Pulmonary rehabilitation in neuromuscular disease

Han Eol Cho

Department of Rehabilitation Medicine, Gangnam Severance Hospital

Rehabilitation Institute of Neuromuscular Disease, Yonsei University College of Medicine

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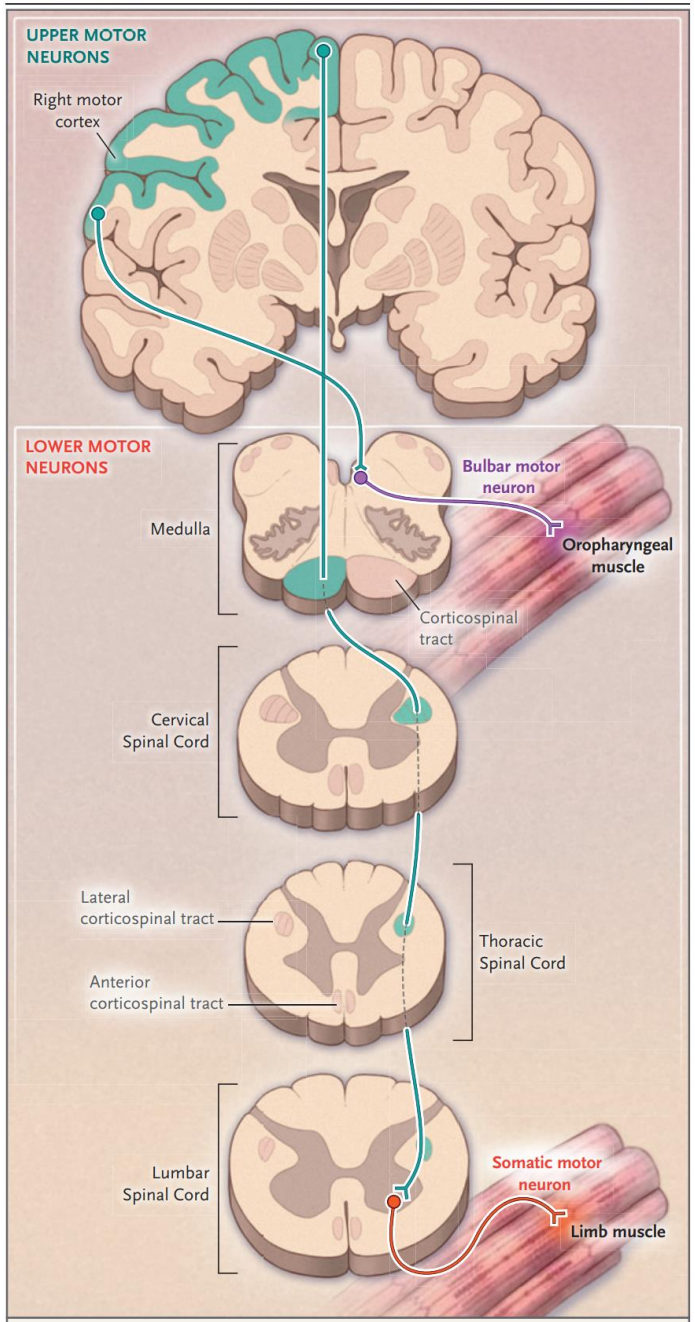
What is neuromuscular diseases?

What is neuromuscular disease?



What is neuromuscular disease?

