

### IMPORTANT

It is common for people exhibiting respiratory difficulty to be treated with supplemental oxygen.

If you are sent to the emergency room with respiratory complications, be prepared to communicate to the health-care team that you have a **progressive neuromuscular disorder** and that receiving supplemental oxygen without ventilatory assistance can worsen the situation.

There may be other additional conditions (ex. pneumonia) that necessitate the use of supplemental oxygen. The medical team must closely monitor your CO<sub>2</sub> levels. Supplemental O<sub>2</sub> can be given through the supportive ventilation.

### VENTILATION VS. RESPIRATION

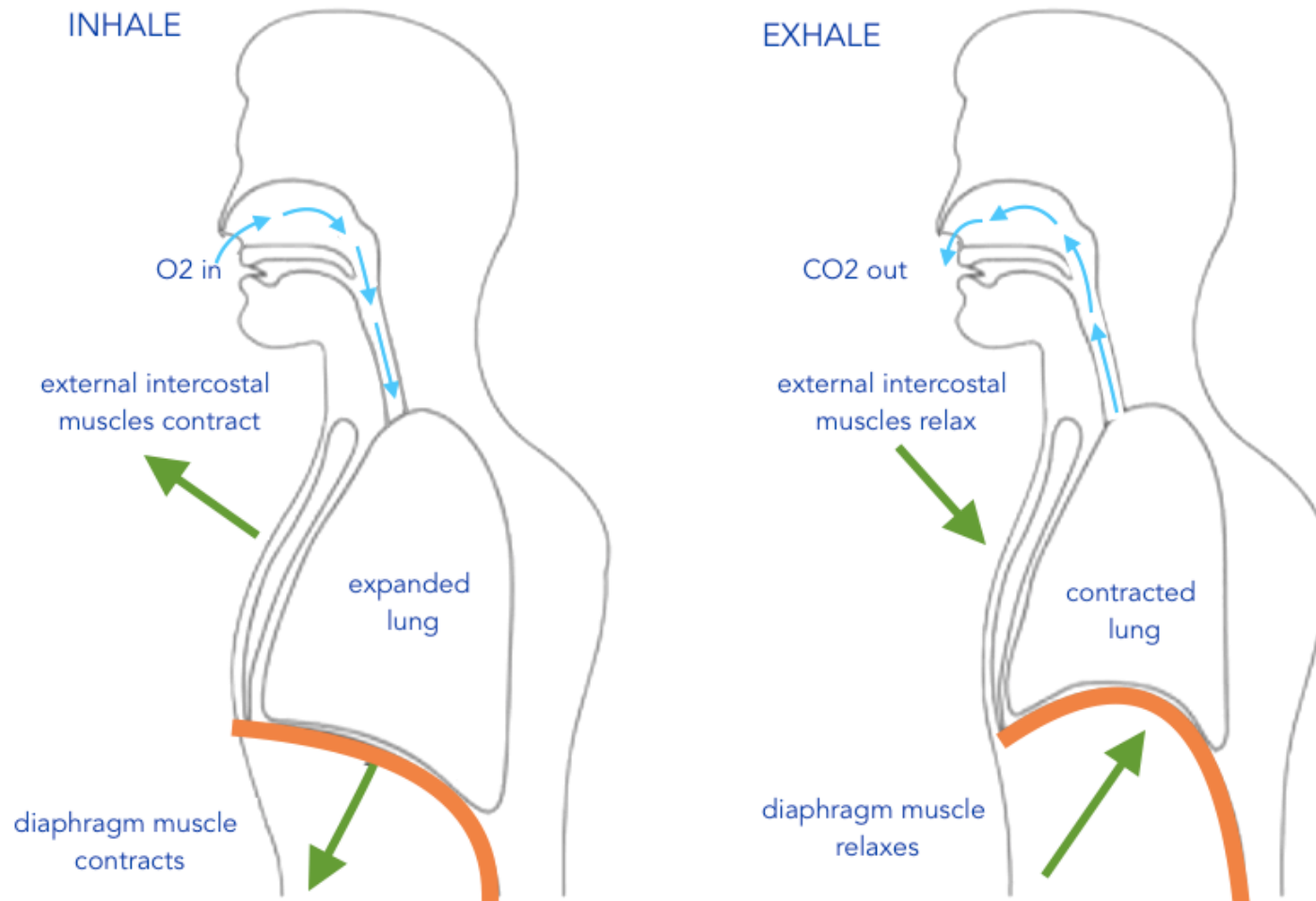
#### VENTILATION

- ★ mechanics of breathing
- ★ voluntary action
- ★ the physical process of inhaling oxygen (O<sub>2</sub>) and exhaling carbon dioxide (CO<sub>2</sub>)

#### RESPIRATION

- ★ takes place in the cells
- ★ involuntary action
- ★ the chemical process where the body breaks down the oxygen (O<sub>2</sub>), so that the cells in the body can use it.

