Case Report

Dry Lung Syndrome: The Positive End of the Oligohydramnios Spectrum

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Oligohydramnios secondary to mid-trimester preterm premature rupture of the membranes can result in a range of abnormalities from functional hypoplasia of the lungs to structural hypoplasia and fetal compression syndrome. Here, we discuss two infants born at 24-week gestation with a history of severe oligohydramnios requiring extensive resuscitation with high ventilation pressures at birth. One had dry lung syndrome (DLS) and after adequate resuscitation did well. The second case had severe pulmonary hypoplasia complicated by recurrent air leaks and succumbed to it. Management of DLS is primarily supportive; these neonates require adequate respiratory support to open the collapsed airway at birth and optimal ventilation in the early neonatal period to facilitate smooth recovery.

Keywords: Dry lung syndrome, oligohydramnios, pulmonary hypoplasia

INTRODUCTION

Mid-trimester preterm premature rupture of the membranes (PPROM) occurs in approximately 0.4%–0.7% of pregnancies.[1] Infants born with PPROM have poor outcomes attributable to the complications of prematurity, sepsis, and consequences of oligohydramnios, that is, “oligohydramnios sequence,” which includes pulmonary hypoplasia (PH) with pulmonary hypertension (PHT) and fetal compression syndrome. However, subset of these patients has only functional lung hypoplasia. This was first described by McIntosh[2] and he coined the term “dry lung syndrome (DLS)”. DLS has been ascribed to airway collapse and external compression secondary to oligohydramnios. Infants with DLS respond well to early high ventilator pressures to open the airways. Here, we describe two cases of oligohydramnios sequence and discuss the pathophysiology behind the spectrum of respiratory outcomes in these infants.

CASE REPORTS

Case 1

A female infant was born at 24-week gestation with a history of leaking liquor for 7 days with amniotic fluid index of 0.84 cm. The antenatal history was suggestive of chorioamnionitis. There were no other significant morbidities during the antenatal period. Course of antenatal steroids was completed 8 days prior to delivery. She was born with Apgar scores of 2 and 3 at 1 and 5 min, respectively. The birth weight was 700 g. She required extensive resuscitation at birth and needed high ventilator support. While on a peak inspiratory pressure (PIP)/positive end-expiratory pressure 30/5 and fraction of inspired oxygen (FiO2) 100%, the blood gas was pH 6.9, pCO2 70, and base excess (BE) 19. Initial chest X-ray (CXR) showed uniform dense opacities in both lung fields suggesting collapsed airway [Figure 1a]. She required high-frequency oscillatory ventilation (HFOV) with mean airway pressure (MAP) of 15 and two doses of surfactant (Survanta®, Abbott Laboratories, Abbott Park, IL, USA). Six hours of infant life showed dramatic improvement with normal blood gases. FiO2 was gradually weaned to 22%, and the chest radiograph revealed well-expanded lung fields with minimal reticulogranular pattern [Figure 1b]. She was successfully extubated to continuous positive airway...
pressure and continued to remain stable with noninvasive ventilation and was discharged home at corrected gestation of 41 weeks. Her follow-up is arranged for growth and neurodevelopmental assessment.

Case 2
A male surviving twin infant of dichorionic diamniotic pregnancy was born at 24-week gestation with an antenatal history of PPROM for 7 weeks. There was antenatal anhydramnios for both twins at 20 weeks with a demise of one of the twin at 23 weeks of gestation. Mother completed a course of antenatal steroids 3 days before the delivery. The baby was born through precipitous vaginal delivery with Apgar scores of 2, 2, and 6 at 1, 5, and 10 min, respectively, and weighed 550 g. At birth, he was intubated and ventilated with pressures of 25/5 and FiO\textsubscript{2} 100%. In the neonatal intensive care unit, the blood gas (pH – 7.068; pCO\textsubscript{2} – 59; and BE – 13.5) showed mixed acidosis and CXR revealed small volume lungs [Figure 2a]. His respiratory support was maximized with HFOV following two doses of the surfactant. The bedside two-dimensional echocardiogram showed severe PHT with pulmonary pressures of 35 mmHg, this was managed with inhaled nitric oxide. His postnatal course was complicated by the recurrent episodes of pneumothorax requiring multiple chest drain insertions [Figure 2b]. In spite of all the above endeavors infant succumbed to respiratory failure secondary to severe PH. Table 1 highlights the comparison between the two cases of oligohydramnios sequence.

