WELCOME!

January 30, 2023

Features and Benefits of ALS
Multidisciplinary Care

Guest Speaker:
Lauren Tabor Gray, SLP, PhD
Assistant Professor Nova Southeastern University
CO-Director, ALS Clinic NSU Neuroscience Institute
Optimizing ALS Care: The Multidisciplinary ALS Clinic

Lauren Tabor Gray, PhD
Assistant Professor
Co-Director, NSU Health ALS Clinic
1/30/2023
Amyotrophic Lateral Sclerosis

A: No
myo: Muscle
*trophic*: nourishment

**Lateral**: the area of nerve cells affected (both upper *and* lower motor neurons)

**Sclerosis**: hardening that occurs as motor neurons degenerate
ALS Pathology

UMN
Cortical Motor Neurons
Corticospinal/Bulbar Tracts
Supranuclear Symptoms
- Spasticity
  - Muscle Stiffness
  - Muscle Slowness
  - Hyperreflexia
  - Decreased speed

LMN
Brainstem Cranial Nerve Nuclei
Bulbar Palsy
- Atrophy
  - Flaccid Paresis
  - Decreased Strength
  - Decreased Force
  - Fasciculations

Plowman (2015) JSLHR.
Motor Neuron Diseases

ALS
- Spinal-Onset Variant
- Bulbar-Onset Variant

ALS
- Primary Muscle Atrophy

PMA

PLS
- Primary Lateral Sclerosis

Lower Motor Neuron

Upper Motor Neuron

Swinnen, 2014
What does evidence-based care look like for pALS?
Interdisciplinary & Collaborative

- Patients as Partners
- Friends and Family
- Caregivers
- Primary Care Doctors
- Hospital and Emergency Room
- Palliative and Hospice
- Home and Outpatient therapists
- Home Health agencies
The Mission: Comprehensive Care

- Multidisciplinary clinics
- Multidisciplinary Telehealth appointments
- Mobilize Community Resources: Partnership with ALSA, MDA, Always for ALS, local and national
- Advocacy: Clinical Research, ALSA, ALS TDI, Legislation, Patient Ambassadors (CRLI)
Your Care Team

- Neurology
- Nurse Practitioner
- Nurse/Clinic Coordinator
- Speech Language Pathologist
- Occupational Therapy
- Physical Therapy
- Dietitian
- Social Work/ALSA Liaison
- Assistive Technology Specialist
- Neuropsychologist
- Palliative Care Team
- Research
Impact on Life and Function

- Breathing
- Swallowing
- Quality of Life
- Cognition
- Mobility
- Communication
ALS: Nuts and Bolts

- Important Disease Characteristics influencing care:
  - Onset Type (where symptoms began)
  - Body Mass Index (BMI, changes in weight)
  - Degree of respiratory involvement
  - Family History:
    - ALS, Motor neuron disease
    - Psychiatric illness, dementia
ALS progresses at different rates

### Patient Demographics

<table>
<thead>
<tr>
<th>Predictors</th>
<th>Data</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at onset (years)</td>
<td>66.0</td>
</tr>
<tr>
<td>Diagnostic delay (months)</td>
<td>9.9</td>
</tr>
<tr>
<td>Progression rate (points decrease on ALSFRS-R per month)</td>
<td>1.22</td>
</tr>
<tr>
<td>Forced vital capacity (% of predicted)</td>
<td>65</td>
</tr>
<tr>
<td>Definite ALS (according to El Escorial criteria)</td>
<td>Yes</td>
</tr>
<tr>
<td>Frontotemporal dementia</td>
<td>Absent</td>
</tr>
<tr>
<td>C9orf72 repeat expansion</td>
<td>Absent</td>
</tr>
<tr>
<td>Site of onset</td>
<td>Bulbar</td>
</tr>
</tbody>
</table>
ALS progresses at different rates

### Patient Demographics

<table>
<thead>
<tr>
<th>Predictors</th>
<th>Data</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at onset (years)</td>
<td>64.7</td>
</tr>
<tr>
<td>Diagnostic delay (months)</td>
<td>13.8</td>
</tr>
<tr>
<td>Progression rate (points decrease on ALSFRS-R per month)</td>
<td>0.29</td>
</tr>
<tr>
<td>Forced vital capacity (% of predicted)</td>
<td>75</td>
</tr>
<tr>
<td>Definite ALS (according to El Escorial criteria)</td>
<td>Yes</td>
</tr>
<tr>
<td>Frontotemporal dementia</td>
<td>Absent</td>
</tr>
<tr>
<td>C9orf72 repeat expansion</td>
<td>Absent</td>
</tr>
<tr>
<td>Site of onset</td>
<td>Spinal</td>
</tr>
</tbody>
</table>
A Day in ALS Clinic
Multidisciplinary Visit

Goals for a day in clinic:
1) Assess physical function, QOL, symptom management and disease-related complications.
2) Empower pALS and cALS through open discussions regarding care, care planning, and opportunities for research participation.

