

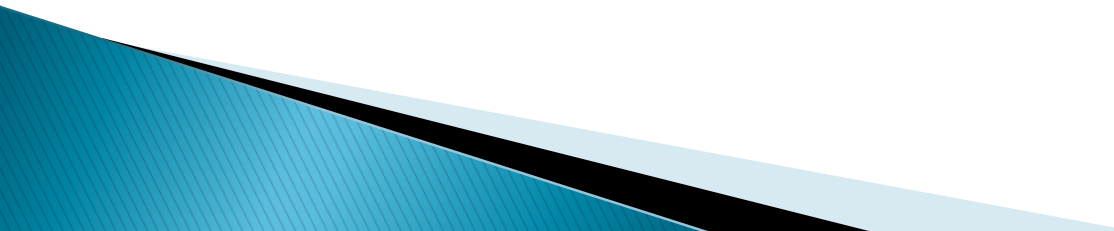
Respiratory Function and ALS

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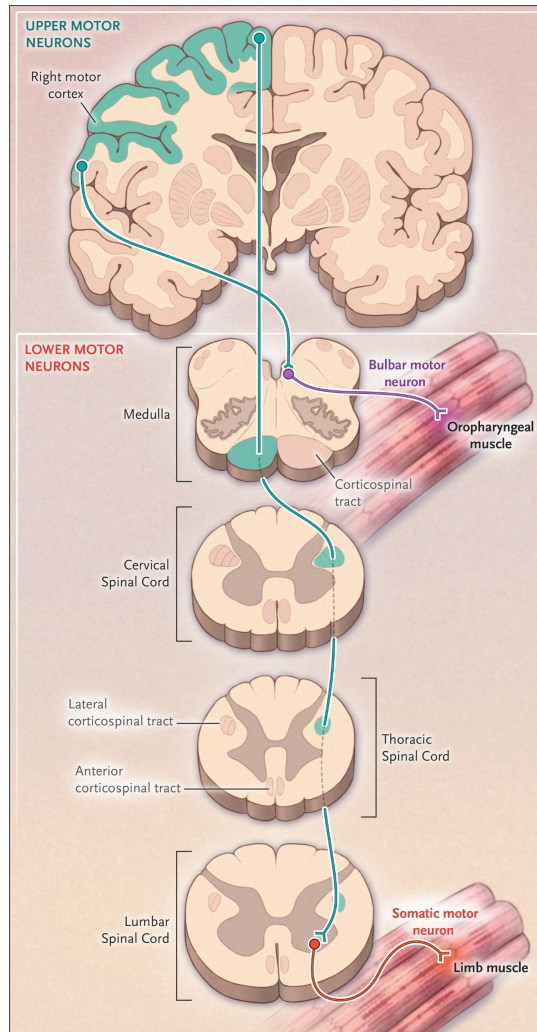
Disclosures

- ▶ None

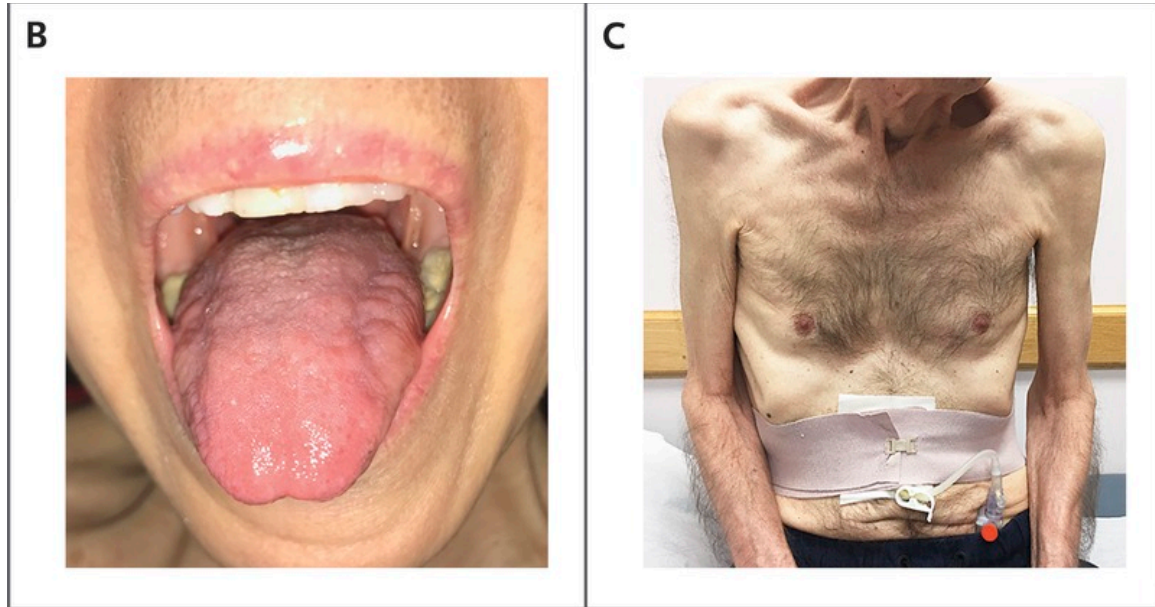
Outline

- ▶ Respiratory muscle overview
 - ▶ Noninvasive ventilation
 - Why, when and which type of PAP machine?
 - ▶ Airway clearance
 - How to augment cough?
 - ▶ Invasive ventilation/ tracheostomy
- 