Neuromuscular disease

are diseases of

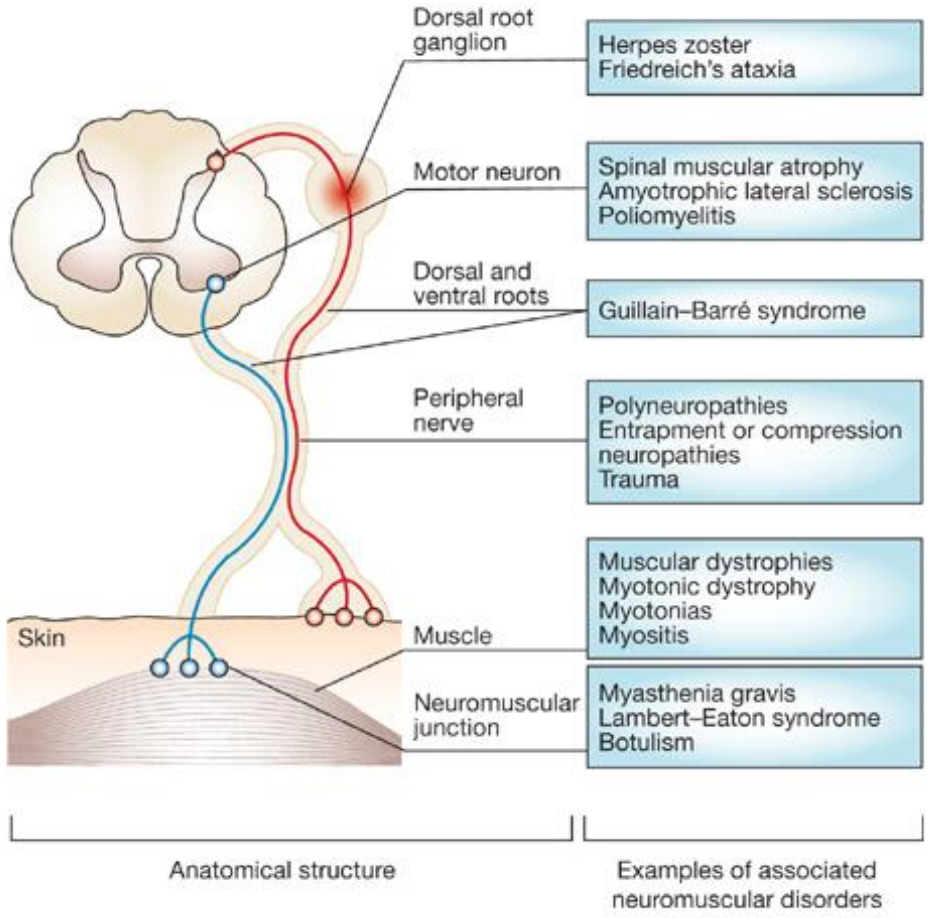
the peripheral nerves, Root, AHC

(neuropathies, GBS, AHC diseases)

the myoneural junctions(myasthenia gravis),

or

the muscles (myopathies) themselves



What is pulmonary rehabilitation?

Pulmonary Rehabilitation

Pulmonary rehabilitation is a comprehensive program designed to help people with lung diseases, such as chronic obstructive pulmonary disease (COPD), asthma, and pulmonary fibrosis, to improve their respiratory function, reduce symptoms, and enhance their quality of life.

Pulmonary rehabilitation typically involves a combination of exercise training, breathing exercises, education on lung disease management, and psychological support. The program is tailored to the individual's specific needs and can be done in both inpatient and outpatient settings.

The exercise component of pulmonary rehabilitation typically involves aerobic exercise, strength training, and flexibility exercises. Breathing exercises, such as pursed lip breathing and diaphragmatic breathing, can help improve breathing patterns and reduce shortness of breath.

Education on lung disease management includes topics such as medication management, oxygen therapy, and nutrition. Psychological support may include counseling and stress management techniques to help individuals cope with the emotional and social impact of their lung disease.

Pulmonary rehabilitation has been shown to improve exercise capacity, reduce symptoms, and enhance quality of life for people with lung disease. It is typically recommended as a part of comprehensive treatment for people with moderate to severe lung disease.

Pulmonary Rehabilitation is different according to pulmonary disease

Obstructive disorders

- Reduction in airflow
- Significant ventilation-perfusion mismatching
- Primarily in impaired oxygenation of the blood

- COPD, Asthma, Bronchiectasis, …

Restrictive disorders

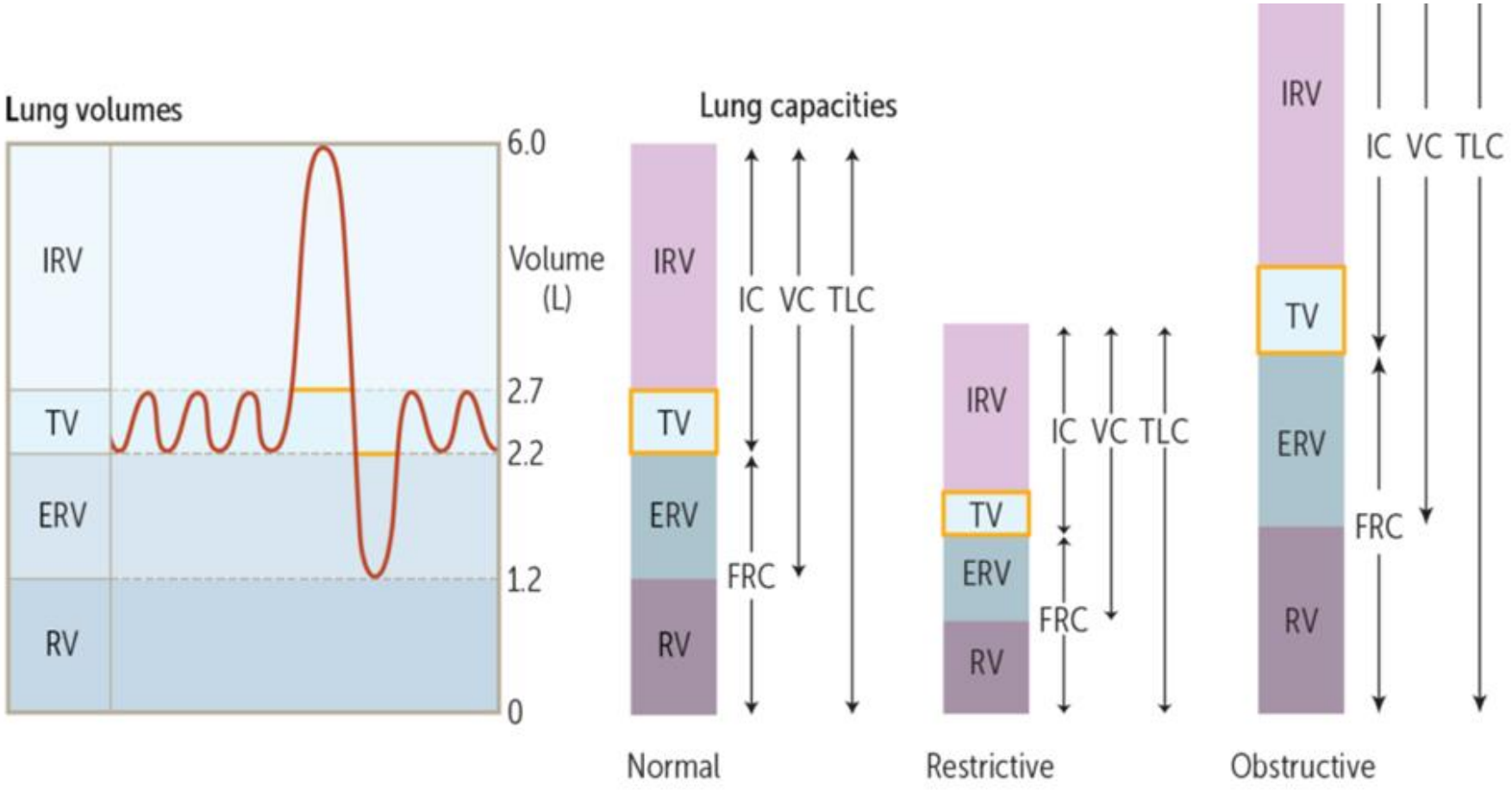
- Reduction in lung volume
- Respiratory muscle dysfunction
- Impaired lung ventilation
- Primarily hypercapnia

