The Challenge of Chronic Lung Disease in a Patient with a Primary Neurodisability

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Speaker Name: Robert Warren, MD

Disclosure of Relevant Financial Relationships
I have no financial relationships to disclose.

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I will not discuss off label use and/or investigational use in my presentation

Presentation Objectives

DEVELOPING THE RESPIRATORY CARE PLAN
It’s All About Secretions
The Pulmonary Composite
Progressive Pulmonary Dysfunction
Chronic Lung Disease of Neurodisability?
Bacterial Colonization of the Airway

Respiratory Therapy Equipment and Devices
The Caregiver: Psychological Characteristics-Education
Case Presentations: Yours and Ours
Schedule

- Pulmonary Dysfunction Concepts - 20 min.
- Q&A – 5 min
- Respiratory Care Plans – 25 min.
- Q&A and BREAK – 15 min
- Emphasis on the Caregiver – 15 min
- Q&A - 5 min
- Cases – Yours and Ours 35 min

Pneumonia in patients with neurodisability


Review article: 77% of deaths were a result of pneumonia

Respiratory problems in children with neurological impairment


“High prevalence of respiratory disease in children with severe disabilities”
- Recurrent aspiration
- Poor cough/airway clearance
- Respiratory muscle weakness
Pediatric Respiratory Reviews


Cerebral Palsy 1-500 live births. “The major morbidity and mortality associated with cerebral palsy relates to respiratory compromise”.

Markers of respiratory exacerbation risk in cerebral palsy


“Respiratory exacerbation found to be associated with GER and higher PaCO2 levels”

CONGENITAL NEUROLOGICAL DIAGNOSES

- Becker Muscular Dystrophy
- Cerebellar Atrophy
- Cervicothoracic Syringomyelia/syrinx
- CHARGE Syndrome
- Congenital Muscular Dystrophy
- Congenital Hypomyelination Syndrome
- CFC Syndrome
- Down Syndrome
- Duchenne Muscular Dystrophy
- Fredericks Ataxia
- Järcho-Levin Syndrome
- Neuronal Migration Disorder
- FSH Muscular Dystrophy
- Cockayne Syndrome
- Thoracic Dystrophy
- Merosin Deficient Muscular Dystrophy
- Myotonic Dystrophy
- Neiman Pick
- Partial Complex IV Mitochondrial Disease
- Pompe Disease
- Soto Syndrome
- Spina Bifida
- Spinal Muscular Atrophy Types I & II
- Undiagnosed Mitochondrial Disorder
- Undiagnosed Neurodegenerative Disorder
- X-linked Myotubular Myopathy
- Bethlehem Myopathy
- Marshall Syndrome
- Metachromatic Leukodystrophy
- Congenital Scoliosis
Acquired Neurological Diagnoses

- NICU
- Traumatic Brain Injury
  - Non-accidental
- Traumatic Brain Injury/
  Spinal Cord Injury

THE PULMONARY COMPOSITE

Refers to a group of clinical characteristics each of which make a contribution to either the accumulation, retention, or production of airway secretions which in turn forms the basis for the progressive pulmonary dysfunction in the neurodisability patient.

Chronic Pulmonary Disease in Neurodisability

IT’S ALL ABOUT SECRETIONS!!

Regardless of which Pulmonary Composite Factor is being considered, the key is how that factor influences either secretion production, accumulation, or retention

- Secretion Production
- Secretion Accumulation
- Secretion Retention
Pulmonary Composite Of Child With Neurodisability

- State of Ambulation
- Seizure Disorder
- Dysphagia with aspiration
- GER with aspiration
- Hypopnea
- Spasticity
- Hypotonia/Ineffective Cough
- Recurrent acute respiratory illness
- Sialorrhea – Excessive oral secretions

The Pulmonary Composite in Clinical Assessment

History – Using the Pulmonary Composite as a Guide

Physical Assessment: Visual observation, listen, auscultation

Laboratory Data:  
- Resting Tidal Volume
- End Tidal CO2
- Pulse Oximetry
- Blood Gas Analysis: ABG – VBG – CBG
- Transcutaneous O2 and CO2 determination

Radiology:  
- Chest imaging
- Sinus imaging

Use of the Pulmonary Composite

Taking the history

1. Ambulation
2. Seizures
3. Dysphagia
4. GER
5. Hypopnea
6. Spasticity
7. Hypotonia
8. Recurrent Respiratory Illness
9. Sialorrhea
Sources of Excessive Airway Secretions

