Chronic Lung Disease in the Patient with Neurodisability: Concepts and Management Strategies

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

Disclosure of Off-Label and/or investigative uses:
I will not discuss off label use and/or investigational use in my presentation

Presentation Objectives

- Understand the pathophysiology of airway secretions in the patient with neurodisability and potential for progressive pulmonary dysfunction
- Understand the potential clinical application of the Pulmonary Composite
- Understand the basic principles of respiratory care medications and devices.
- See, through the use of case presentations, the need for individualization of the respiratory therapy management plan
Admission Criteria for the ACRTDC

- Child followed by Pulmonary Section
- Diagnosis of any type of neuromuscular disease, chronic lung disease and/or syndromes or conditions causing chronic respiratory insufficiency
- Requires ventilator support and/or tracheostomy and/or airway clearance device

MISSION STATEMENT
The Arkansas Center for Respiratory Technology Dependent Children (ACRTDC) is committed to providing specialized care to respiratory technology dependent children and enhancing quality of life through caregiver education, research and advocacy.

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
- Jarcho-Levin Syndrome
- Neuronal Migration Disorder
- Myotonic Dystrophy
- Cockayne Syndrome
- Thoracic Dystrophy
- FSH Muscular Dystrophy
- X-linked Myotubular Myopathy
- Bethlehem Myopathy
- Marshall Syndrome
- Metachromatic Leukodystrophy
- Congenital Scoliosis
- Merosin Deficient Muscular 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
- Acquired Neurological Diagnoses
- NICU
- Traumatic Brain Injury
  - Non-accidental
- Traumatic Brain Injury/Spinal Cord Injury
  - Accidental secondary to non-primary pulmonary condition
Sources of Excessive Airway Secretions

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

Airway Responses

- Decrease in airway sensitivity – Change in irritant fiber response
- Airway Inflammation
- Mucosal gland proliferation and hypertrophy
- Infection leading to airway and parenchymal damage

ORIGINATION OF AIRWAY SECRETIONS

- INTRODUCED into the airway
- PRODUCED in excess in the airway
- RETAINED in the airway
Pulmonary Composite Of Child With Neurodisability

- State of ambulation
- Seizure Disorder
- Dysphagia with secondary aspiration
- GER with secondary aspiration
- Hypopneic Breathing Pattern
- Spasticity
- Hypotonia/Ineffective Cough
- Recurrent acute respiratory illness
- Drooling – Excessive oral secretions

THE PULMONARY COMPOSITE

- Ambulation Retained/Produced
- Seizure Disorder Introduced
- Dysphagia Introduced
- GER Introduced

The Pulmonary Composite

- Hypopnea Retained/Produced
- Spasticity Introduced
- Hypotonia Retained/Produced
- Recurrent Illness Produced
- Excessive oral secretions Introduced
Clinical Application of the Pulmonary Composite

- Development of a SEVERITY SCORE
- Assign “severity score” to each component of the pulmonary composite
- Determine ability to decrease the “severity score” for each component

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

“Working up” the patient: 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
Airway Bacterial Colonization in the Neurodisability Patient

- Retrospective study
- Total of 93 subjects studied

Table 1: Demographics of study population, N=93

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>N (%) unless otherwise noted</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at time of trach, mean (SD), years</td>
<td>3.0 (4.3)</td>
</tr>
<tr>
<td>Length of time with trach, mean (SD) years</td>
<td>6.2 (4.7)</td>
</tr>
<tr>
<td>Sex</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>53 (57.0)</td>
</tr>
<tr>
<td>Female</td>
<td>40 (43.0)</td>
</tr>
<tr>
<td>Race/Ethnicity</td>
<td></td>
</tr>
<tr>
<td>White</td>
<td>58 (62.4)</td>
</tr>
<tr>
<td>African American</td>
<td>26 (28.0)</td>
</tr>
<tr>
<td>Hispanic</td>
<td>9 (9.7)</td>
</tr>
</tbody>
</table>

