Amyotrophic Lateral Sclerosis: Update with a Focus on Diagnosis and Management of Communication Changes

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Disclosures

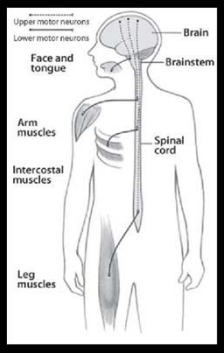
- Financial Disclosure – I receive a salary from Michigan State University.
- No relevant non-financial discloures.

Agenda

- ALS as a neurological condition
  - Motor Neuron Disease - Intro
  - A few of the common types
  - ALS
    - Epidemiology
    - Understanding of the etiology & pathogenesis?
    - Sign & Symptom Presentation
    - Clinical Diagnostic Process
    - Interventions
- Updates and Advances
- ALS a communication disorder
  - Signs & symptoms
  - Communication diagnostic process
  - Interventions

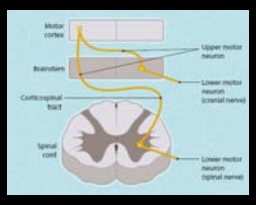
Motor Neuron Disorders

- Group of progressive neurological disorders
- Abnormality/destruction of motor neurons
  - Upper Motor Neuron
  - Lower Motor Neuron



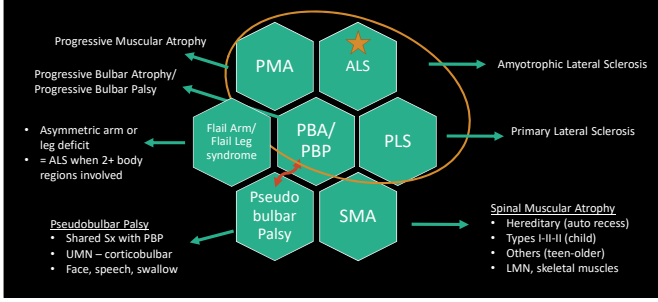
Motor Neuron Disorders

- **Adults** and children
- Men/boys and Women/girls
- familial and sporadic
  - Genetic
    1+ family members
    With ALS or FTD
  - ??? Genetic Environmental Viral ?



Some of the more common MNDs

link to ninds regarding MND



UMN and LMN Signs

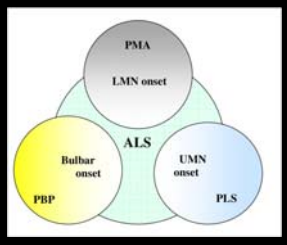
SIGN	Upper Motor Neuron	Lower Motor Neuron
Weakness	Yes	Yes
Atrophy	Disuse atrophy	Yes – marked & early
Fasciculations	No	Yes
Reflexes	Hyper-reflexia	Hypo-reflexia
Tone	Increased	Decreased
Nerve Conduction Velocity	Normal	Abnormal (motor nerves: reduced amplitude and delayed onset)
EMG Denervation Potentials	No	Yes

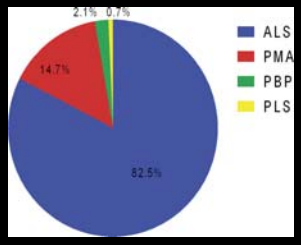
Distinguishing the MNDs

MND Type	UMN Involvement	LMN Involvement	Rate of Progression	Limb, Bulbar, both	Other Features
Amyotrophic Lateral Sclerosis	Yes	Yes	Fast	Can be Both	Respiratory failure; M >W
Primary Lateral Sclerosis	Yes	No	Slow	Can be Both	Normal lifespan
Progressive Muscular Atrophy	No	Yes	Slow	Hands usually, then lower body; trunk	Largely affects Men; progresses to ALS?
Progressive Bulbar Palsy	No/Some	Yes	Slow/fast	Bulbar [limbs but less prominent] .	Considered more rare

Revised El Escorial criteria = No; literature says may have some

Relationships Among





NINDS materials

ALS: Epidemiology

- CDC MMWR 11/23/2018
  - USA Prevelance 01 to 12/2015 = 5.2 per 100K
  - 16, 583 "definite" cases identified
  - 6,250 new "definite" cases 2015
  - Higher prevalence in Midwest likely reflects population demographics (more White)

TABLE. Number and percentage of amyotrophic lateral sclerosis (ALS) cases (N = 16,583) and estimated prevalence, by age group, sex, race and geographic region — National ALS Registry, United States, 2015

Characteristic	Population*	No. (%) cases	Estimated cases per 100,000 population (95% CI)
Age group (yrs)			
18–39	95,782,809	480 (2.9)	0.5 (0.5–0.6)
40–49	41,141,609	1,462 (8.8)	3.6 (3.4–4.1)
50–59	43,712,960	3,214 (19.4)	7.4 (6.9–7.9)
60–69	35,356,070	4,774 (28.8)	13.5 (12.9–14.1)
70–79	19,606,548	3,953 (23.8)	20.2 (19.4–21.3)
≥80	11,892,496	1,522 (9.2)	12.8 (12.3–13.4)
Unknown	—	1,178 (7.1)	—
Sex			
Male	158,138,060	10,098 (60.9)	6.4 (6.2–6.5)
Female	163,280,761	6,458 (38.9)	4.0 (3.9–4.1)
Unknown	—	27 (0.16)	—
Race			
White	243,635,466	13,074 (78.8)	5.4 (5.2–5.6)
Black	44,677,216	1,045 (6.3)	2.3 (2.2–2.5)
Other	—	958 (5.8)	—
Unknown	—	1,503 (9.1)	—
U.S. Census region†			
Midwest	67,907,403	3744 (25.6)	5.5 (5.4–5.6)
Northeast	56,283,891	2881 (17.6)	5.1 (5.0–5.2)
South	121,182,847	5676 (34.2)	4.7 (4.6–4.8)
West	76,044,679	3352 (20.2)	4.4 (4.3–4.5)
Unknown	—	930 (5.6)	—
Total	321,418,821	16,583 (100.0)	5.2 (5.1–5.3)

