

Case report:

Rare case of acardiac Monster in TRAP (Twin Reversed Arterial Perfusion: MRI and USG findings

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Abstract

The twin reversed arterial perfusion (TRAP) sequence is a rare complication of 1% of monochorionic twin gestation with an incidence of 1 in 35,000 births. One twin is structurally normal while the other is abnormal with development of legs and lower body and undeveloped thorax, head and heart. The abnormal twin also referred as Acardiac monster twin receives its blood supply from normal or the pump twin and continues to develop putting the normal fetus at risk from high output cardiac failure. Mortality of the pump twin is 50-75% usually due to the result of heart failure and mortality of the acardiac monster twin is 100%. Our patient was Primigravida who was 29 weeks pregnant with increased uterine height. Ultrasonography and color Doppler study revealed monoamniotic monochorionic twin gestation with one structurally normal fetus and other acardiac cephalic fetus with TRAP sequence. MRI confirmed the diagnosis. The patient was referred to higher centre. Early diagnosis and proper management is vital to save the normal pump fetus

Keywords: Twin Reversed Arterial Perfusion (TRAP), Acardiac monster Twin, Pump twin

Introduction

Twin-reversed arterial perfusion / TRAP sequence is a rare complication seen in 1% of monochorionic twin gestation and has an incidence of 1 in 35,000 births.¹

¹ One of the twin is structurally normal. The other twin is abnormal with undeveloped heart (acardiac), head (acephalic), thorax and upper body and is parasitic to normal or pump twin and receives all its blood supply from it through vascular connections on the surface of placenta. This puts normal pump twin at risk of high output cardiac failure with a mortality rate of 50%^[2]. This article highlights the importance of early diagnosis for proper management and timely intervention to save the normal twin.

Case Report

A 27-year-old Primigravida, with 29 weeks amenorrhea reported for the first time in OPD for

check up. She had relaxed uterus with increased uterine height of 36 weeks and was referred for an ultrasonography scan. The study revealed a monochorionic monoamniotic twin gestation. Normal viable fetus (pump twin) is seen in breech presentation of maturity approx. 28 weeks. No definite anomaly noted in this fetus. Three vessel umbilical cord in pump twin is noted. Mild IUGR was observed. Fetal weight was 1.2 kg.

The second fetus noted as a Monster Parasitic Twin is seen as a malformed incompletely formed fetus with well-formed lower torso, lower limbs, kidneys and malformed thorax with acardia and malformed head with absent upper limbs. Massive diffuse soft tissue edema (5.9 cm) was seen around entire fetus. Double vessel with single umbilical artery is noted in the cord. On color Doppler imaging, the umbilical artery

in the abnormal fetus showed reversal of flow on the spectral. The longest length of Acardiac twin was measured in MRI sequence (19.7 cm) . The formula used for calculating weight in grams of cardiac twin: $(1.2 \times \text{longest length}^2) - (1.7 \times \text{longest length})$. The weight of monster Acardiac twin was 410 gms.

Placenta is single and Seen in upper uterine segment in right anterior wall. Two separate umbilical cords were seen arising from the placenta with vascular anastomosis. Liquor appears adequate. Internal os is closed.

Diagnosis was made of Twin with TRAP sequence of acardiacephalic type.

MRI was performed which confirmed the findings.

The patient was referred to higher specialized centre for management. There attempt was made to embolise the cord of second malformed fetus. The procedure failed and the patient went into preterm labor within few hours. The patient had Normal Delivery and delivered two male fetus. One normal alive fetus weighing 1.4 Kg. Second malformed fetus measuring 20x16 cm weighing 2.2 Kg. Normal infant was shifted to neonatal intensive care unit where it progressed well.

