

**HOSPICE  
OF THE  
WESTERN  
RESERVE**

**ALS  
ASSOCIATION  
Northern Ohio  
Chapter**


## Transitions in the Illness Trajectory of the ALS Patient

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### Collaborative Model

The ALS Association and Hospice of the Western Reserve

1. ALS/MND has a poor prognosis, often 2 to 5 years life expectancy and a relatively rapid decline in function, independence and health.
2. Symptoms are often significantly progressed at the time of diagnosis since the diagnosis is one of exclusion.
3. Many patients are not seen at ALS interdisciplinary (IDT) clinics.
  - a. many ALS IDT clinics are great distances from patients,
  - b. disease progression prevents return to clinic, and
  - c. outpatient model may be ill-equipped to provide appropriate care for complex patient with MND
4. Patient and family often feel isolated and on their own, lack support and trusted guidance.
5. Care providers suffer caregiver stress and burnout through lack of resources for client.
6. Collaborative model fits with Chapter, palliative and hospice care throughout disease process with the goal to improve access to care, treatment of symptoms, and quality of life.



## Amyotrophic Lateral Sclerosis

A = loss  
Myo = muscle  
Trophic = nourishment  
Lateral = areas of the spinal tract  
Sclerosis = scarring

Also known as: Lou Gehrig's Disease/ MND



## Motor Neuron Disease

- a group of progressive neurological disorders
- destroy cells that control voluntary muscle activity
- Eventually, ALL voluntary movement can be lost and can be completely paralyzed.
- Life expectancy is usually 2-5 years, while 10% live 10 years or more.
- Death usually related to respiratory failure and/or pneumonia.




## No test to diagnose

- Diagnosis often takes 8 to 12 months or more depending on initial symptoms.
- Must rule out all other diagnosis such as stroke, spine injuries, nerve injuries, other disease processes such as MS, Bell's Palsy, Myasthenia Gravis, Parkinson's...
- Many people undergo surgery for nerve injuries and present with poor recovery
- Blood work, MRI, CT, Muscle Biopsies, EMG, clinical observations



## Who gets ALS?

- Most commonly diagnosed in mid 50's, but can occur from 20 -80+.
- No boundaries across gender, ethnic or socioeconomic status.
- Slightly more men than women (1.5:1)
- MNDs may be inherited (genetic) or acquired.
- Can occur as
  - Limb onset, (most common- foot drop)
  - Bulbar onset (speech and swallowing affected) or
  - Bi-brachial (very focal spinal onset)- often present as COPD or respiratory failure
- In many cases, the mind remains unaffected.
  - new research focuses on Frontal Temporal Dementia



## Risk Factors

- **Veterans** are twice as likely to develop ALS- unknown reasons, theories include excessive stress, excessive exercise, immunizations, chemical exposure, etc.
- The "**Perfect Storm**": Combination of specific genetic makeup, environmental influence and chance. (known cases of identical twins in which one twin developed ALS and the other did not).
- **Genetic forms** account for approximately 10% of the cases- almost 30 genes identified- learning more everyday.
- Most cases are **sporadic**- they come out of the blue with no other family links.
- Football players may appear to be at greater risk but may be related to **frequent concussion injuries**- research currently underway to distinguish symptoms and physical damage within the brain and spinal column.
- Oxidative stress and free-radicals as well as proteinopathies are thought to be related causality agents.
- Current research looking a **blue-green algae blooms** and cyanobacteria which is thought to be neurotoxic.



## How many people live with ALS?

- Thought to affect ~30,000 Americans at any given time
  - No mandatory reporting system at the state or federal level
  - There is now a National ALS Registry where patients can self-enroll
  - Every 90 minutes, a person is diagnosed with ALS, while another loses his/her battle
  - In northern Ohio, the Chapter follows ~450 persons each year (33 counties).
- First drug approved by the FDA in 1993 (Riluzole) slows the rate of disease 2-3 months in some people.
- Second drug approved in May 2017- (Radicava) began use August 2017. Slows progression in some people with ALS. Expected to cost \$146,000 per year.



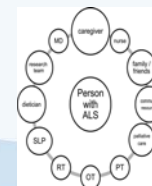
## Medicare and Social Security Benefits

- Advocacy efforts by The ALS Association have helped to provide **Presumptive Social Security Disability Benefits** (don't have to prove disability) and **Medicare** health insurance access at 6 months if younger than age 65 provided SS credits have been earned.
- Advocacy efforts also helped to provide **VA Service Connected** benefits to eligible veterans. Spousal compensation also available after the death of the veteran.
- Some public and state employee plans have allowed access to Medicare benefits at the time of qualifying (i.e., at 6 months).



