OCHSNER REGIONAL NEUROSCIENCES SYMPOSIUM

2026

ALS CLINIC ROUNDTABLE DISCUSSION

LEARNING OBJECTIVES

- Understand the concept and goals of multidisciplinary care.
- Understand the complexity of ALS care and management.
- Understand the vital roles of the ALS multidisciplinary team members including specialties outside of neurology.
- Be familiar with the clinical assessment tools used to monitor individuals with ALS.

DISCLOSURES

- Kristin M Johnson, DO
  - Argenx Advisory Board for CIDP
  - Argenx Speaker Bureau for Myasthenia Gravis
  - Amicus Advisory Board for Pompe Disease
- Stephen Kantrow, MD
  - No disclosures
- Mahmoud Sarmini, MD
  - No disclosures
- Dewitt Harrison, DO
  - No disclosures
- Gabi Diaz, PT
  - No disclosures
- Hayley Montgomery, OT
  - No disclosures
- Alicia Cantrell, SLP
  - No disclosures
- Deborah Ross, RDN
  - No disclosures
- Jena' Hampton, RN, BSN, BS
  - No disclosures

MULTIDISCIPLINARY CARE CLINICS (MDCS)

- Initial origins were in hematology and oncology in the 1950s focusing on hemophilia.
- The ALS Association began certifying centers for multidisciplinary care in 1989
- MDCs are composed of various medical specialty clinicians, nurses and allied health professionals who come together to assess patients at a shared clinic visit to provide patient centered care for complex conditions
- Benefits:
  - Comprehensive care coordination at one location fostering a holistic approach
  - Optimized treatment plans by having multiple experts collaborate at the time of care.
  - Reduce time between recognition of a problem and initiation of management/treatment
  - Improved patient experience by minimizing travel to multiple appointments thereby accomplishing more in less time
  - Reduce complications with real time collaboration amongst care team members
- Goals:
  - Improve quality of life
  - Improve patient outcomes
  - Enhance communication
  - Accelerate treatment plans for complex conditions

ALS MULTIDISCIPLINARY CARE CLINICS

- Certified Treatment Centers of Excellence, as designated by the ALS Association, are considered vital to managing the disease's complexity
- Criteria for these centers is specifically aligned with the American Academy of Neurology Practice Parameters and Quality Measures for ALS
- Primary goals are to provide symptomatic and palliative care to improve quality of life and survival for people with ALS
- Care provided is patient focused, but incorporates family/caregiver as part of the team
- Provide education
- Increase assess to research opportunities
- Assist with access to other organizational resources
- Minimize hospitalizations
- Establish goals of care

OCHSNER ALS MULTIDISCIPLINARY CLINIC

- ALS Certified Center of Excellence for 10 years
  - Audited yearly by the national ALS Association
- Valued collaborations with other organizations promoting quality of care and research in ALS
  - ALS Association of Louisiana/Mississippi
  - Team Gleason Foundation
  - NEALS (Network of Excellence for ALS)
  - Muscular Dystrophy Association
- Clinical Trials and Expanded Access Programs
- Community Engagement and philanthropy
- Participate in educational opportunities:
  - Improve recognition of potential ALS
  - Expedite referral to ALS specialists
  - Expand knowledge of ancillary providers in ALS management

OCHSNER ALS MULTIDISCIPLINARY CLINIC VISITS

- Pre Clinic Call
  - Clinic coordinator contacts the patient to explain the process, screen for new concerns, answer questions, help prepare necessary paperwork and medical records
- Visits
  - First visit (4 hours) – comprehensive baseline assessments with each specialist, initial care plan developed
  - Subsequent visits (3-4 hours) – focus on tracking progression, addressing new symptoms, adjusting the care plan
- Post Visit Wrap Up Meeting
  - Coordinate care and develop integrated recommendations
- Between visits
  - Clinic coordinator maintains contact with patient until care plan goals are completed
  - Clinic coordinator assists with triaging issues between clinic visits that require immediate attention from various team members

OCHSNER ALS MULTIDISCIPLINARY CLINIC TEAM

- Clinic Coordinator/Nurse
- Medical Assistant
- Neurologist
- Pulmonologist
- Physical Medicine and Rehabilitation Specialist
- Palliative Medicine Specialist
- Physical Therapist
- Occupational Therapist
- Speech Therapist
- Clinical Dietitian
- Respiratory Therapist
- Social Worker
- ALS Association Liaison
- Neuropsychologist*
- Geneticist*

*not part of routine MDC visits

PRESENT TODAY ARE SOME OF THE MEMBERS OF THE OCHSNER ALS MULTIDISCIPLINARY TEAM

TAKE A CLOSER LOOK AT HOW EACH OF US BRING OUR AREAS OF EXPERTISE TO THE MDC SETTING

NEUROLOGY – KRISTIN M JOHNSON, DO

- Initial role is recognition, diagnosis and referral to an ALS Multidisciplinary Clinic
- Education:
  - How the diagnosis is determined given it is a diagnosis of exclusion
  - Genetic vs spontaneous onset; initiate genetic testing and genetic consultation after discussion
- Monitor clinical exam changes, reported symptoms and ALS Functional Rating Scale from visit to visit to gauge rate of progression and determine appropriate recommendations/interventions
- Review currently available medication options to slow disease progression(riluzole and edaravone), prescribe as appropriate
- Review appropriate therapies for symptom management (such as pseudobulbar affect), prescribe as appropriate
- Provide resources if interested in participation in clinical trials
- Collaborate with members of the ALS Multidisciplinary team as well as supportive organizations including the ALS Association to provide evidence-based care and access to equipment to improve quality and quantity of life.

