Prenatal Diagnosis and Management of Twin Reversed Arterial Perfusion (TRAP) Syndrome

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Introduction

Twin reversed arterial perfusion (TRAP) syndrome, also known as acardiac twinning, is a rare obstetric condition unique to monochorionic twin gestations. This condition complicates approximately 1/35,000 pregnancies and occurs in 1/100 monochorionic twin gestations and 1/30 monochorionic triplet gestations [1]. TRAP syndrome results from umbilical artery-to-artery anastomosis between twins in multiple pregnancies. This is most commonly found in monozygotic twin gestations with a single placenta and is also accompanied by venous-to-venous anastomosis [2,3]. The severity of this syndrome makes it a lethal anomaly for the acardiac twin. A wide range of gestational and perinatal complications have been noted, including polyhydramnios, premature labor, cardiac insufficiency and intrauterine death of the pump twin [3,4].

Here, we report one case of TRAP syndrome associated with death of the pump twin at 28 weeks of gestation.

Case Report

A 32-year-old, gravida 1, para 0, Taiwanese woman presented with suspected intrauterine fetal demise of one twin at a local medical clinic. Prior to this pregnancy, she had used no contraception and had taken no medications or drugs, and there was no family history of twins or fetal anomalies. She was a nonsmoker and received regular prenatal examinations. Detailed ultrasound examination revealed a twin pregnancy with a single posterior placenta. Markedly different sized umbilical cords were also observed (Figure 1). No separating membranes were noted. One of the twin fetuses was grossly malformed, amorphous, and acardiac. There were no upper limbs and only one lower limb (femur length, 27 mm) (Figure 2). No umbilical vessel retrograde blood flow of the acardiac twin was found by color flow Doppler ultrasound examination. Four-dimensional ultrasound examination revealed an acardiac twin with one lower limb, suspicious for TRAP syndrome (Figure 2). The karyotype showed a normal male (46,*XY*). Fetal echocardiography was performed and showed a normal heart without signs of cardiac failure in the pump twin. The femur length in the pump twin was small for gestational age (femur length, 33 mm). Unfortunately, intrauterine fetal demise of the pump twin was noted at 28 weeks of gestation. Entanglement of the cord was noted after delivery (Figure 3).

Discussion

The pathogenesis of TRAP syndrome is unknown, but current theories include vascular anastomosis to be the primary abnormality with resultant early tissue hypoxia in the recipient twin leading to secondary atrophy of the heart and other organs [5]. Others believe a primary defect in cardiac embryogenesis with twin reversal arterial perfusion allowing survival of the affected twin. Chromosomal disorders have also been suggested [5].

Acardiac twins can be diagnosed antenatally on ultrasound by the absence of cardiac pulsation and poor definition of the trunk, head and upper extremities, and deformed lower extremities [6]. However, misdiagnosis of TRAP syndrome can occur on prenatal

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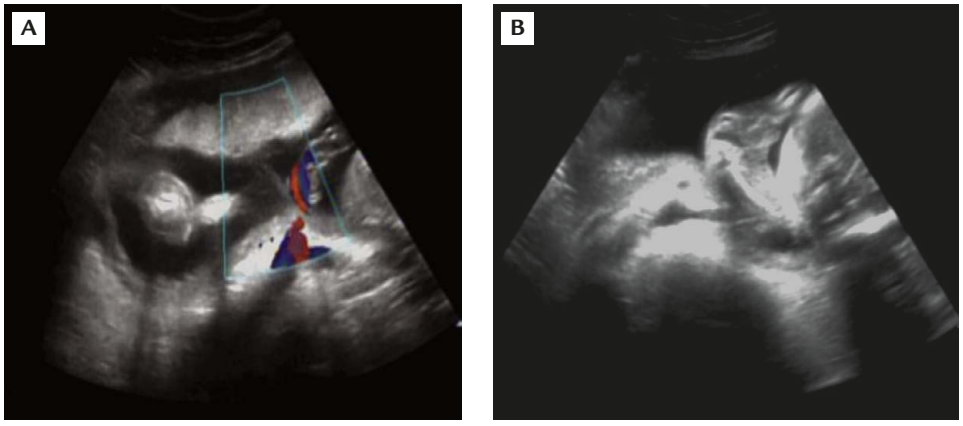

Figure 1. (A, B) Ultrasound shows markedly different sized umbilical cords, which may be due to them being inserted close together. No umbilical vessel retrograde blood flow of the acardiac twin is found by Doppler ultrasound.

