



**IPRC**  
International  
Pediatric Rehabilitation  
Collaborative

# Problem-solving Respiratory Issues in Children With Neuromuscular Disease

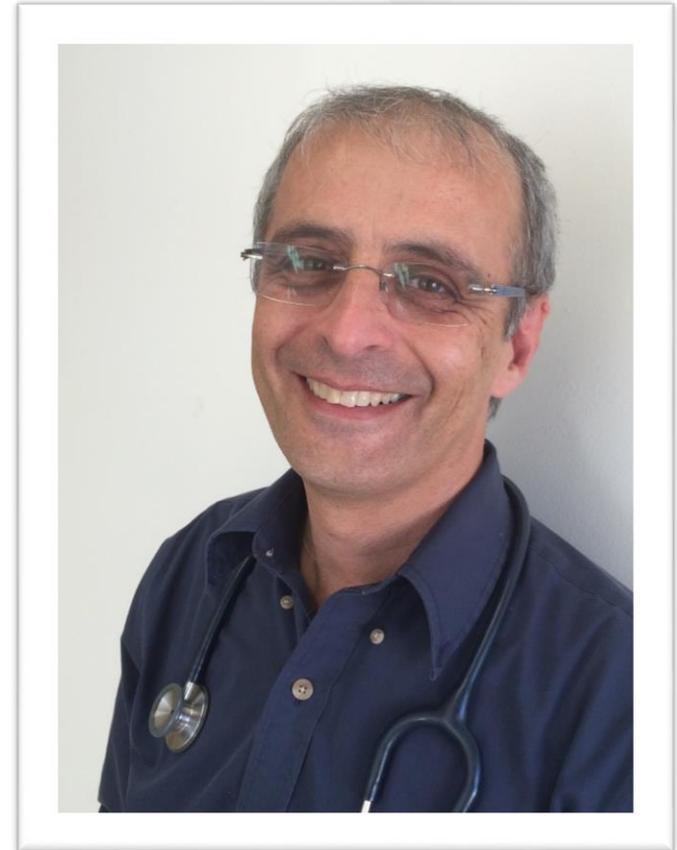
December 13, 2018  
Eliezer Be'eri, M.D.



# About Our Presenter



Eliezer Be'eri, M.D.  
Alyn Rehabilitation Hospital  
Jerusalem, Israel  
[ebeerer@alyn.org](mailto:ebeerer@alyn.org)



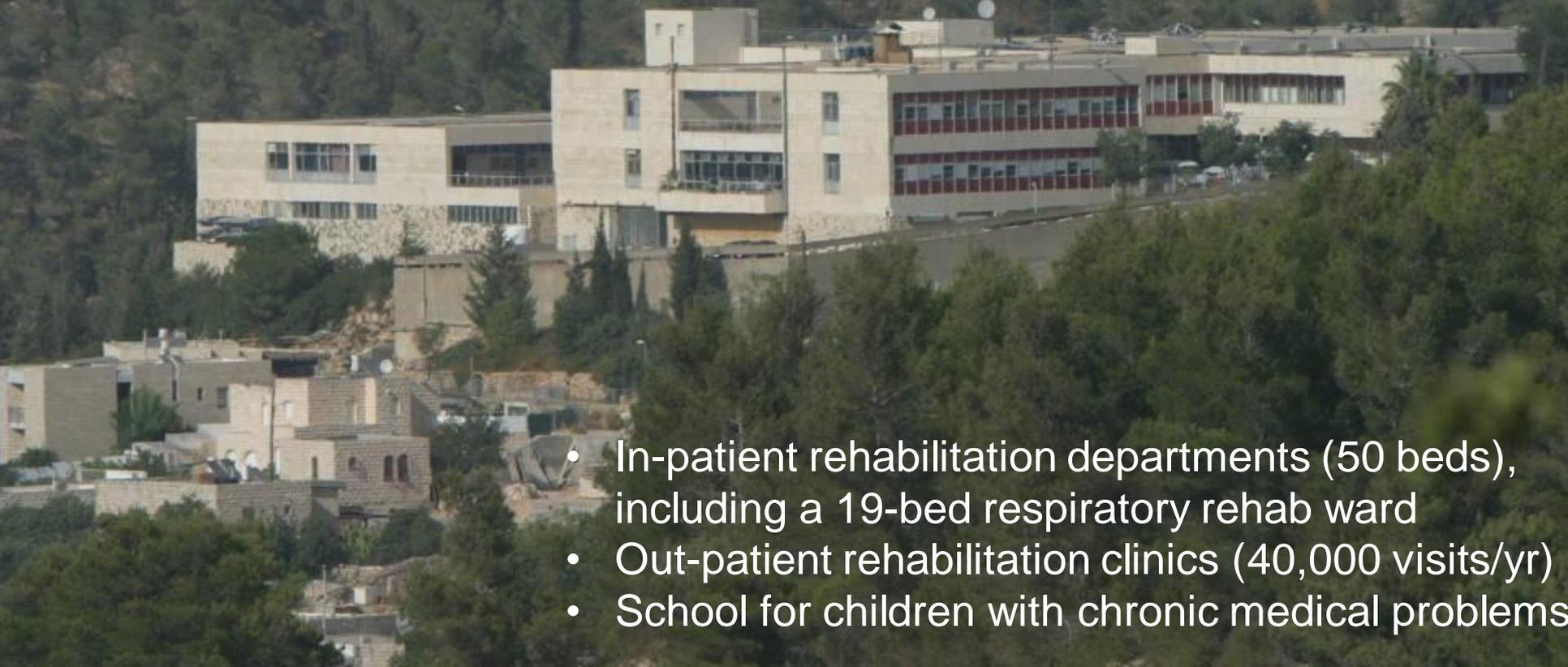
# Objectives



- Appreciate the major psychosocial issues that can undermine successful rehabilitation of a child on mechanical ventilation, if not identified and addressed at the outset of the rehab program,
- Identify the pathophysiology of neuromuscular respiratory failure and how that influences strategies for ongoing mechanical ventilation and for mechanical ventilation weaning,
- Discuss the indications, contraindications, benefits, and drawbacks of noninvasive ventilation versus tracheostomy ventilation,
- Learn strategies to optimize speech in the ventilated patient, and
- Understand strategies to diagnose and manage respiratory problems in the child with Cerebral Palsy.

# Shalika Pediatric Rehabilitation Hospital

Jerusalem, Israel



- In-patient rehabilitation departments (50 beds), including a 19-bed respiratory rehab ward
- Out-patient rehabilitation clinics (40,000 visits/yr)
- School for children with chronic medical problems

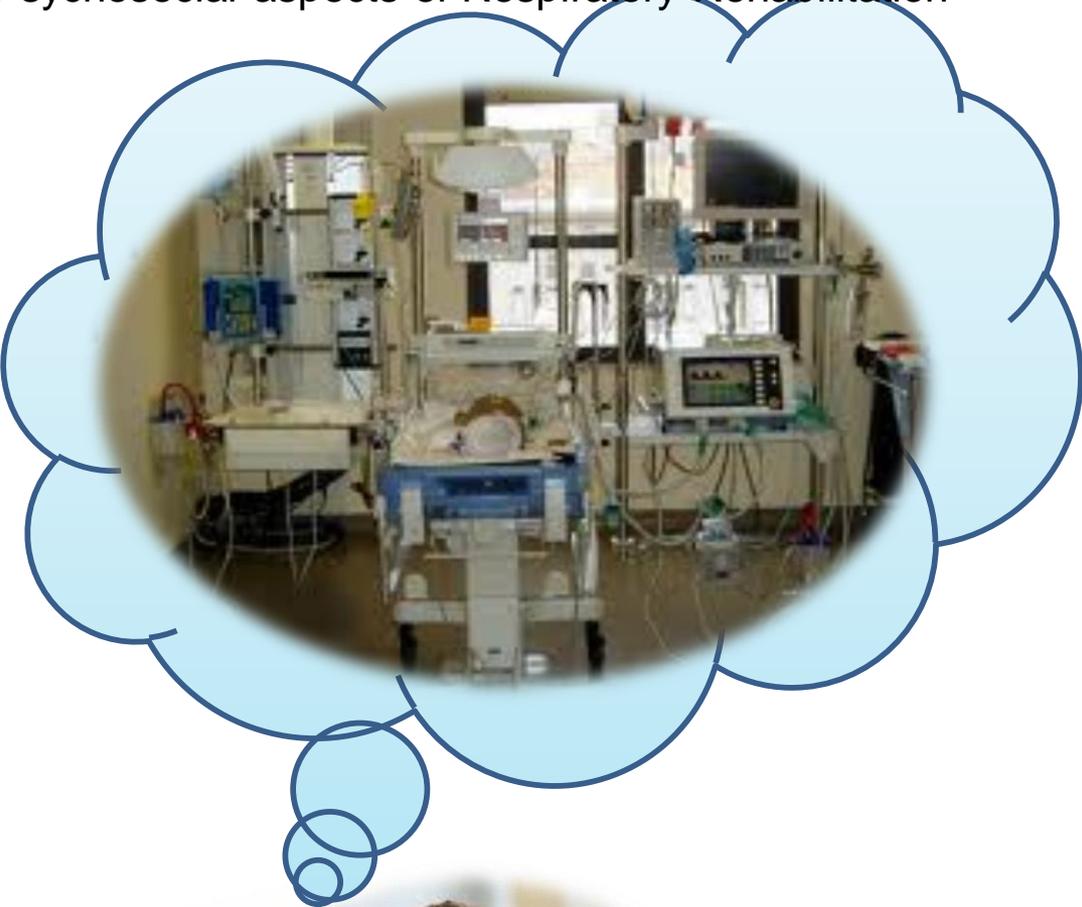


# Psychosocial Aspects of Respiratory Rehabilitation



The biggest barrier to discharge home and participation in society of ventilated children:

## Parental Psychological Paralysis



## Enabling Participation

A picnic at the zoo

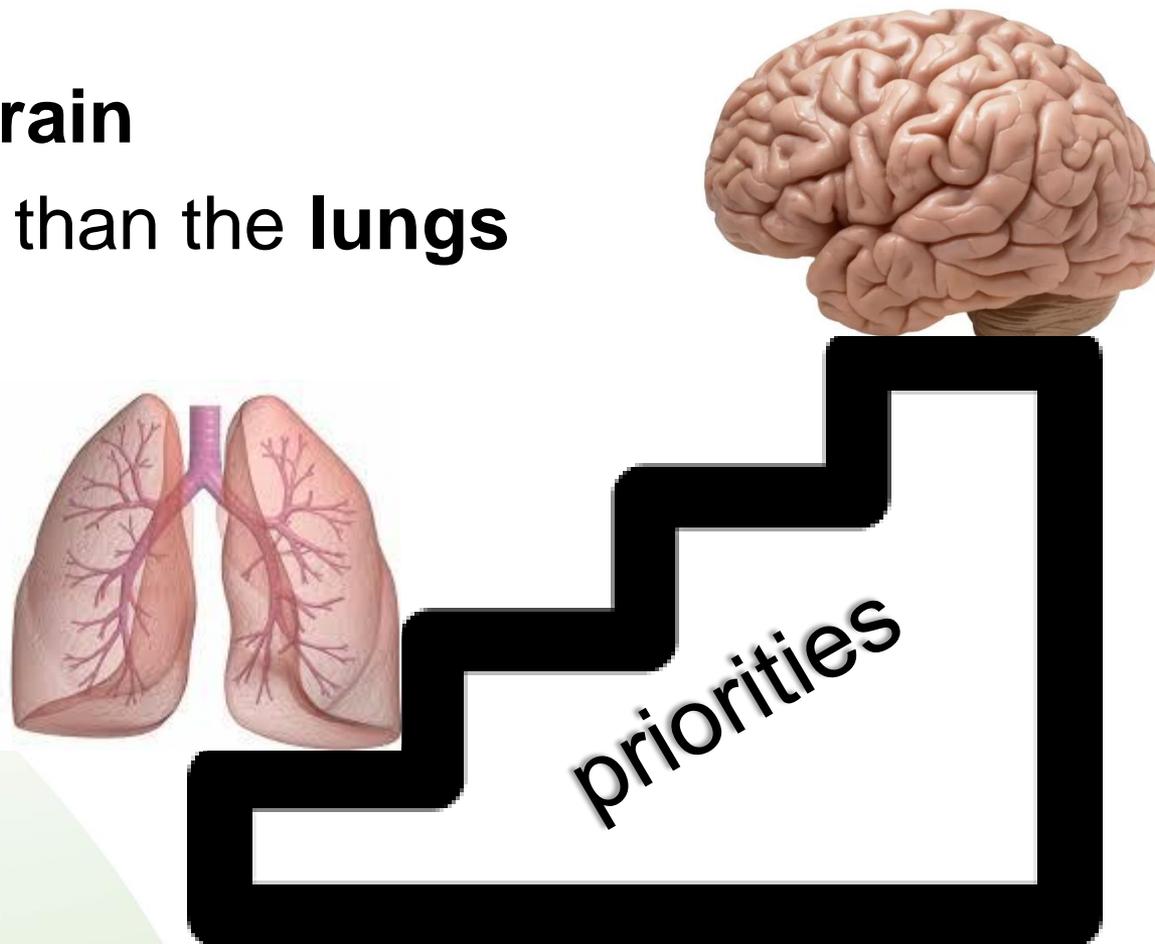


Hydrotherapy



## Enabling Participation

Explain that the **brain** is more important than the **lungs**

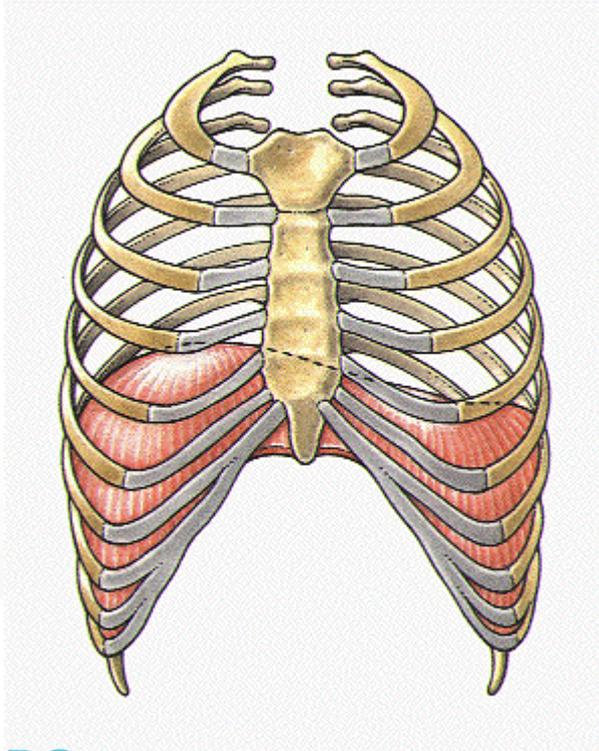




# Pathophysiology of Neuromuscular Respiratory Failure



## INSPIRATORY and EXPIRATORY muscles work differently

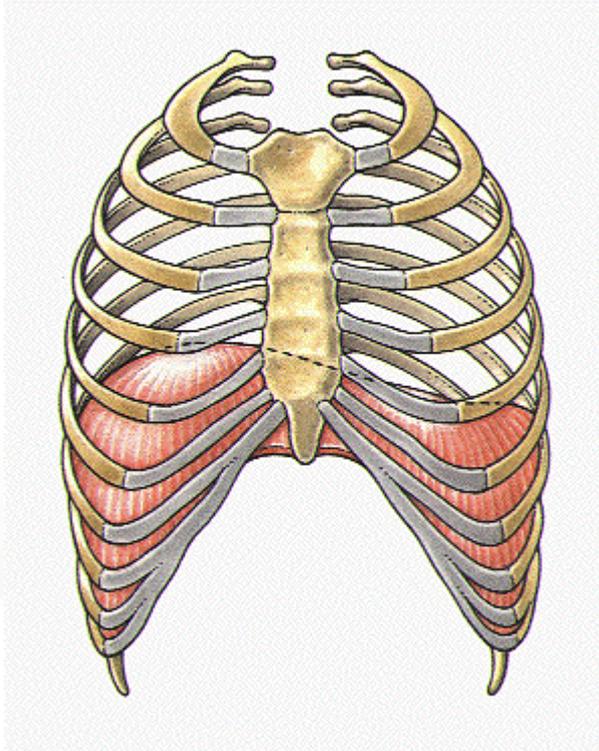


### Diaphragm:

- Inspiration only
- Innervation: C 3,4 and 5  
(keeps the diaphragm alive)
- Efficiency is dependent on:
  - 1) stable rib cage (strong intercostals) otherwise get paradoxical breathing