CLINIC COORDINATOR/NURSE – JENA' HAMPTON, RN, BSN, BS

- Role Overview
  - Central coordinator for patients
  - Integrates care across all disciplines, primary point of contact for patients, caregivers, and team
- Key Responsibilities
  - Clinic Management: Ongoing assessment, progression tracking, symptom identification
  - Therapeutics: Manage ALS Treatment (Riluzole, Radicava) and adherence
  - Care Coordination: Align neurology, respiratory, rehab, nutrition, and support services
  - Education & Support: Equp pALS/cALS; guide advance care planning
  - System Role: Optimize clinic flow, documentation, and research participation
- Strategic Value
  - Reduces fragmentation of care
  - Enables early intervention
  - Improves patient and caregiver experiences
  - Support quality, safety and outcomes
- Bottom Line
  - The RN is the operational and clinical hub- transforming complex ALS care into coordinated, patient-centered delivery.

PULMONOLOGY – STEPHEN KANTROW, MD

- Respiratory muscle weakness affecting sleep?
  - Difficulty breathing while lying on back
  - Poor sleep quality
- Abnormal use of respiratory muscles?
  - Accessory muscles of respiration in the neck
  - Abdominal movement during inspiration
- Is the cough strong enough?
  - Peak cough flow
  - Observed cough
  - Difficulty clearing secretions
- Can we prevent secretions that may be aspirated?
  - Saliva, allergic rhinitis

- Review pulmonary function tests
  - Spirometry
  - Maximum inspiratory pressure
- Insurance thresholds for coverage of Non-Invasive Ventilation
  - Forced Vital Capacity < 50%
  - Maximum Inspiratory Pressure - weaker than –60 cmH2O

- **How rapidly progressive is this version of ALS?**

PALLIATIVE MEDICINE - DEWITT HARRISON, DO

- Palliative care is an extra layer of support for patients with a serious illness
- In palliative care clinic, we focus on three things
  - Symptom management (Pain, nausea, fatigue, Depression, Anxiety)
  - Communication
  - Planning/Discussing future treatment decisions
    - Where most of our focus is spent in ALS clinic
    - Who do you trust to make decisions on your behalf?
    - Willingness to undergo artificial life support?

PHYSICAL MEDICINE & REHABILITATION - MAHMOUD SARMINI, MD

- Physical Medicine & Rehabilitation: an integral part of ALS multidisciplinary Clinic.
- ALS is predominantly a neurological motor dysfunction, but is also a multi-system disorder.
- Focus of PM&R is mostly:
  - Treating pain (highly prevalent)
  - Improving quality of life.
  - Assisting in prescribing adaptive devices
  - Initiating referrals for interventions such as axial or peripheral joint injections.
- Pain in patients with ALS can be primary or secondary:
  - Primary pain: may include muscle cramps, spasticity, neuropathic pain (uncommon in ALS).
  - Secondary pain: may include pain from chronic low back pain, chronic neck pain, peripheral joint pain.. aggravated by weakness and immobility in ALS patients.
  - Central sensitization: proposed as an additional mechanism

PHYSICAL THERAPY - GABI DIAZ, PT

- Primary role is to assess for deficits with gross mobility (transfers, walking, stairs, wheelchair mobility)
- Screen for orthotics (ex: AFO) and assistive devices (walker/rollator, cane, etc.)
- Determine need for more supportive equipment including power wheelchairs, hospital beds, and Hoyer lifts
- Education (with coordination from OT) on the following topics:
  - Adaptive strategies
  - Transfer training safety
  - Energy conservation
- Review patient's functional goals and provide any resources to maximize independence while keeping safety at the forefront

OCCUPATIONAL THERAPY - HAYLEY MONTGOMERY, OT

- Assess for deficits in Activities of Daily Living (bathing, dressing, toileting, grooming)
- Assess for deficits in Instrumental Activities of Daily Living (medication management, preparing meals, household maintenance, etc)
- Identify significant changes in upper extremity strength, ROM, coordination
- Provide education and recommendations for supporting functional independence
  - Durable Medical Equipment (examples: bedside commode, shower chair, tub transfer bench)
  - Adaptive Equipment (examples: dressing equipment, toileting aids, feeding equipment, splints)
  - Coordinate with PT to educate in energy conservation and safety strategies with functional mobility

