

Welcome!
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Create Your ALS Care Team

Nicole Yarab, RN, BA
The ALS Association

Attendees are advised that portions of this webinar will be recorded for later viewing in our archives. If you would like to review the recording, please refer to our website, for information (www.alsa.org).

Hosted by:
The ALS Association
National Office-Care Services





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Nicole Yarab, RN, BA
Senior Director, Clinical Programs
The ALS Association

Disclosures

- Employee of The ALS Association, National Office
- No conflicts of interest to report

Objectives

At the end of this presentation, you will be able to:

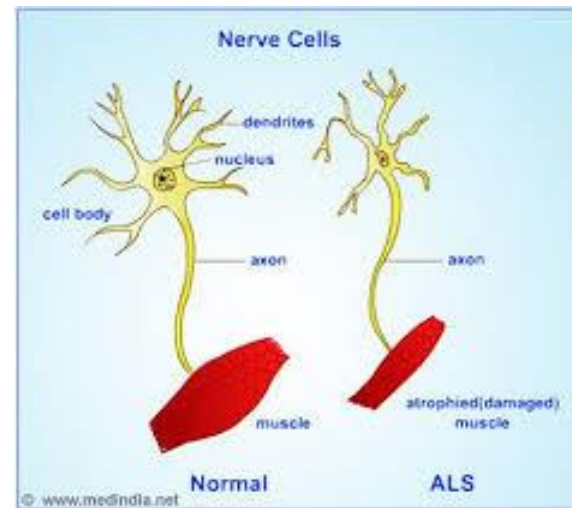
- Discuss the multidisciplinary approach to the care management of a person living with ALS
- Identify key members of the ALS healthcare team needed to maximize quality of life and symptom management
- Discuss the importance of early intervention in the management in ALS

Outline

- 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
- Causes severe muscle weakness and the loss of the ability to:
 - Speak
 - Swallow
 - Move
 - Breathe



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
- Friends
- Physician
- Various healthcare professionals
- Community support

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

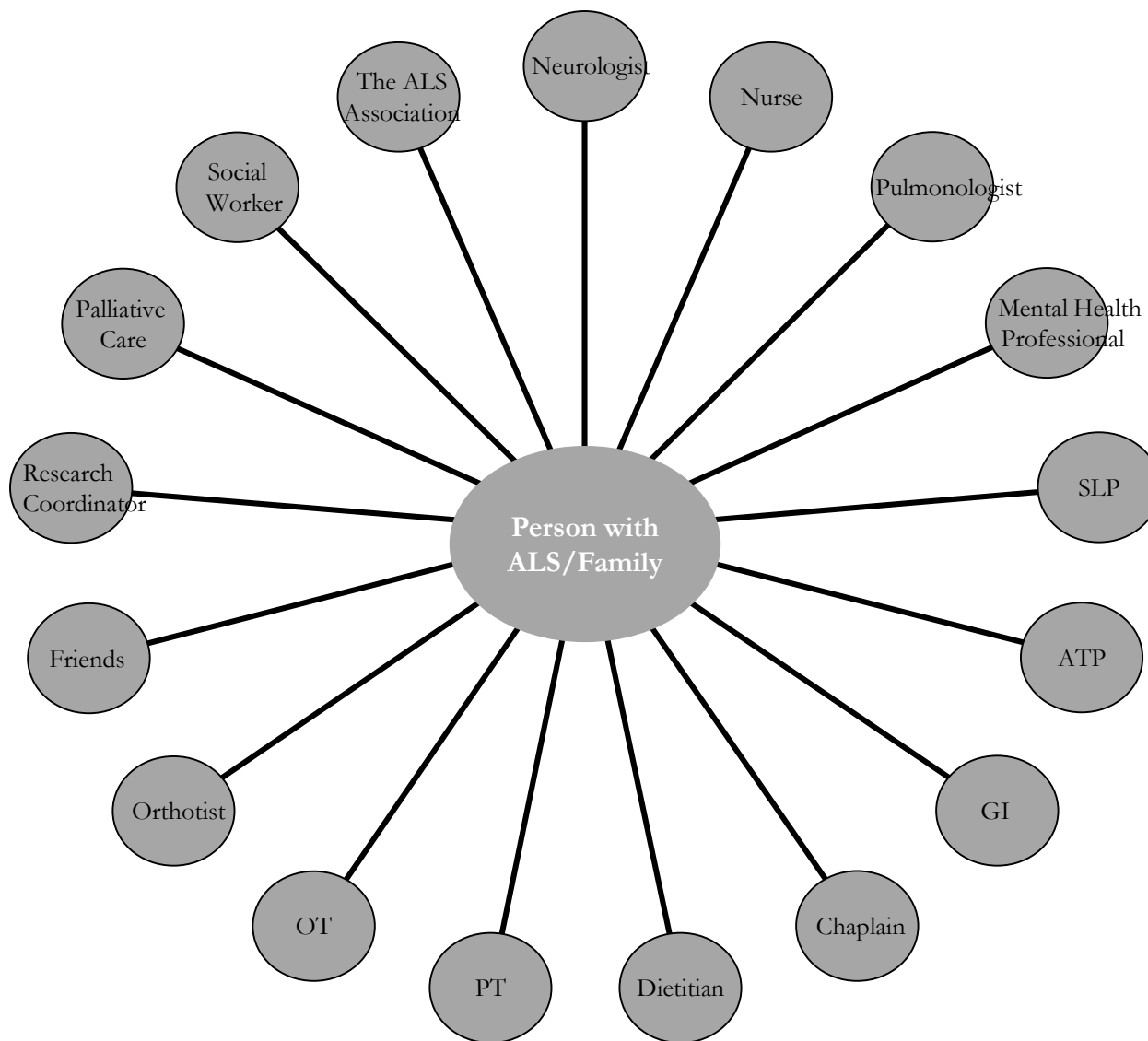
- Provide best-practice, multidisciplinary care to people living with ALS and their families
- Focus on independence and quality of life



