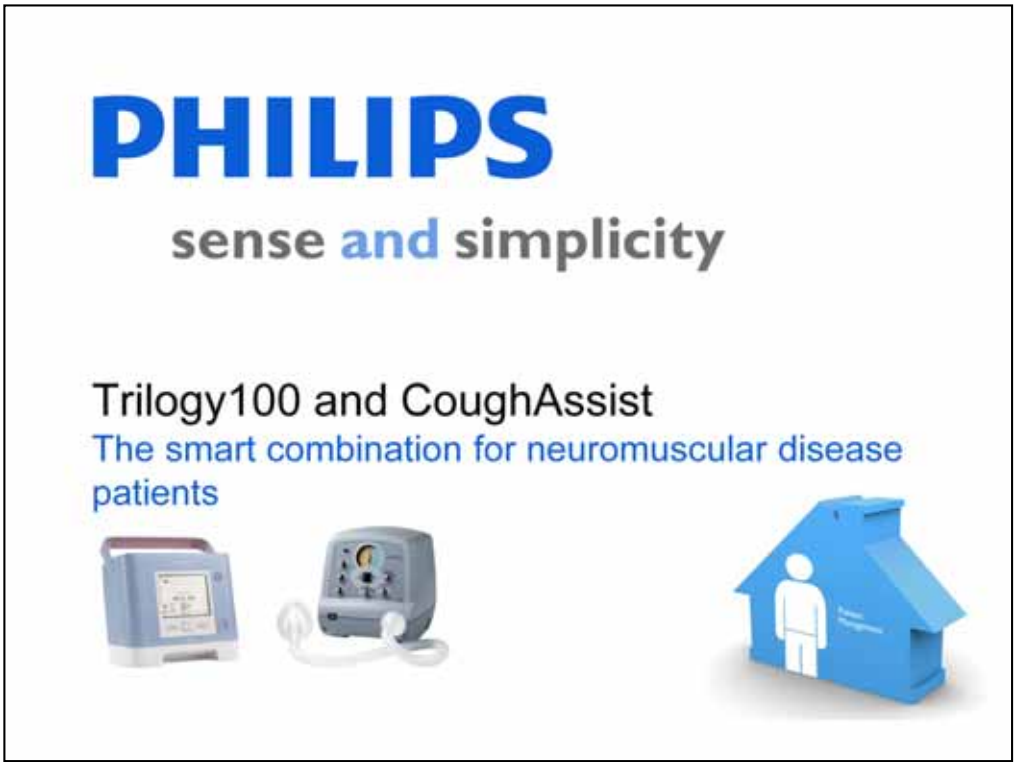
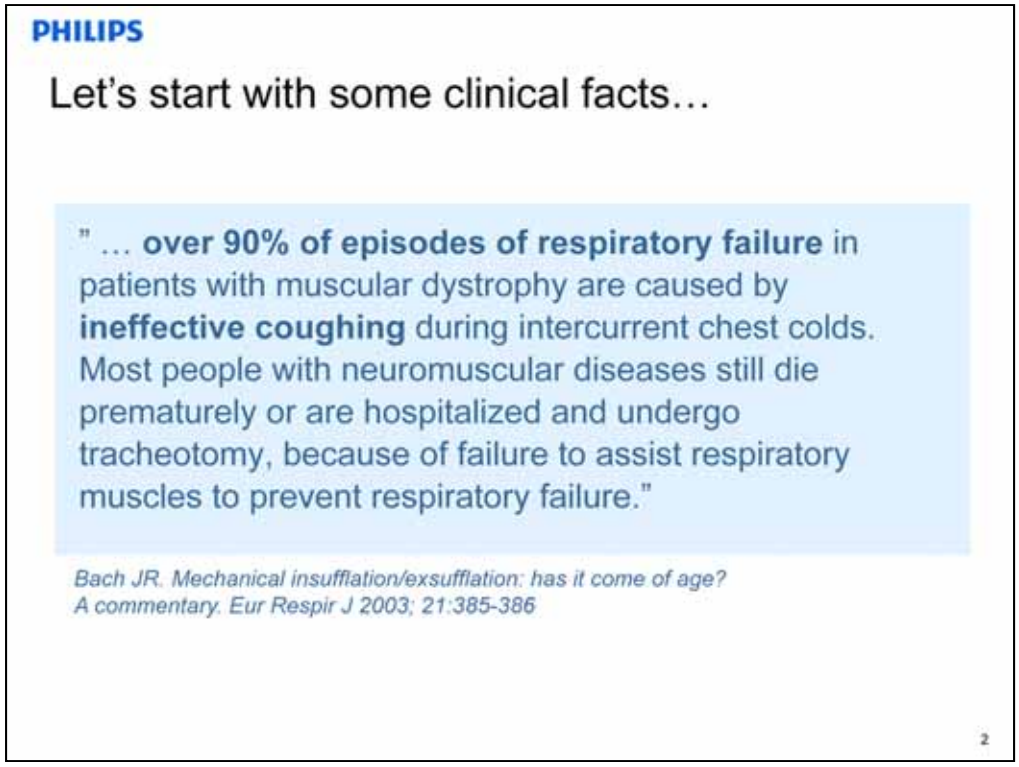


Neuromuscular diseases (NMDs) include both hereditary and acquired diseases of the peripheral neuromuscular system. They are diseases of the peripheral nerves (neuropathies and anterior horn cell diseases), the myoneural junctions (myasthenia gravis), or the muscles (myopathies) themselves. The pathology processes affecting the nerves and muscles can be anatomic, metabolic, electrical, inflammatory or immunologic, as well as idiopathic (unknown cause). These conditions cause muscle weakness, including respiratory muscles, but do not usually affect sensory functions.



