Penn Medicine
Lancaster General Hospital

Department of Pulmonary Medicine

Neonatal Acute Disease Identification and Management

+ Synopsis of Multi-system Inflammatory Syndrome

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Disclosure

- Mr. Pavlichko has no real or perceived conflict of interest related to this presentation
- Off-label and/or investigational use of respiratory equipment/medications may be included in this presentation

Learning Objectives

- The learner will identify key characteristics of common neonatal diseases
- The learner will identify evidence-based treatment strategies directly related to specific neonatal disease states
- The learner will be able to discuss pathophysiology, identification, and management of COVID related Multi-system Inflammatory Syndrome.

Infant disease process

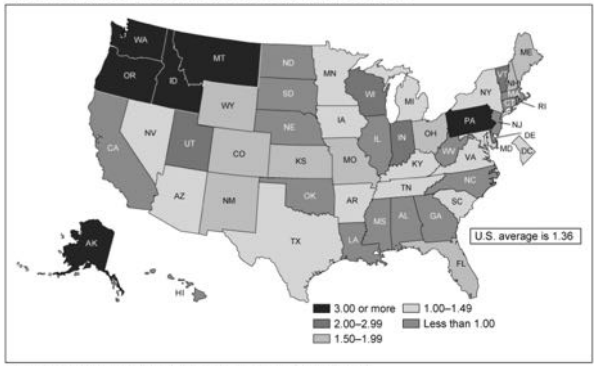
- Understanding term and preterm neonatal disease processes is important to the respiratory therapist
  - Assessment = troubleshooting, plan of care
  - It enables better understanding of therapies
  - Enables us to anticipate patient needs
  - Makes us an essential part of the health care team

- Structure of disease learning
  - Pathophysiology (what is wrong)
  - Clinical Features (what do we see)
  - Management (how do we fix)

Why is this important?

- "What you do in the first few minutes in an emergency for a newborn will set the tone for the rest of their life" – David Sorrentino, MD
- Data:
  - 5,273 emergency departments in the US – Emergency Medicine Network
  - 2,428 hospitals offer obstetrical services – March of Dimes
  - 220 total children's hospitals - CHA
  - 90% of all children emergency cases happen outside of a children's hospital – Chamberlain, et. al.
  - In 2012, 1.36% out of hospital births – CDC
    - 3.1% in PA

Figure 3. Percentage of births occurring out-of-hospital, by state: United States, 2012



NOTE: Access data table for Figure 3 at: http://www.cdc.gov/nchs/data/databriefs/db144_table.pdf#1.
SOURCE: CDC/NCHS, National Vital Statistics System, birth certificate data.

Mutually Beneficial

- Community Hospitals
  - Keep neonates with moms
    - Patient development
    - Family development
  - Financially responsible
    - Institution
    - Socially

- Children's Hospitals
  - Resources for difficult cases
  - Resources are limited
    - Unable to care for common neonatal issues due to demand
    - Don't want your TTN patients
    - But also don't want "train wrecks"

Delivery Room!

Scenario 1

- 26 3/7 week gestation
- Vaginal birth, no prenatal care, no prenatal steroids, G7P5

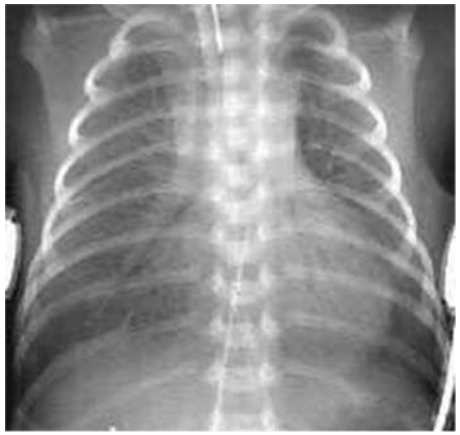
- Delivered
  - Placed in warmer in Neo Wrap (plastic)
  - No cry apnea, floppy, HR < 60, monitor applied
  - Deliver PPV 20/5 30%, good chest rise, HR > 100
  - Saturations within NRP range for time of life
  - No spontaneous respirations

