An Overview of Bronchopulmonary Dysplasia and Chronic Lung Disease in Infancy
Housekeeping:

- I have no financial disclosures
- Learning objectives:
  - Develop an understanding of bronchopulmonary dysplasia (BPD)
  - Understand the multisystem nature of BPD
  - Begin to appreciate some of the feeding challenges among infants with BPD
  - Begin to appreciate the complexity of management of an infant with BPD with or without a tracheotomy
Bronchopulmonary Dysplasia:

BPD refers to a chronic respiratory condition that develops in infants treated for prematurity or any critical illness in the newborn period. It is a non-specific term and can be used interchangeably with the term neonatal chronic lung disease. While it is most often associated with the complications of and treatment for premature birth and lung immaturity any severe neonatal illness can result in the development of BPD/CLDI.
Bronchopulmonary Dysplasia:

- First described in 1967, BPD has evolved over time due to improved understanding and treatments:
  - Surfactant therapy
  - Maternal corticosteroid administration
  - Lung protective mechanical ventilation
  - Other factors
- This makes it very difficult to compare infants born during different time periods
- It is difficult to strictly define an evolving disorder and this makes randomized studies difficult as well
Bronchopulmonary Dysplasia:

• Incidence of BPD varies with birth weight and gestational age
• Some definitions:
  – Low BW (LBW) – BW < 2500g
  – Very low BW (VLBW) – BW < 1500g
  – Extremely low BW (ELBW) – BW < 1000g
The Normal Lung:

- 3 Basic Compartments:
  - Airways: Conduits for breathing
  - Alveoli: Site of gas exchange
  - Blood vessels: Gas transport
- Developmental factors: Preterm lung not ready to support air breathing
Bronchopulmonary Dysplasia:

- **Old BPD:**
- **Lung injury:**
  - Lung scarring
  - Obstructive physiology
  - Wheezing
  - Inhaled medications
  - Less common today due to improved neonatal care
Bronchopulmonary Dysplasia:

- **New BPD:**
- Arrested lung development:
  - Less scarring
  - Restrictive physiology
  - Tachypnea
  - “No nebs”
  - Rare over 1200g
Pathogenesis of BPD:

• Multifactorial including:
  – Antenatal inflammation
  – Lung immaturity
  – Oxygen toxicity
  – Barotrauma
  – Inadequate repair
  – Abnormal lung growth
  – Persistence of injurious stimulus

  Non-specific pulmonary response to injury
Suggested Classification of BPD:

Gestational age:  
<32wk  
>32wk

Time of assessment:  
36 wk PMA  
29-55 days PNA

Treatment with oxygen for at least 28 days PLUS:

Mild BPD:  
Room air  
Room air

Moderate BPD:  
<30% FiO₂  
<30% FiO₂

Severe BPD:  
>30% FiO₂  
>30% FiO₂

This scheme is fairly simplistic but is at least physiologic  
Respiratory symptoms are assumed to be present
The Incidence of BPD is Related to Birth Weight:

- Decreased birth weight results in increased BPD
- Infants with BW < 1250g account for 97% of BPD
- General statistics:
  - BW 1251-1500 6%
  - BW 1001-1250 14%
  - BW 751-1000 33%
  - BW 501-750 46%
- In one study of over 18,000 VLBW infants, 25% required oxygen at 36 weeks post-menstrual age
- Infants with tracheotomies are a small percentage of these small percentages
So what does this mean for us?