## INSPIRATORY and EXPIRATORY muscles work differently

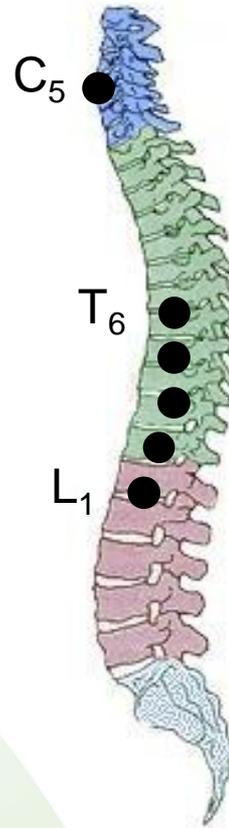
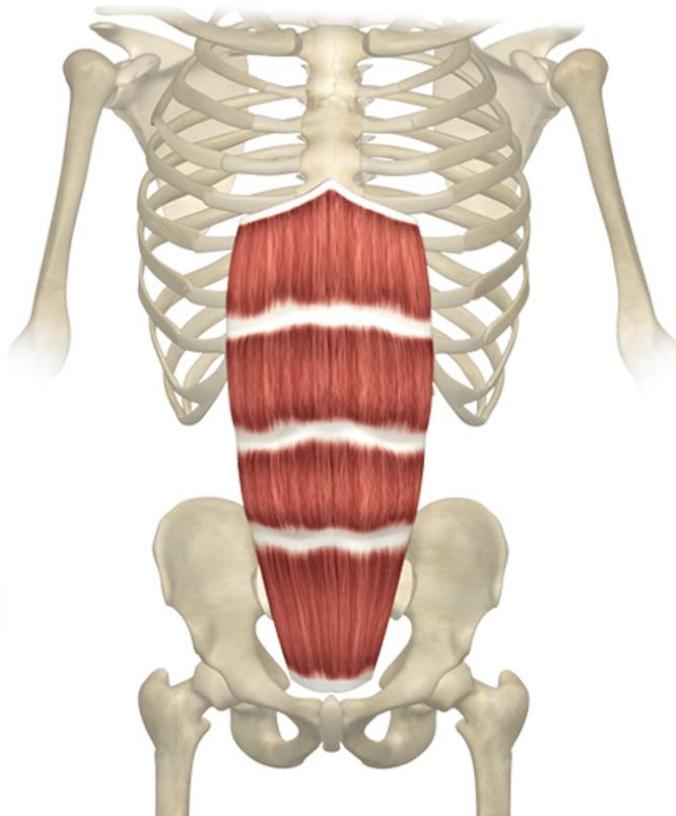


### Diaphragm:

- Inspiration only
- Innervation: C 3,4 and 5  
(keeps the diaphragm alive)
- Efficiency is dependent on:
  - 1) stable rib cage (strong intercostals) otherwise get paradoxical breathing
  - 2) Optimal starting position (determined by abdominal pressure)



## INSPIRATORY and EXPIRATORY muscles work differently

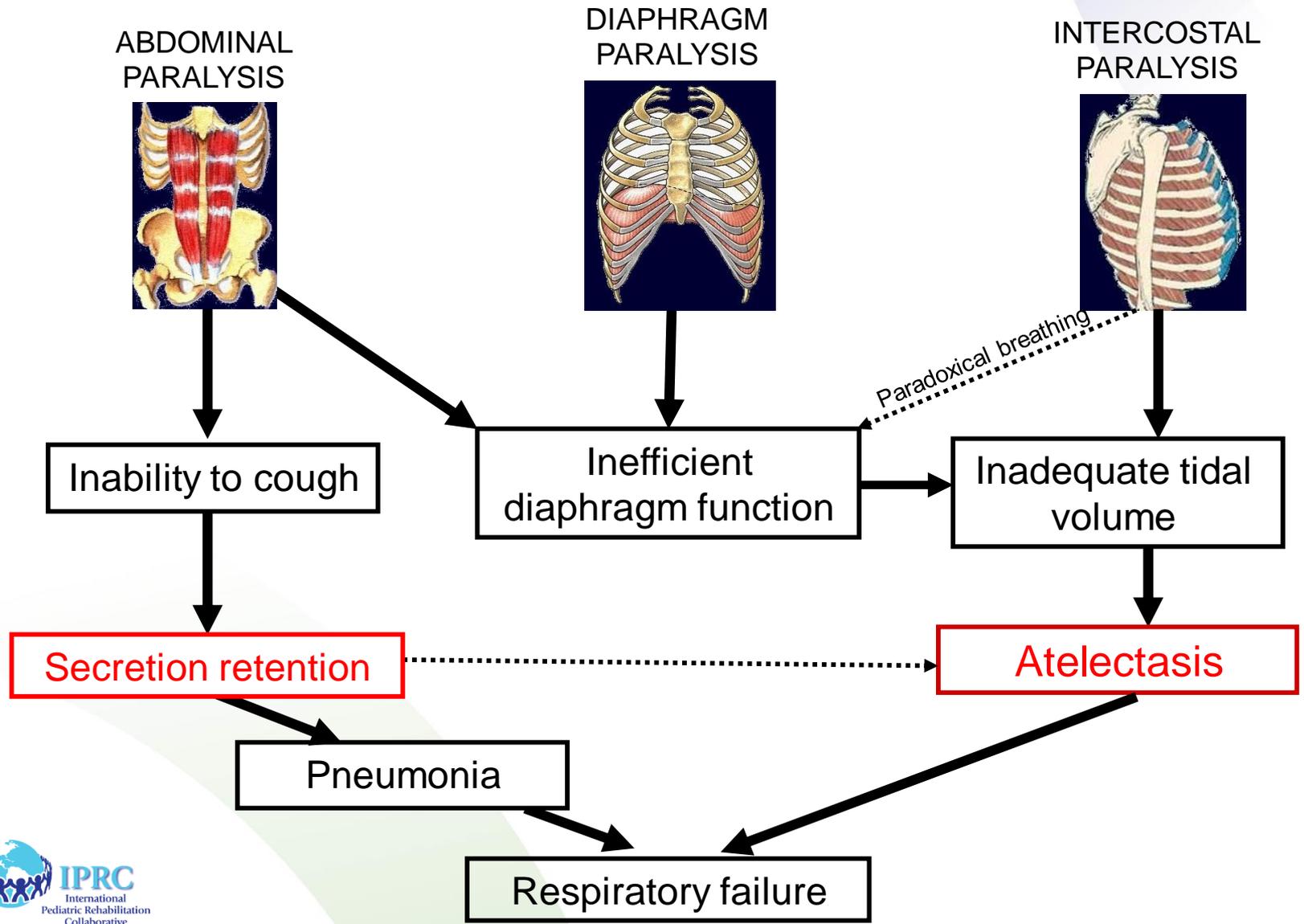


### Abdominal Muscles:

- Expiration (coughing)
- Maintains normal intra-abdominal pressure
  - keeps diaphragm in optimal starting position
- Innervation: T6 – L1



