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Case Report

# TWIN REVERSED ARTERIAL PERFUSION SEQUENCE (TRAP SEQUENCE). THE ACARDIAC /ACEPHALIC TWIN

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## ABSTRACT

**Twin-Reversed Arterial Perfusion (TRAP sequence)** is a rare complication of monochorionic twins (MC, twins sharing one placenta). TRAP sequence is known as acardius or chorioangiopagus parasiticus. It occurs in 1% of monochorionic twin pregnancies and in 1 in 35,000 pregnancies.

The risk of recurrence was estimated 1:10,000. TRAP sequence is characterized by a structurally normal pump twin perfusing an anomalous twin. In TRAP syndrome, there is mortality and deformities in both twins.

The acardiac/acephalic twin is a parasitic twin that fails to develop a head, arms and a heart. The parasitic twin is with or without legs, receives its blood supply from the host twin (Pump twin). In TRAP sequence, one twin is usually developmentally normal (pump twin) and the other twin has a serious condition, either missing a heart (acardiac) or a head (acephalic) or both, that prevents it from surviving on its own. The acardiac/acephalic twin receives all of its blood from the normal or "pump" twin. The abnormal fetus does not have a heart; all of its blood must come from the normal twin. And it is kept alive by the blood which is pumped from the normal twin through the placenta. The term "reversed perfusion" is used to describe this scenario because blood enters the acardiac/acephalic twin through reversed flow through its umbilical artery and exits through the umbilical vein, which is opposite to the normal blood supply of the fetus.The acardiac twin loses direct vascular connection with the placental villi and receives its entire blood supply from the pump twin [1,2,3,4].

Proper timing of the delivery is of prime importance for the survival of the normal fetus. The emphasis is placed on close sonographic monitoring from early antenatal diagnosis. The mortality of the acardiac twin is 100%, and the perinatal mortality of the pump twin is reported to be around 50%. The mortality rate of the pump twin appears to correlate with the size of the acardiac twin and primarily due to congestive heart failure.

**We present such a case of TRAP sequence because of its rarity.** TRAP sequence, was diagnosed by ultrasound at the obstetrics and gynecology department at KAMS & RC (Kamineni) and it was closely followed up until delivery. Clinical presentation, ultrasound findings, outcome and management were reviewed in present article.

**KEYWORDS:** Monochorionic twins; Pump twin; Reverse flow; Acardiac / Acephalus.

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## INTRODUCTION

Acardiac Twin or TRAP Sequence is a very rare problem, with an incidence of, 1 in 35,000 pregnancies**,** occurring in approximately 1% of monochorionic twins. One twin is structurally normal and is referred to as pump twin because it pumps blood to other which is abnormal consisting of legs and lower body but no upper body, head or heart. Abnormal twin is therefore referred as Acardiac twin. The acardiac twin has no chance of survival. This pattern of development may be explained by the mechanism of perfusion of the acardiac twin. The deoxygenated blood from the normal twin enters the abdomen of the acardiac fetus, allowing for some development of the lower body and extremities. Once blood reaches the upper half of the body, oxygen saturation is extremely low,

halting development of this area. The acardiac twin is thus, a parasite. It requires blood pumped from the normal twin to keep developing, putting the pump fetus at risk of high output cardiac failure. The risk is directly dependent on the size of the acardiac twin and the mortality rate for the normal (pump) twin is approximately 50%. The majority of pump twins are congenitally normal. This article is to highlight the acardiac /acephalic twin (Twin-Reversed Arterial Perfusion sequence or TRAP) and its clinical variable presentation.

## CASE REPORT

A 25 years old primigravida was 36 weeks of gestation with ultrasound report performed at a peripheral hospital which revealed a twin pregnancy. She was examined in our hospital Kamineni Academy of Medical sciences & Research Centre in the OBG department. Her clinical examination revealed uterus size of 36 weeks of gestation with multiple fetal parts. Ultrasonographic evaluation revealed twin pregnancy with monochorionic placenta.

