Breathing Strong All Day Long

2017 Annual Cure SMA Conference
Walt Disney World, Orlando Florida
June 29-July 2, 2017
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Objectives

• Understand how SMA affects breathing
• Learn techniques to help your child’s breathing
• Understand Non-Invasive and Invasive ventilation
• Understand the impact a cold can have on breathing
• Understand that even with the therapies available now and in the future we need to focus on providing the care these children need now
Encompasses:

• Diagnosis
• Respiratory Care
• GI and Nutrition
• Orthopedic Concerns
• Palliative Care

SMA type I / Nonsitters

• Weak intercostal muscles
• Chest wall: very soft and flexible during the first year of life
• Diaphragm: easily fatigued, the primary muscle for breathing
• Other complications:
  • Dysautonomia
  • Dysphagia with aspiration
  • Scoliosis, joint contractures
  • Fatty acid oxidation metabolic disorder
  • Poor bone quality – increased fracture risk
  • Intermittent gastroparesis
SMA type II / Sitters

• Range of respiratory muscle weakness
  • Weak intercostal muscles
  • Chest wall: rib collapse over time (parasol deformity)
  • Diaphragm: fatigue and the primary muscle for breathing

• Other complications:
  • Some develop dysphagia – can occur in teens
  • Scoliosis, joint contractures
  • Fatty acid oxidation metabolic disorder
  • Poor bone quality – increased fracture risk
  • Chronic pain
Chest Wall Changes

Normal

SMA 1 and some type 2

SMA type III / Walkers

- Generally normal pulmonary function tests
- At risk for:
  - Obstructive sleep apnea
  - Respiratory muscle weakness in adolescence and adulthood
  - Respiratory compromise with anesthesia, narcotic use, illness
- Other complications:
  - Obesity
  - Scoliosis, joint contractures
  - Mild fatty acid oxidation metabolic disorder
  - Chronic pain
Results of Respiratory Muscle Weakness in SMA

1. Difficulty coughing and mobilizing secretions
2. Small shallow breaths during sleep: hypoventilation
3. Chest wall and lung underdevelopment
4. Recurrent infections that contribute to muscle weakness.

Normal breathing

Respiratory and bulbar muscle weakness

REM related sleep disordered breathing

Ineffective cough reduced peak cough flows

NREM and REM sleep disordered breathing

Swallow dysfunction

Chest infections

Daytime ventilatory failure

Physical examination

Pulmonary function, peak cough flow, respiratory muscle strength

Chest xray, Sleep study

Swallow function evaluation

Airway clearance with cough assistance

Nocturnal non-invasive ventilation

Nocturnal or continuous non-invasive ventilation

Death

FRC Relative to Position for Diaphragm Dependent Breathers

From Nunn’s Applied Respiratory Physiology, 2000
Measurements of Breathing Muscle Function

**Everyone**
- $O_2/CO_2$ analysis
  - Blood gas
  - Blood bicarbonate level
  - Pulse oximetry
  - Exhaled CO2
- Chest xray
- Overnight oxygen saturation and carbon dioxide levels
- Sleep Study

**Stronger children**
- Pulmonary function testing
  - Spirometry
  - Lung volume
- Respiratory muscles tests
  - Maximal Expiratory Pressure
  - Maximal Inspiratory Pressure
- Cough Peak Flow
Breathing is Affected by Secretions

- Quantity of secretions
  - Normal versus increased amount
  - Ability of cough to remove a usual amount of secretions

- Quality of secretions
  - Normal (watery) versus sticky
  - Infection (viral versus bacterial)
Pulse Oximetry

- Normal is ≥ 95% or greater
- < 95% suggests:
  - Mucus plugging (needs more airway clearance)
  - Shallow breathing (awake and/or asleep)
Other Factors Affecting Airway Clearance

• Aspiration
  Impaired swallowing
  Impaired protective reflexes

• Gastroesophageal reflux
  Decreased lower esophageal muscle tone
  Scoliosis can make the esophagus pass through the diaphragm at a bad angle, leading to reflux
  Poor stomach emptying
Respiratory Symptom Progression and Interventions

- Normal (asymptomatic)
  - No treatment needed

- Weakened cough
  - Assist in airway clearance (required with viral illnesses)
    - Assisted cough
    - Chest physiotherapy
  - May require ventilation (if low $O_2$ levels or in respiratory distress)

- Nighttime symptoms
  - BiPAP or volume ventilator when asleep

- Daytime symptoms
  - Continuous ventilation
  - Non-invasive ventilation versus tracheostomy (in select patients)
Breathing Support Approaches

• **Non-invasive ventilation**
  
  BiPAP or Ventilator
  
  Interface: Mask over the nose

• **Invasive ventilation**
  
  Ventilator
  
  Interface: Tracheostomy
GOALS OF VENTILATORY SUPPORT

- Improve oxygen and carbon dioxide levels
  Ventilation and Oxygenation

- Muscle rest

- Treatment of pectus excavatum

- Compensate for worsening strength when having a respiratory illness

- Quality of life and comfort

- Preventative care vs acute care
Chest Wall Development After NIV

6 mths

18 mths

Courtesy of A. Simonds, Royal Brompton Hospital, UK
Positive Pressure Ventilation Devices

1. Bilevel positive airway pressure devices
   Non-invasive only

2. Home mechanical ventilators

CPAP is NOT INDICATED for Neuromuscular hypoventilation
Positive Pressure Ventilation

• Goals during **sleep**:  
  – Respiratory muscle rest  
  – Synchronization  
  – Chest wall expansion

• Recommended modes:  
  – PC (Pressure control) guaranteed inspiratory time with back up rate  
  – ST (spontaneous timed) with back up rate  
  – AVAPS (average volume assured pressure support) targeted tidal volume within IPAP range

