

PSPCAN

PROGRESSIVE SUPRANUCLEAR PALSY
SOCIETY OF CANADA

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)

A Comprehensive Guide to Signs, Symptoms,
and Management Strategies



Progressive Supranuclear Palsy (PSP)

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

Signs and Symptoms

- Early onset gait and balance problems
- Clumsy, slow, or shuffling gait
- Lack of coordination
- Slowed or absent balance reactions and postural instability
- Frequent falls (primarily backward)
- Slowed movements
- Rigidity (generally axial)
- Vertical gaze palsy
- Loss of downward gaze is usually 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,” when patient jumps up quickly from seated position, often falling back in chair

Corticobasal Degeneration (CBD)

- Rare neurodegenerative disease that affects the cortex (frontoparietal) and basal ganglia
- Originally described in 1968 by Drs. Rebeiz, Kolodny, and Richardson with earliest descriptions by Charcot (1888)
- 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 (PD), progressive supranuclear palsy (PSP), Alzheimer's, primary progressive aphasia, and frontotemporal dementia
- Average age of onset usually between 60 and 80 years
- Prevalence unknown; estimated to be less than one case per 100,000 people
- Slightly more common in women
- Life expectancy is seven to 10 years following symptom onset
- No known cure or medications to slow disease progression

Signs and Symptoms

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

Multiple System Atrophy (MSA)

- Rare neurodegenerative disease that affects multiple systems, particularly motor and autonomic nervous systems
- First described in the 1960s as Shy-Drager syndrome
- Pathologically classified as a synucleinopathy (accumulation of the protein alpha synuclein)
 - Sub-classified by three core clinical features: parkinsonian, autonomic, and cerebellar
 1. MSA-P (parkinsonian)-striatonigral degeneration implies parkinsonism with some degree of cerebellar dysfunction
 2. MSA-A (autonomic)-Shy-Drager syndrome reflects a predominance of autonomic failure
 3. MSA-C (cerebellar): olivopontocerebellar atrophy indicates primarily cerebellar defects with minor degrees of parkinsonism
- Three to four cases per 100,000 people
- Average age of onset is usually after 50 years
- More common in men
- Life expectancy is seven to 10 years after symptom onset
- No cure or medications to slow disease progression

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 swallowing 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

THE ROLE OF SOCIAL WORKERS

In order to successfully assess and treat people struggling with PSP, MSA, or CBD, social workers need to understand the symptoms, progression, and challenges these diseases pose. Social workers' understanding of their clients' biopsychosocial needs enable them to play a key role ensuring that patients and their families receive the best care, empathy, and guidance these chronic, progressive neurodegenerative diseases require. Social workers can build ongoing, long-term relationships with their clients as well as provide valuable services to patients and families around specific needs or at periodic intervals, as particular challenges and changes develop.

PSYCHOLOGICAL SUPPORT

Sadness and depression are intrinsic biochemical responses to these diseases, normal responses to the losses both patients and their families face. In addition, fear, uncertainty, and frustration are normal reactions to living with a neurodegenerative disease. It is important for social workers to help patients and family members to:

- Express emotions such as sadness, anger, worry, and frustration
- Build a support team with healthcare providers, family, friends, volunteers, and clergy
- Reach out to support groups, peer networks, or one-on-one peer support in order to gain knowledge, information, and resources; share feelings and experiences; receive understanding and encouragement
- Develop attitudes and habits 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 CONNECTIONS

Patients and families often feel, and actually are, alone and isolated. This is a result of 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 other difficulties brought on by PSP, MSA, and CBD. The social worker will help patients and families to:

- Prioritize their needs
- Adapt their lifestyles 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, and 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 gain access to community resources, services, and programs

EDUCATION AND RESOURCES

Management of these diseases requires input from multiple disciplines and community resources including physical therapy, speech therapy, occupational therapy, social work, psychiatry, and nutrition. Social workers can facilitate referrals, communication, and care coordination among multidisciplinary professionals and settings and educate family members and healthcare workers about the diseases and the needs of patients and families.