Let's start with some clinical background and facts.

At present, the great majority of morbidity and mortality of NMD patients is due to respiratory complications of muscles weakness. That is why pulmonary management is the most critical aspect of treating NMDs – which is clearly illustrated by these quotes from Bach.

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" Neuromuscular diseases (NMD) cause **muscle weakness that can affect […] breathing, coughing** […], all of which can cause disability and compromise quality of life. […]. **Weakness of breathing and coughing muscles, however, continues to be the major cause of repeated hospitalizations and loss of life**."

Bach JR. Extract from the preface of the book "Management of Patients with Neuromuscular Disease"

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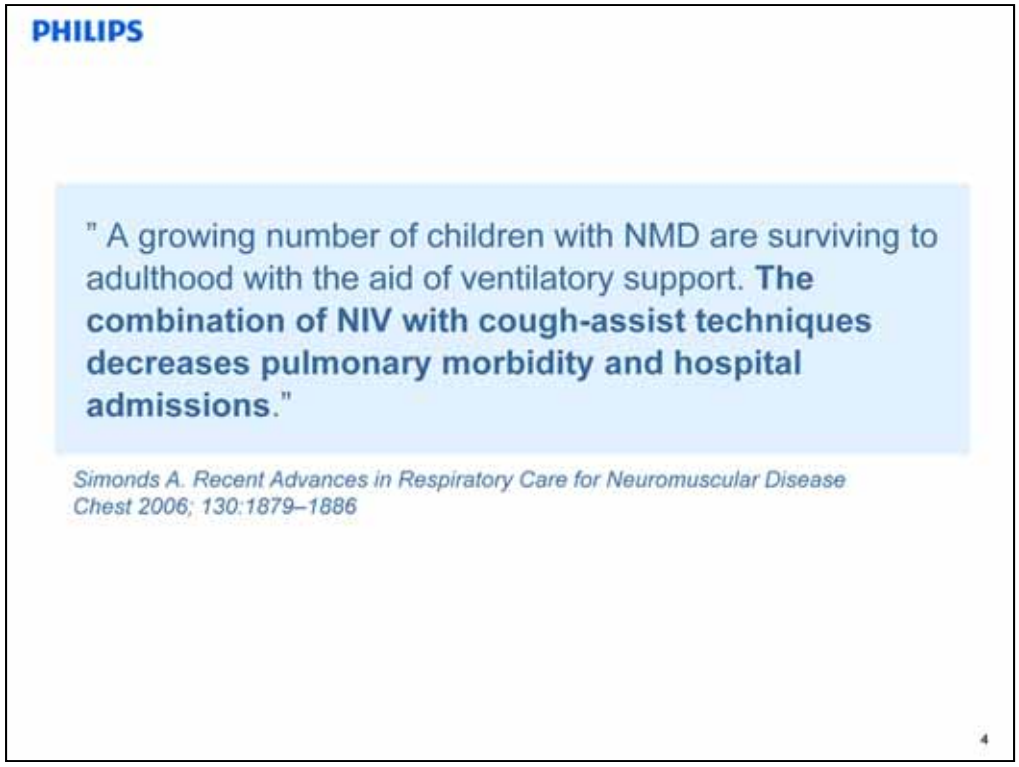


Anita Simonds mentions in her article “Recent advances in respiratory Care for NMD” published in Chest in 2006 that [quote], emphasizing the need to combine coughassist and ventilation techniques to efficiently aid the respiratory muscles.

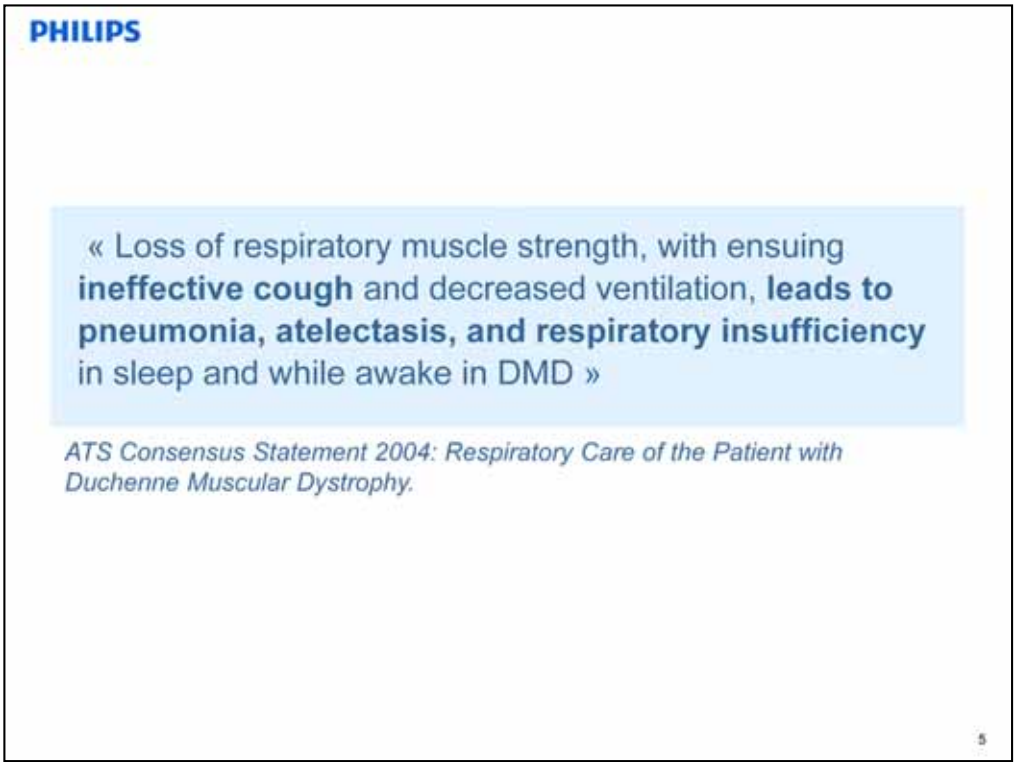
ADDITONAL NOTE: Inspiratory and expiratory muscle aids are devices and techniques that involve the manual and/or mechanical application of forces to the body or intermittent pressure changes to the airway to assist inspiratory and expiratory muscle function.

