

Case Series of Twin Reversed Arterial Perfusion (TRAP) Sequence at GGH, Kakinada, A.P.

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Abstract: *Twin Reversed Arterial Perfusion (TRAP) sequence is associated with poor prognosis, seen only in monochorionic monozygotic twin pregnancies. The incidence is one in 35,000 births and one in 100 monozygotic twin pregnancies. This entity is characterized by a recipient fetus exhibiting lethal anomalies including acardia and a donor fetus with complications like heart failure or premature labor caused by polyhydramnios. Herein, we report 4 cases of TRAP sequence presented to the radiology department for routine obstetric ultrasound.*

Keywords: TRAPS, ACARDIAC TWIN

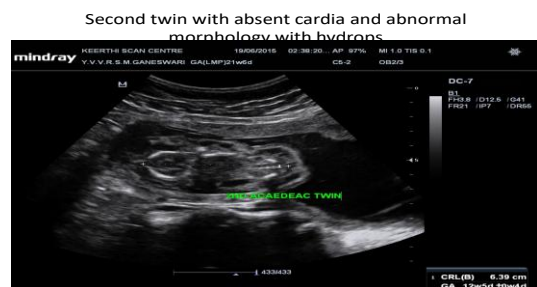
1. Introduction

Twin Reversed Arterial Perfusion (TRAP) sequence is a phenomenon with poor prognosis which is seen in 1/100 of monochorionic twin pregnancies. In TRAP sequence, there is a non-viable, acardiac fetus that has multiple anomalies and a donor fetus that feeds this fetus via vascular anastomosis in the placenta in the form of direct communication of umbilical artery of donor twin to the umbilical artery of recipient twin, so impure blood goes in a reverse direction to supply co-twin causing multiple anomalies including absent heart, head, upper limbs and gross body edema. The abnormal twin has no direct vascular communication with the placenta. Mortality is 100% in acardiac twin. Mortality of donor twin is around 50%, and death is usually due to heart failure and sometimes prematurity caused by polyhydramnios.

2. Case Presentation

1. A 22 year old primi came for follow up antenatal scan at 13 wks gestation. Previous scan was at 8 weeks gestation suggestive of monochorionic twin gestation with one live normal fetus and other twin with no cardiac activity diagnosed as non viable gestation.

At 13 weeks follow up scan normal viable 1st Twin showed adequate interval growth with no obvious fetal abnormalities and 2nd twin showed abnormal morphology with generalised body edema/hydrops with small head, ill developed upper extremities, absent heart, deformed and relatively well developed lower extremities with sluggish movements suggesting acardiac twin.



Aborted specimen with both normal and acardiac twin

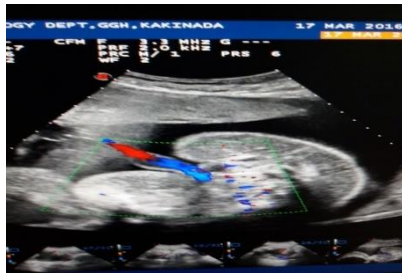


Under the light of these data, twin-twin transfusion syndrome and TRAP sequence were considered in the radiologic differential diagnosis of this complicated monochorionic–monoamniotic twin pregnancy. Chorionicity, presence of hydrops, acardiac twin on USG were consistent with TRAP sequence.

2. A 21-year-old pregnant woman (primi) with 23 weeks of gestational age was referred to our hospital for routine ultrasonography. On obstetric ultrasound it's a triploid gestation with two normal live fetuses of 22- 23 weeks gestation and one abnormal amorphous fetus with no cardia, a large cystic structure noted in the region of head, malformed lower limbs, absent upper limb and diffuse body edema. There are two separate placentae with intertwin membrane with one normal viable twin and acardiac twin in one amniotic sac with both umbilical cords inserted at single site of placenta and one normal live fetus in another amniotic sac with separate placenta. Umbilical cord of acardiac twin shows single umbilical artery with reversal of flow.



Amorphous cardiac fetus with cystic structure in the region of head with absent cranium and malformed limbs.



Acardiac acephalus twin with single umbilical artery.



Aborted specimen showing amorphous mass with absent cranium, upper limbs with malformed lower limbs



X ray of the specimen showing absent skull vault, facial bones, ribs and upperlimbs with malformed lowerlimbs.



