Differential Diagnosis and Management of Respiratory Distress

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The speaker has signed a disclosure form and indicated she has no significant financial interest or relationship with the companies or the manufacturer(s) of any commercial product and/or service that will be discussed as part of this presentation.

Session Summary
This presentation will provide an overview of airway issues, diseases, mechanical, structural, obstructive, and iatrogenic causes of neonatal respiratory distress. The speaker will review the key characteristics, stabilization, and treatment options for the conditions discussed.

Session Objectives
Upon completion of this presentation, the participant will be able to:
- understand the phases of fetal lung disease;
- discuss the physiology related to neonatal respiratory disorders;
- identify management strategies and apply them to neonatal illness;
- recognize respiratory emergencies and emergent management.

Test Questions
1. At the end of which stage is the lung considered viable?
   a. Embryonic
   b. Pseudoglandular
   c. Canalicular
   d. Saccular
   e. Alveolar
   f. Vascular

2. What is the most common associated anomaly with TEF and EA?
   a. Renal
   b. Cardiac
   c. Vertebral
   d. Limb
   e. CNS
3. Which direction is blood shunted in babies with PPHN?
   a. Right to left
   b. Left to right
   c. Bidirectional

4. You are called to evaluate an intubated 4-day old infant born at 25 weeks’ gestation. Oxygen saturation is 50% and the heart rate is 75 beats per minute and the baby is pale and cyanotic. The ETT has been suctioned with no occlusion. You bag by hand with a Pip of 30 without chest wall rise. What intervention will you do next?
   d. Administer an IV caffeine bolus
   e. Continue to hand bag with a higher pressure
   f. Obtain an emergent chest x-ray
   g. Remove the ETT and bag-mask ventilate
   h. Start chest compressions

5. Which of the following is true about self-inflating bags?
   a. It cannot reliably deliver free flow 100% oxygen
   b. It does not have a safety pop-off valve
   c. It does not fill spontaneously after it is squeezed
   d. It requires a gas source to inflate
   e. It requires a tight seal to maintain inflation of the bag

References


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Part 1 Developmental Disorders of the Respiratory System
Part 2 Respiratory Diseases
Part 3 Basic Management of the Airway
Part 4 Respiratory Emergencies

Part 1 Lung Development and Maturation
- Embryonic Period
- Pseudoglandular Stage
- Canalicular Stage
- Saccular and Alveolar Stages

What is a differential diagnosis?
Differential diagnosis

- Think of problems by system (ex. respiratory, cardia)
- View babies by system **symptoms**
- A diagnosis is what is known or suspected enough to consider for treatment, or has been diagnosed (x-ray – RDS, ABG – acidosis)
- Diagnosis leads to management
- Neonates often have concurrent diagnoses (RDS & Sepsis)
- Classic example – Respiratory versus Cardiac

Gestational Phases of Lung Development

- **Beginning – Embryologic and Pseudoglandular**
- **Middle – Canalicular and Saccular**
  - Capable of gas exchange
  - Pneumatocytes (Type I & II)
  - Capillary beds
- **Later – Alveolar and microvascular**

Stages of Lung Development (Kajekar et. al)

Branching Development of the Human Lung

Embryonic Lung Development

Embryonic Phase

- Begins with groove in ventral lower pharynx
- 2 bud form and develop asymmetrically
- Subdivides into 2 bronchi
- Disorders during this phase:
  - Atresias (laryngeal, esophageal, tracheal_
  - Bronchogenic cysts
  - TEF
  - Pulmonary sequestration
Pseudoglandular (5-17 weeks)
- Disorders during this time
  - Renal agenesis (pulmonary hypoplasia)
  - CCAM
  - Pulmonary lymphangiectasis
  - CDH
  - Tracheomalacia/bronchomalacia

Canalicular (16-26 weeks)
- Renal dysplasia and pulmonary hypoplasia
- Alveolar capillary dysplasia
- Surfactant deficiency

Saccular (24-38 weeks)
- Oligohydramnios and pulmonary hypoplasia
- Alveolar capillary dysplasia
- Surfactant deficiency

Alveolar (36 weeks+)
- Lobar emphysema
- Pulmonary hypertension
- Surfactant deficiency

Vascular 36 + weeks
- Derived from 6th aortic arch during development

Phases of Lung Development
QUESTION

At the end of which stage is the lung considered viable?
A. Embryonic
B. Pseudoglandular
C. Canalicular
D. Saccular
E. Alveolar
F. Vascular

Stages of Lung Development (Kajekar et. al)

How will you ever remember the stages of lung development?

Each Person Can Study Alone Vigorously

From Lucki Jain

- Each Embryonic (5 weeks)
- Person Pseudoglandular (15 weeks)
- Can Canalicular (25 weeks)
- Study Saccular (35 weeks)
- Alone Alveolar (3-5 years)
- Vigorously Vascular (3-5 years)

Case Study

A G2P1→2 28 year old woman gives birth to a 38 week baby boy weighing 3,200g by NSVD; prenatal history is uncomplicated. Fundal height consistently agreed with dates throughout the pregnancy.

