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Dysphagia in Neurodegenerative Disease

Focus on Amyotrophic Lateral Sclerosis & Parkinson's Disease

Amyotrophic Lateral Sclerosis (ALS)

- "A" means no or negative.
- "Myo" refers to muscle
- "Trophic" means nourishment
  - So, "Amyotrophic" = "No muscle nourishment"
  - When a muscle has no nourishment, it "atrophies."
- "Lateral" identifies the areas in a person's spinal cord where portions of the nerve cells that nourish the muscles are located.
  - As this area degenerates it leads to scarring or hardening ("sclerosis") in the region.

ALS

- ALS involves both upper and lower motor neurons
- Presents as an idiopathic, progressive degeneration of anterior horn cells and their associated neurons resulting in progressive muscle weakness, atrophy, and fasciculations.

ALS

- There is degeneration of:
  - The lateral corticospinal tracts
  - Anterior horn cells
  - Anterior roots and peripheral nerves
  - Nuclei of cranial nerves V, VII, IX, and XII
  - Corticobulbar tracts

ALS

- Prevalence of ALS in the United States is estimated at 25,000-30,000 cases
- The incidence of ALS is two per 100,000 people.
- Approximately 5,600 people in the U.S. are diagnosed with ALS each year.

ALS

- In the United States, 90–95% of ALS cases are sporadic.
  - Military personnel are more likely to develop ALS than the general population.
- 5% are familial ALS
  - FALS is linked to a genetic defect on chromosome 9.

ALS

- Causes
  - Not completely understood.
  - Researchers and physicians suspect viruses, neurotoxins, heavy metals, DNA defects (especially in familial ALS), immune system abnormalities, and enzyme abnormalities.

ALS

- The clinical picture in all stages of ALS can vary and depends on the areas of the nervous system involved in each patient.
- Overall, the disease tends to be insidious and progressive with asymmetric weakness and atrophy.

ALS

- Three types:
  - Bulbar
    - Most rapidly progressive form
    - Involves muscles innervated by cranial nerves involved in speech and swallowing
  - Spinal
    - Progresses more slowly
    - Involves upper and lower extremities controlled by spinal nerves
  - Mixed
    - Includes symptoms of both bulbar and spinal forms

ALS Diagnosis

- Nerve conduction velocity (NCV) and electromyography (EMG) help diagnose nerve and muscle disorders.
  - NCV is administered before EMG and measures the speed at which nerves transmit electrical signals.

ALS Treatment

- There is no cure.
- Riluzole (Rilutek®) is one of the few drugs effective against ALS and may prevent progression and prolong life for a few months or so.
- Treatment focuses on relieving symptoms and maintaining an optimal quality of life.

ALS: Prognosis

- ALS is a terminal illness.
  - 50% of patients die within 3 years of diagnosis
  - 20% live 5 years
  - 10% live 10 years

ALS Severity Scale

(Hillel et al., 1989)

- Used to rate function in four areas:
  - Speech
  - Swallowing
  - Lower Extremities
  - Upper Extremities

ALS Severity Scale

(Hillel et al., 1989)

- Swallowing subscale:
  - 10 Points
    - 9-10 Normal Eating Habits
      - Normal-to-nominal abnormality
    - 7-8 Early Eating Problems
      - 8: Isolated choking episodes; regular diet
      - 7: Prolonged meal times or smaller bite size
    - 5-6 Dietary Consistency Changes
      - 6: Soft diet
      - 5: Liquefied diet
    - 3-4 Needs Tube Feeding
      - 4: Supplemental tube feedings
      - 3: Tube feeding with occasional po nutrition
    - 1-2 NPO
      - 2: Secretions managed with suction or medication or both
      - 1: Aspiration of secretions

Dysphagia in ALS

- Onset is going to vary depending upon type of ALS.
  - May be initial symptom in individuals with bulbar onset ALS.

Dysphagia in ALS

- Dysphagia management may be affected by co-occurrence of mild cognitive impairment or fronto-temporal dementia.

Dysphagia in ALS

- Speech-language pathologists often work as part of a multi-disciplinary team that may include:
  - Neurologists
  - Respiratory Therapists
  - Pulmonologists
  - Rehab Specialists: PT, OT, KT
  - Social Workers
  - Mental Health

Dysphagia in ALS

- Oral Phase Dysphagia
  - Lingual weakness is often more pronounced than jaw or lip weakness (Weikamp et al., 2012)
    - Tongue strength has been found to be a prognostic indicator of survival in ALS.