- Neuromuscular disorders, Kyphoscoliosis, Interstitial lung disease, Marked obesity, …

What kind of pulmonary rehabilitation does NMD patients needed?

Feature of pulmonary function of NMD

Feature of pulmonary function of NMD



IRV = inspiratory reserve volume FRC = functional residual capacity

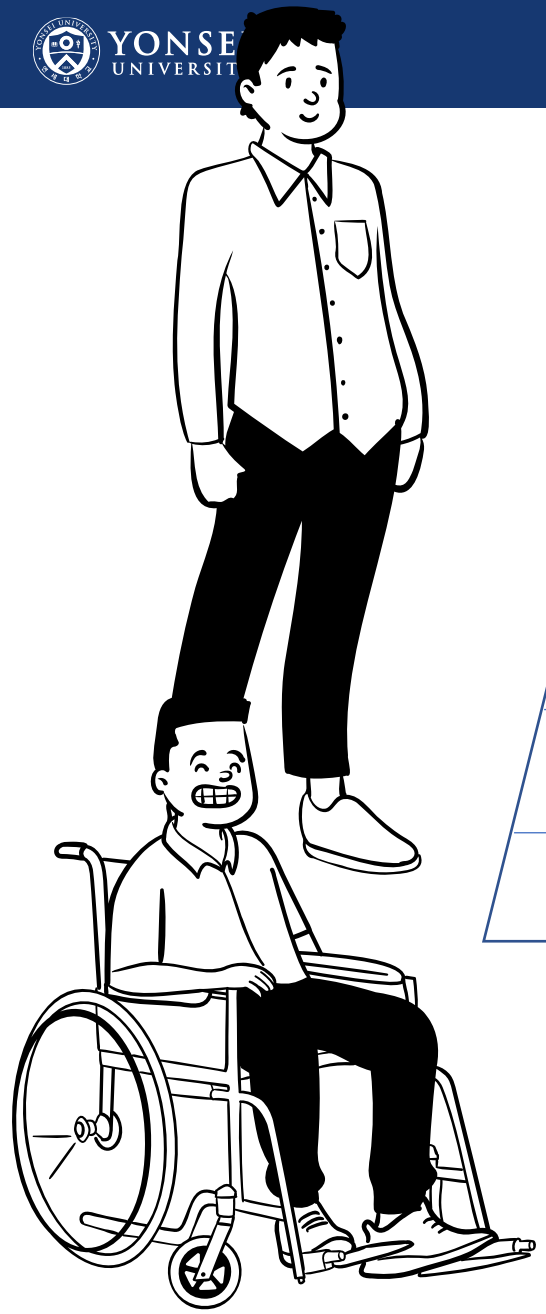
Summary of lung function of NMD

- Neuromuscular disease
  - → Muscle weakness
  - → Weakness in both inspiration and expiration
  - → Assistance required

- Inspiration assist : Mechanical ventilator
- Expiration assist: Assistive coughing methods / Cough machine

Comparison of use of ventilator

	Internal medicine patients	Pulmonary rehabilitation patients
Type of disease	ARDS, pneumonia, lung cancer, ….	Neuromuscular disorders (ALS, DMD, SMA, GBS…), Cervical cord injury….
Problematic areas (treatment targets)	Lung	Muscle
Reasons to use ventilator	Assist ventilation until lung disease is treated (Recovery)	Assist with lack of muscle function (Support)
Treatment goal	Wean ventilator	Avoid exhausting respiratory muscles
Duration of ventilator use	Short term	Long term



Pulmonary rehabilitation of Neuromuscular disease

Patient Evaluation

Vital capacity (VC)

- VC in sitting, supine, with braces on and off
- Diaphragmatic weakness : VC sit >> VC sup

Peak cough flow (PCF)