SPEECH THERAPY - ALICIA CANTRELL, SLP

- Primary role is to assess speech, communication, and swallowing function
- Educate about changes in speech clarity and volume, breath support for speech, and swallow function
- Educate on compensatory strategies for speech and swallowing
- Assess and educate the need for Augmentative-Alternative Communication (AAC) to supplement speech as intelligibility declines and speech becomes nonfunctional. Discuss the options of AAC as the disease progresses.
- Educate on voice preservation (voice banking and message banking) in early phase of disease
- Complete a clinical swallow evaluation in the clinic and discuss diet recommendations and swallowing compensatory strategies as needed.
- Recommend further testing for swallow function such as MBSS or FEES as needed. Review patient's goals for alternative means of nutrition if needed

CLINICAL DIETITIAN - DEBORAH ROSS, RDN

- Primary Role is to develop a personalized, nutrition-based treatment plan, with the goal to help a person manage their health conditions and enhance their well-being through nutrition provided in partnership with the health care team.
- This includes assessing a person's health status, medical history and eating patterns; identifying any dietary issues; and creating a personalized nutrition plan.
- Provide nutrition support based on individual needs, including family/friends in discussions as indicated. This may include modified textures, increased calorie/protein needs, nutrition education, enteral nutrition.
- In the course of follow-up clinic visits, making adjustments to the nutrition plan as indicated by changes in condition.

CASE ONE

72 YEAR OLD FEMALE, 3RD CLINIC VISIT

72 YEAR OLD FEMALE

- HPI:
  - Initially presented early 2025 with hoarse voice which progressively worsened to more notable dysarthria with increasing difficulty swallowing. Concerns included taking longer to complete a meal and 15# weight loss. No improvement with speech therapy. Late 2025 she developed left leg weakness, slow gait, declining balance, difficulty with stairs. Referred to neurology and diagnosed with ALS by clinical exam with supportive findings on EMG and an elevated neurofilament light chain. Subsequently referred to ALS clinic. Now on liquid riluzole and edaravone.
- Current concerns since last visit:
  - Worsening left arm and leg weakness
  - Increasing difficulty swallowing with episodes of choking on liquids and solids
  - Worsening dysarthria, requires repetition, unable to amplify voice
  - Excess saliva and drooling, not controlled by scopolamine patch
  - Unsteady gait with limited ability to use a walker
  - Increasing shortness of breath, including while speaking, weakened cough
  - Some increased crying and laughing but not felt to be bothersome. Does not report depressive symptoms
  - Pain in the right hip which has increased since her last visit

ALS FUNCTIONAL RATING SCALE

Item 1: SPEECH

4 Normal speech process

3 Detectable speech disturbance

2 Intelligible with repeating

1 Speech combined with non-vocal communication

0 Loss of useful speech

Item 2: SALIVATION

4 Normal

3 Slight but definite excess of saliva in mouth; may have nighttime drooling

2 Moderately excessive saliva; may have minimal drooling (during the day)

1 Marked excess of saliva with some drooling

0 Marked drooling; requires constant tissue or handkerchief

Item 3: SWALLOWING

4 Normal eating habits

3 Early eating problems – occasional choking

2 Dietary consistency changes

1 Needs supplement tube feeding

0 NPO (exclusively parenteral or enteral feeding)

Item 4: HANDWRITING

4 Normal

3 Slow or sloppy: all words are legible

2 Not all words are legible

1 Able to grip pen, but unable to write

0 Unable to grip pen

Item 5a: CUTTING FOOD AND HANDLING UTENSILS Patients without gastrostomy use 5b if >50% is through g-tube

4 Normal

3 Somewhat slow and clumsy, but no help needed

2 Can cut most foods (>50%), although slow and clumsy; some help needed

1 Food must be cut by someone, but can still feed slowly

0 Needs to be fed

Item 5b: CUTTING FOOD AND HANDLING UTENSILS gastrostomy present and is the primary method (more than 50%) of eating

