On the Yellow Brick Road
PROGRAM BUILDING
for
ALS Care
Dr. Kevin White, MD
Iliene Page, ARNP-C
Catherine Wilson, PSYD, ABPP
Integration of Medical, Nursing & Psychology in the Care of Veterans with Amyotrophic Lateral Sclerosis

Michael Bilirakis VA
Spinal Cord Injury Center
James A. Haley Veterans’ Hospital
Tampa, FL
ALS Overview

- Progressive and terminal degenerative motor neuron disease.
  - Upper motor neuron (UMN): Spasticity, and hyper reflexivity
  - Lower motor neuron (LMN): muscle weakness and wasting

- Dysphagia
- Cognitive Deficits*
- Ambulation difficulties
- Para/Tetraplegia
- Diaphragmatic Weakness
- Difficulties coughing/managing oral secretions

Can lead to complete dependence for ADLs
Majority of patients perish due to respiratory compromise
ALS Epidemiology

[De Carvalho & Swash 2011; Chiò et al. 2013; Cronin et al. 2007; ALS Association 2014; Weisskopft et al 2005; Strong 2003; Mehta et al 2014]

- **Prevalence & Incidence:**
  - 3.9 new cases per year per 100,000 (US)
  - 14 new cases diagnosed each day
  - 30,000 people living with the disease

- **Gender Differences:** Males > Females (1.5-2 : 1)

- **Racial Differences:** Whites (Non Hispanics) > Nonwhites (1.6:1)

- **Median age of onset:** 55-65 years old
ALS Survival

[Schmidt et al. (2006); Saeed et al. (2009)]

- **Median survival:**
  - 2-5 years from onset of symptoms [Strong 2003; Weisskopf et al., 2005]
  - 1-3 years after diagnosis [Weisskopf et al 2005]

- About 15% live beyond 5 years from diagnosis
- About 5% live beyond 10 years.

- **Long-term survival is associated with:**
  - Younger age at onset
  - Being male
  - Limb (rather than bulbar) symptom onset
ALS Phenotypes

[Beghi et al., 2006; Mitsumoto 2009; Chio et al., 2011; Swinnen & Robberecht, 2014]

- **El Esactorial System (1990):**
- Other Distinctive Types:
  - Classic (Charcot’s Type)
  - Bulbar
  - Flail Arm
  - Flail Leg
  - Pyramidal
  - Respiratory
  - Pure Upper Motor Neuron
  - Pure Lower Motor Neuron
  - Primary Lateral Sclerosis
ALS Etiology
(Ahmed et al 2011; De Carvalho & Swash 2011)

- **Familial ALS (5-10% of cases):** Genetic etiology suspected
  - 12+ genetic mutations implicated [National Institute for Neurological Disorders and Stroke]

- **Sporadic ALS (90% of cases):** Largely unknown etiology

- **Environmental factors suspected to increase risk:** [Beghi et al, 2006]
  - Smoking
  - Excessive physical activity
  - Dietary factors
  - Changes in immunity
  - Exposure to toxins (heavy metals, solvents)
  - Exposure to electromagnet currents
  - Mechanical trauma (head injury)
  - Correlated with cancer diagnosis
ALS Treatment
[Riviere et al. 1998; Strong 2003]

• **No cure**

• **Medications:** *Riluzole* (Antiglutaminergic agent)

• **Symptom management**
  – Pain, oral secretions, spasticity, etc.

• **Emphasis on multidisciplinary rehabilitation model of care**
  – Neurology    Social Work    Speech Therapy
  – Pulmonology  Nursing       Vocational Rehabilitation
  – Psychiatrist Nutrition    Physical Therapy
  – Psychology   Palliative Care  Occupational Therapy
  – Pharmacist
Veterans with ALS: Greater Risk??
[Allen et al. 2008]

- **2003:** Veterans deployed to the Persian Gulf War at x2 risk
  [Horner et. al, 2003; Haley 2003]
  - National Registry of Veterans with ALS established

- **2006:** Military service viewed as a risk factor for ALS
  [Weisskopf et al.]

