Multidisciplinary Care in Amyotrophic Lateral Sclerosis

HUNED PATWA MD
VA CT HEALTHCARE SYSTEM
YALE UNIVERSITY

PVA SUMMIT 2012
Disclosures

- This continuing education activity is managed and accredited by Professional Education Service Group. The material presented in the activity represents the opinion of the faculty. Neither PESG, nor any accrediting organization endorses any commercial products displayed in conjunction with this activity.

- Commercial support was not received for this activity.
Disclosures

• Huned Patwa MD has no financial interest or relationship to disclose
• CME Staff Disclosure
• Professional Education Service Group staff have no financial interest or relationship to disclose
Learning Objectives

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

A. Recognize clinical signs and symptoms of Amyotrophic Lateral Sclerosis (ALS)

B. Identify and understand the role of key team members in an ALS multidisciplinary clinic (MDC)

C. Recognize the value of multidisciplinary care for ALS.

D. Be familiar with Practice Parameters on the care of patients with ALS.
ALS From Charcot....

- A-without
- Myo-muscle
- Trophic-nourishment
- Lateral-lateral corticospinal tracts
- Sclerosis-gliosis on pathology
ALS From Charcot to Gehrig
Diagnosis of Amyotrophic Lateral Sclerosis

**UPPER MOTOR NEURON**
- Spasticity
- Hyperreflexia
- Pathological reflexes

**LOWER MOTOR NEURON**
- Weakness
- Atrophy
- Hyporeflexia
- Fasciculations
Pathology
ALS Epidemiology

- Incidence 2 per 100,000
- Prevalence 5-7/100K
- 60% men
- Average age 55y
- 90% sporadic
- 10% genetic
- 50% die in 30 months
- 20% between 5-10y
Diagnosis - El Escorial / Awaji Criteria
ALS Phenotypes

- Classic “Charcot”
- Bulbar
- Flail arm
- Flail leg
- Pyramidal
- Respiratory
- Pure lower motor neuron
- Pure upper motor neuron
<table>
<thead>
<tr>
<th>Clinical Manifestations</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Weakness</td>
<td>No bowel/bladder</td>
</tr>
<tr>
<td>Atrophy</td>
<td>Pain</td>
</tr>
<tr>
<td>Spasticity</td>
<td>Pseudobulbar Affect</td>
</tr>
<tr>
<td>Dysarthria</td>
<td>Cognitive</td>
</tr>
<tr>
<td>Dyspnea</td>
<td></td>
</tr>
<tr>
<td>Dysphagia</td>
<td></td>
</tr>
<tr>
<td>Cramps</td>
<td></td>
</tr>
</tbody>
</table>
Progressive Loss of Motor Neurons

Bromberg J of Neurol Sci Aug 2008
Cognitive Problems

- Spectrum of disease from ALS → ALS with dementia → Frontotemporal Lobar dementia (FTLD)
- Linked by a TDP-43 proteinopathy
- 15% of patients
- Characterized by executive dysfunction, disinhibition, impulsivity, apathy, decision making, decreased insight
- Optimal screening is not known-verbal fluency, naming, word generation..
- Care planning, compliance with interventions and end of life decision making
Survival in ALS

Kollewe J of Neuro Sci Dec, 2008
Genetics

- **SOD1**
  - 20% familial
  - Gain of function
  - Protein aggregates
- **TARDBP**
  - TDP-43
  - RNA processing
- **FUS/TLS**
- **Ubiquilin-2**
  - UQLN2
  - Clearance of protein degradation
- **C9ORF72**
  - 37% of fALS
  - 6% of sALS
Mechanism of Disease

Kiernan-Lancet Neurology, 2011
Timeline of Increased Risk of ALS in Veterans

- 1991 Persian Gulf War
- 2003 Horner-DVA/DOD-2x increase in ALS, greatest in Air Force and Army
- 2003 Haley-Perot Found-similar conclusion
- 2004/2008-Research Committee on Gulf War Illness determined “most serious condition reported to affect Gulf War veterans at a higher than expected rate is amyotrophic lateral sclerosis.”
- 2005 Weiskopf-Harvard-ALS associated with any history of military service
- 2006 IOM-reviewed all evidence
- 9/23/2008-presumption of service connection for ALS
Conclusions:
- Two Class II studies and one Class III study show that multidisciplinary clinics specializing in ALS care are probably effective in several ways: increased use of adaptive equipment; increased utilization of riluzole, PEG, and NIV; improved quality of life; and lengthened survival. However, one Class II study with low use of treatments found no survival benefit.

Recommendations:
- Specialized multidisciplinary clinic referral should be considered for patients with ALS to optimize health care delivery (Level B) and prolong survival (Level B), and may be considered to enhance quality of life (Level C).
What is MDC

- There is no curative tx for ALS
- Care is focused on symptomatic, rehabilitative and palliative care services
- MDC works as a coordinated and organized team to deliver this care efficiently and effectively
Characteristics of MDC

- Concentrates expertise and resources
- Designed to patient centered
- Flexibility
- Responsiveness
- Maximize activity and participation
Multidisciplinary Model

- Physical Therapy
- Occupational Therapy
- Speech Therapy
- Psychology
- Nursing
Interdisciplinary Model

Physical Therapy  Occupational Therapy

Speech Therapy  Psychology

Nursing

MD
The Multidisciplinary Clinic Approach

Concept: Patient is “in control” and approach is “pro-active”.

