Pulmonary Impact and Management of Bulbar Dysfunction

Oscar Henry Mayer, MD
Division of Pulmonology
The Children's Hospital of Philadelphia
Professor of Clinical Pediatrics
The Perelman School of Medicine
at the University of Pennsylvania

May 8, 2025

Disclosures

- Active Consultant, Scientific Advisory Board member or Lecturer
  - Hoffman-La Roche
  - Scholar Rock
  - Sanofi
  - Astellas
  - Catalyst Pharmaceuticals
  - Novartis

Outline

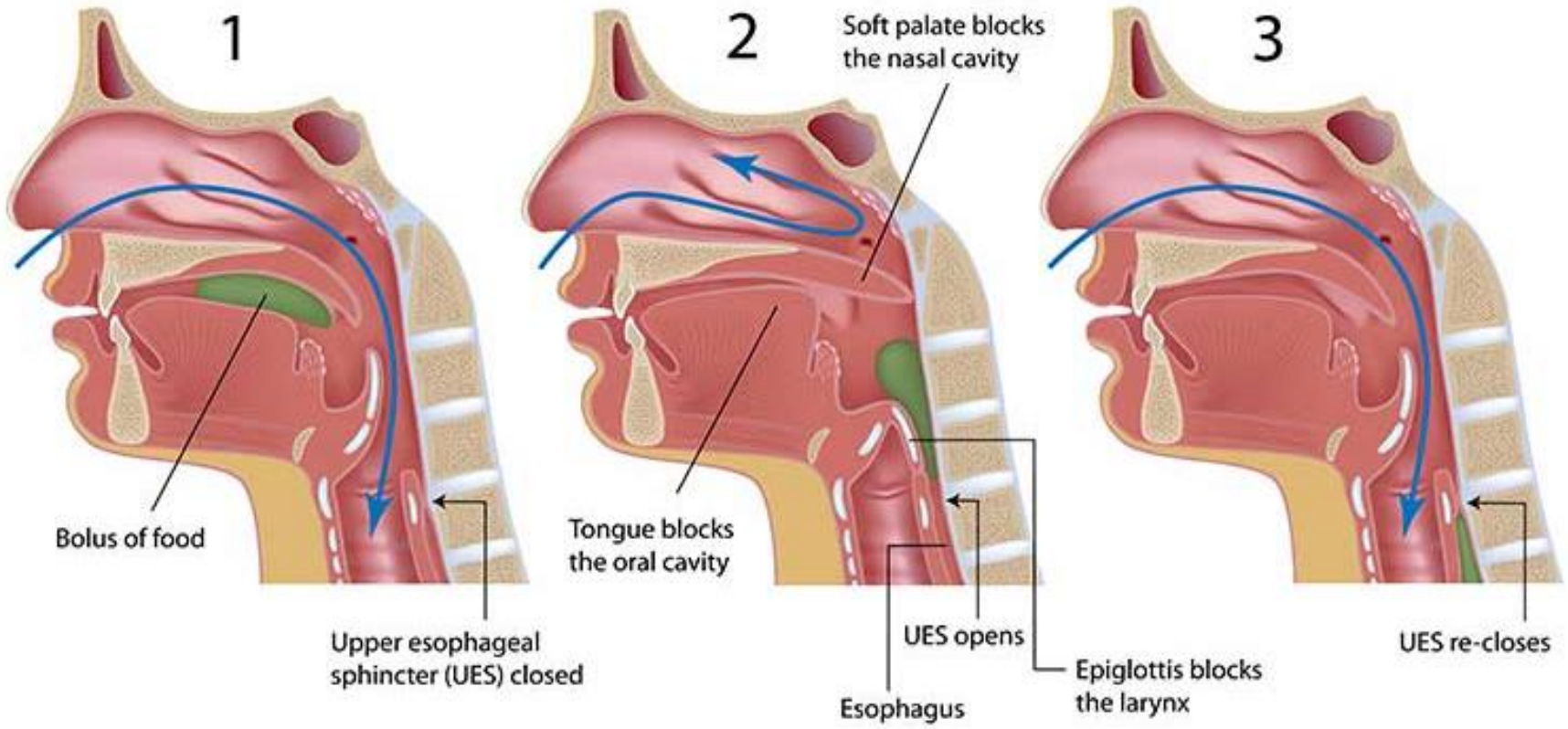
- Why is bulbar function important to a pulmonologist?
- Impact of inadequate bulbar function
- How to support bulbar dysfunction
- Management principles & challenges

Importance of bulbar function to a pulmonologist?

