Thoraco-Amniotic Shunting for Fetal Pleural Effusion â A Case Series

Abstract:

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Abstract
Fetal pleural effusion is a rare occurrence, with an incidence of 1 per 10-15,000 pregnancies. The prognosis is related to the underlying cause and is often poor. There is increasing evidence that in utero therapy with thoraco-amniotic shunting improves prognosis by allowing lung expansion thereby preventing hydrops and pulmonary hypoplasia. This is a review of all cases of fetal pleural effusion managed over an eight year period the National Maternity Hospital Dublin. Over the nine year period there were 21 cases of fetal pleural effusion giving an overall incidence of 1 per 9281 deliveries. Of these, 15 underwent thoraco-amniotic shunting. There were associated anomalies diagnosed in 5(33%) of cases. The overall survival in our cohort was 53%. The presence of hydrops was a poor prognostic factor, with survival in cases with hydrops of 33% (3/9) compared to 83% (5/6) in those cases without associated hydrops.

Introduction
Fetal pleural effusion is a rare occurrence, with an incidence of 1 per 10-15,000 pregnancies [1]. The associated prognosis depends primarily on the underlying aetiology, which is varied. Fetal pleural effusion may occur in association with structural congenital malformations [2] in particular primary lung lesions, such as bronchopulmonary sequestrations or congenital cystic adenomatoid malformations [3,4]. It has also been reported with a variety of cardiac anomalies [5]. There is an established association with chromosomal anomalies, particularly Trisomy 21 [6], and various genetic syndromes [7,8]. It may occur as part of the overall manifestations of fetal hydrops. A primary hydrothorax or chylothorax is felt to be secondary to a developmental anomaly of or laceration to the fetal thoracic duct [9,10], allowing an accumulation of lymphatic fluid within the fetal pleural cavity. The diagnosis of fetal pleural effusion is easily made on antenatal ultrasound, where it appears as a unilateral or bilateral sonolucent area within the fetal chest. Moderate and severe effusions may have the effect of a space occupying lesion, resulting in mediastinal shift and potentially impeding lung development. Severe bilateral pleural effusions result in superior vena cava compression and culminate in generalised hydrops fetalis [1]. Detailed ultrasound including fetal echocardiography and antenatal karyotyping will often reveal the underlying aetiology. Primary hydrothorax is then often a diagnosis of exclusion.

The natural history of fetal pleural effusion is varied and depends primarily on the underlying pathology. Spontaneous resolution has been reported to varying degrees [11]. In a review of 204 cases published in 1998, spontaneous regression was observed in 22% of cases [12]. The cases that were most likely to regress spontaneously presented more often in the second trimester, were more likely to be unilateral and not associated with fetal hydrops. Progressive accumulation of fluid within the pleural space leads to worsening fetal hydrops and potentially intrauterine demise. Pulmonary hypoplasia is likely to result from moderate or severe effusions impeding neonatal resuscitative measures. There is now increasing evidence that in utero therapy improves the prognosis in cases of fetal pleural effusion by allowing lung expansion thereby preventing progressive hydrops and pulmonary hypoplasia [13]. Because fluid reaccumulates rapidly following aspiration, long term drainage via fetal thoraco-amniotic shunting has been shown to confer a greater chance of survival and has been proposed as a more appropriate procedure [14]. We aimed to review our experience of fetal thoraco-amniotic shunting over an 8 year period at the National Maternity Hospital Dublin.

Methods
This is a review of all cases of fetal pleural effusion managed over an eight year period from January 2002 to January 2010 at the National Maternity Hospital Dublin which is a tertiary level referral institution delivering approximately 9000 women annually. It is the only centre in the Republic of Ireland performing thoraco-amniotic shunting. All cases of suspected fetal pleural effusion are reviewed by one of five fetal medicine specialists and have detailed ultrasound including fetal echocardiography and antenatal karyotyping performed. Cases of mild to moderate effusions occupying less than 50% of the chest cavity are managed conservatively. Thoraco-amniotic shunting, when deemed necessary, is performed after a qfPCR result has excluded Trisomy 13, 18 and 21, unless the fetal clinical status warrants immediate intervention. Shunting is performed under ultrasound guidance using a 14G trocar and a Rocket pigtail catheter (Rocket, London, UK). All procedures are performed by one of two operators under local analgesia with antibiotic prophylaxis.

Ultrasound is performed 24 hours following shunt insertion and weekly thereafter. Inductions of labour and caesarean delivery are performed for obstetric indications only. All livebirths have the shunt(s) clamped immediately following delivery and have a chest X- ray within the first hours of life. Details recorded included maternal demographics, gestation at diagnosis, laterality, associated ultrasound findings, gestation at shunt insertion and maternal and perinatal outcome.

Results
Over the nine year period there were 21 cases of fetal pleural effusion giving an overall incidence of 1 per 9281 deliveries. Of these, 15 underwent thoraco-amniotic shunting. The mean maternal age was 30.8 yrs, with a range from 23 â 40yrs. Two thirds of the cases (10/15) were external referrals. The majority were an incidental finding on routine ultrasound or had been diagnosed due to associated polyhydramnios. None of the cases had any previous history of note. All cases underwent antenatal karyotyping. The median gestation at diagnosis 28 weeks (range 22 â 32.2 weeks), with a median gestation at shunt insertion of 29.8 weeks (range 24.5 â 32.4 weeks). The median shunt to delivery interval was 4.7 weeks (range 0.4 -10.1 weeks).