The **diaphragm** is a muscle. It moves the lungs, which then pull O<sub>2</sub> in, push CO<sub>2</sub> out. Supplemental O<sub>2</sub> is NOT for a person living with ALS. When it is done through a nasal cannula ONLY then the additional O<sub>2</sub> is being pushed **in** but there is no compensation made to then push that extra CO<sub>2</sub> **out**. Unless there is an underlying condition, a pALS doesn't have a problem with their lungs – it is the **diaphragm** that is weakened.



People living with ALS (and other neuromuscular disorders and/or diaphragm weakness) require VENTILATORY support.

## RESPIRATORY TERMS

*Assisted Ventilation* – a mechanical means used to assist or replace spontaneous breathing. This can be done through an invasive or non-invasive measure.

*IV* – invasive ventilation (through a tracheotomy)

*NIV* – non-invasive ventilation (mask, sip n' puff, nasal pillows, etc.)

Vent types most commonly used in ALS:

1. Trilogy [www.respironics.com](http://www.respironics.com)
2. LTV [www.carefusion.com](http://www.carefusion.com)

There are stand-alone machines – biPAP, CPAP (not for ALS), AVAPS, etc. but the functions they perform are all included within the most frequently used machine, the Trilogy 100 ventilator.

★ The TRILOGY can be used with both NIV & IV interfaces

### TRILOGY 100 VENTILATOR

*AVAPS* (Average Volume Assured Pressure Support) - automatically adapts to disease progression and changing needs while maintaining a target tidal volume, and has been shown to reduce elevated CO<sub>2</sub> levels.

*AVAPS-AE* offers additional automatic adjustment of *EPAP* (Expiratory Positive Airway Pressure) and a backup breath rate to meet the needs of the most challenging patients, including those needing longer expiratory times to reduce the effects of flow limitation and air trapping.

*BiPap* (Bi-level Positive Airway Pressure) - provide a variable inspiratory pressure (inhale) and a lower expiratory pressure (exhale).

All settings within the trilogy, programmed by respiratory team.

**Cough Assist Machine** - helps to clear secretions from the lungs by helping you with your breathing. When you breathe in (inspiration), the machine gives you air (positive pressure) to help expand your lungs. When you blow out (expiration), the machine creates a sucking force (negative pressure) that pulls the air out of your lungs. This rapid change in pressure during the two phases of breathing helps make your cough stronger and more effective.

**Portable Suction Machine** - Many times, people with ALS also have weakness in their tongue muscles. This weakness makes it impossible to move phlegm to the front of the mouth so that it can be spit out. A portable suction machine can be used at home. It can help to help suck up saliva or secretions that stay in the back of the throat.

**Ambubag** - device used to manually give air or oxygen to a person under pressure. It is an important item to become familiar with in case of **emergency**. Respiratory therapists also often use it to teach "breath-stacking techniques".

**Breath Stacking** – an exercise to help maintain strength, or slow loss of strength, of muscles used for breathing & keep lungs clear of secretions.

possible benefits

- ❖ Keep the lungs clear of secretions
- ❖ Help to prevent chest infections from developing
- ❖ Keep lungs and chest wall flexible
- ❖ Make it easier to cough effectively
- ❖ Improve voice strength

## RESPIRATORY MEASUREMENT TERMS

**FVC** – **Forced Vital Capacity** – measures lung function – the amount of air pushed out of the lungs as quickly as possible. It should be measured **both sitting up and laying down**.

**SVC** – Slow Vital Capacity – measures lung function - the amount of air pushed out of the lungs as slowly as possible, with minimum effort.

**MIP** - maximum inspiratory pressure

**MEP** - maximum expiratory pressure

**SNIP** - sniff nasal inspiratory pressure