Substantial Growth in Research Past Decade

Mostly re: genetics and pathogensis, some drug Tx

- National ALS Registry - 2008
  - https://www.cdc.gov/als/
  - Culls databases to ID cases
    - Medicare
    - VA Health Admin
    - VA Benefits Admin
  - Self-enrollment – web portal
  - Biorepository → 2014

Understanding of the

- Familial ALS → 5% to 10% cases
- Sporadic ALS

genes → environment → time

Classification/Level of Certainty	Family History
Definite	≥ 2 First- or second-degree relatives with ALS (or FTD)
	≥ 1 Relative with ALS and gene-positive cosegregation
Probable	1 First- or second-degree relative with ALS
Possible	Distant relative (third degree or beyond) with ALS
	Sporadic ALS patient with no family history but positive for a FALS gene
	≥ 1 First- or second-degree relative with confirmed frontotemporal dementia
1st degree relative: parents, children, siblings 2nd degree relatives: grandparents, aunts, uncles	

50%

From Boylan K. 2016. Familial ALS. *Neurol Clin, 33*(4): 807-830

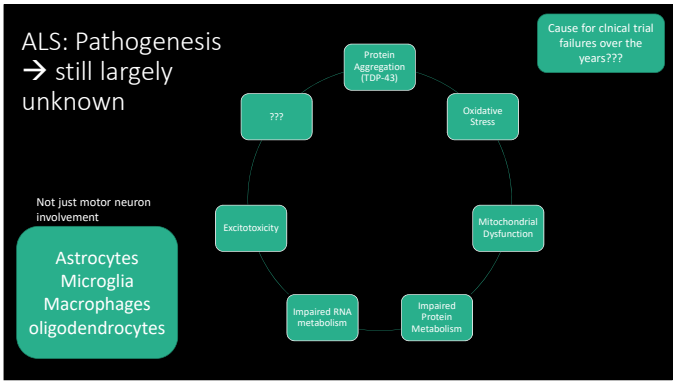
Understanding of the Etiology?

- Less advancement beyond genetic basis
- Assume environmental exposure and genetic risk interplay
- No irrefutable links of environmental factors and ALS

- Suspected/possible – but not confirmed
  - Athletic activity
  - Military service
  - Head trauma
  - Heavy metal and lead exposure
  - Electromagnetic field exposure
  - b-N-methylamino-L-alanine (cyanobacteria neurotoxin)

Gene ←→Time← → Environment Model (Al-Chalabi et al., 2013)

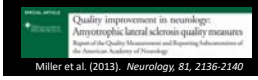
ALS: Pathogenesis → still largely unknown



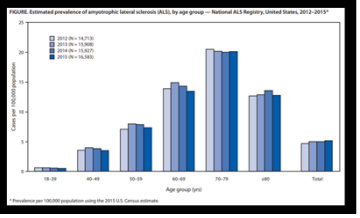
ALS:

- Increased likelihood for
  - Male [1.6 : 1 for M : F]
  - White [5.5 per 100K vs 2.3 per 100K for Black]
  - Non Hispanic
  - ≥ 60 years

- ~50% die within 2-5 years; ~20% live 5-10 years; and ~10% live years+
- Respiratory failure
- 20% - 50% with cognitive dysfunction or Dx FTD
- Familial ALS = shorter life expectancy on average



Miller et al. (2013). *Neurology, 81, 2136-2140*



ALS – phenotypic heterogeniety

Motor Signs

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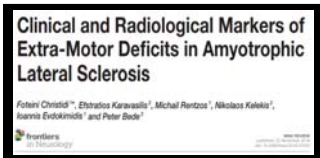
- Variable UMN/LMN involvement
- Variable site of onset in the body
- Variable rate of progression

- Causative genes not distinctly related to motor phenotype

- But → focal onset then progressive spread of motor symptoms
  - → usually along neuroanatomic pathways



- Cognitive
  - Link/continuum with Frontotemporal Dementia (FTD)

Links

TDP-43 protein in nearly all ALS cases and ~50% FTD

Hexanucleotide G:C repeat expansion of chromosome 9 open reading frame 72 gene (C9orf72) →
most common known genetic cause of ALS, accounting for 30% to 40% of familial ALS, and it also causes frontotemporal dementia (FTD). [Oskarrson et al., 2018]

ALS phenotype – beyond motor

- Sensory
  - Degeneration and malfunction of sensory neurons in ALS, particularly those wit SOD1 mutation [Pugdahl, et al., 2007; Pradat & El Mendili, 2014]
  - Sensory 'dying off' may preced motor deficits in ALS (Vaughan, et al., 2015)
  - Distal small-fiber neuropathy [Weis, et al. 2011; Truini, et al., 2015] – numbness, pain usually in hands, feet

- Behavioral Deficits
  - Apathy = most commonly reported [Dimensional Apathy Scale, specific for ALS, Radakovic, et al., 2016]
  - Disinhibition [Terada, et al., 2011]
  - Hallucinations – more associated with C9orf72 genotype [Snowden, et al., 2012]
  - Pseudobulbar affect [Bede, et al., 2018]


Clinical and Radiological Markers of Extra-Motor Deficits in Amyotrophic Lateral Sclerosis

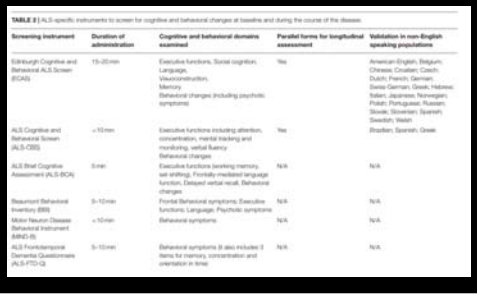


TABLE 2 | ALS-specific instruments to screen for cognitive and behavioral changes at baseline and during the course of the disease.