Figure 1: Outcomes in 15 cases of thoraco-amniotic shunting for fetal pleural effusion

IUD : Intrauterine demise
NND : Neonatal death

The most common associated complication was preterm delivery. There was preterm rupture of membranes less than 37 weeks gestation in 6/13(46 %) of ongoing pregnancies, and overall 10/13(77%) delivered less than 37 weeks. There were two cases of shunt blockage or displacement. One case was 4 weeks following shunt insertion and the other 6 weeks following insertion. Neither case required shunt reinsertion. Overall 13/15(87%) of cases had bilateral thoraco-amniotic shunting performed; of these the majority (8/13) had both shunts inserted on the same day.

Fetal anomalies
Overall there were associated fetal anomalies diagnosed in 5(33%) of cases. There was one abnormal result confirmed on full karyotype leading to a neonatal death. In this case a normal qfPCR result had been confirmed prior to shunt insertion. There was one case of an associated structural malformation diagnosed prior to shunt insertion. This was a case of bronchopulmonary sequestration. There was associated hydrops fetalis and tense polyhydramnios. Shunt placement at 25 weeks and 4 days gestation resulted in good lung expansion however premature labour and delivery occurred 2 days later and the infant died at 12 hours of age. The third fetal anomaly was in a case of a consanguineous relationship in which there were associated findings of fixed flexed limbs and talipes. Despite extensive counselling of the possibility of an underlying genetic pathology the couple were keen for in utero therapy. An intrauterine demise occurred three days following shunt insertion. Post mortem examination was refused on cultural grounds.

Gestation = weeks + days; NND: Neonatal death; KMS: Kasabach Merrit Syndrome

There were two cases in which further associated anomalies came to light following delivery. The first was a case of Noonanâ s syndrome. In this case there had been an increased first trimester nuchal translucency of 5mm. Chorionic villous sampling confirmed a normal male karyotype. Bilateral hydrothoraces were diagnosed at 32 weeks and bilateral shunts were inserted at 32 weeks and 3 days gestation. Delivery occurred 4 days later following preterm rupture of membranes and placental abruption. Neonatal death occurred at 3 days of life. Postmortem findings confirmed a cardiac anomaly and Noonanâ s syndrome. The second was a case of Kaposiform haemangioendothelioma (Kasabach Merrit Syndrome), which resulted in a neonatal death at six weeks.

Figure 2: Transverse section through the fetal thorax at 29 weeks gestation demonstrating bilateral fetal pleural effusions (1 â Moderate left sided pleural effusion; 2 â Severe right sided fetal pleural effusion; 3 â Compressed fetal lungs bilaterally; 4 â Skin oedema;5 â Fetal heart)

Perinatal deaths
There were 2 cases (14%) of intrauterine demise following shunt insertion. In addition to the above mentioned case a second intrauterine demise occurred in a case with severe associated hydrops fetalis 3 days after shunt insertion. There were 5 neonatal deaths. Four of these had associated anomalies as discussed above. The fifth case was a diagnosis of fetal hydrops at 31 weeks gestation. Bilateral chest shunts were inserted 6 days later. Spontaneous onset of preterm labour occurred at 32 weeks and 2 days gestation. This infant died at 5 weeks of age.

Figure 3: Transverse section through the fetal thorax in the same case as Image 1 three days following bilateral thoracoamniotic shunting (1 â Right sided thoracoamniotic shunt in situ; 2- Left sided thoracoamniotic shunt with free end in amniotic cavity; 3 â Bilaterally expanded lungs)

Survival
The overall survival in our cohort was 53%. The presence of hydrops was a poor prognostic factor, with survival in cases with hydrops of 33% (3/9) compared to 83% (5/6) in those cases without associated hydrops. Excluding cases with associated anomalies as discussed above the overall survival was 80%.

Discussion
Fetal pleural effusion is a rare finding which, when severe, is usually associated with poor perinatal outcome. In our series of fifteen cases the overall survival was 53% and 83% in those cases without associated hydrops fetalis. These results would lend support to the growing body of evidence that outcome in cases of severe fetal pleural effusion can be improved with thoraco-amniotic shunting.

The most common complication in our series was preterm rupture of membranes and preterm delivery occurring 46% and 77% of cases respectively, which is consistent with other reports in the literature [15]. Indeed, prematurity is the most common complication of this procedure reported in all series to date and patients should be counselled appropriately regarding the risks and long term outcome of infants born prematurely as a result of chest shunt placement. Nonetheless, this must be weighed against the likely poor outcome associated with conservative management of such cases. In our series the mean shunt to delivery interval was over 4 weeks, a period of gestation which can vastly improve long term survival rates at the extremes of prematurity. Also, it is likely that even in cases where premature delivery occurs following shunt placement, neonatal resuscitative measures are greatly enhanced by the lung expansion provided by shunt placement. This series emphasises the important role of fetal thoraco-amniotic shunting in dramatically improving survival in fetuses with apparently isolated bilateral pleural effusions.

Our series also again highlights the importance of counselling patients that primary hydrothorax remains largely a diagnosis of exclusion, and that other causes of fetal pleural effusion may only come to light following delivery which may drastically alter the prognosis regardless of the anatomical success of the thoraco-amniotic shunt. Our rate of associated anomalies of 33% is similar to other series [13]. In conclusion we report our results from a single Irish centre, with our results to date in keeping with the most recent international published series. [16]

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Comments: