What Every...

Social Worker
Physical Therapist
Occupational Therapist
Speech-Language Pathologist

Should Know About:

Progressive Supranuclear Palsy (PSP)
Corticobasal Degeneration (CBD)
Multiple System Atrophy (MSA)
Progressive Supranuclear Palsy (PSP)

- Originally described as Steele-Richardson-Olzewski Syndrome in 1964
- Rare neurodegenerative brain disease which is the most common parkinsonian disorder after Parkinson’s disease (PD)
- Often mistakenly diagnosed as PD due to similarity of early symptoms
- Presents with early postural instability, supranuclear gaze palsy (paralysis of voluntary vertical gaze with preserved reflexive eye movements), and levodopa-non-responsive parkinsonism
- Onset of symptoms is typically symmetric
- Pathologically classified as a tauopathy (abnormal accumulation in the brain of the protein tau)
- 5–7 cases per 100,000 people
- Slightly more common in men
- Average age of onset is between 60–65, but can occur as early as age 40
- Life expectancy is 5-7 years following symptom onset
- No cure or effective medication management

Corticobasal Degeneration (CBD)

- Originally described in 1968 by Rebeiz, Kolodny, and Richardson; with earliest descriptions by Charcot (1888)
- Rare neurodegenerative brain disease that affects the cortex (fronto-parietal) and basal ganglia
- Pathologically classified as a tauopathy (accumulation of the tau protein in the brain)
- Onset of symptoms is markedly asymmetrical
- Diagnosis is difficult because clinical features often overlap with Parkinson’s disease, progressive supranuclear palsy (PSP), Alzheimer’s, primary progressive aphasia, and fronto-temporal dementia
- Average age of onset usually between 60-80
- Prevalence unknown, estimated less than 1 case per 100,000 people
- Slightly more common in women
- Life expectancy is 7-10 years following symptom onset
- No known cure or medications to slow disease progression

Multiple System Atrophy (MSA)

- First described in the 1960’s as Shy-Drager Syndrome
- Rare neurodegenerative disease that affects multiple systems, particularly motor and autonomic nervous systems
- Pathologically classified as a synucleinopathy (accumulation of the protein alpha synuclein)
- Sub-classified by 3 core clinical features – parkinsonian, autonomic, and cerebellar:
  - MSA-P (parkinsonian): Striatonigral degeneration implies parkinsonism with some degree of cerebellar dysfunction
  - MSA-A (autonomic): Shy-Drager Syndrome reflects a predominance of autonomic failure
  - MSA-C (cerebellar): Olivopontocerebellar atrophy indicates primarily cerebellar defects with minor degrees of parkinsonism
- 3-4 cases per 100,000 people
- Average age of onset is usually after 50
- More common in men
- Life expectancy is 7-10 years following symptom onset
- No known cure or medications to slow disease progression
In both assessment and treatment, helping people who are affected by a degenerative movement disorder is a “call to action” for the fundamental core of social work - person-in-environment, or the biopsychosocial perspective. In order to understand the role of social work with PSP, MSA, and CBD, we will look at “what every social worker should know” in each of the domains.

**Biological**

The Social Worker needs to understand the symptoms, progression, and unique challenges posed by chronic, progressive neurodegenerative disease in order to:

- Offer the best care, empathy, and guidance to patients and families
- Facilitate referrals, communication, and care coordination among multidisciplinary professionals and settings
- Educate family members and healthcare workers in community settings about these diseases and the needs of patients and families

**Psychological**

Depression is biochemically (intrinsic) part of these diseases. Sadness and depression, on the part of patient and family, are also normal reactions (extrinsic) to the losses brought on by the disease. In addition, fear, uncertainty, and frustration are normal, expectable reactions to living with a neurodegenerative disease. It is important for the social worker to help patients, as well as family members, to:

- Express emotions such as sadness, anger, worries, frustration
- Build a support team: healthcare providers, family, friends, volunteers, clergy
- Reach out for support from support groups, peer networks, or one-on-one peer support in order to gain knowledge, information, resources; share feelings and experiences; receive understanding and encouragement
- Develop an attitude and modus operandi of flexibility, persistence, and adaptation
- Cultivate a focus not on “cure,” but rather, on living the best life possible and creating new meaning

