

**“I Need to Cough”**

**Ways to Keep Your Airways Clear**

**2018 Annual Cure SMA Conference**



**Make today a breakthrough.**

# Presenters

## Richard Kravitz, MD

Duke University School of Medicine  
Duke University Medical Center  
Durham, North Carolina



## Jane Taylor, MD

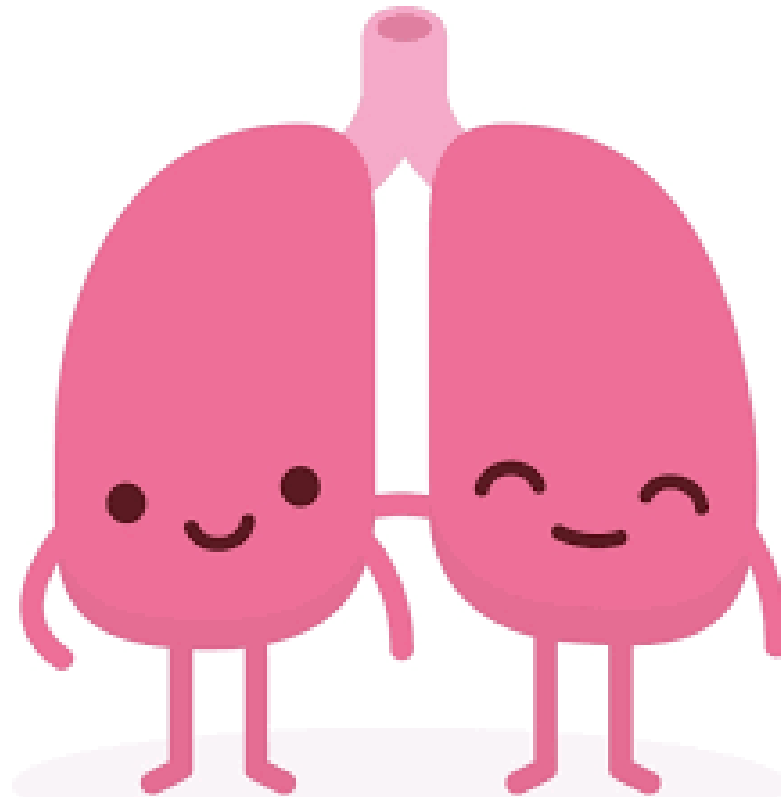
University of Missouri-KC  
School of Medicine  
Children's Mercy Kansas City  
Kansas City, Missouri



# Goals of This Session

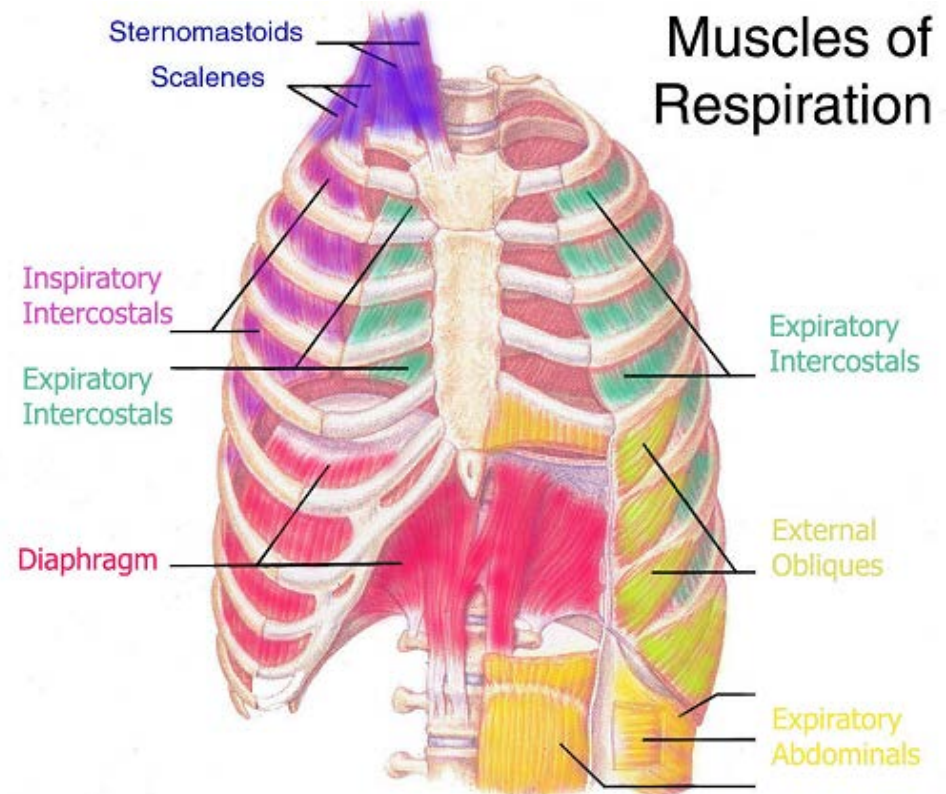
- Basics of respiratory dynamics / How the lungs keep themselves clean
- How SMA affects the respiratory system
- Assessing and monitoring your respiratory status
- Optimizing airway clearance
- Treating acute respiratory illness
- Maintaining wellness
- Preparing for the present and future