Screening instrument	Duration of administration	Cognitive and behavioral domains examined	Parallel forms for longitudinal assessment	Validation in non-English speaking populations
Edinburgh Cognitive and Behavioral ALS Screen (ECAS)	15–20 min	Executive functions, Social cognition, Language, Visuoconstruction, Memory, Behavioral changes (including psychotic symptoms)	Yes	American-English, Belgium, Chinese, Croatian, Czech, Dutch, French, German, Swiss-German, Greek, Hebrew, Italian, Japanese, Norwegian, Polish, Portuguese, Russian, Slovak, Slovenian, Spanish, Swedish, Welsh
ALS Cognitive and Behavioral Screen (ALS-CBS)	~ 10 min	Executive functions including attention, concentration, mental tracking and monitoring, verbal fluency, Behavioral changes	Yes	Brazilian, Spanish, Greek
ALS Brief Cognitive Assessment (ALS-BCA)	5 min	Executive functions (working memory, set-shifting), Frontally-mediated language function, Delayed verbal recall, Behavioral changes	N/A	N/A
Beaumont Behavioral Inventory (BBI)	5–10 min	Frontal Behavioral symptoms, Executive functions, Language, Psychotic symptoms	N/A	N/A
Motor Neuron Disease Behavioral Instrument (MND-BI)	~ 10 min	Behavioral symptoms	N/A	N/A
ALS Frontotemporal Dementia Questionnaires (ALS-FTD-Q)	5–10 min	Behavioral symptoms (it also includes 3 items for memory, concentration and orientation in time)	N/A	N/A

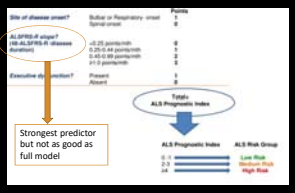
Relevance to Clinical Care

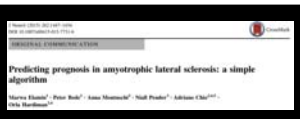
- Cognitive-Behavioral deficits → Tx adherence, making informed decisions, ability to learn/use assistive devices [Christidi, et al., 2018]
- Cognitive impairment
  - negative prognostic indicator linked to survival [Elamin, et al., 2015]
  - Increased care-giver burden [Andrews, et al., 2017]
  - Reduced QOL [Bock, et al., 2017]

→ Inclusion of neuropsychologist as core members of ALS multidisciplinary care teams [Hardiman, 2012]

ALS: Prognosis – most recent modeling

- ALS Prognostic Index (API)




Predicting prognosis in amyotrophic lateral sclerosis: a simple algorithm

- Validity established in model training (n=117) and testing (n=87) set; then externally validated on another n=122.
- Risk Group classification →
  - predicted survival time
  - Predicted poor prognosis (<25 month survival from Sx onset)
  - Predicted good prognosis (>50 month survival from Sx onset)

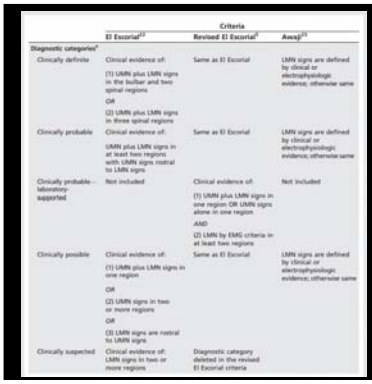
ALS: Diagnostic Process

- Remains a clinical task –ruling out
  - History → progressive weakness; usually focal, usually painless
  - Examination → UMN/LMN involvement usually focal, spreading to 1+ areas
  - Exclusion of other diseases
    - Imaging
    - Blood work
  - Electromyography and nerve conduction studies often used
  - Muscle biopsy
  - Less often – spinal fluid analysis
- Standard of Care
  - Guided by 2009 American Academy of Neurology (AAN) practice parameters
  - Informed by other guidelines and more recent literature
  - MULTIDISCIPLINARY TEAM CARE – all agree this is essential

Diagnosis of ALS Requirements - AAN

1. Signs of LMN – by clinical, electrophysiological, neuropathological
2. Signs of UMN – by clinical
3. Progression from within a region to other region(s)
4. Absence of electrophysiological evidence of other disease to explain clinical/electrophys signs
5. Absence of neuroimagining evidence of other disease to explain clinical/electrophys signs

- Clinical features required
  - Signs of LMN degeneration in ≥1 body region (bulbar, cervical, thoracic, lumbosacral)
  - Signs of UMN degeneration in ≥1 body region
  - Occurrence of LMN and UMN signs + progression to other region(s) determines certainty of the ALS diagnosis




Consensus Guidelines for Diagnosis
El Escorial revisited: Revised criteria for the diagnosis of amyotrophic lateral sclerosis

ALS Multidisciplinary Teams

- Typical Composition
  - Neurologist
  - SLP
  - PT
  - OT
  - Nutrition/Dietetics
  - Respiratory therapy
  - Social Work
  - Genetic Counselor
  - Neuropsychologist

http://www.alsa.org/community/

ALS Multidisciplinary Team Care vs … community care

- Longer survival
- Improved QOL
- Improved access to therapies
- Greater patient satisfaction

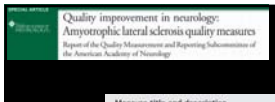
EFNS Task Force. (2012). Eur J Neurolo, 19; 360-375

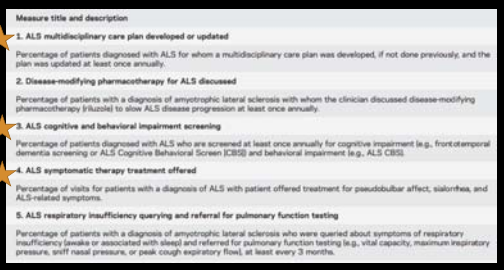
Rodriquez et al. (2011). Neurologia; 26; 455-460

Miller et al, (2006). Amyotroph Lateral-Scler, 7, 56-57

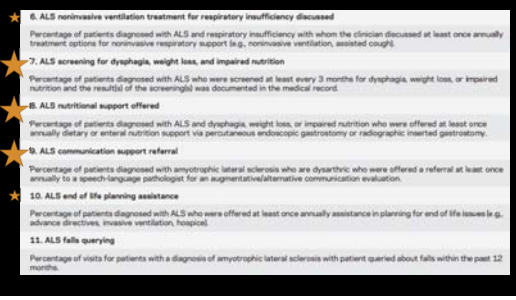
Riemenschneider et al. (2013). Ann Neurol, 65, 524-528

