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**Interdisciplinary Care of the Patient with
Amyotrophic Lateral Sclerosis –
Respiratory Therapy Care**

Disclosure

- I have the **following relevant financial relationship(s)** in the products or services described, reviewed, evaluated or compared in this presentation.
- University of Louisville Office of Continuing Medical Education and Professional Development
 - Financial compensation for speaking
- Frazier Rehab Institute
 - Employee
- I have **no relevant nonfinancial relationship(s)** to disclose.

Objectives

- Basic understanding of breathing tests which qualify patients for NIV (noninvasive ventilation)
- Basic understanding of NIV
- Acquire knowledge of various ventilation options in regards to the patient with ALS

Respiratory Assessment

- Objective data
 - Oxygen saturation & respiratory rate
 - Breathing tests –FVC, MIP, MEP, SNIP
 - Auscultation (listen to breath sounds)
 - Accessory muscle use
 - Swelling
- Subjective data
 - c/o shortness of air (SOA)
 - c/o cough, congestion & choking

Respiratory Assessment

- Screening for sleep apnea – hours asleep, awaken SOA, morning headache, snoring, orthopnea (SOA that occurs when lying flat)
- Pulmonary hygiene – use of Cough assist device
- Assistive coughing (Abdominal thrust maneuver) – use when choking or foreign body or aide in clearing congestion
- Breathing retraining techniques
- Signs & symptoms of pulmonary infections
- Smoking cessation
- Use of respiratory medications and oxygen use
- Flu and Pneumovax vaccines
- End of life issues – wishes regarding trach tube with invasive ventilation & prolonged use of mechanical ventilation

Breathing Tests

- FVC (Forced Vital Capacity)
 - maximal amount of air exhaled after maximal inspiration
 - volume test – *Liters*
 - compared to % of predicted
 - 50% or < of predicted indication for noninvasive ventilation (NIV)
- Decline guides ongoing treatment
- Is a significant predictor of survival (Miller et al., 2009, p. 1221)



Breathing Tests

- MIP (Maximal Inspiratory Pressure)
- MEP (Maximal Expiratory Pressure)
- Pressure tests – *cm H₂O*
- Reflects strength of breathing muscles
- Effort dependent & technician dependent
- MIP <60 cmH₂O with orthopnea indication for NIV (Miller et al., 2009, p. 1223).



Breathing Tests

- SNIP test – sniff-nasal-inspiratory pressure
 - <30 cmH₂O median survival was months
 - More reliable in later stages than FVC & MIP (Miller et al., 2009, p. 1221).
- Conventional PFT- Pulmonary Function Test

