It Takes a Village: The Multidisciplinary Approach to ALS Care

September 24, 2018

Nicole Yarab, RN, BA
Senior Director, Clinical Programs
Disclosures

- Presenter has no financial or non-financial interest to disclose.
Learning Outcomes

At the conclusion of this activity, the participant will be able to:

• Discuss the importance of early intervention in the management of ALS

• Identify key members of the ALS healthcare team needed to maximize quality of life and symptom management.

• Describe at least two benefits of the multidisciplinary ALS clinic model

• Identify two essential elements of proactive ALS care
Outline

• What is ALS?
• Multidisciplinary care
• Symptom management
• Roles of the various care team members
• Key treatments and interventions
• Final thoughts
What is ALS?

• First identified in 1869 by Charcot, French neurologist

• Progressive, neurodegenerative disease

• Affects motor nerve cells in the brain and spinal cord:
  – Upper motor neuron: spasticity
  – Lower motor neuron: muscle weakness and atrophy
WHAT IS ALS?
amyotrophic lateral sclerosis | noun

ALS IS A PROGRESSIVE NEURODEGENERATIVE DISEASE THAT AFFECTS NERVE CELLS IN THE BRAIN AND SPINAL CORD.

ALS usually strikes people between the ages of 40 and 70, and approximately 20,000 people in the U.S. have the disease at any given time.

Although there is not yet a cure or treatment that halts ALS, scientists we fund through our global research program have made significant progress in understanding what causes ALS. But their work is not done. Together, we work toward a cure.

FACTS ABOUT ALS

- 5,000 people are diagnosed per year
- 10% of cases are inherited through a mutated gene
- 2-5 years is the average life expectancy
- Every 90 minutes someone is diagnosed and someone passes away
- $250,000 is the estimated out-of-pocket cost for caring for a person with ALS
- Only 3 drugs are currently approved by the U.S. FDA to treat ALS (Riluzole, Nuedexta, and Radicava)
- $2 billion is the estimated cost to develop a drug to slow or stop the progression of ALS
- There is NO CURE for ALS

SYMPTOMS

Progressive loss of muscle control

ALS gradually prohibits the ability to:
- Speak
- Swallow
- Walk
- Grasp objects
- Move
- Breathe

DIAGNOSIS

Difficult to diagnose

- ALS is often diagnosed by ruling out other diseases, which may take months or years

MILITARY

Veterans are 2x as likely to get ALS

- ALS affects veterans who served in peacetime and war
- ALS impacts veterans, regardless of the branch of service or the war they served in

The ALS Association Core Values: COMPASSION - INTEGRITY - URGENCY
Presentation and Diagnosis

• Gradual onset of symptoms
• Progression very individualized
• Difficult to diagnose- no one diagnostic test for ALS
• ‘Clinical diagnosis’
• Rule-out other treatable conditions
• Second opinion with neuromuscular/ALS specialist
• Multidisciplinary care recommended
Management of ALS, Related Symptoms and Potential Complications

- Pseudobulbar affect (PBA)
- Mood
- Cognitive changes
- Excessive saliva
- Speech changes
- Communication challenges
- Difficulty swallowing
- Shortness of breath
- Difficulty coughing/clearing secretions
- Spasticity
- Muscle cramps
- Edema
- Weight Loss/Malnutrition
- Decreased fluid intake/dehydration
- Constipation
- Weakness
- Contractures
- Pain
- Mobility Issues
- Person with ALS
- Family members
- Friends
- Physicians
- Various healthcare professionals
- Veterans Administration
- Veterans service organizations
- Nonprofit organizations
- Other community and governmental organizations
Multidisciplinary ALS Clinics - WHY?

- Research shows that individuals treated in a multidisciplinary ALS clinic setting live an average of 12 months longer.

- The **American Academy of Neurology (AAN) Practice Parameter Update (2009)** cited an Italian study that showed that people with ALS who attended specialty ALS clinics vs. a general neurology clinic had fewer hospitalizations and were more apt to utilize:
  - Riluzole
  - Feeding tube
  - NIPPV (Noninvasive Positive Pressure Ventilation, i.e.-BiPAP)
Multidisciplinary ALS Clinics - WHY?