# Pathophysiology of NM Respiratory Failure

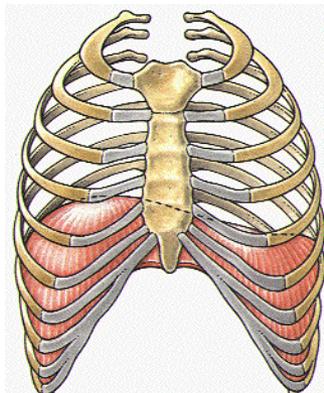




# Clinical Diagnosis and Treatment of Neuromuscular Respiratory Failure

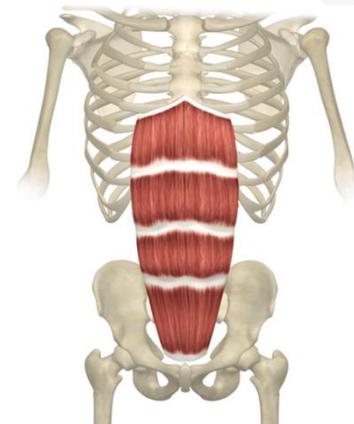


## The clinical presentations of INSPIRATORY muscle failure and of EXPIRATORY muscle failure are different



### Inspiratory Muscle Failure:

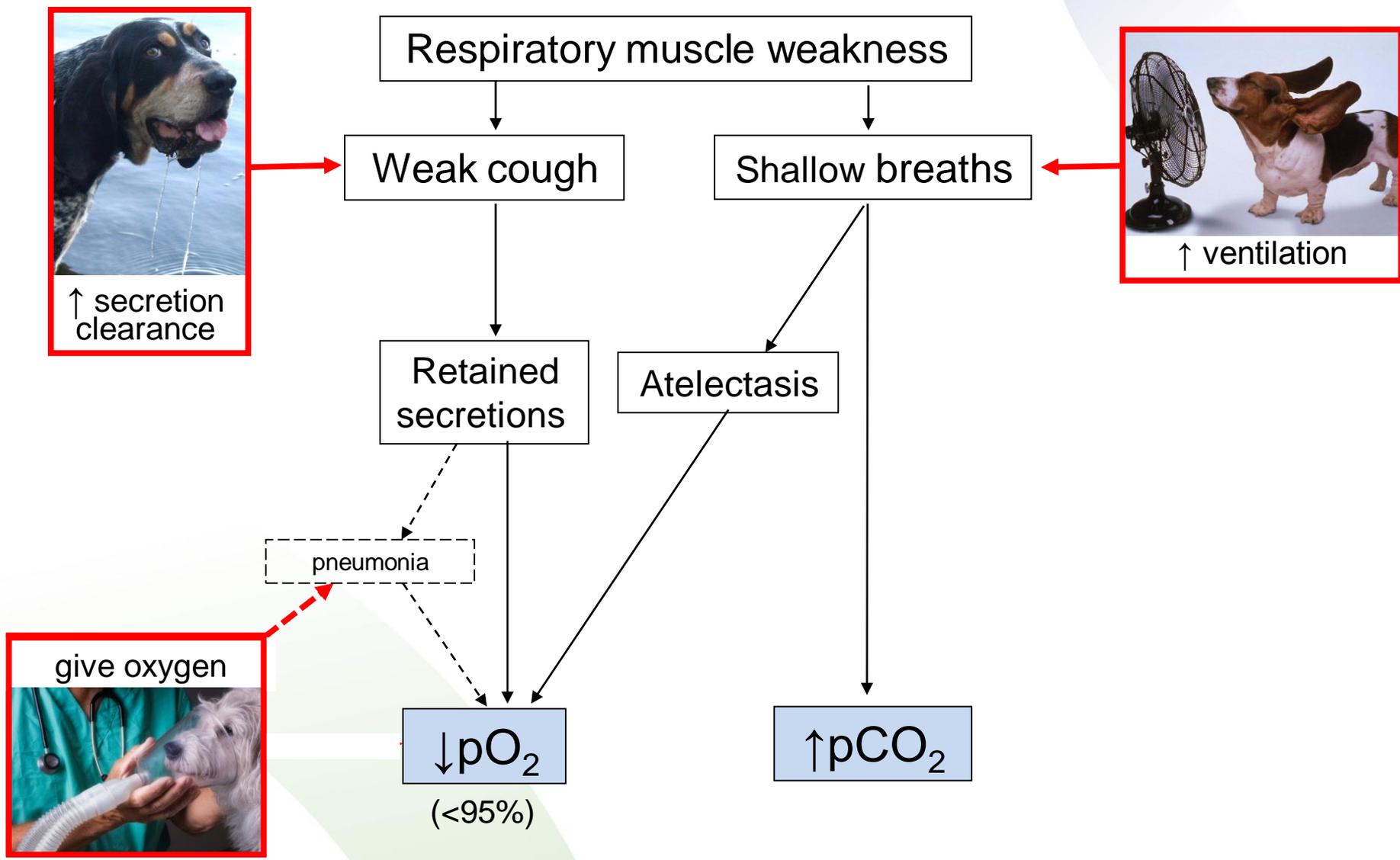
- Pathology: inadequate tidal volume
  - Symptoms: Gradually worsening hypercapnea
    - morning headache
    - decreased intellectual functioning
    - Drowsy
- (↓ in oxygen saturation occurs late)
- *Gradual deterioration*



### Expiratory Muscle Failure:

- Pathology: Ineffective cough
  - Symptoms: Acute pneumonia or atelectasis after “trivial” respiratory infection
- (sudden ↓ in oxygen saturation)
- *Sudden death*

Treat the neuromuscular cause, not the hypoxia symptom





# Strategies for Improving Ventilation





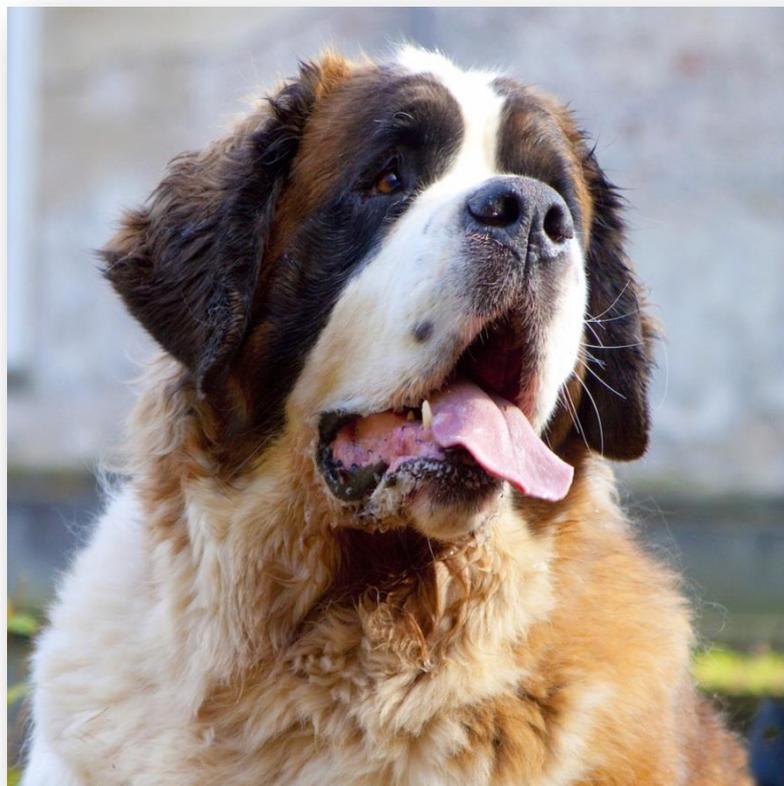
The ventilation strategy for treating neuromuscular respiratory failure is different from the strategy for treating parenchymal respiratory failure

## Parenchymal ventilation



Small, rapid breaths (to avoid barotrauma)  
SIMV mode

## Neuromuscular ventilation



Deep, slower breaths (to prevent atelectasis)  
Assist Control mode



# Strategy for Weaning Neuromuscular Patients from Ventilation



Short periods of intense effort (disconnection from the ventilator), so as to strengthen respiratory muscles



## “The Effect of Tidal Volumes on the Time to Wean Persons With High Tetraplegia From Ventilators.”

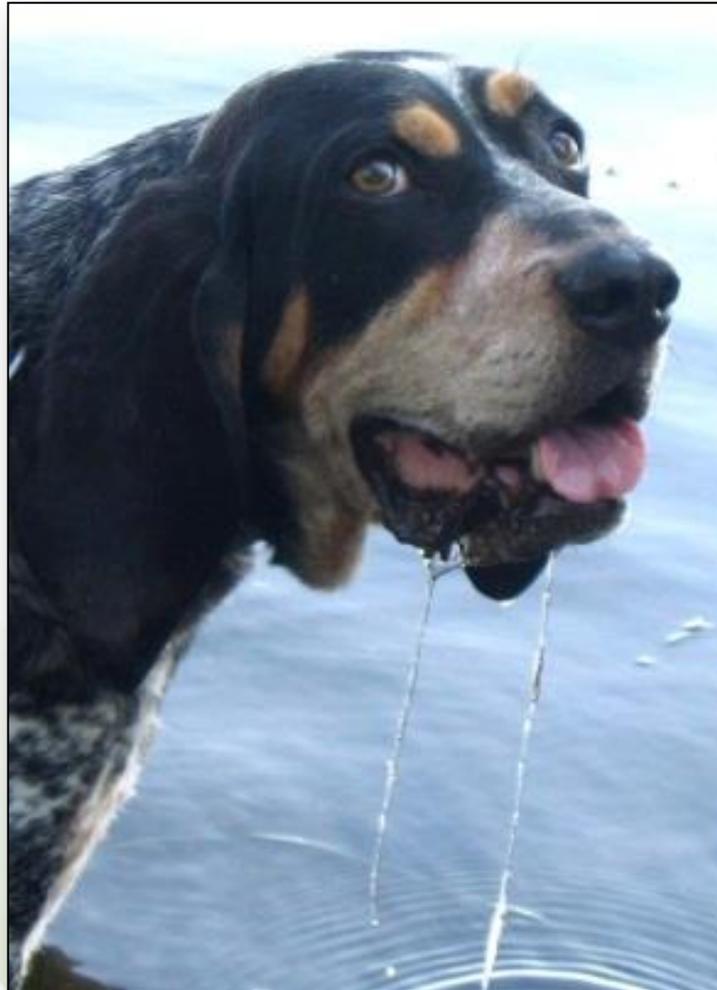
Peterson WP et al. *Spinal Cord*. 1999



- Retrospective review of 42 ventilated high SCI patients (C3/C4)
- 23 patients treated with “low Vt” ventilation (<20ml/Kg)
  - Atelectasis: 32% on admission, 52% after 2 weeks
  - Time to wean: 58 days
- 19 patients treated with “high Vt” ventilation (>20ml/Kg)
  - Atelectasis: 84% on admission, 16% after 2 weeks
  - Time to wean: 37 days
- High Vt ventilation resulted in less atelectasis ( $p=0.01$ ) and quicker weaning ( $p=0.02$ )



