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Case Report

# Acardiac Twins: A Rare 'TRAP' Case

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**Abstract**

Acardiac twinning, a rare congenital anomaly of monozygotic twin pregnancy, often results from abnormal placental anastomosis. This leads to reverse arterial perfusion with complex pathophysiology. Among the twin, acardiac twin is one of the twins that fails to develop head, arms and heart and gets its entire blood supply from structurally normal pump twin. The term reversed perfusion is used to describe this condition because blood enters the acephalic twin through umbilical artery and exit through umbilical vein which is opposite to the normal supply. The acardiac twin loses direct vascular connection with placental villi and receives its entire blood supply from pump twin. Here we are presenting a primigravida of 26 weeks with monochorionic diamniotic twin gestation. One of the twins was acardiac acephalous and the other pump twin with hydrops.

**Keywords:** Twin Reversed Arterial Perfusion Sequence; Acardiac Twin; Monochorionic Twin.

## Introduction

Multiple pregnancies accounts for 1.5% of all pregnancies, with approximate perinatal morbidity and mortality of 10% [1]. Multiple pregnancy is complicated by congenital malformations twice as often as with singletons. Certain malformations, such as conjoined twins and chorioangiopagus parasiticus due to twin reversed-arterial-perfusion (TRAP) sequence are unique to multiple pregnancy. TRAP sequence is an extremely rare anomaly with an overall incidence of 1 per 35,000 births, amounting to an average risk of 1% among monozygotic twins [2]. Other names given are holocardius, hemicardius, fetus amorphous.

Occurrence of acardiac twin is due to twin reversed arterial perfusion sequence (TRAP) occurring early in embryogenesis. There is vascular communication between the twins in monozygotic twins. The vascular communication in acardiac twin is different, in that, the acardiac twin receives blood supply from other twin-pump through umbilical artery. The blood in the umbilical artery is mostly deoxygenated. Hence it leads to secondary organ atrophy [3]. Upper body does not develop at all, hence missing heart and head. All the blood supply to the acardiac twin is derived from the pump twin. The acardiac twin develops only lower part of body or just a mass of tissue (fig. 1). Hence the mortality of acardiac twin is 100%. The pump twin suffers congestive cardiac failure and hydropic changes due to pumping blood to the acardiac twin. Mortality for pump twin is 50-70%. However, early identification and follow up with treatment improve the survival rate of pump twin.

## Case Report

A 22 year old primigravida admitted to our hospital at 26 weeks with complaints of pain in abdomen and leaking per vaginum since 6hours. Her examination revealed a uterus size of 26-28 weeks. Per vaginum examination confirmed a frank leak with well effaced cervix with 4cm dilatation and absent membranes. USG done on the same day reported twin gestation with a single placenta and a thin membrane separating the two foetuses–monochorionic diamniotic twin. Twin A of 26weeks size, normal development but with hydropic changes, grossly

increased liquor (28cm). Twin B, a malformed fetus with lack of head, thorax, upper extremities and only abdomen and lower extremities formed with multiple intra abdominal cystic structures – acardiac twin.

Within few hours patient delivered (fig.2), twin A, an alive female baby of 1100 gms, followed by the acardiac twin of 800 gms. The first twin was found to be normal without any external abnormalities. It was shifted to neonatal care unit, but perinatal mortality occurred. The acardiac twin had lower limbs and the lower trunk only. Both feet showed equinovarus deformity. Placenta was 450 gms with two umbilical cords. The normal twin cord was long and edematous and had three vessels. The acardiac twin had short thin cord. Both twins shared same placenta.