ALS Team Quality Measures – SLP related





ALS Team Quality Measures – SLP related



Primary Areas of ALS Disease Management

- Disease modifying - pharm
- Symptomatic Treatment
- Palliative Care
  - Respiratory
  - Nutrition/Hydration
  - Communication
  - Mobility
  - ADLs

- Riluzole → 1995 as Tx for ALS
  - Survival benefit ~3 (maybe 6) months [Lacomblez, et al., 1996; Bensimon, et al., 1994]
  - No discernable effect on QOL or function
  - Per American Academy of Neurology → "strong evidence" that it should be offered to slow disease progression (Level A)
  - Generally well tolerated
  - Cost: depends…$1200/month no coverage; $20-$200 copays with coverage; Medicare and VA cover

Fatigue
Nausea
Elevated liver enzymes
Tiredness
Stomach/Abdominal Pain

Primary Areas of ALS Disease Management

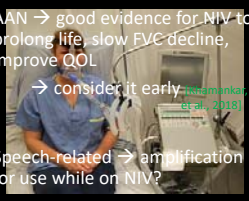
- Disease modifying
- Edaravone (Radicava®) → 2017 FDA approval
  - IV administration
  - 2week daily dose → 2 week off
  - Then repeated (weekends off) – how many cylces?
  - Can be taken with Riluzole
- 2001 Japanese approval for stroke → free radical scavenger
- In ALS application
  - slowed decline of ALSFRS-R by 33% over 6 months
  - Similarly slowed decline in QOL
  - Respiratory measures trended similarly but not significant
- High cost – estimated at $145,000/year; VA, many insurance companies limit coverage

Primary Areas of ALS Disease Management

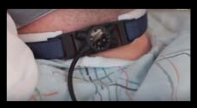
- Disease modifying
- Several other on going trials
- https://clinicaltrials.gov/

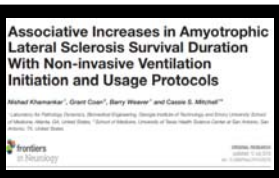
Symptomatic Treatment

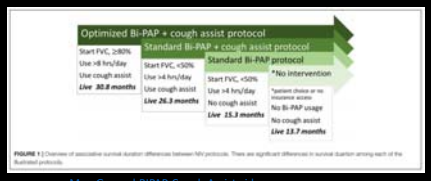
- Ventilatory support
  - Assisted ventilation → usually noninvasive (NIV), bilevel positive airway pressure (BIPAP)
  - Preset inspiratory and expiratory pressures delivered via mask
  - Trials =
    - Substantially prolong life [Radunovic, et al., 2017; Burkhardt, et al., 2017]
    - Improve QOL [Berlowitz, et al., 2016]
  - Noninvasive often eventually fails – decision about invasive (trach)
- AAN → good evidence for NIV to prolong life, slow FVC decline, improve QOL
  → consider it early [Miyamanka, et al., 2018]
- Speech-related → amplification for use while on NIV?



Symptomatic Treatment







MassGeneral BiPAP-Cough Assist video

Symptomatic Treatment

- Ventilatory support
  - Phrenic Nerve Stimulators = not effective per RCTs [Gonzalez-Bermejo, et al., 2016; DiPALS Writing Committee, 2015]

Pseudobulbar affect treatment

- Pharmaceutical intervention
  - Dextromethorphan + quinidine → FDA approved
  - Oral med
  - Various side effects possible (diarrhea, stomach pain, cough, dry eyes, muscle spasms, etc.
  - AAN identifies "good' evidence for its use in people with ALS who have pseudobulbar affect

Symptomatic Treatment

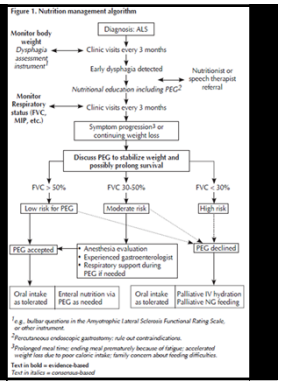
- Pseudobulbar affect
  - 25%-50% of patients with ALS
  - Nuedexta (DMQ) = dextromethorphan + quinidine
    - Approved 2011 for ALS
    - Interestingly, pt's reported improved speech


Enhanced Bulbar Function in Amyotrophic Lateral Sclerosis: The Nuedexta Treatment Trial

- Center for Neurologic Study – Bulbar Function Scale (CNS-BFS)
  - Speech
  - Swallowing
  - Salivation
- All domains improved on Nuedexta trial
- More to come

Symptomatic Treatment

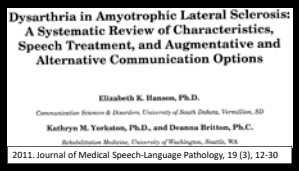
- Nutrition Support
  - AAN → if impaired oral intake, consider enteral nutrition with PEG
    - stabilize body weight- good evidence
    - Prolong survival – good evidence
    - Do not use Creatine – not effective with strong evidence
    - Do not use high dose vitamin E – not effective with good evidence



ALS: Speech Features

- Quite variable
  - UMN vs. LMN involvement
  - Specific speech sub-system involvement
  - Other impacts
    - General fatigue
    - Mood, apathy
    - Pseudobulbar affect
    - Ventilatory support needs

Academy of Neurologic Communication Disorders and Sciences (ANCDS)

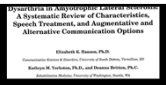

Dysarthria in Amyotrophic Lateral Sclerosis: A Systematic Review of Characteristics, Speech Treatment, and Augmentative and Alternative Communication Options

Elizabeth K. Hanson, Ph.D.

Kathryn M. Yorkston, Ph.D., and Deanna Britton, Ph.C.

2011. Journal of Medical Speech-Language Pathology, 19 (3), 12-30

- Appearance of Speech symptoms?
  - Wide variation
  - 33 months before Dx – 60 months post Dx
- Type of 1st Speech Symptoms – Varies
  - Laryngeal early in many studies [Watts & Vanryckeghem, 2001]
    - Acoustic may precede perceptual [Tomik et al, 2007]
  - Velopharyngeal in some
  - Oral Articualtory precision in some


Dysarthria in Amyotrophic Lateral Sclerosis: A Systematic Review of Characteristics, Speech Treatment, and Augmentative and Alternative Communication Options

and


Dysarthria in amyotrophic lateral sclerosis: A review

BARBARA TOMIK & ROBERTO J. GUILOFF

Amyotrophic Lateral Sclerosis. 2010; 11: 4-15

- Harsh, strain, breathy
- articulatory imprecision
- Hypernasality, nasal emission
- Slowed speaking rate
- Increased pause frequency, duration
- Monopitch
- Monoloudness
- Maximum performance – DDK
- Reduced intelligibility
- Etc.