- Ineffective cough
- Airway hyposensitivity
- Aspiration
- Dysphagia
- GER
- Airway inflammation
- Infection
- Mucosal gland proliferation and hypertrophy

Evolution Of Chronic Lung Disease In Children With Neurodisability

- Impaired cough and retained secretions
- Aspirated secretions from dysphagia and GER
- Respiratory infection / atelectasis
- Bronchiectasis / pulmonary dysfunction
- Diminished oxygenation
- Alveolar hypoventilation and CO₂ retention

Airway Bacterial Colonization in the Neurodisability Patient

- Retrospective study
- Total of 93 subjects studied

Table 3: Prevalence of pathogens isolated from respiratory cultures of all subjects at any time since trach placement, N=93

<table>
<thead>
<tr>
<th>Organism</th>
<th>Prevalence</th>
<th>N (%)</th>
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</thead>
<tbody>
<tr>
<td>Pseudomonas aeruginosa</td>
<td>84</td>
<td>90.3</td>
</tr>
<tr>
<td>Stenotrophomonas maltophilia</td>
<td>72</td>
<td>77.4</td>
</tr>
<tr>
<td>Serratia marcescens</td>
<td>62</td>
<td>66.7</td>
</tr>
<tr>
<td>Moraxella catarrhalis</td>
<td>57</td>
<td>61.3</td>
</tr>
<tr>
<td>MRSA</td>
<td>52</td>
<td>55.9</td>
</tr>
<tr>
<td>MSSA</td>
<td>43</td>
<td>46.2</td>
</tr>
<tr>
<td>Haemophilus influenza</td>
<td>41</td>
<td>44.1</td>
</tr>
<tr>
<td>Group B streptococcus</td>
<td>32</td>
<td>34.4</td>
</tr>
<tr>
<td>Streptococcus pneumoniae</td>
<td>26</td>
<td>28.0</td>
</tr>
<tr>
<td>Acinetobacter baumanii</td>
<td>24</td>
<td>25.8</td>
</tr>
<tr>
<td>Acinetobacter calcoaceticus</td>
<td>13</td>
<td>14.0</td>
</tr>
<tr>
<td>Serratia liquefascins</td>
<td>9</td>
<td>9.7</td>
</tr>
<tr>
<td>Haemophilus parainfluenza</td>
<td>7</td>
<td>7.5</td>
</tr>
</tbody>
</table>

Organism Prevalence and placement of tracheostomy

**Figure 1**: Organism Prevalence in 2010 based on length of time with tracheostomy (N=90)

Chronic Lung Disease of Neurodisability

A real entity?

If so, what is the etiology?
Identifying the Presence of a Specific Chronic Lung Disease of Neurodisability
IRB study 203740 Warren/Jones

Study: Patients with primary neurodisability diagnosis and NO pulmonary disease at initial diagnosis.

Hypothesis: Pulmonary Composite factors produce chronic airway secretion overload with resultant progressive chronic lung disease over time.

Study Group: 136 patients age range 4 months to 18 years. Multiple neurodisability diagnoses represented.

Methodology:
Identify pulmonary composite factors at time of first visit to the pulmonary clinic.
Follow factor persistence and accumulation over time.
End points: Progression to pulmonary insufficiency and mechanical ventilation or end of study period whichever comes first

Methodology – Data Points:
1. First visit to pulmonary clinic
2. First increase in respiratory care plan
3. Second increase in respiratory care plan
4. Initial placement on ventilator support OR end of study period
Identify Pulmonary Composite Factors at each data point
Identifying the Presence of a Specific Chronic Lung Disease of Neurodisability
IRB study 203740

Clinical Questions:
- Role of pulmonary factors individually and collectively
- Individual contribution in order of prominence
- Relationship of number of factors and severity to speed of progression of disease

Preliminary Data
DATA BASE: 135 patients

- 59 subjects analyzed at this time
- 11 have progressed to ventilation
- 33 with multiple increases in RT plans
- 15 without increases in RT plans

