CASE IN IMAGES OPEN ACCESS

Acardiac twin: Conservative management

Deepa V Kanagal, Mangala Srikantaswamy, Rashmi M Devaraj, Maitri M Kulkarni, Mahendra Gangadhariah, Lokeshchandra C Hanumanalu

ABSTRACT

Introduction: Acardiac twin is a rare anomaly occurring in 1 in 35,000 pregnancies. The outcome is fatal for the acardiac twin with a 50–75% mortality of the normal pump twin. Management options include conservative management, medical therapy, occlusion of connecting vessels and selective termination of the acardiac twin. The Management of a twin pregnancy complicated by an acardiac twin is a challenge because the continuous growth of the acardiac fetus is deleterious to the healthy pump twin leading to cardiac insufficiency, polyhydramnios, prematurity and even death of the normal twin. Case Report: We report three cases of acardiac twinning which presented with different manifestations, acardius acephalus, acardius myelocephalus and acardius amorphous. All the three cases were managed conservatively with good outcome of the pump twin in all the cases. We present the cases with review of literature. Conclusion: Conservative non-intervention may be appropriate in developing countries with low resources.

Keywords: Acardiac twins, Twin pregnancy, Conservative management

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INTRODUCTION

Acardiac twinning or Twin Reversed Arterial Perfusion (TRAP) is a rare anomaly occurring in monozygotic multiple pregnancies with an incidence of 1% and in 1 in 35,000 pregnancies [1]. The mortality in recipient acardiac twin is 100% whereas the normal donor twin is also at an increased risk of morbidity and mortality. This is a report of three cases of acardiac twins with different presentation. One was acardius acephalus, another was acardius myelocephalus and the third one was acardius amorphous. All cases resulted in live birth of the normal twin.

CASE REPORT

Case 1: A 30-year-old third gravida with previous two uneventful vaginal deliveries came to us for the first time at 34 weeks of gestation with twin pregnancy in active labor. She delivered a normal female baby of

1.7 kg followed by a malformed, dead acardiac fetus of 900 g (Figure 1). The acardiac fetus had a grossly malformed upper body without upper limbs head and neck (acardius acephalus) (Figure 2A). The trunk was partially developed. The lower limbs were developed but had bilateral congenital Talipes equinovarus deformity and syndactyly (Figure 2B). External genitalia resembled female (Figure 2C). There was a single placenta with two cords , one small and thin belonging to the acardiac fetus. (Figure 3). Autopsy showed the absence of neural tissue, (Figure 4) absence of lungs, incomplete diaphragm,

Figure 1: Normal fetus and acardiac (Case 1).

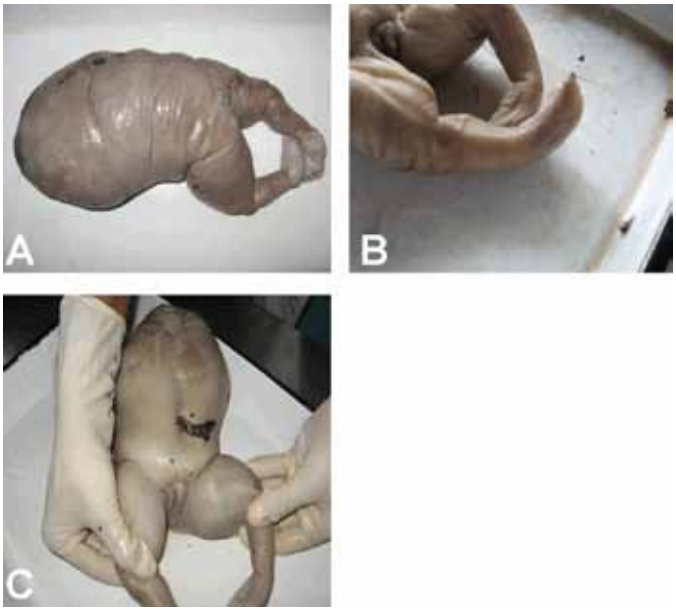

Figure 2: (A) Acardius acephalus (Case 1), (B) Bilateral Congenital Talipes equinovarus (Case 1), (C) Female Genitalia (Case 1).

