

Breathing is Everything (when you can't)

Respiratory Care for People with ALS

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Penn Medicine

Jay and Randy Fishman

2015



Penn Medicine

Fishman Program for Home Assisted Ventilation

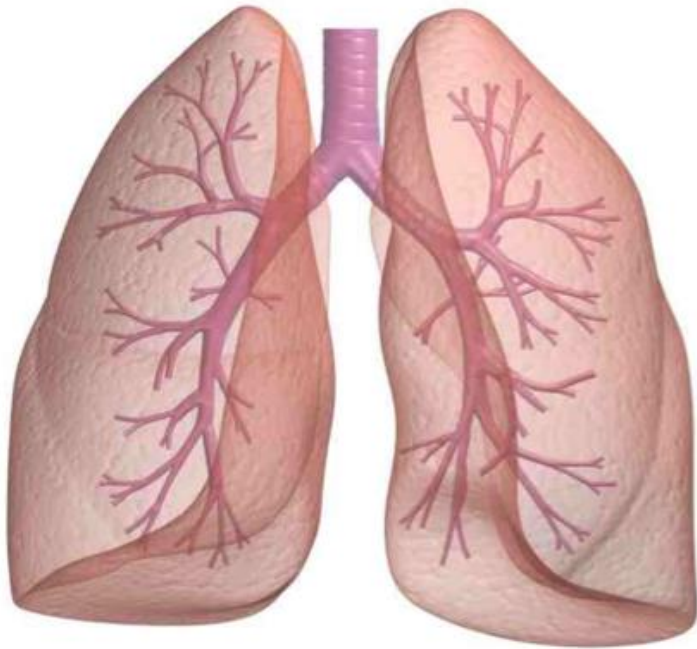
2017



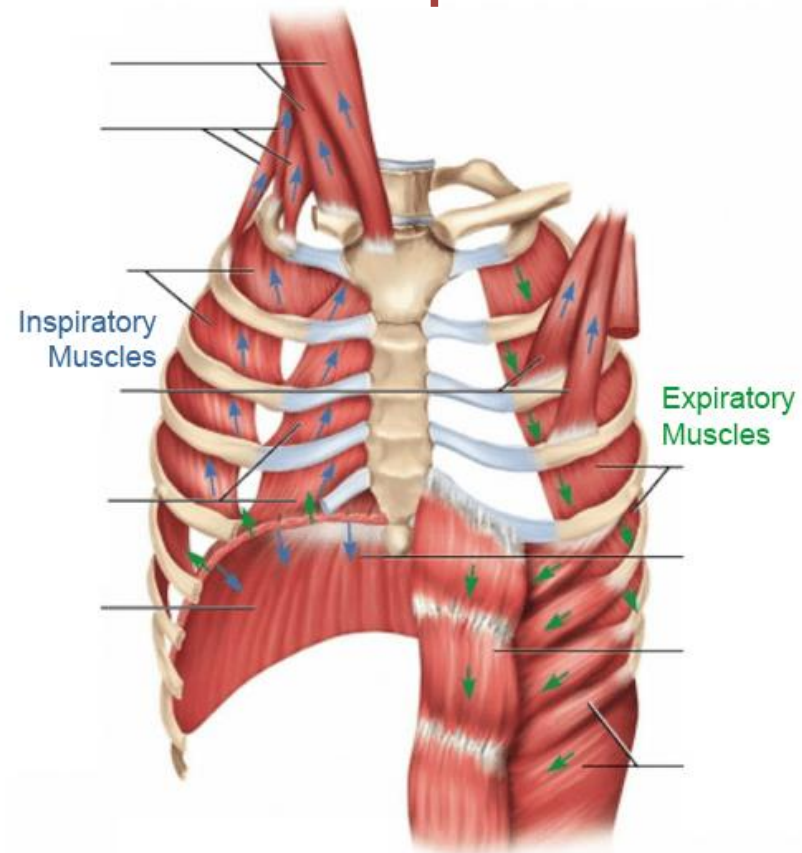
Penn Medicine

The Two Vital Organs of Respiration

Lungs



Respiratory Pump



Respiratory Muscle Failure

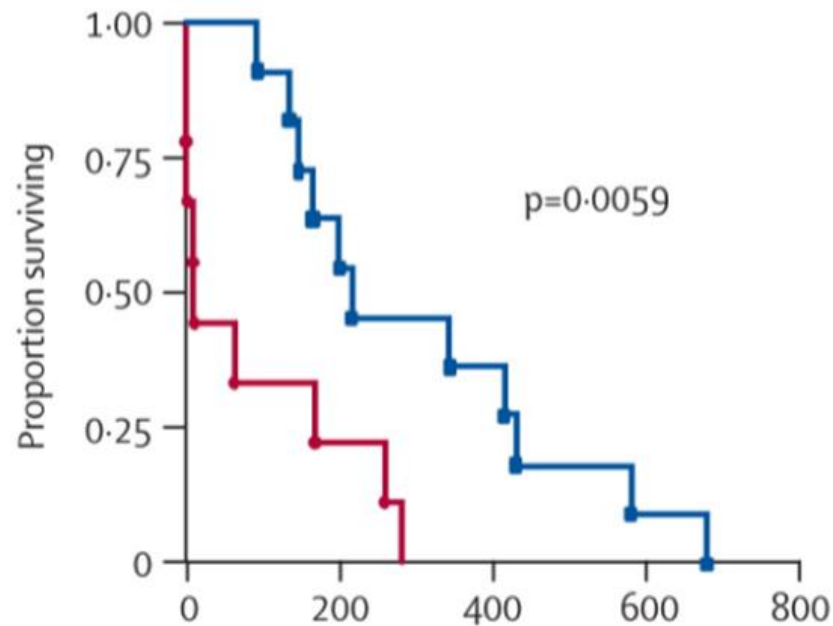
- Breathlessness
- Impaired Sleep
- Weak Cough
- Brain Fog, Confusion, Lethargy