Table 2: Underlying diagnosis that led to tracheostomy, N=93

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Prevalence N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cerebral Palsy</td>
<td>24 (25.8)</td>
</tr>
<tr>
<td>Spina Bifida</td>
<td>5 (5.4)</td>
</tr>
<tr>
<td>Moebius Syndrome</td>
<td>3 (3.2)</td>
</tr>
<tr>
<td>Laryngomalacia</td>
<td>3 (3.2)</td>
</tr>
<tr>
<td>Head Injury</td>
<td>3 (3.2)</td>
</tr>
<tr>
<td>Diaphragm Paralysis</td>
<td>2 (2.2)</td>
</tr>
<tr>
<td>Mandibular Agenesis</td>
<td>2 (2.2)</td>
</tr>
<tr>
<td>Muscular Dystrophy</td>
<td>2 (2.2)</td>
</tr>
<tr>
<td>Organism</td>
<td>Prevalence</td>
</tr>
<tr>
<td>--------------------------------</td>
<td>------------</td>
</tr>
<tr>
<td>Pseudomonas aeruginosa</td>
<td>84</td>
</tr>
<tr>
<td>Stenotrophomonas maltophilia</td>
<td>72</td>
</tr>
<tr>
<td>Serratia marcescens</td>
<td>62</td>
</tr>
<tr>
<td>Moraxella catarrhalis</td>
<td>57</td>
</tr>
<tr>
<td>MRSA</td>
<td>52</td>
</tr>
<tr>
<td>MSSA</td>
<td>43</td>
</tr>
<tr>
<td>Haemophilus influenza</td>
<td>41</td>
</tr>
<tr>
<td>Group B streptococcus</td>
<td>32</td>
</tr>
<tr>
<td>Streptococcus pneumoniae</td>
<td>26</td>
</tr>
<tr>
<td>Acinetobacter baumanii</td>
<td>24</td>
</tr>
<tr>
<td>Acinetobacter calcoacetiuls</td>
<td>13</td>
</tr>
<tr>
<td>Serratia liqueficans</td>
<td>9</td>
</tr>
<tr>
<td>Haemophilus parainfluenza</td>
<td>7</td>
</tr>
</tbody>
</table>

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>N (%)</th>
<th>Mean time to isolation, Month (SD)</th>
<th>Mean time to isolation, Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>MSSA</td>
<td>22</td>
<td>5.2 (10.3)</td>
<td>0.2 – 41.7</td>
</tr>
<tr>
<td>MRSA</td>
<td>19</td>
<td>6.5 (7.5)</td>
<td>0.2 – 26.4</td>
</tr>
<tr>
<td>Pseudomonas aeruginosa</td>
<td>37</td>
<td>7.8 (7.2)</td>
<td>0.2 – 25.1</td>
</tr>
<tr>
<td>Stenotrophomonas maltophilia</td>
<td>34</td>
<td>11.0 (10.6)</td>
<td>0.2 – 43.9</td>
</tr>
<tr>
<td>Serratia marcescens</td>
<td>22</td>
<td>13.3 (11.7)</td>
<td>0.1 – 38.4</td>
</tr>
<tr>
<td>Moraxella catarrhalis</td>
<td>24</td>
<td>16.3 (14.7)</td>
<td>0.1 – 44.3</td>
</tr>
<tr>
<td>Haemophilus parainfluenza</td>
<td>17</td>
<td>16.4 (16.1)</td>
<td>0.1 – 44.4</td>
</tr>
</tbody>
</table>

Table 4: Mean time to isolation after trach for selected organisms, N=45 subjects with respiratory culture after 2006

Figure 1: Organism Prevalence in 2010 based on length of time with tracheostomy (N=90)
Identifying the Presence of a Specific Chronic Lung Disease of Neurodisability
IRB study 203740

Study: Cohort of patients with primary neurodisability diagnosis with NO pulmonary disease at the time of initial diagnosis

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

Study Group: 136 patients age range x to x and with multiple congenital and acquired neurodisability diagnoses

Methodology:
Identify factors at time of initial presentation 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

Clinical Questions:
Role of pulmonary factors individually and collectively
Individual contribution in order of prominence
Results indicate that resilient characteristics may derive from common and normative measures of coping: the Family Crisis Oriented PersonalScales (FACES) and the Coping Health Inventory for Parents (CHIPS).

Parents (CHIPS) of children who have severe neurodisability that require chronic care were seen in the Arkansas Center for Respiratory Technology Dependent Children outpatient clinic. All children receiving care at the Arkansas Center for Respiratory Technology Dependent Children had chronic pulmonary symptoms that required daily respiratory plans and all had either acquired or congenital neurodisabilities. Caregivers provided informed consent and completed the Big 5 Questionnaire (BFQ) to measure personality traits; the Connor Davidson measure of resilience (Davidson, 1999), and general physical health and wellness behaviors (Booth Kewley & Vickers, 1994). More recently, the Big Five personality can be used as a reliable predictor of functioning in important domains of life such as mental health (Aldao, 2011), and psychological distress are commonplace. Furthermore, the literature suggests that the Big Five personality factors (John & Srivastava, 1999), can be a perfect reflection of what is typically found in trait resilience: high extraversion, low neuroticism, and agreeableness. The undercontrolled group is high on openness, conscientiousness, and emotional stability (as measured by the CHIPS; M = 25.14, SD = 8.12). No other differences were found between the caregiver personality prototypes on the other coping measures.