There are three major goals of such intervention:

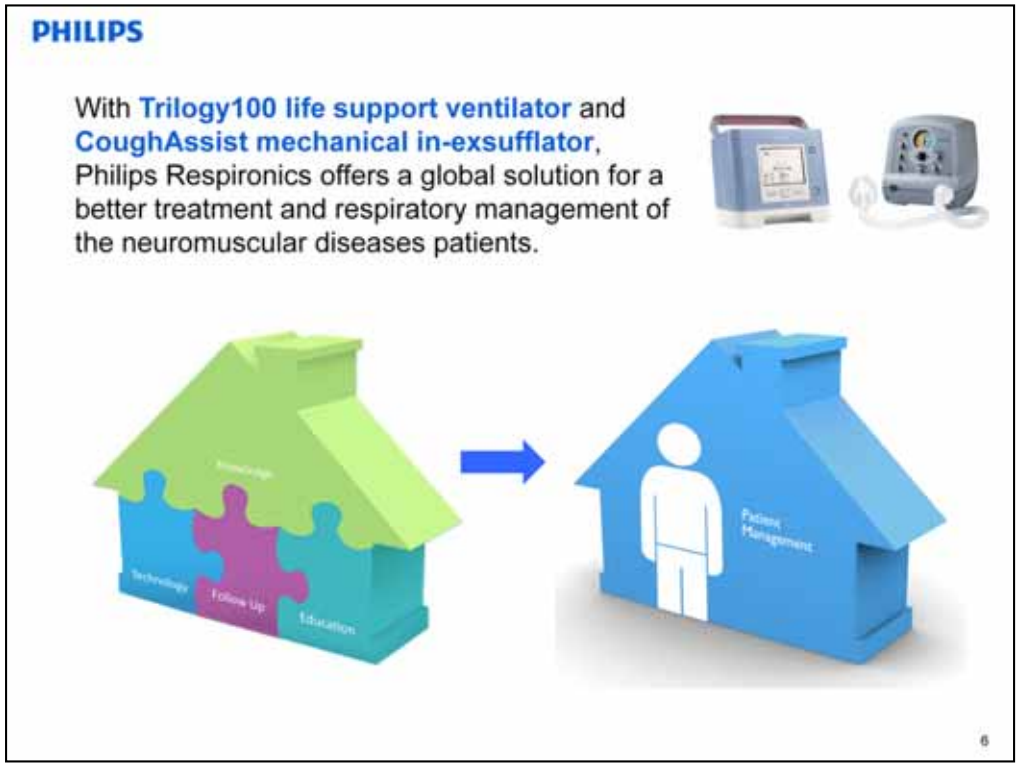
-To maintain lung and chest wall elasticity and to promote normal lung and chest wall growth for children

-To maintain normal alveolar ventilation

-To facilitate airway clearance



From the ATS consensus statement in 2004, we can see that not only the loss of respiratory muscle strength provokes hypoventilation, but also inefficient airway clearance, both phenomena leading to “pneumonia, atelectasis, and respiratory insufficiency in sleep and while awake in DMD”.



From the previous slides and quotes, we see that a global respiratory management has to be considered for the NMD patients.

With the Trilogy100 life support and mechanical in-exsufflator CoughAssist, Philips Respironics is able to offer a global solution for a better treatment and respiratory management of these patients.

We offer a solution built around the three items Technology, Follow Up and Education, along with the knowledge of the physician/clinician to effectively and efficiently manage their patients.

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Improve patient clinical condition	Prevent exacerbation and chest infections	Provide optimal therapy
Technology	**Follow Up**	**Education**
• High performance • Effective ventilation • Efficient airway secretion removal	• Monitor ventilation and airway secretion encumbrance • Assess efficacy & adjust therapy when necessary • Follow disease progression	• Properly use the equipment • Understand the technology

7

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Trilogy100

8

Trilogy presentation

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Technology *is critical for successful therapy*

"**Making the right choice is important**, since performance and patient comfort vary among devices. **Patient-ventilator synchrony is a key to the success** of long-term Home NIV as dysynchrony causes patient intolerance."

Performance Characteristics of 10 Home Mechanical Ventilation Pressure-Support Mode - Comparative Bench Study
Anne Battisti et al - Chest 2005

"The performance of the ventilators showed great variability, and depended upon the type of trigger (flow or pressure), type of circuit and patient profile.

Differences were observed between the preset and measured airway pressure and between the tidal volume measured by the ventilator and on the bench.

Leaks were associated with an inability to detect the patient's inspiratory effort or auto-triggering."

Performance of ventilators for non invasive positive-pressure ventilation in children
B. Fauroux et al - ERJ 2008

9

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Trilogy100 *features*

Unique features
for unique patients

Because everyone
is different.