  - Intubated at 3 minutes of age – 2.5 ETT secured @ 7cm lip

- Taken to the NICU for further evaluation

Continued 1

- NICU
  - Admitted, regular labs, UAC/UVC etc.
  - Ventilator – PC SIMV 18/5, f 40, Ti 0.35, PS +6, 40%
  - ABG 7.18/68/60/-4
  - CXR – hazy, ground glass, air bronchogram, ETT ok
  - Saturations = 84% with increased oxygen requirements



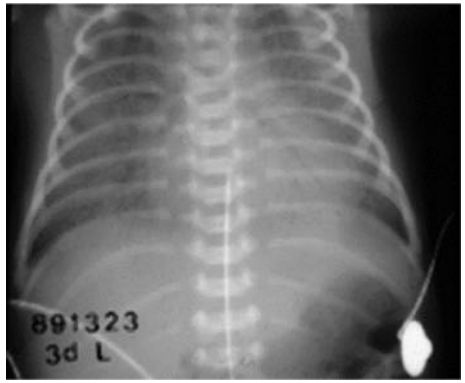
- Intervention?
  - Surfactant
    - Curosurf 2.5 ml/kg
    - Survanta 4 ml/kg

Respiratory Distress Syndrome (RDS)

- Pathophysiology
  - Formally called Hyaline Membrane Disease
  - Caused by immature lung anatomy and inadequate pulmonary surfactant
    - Starts around 20 weeks
    - Reduces surface tension
    - Lung compliance issues
    - Prematurity decreases quantity and quality of surfactant
      - Directly related to severity
  - Inflammation & Pulmonary Edema
    - VQ mismatch
    - Atelectasis & hyperinflation
    - Poor gas exchange
    - Hypoxemia
  - Complications in survivors of RDS are significant
    - PDA, Pulm. hemm., BPD, sepsis
    - Neurofunction
    - Slightly higher in males

Respiratory Distress Syndrome (RDS) con't

- Clinical Features
  - Inc. WOB, rales, tachypnea
    - Grunting, retractions, cyanosis
  - Clinical Course
    - Atelectasis
    - FRC
    - VQ mismatch
    - edema
    - cell injury
    - hypoxemia
    - Acidemia
    - Increased need for ventilation
    - Repeat…
  - Chest x-rays
    - Low lung volumes
    - Diffuse reticulogranular pattern
    - Air bronchograms
  - Labs
    - Hypoxemia
    - Hyponatremia – attentive fluid management



Respiratory Distress Syndrome (RDS) con't

- Management
  - Antenatal
    - Stop preterm births
    - Increase lung development
      - L/S Ratio >2 good
      - Glucocorticoids
  - Postnatal
    - Surfactant replacement
    - MV, CPAP, PEEP, O2, HFNC
      - Lung protective strategies – don't over do it!!
    - Patience
  - Treat Co-morbid Conditions
    - Air leaks, infection, ICH, PDA
    - Skin
    - Electrolytes and nutrition
    - PPHN – iNO?

Scenario 2

- 39 1/7 week gestation
- Primary C-section for breech presentation G3P2, GBS –, SSRI

- Delivered
  - Infant no cry, delayed cord clamping
  - HR > 100, dry and stimulated, bulb suction for lg. amt clear fluid (mouth and nose)
  - Spont. resp., cry with stimulation
  - APGAR 7, 9
  - Infant slow to transition, color poor, some slight retractions, flaring
  - Patient requires +5 NCPAP, 30% FiO2