Respiratory Muscle Failure in ALS

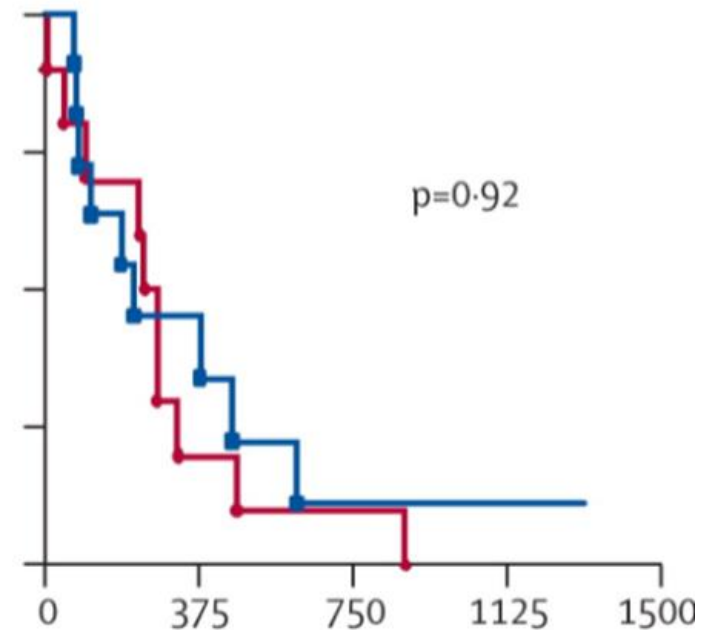
- Breathlessness
- Impaired Sleep
- Weak Cough
- Brain Fog, Confusion, Lethargy
- **Death**

Noninvasive Ventilation Prolongs Survival

2006 Randomized trial of **NIV** vs. **then standard care** for ALS



Normal or mildly impaired bulbar dysfunction



Severe bulbar dysfunction

-- Bourke SC et al. Lancet Neurology. 5:140, 2006.

Noninvasive Ventilation for ALS

- The **good** news:

The state of the art of NIV has advanced dramatically since 2006

Noninvasive Ventilation for ALS



Noninvasive Ventilation for ALS



Noninvasive Ventilation for ALS



Noninvasive Ventilation for ALS



Noninvasive Ventilation for ALS



Noninvasive Ventilation for ALS



Noninvasive Ventilation for ALS

- The **bad** news:

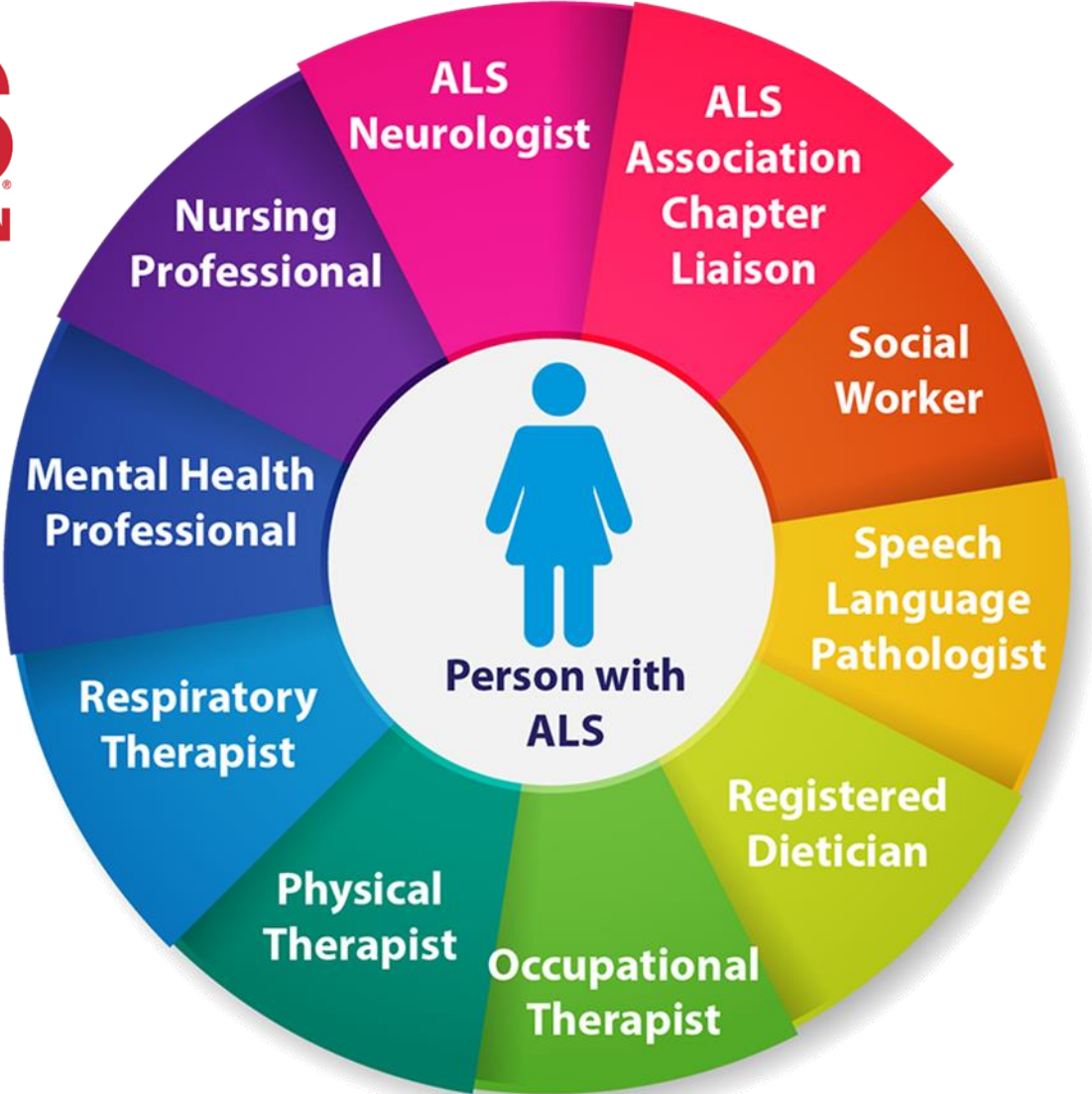
Only a small fraction of people with ALS in the US have access to optimal respiratory care for ALS.

