

Acardius Acephalus - Twin Reversed Arterial Perfusion Sequence: A Rare complication of Monochorionic Multiple pregnancy

¹Dr Abhishek Mishra, ²Dr Shrestha Aggarwal, ³Dr Birma Ram Hudda

^{1,3}Radiologist, ²Feto-maternal Specialist
^{1,3}AFMS, ²SFMC Bathinda

Abstract: Seen with Monochorionic twin pregnancy, Twin Reversed arterial Perfusion sequence is a very rare condition often considered to have a poor prognosis. The usual incidence is one in 35000 births. Typical findings are of a foetus that continues to grow despite an absent or a rudimentary heart. Often such a foetus has lethal anomalies in association. The acceptable causes of mortality in the Pump twin is due to Premature labour or cardiac failure induced by Polyhydramnios or due to Hypoxia induced IUGR as a result of aberrant vascular anastomosis. Herein, we are reporting a case of complicated Dichorionic-Triamniotic Intrauterine pregnancy presented with this syndrome emphasizing the need for an early diagnosis to institute early corrective measures by using minimally invasive techniques.

Keywords: Twin Reversed Arterial Perfusion, Monochorionic, Acardius

Introduction

Seen in about 1 in 100 Monochorionic twin pregnancies, this syndrome is one of the rarest complication of Multifetal pregnancy that often has a poor prognosis and carries significant antenatal and perinatal mortality. It is characterised by an Acardiac foetus also known as Acardiac Parabolic twin that despite not having a functional heart, continues to grow. This Acardiac foetus often has lethal anomalies in association. There is a pump foetus as well that feeds this Acardiac foetus through various vascular anastomosis through placenta. The pump twin is anatomically normal. The usual incidence of this syndrome is about 1 in 35000 births ⁽¹⁾. The acardiac twin has 100 % mortality whereas the perinatal mortality in pump foetus ranges from 35 to 55% ⁽²⁾. The perinatal mortality in Pump twin is usually because of Premature labour or cardiac failure induced by Polyhydramnios or due to Hypoxia induced IUGR as a result of aberrant vascular anastomosis. Herein we are reporting USG and Color Doppler findings in a case of Complicated Dichorionic-Triamniotic Intrauterine pregnancy presented with this syndrome thus there by emphasizing the need for an early diagnosis to institute early corrective measures.

Case Report

A 25 year old Primigravida of Asian ethnicity and spontaneous conception referred to the Department of Radio-diagnosis of Military Hospital Bathinda at 21 weeks 04 days of gestation (as per LMP) for a targeted anomaly scan, subsequently the patient was reviewed by foetal medicine specialist at Sanjeevni Fetal Medicine Centre, Goniana Road, Bhatinda, (Punjab, India). On detailed history she didn't reveal any personal or family history of a significant medical comorbidity or a congenital condition. She was a non-smoker and a non-alcoholic. On a detailed evaluation, pregnancy was confirmed on ultrasound as Intrauterine Dichorionic-Triamniotic triplet pregnancy with triplets A and C as a Monochorionic pair. With this monochorionic-diamniotic pair, placenta was seen posteriorly in upper segment with central insertion of umbilical cord.

Twin A was localized on maternal right where as Twin C was seen lying over the internal Os. Twin A showed cephalic presentation, spontaneous foetal movements and a rhythmic cardiac activity with a foetal heart rate of 136bpm. On detailed evaluation, no gross congenital anomaly was seen in Twin A, however amniotic fluid volume was excessive with largest vertical pocket measuring about 14cm suggesting polyhydramnios (Fig- 1). No features of hydrops were seen at the time of scan. The biometric analysis by USG suggested gestational age to be 20 weeks 04 days.

Twin C of this monochorionic pair appeared to be severely malformed on USG with poorly formed cranial structures and short malformed spine. The brain parenchyma appeared dysgenetic with features suggesting Holoprosencephaly (Fig-2a). There is associated cystic swelling in soft tissues of rudimentary neck (Fig-2b). The foetal thorax appeared rudimentary and couldn't be clearly delineated from abdominal structures. Foetal cardia was and upper extremities were appeared rudimentary (Fig -3a). The lower extremities were well formed and showed spontaneous movements despite not having a functional cardia. Thus, Twin C was coined an Acardiac twin (Acardius acephalus type). Umbilical cord showed central insertion in anterior abdominal wall with umbilical arteries showing reversed perfusion (Fig-3b). Twin B didn't show any obvious congenital anomaly and showed normal foetal movements and cardiac activity with a foetal heart rate of 140bpm.

Discussion

Twin Reversed Arterial Perfusion sequence is a very rare complication of Monozygotic Monochorionic pregnancy. Two theories are proposed to explain the pathogenetic mechanisms of this syndrome. The widely acceptable theory is that primarily an aberration of vascular anastomosis exists with a secondary developmental defect in foetal cardia ⁽³⁾. Another theory proposes that a primary abnormality in cardiac organogenesis predisposes to secondary alterations in foetal vasculature and flow patterns ⁽⁴⁾. Ultrasonic demonstration of Acardia or a rudimentary heart supports this theory.

Antenatal diagnosis can be made correctly on Ultrasound if a foetus in a monochorionic twin gestation continues to grow and shows spontaneous movements in absence of a functional heart. The pelvis and lower extremities are well-developed as compared to

cranial structures and upper extremities due to preferential perfusion. Demonstration of an arterial flow entering into Umbilical arteries of Acardiac fetus makes the sonological diagnosis certain.

Based on morphological features, these Acardiac foetuses can be broadly classified into three types ^(2,5).