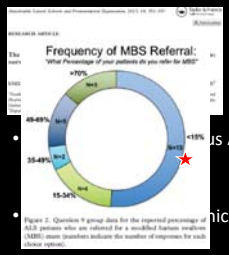
- Aerodynamic, kinematic, acoustic changes – many
  - Reduced articulator velocities and displacements
  - Increased nasal air flow
  - Reduced oral air pressure
  - Reduced vowel space
  - Increased vowel duration
  - Flattened F2 slope
  - Etc.

Classically described as mixed flaccid-spastic dysarthria

- Speech Intelligibility relative to speech changes
  - Physiologic correlates – many
    - Reduced articulator velocity [Yunusova, et al., 2010]
    - Flattend F2 slope [Kent et al., 1989]
    - Reduced vowel space [Turner et al., 1995]
    - Speaking rate [Ball et al., 2001]
    - Etc.

- <u>Of Note:</u>
  - Speech intelligibility reduction not usually an early bulbar symptom
  - Often with good intelligibility well beyond bulbar symptom presence
  - Once SI decrease starts, often a rapid intelligibility decline
  - Speaking rate at or below 120 wpm predicts decline in SI within a few months [Green et al., 2013] and should prompt AAC referral [Ball et al., 2002] if it hasn't happened already

Bulbar Dysfunction Diagnostic – What are people doing?



- …us ALS
- …nical

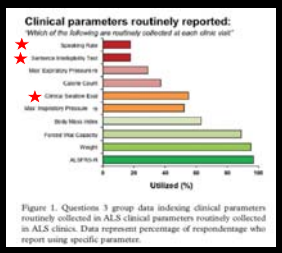


Figure 1. Questions 3 group data indexing clinical parameters routinely collected in ALS clinical parameters routinely collected in ALS clinics. Data represent percentage of respondentage who report using specific parameter.

Authors note: 85% pts have dysphagia → 45% don't routinely do CSE, 73% don't routinely do MBS → citing "clinical assessment alone is enough," "basing Tx decisions on pt report alone," "immediate PEG if choking or weight loss," "don't need it since dysphagia is expected."

Best Practice – Bulbar Function Diagnosis


PROVISIONAL BEST PRACTICES GUIDELINES FOR THE EVALUATION OF BULBAR DYSFUNCTION IN AMYOTROPHIC LATERAL SCLEROSIS

- Response to the Plowman et al., 2017 survey results
- Convened working group focused on – clinical speech evaluation, AAC evaluation & swallow evaluation
- Goals:
  1. Standardize bulbar data collection across sites
  2. Develop referral guidelines for speech, AAC, swallow
  3. Establish common data elements for speech, AAC, swallow evaluations

Muscle & Nerve, epub ahead of print

Speech Referral Guidance

1. Initial clinic visit – speech evaluation
2. At all visits – gather this
3. Follow-up visits – speech suggested as integral component but frequency/ duration may vary
   - Patient needs
   - Clinic resources
   - Etc
4. All patients with atypical oral motor exam – Otolaryngology referral

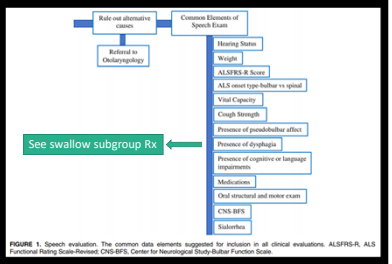


FIGURE 1. Speech evaluation. The common data elements suggested for inclusion in all clinical evaluations. ALSFRS-R, ALS Functional Rating Scale-Revised; CNS-BFS, Center for Neurological Study-Bulbar Function Scale.


ALS Functional Rating Scale Revised (ALS-FRS-R)

link to on-line ALSFRS

What's a meaningful ALSFR-R change?

- 90% clinicians (n = 65) indicated a 20% change as meaningful (i.e., 4 point change) [Castrillo-Viguera et al., 2010]

A moment on CNS-BFS


Assessment of bulbar function in amyotrophic lateral sclerosis: validation of a self-report scale (Center for Neurologic Study Bulbar Function Scale)

- This group deployed the CNS-BFS in the Nuedexta trial – [Smith et al., 2017]
- It was more sensitive to Tx effect than other bulbar measures (speaking rate, swallow)
- Here they validate the scale

- N=120 at 7 sites; 60 were from Nuedexta trial
- Clinic judgement of
  - normal or abnormal speech;
  - clinical swallow assessments (duration measures)
  - Salivation – normal or abnormal
- Patients
  - CNS-BFS
  - VAS for speech, swallow, salivation
- Trained evaluator – ALSFRS-R

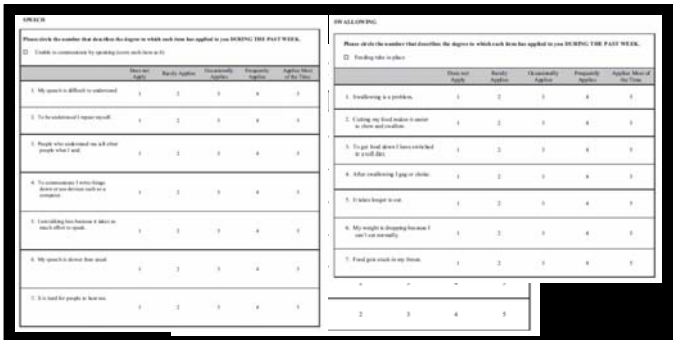
CNS-BFS

- 21 questions
- Self-administered
- 3 domains
  - Speech
  - Swallowing
  - Salivation