- People with ALS can maintain independence longer and enjoy improved quality of life when provided with options for:
  - symptom management
  - assistive and adaptive equipment
  - education
  - care services
  - emotional support

- Due to the progressive nature of ALS and the constant need to adapt to change, regular 3-month follow-ups are recommended
Goals of Multidisciplinary Care Model

• Provide best-practice, multidisciplinary care to people living with ALS and their families
• Focus on independence and quality of life
• Coordinated, streamlined care
What to Expect at a Multidisciplinary ALS Clinic

• “One Stop Shop” approach whenever possible

• Coordinated care

• Mutual respect and active collaboration between health care providers and person with ALS in their care plan

• May have opportunity to meet other people living with ALS and their families

• May have opportunities to participate in some form of ALS research
The ALS Multidisciplinary Care Team

- The ALS Association
- Neurologist
- Nurse
- Pulmonologist
- Mental Health Professional
- SLP
- ATP
- GI
- Chaplain
- Dietitian
- PT
- OT
- Orthotist
- Friends
- Research Coordinator
- Palliative Care
- Social Worker
- Person with ALS/Family
Neuromuscular Specialist (MD/DO)

• Expert in diagnosis, evaluation and management of ALS
  – Typically a neurologist
  – May be a physiatrist in some settings

• Observe and assess for:
  – Fasciculations (twitching)
  – Brisk reflexes
  – Spasticity (stiffness/slowness)
  – Atrophy
  – Muscle strength
Neuromuscular Specialist (MD/DO)

• Educates about the disease, risks/benefits of medications and procedures used for treatment and symptom management

• Discusses alternative, unproven or unorthodox treatments, including potential risks/benefits

• May provide ALS research updates and information about/opportunities to participate in research
Neuromuscular Specialist (MD/DO)

- Reviews patient history, performs neurological exam and related work-up, diagnoses and treats

- Develops care plan in conjunction with person living with ALS/family
  - ideally with other multidisciplinary team (MDT) members and care providers

- Recommends and prescribes appropriate therapy, interventions, equipment and technology in collaboration with MDT

- Provides maximum symptomatic management and proactive treatment plan options that may include:
  - Palliative measures
  - Gastrostomy tube (PEG or RIG)
  - Non-Invasive Positive-Pressure Ventilation (NIPPV)
  - Tracheostomy/invasive ventilation
ALS Treatment

• Medications
  – Slow disease progression
  – Symptom management

• Therapy

• Assistive devices/technologies

• Supportive care
ALS Treatment

Riluzole

First FDA-approved drug for the treatment of ALS (1995)
  o Oral medication taken twice a day
  o Modest benefit
  o Studies show that it can prolong life by 2-3 months
  o Does not treat symptoms of ALS

*Possible side effects:* elevated liver enzymes (monitor), increased fatigue, stomach upset

➢ Tiglutik- (*ITF Pharma*) thickened liquid riluzole
  – FDA-approved Sept. 2018
  – estimated availability mid-October 2018
ALS Treatment

- FDA approved in USA in 2017, based on trials conducted outside of the USA
- Was used in Japan to treat stroke and approved for ALS in Japan & South Korea in 2015
- IV infusion
- May slow decline in physical function
- For up-to-date FAQs and webinars: www.alsa.org/research/radicava/
Pseudobulbar affect (PBA)

- Condition characterized by uncontrollable outbursts of laughing or crying
- Often exaggerated compared to one’s emotional state

**Treatment options may include:**

- Antidepressants (tricyclics, SSRIs)
- Nuedexta® - a combination of dextromethorphan and quinidine was FDA-approved in 2010 for PBA

*Some people opt for no treatment*
Saliva management

• **Drooling, excessive salivation**

  **Treatment options may include:**
  - Medications: e.g.- Glycopyrrolate, hyoscyamine, scopolamine patch, atropine drops
  - Portable oral suction machine
  - Botox injections into salivary glands
  - Radiation of salivary glands

• **Thick mucus/secretions**

  **Treatment may include:**
  - Increase fluid intake
  - Guaifenesin
Nurse/ Clinic Coordinator

• Coordinates multidisciplinary team and ALS clinic operations

• Point person for patient/family

• Educates about disease, managing symptoms and various aspects of living with ALS

• Coordinates appointments and referrals for community services, equipment, etc.