# Respiratory Physiology (Welcome to RT School)



# The Muscles of Respiration

- **Inspiration**
  - Muscles that expand the thoracic cavity
    - Diaphragm
    - External intercostals
    - Accessory muscles
- **Expiration**
  - Muscles that compress the thoracic cavity
    - Mainly elastic recoil of chest wall – no muscle involvement
    - Internal intercostals
    - Abdominal muscles



Mechanics of respiratory muscles - ScienceDirect

# The Muscles of Respiration

## Muscles of inspiration

### Accessory

Sternocleidomastoid  
(elevates sternum)

Scalenes Group  
(elevate upper ribs)

Not shown:  
Pectoralis minor

### Principal

External intercostals  
Interchondral part of  
internal intercostals  
(also elevates ribs)

Diaphragm  
(dome descends, thus  
increasing vertical  
dimension of thorac  
cavity; also elevates  
lower ribs)



## Muscles of expiration

### Quiet breathing

Expiration results from  
passive, elastic recoil  
of the lungs, rib cage  
and diaphragm

### Active breathing

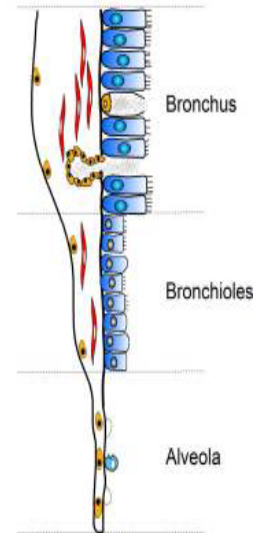
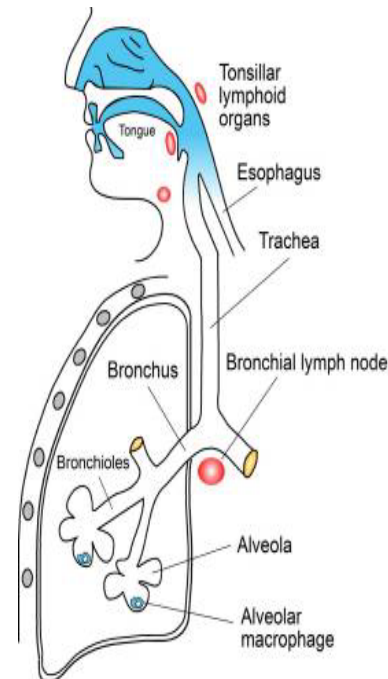
Internal intercostals,  
except interchondral  
part (pull ribs down)

Abdominals  
(pull ribs down,  
compress abdominal  
contents thus pushing  
diaphragm up)

Note shown:  
Quadratus lumborum  
(pulls ribs down)