Peak Cough Flow

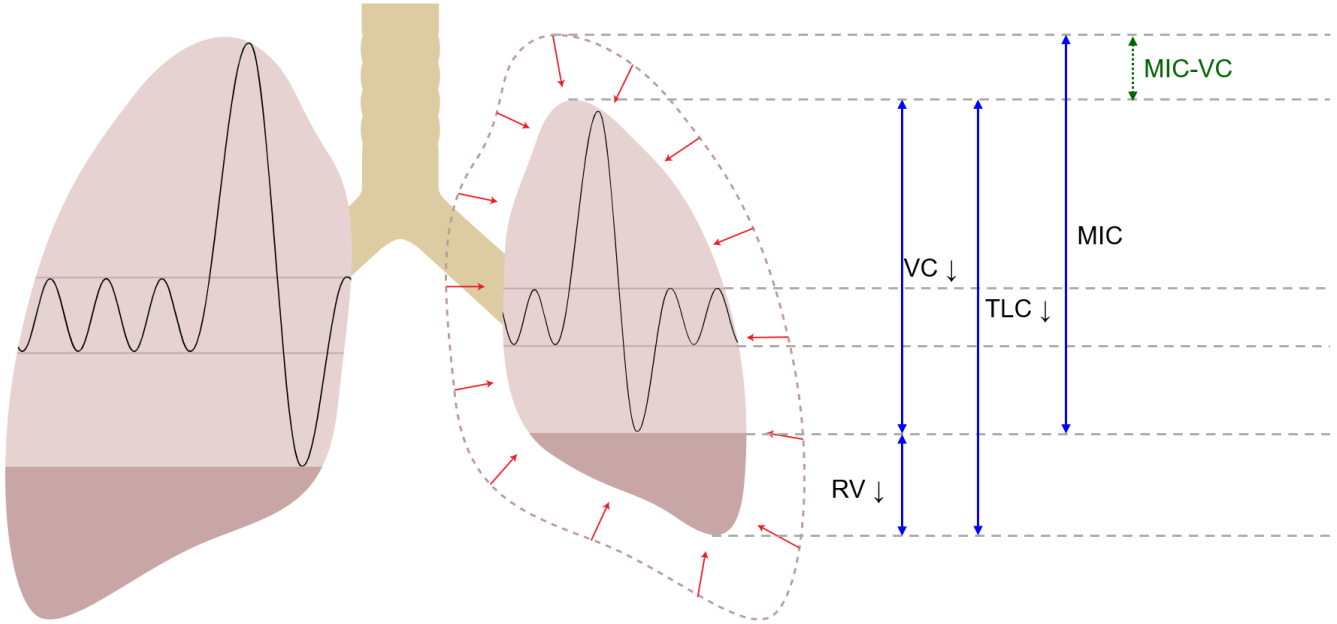
1. Take a deep breath
2. Cough (Without any assist)

- Ability to eliminate secretions
- Normal cough : precough inspiration to 85-90 % of TLC
- VC < 1500 ml - insufflate before assisting a cough
- Normal : 360 - 720 L/min (6 – 12 L/sec)

Maximal respiratory pressure

- Maximum inspiratory and expiratory pressure (MIP and MEP)
- Evaluation for respiratory muscle weakness
- Every 6-12 months interval
- Every 1- 3 months in rapid progressive diseases (ALS)

Maximal insufflation capacity



- Maximum volume of air that can be mechanically delivered to a patient's lung
- None of us can cough with lung empty.
- The greater the MIC, the greater the patient's ability to cough.

Nocturnal CO_2

- The most important value in patients with restrictive lung disease (RLD)
- The most sensitive test to determine if there are respiratory problems in RLD patients.

Indications of Nocturnal monitoring

- Symptoms of lung underventilation
- Supine VC much less than sitting VC
- Two or more pillows for sleep
- VC is under 40% of the predicted normal
- Daytime end-tidal CO2 levels > 44 mmHg
- Daytime SaO2 < 95%
- Frequent nocturnal arousals for small children

Inspiratory muscle assistance

INSPIRATORY RESISTIVE EXERCISE

- Increase exercise capacity by reconditioning the inspiratory muscles

- Several devices are commercially available

- Typical training
- Begin at 25 % to 35 % of the maximal pressure
- Duration : 15 to 30 minutes
- Frequency : twice a day
- Complete two 15 to 30 minute sessions a day
- - increase the resistance