- Safe and effective breathing - ”Traffic Control”

Importance of bulbar function to a pulmonologist?

- Safe and effective breathing - ”Traffic Control”



https://www.ausmed.com/learn/articles/dysphagia

Importance of bulbar function to a pulmonologist?

- Safe and effective breathing - ”Traffic Control”
- Critical in overall secretion control
  - Reducing risk of aspiration
    - Processing oronasal secretions
    - Moving to esophagus
  - Preventing / minimizing penetration
    - Recognizing material in posterior pharynx
    - Closing vocal cords
  - Removing aspirated material
    - Coughing

Impact of inadequate bulbar function

- Supraglottic/Glottic
  - Aspiration – inadequate processing of pharyngeal secretions
    - Micro
      - Small volume repeated episodes
      - Chronic inflammation
    - Macro
      - Larger volume single / small number of episodes
      - Lower respiratory tract obstruction / acute inflammation / infection
- Subglottic
  - Inadequate clearance – Cough and vocal cord adduction
    - Removing aspirated material
    - Removing infectious material

Impact of Aspiration - Microaspiration

- Signs of microaspiration
  - Persistent airway inflammation
    - What is being aspirated?
      - Acid vs milk vs water
    - Confused with asthma
    - Wet cough due to mucus production in the lungs
  - Recurrent respiratory symptoms/illnesses
    - With or without fever
  - Chest CT Scan - Interstitial lung disease
    - Can be diffuse
    - Bronchiectasis
  - Supplemental oxygen requirement

Impact of Aspiration - Microaspiration

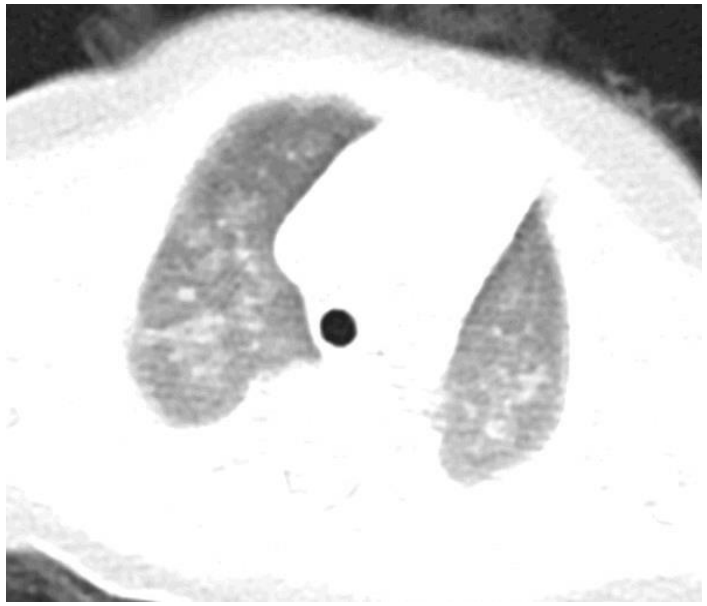
- Evaluations for microaspiration
  - Broad, long-term assessment
    - Pattern of symptoms
    - Character of illnesses
    - Hard to identify an episode – low impact from a single microaspiration event
  - Bypass swallow / stomach processing – Nasogastric Tube
    - Evaluate for symptom resolution over time
  - Empiric therapy for gastroesophageal reflux is not enough
    - Reduce acid content
    - Upper airway secretions

