CASE REPORT

Two Cases of Twin Reversal Arterial Perfusion Sequence

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Abstract

Aim: To describe two cases of late presentation of acardiac twin in monochorionic gestation, which was earlier diagnosed as a vanishing twin in first trimester scans and later on found to be an acardiac twin (TRAP sequelae) on follow-up scans.

Background: Monozygotic twinning is complicated by anomalous vascular connections in the placenta resulting in clinical syndromes such as twin-twin transfusion syndrome (TTTS), twin anemia-polycythemia sequence (TAPS), and twin reversed arterial perfusion sequence (TRAPS; acardiac twinning). Twin reversed arterial perfusion sequence is a condition arising due to paradoxical retrograde transfusion by a structurally normal "pump" twin, there is a disruption of organ development in the perfused twin.

Case description: Twenty-one-year-old G2A1 who was initially diagnosed with vanishing twin in the 14-week scan, which was later on found to be acardiac twinning at 29 weeks and presented with PPROM. She went into spontaneous labor and there was a dilemma regarding delivering her vaginally or by cesarean section. The second case of primigravida, initially diagnosed as single fetal demise at 12 weeks and found to have TRAP sequelae later on at 31 weeks, was admitted and underwent elective LSCS at 32 weeks on maternal request.

Conclusion: This lays down the importance of early diagnosis with dedicated ultrasound and management at tertiary care centers for better perinatal outcomes.

Clinical significance: The phenomenon of the "vanishing twin" refers to the occurrence of a single fetal loss in a twin gestation during the first trimester. Such cases seen in the first trimester may be a case of acardiac twins, which might become evident later in a growth scan. Hence, it is important to be alert to the possibility of missing an acardiac twin since it is associated with 50% mortality of the pump twin.

Keywords: Acardiac twin, Monochorionic, Perinatal, Twin reversed arterial perfusion, Vanishing twin.

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Background

Monozygotic twins arise from a single fertilized zygote that shares the same genetic heritage but is not completely identical. In monozygotic twinning, a group of fetal syndromes arises due to the anomalous vascular connections in the monochorionic placenta. These chronic feto-fetal connections may result in clinical syndromes such as TTTS, TAPS, and TRAPS; (acardiac twinning).[1]

Twin reversed arterial perfusion sequence was first documented in 1533; and detected by ultrasound in 1978. This is a condition arising due to paradoxical retrograde transfusion by a structurally normal "pump" twin, there is a disruption of organ development in the perfused twin.[1,2] Hence, the transfused twin receives blood from an artery (deoxygenated blood) instead of a vein giving rise to reversed perfusion which can be picked up through Doppler.

The blood reaches the acardiac twin through the umbilical vessels and preferentially goes to the iliac vessels. Thus, the lower body has a preferential growth pattern compared to the disrupted growth of the upper body. In these cases, the twin with a failed head growth is called acardiac acephalous; a partially developed head with identifiable limbs is called acardiac myelacephalus, and failure of any recognizable structure to form is called acardiac amorphous. Due to the anomalous connection, the donor twin is in a hyperdynamic state, which results in high output cardiac failure exposing it to increased perinatal risks.

Case Description

We report a case of 21-year-old G2A1, referred as a case of MCMA twin with a single live fetus at 30 weeks 5 days, in view of PPROM, in February 2021 to our institution.

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She conceived spontaneously, was having regular antenatal care from the local hospital, and was detected to have MCMA twin gestation with a single live fetus at 14 weeks and miscarriage of second twin (vanishing twin). On TIFFA at 20 weeks 1 day, it was detected that the second twin was acardiac. She continued antenatal care at the local hospital and a growth scan at 29 weeks 3 days suggested an acardiac twin larger than the first twin (1.37 kg). She developed PPROM at 30 weeks 5 days and was referred here.

On admission, her general examination was normal and the uterus was corresponding to 34 weeks with PPROM. She was started on prophylactic antibiotics and corticosteroids for fetal lung maturity. A Follow-up scan at 31 weeks and 1 day showed the first twin with a weight of 1.98 kg, with no Doppler abnormalities and the second twin was acardiac, larger than the first. It was decided to give magnesium sulfate for neuroprotection and do elective LSCS at 32 weeks. However, she went into spontaneous labor at

31 weeks and 5 days and reported to the labor room with a cervical dilatation of 8 cm. Initially, there was a dilemma regarding keeping her for vaginal delivery as the first twin was cephalic and the head was at 0 station. But it was decided for emergency LSCS, anticipating difficulty in delivery of the larger second acardiac amorphous twin.

LSCS was done and delivered a first twin of weight 1.475 kg as breech and the amorphous second twin had a partially developed head with identifiable limbs and external genitalia; acardiac myelacephalus of weight 2.8 kg (Fig. 1). The placenta was MCMA type with the anomalous vascular connection between donor and pump twin. The first twin was admitted to neonatal ICU (NICU) and monitored for signs and symptoms of heart failure and discharged from the NICU on day 9 in good health.

The second case was a 24-year-old primigravida, referred to as a case of TRAPS complicating monochorionic twin gestation at 31 weeks 6 days to our institution in July 2021.

She was on regular ANC; was detected to have a single live fetus and a second non-viable twin in an NT scan done at 12 weeks. Later TIFFA showed growth in one fetus with the second fetus having crowded parts and no cardiac activity. When her growth scan was done at 31 weeks and 5 days; it showed mild polyhydramnios of the pump twin and second twin as a mass with no definite cardiac activity and minimal blood flow seen in the abdominal region suggestive of acardiac twin pregnancy (TRAPS) and was referred here.

