Fetoscopic approach to amniotic band syndrome

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A B S T R A C T

Background: Amniotic band syndrome (ABS) is an uncommon complication of pregnancy that can result in fetal demise.

Methods: We present our experience with fetoscopic amniotic band release.

Results: Five patients underwent fetoscopic amniotic band release for preoperatively diagnosed ABS involving at least one extremity. Four of five patients were found to have involvement of the umbilical cord at the time of fetoscopy. One of these four did not have the band released and underwent fetal demise at 24 weeks. All four survivors had good functional outcomes of affected limbs. Two patients developed membrane separation and had preterm deliveries at 32 weeks gestation whereas the other two carried to term. No maternal complications were noted.

Conclusions: Fetoscopic amniotic band release is safe. Umbilical cord involvement is difficult to assess preoperatively, but when it is present should be treated to reduce the risk of fetal demise.

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Amniotic band syndrome (ABS) is a rare in utero condition with an incidence of 1:1200 to 1:15,000 [1]. It is likely underreported, as detection is difficult with current radiographic modalities. More cases are likely to be encountered in the future due in large part to advancements in fetal imaging. While the cause is unknown, some speculate that either a primary defect or an early disruption of the amnion results in mesodermal matrix production [2]. Others believe that an ischemic event leads to band development [3].

The natural history of ABS is variable and so are its consequences. Complications arising from ABS are highly dependent on the anatomic location. Bands limited to an extremity can result in disfigurement, motor and sensory dysfunction, pseudosyndactyly and limb amputation [4,5]. Bands that involve the head can lead to craniofacial clefts and anencephaly. Broad bands can lead to significant chest and abdominal wall deformities that are uniformly fatal [6].

Successful in utero treatment of ABS via fetoscopic band release has been described [7,8]; however, experience with fetoscopic amniotic band release is limited to case reports [7–14]. We present our experience of five patients who underwent fetoscopic amniotic band release.

1. Materials and methods

Following IRB approval, we performed a retrospective review of patients evaluated for ABS at our Fetal Treatment Center from 1997 to 2012. Fetuses between 18 and 30 weeks gestation with isolated limb involvement were considered for fetoscopy. Fetal contraindications included abnormal karyotype and further structural abnormalities. Maternal contraindications included a shortened cervix (<2 cm), frequent contractions, membrane rupture, and uncontrolled comorbidities. Suitable candidates underwent fetoscopy under the auspices of standard-of-care at our Fetal Treatment Center. All cases were reviewed in a multi-disciplinary conference held on a weekly basis.

Those who underwent fetal intervention were reviewed. Details of the prenatal course, fetal procedure, and the postnatal course were obtained. Amniotic bands were demonstrated with preoperative fetal ultrasonography. Edema and vascular flow across the constrictive bands were evaluated in all cases.

Selective photocoagulation of amniotic bands was performed using a neodymium:yttrium–aluminum–garnet (Nd:YAG) laser. Other modes of band lysis included Bugbee fulgurating diathermy electrode (5–15W) and mechanical lysis with a miniature 1 mm grasper. Laparoscopic hook cautery and Metzenbaum scissors were options if a second cannula was placed. Postoperative ultrasounds were routinely obtained on postoperative day 1 and again confirmed on follow-up 1 week later.

2. Results

Between 1997 and 2012, 38 patients were referred to our Fetal Treatment Center with the diagnosis of ABS. Twenty-eight were confirmed to have ABS, five of whom underwent fetal intervention and were included in this series. The details of each case are highlighted in Table 1.
At referral, the average maternal age was 29.8 ± 5.0 years and the mean gestational age was 21.1 ± 2.9 weeks. At intervention and delivery the mean gestational age was 23.0 ± 2.2 weeks and 36.1 ± 4.5 weeks, respectively.

In all patients who underwent fetoscopy, band involvement was isolated to limbs. Four fetuses had a single limb involved and one had two limbs involved. Preoperative sonography demonstrated edema distal to constrictive bands in all six extremities. Arterial flow was preserved in all limbs, and venous flow past the constrictive band was evident in three. Preoperative umbilical cord involvement was suggested in one case.

Intra-operatively, amnioexchange was required in three cases. Umbilical cord involvement was evident in four of five cases and umbilical bands were taken down in three of these four cases. Postoperative oligohydramnios was evident in three cases and complications included chorioamniotic separation (n = 2), preterm premature rupture of the membrane (n = 1), and preterm birth (n = 2). Postoperative ultrasonography 1 week following band lysis demonstrated arterial and venous flow across all six constrictive bands that were treated with improved edema in two cases.