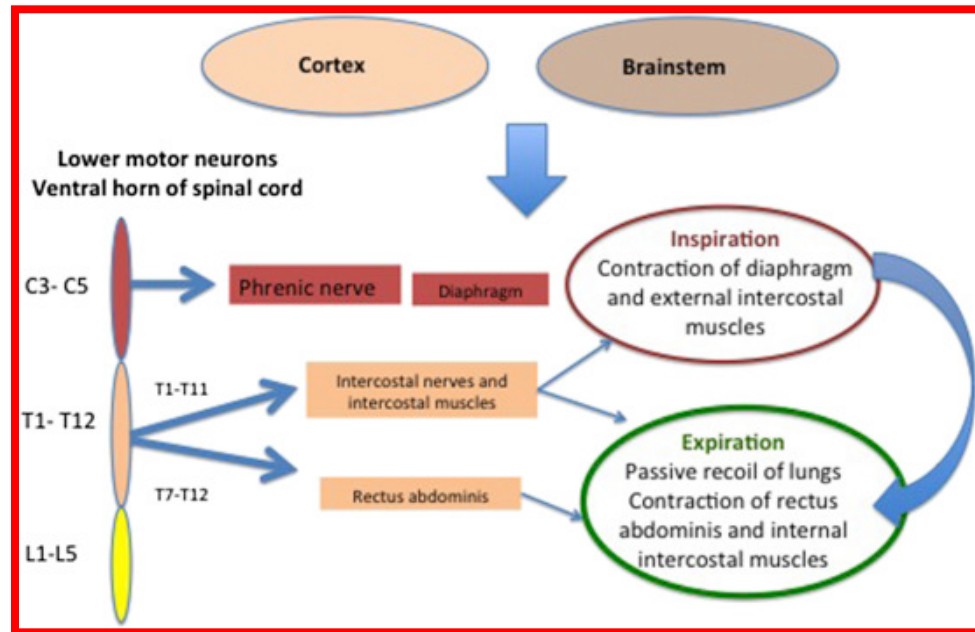
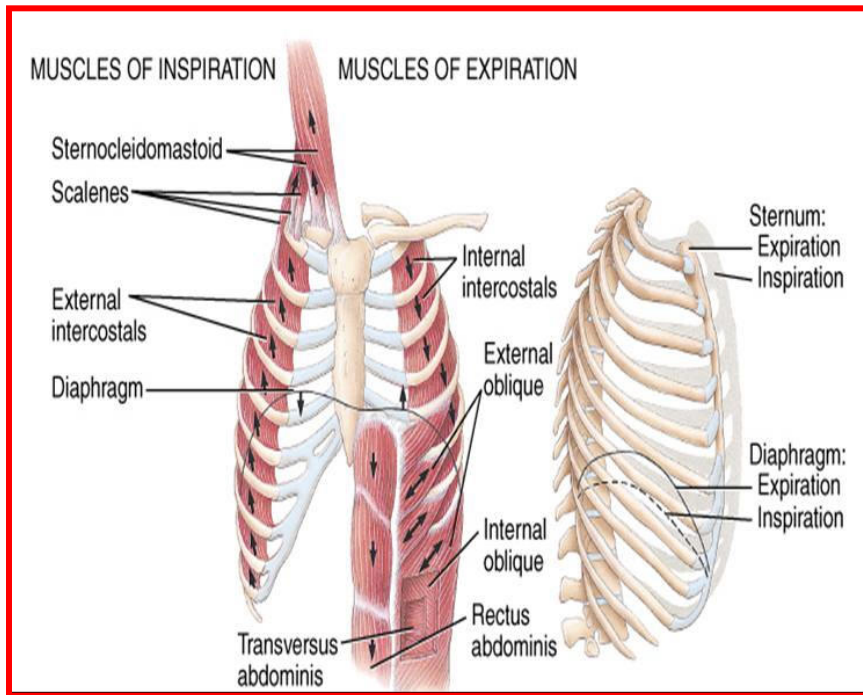
ALS is a progressive disease



ALS involves muscles in multiple regions of the body



Respiratory muscles are almost always involved

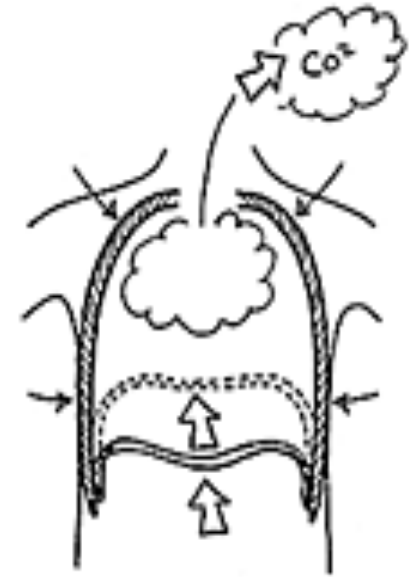


Respiratory limitations in ALS

Muscle Weakness	Shallow breathing	Low O ₂ levels	Poor cough	Risk for aspiration
Inspiratory Muscles	+	+	+	
Expiratory Muscles			+	
Upper Airway Muscles			+	+

There are many clues pointing to weak breathing muscles

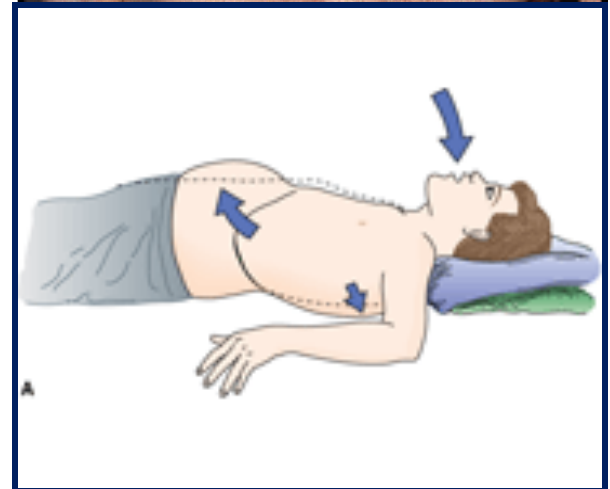
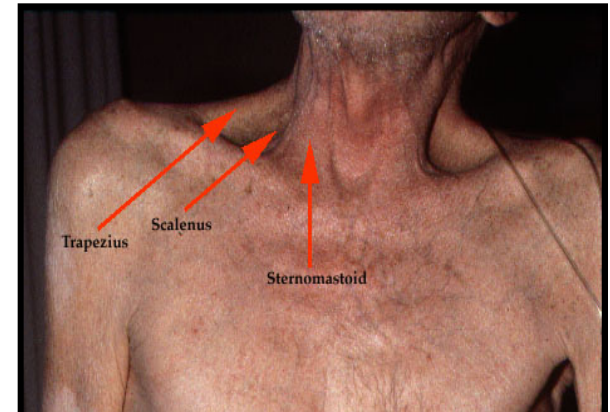
- ▶ Trouble lying flat in bed
- ▶ Disturbed sleep
- ▶ Daytime sleepiness
- ▶ Morning headaches
- ▶ Sleep related chokes
- ▶ Night sweats, bedwetting
- ▶ Trouble clearing phlegm