A routine post-date non-stress test one week before delivery was reactive. An abdominal ultrasound obtained at the same time revealed an amniotic fluid index of 26. At delivery, the infant was vigorous and had Apgar scores of 9 at one minute and 9 at five minutes. Initial physical examination at one hour of life was remarkable only for a moderate increase in white oral secretions, which cleared with suctioning. The patient passed a meconium stool in the first six hours of life and tolerated his first feeding with minimal coughing and sneezing.
At approximately 8 hours of life, the infant was noted to have increased frothing and coughing after breastfeeding, and his abdomen was mildly dilated. Crackles were heard all throughout the lung fields. Vitals signs were all within normal limits. A catheter was gently passed into the esophagus and met resistance.

What is the diagnosis?

What is the significance of the AFI?

- Fetus swallows 100/mls/kg of amniotic fluid every day
- Amniotic fluid is produced by fetal kidneys

Based on the history, you suspect this baby has:

A. Congenital Diaphragmatic Hernia
B. Respiratory Distress Syndrome
C. Tracheoesophageal fistula

Bonus – During which phase on development did this disorder originate?

Tracheoesophageal Fistula

- **Tracheoesophageal Fistula**
- A Tracheoesophageal Fistula (TEF) is an abnormal passage between the trachea and esophagus
- 1: 3000
- Males
- Usually associated with Esophageal Atresia
- Incomplete division of the foregut during 4th week of gestation
- Septum between trachea and esophagus is defective

What diagnostic study would you order?

A. PA and lateral chest X-ray & KUB
B. Upper GI with Contrast
C. Abdominal CT
After ABCs, what is your priority for managing this infant?

A. Septic work-up  
B. Prevent aspiration  
C. Complete physical exam

What is the most common associated anomaly with TEF and EA?

A. Renal  
B. Cardiac  
C. Vertebral  
D. Limb  
E. CNS

Respiratory Disorders of Embryonic Phase

• Atresias (laryngeal, esophageal, tracheal)  
• Bronchogenic cysts  
• TEF  
• Pulmonary agenesis

Pulmonary Hypoplasia

• Maternal history  
• Most common is secondary and due to Restrictions is fetal breathing OR Restrictions in lung growth  
***Usually vascular in origin  
• Due to deficient development of the lung parenchyma→↓number of distal airways, alveoli, pulmonary vessels  
• Primary – caused by intrinsic lung development failure  
• Secondary – space occupying lesions compressing lungs/preventing normal growth

What are some causes of secondary lung hypoplasia?

• Cardiomegaly  
• Abnormal diaphragmatic activity (CNS or PNS)  
• Congenital Diaphragmatic Hernia
Disorders of Pseudoglandular Period

- Renal Agenesis
- CCAM
- CDH
- Tracheomalacia/bronchomalacia

Congenital Diaphragmatic Hernia

- Developmental defect during diaphragm formation
- Allows herniation of abdominal contents into thoracic cavity
- Can be isolated or associated with other anomalies
  - *Pulmonary insufficiency and pulmonary hypertension secondary to pulmonary hypoplasia*

Management of CDH

- Delivery in a proficient center
- Intra-disciplinary team with a coordinated approach
- Lower PIP/gentler ventilation/permissive
- HFOV
- ECMO
  - *Delayed surgical repair*
- Sometimes used by not yet proven beneficial: Surfactant, iNO
- Associated with sensorineural hearing loss

Martin, Fanaroff, Walsh, 2015
You are called to the delivery room to evaluate a full-term male infant who has just been born by virginal birth. You observe that the infant is in severe respiratory distress without breath sounds on the left side. The infant’s abdomen is scaphoid in appearance.

What is the most appropriate initial management of this infant?
A. Insert a chest tube to evaluate air from the pleural space
B. Intubate the infant to minimize inflation pressure
C. Place an umbilical venous line and start PGE1
D. Provide bag-mask ventilation to re-inflate the collapsed lung

Acquired Pulmonary Diseases
- MAS
- Neonatal Pneumonia
- TTN
- Aspiration Syndromes
- Pulmonary Air Leak Syndromes
- Rib Cage Abnormalities
- Phrenic Nerve Injury

More Respiratory Disorders of Development
- Canicular Phase (16-26 weeks)
  - Surfactant Deficiency
- Saccular Phase (24-38 weeks)
  - Oligohydramnios
  - Surfactant deficiency
- Alveolar Phase (36+ weeks)
  - Lobar emphysema
  - Surfactant deficiency
- Vascular Phase

Part 2 Respiratory Diseases
- Meconium Aspiration
- Transient Tachypnea
- Pneumonia
- Respiratory Distress Syndrome
- PPHN
Meconium Aspiration Syndrome

- Population
- Pathophysiology
- Clinical Presentation
- Differential Diagnoses
- Diagnostics
- Management
- Pharmacology
- Future trends

Transient Tachypnea

- Population
- Pathophysiology
- Clinical Presentation
- Differential Diagnoses
- Diagnostics
- Management
- Pharmacology
- Future trends

