ALS and Symptom Management

Information for the Healthcare Provider
Part I

What is ALS?

- Amyotrophic Lateral Sclerosis, Lou Gehrig’s Disease
- Progressive, fatal neurodegenerative disorder
- Death of motor neurons cause progressive muscle weakness, atrophy and paralysis of the voluntary muscles

Motor Neuron

- Motor neurons reach from the brain to the spinal cord and from the spinal cord to the muscles throughout the body.
- When the motor neurons die, the ability of the brain to initiate and control muscle movement is lost.

History of ALS

- Charles Bell (1830): case of middle aged woman with progressive paralysis of limbs and tongue, preservation of sensation.
- Charcot (1874) established clinical entity of Amyotrophic Lateral Sclerosis (ALS)

Epidemiology of ALS

- Incidence of ALS 2/100,000 population per year
- Each year 5,600 cases are diagnosed in U.S.
- 30,000 Americans may have the disease at any given time
### Epidemiology
- Clusters: In the 1940s-Very high incidence of a fatal variant on West Pacific Island of Guam. (ALS,PD).
- ALS is not contagious.
- Throughout the world with no racial, ethnic or socioeconomic boundaries

### Risk Factors Linked to ALS
- Aging
- Gender
- Genetics (10% of ALS is familial)
- Military service
- Other potential factors
  - Smoking
  - Viral infections
  - Exposure to environmental toxins
  - Head and Spinal Trauma

### Statistics
- Every 90 minutes someone is diagnosed with ALS and every 90 minutes someone dies of ALS
- 1 in every 50 families is affected by ALS
- 300,000 people alive today in the U.S. will develop and die with ALS
- Types: Sporadic (90%), Familial (10%)

### Life expectancy
- Average: 2-5 years after diagnosis
- Slow progression: 15% to 25% with ALS live 10 years after they first notice initial symptoms
- Rapid progression: respiratory failure within a year which is unusual.

### Types of Onset
- Limb Onset (Upper or Lower)
  - Fasciculation/Twitching of muscles
  - Drop foot (tripping over carpet, etc.)
  - Weak Hand (carpal tunnel)
- Bulbar Onset
  - Problems with speaking, chewing and swallowing
- Respiratory Onset
  - Problems with shortness of breath, sleep, fatigue
- Trunk Onset
  - Unable to hold body upright
- FTD
  - Word finding, poor decision making
ALS Diagnosis

- No clinical or laboratory test to identify ALS; diagnosis is generally made through a careful examination of medical history and neurological examination.
- Routine tests to assist in establishing the diagnosis of ALS include: EMG, NCV, muscle biopsy, X-rays, blood tests, urine tests, spinal taps and pulmonary function tests.

Establishing an ALS diagnosis

- El Escorial World Federation of Neurology Criteria for Diagnosis of ALS
- History and neurological examination searching for clinical evidence of UMN and LMN signs in 4 regions [brainstem, cervical, thoracic, or lumbar spinal cord] of the central nervous system [CNS].
- Neurophysiological, neuroimaging and laboratory studies as clinically indicated, to exclude other disease processes.

Diagnostic criteria

The presence of:

1) Evidence of lower motor neuron (LMN) degeneration (clinical, electrophysiological or neuropathologic examination)
2) Evidence of upper motor neuron (UMN) degeneration (clinical examination, and
3) Progressive spread of symptoms or signs within a region or to other regions, (history or examination)

Diagnostic criteria—con’t

The absence of:

1) Electrophysiological and pathological evidence of other disease processes that might explain the signs of LMN and/or UMN degeneration, and
2) Neuroimaging evidence of other disease processes that might explain the observed clinical and electrophysiological signs.