4 Normal

3 Clumsy, but able to perform all manipulations independently

2 Some help needed with closures and fasteners

1 Provides minimal assistance to caregiver

0 Unable to perform any aspect of task

ALS FUNCTIONAL RATING SCALE

Item 6: DRESSING AND HYGIENE

4 Normal function

3 Independent and complete self-care with effort or decreased efficiency

2 Intermittent assistance or substitute methods

1 Needs attendant for self-care

0 Total dependence

Item 7: TURNING IN BED AND ADJUSTING BED CLOTHES

4 Normal function

3 Somewhat slow and clumsy, but no help needed

2 Can turn alone, or adjust sheets, but with great difficulty

1 Can initiate, but not turn or adjust sheets alone

0 Helpless

Item 8: WALKING

4 Normal

3 Early ambulation difficulties

2 Walks with assistance

1 Non-ambulatory functional movement

0 No purposeful leg movement

Item 9: CLIMBING STAIRS

4 Normal

3 Slow

2 Mild unsteadiness or fatigue

1 Needs assistance

0 Cannot do

Item 10: DYSPNEA

4 None

3 Occurs when walking

2 Occurs with one or more of the following: eating, bathing, dressing (ADL)

1 Occurs at rest: difficulty breathing when either sitting or lying

0 Significant difficulty: considering using mechanical respiratory support

Item 11: ORTHOPNEA

4 None

3 Some difficulty sleeping at night due to shortness of breath, does not routinely use more than two pillows

2 Needs extra pillows in order to sleep (more than two)

1 Can only sleep sitting up

0 Unable to sleep without mechanical assistance

ALS FUNCTIONAL RATING SCALE

Item 12: RESPIRATORY INSUFFICIENCY

4 None

3 Intermittent use of BiPAP

2 Continuous use of BiPAP during the night

1 Continuous use of BiPAP during day & night

0 Invasive mechanical ventilation by intubation or tracheostomy

Total: 30

72 YEAR OLD FEMALE – NEUROLOGY ASSESSMENT

Exam (pertinent findings):

- Cognitive: normal
- Speech: severely dysarthric, but fluent with appropriate responses
- Cranial nerve exam: moderate to severely weak tongue, significant atrophy and fasciculations; remainder of CN are normal.
- Gait: slow, unsteady, unable to ambulate without walker
- Tone: normal
- Coordination: LUE pronator drift, no ataxia in the upper or lower limbs
- Sensory: normal exam
- Strength:
  - Neck: normal
  - RUE: Delt 4/5; WE 5-/5; remainder normal
  - LUE: Delt 3/5; Bi, Tri, WE, WF, FF, 4/5; FE and IO 3/5;
  - RLE: HF, HAbd, DF 4/5; remainder normal
  - LLE: HF, DF, PF 3/5; Habd, KF 4/5; remainder normal
- Reflexes:
  - BUE: normal
  - BLE: hyper-reflexic
  - Jaw Jerk: absent

72 YEAR OLD FEMALE – NEUROLOGY ASSESSMENT

- ALS: Review liquid riluzole and liquid edaravone, tolerating well so continue both to slow disease progression, CBC and CMP to monitor nutrition, hydration, and potential medication side effects, field any new questions including any questions regarding research in ALS
- Progressive weakness: continue use of walker in combination with AFO, continue use of portable power wheelchair for longer distances, when fatigued or concerns related to difficult terrain, take note of increasing hand weakness interfering with daily activities; collaborate with PT and OT on care plan
- Progressive dysphagia: increased time to eat, weight loss and early signs of neuromuscular respiratory weakness noted, discussed plans for feeding tube placement; collaborate with pulmonology, ST and dietician on a care plan
- Progressive dysarthria: discussed augmentive/alternative communication, currently using iPad and phone with speech assistance app and completing voice preservation; collaborate with ST on care plan
- Progressive dyspnea, weakened cough, difficulty with secretion management, no clear orthopnea: discuss other medications such as atropine drops, glycopyrrolate, and botulinum toxin as options for management of excess oral secretions; collaborate with pulmonology on a care plan
- Early signs of pseudobulbar affect: discussed options for management including sertraline, dextromethorphan/quinidine sulfate, or no treatment.
- Musculoskeletal pain: review that while pain is not a direct result of ALS, there can be secondary pain with disease progression as well as pain from pre-existing conditions; collaborate with PT, OT and PM&R to manage musculoskeletal issues
- Goals of care review: collaborate with the MDC team, in particular palliative medicine

72 YEAR OLD FEMALE - PULMONARY

- Weak cough – using mechanical cough device for help clearing secretions
  - Peak Cough Flow 78 lpm (consider if < 300 lpm)
- Prevent secretions that may be aspirated
  - Treat excessive saliva – glycopyrrolate
  - Treat allergic rhinitis – fluticasone and azelastine spray
- Risk assessment for gastrostomy in interventional radiology
  - Orthopnea? - would plan non-invasive ventilation during the procedure
  - Saliva – all are pretreated with glycopyrrolate for the procedure to avoid stimulating salivation
- Schedule pulmonary function testing in lab to include spirometry, maximum inspiratory pressure to evaluate/qualify for non-invasive ventilation
  - Forced Vital Capacity was 78% of predicted 3 months ago (~ one year after disease onset)

72 YEAR OLD FEMALE - PALLIATIVE CARE

- Who is she outside of ALS
  - She lives at home with her husband of 22 years
  - Loves to attend pelicans/saints games
  - Loves to gather with her family; grandchildren call her MawMaw
  - Dignity Therapy Questions:
    - Tell me a little about your life history, particularly the parts that you either remember most, or think are the most important. When did you feel most alive?"
      - "The day I married their dad. 1971, May 29. Justice of the peace.

