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Limb amputation in amniotic band syndrome: serial ultrasonographic and Doppler observations

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ABSTRACT

A 28-year-old woman, gravida 7, para 6, was referred at 21 weeks' gestation to our ultrasound unit because of bilateral fetal lower limb edema diagnosed previously. Ultrasonography showed a constriction ring around both legs, and, with the aid of color Doppler, arterial flow was demonstrated in both legs beneath the constriction ring. Subsequent examinations during the 24th and 28th weeks revealed absence of flow below the constriction ring of the right leg and decreased flow in the left leg. This was followed by the gradual bending, breaking and resorption of the tibia and fibula of the right leg. Between weeks 30 and 34, gradual shrinkage of the remains of the right leg beneath the knee was recorded by serial ultrasonic observations. At the 38th week of gestation, a male infant was born by normal vaginal delivery. Examination at birth revealed amputation of the right leg below the knee, with a denuded end of the stump. There was a partial amputation of the left leg below the knee, with tissue continuity being maintained by the posterior neurovascular bundle, and a posterior strip of skin. The left foot was extremely edematous, with an area of necrosis dorsally.

This case afforded us the opportunity of in utero *following of natural limb amputation in the amniotic band syndrome.*

INTRODUCTION

Amniotic band syndrome, or amniotic band disruption complex, is a common cause of miscellaneous fetal malformations involving the limbs, trunk and craniofacial region. It occurs in approximately 1 in 1200 to 1 in 15 000 live births[1]. The clinical manifestations of the syndrome vary from death secondary to severance of the umbilical cord or anencephaly, to extremity amputations, (minor) facial clefts, or lymphedema.

Repeated ultrasonic examinations afforded us the opportunity of observing the *in utero* process of limb strangulation and subsequent amputation in a fetus with the amniotic band syndrome.

CASE REPORT

A 28-year-old woman, gravida 7, para 6, with six healthy children was referred to our ultrasound unit because of bilateral fetal lower limb edema diagnosed previously. Her past obstetric history was uneventful.

On ultrasound examination at 21 weeks' gestation, both feet of the fetus were markedly swollen and enlarged owing to severe edema (Figure 1). The two legs were very close to

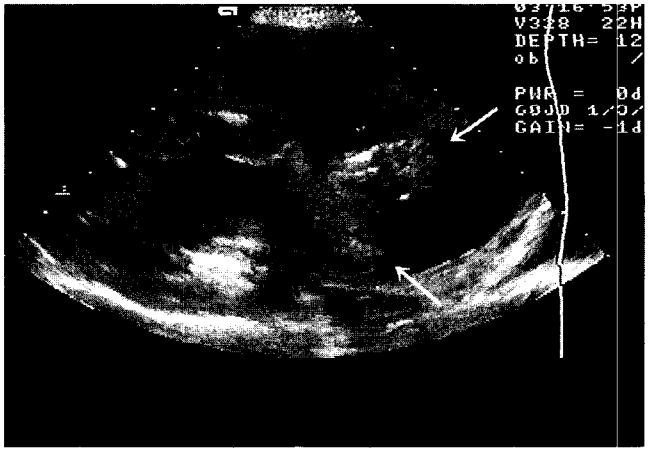

Figure 1 Fetal lower limbs at 21 weeks' gestation are closely apposed, enlarged and swollen

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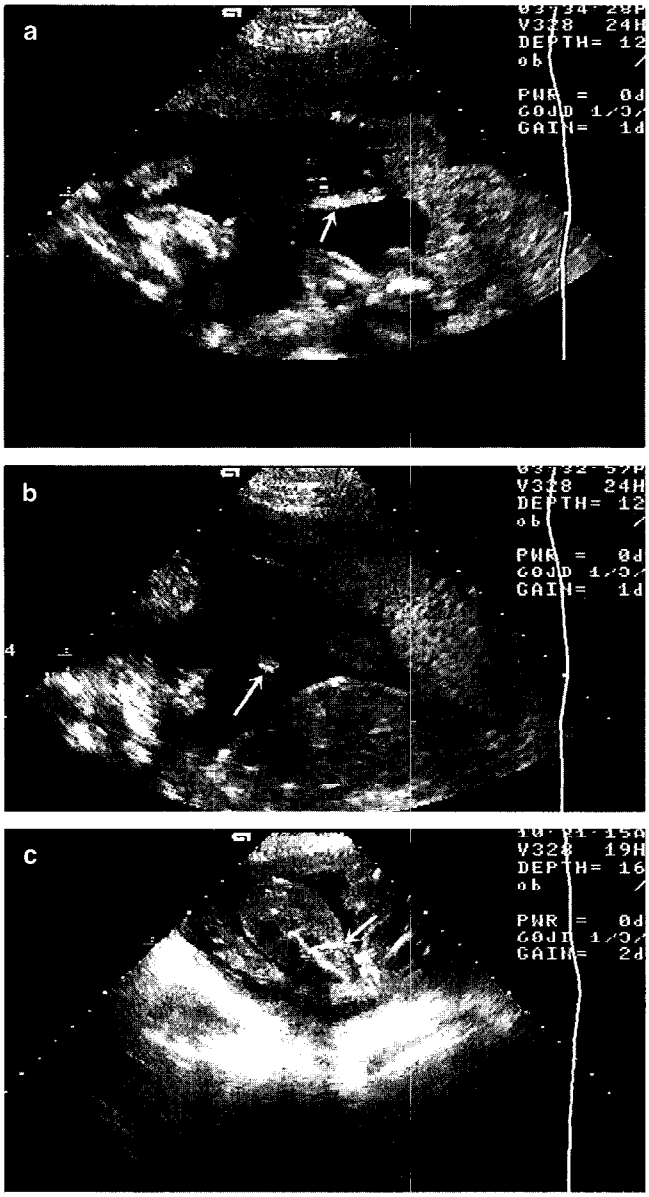