Diagnostic criteria

**Definite ALS:** Progressive disease with
- Upper & lower motor neuron signs in bulbar & 2 spinal regions, or
- Upper & lower motor neuron signs in 3 spinal regions

**Probable ALS:** Progressive disease with
- Upper & lower motor neuron signs in 2 regions
  and
- Upper motor neuron signs in a region rostral to the lower motor neuron signs

Differential diagnoses that can lead to ALS

- PLS: Primary Lateral Sclerosis (Upper Motor Neuron Only)
- PMA: Progressive muscular atrophy (lower motor neuron only)
  - BAD: Brachial Amyotrophic Dislegia
- PBP: Progressive Bulbar Palsy
Principles of ALS Management

- High priority on patient self-determination or autonomy in the therapeutic relationship
- Patients and families need information that is timely and well in advance of major management crossroads.
- Health care professionals should address the full continuum of care for the ALS patient
- Advanced Directives should be introduced and reevaluated no more than every 6 months

Nutrition/Dysphagia

- ALS has a hypermetabolic nature—weight loss in ALS results from loss of muscle mass, decreased intake and energy cost of activities
- Suboptimal caloric and fluid intake causes a worsening of muscle atrophy, weakness, and fatigue.
- Common symptoms include jaw weakness and fatigue, drooling, choking on fluid and food, and slow eating.
- Modified barium swallow test indicates a high risk for aspiration and subsequent pneumonia.

Dietary Modification

- Consult by SLP and Dietitian
- Liquids: Thick It/Thicken Up/Simply Thick
- Food Consistency: I Can’t Chew Cookbook by J. Randy Wilson (Soft and pureed ideas)
- Increasing Calories/Protein: Ensure/Boost/Carnation Instant Breakfast

Treatment

- Rilutek
  - Currently the only FDA approved drug to slow progression of the disease
  - Can extend life expectancy average of 3 months
  - Average cost: $1000 per month

- Symptom Management
  - Providing management of the patient’s symptoms rather than curing the disease
  - Quality of life

When is a PEG indicated in ALS?

- The presence of inadequate oral intake and diminished quality of life due to choking rather than the result of a swallowing study.
- When FVC falls to 50% of predicted
- Not in the preterminal phase.
- Symptomatic dysphagia with accelerated weight loss due to insufficient caloric intake, dehydration, or ending meals prematurely.
**Early indications of respiratory insufficiency?**
- Dyspnea on exertion
- Supine dyspnea
- Marked fatigue
- Disturbed sleep (frequent nocturnal awakenings, excessive daytime sleepiness), morning headaches

**Does noninvasive ventilation improve respiratory function or increase survival?**
- Noninvasive ventilation improves the symptoms of hypoventilation, thereby improving quality of life and increasing survival of patients with ALS. *Aboussouan*, 1997.
- Invasive ventilation increase survival more effectively but with a greater financial and care burden. *Cazzoli PA. J Neurol Sci 1996;139*
- Loss of bulbar muscle tone and difficulty clearing secretions reduce tolerance of noninvasive ventilation and invasive ventilation should be considered.

**Tracheostomy Invasive Ventilation**
- Patients that want long term ventilator support
- Both patient and family must understand the benefits and burdens of this decision (cost, amount of care needed, possible nursing facility placement)
- Studies show that TIV is effective in preserving QOL for patients with ALS but with greater burden to the caregiver

**What is the best test of detecting early signs of impending respiratory failure?**
- Maximal inspiratory pressure is the most sensitive
- Erect sitting FVC, and possibly supine FVC
- Nocturnal oximetry in evaluating nocturnal hypoventilation
- Full polysomnogram is rarely needed.
- A decrease in VC to 50% of predicted: Respiratory symptoms.
- A VC less than 1 L (or less than 25 to 30% of predicted) indicates significant risk of impending respiratory failure or death.

**Respiratory Guideline**
- In accordance with the principle of patient autonomy, physicians should respect the right of the patient with ALS to refuse or withdraw any treatment, including mechanical ventilation.
- When withdrawing ventilation, use adequate opiates and anxiolytics to relieve dyspnea and anxiety.
Symptom management

- Sialorrhea
- Pseudobulbar affect
- Cognitive and Behavioral Impairments
- Depression/Anxiety
- Spasticity and Cramps
- Fatigue
- Dysarthria
- Mobility issues

Sialorrhea

- Pharmacologic Intervention
  - Robinul (Glycopyrrolate)
  - Amitriptyline
  - Scopolamine
  - Atropine
  - Botox (Type B)

- Nonpharmacologic treatments
  - Suction machines
  - MIE

Clinical complaint: dribbling (sialorrhea)

- Initial intervention
  - Evaluate swallowing
    - Nasogastric tube
    - Suction

- Pharmacologic
  - Ondansetron
  - Metoclopramide
  - Nortriptyline

- Nonpharmacologic
  - Suction
  - MIE

Figure 1. Algorithm for assessment and management.