Primary Lateral Sclerosis and Amyotrophic Lateral Sclerosis: The “Ups” and “Downs” of Motor Neuron Disease

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Disclosures

Drs. Howard and Ma have no financial conflicts of interest to disclose.

PESG and PVA staff have no interest to disclose.

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Learning Objectives

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

1. Describe the distinguishing features of PLS/ALS from traditional ALS.

2. Describe the prognosis for an individual with PLS/ALS and how it differs from typical ALS.

3. State two unique considerations for rehabilitation of the person with PLS/ALS.
Phenotypic Variability in ALS

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NEUROMUSCULAR NEUROLOGIST

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Topics

- ALS history and terminology
- Diagnosis
- Variability:
  - Progression
  - Location of onset
  - Age and penetrance
  - UMN vs LMN (PLS?)
  - Genetic
- Nonmotor manifestations
The primary motor system
Terms

Motor neuron disease

ALS
- PMA
- Classic
- PLS

Other
- SMA
- Kennedy
Jean-Martin Charcot (1825–1893)

Amyotrophic lateral sclerosis:
“No nourishment to muscles and hardening of the lateral spinal cord”
ALS diagnosis criteria

When diagnosing ALS, the Awaji-shima consensus recommendations look for either “clinical” or “electrophysiological” evidence of ...

| Clinically definite * | UMN + LMN signs in bulbar region + ≥ 2 spinal regions; or  
<table>
<thead>
<tr>
<th></th>
<th>UMN + LMN signs in 3 spinal regions*</th>
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<tbody>
<tr>
<td>Clinically probable</td>
<td>UMN + LMN signs in ≥ 2 spinal regions and “with some UMN signs necessarily rostral to (above) the LMN signs”</td>
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</table>
| Clinically possible   | UMN + LMN signs in 1 spinal region; or  
|                       | UMN signs in ≥ 2 spinal regions; or  
|                       | LMN signs are found rostral to UMN signs,  
|                       | ONLY AFTER the appropriate neuroimaging and laboratory test are performed to exclude other possible differential diagnosis that may mimic ALS |

*Spinal regions: upper limbs/ lower limbs/ thoracic/ bulbar

Rate of progression

Swinnen
Predictors of prognosis

Factors Influencing the Rate of Progression in ALS

<table>
<thead>
<tr>
<th>Factor</th>
<th>Associated with Longer Survival</th>
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<tr>
<td>Phenotype</td>
<td>Flail limb variant, LMN-predominant disease, UMN-predominant disease, prolonged interval to diagnosis</td>
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<td>Demographic features</td>
<td>Younger age at diagnosis</td>
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<table>
<thead>
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<th>Phenotype</th>
<th>Associated with Shorter Survival</th>
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<tr>
<td>Phenotype</td>
<td>Bulbar onset ALS, respiratory onset ALS, cognitive impairment, impaired nutritional status, neck flexor weakness</td>
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<tr>
<td>Demographic features</td>
<td>Older age at diagnosis, lower economic status, smoking</td>
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</table>

Simon 2014
Location of onset

a. Spinal onset
b. Bulbar onset
c. Progressive muscular atrophy
d. Primary lateral sclerosis

e. Pseudopolyneuritic ALS
f. Hemiplegic ALS
g. Flail arm syndrome
h. Flail leg syndrome

Swinnen 2014
SPINAL onset

- Most common phenotype
- Asymmetric onset in limb
- Painless weakness
- Spread to other regions
- Rule out MMN, IBM
BULBAR onset

- 20% of ALS patients
- “Bulbar” refers to the medulla
- Dysarthria, dysphagia and tongue fasciculations
- Jaw jerk, palmomental reflex
- Pseudobulbar affect
- Rule out Kennedy disease
Variations in age and penetrance

- Usually starts in 5\textsuperscript{th}-6\textsuperscript{th} decade
- Juvenile ALS starts <25 yo
- Younger patients bias UMN phenotype

- Patients carrying mutations in the same ALS-associated allele may vary in onset age, location, motor neuron type
Juvenile ALS

- Hereditary (autosomal recessive or dominant)
- Manifests before age 25
- Slowly progressive - survival often greater than 20 years
- Mutations in FUS, alsin, senataxin, and SPG11 genes
UMN vs. LMN

PMA: progressive muscular atrophy
LMN: lower motor neuron
UMN: upper motor neuron
PLS: Primary lateral sclerosis
The primary motor system

**LMN lesion:**
- Weakness
- Atrophy
- Fasciculations

**UMN lesion:**
- Spasticity
- Hyperreflexia
- Slow mov’t
LMN predominant

- Preferential involvement of the lower motor neuron
- Progressive muscular atrophy (PMA) accounts for 5% of patients with motor neuron disease
UMN predominance