Figure 2 (a) Thick amniotic band; (b) cross-section of the amniotic band encircled by the umbilical cord; (c) amniotic band cutting right fetal leg

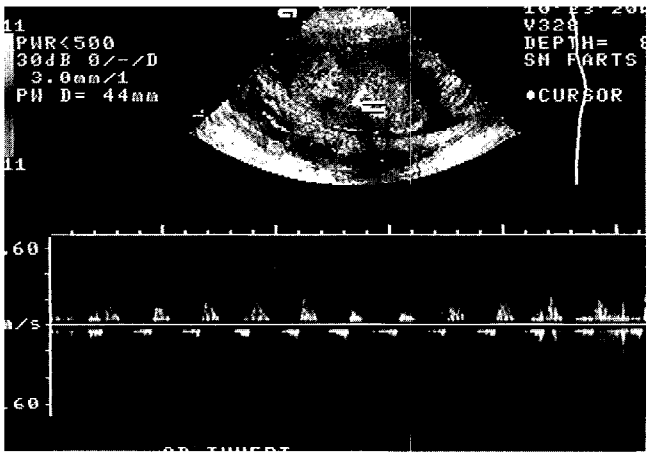

Figure 3 Right fetal leg. Pulsed Doppler demonstrating arterial flow beneath the constriction ring. Arterial flow could not be demonstrated at 24 and 28 weeks' gestation.

each other and showed very little movement. Figure 2 shows views of the amniotic band originating from the fetal membrane and surrounding both legs.

On the basis of these findings and the presence of amputated fingers in both hands, a diagnosis of amniotic band syndrome, with bilateral constriction ring and feet lymphedema was made. With the aid of color Doppler, arterial flow was demonstrated in both legs beneath the constriction ring (Figure 3). Later examinations during the 24th and 28th weeks revealed absence of flow below the constriction ring of the right leg and decreased flow in the left leg. This was followed by the gradual bending of the tibia and fibula of the right leg (Figure 4). Subsequently, autoamputation of the right leg below the knee occurred.

Serial ultrasonic observations between weeks 30 and 34 showed gradual shrinkage of the remains of the right leg below the knee. At the 38th week of gestation, a male infant was born by a normal vaginal delivery. Examination at birth revealed amputation of the right leg below the knee, with a denuded end of the stump (Figure 5). There was a partial amputation of the left leg at the mid-tibial level. Tissue continuity in the lower left leg was maintained by the posterior neurovascular bundle and a posterior strip of skin. The left foot was extremely edematous, with an area of necrosis on the dorsum (Figure 6). There was syndactyly of both hands, with partial amputation of two

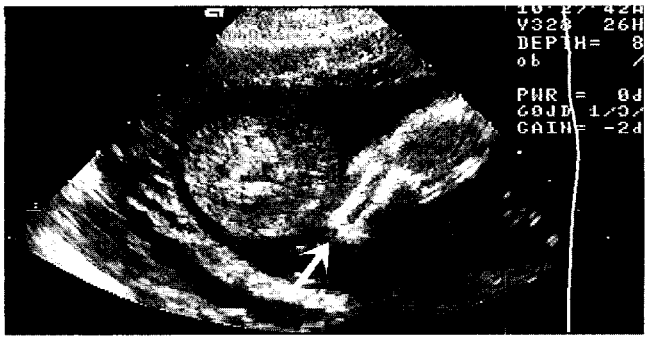

Figure 4 Right fetal leg showing swelling of the foot and deformity shortly before autoamputation occurred

