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

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Abstract

Pulmonary hypoplasia (PH) is a rare, congenital anomaly involving reduced or incomplete development of lung tissues and vasculature. Approximately 33% of pregnancies complicated by oligohydramnios are associated with PH and the estimated perinatal mortality risk is 70%. The management of affected neonates is challenging. Clinicians must balance the need to provide adequate respiratory support to achieve proper ventilation/perfusion matching yet do so in the gentlest manner possible to avoid ventilator-induced lung injury and associated comorbidities. This presentation describes a case report of a preterm infant born with significant PH attributed to PPROM and severe oligohydramnios, as well as a discussion of the importance of multi-disciplinary collaboration in optimizing prenatal counseling, team readiness, and coordination of care.

Introduction/Clinical Course Summary

Prenatal history is significant for PPROM at 18 weeks' gestation with an hydramnios resulting in pulmonary hypoplasia. The infant was delivered at 28 weeks' gestation and remained hospitalized for close to 150 days requiring multiple respiratory support modalities including HFJV, SIMV-PRVC, CPAP, HFNC, and LFNC. Pertinent medications given during hospital course included 3 doses of exogenous pulmonary surfactant (with first dose administration in the delivery room), inhaled Nitric Oxide, dexamethasone, and antibiotics for GBS positive culture. Co-morbidities included PPHN, pneumothorax, PDA, BPD, bilateral grade 1 IVH, and poor growth with requirement of G-tube placement.

Suggested Process Improvement

When the delivery of an infant with PH is anticipated, a multidisciplinary team approach should be organized. Best practice encourages neonatal clinicians to allocate time for antenatal family counseling (Momin et al., 2024). Immediately preceding the birth of the infant, pre-delivery team briefings are necessary to guide the planning and anticipated resuscitation needs in the delivery room. Structured huddles improve confidence of the staff (Thomas et al., 2024), as well as improve teamwork, communication, and patient safety (Momin et al., 2024). When anticipating the delivery of an infant with PH, the resuscitation is often extensive, as is the equipment needed for delivery. To ensure consistent performance, a customizable template to aid in team planning and preparations is presented (Figure 1). This customizable guide calls for pre-delivery surfactant preparation with the understanding that it is reasonable for skilled teams to administer the first dose of surfactant in the delivery room. Zhang et al. (2024) concluded that providing surfactant within 30 minutes of birth significantly reduced the severity and incidence of RDS, which in turn reduced the risk of pneumothorax and neonatal mortality (Zhang, et al., 2024). A published cost comparison of prophylactic surfactant administration compared to standard treatment is also presented (Yao et al., 2022). Continued efforts in neonatal respiratory support strategies, antenatal interventions, and individualized treatment approaches will be crucial in mitigating the effects of pulmonary hypoplasia and improving the prognosis for affected infants. Utilizing the tool provided as a framework to organize a pre-delivery team huddle when anticipating the high-risk delivery of an infant with suspected pulmonary hypoplasia will be

beneficial in setting expectations, ensuring adequate preparation, fostering effective teamwork, and will ultimately result in the best outcome for the neonate.

Figure 1: Pre-Delivery team briefing template

Role:	**LIP/APP**	**RT**	**Delivery RN**

Responsibilities when notified of plan to deliver:	• Review maternal and fetal history (including fetal scans with interpretation of severity of pulmonary hypoplasia).
	• Antenatal discussion with parents and OB providers (if hasn't already happened during antepartum stay).
	• Place initial orders (using 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)
		  ○ ETT (size based on EFW)
		  ○ Intubation supplies
		• Remove surfactant from fridge (if able) 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
			  ○ Pulse oximeter probe and monitor
			  ○ EKG leads
			  ○ Chemical warming mattress
			  ○ Plastic bag or wrap and hat
			• Remove surfactant from fridge if RT unable, give to RT for administration

Team briefing discussion
Participants: Neonatologist, Neonatal Nurse Practitioner, lead RN, admit/delivery RN, RT, resuscitation team members if applicable (medications, recorder, etc.)

- Gestational age: _____
- Estimated Fetal Weight: _____g
- Review significant maternal history
    - Length of rupture
    - Medications received…antenatal corticosteroids, antibiotics, magnesium, etc.
    - Chorioamnionitis or other infection risk identified (GBS, STI, etc.)
- Discuss severity of pulmonary hypoplasia if known/antenatal testing completed
- Review DR resuscitation plan
    - Anticipated need for airway – RT to prepare ETT (size based on EFW), and intubation supplies. Prepare for Surfactant 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 hospital policy)

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

Figure 2. Initial Chest X-Ray, Day 1 of Life

FINDINGS: Lines and Tubes: Endotracheal tube tip 10 mm superior to the carina. UVC at the inferior atrial caval junction. UAC at T8. Chest: Low lung volumes with diffuse granular pulmonary opacity and patchy opacities in the right lung. No large effusion or pneumothorax. Cardiothymic contours grossly unremarkable. Abdomen: No dilated bowel loops, pneumatosis or portal venous gas. Reprinted with permission.

References

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