

Fetal Pulmonary Sequestration Presenting as Unilateral Idiopathic Hydrothorax A Case Report

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A case of fetal pulmonary sequestration presented as idiopathic unilateral hydrothorax. (J Reprod Med 1995; 40:319-322)

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Introduction

Fetal pleural effusion occurs in about 1 in 15,000 pregnancies.¹ Differential diagnosis includes immune or nonimmune hydrops, with chylothorax the most common, as well as abnormalities of lung or chest development. Whether or not a mass is visualized in the thoracic cavity, one must consider the possibilities of diaphragmatic hernia, cystic adenomatoid malformation, mediastinal teratoma, neuroblastoma and pulmonary sequestration.

The association of fetal pleural effusion with a perinatal mortality rate of 50–82%,² even in the absence of other fetal abnormalities or defects, makes a compelling plea for intervention on the behalf of a fetus with this condition. Many of these neonates die of the sequelae of pulmonary hypoplasia. Therefore, intrauterine drainage of the effusion may be attempted to promote better intrauterine pulmonary development and to improve visualization of intrathoracic organs.

Case Report

A 30-year-old, white woman, gravida 2, para 1001, had her estimated date of confinement determined by 10.1-week crown-rump length ultrasound evaluation. She had an unremarkable prenatal course until 33 weeks' gestation, when she was noted to have a fundal height of 36 cm. Ultrasound evaluation revealed a single intrauterine gestation with the size consistent with the dates. A large fetal left pleural effusion was seen displacing the fetal heart to the right and compressing the right lung (Figure 1A). Severe hydramnios was also noted. There was no evidence of diaphragmatic hernia or other structural abnormality.

At this time the differential diagnosis included chromosomal abnormalities, viral infection, fetal-maternal hemorrhage, and chylothorax or hydrothorax of undetermined etiology. The patient had declined α -fetoprotein screening; she now consented to fetal blood sampling. Maternal serum studies, including liver function tests, uric acid, electrolytes, complete blood count, toxoplasmosis and parvovirus titers, were all unremarkable (Table I). The Kleihauer-Betke test was negative for fetal cells. At 34.4 weeks' gestation the amniotic fluid index was 32. Therapeutic amniocentesis yielded 1,700 mL of amniotic fluid. At 35.1 weeks' gestation, repeat amniocentesis, yielding 2,300 mL of amniotic fluid, produced a final amniotic fluid index of 21 cm. Thoracocentesis was performed, yielding a total of 117 mL of pleural effusion fluid, after fetal paralysis



Figure 1
(A) Left fetal pleural effusion. (B) Complete expansion of the left lung; heart returned to normal position.

was accomplished with pancuronium, given intravenously via the umbilical vein (Table II). Following thoracocentesis, the left and right lungs were seen to be completely expanded, and the heart returned to the normal position (Figure 1B). The pleural effusion was confirmed to be left unilateral. The appearance of fetal lung parenchyma was homogeneous, without evidence of cystic adenomatoid malformation or another mass. The fetal mediastinum was without abnormality. Two-dimensional and Doppler echocardiography revealed a structurally and functionally normal heart. Doppler flow studies of the umbilical artery were normal. No other structural anomalies were seen.

Estimated fetal weight was 2,238 g, and fetal biometry was indicative of mild asymmetric growth impairment. Daily serial ultrasonography revealed gradual partial (50%) reaccumulation of hydrothorax. Mild hydramnios persisted, with an amniotic fluid index of 21 cm. At 35.4 weeks' gestation the patient arrived at the obstetrics department with spontaneous rupture of the membranes and progressed to vaginal delivery of a live female infant

with Apgar scores of 6 at one minute and 8 at five minutes and weighing 2,281 g. Arterial cord blood gas revealed a pH of 7.30.

A chest radiograph of the infant revealed a marked pleural effusion on the left, with the mediastinum and heart shifted to the right. Thoracocentesis was performed on days 1 and 3 of life, yielding 60 mL of fluid on each occasion; the second required placement of a thoracostomy tube for pneumothorax. The infant was placed on nasal continuous positive airway pressure, with a marked improvement in symptomatology and hypercapnia. Ultrasound and computed tomography scans revealed a mass lesion in the left lower lobe of the chest (Figure 2). At 17 days of life the infant underwent thoracotomy with resection of the left lower lobe and left lingula, which were nonaerated and without evidence of communication to the main bronchus. The thoracotomized specimens measured $3.2 \times 3.2 \times 2.0$ cm and $3.6 \times 2.8 \times 1.2$ cm, respectively, and were initially judged to represent cystic adenomatoid malformation. The infant did very well after surgery and was discharged at age 25 days in good condition.

The pathologic specimens were reviewed by pathologists from three institutions, with the final diagnosis determined to be extralobar pulmonary sequestration, with an associated hypoplastic lobe versus a left lower lobe separated from the tracheobronchial tree during early development.