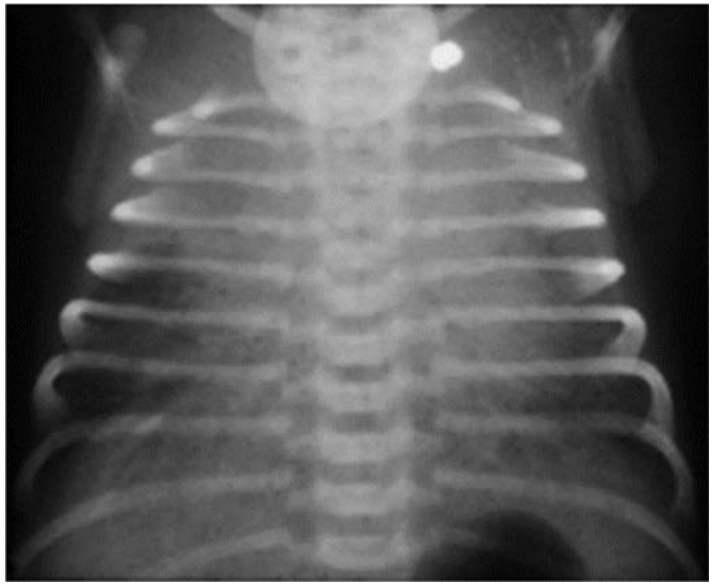
- Taken to the NICU for further evaluation

Continued 2

- NICU
  - Admitted
  - FiO2 weaned quickly, patient comfortable
  - CBG 7.32/50/35/-2
  - CXR – hazy

- 2 hours later
  - Increased retractions and flaring over time
  - +5 NCPAP @ 65%

- Likely Diagnosis?



Transient Tachypnea (TTN)

- Pathophysiology
  - Delayed reabsorption of fetal lung fluid – transient pulmonary edema
  - Obstructive abnormality – “wet lung”
  - Term or near term with low APGARs
    - C-section, narcotics, SSRI, nuchal cord
  - Onset: 2-6 hours after birth
- Clinical Features
  - Hypoxemia, inc. WOB
  - Tachypnea (duh)
  - Diagnosis of exclusion
    - Rule out other causes
    - Could appear as RDS
  - CXR
    - Streaky infiltrates from hilum, Air-trapping, fluid fissures, cardiomegaly

Transient Tachypnea (TTN) con't

- Management
  - Supportive care
  - O2 therapy, keep > 90%
  - Positive Pressure
  - Usually resolves in 72 hours with just supportive care
  - Feeding?
  - Fluid restriction?
  - Diuretics?

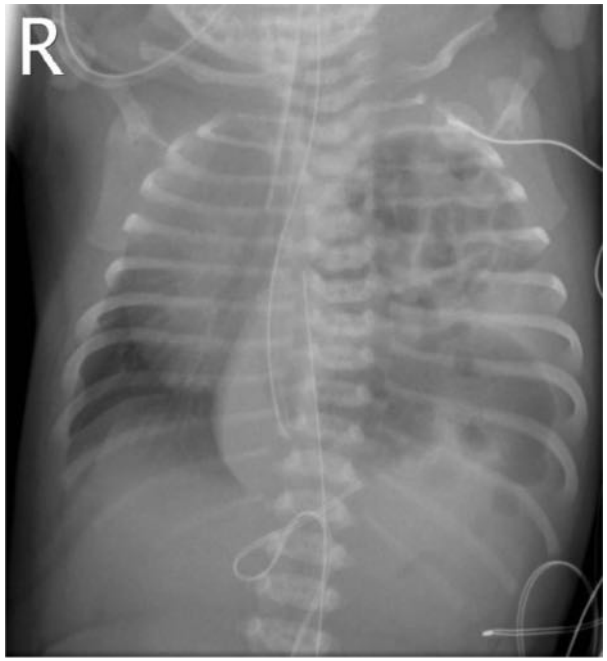
Scenario 3

- 39 5/7 week gestation
- Vaginal birth, no prenatal care, Primagravida, polyhydramnios
- Non-reassuring heart tones
- Ultrasound reveals possible chest abnormality
- Delivered
  - No cry apnea, floppy, cyanotic, HR < 60, monitor applied
  - Scaphoid abdomen
- Intervention?
  - Intubation? FiO2? EtCO2?
  - Gastric Tube?
  - CPR? (HR 50)
- What are you suspecting?
- Taken to the NICU for further evaluation