- Most eventually develop LMN signs
- Frank incontinence is rare
- Babinski reflex tends to be preserved

Ravits 2009
Primary lateral sclerosis (PLS)

- 5% of ALS patients
- Often symmetric lower limb onset
- Slower progression
- Sparing of respiratory function
- Less weights loss
- Rule out MS, HSP
Does primary lateral sclerosis exist?
A study of 20 patients and a review of the literature

Nadine Le Forestier,1 Thierry Maisonneuve,2 Ambre Piquard,1 Sophie Rivaud,3 Lise Crevier-Buchman,4
François Salachas,1 Pierre-François Pradat,1 Lucette Lacomblez1,5 and Vincent Meininger1

Pringle Criteria:

Clinical
1. Insidious onset of spastic paresis, usually beginning in lower extremities but occasionally
   bulbar or in an upper extremity.
2. Adult onset, usually fifth decade or later.
3. Absence of family history.
4. Gradually progressive course (i.e. not step-like).
5. Duration ≥3 years.
6. Clinical findings limited to those usually associated with corticospinal dysfunction.
7. Symmetrical distribution, ultimately developing severe spastic spinobulbar paresis.
# Does PLS exist

Table 4 *Electrophysiological and histological motor results*

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</table>

*Number of electrophysiological studies carried out.*
ALS genetics

Renton 2013
Autosomal dominant
Copper-zinc (Cu,Zn) superoxide dismutase type 1 (SOD1)
chromosome 21q22
SOD1$^{G93A}$ Current mouse model for ALS
Reactive oxygen species

Mitochondrial Respiration → $O_2$ → $O_2^-$ → SOD → $O_2 + H_2O_2$

Reaction:
- Fe-S Clusters
- Fe-S Clusters
- Protein Thiols

1. Catalase
2. GPx
3. Prx

$O_2 + H_2O_2$ → $H_2O + O_2$ → $\cdot OH$

Fenton Reaction:
- Nucleic Acids
- Lipids
- Amino Acids

1. GSH
2. AA

$H_2O$
LOSS OF FUNCTION? Mutations in SOD1 reduce total SOD activities only by 30-60%

GAIN OF FUNCTION? Mutant SOD1 mice develop ALS but SOD1 knockouts don’t
Over 140 mutations of the SOD1 gene have been reported

- **A4V**: LMN, rapid progression
- **I113T**: UMN+LMN, Low penetrance
- **A4T**: LMN, legs > arms
- **H46R**: Distal leg, very slow (12yr)
- **A89V**: Painful sensory involvement
- **G93C**: Bulbar, very slow (13yr)
Nonmotor: Cognitive manifestations

- Up to half of ALS patients have cognitive deficit
- Up to 25% of ALS patients meet criteria for frontotemporal dementia
- Mostly behavioral variant (rather than language)

FTD bv
- Apathy
- Disinhibition
- Stereotyped behavior
- Compulsivity
- Executive dysfunction
- Psychosis
C9orf72

A. GWENT#1

B. DUTCH#1

- ALS
- FTD
- ALS-FTD
Patients with frontotemporal dementia (FTD) with no known diagnosis of ALS or family history of ALS were clinically and electrophysiologically assessed for the presence of ALS.
Are amyotrophic lateral sclerosis patients cognitively normal?

C. Lomen-Hoerth, MD, PhD; J. Murphy, PhD; S. Langmore, PhD; J.H. Kramer, PsyD; R.K. Olney, MD; and B. Miller, MD

“Word generation, a simple frontal task that takes <2 minutes was tested in 100 consecutive patients with ALS seen in the authors’ multidisciplinary clinic”

Table 2 Patient characteristics of 44 ALS patients who underwent detailed neuropsychological testing

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>ALS with FTLD</th>
<th>ALS without FTLD</th>
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<tbody>
<tr>
<td>No. of subjects</td>
<td>23</td>
<td>21</td>
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<td>Age, y</td>
<td>65, range 42–80</td>
<td>54, range 37–81</td>
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<td>Sex</td>
<td>14M, 9F</td>
<td>13M, 8F</td>
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<td>Site of onset</td>
<td>11 bulbar, 12 limb</td>
<td>8 bulbar, 13 limb</td>
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<td>Family history</td>
<td>6 dementia, 2 PD, 4 ALS</td>
<td>3 ALS, 1 PD</td>
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<tr>
<td>FVC</td>
<td>66% predicted, range 0°–117</td>
<td>95% predicted, range 32–157</td>
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<tr>
<td>ALSFRS score</td>
<td>34, range 17–46</td>
<td>37, range 22–47</td>
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</tbody>
</table>
C9orf72 genetics