The Twin A was with breech presentation, normal fetal anatomy and with normal liquor volume. Grossly abnormal anatomy was noted in Twin B. There was no definition of fetal skull above the thorax and the spine ended abruptly. Ultrasound findings were consistent with TRAP sequence which was confirmed by MRI. **(Fig-1)**

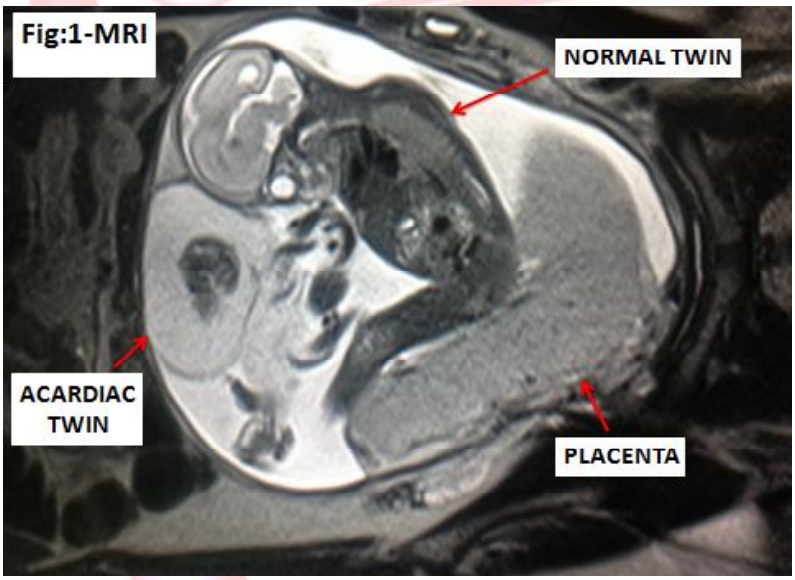


**Fig. 1:** MRI Scan Showing Twin Reversed Arterial Perfusion Sequence (TRAP sequence).

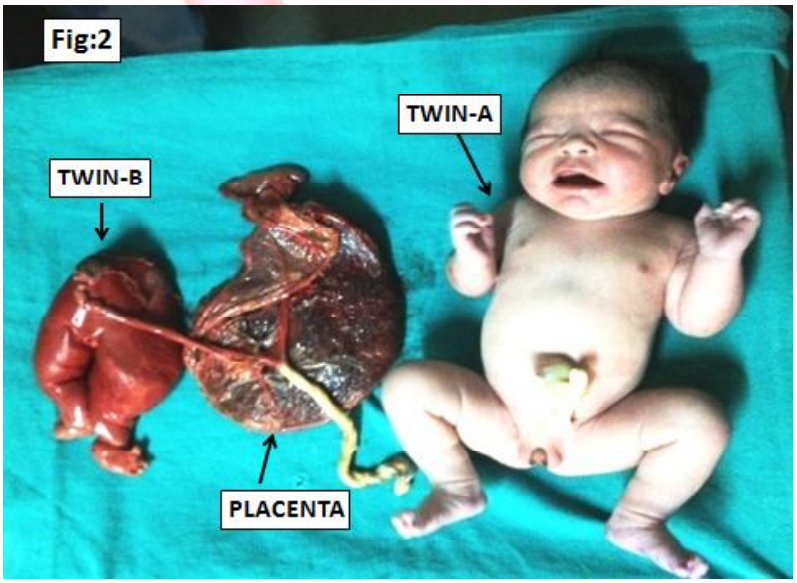


**Fig. 2:** Showing Twin Reversed Arterial Perfusion Sequence (TRAP sequence) foetuses with Placenta.

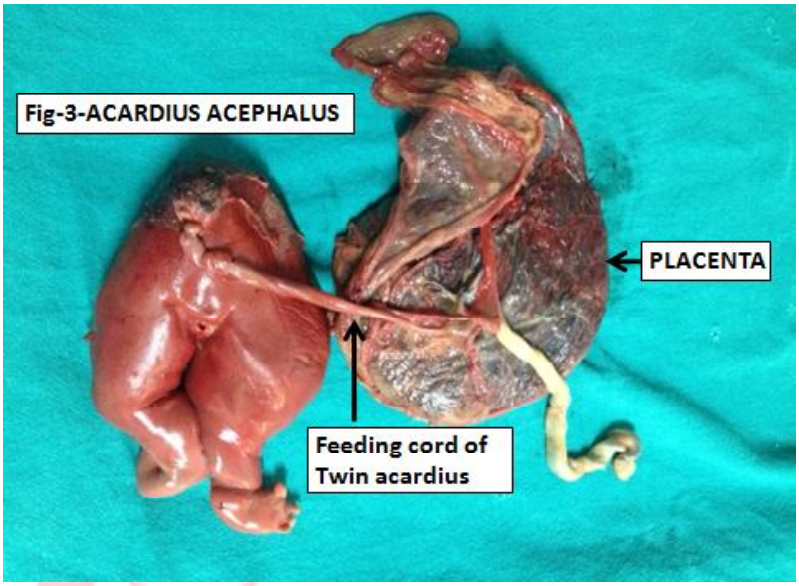


**Fig. 3:** Showing Acardius Acephalus foetus with Placenta.

## OBSERVATIONS & RESULTS

Cesarean section was done. First baby or twin A was female and was to be found normal without any external malformation with weight of 2.3 kg and admitted in neonatal intensive care unit**. (Fig-2)**

