

## Case Report

### A rare case of acardiac twin pregnancy

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**Abstract:** Acardiac Twinning or TRAP sequence is a rare complication of monochorionic twin pregnancy. We present a case of acardiac twin with late presentation. This case report highlights the need for correct and early ultrasonographic diagnosis of chorionicity and detailed ultrasonographic follow-up of twin pregnancies.

**Keywords:** Acardiac Twin, Twin Reversed Arterial Perfusion, Monochorionic Twin, Ultrasound

#### INTRODUCTION

Acardiac twinning also referred to as the twin reversed arterial perfusion sequence (TRAP) represents an extreme manifestation of the monochorionic twin pregnancy and has an incidence of approximately 1 in 35,000 deliveries [1]. 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 communication with placenta. The acardiac twin presents with many bizarre anomalies thought to be due to low oxygen tension and dramatic alterations in the fetal physiology. Complications in the pump twin of an acardiac fetus include congenital heart failure, polyhydramnios, preterm labor and death in 50-70% of the cases. We present a case of acardiac –acephalus twin which was initially mistaken for a dead anomalous fetus.

#### CASE REPORT

A 24 year old 3<sup>rd</sup> Gravida having two full term vaginal deliveries with both baby living was referred to us at 30 weeks gestation as twin pregnancy with one live and one dead fetus along with polyhydramnios. Her clinical examination revealed a term size uterus with No fetal heart sound audible by stethoscope. She was admitted for conservative treatment. Later Ultrasonography with Colour Doppler showed two fetuses inside the uterus, only single placenta with absence of amniotic membranes suggestive of monochorionic monoamniotic twin pregnancy. The first fetus was apparently normal but without any cardiac activity. The second fetus was also dead (26 weeks gestational age) but showed gross congenital malformations in the form of absence of fetal cranium replaced by soft tissue density mass lesion, spinal deformity, gross edema of limbs, abdomen and body. After blood coagulation profile was done, labour was induced. She delivered subsequently, first twin, F1

which was stillborn and weighed 475gms, F2 delivered after 2hrs and weighed 450gms. whose only recognizable parts were lower limbs that too deformed. On gross examination the proximal part of the fetus was covered with a gelatinous swollen skin and no heart or lung like structure. There were no upper limbs or head and neck. (Figure 1). Radiograph of the fetus showed normal skeletal features below the umbilicus but no bony shadow was present in the upper part of the body. A single placenta was delivered with single amniotic membrane and two cords inserted side by side, one of them was very small and attenuated which belonged to the acephalus twin. Postnatal period was uneventful and she was discharged on 4<sup>th</sup> postnatal day.



Fig-1: Showing the Acardiac-acephalus twin fetus

## DISCUSSION

Acardiac twinning is a rare congenital anomaly characterized by formation of a malformed fetus with an absent or rudimentary (but nonfunctional) heart. 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, while an *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-amorphus* fetus, has the most severe malformation and lacks all cephalic and truncal differentiation [2].

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 [3]. This flow pattern has been termed "twin reversed-arterial-perfusion" (TRAP) 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. The twin with cardiac activity may be hydrotic or malformed in 9% of cases. This twin is often at high risk for congestive cardiac failure due to increased cardiovascular demand, resulting in preterm delivery. Perinatal mortality for the pump twin has been estimated to be up to 55%, while it is usually fatal for the acardiac twin [4].

Twin reversed arterial perfusion (TRAP) 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 [5]. 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 [6]. The other therapy is fetoscopic placental laser surgery directed at the vascular connection between the twins. Recently High intensity focused ultrasound (HIFU) has been described as a non-invasive treatment for occlusion of blood flow in the acardiac twin [7]. No comparative studies with HIFU exist.

## CONCLUSION

This case demonstrates the importance of correct early diagnosis of acardiac twinning to avoid late complications. Referring every twin pregnancy for a detailed early ultrasound to a specialist centre can avoid diagnostic and therapeutic difficulties later in the pregnancy.

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