Resources to consider include:

- Associations like the PSP Society of Canada
- 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

While many of the signs and symptoms of PSP, CBD and MSA appear similar, understanding the issues specific to each disease can help physical therapists develop and implement more successful treatment strategies.

Progressive Supranuclear Palsy (PSP)

Patients will benefit from continual exercise, physical activity, and social engagement throughout the course of the disease, including appropriate group exercise classes. Caregivers should be educated about the likelihood of the patient's increased movement impulsivity and decreased safety judgment as the disease progresses.

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

The acronym "FIGS" will help to differentiate PSP from Parkinson's disease (PD):

F = Frequent, sudden falls (generally posteriorly; occurring early in the disease)

I = Ineffective medication (antiparkinsonian meds do not work)

G = Gaze palsy (vertical)

S = Speech and swallow changes

Treatment Strategies

- Patient and caregiver education about the 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, including tracking, searching for objects, reading words placed at various heights on paper on wall
- Aerobic, strength, and balance exercises along with fall prevention training
- Gait training, with a focus on large steps with adequate foot clearance and heel strike
- Appropriate assistive devices: generally swivel-wheeled rollators with brakes work well; wheelchairs or scooters will eventually be needed
- Teach safe turns, i.e., U-turns in open spaces, avoiding 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

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

Research Articles

Suteerawattananon M, MacNeill B, Protas EJ. Supported treadmill training for gait and balance in a patient with progressive supranuclear palsy. *Phys Ther* 2002 May; 82(5):485-95.

Steffen TM, Boeve BF, Mollinger-Riemann LA, Petersen CM. Long-term locomotor training for gait and balance in a patient with mixed progressive supranuclear palsy and corticobasal degeneration. *Phys Ther* 2007 Aug; 87(8):1078-87.

Zampieri C, Di Fabio RP. Balance and eye movement training to improve gait in people with progressive supranuclear palsy: quasi-randomized clinical trial. *Phys Ther* 2008 Dec;88(12):1460-73.

Nicolai S, et al. Improvement of balance after audio-biofeedback. A six-week intervention study in patients with progressive supranuclear palsy. *Z Gerontol Geriatr*. 2010 Aug;43(4):224-8.

Winter Y, et al. Health-related quality of life in multiple system atrophy and progressive supranuclear palsy. *Neurodegener Dis*. 2011;8(6): 438-46.

Corticobasal Degeneration (CBD)

CBD presents with an “alien limb” phenomenon – arms and legs that seem to move on their own. Patients also experience increasingly worse apraxia and contractures. Encourage continual exercise, physical activity, and social engagement throughout course of the disease, with appropriate group exercise classes. While the literature on rehabilitation for CBD is limited, it suggests that PT plays a role in managing apraxia, balance, and gait and will be needed at various times throughout the course of the disease.

Treatment Strategies

- Patient and caregiver education about the disease
- Aerobic, therapeutic, and balance exercises along with fall prevention training
- Encourage use and exercise of affected limb
- PROM, positioning, and bracing to prevent contractures
- ADL training, adaptive devices, energy management, and fall prevention
- For parkinsonian-type gait, focus on large steps and heel strike
- Teach safe turns, i.e., U-turns in open spaces, avoiding 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
- Caregiver training in assistance techniques
- Home modifications and adaptive equipment/devices
- SLP consultation
- OT consultation
- Dystonia and blepharospasms may be helped with Botox

Research Articles

Kawahira K, Noma T, Iiyama J, Etoh S, Ogata A, Shimodozono M. Improvements in limb kinetic apraxia by repetition of a newly designed facilitation exercise in a patient with corticobasal degeneration. *Int J Rehabil Res* 2009 Jun; 32(2):178-83.

Stamenova V, Roy EA, Black SE. A model-based approach to understanding apraxia in Corticobasal Syndrome. *Neuropsychol Rev*. 2009 Mar;19(1):47-63.

Multiple System Atrophy (MSA)

Patients will benefit from ongoing exercise, physical activity, and social engagement throughout the course of the disease, including appropriate group exercise classes. While the literature on rehabilitation for MSA is limited, it suggests that PT plays a role in managing balance and gait. PT is likely to be needed at various times throughout the course of the disease.

