

MEDICAL INTELLIGENCE



LONG-TERM IN UTERO DRAINAGE OF FETAL HYDROTHORAX

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FETAL hydrothorax has a perinatal mortality of 57 to 100 percent. Mortality is higher when the hydrothorax occurs in association with generalized hydrops than when it is an isolated finding.^{1,2} Intrathoracic compression of the developing lung produces pulmonary hypoplasia, which is the main cause of perinatal death in fetuses with hydrothorax.^{1,2} Large pleural effusions may cause hydramnios by interfering with fetal swallowing and hydrops by vena caval obstruction and cardiac compression.^{3,4} Chylothorax, the most common cause of hydrothorax in neonates,⁵ can be diagnosed by demonstrating chylomicrons in pleural fluid after alimentation; however, the differentiation of this entity from other causes of hydrothorax during intrauterine life is difficult. Although some investigators have claimed that this diagnosis can be made during fetal life by the demonstration of high mononuclear-cell counts in aspirated pleural fluid,^{6,7} they have not confirmed chylothorax postnatally. Despite detailed investigation with ultrasound and fetal blood sampling, the cause of nonimmune hydrops that is not associated with cardiac abnormalities or dysrhythmia remains unexplained in more than 50 percent of cases.⁸ Congenital chylothorax has thus been suggested as the primary cause of many cases of unexplained hydrops.²

The aspiration of fetal hydrothoraxes facilitates neonatal resuscitation if performed immediately before delivery.⁹ Because the fluid reaccumulates in the fetus within 6 to 48 hours after aspiration,^{4,8} long-term drainage is required to prevent pulmonary hypoplasia and to reverse hydrops and hydramnios. Harrison et al. found that relief of experimental intrathoracic compression allowed lung growth and neonatal survival in lamb fetuses, whereas continued compression led to fatal pulmonary hypoplasia.¹⁰ We inserted pleuroamniotic shunts for long-term drainage in eight human fetuses, and found that hydramnios resolved in

six and hydrops, initially present in five, disappeared in three. Six infants survived, five of whom had no respiratory difficulties at birth. Lung reexpansion was seen after in utero shunting in the six survivors, but not in the two who died.

METHODS

Eight women with singleton pregnancies were referred with unexplained fetal pleural effusions, which had been detected during an investigation for hydramnios. The assessment included detailed ultrasonography and fetal echocardiography to exclude other abnormalities. Samples of fetal blood were obtained either fetoscopically¹¹ or by ultrasound-guided aspiration¹² and were investigated for cytogenetic, hematologic, and virologic abnormalities.⁸

Thoracentesis was performed under ultrasound control with use of a 20-gauge needle.¹³ Pleuroamniotic shunts were established with double-pigtail nylon catheters (Rocket of London, United Kingdom), with external and internal diameters of 0.21 and 0.15 mm, respectively. A 15-cm metal trocar and cannula were introduced transamniotically under ultrasound visualization through the fetal midthoracic wall into the effusion (Fig. 1). An introducer rod was then inserted to position one end of the catheter within the pleural cavity. The cannula was then withdrawn into the amniotic cavity, and the other end placed outside the chest wall.

RESULTS

All patients had gross fetal pleural effusions with hydramnios. Five had hydrops fetalis (Table 1). In the three with unilateral effusions, there was deviation of the heart and mediastinal shift to the contralateral side. One fetus (Patient 3) had other abnormalities (i.e., trisomy 21 and an atrioventricular defect) that were not discovered until after the shunt had been inserted. The karyotype was determined several days later from lymphocyte culture, and the cardiac defect became apparent after removal of the pleural fluid allowed restitution of normal mediastinal topography.

The pleural fluid reaccumulated one and seven days after aspiration in the two patients (Patients 1 and 4, respectively) who underwent aspiration as an initial procedure.

Twelve pleuroamniotic shunts were placed. One was noted to be free in the amniotic cavity a week later, and a second shunt was inserted. Eleven functioned until delivery (median, 2.5 weeks; range, 1 to 14; Table 1). In six fetuses, the pleural effusions almost completely resolved, except for a rim of fluid due to the presence of the coiled catheter (Fig. 2), and hydramnios disappeared. No reaccumulation of fluid was seen on serial scans. Three of the six had hydrops, which resolved after the insertion of the shunt. All six infants survived, and five had no respiratory difficulties at birth. One required ventilation before spontaneous respiration was established.

