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Management of Less Common Neurological Disorders

By, Amber L. Ward

Background:

• I work with many “orphan”, rare and uncommon disorders
• Use this webinar later as a reference- I will move fast
• Consider treatment for progressive vs. non-progressive neurological disorders
Learning Objectives:

• Recall 3 characteristics of each uncommon diagnosis presented.
• Identify 2 precautions for treatment of clients with each disorder.
• Plan 3 treatments and 3 goals for a client with a progressive or non-progressive neurological disorder.

Rare diseases covered:

• Multiple (Multi) System Atrophy (MSA)
• Progressive Supranuclear Palsy (PSP)
• Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)
• Multifocal Motor Neuropathy (MMN)
• Spastic Paraparesis
• Dermatomyositis/Polymyositis
• Inclusion Body Myositis (IBM)
• Post-Polio Syndrome
Multiple (Multi) System Atrophy (MSA)

• Degeneration of nerve cells, lifespan average about 8 years
• Often called Parkinson’s Plus
• Onset 50’s-60’s
• Problems with:
  – Movement (slowness/rigidity)
  – Balance
  – Poor coordination/ataxia
  – Cognitive changes in about 20%
  – Autonomic functions (bladder/bowel issues and control, hypotension, temperature regulation, swallowing, anhidrosis)
  – Speech, swallowing

• No known cause, high levels of protein alpha-synuclein seen in nerve cells (basal ganglia, cerebellum, brain stem)

MSA- OT issues

• Precautions
  – Fall risk (balance, hypotension)
  – Heatstroke from anhidrosis/temperature regulation
  – Safety- cognition, urinary urgency, balance, communication, swallowing

• Treatments
  – Managing ADL/IADLs with ataxia, balance/coordination challenges
  – Strategies to manage bradykinesia, rigidity, lack of initiation
  – Fine motor adaptations- writing, buttons
  – Turning in bed, bed mobility

• Goals
  – Home safety (physical and cognitive)
  – Balance/coordination during occupations
  – Performance with AE or adaptive techniques of all occupations
  – Transfers, bed mobility (AE, techniques)
Progressive Supranuclear Palsy (PSP)

- Abnormal Tau protein in nerve cells, some genetic, most sporadic
- Onset generally in 60’s, dependent for care within 3-4 years, may live 5-9 years after onset with good care, death generally aspiration pneumonia

- Problems include:
  - Balance, falls - backward mostly
  - Lurching/unsteady gait
  - Bradykinesia, clumsiness, stiffness
  - Vision - difficulty looking downward, diplopia, blurred
  - Speech, swallowing changes
  - Cognitive, personality/mood, behavior changes

<table>
<thead>
<tr>
<th>Gait &amp; station</th>
<th>PSP</th>
<th>Parkinson’s Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Speech</td>
<td>Early falls</td>
<td>Normal gait initially</td>
</tr>
<tr>
<td>Face</td>
<td>Startled</td>
<td>Masked</td>
</tr>
<tr>
<td>Tremor</td>
<td>&lt;10%</td>
<td>&gt;90%</td>
</tr>
<tr>
<td>Rigidity</td>
<td>Axial</td>
<td>Limb</td>
</tr>
<tr>
<td>Symmetry</td>
<td>Symmetric</td>
<td>Asymmetric (unilateral) onset</td>
</tr>
<tr>
<td>Response to L-DOPA</td>
<td>Little</td>
<td>Excellent</td>
</tr>
</tbody>
</table>

PSP- OT issues

- Precautions
  - Balance, falls/tripping (both from balance and inability to look down)
  - Safety - cognition, balance
  - Swallowing

- Treatments
  - Safety strategies, move obstacles, avoid stairs
  - ROM - stiffness
  - Vision - prism glasses, compensation, patching
  - Balance management with ADL/IADL tasks

- Goals
  - Home safety - meds, trip hazards, falls, etc.
  - Safe swallowing, snack prep, self-feeding
  - ADL performance with stiffness/clumsiness, strategies
  - Pt/family education on ROM, safety, cognition, ADL strategies
  - Visual strategies/compensation
Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

- Immune disorder of peripheral nervous system (abnormal immune response, attacks myelin sheath)- related to Guillain-Barre Syndrome- chronic version
- No particular risk factors, no genetic links known
- Problems include:
  - Fatigue, muscle aches/pain
  - Loss of reflexes, autonomic system dysfunction
  - Progressive weakness, sensory changes (numbness, tingling, burning) in arms, legs
- May have relapses with full recovery or partial/no recovery
- Treatment may include corticosteroids, immunosuppressant drugs, plasma exchange, and/or intravenous immunoglobulin (IVIG)- 15% respond to no treatment

