

Spectrum of progression of TRAP sequence (acardiac anecephalic twin): a rare case report

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Abstract

TRAP (Twin reversed arterial perfusion) sequence is one such sequelae of monochorionic diamniotic multifetal pregnancy wherein one twin acts as a pump twin or perfusionist and second twin as an amorphous acardiac structure connected to this pump through an abnormal placental vascular anastomosis in reverse direction. This is an extremely rare complication occurring in 1 per 35,000 births. The acardiac twin which has either an absent, rudimentary, or non-functioning heart due to Compression of the cephalic pole of the embryo inhibiting curving and fusion of the primitive heart tube is the basic cause resulting in non-formation of the heart and other organs. However, whether this TRAP sequence is the cause or effect of acardia is still unknown. The mortality of pump twin is 50-75% and acardiac twin is 100%. 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. Serial vigilant sonography and doppler evaluation is the "mainstay" in early detection of TRAP sequence. With advances in foetal medicine, many treatment options are available with minimally invasive techniques. We here presented one such rare case in which the diagnosis was reached quite early through sonography with doppler flow studies and it reemphasize the importance of ultrasonography as a simple but adequate tool not only in the diagnosis and follow-up but also in management of such a case.

Keywords: Acardiac/Anecephalic; Monochorionic twins; Pump twin; twin reversed arterial perfusion

Introduction

The incidence of congenital anomalies is more in monochorionic multifetal pregnancy. TRAP (Twin reverse arterial perfusion) sequence is one such sequelae wherein one twin acts as a pump twin or perfusionist and second twin as an amorphous acardiac structure connected to this pump through an abnormal placental vascular anastomosis in reverse direction [1].

This is an extremely rare complication occurring in 1 per 35,000 births [1, 2, 3]. The risk of recurrence of acardiac twins was estimated to be 1 in 10,000 deliveries [2]. Twin reversed arterial perfusion (TRAP) sequence, also known as acardia, in which one twin has an absent, rudimentary, or non-functioning heart in which the arterial blood flows in a retrograde direction

from the pump twin to the recipient acardiac twin through a common placenta [4]. Compression of the cephalic pole of the embryo inhibiting curving and fusion of the primitive heart tube is the basic cause resulting in non-formation of the heart and other organs. However, whether this TRAP sequence is the cause or effect of acardia is still unknown. The pump or donor twin may develop cardiac failure because of the anomalous perfusion circuit. Polyhydramnios is significantly associated with presence of renal tissue in the acardiac twin. An acardiac pump twin weight ratio (> 50%) is associated with the development of polyhydramnios and preterm labour. Identified high-risk factors for poor obstetrical outcome include: acardiac anecephalic, polyhydramnios, acardiac twin with ears, and pump twin cardiac failure [5]. 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.

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Case Report

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 twin. Several intervention techniques including hysterotomy with selective delivery of acardiac twin, umbilical cord block using coils, laser coagulation, alcohol ablation, bipolar coagulation and radio-

frequency ablation are employed to save the pump twin [6, 7, 8].

We present one such case in which the diagnosis was reached quite early in second trimester through antenatal check-ups and sonography with Doppler flow studies. This allowed the parents to choose option of termination of pregnancy to avoid adverse physical and psychological sequel for both mother and foetus.

Case Report

A 25-year-old, third gravida spontaneous conception, under treatment for hypothyroidism reported to us after two months of amenorrhoea. She had one normal delivery 5 years back and one missed abortion 2 years back. At 8 weeks of gestation sonography showed twin pregnancy with single yolk sac, one live foetus and other foetus showing no cardiac activity. Patient and relative counselling done with close follow up advised to them. They reported to us after one month with sonography report showed twin A normal gestational growth 13 weeks 2 days but twin B continued to show absent cardiac activity and growth up to 11 weeks 5 days. The unusual development raised suspicion which was confirmed by level 3 scan with detection of an abnormality in twin B which appeared like Cystic hygroma (Figure 1).



Figure-1: Gross photograph showing Twin B which appeared like cystic hygroma

Patient and relative were counselled about anomaly of twin B. They decided to have a repeat scan and second opinion. Patient had lost to follow up with us after this.

She had again reported to us after one month, sonography at this stage showed twin A 19 weeks in gestation with polyhydramnios with no anomaly. Twin B corresponded to 18 weeks in gestational growth with Acardia, marked swelling of whole body and poorly defined upper half of body morphologically with scanty liquor with thin wispy intertwining membrane (Figure 1). The Doppler study revealed grossly resistant flow in descending aorta of this anomalous foetus with a communicating artery between two showing reverse Doppler flow (Figure 2). This raised the possibility of TRAP sequence.