- - She wants her children to learn how to cope with her ALS progression when she can no longer care for herself
- - Patient has established general power of attorney that includes healthcare decisions naming her daughter,
- - Strong religious faith, member of local Catholic church
- - Family has experience with medical decision-making through her children's father's end-of-life care in March 2025 due to sepsis. Recalls a positive experience with palliative care in the hospital

72 YEAR OLD FEMALE – PHYS. MED. & REHAB.

- Symptoms:
  - No significant neck pain
  - Mild non-radicular low back pain
  - Mild bilateral hip pain.
  - Painful cramps in BLE, mostly at night.
- Physical Exam::
  - Diffuse weakness
  - No increased tone.
  - Knee crepitus.
- Imaging studies:
  - MRI of cervical spine (9/2025): Mild-to-moderate DJD, facet arthropathy, mild-to-moderate foraminal and spinal canal stenosis at C3-6.
  - X-rays of lumbar spine (4/2026): unremarkable
  - X-rays of hips (4/2026): unremarkable
- Treatment:
  - Tylenol p.r.n.
  - Baclofen p.r.n.
  - Lidocaine patches p.r.n.
  - Diclofenac patches p.r.n.

72 YEAR OLD FEMALE - PT/OT

Physical Therapy

- Strength, ROM, and Mobility
  - Strength – grossly 4/5 in BLE
  - Mobility – using SBQC for gait, transfers need CGA-min A
- Outcome Measures (balance)
- Problems reported: fatigue with ADLs, balance issues
- Placed a referral to outpatient neuro PT services for focus on balance training
- Provided education re: energy conservation

Occupational Therapy

- Maintained modified independence with ADLs/IADLs
  - Adaptive techniques for managing fatigue levels
- Progressive, but slow decline in LUE strength, AROM, coordination
  - Left thenar atrophy
- Current equipment needs – standard shower chair and grab bar (walk-in shower)
- Education in energy conservation
- No recommendation for outpatient or home health OT at this time

72 YEAR OLD FEMALE – SPEECH THERAPY

Motor speech skills assessment: Severe mixed spastic-flaccid dysarthria

Oral Motor/Cranial Nerve Exam and Respiration/Phonation Assessment: Severe oral-motor weakness, reduced breath support for speech

Communicative Effectiveness Survey: Family and patient reported less effectiveness (50% - 1, 50% - 2/3 with familiar people)

Use of AAC and compensatory speech strategies: Speech Assistant AAC app in addition to speaking and writing. Voice banking with Team Gleason.

Eating Assessment Tool (EAT-10): 16/40. Increased difficulty swallowing solids, liquids, and pills reported.

Clinical Swallow Evaluation: Coughing when drinking water, water came through her nose. Severe oral-pharyngeal dysphagia. Getting a peg tube.

ALS Speech Severity Scale: **Stage 4:** Use of Alternative/Augmentative Communication (AAC): Intelligibility problems need to be resolved by writing or a spokesperson. The speaker may initiate communication nonverbally. **ALS Severity Scale: 3 or 4**

Recommendations: Continue using AAC. No further ST needs, using strategies independently. Monitor for need of Speech Generating Device with eyegaze access method. Use peg tube for primary means of nutrition with oral pleasure feedings.

72 YEAR OLD FEMALE - DIETITIAN

- 10/15/2025 initial ALS clinic visit; pt was doing her own grocery shopping and cooking. stated she had a good appetite, but endorsed unintentional weight loss.
- 01/14/2026 2nd CV: pt with continuing unintentional weight loss despite stating "I have a good appetite". Gastrostomy placement recommended. G-tube was placed with NOMH IR DOSC on 4/13/2026.
- With unintentional weight loss secondary to difficulty swallowing and dx of ALS, pt's primary source of nutrition will be via G-tube.
- Recommendation for enteral nutrition Isosource 1.5 (or equivalent) made based on her calorie needs: 1400-1600 kcal (35-40 kcal/kg/day); protein needs: 60 gm (1.5 gm/kg/day); fluid needs: ~1500 ml (1 ml/kdal)
- 4-250 ml cartons Isosource 1.5 daily will provide 1500 kcal, 68 gm protein, 15.2 gm dietary fiber, 764 ml free water + additional water from flushes

- Weight history:
  - 04/08/2026: 40 kg (88 lb 2.9 oz) - standing scale
  - 01/14/2026: 44.1 kg (97 lb 3.6 oz) - standing scale
  - 10/15/2025: 45.5 kg (100 lb 1.4 oz) - standing scale