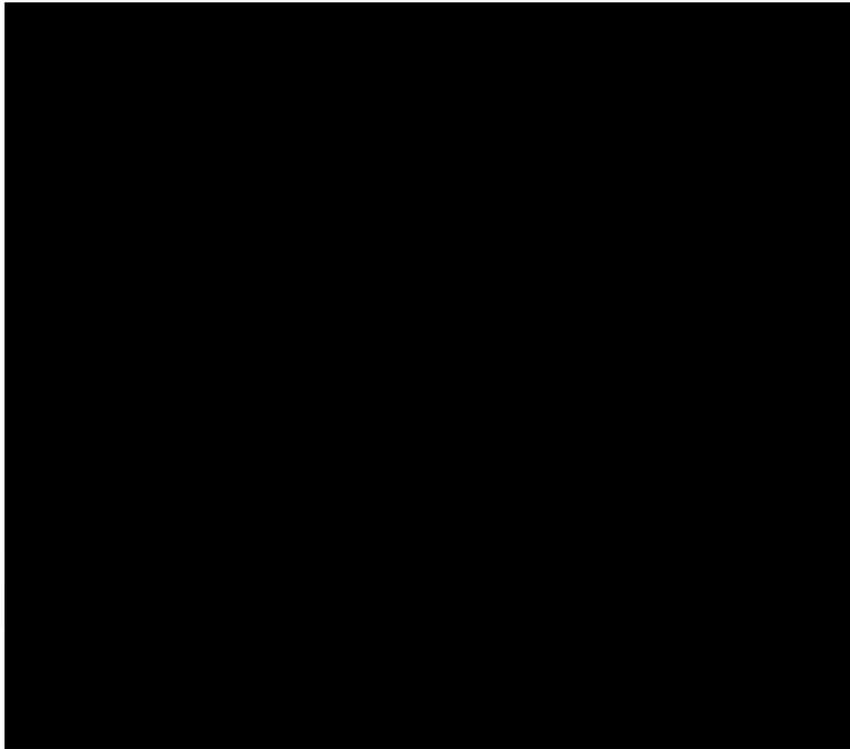
# Strategies For Improving Secretion Clearance



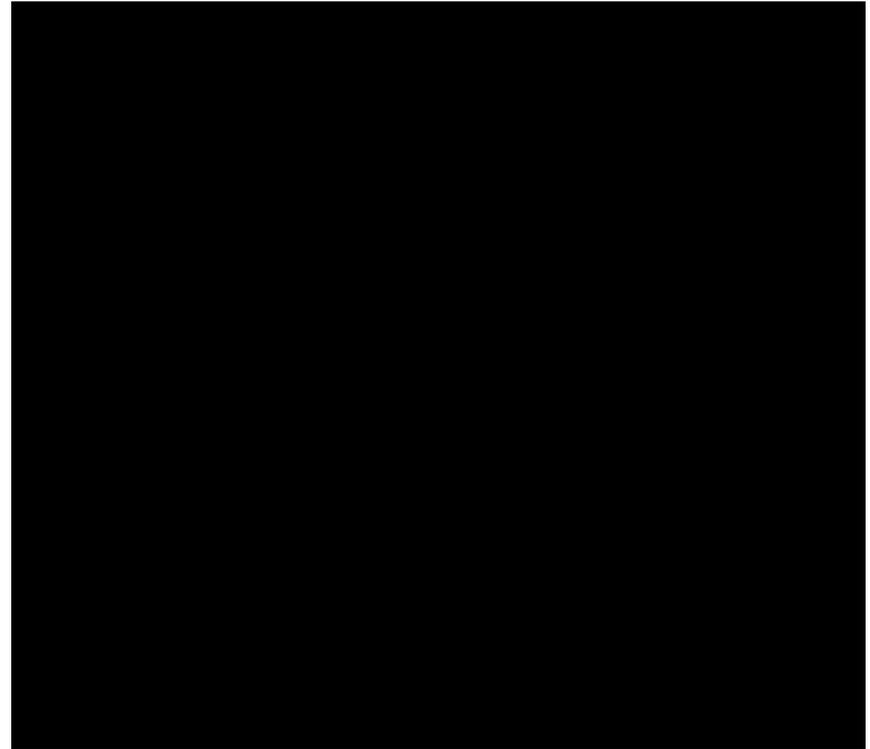


# Coughing is better than Catheter Suction

Catheter suction



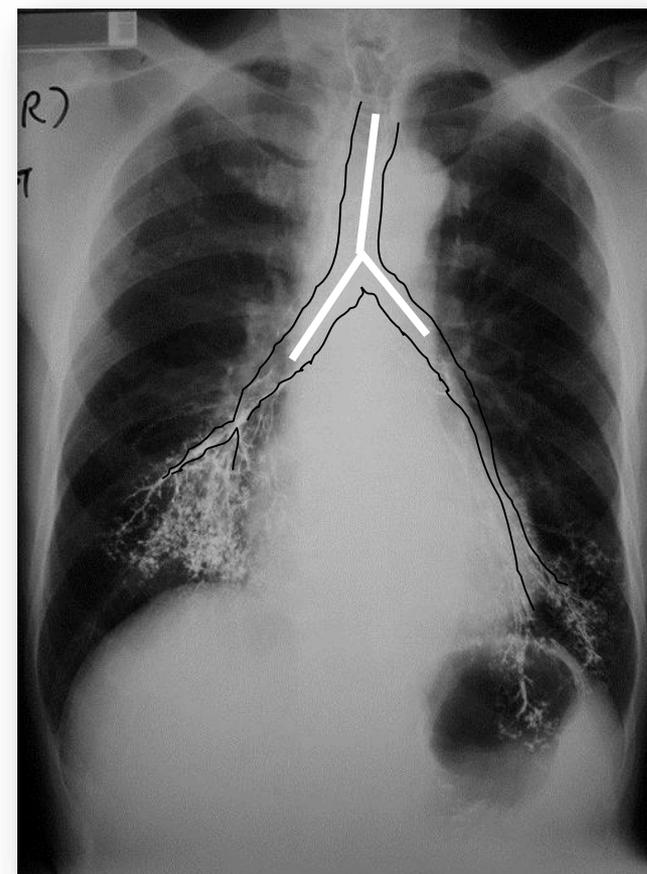
Cough





# Limitations of Catheter Suction

- Limited to trachea and mainstem bronchi
- Catheter suction misses the left lung 92% of the time
- 70% of pneumonia in SCI occurs in left lung, only 30% in right lung  
(*Fishburn, 1990*)





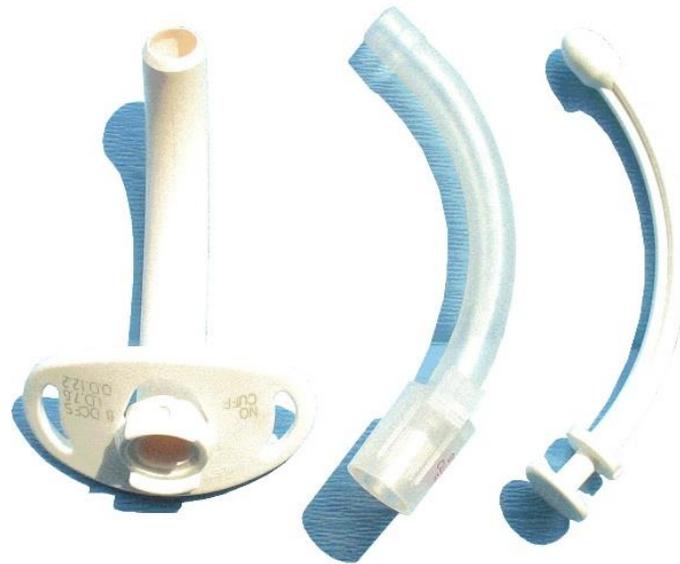
# Mechanical Inexsufflation

- Simulates a natural cough by alternating positive pressure insufflation with negative pressure exsufflation
- Can generate effective cough flows (>160 l/min) in paralyzed patients
- Generates airflow from 5<sup>th</sup> generation bronchi, bilaterally
- May be attached to facemask or ET tube
- Completely noninvasive





# Troubleshooting Tracheostomy





# Types of Tracheostomy Tubes

## Cuffed



- Protection from aspiration
- Best ventilation
- Precludes speech

## Uncuffed



- Fewer traumatic complications (scarring)
- Have to manage air-leak
- Facilitates speech