**Social and Family**

Often the patient and family feel, and actually are, alone and isolated. This is understandable when we consider the myriad of physical challenges (loss of balance and difficulty with mobility), emotional changes (loss of interest in previously enjoyable activities), self-consciousness (difficulty swallowing, slurred speech), and family impact of PSP, MSA, and CBD. The social worker will help the family to:

- Prioritize their needs
- Adapt their lifestyle to the changes incurred by living with someone with a progressive neurological disorder
- Cope with caregiver stress
- Build or maintain good communication about the impact of living with the disease throughout its progression
- Resolve conflict in the family around family members’ roles, ideas about care, or degrees of acceptance of the disease
- Plan for the future, including understanding home care and housing, advance directives, insurance issues, and other concrete needs
- Learn about and access community resources, services, and programs

**Education and Resources**

Management of these diseases requires care from multiple disciplines and multiple community resources including physical therapy, speech therapy, occupational therapy, social work, psychiatry, and nutrition. Concrete resources include:

- Associations like CurePSP
- Home safety evaluation and modification
- Workplace accommodation
- Social Security disability
- Legal assistance
- Exercise classes and videos
- Prescription drug programs
- Transportation
- Support groups
- Advance directives
- Caregiver support
- Home rehabilitation and home health agencies
- Adaptive and mobility equipment
- Respite
- Hospice

Social workers can offer service through an ongoing, long-term relationship with those affected by the diseases. They also provide a valuable service to patients and families around a specific need, or at periodic intervals, as particular challenges and changes arise.
Progressive Supranuclear Palsy (PSP)

**Signs and Symptoms**

- Early onset gait and balance problems
- “Clumsy-type” gait or slow, shuffling gait
- Incoordination
- Slowed or absent balance reactions / postural instability
- Frequent falls (most often backward)
- Slowed movements
- Rigidity (generally axial)
- Vertical gaze palsy
- Generally downward gaze loss is first
- Abnormal eyelid control
- Decreased blinking with “staring” look
- Blepharospasms (involuntary eyelid spasms)
- Double vision
- Dystonia - commonly at neck and hands into flexion, but can also be into extension at neck
- Speech and swallowing changes
- Subcortical dementia (personality changes, slowness of thought)
- “Rocket sign” - patient jumps up quickly from seated position, often falling back in chair

**Treatment Strategies**

- Patient and caregiver education about disease
- Caregiver training in assistance techniques
- Dystonia – Botox (except for antecollis), stretching, positioning / bracing
- Blepharospasms – Botox, eye crutch
- Double vision – Prism glasses
- Eye movement exercises - tracking, searching for objects, reading words placed at various heights on paper on wall
- Aerobic, strength, and balance exercises along with fall prevention training
- Encourage continual exercise/physical activity and social engagement throughout course of the disease – look to group exercise classes appropriate for patient
- Gait training - focus on large steps with adequate foot clearance and heel strike
- Appropriate assistive device - generally swivel-wheeled rollators with brakes work well, eventual need for wheelchair or scooter
- Teach safe turns - u-turns in open spaces, avoid pivot turns or crossing one foot over the other to turn
- Heel wedge in or on shoe to shift weight anteriorly (may help reduce posterior LOB)
- Home modifications
- Adaptive equipment / devices
- Education of caregiver on likelihood of increased movement impulsivity and decreased safety judgment with disease progression

**Compensatory Tips**

- Wide, staggered stance with ADL performance
- Scanning environment before walking
- Tilting head down to assist with looking down
- Avoid bending low and standing up quickly to prevent posterior LOB

Remember the acronym “FIGS” to assist with differentiating PSP from Parkinson’s disease (PD):

- F = Frequent, sudden falls (generally posteriorly, occur early in disease)
- I = Ineffective medication (anti-Parkinsonian meds do not work)
- G = Gaze palsy (vertically)
- S = Speech and swallow changes

While the literature on rehabilitation for PSP is limited, it suggests that PT plays a role in managing balance and gait. Remember, PT will be needed at various times throughout the course of the disease.