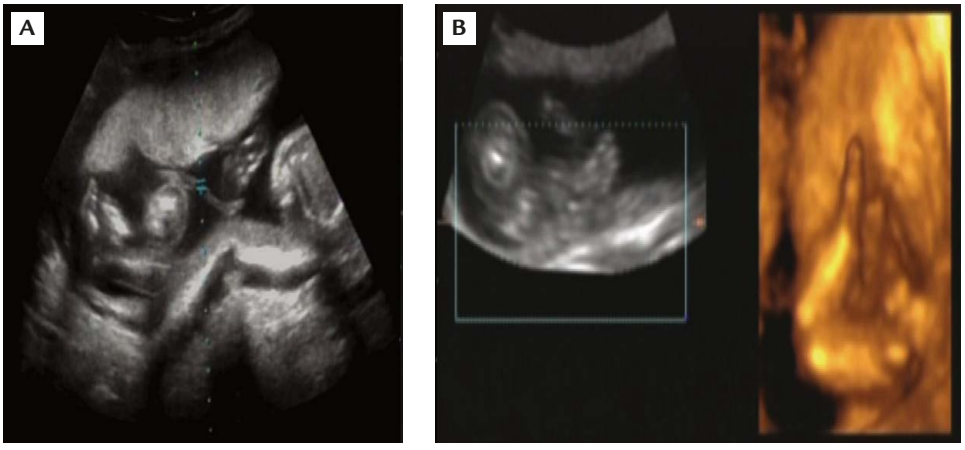

Figure 2. (A) Ultrasound shows twin pregnancy with the normal twin at about 20 weeks' gestation and an amorphous acardiac twin with one lower limb (femur length, 27 mm), and a single placenta in a singleton sac. (B) Four-dimensional ultrasound reveals a lower limb.

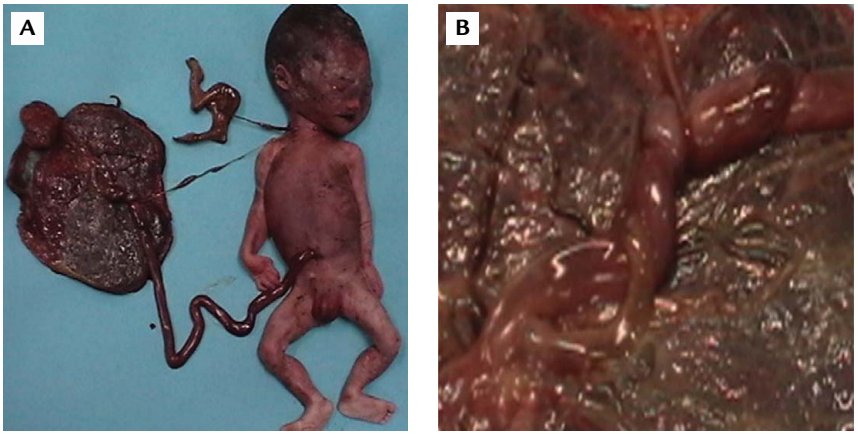

Figure 3. (A) Intrauterine fetal demise of the pump twin at 28 weeks of gestation. (B) Entanglement of the cord and discordance in the size of the two cords, with insertion close together, are noted.

ultrasound, because the acardiac fetus may be thought to be anencephaly, cystic hygroma, conjoined twins, twin demise, or intra-amniotic or placental tumors. The finding of retrograde blood flow in an acardiac twin by color flow Doppler ultrasound examination confirms the diagnosis [7,8]. Lately, four-dimensional ultrasound has also been used as an adjunct to confirm the diagnosis and to establish the extent of fetal malformations. However, views are usually limited by the size and atypical grotesque appearance of the acardiac twin [9].

Although no umbilical vessel retrograde blood flow of the acardiac twin was found by color flow Doppler ultrasound examination in our case (possibly because the umbilical cord was too thin), four-dimensional ultrasound images helped us to diagnose the TRAP syndrome.

The reported fetal/neonatal mortality of the pump twin is extremely high (50–75%) and is thought primarily to be because of increased cardiac demands on the pump twin in an effort to perfuse its acardiac sibling [6]. Poor prognostic factors for the pump twin include acardiac-to-pump twin weight ratio of >70%, rapid growth of the acardiac twin, congestive heart failure or hydrops in the pump twin, polyhydramnios, certain morphologic characteristics of the acardiac twin such as acardius anceps and presence of arms, a small resistance index difference of <0.20 on Doppler studies of the umbilical artery of both the acardiac and pump twins, and delivery before 32 weeks [6].

The goal of treatment is to interrupt the vascular communication between the pump and recipient twin. If available, ultrasound-guided radio-frequency ablation [10,11] or laser coagulation [11] of the intrafetal vessels should be the first line of treatment based on currently available experience. If this is not possible, chemosclerosis of the intrafetal arterial vessels with alcohol should be attempted [12].

Conservative management is recommended if none of the above features is present, because spontaneous cessation of blood supply to the acardiac twin occurs in many cases [6]. Despite a persistent blood supply from the pump twin, the size of the acardiac twin remains significantly smaller than the size of the pump twin. Even if deterioration is subsequently detected, conservative management is still recommended, because treatment could be carried out later in gestation. One exception to this conservative approach, as in our case, may be the treatment of an acardiac twin in a monochorionic-monoamniotic pregnancy. In such cases, ablation of the acardiac twin's vasculature together with transection of the umbilical cord to prevent death of the pump twin due to entanglement of the cord should be considered.

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