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Shunt-based interventions: Why, how, and when to place a shunt

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A B S T R A C T

The broad categories of surgical fetal therapy can be separated into either open surgical techniques or minimally invasive endoscopic/ultrasound-guided techniques that require only puncture of the uterus with single or multiple small ports. Benefits of fetoscopic or ultrasound-guided fetal intervention include decreased uterine irritability, decreased incidence of preterm labor, and avoidance of risks associated with hysterotomy and commitment to cesarean delivery for future pregnancies. Fetal abnormalities potentially amenable to ultrasound-guided drainage techniques include thoracic fluid-filled lesions and lower urinary tract obstruction.

1. In-utero shunting for thoracic pathologies

The fetal thoracic cavity represents a closed space that has minimal capacity to expand in response to space-occupying masses. Masses such as large congenital pulmonary airway malformations (CPAMs), or fluid accumulation such as pleural effusions, may result in intrathoracic compression of the developing lungs, mediastinal structures, and heart, resulting in lung hypoplasia and fetal hydrops (Fig. 1A). The risk for pulmonary hypoplasia or cardiovascular impairment is related to the timing and degree of compression of these structures. Drainage of fluid-based lesions via shunt placement or thoracentesis leads to a decrease in volume of the lesions with beneficial effects on fetal hemodynamics, lung growth, and long-term survival (Fig. 1B).

2. Pleural effusions

Fetal pleural effusions (PEs) can present as part of a more generalized picture of non-immune fetal hydrops or as an isolated sonographic finding. They are divided into primary and secondary causes. Primary PE is a lymphatic malformation whereas secondary PE is usually due to associated anomalies such as aneuploidy, cardiac defects, anemia, or infections. Primary PE occurs in ~1 in 12,000 pregnancies with a 2:1 male:female ratio [1]. Gestational age at presentation is generally <32 weeks and the presence of hydrops is associated with high perinatal mortality ranging from 36% to 40% [2–6]. Isolated PE in the absence of fetal hydrops still poses a threat to the developing fetus by acting as a space-occupying lesion. The risk for pulmonary hypoplasia or cardiovascular impairment is directly related to the effusion volume. The laterality of the PE does not appear to influence outcome, but bilateral effusions may result in greater pulmonary and cardiac compression [6].

3. Selection criteria for treatment of fetuses with pleural effusion

Selection criteria for PE treatment by thoracoamniotic shunting (TAS) requires a primary PE etiology, normal karyotype, absence of other significant congenital anomalies, negative viral studies, normal echocardiogram, rapid recurrence of the effusion after thoracentesis, and gestational age of <32 weeks [1,3,7]. However, there may be a role for shunting up to 37 weeks to improve postnatal resuscitation efforts [4]. Minimal evaluation should include confirmation of a primary lymphocytic effusion by cell count (>95% mononuclear cells) and polymerase chain reaction-based infectious studies to rule out parvovirus, cytomegalovirus, herpes virus, and toxoplasmosis from the effusion fluid obtained at amniocentesis or initial diagnostic thoracentesis.

4. Thoracentesis and thoracoamniotic shunt technique

Thoracentesis utilizes a 20–22-gauge spinal needle under

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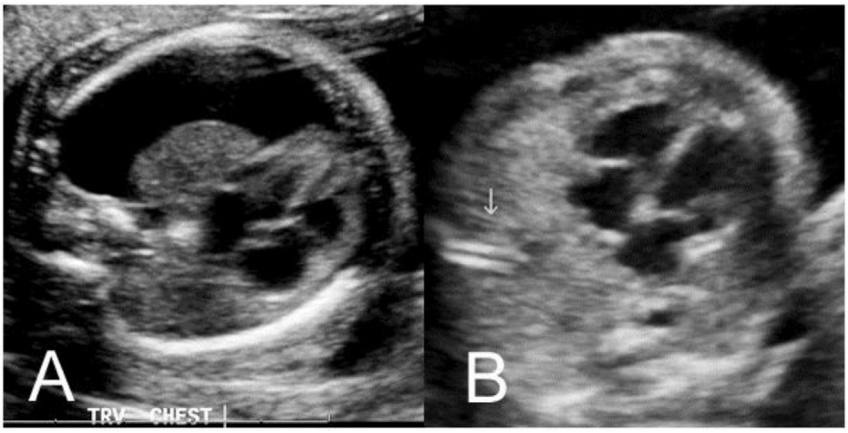

Fig. 1. (A) Severe left-sided pleural effusion with hydrops. (B) Effusion following thoracoamniotic shunt placement (arrow) and re-expansion of both lungs.