# Speech Valves

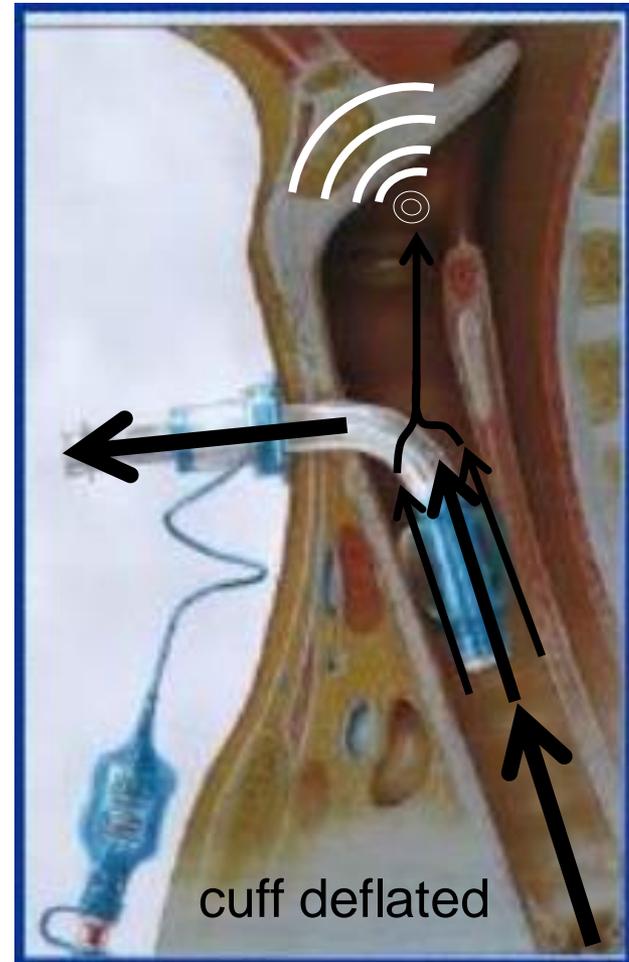
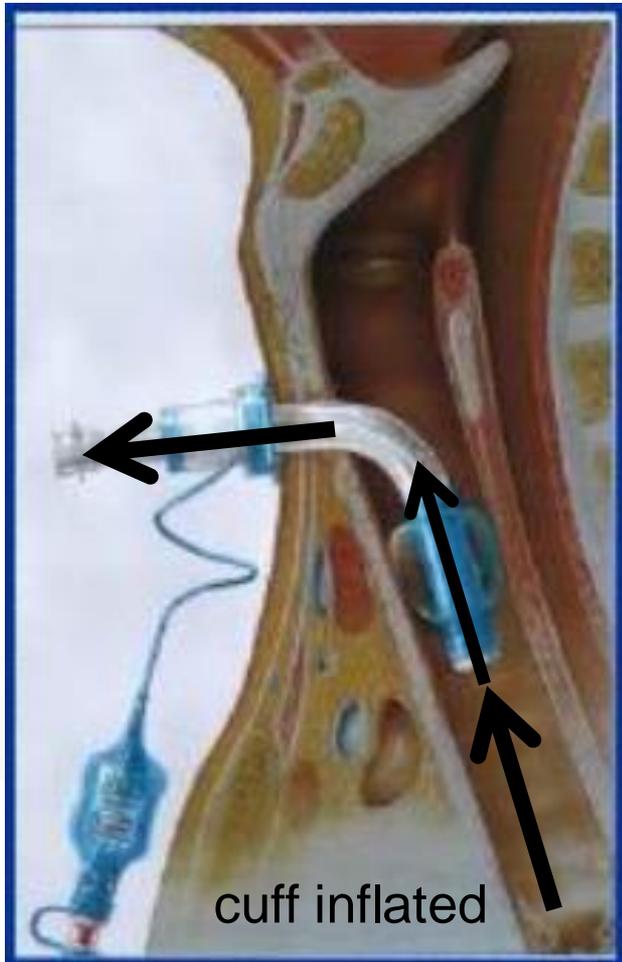




# Speech in the Non-Ventilated Patient With a Tracheostomy

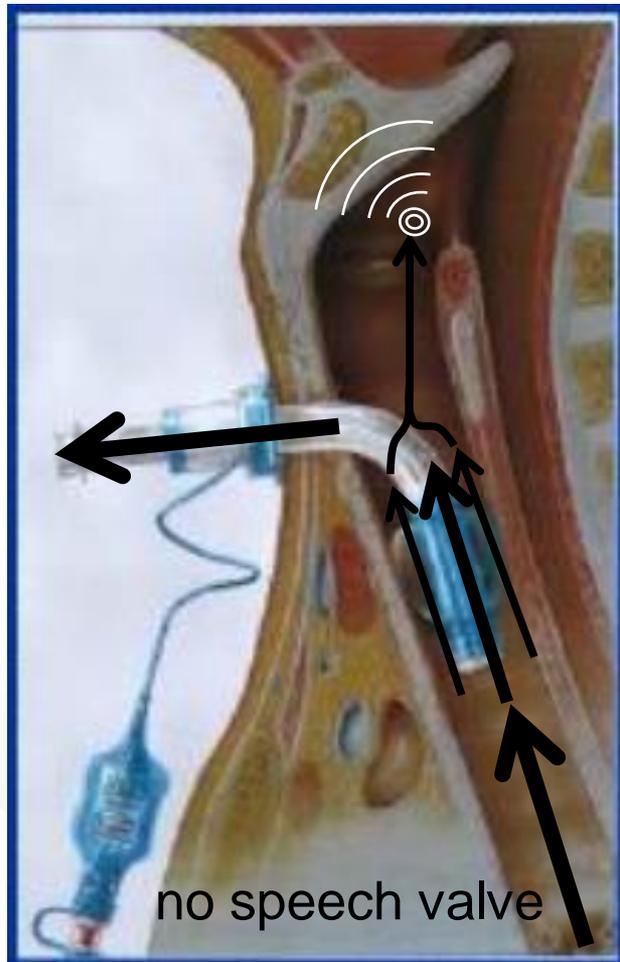


Phonation during natural breathing occurs only during EXPIRATION and with cuff deflated

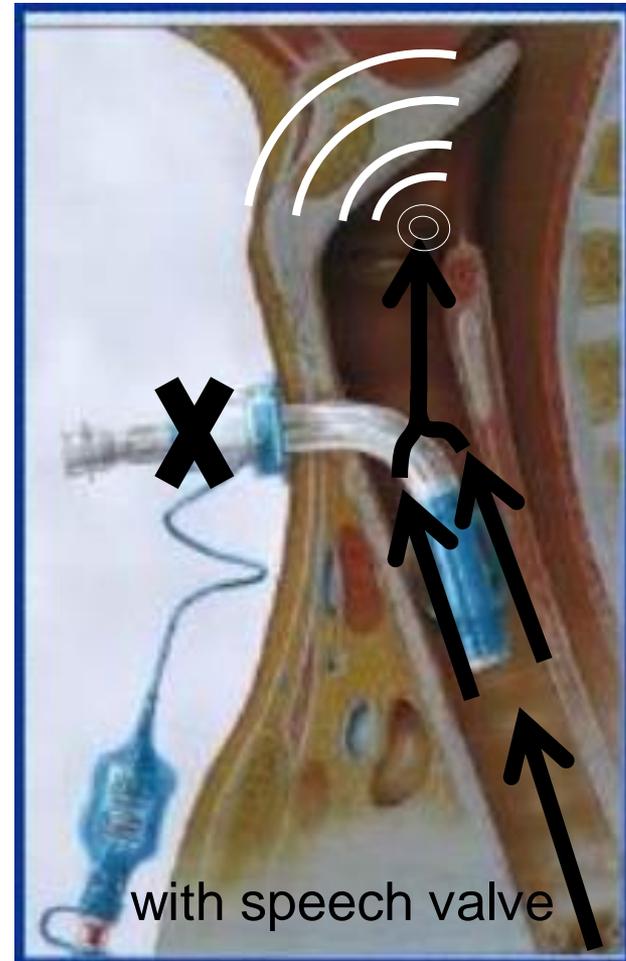




Speech valve prevents exhalation through the cannula, forcing more leak around cannula



no speech valve



with speech valve



# Speech Valve

## Benefits

- Voice production
- May partially enable valsalva (increases trunk stability and limb strength)
- More natural swallowing?