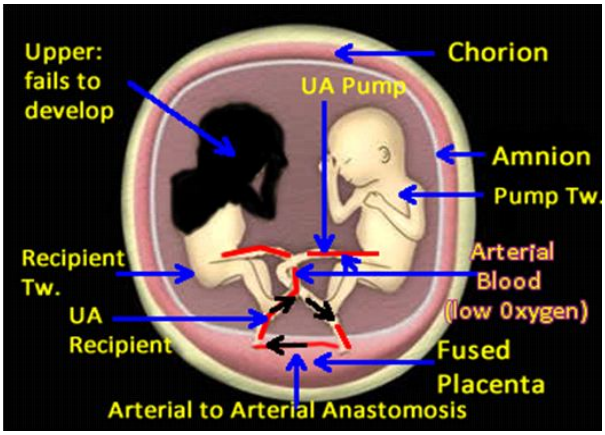


**Fig. 1:** Schematic diagram; TRAP sequence

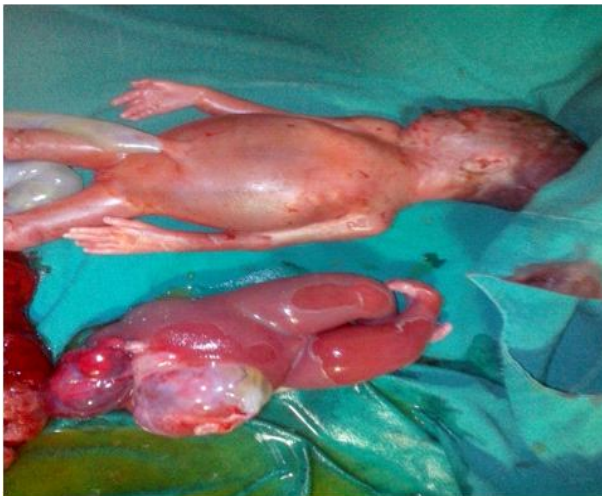

**Fig. 2:** Pump twin and Acardiac twin.

## Discussion

There are vascular connections in monozygotic twins. Twin to twin transfusion syndrome (TTS) is one of the manifestations affecting up to 15% of monochorionic twins. In this condition there is disproportionate blood supply between the twins. Mortality is high without treatment.

TRAP sequence is one rare occurrence. Here the pump twin pumps blood in reversed way, through the umbilical artery, in the acardiac twin. The acardiac twin suffers 100% mortality. Acardiac twin is classified according to the degree of cephalic and truncal maldevelopment [4].

1. Acardius-acephalus: Absent cephalic structures. Head and upper extremities are lacking. It is most common variety. This is the type seen in the present case.
2. Acardius-anceps: Some cranial structures and neural tissue is present. The body and extremities are also developed. It is highly developed form.
3. Acardius-acromus: Cephalic structures present but no truncal development seen. The umbilical cord is attached to the head. It is the rarest form of acardia.
4. Acardius-amorphous: No distinguishable cephalic or truncal structure. It is least developed and not recognizable as a human form with minimal development. This differs from teratomas only by its attachment to an umbilical cord.

A late separation of the embryonic cell mass results in a monochorionic twin pregnancy in monochorionic pregnancies, anastomotic vessels are established connecting the two circulations. Retrograde perfusion via the anastomotic channel prevents the normal cardiac development due to lack of sufficient oxygenated blood. The heart, if develops, is either tubular or completely infantile. Thus, the acardiac fetus becomes dependant on the perfusion of the "pump" twin.

The pathogenesis in TRAP sequence include [3, 4]:

1. Deep placental anastomoses in early embryogenesis cause malformation of the acardiac twin. The early pressure flow in one twin exceeds that of other leading to the reversal of flow in the umbilical artery of the co-twin.
2. A primary defect in embryogenesis in one twin leads to failure of cardiac development. The normal twin then perfuses the acardiac twin via artery- artery anastomoses. The anastomoses are not responsible for the cardiac anamoly but are established as a result of it.

Diagnosis of cardiac twin should be made early by ultrasound and Doppler by recognizing the absence of heart and reversal of flow in umbilical artery. Once diagnosed it should be followed up to weight-ratio of twins, changes in the pump twin like cardiac failure and polyhyrdamnios. Preterm delivery is strongly

associated with the development of hydramnios and congestive heart failure in the pump twin [5]. Conservative management in normal twin only if acardiac twin is small in size. Invasive treatment is required when pump twin is having early signs of heart failure to improve outcome [6,7].

*Treatment*

Each pregnancy has to be assessed individually. Minimal invasive procedures like percutaneous insertion of helical coil to induce thrombogenesis in single umblical artery of acardiac twin can be done [8,9]. Blocking the vessels by coagulation using Nd:Yag laser and radiofrequency ablation [10] under ultrasound guidance are now the first line treatment.

## Conclusion

Diagnosing of acardiac twin can be made in first trimester itself by USG and Doppler. Early diagnosis of chorionicity of twin pregnancy helps in improving the survival of the pump twin. Prevention of preterm labour and diagnosing cardiac failure in the pump twin is very important. First line of treatment is by blocking the vessel of acardiac twin by blocking the vessel of acardiac twin by radiofrequency ablation by ultrasound guidance. Treatment at appropriate time improves the survival of the pump twin.

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