- Compact & easy to use
- Auto-TRAK and AVAPS
  Respironics proven technology
- Non Invasive & invasive capabilities
- Volume & pressure modes
- For both children (> 5 kg) and adults
- Up to 8 hours battery capacity
- Weighs less than 5 kg

10

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Trilogy100 *features*

Removable air path
To meet high level disinfection standards

Battery
3-4hrs Detachable Lithium Ion battery
+3-4hrs Internal battery
External battery connector

Oxygen
O_2 valve port (up to 15lpm)

Remote Alarm / Nurse call connector

11

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Trilogy100 *features*

Display options
Limited patient use
Detailed clinical use

Data Management
Serial connector to PSG
1GB SD card

12

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Trilogy100 *configuration*

Delivery		Invasive & Non Invasive	
Application		Adults & Children	
Ventilation modes		Pressure	Volume
Therapy modes		CPAP S ST PC PC-SIMV	AC VC SIMV
Circuit configurations	Passive exhalation port 	Auto-Trak AVAPS ✓	Auto-Trak ✓
	Active exhalation valve 	✓	✓

13

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Respironics *technology*

Respironics well known technology in leak circuit configuration

Single limb circuit	Connects directly to a - mask with integrated leak - leak & a tracheotomy
	Only one tube - easy to connect - easier to clean - less noise - safer
Advanced leak compensation	In pressure & volume - Auto-Trak - AVAPS in pressure modes - Estimated Ve & Vte - Minimum PEP of 4 cmH_2O

14

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Trilogy100

technology summary

Respironics *proven technology*

Performance	Versatility	Simplicity
• Advanced leak compensation	• Configurable to meet patient ventilation needs	• Intuitive operation
• Auto-Trak algorithm	• Invasive & non invasive	• Patient friendly
• AVAPS hybrid ventilation	• Power options	• Easy to maintain
• DirectView management software	• Diagnostic interfacing	• Safe

15

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Enhanced *clinical assessment*

Polysomnographs

- Ventilation curves from the Trilogy100 in addition to the PSG signals
- Assess ventilation efficacy to adjust parameters and provide accurate diagnosis

DirectView

- Manage patient ventilation therapy

16

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DirectView *data management software*

Is my patient compliant?

1 yr compliance data

How does patient ventilation progress?

30 days daily trends

1 year long term trends

Look at trends first to then zoom in abnormal periods

72 hrs wave forms:

- Pressure
- Flow
- Leak
- Tidal Volume
- BPM

To get the values

1 yr Statistics

17

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CoughAssist

18

CoughAssist presentation

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Technology *is critical for successful therapy*

CoughAssist has been clinically proven to:

- Increase peak cough expiratory flow by more than fourfold

McCool DF, Nonpharmocologic airway clearance therapies: AACP evidence-based clinical practice guidelines. Chest. 2006; 129:250-259.

- Reduce recurrent respiratory infections in patients with respiratory weakness from neuromuscular disease

Chatwin M, Cough augmentation with mechanical insufflation/exsufflation in patients with neuromuscular weakness. Eur Respir J. 2003;21:502-508.

Miske LJ, Use of the mechanical in-exsufflator in pediatric patients with neuromuscular disease and impaired cough. Chest. 2004;125:1406-1412.

19

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Airway Secretion Clearance

Loosening

Devices that help loosening secretions in case of thick phlegm so that the patient **who can cough** can more easily clear his secretions

Assisted Cough

Replace a cough when **patients cannot cough** whether the secretions are thick or loose, it does not change the structure of the phlegm but just makes a cough

Mainly, **Neuromuscular Diseases** (Muscular Dystrophy (Duchenne), Myasthenia gravis, Poliomyelitis, Amyotrophic Lateral Sclerosis (ALS), Spinal Muscular Atrophy (SMA)) and **Spinal Cord Injuries**

MI-E is the only current therapy that mechanically assists the expulsive phase of coughing, which is critical in airway clearance.

MI-E is able to clear medium and small bronchi in addition to central airways and both the left and right airways.

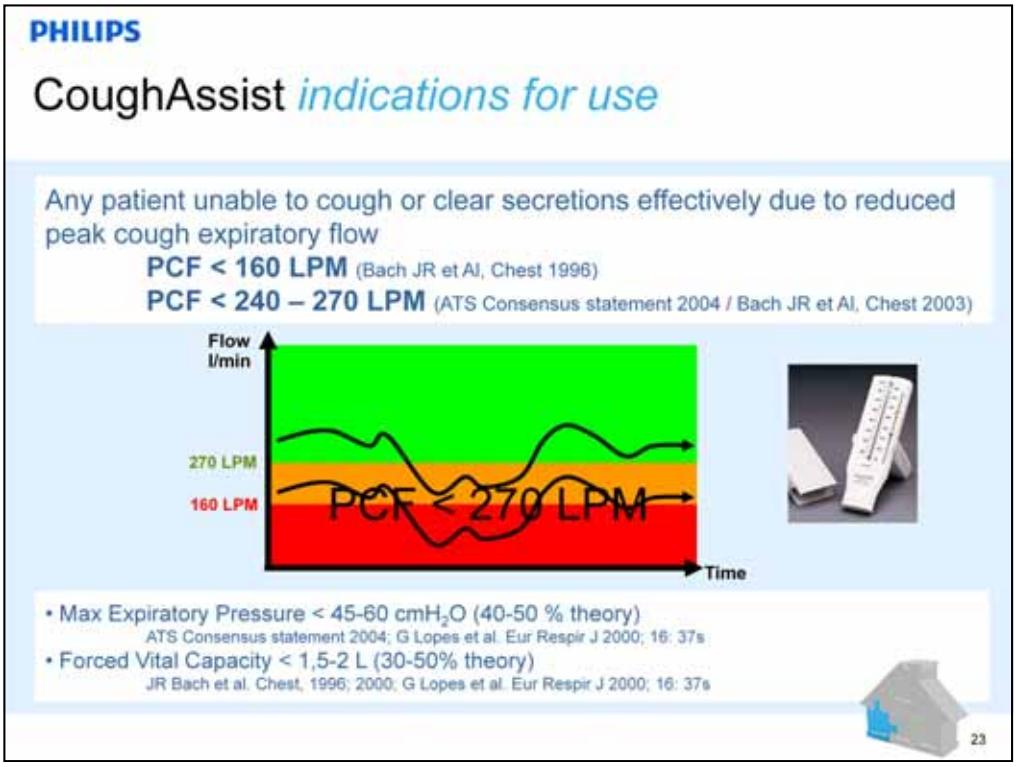
Inhale pressure control knob = varies the inspiratory pressure between 50-100% of the exhale pressure