On admission her general examination was normal, uterus was 34 weeks in size. A repeat ultrasonogram from our institution at 32 weeks showed a normal twin with a growth of 31 weeks and weight of 1836 gm, normal Doppler, and an amorphous mass with fetal skeleton and extensive subcutaneous edema; approximately 2 kg. There were vessels noted entering into acardiac twin with umbilical artery waveform suggestive of TRAPS. LSCS was planned for 34 weeks. However, on maternal request, she underwent LSCS and delivered the first twin male baby of weight 2.3 kg as breech and a second twin; acardiac acephalous of weight 2.4 kg (Fig. 2). The placenta was MCDA type. The first twin was admitted to NICU and discharged on POD-8, in good health.

Discussion

Twin reversed arterial perfusion sequence affects 1 in 35,000 pregnancies.[1] The acardiac twin gets partially transfused from the normal twin through the artery-to-artery or vein-to-vein anastomosis existing in the shared placenta. TRAP sequences might be seen associated with a single umbilical artery of acardiac twin in two-thirds of the cases or with chromosomal anomalies of acardiac twin in the other one-third of cases.

A high amount of suspicion should arise in the absence of identifiable cardiac motion, disrupted development of lower extremities, and conspicuous subcutaneous edema in one twin.[2] Color Doppler will confirm the diagnosis with reversal of blood flow (deoxygenated blood through the artery) within the abnormal fetus.[2]

Pump twins of TRAP sequelae are at risk of cardiac failure, cerebral ischemic sequelae, preterm birth, and *in-utero* death. In more than half of the cases, there is the spontaneous cessation of blood flow to the acardiac twin; such sudden flow arrest was associated with subsequent death or neurological injury in 85% of the normal twin.[3] The mortality rate was 50% in pump twin, mainly due to prematurity or from a prolonged high output state leading to cardiac failure.

The size of the acardiac fetus is a major determinant in the case of many TRAP sequences. The larger the size of the acardiac twin, the worse the perinatal outcome of the pump twin. TRAP sequence with pump twin larger than acardiac twin can be managed conservatively. The size of the acardiac twin can be estimated as the volume of an ellipse; length * width * height * 3.14/6.[1] When the volume of the acardiac twin is more than 50% of the donor twin, early intervention should be planned to improve the outcome for the donor twin.

Clinical Significance

Twin reversed arterial perfusion sequence can be confused with the phenomenon of "vanishing twin" which is seen in 21.2% of all multiple gestations, but has no detrimental impact on the surviving fetus as compared to TRAP which is associated with 50% mortality of the pump twin.[4]

Conclusion

With the present era of good neonatal care and better radiological aids, the survival chance of pump twins has increased with meticulous planning of antenatal follow-up, need for radio-frequency ablation, and plan of delivery. In cases like this, if we put emphasis on first and second-trimester screening similar to

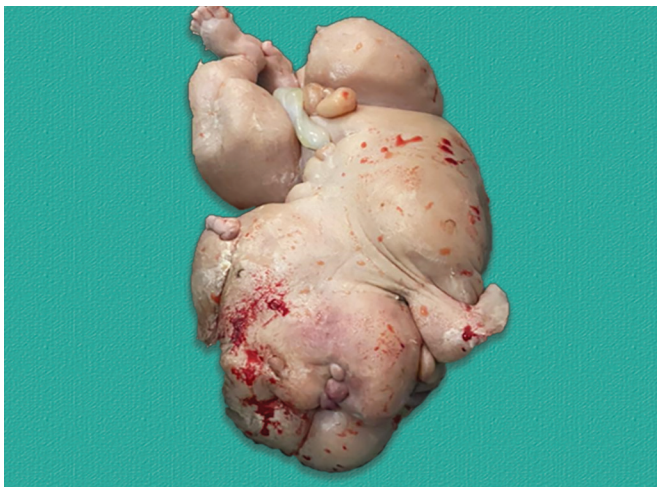

Fig. 1: Acardiac second twin with developed lower limb and underdeveloped upper part: Acardiac myelacephalus

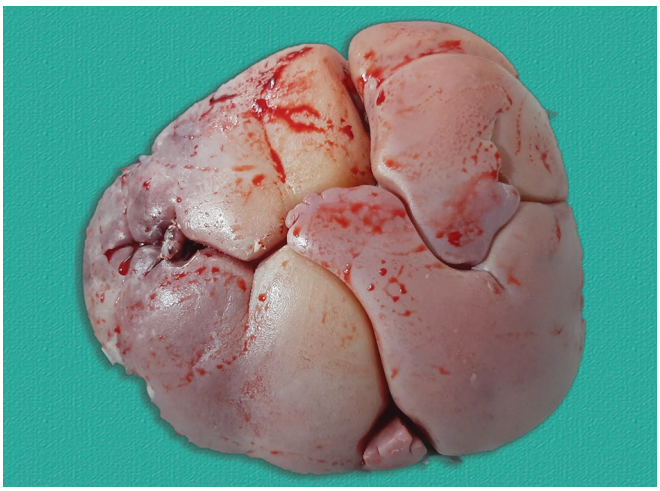

Fig. 2: Acardiac second twin with failed head growth: Acardiac acephalous

the model of an inverted pyramid of antenatal care, it can lead to early diagnosis and timely management for a better perinatal outcome.

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