CIDP- OT issues

- Precautions
  - Over heating/pushing too hard can cause relapse
  - May be not used to sensation loss/weakness for certain tasks- safety
  - Safety with sensory- hot, cold, sharps
- Treatments
  - Gaining strength and endurance if possible
  - ADL/IADL performance, adaptations, safety, AE, compensation as needed
  - ADL transfers, bed mobility
- Goals
  - Pt/family education- sensory/safety issues
  - Increase strength (as possible)
  - Increase ADL performance (rehabilitation or compensation)
Multifocal Motor Neuropathy (MMN)

- Abnormal immune response - dramatic improvement with IVIG
- Problems include:
  - Progressive, asymmetric muscle weakness, distally primarily
  - Hand weakness or wrist drop usually first symptom
  - Lack of atrophy/reflex changes in proportion to weakness
  - Cramping, fasciculations

MMN - OT issues

- Precautions
  - Walking - tripping, fall risk
  - Hand use - may drop items
  - Fluctuating abilities depending on IVIG cycle
- Treatments
  - AE, adaptive techniques for ankle/hand weakness with occupations
  - Home safety - falls, balance, during functional tasks
  - Ways to avoid cramps during functional tasks - alter task performance
  - Strengthening (potentially, with IVIG improvement)
- Goals
  - Mod I with ADLs/IADLs
  - Pt/family education on home safety
  - Mod I, education with AE use (writing, buttons, etc.)
  - Increase in strength, education on home ex program (as possible)
Spastic Paraparesis/Hereditary Spastic Paraplegia (HSP)

- Genetic disorder - age of onset, progression and severity vary widely - 70 different forms
- Average age of onset is 24
- Slowly progressive degeneration of upper motor neurons in brain, spinal cord
- Problems include:
  - Spasticity/weakness of hip, leg muscles
  - Balance issues
  - Clonus, muscle spasms
  - Bladder hyperactivity or frequency

HSP- OT issues

- Precautions
  - Balance/falls
  - Decreased safety with mobility
- Treatments
  - Home safety/management
  - ADL/IADL adaptations with spasticity, hip/leg weakness
  - AE use, training
  - Potentially strengthening/maintaining ROM/strength
- Goals
  - Mod I with ADL/IADL
  - Maintain strength/ROM (immobility from spasticity weakness/stiffness)
  - AE training, use
  - Pt/family education on safety, falls, mobility, AE, ROM, ex program
Polymyositis
• Connective tissue disease of the muscles, proximal more than distal
• Potentially autoimmune, inflammatory myopathy
• Problems include:
  – Progressive, symmetrical proximal muscle weakness
  – Dysphasia
  – Muscle tenderness, aches
  – Fatigue
• Most common in 40’s to 50’s
• Corticosteroids, IVIG

Dermatomyositis
• Possible autoimmune disorder, inflammatory process
• Polymyositis plus skin rash
• Tx: Corticosteroids, IVIG, creams for skin
• Problems include:
  – Chronic muscle inflammation with muscle weakness
  – Rash accompanies or proceeds muscle weakness
  – Swallowing, breathing if those muscles affected
Dermatomyositis/Polymyositis- OT issues

- **Precautions**
  - Swallowing precautions
  - Falls
  - Potential for pressure injuries on areas already irritated by rash (DM)

- **Treatments**
  - Adaptations for progressive weakness with mobility, ADL, IADLs
  - Swallowing management, food consistencies, prep
  - Home safety, falls
  - Fatigue/SOB management with energy conservation/simplification of tasks

- **Goals**
  - Mod I with ADLs as long as possible, then as much independence as able with AE and adaptive techniques- serial therapy
  - Home safety education
  - Mod I to manage swallowing precautions

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Inclusion Body Myositis (IBM)

- Inflammatory, autoimmune, genetic in some cases

- Presents in adulthood >50, primarily in men, slow progression

- Problems include:
  - Progressive muscle weakness/atrophy- first is wrists/hands, quads, anterior tibial
  - Tripping, falling
  - Muscle pain/tenderness
  - Dysphagia
IBM- OT issues

- Precautions
  - Fall risk, stuck in lower chairs
  - Drop items (hot coffee)
  - Swallowing

- Treatments
  - Adaptation for hand/finger flexor weakness with daily tasks
  - Balance during occupations
  - Home safety
  - AE and adaptive techniques for ADL/IADL tasks
  - Modify diet, appropriate food prep

- Goals
  - Mod I with ADL/IADL tasks (as long as possible with AE/techniques)
  - Home safety assessment and training, falls training
  - Education on AE
  - AROM
  - Independent management of swallowing issues, food/drink choices