Unequal pressures may be prescribed to maximize the exsufflation phase while minimizing the stretch to the intercostal muscles during the insufflation phase.

Inhale flow control knob = adjust to low (3.3 LPS) or high (10 LPS)

→set as per patient comfort

→lower setting may be preferable for patients with flaccid upper airways (i.e. ALS

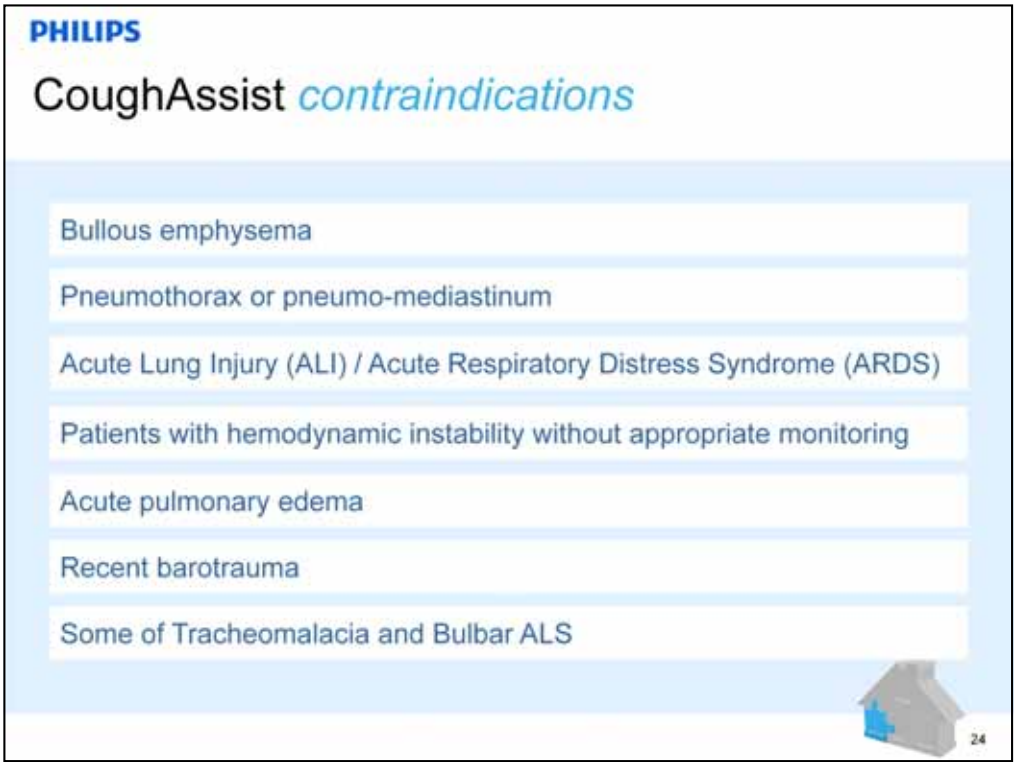


CA is to be used when inspiratory muscles and expiratory muscles do not provide an efficient cough and other techniques are inefficient, or balance action/fatigue is too high.

160 LPM is the minimum PCF value needed to clear airway secretions from central airways. In patients with unassisted PCF<160 LPM, mechanical assisted cough is required.

But a PCF threshold of 160 LPM do not guarantee adequate airway clearance in case of sudden respiration infection. Indeed, these patients are likely to suddenly deteriorate in case of acute viral illness (acute viral illness is associated with a reduction of vital capacity). In this case, PCF decreases below 160 LPM, the patient decompensates and has to be admitted to ICU.

This is why a PCF value of 270 LPM has been identified as a threshold for patients to be taught assisted cough techniques. In case of a sudden deterioration of the respiratory function, the patient stays above the critical threshold of 160 LPM.



-Bullous emphysema: emphysema associated with large bullae, type of COPD involving damage to the air sacs (alveoli) in the lungs (destruction of lung tissue).