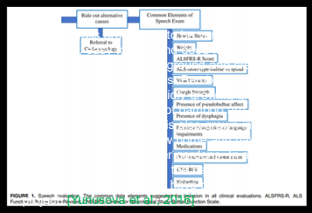
- 7 questions per domain
- Scaled 1-5 (6 on speech items if unable to speak)
- Score range
  - Low of 21 (no bulbar Sx)
  - High of 112

- CNS-BFS (and ALSFRS-R) highly predictive of clinician Dx

- CNS-BFS stronger correlations than ALSFRS-R and patient VAS with timed reading and swallowing



Back to Provisional Guideline: common elements



- Speech Assessment to include
  - Spontaneous sample
  - Reading passage – either/or
    - Rainbow
    - Bamboo
  - Sequential Motion Rate ("puhtuhkuh")
  - Max sustained /a/

- Clinician Rating of dysarthria severity (0=normal; 4= severe)

- Speaking rate (wpm)

- Identification of speech subsystems involved (respiratory, phonatory, articulatory, resonatory)

- Estimated time = 8-10 min

Pattee et al., 2018

AAC SubGroup Recommendations

- Early AAC exposure and training emphasized
  - Start before overt bulbar Sx →

    "The AAC evaluation should, therefore, be recommended at the time of diagnosis, regardless of whether speech impairment exists." (Pattee et al., 2018, p2)

  - Initial clinic visit →
    - introduce concept of AAC, broad definition of AAC
    - Arrange referral for AAC evaluation – should be ongoing, repeated as needed as abilities and needs change

Swallowing SubGroup

- SLP Swallow Screen
  - Testing/information in 5 domains
    - Patient-report outcomes
    - Diet intake
    - Pulmonary function and airway defense
    - Bulbar function broadly
    - Dysphagia/aspiration screen
  - All patients should undergo swallow screen → failed screening results in referral for comprehensive dysphagia evaluation → → VFSS an important component
    - Define issues
    - Assess strategy effectiveness

Swallowing SubGroup – specific tools recommended

DOMAIN	TOOL	SOURCE
Pt-report measure	Eating Assessment Tool - 10	Belafsky et al., 2008
Diet intake	ALS Severity Scale – Swallowing Subscale	Hillel et al., 1989
	Neuromuscular Disease Swallow Status Scale (NdSSS)	Wada et al., 2015
Pulmonary Function/airway defense	Forced Vital Capacity (FVC) Cough Test	Plowman et al., 2016
Bulbar function	Oral Motor Exam Iowa Oral Performance Instrument (IOPI)	Hiraoka et al., 2017
	CNS-BFS	Smith et al., 2018
Dysphagia – Aspiration Screen	Yale Swallow Protocol	Leder & Suitor, 2014

[information extracted from Table 1 in Pattee et al. (2018)]

EAT-10

Circle the appropriate response	0 = No problem				4 = Severe problem
1. My swallowing problem has caused me to lose weight.	0	1	2	3	4
2. My swallowing problem interferes with my ability to go out for meals.	0	1	2	3	4
3. Swallowing liquids takes extra effort.	0	1	2	3	4
4. Swallowing solids takes extra effort.	0	1	2	3	4
5. Swallowing pills takes extra effort.	0	1	2	3	4
6. Swallowing is painful.	0	1	2	3	4
7. The pleasure of eating is affected by my swallowing.	0	1	2	3	4
8. When I swallow food sticks in my throat.	0	1	2	3	4
9. I cough when I eat.	0	1	2	3	4
10. Swallowing is stressful.	0	1	2	3	4
				Total EAT-10:	

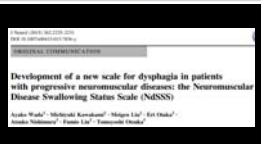
Belafsky PC, Mouadeb DA, Rees CJ, Pryor JC, Postma GN, Allen J, and Leonard RJ. Validity and reliability of the Eating Assessment Tool (EAT-10). Ann Otol Rhinol Laryngol 117: 919-924, 2008

ALS Severity Scale (ALSSS; Hillel et al., 1989)

Swallowing	Ability	Points
normal eating habits	normal swallowing	**10**
	nominal abnormality	**9**
early eating problems	minor swallowing problems	**8**
	prolonged time or smaller bite size	**7**
dietary consistency changes	soft diet	**6**
	liquefied diet	**5**
needs tube feeding	supplemental tube feedings	**4**
	tube feeding with occasional oral nutrition	**3**
no oral feeding	secretions managed with aspirator and/or medications	**2**
	aspiration of secretions	**1**

Neuromuscular Disease Swallow Status Scale

Level 1	Tube feeding with saliva suctioning in the oral cavity necessary. A patient can neither discharge nor swallow saliva
Level 2	Tube feeding without suctioning. Although a patient cannot take anything by mouth, can discharge and/or swallow saliva
Level 3	Tube feeding with occasional oral intake. A patient sometimes take orally for the fun, not for nourishment
Level 4	Totally orally fed and tube-free with supplemental nutrients, such as enteral solution. A patient usually take supplemental nutrients by mouth although don't take general food
Level 5	Totally orally fed with easy-to-swallow food and supplemental nutrients, such as enteral solution. A patient sometimes/often take supplemental nutrients by mouth
Level 6	Totally orally fed with only easy-to-swallow food. A patient eat foods processed in a mixer and drink thicken water
Level 7	Totally orally fed with no difficulties. A patient eat without something difficult to eat
Level 8	Totally orally fed with no restrictions. A patient eat all kinds of food

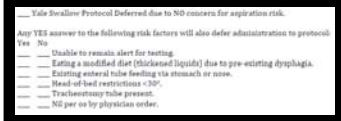


ORIGINAL COMMUNICATION

Development of a new scale for dysphagia in patients with progressive neuromuscular diseases: the Neuromuscular Disease Swallowing Status Scale (NdSSS)

Yale Swallow Protocol –Suiter, Sloggy & Leder, 2014

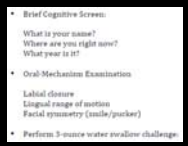
1. Exclusion Criteria



Yale Swallow Protocol Deferred due to NO concern for aspiration risk.