Fetal hydrothoraxes did not resolve in two patients (Patients 6 and 7), both of whom had hydrops. Since massive hydramnios precluded shunt insertion at 30 weeks' gestation in Patient 6, bilateral thoracenteses were performed, but fluid reaccumulated in the next 24 hours. During the next week, a total of 2.25 liters of amniotic fluid was removed in two amniocenteses. We were then able to use the trocar to reach the nearest side of the fetal chest for a unilateral shunt insertion.

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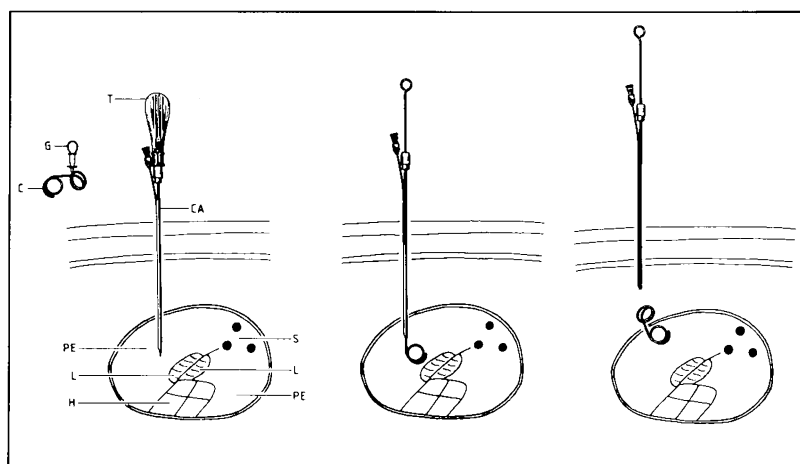


Figure 1. The Technique of Pleuroamniotic Shunt Insertion.

The instruments consist of a double-pigtail catheter (C) of 6-French nylon tubing with radiopaque stainless-steel inserts at each end and lateral holes around the coil, a guidewire (G) for straightening, and the trocar (T) and cannula (CA) with two introducers. The trocar and cannula are inserted transabdominally and transamniotically into the pleural effusion (PE) (left panel). The guidewire is then straightened, the catheter introduced into the cannula, and the wire removed. A short introducer rod is then inserted to position half the catheter within the fetal hemithorax (center panel). The cannula is then withdrawn into the amniotic cavity, and a long introducer rod is inserted to position the other half of the shunt in the amniotic cavity (right panel).

H denotes heart, S spine, and L lungs.

This produced a minor decrease in the effusion on that side only. Spontaneous vaginal delivery occurred a week later, but the neonate's course was complicated by birth asphyxia, bilateral pleural effusions, and persistent fetal circulation. The infant died on day 19, and permission for a postmortem examination was denied.

Only left-sided shunt insertion was possible initially in Patient 7 because of the fetal position; simple aspi-

tensive fibrinous exudates and adhesions were found within the pleural cavities.

Five infants were delivered vaginally, and three by cesarean section. The shunts were removed after birth in five of the six surviving infants. Pleural effusion recurred in one, who required a postnatal thoracentesis before the condition resolved. In another the shunt was not seen externally at delivery, and a chest film showed it to be just under the chest wall, having pre-

ration was done on the right. The left hemithorax, which had partially drained after the shunt insertion, refilled subsequently. An analysis of the fluid reaspirated from the left side showed fetal squames, confirming that amniotic fluid had entered the chest through the valveless shunt, possibly because of a decrease in intrathoracic pressure caused by the right-sided thoracentesis. Fluid reaccumulated on the right side over the next two weeks, producing mediastinal shift. Right-sided shunting was then performed, but it only partially reduced the effusion. Neither lung reexpanded to fill its hemithorax. Preterm labor occurred, and a cesarean section was performed a week later, but the infant had severe ventilatory difficulties and died 40 minutes after birth. Post-mortem study confirmed the presence of bilateral pulmonary hypoplasia with a ratio of lung weight to body weight¹⁴ of 0.0075 to 1. Ex-

Table 1. Results of Pleuroamniotic Shunting in Eight Patients with Fetal Hydrothorax.*