Continued 3

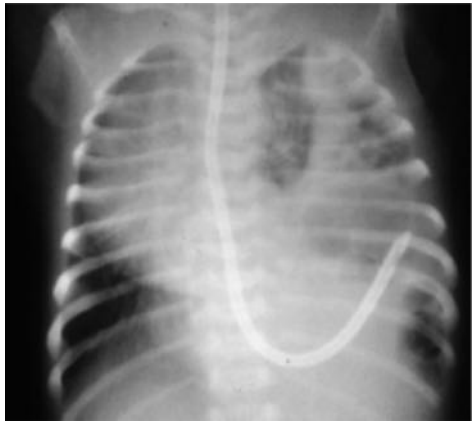
- NICU
  - Admitted, regular labs, UAC/UVC etc., CPR ceased HR > 100
  - Ventilator – PC SIMV 22/6, f 40, Ti 0.35, PS +8, 80%
  - ABG 7.00/98/42/-8
  - CXR – bowel gas pattern in chest, ETT ok?
  - Poor, asymmetrical chest rise

- Intervention?
  - HFOV?
    - Settings?
  - Stabilization
  - Surgical Consult
  - Permissive hypercapnia



Congenital Diaphragmatic Hernia (CDH)

- Pathophysiology
  - 1:4000 live births
  - Usually diagnosed in utero via ultrasound
    - 83% occur on L. side
    - 15% R. side – Liver herniation – poor prognosis
    - 1-2% bilateral
  - Usually associated with other malformations (genetic eval)
    - Cardiac anomalies 50%
    - Chromosomal abnormalities 30-50%
    - Neural Tube defects 50%
  - Pulmonary Hypoplasia can be significant
    - Loss of pulmonary mass
    - Surfactant system dysfunction
    - Pulmonary Hypertension

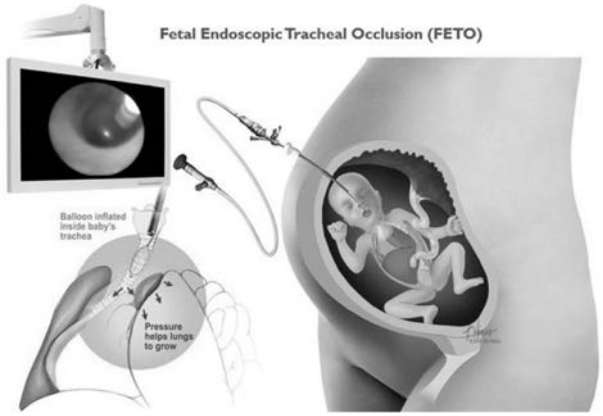


Congenital Diaphragmatic Hernia (CDH) con't

- Clinical Features
  - Cyanosis
  - Distress
  - "scaphoid abdomen"
  - Bowel sounds in chest
  - Distant or absent heart and breath sounds
  - CXR:
    - "hey, is that intestine!?"

Congenital Diaphragmatic Hernia (CDH) con't

- Management
  - Fetal surgery
    - Utero tracheal occlusion - investigational
  - INTUBATION!!
  - NO BMV, N-G tube placement
  - Resuscitate before cord clamping
  - Stabilize ⇨ Transport ⇨ Surgery ⇨ ECMO
    - Or…deliver at appropriate center



Scenario 4

- 41 5/7 week gestation
- Vaginal birth, prenatal care, Primagravida, GBS –
- Mom ruptured, thick particulate meconium present
- What should you anticipate?
  - Intubation?
  - Refractory Hypoxia?
  - Airway obstruction?
  - Respiratory depression? Why does MAS happen?
- Delivered
  - No cry, apnea, floppy, meconium stained skin & tongue, monitored applied
  - HR < 100, airway cleared, PPV
  - HR improves, still apneic, tone depressed
  - Intubation 3.5 @ 9 lip
- Taken to the NICU for further evaluation