- 2-20 repeats are commonly seen in the healthy population
- 20-few hundred repeats are not often seen in either patient or healthy groups (intermediate lengths), and confer uncertain risks
- >few hundred repeats are pathogenic
1. Reduced C9orf72 protein and function

Repeat containing C9orf72 DNA

2. RNA toxicity: sequestration of RNA-binding proteins

(GGGGCC)_n sense RNA

(GGCCCC)_n antisense RNA

3. Translation into toxic DPR proteins

Nucleus

Cytoplasm

Neuron

Rohrer 2015
Nonmotor : ALS-PDC

GUAM
Lumping vs. Splitting
“the prognosis, up to the present, is of the gloomiest.......the verdict we will give such a patient tomorrow will not be the same we must give this man today.” - Charcot (150 years ago)
1. C9orf72 expansions in frontotemporal dementia and amyotrophic lateral sclerosis. 

2. RNA targeting therapeutics: molecular mechanisms of antisense oligonucleotides as a therapeutic platform. 

3. Antisense oligonucleotide therapeutics for inherited neurodegenerative diseases. 

4. The overlap of amyotrophic lateral sclerosis and frontotemporal dementia. 

5. Length of normal alleles of C9ORF72 GGGCC repeat do not influence disease phenotype. 


7. C9orf72; abnormal RNA expression is the key. 

8. Mutations in Cu/Zn superoxide dismutase gene are associated with familial amyotrophic lateral sclerosis. 


14. ALS: A bucket of genes, environment, metabolism and unknown ingredients S0301-0082(15)30090-3 Monica Zuﬁr´ia Francisco Javier Gil-Bea

15. ALS: Recent Developments from Genetics Studies Curr Neurol Neurosci Rep (2016) 16: 59 Martine Therrien1 & Patrick A. Dion2 & Guy A. Rouleau2


17. Autophagy in motor neuron disease: Key pathogenetic mechanisms and therapeutic targets Molecular and Cellular Neuroscience 72 (2016) 84–90 Maria Sara Cipolat Mis 1, Simona Brajkovic 1, Emanuele Prattini, Alessio Di Fonzo, Stefania Corti


19. Impaired Autophagy and Defective Mitochondrial Function: Converging Paths on the Road to Motor Neuron Degeneration doi: 10.3389/fncel.2016.00044 Brittany M. Edens1,2, Nimrod Miller 1,2 and Yong-Chao Ma

20. There has been an awakening: Emerging mechanisms of C9orf72 mutations in FTD/ALS Aaron D.Gitler a,n, HitomiTsuiji
Bend and Not Break: Rehabilitation of Upper Motor Neuron- Variant ALS

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From diagnosis to management

- Charcot describes ALS in 1874
- PM&R Specialty founded in 1947
- ALS becomes a chronic illness in ?

Diagram:
- A timeline from 1874 to 1947 and beyond.
The Case of the Tin Man

- 63-year old retired welder presented to clinic
- Symptom onset 7 years prior to presentation
  - Difficulty kick starting motorcycle
  - Speech changes
- Boss thought he was intoxicated (poor balance, dysarthria)
• Fragmented care at outside clinics
• Frequent falls, using trekking poles for balance
• Mood swings - inappropriate laughing and crying
• Withdrawing from social situations due to communication barrier (son has to “translate”)
• Difficulty swallowing, but no weight loss
• Leg pain from spasticity
• Service connection turned down due to “PLS”
Motor neuron diseases are thus categorized by the location of the diseased cells. Diseases that only affect the lower motor neurons are not relevant to this discussion. ALS is a disease process that involves both the lower and the upper motor neurons. PLS, by contrast, is a disease process that involves only the upper motor neurons.

- Is this accurate?

- Should this Veteran receive care in the ALS clinic?
Functional presentation PLS/ALS- how is this different from ALS?

Rehabilitation Interventions for PLS/ALS
- Spasticity
- Gait
- Bladder urgency
- Pseudobulbar symptoms

Community Resources for PLS
- Preserved muscle strength
  - Weakness of proximal hip girdle and hands
  - Usually >3/5 MRC
- Stiffness interfering with gait
- Hyperreflexia
- Slower progressing, longer disease duration

“[PLS] usually begins slowly and insidiously,. . .with some sense of weight, dragging and slight feebleness in one or the other leg, without pain. . . .. The condition progresses just as slowly as it commenced; the legs become stiffer and heavier, the gait progressively more labored, dragging, and distinctly spastic; occasional muscular cramps and contraction of the legs may occur, but nothing else. . .”