Impact of Aspiration – Macroaspiration

- Signs of macroaspiration
  - Acute airway obstruction
  - Acute infection
  - Localized based on position and the dependent part of the lung
    - Upright – Lower lobes
    - Supine – Posterior regions Upper and Lower lobes
  - Determined in part by cough effectiveness
    - Ability to cough
    - Ability to recognize the need to cough

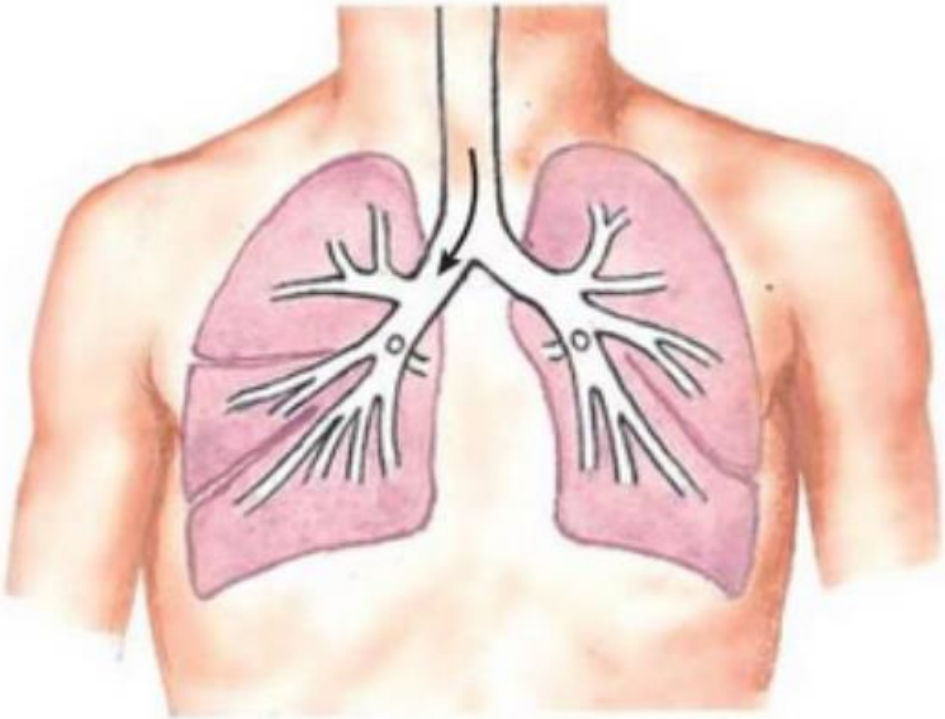
Impact of Aspiration – Macroaspiration

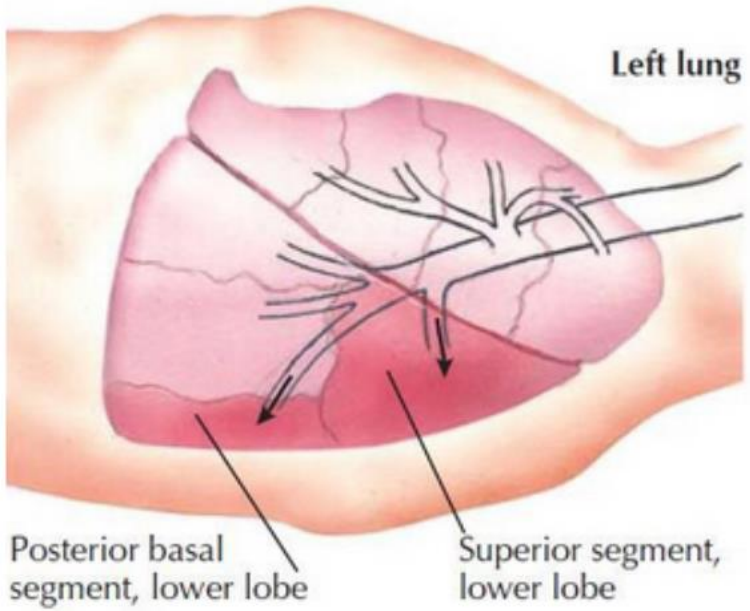
- Evaluation for aspiration
  - Chest radiography is often diagnostic
  - Chest CT
    - Volume loss – Atelectasis
    - Bronchiectasis – repeated episodes