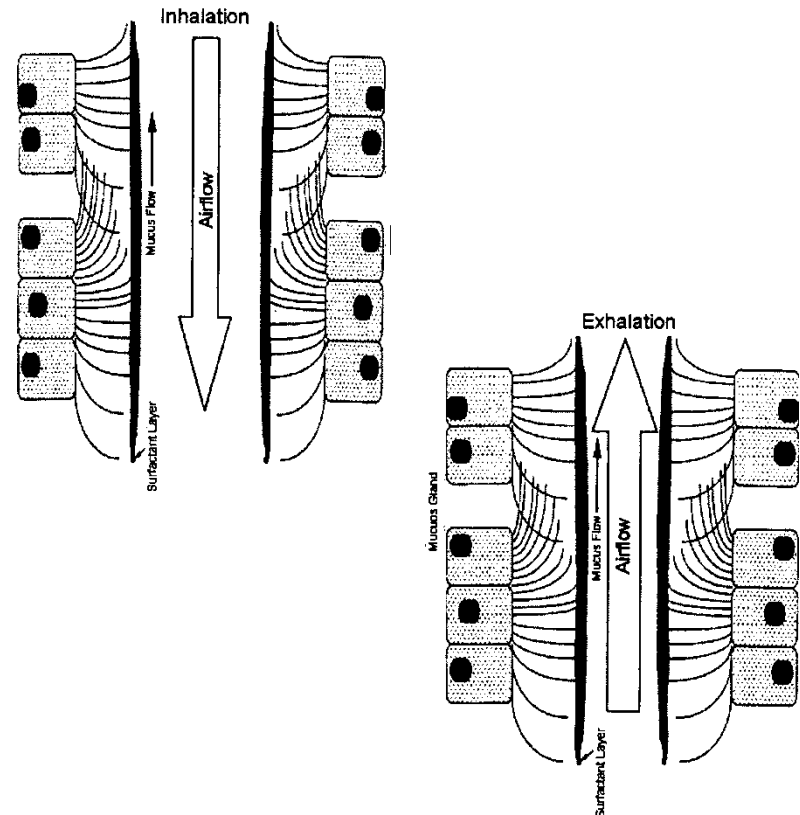
# Normal Airway Clearance

- Conducting airways
  - Mucociliary elevator
  - Airflow
  - The part most affected in SMA
    - Cephalad airflow bias
    - Cough
- Respiratory zone
  - Chemical absorption
  - Alveolar macrophages
  - Less of an issue in SMA



# Cephalad Airflow Bias

- Decreased airway diameter during exhalation results in increased flow velocity
- Increased airflow velocity shears secretions and drives material in direction of flow
- Present in large and small airways but is the primary mechanism of transport in smaller airways



## Consensus Statement for Standard of Care in Spinal Muscular Atrophy

Ching H. Wang, MD, PhD, Richard S. Finkel, MD, Enrico S. Bertini, MD, Mary Schroth, MD, Anita Simonds, MD, Brenda Wong, MD, Annie Aloysius, MRCSLT, HPC, Leslie Morrison, MD, Marion Main, MCSP, MA, Thomas O. Crawford, MD, Anthony Trela, BS, and Participants of the International Conference on SMA Standard of Care

### Encompasses:

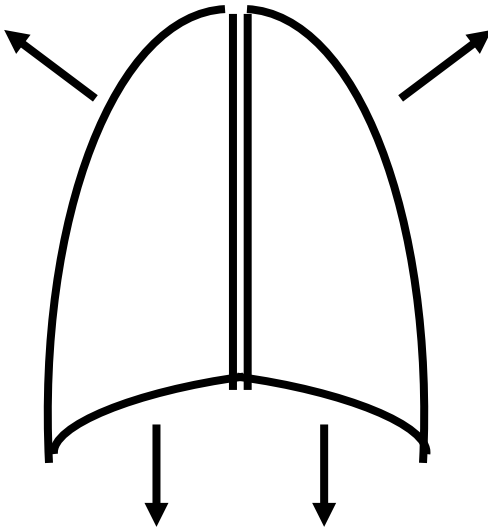
- **Diagnosis**
- **Respiratory Care**
- **GI and Nutrition**
- **Orthopedic Concerns**
- **Palliative Care**