Discussion

Pulmonary sequestration is a rare congenital abnor-

Table I Results of Percutaneous Umbilical Blood Sampling

Parameter	Finding
Karyotype	46,XX, without abnormalities
Fetal CBC	
WBC	6,200/cm
Hemoglobin	12.3 g/dL
Hematocrit	35.9%
Platelet count	Adequate
Cord gas	
pH	7.37
pCO ₂	43
pO ₂	33
Fetal liver function	
Total protein	3.4
Albumin	2.5
Aspartate aminotransferase	16
Alanine aminotransferase	2
Cytomegalovirus titers	Negative
Parvovirus	Negative

mality with a broad definition encompassing "all congenital lung anomalies in which there is abnormal connection of one or more of the four major components of lung tissue, namely tracheobronchial airway, lung parenchyma, arterial supply and/or venous drainage."³ Most are detected neonatally or during childhood, suspected only after repeated respiratory tract infections. If asymptomatic, they may be discovered only incidentally well into adulthood.

Approximately 75% of all sequestrations are of the intralobar variety.⁴ Intralobar sequestration is seen more commonly than extralobar sequestration and features a nonfunctional mass of lung parenchyma, encapsulated by the same visceral pleura as the surrounding normal lung parenchyma but lacking communication with the tracheobronchial tree and with an aberrant blood supply from the descending thoracic aorta, upper abdominal aorta, and celiac, splenic and/or other arterial supply. However, it has venous drainage via the pulmonary veins. Intralobar sequestration is less likely to be seen in conjunction with other congenital anomalies than is the extralobar variant. Its most common manifestation is (repeated) respiratory tract infections.

By contrast, extralobar sequestration has an incidence of approximately 25% of classic pulmonary sequestrations, is covered by its own visceral pleura and is often (65% of cases) found along with other congenital abnormalities, such as cardiac anomalies; gastrointestinal anomalies, such as diaphragmatic hernia; or less serious abnormalities, such as accessory spleen. Extralobar masses also re-

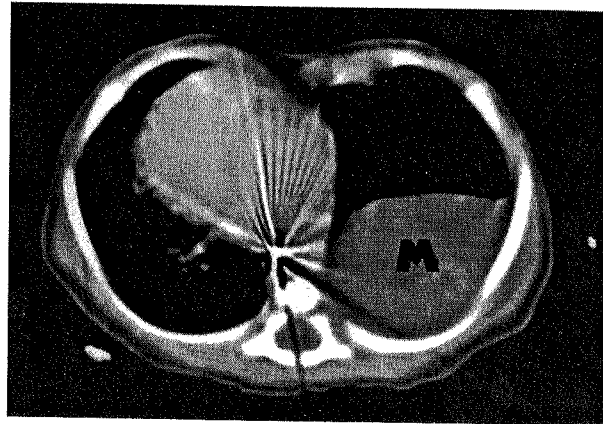


Figure 2
Computed tomographic scan; note the mass (M).

ceive an anomalous arterial supply; however, their venous drainage is usually via systemic veins, such as the azygous, hemizygous and portal systems.³ Indications of the condition may include polyhydramnios, pleural effusion or hydrops prenatally and neonatal anasarca postnatally. Other postnatal manifestations of extralobar pulmonary sequestration include cyanosis, dyspnea, respiratory distress, hemoptysis, hemothorax and feeding difficulties, which may be reflections of pulmonary hypoplasia, congestive heart failure and/or pulmonary hypertension.

According to a recent review of antenatal diagnosis of pulmonary sequestration, "the prognosis for a fetus with a sequestration appears unpredictable."⁵

In their series, mortality was 50% before or shortly after birth. In the case presented above, the duration of the hydrothorax was unknown. Depending upon the severity of the condition, pulmonary hypoplasia or, in the worst case scenario, neonatal death is the anticipated outcome.

We performed fetal thoracocentesis in order to decompress the lungs, thus affording a better evaluation of the intrathoracic structures and enabling us to establish a diagnosis. In addition, it was reasoned that expansion of the fetal lung should be helpful in preventing pulmonary hypoplasia. We intended to perform serial thoracocentesis weekly as warranted, based on fluid reaccumulation. The fact that the first postnatal thoracocentesis yielded only half the volume drained prenatally indicates that more intrathoracic space was available for lung expansion. Therefore, the potential existed for complete resolution of the condition with serial prenatal

Table II Fetal Left Pleural Effusion Fluid Obtained by Thoracocentesis

Parameter	
Protein	1.79 g/dL
Triglyceride	6 mg/dL
Cell count	
RBC	12 intact
WBC	7
Mature neutrophils	12%
Lymphocytes	18%
Eosinophils	2%
Macrophages	68%
Electrolytes	
Sodium	136 mmol/L
Potassium	3.9 mmol/L
Chloride	109 mmol/L
Glucose	80 mg/dL

thoracocenteses had the fetus remained undelivered. One may argue that this condition might be self-limited and therefore not justify intervention. We believe, rather, that intervention in selected cases may be not only justifiable but potentially life saving. Because of the rarity of this condition and considering the ethical issues, randomized clinical studies have not heretofore been undertaken (nor are they likely to be).

We conclude that with careful evaluation on a case-by-case basis, similar treatment should be offered to these patients. We have demonstrated the possibility of an improved postnatal outcome with early diagnosis and prenatal intervention, including therapeutic amniocentesis and fetal thoracocentesis. We would also urge that physicians encountering such patients report the results of their experiences to contribute to the general understanding of the pathophysiology of this condition as

well as the sequelae of various management schemes.

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