1. Acardius Acephalus: most common type, with absent head and rudimentary thorax and upper limbs. Pelvis and lower extremities are well-developed. It constitutes about 60-75% of overall cases.
2. Acardius Anceps: this type constitutes about 20% of all cases in which head is partially formed and body and extremities are developed.
3. Acardius Acornus: only head of the fetus has developed. Its very rare and consist of about 10% of overall cases.
4. Acardius Amorphous: characterised by a poorly differentiated mass of tissue with no identifiable foetal organ.

Our case belongs to the first set of group.

Management

The perinatal mortality for Acardiac twin is 100%. The perinatal mortality of the pump twin varies between 35% to 55% ⁽⁶⁾ and depend on various factors like preterm delivery induced by polyhydramnios, cardiac failure or hydrops in pump twin induced by continuous shunting of blood to acardiac twin which increases the load on heart of pump twin. Another reason is hypoxia induced IUGR in pump twin due to recirculation of deoxygenated blood to pump twin through aberrant vascular anastomosis.

The management of TRAP sequence is aimed at salvage of pump twin through minimally invasive techniques like cord occlusion and intra-foetal ablation using laser coagulation aimed at discontinuation of the blood flow to the Acardiac twin.

Conclusion:

Twin Reversed Arterial Perfusion Sequence is a very rare complication of Monochorionic twin pregnancy and carries a poor prognosis. Early diagnosis with typical ultrasound and color Doppler findings helps in providing immediate treatment aimed at salvaging the pump twin.

References

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Figure 1: Polyhydramnios with Triplet A

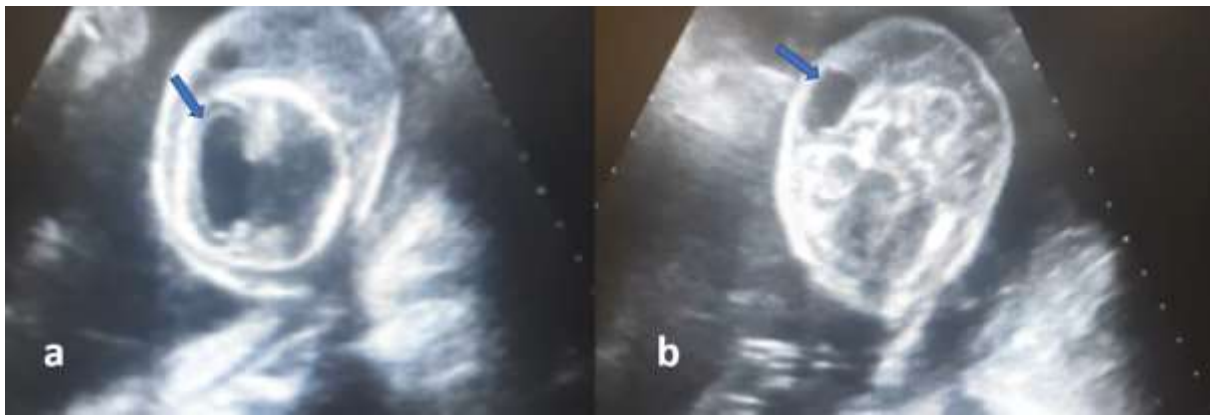


Figure 2: (a) Rudimentary cranial structures in Triplet C. (b) Soft tissue edema witha cystic swelling in the rudimentary foetal neck in triplet C.

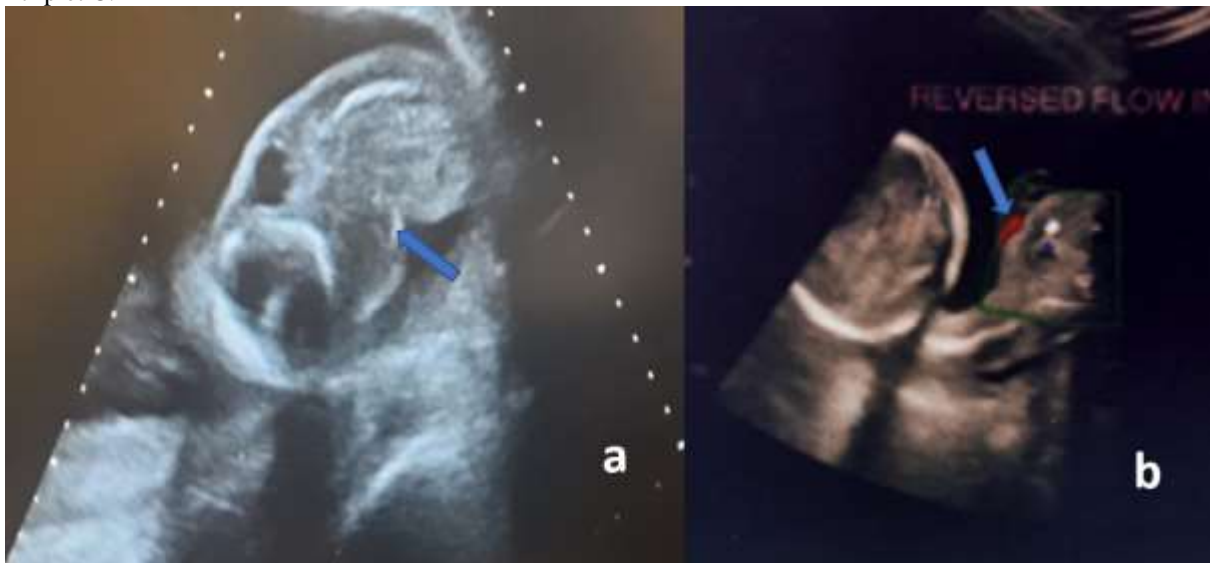


Figure 3: (a) Triplet C showing rudimentary thorax and spine with non-visualization of foetal cardia and upper limbs. (b) Doppler imaging showing reversed perfusion in the umbilical arteries of triplet C