# SMA Breathing Basics

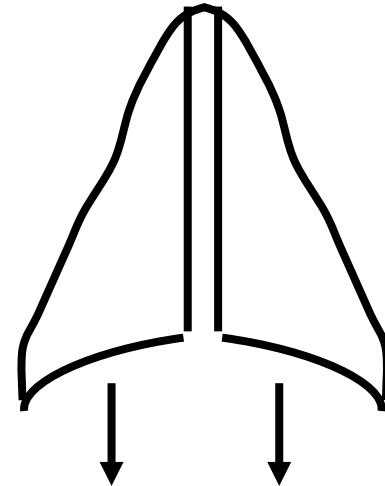
- SMA type I and II
  - Weak muscles between the ribs
  - Diaphragm: primary muscle used to breath
  - Cough is weak in type I and more variable in type II
- SMA type III
  - Cough is usually well preserved
  - May get weaker with age
- All coughing is weaker when ill

# Chest Wall Changes

Normal



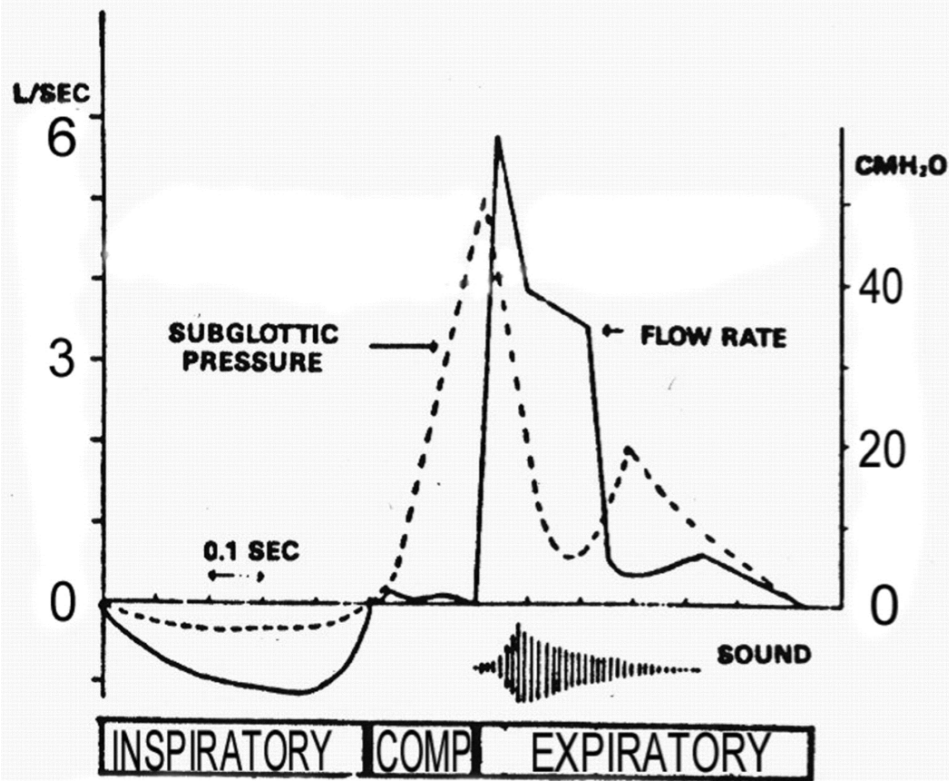
SMA



# Respiratory Symptom Progression

- Normal (asymptomatic)
- Weakened cough
  - Usually adequate when well
  - Problems develop with infections
  - Can lead to (recurrent) pneumonia/bronchitis
- Nighttime symptoms
- Daytime symptoms

# Cough Mechanism



1. Stimulus
2. Deep inhalation
3. Closure of vocal cords
4. Compression of the chest against closed vocal cords
5. Vocal cords open to release air = audible cough

# What Makes a Cough Weak?

- Inadequate inspiration (need to get to 85-90% of Lung Capacity)
  - Inherent muscle weakness
  - Muscle fatigue
  - Scoliosis
  - Chest wall stiffness
- Incomplete closure of vocal cords
  - Not usually a SMA problem unless a tracheostomy is needed
- Inadequate expiration (need cough flow rate > 6L/min)

Same problems with inspiration

**PLUS**

Diminished Inspiratory Capacity → Mechanical disadvantage

- Muscles not at point of maximal capacity for active contraction
- Lung not at point of maximal (passive) elastic recoil
- Airway diameter not fully maximized