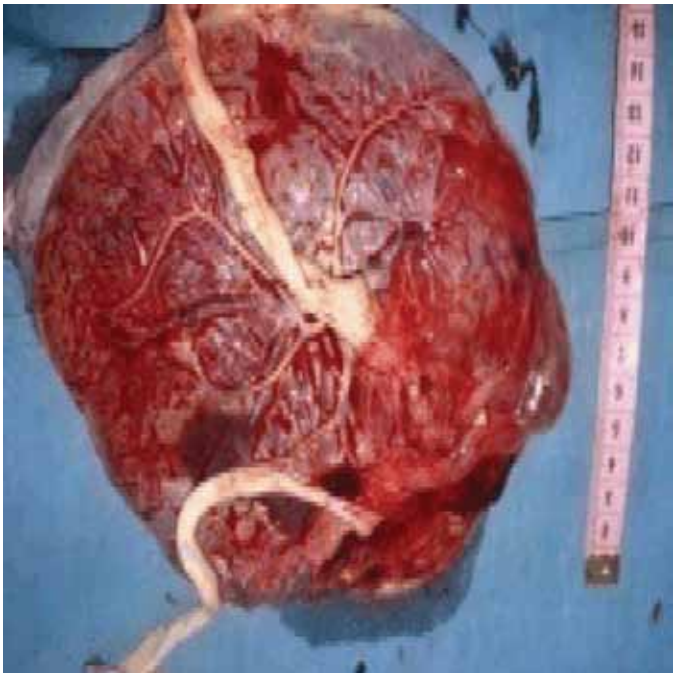

Figure 3: Single placenta with 2 cords (Case 1).

Figure 4: Absent neural tissue (Case 1).

rudimentary cardia, liver tissue, uterus, fallopian tube, ovary, kidney and ureters. (Figure 5). The normal twin was fine at the time of discharge three days later.

Case 2: A 26-year-old second gravida with previous normal delivery came with 24 weeks pregnancy. On examination, the uterus was of 28 weeks size. Ultrasound revealed twin pregnancy with one normally formed fetus of 24 weeks and another fetus with a partially formed head with hydrocephalus and enlarged lateral ventricle (Figure 6A–B). The spine was partially seen and the lower limbs

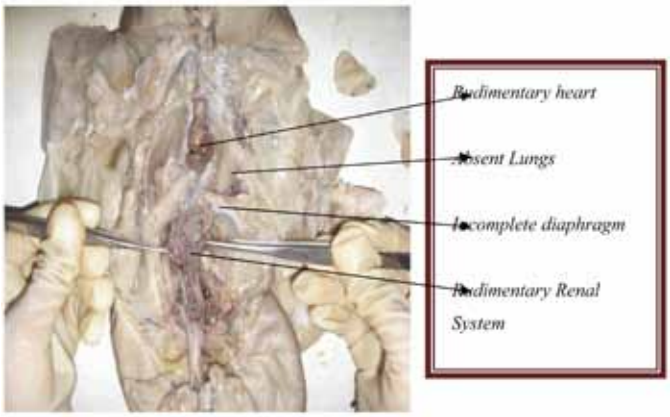

Figure 5: Thoraco-abdominal autopsy (Case 1).

Figure 6: (A) Hydrocephalus (Case 2). (B) Enlarged lateral ventricle (Case 2).

were recognizable. No other anatomical structure was made out. Gross abdominal edema and ascites was noted (Figure 7). The placenta was a single large mass. Color Doppler of umbilical artery of the abnormal fetus showed reversal of flow thus confirming the diagnosis of acardiac pregnancy. The parents were counseled regarding the prognosis of the surviving twin, options of conservative and invasive management and non availability of invasive management in our center. After obtaining their consent, the lady was managed conservatively with regular follow-up and serial ultrasound examination. At 36th week of gestation, she developed polyhydramnios and pre-eclampsia and an emergency cesarean section was done. A borderline term male fetus of 2 kg weight with normal morphology was delivered by breech extraction. The other twin was acardiac weighing 750 grams with a partially developed head and face. Lower limbs could be made out. (acardius myelocephalus) (Figure 8A–B). External genitalia were poorly developed. Autopsy was not done for the acardiac fetus. The normal twin fared well and was discharged on seventh postoperative day.

