Welcome!



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Respiratory Challenges and Options in ALS

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Understanding breathing and cough impairment in ALS patients





Introduction



Amyotrophic lateral sclerosis (ALS)

- In the U.S. ALS incidence rate is approximately 2/100,000 and ALS prevalence is approximately 6/100,000
- The majority of ALS cases are the result of an unknown cause
- Between 2015 and 2040 the number of ALS cases globally is expected to increase by 69%
- ALS patients suffer a steep decline in lung function approximately 2 3% per month
- Most ALS patients will face some degree of respiratory insufficiency or failure throughout their disease progression





Breathing mechanics

The basic mechanics of breathing

Understanding the mechanics of natural breathing.

Inhalation:

- 1. Requires active effort from the body
- 2. Diaphragm and intercostal muscles contract
- 3. Thoracic volume increases, decreasing alveolar pressure
- 4. Air rushes in from the atmosphere because alveolar pressure is less than atmospheric pressure



The basic mechanics of breathing

Understanding the mechanics of natural breathing.

Exhalation:

- 1. Diaphragm and intercostal muscles relax, returning to their original position
- 2. Lung recoils to its original size and shape
- 3. Thoracic volume decreases, increasing alveolar pressure
- 4. Air rushes out of the lungs because alveolar pressure is greater than atmospheric pressure
- 5. Diaphragm and intercostal muscles are at rest at the end of exhalation
- 6. Atmospheric and alveolar pressure are equal, resulting in no airflow



What is a cough?

- A reflex that clears the airways of foreign particles and irritants
- Coughing can be voluntary or involuntary
- Coughing is important in maintaining respiratory health
- An ineffective cough increases risk of infection
- Repetitive coughing can be a sign of a serious condition



The basic mechanics of a cough

Understanding the mechanics of natural coughing is important. There are 3 phases to a cough:

Inhalation phase:

- Individual takes a large, deep breath, to generate the volume or air needed to clear secretions
- Inspiratory effort comes close to the individual's maximal inspiratory capacity (MIC) and total lung capacity (TLC)





The basic mechanics of a cough

Understanding the mechanics of natural coughing is important. There are 3 phases to a cough:

Compressive phase:

- Glottis closes
- Abdominal muscle contracts causing thoracic compression
- Compression cause a large pressure increase (≈ 100 cmH₂O)





The basic mechanics of a cough

Understanding the mechanics of natural coughing is important. There are 3 phases to a cough:

Explosive phase:

- Alveolar pressure is released by the opening of the glottis in a sudden and explosive manner
- Airway secretions are cleared from the central airways
- Cough strength is typically measured using peak cough flow (PCF)
- Effective PCF in a healthy adult is typically between 360 1000 L/min



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Topic 3 Airway clearance components

Why is a cough so important?

- Our lungs are naturally resistant to poor quality air in the atmosphere
- Our natural resistance is tanks to two connected mechanisms:
 - Mucocillary escalator, which uses mucus to trap toxins and move them to the central airway
 - Coughing, which clears the toxins that have been transported to the central airway
- Airway clearance therapies (ACTs) have been created to replicate these mechanisms for those unable to naturally achieve one or both
 - Proximal airway therapy augments the cough mechanism, such as MI-E
 - Peripheral airway clearance therapy augments mucocillary escalator



The mucociliary escalator

- The mucociliary escalator uses mucus to trap toxins and move them to the central airway:
 - Mucocillary escalator, which uses mucus to trap toxins and move them to During inhalations, mucus traps impurities
 - The impurities are transported to the central airways by a large collection of tiny hairs, called cilia
 - The cilia beat, transporting the impurities to the central airway
- Examples of peripheral ACT often incorporate oscillatory airflow which loosens secretions from the airway wall and makes mucus less sticky and easier to evacuate by coughing. This oscillatory airflow effect may be created by use of:
 - An inflatable vest (externally created)
 - MetaNeb, IPV, and CoughAssist oscillatory feature (internally created)
 - Oscillatory PEP valves (patient effort dependant)





Measuring respiratory muscle strength

Peak cough flow

- One way a healthcare provider can measure cough effectiveness is by using a simple test called peak cough flow (PCF)
- Healthy individuals will produce a PCF between 360 and 1000 L/min
- 160 L/min is the minimum PCF needed to clear airway secretions
- Individuals diagnosed with a neuromuscular disease will have their PCF monitored
- A PCF of less than 270 L/min is an indication that assistance Is required
- However, patients with a PCF between 270 L/min and 360 L/min should still be provided with cough assistance





Available treatment options

Bi-level therapy devices

A blower within the device delivers a prescribed amount of air through a mask into your lungs. The level of pressure will be higher when you inhale and will decrease as you exhale.