# Assessment and Monitoring of Respiratory Status

- Medical History
- Physical Exam
- Pulmonary Function Tests
  - Spirometry
    - Both upright and supine
  - Lung volumes
  - Muscle force testing (MEP and MIP)
  - Cough Peak Flow
- Oxygen saturation
- Carbon dioxide levels

# Cough Peak Flow Maximal Expiratory Pressure

- Measure respiratory muscle strength
- CPF Ranges:
  - Normal cough >360 L/min
  - Adequate cough >270 L/min
  - Low cough <160 L/min
- MEP Ranges:
  - Adequate cough > 60 cm H<sub>2</sub>O
  - Weak cough < 45 cm H<sub>2</sub>O



## OTHER FACTORS AFFECTING AIRWAY CLEARANCE

- Nutrition
- Sleep
- Scoliosis
- Aspiration
- Gastroesophageal reflux
- Constipation

# Assessment and Monitoring of Respiratory Status – Additional tests

- Chest x-ray
- Swallow study (with speech consultation)
- Reflux evaluation
- Polysomnography (sleep study)
- Scoliosis x-rays

# Results of Respiratory Muscle Weakness in SMA

- Difficulty coughing
- Small shallow breaths during sleep: hypoventilation
- Chest wall and lung underdevelopment (type I and weak type II)
- Difficulty getting over respiratory infections that contribute to muscle weakness

# Chronic Management Goals

- Normalize oxygen saturation and CO2 levels
- Improve sleep
- Facilitate care at home
- Have a plan for illnesses
- Decrease hospitalizations and PICU stays
- Decrease burden of illness on families

# Interventions to Aid Airway Clearance

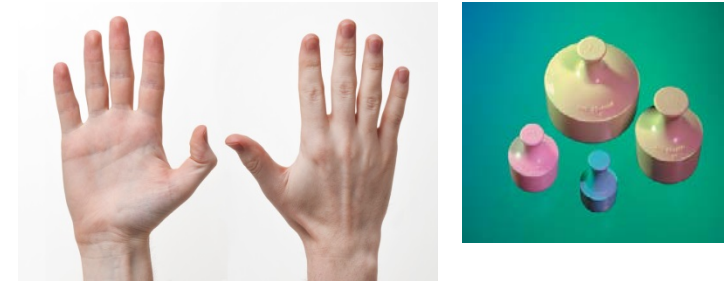
- **Cough augmentation – Priority #1**
  - Inspiratory phase; contraction phase; expiratory phase
- **Mucociliary clearance** (mechanical, if needed)
  - Manual chest physiotherapy
  - Intrapulmonary percussive ventilation (IPV)
  - High frequency chest wall oscillation (***The Vest***)
  - (bronchoscopy)
- **Mucociliary clearance** (medical, if needed)
  - Albuterol, Pulmozyme, 3% hypertonic saline, Mucomyst
  - Avoid drying agents (if possible)
  - Asthma medications in select cases

# Factors Affecting Secretions

- Quantity of secretions
  - Normal versus increased amount
  - Ability of cough in removing this “normal” amount of secretions
- Quality of secretions
  - Normal (watery) versus sticky
  - Infection (viral versus bacterial)

# Mechanical Mobilization of Secretions

- Helpful in SMA
  - Manual chest physiotherapy
  - High frequency chest wall oscillation (**The Vest**)
  - Intrapulmonary percussive ventilation (IPV)
  - [Bronchoscopy]



- Less helpful in SMA
  - Positive Expiratory Pressure
  - Acapella / Flutter



- Must be followed by cough augmentation

# Improving a Cough

- **Inspiratory** component
  - Glossopharyngeal breathing (breath stacking) (II, III)
  - Positive pressure assist (ambu bag) (I, II, III)
  - Intermittent positive pressure ventilation (IPPV) (I, II, III)
  - Mechanical ventilator (I, II, III)
- **Expiratory** component
  - Abdominal/chest wall thrust (less effective with scoliosis)
  - Most useful in SMA II and III
- **Combined**
  - Mechanical in-exsufflator (aka Cough Assist)** (I, II, III)