Discussion

Twin reverse arterial perfusion syndrome (TRAP) is a type of twin to twin transfusion which occurs in monochromic gestation. There are two accepted theories about the pathogenesis of the TRAP sequence. The popular theory suggests that perfusion of the recipient twin occurs by reversal of flow through the umbilical vessels of the normal pump twin mainly through vascular anastomosis in the placenta. This blood entering the recipient twin is under reduced oxygen tension. The desaturated blood that enters the body of the affected twin allows some development of lower body and lower extremities but

by the time the blood reaches the upper half of the body the oxygen saturation is too low causing disruption of organ morphogenesis and poor development of heart, head and upper torso^[3] The second theory suggests that there is primary failure in early organogenesis with cardiac dysmorphogenesis^[3]

Twin reversed arterial perfusion (TRAP) sequence is classified according to the degree development of the parasitic twin.^[4]

1. A cardiusacephalus, where head & upper extremities are not developed. It is most common variety and is seen in present case.
2. Acardiusanceps where brain and some cranial structure brain tissue is present. This is considered as most developed type.
3. Acardiusacormus is the rarest form. It has cephalic structure but no truncal structures are present.
4. Acardiusamorphous is minimally developed fetus with no distinguishable structure. Only the presence of umbilical cord differentiates it from Teratomas.^[5]

On ultrasonography the present case showed a monochromic, monoamniotic single placenta. One of the twin (pump twin) was structurally normal and the other twin was incompletely developed with well formed abdomen, lower limbs, kidneys and malformed thorax with no heart and the absence of identifiable cardiac pulsation. Fetal head could not be identified with absent upper limbs. This twin (recipient twin) was Acardiac cephalic type. The parasitic or recipient twin had gross edema of the skin. The presentation varied as there was no polyhydramnios. The pump twin was complicated by mild intrauterine growth retardation. All the abnormalities in the parasitic twin were well demonstrated on Ultrasonography and Color

Doppler.MRI also gave over view of the normal structural anatomy of pump twin and recipient twin .This confirmed all the findings observed on USG. In the TRAP sequence, the normal twin ‘pumps’ or ‘donates’ blood to the abnormal recipient twin, through abnormal artery-to-artery or venous-to-venous communications in the placenta^[6] There is a reversal of flow in the recipient twin, from the abnormal anastomosis to the umbilical artery. This finding can be confirmed by pulsed Doppler of the umbilical artery of the recipient twin, which revealed reversal of flow on the spectral waveform. In 75% of cases, the umbilical cord of the recipient twin contains a single umbilical artery^[5].In our case the recipient twin had single umbilical artery which was well demonstrated by color Doppler.MRI also shows two vessels in the cord of the parasitic twin. The difference between the pump twin and the acardiac twin resistance index (RI) of morethan 0.2 is associated with a good prognosis^[7].In our case we were not able to demonstrate it.Sherer used Doppler velocimetry of the umbilical cord in the TRAP syndrome and reported a markedly abnormal peak systolic to end diastolic velocity (S/D) ratio^[8]. Our case, failed to demonstrate it.

Other main complication like hydramnios and congestive heart failure in the normal twin were not present in our case. Preterm delivery is another complication, in our case preterm delivery was due to the intervention. Increasing perinatal morbidity and mortality has been associated with the relative size of the affected twin. Moore assessed the perinatal outcome by correlating its Twin Weight ratio.^[9] He predicted that if theacardiac recipient-to-pump twin weight ratio is more than 0.5,the adverse pregnancy outcome is 64% . If this weight ratio is greater than 0.7, the adverse pregnancy outcome is 90%This

evaluation is helpful in deciding the management of the case.In our case the Twin weight ratio was 0.3.This weight ratio predicts an favourable outcome. There were no complications in pump twin and the patient was referred higher hospital well equipped with proper neonatal facilities for management ..

The 3D sonography is a better modality to evaluate and assess both the twins and also estimate the body volume and size of parasitic twin^[10].MRI is also helpful in selected cases^[11].In our institute 3D ultrasonography was not available hence limited sequence MRI was done.MRI gave us the overview of both healthy and Acardiac monster. It was also helpful in measuring the longest length of Acardiac twin which was used to calculate Twin weight ratio.