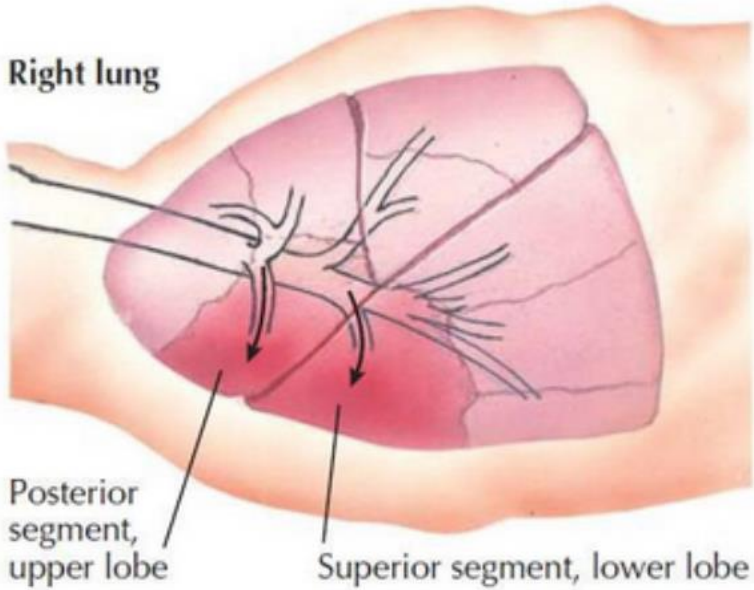





Aspiration and Position







https://medicinespecifics.com/lung-lobes-most-commonly-affected-by-aspiration-upright-vs-supine/

Treatment Strategies

Parachute use to prevent death and major trauma related to gravitational challenge: systematic review of randomised controlled trials

Gordon C S Smith, Jill P Pell

BMJ VOLUME 327 20–27 DECEMBER 2003 bmj.com

Parachutes reduce the risk of injury after gravitational challenge, but their effectiveness has not been proved with randomised controlled trials

How to Support Bulbar Dysfunction

- Reduce the risk of aspiration
  - Minimize presence of upper airway secretions / material
  - Decrease production of secretions
  - Reduce gastroesophageal reflux
  - Make secretions easier to clear
- Improve function
  - Improve swallow function to handle
    - Normal secretion volume
    - Increased secretion volume
  - Support swallowing
    - Suctioning
  - Support / replace cough

Reducing Risk of Aspiration

- Decrease production – making thin secretions less
  - Oral
    - Anticholinergic
      - Glycopyrrolate
      - Atropine
      - Scopolamine
  - Nasal
    - Adrenergic
      - Oxymetolazine
    - Antihistamine
    - Antiinflammatory

Reducing Risk of Aspiration

- Decrease secretion production / stimulation
  - Gastroesophageal reflux – Increase saliva production
    - Esophageal acid contact
    - Posterior pharyngeal contact
    - Physical esophageal stimulation
  - Oral stimulation

A Fool’s Errand??

A Fool's Errand??

- Using medications to overcome physical dysfunction
- Challenges:
  - Getting the optimal effect
  - Maintaining the optimal effect – Being dynamic
    - Resolution – de-escalation
    - Progression – escalation
  - Minimizing side effects
    - Anticholinergic – over drying, urinary retention
    - Mucosal drying / trauma
    - Mucus viscosity – making secretion clearance worse
    - Saline – over hydrating mucus and increasing volume