## Contraindications

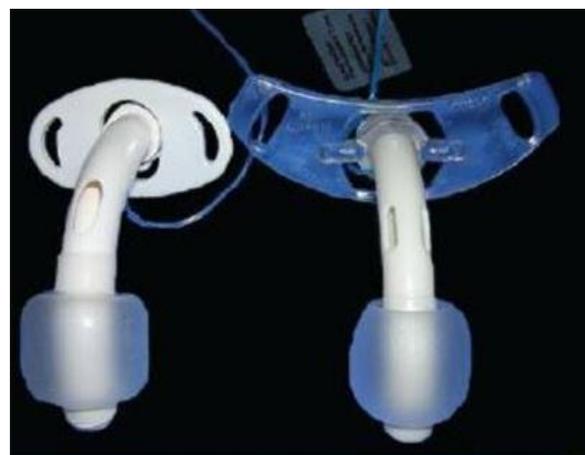
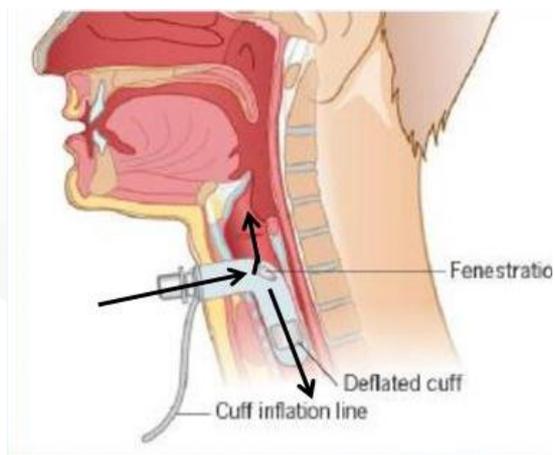
- No air leak around cannula (small children)
- Inflated cuff
- Upper airway obstruction





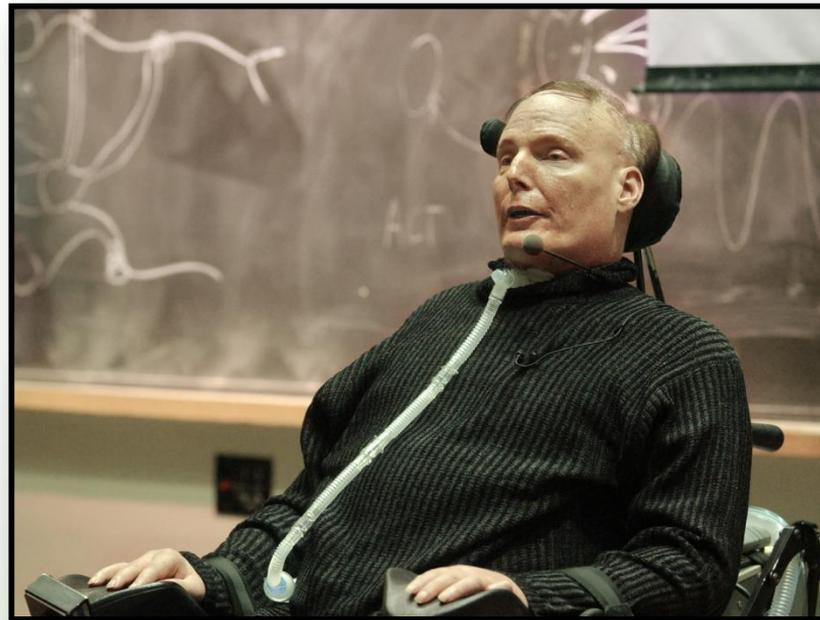
# To Improve Natural Speech:

- Use non-cuffed cannula instead of cuffed
- Try smaller cannula
- Use speech valve if possible
- Laryngeal vibrator
- Try fenestrated cannula



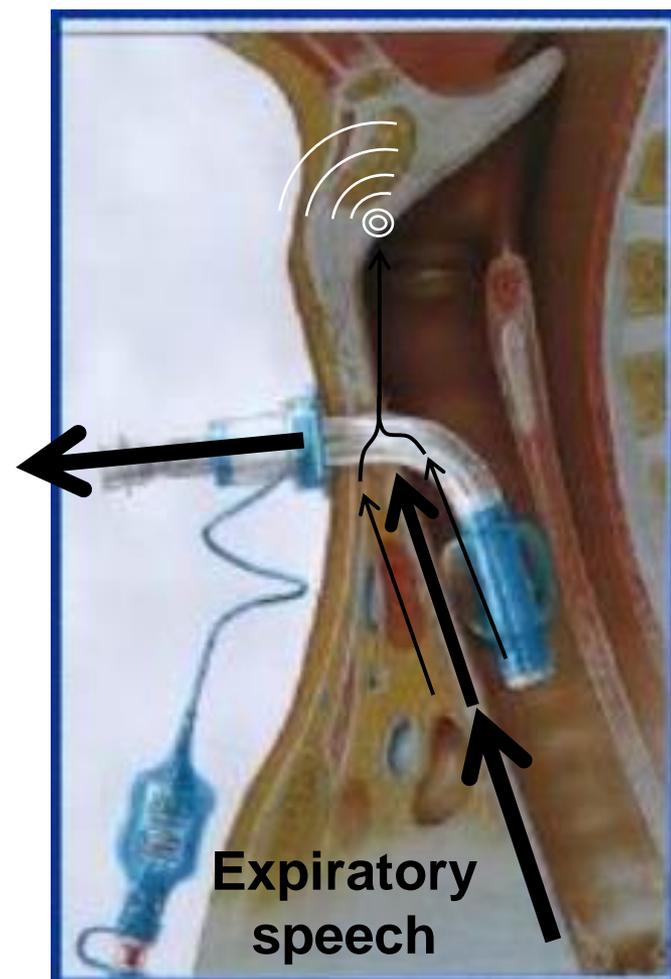
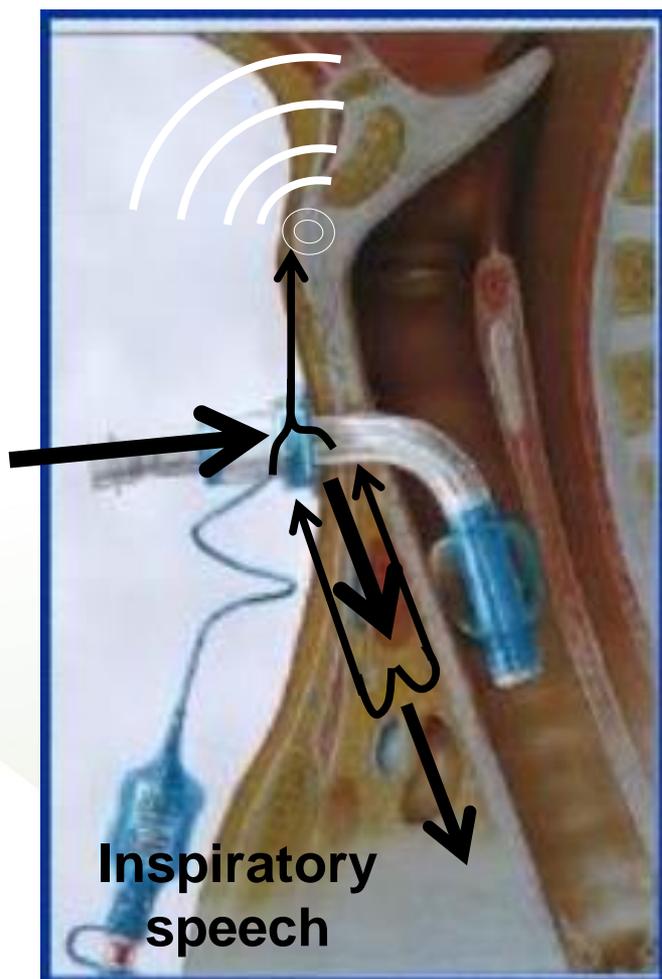


# Speech in the Ventilated Patient





Phonation during mechanical ventilation occurs loudest during INSPIRATION, weakly during expiration, and only with cuff deflated