Logistics: Visits typically occur every 3-6 months and last for 3.5 hours to see multiple providers.
Neuromuscular Disease Specialist/Neurologist (MD)

- Assesses:
  - Muscle twitching
  - Muscle cramping
  - Muscle strength
  - Reflexes

- Diagnosis
- Head to toe evaluation
- Puts the puzzle pieces together
- Medication management
- Alternative/off label treatments
- Advanced Care Planning
## Supplements and Alternative Off-Label Treatments

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Podcast</th>
<th>Mechanism</th>
<th>Pre-Clinical</th>
<th>Cases</th>
<th>Trials</th>
<th>Risks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ketogenic Diets (2021)</td>
<td>B</td>
<td>C</td>
<td>D</td>
<td>U</td>
<td>D</td>
<td></td>
</tr>
<tr>
<td>Vitamin C (2021)</td>
<td>C</td>
<td>C</td>
<td>B</td>
<td>F</td>
<td>B</td>
<td></td>
</tr>
<tr>
<td>Melatonin (2021)</td>
<td>A</td>
<td>C</td>
<td>B</td>
<td>U</td>
<td>B</td>
<td></td>
</tr>
</tbody>
</table>
Respiratory Management

- Improve ventilation

Shortness of Breath

NIVV: Noninvasive Volume Ventilation

Allison, M., 2020
Respiratory Management

- Improve weak Cough
- Aide in managing secretions
- Prevent respiratory infections

Weak Cough/ Managing Secretions
Cough Augmentation

- Treatment protocol:
  - 3-5 coughs
  - Performed 3x/day

Cough Assist Device

Oral Suction Device

- Use as needed
- Can be combined with medication:
  - Atropine drops
  - Scopolamine patch
  - Botox
  - Various other meds.
Invasive Ventilation: Tracheostomy

Respiratory Support when NIVV is no longer supportive and/or secretions unmanageable
Speech Language Pathologist (SLP)
Swallowing difficulty (dysphagia) & Airway Protection
Swallowing and Breathing: Airway Clearance and Cough Augmentation
Respiratory Muscle Strength Training

Tabor et al., 2016; Plowman et al., 2015; 2019
Speech-Language Pathologist (SLP)

Communication Evaluation

Voice Preservation & Communication Strategies
- Voice & Message Banking
- Compensatory Strategies, Energy Conservation

Alternative and Augmentative Communication (AAC)
- Low-tech Communication
- High-tech Communication

Costello, J. 2011
**Dietitian**
- Nutrition Support and Management
- Feeding Tube Placement

- Body Mass Index (BMI) a significant predictor of disease progression and survival
- Low BMI associated with faster progression

Common Recommendations
- Extensive education to prevent weight loss
- Smaller, more frequent meals
- Snack often
- High calorie sauces, condiments, EVOO
- Benecalorie, oral supplements
- Feeding tube placement

Wang et al 2017; Deport et al 1999
Physical and Occupational Therapy

“DME”: Durable Medical Equipment

Home Exercise Programs

Fall prevention
Social Worker, ALSA Liaison

- The Resource Gurus
- Insurance, home health, DME

ALSA Registry

https://www.als.org/advocacy/als-registry
Neuropsychology

Address the emotional, cognitive and quality of life issues that arise when confronting the challenges of living with ALS

Apathy
Anxiety
Depression
Behavior

Wooley & Rush, 2017
Palliative Care

• Extra layer of support and care throughout the duration of disease
• Appropriate at any stage of the disease
Palliative Care

Helpful at *any time* throughout the disease for pALS and cALS

**In Common**
- Comfort care
- Reduce stress
- Offer complex symptom relief related to serious illness
- Physical and psychosocial relief

**Palliative Services**
- Paid by insurance, self
- Any stage of disease

**Hospice Services**
- Paid by Medicare, Medicaid, insurance
- Prognosis 6 months or less

Vitas Healthcare
Best Management = TEAM approach

- Life planning, Advanced Directives
- Access to support groups
- Mental Health counseling

Pharmacologic Management

- Respiratory: Breath stacking/Respiratory strength training (early); NIV, Cough Assist
- Swallowing: dietary modification, secretions
- Nutrition: Oral supplements, PEG
- Mobility: Passive ROM/Stretching, DME

- FDA Approved Medications
- Alternative off-label treatments
- Experimental Drugs
- Clinical Trials
- Patient and caregiver education

- Life planning
- Individualized patient care
- Mental Health counseling
- Resources to help
- Support groups

- Caregiver Burden
- Disease Progression
- Symptoms
- Quality of Life
## Multidisciplinary Care Team Approach

<table>
<thead>
<tr>
<th>Specialist</th>
<th>Role</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Neurology/ NP</strong></td>
<td>Diagnosis, Medication management</td>
</tr>
<tr>
<td><strong>SLP</strong></td>
<td>Swallowing, Airway protection, Secretions, Communication</td>
</tr>
<tr>
<td><strong>OT/PT</strong></td>
<td>Fine motor skills (feeding), mobility, fall preventions home exercise program</td>
</tr>
<tr>
<td><strong>Respiratory</strong></td>
<td>Cough Augmentation, Respiratory therapy</td>
</tr>
<tr>
<td><strong>Social Work</strong></td>
<td>Service provision and setup, Insurance, Resources</td>
</tr>
<tr>
<td><strong>Dietitian</strong></td>
<td>Nutrition and weight Management, Alternative Feeding (PEG)</td>
</tr>
<tr>
<td><strong>Nurse</strong></td>
<td>Vitals, Clinic coordination, Patient Advocacy</td>
</tr>
<tr>
<td><strong>Palliative Care</strong></td>
<td>Additional support layer, Life decision-making, Hospice consult</td>
</tr>
<tr>
<td><strong>Neuropsychology</strong></td>
<td>Emotion and well-being, changes in thinking, behavior, memory</td>
</tr>
</tbody>
</table>
Clinical Research Opportunities

Interventional

Observational

Biobank
Goals of Exceptional Multidisciplinary ALS Care

1. Comprehensive, coordinated care: family, caregivers, friends, ALS clinic team

2. **Proactive** intervention vs. **Reactive** intervention
   - Optimize Symptom Management
   - Energy Conservation
   - Prophylactic decision-making
   - Patient and Caregiver Education

3. Provide the opportunity to participate in experimental research, clinical trials, observational studies or biobanks
Thank you
Lauren.taborgray@nova.edu