Noninvasive Ventilation for ALS

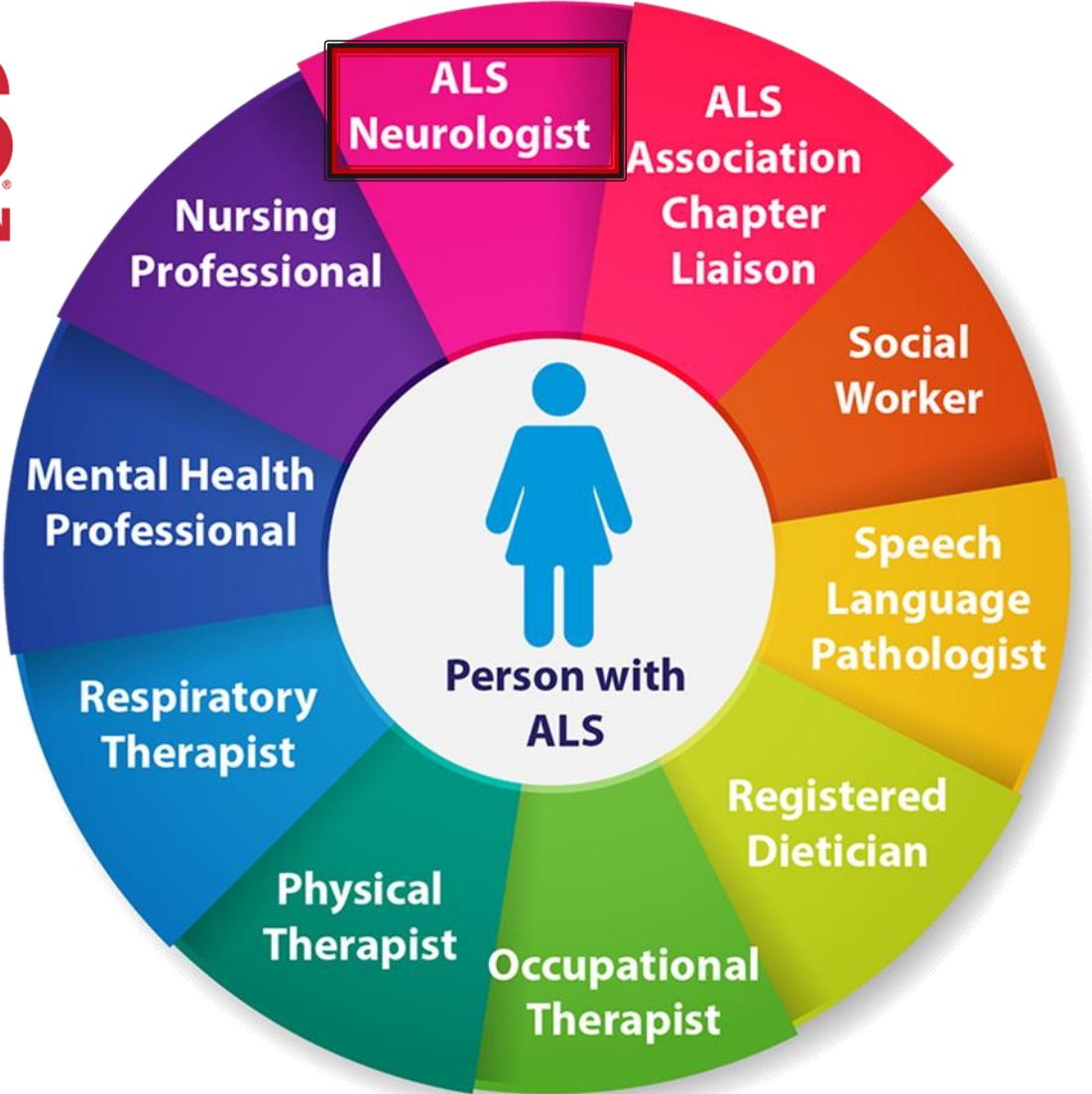
2019 Analysis US National PRO-ACT clinical trial participant database

- NIV was used at the time of **only 52.5% of office visits** when the Forced Vital Capacity (FVC) was less than 50% predicted.
- **15%** of study participants who initiated NIV **subsequently abandoned treatment.**