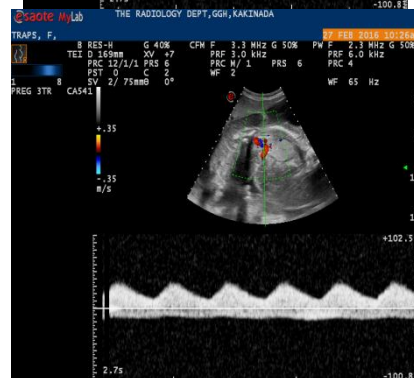
Post abortal placental specimen with contrast (iopomidol) injected into umbilical artery of normal fetus shows direct communication of umbilical artery of acardiac twin denoting

twin reversal arterial perfusion with out any intervening capillary network in the placenta.

3. A 22-year-old pregnant woman (G2P1L1) with 26 weeks of gestational age was referred to our department for ultrasonography. On obstetric ultrasound monochorionic monoamniotic twin gestation with one normal fetus of 26 weeks and second fetus with large deformed body, absent heart and cranium. On Doppler study of umbilical artery of abnormal twin showing reversal of flow.



Abnormal foetus with absent cardia



Doppler sonography showing reversal of flow in the abnormal fetus

Under the light of these data, twin-twin transfusion syndrome and TRAP sequence were considered in the radiologic differential diagnosis of this complicated monochorionic–monoamniotic twin pregnancy. Chorionicity, presence of hydrops, acardiac and acephalic twin and Doppler USG findings were consistent with TRAP sequence.

4. A 20-year-old pregnant woman (G2P1L1) with term gestation was referred to our department for follow up ultrasonography and Doppler examination with prior diagnosis of twin gestation with one abnormal twin. As

Patient refused for termination at the time of diagnosis, pregnancy was continued and referred to ggh,Kakinada for institutional management.

On obstetric ultrasound monochorionic monoamniotic twin gestation with one live foetus of 34 weeks and other abnormal foetus with absent cardia, diffuse body wall edema with absent cranium was found. Normal live twin show normal S/D ratio in umbilical artery doppler study. Woman was delivered spontaneously at 37 weeks gestation a live normal male baby of weight 3 kgs. There is difficulty in delivering the second abnormal twin and emergency LSCS was done and delivered large abnormal acardiac second twin.



Gross specimen showing large amorphous featureless mass with absent cranium, upper limbs and malformed lower limbs

3. Discussion

TRAP sequence represents a variant of conjoined twins in which chorionic circulation is shared. Organogenesis defect due to anastomosis in the placenta during the early embryonic period is suggested in the pathogenesis of TRAP sequence. In this entity placental blood circulation between acardiac fetus and donor fetus is provided by artery-artery and vein-vein anastomosis. The acardiac twin survives through these anastomoses, it uses blood which has poor oxygen, coming from cardiac twin flows reversely in arteries of the acardiac fetus and normal tissues of this fetus are partially atrophic. Atrophy is higher in the cranial parts as the caudal part uses blood predominantly.

Antenatal diagnosis of acardiac twin may be made by ultrasonography by the absence of the heart despite the presence of trunk and extremity movements with multiple anomalies. On doppler ultrasonography placental vascular anastomosis with reversal flow to the recipient twin can be demonstrated. Prenatal diagnosis of an acardiac fetus may be made with ultrasonography at the end of the first trimester.

An acardiac fetus is clinically divided into two types. Pseudocardiac is the presence of cardiac structures, although rudimentary. Halocardia is the absence of cardiac structure development. It is widely classified into four subgroups morphologically.

- 1) Acardiac acephalic: the fetus has developed pelvis and lower extremities. Head, arms, and thoracic organs are absent. This is the most common type with a frequency of 60 - 75%.

- 2) Acardiac aniceps: body and extremities have developed. Head and face are partially formed. This type consists of approximately 20% of all cases.
- 3) Acardiac acormus: only the head of the fetus has developed. It is quite rare and consists of approximately 10% of all cases.
- 4) Acardiac amorphous: the fetus has no identifiable organs. It is as an amorphous tissue mass, and it consists of approximately 5% of all cases.

Mortality is 100% for acardiac twin. Mortality of pump twin is around 50%, and death may usually be due to heart failure and sometimes prematurity caused by polyhydramnios. Chromosomal anomalies that are reported as 9% should be excluded for management of TRAP sequence. Significance of early diagnosis is clear both for determining the therapy and also timely terminating the pregnancy, which has a high mortality. Continuance of the growth of the acardiac twin, presence of polyhydramnios, cardiomegaly and pericardial effusion are poor prognostic factors. On color Doppler ultrasonography, low pulsatility index in the umbilical artery of the donor twin indicates poor prognosis. Options for treatment of TTTS include serial amnioreduction, laser photocoagulation of communicating vessels, septostomy, and termination.

In conclusion, TRAP sequence is a complication that is seen in monochorionic twin pregnancies and it has a poor prognosis. Selection of the proper treatment modality by making the diagnosis with typical ultrasonography and Doppler findings is of great importance.