Case 3: A second gravida of 21 years of age with a previous cesarean section presented at 24th week of gestation with twin pregnancy. Ultrasonography showed the first twin to be an acardiac amorphous mass without any recognizable structures. Only the spine was made out throughout its length (Figure 9). The second twin was normally developed. There was a single placenta (Figure 10). After explaining about the treatment options, the lady opted for conservative management in view of the cost and non-availability of facilities for invasive management at our hospital. She was followed-

Figure 7: Abdomen of fetuses, ascites in acardiac twin and common placenta (Case 2).

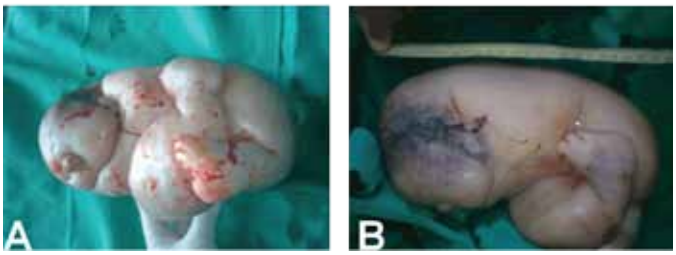

Figure 8: (A) Acardius myelocephalus (Case 2). (B) Partially developed head (Case 2).

Figure 9: Acardius amorphous with spine (Case 3).

up by serial ultrasound examinations. The growth of the normal fetus was satisfactory without any complications. The patient had regular antenatal check ups and was treated for anemia. She went into labor at 36th week of gestation. An emergency cesarean section was done in view of previous cesarean section. The first twin was an amorphous soft, globular mass without any recognizable structure and was weighing 700 g (acardius amorphous) (Figure 11). The second twin was a normal female of weight 2.1 kg. The postoperative period was uneventful. The mother and the normal twin were discharged on the eighth postoperative day.

Figure 10: Both fetuses with single placenta (Case 3).

Figure 11: Acardius amorphous specimen (Case 3).

DISCUSSION

Multiple pregnancy accounts for 1.5% of all pregnancies with a perinatal morbidity and mortality of 10% [2]. Acardiac twinning or TRAP sequence is a rare congenital anomaly of monozygotic multiple pregnancy due to abnormal placental anastomosis characterized by formation of a malformed fetus with an absent or rudimentary heart (acardius) and other structures. There is usually a normally formed donor twin who may have features of heart failure. Acardiac fetuses were first described by Beneditti in 1533 [3]. Acardiac acephalic variety is the most common type of acardiac fetus.

It has been hypothesized that the TRAP sequence is caused by a large artery to artery placental shunt often accompanied by a vein to vein shunt. Within the single shared placenta, arterial perfusion pressure of the donor twin exceeds that of the recipient twin who thus receives reverse blood flow of deoxygenated arterial blood from its co-twin. This used blood reaches the recipient twin through its umbilical arteries and preferentially goes to its iliac vessels. Thus only the lower body is perfused and disrupted growth and development of the upper body results [4].

An acardiac twin should be suspected in all monochorionic, malformed fetuses with cystic hygroma, generalized edema and an absent cardiac pulsation with a non-functioning heart. Also, an ultrasonography finding of twins revealing discordant or grotesque malformation along with reverse flow in the umbilical artery is usually diagnostic of an acardiac twin [5]. This can be diagnosed in first trimester by vaginal scanning and color Doppler sonography. Serial ultrasonography is indicated to assess such twin pregnancies.

Failure of the head growth is called acardius acephalus, a partially developed head with identifiable limbs is acardius myelocephalus and failure of any recognizable structure to form is acardius amorphous [6]. Acardiac twins can also be classified as follows:

Acardius anceps – when head is poorly formed,
Acardius acephalus – if the head is absent
Acardius acormus – when only head is present.

Based on the development of heart, acardiac twins can be classified as hemiacardius when heart is incompletely formed and holoacardius if the heart is absent [7]. The prominent features of the recipient twin are total or partial absence of cranial vault, holoprosencephaly, absent facial structures, anophthalmia, microphthalmia, cleft lip, cleft palate, absent or rudimentary limbs, lungs, heart, liver and gallbladder, diaphragmatic defects, esophageal atresia, ventral wall defects, ascites, edema of skin and single umbilical artery.

Management of twin pregnancy with an acardiac fetus is a challenge as the continuous growth of the acardiac fetus is deleterious to the healthy pump twin. It can lead to cardiac insufficiency, polyhydramnios, prematurity and even death of the structurally intact twin in up to 50% of cases [8].