## What is affected?

- Mobility
- ADLs/Independence
- Speech/swallowing
- Nutritional Needs
- Respiratory Support
- Social Work/Psychosocial Adjustment
- Further detailed needs often include:
  - Secretion Management
  - Constipation/Diarrhea
  - Frontal Temporal Dementia
  - Pseudo-Bulbar Affect
  - Anxiety and Breathing
  - Skin Integrity, Contractures, Cramps/Spasms



## Case Overview

- Mrs. Jones is a 71 year old who was referred to the ALS association after confirmation today of the diagnosis of ALS after 8 months of evaluation. She initially presented to her PCP with complaints of slurred speech, weakness, and excessive saliva with nighttime drooling. She complained of being easily fatigued and reported that climbing steps is hard, with her slow and feeling as though she is at times "climbing a mountain" with shortness of breath causing her to pause. She had EMG testing and was told she has ALS.



## Communication options

(when speech is not intelligible)



### Intact Hand Function

- Low tech: pen/paper (fatter marker preferred over pen), dry erase board, electronic LCD board, paper letter board,
- High tech: smart phones, iPad and/or tablet with text-to-speech apps, laptop computers, notes apps or text apps
- Insurance funded speech generating devices must be obtained prior to hospice enrollment and generally are reserved when hand function no longer functional for access.



## Communication Options

Poor hand control

- Adapted devices to hold marker/pen and stylus.
- Laser pointer mounted on eye glasses or hat/headband with paper letter board.
- Windows tablet computer with low cost speech software using alternative mouse devices such as headmouse and eye gaze devices as add-on.
- Insurance funded speech generating devices must be obtained prior to hospice enrollment





## Communication Options



Poor hand control, poor head control, poor eye control (eyeglass interference, medication interference, arousal state interference)

- Partner assisted auditory scanning methods
- Forced choice: always allow for "I don't know" response" not only yes/no




## Communication Options

**Website links for further self study**

Laser Pointer with head movements:  
<https://www.youtube.com/watch?v=AooDQOzdOvE>

Partner Assisted Auditory Scanning with no hand, head or neck control:  
<https://www.youtube.com/watch?v=pLb6-Oi3uR0>

FREE Communication Boards for adults and children:  
<http://www.amyspeechlanguagetherapy.com/communication-boards.html>

Where to purchase the laser seen in the picture and on the video:  
<http://store.lowtechsolutions.org/testimonials/>




## Transition #2: "From Unknown to Known" *Palliative Care*





## ALS Case: Progression of Disease

- Mrs. Jones' disease has progressed. She has had increased dysphagia, weight loss, dyspnea and her cough is weak. Her speech has changed and sometimes she needs to use the writing board as it is becoming more difficult for her family to understand her. She has marked drooling which is distressing to her. She is only able to eat small amounts of soft/pureed foods. She feels weak and clumsy and is now using a 4 pronged cane to ambulate. She feels short of breath with minimal exertion, she has been using a BiPAP intermittently. She is referred to palliative care for symptom management/goals of care.



## Care Options

1. Riluzole 50 mg PO q 12 hrs
2. Multidisciplinary Care
3. Early use of Noninvasive ventilation
4. Timing of a feeding tube
5. Managing symptoms

## Supporting Quality of Life

- Functional Support
- Respiratory Symptoms
- Discomfort
- Nutrition
- Psychosocial
- Spiritual




## Symptom Management


- Mobility
- Pain Management
- Mobilize Secretions
- Supportive Orthotics






## Emotional Care


- Reframing hope
  - Hope for a cure
  - Search for information
  - Acceptance
  - Living in the moment
  - Self Transcendence
- Shifting concern of being a "burden"



## ALSFRS -R

Monitor function status in patients with ALS

Measure	Points
Speech	0-4
Salivation	0-4
Swallowing	0-4
Handwriting	0-4
Cutting food and handling utensils;	0-4
Dressing and hygiene	0-4
Turning in bed and adjusting bed clothes	0-4
Walking	0-4
Climbing stairs	0-4
Breathing	0-4



## Serious Illness Conversations

*What are your most important goals as you journey through your illness?*

- Patient's Values
- Patient's Quality of Life
  - Feeding Tube Placement?
  - Ventilation goals?