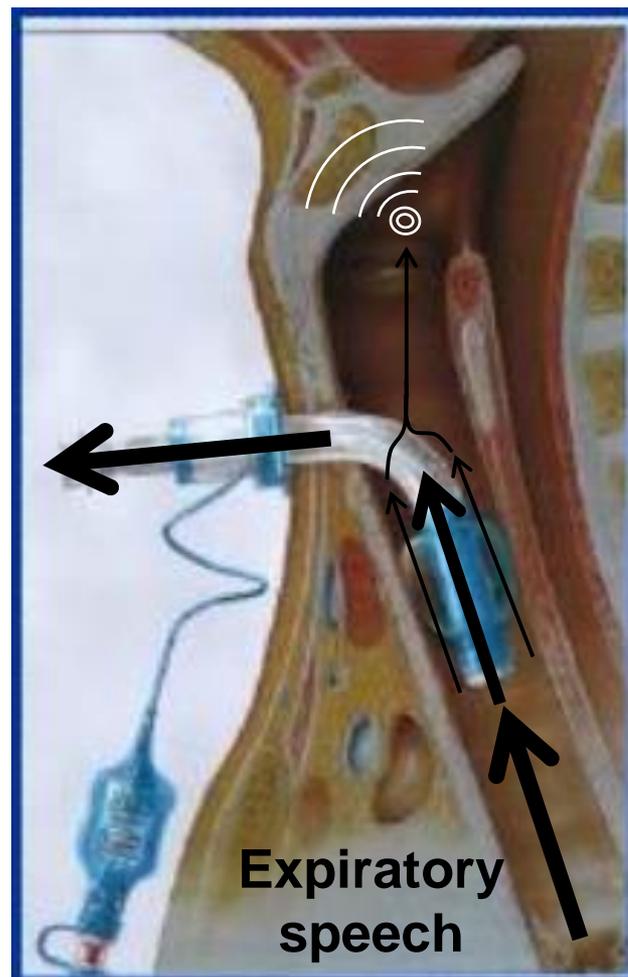
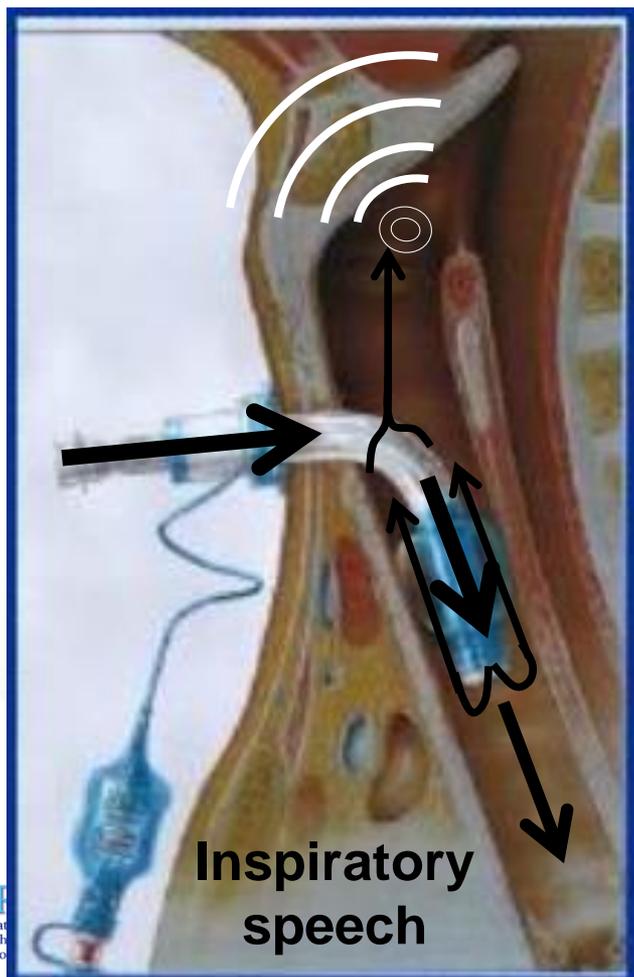


To improve  
ventilation  
speech

1. Don't use cuffed cannula
2. Decrease cannula size
3. Speech valve (expiratory speech)
4. Change parameters:
  - Longer inspiratory time (inspiratory speech)
  - Higher inspiratory pressure or volume (inspiratory speech)
  - Higher PEEP (expiratory speech)



- Hoit et al *Am J Speech-Lang Pathol* 1997
- Hoit et al *Chest* 2003
- Shea et al *J Speech Hear Res* 1994





# Noninvasive Ventilation





# Noninvasive Ventilation Advantages

- Better speech
- No risk of sudden death
- No increased secretions
- Psychological: gradual, *acceptable*
- Cosmetic





# Noninvasive Ventilation Disadvantages

- Labor intensive
  - Doctors
  - Parents
- Fussy (lots of possibility for error)
- Pressure sores
- Conjunctivitis
- Dry mouth
- Abdominal distension
- Full face mask may prevent speech
- Deformation of the maxilla



**The biggest hurdle: lack of a “philosophy of noninvasion” in acute care facilities.**



- Duchenne Muscular Dystrophy
- 24-hour vent dependent (3 years)
- Daytime mouthpiece ventilation
- Nighttime facemask ventilation





# Siblings with Nemaline Myopathy

Both fully vent dependent since age 2



Both facemask and tracheostomy are options



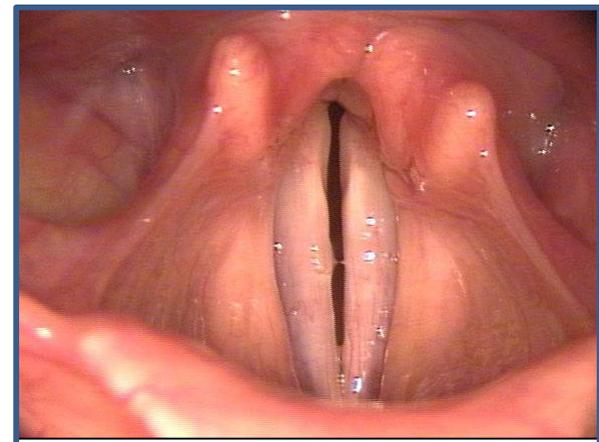
# Contraindications to Noninvasive Ventilation



Uncooperative or hyperactive child



Incompetent or unmotivated parents



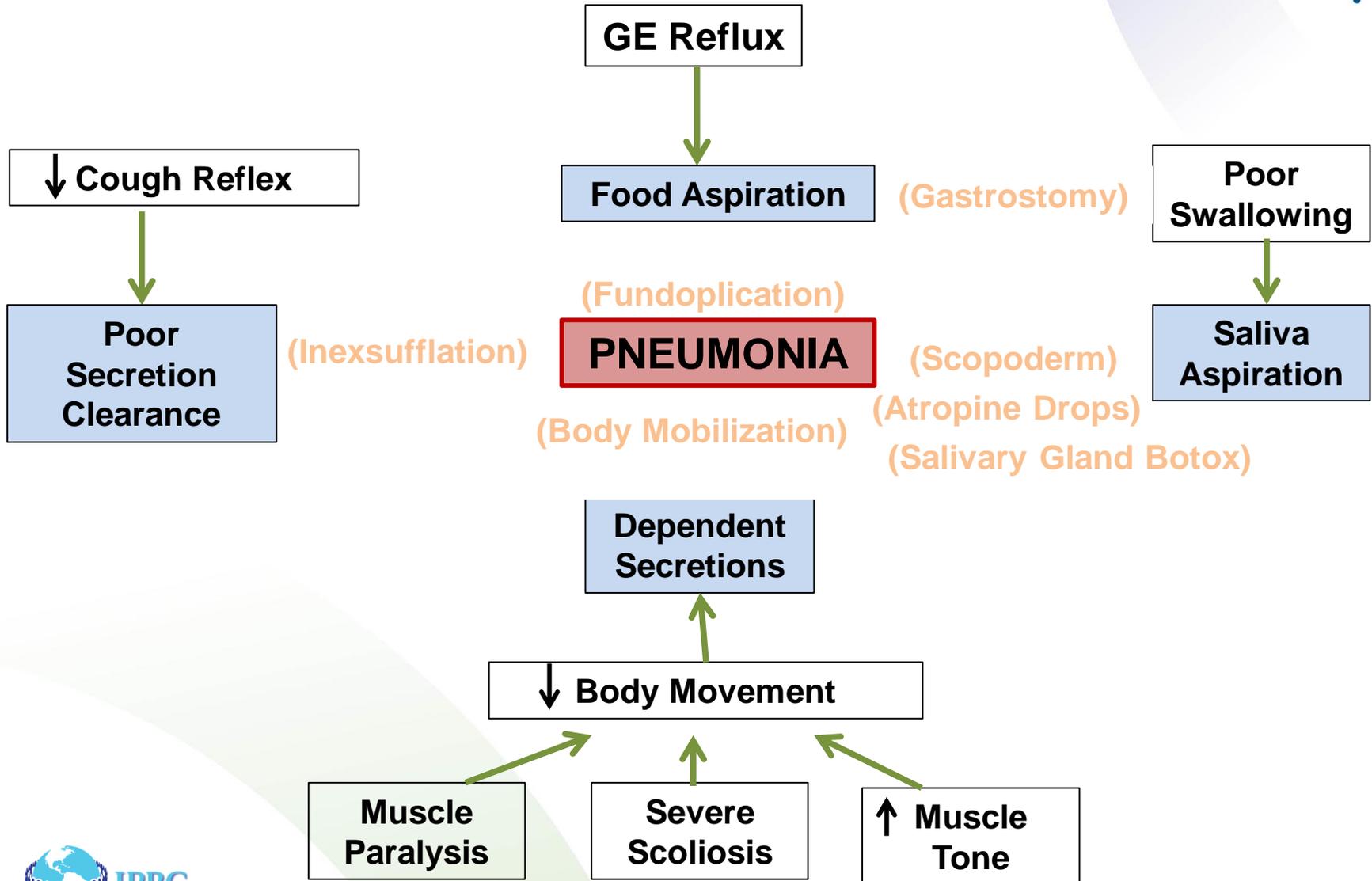
Airway obstruction:

- Vocal cord paralysis
- Poor secretion control

# Understanding Respiratory Pathology in Cerebral Palsy



# Pneumonia and its Prevention in CP





# Conclusions

- Neuromuscular disease has a unique impact on respiratory function, and should be managed with appropriate strategies
- Effective respiratory rehabilitation requires an integrated, holistic and multi-disciplinary approach



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A certificate of attendance for today's presentation may be requested via the survey.

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# Thank you!



I welcome any comments and questions:

Eliezer Be'eri, MD  
ebeerer@alyn.org