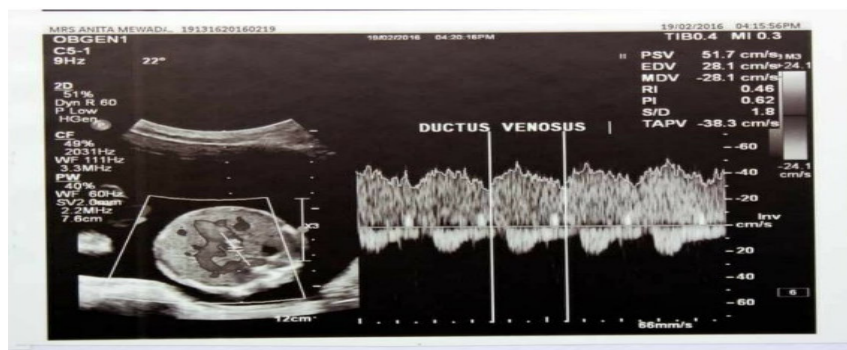


Figure-2: Gross photograph showing a communicating artery between twin foetuses with reverse doppler flow

Case Report

This pregnancy was terminated as per patients wish by medical method of MTP. Before beginning with the process of MTP, amnioreduction was carried out after obtaining required consent from patient and her husband. She aborted both foetuses within 22 hours after induction of abortion. Anatomical findings at abortion were as follows: -

Twin A- female foetus, weight 410 grams, extremely pale, no obvious external anomaly

Twin B- indeterminate sex, weight 280 grams, dark pink in colour, amorphous mass, red oedematous soft skin with well-developed lower limbs with talipes equino varus defect of feet with poorly developed cephalic pole (face, neck, thorax and upper limbs).

Placenta- single large 16 by 13 cm. with no abnormality on both surfaces except that cord of twin B was replaced by a tubular structure of 8 cm long with only two vessels in it without Wharton's jelly which was attached directly to placenta with anastomosis with twin A arterial vessel. The sac was single with thin intertwine membrane.

Autopsy was refused by patient, so internal details couldn't be ascertained.

This case made us conclude with final diagnosis of monochorionic diamniotic twin pregnancy with TRAP sequence of Acardiac Anceps type with arterio-arterial twin to twin anastomosis.

Discussion

The overall incidence of monozygotic twin pregnancy is at 3.5 per 1000 live births [9]. Monozygotic twin of monoamniotic monochorionic (Mo-Mo) type and of diamniotic monochorionic (Di-Mo) type result if twinning occurs within 4-8 days or after 8 days of fertilization. Both these types have a great risk of Twin to Twin Transfusion Syndrome, discordant twins, cord entanglements and ultimately perinatal mortality as high as about 25-30%. TRAP sequence is one form of twin to twin transfusion syndrome occurring very rarely in 1 in 35,000 births [1, 2] where one of the twins is acardiac and gets perfused by second healthy pump twin with deoxygenated blood via abnormal vascular anastomosis on the surface of placenta in reverse direction [10].

The mortality of pump twin is 50-75% and acardiac twin is 100%. Benedetti reported first case in 1533 and Grunewalelun defined the whole sequence in 1992 [2].

The acardiac twin suffers grossly from reduction type of anomalies in upper part of the body. The donor twin suffers from perinatal mortality in 50-75% cases due cardiac failure. The larger the size of acardiac twin, the higher is the risk of cardiac failure of pump twin [10].

The retrograde flow from umbilical artery of pump twin to iliac arteries of acardiac twin results in preferential caudal perfusion and development of lower half of body. TRAP sequence may be Hemicardius (imperfectly developed heart) or Holocardius (absent heart).

There are four types of TRAP sequence:

- a) Acardius Acephalus- Absent head +Lack of upper extremities + Absent heart
- b) Acardius Anceps- Some cranial & brain tissue & body extremities are present + absent heart
- c) Acardius Acormous---Cephalic structure present + cord attached to head + absent trunk---very rare
- d) Acardius Amorphous--- no recognizable human form

The reverse flow in TRAP sequence means the acardiac twin receives blood through umbilical artery from the pump twin and returns this blood not to placenta but to the pump twin circulation through umbilical vein directly [1, 2].

With advances in foetal medicine many treatment options are available with minimally invasive techniques. These are intrauterine ablation of anastomosis by laser/unipolar or bipolar diathermy, ligation of vascular link, injection of alcohol in the linking vessels, radio frequency ablation of anastomosis, cord coagulation, embolization and intrafetal laser therapy [10]. These aim at improving survival of pump twin.

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But limited availability of centres for these techniques and also skills makes it difficult to undertake these procedures. On this backdrop, the best strategy remains always early diagnosis and early prophylactic intervention to avoid technical difficulties at late interventions. Early pickup of cases of TRAP sequence by sonography, will allow the clinician to choose appropriate method of foetal reduction and allow the normal growth of pump twin to be beyond 36 weeks without any compromise [1, 2, 10, 11].



Figure 3: Gross photograph showing Twin A grossly normal foetus and Twin B replaced by amorphous structure with double vessels cord of Twin B attached directly to placenta and anastomose with arterial vessels of Twin A

Conclusion

Thorough serial vigilant sonography and Doppler evaluation is the “mainstay” in early detection of TRAP sequence. Single yolk sac with twin foetal poles with absent cardiac activity in one with very thin i.e. <2 mm thick interfoetal membrane with acardiac foetus showing competitive growth with normal twin serially, should raise an alarm for this condition. In the present case, with early diagnosis of TRAP sequence, patient was able to decide the fate of pregnancy quite early. This helped the family to avoid financial burden and psychological sequelae too.

Author’s contribution:

Dr. Shweta Patel is the corresponding author who wrote the manuscript. **Dr. Manik Sirpurkar** had firstly saw the case and helped in preparation of manuscript. **Dr. Anjali Kanhere** helped in the preparation and proof reading of the case report.

Consent. Informed consent was obtained from patient prior to the preparation of the manuscript.

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