Case Report

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Management of Acardiac Twins: Does Conservative Approach Deserves Consideration? A Case Report

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Abstract

Acardiac twinning, a rare congenital anomaly of monozygotic twin pregnancies, often leading to abnormal placental vascular anastomosis. This in turn results in twin reversed arterial perfusion (TRAP) with complex pathophysiology. Current information on early diagnosis and treatment for the salvation of the pump twin is based mainly on various individual case reports. Here, we report a case of monozygotic twin complicated with TRAP sequence, which was diagnosed early, managed conservatively and the outcome was excellent. With the availability of various sophisticated studies for antepartum fetal surveillance, outcomes in expectantly managed cases are getting better. So, in acardiac twins aggressive interventions should be used cautiously and should be reserved only for some really indicated cases.

Key words: Acardiac twin, Conservative management, Monochorionic twins, Twin pregnancy, Twin reversed arterial perfusion sequence

INTRODUCTION

Acardiac malformation also known as twin reversed arterial perfusion (TRAP) sequence, is a unique complication of monochorionic pregnancies. It is characterized by paradoxical retrograde perfusion of abnormal twin by structurally normal pump twin through a single artery-artery anastomosis.[1] It is a very rare condition. The reported incidence being 1 in 35,000 deliveries and 1 in 100 monozygotic twins.

Optimal management of acardiac twin pregnancies is controversial. The reported fetal/neonatal mortality of the pump twin is extremely high (50-60%). It is thought primarily to be due to increasing in cardiac demands of the pump twin in an effort to perfuse its acardiac sibling. Data suggest 50% mortality rate in pump twin with expectant management.

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CASE REPORT

A 25-year-old G2P1L1 with previous cesarean section done referred to our tertiary care center at 10 weeks of gestation as a case of monochorionic and monoamniotic (MCMA) twins with single fetal demise. She had excellent dates. Her family history and past history were absolutely unremarkable for twins or perinatal issues. All routine investigations were done and found to be within normal limits. Since then the patient had regular antenatal check-ups from our institute. During routine antenatal care at 20 weeks of gestation, the abdomen was enlarged to around 24 weeks. We got an ultrasound (USG) scan done. To our surprise, USG revealed a twin pregnancy with a single placental mass, and there was no dividing membrane (MCMA). There was appropriate for dates twin without any obvious anomalies, which was associated with an acardiac, acranial amorphous twin (Figure 1). At that time size of the cardiac twin was almost same as the pump twin. But, pump twin showed no signs of decompensation. We discussed in detail with parents about the condition, and we also counseled them about the risks and benefits of the expectant management and surgical intervention.

Management and Outcome

Pregnancy was followed up carefully with serial USG studies, Doppler flow studies, non-stress test (NST),

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biophysical profile (BPP), and fetal echocardiography. USG with Doppler study was done once in 2 weeks. Echocardiography was done at 24, 28, and 32 weeks of gestation. We looked closely for the presence of any signs of decompensation like pericardial or pleural effusion, or ascites. Serial USG scan revealed that acardiac twin was constantly increasing in size. USG at 32 weeks of gestation showed the acardius which had increased in size relative to the pump twin, who had maintained an appropriate weight for gestational age with an estimated fetal weight of 1.960 kg. But, pump twin showed no signs of decompensation. The patient was admitted at 30 weeks of gestation. Alternate day NST was done, which was found to be reactive. Two doses of betamethasone were given 24 h apart for fetal lung maturity.

At 34 weeks of gestation, the patient presented with preterm premature rupture of membranes. Emergency lower segment Cesarean section was done. Pump twin weighed 1.9 kg with APGAR 1'9. Acardiac twin weighed 2.9 kg, its head, and upper extremities were absent. It had well-developed lower limbs (Figure 2). Pump twin was apparently alright, and it showed no apparent anomalies (Figure 3). The baby was kept in NICU for 12 h for observation. As there were no signs of failure, it was managed on mother side only.

Postnatally pump twin was healthy. Both mother and baby were discharged on day 6.

DISCUSSION

Acardiac twinning or TRAP sequence is a rare congenital anomaly of monozygotic multiple pregnancy. This occurs secondary to abnormal placental anastomosis which is characterized by formation of a malformed fetus with an absent or rudimentary heart (acardius) and other structures.[1]

The pathogenesis in TRAP sequence is, there will be a defect in embryogenesis in one of the twin leads to failure of cardiac development. The normal twin then perfuses the acardiac twin via placental artery-artery anastomoses.[2] It should be noted that the cardiac anomaly is not secondary to anastomoses, but they are established as a result of it.[3,4]

Within the placenta, which is singe and shared by both the twins, arterial perfusion pressure of the donor twin exceeds that of the acardiac twin who thus receives reverse blood flow of deoxygenated arterial blood. This used blood reaches the recipient twin through its umbilical arteries, and it preferentially goes to iliac blood vessels. Thus, only the lower extremity is perfused and disrupted growth and development of the upper extremity results as occurred in the present case.[5]