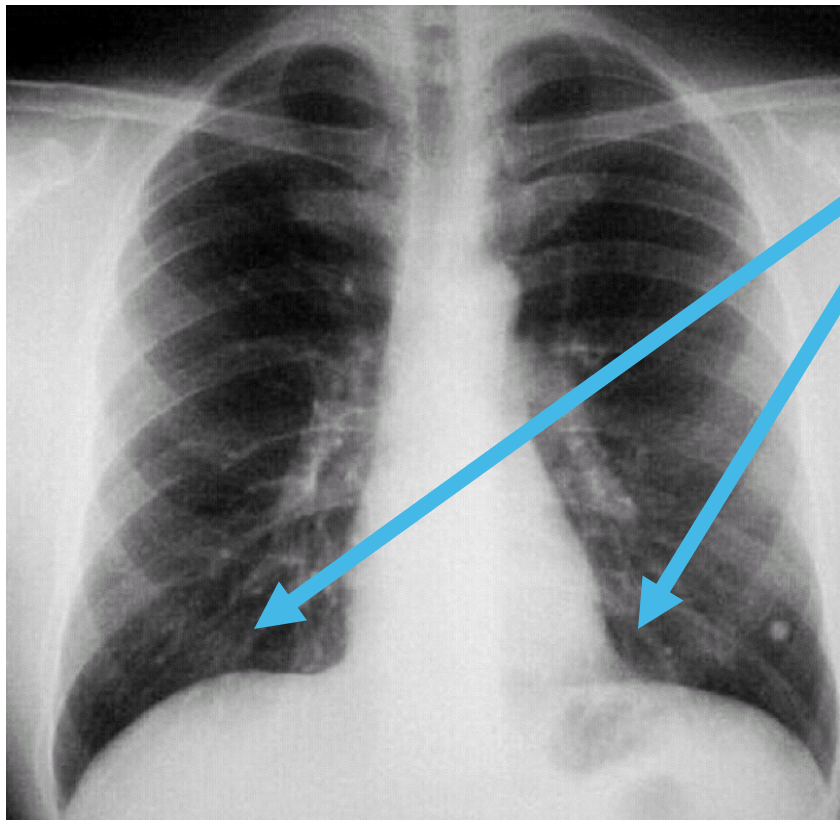


When the diaphragm contracts upwards it forces air from the lungs.

Diaphragm weakness tends to mirror that of peripheral muscles

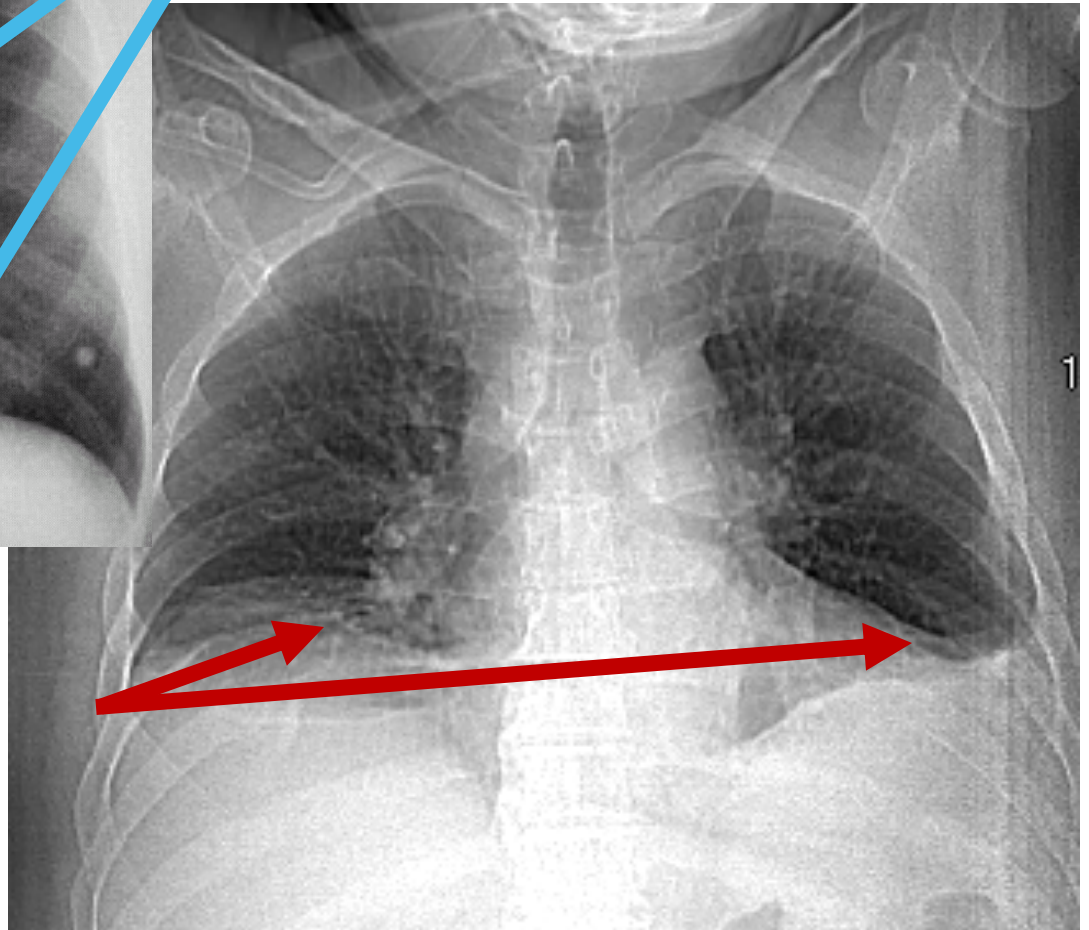
Exercise/ work related shortness of breath
ADLs become harder
Trouble coughing up phlegm
Trouble lying flat with sleep
Resting shortness of breath





Normal

Small lung
fields,
atelectasis



Patients with bulbar onset have weak upper airway muscles

- ▶ Trouble swallowing (dysphagia), choking (aspiration), speech (dysarthria), hoarseness (vocal cord weakness)

How to measure the strength of the breathing muscles?