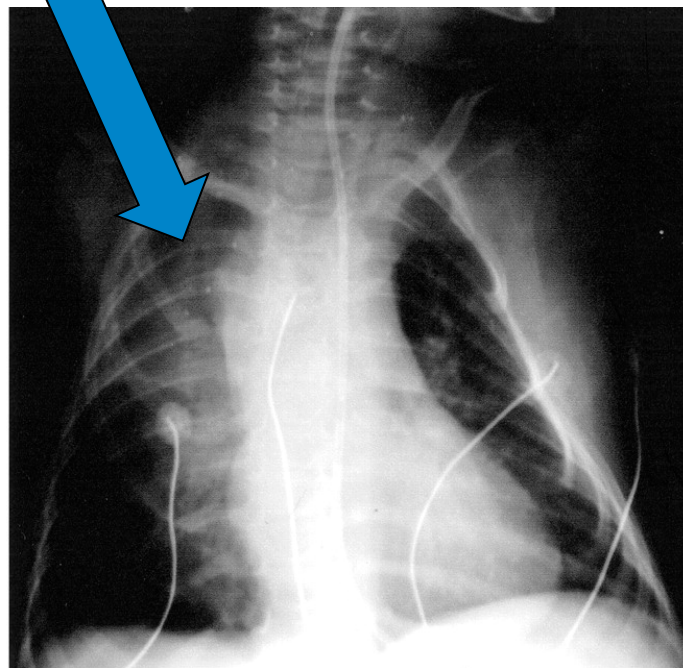
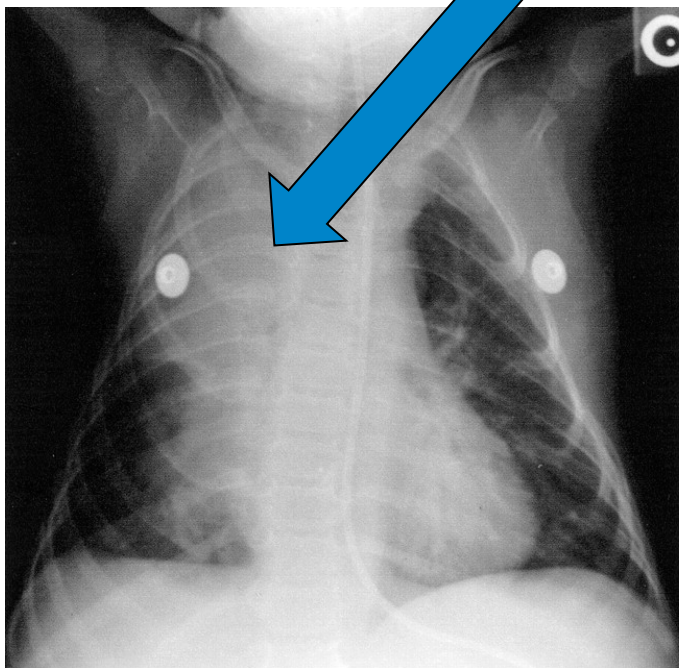
# Mechanical In-exsufflator (The Cough Assist™)

- Well tolerated:
  - No gastroesophageal reflux or aspiration
  - No pneumothorax (collapsed lung)
- Main complaints: “I just don’t like doing it!”
  - Interface discomfort
  - Pressures uncomfortable
  - Chest/abdominal discomfort
  - No perceived benefit
- **Most effective when used regularly**  
(versus only when ill)
  - Takes some practice, but few complaints
  - Once used to it, most kids will start to ask for it when ill!