What to Expect at an 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 Care Team



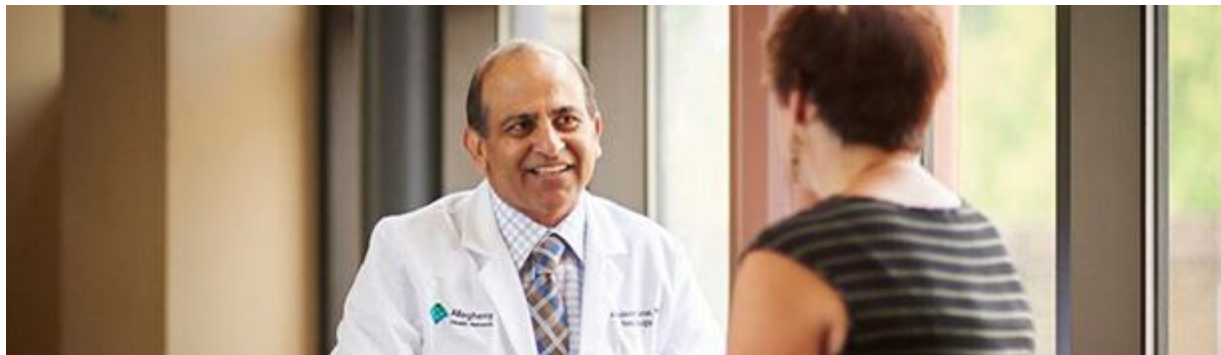
Neurologist

- Expert in diagnosis, evaluation and management of ALS
- Observe and assess for:
 - Fasciculations (twitching)
 - Brisk reflexes
 - Spasticity (stiffness/slowness)
 - Atrophy
 - Muscle strength



Neurologist

- 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

Neurologist

- Reviews patient history, performs neurological exam and related work-up, diagnoses and treats
- Develops care plan in conjunction with individual 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

Riluzole



FDA-approved drug for the treatment of ALS

- Oral medication taken twice a day
- Modest benefit
- Studies show that it can prolong life by 2-3 months
- Does not treat symptoms of ALS

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

ALS Treatment

1st Drug Approved to Treat ALS in 22 Years!



- Was used in Japan to treat stroke and approved for ALS in Japan & South Korea in 2015
- Manufactured in Japan by Mitsubishi Tanabe and distributed by MT Pharma America in the U.S.
- Drug expected to be available in U.S. in August
- Infusion drug

For More Information About Radicava

- www.Radicava.com: provide email and MT Pharma will send you updates
- For up-to-date FAQs and webinars:
www.alsa.org/research/radicava/
- Visit company website: www.mt-pharma-america.com/
- Contact **Searchlight Support program**:
1-844-SRCHLGT or 1-844-772-4548
 - for case management, bridge program and clinical educator support

ALS Treatment

- Symptom management
- Therapy
- Assistive devices/technologies
- Supportive care

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 with Yankauer catheter
- 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
- Contact 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 Chapter liaison to support continuity of care in between visits



Respiratory Therapist or Pulmonologist

- Educates people with ALS/families about how ALS effects breathing and the importance of early intervention
- 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, 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*