- ▶ Pulmonary function test:
 - Spirometry: FVC is most commonly used
 - Maximum inspiratory and expiratory pressures
 - Sniff nasal inspiratory pressure (SNIP)
- ▶ Overnight oximetry
- ▶ Arterial blood gas
 - Low O₂ and high CO₂ levels are late findings

Pulmonary Function Test

- ▶ Forced vital capacity and/ or vital capacity
 - ▶ Upright and supine
 - ▶ Predicts diaphragm weakness
 - ▶ <50% – predictor of failing respiratory muscles

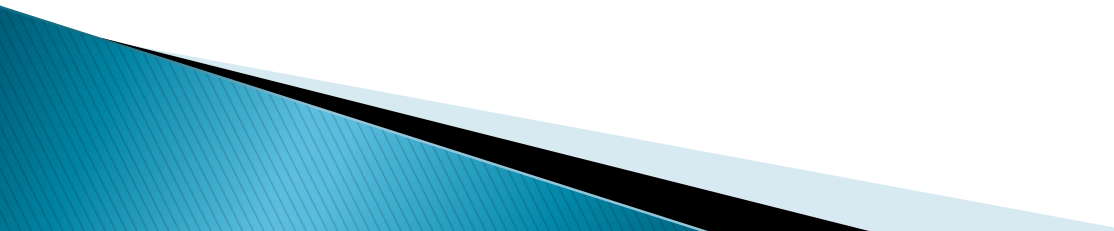


Sniff nasal inspiratory pressure

- ▶ Can be used in patients with bulbar dysfunction
- ▶ $SNIP < 40 \text{ cm H}_2\text{O}$ correlates with low O_2 levels at night



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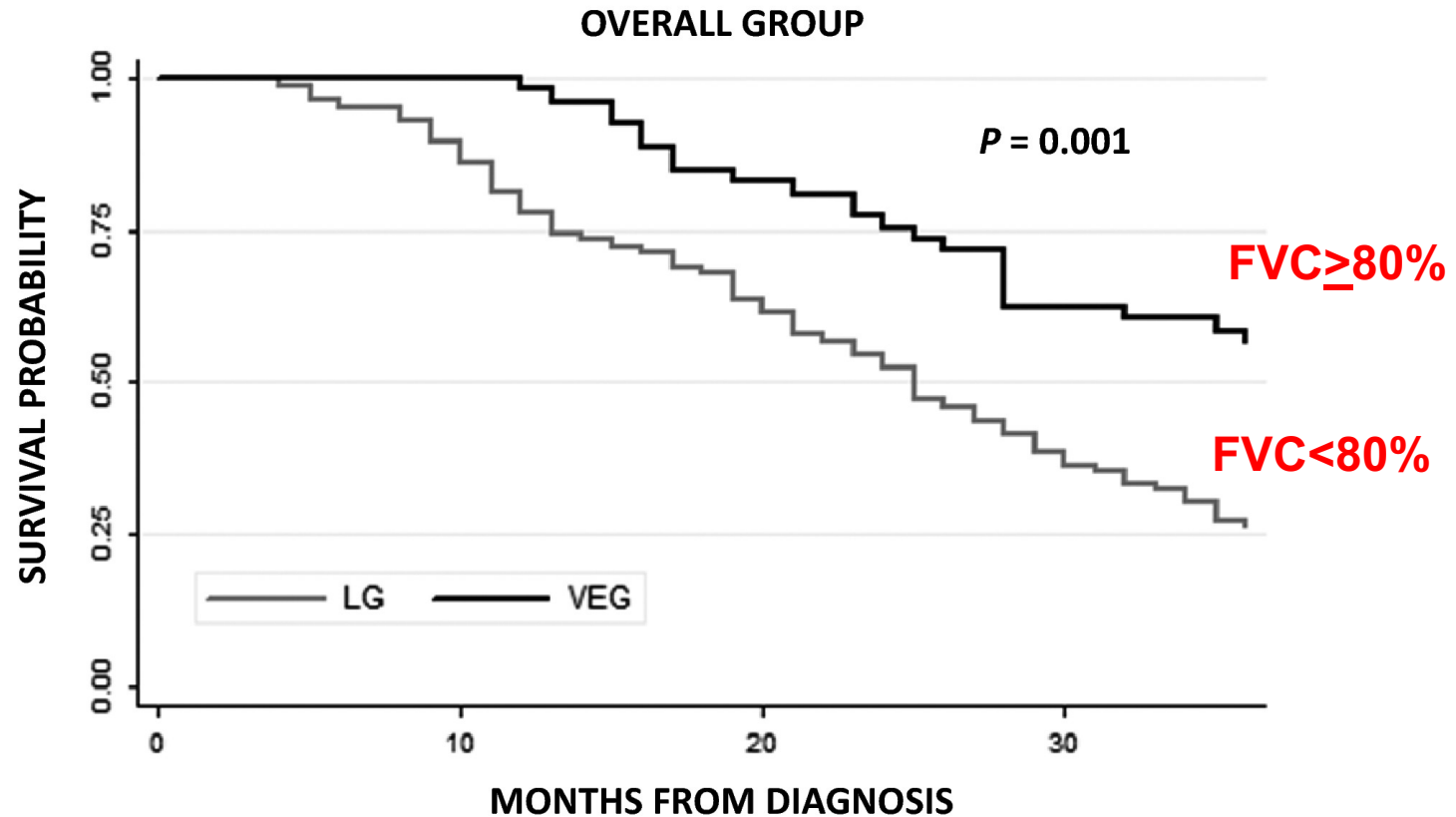
Why NIV and not Oxygen?

- ▶ NIV improves oxygen and decreases CO₂ levels and rests the breathing muscles
 - Improves survival and quality of life
 - Improves dyspnea
 - Impacts positively rate of FVC decline
- ▶ Oxygen builds up CO₂ which acts like narcotic
 - Not different than room air to improve refractory breathlessness in patients with normal oxygen levels ⁽¹⁾