• Collaborates with ALS Association Chapter liaison to support continuity of care in between visits
Respiratory Therapist or Pulmonologist

- Educates people with ALS/families about how ALS affects breathing and the importance of early intervention
  - Respiratory muscle weakness; not gas exchange issue

- Evaluates respiratory status
  - Forced or Slow Vital Capacity (FVC/SVC)
  - Negative Inspiratory Force (NIF) or Maximum Inspiratory Pressure (MIP)
  - Pulse oximetry

- Collaborates with neurologist to address issues related to pulmonary diseases which may worsen due to ALS
Respiratory Therapist/Pulmonologist

• Discusses non-invasive and invasive support options, as appropriate

• Educates on how to use tools/equipment
  – e.g.- breath stacking, Cough Assist machine, non-invasive positive pressure ventilation (NIPPV)

• Works with team to ensure that home health providers meet respiratory needs
Shortness of Breath & Airway Clearance Issues

• Diaphragm and accessory muscles of breathing become progressively weaker

• **Treatment options may include:**
  - Breath stacking
  - Non-Invasive Positive-Pressure Ventilation (NIPPV)
  - Insufflator-Exsufflator (Cough Assist)
  - Oral Suction
  - Tracheostomy/invasive ventilation
  - Palliative measures

❖ As respiratory numbers decline, continued discussions about:

*Quality of Life*   *Personal goals of care/Advanced directives*
*Respiratory support*  *Feeding tube*   *Equipment needs*
*Finances*  *Lifestyle changes*   *Coping/support system*
*Palliative measures vs. surgical intervention*
Dysarthria

- Progressive deterioration of speech
  - Changes in voice pitch and tone
  - Slurred or slow speech
  - Stiff/spastic speech
  - Worsens with fatigue/time of day
  - Communication challenges

Treatment plan:
  - SLP consultation
  - Energy conservation
  - Compensatory strategies
  - Early: voice amplifier, voice banking
  - Later: augmentative and alternative communication (AAC)
Speech-Language Pathologist

• Assesses speech

• Educates about changes in speech

• Offers compensatory strategies
  – energy conservation
  – planning for communication early in the day

• Discusses options for communication and voice banking
Speech-Language Pathologist/ATP

- Evaluates and recommends appropriate rapid access and high-tech communication (AAC) options
Speech-Language Pathologist

- Assesses swallowing
- Educates about changes
- Recommends strategies and modifications
  - smaller bites
  - changes in food consistency
  - chin tuck
- Recommends best consistencies for safest swallow
- Educates about feeding tubes/enteral nutrition as appropriate
Dietitian

• Evaluates weight and current nutritional status

• Collaborates with the Speech-Language Pathologist to maximize hydration and nutrition

• Educates patient and family on:
  o changes in nutrition seen in ALS
  o guidelines for caloric intake
  o importance of maintaining weight/good nutritional status
  o feeding tubes/tube feedings
Weight Loss and Malnutrition

- Monitor weight closely
- Goal= maintain weight
- Monitor respiratory status regularly

❖ Correlation between significant weight loss and drop in breathing numbers - rapidity of progression

Treatment:
- Early intervention is key
- Smaller, more frequent meals
- Nutrition supplements
- Gastrostomy tube (PEG/RIG)
Constipation

- Frequent problem
- Contributing factors: dysphagia, decreased mobility

**Treatment:**
- Dietitian consult
- Increase fluid intake (orally or via PEG)
- Prune juice
- Fiber-rich foods/supplements
- Stool softener
- Bulk laxative
- Bowel training regimen
Occupational Therapist

• Evaluates upper extremities (UE) for:
  o Pain
  o Weakness
  o Edema
  o Range of motion (ROM) limitations