Post-Polio Syndrome

- Generally 35 years after having Polio

- Problems include:
  - Progressive muscle and joint weakness/pain
  - General fatigue and exhaustion after minimal activity
  - Muscle atrophy
  - Dysphagia, shortness of breath
  - Decreased tolerance of cold temperatures
  - Cognitive problems- concentration and memory
  - Depression or mood swings
Post Polio Syndrome- OT issues

• Precautions
  – Falls
  – Safety (cognitive, physical)

• Treatments
  – Adapting from how they have “always done it”
  – Integrating new mobility, AE into home
  – Energy conservation, work simplification, alternative task performance
  – Cognitive/physical management of home tasks, safety
  – ROM, pain management

• Goals
  – Stay at current ADL/IADL level with new AE/techniques
  – Education on AE, adaptive techniques
  – Home safety assessment
  – Balance, mobility training with occupations
  – ROM WFL, manage/decrease pain

Progressive vs. Non-progressive Treatment

<table>
<thead>
<tr>
<th>Treatment/goal</th>
<th>Progressive</th>
<th>Non-progressive/Traditional OT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Strengthening</td>
<td>Generally not possible, just stay as active as possible</td>
<td>Generally possible, see improvements</td>
</tr>
<tr>
<td>ADLs, IADLs</td>
<td>Will need to adapt over time, AE, possible home renovation</td>
<td>Task performance at current level</td>
</tr>
<tr>
<td>Home safety</td>
<td>Continuous need due to changes, ramp on home</td>
<td>Need for current level only</td>
</tr>
<tr>
<td>Fatigue/endurance</td>
<td>May fluctuate significantly with treatments, days, times, plan strategies for the bad times</td>
<td>Generally stable</td>
</tr>
<tr>
<td>Pain management</td>
<td>May fluctuate, watch the client for over-working</td>
<td>Generally consistent, specific management</td>
</tr>
<tr>
<td>Range of motion</td>
<td>May start as AROM, then supported AROM, then AAROM, then PROM, teach caregivers too</td>
<td>Generally stable, increases with therapy</td>
</tr>
<tr>
<td>Mobility</td>
<td>Transition over time from cane/walker to wheelchair/scooter</td>
<td>Generally stable or improving</td>
</tr>
<tr>
<td>Leisure</td>
<td>May need new hobbies or alternate methods</td>
<td>Generally stable</td>
</tr>
<tr>
<td>Exercise</td>
<td>Sub-maximal, watch fatigue, no sore muscles, daily routine may be enough</td>
<td>Appropriate, expect improvement</td>
</tr>
</tbody>
</table>
What to Expect

• Decline in functional performance – THINK & PLAN AHEAD
  – Think about decline, make appropriate recommendations with future needs in mind
• Expect inconsistent function throughout day due to
  • Fatigue
  • Cognitive changes
• Impact of diagnosis on family & client/family roles
• Limited medical interventions
• Psychological issues – progressive disease with end-of-life issues
  • Anxiety
  • Depression

General Role of Occupational Therapy

• Assist in managing symptoms
• Maintain function
• Optimize quality of life
• Typical to use compensation and adaptations
• Recognize strengths client’s bring
• Often serial therapy- short sessions to solve problems
• Therapy can be non-traditional
• Can be home or community based
Treatment/Goal Ideas

- Eating, grooming, dressing, bathing, toileting
- Emergency access to phone/door, safety
- SRM, AROM, AAROM, PROM with family
- Adaptive techniques and equipment use
- Energy conservation, work simplification
- Set up/transfers to toilet, bath, bed
- Leisure
- IADL's
- Writing, holding books, computer access, phone access
- School/work tasks
- Positioning, bed/chair set up

Goal Setting

- Generally little to no improvement in strength or fine motor skills. Focus on function/ADL's.
- Client centered- what is worth the energy to be independent?
- Be creative
- Adaptive equipment use, adaptive techniques
- Patient and family education
- Improve function or safety, not necessarily FIM level
Writing Sample Goals:

• Client will demonstrate increased safety awareness to modified independent level with transfers in 3 weeks.

• Client will perform LB dressing at mod I with adaptive equipment within 2 weeks.

• Client and caregiver will demonstrate proficiency to independent level with UE ROM home program within 3 weeks.

• Client will express decrease in right shoulder pain to 2/10 within 2 weeks.

• Client will make appropriate food/drink choices 100% of the time when given 3 options.

Helpful sites and references:

• www.rarediseases.org
• www.orpha.net
• www.checkorphan.org
• www.mda.org (for myositis, IBM)
Thanks for your time!!

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