Pulmonary Hypoplasia from Prolonged Prelabor Rupture of Membranes: A Case Report

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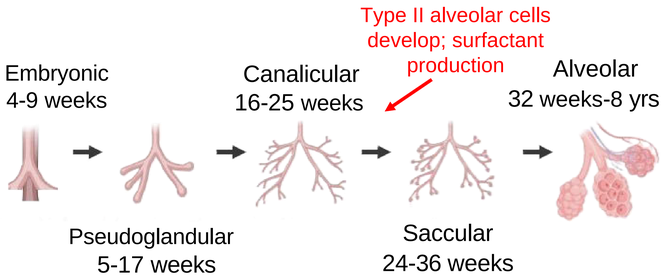
PURPOSE

To present a case study of a neonate affected by pulmonary hypoplasia (PH). Pathophysiology, clinical manifestations, a pre-delivery huddle tool, and implications for practice are explored.

BACKGROUND

- Rare congenital anomaly characterized by reduced or incomplete development of lung tissue and vasculature
- Develops in 33% of pregnancies with oligohydramnios
- Estimated perinatal mortality risk is 70%
- Management is challenging → balancing respiratory support to achieve adequate ventilation/perfusion while avoiding ventilator-induced injury and associated comorbidities

PATHOPHYSIOLOGY



- Total lung volume and postnatal lung function impacted
- Reduction in lung cells, airways, alveoli, and corresponding pulmonary vasculature→ severely limits pulmonary mechanics and gas exchange after birth
- Prolonged prelabor rupture of membranes (PPROM) exacerbates PH → loss of amniotic fluid results in insufficient volumes for ongoing fetal lung growth
- Severity of hypoplasia is inversely proportional to gestational age at time of rupture
- PPROM during canalicular phase compromises surfactant production and exacerbates hypoplasia

COMMON CLINICAL FINDINGS

Prenatal
- Decreased alveolar counts
- Low lung to body weight ratio
- Small, "bell-shaped" chest
- Low lecithin-to-sphingomyelin (L/S) ratio

Laboratory
- Respiratory acidosis
- Hypoxemia
- Hypercarbia

Postnatal
- Respiratory distress
- Surfactant deficiency
- Cyanosis
- Low oxygen saturation despite increasing FiO2

Radiographic
- Reduced lung expansion
- Atelectasis
- Air bronchograms
- Reduced pulmonary vascular markings

CASE PRESENTATION

Prenatal history: Significant for PPROM at 18 weeks gestation → severe oligohydramnios → pulmonary hypoplasia; Mother treated with 7-day course IV ampicillin and cefazolin and betamethasone x 2 doses (27 days prior to delivery); rescue steroid dose not given due to precipitous delivery

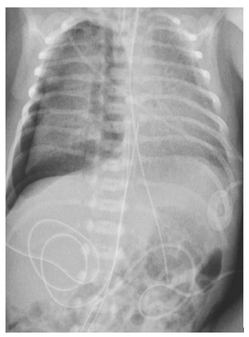
Delivery: 28 1/7 weeks gestation via C/S. Apgars 4, 7, and 8 at 1, 5, and 10 min. Required CPAP, PPV, and intubation in the delivery room (DR) with 100% FiO2 due to persistent low oxygen saturations, following NRP guidelines. Surfactant dose #1 (Poractant alfa 2.5 mL/kg) in DR

Hospital Course:

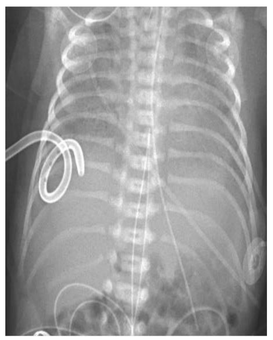
Day	Respiratory Support	X-ray interpretation	Lab Results	Medications/Treatments
1	High Frequency Jet Ventilator (HFJV) FiO2 100%	Consistent with pulmonary hypoplasia and respiratory distress syndrome (see images below)	Uncompensated respiratory acidosis Hypoxemia Blood culture sent	Nitric Oxide (iNO) 20ppm Surfactant dose #2 Ampicillin 50mg/kg Gentamicin 5 mg/kg
2	HFJV (max settings) FiO2 100%	R side pneumothroax	Blood culture positive for GBS Agalactiae	Chest tube placement Surfactant dose #3 Penicillin G 250,000 units/kg/d
6	SIMV-PRVC FiO2 60%		Respiratory acidosis resolved	Chest tube removed iNO discontinued
7+	Bubble CPAP 7 FiO2 50% Bubble CPAP 5-10 FiO2 30-50% over next 4.5 months	Improved aeration, persistent infiltrates with slightly reduced densities compared to previous images		DART course x 2 Inhaled corticosteroids Ibuprofen for PDA closure Multivitamin Ferrous sulfate

- Weaned to HFNC at 4.5 months → LFNC at 6 months of age
- Discharged home on LFNC
- Additional pertinent comorbidities during inpatient stay:
  - Hyperbilirubinemia requiring phototherapy
  - Oral candidiasis requiring topical antifungal treatment
  - Severe protein-calorie malnutrition requiring gastrostomy tube placed for home nutrition support

Day 1 - Admission



Day 2 - Pneumothorax



Day 2 – Chest tube

PRE-DELIVERY HUDDLE TOOL

Responsibilities Upon Notification of Plan to Deliver

Provider	RT	Delivery RN

• Review maternal and fetal history
• Review fetal scans with interpretation of PH severity
• Antenatal discussion with parents and OB providers
• Place initial orders using Estimated Fetal Weight (EFW)
  ○ Fluids
  ○ Surfactant dose
  ○ Initial HFJV settings
• Lead team briefing (see below)
	• Set up respiratory equipment in admission room (HFJV, iNO?)
	• Gather supplies needed in delivery room (DR)
	  ○ Endotracheal Tube (size per EFW)
	  ○ Intubation supplies
	• Remove surfactant from fridge and bring to DR
	• Blender set per protocol
	• Ensure PPV device present and functioning with appropriate mask size
		• Delivery bed ready following NRP
		• Pre-warmed
		• Stethoscope
		• Pulse oximeter probe and monitor
		• EKG leads
		• Chemical warming mattress
		• Plastic bag or wrap and hat
		• Verify equipment preparation

Team briefing discussion

Participants: Neonatologist, NNP/APP, lead RN, admit/delivery RN, RT, resuscitation team members if applicable

- Gestational age: _____
- Estimated fetal weight: _____g
- Review significant maternal history
  - ○ Length of rupture
  - ○ Medications received: antenatal corticosteroids, antibiotics, magnesium, tocolytics, etc.
  - ○ Chorioamnionitis or other infection risk (GBS, STI, etc.)
- Discuss severity of PH if known/antenatal testing completed
- Review DR resuscitation plan
  - ○ Anticipated need for airway – RT to prepare ETT, intubation supplies, and surfactant for administration in DR.
- Assign team members and delegate roles
- Alert x-ray of anticipated need for STAT imaging in the DR following intubation/prior to surfactant administration (per policy)

*Pre-delivery briefing expected to take less than 5 minutes.

DISCUSSION

- Early identification of infants with pulmonary hypoplasia allows for adequate preparation, including comprehensive antenatal counseling, multidisciplinary collaboration, and delivery team briefing.
- Future advancements in fetal MRI may help assess lung maturity in infants with PH.
- Resuscitation of an infant with PH is often extensive, as is the equipment needed for delivery.
- Structured huddles improve confidence, teamwork, communication, and patient safety.
- A customizable pre-delivery template is recommended to aid in preparation and ensure consistency.