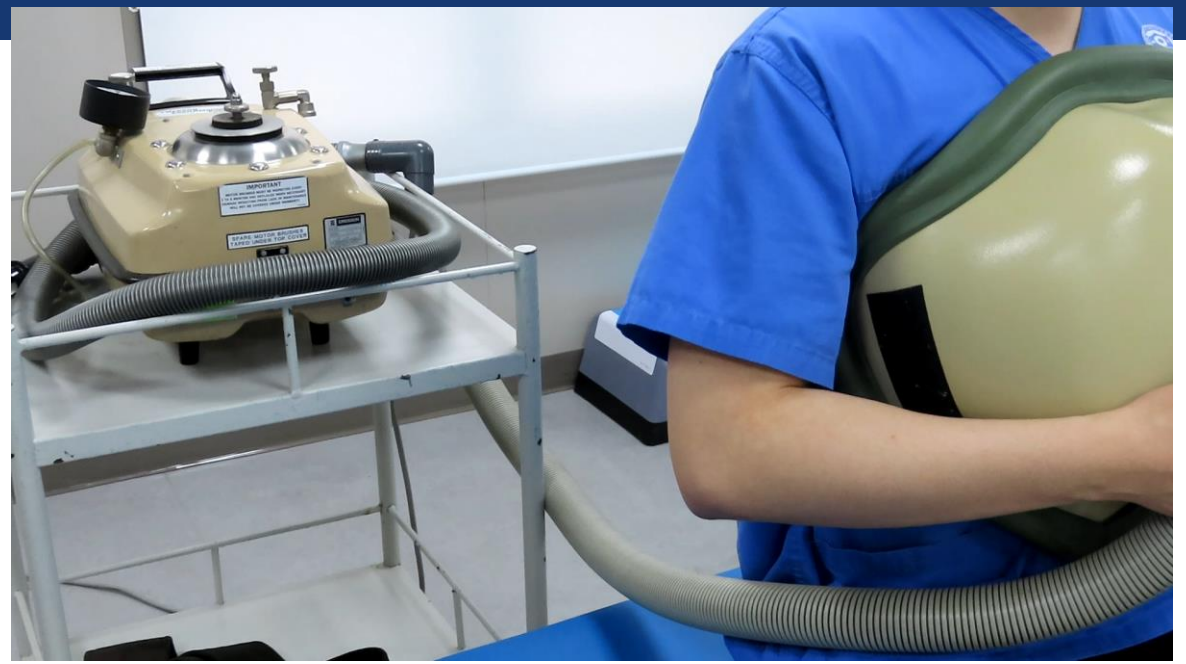
Neuromuscular disease (NMD) patients often experience significant muscle weakness, which makes resistive training inappropriate for them

=> Mechanical ventilator!

Various types of ventilators

- **Body ventilators**
  - Negative body ventilators
    - Iron Lung, Porta-lung, Chest shell
  - Positive body ventilators
    - Intermittent abdominal pressure ventilator(pneumobelt)
  - Rocking Bed
    - Head up (negative)/ down (positive)
- **Positive pressure ventilator**

Body ventilator




PLV-100

Ventilator의 종류

- Body ventilators

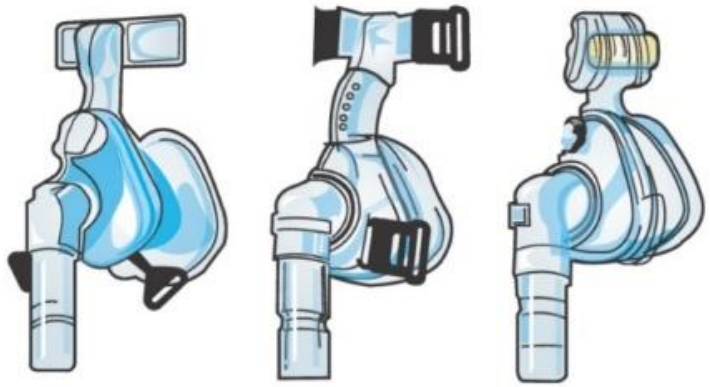
- **Positive pressure ventilator**
- Volume-limited ventilator (LTV series, CARAT, LEGENDAIR, PB560, Astral and Trilogy)
- Pressure- limited ventilator (BiPAP)

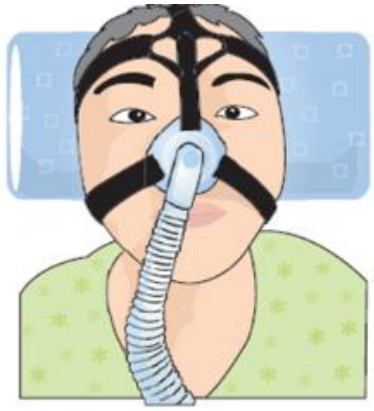
How to use ventilator?

Noninvasive ventilation VS **Invasive ventilation**

Mask Interface

- Nasal interface; nasal mask, nasal pillows
- Face masks: Oronasal masks, total full-face masks
- Hybrid
- Mouthpiece /lipseal



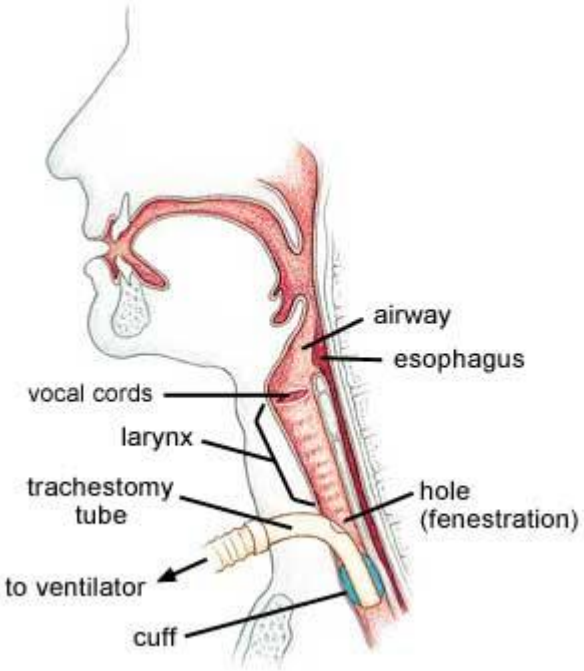


AF531 with CapStrap

Mouthpiece

- when awake is a reasonable option
- An angled mouthpiece or straw-type mouthpiece
- supported by a flexible arm placed near the mouth so that patients can access it with the lips and mouth as desired