## Transition #3 Known to Unknown-Hospice: Less than 6 months

- Prognostication: Fatal Disease Typically 2-5 years, (3-5 years)
- Age of onset: some data suggests 50 years and younger average survival >3 years
  - Age 80 or older average survival just over 2 years
  - Evaluated by using functional scales (ALSFRS-R score)=subjective
  - Loss of muscle mass
  - Inflammatory markers CRP : Elevated suggestive of progressing faster
  - Death 2/2 to respiratory failure



## ALS: Hospice Eligibility

- Impaired Cognitive function
  - Attention changes, decreased coherent thoughts, changes in memory, signs of disorientation, decreased ability to take on simple/multiple tasks and daily routines. Cognitive changes
- Impaired Neuromuscular function
  - Dysphagia
  - Impaired muscular strength: hypotonia, hypertonia, spasticity, involuntary movements
  - Impaired muscle strength



## ALS : Hospice Eligibility

- Impaired Respiratory Function : Vital capacity less than 30% of normal. Dyspnea at rest. Use of BiPAP, Impaired cough
- Impaired Mobility : Progressive decline
- Functional Changes: PPS, Progressive dependence in ADL, Impaired movements, Falls
- Nutritional changes: Dehydration, BMI , Weight loss
- Recurrent infections, aspiration pneumonia



## What to expect: Anticipatory Grief

- Anticipatory Grief: Family and patient, bereavement support
  - "Anticipatory grief can be a big part of the ALS process for diagnosis to the end-of-life decisions and loss. ALS is a disease of continually diminishing capacities. This continual change is a hardship on the person living with ALS as well as on caregivers and loved ones".
  - "The anticipatory grief did not necessarily alleviate our grief as we approached the end of life, but I think it helped us open up communication channels to discuss changes in caregiving needs and plan for end of life wishes and decisions as well as allowed us to establish support networks (e.g., hospice team relationships). These preparations were invaluable to us as we traversed the end-of-life stages and the time immediately following the loss of my mom". Jacqueline Tripi Pfadt daughter of person with ALS



## Coping w/ Anticipatory Grief

- Walks
- Journaling
- Plan for the future
- Talk to someone-Counselor, Social worker, Spiritual Care
- Do things you want to do now-postpone chores
- Make changes only as needed, put off major decisions when possible
- Spend time with loved ones
- Seek help
- Spiritual Assistance



## Hospice: Spiritual Support

- Spirituality: Personal quest to find meaning and purpose in life and a relationship with something greater than oneself.
  - May be found in connections, relationships, faith, religion and meanings that give life passion, commitment and hope.
  - Tapping our inner spirituality differs for each person. Importance is giving meaning and purpose to one's life.
- \*\*\*Spiritual care is a part of the hospice interdisciplinary team
  - Methods to connect with Spirituality : Ritual, Meditation, Relaxation techniques, Legacy building, Life review



## Hospice: Legacy Building

- Distribution of sentimental/personal effects
- Journaling
- Cards, Letters, Blogs
- Sharing pictures
- Writing poetry, Music, Art
- Video
- Planning memorial, personal statements



## Hospice : What to expect

- How much information does the patient want to know
- Common concerns:
  - What does death look like in ALS
  - Symptom management
  - Withdrawal of life sustaining measures
  - Palliative Sedation



## ALS: Case: Final days

- Mrs. Jones is a 71 year old with 2 year history of ALS> She was admitted to hospice level of care 7 months ago and has declined slowly since. She was able to walk with her walker however over the last 3 weeks is no longer able to do so independently and is now only able to ambulate a few steps with help. She has a feeding tube and feels bloated much of the time. She c/o constant dyspnea and has become dependent on her BiPAP only taking it off for less than 5 minutes at a time. She has had continued difficulty with sialorrhea despite medications. She is concerned about becoming burdensome to her family and says she is tired. "I do not want to live like this" She asks if she were to leave the BiPAP off, what would happen.



## Transitions ALS: Final-Withdrawing of Life-Sustaining Therapies

- HPNA Position Statement:
  - All life sustaining therapies may be withheld or withdrawn. There is no difference, ethically or legally between the decision to not initiate a treatment that may not be beneficial or stop or remove a treatment that is not beneficial and/or **no longer wanted**



## Transitions ALS: Peaceful Death

- HWR Practice: Withdrawal of External Respiratory Support
- Requires TDT assessment prior
- Atmosphere, Environment, Education/Expectation, Support
- Pre-medicate: Requires both Opioid and Anxiolytic, consider also phenobarbital.
- Start of procedure: Morphine, Midazolam
  - Intent is to relieve symptoms not to hasten death.



## Peaceful Death

- Mrs. Jones died peacefully in her home surrounded by her family, minister, favorite music approximately 45 minutes after withdrawal of his BiPAP. Family was grateful for this final gift.