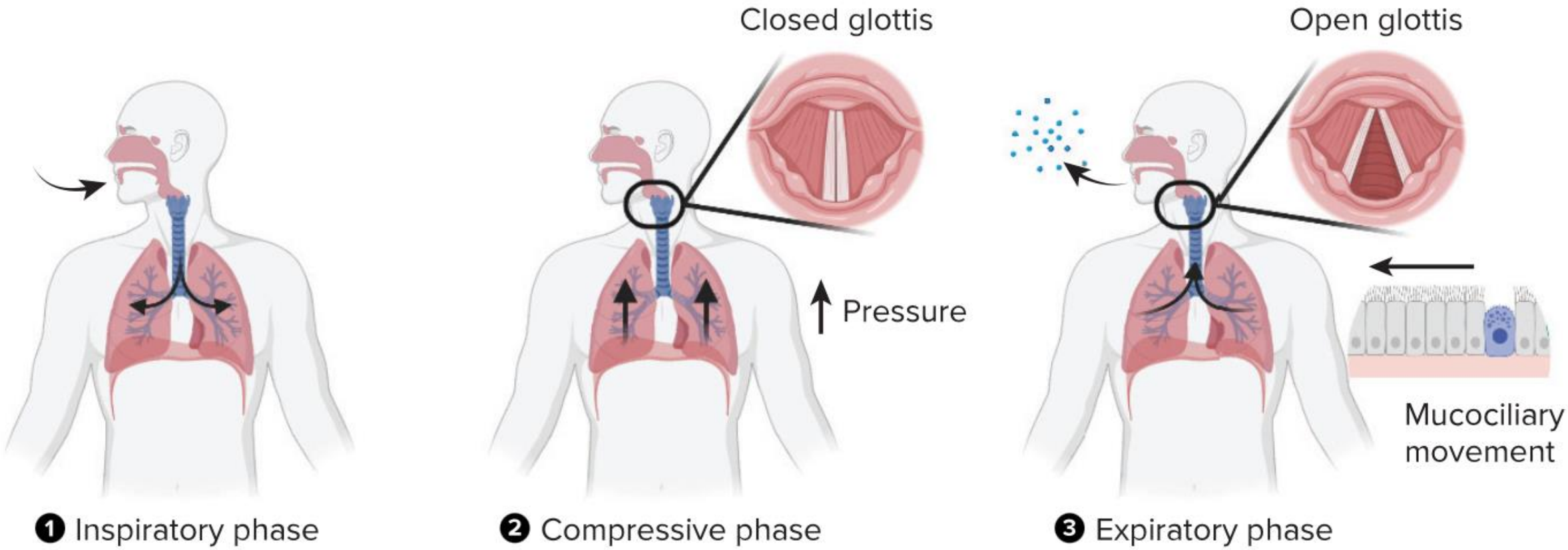
Reducing Risk of Aspiration

- Improve function
  - Improve swallow function to handle
    - Normal secretion volume
    - Increased secretion volume
  - Support swallowing
    - Suctioning
- Removing secretions
- Safe oral feeding / G Tube supplementation

Removing Secretions

- Cough or suction
- Cough phases
  - Deep inspiration
  - Forceful exhalation
  - Glottic closure
  - Glottis opens expulsive expiratory flow
- Absence of either step causes impaired airway clearance

Cough Phases



https://knowablemagazine.org/content/article/health-disease/2023/new-secrets-of-cough-reflex

What is a normal cough?

- Peak cough flow
  - >160 L/s – predicted successful extubation (Bach, et.al)
  - >270 L/s – “predicts” a PCF > 160 L/s during illness
- Unclear how applicable to young pediatric population
- Management of acute illness
  - Duration of acute illnesses
    - <7 days
    - >14 days

Supplementing Airway Clearance

- Inspiratory Assistance
- Expiratory Assistance
- Inspiratory / Expiratory Assistance
- Other

Inspiratory Assistance

Intermittent Positive Pressure Breaths (IPPB)

- Apply inspiratory pressure
  - Hyperinflate lungs
  - Increase airway caliber
- Passive recoil with no expiratory pressure
- Can be used with nebulization

Alpha 200®; Salvia Lifetec

Inspiratory Assistance

- Breath Stacking
  - Oronasal Mask
  - One way valve
- Glossopharyngeal Breathing
  - Inspiration augmented by pharyngeal muscles