# The Cough Assist™



# Chest radiograph of 22-month-old with SMA type I

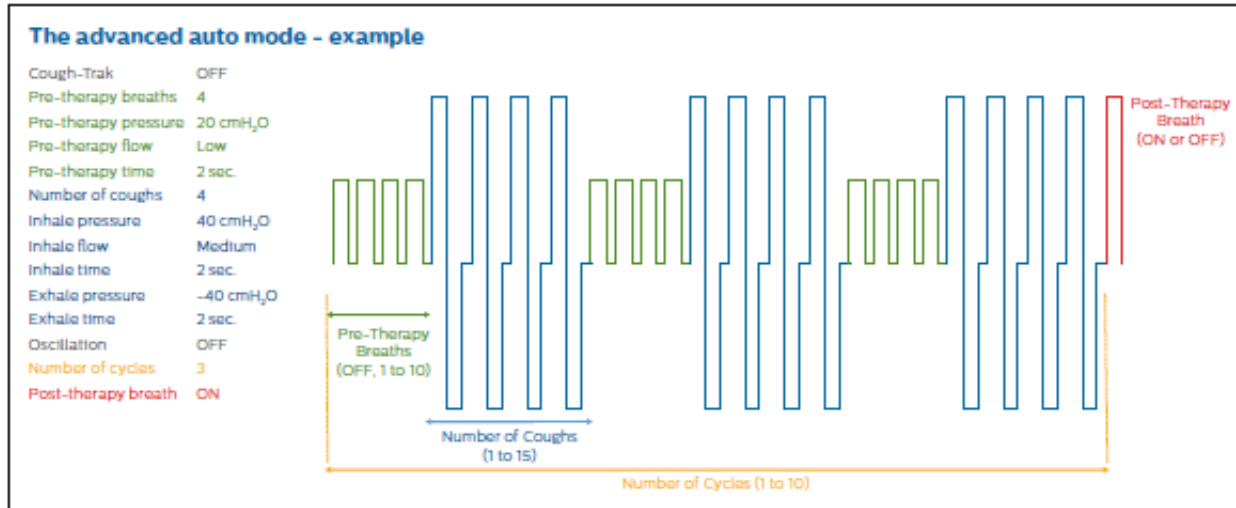


# The Cough Assist™

- Settings for use:
  - Inspiratory:
    - visually good lung expansion (up to +40 cm H<sub>2</sub>O)
    - Over 2-3 seconds
  - Expiratory:
    - start at -15 cm H<sub>2</sub>O (up to -40 cm H<sub>2</sub>O)
    - Over 2 seconds
  - Pause: 1-2 seconds between cycles
  - Optimal pressures +/-40 cm H<sub>2</sub>O
  - Manual versus Automatic versus Cough Trak
  - Oscillatory settings (T70 / E70)
    - Amplitude (between 1-10)
      - Start at a low amplitude and titrate up as tolerated
    - Frequency (1-20 Hz)
      - Start at a higher frequency and titrate down as tolerated
    - Inspiratory and/or Expiratory

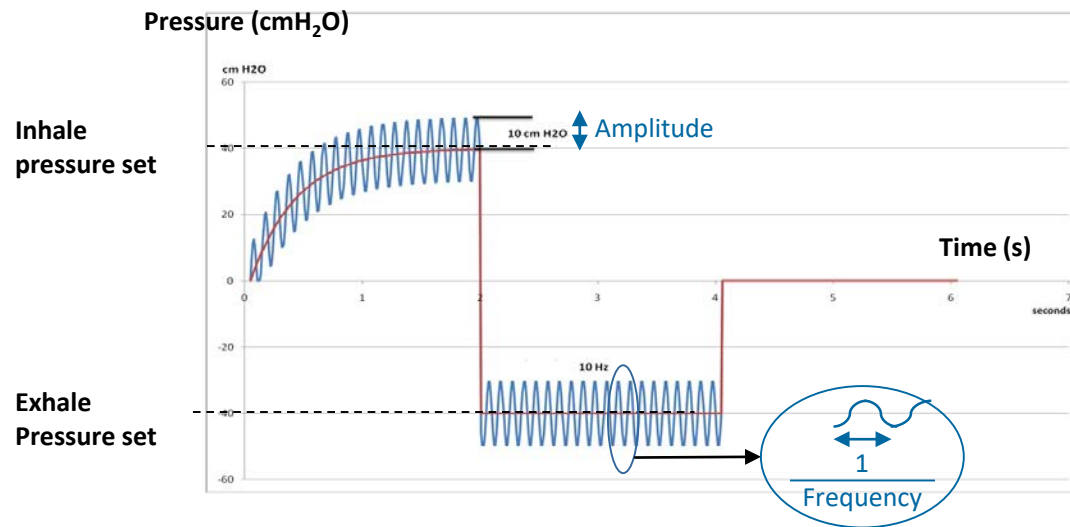
# The Cough Assist™

- Cough Assist



# The Cough Assist™

- Oscillatory Cough Assist



# The Cough Assist™

- Number of cycles:
  - 3-5 breaths/cycle
  - 3 to 5 cycles per treatment (as tolerated)
- Frequency of use:
  - When well: once daily to twice a day (as needed)
  - When ill: as often as necessary
- Suction
  - Have the suction machine on / ready
  - Suction in-between cycles and as needed
  - Avoid deep suctioning
    - Laryngospasm
    - Risk of vasovagal event
    - Risk of inflammation/scarring the back of throat / airway