• Backup respiratory rate required
Non Invasive Bilevel Positive Airway Pressure

- IPAP: 14-20 cm of H₂O
- EPAP: 4-6 cm of H₂O
- Respiratory Rate: high enough to capture breathing efforts and rest.
- Inspiratory Time: depends on age and set respiratory rate
- Rise time: time between exhalation and rise to peak inspiratory pressure (IPAP)
Invasive Bilevel Positive Airway Pressure with Home Mechanical Ventilator

• Ventilation is more efficient
• Chest wall rise on inhalation (resting/sleep)
• Modes:
  – Assist control (Every breath is the same)
    • Pressure or volume ventilation
  – Synchronized intermittent mechanical ventilation (SIMV)
    • Pressure or volume ventilation
    • May not be tolerated as well during sleep due to hypoventilation – triggering breath is work
Breathing Assist (Interfaces)

• Non-invasive Ventilation
• Invasive Ventilation

*Pictures were removed until consent is obtained.*
Nasal Masks

Respironics Wisp Pediatrics

Infant to 2 yo:
- ResMed Pixi
- Sleepnet MiniMe 2
- ResMed Wisp
- Fisher & Paykel Eson
- Fisher & Paykel Zest Q Petite

Over 2 years old:
- AG Industries Nonny, Size Small Child, AG-PEDKIT-S

*Pictures were removed until consent is obtained.*
Positive Pressure Ventilation
Non-Invasive

• Advantages
  – Non-invasive
  – Reversible
  – Safe
  – Effective
  – Preserves (natural) airway protection mechanisms
  – Decreased risk of infection
  – Comfort
  – Preserves speech
  – Less swallowing issues
NIPPV Complications

- Nasal/oral dryness
- Bloody nose
- Nasal congestion
- Sneezing
- Runny nose
- Sinus Infection

- Claustrophobia
- Mask irritation
- Swallowing air
- Mouth leak
- Facial deformities
Invasive Respiratory Care

- Intubation (in an acute illness)
- Tracheostomy with ventilator support

*Pictures were removed until consent is obtained.*
Tracheostomy: Indications

• NIPPV no longer effective at treating hypoventilation

• NIPPV not tolerated by the patient

• Excessive oral secretions puts patient at risk for aspiration

• Care for patient with NIPPV beyond the capabilities of the family

• Resources for outpatient management with NIPPV not available in the community

• Failure to extubate a patient who has been intubated (for whatever reason). This assumes proper methods of extubation were employed
Tracheostomy: Advantages for the Patient

- Allows for assisted ventilation
- Airway is protected from aspiration
- Increased ease in suctioning excessive secretions (if needed)
- No NIPPV mask pressure sores or facial remodeling
- Treats obstructive sleep apnea (if present)
- In select cases, may be a temporizing measure until extubation is possible
- Reversible (trach can come out)
Tracheostomy: Complications

- Infections
- Bleeding
- Irritation/granulation tissue
- Accidental dislodgement
- Skin yeast infection
Key Points Regarding a Tracheostomy

• Elective tracheostomy is better than emergency tracheostomy
• Long-term treatment plan is best determined in times of health. The middle of an acute decompensation (especially one which was predictable) is not the best time to be making these types of decisions
• Quality of life should be considered in the decision making
• Reversibility
How the Common Cold Makes Children with SMA Sick

• Increased secretions
  Worsening of (already impaired) swallowing function
  Increased risk of aspiration

• Weaker 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 ($\text{SaO}_2 \geq 95\%$) \textbf{Avoid supplemental $O_2$!}
  - 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)
  - \textit{Watch out for worsening nocturnal hypoventilation}
  - \textit{Good nutrition}
A moment about Oxygen
Issues to Address at Follow up Appointments

- Pulmonary
- Sleep
- GI status
- Well child care
Palliative Care
Quality of Life Care

• Recognizes that SMA can be a life threatening disease

• Should be part of the care of the disease process

• Goals
  Provide symptom relief: pain, dyspnea, agitation, nausea, anxiety
  Provide psychological, social and spiritual support for patient and family

• Palliative care can be in the hospital, in the home, or both

• *Palliative care is not giving up; it is just changing the focus of therapy*
Well Child Care

• Routine health care

• Immunizations
  Standard immunizations
  Influenza vaccination yearly
  Pneumococcal vaccination
  Palivizumab (Synagis)

• Proper nutrition
  Obesity versus malnutrition
Equipment Recommended for the Home

- Mechanical in-exsufflator (The Cough Assist)
- Ambu bag
- Pulse oximeter
- Chest cups or high frequency chest wall oscillation (if needed)
- Suctioning equipment
- Nebulizer (if needed)
- BiPAP or volume ventilator (when needed)
MULTIDISCIPLINARY Health Care Team

- Pulmonologist
- Neurologist
- Cardiologist
- Orthopedist
- Sleep physician
- 1º Care physician
- Nursing
- Nutritionist
- Respiratory therapist
- Physical therapist
- Occupational Therapist
- Speech Therapist
- Social work
- Palliative care
Things to Remember

- Every child is unique
- You are your child’s best advocate
- The decisions you make regarding your child’s care are the correct ones for your family
- You are part of team caring for your child (actually, you are the captain, so the call is yours), but 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…
SMA Conference Survey

Please complete your conference survey at this link:
https://www.surveymonkey.com/s/2017SMAAnnualConference

Or fill out the paper survey in your conference folder.

- All participants who complete a survey by 10:30 am on Sunday June 19th, will have their name entered into a raffle for a brand new iPad!
- The winners will be drawn and announced on Sunday, June 19th at the Closing General Session/It’s a Wonderful Life.