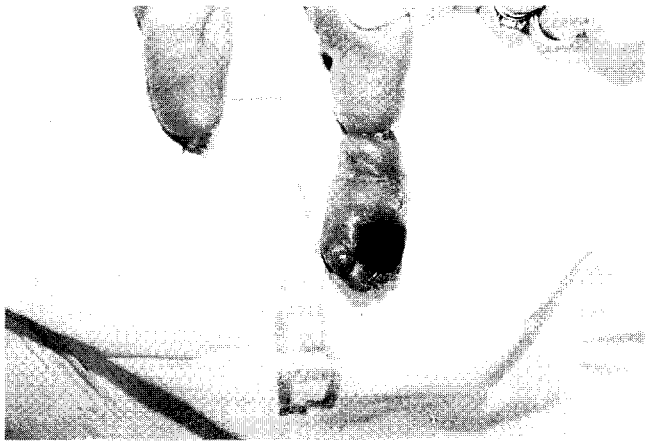

Figure 5 Both legs after delivery. Right leg: below-knee amputation; left leg: almost complete below-knee amputation; tissue continuity maintained by the posterior neurovascular bundle and the posterior strip of skin

Figure 6 Left fetal leg after delivery. There is an almost complete below-knee amputation, at the mid-tibial level. Tissue continuity is maintained by the posterior neurovascular bundle, and the posterior strip of skin. The left foot is extremely edematous, with an area of necrosis on the dorsal surface

fingers in each hand. Pathological examination of the placenta and amnion was normal.

DISCUSSION

To the best of our knowledge, this is the first report of the *in utero* observation of the natural history of limb reduction in the amniotic band syndrome. A fetus was observed during the process of limb strangulation and ultimately amputation. With the aid of color Doppler, the arterial blood flow before and during the amputation was evaluated.

Amniotic band syndrome is also known as amniotic band disruption complex, amniochorionic mesoblastic fibrous strings, or congenital constriction band syndrome. The etiology of the syndrome is under debate. Most authors agree with the hypothesis suggested by Torpin[1,2], that rupture of the amnion leads to entanglement of fetal tissue by fibrous bands originating from the fetal membrane.

Because amniotic band syndrome occurs far more frequently in monozygotic than in dizygotic twins, it has been suggested by a number of researchers[3–7] that the syndrome may result from a teratogenic event. Streeter[3] explained that the amniotic band syndrome is caused by developmental problems that occur during the formation of the amniotic cavity and the laying down of the germ disk.

Recently, Bamforth[4] reinforced Streeter's hypothesis by showing that, in a number of cases of amniotic band syndrome, most defects were explicable in terms of interference with neuropore closure. The teratogenic theory may be supported by cases of amniotic band syndrome involving monozygotic twinning, in which the amniotic bands may be caused by a common early insult[5–10]. Lockwood and colleagues[8] suggested that hemorrhages precede some of the anomalies associated with the amniotic band syndrome. Another explanation presumes multifactorial or polygenic inheritance[11].

Limb amputations may also result from teratogenic or genetic causes, although these may be differentiated from amputations resulting from the amniotic band syndrome by ultrasonic and physical examinations. In our case, the existence of a constriction ring with edema and the typical soft tissue denudation at the end of the stump was convincing evidence for the amniotic band syndrome. The asymmetric amputation added further proof to the diagnosis in our case, because genetic or teratogenic insult would be expected to have caused bilaterally symmetrical amputations. As stated above, the Torpin[1,2] hypothesis suggests that the specific fetal malformations in the amniotic band syndrome are determined by the timing of the fetal membrane disruption and the position of the attachment of the fibrous band to the fetus. A number of authors have postulated that the earlier the offence occurs, the more severe the injury to the fetus[8,9,12–14]. Fetal membrane disruption in the first weeks of pregnancy results in craniofacial and visceral defects[12,13], whereas during the second trimester it causes constriction rings and limb amputations[14]. This hypothesis lacks the proof of serial ultrasonic examinations of the process as it occurs.

Recently, a description of consecutive ultrasonic observations of amniotic band syndrome in the first trimester was published[15]. The authors described a case of amniotic band syndrome that was observed by sequential ultrasonographic examinations from the late embryonic to the early fetal period. Laberge and co-workers[16] described a case in which, on ultrasonography, an 18-week-old fetus was found to have his left hand attached to an amniotic band. The child was born with typical anomalies of the amniotic band syndrome. Previous reports of the ultrasonographic prenatal diagnosis of the amniotic band syndrome, involving pregnancies in the second or third trimesters, reported anomalies that were already present when they were diagnosed by ultrasonography[17–20]. Authors who reported on the amniotic band syndrome associated with amniocentesis[18–20] have assumed the sequence of events but lacked direct evidence, namely sequential ultrasonographic examinations before, during and after the injury to the fetus.

We have had the opportunity of following the natural history of limb amputation in amniotic band syndrome. A fetus was observed *in utero* during the process of limb strangulation and, later, amputation. With the aid of color Doppler, the blood flow was evaluated before and during the edema that preceded the amputation, and we were able to pinpoint the moment when the flow in one limb ceased. Our case provides support for Torpin's[1,2] hypothesis, although it seems likely that the etiology may vary from case to case.

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