- **2008:** VHA ➔ “Presumption of Service Connection for ALS”

- **2010:** National ALS Registry launched

- **2011:** VHA ➔ Automatic 100% Service-Connection for ALS

- **Paralyzed Veterans of America:** Instrumental role toward automatic 100% service connection, housing and vehicle grants.
Location of Veterans with ALS
Veterans with ALS in FL / VISN 8

- 1,521 Veterans with ALS served by VHA in 2014

- JAHVH
  - FY 2013: 136 patients
  - FY 2014: 151
  - FY 2015: 163
  - FY 2016: 153
Task Force Recommendations

- ALS diagnosis should be confirmed or excluded as quickly as possible
- Structure ALS services using established principles of ALS management
- Diagnosis communication: understandable, empathetic, and supportive
- Veteran with ALS should be seen by an ALS interdisciplinary team
- Create effective interdisciplinary care processes for Veterans with ALS
- Interdisciplinary team should include the Patient’s Primary Care Provider
Task Force Recommendations (con’t)

• Need to address at each visit - strength, pain/spasticity, swallowing, breathing, communication, sleep, emotional needs, anticipate issues such as feeding tube and end of life issues

• Co-management of care should be used to balance the need for accessible, local care with the need for interdisciplinary ALS specialty care.

• Management and coordination should address durable medical equipment, community collaboration, caregiver issues, and standardized assessment.

• Educate clinical staff members caring for Veterans with ALS on best practices and evidence-based treatment as well as ways to support the needs/options for Veterans and caregivers.
Growth of ALS Population

- New ALS
- Death
- Total # of ALS Patients by the end of FY

<table>
<thead>
<tr>
<th>Year</th>
<th>New ALS</th>
<th>Death</th>
<th>Total # of ALS Patients by the end of FY</th>
</tr>
</thead>
<tbody>
<tr>
<td>2007</td>
<td>22</td>
<td>10</td>
<td>22</td>
</tr>
<tr>
<td>2008</td>
<td>23</td>
<td>2</td>
<td>23</td>
</tr>
<tr>
<td>2009</td>
<td>26</td>
<td>13</td>
<td>39</td>
</tr>
<tr>
<td>2010</td>
<td>41</td>
<td>15</td>
<td>56</td>
</tr>
<tr>
<td>2011</td>
<td>38</td>
<td>22</td>
<td>65</td>
</tr>
<tr>
<td>2012</td>
<td>22</td>
<td>26</td>
<td>81</td>
</tr>
<tr>
<td>2013</td>
<td>28</td>
<td>28</td>
<td>87</td>
</tr>
<tr>
<td>2014</td>
<td>34</td>
<td>34</td>
<td>105</td>
</tr>
<tr>
<td>2015</td>
<td>56</td>
<td>38</td>
<td>123</td>
</tr>
</tbody>
</table>
ALS Challenges in Treatment

• ALS is a complex, low incidence, progressive [and terminal] disease with many physical and emotional considerations

• CHALLENGES:
  – Few experienced health care providers or specialty programs
  – Need for extensive multi/interdisciplinary specialty services: move to more transdisciplinary team
  – Often there is no “conductor” of care to drive and coordinate services
  – Small delays in service delivery can have huge negative impact
  – Few resources for family education, networking, and support
  – INCREASING number of Veterans entering the VHA system
Respiratory complications in ALS patients

1. Diaphragmatic weakness.
2. Inability to handle oropharyngeal secretions.
3. Difficulty clearing respiratory secretions.
4. Ineffective cough.
5. Sleep-disordered breathing and nocturnal hypoventilation.

Since ALS overwhelmingly brings death via respiratory compromise, Pulmonary function Tests (PFTs) should be particularly appropriate predictors of individual survival.
Vital Capacity & ALS

- **Forced Vital Capacity (FVC)** is a significant predictor of survival, and <50% of the predicted value has been shown to be associated with poor prognosis.
- Supine FVC may be a better predictor of diaphragm weakness than erect FVC.
- Supine FVC closely correlates with transdiaphragmatic pressure (Pdi), and a supine FVC < 75% reliably predicts an abnormally low Pdi.

Management:

- “Early” intervention with Noninvasive ventilation (NIPPV) and use of NIPPV >4 hours/day have been associated with greater survival & a slower rate of FVC.

- NIPPV can prolongs survival in ALS patients with normal to moderately impaired bulbar function, but only improve *quality of life* in patient with *severe* bulbar impairment.

The use of tracheostomy ventilation in ALS patients is varies widely by country, from 1.4–15% in USA, to a majority of patients in Japan.

Annual cost of home ventilation of ~ $150,000.

Following tracheostomy, median survival ranges from <12 months to 37 months.