**Research Articles**


Nicolai S, et al. Improvement of balance after audio-


Corticobasal Degeneration (CBD)

Signs and Symptoms

Initially:
- Asymmetrical presentation - symptoms begin on one side and always remain worse
- Slowness and stiffness
- Shakiness
- Clumsiness in UEs or LEs
- Dysphasia, dysarthria, and dysphagia
- Changes in gait and balance
- Mild memory or behavior problems

With disease progression:
- Increased parkinsonian symptoms (rigidity, bradykinesia, postural instability)
- Tremor
- Myoclonus
- Dystonia
- Blepharospasm (involuntary eyelid spasm)
- Sensory loss
- Increasing speech and swallowing difficulty
- Mild to moderate cognitive impairments
- Frontal dementia
- “Alien Limb” phenomenon – limb seems to move on its own and patient - patient has difficulty controlling this, can combine with a sense that the limb is not their own
- Worsening apraxia and contractures

Treatment Strategies

- Patient and caregiver education about disease
- Aerobic, therapeutic and balance exercises along with fall prevention training
- Encourage use and exercise of effected limb
- With disease progression PROM, positioning, and bracing to prevent contractures
- ADL training – adaptive devices, energy management, fall prevention
- For parkinsonian-type gait, focus on large steps and heel strike
- Teach safe turns - u-turns in open spaces, avoid pivot turns or crossing one foot over the other to turn
- Break down tasks into smaller steps
- Appropriate assistive devices - swivel-wheeled rollators with brakes work well only in early stages, apraxia or alien limb often interfere with AD use
- Eventually unable to ambulate due to progressive rigidity / cognitive changes – caregiver assisted mobility with wheelchair
- Caregiver training in assistance techniques
- Home modifications and adaptive equipment / devices
- SLP consultation
- OT consultation
- No treatment for “alien limb”
- Dystonia and blepharospasms may be helped with botox
- Encourage continual exercise/physical activity and social engagement throughout course of the disease – look to group exercise classes appropriate for patient

While the literature on rehabilitation for CBD is limited, it suggests that PT plays a role in managing apraxia, balance and gait. Remember, PT will be needed at various times throughout the course of the disease.

Research Articles


Multiple System Atrophy (MSA)

Signs and Symptoms

- Rigidity
- Action tremor – irregular, jerky, myoclonic movements
- Bradykinesia
- Freezing of gait
- Early postural instability and falls
- Orthostatic hypotension
- Erectile dysfunction
• Bladder dysfunction: urgency, frequency, incontinence
• Constipation
• Speech and swallow difficulties – mixed dysarthria tends to emerge earlier in MSA than PD, is more severe, and deteriorates more rapidly
• REM Behavioral Disorder (RBD) – acting out dreams while sleeping due to lack of atonia
• Gait and limb ataxia
• Nystagmus and jerky pursuit
• Antecollis
• Difficulty with thermoregulation
• Cognitive impairment is typically mild

Treatment Strategies

• Patient and caregiver education about disease

For bradykinesia and rigidity:
• Levodopa and Dopamine agonists may help initially
• Aerobic and flexibility exercises; large movements

For orthostatic hypotension:
• Fludrocortisone, midodrine
• Increase salt in diet
• Rising slowly and resting after position changes
• Support stockings
• Elevate head of bed
• Smaller, more frequent meals
• Avoid increased time in motionless positions
• Avoid warmer temperatures
• Avoid Valsalva-provoking maneuvers

For meal time:
• SLP and OT consultations
• Upright posture in chair
• Alternate food and liquid swallows
• Softer foods
• Adaptive devices to help make self-feeding easier and safer

For antecollis:
• Stretching and positioning / bracing
• Tilt wheelchair
• Botox not often used due to possibility of further speech and swallow problems occurring

For parkinsonian-type gait:
• Focus on large steps and heel strike
• Teach safe turns - u-turns in open spaces, avoid pivot turns or crossing one foot over the other to turn
• Appropriate assistive devices - generally swivel-wheeled rollators with brakes work well, eventual need for wheelchair or scooter

For gait and balance:
• Therapeutic exercise, balance exercise, and fall prevention training
• Encourage continual exercise/physical activity and social engagement throughout course of the disease – look to group exercise classes appropriate for patient

For freezing of gait:
• Don’t “fight” the freeze
• At the first sign of shuffling or freeze, remember the 4 S’s:
  - Stop
  - Sigh – take a deep breath
  - Shift – weight side-to-side
  - Step – take a large step
• Count “1 and 2 and 3...” and then step
• Imagine stepping over something on the ground and then actually step over it
• Place all of your weight on one leg while you swing the other leg back and forth a few times. On the last swing forward, take a step

Other tips:
• Caregiver training in assistance techniques
• Home modifications and adaptive equipment / devices

While the literature on rehabilitation for MSA is limited, it suggests that PT plays a role in managing balance and gait. Remember, PT will be needed at various times throughout the course of the disease.

Research Articles


Progressive Supranuclear Palsy (PSP)

Signs and Symptoms

- Early onset gait and balance problems
- “Clumsy-type” gait or slow, shuffling gait
- Incoordination
- Slowed or absent balance reactions / postural instability
- Frequent falls (most often backward)
- Slowed movements
- Rigidity (generally axial)
- Vertical gaze palsy
- Generally downward gaze loss is first
- Abnormal eyelid control
- Decreased blinking with “staring” look
- Blepharospams (inability to open eyelid)
- Double vision
- Dystonia
- Commonly at neck and hands into flexion, but can also be into extension at neck
- Speech and swallowing changes
- Subcortical dementia (personality changes, slowness of thought)

Disease Progression:

- Motor: balance decreases and rigidity increases so that walking becomes increasingly difficult
- Vision: visual acuity remains but tracking ability deteriorates both vertically and horizontally; inability to maintain eye contact during conversation; abnormal eyelid movement increases
- Cognition: difficulty with multi-tasking, executive function and attention, apathy, emotional lability

Treatment Strategies

- Patient and caregiver education about disease
- Caregiver training in assistance techniques
- Double vision - prism glasses
- Fall prevention training
- Focus on the importance of always scanning the environment (via a downward head tilt) during all functional mobility and ADL’s both in the home and outside in the community
- Appropriate assistive devices - generally swivel-wheeled rollators with brakes work well

- Teach safe turns during ADL training (for example in the kitchen during meal prep and in the bathroom during self-care)
- Using u-turns in open areas in the home or when out in the community
- Always avoiding pivot turns or crossing one foot over the other to turn; instead move right foot first to turn right, left to turn left
- Home modifications / adaptive techniques / compensatory strategies / AE / DME

Compensatory Tips:

- Always use a wide and staggered stance while performing all ADL tasks
- Use of a shower bench with a back and grab bars to eliminate LOB and to promote good posture and energy conservation
- Use of a hand held shower to eliminate turning while removing soap / shampoo
- Use of a long handled sponge to eliminate bending over and potential LOB
- Use of liquid soap vs. a bar of soap to compensate for decreased coordination
- Installing a non-skid surface into tub / shower (non-skid strips)
- Tilting head down to assist with lack of downward eye gaze during all functional mobility and ADL’s especially eating
- Always dress in a seated position to eliminate LOB
- Raising the height of plate / bowl to face level during meals to compensate for lack of downward eye gaze
- Use of rocker knives, deep spoons and food guards to assist with improving self-feeding skills
- Reduce background distractions
- Break down tasks to one step at time
- Allow for increased response time
- Try to face the patient when communicating and restate for clarification during conversations
- PT and SLP consultations
Corticobasal Degeneration (CBD)

Signs and Symptoms

Initially:
- Slowness and stiffness
- Shakiness / UE / LE clumsiness
- Dyphasia and dysarthria and dysphagia
- Changes in gait and balance
- Mild memory or behavior problems

With disease progression:
- Increased parkinsonian symptoms (rigidity, bradykinesia, postural instability)
- Tremor / myoclonus / dystonia
- Sensory loss
- Increasing speech and swallow difficulty
- Cognitive impairments / frontal dementia
- “Alien Limb” phenomenon

Treatment Strategies

- No known cure, no medications to slow disease progression
- Unfortunately there is no treatment for “alien limb”
- Avoid hand flexion / resistive exercise (ex. theraputty / foam squeezes) - instead encourage hand / wrist extension to maintain an open hand for functional tasks
- Role of exercise with this disease is prevention of contractures and disuse atrophy rather than improving coordination - there is no literature to support improvement in FMC via performing FMC activities in the clinic or in the home
- Educate / train patient and caregiver on how to improve safety during ADL’s via modifications to the patient’s routine / way of performing tasks as well as to their home environment