- **Bi-level S (spontaneous)** responds when you start to inhale and exhale. There is no automatic delivery of a breath if you do not inhale
- **Bi-level S/T (spontaneous/timed)** responds when you start to inhale and exhale. If you do not start inhaling within a set time, the device automatically delivers a breath
- Respironics Average Volume Assured Pressure Support (AVAPS) feature guarantees a desired volume of air to the lungs.



Pressure or volumecontrolled ventilators

Pressure - or volume-controlled ventilators deliver a preset pressure or volume of air when you inhale. These ventilators can deliver higher volumes and pressure than bi-level units. They also have additional alarms and internal batteries.



Secretion management

Secretion management therapy such as use of CoughAssist from Philips Respironics can supplement your ventilation therapy to increase the effectiveness of your cough and clear secretions from your lungs and trachea. This small electrical machine first delivers a large volume of air, then quickly reverses air flow to pull out secretions. This will keep your airways clear and help reduce the chance of respiratory infection.



Humidification

A humidifier adds moisture to the air that is being delivered to you from your ventilator. It can help alleviate most nasal problems. There are two types of humidification - heated and unheated. Heated humidifiers do not actually 'heat' the air. Instead, they increase the moisture content of the air that is delivered to you. Humidifiers can help to reduce some of the side effects of NW therapy, such as dry nasal passages.





What is mechanical ventilation and why is it needed?

Key points

From hospital to home or clinicians to caregivers, transitions can be challenging.





Ventilation assistance

For some people, the routine process of taking air in and out is far from simple. When your breathing is interrupted - either because your muscles are too weak to pull air into your lungs or because your airways are blocked - you may experience a variety of symptoms.

Typical symptoms may include fatigue, shortness of breath and difficulty lying flat. When you wake up, you may have a headache or feel confused, disoriented and anxious. You may also lose your appetite and develop a weak cough.

To help, your doctor may recommend assisted ventilation. This will come in the form of mechanical ventilation, which is used to support or replace a person's natural breathing when it's not possible for them to breathe themselves.



How does breathing work?

Your lungs are vital organs that bring fresh oxygen into your body and remove carbon dioxide. This process known as ventilation.

- 1. When you breathe in, your diaphragm tightens and flattens, allowing you to take air into your lungs.
- 2. The air passes through the mouth and into the trachea. The trachea is divided into two smaller air passages called bronchial tubes. One leads to the left lung, the other to the right.
- 3. The bronchial tubes divide into yet smaller air passages called bronchi, and then into bronchioles.
- 4. At the end of the bronchioles are air sacs called alveoli. When you breathe in, the alveoli expand as air rushes in.
- 5. Tiny blood vessels that surround each alveoli transfer oxygen from the inhaled air to the blood, and also allow carbon dioxide and waste gases to be exhaled.



Patients with neuromuscular diseases

When neuromuscular disease affects the nerves that control the respiratory muscles, breathing becomes strained and weak.

- 1. You may become short of breath when you exert yourself or while you're lying in bed. This "breathlessness" is an important indication of trouble.
- 2. You may need to prop yourself up on pillows or even try to sleep sitting up in a chair.
- 3. You might also find it's difficult to cough and to control swallowing.

These symptoms can lead to infection or to blocked airways. It's important to contact your doctor as soon as you experience these symptoms so that further complications will not occur.



Ventilation for patients with neuromuscular diseases

Although your respiratory muscles might be severely weakened, there are ways to maintain or improve your quality of life.

Noninvasive ventilation can help patients to be more independent and reduces the likelihood that the patient will need to be hospitalized for breathing-related episodes. Many ALS patients, for example, go to school, work or travel and enjoy the company of friends. In fact, ALS patients who use NIV to treat the respiratory insufficiency have significantly better survival advantage than other patients who received standard care.

When deciding on the appropriate level of respiratory support, there is no right or wrong decision. It's a personal choice. Your health care team can help you and your family plan as your needs change.



Key points

Ventilation is essential for life. For some people natural ventilation and oxygenation is limited or impossible, so mechanical ventilation in used to:

- provide or improve ventilation,
- improve oxygenation,
- improve lung mechanics,
- improve patient comfort, and
- minimize any associated complications.