PATIENT No.	LOCATION OF FETAL HYDROTHORAX	GESTATIONAL AGE		PREVIOUS NORMAL SCAN	HYDRAMNIOS	HYDROPS	FETAL KARYOTYPE	SHUNT INSERTION		NEONATAL RESPIRATION
		AT DIAGNOSIS	AT DELIVERY					LOCATION	GESTATIONAL AGE	
		weeks							weeks	
1	Left	25	39	Yes	+	—	46,XY	Left	27	Good
2	Left and right	26	32	No	++	+++	46,XY	Left	28	Good
								Right	28	
3	Right	32	34	Yes	++	—	47,XX,+21	Right	32	Good (Down's syndrome and cardiac defect)
4	Right	20	39	Yes	+	—	46,XY	Right	25	Good
5	Right larger than left	34	39	Yes	+++	+++	46,XY	Right	35 (fell out)	Good
								Right	36	
6	Left and right	25	32	No	+++	++	46,XY	Right	31	Poor (neonatal death on day 19)
7	Left and right	21	32	No	++	+	46,XY	Left	29	Poor (neonatal death <1 hr)
								Right	31	
8	Left and right	31	34	No	++	++	46,XX	Left	32	Good
								Right	32	

*+ denotes mild, ++ moderate, +++ severe, and — absent.

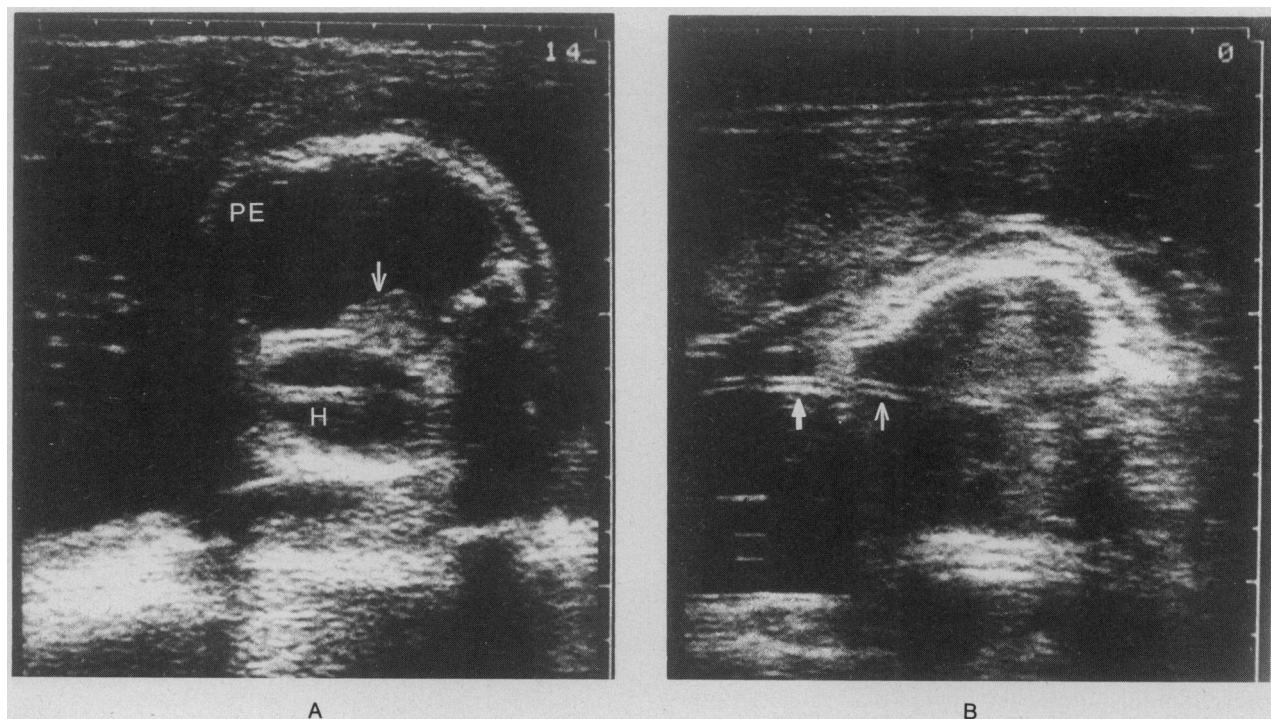


Figure 2. Ultrasound Cross-Sectional Views of the Fetal Chest.