-Wilhem Erb, 1902
PLS: Common Symptoms

- Speech changes (95%)
  - Strained/nasal speech, slow and monotone speech
- Pseudobulbar involvement (85%)
- Cognitive impairment (80%)
- Urinary urgency (70%)
- Dysphagia without weight loss (60%)
- Severe spasticity (30%)

Le Forestier, 2001
### Table 1 Clinical characteristics of the patients

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex</th>
<th>Age at disease onset (years)</th>
<th>Duration of disease (years)</th>
<th>Follow-up period (months)</th>
<th>Site of disease onset</th>
<th>Pseudobulbar syndrome</th>
<th>Tetrapyrimal syndrome</th>
<th>Urinary urgency</th>
<th>Prefrontal and/or premotor dysfunction</th>
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B = bulbar; LLr/l/s = right/left/both lower limbs.
NEALS PLS cohort

- 20 NEALS consortium sites did chart reviews and identified 233 UMN-predominant patients
  - Mean disease duration: 7 years (!)
- Most common symptoms:
  - Spasticity (93%)
  - Abnormal gait (94%)
  - Dysarthria (68%)
  - Dysphagia (55%)
  - Pain (43%)
  - Dyspnea (28%)
  - Prevalence of urinary symptoms not reported
- Spirometry performed on 110 of these patients, mean FVC 73% of predicted, NIPPV used in 17% of patients
- Feeding tube placed in 8%

Fournier et al, 2016
Spasticity Management

Staple interventions:

- **Therapy**
  - Stretching
  - Splinting
  - Aerobic exercise*

- **Modalities**
  - Heat
  - Ice
  - Vibration
  - Electrical Stimulation
  - Aquatic therapy

*Drory, 2001
Spasticity Management

**Oral medications**
- baclofen*
- tizanidine*
- dantrolene
- gabapentin
- benzodiazepines

[Botox]
Spasticity Management

- **Intrathecal Baclofen Pump**
  - First described for use in PLS in 2005*
  - Trial important to gauge effect
  - >95% global satisfaction in >10-year follow-up**

*Buzetti Milano, 2005
**Mathur, 2014
Gait Management in PLS

- Spasticity management
- Maintain some stiffness for transfers
- Assistive device to mitigate fall risk
- Consider power mobility
Bladder dysfunction

- Prevalence in PLS: 50% -70%
  - Classic ALS: 4-40%*
- Urinary urgency + mobility limitations = misery!
- Pathophysiology not clearly understood
  - Detrussor hyperreflexia common
  - EUS sphincter dysfunction common

Arlandis, 2016
Bladder dysfunction

- Diagnostic interventions
  - Check PVR to exclude poor emptying
  - UA & Cx to exclude infection
- Treatment
  - Timed voids
  - Medications
  - Condom catheter
  - Indwelling foley
Mood lability

Not necessary to treat unless symptoms are disturbing to the individual

Treatment options
- TCA
- SSRI
- Nuedexta
  - Possible palliation of bulbar function: speech, swallow, salivation (Smith, 2016)
Spastic Dysarthria

**Spastic dysarthria example**

- Excess and equal stress
- Slow speaking rate
- Short phrases
- Imprecise articulation
- Hypernasality
- Strained strangled vocal quality
Spastic Dysarthria: Treatment

- May benefit from relaxation exercises
- Train to speak slower (due to extra time required to reach motor speech target)
- Train communication partners on communication strategies / environmental modifications
  - face each other when speaking, reduce room noise, allow person w/ PLS extra time for communication
- Evaluate/train AAC if dysarthria is severe
- Management of concurrent PBA (Nuedexta)
Dysphagia

- NEALS cohort:
  - High prevalence of dysphagia (55%)
  - Low prevalence of feeding tube placement (8%)
- Quarterly FVC and weight
Pain management

- 43% prevalence in NEALS report
  - 50% reported prevalence in classic ALS
- Pain often related to spasticity in UMN-predominant patients
Community Resources for PLS

Paralyzed Veterans of America

ALS Association

Spastic Paraplegia Foundation, Inc.

MDA Muscular Dystrophy Association
Conclusion

1. Motor Neuron diseases exist along a spectrum
2. PLS classically presents as slower progressing, severe spasticity, mild weakness, and hyperreflexia
3. Bulbar and bladder dysfunction more common than classic ALS
4. Aggressive spasticity management is critical to enhance function

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RECENT ACTIVITY

Maryann Hibbs 4 weeks ago

is anyone willing to share your current practice. SOP, or policy regarding ventilator patients in your extended areas. ALS
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