72 YEAR OLD FEMALE – RN CLINIC COORDINATOR

- RN Clinical Impression
  - Rapid disease progression with high-risk for respiratory compromise, aspiration, malnutrition, and falls
- Priority Interventions
  - Airway/Breathing:
    - Schedule Pulmonary function testing (FVC/MIP)
    - Forward orders for NIV, cough assist, suction if qualify
  - Swallow/Nutrition:
    - Schedule swallow study (MBSS/FEES)
  - PEG tube discussion/referral
    - Schedule for CT of abdomen
    - Contact IR to schedule consult and procedure
    - Send orders for Home Health for in-home assessment and education
    - Coordinate with pharmacy RD/RN for bedside teaching
  - Dietitian for caloric optimization
    - Submit orders for enteral nutrition to specialty pharmacy
    - Follow up with patient to ensure tolerating formula
  - Communication:
    - Encourage patient to utilize AAC device
    - Follow up with patient and caregiver regarding need for eye gaze
    - Caregiver communication strategies
  - Mobility/Safety:
    - Follow up with patient regarding HH PT/OT
      - Once PEG Tube healed switch to Outpatient PT/OT
    - Fall prevention & home safety
    - Follow up with patient to verify receipt of shower chair order via ALSA and grab bars installed
  - Secretion Management:
    - Follow up with patient regarding medication effectiveness (glycopyrrolate/atropine)
    - Discuss with Neurologist & Pulmonologist if oral medication are not effective
    - Consider botulinum toxin referral
      - Place prior authorization order for pre-service & coordinate with MA for scheduling
  - Pain Management:
    - Follow up with patient regarding pharmacologic plan

CASE TWO

55 YEAR OLD FEMALE, 5TH CLINIC VISIT

55 YEAR OLD FEMALE

- HPI:
  - Onset winter 2022 with left hand weakness and muscle atrophy. Initially with C4-6 neck surgery in 2023 followed by cubital tunnel release without improvement. Later in 2023 the right hand was becoming weaker with atrophy developing. She was again referred to surgery and underwent C7-T2 surgery. In the winter of 2023 she developed left foot drop. She was referred to neurology to be evaluated. ALS was diagnosed based on clinical exam findings and a referral was placed to ALS clinic. Since then she has been taking riluzole and oral edaravone.
- Current concerns since last visit:
  - Significant weakness in both hands, weakness in the arms, bilateral foot drop but AFOs help as well as walking sticks
  - Reports voice is getting weaker, occasionally episodes of voice giving out
  - Is focusing more on chewing, but has made no adjustments in her diet and does not report concerns related to choking or weight loss
  - Sleep can be disrupted by need to use the bathroom and restless leg symptoms
  - Reports episodes of laryngospasm becoming more frequent, identifies any irritation is a trigger including certain foods/spices. Has found lorazepam helpful
  - Voices frustration with her changing abilities but does not report depression

ALS FUNCTIONAL RATING SCALE

Item 1: SPEECH

4 Normal speech process

3 Detectable speech disturbance

2 Intelligible with repeating

1 Speech combined with non-vocal communication

0 Loss of useful speech

Item 2: SALIVATION

4 Normal

3 Slight but definite excess of saliva in mouth; may have nighttime drooling

2 Moderately excessive saliva; may have minimal drooling (during the day)

1 Marked excess of saliva with some drooling

0 Marked drooling; requires constant tissue or handkerchief

Item 3: SWALLOWING

4 Normal eating habits

3 Early eating problems – occasional choking

2 Dietary consistency changes

1 Needs supplement tube feeding

0 NPO (exclusively parenteral or enteral feeding)

Item 4: HANDWRITING

4 Normal

3 Slow or sloppy: all words are legible

2 Not all words are legible

1 Able to grip pen, but unable to write

0 Unable to grip pen

Item 5a: CUTTING FOOD AND HANDLING UTENSILS Patients without gastrostomy use 5b if >50% is through g-tube

4 Normal

3 Somewhat slow and clumsy, but no help needed

2 Can cut most foods (>50%), although slow and clumsy; some help needed

1 Food must be cut by someone, but can still feed slowly

0 Needs to be fed

Item 5b: CUTTING FOOD AND HANDLING UTENSILS gastrostomy present and is the primary method (more than 50%) of eating

4 Normal

3 Clumsy, but able to perform all manipulations independently

2 Some help needed with closures and fasteners

1 Provides minimal assistance to caregiver

0 Unable to perform any aspect of task

ALS FUNCTIONAL RATING SCALE

Item 6: DRESSING AND HYGIENE

4 Normal function

3 Independent and complete self-care with effort or decreased efficiency

2 Intermittent assistance or substitute methods

1 Needs attendant for self-care

0 Total dependence

Item 7: TURNING IN BED AND ADJUSTING BED CLOTHES

4 Normal function

3 Somewhat slow and clumsy, but no help needed

2 Can turn alone, or adjust sheets, but with great difficulty

1 Can initiate, but not turn or adjust sheets alone

0 Helpless

Item 8: WALKING

4 Normal

3 Early ambulation difficulties

2 Walks with assistance

1 Non-ambulatory functional movement

0 No purposeful leg movement

Item 9: CLIMBING STAIRS

4 Normal

3 Slow

2 Mild unsteadiness or fatigue

1 Needs assistance

0 Cannot do

Item 10: DYSPNEA

4 None

3 Occurs when walking

2 Occurs with one or more of the following: eating, bathing, dressing (ADL)

1 Occurs at rest: difficulty breathing when either sitting or lying

0 Significant difficulty: considering using mechanical respiratory support

Item 11: ORTHOPNEA

4 None

3 Some difficulty sleeping at night due to shortness of breath, does not routinely use more than two pillows

2 Needs extra pillows in order to sleep (more than two)

1 Can only sleep sitting up

0 Unable to sleep without mechanical assistance

ALS FUNCTIONAL RATING SCALE

Item 12: RESPIRATORY INSUFFICIENCY

4 None

3 Intermittent use of BiPAP

2 Continuous use of BiPAP during the night

1 Continuous use of BiPAP during day & night

0 Invasive mechanical ventilation by intubation or tracheostomy

Total: 33

55 YEAR OLD - NEUROLOGY ASSESSMENT

Exam (pertinent findings)