Four of five fetuses progressed to viable births whereas one underwent in utero fetal demise 32 days after extremity band lysis but no treatment for a band involving the umbilical cord (see case 1 below). Postnatal follow-up was available for three of four patients with a median length of 3.0 ± 9.5 months (range = 2–19 months). One patient had soft tissue reconstructive surgery at 3 months of life. All limbs had preserved form and function.

3. Case presentations

3.1. Case 1

A 37-year old gravida 3, para 0 presented at 16 4/7 weeks gestation with suspicion for ABS on a screening obstetric ultrasound. An amniocentesis indicated a normal male karyotype and no other fetal anomalies were identified. The soft tissue bands seen on ultrasound involved the proximal left ankle circumferentially and the hallux of the right heel both with marked distal edema. There was clubbing of the left foot and amputations of three digits on the right foot. Duplex imaging demonstrated preservation of arterial and venous flow in both extremities. Additionally, umbilical cord constriction was suggested by variceal dilation of the umbilical vein adjacent to the band. No other anatomic abnormalities of the fetus were detected.

The patient was taken to the operating room at 19 3/7 weeks gestation for fetoscopic band release. Under spinal anesthesia, a small maternal laparotomy was made in the left lower quadrant. As the uterus was tightly contracted, general anesthesia was induced to promote uterine relaxation and a single 3 mm cannula was placed. The amniotic fluid was murky, and, therefore, amnioexchange was performed. A combination of Bugbee electrode, laser ablation and mechanical lysis was used to release the amniotic bands involving both lower extremities. These instruments were passed through the biopsy channel of the fetoscope. The amniotic band involving the umbilical cord was seen but not lysed in attempt to prevent iatrogenic cord injury and fetal demise. Postoperative ultrasonography 2 weeks following the procedure demonstrated oligohydramnios and new onset ascites. A fetal echocardiogram demonstrated cardiomegaly although middle cerebral artery Doppler waveforms did not suggest anemia. Thirty-two days following fetal intervention, at 24 weeks gestation, ultrasonography showed fetal demise.

3.2. Case 2

A 26-year old gravida 5, para 4 was diagnosed with ABS on routine obstetric ultrasound at 22 weeks gestation. The band involved the right lower extremity. It entangled the right ankle just distal to the tibial and fibular epiphysis (Fig. 1). The foot distal to the constrictive band was edematous with arterial but no venous flow. An amniocentesis indicated a normal female karyotype and no other fetal anomalies were identified.

At 24 5/7 gestation, a maternal laparotomy with fetoscopy was performed. After spinal anesthesia was induced, two 3 mm cannulas were placed. Amnioexchange allowed identification of a band involving the right lower extremity, and a single attempt to mechanically disrupt the band was made. At this point, on closer inspection of the amniotic sac, chorioamnionic separation was noted and the procedure was aborted. Despite incomplete lysis of the amniotic band, arterial and venous flow was observed distal and proximal to the affected area on postoperative sonography, although edema persisted. The postoperative course was complicated by preterm delivery at 32 weeks gestation. At 3 months post-delivery, further reconstruction was required but the limb was salvaged and functional (Fig. 2A).

3.3. Case 3

A 27-year old gravida 1, para 0 presented for routine obstetric ultrasound at 19 4/7 weeks gestation when ABS was suspected because of a single amniotic band constricting the left forearm. Edema was noted distal to the band and arterial flow was preserved although there was no evidence of venous flow. No other fetal anomalies were identified and an amniocentesis indicated a normal female karyotype.

At 24 5/7 weeks gestation, under spinal anesthesia, a maternal laparotomy with fetoscopy, using a single 3 mm cannula, was performed. At the time of fetoscopy, the amniotic band appeared to involve the umbilical cord as well. A laser and grasper were used to successfully release the amniotic band from the left forearm. Given our prior experience with fetal demise after not releasing an umbilical cord band, in this case the umbilical cord band was released. Postoperative imaging indicated resolution of edema and re-establishment of venous flow across the constriction in addition to increased movement of the left hand. The fetus delivered at 40 weeks gestation without any postnatal sequelae. At 16 months...
follow-up, there was no evidence of residual edema and neurologic function was completely intact (Fig. 2B).