Disadvantage of Tracheostomy

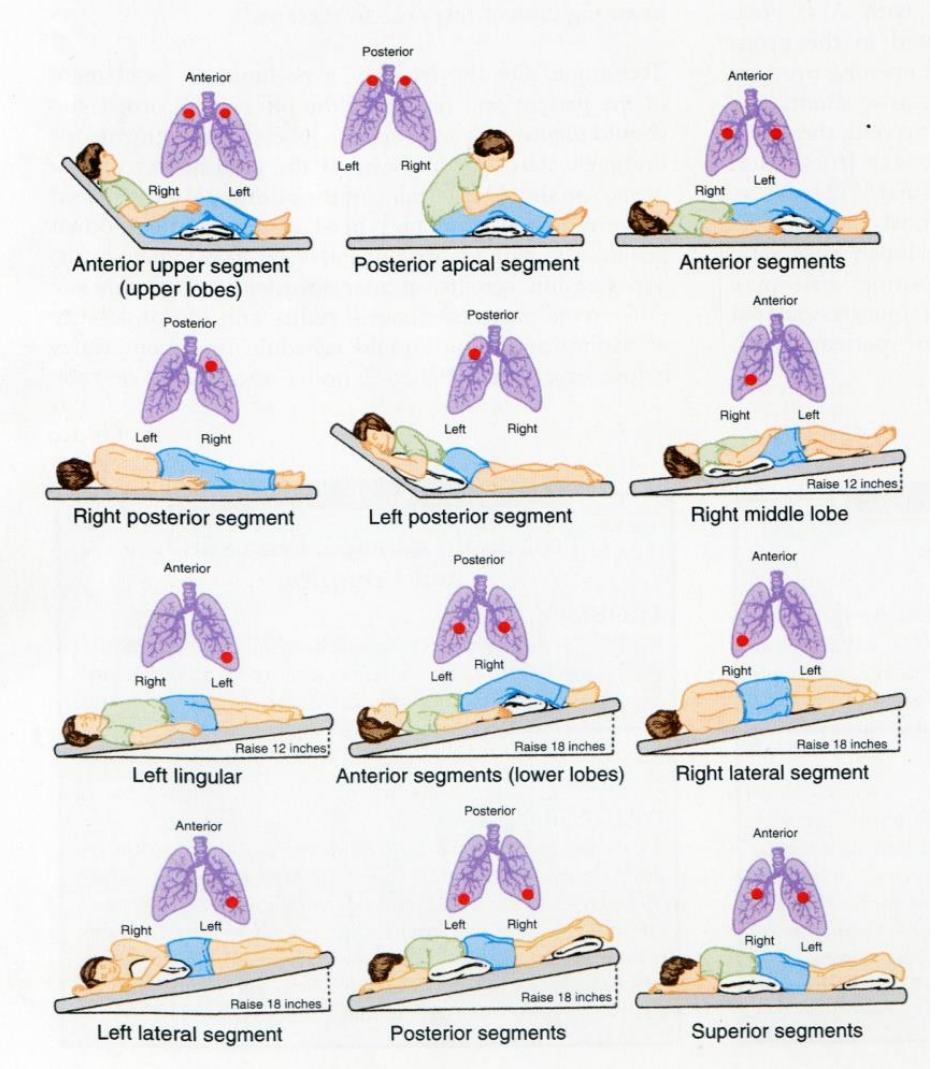


- Profuse secretions
- Speaking or eating problem
- Pathogenic bacterial colonization
- Direct access of bacteria
- Granulation tissue

Airway secretion management

Postural Drainage

- Use gravity to help move airway secretion
- Changing position to make each lung segment's airway is aligned perpendicular to the central airway.
- Mucus must be diluted by systemic hydration and airway hydration.



Percussion and Vibration

Increase the effect of sputum excretion

Percussion - to release sputum from the bronchi

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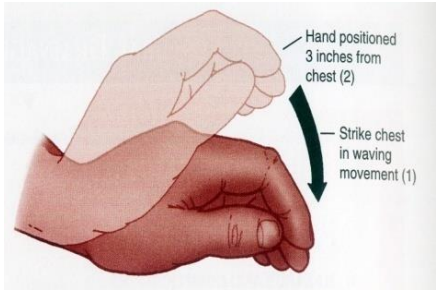
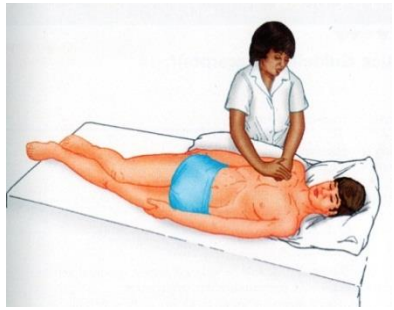




Vibration – movement of sputum into the central airway; applied when exhaling

High-Frequency Chest Wall Oscillation

- Composition

(1) an air vibration generator

(2) an inflatable vest

- The high-speed vibration of the chest wall is caused by the air vibration generator.

- Treatment for 10 to 30 minutes at a frequency of 5-25 vibrations per second

Cough assistance

Cough

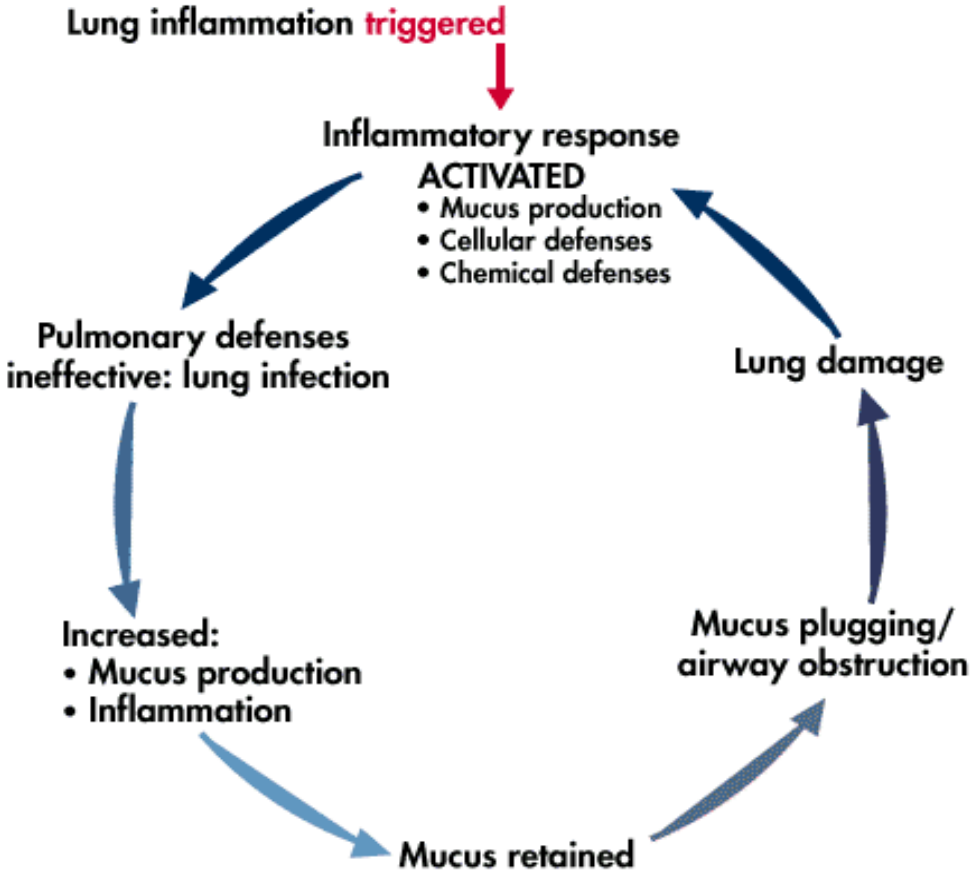
- helps protect your airway and lungs against irritants
- Normal PCF : 360 - 720 L/min (6 – 12 L/sec)

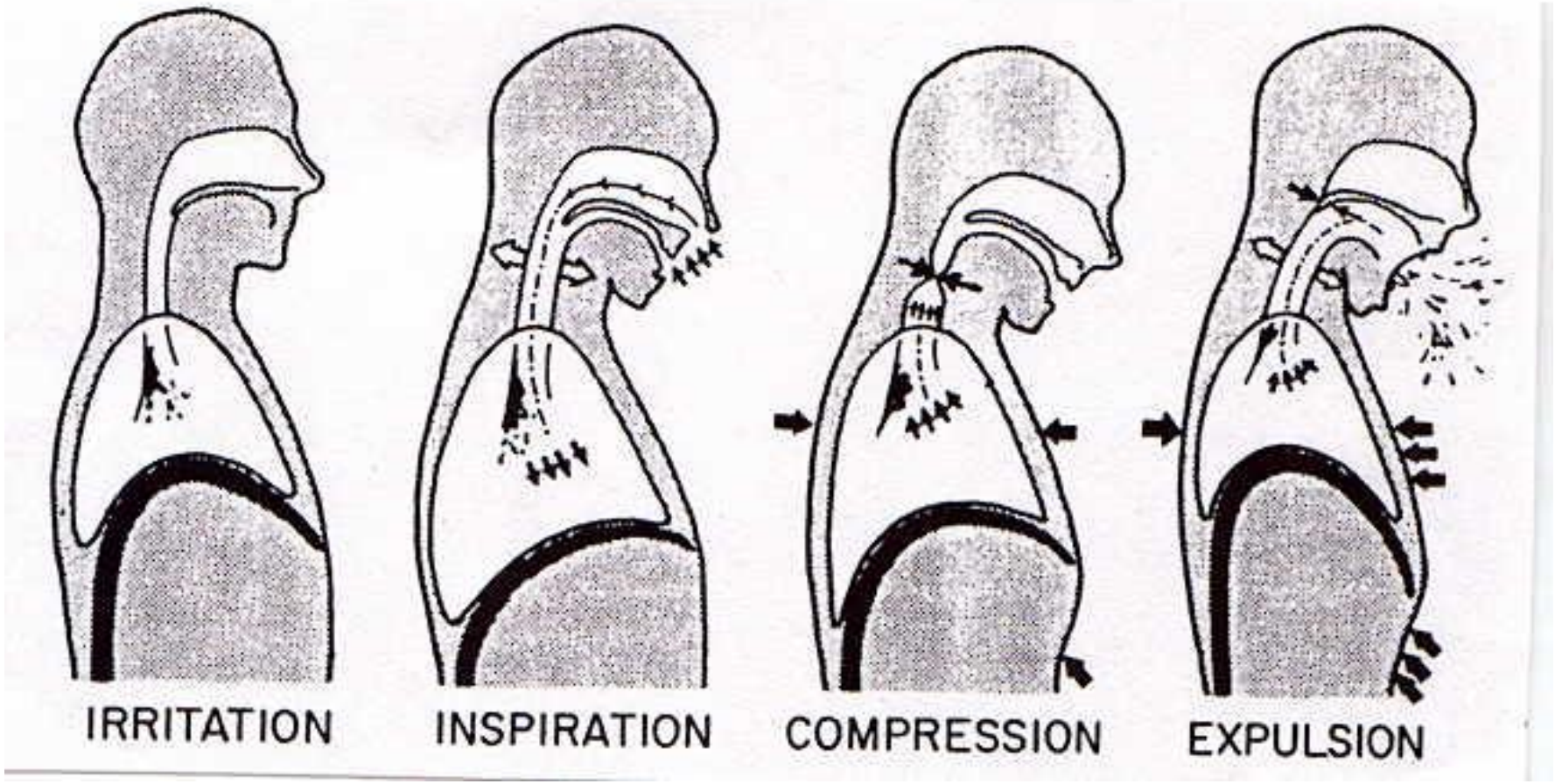
✓ Assisted PCF < 160 L/min
- severe throat muscle weakness
- scarring of the voice box and vocal cords
- narrowing of the airway
- severe coincident airways disease (asthma or severe COPD)

✓ Assisted PCF below 250 L/min
- high risk during colds
- colds can decrease APCF below 160 L/min

AIRWAY SECRETION CLEARANCE

- Vicious Cycle of Poor Airway Clearance




IRRITATION
INSPIRATION
COMPRESSION
EXPULSION

Manually assisted cough

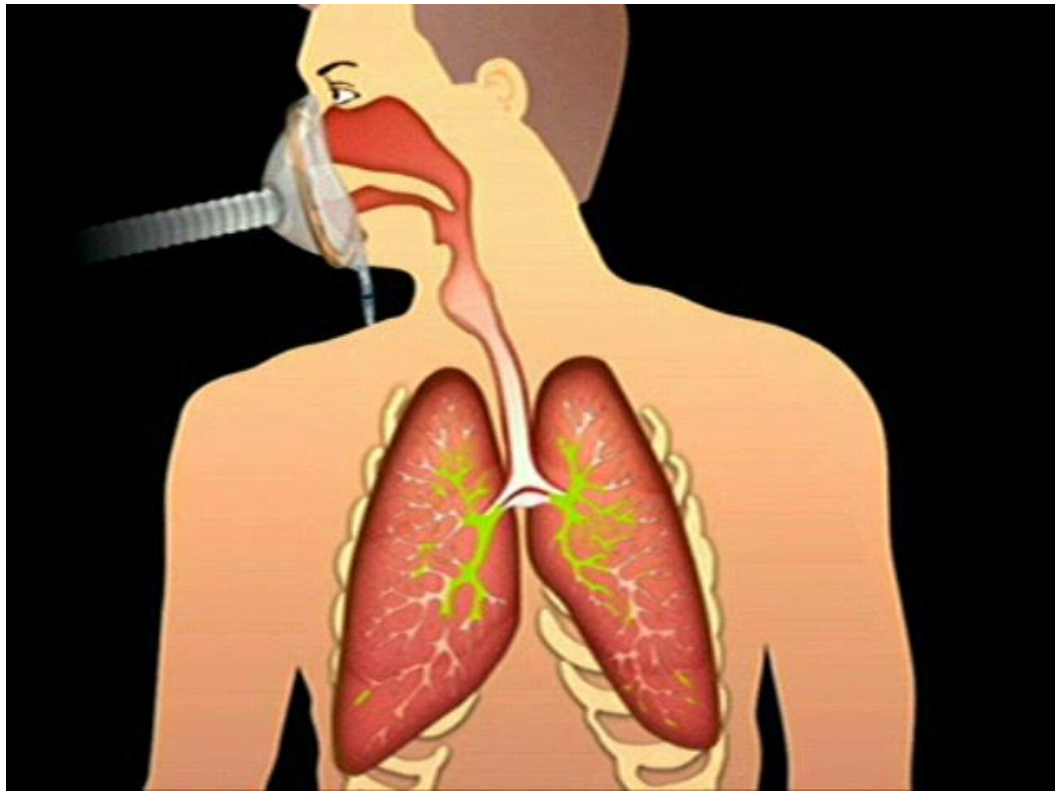
Manually assisted cough

Full assisted Peak Cough Flow

1. Take a deep breath
2. Airstacking with ambu bag
   (Maximal Insufflation Capacity)
3. Cough with abdominal thrust

Mechanical In-Exsufflator

- Especially useful for patients with neuromuscular diseases.
- Principle similar to vacuum cleaner (using suction)
- After injecting enough air by applying positive pressure to the lungs, instantaneous negative pressure is applied alternately to generate strong expiratory force.
- The sputum can be easily removed through the mouth-nose mask without a tracheostomy tube.
- More effective than suction using a suction tube even in patients with a tracheostomy tube



Mechanical In-Exsufflator

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COLLEGE OF MEDICINE