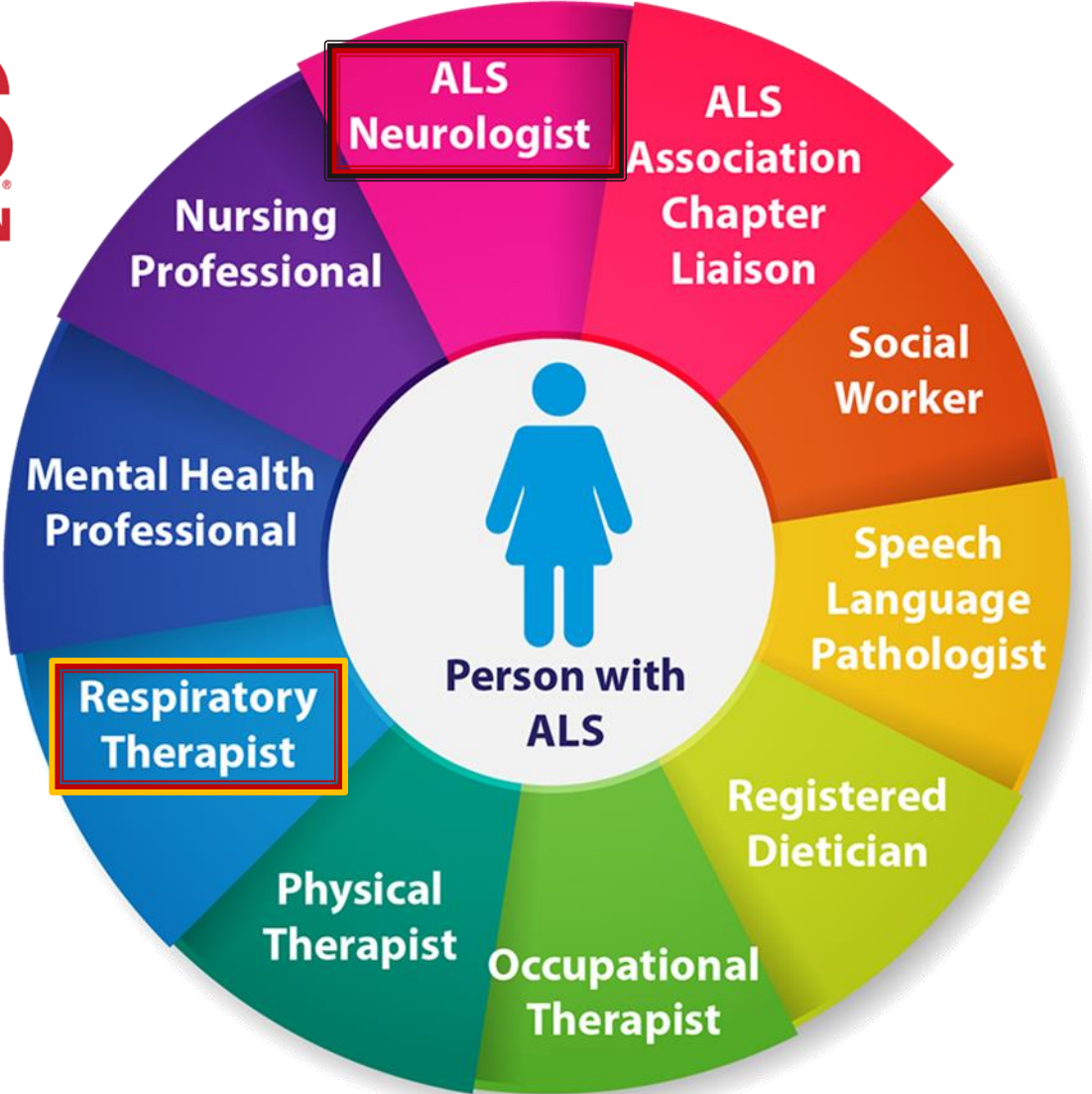
Conventional Care Model for ALS



Conventional Care Model for ALS



Conventional Care Model for ALS



Conventional Respiratory Care for ALS

- A neurologist submits an order for a bilevel device or a ventilator to a durable medical equipment company.
- A respiratory therapist sets up a device in the person's home adjusted to comfort.
- When the machine no longer provides adequate support, the person is advised to consider tracheostomy or hospice.
- Alternately, the person is emergently admitted for acute respiratory failure to an ICU and tracheostomy is considered at that time.

Conventional Respiratory Care for ALS

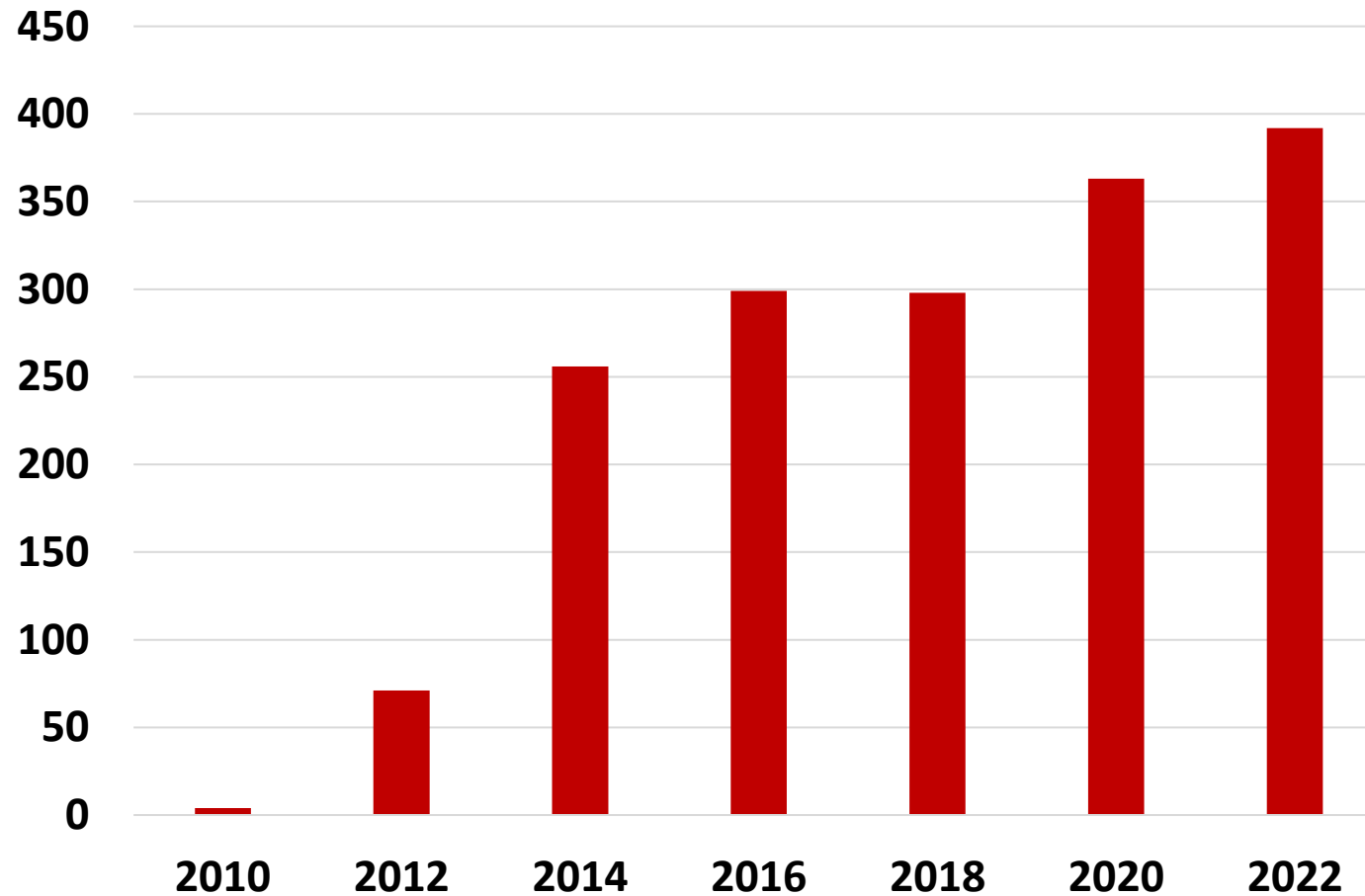
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Noninvasive Ventilation

