attention to "red flags" that potentially indicate premature dementia such as history of cerebrovascular disease or head injury, sleep disorders, or vitamin B12 deficiency
- Documenting a historical baseline of function from family members of caregivers
- Comparing current functional level with baseline
- Noting dysfunctions that are common with age and also with possible emerging dementia
- Reviewing medications and noting those that could impair cognition
- Obtaining family history, with particular attention to a history of dementia in first-degree relative
- Noting other destabilizing influences in patient's life such as leaving family, death of a loved one, or constant turnover of caregivers, which could trigger mood disorders
- Reviewing the level of patient safety gleaned from social history, living environment, and outside support
- Continually "cross-referencing the information with the criteria for a dementia diagnosis"
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, neglect, and caregiver burnout
- Cholinesterase Inhibition and Memantine
- Pharmacologic and behavioral interventions
- Palliative Care
- End of Life Care
Behavioral and Psychological Symptoms of Dementia (BPSD)

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

- 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 Approaches

- Familiar environment—avoid frequent moves
- Soft lighting
- Calm colors
- Places to walk
- Access to outdoor spaces
- Home-like environment
- Low stimuli—minimize background noise
- Time out space

- Individualized Care Planning
- Careful analysis of care interactions
- Meaningful activity
- Music Therapy
- Exercise
- Snoezelen
  (multisensory stimulation program)
- Aromatherapy
- Yoga
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 that they cannot understand you
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 events¹

- 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>
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<tr>
<td></td>
<td>• trazodone</td>
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<thead>
<tr>
<th>Target Symptoms</th>
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</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 use in predictable situations)</td>
<td>Anxiolytics:</td>
</tr>
<tr>
<td></td>
<td>• lorazepam</td>
</tr>
<tr>
<td></td>
<td>• oxazepam</td>
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</tbody>
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Antipsychotics

- Class of meds used to treat psychosis and other mental or emotional conditions; delusions, hallucinations, agitation, paranoia
- Block release of dopamine in the brain
- Typical (conventional) or atypical
- Typical are not selective and also block receptors in other areas of the brain which may produce unwanted side effects
- Atypical cause fewer acute or chronic extra-pyramidal symptoms (EPS)
- Atypical antipsychotics result in improvement in mood and cognition compared to typical antipsychotics
Side effects of antipsychotics

- Parkinsonism
- Dystonia - abnormal face and body movements
- Akathisia (restlessness)
- Tardive dyskinesia (long term)
- Exacerbated by drug holiday regime
- More common in females
- Worsened in response to reducing drug
- Irreversible (denervation supersensitivity)

Many undesirable side effects (e.g., constipation, metabolic syndrome, lactation, and retrograde ejaculation)
Cognitive Enhancers

- Cholinesterase Inhibitors; Aricept, Exelon, Razadyne

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

- Herbal Supplements/Vitamins
  - Ginkgo Biloba

- Research
  - Anticholinergics
  - Nicotine
  - Homocysteine
  - Huperzine A
  - NSAIDS
  - Beta Amyloid and Tau protein antagonists
  - Vaccination trials
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