PERSISTENT TACHYPNEA OF NEWBORN

Dr. Sharada
DNB PG, NICU
Dr. Mehta’s hospitals
- Born to 31 yrs, G2A1 mother
- Non consanguinous
- No antenatal risk factors, no fever
- ARM done, clear liquor, not foul smelling
- Delivered by LSCS at 40 completed weeks in view of non progression of labour
- Cried soon after birth
- Apgars of 7/10, 8/10
Baby had respiratory distress since birth
started on hood O2, IV fluids and IV antibiotics
CXR showed minimal streaking
Probable TTNB
- Echo done was normal
- Continued to have distress at D4 of life
- Required an FiO2 of 0.5
- Referred to Mehta NICU
- On examination had a RR-80/min, minimal retractions
- RD score of 4/10
- SpO2 of 95% with FiO2 of 0.6
- ABG: pH-7.230 PCO2- 61.5 PO2-121.5
- CXR showed increased peribronchial infiltrates
Initially baby was kept NPO
No H/O pooling of secretions/frothing from the mouth
No H/o cough, no grunt/stridor
Suspicion of aspiration pneumonitis
Congenital pneumonia
Sepsis screening was negative
ENT surgeon’s opinion obtained
ENT normal
Baby was started on paladai feeds with minimal O2 support (FiO2- 0.2) from day 15 of life.

breast feeds with minimal O2 support from day 20 of life.

O2 requirement decreased with time

TORCH screening done was negative

Repeat CXR showed radiolucent opacities in the peribronchial regions.
His ABG showed high pCO2 with normal SaO2

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In view of persistent respiratory distress and alert child

HRCT was planned
Mediastinum:
The trachea and major bronchi are normal.
No significant hilar / mediastinal lymphadenopathy.
The thoracic esophagus is normal. The gastroesophageal junction is normal.

Lungs and pleura:
Patchy areas of ground glass opacification with septal thickening are noted scattered in bilateral lung fields.
No pleural thickening / nodularity / effusion on either side.

Thoracic cage and extra thoracic soft tissues: appear normal.

Quality is our Image
IMPRESSSION:

- Patchy areas of ground glass opacification with septal thickening scattered in bilateral lung fields; possibilities include: surfactant deficiency / venous obstruction / congenital alveolar proteinosis.

Please correlate clinically and with HPE.
- Baby was given a trial of oral erythromycin
- We were able to wean him of supplemental oxygen by D25 of life
- At discharge,
  - he had tachypnea
  - maintaining SpO2 of 95% in room air
discussion
differential diagnosis of persistent tachypnea

- ASPIRATION PNEUMONIAS
- CONGENITAL PNEUMONIAS
- INTERSTITIAL LUNG DISEASE
ASPIRATION PNEUMONIA

- Aspiration of amniotic fluid
- Aspiration of meconium
- Aspiration of blood
- Aspiration of vernix caseosa
AMNIOTIC FLUID ASPIRATION PNEUMONIA

- **Results**: A large number of cases were term infants (82.1%) and infants born by caesarean section (85.7%). Soon after birth, the amount of amniotic fluid sucked out from airway below the vocal cord was $16.0 \pm 10.1$ mL. Compared with pre-SRT, the oxygenation index ($8.0 \pm 9.6$ vs. $18.9 \pm 7.3$) showed a significant improvement at 12 h after SRT ($P<0.001$). Furthermore, most cases showed radiological improvement for aeration at 12 h post-treatment.

- **Conclusion**: AFAP may be an important cause of serious respiratory distress in near-term and term infants, and SRT seems to be an effective adjuvant therapy in mechanically ventilated neonates with severe AFAP.

Clinical findings of severe amniotic fluid aspiration pneumonia and effects of surfactant replacement therapy Sang Woo Park, M.D., Chun-Soo Kim, M.D., Sang-Lak Lee, M.D. and Tae-Chan Kwon, M.D. Department of Pediatrics, Keimyung University School of Medicine, Daegu, Korea.
Blood aspiration syndrome as a cause of respiratory distress in the newborn infant.

Early-onset respiratory distress and a radiographic appearance of an aspiration syndrome occurred in three neonates who had not passed meconium before delivery. In each case there was evidence of inhalation of blood, associated with very high plasma protein concentration in lung fluid. Blood aspiration syndrome is a distinct diagnostic entity that can result in significant respiratory distress in the neonate.

