ALS Multidisciplinary Care

Guest Speaker:
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and Clinical Research Center
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Holy Cross Health

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Optimizing ALS Care: the Multidisciplinary Clinic

Lauren Tabor Gray, PhD
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Holy Cross Health
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Motor Neuron Disease

ALS
- Spinal-Onset Variant
- Bulbar-Onset Variant

ALS

PMA
- Primary Muscle Atrophy

PLS
- Primary Lateral Sclerosis

Lower Motor Neuron
Upper Motor Neuron

Swinnen, 2014
Amyotrophic Lateral Sclerosis

- **A**: No; **myo**: Muscle; **trophic**: nourishment
  - No Muscle Nourishment

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

- **Sclerosis**: or hardening that occurs as the 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.
What does evidence-based care look like for pALS?
- 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 MGH, ALSA, MDA, Always for ALS
- Advocacy: Clinical Research, ALSA, ALS TDI, Legislation, Patient Ambassadors (CURLI)
The 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
Goals of Multidisciplinary ALS Clinic Visits

1. Manage Symptoms
2. Slow Disease Progression
3. Maintain Quality of Life
4. Coordination of Care
5. Reduce Caregiver Burden
6. Research Opportunity
Manage Symptoms

- Slow Disease Progression
- Maintain Quality of Life
- Reduce Caregiver Burden

- Rx: Nuedexta, baclofen, pain & sleep meds
- Respiratory (early): Breath stacking/Respiratory muscle strength training
- Secretion Management: rx, botox, radiation
- Nutrition: Oral supplements, PEG
- Mobility: Orthotics, Botox, Passive ROM/Stretching, DME

- Riluzole (+ ~89 days to NIVV/IV)
- Edaravone (33% slowing in subgroup)
- NIVV/cough assist
- Patient and caregiver education
- Clinical Trials

Life planning and decision making, Advanced Directives
Access to support groups
Respite care
Mental Health counseling

- Life planning and decisions
- Individualized care, patient navigates journey
- Mental Health counseling
- Resource Identification
- Support groups

ALS

Resource Identification
Support groups
ALS: Nuts and Bolts

**Important Disease Characteristics influencing care:**
- Onset Type (where symptoms began)
- Body Mass Index (BMI)
- Degree of respiratory involvement
- Family History:
  - ALS, Motor neuron disease
  - Psychiatric illness, dementia
Impact on Life and Function

Breathing

Swallowing

Quality of Life

Cognition

Mobility

Communication
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>

![Graph showing survival probability over time since onset (months) for average patient and current patient.](image)
ALS progresses at different rates

### Patient Demographics

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<th>Data</th>
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<tbody>
<tr>
<td>Age at onset (years)</td>
<td>64.7</td>
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<tr>
<td>Diagnostic delay (months)</td>
<td>13.8</td>
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<tr>
<td>Progression rate (points decrease on ALSFRS-R per month)</td>
<td>0.29</td>
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<tr>
<td>Forced vital capacity (% of predicted)</td>
<td>75</td>
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<tr>
<td>Definite ALS (according to El Escorial criteria)</td>
<td>Yes</td>
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<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>
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</tbody>
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A Day in ALS Clinic
Neuromuscular Disease Specialist (MD)

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

- Diagnosis
- Head to toe evaluation
- Puts the puzzle pieces together
- Medication management
- 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>
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</thead>
<tbody>
<tr>
<td>Ketogenic Diets (2021)</td>
<td>B</td>
<td>C</td>
<td>C</td>
<td>U</td>
<td>D</td>
<td></td>
</tr>
<tr>
<td>Vitamin C (2021)</td>
<td>C</td>
<td>C</td>
<td>E</td>
<td>F</td>
<td>R</td>
<td>B</td>
</tr>
<tr>
<td>Melatonin (2021)</td>
<td>A</td>
<td>C</td>
<td>E</td>
<td>U</td>
<td>R</td>
<td>B</td>
</tr>
</tbody>
</table>
Shortness of Breath

NIVV: Noninvasive Volume Ventilation
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

Oral Suction Device

- Use as needed
- Can be combined with medication:
  - Atropine drops
  - Scopolamine patch
  - Botox
  - Various other meds.
Respiratory: Cough Augmentation

Respiratory Muscle Strength Training

Tabor et al., 2016; Plowman et al., 2015; 2019
Invasive Ventilation: Tracheostomy

Respiratory Support when NIVV is no longer supportive and/or secretions unmanageable
Speech Language Pathologist (SLP)
Swallowing difficulty (dysphagia) & Airway Protection
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:
- Smaller, more frequent meals
- Snack often
- High calorie sauces, condiment, 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
Social Worker, ALSA Liaison
(aka: The Glue)

- 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
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
# Multidisciplinary Care Team Approach

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

Interventional

Observational

Biobank
Goals of ALS Care

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

Proactive intervention vs. Reactive intervention
  • Energy Conservation
  • Prophylactic decision-making
  • Patient Education
Summary

Multidisciplinary clinics can improve QOL, symptom management, disease-related complications and survival.

Visits typically occur every 3-6 months and last for 3.5 hours to see multiple providers.

Empowering pALS and cALS through research design and participation.

Clinical Research Learning Institute® (CRLI)
“We’ve been wrong about what our job is in medicine. We think our job is to ensure health and survival. But really it is larger than that. It is to enable well-being.”

-Dr. Atul Gawande, Being Mortal: Medicine and What Matters in the End

Thank you
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