### DISCUSSION
Normal fetal lung growth depends on several mechanical factors such as adequate lung fluid volume, normal fetal breathing movements, and optimal intrathoracic and intrauterine space. The fetal lung secretes fluid by active transport across the pulmonary epithelium and this fluid serves as an internal stent for the lung and maintains the positive intrapulmonary...
pressure.[1] Fetal breathing movements regulate the volume of the fluid by modulating the resistance offered to the outflow of the lung fluid. The distending pressure generated by the lung fluid and cyclic stretch of the lung are the two major determinants of normal fetal lung development.

There is a known association between oligohydramnios and PH but the mechanism is not clear. It has been postulated that (1) oligohydramnios reduces the size of the intrathoracic cavity limiting the space available for the lung growth due to the pressure exerted on the fetal chest and abdomen; (2) hampering the fetal breathing movements by prolonged thoracic compression; and (3) reduced intrapulmonary pressure due to the increased efflux of lung fluid from the airways to the amniotic space.[4]

Severe oligohydramnios of ≥14 days with onset of PPROM before 24-week gestation is associated with PH in 25% of the cases.[3] Early PPROM (defined as the rupture of fetal membranes prior to 28 weeks of gestation) disrupts the pseudoglandular (6–16 weeks) or canalicular phase (16–25 weeks) of the lung development.[1] Oligohydramnios may manifest in a wide spectrum of abnormalities ranging from DLS to PH or fetal compression syndrome. At the severe end of the spectrum are infants with Potter sequence and skeletal deformities.[6] This group of infants have very high mortality. In a study by Thibeault et al.,[7] 86% of the neonates with joint contractures and need for ventilation died, compared to 52% of those without skeletal deformities. At the milder end of the spectrum is functional hypoplasia, which occurs once the lung has leaked dry of the normal volume of the lung fluid and collapsed. McIntosh.[2] discussed four neonates with antenatal history of PPROM, who had severe respiratory distress at birth but responded rapidly to high ventilation pressures during resuscitation and improved over the next 24–48 h of life. This was an unexpected clinical outcome for infants expected to have PH secondary to severe oligohydramnios.

The incidence of DLS is approximately 3.5% compared to PH, which varies from 7.5% to 9.6% in infants with oligohydramnios sequence.[2,8]

In the extremely preterm infants described both had clinical presentation suggestive of progressive hypercarbia and hypoxia. Giving surfactant makes a dramatic improvement in the acid base status of an infant with respiratory distress syndrome (RDS) but in infants with DLS due to the collapse of the airway higher pressures are needed to improve the air entry and thereby oxygenation. In the background of oligohydramnios, the early usage of high PIP will inflate the lung and overcome the functional hypoplasia.[3] In the two cases described, the mothers had received a complete course of antenatal steroids, it is expected that the severity of RDS would be attenuated. The surfactant was given to both the cases, but the response in the first case was predominantly after the increase in pressures. The CXR for the first case showed a predominantly collapsed lung with mild granularity and the white out lung improved rapidly with high MAP during the first 24–48 h of life. The chest radiograph for the second case demonstrated small volume lungs and a bell-shaped chest. In the absence of clinical response to high pressure ventilation with surfactant therapy and recurrent pneumothoraces a diagnosis of moderate-to-severe PH was considered. This infant had progressive deterioration and eventual demise. None of our case had sepsis. Both the cases discussed fall in the mild-to-moderate range of the spectrum of oligohydramnios sequence and did not demonstrate the classic facial or skeletal features of Potter sequence as seen in infants on the severe end of the spectrum.

**Conclusion**

The management of fetuses complicated with antenatal oligohydramnios should ideally be at a tertiary neonatal center with close monitoring. An aggressive respiratory support with high MAP is crucial to management of the respiratory failure. Although it is difficult to predict whether the newborn infant has DLS or PH at birth, the rapid and sustained clinical improvement will indicate the possibility of DLS.

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**Conflicts of interest**

There are no conflicts of interest.

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