Acardiac Twin: A rare Complication of Monochorionic Monoamniotic Twin

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Abstract
Acardiac malformation, also known as twin reversed arterial perfusion (TRAP) sequence, is a unique complication of monochorionic twinning, occurring with a reported incidence of 1 in 35,000 deliveries. It is characterized by lack of heart development associated with a spectrum of malformations and anomalies in one of the twins, which is perfused in a paradoxical retrograde fashion by a structurally normal ‘pump’ twin through a single artery-to-artery anastomosis. There is usually a normally formed donor twin who has features of heart failure as well as a recipient twin who lacks a heart (acardius) and various other structures. It is caused in the embryo by a large artery-to-artery placental shunt, often also accompanied by a vein-to-vein shunt. The common complications include congestive cardiac failure in the pump twin, polyhydramnios and preterm delivery.

Keywords
Acardiac twin; Mono chorionic twins; TRAP

Introduction
Multiple pregnancy account for 1.5% of all pregnancies, with approximate perinatal morbidity of 10% (Hrubec and Robinette, 1984). Acardiac twining also referred to as the twin reversed arterial perfusion sequence (TRAP) represents an extreme manifestation of the twin transfusion syndrome and has an incidence of 1 in 35,000 deliveries (Chanthasenanant and Pongrojpaw, 2005) amounting to an average risk of 1% among monozygotic twins. The acardiac twin is transfused by the normal co twin by means of reversal of circulation through large vein to vein and artery to artery anastomosis and has no direct connection with placenta.

The acardiac twin presents with many bizarre anomalies thought to be due to low oxygen tension and dramatic alterations in fetal physiology. Complication in pump twin include congestive heart failure, polyhydramnios, preterm labour and death in 50–57% of all cases.

We present a case of acardiac-acephalus twin diagnosed at 18 weeks of pregnancy on ultrasonography.

Case Report
A 26 year old 2nd gravida having one full term vaginal delivery with one living baby came for antenatal registration at 18 weeks of pregnancy. Her past history and family history were unremarkable for twins or perinatal issues. Investigations included completer blood count, urinalysis, syphilis and blood glucose measurement. An ultrasound report done documented a monozygotic twin pregnancy with one normal fetus of 18 weeks and other showing normal amniotic fluid in amniotic sac with a large soft tissue mass with deformed skeletal elements. Head and neck, heart, viscera and upper limbs were not detected; only a small lower lumbar vertebral column and a small hemipelvis with full formed right lower limb and short femur on left side were seen suggestive of acardiac acranial amorphous twin. The acardius had a 2 vessel cord with TRAP via the pump twin with anastomosis within placenta (Figure 1).
Figure 1
Her clinical examination revealed a 22–24 weeks size uterus with multiple fetal parts. Patient was counseled and opted for termination of pregnancy. Pregnancy was terminated and she aborted a congenially malformed abortus by breech and a second abortus by vertex which externally appeared normal.

The only recognizable parts in the malformed twin were lower limbs that too deformed. On gross examination the proximal part of the fetus was a multilocular cyst filled with a transparent fluid covered with a gelatious swollen skin and no heart or lung like structure. There was no upper limb or head and neck (Figure 2). Patient refused autopsy. Single placenta was delivered with single amniotic membrane with 2 cords inserted side by side; one of them was very small and attenuated which belonged to acephalus twin.

Figure 2
Discussion
Twin reversed arterial perfusion (TRAP) first defined by Greenwald in 1942, is probably responsible for acardiac fetus as one of the forms of twin to twin transfusion syndrome. Acardiac twinning is a rare congenital anomaly characterized by formation of a malformed fetus with rudimentary (but non functional) heart.

Acardiac anomaly usually occurs in monozygotic twins, although there are a few reports of dizygotic twins with a fused placenta. It is more common in female twins, and because the disorder is monozygotic, the twins are usually of same gender. The etiopathogenesis of this anomaly is abnormal placental vascular communication between the twins, leading to imbalance of interfetal circulation. Reversed blood flow in the umbilical artery of the acardiac twin causes atrophy of the heart and other organs (Chen et al., 1997). This flow pattern has been termed “twin reversed-arterial perfusion” sequence in which the lower body of the fetus receives blood with more oxygen saturation and nutrients than the upper body, leading to maldevelopment of the head, neck and upper extremities as seen in our case.

A variety of acardiac twins have been described based on the degree of cephalic and truncal maldevelopment. The acardiac-acephalus fetus has no cephalic development, whereas acardius-anceps fetus has some cranial structures and/or neural tissue development. The acardius-acormus fetus has cephalic structures with limited or no truncal development. The fourth type, the acardius-amorphous fetus, has the most severe malformation and lacks all cephalic and truncal differentiation (Hanafy and Peterson, 1997).

An acardiac twin must be suspected in all monochorionic, malformed fetuses with cystic hygroma, generalized edema, and an absent cardiac pulsation with a nonfunctioning heart. Similarly, an ultrasonography finding of twins revealing discordant or grotesque malformation along with reverse flow in the umbilical artery is usually diagnostic of an acardiac twin (Ishimatsu et al., 1993).

Termination of pregnancy should be considered if acardiac twinning is diagnosed early in the pregnancy. However, early recognition and active management of the TRAP syndrome with maternal digoxin and indomethacin therapy can salvage the pump twin.

Twin to twin transfusion syndrome (TTTS) cannot be prevented but an early diagnosis of this disorder in an identical twin pregnancy can possibly save one or both babies. This can be detected in the early stage of pregnancy by
ultrasound scanning and Doppler velocimetry (Dashe et al., 2001).

Currently to stop the blood flow to the acardiac twin, a high energy radiofrequency ablation is utilized to destroy the blood vessels and surrounding tissues at the site where they enter the acardiac twin. The other therapy is fetoscopic placental laser surgery directed at the vascular connection between the twins (Tsaok et al., 2002).

Conclusion

Conservative treatment is best suited for salvation of the pump twin when the acardiac twin is less than one fourth of weight of pump twin and no signs of impending heart failure. Invasive intervention is justified when acardiac twin exceeds 70% of that of pump twin; however the prognosis appears to be poor. Intrefetal ablation is the treatment of choice because it is simpler, safer and more effective than cord occlusion techniques.

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