Inspiratory Assistance

Expiratory Assistance

Manually Assisted Cough

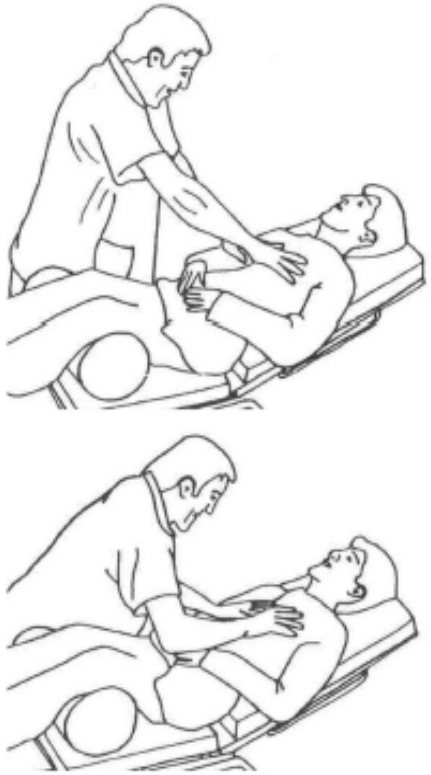


Fig. 1. Manually assisted cough via thoracic compression.

Fig. 2. Manually assisted cough via abdominal-thoracic compression.

Expiratory Assistance

	Duchenne or Becker Muscular Dystrophy	Type-2 Spinal Muscular Atrophy	Other Neuromuscular Diseases
Male/female	127/0	14/12	18/8
Age (mean ± SD y)	22.2 ± 7.1	26.4 ± 9	33 ± 15
BMI (mean ± SD kg/m^2)	17.2 ± 5	19.6 ± 7.6	18 ± 3.9
VC (mean ± SD mL)	1,191 ± 890	1,644 ± 1,005	1,348 ± 719
VC (mean ± SD % predicted)	24 ± 17	38 ± 22	29 ± 15
MEP (mean ± SD cm H_2O)	26 ± 18	39 ± 16	42 ± 20
Unassisted PCF (mean ± SD L/min)	163 ± 81	198 ± 78	199 ± 84
PCF with thoracic MAC (mean ± SD L/min)	209 ± 71*	225 ± 73*	197 ± 78
PCF with abdominal-thoracic MAC (mean ± SD L/min)	210 ± 70*	245 ± 73*†	197 ± 85

*Assisted peak cough flow (PCF) was significantly greater than unassisted PCF ($P < .001$ via paired t test).
† PCF was greater with abdominal-thoracic manually assisted cough (MAC) than with thoracic MAC ($P < .001$ via paired t test).
BMI = body mass index
VC = vital capacity
MEP = maximum expiratory pressure

Toussaint, *et. al. Respir Care.* 2009.

Inspiratory & Expiratory Assistance

- Breath stacking / Manually Assisted Cough
- Mechanical In-Exsufflator (Cough Assist®)

Inspiratory & Expiratory Assistance

- Cough Assist
  - Cycle
    - Pi 15-50 cm H2O
    - Hold at target Pi for 1-2 minutes
    - Pe 15-50 cm H2O
  - 5 Cycles
  - Suction
  - 5 Sets
  - Oscillation