Cluster analysis of BFQ yielded 3 cluster prototypes: 22 resilient caregivers were characterized as resilient, 17 were an overcontrolled prototype (N = 22), an overcontrolled prototype (N = 22) or as an undercontrolled prototype (N = 19), based on an item between the caregiver personality prototypes on the other coping measures.

Overcontrolled caregivers had the highest average score averages on Extraversion. Overcontrolled caregivers had the highest average score averages on Neuroticism, and the lowest average score on Agreeableness. Overcontrolled caregivers had a significantly higher score on the Davidson measure of resilience (M = 3.65, SD = 1.00) than overcontrolled caregivers (M = 3.23, SD = .87; respectively). Resilient caregivers were significantly more likely to cope by maintaining their self esteem. However, resilient caregivers may not likely than overcontrolled caregivers to have supportive social relationships that help them cope and that maintain their self esteem. Resilient caregivers were characterized by higher Neuroticism and lower Extraversion, and significantly higher scores on the Davidson measure of resilience (M = 3.65, SD = 1.00) than overcontrolled caregivers (M = 3.23, SD = .87; respectively). Resilient caregivers were characterized by higher Neuroticism and lower Extraversion, and significantly higher scores on the Davidson measure of resilience (M = 3.65, SD = 1.00) than undercontrolled caregivers (M = 3.00, SD = .73). No other differences were found between the caregiver personality prototypes on the other coping measures.

Discussion

Although the study is limited by its cross sectional nature and its small sample size, the findings provide initial evidence to support the hypothesis that personality styles of caregivers may be associated with coping strategies and mental health outcomes. The results indicate that caregivers who have resilient personality styles may be less likely to have anxiety issues. Caregivers who are more overcontrolled may be particularly predisposed to distress and lack effective problem solving orientation. The results also suggest that caregivers who have resilient personality styles may be less likely to have anxiety issues. Caregivers who are more overcontrolled may be particularly predisposed to distress and lack effective problem solving orientation. The results also suggest that caregivers who have resilient personality styles may be less likely to have anxiety issues. Caregivers who are more overcontrolled may be particularly predisposed to distress and lack effective problem solving orientation. The results also suggest that caregivers who have resilient personality styles may be less likely to have anxiety issues. Caregivers who are more overcontrolled may be particularly predisposed to distress and lack effective problem solving orientation.
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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: In-exasufflator, high frequency oscillation, intrapulmonary percussionator
4. Ventilator support
### 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

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 ACRTDC
Special considerations

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

Skills Validation Quality Improvement Project

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

- Notification of the caregiver prior to clinic visit
- The caregiver must bring their 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

- 81% of participants had deficiencies in some area of home equipment use (set up, settings, technique, etc.)
- 81% would like to see SV for all respiratory equipment
- 45% reported an increase in confidence and decrease in anxiety when using equipment

Home Care

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

Respiratory Care Management
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

Case Study: 9 month old Caucasian male
Diagnosis: Neurodisability secondary to Kernicterus

- **Pulmonary Composite Factors**: Seizures, Dysphagia, Drooling, GER, Infective 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

What is your plan for this patient?

- Medications?
- Devices?
- Frequency?
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

What is your plan for this patient?

- Medications?
- Devices?
- Frequency?
- Ventilation?

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
What is your plan for this patient?

- Medications?
- Devices?
- Frequency?
- Ventilation?

Case Study: 16 year old Caucasian male
Diagnosis: Ulrich’s Muscular Dystrophy

- **Pulmonary Composite Factors**: non-ambulatory, hypotonia, hypopneic breathing pattern
- **Pulmonary Symptoms**: the need to create an aggressive pre-op respiratory care plan for spinal surgery to improve VC
- **Challenges to the Respiratory Management Plan**: Currently on non-invasive ventilation for 8 hrs/night, albuterol MDI followed by cough assist BID

What is your plan for this patient?

- Medications?
- Devices?
- Frequency?
- Ventilation?