Meconium Aspiration (MAS)

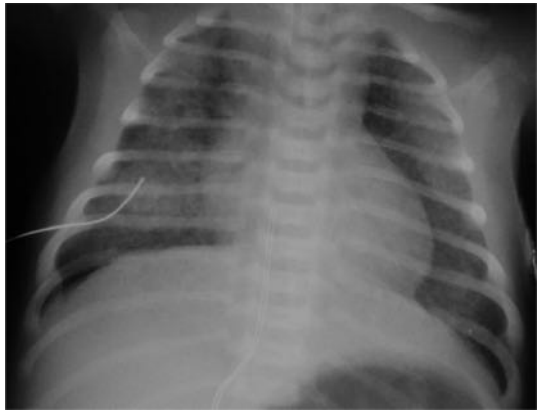
- Pathophysiology
  - 2-10% of infants born with meconium stained fluid
  - Birth depression
    - Why?

  - Pulmonary disease
    - Airway obstruction – “duck bill valve”
    - Inflammation
    - Infection
    - Inactivation of surfactant
    - Hypoxemia, academia, PPHN – VQ mismatch

Meconium Aspiration (MAS) con't

Clinical Features

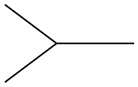
- Staining, peeling skin, decreaed vernix
- Inc. WOB, cyanosis, grunting
- Barrel shaped chest
- Hypoxemia, acidemia
- Chemical pneumonitis
- Pneumo ~ "ball-valve effect"



- CXR
  - Air trapping, hyperventilation
  - Bilateral infiltrates, R > L
  - Areas of atelectasis
- Long Term
  - Usually fine
  - Some neuro issues
  - Airway reactivity
- Other differential
  - TTN, Delayed transition (PDA, PFO), pneumonia, PPHN, Pulm Edema, Airleaks,

Meconium Aspiration (MAS) con't

- Management
  - Prevention:
    - Suction?
    - Identification and earlier delivery
    - Amnioinfusion
  - MV
    - Sedation
    - Surfactant replacement
  - Life saving
    - iNO
    - ECMO

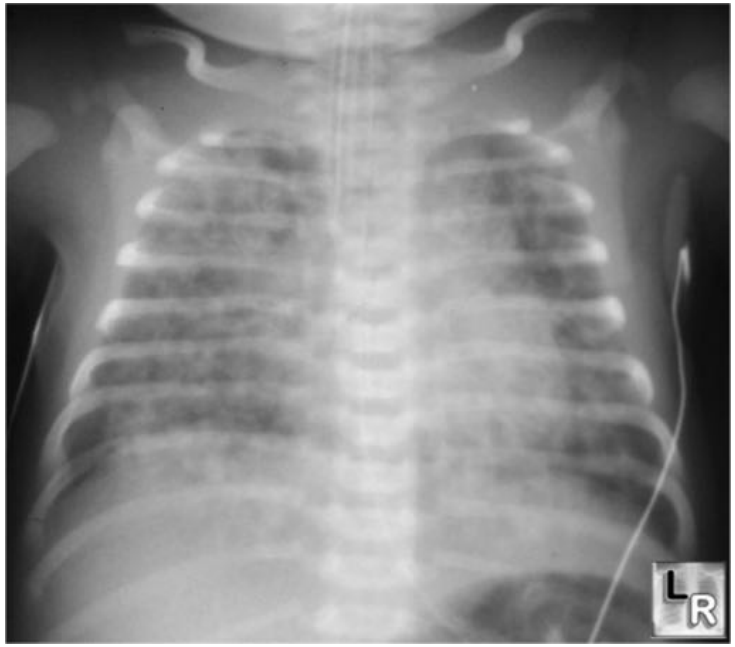


Severe MAS with PPHN

Continued 4

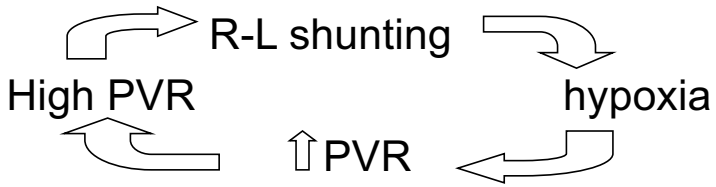
- Continued refractory hypoxemia
  - OI = $\frac{\text{MAP x FiO2}}{\text{PaO2}}$ = 25
  - ECHO ordered
  - Pre and post (92% and 80%)