Recommendations:
- Use of a shower chair with a back and a hand held shower head to eliminate turning while bathing and to improve energy conservation
- Grab bar installation to decrease risk of falls
- With disease progression – PROM, positioning, and splinting to prevent contractures if necessary; consider a hand therapist consultation for custom splint fabrication

Cognitive / Psychosocial Strategies

- Break down tasks in to smaller steps / keep instructions simple
- Reduce background noise / eliminate distractions
- Stress can increase symptoms - in stressful situations (e.g. out in the community) be sure to simplify tasks

Multiple System Atrophy (MSA)

Signs and Symptoms

- Rigidity
- Action tremor – irregular, jerky, myoclonic
- Bradykinesia
- Freezing of gait / gait and limb ataxia
- Early postural instability and falls
- Orthostatic hypotension
- Erectile dysfunction
- Bladder dysfunction / constipation
- Speech and swallow difficulties – mixed dysarthria tends to emerge earlier MSA than PD, is more severe, and deteriorates more rapidly
- REM Behavioral Disorder (RBD) – acting out dreams while sleeping due to lack of atonia
- Nystagmus and jerky pursuit
- Antecollis
- Difficulty with thermoregulation
- Cognitive impairment is typically mild
Treatment Strategies

• No medications to slow disease progression
• Patient and caregiver education about disease

For bradykinesia and rigidity:
• Levodopa and dopamine agonists may help initially
• Avoid hand flexion / resistive exercise (e.g. theraputty / foam squeezes). Encourage hand / wrist extension instead

For orthostatic hypotension:
• Fludrocortisone, midodrine
• Increase salt in diet
• Rising slowly and resting after position changes
• Support stockings
• Elevate head of bed
• Smaller, more frequent meals
• Avoid increased time in motionless positions
• Avoid warmer temperatures
• Avoid valsalva-provoking manuevers

Compensatory strategies for self-feeding skills with tremor:
• Use of adaptive devices (adaptive utensils and plate guards)
• Non-skid mat (dycem) to prevent the plate / bowl from sliding
• Arm Position - anchor elbow on table, then lower arm down to plate / bowl to retrieve food and then back up to mouth (never taking elbow up / off of the table – use your arm as a lever)
• Use deep spoons to assist with scooping
• Elevate the height of the dish / bowl

Functional mobility in the home bedroom:
• Use a bedrail to assist with difficulty rolling
• Keep floors clutter free and walkways well lit
• Wear warmer pajamas and use less blankets to decrease likelihood of getting tangled up in bed
• Use a satin pillowcase / sheet under the bottom of the patient to assist with improving bed mobility
• Always sit as much as possible when dressing; this will help to decrease risk of falls (hypotension)
• Always dress the most affected limb first

Functional mobility in the home kitchen:
• Never overreach - get as close as you can to the object that you are reaching for (e.g. in cabinets)
• Always support yourself with one hand when reaching (e.g. countertop)
• Always stand to the side of the dishwasher / oven / refrigerator when opening
• Make u-turns in open spaces around the house (swivel-wheeled rollators with brakes work well)
• Avoiding pivot turns or crossing one foot over the other to turn; instead move right foot first to turn right, left to turn left
• Use a wide staggered stance when performing activities (this helps to reduce LOB and retropulsion)
• Side step when working at the counter-space or sink

Occupational therapy may improve functional abilities in patients with mild to moderate MSA.

Research Articles

Progressive Supranuclear Palsy (PSP)

Speech Pathology Considerations

• Management of swallowing and speech disorders in PSP requires changing intervention strategies as the disease progresses
• Changes in swallowing and speech often occur early in PSP, with typically more profound and rapid deterioration in function as compared with Parkinson’s disease (PD)
• Index of suspicion for dysphagia in PSP should be high, as it is one of the most common causes of mortality, with recurrent respiratory infections and respiratory deaths more common in PSP than PD
• Early swallowing evaluation and frequent monitoring of swallowing function allow for problem anticipation and use of supportive measures
• Goal of swallowing therapy is to minimize complications such as aspiration pneumonia and malnutrition through early symptomatic treatment
• No efficacious approach to speech therapy has been documented for this patient population
• Speech evaluation and re-evaluation may help to classify motor speech impairment type and clarify the neurodegenerative process
• Therapy program should be simple and enhance functional communication as quickly and efficiently as possible vs. just speech outcomes alone
• Strategies to support communication should be implemented proactively as circumstances allow in advance of anticipated functional decline