Any YES answer to the following risk factors will also defer administration to protocol:
Yes No
___ ___ Unable to remain alert for testing.
___ ___ Eating a modified diet (thickened liquids) due to pre-existing dysphagia.
___ ___ Existing enteral tube feeding via stomach or nose.
___ ___ Head-of-bed restrictions <30°.
___ ___ Tracheostomy tube present.
___ ___ NS per os by physician order.

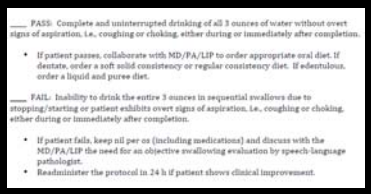
2. Administration



- Brief Cognitive Screen:
  - What is your name?
  - Where are you right now?
  - What year is it?
- Oral-Mechanism Examination
  - Labial closure
  - Lingual range of motion
  - Facial symmetry (smile/pucker)
- Perform 3-ounce water swallow challenge.

Yale Swallow Protocol –Suiter, Sloggy & Leder, 2014

3. Results/Rx



___ PASS: Complete and uninterrupted drinking of all 3 ounces of water without overt signs of aspiration, i.e., coughing or choking, either during or immediately after completion.

- If patient passes, collaborate with MD/PA/LIP to order appropriate oral diet. If dentate, order a soft solid consistency or regular consistency diet. If edentulous, order a liquid and puree diet.

___ FAIL: Inability to drink the entire 3 ounces in sequential swallows due to stopping/starting or patient exhibits overt signs of aspiration, i.e., coughing or choking, either during or immediately after completion.

- If patient fails, keep nil per os (including medications) and discuss with the MD/PA/LIP the need for an objective swallowing evaluation by speech-language pathologist.
- Readminister the protocol in 24 h if patient shows clinical improvement.

Swallowing SubGroup

- Education of patient/caregiver → stressed
  - Feeding tube role for nutrition/hydration supplementation
  - Need for good oral hygiene – linking to aspiration pneumonia
  - Various swallow maneuvers as needed
  - Modifications to food textures, other diet modifications
  - Pulmonary hygiene, cough, basic life saving techniques

Provisional Guidelines – what about cognitive & language?

- They didn't specify a tool.
- But other pubs and authors heavily using
  - ALS Cognitive Behavioral Screen (ALS-CBS)
  - ALS Caregiver report of behavior

Woolley SC, York MK, Moore DH, et al. Detecting frontotemporal dysfunction in ALS: utility of the ALS Cognitive Behavioral Screen (ALS-CBS). Amyotroph Lateral
Scler 2010;11:303–311

ALS Cognitive Behavioral Screen – Cog Section

(ALS-CBS; Woolley et al., 2010)

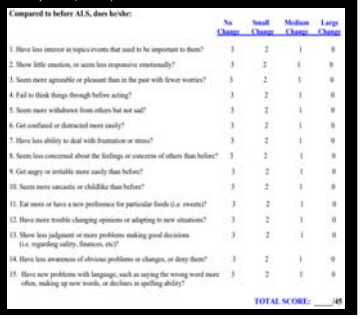
- Four subsections
  - Attention
    - 2 & 3 step commands
    - Mental Counting
    - Eye-mvmnt →saccades
  - Concentration → digit span reverse order
  - Tracking/Monitoring
    - Months backward
    - Alphabet
    - Letter-number alternation
  - Initiation and Retrieval – verbal fluency → words w/letter 'f'
- Cognitive Section (direct pt. screening)

Suggested Cognitive Cutoff Scores	Total: 20
Normal Scores	≥16
Suspected Impairment (2-3 SD below mean)	12-15
Probable Impairment (>3 SD below mean)	≤ 11
ALS-FTD High Suspicion (10 SD below mean)	≤ 5

- It is a screen, not a full neuropsych assessment
- Cog score moderately correlated with education level → consider in interp of patients with limited education

ALS Cognitive Behavioral Screen – Behavioral Section (ALS-CBS; Woolley et al., 2010)

- 15 items
- Changes since disease onset
- 0-3 score per item
  - "no change"
  - "small change"
  - "medium change"
  - "large change"
- 0-45 total score



Compared to before ALS, does he/she:	No Change	Small Change	Medium Change	Large Change
1. Have less interest in topics/events that used to be important to them?	3	2	1	0
2. Show little emotion, or seem less responsive emotionally?	3	2	1	0
3. Seem more agreeable or pleasant than in the past with fewer worries?	3	2	1	0
4. Fail to think things through before acting?	3	2	1	0
5. Seem more withdrawn from others but not sad?	3	2	1	0
6. Get confused or distracted more easily?	3	2	1	0
7. Have less ability to deal with frustration or stress?	3	2	1	0
8. Seem less concerned about the feelings or concerns of others than before?	3	2	1	0
9. Get angry or irritable more easily than before?	3	2	1	0
10. Seem more sarcastic or childlike than before?	3	2	1	0
11. Eat more or have a new preference for particular foods (i.e. sweets)?	3	2	1	0
12. Have more trouble changing opinions or adapting to new situations?	3	2	1	0
13. Show less judgment or more problems making good decisions (i.e. regarding safety, finances, etc)?	3	2	1	0
14. Have less awareness of obvious problems or changes, or deny them?	3	2	1	0
15. Have new problems with language, such as saying the wrong word more often, making up new words, or declines in spelling ability?	3	2	1	0
			TOTAL SCORE:	/45

SLP Intervention

- Limited (no?) change over the years in terms of approach
- Primarily compensatory in nature
- Very little investigation of SLP intervention effectiveness
- Low level evidence…mostly
  - Expert opinion
  - Case reports and case series
  - Very few and uncontrolled trials

[Tomik & Guiloff, 2010; Hanson, Yorkston & Britton, 2011]

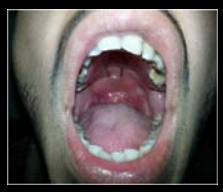
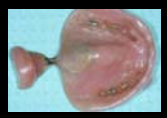
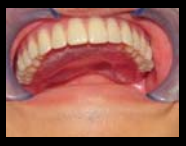
General Approach

- Optimize speech for as long as possible, including working with partner and family
- Plan for long term communication beyond useable speech, i.e., AAC – including voice banking, message banking
- Complete or help ensure AAC eval is done and Rx made
- Continue to follow regularly to adjust to the patient's changing needs

A few items that have been reported or evaluated

- What's been or is tried?
- Evidence?