NIV Publications





Proposed New Care Model for ALS

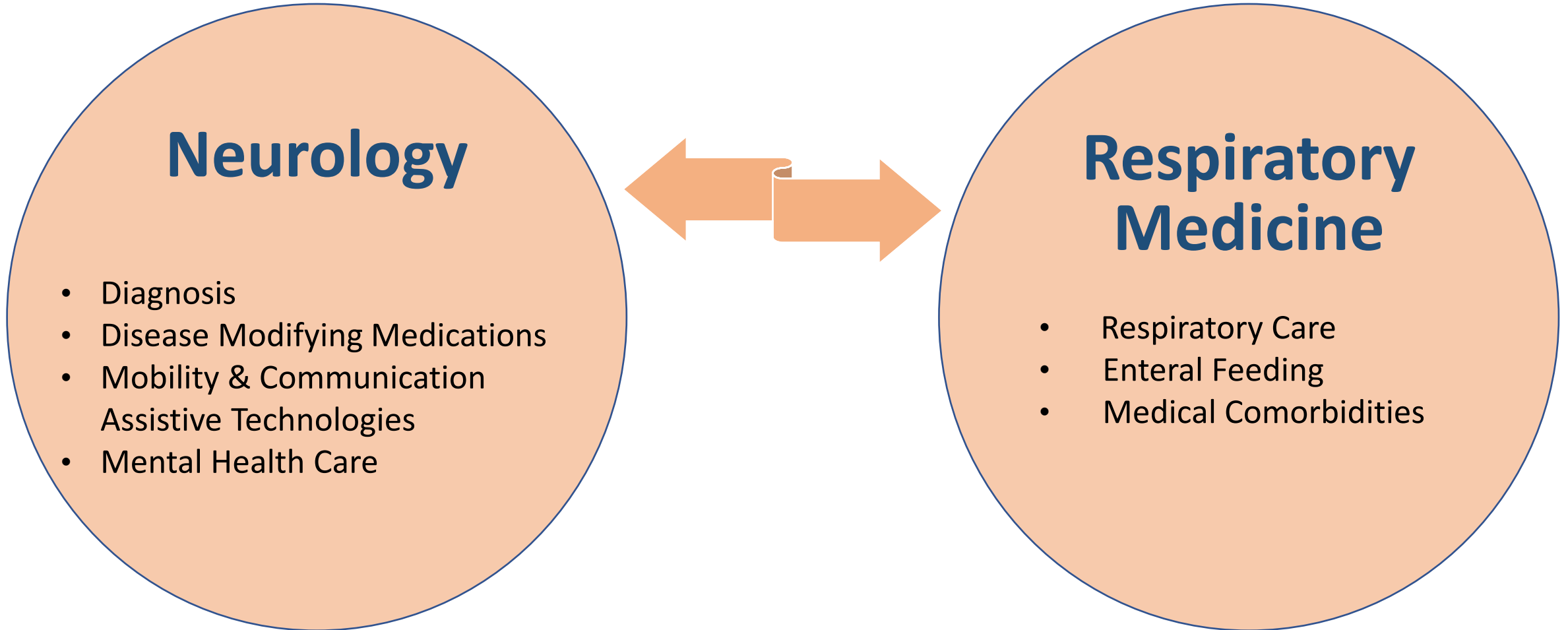
Neurologist and a physician specialist in chronic respiratory failure collaborate in co-principal care from diagnosis to death.

Proposed New Care Model for ALS

Neurologist and a physician specialist in chronic respiratory failure collaborate in **co-principal care** from diagnosis to death.

Proposed New Care Model for ALS

Co-principal care model



Goals for ALS Respiratory Care

- Enable restful, restorative sleep
- Assure comfortable breathing when awake
- Maintain normal gas exchange night and day
 - $\text{SpO}_2 \geq 92\%$; PCO_2 35 – 45 mmHg

Optimal Respiratory Care for ALS

1. Early referral to a physician specialist in chronic respiratory failure.
2. Early initiation of noninvasive ventilation and cough assistance.
3. Physician-directed, ongoing optimization of home respiratory care guided by quantitative monitoring.
4. Early initiation of tube feeding.
5. Advanced planning for home care of respiratory infections.
6. Option for palliative withdrawal of life support.

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Optimal Respiratory Care for ALS

Ideally start assisted ventilation for FVC < 80% with early symptoms of respiratory pump failure.