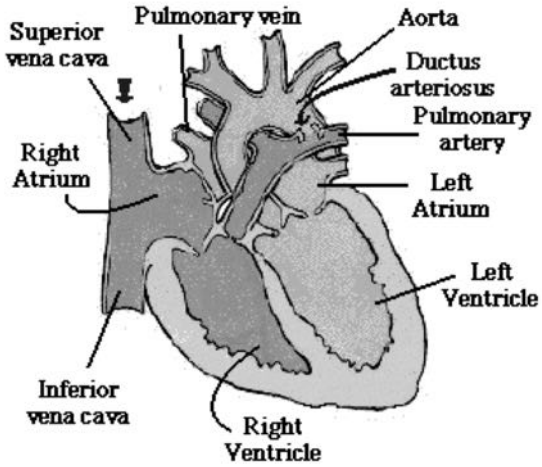
- ECHO diagnosis - PPHN



Persistent Pulmonary Hypertension (PPHN)

- Pathophysiology
  - Poor transition from fetal to neonatal circulation
  - ⇧ PaO2 = ⇩ PVR
    (did not happen)
  - Due to underdevelopment
    of pulmonary vasculature





Blood just not getting to the lungs

Persistent Pulmonary Hypertension (PPHN) con't

- Clinical Features
  - Near term infants > 34 weeks with worsening cyanosis
    - Tachypnea, WOB
    - CBC: anemia, polycythemia, infection?
    - ABG: acidosis, hypoxemia
    - Pre/Post ductal pulse ox difference
    - CXR – Normal, slightly enlarged heart
  - Underlying disorders
    - MAS, pneumonia, RDS, CDH, etc.
  - Hyperoxia test
  - ECHO
    - severity of PPHN
    - Ensure not ductal dependent lesion or abnormality

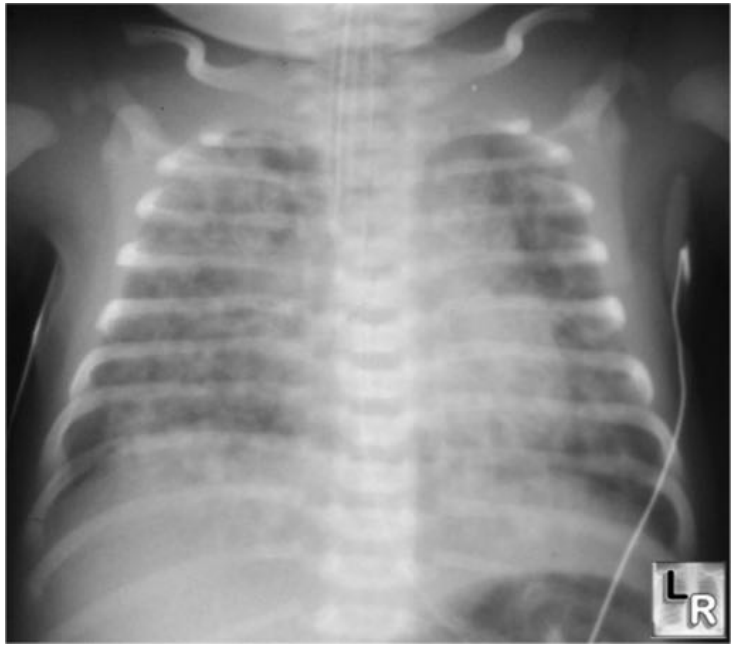
Persistent Pulmonary Hypertension (PPHN) con't