Prosthetics – velopharyngeal, palatal lowering

Prosthetics – data regarding ALS?

- Esposito et al. (2000) – case series
- N=25, all ALS
- Palatal lift = 25
- Palatal augmentation = 10
- Measures:
  - "Intelligibility" – hypernasality, articulation (unclear how they did this)
  - Interview re: benefit
- 84% = reduced hypernasality
- 100% = pt/family report of benefit ["easier to speak," "worth the effort"]
- Of 10 with palatal augmentation = 60% perceived benefit to articulation

Prosthetics – data regarding ALS?

- Watanabe et al. (2012) – single case
- ALS
- Several types of prosthesis attempted over 13 months
- Unclear how they tracked speech
- Concluded: prosthesis was beneficial but only early on
- Decker et al. (2012) – single case
- ALS
- Palatal lift
- "The patient stated that her team of speech therapist, neurologist, otorhinolaryngologist and dentist noticed a better pronunciation with significant decreased hypernasality." p. 561
- Also reference to Nasometer with improvement noted

Prosthetics – data regarding ALS?

- Not much to hang your hat on

Exercise to strengthen articulators? No.

- Dworkin & Hartman (1979) –
  - fairly intense tongue strengthening in single case
  - no improvement
- Watts & Vanryckeghem (2001)
  - Single case – female with ALS
  - LSVT to address voice (without improvement)
  - Oral motor movements and strengthing → intelligibility decline continued

Voice Amplification?

Voice amplification video Boston Childrens

- Often hear that it won't help
- Will only make unclear speech louder?
- Anecdotal evidence from patients
- Most focus on the reduction in "effort' that might happen with amplifier

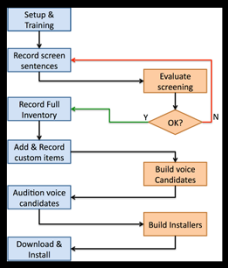
Banking

- Voice Banking
- Message Banking

Voice Banking

- Recording a relatively lengthy set of words or sentences
- These are then used synthesize your voice
- produce unique output; not the recorded input

Generally the flow is something like this https://www.modeltalker.org/



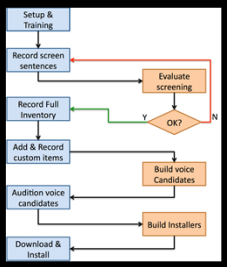
Voice Banking

- ModelTalker – for example
  - Create your synthetic voice
  - Loadable/useable in various apps and speech generating devices

(e.g.,, TherapyBox apps such as predictable)

$100

Generally the flow is something like this https://www.modeltalker.org/



Message Banking

- TobiiDynavox Message Banking
- Patients own voice
- Recordings of phrases, sentences, meaningful sayings wanted for subsequent import into speech generating device or apps

Boston Childrens Message vs Voice Banking

- Message Banking app

And a different type of Banking - Legacy

- http://www.recordmenow.org/
- Video and/or audio
- To leave a lasting set of messages, stories, etc. for loved ones.

Strategies talked about but not evaluated specific to ALS

- Communication Environment

Indicates reported in Murphy (2004) as useful by patients and families

  - Background noise reduction
  - Face-to-face when possible
  - Select environments (e.g., quieter restaurant)
  - Window up in car if needed
  - Determine whether eating and speaking are compatible – avoid the combo if needed

Strategies talked about but not evaluated specific to ALS

- Speech Production
  - Some over enunciation – if it is not fatigue inducing
  - Slighting of consonants – avoid if possible
  - Monitor phrase length – avoid speaking on residual air
  - Many think about slowing down, but most often they are already slow
  - Talking louder often just increases fatigue
  - Repeat if needed
  - Verbal spelling
  - Emphasizing key words

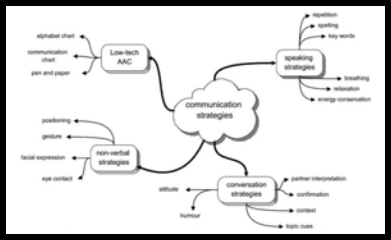
Strategies talked about but not evaluated specific to ALS

- Conversation Strategies & Communication
  - Understand your best times of day → fatigue/energy → plan important conversations accordingly
  - Partner assist/interpretation — Partner prediction video
  - Topic cuing
  - Facial expression, eye contact, gestures
  - Look for indications of understanding or lack thereof → repair strategies
  - Low tech assists – pointing, alpha board, writing, various pic boards

Strategies talked about but not evaluated specific to ALS

- Partner Training
  - Awareness of emotional lability – discuss how to handle (e.g., topic change, rib nudge, nothing)
  - Partner prediction – potentially helpful, potentially not — Partner prediction video
  - Don't talk over
  - Talking louder to me doesn't help
  - Ask before you decide to speak for me
  - I'll let you know if I am too frustrated and want to stop a conversation.
  - Acknowledge when you don't understand

Murphy, 2004 – 15pts/partners; self report of what they tried to facilitate communicatin



Sialorrhea treatment

- Sialorrhea in ALS = not increased saliva production; decreased swallowing of saliva
- Anticholinergic medications – usually tried 1st; various outcomes.
  - Some suggest scopalimine patches [McGeachan et al. 2013] – but various side effects
  - About 33% of ALS patients do not respond to anticholinergics
  - And even for those who have initial response, often not safe or becomes in effective over time

Sialorrhea treatment

- Botox – ANN good evidence
  - Parotid and submandibular glands
  - Sialorrhea decreases about 3-7 days post injection
  - max reduction at 2-4 weeks
  - Typical effect for 3.5 months but quite variable

Sialorrhea treatment

- Radiation to submandibular glands – AAN weak evidence
  - Electron based better than photon based
  - Various dosing schedules attempted
  - 4-6 months benefit reported in some studies
  - Comparison to Botox → not enough to draw conclusion regarding superiority of one over the other

All Done!

- Questions & Comments?