# The Cough Assist™

- Various interfaces acceptable (full face mask; mouth piece; endotracheal tube; tracheostomy tube)
- Observe for transient O<sub>2</sub> desaturations with use (suggests mucus mobilization)



# Pulse Oximetry

- Normal is  $\geq 95\%$  or greater
- $< 95\%$  suggests:
  - Mucus plugging (needs more airway clearance)

Shallow breathing (awake and/or asleep)

Stronger patients  
May not need this  
monitoring



# How the Common Cold Makes Children with SMA Sick

- **Increased secretions**

- Worsening of (already impaired) swallowing function
- Increased risk of aspiration

- **Weakened cough**

- Diminished ability to clear secretions
- Increased risk for mucus plugging, atelectasis, and pneumonia
- Hypoxia

- **Further weakening of muscle strength**

- Vicious cycle leading to more aspiration and further weakening of an already diminished cough
- Higher risk for inadequate nighttime breathing and hypoxia

# How to Treat the Common Cold

- **Treat the increased secretions**
  - Good hydration
  - Suctioning (if necessary)
  - Avoid drying agents/anti-histamines (if possible)
- **Treat the weakened cough**
  - Increased use of cough assisting maneuvers (i.e.: the Cough Assist)
  - You can do this as often as needed (up to every 10-15 minutes)
  - Maintain normal oxygenation ( $\text{SaO}_2 \geq 95\%$ ) **Avoid supplemental  $\text{O}_2$ !**
  - Assisted ventilation may be needed
  - Judicious use of antibiotics
- **Treat the worsened muscle strength**
  - The patient may require increased use of NIPPV (both while awake and asleep) possibly this should be done in the hospital
  - ***Watch for worsening nocturnal hypoventilation***

# Don't Forget – Routine Medical Care

- Routine health care
- Immunizations
  - Usual immunization
  - Influenza vaccination
  - Pneumococcal vaccination
  - Palivizumab (Synagis)
- Proper nutrition
  - Obesity versus malnutrition
  - Oral versus tube (versus both)

# ISSUES TO ADDRESS AT FOLLOW UP APPOINTMENTS

- Pulmonary (Do I need more support?)
- Sleep (Do I need a Sleep Study?)
- Nutrition and GI status (Should I see a Dietitian?)
- Well child care (Are my shots up to date?)

# MULTIDISCIPLINARY HEALTH CARE TEAM (It Takes a Village...)

- Pulmonologist
- Neurologist
- Orthopedist
- 1<sup>o</sup> care physician
- Sleep physician
- Nursing
- Nutritionist
- Respiratory therapist
- Physical therapist
- Occupational Therapist
- Speech Therapist
- Social work
- Palliative care

# Recommended Home Equipment

- Mechanical in-exsufflator (Cough Assist™)
- Pulse oximeter (for monitoring)
- Suctioning equipment
- Ambu bag (if needed)
- Chest cups or high frequency chest wall oscillation (if needed)
- BiPAP or ventilator (if needed)
- Nebulizer (if needed)

# Going to the ER or having surgery?

- **Bring your respiratory equipment:**
  - Cough Assist with tubing
  - BiPAP with mask and tubing or ventilator
  - Suction
- Tell your SMA team!
- If you have one, bring your written sick plan or passport
- Bring your advance directives

# How Does Spinraza Change All This?

- Real answer: We don't know. Let's find out together.
- Don't stop respiratory therapies abruptly, you may still need them even if you are getting stronger.
- Talk to your SMA team about your choices.

# Things to Remember

- Every person is unique
- You are your/your child's best advocate
- The decisions you make regarding your/your child's care are the correct ones for your family
- **You are the captain** of the team caring for you/your child, so the call is yours. Don't be afraid to ask for input and be open to your health care team's recommendations
- You are not alone. There are many support groups out there to help you. Get involved!

Remember, **ANYTHING** is Possible...