While the second baby, twin B was Acardiac/ Acephalus. Twin B was with absent development of cephalic pole, heart and upper limbs. It had well-formed lower limbs and the lower trunk which was normal. Both feet showed equinovarus deformity. Weight of the second baby was 400gms**. (Fig-2)**

Placenta was 500 grams. Two Umbilical cords were present. The normal twin cord was long and edematous, twin B had a short cord and both sharing the same Placenta**. (Fig-2)** Patient was transferred to ward in a satisfactory condition and was discharged from hospital on 5th postpartum day with a healthy female baby. Patient was given appointment for six weeks in postnatal clinic.

**Anatomical Examination** of the Acardia acephalus fetus:

The fetus acardius was covered with reddish brown (darker than that of normal twin) skin. The skin was soft and edematous. Both the lower limbs were near normal except for equinovarus deformity of the foot. There were no upper limbs, no head and the top of the fetus coming to a blunted end. External genitalia were ambiguous (**Fig-3)**

## DISCUSSION

Twinning is the most common type of multiple gestations. Twin reverse arterial perfusion syndrome (TRAP) was first defined by Grunewald in 1942. The acardiac fetus or Chorioangiopagus parasiticus or Trap Syndrome is one of the forms of twin to twin transfusion (TTT).

TRAP syndrome occurs in monochorionic gestation form. In which case one twin has Acardia (the recipient) with no heart and the other is structurally normal (pump or donor twin). Due to the absence of heart, in the acardia twin, the pump twin supplies deoxygenated blood via vascular anastomoses to the acardiac twin. Acardia twin does not send blood to placenta and all its blood comes from and goes back to the circulation of the pump twin, through the vascular connections on the surface of the shared placenta.

It is therefore structurally normal twin perfusing an anomalous recipient twin via an artery-to-artery anastomosis in a reverse direction**.** The reversed flow is through its umbilical artery and exits through the umbilical vein, which is opposite to the normal blood supply of the fetus.Deoxygenated, low-pressure blood from the pump twin, which would normally return to the placenta, instead flows directly to the acardiac twin, resulting in a wide array of structural abnormalities, caused by arterio-arterial and veno-venous placental anastomoses. The acardiac twin is usually grossly abnormal with severe reduction anomalies of

the upper part of the body **as seen in our present case**. Mortality of the pump twin is 50-75% usually due to the result of heart failure and of the acardiac is 100%.

The upper half of the body of an acardiac twin is extremely poorly developed and, sometimes, not developed at all. Head, cervical spine and upper limbs are usually absent. In contrast, the lower half of the body, although malformed, is better developed. The risk is directly dependent on the size of the acardiac twin: the higher the weight of the acardiac twin, the higher the risk of cardiac failure and death for the normal twin.Therefore classical TRAP/Acardius sequence is due to a retrograde flow from the umbilical arteries of the pump twin to the iliac arteries of the acardiac twin resulting in preferential caudal perfusion, which results in acardius/ acephalic fetus allowing for some development of the lower body and extremities. It is thought to be a result of an early embryopathy. **(Fig-4)**

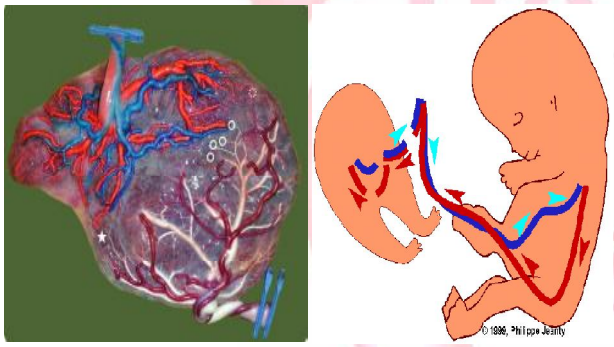

**Fig .4:** Arterio-arterial and veno-venous Placental Anastomoses of the Twins.

Twin reversed arterial perfusion (TRAP) sequence, also known as acardia, is a rare anomaly unique to multiple gestation in which one twin has an absent, rudimentary, or nonfunctioning heart. Schatz (1898) classified acardia into two main groups: hemiacardius (imperfectly formed heart) and holoacardius (absence of heart).