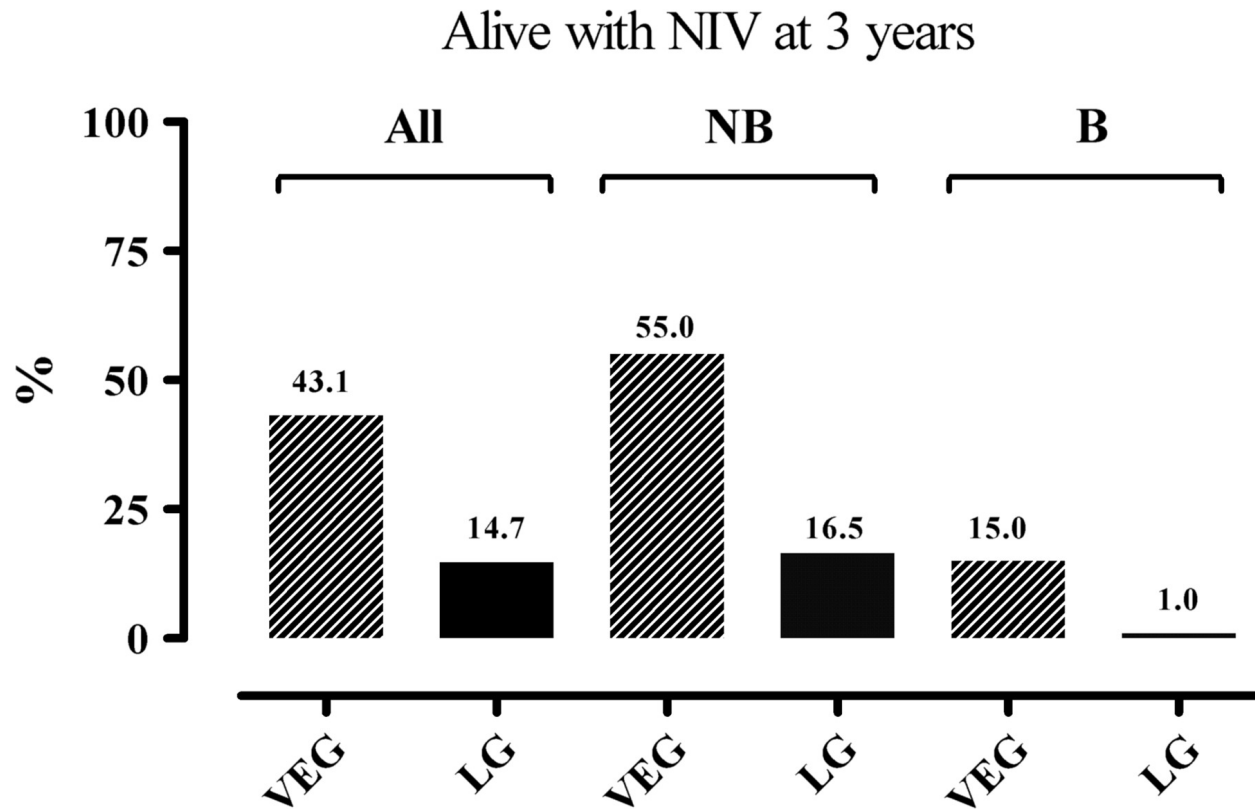
When to start PAP Therapy?

- ▶ Medicare guidelines:
 - ▶ Symptoms
 - ▶ orthopnea, morning headaches, sleepiness
 - ▶ Pulmonary Function Test:
 - ▶ FVC $< 50\%$ predicted (upright or supine)
 - ▶ MIP ≤ 60 cmH₂O (upright or supine)
 - ▶ Sniff nasal pressure < 40 cmH₂O (upright or supine)
 - ▶ Arterial Blood Gases
 - ▶ PaCO₂ > 45 mmHg
 - ▶ Overnight oximetry: > 5 minutes SpO₂ $\leq 88\%$

FVC 80% – Is sooner better?



Survival improved in all groups



Different Types of Respiratory Assist Devices

Can be prescribed without need for a sleep study

- ▶ Traditional bilevel/ fixed pressure devices:
 - ▶ BiPAP® (Philips Respironics)
 - ▶ VPAP® (ResMed)
- ▶ Auto-titrating NIV devices:
 - ▶ BiPAP® AVAPS (Phillips Respironics)
 - ▶ iVAPS® (ResMed)

Which machine to use when support is needed for 10–12 hours?

- ▶ Switch to noninvasive ventilators
 - Trilogy (Phillips Respironics)
 - Astral (ResMed)
- ▶ Advantages:
 - Internal and external batteries
 - Unique feature of mouthpiece ventilation
 - Can be used for invasive ventilation via tracheostomy



Chest 2019; 155: 401

Many different types of masks out there



Full face mask vs. nasal mask vs. nasal pillows

Getting started on NIV

- ▶ Outpatient/ home setting under the pulmonary clinician direction:
 - **Trained, experienced RRT**
 - Start low and increase once patient tolerates initial pressures/volumes
 - Mask should fit without too much pressure on the face/head

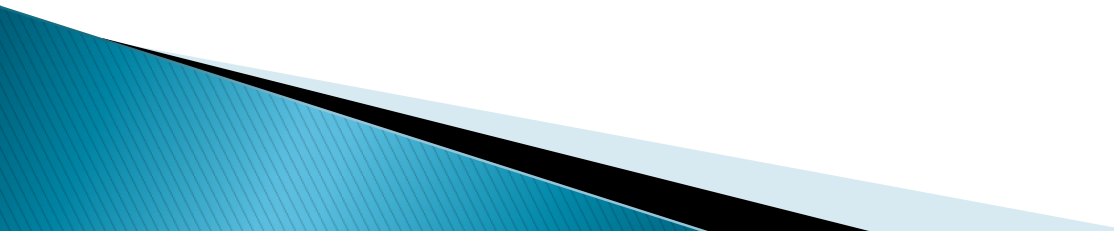
Tips to improve compliance

- ▶ Education and desensitization
- ▶ Chin straps with nasal pillows
- ▶ Heated humidity and heated wire circuit for airway dryness
- ▶ Elevate tubing on a headboard or hose lift
- ▶ Nasal bridge and/ or strap padding for discomfort
- ▶ Zinc oxide or steroid cream for rash
- ▶ Nasal saline spray, nasal steroid, nasal strips for nasal congestion

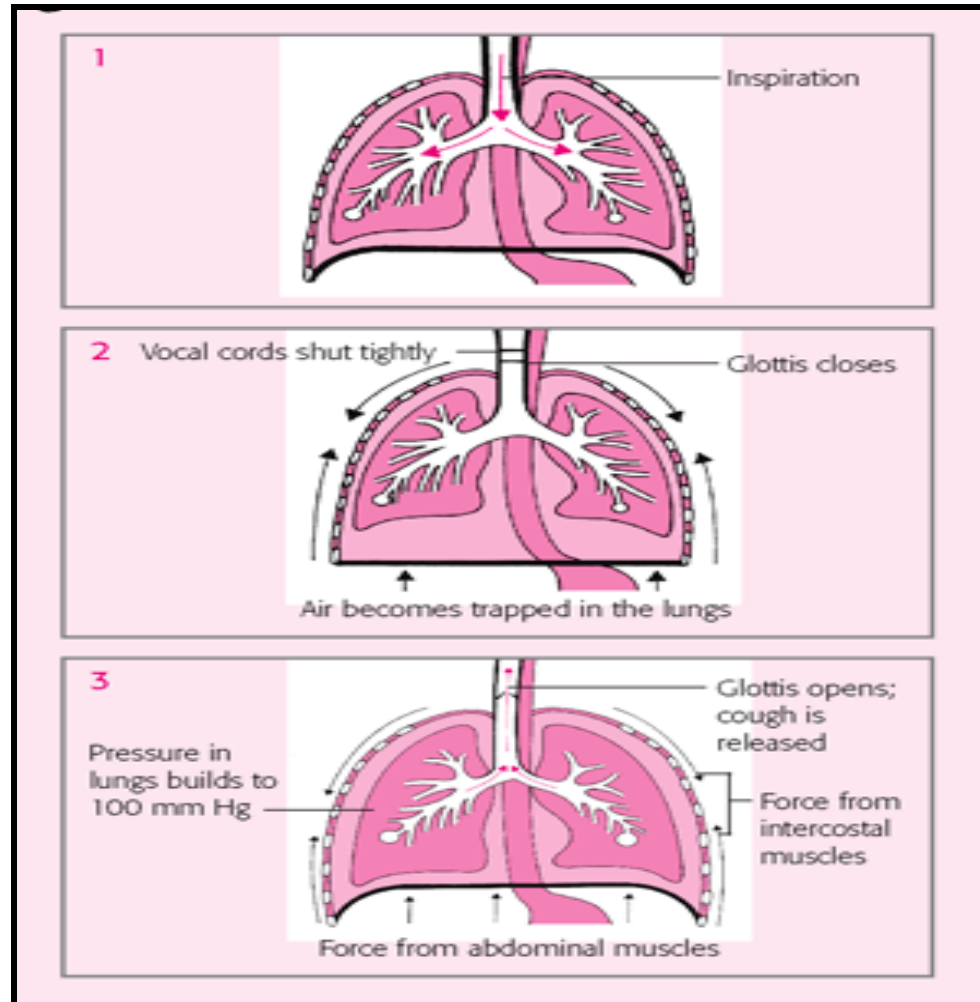
Monitoring is crucial

- ▶ Some PAP adjustments could be done remotely
 - Symptoms
 - Machine downloads:
 - Total use, pressures, volumes, percent triggered breaths, mask leak, etc.
 - Overnight oximetry
 - Daytime blood gas

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Cough is a complex process



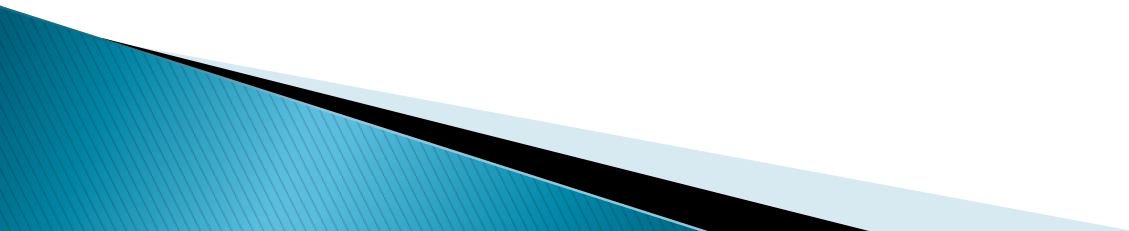
How to augment cough?

- ▶ Manual cough assist
- ▶ Suction machine



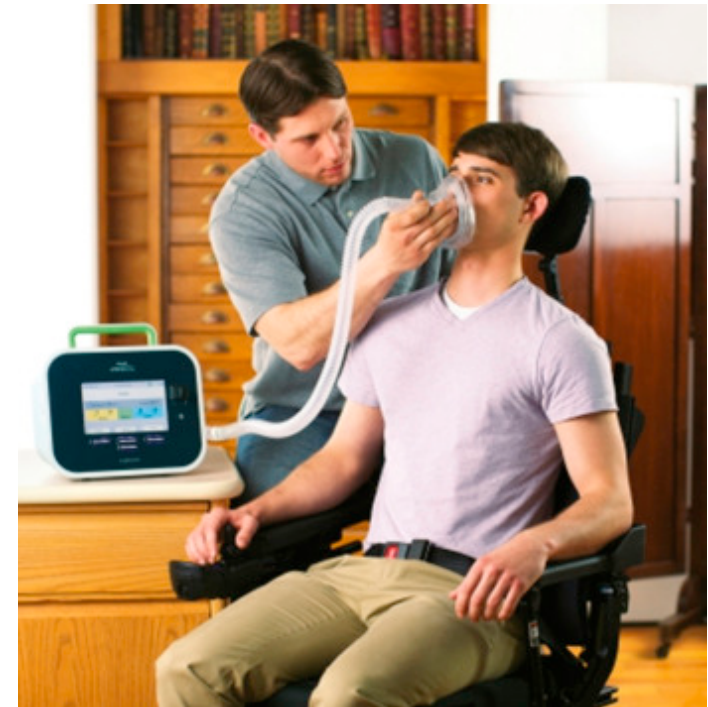
Breath stacking

<https://www.youtube.com/watch?v=JlgeRoI5vCw>

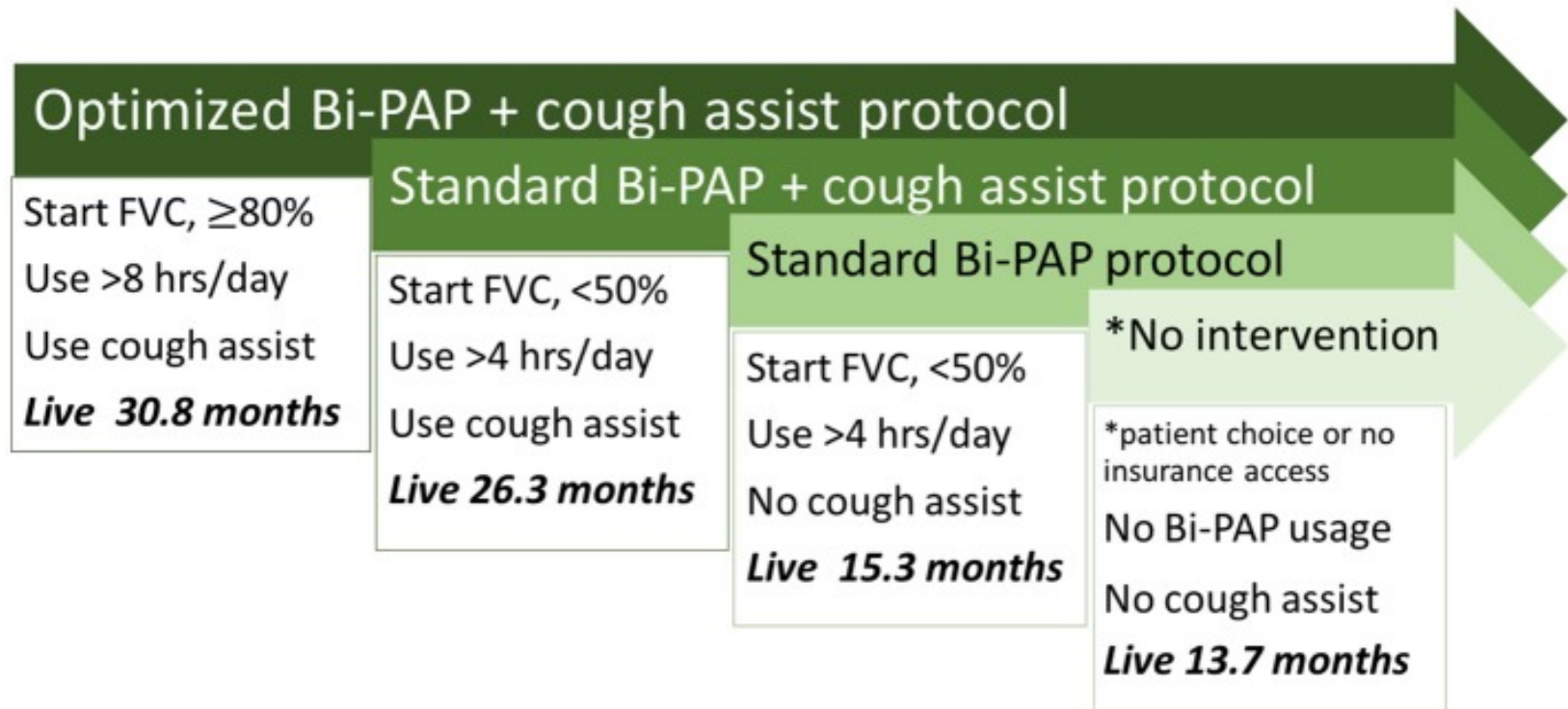


Mechanical Insufflation–Exsufflation Device

- ▶ **Trained, experienced RRT**
- ▶ Augments secretion clearance
- ▶ Prevents intubations, hospitalizations and tracheostomy
- ▶ Challenge:
 - Bulbar dysfunction



NIV and cough assist are better together

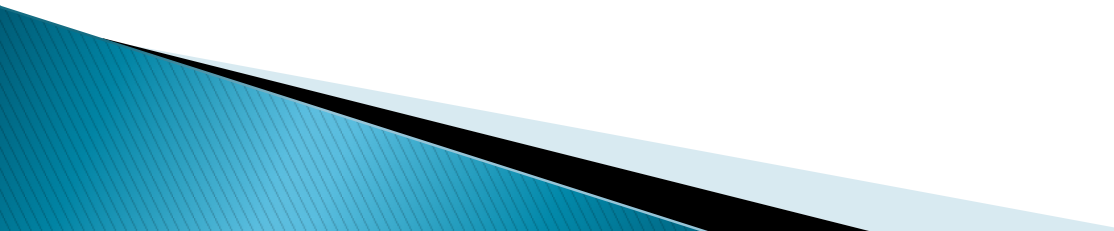


High frequency oscillatory ventilation

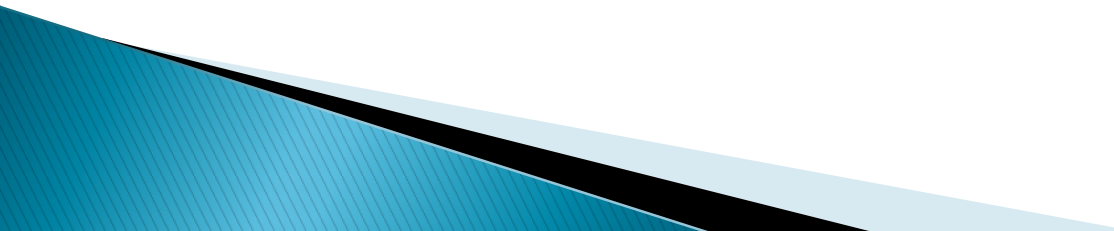
- ▶ Can reduce pneumonia and hospitalizations
- ▶ May improve breathlessness



Many options are available to control oral secretions

- ▶ Scopolamine patch or oral
 - ▶ Glycopyrrolate
 - ▶ Amitriptyline
 - ▶ Atropine drops
 - ▶ Botulinum toxin injection (Botox)
 - ▶ High beam radiation to the salivary glands
 - ▶ Surgery
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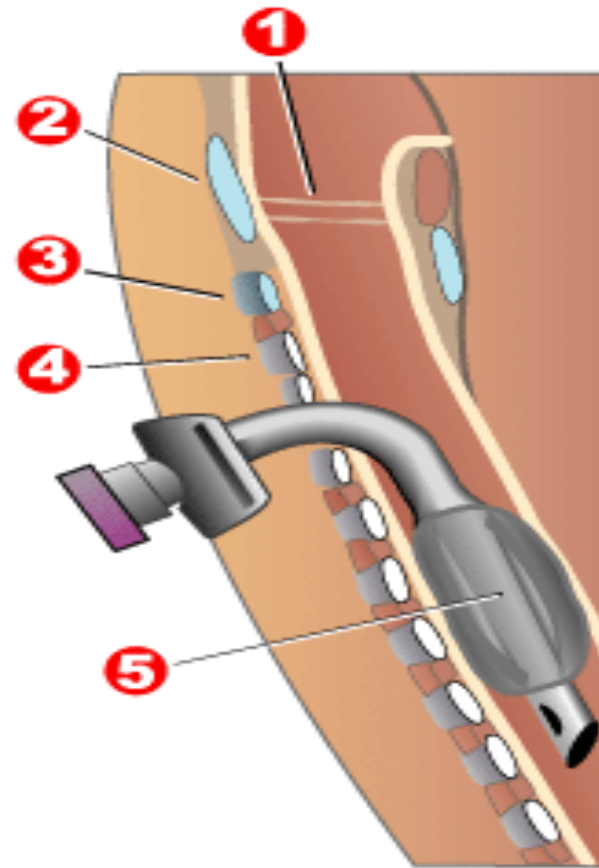


The ALS Association helped actor James Woods prepare for his starring role as a college professor with ALS on the February 2 episode of NBC's "ER."