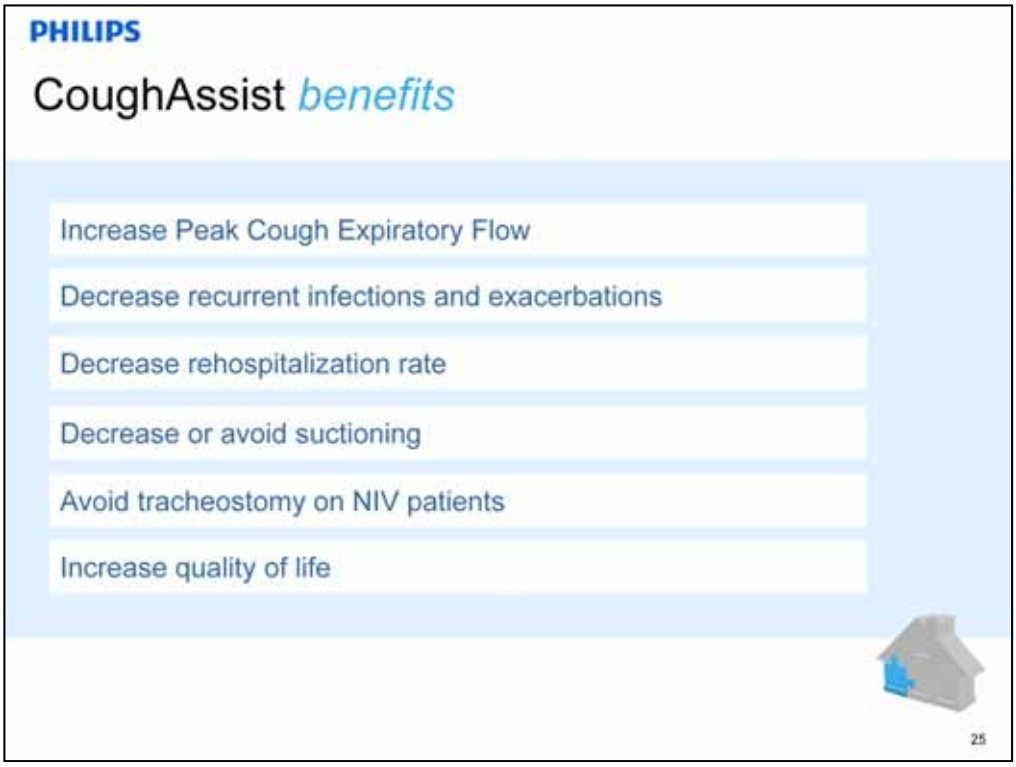
-Pneumothorax is caused by accumulation of air or gas in the pleural cavity, occurring as a result of disease or injury.

-Pneumomediastinum is a condition in which air is present in the mediastinum (structure of the thorax).

-Acute pulmonary edema is swelling and/or fluid accumulation in the lungs.

-Tracheomalacia: disease affecting the upper airways structure, becoming flaccid, soft – possibility to collapse if high negative pressure is applied.

-Bulbar patients cannot control their glottis, possibility of collapsing the upper airways



Endotracheal suctioning is a component of bronchial hygiene therapy and mechanical ventilation and involves the mechanical aspiration of pulmonary secretions from a patient with an artificial airway in place - **Invasive procedure, very uncomfortable for the patient** - Removes secretions but does not loosen them.

Suctioning (Invasive Secretion Clearance) may:

-Damage tracheal mucosa (tracheal bleeding), impair ciliary function or destroy cilia

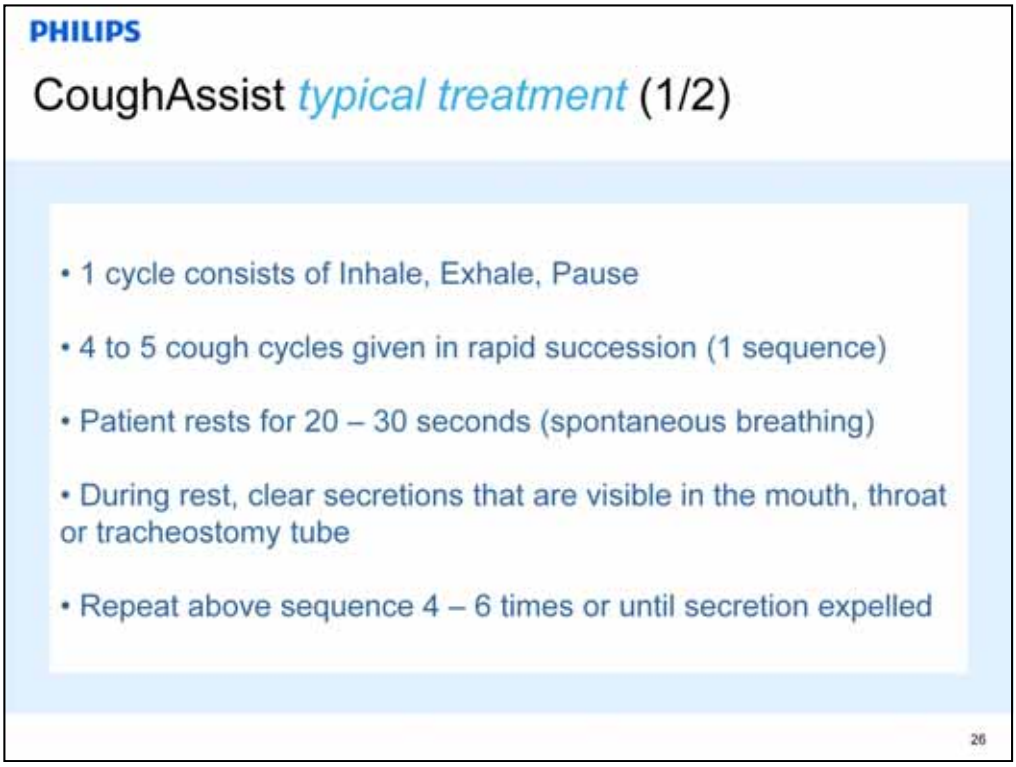
-Clear left lung poorly due to angle of left bronchus

-Increase production of secretions

-Exert vagal response and decrease heart rate

-Increase intracranial pressure

-Cause hypoxemia



A CA cycle is made up of an inhalation phase, an exhalation phase and a pause phase.

One treatment consists of about 5 cycles of MI-E or MAC (Mechanical Assisted Cough, i.e. use of the CA in combination with abdominal thrust with the exsufflation) followed by a short period of normal breathing or ventilator use to avoid hyperventilation.

When delivered via a tracheostomy tube, the cuff if present should be inflated. Briefly deflate cuff to obtain secretion from above.

1. Inform patient about CoughAssist prior to use
2. Have the head of the patient on a support and patient a little bit leant backwards to open the chest and thoracic cage
3. If face mask, apply it securely
4. Adjust mask to eliminate leak
5. Coordinate with patient breathing pattern

For the first session, it is important to start with low pressures, non therapeutic pressures.

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CoughAssist *typical treatment* (2/2)

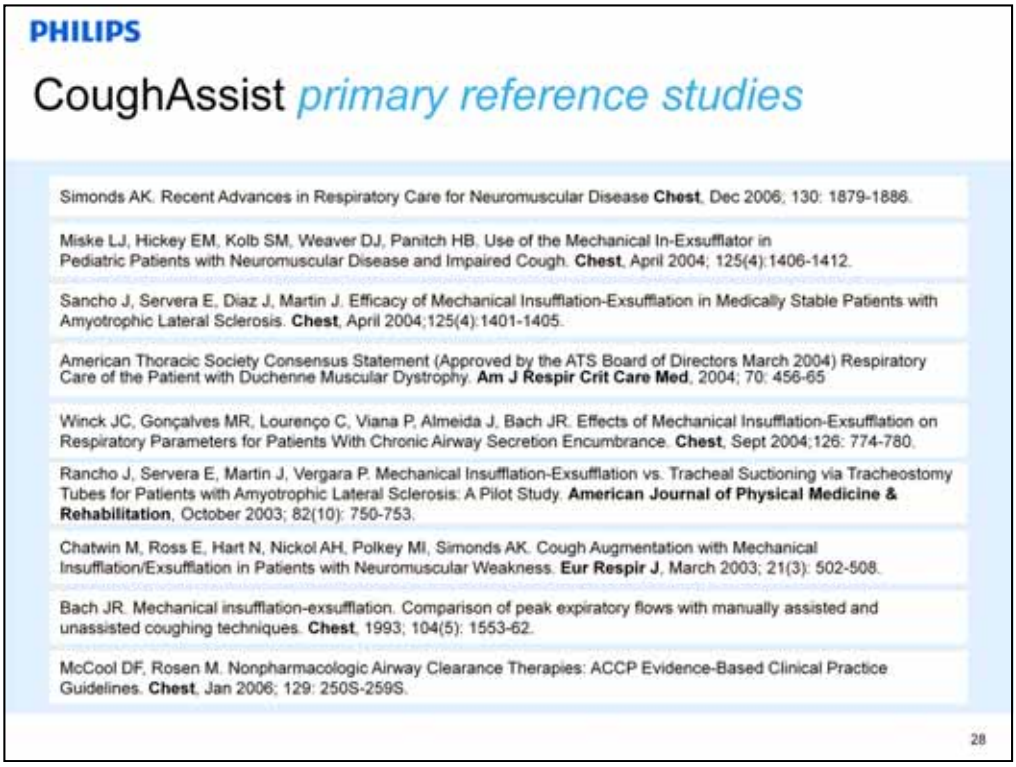
- Possibility to use abdominal and chest compression during expiration
- Typical pressure settings: +40cmH_2O/-40cmH_2O, but to be adjusted to the individual patient's needs
- Frequency of sessions according to case history
- Best before meals and at bedtime
- Home pulse oximetry is useful to monitor the effectiveness of airway clearance

27

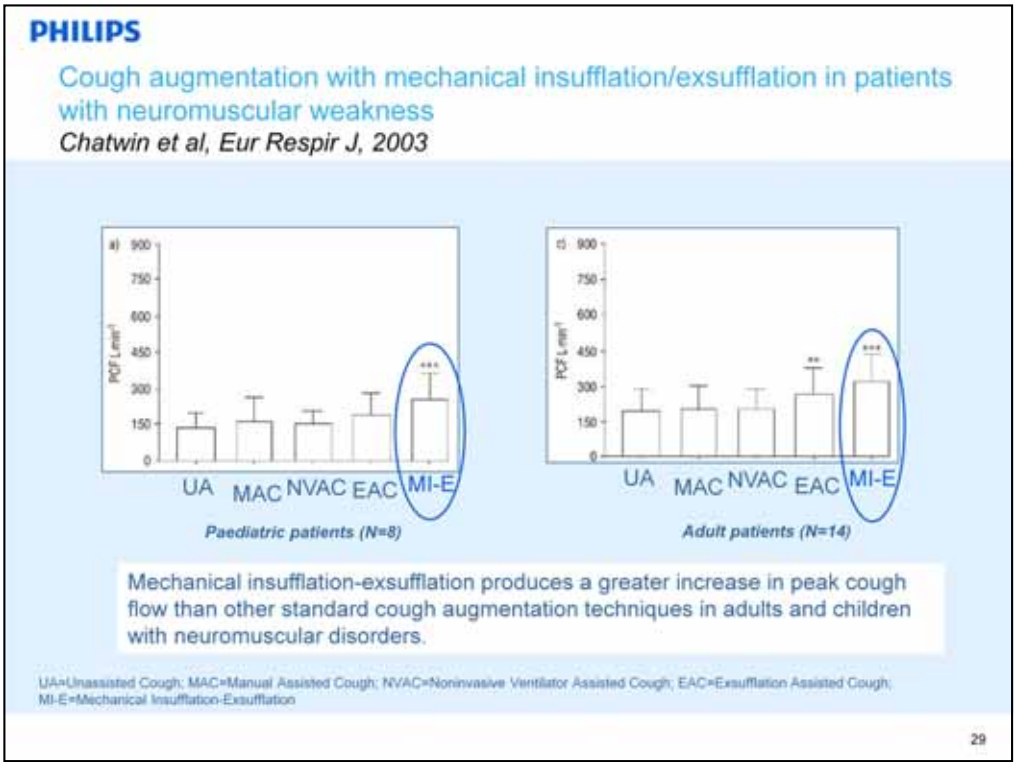
Unless contraindicated, an assisted cough maneuver will usually increase the PCF by 20%. The assisted cough maneuver is applied when lungs are full at the onset of the CA negative pressure.