**It is classified according to the degree of cephalic and truncal maldevelopment [5,6].**

1.The first type is Acardius acephalus, where no cephalic structures present.( head & upper extremities are lacking. It is most common variety. (**Seen in present case).**

2.The second is Acardius anceps where some cranial structure and neural tissue or brain tissue is present. The body and extremities are also developed. It is highly developed form.

3. The third is Acardius acormus with cephalic structure but no truncal structures are present.i.e. Head without a body. The umbilical cord is attached to the head. It is rarest form of Acardia.

4. The fourth type is Acardius amorphous with no distinguishable cephalic or truncal structure. It is least developed and not recognizable as human form with minimal development. This differs from Teratomas only by its attachment to an umbilical cord.

**Epidemiology [7,8]:**

Acardiac monsters have been known since 1533when the condition was first described by Benedetti. Acardiac always occur in a multiple birth and two third are diamniotic. The occurrence rate is estimated to be less than one in 340 deliveries, 1% of monozygotic twin pregnancies and 1 in 30 monozygotic triplets. Acardiacs were predominantly female .Two-third of acardiac fetuses is Acardiac acephalic, which is the most common variety among the various types of acardiac mentioned in the literature.

**The present case in our study was a female with monochorionic, diamniotic placenta and was Acardiac acephalic type which is most common variety.**

A late separation of the embryonic cell mass results in a monochorionic-amniotic twin pregnancy. Anastomosis of vessels establishes a connection between the 2 circulations. Retrograde perfusion interferes with normal cardiac development, which rarely goes beyond the stage of tubular heart. Thus the acardiac fetus becomes dependent on the perfusion of the “pump” twin [9].

**The TRAP sequence is with two main pathogenetic hypotheses.**

1. Deep placental anastomoses in early embryogenesis causes malformation of the acardiac twin. The early pressure flow in one twin exceeds that of other and leads to reversed circulation in the twin who exhibits perfusion.

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 anomaly [10].

**Treatment**:

Therapeutic options targeted at interrupting the vascular anastomosis between the twins under ultrasound guidance using fetoscope. Several different techniques have been used to treat TRAP sequence by interrupting the connection between the acardiac twin and the pump twin to increase the chances that the pump twin will survive. These techniques include cord occlusion by embolization, ligation, laser photo-coagulation, monopolar and bipolar diathermy. Intrafetal ablation has also been performed by alcohol injection, monopolar diathermy, interstitial laser, and radiofrequency ablation (RFA) [11]. **(fig-5)**

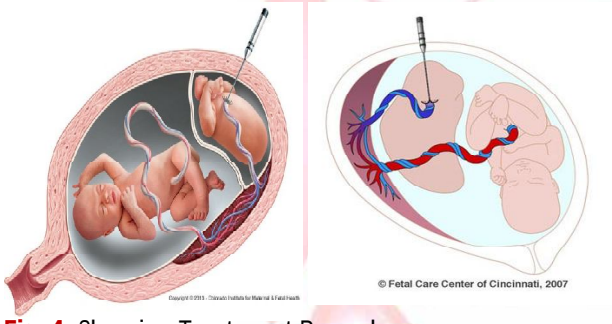


**Fig .4:** Showing Treatment Procedure.

## CONCLUSION

Accurate antenatal diagnosis is essential to improve the prognosis of this rare entity of TRAP sequence**.** Improved imaging techniques like 2D ultrasonography, 3D ultrasonography and transvaginal Doppler ultrasonography have made the diagnosis of Acardia possible even in the first trimester of pregnancy by detecting inversion of vascular flow in the recipient acardiac fetus. Early diagnosis may help to reduce the risk of such complications [12]. There could be 95% survival in the pump twin with an average age at delivery between 36 and 37 weeks. No significant neurological abnormalities have been identified in these children so far.

It is also important to exclude a chromosomal abnormality prior to offering a fetoscopic procedure in TRAP sequence since the incidence of chromosomal abnormality in the pump twin may be as high as 9 percent.

**Conflicts of Interests: None**

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