- Cognitive: normal
- Speech: mildly dysarthric, easily understandable
- Cranial nerve exam: normal
- Gait: assisted by bilateral AFOs, slightly unsteady
- Tone: normal
- Coordination: no ataxia in the upper or lower extremities
- Sensory: normal exam
- Strength:
  - Neck: normal
  - RUE: Tri, WE, WF, FE 4/5; IO 3/5, remainder 5/5
  - LUE: Delt, Bi, Tri, WF, FF, 4/5; WE, FE 3/5; and IO 1/5;
  - RLE: DF 3/5; PF 4/5; remainder normal
  - LLE: HF, KF 4/5; DF 0/5, PF 3/5; remainder normal
- Reflexes:
  - RUE: normal
  - LUE: hyper-reflexic, with positive Hoffman's reflex
  - BLE: hyper-reflexic
  - Jaw Jerk: absent

55 YEAR OLD – NEUROLOGY ASSESSMENT

- ALS: Review liquid riluzole and liquid edaravone, tolerating well so continue both to slow disease progression; CBC and CMP to monitor nutrition, hydration, and potential medication side effects; field any new questions including any questions regarding research in ALS
- Progressive weakness: Encouraged continued use of AFOs, but increasing hand weakness noted which could eventually interfere with holding a cane or walker; collaborate with PT and OT
- Early voice changes reported: No current communications issues; collaborate with Speech Therapy
- Early chewing difficulties: Denies any need for food modification or concerns regarding weight loss; collaborate with Speech Therapy and Dietician
- Difficulty falling asleep: does not feel night time bathroom breaks warrant intervention, but is agreeable to ropinirole for RLS management and will continue amitriptyline for insomnia; collaborate with PM&R to manage
- Laryngospasm: increasing in frequency, but episodes respond to lorazepam; collaborate with pulmonology to manage
- Mood changes: increasing frustration due to changing abilities with muscle weakness, though denies depression; collaborate with palliative medicine
- Goals of care: collaborate with the MDC team, in particular palliative medicine

55 YEAR OLD FEMALE - PULMONARY

- Disease onset 2022
- Episodes of laryngospasm
- No orthopnea
- Respiratory rate 12
- Mild accessory muscle use with deep breath, normal abdominal movement
- Forced vital capacity 73% (at 4 years post onset)
- Peak cough flow 193 lpm
- Observed cough weak

- Ordered a mechanical cough assist device
- Avoid laryngospasm triggers – spicy food, treat with lorazepam if severe

55 YEAR OLD FEMALE - PALLIATIVE CARE

Who is she outside of her diagnosis?

- Retired Elementary Math Teacher
- Grandkids very important, she goes by "big momma"
- Traveled to see as many MLB parks as possible, saw Fenway park shortly after ALS diagnosis

- Went on a church group trip to Greece in January ~ 6 months after ALS diagnosis, Followed the footsteps of Paul's missionary journey
- "What have you learned about life that you would want to pass along to others? What advice or words of guidance would you wish to pass along to your (son, daughter, husband, wife, parents, others)?"
  - Remove the illusion of time. Prioritize, do what matters. Make the moment count. Don't worry about work. When you leave work you are replaced the next day. But your close group, your memories, things you have done, those are the things that matter. Also matters (maybe more) the things you haven't done. Try to live with no regrets, just memories.

PALLIATIVE CARE – GOALS OF CARE/ADVANCED CARE PLANNING

- - Goals include maintaining quality of life and continuing activities such as gardening, fishing, and attending baseball games
- - wants care as long as they have quality of life, but don't want life-sustaining measures if unable to communicate or participate in their own life
- - Expressed willingness to continue living as long as she maintains awareness of loved ones and some form of communication (smile, nod, wink)
- - Does not want prolonged life if she loses recognition of family or ability to interact meaningfully
- - Patient and husband have ongoing discussions about preferences and have experience with advance care planning
- - Living will to be drafted with their family attorney, reflecting communication-based quality of life criteria

72 YEAR OLD FEMALE – PHYS. MED. & REHAB.

- Symptoms:
  - H/O C4-6 ACDF 2023, C7-T1 laminectomy 1/2024
  - Chronic low back pain with right radiculopathy. Well-controlled.
  - Chronic left hip pain
  - No neck pain.
  - Painful cramps in BLE, several times per day, not painful.