Dysphagia in ALS

- Pharyngeal phase
  - Reduced laryngeal elevation
  - Reductions in pharyngeal constrictor movement
- Cervical esophageal
  - UES involvement

Dysphagia in ALS

- Initial complaints may be of occasional choking with liquids or solids.
- Individuals may complain of difficulty chewing hard, crunchy foods.
- Our role:
  - Counseling regarding safer food choices.
  - Instrumental swallow assessment if indicated.

Dysphagia in ALS

- Depending upon individual's readiness, may discuss need to consider feeding tube placement.
  - Timing of tube placement is important
    - Weight loss of 10% or more from premorbid weight (Anderson et al., 2005)
    - Vital capacity should be >50% of predicted value (Miller et al., 2009)
  - Encourage continued p.o. intake

Dysphagia in ALS

- Individuals may complain of difficulty taking liquids from a cup
  - Many choose to use a straw instead.
  - May recommend metered dose type cup to control bolus flow and size.

Dysphagia in ALS

- Sialorrhea may be a complaint
  - Botox appears to be safe and effective.

Stokholm et al., 2013

Dysphagia in ALS

- Recommend regular follow-up with individuals
  - Our role is to continue to make recommendations for diet modifications
    - May suggest liquefying foods or adding gravies/sauces to foods
    - Suggest altering diet consistency
    - Suggest smaller meals distributed throughout the day.

Dysphagia in ALS

- Exercise-based programs
  - Very little empirical evidence that strength training improves or maintains function in the limbs
  - Some evidence that respiratory training may help maintain respiratory function
  - To date, there is no empirical evidence to support exercise-based program for treatment of dysphagia in individuals with ALS.

Parkinson's Disease

- One million people in the U.S. have Parkinson's Disease
- Approximately 60,000 new cases of Parkinson's disease are diagnosed in the U.S. each year.
- The incidence of Parkinson's disease increases with age, but 4% of cases occur before the age of 50.
  - Statistics from Parkinson's Disease Foundation

Parkinson's Disease

- Results from damage to the substantia nigra, which produces dopamine.
- Reduction in dopamine results in an imbalance in neurotransmitters
  - Too much acetylcholine; too little dopamine
- Characteristics include:
  - Rigidity -Tremor
  - Bradykinesia -Disturbed Postural Reflexes

Parkinson's Disease

- Difficulty can occur in any phase of swallowing
  - Oral
  - Pharyngeal
  - Esophageal

Parkinson's Disease

- Oral Phase
  - Difficulties with this phase usually occur first.
  - May include:
    - Difficulty chewing
    - Tongue pumping behavior
      - Non-propulsive back and forth motion of the tongue
    - Anterior spillage of material
    - Drooling
      - Due to reduction in frequency of swallowing
      - Up to 55% of patients with Parkinson's report xerostomia

Tjaden, 2008

Parkinson' Disease

- Pharyngeal Phase:
  - Delayed initiation of the pharyngeal swallow
  - Decreased tongue base retraction
  - Decreased pharyngeal contraction
  - Decreased hyolaryngeal excursion
    - Decreased airway protection during the swallow

Parkinson's Disease

- Esophageal Phase:
  - Decreased UES opening
  - Decreased esophageal motility
  - Patients complain of feeling as if food gets stuck

Sung et al., 2010

Parkinson's Disease

- Co-occurrence of dementia can affect swallow safety
  - Impulsivity
  - Lack of awareness of swallowing difficulty

Parkinson's Disease

- Dysphagia Treatment
  - Lee Silverman Voice Treatment
  - Diet Modification
  - Rehabilitative Exercises
    - EMST
    - Lingual strengthening
  - Compensatory Strategies

Parkinson's Disease

- Sialorrhea Treatment
  - Dopaminergic drugs may have some benefit
  - Botox injections to the salivary glands appear to be most effective.

Srivanitchapoom et al., 2014

Conclusions

- Treatment of dysphagia in patients with neurodegenerative disease requires a unique approach and depends upon the neurological underpinnings of the disease.
- Clinicians must have a thorough understanding of the disease causing the dysphagia in order to formulate an appropriate treatment plan.