Neurologist/
Neuromuscular ”

Physical Therapist
Occupational Therapist

Psychologist

Speech Lang/ Swallow
Nutritionist
Aug Comm Specialist

Respiratory Therapist

Physician Associate

Social Worker
Chaplain
Benefits Specialist
ALSA Pt Care Coordinator

Each Team Member has input and a consensus recommendation for management is made on a linked note, including date for next visit. Any assistive devices, etc. that can be issued are given at visit!
Team Members

- Neurologist/Physiatrist
- Nurse/PA, Case Manager
- Occupational Therapist
- Physical Therapist
- Social Worker
- Psychologist
- Respiratory Therapist
- Hospice/Palliative Care
- Nutrition

- Patient Care Org
  - PVA
  - ALSA
  - MDA
Studies on MDC

- **Traynor (2003)**
  - 7.5 months longer survival

- **Chio (2006)**
  - Improved survival (1080 vs 775 days)
  - Decreased hospitalization frequency and decreased LOS

- **Zoccollela (2007)**
  - No significant difference in median survival

- **Van Den Berg (2005)**
  - Improved QOL on SF-36 mental health domains
ALS MDC: Effect on Survival

Figure 2  Survival of Irish ALS patients with bulbar onset disease according to the clinic type attended, 1996–2000.

Figure 3  Survival of Irish ALS patients attending the multidisciplinary clinic compared to patients prescribed riluzole and attending a general neurology clinic, 1996–2000.

Cost of Care

VHA ALS Patient Cost Profile by Multi-disciplinary Clinics (FY10)

<table>
<thead>
<tr>
<th></th>
<th># ALS Patients</th>
<th>Total Cost</th>
<th>Total Cost per Patient</th>
<th>Inpatient Cost</th>
<th>Inpatient Cost per Patient</th>
<th>Outpatient Cost</th>
<th>Outpatient Cost per Patient</th>
<th>Fee Expenditure</th>
<th>Fee Expenditure per Patient</th>
</tr>
</thead>
<tbody>
<tr>
<td>Multi-Disciplinary Clinics</td>
<td>480</td>
<td>$14,642,542</td>
<td>$30,505</td>
<td>$5,418,515</td>
<td>$11,289</td>
<td>$7,751,007</td>
<td>$16,148</td>
<td>$1,473,020</td>
<td>$3,069</td>
</tr>
<tr>
<td>General Clinics</td>
<td>3060</td>
<td>$92,480,523</td>
<td>$30,222</td>
<td>$31,408,990</td>
<td>$10,264</td>
<td>$44,162,686</td>
<td>$14,432</td>
<td>$16,908,847</td>
<td>$5,526</td>
</tr>
<tr>
<td>Multi-Disciplinary Clinics (PRP)</td>
<td>348</td>
<td>$14,642,542</td>
<td>$42,089</td>
<td>$5,418,515</td>
<td>$15,757</td>
<td>$7,751,007</td>
<td>$22,260</td>
<td>$1,473,020</td>
<td>$4,234</td>
</tr>
<tr>
<td>General Clinics (PRP)</td>
<td>2339</td>
<td>$92,480,523</td>
<td>$39,531</td>
<td>$31,408,990</td>
<td>$13,426</td>
<td>$44,162,686</td>
<td>$18,878</td>
<td>$16,908,847</td>
<td>$7,228</td>
</tr>
</tbody>
</table>
Limitations

- Methodologically weak studies
- Lacks comparison of different intensities or content of care
- Variable outcomes
- Costs benefit analysis was very limited
- Caregivers and needs were not addressed
- Lack of palliative care
Measuring Outcomes

- Evidence based practice vs practice based evidence
- Outcome measures
  - Survival
  - QOL
  - Caregiver burden
  - Quality Measures
- Clinical Practice Improvement
  - observational
Establishing the Program

Location: Spinal Cord Clinic

Participants:
- Clinician (MD + PA): SCI, Neurology, Palliative Care
- OT, PT, Social Work, Clinic Coordinator/Health Tech
- Speech and Swallowing, Pulmonary, Nursing
- Case Manager

Clinic schedule
- Locate patients
- Create documentation, notes
- Make connection to service officers
- Connect with PVA, ALSA and MDA
- Connect with nearest ALS Clinics
Clinic Visits

- History
- Symptom review, patient report using ALS FRS
- Physical Exam (vitals, especially weight)
- Review of needs, new problems, anticipate
- Lab and other tests
- Advanced care planning: ongoing discussions about what’s happening, what to expect, ventilatory or nutritional interventions
1. Speech
   Normal speech processes - 4
   Detectable speech disturbance - 3
   Intelligible with repeating - 2
   Speech combined with nonvocal communication - 1
   Loss of useful speech - 0