The most common cause of death is respiratory tract infection.
Summary

• Respiratory failure is the main cause of morbidity and mortality among ALS patients.
• Forced Vital Capacity (FVC) is a significant predictor of survival.
• Noninvasive ventilation is a vital tool in management of ALS-induced hypoventilation.
• Tracheostomy and mechanical ventilation is suitable alternative for selected ALS patients, but at high annual cost.
• Determining ALS clinical phenotypes can be vital in relation to clinic-pathological correlations, therapeutic trial design & practice benchmarking.
Developing an ALS program

Based in a Spinal Cord Injury Service
on Rehabilitation Nursing Units
Henderson defined nursing

• "The unique function of the nurse is to assist the individual, sick or well, in the performance of those activities contributing to health or its recovery (or to a peaceful death) that he would perform unaided if he had the necessary strength, will or knowledge." 1966

• But she went on to say that the nurse makes the patient independent of him or her as soon as possible.
Try Abiding Instead of Hiding: An Approach to End-of-Life Care


What we can offer when we can’t offer CURE
What is different about VA and ALS?

- 100% SERVICE CONNECTED
- BENEFITS
- MONEY
- ADAPTIVE HOUSING GRANT
- AUTO GRANT
- EQUIPMENT
- TEAM INTRODUCTION AND FOLLOW-UP
What is different about ALS?
Goals of Program

- Manage symptoms of ALS
- Provide specialized equipment in a timely manner
- Facilitate access to needed services
- Achieve patient and family satisfaction
- Provide Telehealth clinic visits using veterans
  - Home computer to link to the hospital
- Offer education to patients and families about disease progression
ALS Program Process

Person may have ALS → Eligibility for VA care determined → Evaluated by Neurology → ALS diagnosis confirmed → Scheduled for SCI admission

- Primary Care Physician
- Pulmonologist
- Psychologist
- Palliative Care Physician
- Nurses

Inpatient Evaluation 4-5 days

- Physical Therapist
- Occupational Therapist
- Speech Therapist
- Dietitian
- Vocational Rehab Therapist

Discharge

Weekly ALS Team Meetings

Follow-up outpatient care

- Primary Care Physician
- Pulmonologist
- Psychologist
- Palliative Care Physician
- Nurses

- TeleHealth
- Home Care

As needed
Team Approach

- Best delivered by multidisciplinary team (Physicians-Neurologists & Pulmonologists, Occupational, Physical, and Speech Therapists, Nutritionists, Social Workers, Psychologist, Home care and Hospice Nurses)
- Medications can be prescribed to treat fatigue, muscle cramp and stiffness, excessive saliva, pain, depression, sleep disturbance.
TEAM BUILDING
TEAM BUILDING

- Forming
- Storming
- Norming
Our Team

Neurologist
Pulmonologist
Nurse Practitioner
Psychology
Nurses
Respiratory Therapist
Dietitians
Speech Pathologist
Social Work
(local/distant)
Occupational Therapist
Physical Therapist

Adaptive Technologist
Palliative Care MD
Vocational Rehabilitation
Recreational Therapist
SCI Physicians
Pharmacist
Meet Our Team
A Multidisciplinary Approach
Developing into a Transdisciplinary Team
Lou Gehrig's Disease

Difficulty Breathing
Voice Change, Weight Loss
Difficulty Swallowing
Slurring of Words, Muscle Weakness
Muscle Cramps, Paralysis

CASE MANAGEMENT
Educating client

- Durable Mechanical Equipment
- Adaptive equipment/Devices
- Assistive Technology
- Mobility Interventions
- Environmental modifications
- Exercise
- Splints
Terminal disease

• Health and wellness
  • Attention to routine health
  • Length of life varies
  • Common age related issues
  • Enlarged prostate
  • Low Vitamin B 12 or D
  • Low red blood cell folate

• Symptom control
  • Cramps/spasm
  • Sialorrhea
  • Pseudobulbar affect
  • Breathing
  • Equipment – moving target
  • Exercise not to exhaustion
  • Rest is restorative
We SUPPORT all paths to live with ALS

Medication, equipment

Noninvasive breathing machines

Tracheostomy and Ventilator
Lessons Learned

- Early education: medication, adaptive equipment, assistive technology addressed by all team members
- Feeding tube placement early with post op extended monitoring
- Psychology: sessions with other disciplines
- Staff more open to personal issues
DESPITE THE SIGNIFICANT STRESSORS AND CHANGES PRECIPITATED BY ALS, WHAT MAY \textit{PROTECT} INDIVIDUALS FROM DEPRESSION?
Resilience Factors:

(Connor & Davidson 2003; Min et al. 2013)

- Active Coping Strategies
- Sense of worth and purpose
- Social Support
- Spirituality
- Internal Locus of Control
- Self-Efficacy
- Planning
- Problem-Solving
- Use of Humor
- Cognitive Flexibility
- Hope
Majority of patients show successful adjustment post-SCI (Kilic 2013)

Inversely related to distress* and depression** (Ziaian 2012; Catalano et al. 2011)

Positively correlated to life satisfaction** (Catalano et al. 2011), and quality of life*

“Buffers” perception of stress and depressive symptoms** (Catalano et al. 2011; Min et al. 2013)

Protects against psychopathology and is associated with favorable response to MH treatment (Min 2013)
• One factor of the construct of subjective well-being [and subsumed under the larger construct of QOL]

• SWL viewed as a vital component of QOL in individuals adjusting to acquired disability (Hernandez, Elliott 2014)

• No studies examining satisfaction with life in ALS → QOL more frequently cited

**QOL in ALS:**

– QOL as the most important criterion for decisions regarding life-sustaining treatment [Kurt 2007]

– Significant others underestimate QOL [Kubler 2005]
  - 60% caregivers not in favor of ventilation; 97% of patients were [Trail et al. 2003]

– Negatively related to Depression [Badger 2001]

– Anticipated poor QOL leading reason for requests for PAS [Bascom 2002]
Depression and ALS

- Illness progression not associated with distress or depression [Rabkin 2000]
- Caregivers as likely to be clinically depressed as patients [Rabkin 2000]
- High concordance of distress and depression between patients and caregivers [Rabkin 2000]

- Protective factors
  - Religious faith
  - Family/social support
- Risk Factors
  - Illness symptoms
  - Suffering and pain
  - Perceived family burden
- Desire for hastened death [Rabkin 2000]
  - Depressive symptoms
  - Hopelessness
  - Suffering
  - Fatigue
DEPRESSION VS. DISTRESS: VETERANS WITH AMYOTROPHIC LATERAL SCLEROSIS
Study Aims

- **Determine whether our sample of individuals with ALS have higher rates of depression** [vs. are symptoms reflective of distress??]

- **Identify predictive and mediating relationships** among distress, depression, satisfaction with life, and resiliency

- **Examine the stability of these factors over time**

- **Examine medical variables (ALS Phenotype)** and their relationship with distress, depression, life satisfaction, and resilience.
Psychotherapy in ALS [Kurt 2007]

• No specific psychotherapeutic intervention for ALS or other motor neuron disorders

• Implications for treatment
  – Lower distress had longer survival time

• Not all ALS treatment is multidisciplinary

• Germany survey [Kurt 2007]
  – German neurological centers
  – 3/20 didn’t answer “no time”
  – MH issues handled by neurologists via medications or referral in “extreme cases”
  – 11/20 no psychologist employed
  – 3/9 centers with psychologists offered long term psychological care

• 1/11 patients with severe depressive symptoms provided with psychotherapy on a regular basis
Depression and ALS

• Lower rates than other motor neuron disorders [Taylor et al. 2010]
  – PD: 45%
  – HD: 50%
  – ALS < 20% (severe depression)

• Depression not associated with ventilation status

• Not associated with progression of disease [nor increased distress 5.5 months posttest] [Rabkin 2000]
Prevalence of Depression for ALS Patients

MDD or PHQ-2
- 2011: 33%
- 2012: 23%
- 2013: 13%
- 2014: 21%
- 2015: 16%

PHQ-9 (> 10)
- 2011: 20%
- 2012: 17%
- 2013: 10%
- 2014: 3%
- 2015: 6%
Patient Distress Changes

<table>
<thead>
<tr>
<th></th>
<th>At Start</th>
<th>1Y</th>
<th>2Y</th>
</tr>
</thead>
<tbody>
<tr>
<td>Global Bulbar</td>
<td>4.6</td>
<td>4.2</td>
<td>4.3</td>
</tr>
<tr>
<td>Global Cervical</td>
<td>4.4</td>
<td>4.3</td>
<td>4.0</td>
</tr>
<tr>
<td>Global Lumbar</td>
<td>5.1</td>
<td>5.5</td>
<td>4.0</td>
</tr>
<tr>
<td>Flail Arm</td>
<td>4.5</td>
<td>4.0</td>
<td>2.0</td>
</tr>
<tr>
<td>Flail Leg</td>
<td>5.6</td>
<td>4.7</td>
<td>3.6</td>
</tr>
<tr>
<td>PLS</td>
<td>4.3</td>
<td></td>
<td>3.6</td>
</tr>
</tbody>
</table>
Spouse/Significant Other Distress Changes