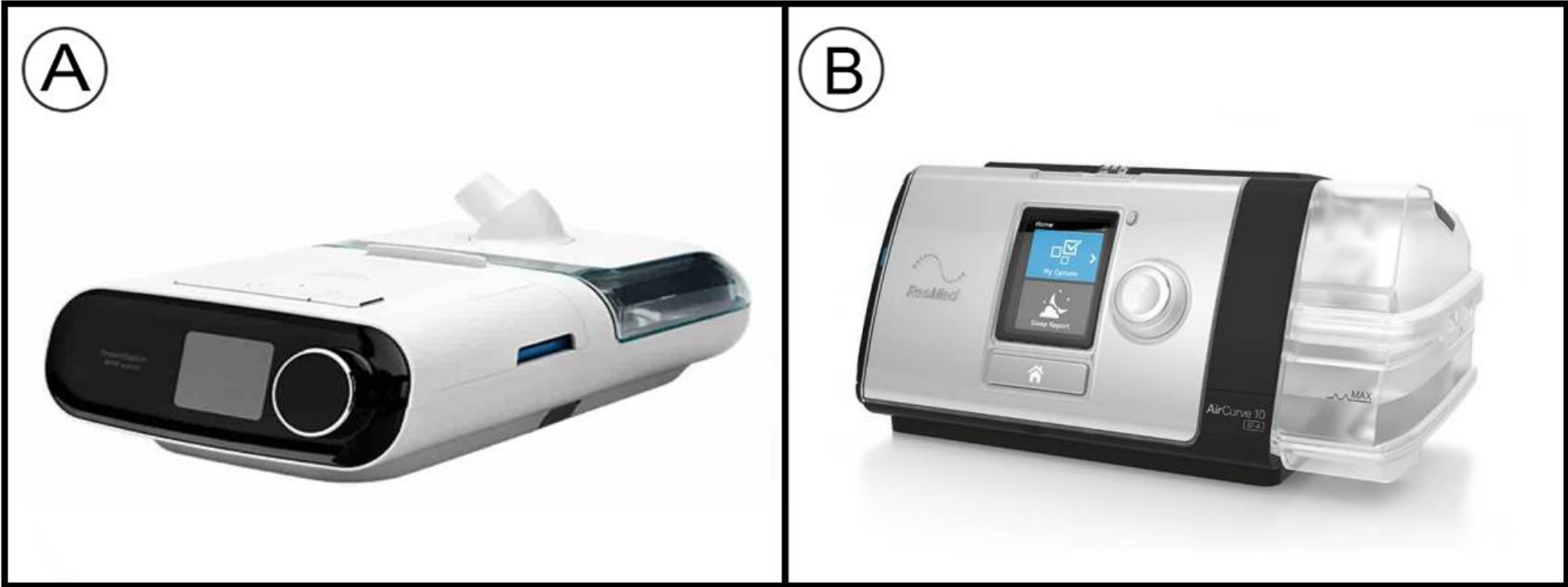
- Uncomfortable breathing
- Interrupted, unsatisfying sleep
- Orthopnea

Optimal Respiratory Care for ALS

Devices for assisted ventilation

- Bilevel device or ventilator with a heated humidifier
- Face mask
- Pulse oximeter

BiLevel Devices



Home Ventilators



Face Masks



Heated Humidifier



Pulse Oximeter



Nonin Go2



Wellue
O2 Ring



Optimal Respiratory Care for ALS

Ideally start assisted ventilation for FVC < 80% with early symptoms of respiratory pump failure.

- Uncomfortable breathing
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Early start to assisted airway clearance

- Persistent chest congestion
- Cough peak flow < 260 L/minute

Optimal Respiratory Care for ALS

Devices for assisted airway clearance

- Cough Assist Device
- Suction machine
- Nebulizer

Airway Clearance Devices



Cough Assist Device



Suction Device



Hand-held Nebulizer

VOCSN Multifunction Device



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In-Office Ventilator Titration



Optimal Respiratory Care for ALS

- Frequent reassessment and adjustment of respiratory assist devices.



Transcutaneous CO2 Monitor

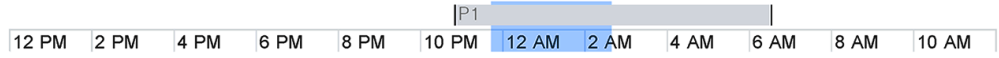


Home Device Telemonitoring



AirView™ Detailed report

01/05/2022



Settings

Circuit type: **Single with leak** | Mode: **STSV** | PIP: **10 cmH2O** | EPAP: **4 cmH2O** | Resp. rate: **15 breaths/min**

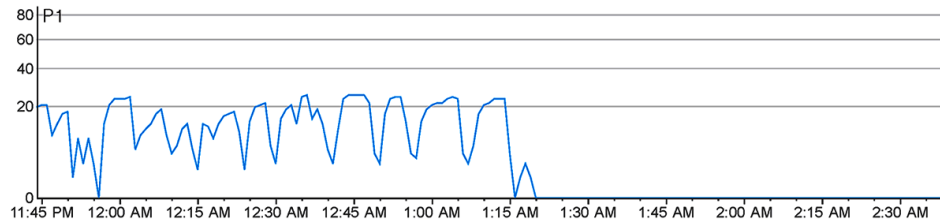
P2 (STSV)

Usage

9 hrs 34 mins total | P1: 0 hrs 0 mins | P2: 9 hrs 34 mins | P3: 0 hrs 0 mins | P4: 0 hrs 0 mins