Preliminary Data

Prevalence of Factors

Percent Prevalence

Percent Prevalence
Preliminary Data

Number of Pulmonary Composite Factors

- Ventilated
- Increased RT
- No Changes

Preliminary Data

Speed of Progression

- Ventilated
- Increased RT
- No Changes

BREAK

FIRST BREAK  FOR Questions/Answers/Other
The Individual Respiratory Care Plan

Step 1: Patient Assessment
Step 2: Medication/Device selection
Step 3: Determine Frequency of Therapy
Step 4: Ongoing Evaluation

Creating The Respiratory Care Plan: Individualization Is The Key

1. Selection of aerosol medications: Bronchodilators, decongestants, anti-inflammatories, mucolytics, antibiotics
2. Delivery systems: MDI & spacer or updraft nebulizer possibly including use of hyperinflation delivery
3. Assessment for airway clearance devices: Manual In-exsufflation, high frequency oscillation, intrapulmonary percussive ventilation
4. Ventilator support
Background

- Penny M. Overgaard, RN; Peggy J. Radford, MD, “High Frequency Chest Wall Oscillation Improves Outcomes in Children with Cerebral Palsy”, Chest. 2005;128
- Kathryn Fitzgerald, Jessica Dugre, Sobhan Pagala, Peter Homel, Michael Marcus, and Mikhail Kazachkov “High-Frequency Chest Wall Compression Therapy in Neurologically Impaired Children”, Respir Care, January 2014 59:1 107-112

Bronchodilators

Albuterol (Proventil, Ventolin, Proair)
Dosage: MDI – 2 puffs QID
Nebulizer – 0.03 mL / kg QID

Levalbuterol (Xopenex)
Dosage: MDI – 2-4 puffs Q 6 hours
Nebulizer - 0.63 mg every 6 hours

Ipratropium Bromide (Atrovent)
Dosage: MDI - 2 to 4 puffs QID
Nebulizer – 250 to 500 mcg QID

Anti-inflammatory

Fluticasone Propionate (Flovent)
Dosage: 88mcg to 880mcg BID

Budesonide (Pulmicort Respules)
Dosage: 0.25mg to 0.5mg BID nebulized

Beclamethasone Dipropionate (Qvar)
Dosage: 80mcg to 160mcg BID
Mucolytics

**Acetylcysteine** *(Mucomyst)*
Dosage: 2 mL of 10% or 20% in 3mL of saline. Always give bronchodilator concurrently

**Dornase alfa** *(Pulmozyme)*
Dosage: 2.5 mg/unit dose

**Sodium Bicarbonate** *(Na HCO3 8.4%)*
Dosage: 1 mL in 3 mL normal saline given up to 4 times a day

Antibiotics

**Tobramycin Inhalation (Tobi)**
Trade name: TOBI
Dosage: 300 mg nebulized BID

**Tobramycin** for injection
Dosage: 80 mg nebulized BID (albuterol given prior)

**Ceftazadime** *(Fortaz)*
Dosage: 1 or 2 grams nebulized BID

Non- Aerosol Medications

- Glycopyrrolate
- Scopolamine
- Botulinum toxin
- Ipratropium Bromide
Special Considerations

- Delivery Devices
  - Nebulization vs metered dose inhaler
  - Hyperinflation techniques
  - Holding chambers
  - Tracheostomy considerations

- Order of medications
- Financial/insurance

Delivery Devices

Order of Medications

- Bronchodilators
- Mucolytics
- Anti-inflammatories
- Antibiotics
Mucus Mobilization
Airway Secretion
Clearance

- Manual physical therapy techniques
- Mechanical devices
  - Vibrators
  - HFCWO
  - IPV
  - M/I-E

Airway clearance devices used by neurodisability patients

Special considerations
- Order of therapies
- Tracheostomy considerations
- Financial / insurance barriers
- Caregiver limitations
- Patient tolerance
- Ability to clear secretions after secretion mobilization techniques
Home Care

- Establish daily respiratory care plan
- Interaction with DME-Home Nursing
- Develop stable lines of communication

Psychological Questionnaires

- Big Five Inventory BFI (Personality)
- Social Problem Solving Inventory SPSI-R
- Connor-Davidson Resilience Scale CD-RISC
- Family Crisis Oriented Personal Scale F-COPES
- Coping Health Inventory for Parents CHIP
- Personal Health Questionnaire PHQ
- Short Form-12 Health Survey SF-12v2

BIG FIVE PERSONALITY INVENTORY

Classified caregivers into three groups:

1 – Resilient
2 - Overcontrolled
3 - Undercontrolled
Caregiver Prototypes

Resilient Characteristics

- Extrovert
- Openness
- Conscientiousness
- Agreeableness

Overcontrolled Characteristics

- More Introverted
- High Neurotic Tendencies

Undercontrolled Characteristics

- Tendency to Extraversion
- High Neurotic Tendencies
- Less Agreeableness
- Less Conscientiousness
Skills Validation Quality Improvement Project - Farrah Jones, RRT, CRA

ASSESSING THE CAREGIVER

Presently ongoing in caregivers of children with a primary neurodisability who have secondary acute and chronic pulmonary symptoms

Skills Validation Quality Improvement Project

- Notification of the caregiver prior to clinic visit
- Caregiver brings own equipment
- Perform skills validation at the end of the clinic visit
- Remedial education will be provided if needed
- Certificate of completion
- Pre and post surveys for caregiver feedback

Skills Validation Quality Improvement Project Preliminary Results

- 75% of participants had deficiencies in some area of home equipment use (set up, settings, technique, etc.)
- 88% would like to see SV for all respiratory equipment
- 56% reported an increase in confidence in their ability to use equipment
Case Study: 9 month old Caucasian male
Diagnosis: Neurodisability secondary to Kernicterus

- **Pulmonary Composite Factors**: Seizures, Dysphagia, Drooling, GER, Ineffective cough, extreme spasticity
- **Pulmonary Symptoms**: cough, airway secretion accumulation (audible airway noise)
- **Challenges to the Respiratory Care Management Plan**: resistance to all devices to deliver medications and airway clearance devices. Severe aversion to anything touching the face.

Building the plan

<table>
<thead>
<tr>
<th>COUGH</th>
<th>AIRWAY SECRETION ACCUMULATION</th>
<th>AVERSION</th>
</tr>
</thead>
</table>
- Medications
  - Albuterol & Atrovent MDI, Robinul
- Devices
  - CPT, MIE
- Frequency
  - Twice daily
Case study: 19 year old Caucasian female
Diagnosis: Lissencephaly

- **Pulmonary Composite Factors**: non-ambulatory, GER, dysphagia, seizures, hypotonia, hypopneic breathing pattern
- **Pulmonary Symptoms**: chronic hypoventilation, copious amount of thick secretions, colonization with *Pseudomonas aeruginosa*, occasional wheeze
- **Challenges to Respiratory Care Management Plan**: copious amount of secretions, insurance/financial barriers, large patient size, chronic bacteria colonization

Building the plan

**CHRONIC PULMONARY INSUFFICIENCY**  **BACTERIAL COLONIZATION**  **EXCESSIVE ORAL SECRETIONS**

- **Medications**
  - Albuterol, Pulmozyme (Mucomyst), Tobi (Fortaz), all with hyperinflation or IPV. Glycopyrrolate
- **Devices**
  - IPV (vest back-up), MIE
- **Frequency**
  - 2-4 times daily for AWC, PRN for antibiotic
- **Ventilation (continuous)**

Case study: AF 17 year old Caucasian female
Diagnosis: Merosin Deficient Muscular Dystrophy

- **Pulmonary Composite Factors**: GER, dysphagia, non-ambulatory, hypopneic breathing pattern
- **Symptoms**: chronic hypoventilation, but no other respiratory symptoms
- **Challenges to respiratory care management plan**: significant (100 degree) scoliosis with lung restriction, continuous NG tube, no tracheostomy
Building the plan

CHRONIC HYPOVENTILATION   HYPOPNEIC BREATHING
PATTERN 3.5mL/kg

- Medications
  - Albuterol
- Devices
  - MIE
- Frequency
  - As needed
- Ventilation
  - Continuous NIV with rotating mask styles

Some Conclusions

- Control of secretions is key to patient stability
- Caregivers: Key to successful respiratory care
- Multiple components to the respiratory plan
- Daily monitoring is required by the caregiver
- Treatment plans must be flexible
- Communication is paramount
- Patient stability results in caregiver stability

Goals for the Child with Neurodisability

- Provide clinical stability
- Prevent acute exacerbations of illness
- Reduce or halt the progression of pulmonary disease
- Reduce/prevent emergency room visits
- Reduce/prevent hospitalizations for acute pulmonary illness
- Allow the patient to be integrated into the family unit