Goals for mechanical ventilation

The goals of mechanical ventilation are:

- Reduce the work of breathing.
- Increase alveolar ventilation.
- Maintain arterial blood gas (ABG) values within acceptable ranges.
- Improve the distribution of inspired gases.



Delivery of mechanical ventilation

Mechanical ventilation is delivered in one of the following way:

Non-invasive ventilation Delivered to the patient via an interface such as a mask or a tube.

Invasive ventilation Delivered to the patient in one of two ways;

• Endotracheal tube - a tube that is inserted into the mouth or nostrils



Delivery of mechanical ventilation

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Invasive ventilation Delivered to the patient in one of two ways;

- Endotracheal tube a tube that is inserted into the mouth or nostrils
- Tracheostomy tube a tube that is inserted directly into the trachea via a neck incision



The future of respiratory care with Trilogy Evo

The only portable life-support ventilator platform designed to stay with patients and provide consistent therapy and monitoring as they change care environments, and when their condition changes.*



* Valid for 1 year



Expanding ventilatory support with MPV

MPV is a form of ventilation whereby the patient's normal state is disconnected from the ventilator and the patient initiates a breath, as needed, through an oral interface

The 'kiss' trigger with signal flow technology detects when the patient engages and disengages from the mouthpiece to deliver on-demand ventilation

This feature combines with a mouthpiece ventilation (MPV) support system to enhance ease of use

MPV with Trilogy Evo

Optional time-based patient reminder

MPV circuit disconnect alarm

Multiple prescription function

Facilitates independent day and nighttime settings (i.e. MPV • during day, mask ventilation at night)

Kiss trigger

- Unique algorithm for a normally disconnected state •
- Eliminates issues with a traditional flow trigger: •
 - no sensitivity to adjust (mitigates auto triggering) ٠
 - does not require patient effort to generate a breath •
 - important for progressively weaker respiratory muscles •



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User-friendly platform

Consistent technology creates practical benefits for patients and providers

- Easy operation with large touch screen
- Enhanced user proficiency
- Quick learning curve for clinicians and caregivers





Cough therapy

What is MI-E?

- Designed to clear secretions from the lungs by;
 - Applying positive air pressure (insufflation), which introduces a large volume of air into the lungs
 - Quickly shifting to negative pressure, creating a large pressure gradient that rapidly increases expiratory flow rates (exsufflation)
- High expiratory flow rates create the shearing force necessary to clear secretions from the central airways



Secretion clearance

As seen earlier in the module, a cough is made up of 2 components: Mucociliary clearance and cough clearance. If either of these are impaired there are a number of options to support these components.

Mucociliary clearance

- High-frequency chest-wall compression
- Chest Physiotherapy
- Oscillation Devices
- Positive Expiratory Pressure

Cough clearance

- Breathing techniques
- Suctioning
- Manually Assisted Coughing (MAC)
- MI-E technique



The need for MI-E therapy

- An effective cough uses lung volume, a closed glottis, and abdominal compression to create a large pressure gradient to raise expiratory flow rates to a level that will clear airway secretions
- Loss of glottis control, muscle weakness, or both prevents an effective cough
- MI-E can replace or augment an ineffective cough
- MI-E uses positive pressure to create sufficient lung volume. Negative pressure is used to mechanically increase the pressure gradient to raise expiratory flow rates out of the lungs at a rate that is adequate to clear airway secretions
- MI-E can therefore compensate for an inability to close the glottis or create compressive force





The goals of MI-E therapy

The goals of MI-E are:

- 1. Augment an ineffective or weak cough.
- 2. Mobilize and clear airway secretions.
- 3. Prevent lung infections.
- 4. Treat and prevent atelectasis.



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The goals of MI-E therapy

MI-E uses three phases to achieve this:

- 1. Insufflation, which is the application of a large volume of air with the aim of getting as close to the patient's maximal inspiratory capacity (MIC) as possible
- 2. **Exsufflation**, which is the application of negative pressure to increase the expiratory flow rate and clear secretions in the airways
- **3. Pause**, which completes the cycle and provides a break before the next cycle



The goals of MI-E therapy

- One example of treatment settings includes 2 warm up breaths, 4 coughs, repeated once
- Treatments are usually administered 3 times a day (morning, afternoon, evening) before meals



MI-E vs. suctioning

MI-E (left) is clinically proven and better tolerated than endotracheal suctioning (right)

- Expels 26% more mucus
- Improves SpO₂
- Clears airways for longer
- Fewer complications
- Invasive suctioning frequently misses the left lung