Photo © 2005 Warner Bros.

Entertainment Inc.

Tracheostomy and invasive ventilation



When should invasive ventilation be considered?

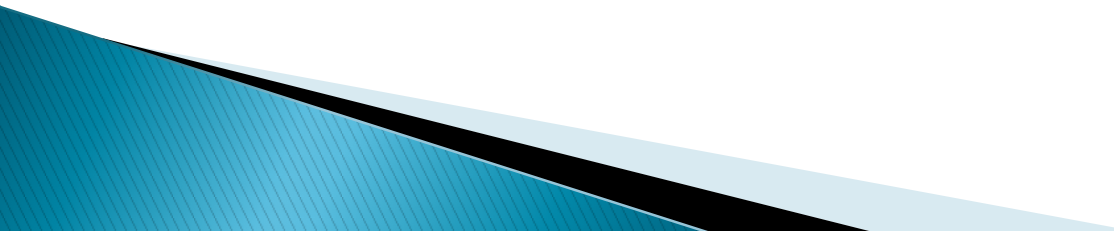
- **Early discussions are crucial**
- Symptoms of respiratory failure associated with:
 - Intolerance of NIV and $FVC < 50\%$
 - Failure of NIV: > 12 hours use and low O_2 saturation

Other facts about invasive ventilation

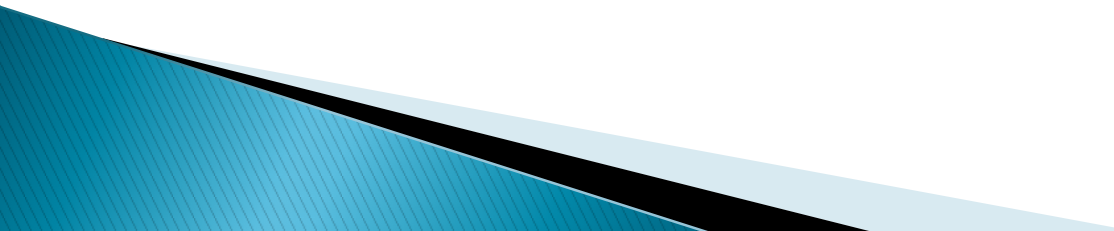
- ▶ Median survival: 1–2 years
- ▶ Quality of life and ability to communicate may wane over time
- ▶ Advance directives preserve patient autonomy
- ▶ Caregivers report poor quality of life and require significant support



What else should we consider?

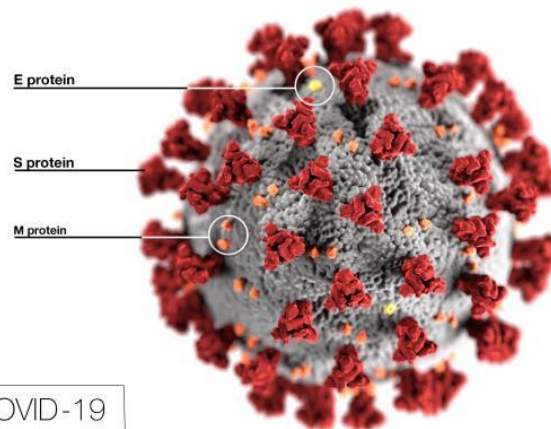
- ▶ Immunizations: Influenza and Pneumococcal vaccines
 - ▶ Gastrostomy for nutritional support
 - ▶ Monitor for aspiration
 - ▶ Treat respiratory infections early
 - ▶ End of life care
- 

Respiratory goals of care

- ▶ Multidisciplinary approach improves outcomes
 - ▶ Early NIV start increases life span
 - ▶ Invasive ventilation if considered requires discussion of all care aspects related to tracheostomy, ventilator, patient and care givers
 - ▶ Palliative care: access to anxiety and pain-relieving medication/nursing assistance at end of life
- 

And the COVID-19 pandemic...

- ▶ Our community prevalence in WI (November 2020) is high
 - Face mask/ facial covering
 - Social/ physical distancing
 - Washing hands
- ▶ Influenza season is here, flu shot is needed
- ▶ Virtual vs. in person clinic evaluations



COVID-19

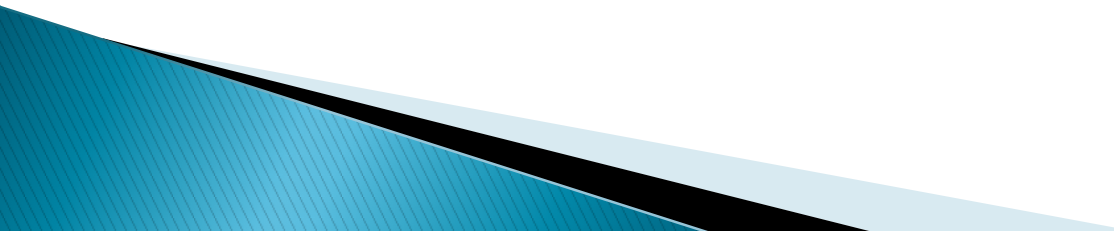


Zablocki VAMC – ALS Center of Excellence

- ▶ Multidisciplinary ALS clinic:
 - Neurologist, nurse, physical therapist, dietician, social worker, respiratory therapist, PAP coordinator and pulmonologist
 - Clinic visit every 3 months
 - In-person or virtual visit



Summary

- ▶ Pulmonary involvement is common in patients with ALS
 - ▶ Early recognition of respiratory manifestations and early interventions improve outcomes
 - ▶ Multidisciplinary team approach and care delivery by experienced healthcare providers result in better care
- 

Thank You

“I might have been given a tough break, but I have got an awful lot to live for.”



Lou Gehrig 1903-1941