<table>
<thead>
<tr>
<th></th>
<th>At Start</th>
<th>1Y</th>
<th>2Y</th>
</tr>
</thead>
<tbody>
<tr>
<td>Global Bulbar</td>
<td>6.464</td>
<td>2.0</td>
<td></td>
</tr>
<tr>
<td>Global Cervical</td>
<td>6.566</td>
<td>6.0</td>
<td>6.0</td>
</tr>
<tr>
<td>Global Lumbar</td>
<td>7.2</td>
<td>5.7</td>
<td></td>
</tr>
<tr>
<td>Flail Arm</td>
<td>7.0</td>
<td>5.0</td>
<td>4.3</td>
</tr>
<tr>
<td>Flail Leg</td>
<td>6.8</td>
<td>6.0</td>
<td>4.5</td>
</tr>
<tr>
<td>PLS</td>
<td>6.0</td>
<td>5.0</td>
<td></td>
</tr>
</tbody>
</table>
One of the first investigations of mental health variables in Veterans with ALS.

- One of the largest sample sizes of ALS patients with focus on psychological variables.
- One of the few published studies looking at psychological variables in ALS over time.
CLINICAL RELEVANCE: MAJOR TAKE-HOME POINTS

• Depression rates in this sample were relatively low and akin to other studies that used diagnostic criteria-based methods.
  – → Use consistent measures/methods to report these rates!

• STILL, depression rates are x2 higher than the general population
  – → Don’t ASSUME, but routinely ASSESS
CLINICAL RELEVANCE:
MAJOR TAKE-HOME POINTS

- Distress was frequently reported in our sample
  - ASSESS and target modifiable stressors in treatment

- RESILIENCE predicted depressive symptoms and mediated the relationship between distress and depression.
  - Identify and target modifiable components of resilience in treatment

- Possible influence on ALS type on depressive symptoms
Transdisciplinary Collaboration In Addressing Complex Clinical Issues Associated with ALS
Learning/Decision Making in Multi & Interdisciplinary Practice: Relay

- Speech Pathology
- Psychology
- Dietary
- Physical Therapy
- Occupational Therapy
- Physician
- Nursing
- Respiratory Therapy

Minimal or no Role-release

Karol RL. Neurorehabilitation 2014; 34: 655-669
Knowledge is Power

- Multi-disciplinary approach
- Educating patient
  - Explain procedure
  - How tube is cared for and tube feeds are given
  - “Patient Rocks” if she accepts
- But done individually not a shared responsibility
Transdisciplinary Team

• changes the relationship between the disciplines and clinical issues.
• do not divide up the person based upon discipline specific areas of expertise as do multi-disciplinary teams
• Incorporates treatment plans, teaching approaches and interventions

(Karol, 1986)
Interdisciplinary Practice

- Medicine
- Nursing
- Occupational Therapy
- Speech Pathology
- Respiratory Therapy
- Physical Therapy
- Psychology
- Dietary
- Biomedical Engineering
- Home Visitation
- Telemedicine

Patient
Transdisciplinary Collaboration In Addressing Complex Clinical Issues Associated with ALS
Transdisciplinary Team

- Issue Focused

- The coordinator facilitates the care and communication among team members

- Requires flexibility, and receptiveness: individual roles less distinct

- Changes the relationship between the disciplines and clinical issues. (Karol, 1986)
Mean Survival ALS Population Per FY

<table>
<thead>
<tr>
<th>Year</th>
<th>Survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>2007</td>
<td>1.5</td>
</tr>
<tr>
<td>2008</td>
<td>2.0</td>
</tr>
<tr>
<td>2009</td>
<td>1.9</td>
</tr>
<tr>
<td>2010</td>
<td>2.0</td>
</tr>
<tr>
<td>2011</td>
<td>1.8</td>
</tr>
<tr>
<td>2012</td>
<td>2.7</td>
</tr>
<tr>
<td>2013</td>
<td>2.8</td>
</tr>
<tr>
<td>2014</td>
<td>5.1</td>
</tr>
</tbody>
</table>
Summary: Focusing on patient issues and practicing role-release may improve survival, shorten hospital stay and improve health-related quality of life even as the search for therapies continues.
Questions?
THANK YOU