Swallowing - Symptoms

• Difficulty looking down at the plate
• Mouth stuffing and rapid drinking
• Tremor or stiffness interfere with self-feeding
• Restricted head and neck posture or hyperextension
• Delayed pharyngeal swallow onset
• Poor cough
• Occasional difficulty opening the mouth
• Patient often lacks awareness of swallowing difficulties

Swallowing - Management

• Clinical swallow evaluation should include mealtime observations and suggestions to promote easier and safer swallowing

• Query the caregiver about swallowing symptoms, as patient may not recognize difficulties
• Family should maintain a journal of observations to help define and adjust management strategies
• VFSS, if conducted, needs to replicate the home eating environment, and rule out non-neurogenic contributions
• Decompensation and aspiration may not be observed in a controlled VFSS environment, thus severity of dysphagia may not be truly appreciated
• Discussions regarding feeding tube options should take place sooner rather than later, and repeated frequently
• Patient and family should agree in advance with a doctor about what is hoped to be accomplished with or without placement of a feeding tube

Swallowing - Treatment Strategies

• Optimize oral hygiene
• Supervision at mealtimes
• Moist, soft, tender and well-lubricated foods; avoid dry, particulate, textured foods
• Keep the plate in the line of vision
• Maintain head in a chin-tucked position
• Restrict liquid and food bolus volumes
• Make sure food is swallowed before taking more
• Put cup and utensils down between bites and sips
• Look for mealtime adaptive devices including cups, plates and utensils to assist with self-feeding
• Medications with a puree consistency
• Ask the neurologist about anti-cholinergic drugs or botulinum toxin for management of secretions

Communication - Motor Speech Symptoms

• Mixed dysarthria typically including hypokinetic and spastic dysarthria, less commonly ataxic features
• Strained voice, impaired speech fluency with slow rate of speech and palilalia (the compulsive repetition of utterances in context of increasing rate and decreasing loudness), and emotional lability
• Language and cognitive deficits including frontotemporal dementia and progressive nonfluent aphasia have been observed and can interfere with therapy efforts
• Progresses to anarthria

Communication - Patient Speaking Strategies

• Use compensatory strategies to enhance intelligibility
• Speech must become a conscious effort
• Breathe first, and speak loudly and slowly
• Keep sentences short
• Repeat entire sentence when necessary vs. isolated word
• Say one sentence at a time without immediate repetition
• Establish the context
• Use gestures
• LSVT® techniques emphasizing increased phonatory effort may be of benefit but difficult to habituate
• Investigate assistive forms of communication considering visual, cognitive, and motor limitations such as a communication board or speech generating device to supplement natural speech
• Simple augmentative communication will be required in later stages
• Consider personal portable amplifier

Communication - Listener Strategies

• Keep comments and questions brief
• Stick with familiar topics, and one topic at a time
• Use “yes / no” question format
• Ask for clarification, “Did you say...?”
• Provide choices to ease decision making

Research Articles


Corticobasal Degeneration (CBD)

Speech Pathology Considerations

• Management of swallowing and speech disorders in CBD requires changing intervention strategies as the disease progresses
• Changes in swallowing and speech often occur early in CBD and are typically more severe and deteriorate more rapidly than Parkinson’s disease (PD)
• Index of suspicion for dysphagia in CBD should be high, as it is one of the most common causes of mortality, with recurrent respiratory infections and respiratory deaths more common in CBD than PD
• Early swallowing evaluation and frequent monitoring of swallowing function allows for problem anticipation and use of supportive measures
• Goal of swallowing therapy is to minimize complications such as aspiration pneumonia and malnutrition through early symptomatic treatment
• No efficacious approach to speech therapy has been documented for this patient population
• Speech evaluation and re-evaluation may help to classify motor speech impairment type and clarify the neurodegenerative process
• Therapy program should be simple and enhance functional communication as quickly and efficiently as possible vs. just speech outcomes alone
• Strategies to support communication should be implemented proactively as circumstances allow in advance of anticipated functional decline

Swallowing - Symptoms

• Impaired self-feeding
• Slow or incomplete chewing
• Oral and swallowing apraxia
• Slowed swallowing movements
• May be aware of swallowing difficulties

Swallowing - Management

• Clinical swallow evaluation should include mealtime observations and suggestions to promote easier and safer swallowing
• Family should maintain a journal of observations to help define and adjust management strategies
• VFSS, if conducted, needs to replicate the home eating environment, and rule out non-neurogenic contributions
• Decompensation and aspiration may not be observed in a controlled VFSS environment, thus severity of dysphagia may not truly be appreciated
• Discussions regarding feeding tube options should take place sooner rather than later, and repeated frequently
• Patient and family should agree in advance with a doctor about what is hoped to be accomplished with or without placement of a feeding tube
Swallowing - Treatment Strategies

- Optimize oral hygiene
- Supervision at mealtimes
- Use less-affected side for self-feeding
- Maintain head in a chin-tucked position
- Avoid highly textured, particulate foods
- Blend multiple consistency foods
- Proceed with caution with thin liquids
- Alternate food and liquid swallows
- Medications with a puree consistency
- Look for mealtime adaptive devices including cups, plates, and utensils to assist with self-feeding
- Consider smaller, more frequent meals to shorten duration of mealtimes
- Ask the neurologist about the role of anti-cholinergic drugs or botulinum toxin for management of secretions

Communication - Motor Speech Symptoms

- Characterized by hypokinetic and spastic dysarthria, as well as progressive apraxia of speech and oral apraxia
- Progressive non-fluent aphasia may be strongly associated with this diagnosis
- Speech is hesitant and halting, with strained voice and slower speech production
- Initially may have intact written language
- Frontotemporal dementia may be present
- Progresses to anarthria

Communication - Patient Speaking Strategies

- Use compensatory strategies to enhance intelligibility
- Speech must become a conscious effort
- Use short phrases and simpler language because of increased errors with increased rate of speech, number of syllables, and complexity of language
- Optimize use of written language
- Use of gestures may be limited by apraxia
- Alert to “yes/no” confusion
- Investigate assistive forms of communication considering cognitive and motor limitations such as a communication board or speech generating device to supplement natural speech
- Simple augmentative communication will be required in later stages

Communication - Listener Strategies

- Eliminate distractions to reduce background noise
- Face the speaker with CBD
- Keep comments and questions brief
- Stick with familiar topics, and one topic at a time
- Use “yes / no” question format
- Ask for clarification, “Did you say...?”

Research Articles


Multiple System Atrophy (MSA)

Speech Pathology Considerations

- Management of swallowing and speech disorders in MSA requires changing intervention strategies as the disease progresses
- Early swallowing evaluation and frequent monitoring of swallowing function allow for problem anticipation and use of supportive measures
- Mixed dysarthria is common and tends to emerge earlier in disease course than in PD (Parkinson disease), is more severe, and deteriorates more rapidly
- Up to 1/3 of individuals with MSA may have laryngeal stridor possibly caused by vocal fold abductor paresis or laryngeal dystonia
- Speech evaluation and re-evaluation may help to classify motor speech impairment type and clarify the neurodegenerative process
- No efficacious approach to speech therapy has been documented for this patient population
- Therapy program should be simple and enhance functional communication as quickly and efficiently as possible vs. just speech outcomes alone
- Strategies to support communication should be implemented proactively as circumstances allow in advance of anticipated functional decline

Swallowing - Symptoms

- Difficulty sitting upright at mealtimes
Swallowing - Management

- Tendency towards bolus holding in oral cavity, and discoordinated oral bolus formation and propulsion
- Pharyngeal weakness and disruption of the cricopharyngeal segment
- Excessive accumulation of pharyngeal secretions
- Vocal fold motion impairment may compromise airway protection later in the disease progression
- May occasionally have tracheotomy
- Cough may also be compromised

Speech-Language Pathologists

Motor Speech Symptoms

- **MSA-P**: hypokinetic dysarthria is expected, sometimes mixed with spastic or hyperkinetic dysarthria, and hypophonia (Parkinsonian)
- **MSA-A**: often ataxic or hypokinetic dysarthria, but may be mixed with spastic dysarthria (autonomic)
- **MSA-C**: ataxic dysarthria is most often expected, or in combination with spastic dysarthria (cerebellar)
- Typically more changes in speech than voice
- Cognitive impairment if present is typically mild