Leak

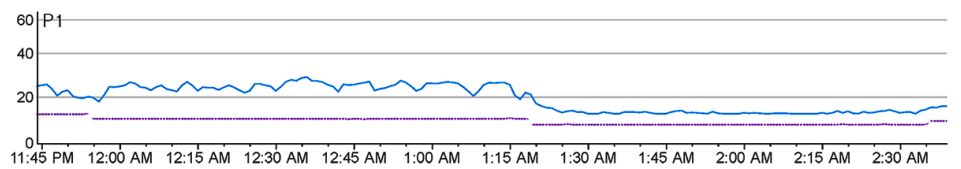
Median: 2.8 L/min



Pressure

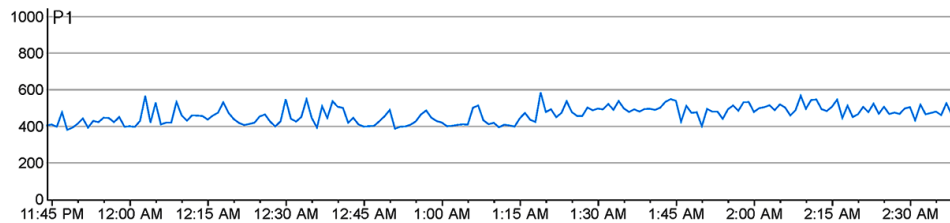
PIP median: 20.6 cmH2O

EEP median: 7.9 cmH2O



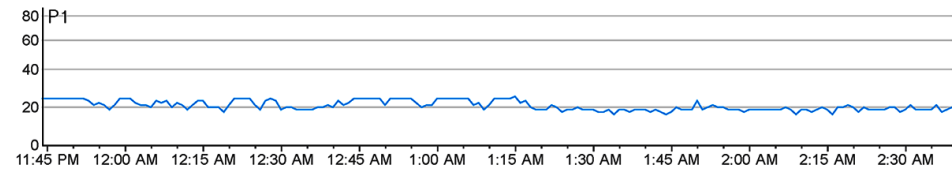
Tidal volume

Median: 465.5 mL



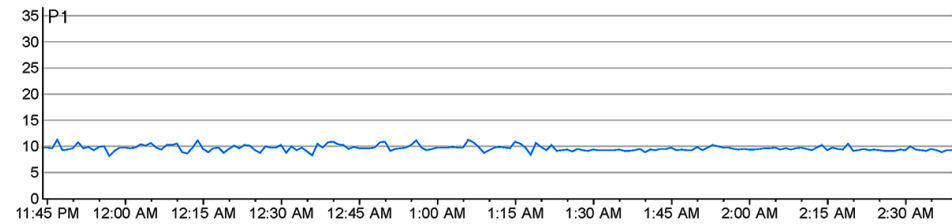
Resp. rate

Median: 20.0 breaths/min



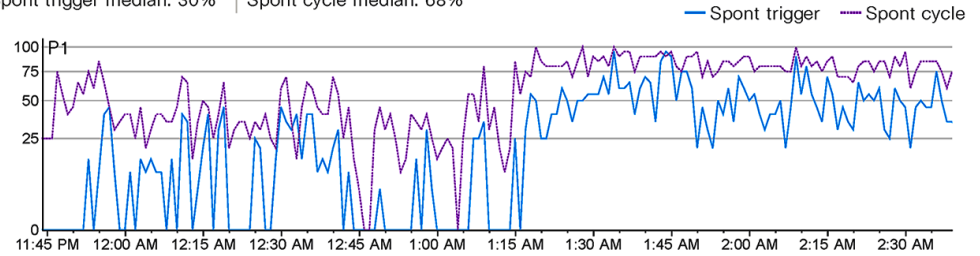
Minute vent

Median: 9.6 L/min



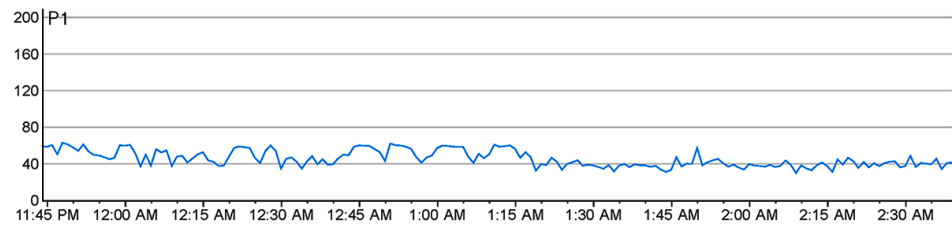
Spontaneous trigger and cycle

Spont trigger median: 30% | Spont cycle median: 68%



RSBI

Median: 43.2 breaths/min/L

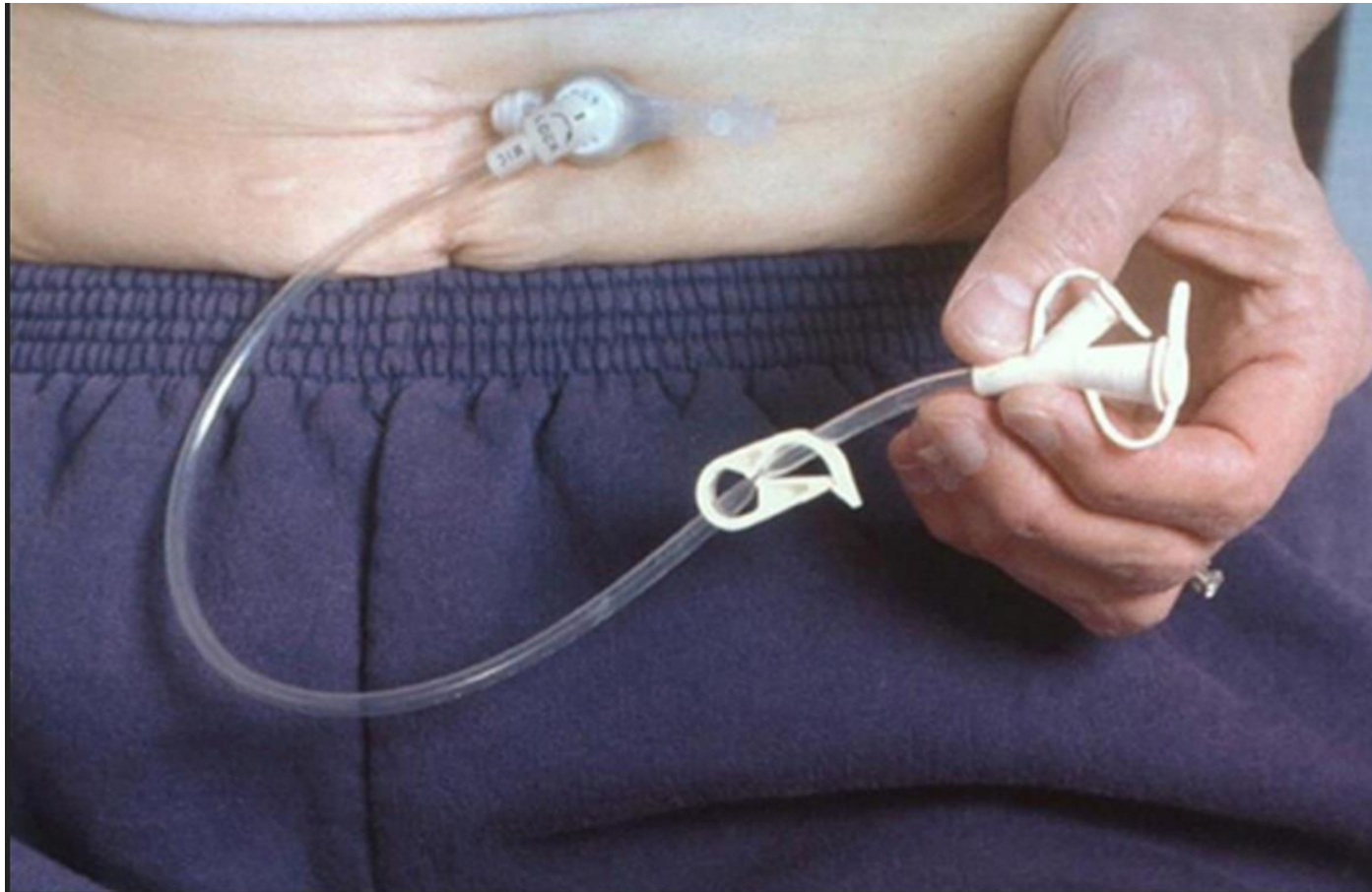


Optimal Respiratory Care for ALS

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Optimal Respiratory Care for ALS