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, i.e., U-turns in open spaces, avoiding pivot turns or crossing one foot over the other to turn
- Appropriate assistive devices; generally swivel-wheeled rollators with brakes work well; wheelchairs or scooters will eventually be needed

THE ROLE OF PHYSICAL THERAPISTS

For gait and balance:

- Therapeutic exercise, balance exercise, and fall prevention training

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 few times. On the last swing forward, take a step.

Other Tips

- Caregiver training in assistance techniques
- Home modifications and adaptive equipment/devices

Research Articles

Landers M, Adams M, Acosta K, Fox A. Challenge-oriented gait and balance training in sporadic olivopontocerebellar atrophy: a case study. *J Neurol Phys Ther* 20Sep;33(3):160-8.

Venglar M. Case report: Tai Chi and Parkinsonism. *Physiother Res* 2005;10(2):116-21.

Wedge F. The impact of resistance training on balance and functional ability of a patient with multiple system atrophy. *J Geriatr Phys Ther* 2008;31(2):79-83.

Winter Y, et al. Health-related quality of life in multiple system atrophy and progressive supranuclear palsy. *Neurodegener Dis*. 2011;8(6):438-46.

THE ROLE OF OCCUPATIONAL THERAPISTS

While many of the signs and symptoms of PSP, CBD, and MSA appear similar, understanding the issues specific to each disease can help occupational therapists develop and implement more successful treatment strategies.

Progressive Supranuclear Palsy (PSP)

Treatment Strategies

- Patient and caregiver education about disease
- Caregiver training in assistance techniques
- Prism glasses for double vision

Fall prevention training

- Focus on the importance of always scanning the environment (via a downward head tilt) during all functional mobility and ADL in both in the home and outside in the community
- Appropriate assistive devices; swivel-wheeled rollators with brakes generally work well
- Teach safe turns during ADL training (i.e., 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 a shower bench with back and grab bars to eliminate LOB and to promote good posture and energy conservation
- Use a hand-held shower to eliminate turning while removing soap/shampoo
- Use a long-handled sponge to eliminate bending over and potential LOB
- Use liquid instead of bar soap to compensate for decreased coordination
- Install nonskid surface into tub/shower (nonskid strips)
- Tilt head down to assist with lack of downward eye gaze during all functional mobility and ADL, 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 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)

The role of exercise with this disease is to prevent contractures and disuse atrophy rather than improve coordination. There is no literature to support improvement in FMC via performing in the clinic or in the home.

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

As the disease progresses

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

Treatment Strategies

- There is no treatment for “alien limb”
- Avoid hand flexion/resistive exercise (e.g., Thera Putty®/foam squeezes); instead encourage hand/wrist extension to maintain an open hand for functional tasks
- Educate/train patient and caregiver on how to improve safety during ADL via modifications to the patient’s routine/performance of tasks as well as to their home environment Recommendations
- Use a shower chair with a back and a handheld showerhead to eliminate turning and conserve energy
- Install a grab bar to decrease risk of falls
- As the disease progresses, employ PROM, positioning, and splinting as necessary to prevent contractures; consider a hand therapist consultation for custom splint fabrication
- ADL training – Adaptive techniques/compensatory strategies, adaptive devices (adaptive utensils), DME (bed rails/shower chairs), energy conservation, fall prevention
- Family/caregiver education on how to assist the patient safely throughout disease
- PT consultation for appropriate assistive device evaluation
- Weighted utensils do not always help with tremor and self-feeding deficits
- Generally, swivel-wheeled rollators with brakes work well in the early stages
- Eventually unable to ambulate due to progressive rigidity/cognitive changes; use caregiver assisted wheelchair

Cognitive/Psychosocial Strategies

- Break down tasks into 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)

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

Signs and Symptoms

- Rigidity
- Action tremor, i.e., 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 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
- 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., Thera Putty®/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 maneuvers

THE ROLE OF OCCUPATIONAL THERAPISTS

Compensatory strategies for self-feeding skills with tremor:

- Use of adaptive devices (adaptive utensils and plate guards)
- Nonskid 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 (using the arm as a lever, never taking the elbow up/off of the table)
- 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 fewer 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 to help decrease risk of falls (hypotension)
- Always dress the most affected limb first

Functional mobility in the home bathroom:

- Grab bar/hand-held showerhead installation
- Remove glass partitions in the shower/tub to make transfers in/out easier and safer (sit and swing technique vs. step in)
- Use a shower bench/tub chair with a back to decrease risk of slipping, promote good posture, and conserve energy
- Always sit as much as possible when bathing; eliminate all turning in the shower
- Avoid extremely hot showers to decrease dizziness/lightheadedness

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)
- Avoid 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

Research Articles

Jain S, Dawson J, Quinn NP, Playford ED. Occupational Therapy in multiple system atrophy: a pilot randomized controlled trial. *Mov Disord.* 2004 Nov; 19 (11) :1360-4.

Progressive Supranuclear Palsy (PSP)

Changes in swallowing and speech often occur early in PSP, typically with more profound and rapid deterioration compared with Parkinson's disease (PD). Management of swallowing and speech disorders requires changing intervention strategies as the disease progresses.

Dysphagia 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 treatment and frequent monitoring of swallowing function allow for problem anticipation and use of supportive measures to minimize complications such as aspiration pneumonia and malnutrition.

No efficacious approach to speech therapy has been documented for this patient population, but speech evaluation and re-evaluation may help to classify motor speech impairment type and clarify the neurodegenerative process. Therapy programs should focus less on speech outcomes alone and be simple and enhance functional communication as quickly and efficiently as possible. Strategies to support communication should be implemented proactively as circumstances allow in advance of anticipated functional decline.

Problems with Swallowing

Patients often lack awareness of swallowing difficulties including:

- 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

Swallowing Management: clinical 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 can be accomplished with or without placement of a feeding tube

THE ROLE OF SPEECH-LANGUAGE PATHOLOGISTS

Swallowing Treatment Strategies

- Optimize oral hygiene
- Supervise mealtimes
- Offer 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
- Give medications with a pureed food
- Ask the neurologist about anticholinergic drugs or botulinum toxin for management of secretions

Speech Problems

Mixed dysarthria, typically including hypokinetic and spastic dysarthria; ataxic features are less common

- 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

Speaking Strategies

Speaking must become a conscious effort and to enhance intelligibility, patients must use compensatory strategies including:

- Breathing first and speaking loudly and slowly
- Keep sentences short
- Repeat entire sentence when necessary instead of isolated words
- 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 such as a communication board or speech-generating device to supplement natural speech, depending on an individual's visual, cognitive and motor limitations
- Simple augmentative communication will be required in later stages
- Consider personal portable amplifiers

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

Goetz CG, Leurgans S, Lang AE, Litvan I. Progression of gait, speech and swallowing in progressive supranuclear palsy. *Neurology*. 2003. 60(6):917-22.

Wenning GK, Litvan I, Tolosa E. Milestones in atypical and secondary Parkinsonisms. *Movement Disorders*. 2011. 26(6):1083-1095.

Corticobasal Degeneration (CBD)

Changes in swallowing and speech often occur early in CBD and are typically more severe and deteriorate more rapidly than Parkinson's disease (PD). Management requires changing intervention strategies as the disease progresses.

No efficacious approach to speech therapy has been documented for this patient population. It may help, however, to evaluate and re-evaluate motor speech impairment type and clarify the neurodegenerative process. Therapy should be simple and enhance functional communication as quickly and efficiently as possible instead of focusing on speech outcomes alone and strategies to support communication should be implemented proactively as circumstances allow in advance of anticipated functional decline.

Swallowing function should be evaluated early and monitored frequently to anticipate problems and provide appropriate supportive measures. Dysphagia is one of the most common causes of mortality, with recurrent respiratory infections and respiratory deaths more common in CBD than PD. The goal of swallowing therapy is to minimize complications such as aspiration pneumonia and malnutrition through early symptomatic treatment.