• Goals:
  o increase ROM
  o reduce pain and/or edema in the UE
  o prevent contractures

• Provides appropriate:
  o orthotics
  o positioning techniques
  o ROM exercises
Occupational Therapist

- Assesses level of independence with activities of daily living (ADLs)
- Offers compensatory strategies to provide optimal performance in:
  - Self-care/ADLs
  - Energy conservation
  - UE home exercise programs
- Offers options for assistive devices/adaptive equipment to assist with ADLs:
  - Feeding
  - Bathing
  - Dressing/grooming
  - Toileting
  - Work-related tasks
  - Driving
Physical Therapist

• Evaluates lower extremities for spasticity, ROM, and mobility

• Addresses fatigue, edema/swelling of limbs, mobility issues

• Evaluates for appropriate exercise program

• Recommends appropriate assistive devices, equipment, and orthotics to optimize safety with functional mobility
Physical/Occupational Therapist

• Recommends appropriate manual or power wheelchair as needed

• Provides resources for dealing with accessibility issues both in and out of the home

• Re-evaluation as needed for comfort and mobility issues as function changes over time
Spasticity

“A condition in which there is an abnormal increase in muscle tone or stiffness of muscle which might interfere with movement, speech or be associated with discomfort or pain” - NINDS Oct. 2011

- Mild to severe
- Can cause:
  - Mobility issues
  - ↑ Fall risk
  - ↑ Difficulty communicating

Factors that worsen spasticity:
- Cold temperature
- Stress
- Fatigue/lack of sleep

Treatment options may include:
- Medications
- Botox
- Stretching/ROM exercises
- Massage

➢ Modify activities to compensate
Dependent edema

• Very common in weak/atrophied limbs

• Affected limb may be cool to touch

• Treatment options may include:
  – Compression stockings
  – ROM exercises
  – Very gentle massage
  – Positioning - elevate limbs
    • utilize power wheelchair features

*Diuretic medications are usually ineffective, generally not prescribed
Social Worker

• Provides emotional support/resources to person with ALS/family

• Provides unique perspective into family dynamics

• Addresses a variety of concerns:
  o relationships
  o workplace issues
  o insurance
  o caregiving
  o financial problems
  o talking to children/family
Social Worker

• Discusses your wishes for current and future care and provides appropriate references/documents

• Seeks out and mobilizes community resources

• Provides information and guidance: eligibility/applying for benefits
The ALS Association Chapter Liaison

• May serve as a liaison between person living with ALS/family and community clinicians
• Empowers people living with ALS and their families by providing information and education
• Provides information and refers to community resources
• Provides information on the National ALS Registry
• Leads ALS support groups
• Consults with clinic team and other community providers to provide appropriate loaner equipment, as needed
• Collaborates with clinicians to support continuity of care
Mental Health Professional

- A psychiatrist, psychologist, social worker or nurse with specialized education and training in psychology and counseling
- Performs psychosocial assessment
- Provides counseling and emotional support
- May perform cognitive/behavioral assessment
Other Providers

- Palliative Care
- Chaplain
- Orthotist
- Durable medical equipment (DME) professional
- Assistive technology professional (ATP)
- Primary care physician
- Gastroenterologist
- PVA representative
Coordinated Care

• ALS is a complex disease
• Over time, increased support required
• More than a physician is needed to optimize care
• Multiple healthcare specialists and community providers involved in care plan
• Can be overwhelming and stressful on the person with ALS and the family
• Ongoing, clear communication and coordination is essential
  – between person with ALS/family and providers
  – between various healthcare professionals
Final Thoughts

• Be proactive, not reactive
• Conserve energy
• Maintain weight
• Adapt activities of daily living
• Maintain a strong support system
• Work closely with the care team
The ALS journey is not an easy one, but each step can be a little easier when it's a team effort.
QUESTIONS?

Nicole Yarab, RN, BA
nyarab@alsa-national.org
www.alsa.org

Connect with your local ALS Association Chapter:
www.alsa.org/community/chapters/

Information and referral email:
alsinfo@alsa-national.org