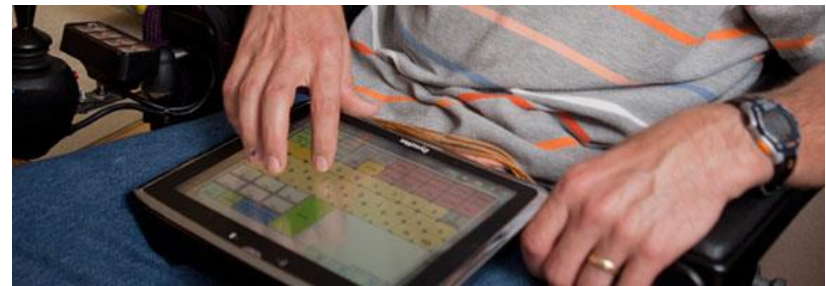
Dysarthria

- 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:
 - changes in nutrition seen in ALS
 - guidelines for caloric intake
 - importance of maintaining weight/good nutritional status
 - 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)



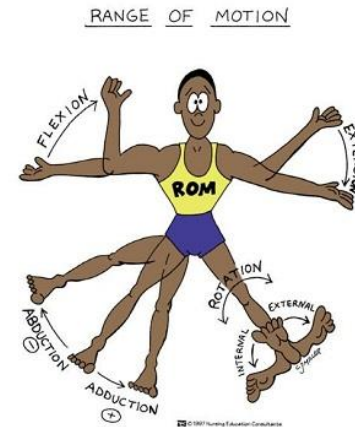
Constipation

- 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 for:
 - Pain
 - Weakness
 - Edema
 - Range of motion (ROM) limitations
- Goals:
 - increase ROM
 - reduce pain and/or edema in the upper extremities
 - prevent contractures
- Provides appropriate:
 - orthotics
 - positioning techniques
 - ROM exercises



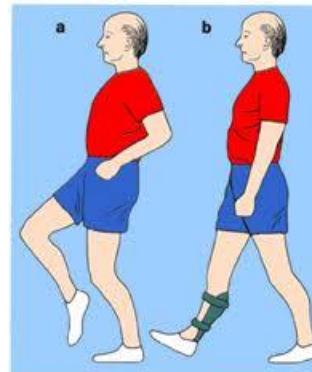
Occupational Therapist

- Assesses level of independence with daily activities
- Offers compensatory strategies to provide optimal performance in:
 - self-care/activities of daily living
 - energy conservation
 - upper extremity home exercise programs
- Offers options for assistive devices/adaptive equipment to assist with: feeding, bathing, grooming, dressing, toileting, work-related tasks and/or 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 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
 - Veins rely on muscles to return blood and fluid to heart
- 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 people with ALS/families



- Provides unique perspective into family dynamics
- Addresses a variety of concerns:
 - relationships
 - workplace issues
 - insurance
 - caregiving
 - financial problems
 - 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 about applying for benefits



The ALS Association Chapter Liaison

- Serves as a liaison between people living with ALS/families and 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
- 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 assessment



Coordinated Care

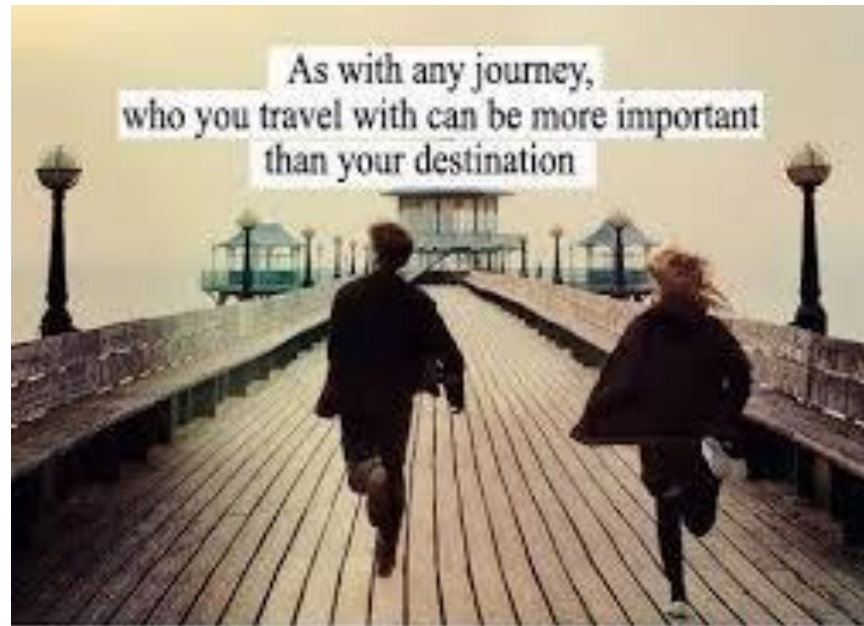
- ALS is a complex disease
- Over time, increased support required
- More than a neurologist is needed
- Multiple healthcare specialists and community providers involved in care plan
- Can be overwhelming and stressful on person with ALS and family
- Ongoing, clear communication and coordination is essential
 - 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 your care team

The journey is not an easy one, but each step can be a little easier when it's a team effort.



*Nicole Yarab, RN, BA
Senior Director, Clinical Programs
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nyarab@alsa-national.org

www.alsa.org