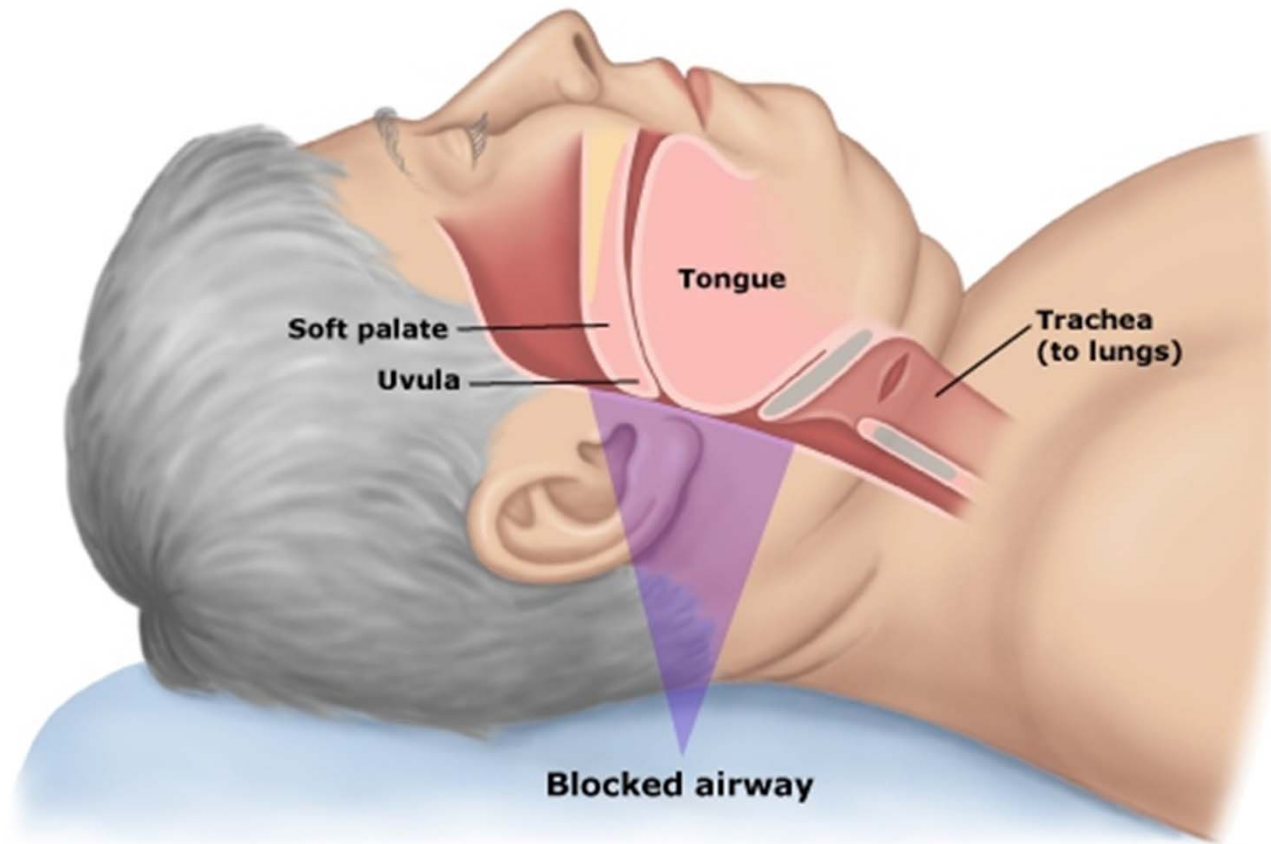


Evaluation of Sleep

- Polysomnography – sleep study
- Diagnosis:
 - obstructive sleep apnea
 - central sleep apnea



Obstructive Sleep Apnea



Noninvasive Ventilation

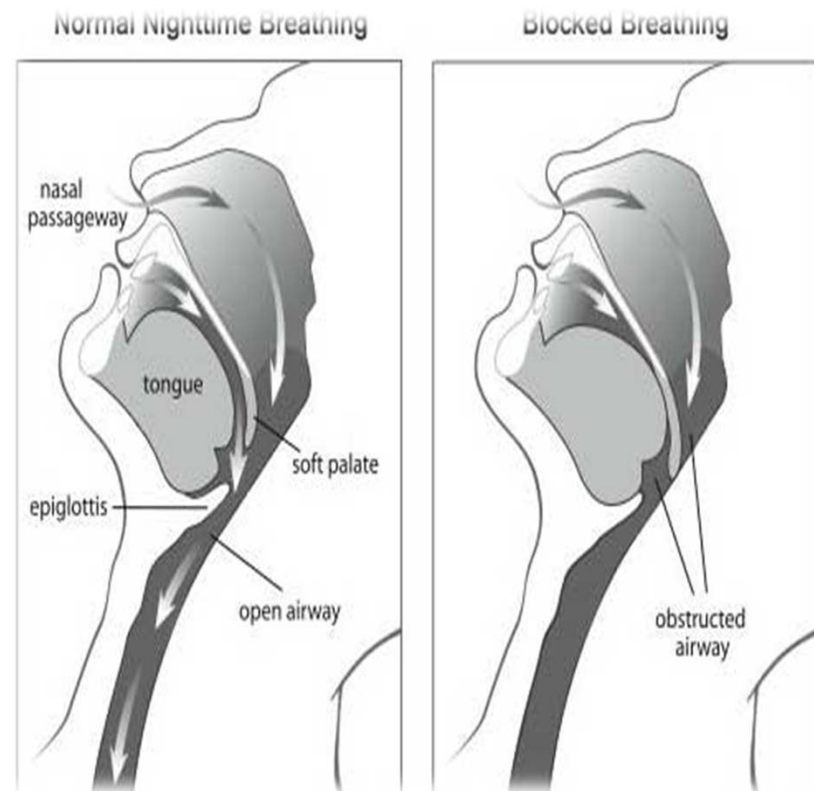


Noninvasive Ventilation



Noninvasive Ventilation

- Cpap (Continuous Positive Airway Pressure) – first line treatment for sleep apnea
- Bipap (Bilevel Positive Airway Pressure)
 - Treatment if noncompliant with Cpap
 - Treatment with **hypoventilation** (typical reason for ALS patient)