• All of the babies we are discussing today have severe BPD until proven otherwise
• Respiratory symptoms include:
  – Tachypnea
  – Labored breathing
  – Cough and wheeze
• All are medically fragile
Reported Risk Factors for BPD and Chronic Lung Disease:

- **Most Significant:**
  - Birthweight & Gestational Age
  - Respiratory Distress Syndrome
  - Severe respiratory illness
  - No antenatal corticosteroids

- **Important:**
  - Aggressive ventilation
  - Exposure to high FiO₂
  - Pulmonary airleak
  - Fluid overload
  - Symptomatic PDA
  - Sepsis
Reported Risk Factors for BPD and Chronic Lung Disease:

• **Other Risk Factors:**
  – White race
  – Male sex
  – Multiple birth
  – Outborn
  – Apgar scores

• Several multi-center studies have confirmed these as key indicators for oxygen dependence at 30 days of life.
Bronchopulmonary Dysplasia:

- **General Comments:**
  - Unresolved neonatal lung injury
  - Most common form of infant chronic lung disease
  - High morbidity and mortality
  
  **BPD is really a systemic illness**
Multi-system Involvement in BPD:

- **Respiratory:**
  - Parenchymal lung disease
  - Large & small airway disease
  - Respiratory control abnormalities
  - Sleep disordered breathing

- **Cardiovascular:**
  - Pulmonary vascular disease
  - Systemic hypertension
  - Structural heart disease
Multi-system Involvement in BPD:

- **Gastrointestinal:**
  - Feeding difficulties
  - Gastroesophageal reflux
  - Failure to thrive

- **Infection/Immunity:**
  - Respiratory viral infections
  - Primary immunologic dysfunction

- **Neurodevelopmental**

- **Psychosocial**
Respiratory Management Principles:

- Must be individualized:
- Tracheotomy:
  - Case-by-case indications
  - Poor correlation with PFT measurements
- Oxygen therapy
- Diuretics
- Airway care:
  - Bronchodilators
  - Anti-inflammatory agents
  - Mucus clearance
Chronic Oxygen Therapy:

- Just enough is better than too little:
  - Weight gain improves if $\text{Sao}_2 > 95\%$
  - Pulmonary hypertension avoided if $\text{Sao}_2 > 92\%$
  - Feeding hypoxemia is relatively common
  - Hypoxemia almost always recurs during illnesses

- The normal impairment in gas exchange during sleep is heightened in the presence of BPD:
  - Eliminate daytime oxygen if $\text{Sao}_2 > 92\%$ and
  - Growth rate is good
  - No pulmonary hypertension

- Overly aggressive oxygen therapy may promote BPD
Carbon Dioxide Retention:

- **Causes of inadequate ventilation:**
  - Too little lung tissue
  - Too much airway obstruction
  - Respiratory inefficiency

- **Elevated carbon dioxide levels result in:**
  - Poor growth
  - Cardiac stress

- **No magic number exists to indicate the need for long term mechanical ventilation with tracheotomy**
Which Infants with BPD get Tracheotomies?

- Handled on a case-by-case basis with variable levels of difficulty:
- **Straightforward indication:**
  - Severe subglottic stenosis or other airway problem
  - Failure to extubate although timing may be difficult
- **Less straightforward indication:**
  - Severe lung disease:
    - Infant is extubated but not thriving
- **Least straightforward indication:**
  - Chronic lung disease with multiple congenital anomalies or CNS injury
Management of the Patient with a Tracheotomy:

- **Patient safety first:**
  - Clinical monitoring
  - Adequate support for discharge
  - Suction, oxygen, airway, pharmacy
  - Other equipment

- **Medical supervision:**
  - Team management in a home ICU
  - Early recognition of problems

- **Team approach**
Management of the Patient with a Tracheotomy:

• In general, the management of a patient with BPD and a tracheotomy is similar to a patient without a tracheotomy except there is a lot more management going on!
Nutritional Management In BPD:

MORE CALORIES!!
- Increased work of breathing
- Hypoxemia and Hypercapnia
- Inflammation
- Feeding dysfunction
- Gastroesophageal reflux
- Intolerance
- Chronic illness

LESS FLUID!!
- Pulmonary edema
- Fluid imbalance
- Cor pulmonale
- Work of feeding
Coordination of Breathing and Swallowing:

- **Breathing is interrupted during nutritive sucking:**
  - Airway closure:
    - Obstructive Apnea
    - Inspiratory and expiratory
  - **Minute ventilation decreases:**
    - Tidal volume & respiratory rate decrease
    - Ventilation decreases inversely to swallow frequency
  - **Respiratory drive decreases:**
    - Decreased CO$_2$ sensitivity
    - Vagally mediated via the laryngeal chemoreceptors
Coordination of Breathing and Swallowing:

- **With maturation:**
  - Swallow limited to expiration
  - Swallow frequency decreases
  - Breathing is interrupted and irregular, but gas exchange remains unaffected

- **Clinical correlates:**
  - Suck-swallow-breathe incoordination
  - Feeding apnea
  - Feeding hypoxemia
Factors at Play in BPD:

- Respiratory factors
- If you can’t breathe, you are not going to want to eat:
  - Tachypnea
  - Increased work of breathing
  - Gas exchange abnormalities
- Non-respiratory factors:
  - Anatomic factors
  - Other medical problems
  - The feeding environment
  - Psychosocial factors
Dysphagia and Chronic Lung Disease: A working list.

- Cardiorespiratory immaturity:
  - Swallowing and apnea and bradycardia
  - Apparent life threatening event (ALTE)
  - Hypoxemia during feeding
- Bronchopulmonary dysplasia:
  - Gas exchange abnormalities
  - Respiratory insufficiency
  - Gastroesophageal Reflux
  - Stooling difficulties
  - Oral aversion
Swallowing and BPD

- Limited data in infants without tracheotomy
- Important themes:
  - Abnormal suck-swallow coordination
    - Disorganized suck-swallow runs
    - Shorter suck-swallow runs
  - Abnormal coordination of breathing and feeding
    - Irregular and interrupted respiration
Swallowing and BPD

• Important themes continued:
  – Increased apneic swallows
  – Maturational abnormalities as well:
    • Emergence of problems beyond 35 weeks PMA
    • Delayed maturation of suck-swallow-breathe coordination
  – The presence of BPD worsens these factors compared with healthy preterm infants
Reflux-Related Apnea:

- Temporal relationship difficult to document:
  - Conflicting literature
  - Spells during GER vs. Spells due to GER

- Regurgitation of material leads to:
  - Triggering of the laryngeal chemoreflex
  - Triggering of nonfeeding swallows
  - Both can influence respiration

- Central, obstructive or mixed apnea can occur

- Most common in young infants
Gastroesophageal Reflux and Oral Aversion:

• Gastroesophageal Reflux:
  – Common and may be severe
  – Medical management of GER may often fail
  – GER may present with atypical symptoms

• Oral Aversion:
  – Often accompanies GER
  – GER treatment often has no effect on aversion
  – A long term problem
Failure to Thrive:

- Increased demands
- Increased losses
- Decreased intake

Pitfalls:
- Hypoxemia:
  - Rapid wean of $O_2$
- Hypercapnia:
  - Need for ventilation
- Underestimation of caloric needs
- Balancing increased need & fluid sensitivity
The Patient with BPD Who is Not Getting Better:

• **Problem list:**
  - Inadequate calories
  - Inadequate oxygenation or ventilation
  - Optimize medical management:
    • Bronchodilators
    • Anti-inflammatory agents
    • Diuretics
  - Unsuspected heart disease
  - Large airway disease
  - Aspiration
Longterm Outcome:

- The data are evolving:
  - High mortality rate in severe BPD
  - Increased incidence of asthma
  - Obstructive lung disease on pulmonary function studies
  - Decreased exercise performance:
    - Out of proportion to measurable cardiopulmonary disease
  - Neurological outcome is very variable
Final Thoughts:

• All critically ill neonates are at risk for developing chronic lung disease (BPD)
• BPD is a syndrome with multisystem impact
• All infants with BPD are at risk for feeding, swallowing and gut motility disorders
• Infants with BPD and tracheotomies are at the severe end of this spectrum
• Lung growth is the only escape, so a safe and effective nutritional plan is critical to success