3.4. Case 4

A 29-year old gravida 4, para 2 presented for routine screening obstetric ultrasound at 20 6/7 weeks gestation. Sonography identified constrictive amniotic bands wrapping around the right hand with pseudo syndactyly of phalanx 2–4. Marked edema was noted in the right hand distal to the band but arterial and venous flow was present. No other fetal anomalies were identified.

At 22 2/7 weeks gestation, the patient was taken to the operating room for fetoscopy under spinal anesthesia where two 3 mm cannulas were used. The amniotic fluid was turbid from the previous RFA, and following amnioexchange, multiple bands were seen deep in the uterine pelvis. Adhesiolysis was performed using hook cautery and Metzenbaum scissors through the second cannula. After satisfactory band release from the extremity, additional bands were identified involving the umbilical cord and these were also lysed. Postoperative imaging indicated improved edema of the left leg, return of venous flow and a large 10.6 × 3.7 cm hematoma adjacent to the soft tissues of the left lower extremity. The patient experienced postoperative oligohydramnios, premature rupture of membranes, and preterm delivery at 32 4/7 weeks gestation. Upon delivery, however, the fetus had full function of the left lower extremity.

4. Discussion

ABS is an uncommon complication of pregnancy. The etiology is unclear. It may be because of spontaneous or iatrogenic membrane rupture or possibly an ischemic event. In our experience, most cases appeared to be spontaneous although clearly one of our cases presented here occurred after RFA for twin-to-twin transfusion syndrome, supporting the theory that amniotic membrane rupture may contribute to the development of ABS. Furthermore, the need for amnioexchange in three out of five cases suggests a possible exudative process, albeit the amniotic fluid could very well have been affected by ischemia that resulted after the amniotic bands had developed.

Patient management and technical strategies for those undergoing fetoscopy continue to evolve. Currently at our institution, all patients receive preoperative tocolysis with a single dose of indomethacin. Spinal anesthesia is generally sufficient. Intra-operatively, ultrasound is used for placental mapping, cannula introduction, fetal heart monitoring, and evaluation of vascular flow across constrictive bands before and after. Operative techniques have also evolved as we generally no longer find it necessary to perform maternal laparotomies. The number of trocars needed, however, continues to vary based on the case. In our series, a second cannula was placed in three cases to provide adequate traction for band lysis. Postoperatively, patients receive tocolysis with indomethacin and magnesium as needed and remain on bed rest overnight. An ultrasound on postoperative day 1 is performed and if reassuring the patient is discharged home with 1 week follow-up. The frequency of subsequent follow-up is dependent on disease severity.

Outcomes from ABS are variable and this is highly dependent on the anatomic location. For fetuses with lesions confined to an extremity (or extremities) the natural history is for the fetal limb to outgrow the band resulting initially in venous congestion, followed by arterial impedance, limb ischemia, and ultimately amputation. In the
success of amniotic band release, but favorable outcomes are realistic. While dreaded obstetric complications, including preterm premature rupture of membranes and preterm birth, are tangible, we had two patients deliver at 40 weeks gestation, emphasizing the feasibility of fetoscopic amniotic band takedown even in cases where the deformity may not be life-threatening. In addition, maternal risk is minimal as we did not have any maternal postoperative complications, which may include bleeding, chorioamnionitis, uterine rupture and pre-eclampsia. Furthermore, no maternal complications with fetoscopic amniotic band release have been reported to date in the literature [12]. Fetal intervention is not without risk, however, and the risk to benefit ratio must be carefully weighed with each patient and family. Each case should be evaluated on an individual basis to determine if fetal intervention is an appropriate therapy.

Our series suggests that not only is fetoscopic amniotic band release possible, but favorable outcomes are realistic. While dreaded obstetric complications, including preterm premature rupture of membranes and preterm birth, are tangible, we had two patients deliver at 40 weeks gestation, emphasizing the feasibility of fetoscopic amniotic band takedown even in cases where the deformity may not be life-threatening. In addition, maternal risk is minimal as we did not have any maternal postoperative complications, which may include bleeding, chorioamnionitis, uterine rupture and pre-eclampsia. Furthermore, no maternal complications with fetoscopic amniotic band release have been reported to date in the literature [12]. Fetal intervention is not without risk, however, and the risk to benefit ratio must be carefully weighed with each patient and family. Each case should be evaluated on an individual basis to determine if fetal intervention is an appropriate therapy.

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