Ventilator Brands – Trilogy & Astral

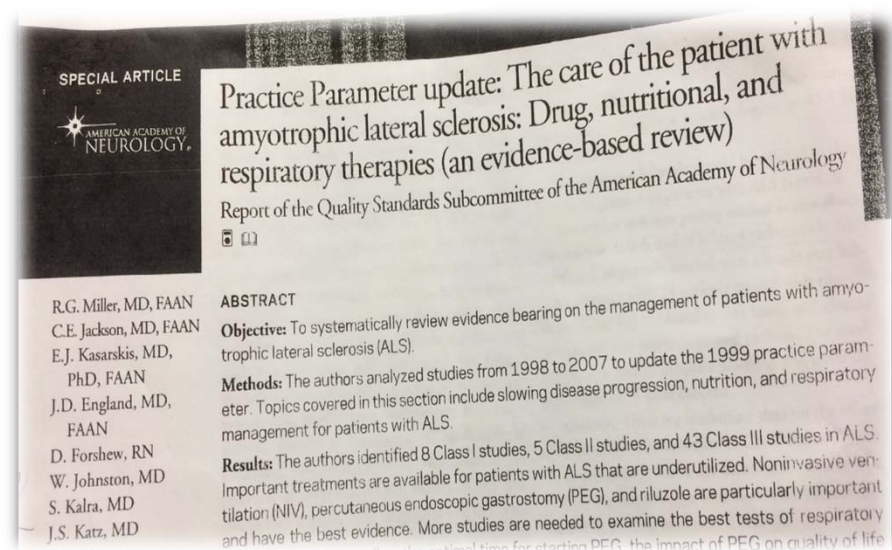
- Ventilator needed as disease progresses
- Can use invasive or noninvasive ventilation
- Advanced modes of ventilation
- More comfortable than Bipap
- Internal & external battery available
- More expensive
- Unavailable for hospice patients



Noninvasive Ventilation Benefits

- Positive impact of quality of life
- Improvement in energy and vitality
- Improvement in shortness of air
- Improvement in daytime somnolence

(Miller et al., 2009, p. 1223)

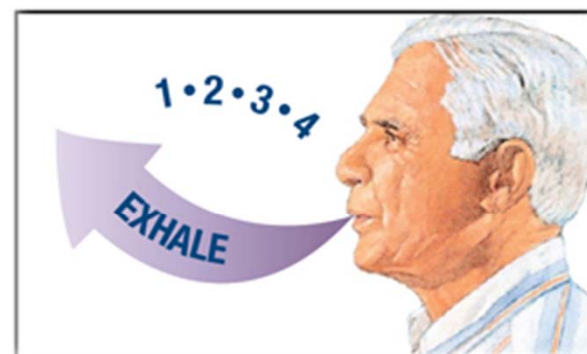
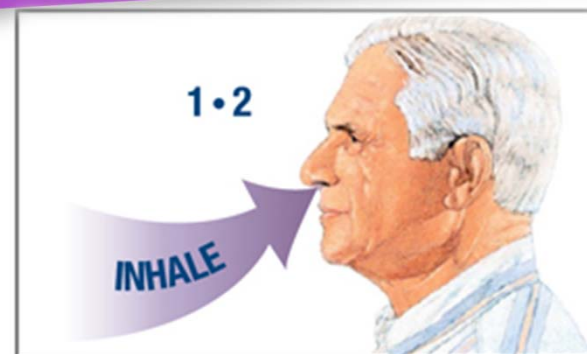


Mouthpiece & Invasive Trach Tube Ventilation



Breathing Retraining Exercises

- Pursed lip breathing – may help anyone who feels shortness of air
- Deep breathing/breath stacking – Incentive spirometer-intermittent use throughout the day
- Diaphragmatic breathing & relaxation exercises



Conclusion

- Most deaths from ALS are due to respiratory failure – (inadequate gas exchange)
- NIV (Noninvasive ventilation) should be considered:
 - FVC <50% predicted (based on height & gender)
 - MIP <-60 cm H₂O
 - SNIP <40 cm H₂O
 - Abnormal nocturnal oximetry
 - c/o orthopnea
- NIV prolongs survival & slows the rate of decline in FVC (Miller et al., 2009, p. 1218)



Thank you!!!!

References

- Miller, R.G., Jackson, C.E, Kasarskis, E.J., England, J.D., ForsheW, D., Johnston, et al. (2009). Practice parameter update: The care of the patient with amyotrophic lateral sclerosis: Drug, nutritional, and respiratory therapies (an evidence-based review). *American Academy of Neurology, 73, 1218-1226.*