- J Pediatr. 2003 Feb;142(2):200-2, Department of Neonatology and Murdoch Children's Research Institute, Royal Children's Hospital, Melbourne, Australia.
CONGENITAL PNEUMONIAS

- Intrapartum pneumonia is acquired during passage through the birth canal.
- Acquired
  - hematogenous or ascending transmission,
  - from aspiration of infected or contaminated maternal fluids
  - from mechanical or ischemic disruption of a mucosal surface that has been freshly colonized with a maternal organism of appropriate invasive potential and virulence.
- Infants who aspirate proinflammatory foreign material, such as meconium or blood, may manifest pulmonary signs immediately after or very shortly after birth.
- Infectious processes often have a honeymoon period of a few hours before sufficient invasion, replication, and inflammatory response have occurred to cause clinical signs.
**Congenital Pneumonia**

- Persistent tachypnea (RR >60/min)
- Expiratory grunting may occur.
- Accessory respiratory muscle recruitment
- Airway secretions may vary substantially in quality and quantity but are most often profuse and progress from serosanguineous to a more purulent appearance.
Interstitial lung diseases (ILDs) in childhood are a diverse group of conditions that primarily involve the alveoli and perialveolar tissues, leading to derangement of gas exchange, restrictive lung physiology, and diffuse infiltrates on radiographs.
INTERSTITIAL LUNG DISEASE

- considered in any infant with a normal birth history
- presents with tachypnea, crackles, hypoxemia, chronic cough, or diffuse infiltrates of unknown etiology.
- in any term or late preterm infant who develops respiratory distress and either fails to respond adequately to respiratory support or cannot be weaned from respiratory support.
- family history of a similarly affected sibling
unique forms of interstitial lung disease in infancy

- Disorders of lung growth and development
- Persistent tachypnea of infancy (PTI)/Neuroendocrine cell hyperplasia of infancy (NEHI)
- Follicular bronchitis/broncholitis
- Cellular interstitial pneumonitis of infancy/Pulmonary interstitial glycogenosis
- Acute idiopathic pulmonary hemorrhage of infancy
- Chronic pneumonitis of infancy/Genetic defects of surfactant function
To assess extent and severity of disease

- Radiographic studies
- Chest radiographs
- High-resolution computed tomography
- Electrocardiogram
- Echocardiogram
- Ventilation perfusion scan
High-resolution computed tomography (HRCT) correlates better with the extent, distribution, and severity of disease than does a chest radiograph and provides guidance for biopsy.
Lung biopsy is the most reliable diagnostic procedure because diagnoses of ILD are based upon histopathologic description. HRCT should be used to identify affected areas for biopsy.
PULMONARY INTERSTITIAL GLYCOGENOSIS

- Also known as Cellular interstitial pneumonitis
- present with tachypnea since birth and diffuse infiltrates of unknown etiology.
- Lung biopsy - interstitial proliferation of bland, nondescript histiocytic type cells and minimal or no inflammation.
- Electron microscopy - cells contained monoparticulate glycogen,
- Most infants remained tachypneic for months, but generally improved over time.
tachypnea, crackles, and hypoxemia in the absence of an underlying disease to explain the symptoms.

- Radiographic findings included hyperinflation on chest radiograph
- HRCT-hyperinflation and ground-glass opacities
- Pulmonary function testing was consistent with air trapping.
Lung biopsy- mild and nonspecific changes, no known pathologic disease process, no interstitial involvement or inflammation, and no infectious organisms.

bombesin immunohistochemistry- hyperplasia of neuroendocrine cells in the distal airways and aggregates of neuroendocrine cells in the lobular parenchyma - consistent finding

A striking feature of NEHI is the discrepancy between the infant's ill-appearance and the lack of significant abnormalities in radiographic studies and biopsies

Patients appear to improve with time, though symptoms may persist for months to years.

Treatment with corticosteroids, bronchodilators, and other agents has been tried, with inconsistent result.

trial of corticosteroids for 6-8 weeks to be considered
Response to steroids may be variable
patients requiring long-term corticosteroid therapy are at risk of developing long term adverse effects
considerable morbidity and mortality associated with many forms of ILD has to be balanced against the treatment risk
# GENETIC ABNORMALITIES OF SURFACTANT FUNCTION