Panel A shows a large, right-sided pleural effusion (PE) with mediastinal shift, displacement of the heart (H), and a markedly compressed right lung (arrow). In Panel B, 24 hours after shunt insertion, the effusion has drained and the lung has reexpanded. Note the intrapleural (thin arrow) and extrapleural (thick arrow) portions of the catheter in situ.

sumably retracted with chest-wall growth and respiratory movements. No surgical intervention was thought necessary. One infant had transient tachypnea after birth due to a small pneumothorax resulting from a delay in the clamping of the drain at delivery. After oral feeding was instituted, pleural fluid was visualized in only one infant (Patient 8), who required unilateral drainage of a pleural effusion at seven days of age. The fluid was opalescent, a characteristic consistent with chylothorax.

DISCUSSION

Pleuroamniotic shunts were inserted in eight fetuses with hydrothoraxes, two of whom died. In the six survivors, shunting produced nearly complete resolution of the pleural effusion and lung reexpansion. The lungs failed to reexpand after shunting in both patients who died. Severe hypoplasia was confirmed in one, but a postmortem study was not performed in the other. The survival rate of 75 percent in the fetuses with hydrops induced by gross hydrothoraxes was better than that previously reported,⁵ especially since five also had hydrops.

The absence of neonatal respiratory difficulty in most of the surviving infants largely reflects the absence of effusion at delivery. Although removal of pleural fluid before birth is known to facilitate neonatal resuscitation,^{9,15} the fact that there is expansion of the lung to refill the hemithorax immediately after

aspiration in the third trimester suggests that marked pulmonary hypoplasia has not occurred and that intrathoracic compression has been relatively brief.⁹ To prevent or ameliorate lung hypoplasia, long-term drainage would be necessary soon after the occurrence of the hydrothorax and would be effective during the second trimester, the critical period for lung development.¹⁶ Long-term drainage of an intrathoracic cyst associated with a type 1 cystic adenomatoid malformation in the second and third trimesters appears to have prevented pulmonary hypoplasia in two reported cases.^{17,18} In our study, the neonates who died of pulmonary hypoplasia (histologically proved in one) had bilateral hydrothoraxes from the 25th week of gestation or earlier. Shunts were inserted much later (at 29 to 31 weeks) because of late referrals. Thus, treatment delays may have deleterious effects on fetal lung growth.

In addition to reducing lung compression, pleuroamniotic shunting reversed hydrops and hydramnios in all patients without severe lung hypoplasia. It is likely that drainage of fetal hydrothorax, by reducing compression on other intrathoracic structures, allows the return of normal swallowing and circulatory function. This minimizes the risks of hypoxia and acidemia associated with hydrops and of preterm labor and premature rupture of the membranes due to hydramnios.

Long-term drainage by serial aspiration has been

attempted in view of the rapid reaccumulation of fluid after thoracentesis.^{4,8} Two to five thoracenteses have been reported to resolve fetal hydrothorax without recurrence,^{3,6,19} but spontaneous resolution may occur in mild cases.² Benacerraf and Frigoletto argue that shunts are prone to blockage because pleural fluid is highly viscous.³ Weiner et al. considered blockage of consecutive shunts in one fetus to be due to cellular debris and the low daily flow rate, but they used 22-gauge catheters.²⁰ None of the larger shunts used in our series became blocked, although one was drawn into the chest and one was pulled into the amniotic cavity.

Patient 3, who had Down's syndrome and a cardiac malformation, illustrates the need to obtain the fetal karyotype as quickly as possible by culture of fetal lymphocytes and the importance of reassessing cardiac structure sonographically after shunting and the return of the heart to its normal position.

The main benefits of pleuroamniotic shunting in fetal hydrothorax appear to be the facilitation of neonatal resuscitation, resolution of hydrops and fetal hydrops, and possibly, if drainage is of long duration, prevention of pulmonary hypoplasia. Since chylothorax in surviving infants carries a good prognosis, this series supports a role for pleuroamniotic shunting to improve neonatal survival in fetal hydrothorax associated with other congenital defects.

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