Problems with Swallowing

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

Swallowing Management

- Clinical swallowing 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 possible with or without placement of a feeding tube

THE ROLE OF SPEECH-LANGUAGE PATHOLOGISTS

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
- Provide medications with pureed food
- Use 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 anticholinergic drugs or botulinum toxin for management of secretions

Speech Problems

- Characterized by hypokinetic and spastic dysarthria, as well as progressive apraxia of speech and oral apraxia
- Progressive nonfluent 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

Patient Speaking Strategies

- Speech must become a conscious effort
- Use compensatory strategies to enhance intelligibility
- 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
- Be alert to “yes/no” confusion
- Investigate assistive forms of communication such as a communication board or speech generating device to supplement natural speech, depending on patients’ cognitive and motor limitations
- Simple augmentative communication will be required in later stages

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...?”
- Ask for clarification, “Did you say...?”
- Provide choices to ease decision making

Research Articles

Josephs KA, Duffy JR. Apraxia of speech and nonfluent aphasia: a new clinical marker for corticobasal degeneration and progressive supranuclear palsy. *Current Opinion in Neurology*. 2008. 21:688-692.

Reich SG, Grill SE. Corticobasal degeneration. *Current Treatment Options in Neurology*. 2009. 11:179-185.

Multiple System Atrophy (MSA)

The management of swallowing and speech disorders in MSA requires changing intervention strategies as the disease progresses. Mixed dysarthria is common and tends to emerge earlier in MSA than in PD, is more severe and progresses more rapidly. No efficacious approach to speech therapy has been documented for this patient population. Up to one-third of individuals with MSA may have laryngeal stridor possibly caused by vocal fold abductor paresis or laryngeal dystonia. Therefore, a therapy program should be simple and enhance functional communication as quickly and efficiently as possible and strategies to support communication should be implemented proactively as circumstances allow in advance of anticipated functional decline. Speech evaluation and re-evaluation may help to classify motor speech impairment and clarify the neurodegenerative process.

Evaluate and monitor swallowing early and frequently to anticipate problems and maximize supportive measures.

Problems with Swallowing

- Difficulty sitting upright at mealtimes
- Tendency towards bolus holding in oral cavity, and disorganized 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 as the disease progresses
- May have tracheotomy
- Cough may also be compromised

Swallowing Management

- Clinical evaluation should include mealtime observations and suggestions for 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 revisited often
- Patients and their families should agree in advance with a doctor about what can be accomplished with or without placement of a feeding tube

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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; blend multiple consistency items
- Use 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 anticholinergic drugs or botulinum toxin for management of secretions
- There is the potential for lowered blood pressure following meal times (postprandial hypotension)

Speech Problems

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

Speaking Strategies

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

THE ROLE OF SPEECH-LANGUAGE PATHOLOGISTS

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

Research Articles

Tada M, Onodera O, Tada M, Ozawa T, Piao YS, Kakita A, et al. Early development of autonomic dysfunction may predict poor prognosis in patients with multiple system atrophy. *Archives of Neurology*. 2007. 64:256-60.

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About The PSP Society of Canada

We are a group of committed volunteers dedicated to growing a national organization serving Canadians affected by PSP, MSA and CBD. All of our board members have personal connections to the illness through our loved ones. Having experienced the frustration felt by so many families affected by rare diseases – difficulty getting a diagnosis, healthcare teams unfamiliar with the illness, lack of information and support – we are passionate about not wanting another family in Canada to face the same challenges.

We have benefitted from relationships and training opportunities with other international PSP charities and rare disease umbrella organizations including CORD (Canadian Organization for Rare Disorders) and Eurordis (Rare Diseases Europe). This has helped equip us in our mission to grow support networks, provide educational materials, connect with healthcare professionals, and stay up to date on current research directions.

The PSP Society of Canada is a federally registered non-profit organization, with an application for charity status pending. Prior to establishing independence as the PSP Society of Canada, we worked closely with the US charity CurePSP, and were previously known as CurePSP Canada. We enjoyed getting to know many of you during this period of our development and look forward to continuing to support you moving forward.

VISION:

Every Canadian has access to support, treatment and, ultimately, a cure for PSP and related illnesses.

MISSION:

Provide information and support to families and care-partners. Collaborate with healthcare professionals to increase awareness and improve treatment. Fund research, leveraging partnerships with other charitable organizations for maximum impact.