Welcome!
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ALS and Breathing: What Works?
Louis Libby, MD
Pulmonary, Critical Care and Sleep Medicine
The Oregon Clinic
Providence Portland Medical Center - Multidisciplinary ALS Clinic

Attendees are advised that portions of this webinar will be recorded for later viewing in our archives. If you would like to review the recording, please refer to our website, for information (www.alsa.org).

Hosted by:
The ALS Association
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Amyotrophic Lateral Sclerosis: Respiratory Management

Louis S. Libby, MD
llibby@orclinic.com
Pulmonary, Critical Care and Sleep Medicine
The Oregon Clinic
Providence Portland Medical Center
Multidisciplinary ALS Clinic

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ALS Management: Summary

- Respiratory complications occur in all ALS patients
- Specific respiratory muscle weakness causes defined problems
- Protocol driven management makes a difference!
Amyotrophic Lateral Sclerosis (ALS)

- Neurodegenerative disease - relentlessly progressive and incurable
- 1-3 cases/100,000
- 10% are familial
- Median survival ~3-5yrs
- Death due to respiratory complications
Healthy motor nerve cell stimulates muscle to contract.

ALS kills motor nerve cell, causing muscles to weaken.

Nervous system.
Respiratory Muscles: 3 groups

- Inspiratory Muscles-ventilatory pump
  - Diaphragm
  - External Intercostals
  - Accessory muscles

- Expiratory Muscles-cough
  - Abdominal muscles
  - Internal Intercostals

- Upper Airway
  - Closure of voice box for cough
  - Airway protection via “gag” reflex
MUSCLES OF INSPIRATION

- Sternocleidomastoid
- Scalenes
- External intercostals
- Diaphragm

MUSCLES OF EXPIRATION

- Internal intercostals
- External oblique
- Internal oblique
- Rectus abdominis
- Transversus abdominis
Inspiratory Respiratory Muscle Fatigue: Clinical Signs

• Shortness of breath with rapid, shallow breathing
• Chest and abdominal respiratory movements not coordinated
• Accessory muscle use
• Orthopnea—short of breath when lay down
• Hypercapnia (elevated CO2 in blood)
  – lethargy, morning headaches, altered thinking/coma
Inspiratory Respiratory Muscle Testing in ALS

- Forced Vital Capacity (FVC)
  - Sitting/supine
- Negative Inspiratory Force (NIF)
- Sniff Nasal Inspiratory Force (SNIF)
- Arterial Blood Gases – CO$_2$ level
Inspiratory Muscle Fatigue: Spirometry - Forced Vital Capacity (FVC)
Inspiratory Muscle Weakness: Negative Inspiratory Force (NIF)
Inspiratory Respiratory Muscle Weakness – Sniff Nasal Inspiratory Force (SNIF)
Inspiratory Respiratory Muscle Weakness: Interventions

- Breath Stacking to avoid stiff lungs/chest wall and pneumonia
- Non-invasive mechanical ventilation (Bipap/AVAPS) to support ventilation and slow down deterioration of respiratory muscle strength
- Diaphragmatic pacemaker to avoid atrophy of diaphragm muscle—controversial
Breath Stacking with Ambu Bag
Respiratory Insufficiency in ALS: Mechanical Ventilation

- Invasive - via Tracheostomy
- Non-invasive Mechanical Ventilation (NIMV)
  - Advantages
    - maintain speech, swallowing
    - relatively comfortable
    - use intermittently
    - maintain defenses against pneumonia
    - Inexpensive and easy to manage
  - Positive pressure with nasal or oral interface
  - Mouthpiece Ventilation (sip and puff mode)
  - Negative Pressure - Biphasic Curass Ventilation
CPAP Vs. Non-invasive Mechanical Ventilation

- CPAP (continuous positive airway pressure)
  - Only for obstructive sleep apnea
- Non-invasive mechanical ventilation
  - BiPAP: bi-level positive airway pressure
    - Also called VPAP
  - AVAPS: average volume assured pressure support
Pressure curve when BiPAP = 10/5 cm H2O; The pressure is higher on inspiration than on expiration, but both pressures are above ambient. In this example IPAP is set as 10 cm H2O and EPAP is set as 5 cm H2O.
BiPAP AVAPS "Average Volume Assured Pressure Support," is a variant of BPAP that automatically adjusts pressure support to meet changing patient needs while maintaining a target tidal volume.
BiPAP Prolongs Survival in ALS*