Speaker with MSA Strategies

- Speech must become a conscious effort
- Emphasis on taking a breath first
- Reduce rate of speech to improve coordination and accuracy of motor speech movements
- Intelligibility drills with exaggeration of articulation movements
- Investigate assistive forms of communication considering motor and cognitive limitations such as a communication board or speech generating device to supplement natural speech
- Simple augmentative communication will be required in later stages
- Consider personal portable amplifier

Listener Strategies

- Eliminate distractions to reduce background noise
- Face the speaker with MSA
- Keep comments and questions brief
- Stick with familiar topics, and one topic at a time
- Use "yes / no" question format
- Ask for clarification, "Did you say...?"
- Provide choices to ease decision making

Swallowing Treatment Strategies

- Optimize oral hygiene
- Positioning—support to sit upright against the chair back
- Maintain head in a chin-tucked position
- Alternate food and liquid swallows
- Restrict bolus volumes
- Moist, soft, tender foods with lubrication; and blend multiple consistency items
- Look for mealtime adaptive devices including cups, plates and utensils to assist with self-feeding
- Consider smaller, more frequent meals to shorten duration of mealtimes
- Ask the neurologist about the role of anti-cholinergic drugs or botulinum toxin for management of secretions
- There is the potential for lowered blood pressure following meal times (post-prandial hypotension)

Research Articles


About CurePSP

CurePSP is the foremost organization dedicated to curing PSP, CBD, and related brain diseases. CurePSP's mission is to increase awareness of progressive supranuclear palsy, corticobasal degeneration, and other atypical Parkinsonian disorders; fund research toward treatment, cure and prevention; educate healthcare professionals; and provide support, information, and hope for affected persons and their families.

CurePSP devotes the vast majority of its funding to research – this research has three goals:

- Identify the cause and development of PSP, CBD and related diseases
- Develop practical diagnostic tests that would be effective in the early stages of the diseases
- Develop treatments that will prevent, slow, halt or even reverse the progression of the diseases

To help achieve these goals, CurePSP funds research programs such as the Charles D. Peebler, Jr. PSP and CBD Genetics Program and the Eloise H. Troxel Memorial Brain Bank at the Mayo Clinic in Jacksonville, Florida. CurePSP also funds investigator-initiated efforts and clinical trials. This is done by awarding grants to the highest quality and most promising research proposals. It is this type of research that will eventually lead to a cure or various treatment options.

CurePSP strives to provide those in need with support and hope. The Foundation encourages and organizes activities that foster face-to-face communication and exchange, while providing comfort and mutual benefit to those who are caregivers, family members, friends, and persons with the diseases. CurePSP’s face-to-face support groups span the United States and are expanding in many more metropolitan areas, while also serving Canada and Australia. In addition to those groups, CurePSP conducts online support groups, which are accessible via the internet and telephone. The Foundation also maintains a directory of peer supporters - volunteers who are available to offer support by phone or e-mail. CurePSP hosts a free, internet-based forum, which provides the public with a place to share their stories, ask questions, and discuss the diseases. Volunteers are a valued and essential part of CurePSP - the input and energy of the organization’s volunteers has helped expand the services CurePSP offers and the geographical areas it reaches. Volunteers are provided with ongoing education and support and can assist CurePSP in a variety of roles.

Raising awareness about the diseases and educating the public is a top priority. The Foundation supplies up-to-date information about PSP, CBD and related brain diseases to patients, caregivers, and healthcare professionals. CurePSP’s Guide for People Living with PSP, CBD and Related Brain Diseases is a major resource to help patients and their families manage the challenges of these diseases. CurePSP hosts free webinars that cover a variety of subjects and are presented by leading movement disorder specialists and other healthcare professionals. The Foundation sponsors educational conferences that provide patients and families with the opportunity to learn about the diseases and ways to manage their difficult challenges. CurePSP also offers free training for healthcare professionals to assist in the treatment of their patients with these diseases. Advocating for those with rare neurodegenerative diseases will lead to better policymaking, so CurePSP actively works with governmental agencies and the pharmaceutical industry on behalf of patient and caregiver needs.