- Management
  - General supportive care
  - OXYGEN!
  - Sedation & minimal handling
  - Control hypoxia and acidosis – MV, HFOV, ECMO
  - Pressors to inc. SVR which will dec. PVR
  - Indocin, ibuprofen
  - iNO
  - ECMO OI>40
  - Sildenafil
  - Prostacyclin?
  - Fix underlying cause

  - 7-10% mortality, increased morbidity

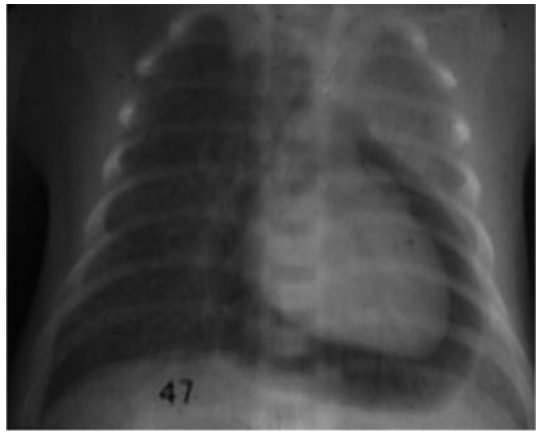
Continued 4

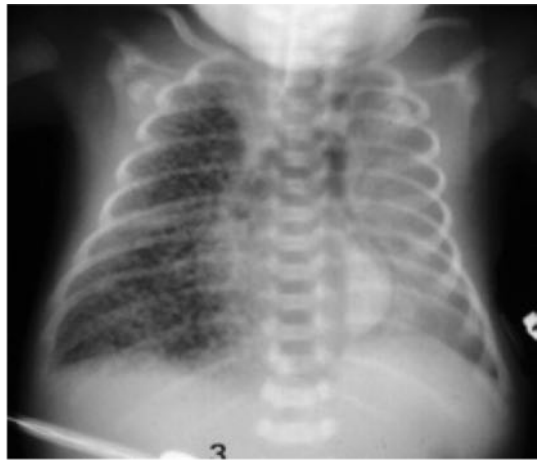
- Suddenly…
- Patient drops saturations

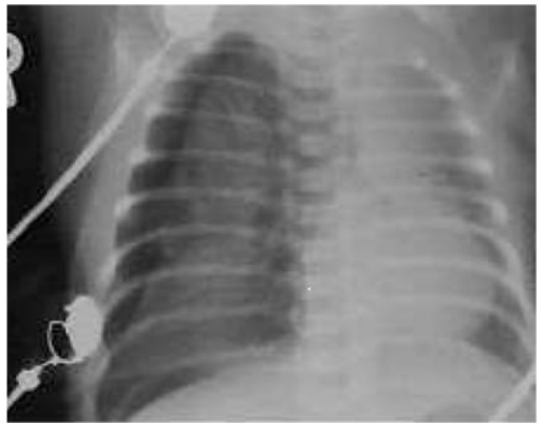


Air leak syndromes

- Pneumothorax
- Pneumopercardium
- Pneumomediastinum
- PIE

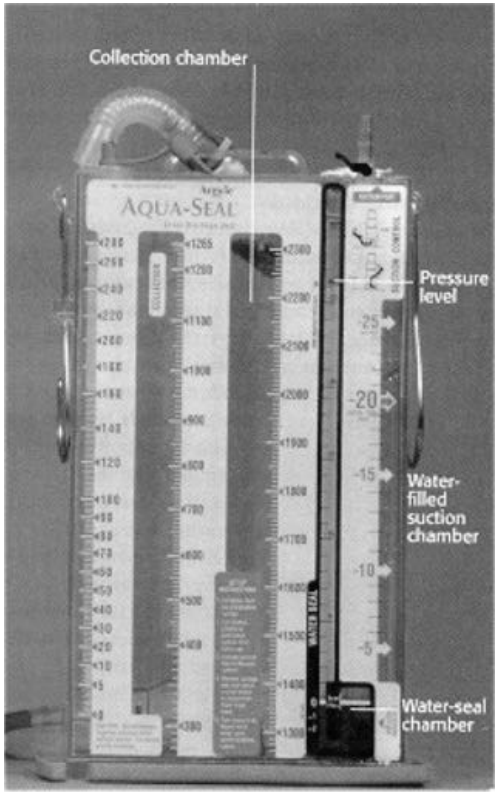






Air leak syndromes

- Management
  - 100% FiO2 – Nitrogen washout
  - Needle aspiration
  - Chest tube
  - Turn down the pressure!