- Early initiation of tube feeding to minimize aspiration.



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Advocacy for ALS Respiratory Care

change.org

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I Support Including ALS Respiratory Research in the "ALS Strategic Plan"



Victory

This petition made change with 695 supporters!



I Support Including Respiratory Research in the "ALS Strategic Plan"



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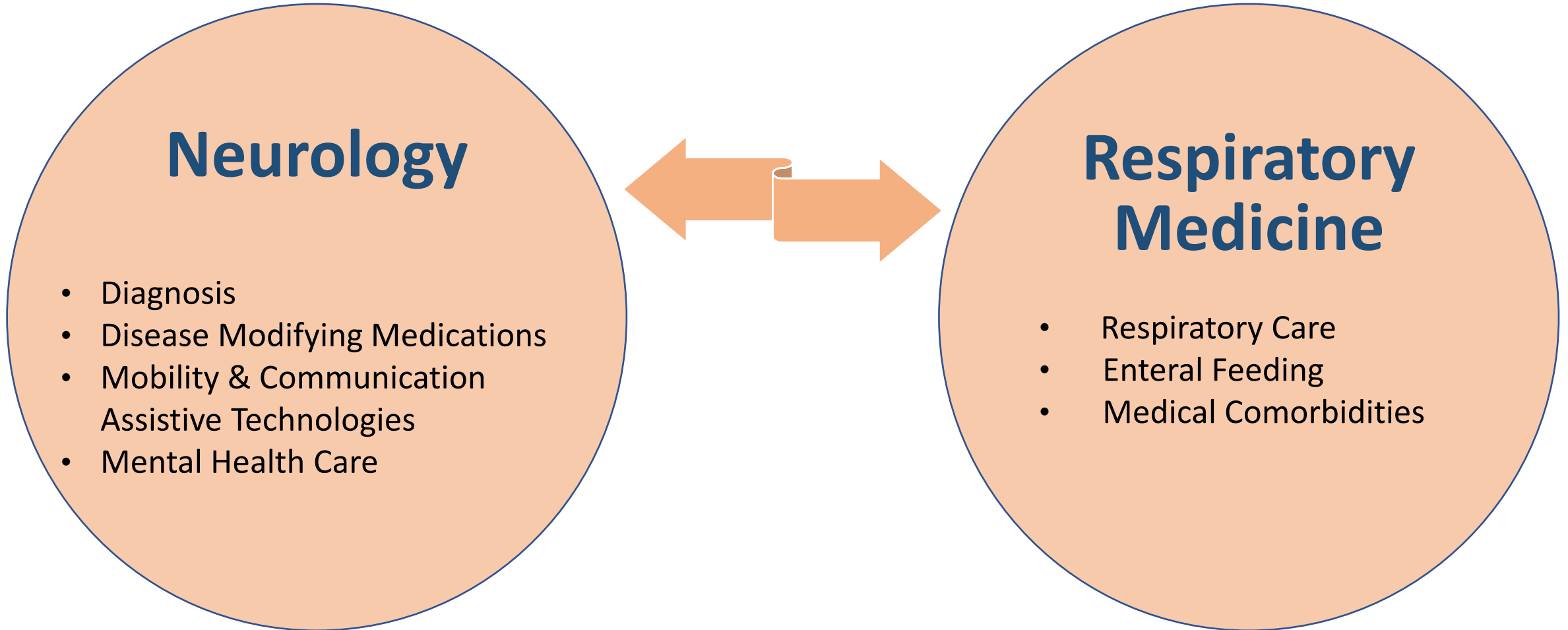
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Advocacy for ALS Respiratory Care

Amyotrophic Lateral Sclerosis: Accelerating Treatments and Improving Quality of Life

MDA Model for Care of Neuromuscular Diseases

Co-principal care



Breathing is Everything