MI-E therapy

As compared to suctioning

• Patients* found MI-E therapy to be less irritating, painful, tiring, and uncomfortable

Understanding breathing and cough impairment in ALS patients

- 89% of patients* found MI-E therapy faster
- 78% of patients* found MI-E therapy more convenient
- 72% of patients* found MI-E therapy more effective

*18 spinal cord injury

27/10/2020





MI-E vs. suctioning

MI-E has been shown to:

- Increase peak cough flow more than 4 times
- Increase survivability
- Decrease hospitalizations
- Decrease recurrent respiratory infection
- Prevent the need for tracheostomy





Enhancing therapy with the oscillations

Clinical aim:

Oscillations help releasing mucus from the bronchial walls, increasing mobilization thus improving bronchial drainage

How:

By applying oscillatory vibrations to the airway

Settings:

- The oscillations can be applied to the inhale, exhale, or both phases
- Frequency : 1-20 Hz
- Amplitude : 1-10 cmH2O

Note: In Advanced Auto mode, when oscillations are enabled in the inhale phase, it also applies to the pre-therapy breaths.

	PHILIPS		
1 : Auto Oscillation	01/02/2020 12:45 PM		
Peak Cough Flow	Settings	Insufflation Volume	
Mode Couph-Trak Inhale Pressure Inhale Flow Inhale Time Exhale Pressure Exhale Ime Pause Time Oscillation Frequency Amplitude	Auto ON 40 cmH20 Low 3 sec 2 0 sec Both 9 9 Hz 5 cmH20	Sec	- +
Finish	Navigate \$	Modify	
	CoughAssist T70		

Oscillations use

Requirements for a comfortable and efficient Mechanical In-Exsufflation:

- 1. Enhanced therapy of CoughAssist T70 Ability of the CoughAssist T70 to enhance mobilization
- 2. Adaptation to patient needs Patient needs may vary over time (more sticky mucus, progress of the disease, winter cold)

Philips Respironics' answer:

- Vibration of the airway
- Painless additional therapy
- Feature can be customizable for each patient

Benefits for the clinicians:

- Can enhance MI-E therapy
- Settings can be adjusted during therapy to match patient's comfort



An example of MI-E settings



- Therapy can be adjusted to the need of individual patient
- Use presets for different therapy needs
- Therapy is best before meals and at bedtime
- Frequency of sessions should be according to the patient's case history
- Adaptation is necessary for invasive use
- Help the patient become familiar with the device

	PHILIPS
3.4	Auto 01/02/2019 12:45 PM
	Peak Cough Flow ***** Vimin Tidal Volume ***** mil -30 1.5 4.0 +30 1.5 cmH20 sec sec sec sec
	Settings Menu A Therapy
	CoughAssist T70



An example of MI-E settings

- Increase pressures as tolerated, 35 to 45 cmH₂O ideally (studies have shown that therapeutic PCF may not be reached with MI-E expiratory pressures less than (+/-40 cmH₂O)
 - The goal of expiratory pressure is to replace a good expiratory cough flow
 - Patient synchronization with the device is key to get the best results
 - Huff when patient can't cough
 - Possibility to use abdominal and chest compression during exsufflation
- Higher pressures are often required when compliance decreases or resistance increases (small tracheal tubes, obesity, scoliosis)



CoughAssist acute care considerations

- Patients on a ventilator consider: 100% oxygen 30 – 60 seconds pre/post cough therapy.
- 2. CoughAssist used invasively? Consider increasing pressure for small ET or trach sizes due to increased resistance.
- 3. Oxygen bleed-in?

Option 1: Patient may wear nasal cannula under the CoughAssist if good seal can be created. Option 2: When using oxygen bleed-in adapter, limit liter flow to < 10 lpm to avoid impacting triggering sensitivity and target inhalation pressure delivery.

- 4. Proper cushion inflation and proper head and neck positioning are critical for a good seal and optimal airway opening.
- 5. Oscillations may enhance secretion removal when used during cough therapy.



Summary

In conclusion, ALS patients may benefit from both ventilation and cough therapy.

- ALS patients should use cough therapy to avoid infections
- Cough therapy can be supported with other techniques to mobilize secretions
- ALS patients use ventilatory assistance to avoid breathlessness and support breathing
- Using Philips Respironics Trilogy Evo platform provides patients consistent therapy as they change environments, helping clinicians and caregivers manage patients with confidence and ease
- Trilogy Evo is further improved by MPV functions
- Combination of mechanical ventilation and cough therapy can help avoid exacerbations