Pressures below 40 cmH2O may be inefficient,

The use of the home pulse oximetry to monitor the effectiveness of airway clearance is a recommendation of the ATS, from the consensus statement from 2004 about the Respiratory Care of the Patient with Duchenne Muscular Dystrophy.



There are lots of clinical studies published about CoughAssist, mainly about NMD, both adults and pediatrics.



This study by Chatwin and coworkers has been very important in the development of the knowledge of the CoughAssist technique in Europe.

It compares different techniques of cough augmentation in patients with neuromuscular weakness, the studied group comprising some adults (31yrs old +/- 13)and some pediatric (14yrs old +/- 2) patients.

Twenty-two patients aged 10–56 yrs (median 21 yrs) with neuromuscular disease and 19 age-matched controls were studied. Spirometry was performed and respiratory muscle strength measured. Peak cough flow was recorded during maximal unassisted coughs, followed in random order by coughs assisted by physiotherapy, noninvasive ventilation, insufflation and exsufflation, and exsufflation alone. Subjects rated strength

of cough, distress and comfort on a visual analogue scale.

- UA = unassisted cough

- MAC = standard physiotherapy assisted cough

- NVAC = cough after inspiration supported by a noninvasive positive pressure ventilator

- EAC = mechanical exsufflation-assisted cough with delivery of negative pressure initiated manually at the end of inspiration

- MI-E = use of the CoughAssist

The greatest increase in peak cough flow was observed with mechanical

insufflation/exsufflation at 235+/-111 LPM. All techniques showed similar patient acceptability. Mechanical insufflation/exsufflation produces a greater increase in peak cough flow than other standard cough augmentation techniques in adults and children with neuromuscular disease.

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Philips Respironics *support*

Expertise

- Clinical specialists & training programs
- Supporting materials & media to
  - Train clinical staff/technicians on the technology (and follow up software for Trilogy100)
  - Support you with the technology in your daily work

Workshops & Symposium

- Learn & share best practice/research

30

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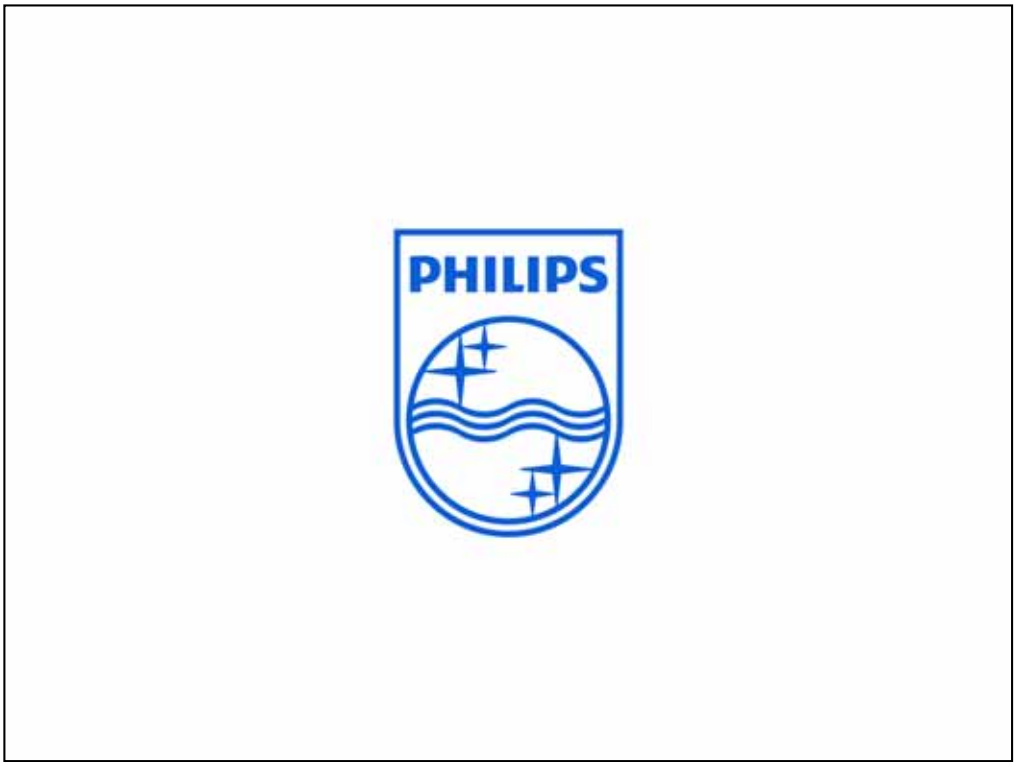
Conclusion *Trilogy100/CoughAssist*

- Full Life Support ventilator featuring Philips Respironics technology
- NIV and IV, Pressure and Volume, for adults and children (> 5 Kgs)

The Smart Combination for the Respiratory Management of the Neuromuscular Disease Patients

- Non invasive and flexible technique
- For adults and pediatrics

31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