Treatment:

Depending upon the gestation age,and the assessment of the pump twin the treatment options range from conservative, minimally invasive and invasive . Different techniques are employed which destructs theanastomotic connection between the acardiac twin and the pump twin . Ligation, embolization, laser photocoagulation, diathermy are being used for cord occlusion .Ablation of Acardiac fetus is being done by alcohol injection, , interstitial laser,monopolar diathermy and radiofrequency ablation^[12] or by selective delivery of acardiac twin .In present case the patient was referred to higher specialized centre for management as facilities for intervention was not available in our hospital.There an attempt was made for cord occlusion which was not successful and the patient went into preterm labour within few hours and delivered normally.The normal twin weighed 1.4 kg and was immediately kept in specialized neonatal care where the infant made good progress.

Conclusion

Dedicated ultrasonography with color Doppler complemented by 3D ultrasound in a monochorionic monoamniotic twin pregnancy is essential for antenatal diagnosis of Twin-Reversed Arterial Perfusion (TRAP) sequence. MRI may also be done for overview of both parasitic and normal pump twin. MRI is helpful in measuring the longest

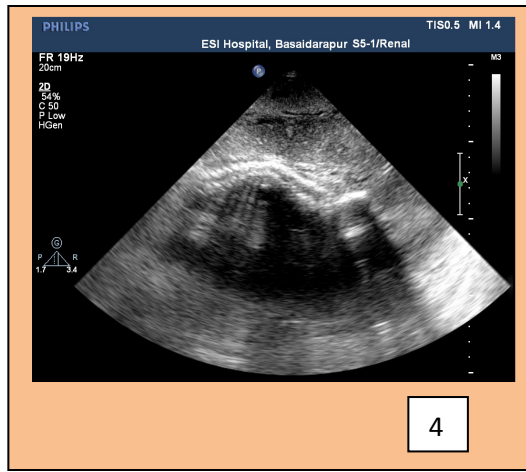
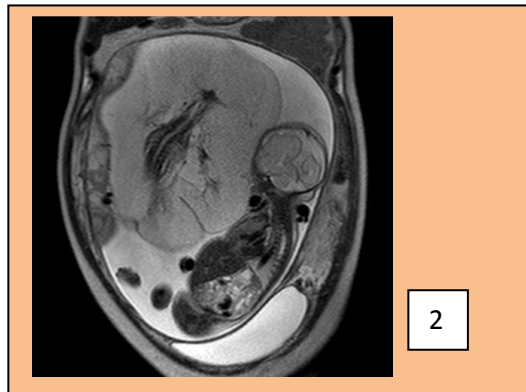
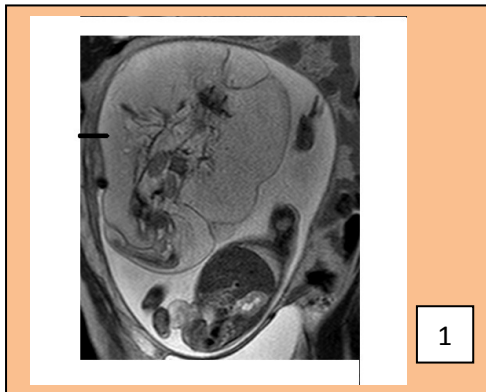
length of Acardiac twin to calculate Twin weight ratio. Proper management and timely intervention in a specialized centre with good neonatal facilities will help in survival of normal pump twin. It is recommended that serial ultrasonography evaluation and minimally invasive techniques may be used for good prognosis.

Fig 1) T2W MRI showing monochorionic monoamniotic twins with parasitic Acardiac twin showing well-formed lower limb and lower torso.

Fig 2) T2W MRI showing acardiaacephalus with massive soft tissue edema in parasitic twin and well formed pump twin.

Fig 3) T2W MRI showing double vessel umbilical cord with single umbilical artery.

Fig 4) USG showing heteroechotexture lesion with spine of Acardiac twin



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