Upper Airway Pathophysiology

Pierre Robin

- Pathophysiology
  - Micrognathia, glossoptosis, cleft soft palate
  - early mandibular hypoplasia
  - Genetic syndromes or not
  - Early mandibular retrognathia – primary anomaly
  - Features:
    - Tongue posterior
    - airway obstruction
    - feeding & pulmonary problems

- <u>Management</u>
  - Proning with head down
  - Nasal Airway
  - Nasal intubation
  - Tracheostomy

Choanal Atresia

Path. and features

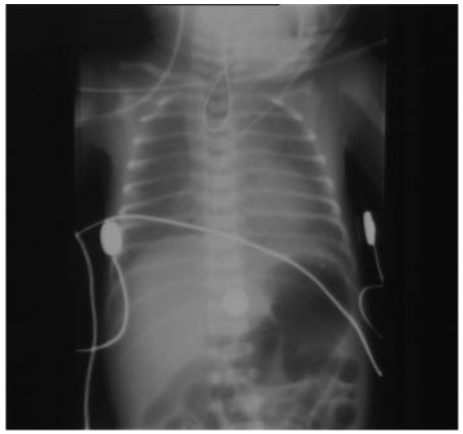
- Micrognathia with nasal airway obstruction
  - Obligate nose breathers
  - Distress and cyanosis relieved with crying
  - Diagnose: catheter to nares to determine
- Rare, 1:5000 live births
  - 2/3 are unilateral
  - Bilateral is life-threatening
  - Usually associated with genetic syndrome

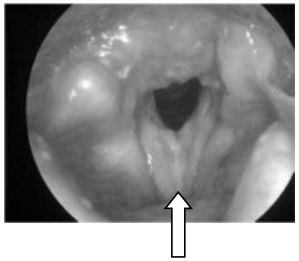
Management

- Support airway
  - Oral pharyngeal airway or ETT
- Laser surgery to nasal passage to make it patent

Structure Disorders

- Tracheomalacia – Tracheal collapse
- Tracheal-esophageal fistula – open connection between trachea and esophagus
- Laryngeal atresia – congenital closure of the larynx
- Laryngomalacia – congenital flaccid larynx, supraglottic structure collapse
- Laryngeal web – failure to recanalize the laryngeal inlet @ ~ 10 weeks GA, membrane occludes inlet
- Intubation Trauma
  - Subglottic stenosis
  - Granuloma formation





Multisystem Inflammatory Disease

Multisystem Inflammatory Syndrome in Children (MIS-C)

- Dr. Julie Fitzgerald – PICU Intensivist – CHOP

Summary

What is already known about this topic?

Multisystem inflammatory syndrome in children (MIS-C) is a rare but severe condition that has been reported approximately 2–4 weeks after the onset of COVID-19 in children and adolescents.

What is added by this report?

Most cases of MIS-C have features of shock, with cardiac involvement, gastrointestinal symptoms, and significantly elevated markers of inflammation, with positive laboratory test results for SARS-CoV-2. Of the 565 patients who underwent SARS-CoV-2 testing, all had a positive test result by RT-PCR or serology.

What are the implications for public health practice?

Distinguishing MIS-C from other severe infectious or inflammatory conditions poses a challenge to clinicians caring for children and adolescents. As the COVID-19 pandemic continues to expand in many jurisdictions, health care provider awareness of MIS-C will facilitate early recognition, early diagnosis, and prompt treatment. – CDC.gov

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