continuous ultrasound guidance with insertion into the lower lateral aspect of the fetal hemithorax in the mid-axillary line. The entrance point is important, as it should be the most dependent portion of the thorax into which the effusion will collect, allowing the maximum amount of fluid to be removed by aspiration. Re-evaluation is performed in 24–72 h and effusions rapidly reaccumulating after drainage will likely benefit from chronic drainage by TAS. The approach to optimal positioning of the shunt is side dependent. For right-sided effusions, the optimal position for shunt placement is the lower third of the chest between the mid-clavicular and mid-axillary lines. For left-sided effusions, optimal placement is the upper third of the chest in the mid-axillary line. This allows for optimal drainage as the heart returns to its normal position and for the lungs to expand to fill the chest. The procedure is performed under intravenous sedation and local anesthesia. The trocar sheath with sharp stylet is then passed through the maternal abdomen into the amniotic fluid space adjacent to the chest. The trocar is advanced until the space between the ribs at the entrance site is located, and, using a gentle twisting action, the trocar is advanced through the chest wall until the tip lies 5–10 mm within the effusion. The sharp stylet is removed and 5–10 cc of effusion fluid aspirated. The double-pigtail shunt is then quickly loaded into the trocar sheath so that the effusion does not drain, making catheter placement more difficult and limiting its ability to resume its original coiled shape. If necessary, it may be helpful to temporarily expand the effusion to facilitate shunt placement by infusing 10–30 cc of warm saline with the shunt loaded in the trocar sheath. Push rods are introduced and slowly advanced for 8–9 cm, displacing the proximal segment of the catheter into the intended cavity. The metal tip and high-density composition of the Rocket catheter optimizes visualization of its entrance into the fluid cavity. The sheath is then withdrawn to ~1 cm from the chest wall. The push rod is then held in place while the outer sheath is pulled ~2–3 cm backward over the push rod through the chest wall into the amniotic cavity, which deploys the central straight portion of the shunt. The distal shunt should then be within the amniotic cavity and, while holding the pushrod in place, the sheath is backed an additional 1–2 cm away from the chest wall. At this point it is important to stop and rotate the sheath away from the insertion site without otherwise moving the tip and then to deploy the remainder of the shunt from the sheath at an angle away from the insertion site. This is an important maneuver as it prevents the distal pigtail from being partially deployed into the effusion which will increase the risk of shunt migration into the chest cavity. Effusion drainage begins rapidly but may take 12–48 h to complete. Prophylactic single-dose intravenous antibiotic therapy is recommended. If polyhydramnios is present, amnioreduction utilizing the trocar sheath can be performed.

5. Complications

The most frequent complications of TAS procedures are related to displacement of the catheter into the amniotic cavity, or less usually into the thoracic cavity [8]. Occlusion from proteinaceous materials or thrombus may occur. Hence, if the effusion fluid on initial aspiration of 2–3 cc appears to have old blood products or particulate matter from the original thoracentesis, we recommend serial aspiration and infusion of 10 cc warm normal saline until the aspiration fluid clears before proceeding with shunt placement. Pregnancy loss is estimated at ~5%. Initial thoracentesis risk for pregnancy loss is estimated at 0.5–1.0%.

6. Outcome

Outcomes following TAS are dependent on selection of fetuses whom will benefit, i.e. fetuses with hydrops or significant risk of pulmonary hypoplasia. Studies have shown that effective drainage leads to improved antenatal lung growth, hydrops resolution, and long-term survival (Table 1). Thompson et al. [2] reviewed 17 neonatal TAS survivors with a mean gestational age of 29 weeks (range: 21–35). Twelve fetuses were hydropic at TAS. Recurrent respiratory symptoms were identified in six infants, not significantly different from a matched control group. Aubard et al. [3] identified hydrops as the only prognostic factor for outcome with survival after TAS, with and without hydrops, of 67% and 100% respectively, whereas without treatment, survival was 21–23% in both groups. Yinon et al. [4] reported on 88 fetuses with large PE, 59 (67.0%) were hydropic, 67 (76.1%) had bilateral effusions and 36 (40.9%) had polyhydramnios. Mean age of shunt placement was 27.6 weeks (range: 18–37), and delivery was 34.2 weeks (range: 19–42). Seventy-four babies (84.1%) were live-born and 52 (70.3%) survived the neonatal period. In 59 hydropic fetuses, 10 (16.9%) died in utero and 18 (30.5%) died after birth for a perinatal survival of 52.5%. However, in 29 non-hydropic fetuses, neonatal survival was 72.4%. Hydrops resolved in 28 fetuses (47.5%) following shunting with 71% survival compared to 35% in 31 fetuses where hydrops persisted. In their 22 neonatal deaths, seven were due to pulmonary hypoplasia, five had genetic syndromes, two had aneuploidy, and one was due to congenital heart disease.

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