*Kleopa et al. J Neuro Science 164;82-88,1999
NIMV in ALS: Randomized/ Controlled Trial - Survival

P = 0.006

*Bourke et al. Lancet Neurol 2006; 5:140-47
NIMV in ALS: Randomized/Controlled Trial-Quality of Life*

*Bourke et al. Lancet Neurol 2006; 5:140-47
NIMV- Mouthpiece Ventilation
NIMV: Advanced Ventilators

• Common Brand names: Trilogy, Astral
• Advantages
  – Many modes- Bipap, AVAPS, Sip and Puff
  – Non-invasive or tracheostomy
  – Robust alarm systems
  – Internal/modular batteries
ALS- weak expiratory muscles and bulbar muscle weakness - Clinical signs

- Dysarthria
- Stuttering cough
- Choking with swallowing
- Coughing after swallowing/meals
- Sensation of tightness/restriction in throat
- Recurrent respiratory tract infections
ALS and inadequate cough*

- Can measure strength of cough - “Peak Cough Flow” (coughing into a peak flow meter)
- PCF < 270L/min – ineffective cough
- Cough requires ability to close voice box

* Bach et al: CHEST 2002;122:92-98
ALS and Inadequate Cough: Options for Rx*

- Breath stacking followed by cough assisted with abdominal thrust
- Mechanical assistance device
  - Cough Assist
  - Settings: +40/ -40cm H20

* Bach et al: CHEST 2002;122:92-98
Cough Assist: Mechanical Insufflator/Exsufflator
ALS Respiratory Complications: Other Issues

- Management of excessive secretions
  - Mucolytics - guaifenesin, NAC, hypertonic saline
  - Drying agents
  - Bronchodilators
  - Cough assist
  - Botox injection salivary glands

- Evaluate for other respiratory disorders
  - COPD, asthma, OSA etc

- Vaccines - respiratory illness

- End of life management - avoid emergency decisions
  - Discuss options early and often
ALS – Respiratory Management: 1988

• Patient diagnosed by neurologist
• Pulmonologist meets patient in ER/ICU in respiratory failure.
• Intubation, initiate mechanical ventilation
• Discuss option of tracheostomy
• Comfort Care or Mechanical ventilation until death
ALS Respiratory Management: Respiratory Protocol Approach-2017*

- Patient education
  - Breath stacking early
  - Assisted coughing with breath stacking or cough assist
    - when peak cough flow <270
  - Salivary aspirator (oral suction) if needed
- NIMV initiated with any of following
  - FVC<50% or decrease >500ml from previous or orthopnea
  - pCO2>45
  - Desaturations at night
- Experienced RT helps with NIMV
  - Multiple interfaces (nasal, oral, facemask) offered and used.
  - BiPAP/AVAPS etc considered

ALS Management: Protocol Approach-Benefits

Protocol resulted in the following benefits*
- Less initiation of ventilation and tracheostomy in ICU
- Increase in use of NIMV
- Improved survival, especially in those without bulbar involvement

Bach et al have reported 20% of patients can use NIMV part time or full time for up to 10 yrs.**

QOL improved in two controlled studies***

** Bach et al: CHEST 2002;122:92-98
*** Lyall et al: Neurology 2001;57:153-6
*** Bourke et al. Lancet Neurol 2006; 5:140-47
ALS Multidisciplinary Clinic: Respiratory Management

- Spirometry, NIF and PCF performed by RT
- Patients asked to cough spontaneously
- Patients asked about dyspnea, orthopnea, morning headaches, sleep quality
- Patients asked about use of breath stacking, cough assist and NIMV
- Download from NIMV evaluated
- Options for end of life discussed proactively
“Fans, for the past two weeks you have been reading about the bad break I got. Yet today I consider myself the luckiest man on the face of this earth.”
ALS Management: Summary

- Respiratory complications occur in all ALS patients
- Specific Respiratory Muscle weakness leads to specific problems
  - Inspiratory muscles- hypercarbic respiratory failure
  - Expiratory muscles- ineffective cough
  - Bulbar muscles- aspiration and ineffective cough
- Protocol driven management makes a difference!
  - Breathe stacking to avoid stiff lungs and chest wall
  - Assisted coughing for airway clearance
  - NIMV for weak inspiratory muscles prolongs life and improves QOL.
  - Avoidance of hospitalizations, emergency tracheostomies