Inspiratory & Expiratory Assistance

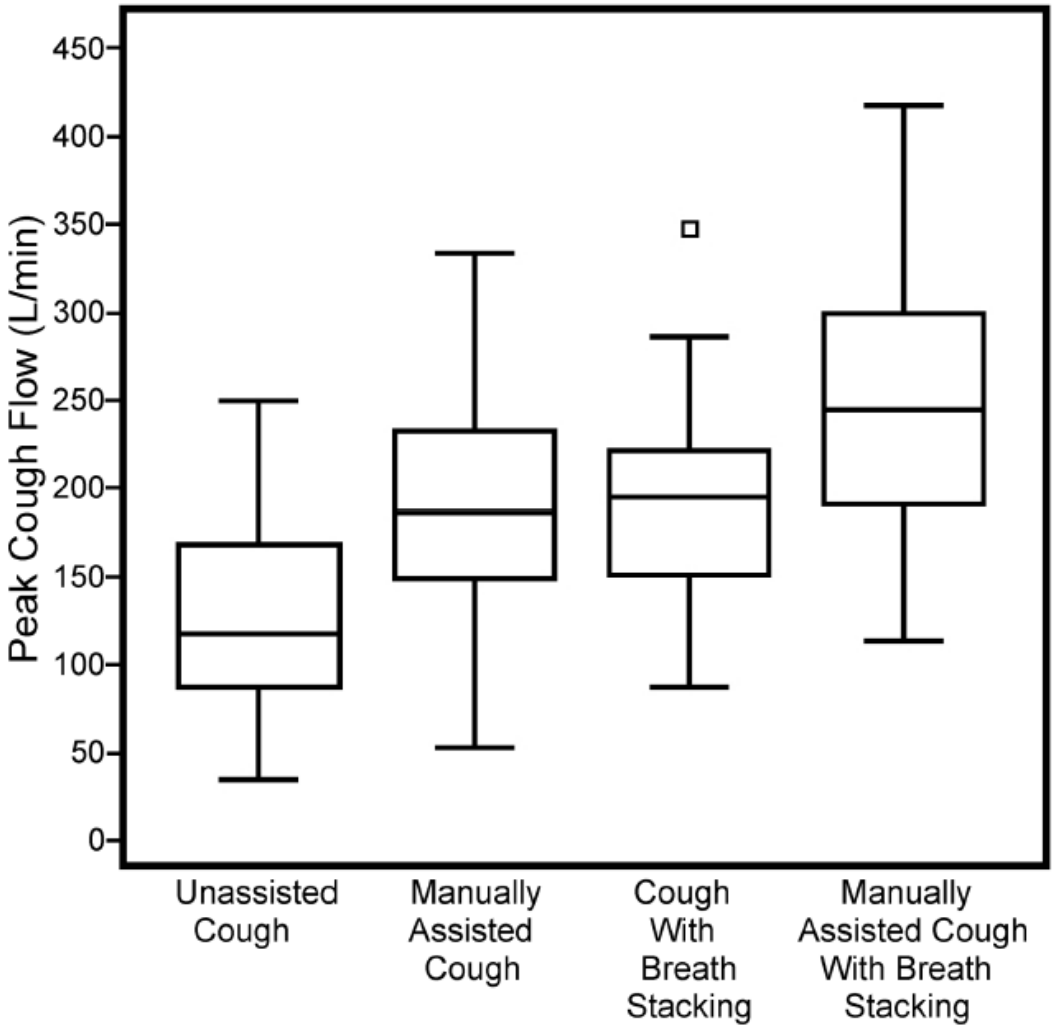
- Cough Assist
  - Cycle
    - Pi 15-50 cm H2O
    - Hold at target Pi for 1-2 minutes
    - Pe 15-50 cm H2O
  - 5 Cycles
  - Suction
  - 5 Sets
  - Oscillation

Synclara®; Hill Rom

Cough Assist®; Phillips Respironics

BiWaze®; ABM Respiratory Care

Inspiratory & Expiratory Assistance



Toussaint, *et. al. Respir Care.* 2009.