- Physical Exam::
  - BLE weakness
  - No increased tone.
  - -ve SLR.

- Imaging studies:
  - CT of cervical spine (5/2025): Prior C4-6 ACDF.. Severe C6-7 DJD. Multilevel facet arthropathy

- Treatment:
  - Tylenol p.r.n.
  - Ibuprofen p.r.n.
  - Cyclobenzaprine p.r.n.
  - Amitriptyline p.r.n. for sleep.

55 YEAR OLD FEMALE - PT/OT

Physical Therapy

- Strength, ROM, and Mobility
  - Strength – initially 5/5 in BLE except left ankle
  - Mobility – maintained independence with gait, transfers
- Outcome Measures (balance and mobility)
- Problems reported: left foot catching occasionally when walking
- Recommendations: left prefab AFO

Occupational Therapy

- Min A for ADLs/IADLs - difficulty with fine motor tasks
  - Adaptive techniques used – clothing without fasteners
- Symptom onset was left hand weakness
  - Progressive decline in UE weakness (distal more than proximal)
- Current equipment needs - Left resting hand splint, Tub transfer bench
- Education in energy conservation
- Outpatient OT completed in 2023, need for additional OT not indicated at this time

55 YEAR OLD FEMALE – SPEECH THERAPY

Motor speech skills assessment: Mild mixed spastic-flaccid dysarthria

Oral Motor/Cranial Nerve Exam and Respiration/Phonation Assessment: decreased sequential and alternating motion rates; mild oral-motor weakness, reduced breath-speech coordination

Communicative Effectiveness Survey: Family and patient reported effectiveness score of 4 (no difficulty)

Use of AAC and compensatory speech strategies: Clear speech strategies trained. Completed voice banking with Team Gleason.

Eating Assessment Tool (EAT-10): 16/40. Increased difficulty swallowing solids and pills reported; uses strategies

Clinical Swallow Evaluation: No objective swallow study recommended - managing with compensatory strategies.

ALS Speech Severity Scale: **Stage 2**: Detectable Speech Disturbance: Speech remains easily understood, but changes are noted by others, especially during fatigue or stress. **ALS Severity Scale: 7 or 8 (decline since last clinic)**

Recommendations: Outpatient ST to educate on speech strategies, The Breather, swallowing precautions/strategies. Use The Breather device 10 sets 2 times per day, if too fatigue, stop using the device

55 YEAR OLD FEMALE - DIETITIAN

- Initial ALS CV pt reported following regular diet, chewing food carefully, more liquid to help with swallowing and taking smaller bites.
- 2/12/25 CV: avoiding gluten and sugar, focusing on gut health. Recommendation made to try plant-based po supplement and increase high calorie po intake. is tracking her weight at home.
- Subsequent CV's: pt continues to state her appetite is "good" and she's "eating more". Has introduced some gluten back into her diet. She and her husband not concerned with weight loss. I again encouraged high calorie po intake.

- Weight history:
  - 11/05/2025: 49.4 kg (108 lb 14.5 oz) - standing scale, BMI=19.3
  - 08/06/2025: 51.9 kg (114 lb 6.7 oz) - standing scale
  - 05/07/2025: 52.5 kg (115 lb 11.9 oz) - standing scale
  - 02/12/2025: 53.6 kg (118 lb 2.7 oz) - standing scale
  - 04/24/2024: 63.5 kg (140 lb) BMI=24.8

55 YEAR OLD FEMALE – RN CLINIC COORDINATOR

- RN Clinical Impression
  - Progressive limb decline with emerging bulbar symptoms → rising risk for airway compromise, falls, and functional loss
- Priority Interventions
  - Airway/Bulbar:
    - SLP referral (voice + baseline swallow)
    - Laryngospasm education (breathing techniques, trigger avoidance)
  - Respiratory Surveillance:
    - Monitor for early respiratory symptoms
  - Mobility/Function:
    - PT/OT: hand splints, ADL adaptations, gait reassessment
    - Fall prevention & home safety
      - Ensure patient are utilizing equipment and following PT/OT recommendations
  - Nutrition (Preventative):
    - Dietitian consult; monitor for subtle dysphagia
  - Sleep Optimization:
    - Address restless legs & nocturia; reinforce sleep hygiene
    - Assess patient taking medication to help alleviate symptoms

AFTER CLINIC DAY

- Potential procedures:
  - Labs
  - Imaging
  - Swallow study
  - Feeding tube placement
  - Tracheostomy placement
- Potential appointments:
  - Voice banking, message banking
  - AAC evaluation
  - Wheelchair assessment
  - Outpatient ST/PT/OT
  - Home ST/PT/OT
- Potential consultations:
  - Neuropsychology
  - Genetics
  - Psychology
- Potential equipment:
  - Grant requests
  - DME prescriptions
  - Equipment loans from supportive organizations
  - Equipment donations from supportive organizations

QUESTIONS?