Neonatal Pneumonia/EOS

- Population
- Pathophysiology
- Clinical Presentation
- Differential Diagnoses
- Diagnostics
- Management
- Pharmacology
- Future trends
Respiratory Distress Syndrome

- Population
- Pathophysiology
- Clinical Presentation
- Differential Diagnoses
- Diagnostics
- Management
- Pharmacology
- Future trends

Lung maturation

- Throughout all of the phases, lungs are in various stages of maturation
  - At 24 weeks airways are completed through terminal bronchioles, gas exchange units are rudimentary
  - At 24 weeks the alveolar membrane enables gas exchange
  - Surface areas are expanding
  - Capillary network is expanding
  - True alveoli by 36 weeks

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Physical Mediators of Lung Development

1. Lung Fluid – helps lungs stretch and grow
2. Fetal Breathing Movements (inspired by?)
3. Peristaltic Airway Contractions
4. Vitamin A Deficiency
5. Glucocorticoids, thyroid, retinoic acid

Back to RDS – What is the Hallmark Pathophysiology?

Surfactant

- Composed of highly organized lipids and surfactant proteins
- Preterm lung is low in mature surfactant
- Molecules are hydrophobic and hydrophilic (opposed at both ends)
- Function of surfactant is to REDUCES SURFACE TENSION AT THE AIR FLUID INTERFACE and increases compliance of the lung and reduces the work of breathing
Surfactant – they will ask you this

*Reduces surface tension at the air fluid interface and increases compliance of the lung and thereby reduces the work of breathing*

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Which of the following is true about surfactant deficient lungs?

A. They are stiff and non-compliant
B. Result in ↑ WOB
C. Result in atelectasis and low lung volumes
D. Alveoli is filled with exudate
E. Gas diffusion is blocked
F. Results in hypoxia, ↑CO2, mixed acidosis

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Types of Surfactant

- Synthetic – ALEC, Exosurf
- Extracted – Survanta (bovine)
  - Curosurf (porcine)
  - Infasurf (calf)
  - Alveofact (bovine)
  - Venticute
  - Surfaxin

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Actions of Surfactant

- Acutely improves lung function (what does this mean to you as an NNP?)
- Surfactant substrate makes better lung surfactant
- Has a prolonged effect

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Surfactant demonstration

- You need a large surface area to allow for gas exchange (How will we accomplish this? – hint is rhymes with creep)
- Surfactant decreases surface tension............
- Type II pneumocytes produce surfactant but also are protective

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More about surfactant
PPHN

Which direction is blood shunted in babies with PPHN?
A. Right to left
B. Left to right
C. Bidirectional

Great website for more details about lung development


Risk Factors

- CDH
- MAS
- Asphyxia
- Surfactant deficiency
- Pneumonia
- Idiopathic

Management

- Delivery room
- Support
- Diagnosis
- PPHN versus cardiac disease
Management Strategies

- Oxygen
- Mechanical ventilation
- ECMO
- Medications
  - iNO
  - Surfactant

Newer Medications

- Sildenafil
- Steroids
- Adenosine
- Milrinone

Other Respiratory Diseases

- BPD
- Interstitial lung disease
- Choanal atresia
- Laryngomalacia
- Tracheobronchomalacia
- Tracheal stenosis
- Vascular airway abnormalities

Part 3 Basic Respiratory Management

- Primary and secondary apnea
- Oxygen
- Properties
- NRP recommendations
- Pulse oximetry
- Continuum of respiratory care
- Suctioning

CPAP

- Physiology of PEEP
- Alveolar Recruitment
- Improving oxygenation

Part 4 Respiratory Emergencies

- Air Leak
- Infections
- Congenital Diaphragmatic Hernia
- Pierre Robin Syndrome
- Abdominal Distention
Resuscitation

- PLEASE STUDY NRP
- Understand the foundational underpinning of ventilation
- PPV as a non-invasive methodology
- Primary apnea is a retrospective concept
- Naloxone is not an emergency drug

Intubation

- Approach to intubation
- Indications for intubation
- Noninvasive ventilation
- Safety concerns
- Technique

Demonstration

This is my trick for success when you are first learning the difficult skill of intubation

Transillumination

Pneumothorax

Collapsed lung
Needle Aspiration

- Indications
- Landmarks and technique

Extrapulmonary Causes of Respiratory Distress

- **Neuromuscular** – CNS, asphyxia, hemorrhage, malformations, drugs, birth injury, muscular dystrophies
- **Obstructive** – thoracic dystrophies, choanal atresias, tracheal stenosis, blocked ETT
- **Diaphragmatic Disorders** – abdominal distention, eventrations
- **Hematologic** – polycythemia, anemia
- **Metabolic** – metabolic acidosis, hypoglycemia
- **Cardiovascular** – ↑ pulmonary blood flow, ↓ pulmonary blood flow, cardiomegaly
- **Miscellaneous** – sepsis, pain, hypothermia, hyperthermia

Martin, Farshoef, Walsh, 2015