<table>
<thead>
<tr>
<th>gene</th>
<th>Type 1 (SFTP-B)</th>
<th>Type 2 (SFTP-C)</th>
<th>Type 3 (ABCA-3)</th>
</tr>
</thead>
<tbody>
<tr>
<td>inheritance</td>
<td>Autosomal recessive</td>
<td>Autosomal dominant</td>
<td>Autosomal recessive</td>
</tr>
<tr>
<td>chromosome</td>
<td>2p12-11.2</td>
<td>8p21</td>
<td>16p13.3</td>
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<tr>
<td>Clinical features</td>
<td>Neonatal respiratory failure and death</td>
<td>Infantile and older chILD with respiratory failure</td>
<td>Neonatal and early infantile respiratory failure</td>
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<td>Lung histology</td>
<td>Alveolar type 2 cell hyperplasia and alveolar proteinosis interstitial infl and fibrosis</td>
<td>Nonspecific interstitial pulmonitis</td>
<td>Alveolar type 2 cell hyperplasia ,accumulation of macrophages and proteinaceous material in airspaces</td>
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Mutations causing surfactant dysfunction

should be considered in infants with

- Severe unexplained lung disease in the newborn period
- Diffuse disease involving the entire lung on HRCT
- Histopathology that demonstrates findings of congenital alveolar proteinosis, desquamative interstitial pneumonia, nonspecific interstitial pneumonia, or chronic pneumonitis of infancy
- Electron microscopy demonstrating abnormal or absent lamellar bodies

Thank you
PATHOPHYSIOLOGY

- structural remodeling of the distal airspaces, leading to impaired gas exchange.
- believed to be the sequela of persistent inflammation;
- tissue injury with aberrant wound healing resulting in collagenous fibrosis
Fibroblasts, which are normally present in the attenuated interstitial spaces between alveoli and surrounding distal airways, play a key role in lung remodeling, which is characterized by proliferation and excessive elaboration of matrix molecules such as collagen.

Fibroblasts also affect remodeling through production of proteases, protease inhibitors, cytokines, and chemokines.
Remodeling of distal airspaces results in hypoxemia. Persistent hypoxemia results in pulmonary hypertension and vascular remodeling, leading to cor pulmonale. The increased work of breathing associated with reduced compliance results in increased energy expenditure, which, combined with the effects of inflammatory mediators, can result in cachexia.
Geographic hyperlucency — bronchiolitis obliterans, bronchocentric granulomatosis

Septal thickening — lymphangiomatosis, hemangiomatosis, microlithiasis

Ground glass opacification — lymphocytic interstitial pneumonitis, desquamative interstitial pneumonitis, hypersensitivity pneumonitis

Lung cysts and nodules — pulmonary Langerhans cell histiocytosis

Consolidation — aspiration, cryptogenic organizing pneumonia (formerly called bronchiolitis obliterans organizing pneumonia)
INTERSTITIAL LUNG DISEASE

- Diffuse developmental disorders
  - Acinar dysplasia
  - Congenital alveolar dysplasia
  - Alveolar capillary dysplasia with pulmonary vein misalignment (This is associated with a poor prognosis.)

- Growth abnormalities
  - Pulmonary hypoplasia
  - Chronic neonatal lung diseases (prematurity-related BPD and acquired chronic lung diseases in term infants)
  - Structural pulmonary changes with chromosomal abnormalities (eg, trisomy 21)
  - Abnormalities associated with congenital heart disease in otherwise healthy children

- Specific conditions with unknown etiology
  - PIG
  - NEHI

- SDMs and related disorders
  - *SFTPB* genetic mutations (PAP as dominant histologic pattern; see below)
  - *SFTPC* genetic mutations
  - *ABCA3* genetic mutations
  - Granulocyte-macrophage colony stimulating factor (GM-CSF) receptor mutations

- Genetic and/or familial disorders
  - SDMs and related disorders
  - Familial hypocalciuric hypercalcemia
  - Lysinuric protein intolerance
  - Farber lipogranulomatosis
  - Hermansky-Pudlak syndrome
ASPIRATION DUE TO VERNIX CASEOSA

*Results:* Our case report was associated with aspiration syndrome caused by an airway obstruction of vernix caseosa: a proteolipid biofilm synthesized by the fetus. A 23-year-old woman normally delivered a mature infant at term. The infantile oral cavity was filled with numerous aggregates of vernix caseosa. Two hours after his birth, the infant died from respiratory insufficiency.

*Conclusion:* Pregnant women with a diffuse pattern of high-level echoes in prenatal ultrasonography, suggesting the presence of massive vernix caseosa, should be transferred to a well-equipped institution that can administer inhaled nitric oxide and extracorporeal membrane oxygenation.

His ABG showed high pCO2 with normal SaO2

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