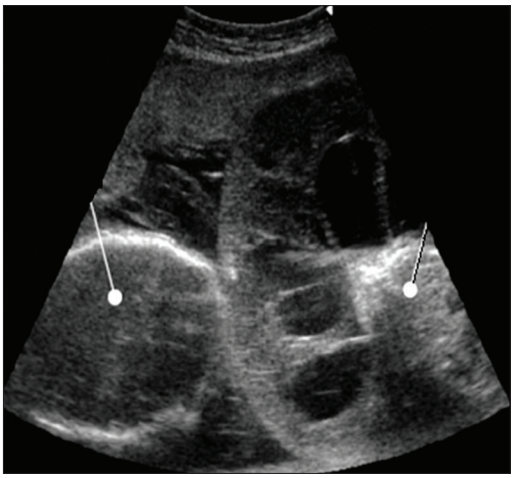

Figure 1: Ultrasound taken at 20 weeks of gestation showing pump twin and acardiac twin.

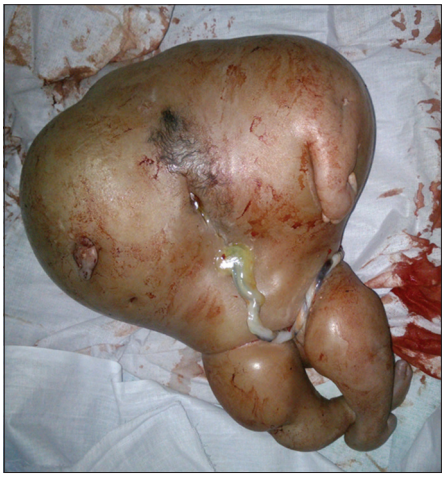

Figure 2: Acardiac twin

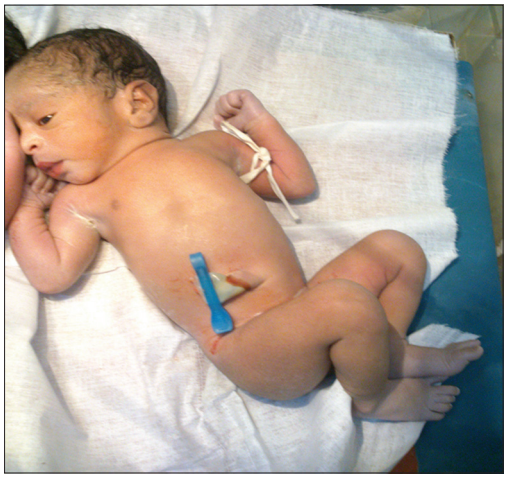

Figure 3: Pump twin

The main differential diagnosis of acardiac twin is the intrauterine death of one of the monochorionic twin, which

will be anomalous. (In the present case, it was misdiagnosed in 10 weeks USG as MCMA twin with single fetal demise). However, the continued growth of the "presumed dead" twin on subsequent scans is often the first clue to the correct diagnosis. The color flow imaging plays a major role in establishing the diagnosis of acardiac twin. This shows the presence of blood flow within the abnormal fetus. This may be evident even during the first trimester.[6]

When blood flow pattern is examined in detail in these cases, it reveals a paradoxical direction of arterial blood flow toward acardiac twin. Lately, three-dimensional USG has also been used for confirmation of diagnosis and also to establish the extent of fetal malformation.[7]

It is clear that TRAP sequence poses substantial risks for the pump twin. Ideally, a diagnostic test would allow us to distinguish between cases that can be safely managed expectantly and those that require intervention. USG with Doppler studies, BPP, NST, and fetal echocardiography play a significant role in deciding the course of treatment.

Even though the pathophysiology of acardiac twin has been clearly resolved and treatment by ablating this vascular connection has been proposed, the availability and difficulty of this mode of treatment and fetal death secondary to the treatment itself have been clearly demonstrated. Invasive treatment should be restricted, only to those pregnancies which would benefit from prenatal intervention like those where the donor twin is at a significant risk of prematurity, cardiac failure or death and should be considered in presence of poor prognostic factors such as polyhydramnios, ascites, large acardiac twin, and rapid growth or evidence of substantial blood flow through the umbilical vessel supplying the acardiac twin.[8]

Sullivan *et al*. advocated expectant management in all cases complicated with TRAP sequence.[9] 90% survival in pump twin were reported in 10 pregnancies with an acardiac twin managed expectantly. They recommended expectant management with close fetal surveillance in all cases, and aggressive interventions should be used cautiously.

The best time to intervene and best mode of intervention are not yet known. With an increase in antenatal diagnosis and with the availability of various sophisticated studies for antepartum fetal surveillance, outcomes in expectantly managed cases are getting better, and they are better than reported.

In the presented case, even though pump twin weighed only 3/4th the weight of acardiac twin it showed no signs of decompensation. The outcome, in this case, was excellent.

CONCLUSION

The most appropriate interventions and management protocols for the various clinical presentations of TRAP Sequence is as yet to be determined, and conservative non-intervention is often appropriate in some carefully selected cases. This case report demonstrates that in TRAP sequence, early diagnosis, close antenatal follow-up can prevent unnecessary interventions without compromising the outcome. Long-term follow-up data on surviving pump twin are lacking. It is anticipated that centers with active study protocols for these conditions will best serve patient care and clinical research needs.

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