Inspiratory & Expiratory Assistance

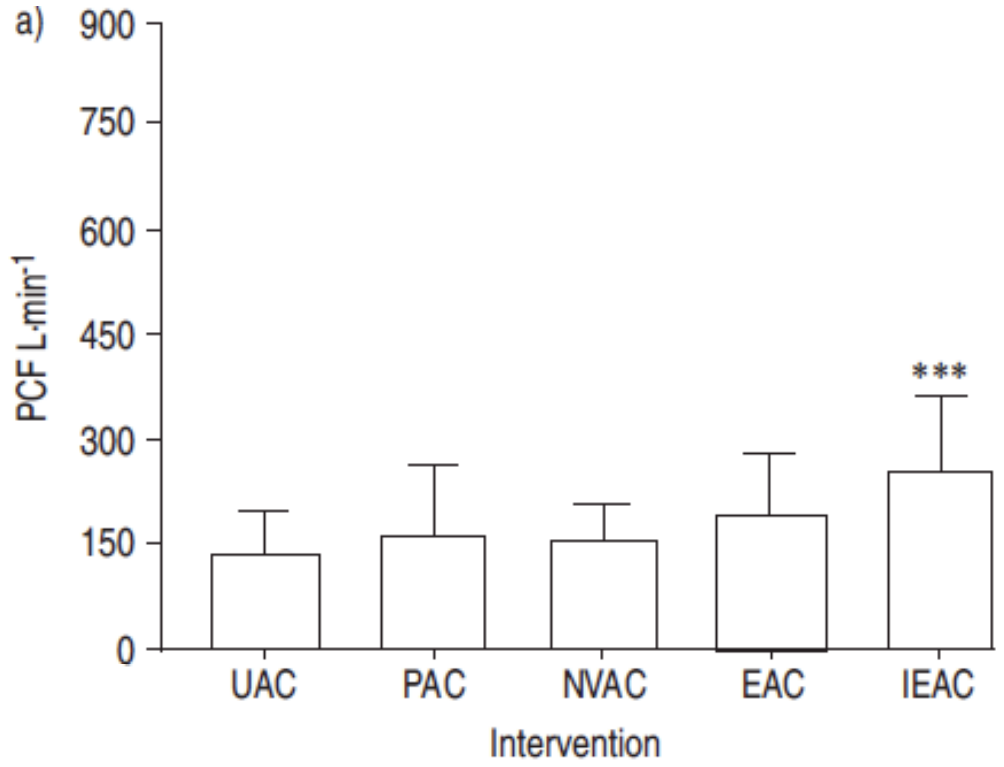


Fig. 2.– Peak cough flow (PCF) in a) paediatric patients, b) paediatric controls, c) adult patients, and d) adult controls. Data are presented as mean±SD. UAC: unassisted cough; PAC: physiotherapy-assisted cough; NVAC: noninvasive ventilator-assisted cough; EAC: exsufflation-assisted cough; IEAC: insufflation/exsufflation-assisted cough. **, ***: $p<0.01$, $p<0.001$ *versus* UAC.

Chatwin, *et. al. Eur Respir J.* 2003.

Mucus Mobilization

- Physiotherapy
  - Intrapulmonary percussive ventilation
  - High frequency chest wall oscillation (Vest®)

Mucus Mobilization

- Physiotherapy
  - Intrapulmonary percussive ventilation
  - High frequency chest wall oscillation (Vest®)
- Phamacologic
  - Mucus hydration
    - Saline – 0.9, 3, 7, 10%
  - Mucolytics
    - Dornase alpha (Pulmozyme®)
    - N-acetylcysteine

Intrapulmonary Percussive Ventilation

Percussionaire Corporation

High Frequency Chest Wall Oscillation

- Creates high frequency pules of air
  - Shears mucus from airways
  - Decrease mucus viscosity
- Distal airway clearance
- Requires intact cough

Baxter Healthcare

How to support the patient?

- Pharmacology
  - Too much mucus
    - Adrenergic agonist – decongestant
    - Antihistamine – allergies
  - Too much saliva
    - Anticholinergic
      - Glycopyrrolate / Atropine
        - Liquid
      - Scolpolamine
        - Patch
    - Botulinum toxin
    - Salivary gland ligation

Management principles and challenges

- Perfection is not practical, manageable is
  - Supplement residual function – Suction / medications
  - Dynamic plan
    - Start / increase therapy early
    - De-escalate / return to baseline therapy promptly
    - Avoid polypharmacy!
- Optimize quality of life
  - Medical stability
  - Reduce risk of infection
  - Optimize speech
  - Minimize drooling
  - Based on parental / patient assessment!