2. Salivation
3. Swallowing
4. Handwriting
5. Cutting Food
6. Dressing and Hygiene
7. Bed Mobility
8. Walking
9. Climbing stairs
10. Dyspnea
11. Orthopnea
12. Respiratory insufficiency
AAN-Practice Parameter

- Breaking the news
- Symptom Management
- Medication
- Algorithm for Respiratory and Nutritional Care
- Palliative Care
- Multidisciplinary Care
Diagnosis

- Confirm diagnosis
- Break the news
- Explains disease, prognosis, treatments (Riluzole information)
- Provides contact numbers/e-mails for questions or concerns
- Information from ALSA and MDA
- Veteran SC information
- Refer to Multidisciplinary Clinic
Riluzole

- Approved by FDA in 1995
- Inhibits glutamate release
- 50mg BID
- Benefit is modest
- Cohort studies suggest longer benefit
- SE-Fatigue and nausea
- Monitor LFTs
Respiratory Management Algorithm

Text in bold = evidence-based
Text in italics = consensus-based

- Diagnosis ALS
  - Symptom evaluation* and PFTs
    - Initiate NIV orientation, Pneumovax and flu vaccine

- Orthopnea or
  - SNP < 40cm or MIP < -60cm
  - Abnl nocturnal oximetry or FVC < 50%
  - Consider NIV
    - NIV tolerated?
      - No
      - Further education regarding documented benefits. Evaluate reasons for noncompliance.
      - Yes
      - Ongoing evaluations and adjustment of pressures
        - Reintroduce NIV
          - Successful
          - Not successful †
            - Hospice referral for palliative care
            - Invasive ventilation

- PCEF < 270 L/min
  - Suction machine
    - Manual assisted cough
    - Mechanical inexsufflator
      - Treat sialorrhea/phlegm

Unable to maintain pO₂ > 90%, pCO₂ < 50mmHg or unable to manage secretions
Measuring Respiratory Function

- **Functional ventilatory capacity (FVC)**
  - Supine FVC better predictor of diaphragm weakness

- **Maximum inspiratory pressure (MIP)**

- **Nocturnal oximetry correlates with survival**:
  - Mean $O_2$ sat $< 93$ mm Hg $\rightarrow$ mean survival of 7 months
  - Mean $O_2$ sat $> 93$ mm Hg $\rightarrow$ mean survival of 18 months

- **Sniff nasal pressure (SNP)**
  - greater predictor of hypercapnea than FVC or MIP
  - more reliably recorded at later stages of ALS

- **Peak cough expiratory flow (PCEF)**

- **Serum bicarbonate and chloride**

- **ABG: Daytime $SpO_2 < 95\%$ not corrected by NIV $\rightarrow$ tracheostomy or death within 2 months**
Nutritional Management Algorithm

**Diagnosis: ALS**
- Clinic visits every 3 months
- Early dysphagia detected
- Nutritional education including PEG
- Nutritionist or speech therapist referral

**Monitor body weight**
- Dysphagia assessment instrument

**Monitor Respiratory status** (FVC, MIP, etc.)
- Clinic visits every 3 months
- Symptom progression or continuing weight loss
- Discuss PEG to stabilize weight and possibly prolong survival

**FVC >50%**
- Low risk for PEG
  - PEG accepted

**FVC 30-50%**
- Moderate risk
  - Anesthesia evaluation
  - Experienced gastroenterologist
  - Respiratory support during PEG if needed

**FVC <30%**
- High risk
  - PEG declined

**Oral intake as tolerated** | **Enteral nutrition via PEG as needed**
|-----------------------------|-----------------------------|

Text in **bold** = evidence-based
Text in **italics** = consensus-based
**Nutritional Management**

**Early disease**
- Weights
- Dysphagia Assessment
  - ALS FRS bulbar questions
  - Prolonged meal time
  - Fatigue

**Signs of dysphagia:**
- PEG education
- Nutrition consult
- Modified textures

**Progression:**
- Worse symptoms
- Continued weight loss

PEG (Percutaneous Endoscopic Gastrostomy)
- Stabilize weight
- Possibly prolong survival
- FVC > 50%
Advanced Care Planning
Palliative Care

- Hospice and Palliative Care--Multidisciplinary Team
- Encourage as early as acceptable to veteran
- PEG and ventilatory support choices
- Bereavement from start to finish and after
- Medicare Hospice Benefit: no housebound requirement
- Flexible source of extra care
- Preparing for difficult situations
Advantages of VA Care

- Integrated Healthcare System
- Clinical Services are available
- Unique Services
  - Palliative Care
  - Home Based Care
- Development of Best Practices
- Opportunities for Clinical Research
http://www.youtube.com/watch?v=g0Q4kIx95aU
National ALS Registry

The National Amyotrophic Lateral Sclerosis (ALS) Registry

Click Here to Enroll

www.cdc.gov/als
(800) 232-4636

ATSDDR
Thank You
Obtaining CME/CE Credit

- If you would like to receive continuing education credit for this activity, please visit
  - http://pva.cds.pesgce.com/