Moore et al. reviewed 49 cases of acardiac twins and reported that perinatal outcome was related to the ratio of weight of the acardiac twin to the weight of the normal twin. They stated that when twin weight ratio which is the acardiac twin weight divided by the normal twin weight was above 70%, preterm labor, hydramnios and congestive heart failure in the pump twin were found and when the weight of the acardiac fetus was less than 25% compared to the pump twin, the prognosis was better

[8].The weight ratio in this study was derived from post delivery weights. They also proposed a second order regression equation to predict weight of the acardiac twins antenatally: Weight (g) = 1.2 x (longest dimension (cm)) 2 – 1.7 x longest dimension (cm). Also the use of abdominal circumference ratios could provide a better approach to establish the differences in body weights [8].

Brassard et al. reported that low pulsatility indices in the umbilical artery perfusing the acardiac twin compared with the pump twin correlated with poor prognosis [9].

Optimal management is controversial. Expectant management versus prenatal intervention is to be debated. Many methods of management have been proposed including termination of pregnancy, serial ultrasound scans to monitor for signs of decompensation, medical management of polyhydramnios or by serial amniocentesis, hysterotomy to remove anomalous twin and invasive procedures. Goal of prenatal treatment is to stop blood flow to the acardiac twin without affecting the pump twin in order to improve its outcome. Platt et al. in 1983 were the first to suggest occlusion of the circulation to the acardiac twin as the definitive treatment to interrupt blood supply to it [10]. Minimally, invasive intervention methods are through cord occlusion techniques or intrafetal ablation. Cord occlusion has been attempted by embolization, cord ligation, laser coagulation, bipolar diathermy and monopolar diathermy while intrafetal ablation is performed with alcohol, monopolar diathermy, interstitial laser and radiofrequency [11].

Tan and Sepulveda recommended intrafetal ablation as the treatment of choice than cord occlusion [11]. They claimed ultrasound guided intrafetal approach to be easier, less invasive and with a higher rate of success than ultrasound and fetoscopy guided cord occlusion procedures.

Invasive treatment should be restricted to those pregnancies which would benefit from prenatal intervention like those where the pump twin is at a significant risk of prematurity, cardiac insufficiency or death and should be considered in presence of poor prognostic factors like polyhydramnios, ultrasound markers of cardiac insufficiency, large acardiac twin and rapid growth of or evidence of substantial blood flow perfusion through the umbilical vessel supplying the parasitic mass [12].

Sullivan et al. advocated expectant management in all cases [13]. They reported 90% survival in pump twin in 10 pregnancies with an acardiac twin managed expectantly. They cautioned against aggressive intervention and recommended expectant management with close fetal surveillance. Stamatian et al. gave an opinion that conservative management is indicated in cases where the acardiac twin is small and when there are no signs of cardiovascular impairment in the pump twin [14]. Serial ultrasound surveillance is important for detecting any worsening of the condition, which may suggest the need for interventions to optimize the pump-twin's chance for survival [15].

CONCLUSION

Expectant management with close antepartum surveillance deserves consideration in cases of monozygotic twins with Twin Reversed Arterial Perfusion sequence. Neonatal mortality of the pump twin diagnosed antenatally may be considerably less than reported. The interruption of vascular communication between the twins is difficult to accomplish. It needs expensive equipment and trained personnel for the procedures. Hence invasive treatment should be considered only in cases with poor prognostic factors. Such pregnancies should be managed by fetal medicine specialists familiar with invasive procedures. Intra fetal ablation procedures are better than cord occlusion techniques. In our cases, all the three pump twins survived with expectant management.

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Author Contributions

Deepa V Kanagal – Substantial contributions to conception and design, acquisition of data, drafting the article, revising it critically for important intellectual content, final approval of the version to be published
Mangala Srikantaswamy – Substantial contributions to conception and design, acquisition of data, drafting the article, revising it critically for important intellectual content, final approval of the version to be published
Rashmi M Devaraj – Substantial contributions to conception and design, acquisition of data, drafting the article, revising it critically for important intellectual content, final approval of the version to be published
Maitri M Kulkarni – Substantial contributions to conception and design, acquisition of data, drafting the article, revising it critically for important intellectual content, final approval of the version to be published
Mahendra Ganagadharaiah – Substantial contributions to conception and design, acquisition of data, drafting the article, revising it critically for important intellectual content, final approval of the version to be published
Lokeshchandra C Hanumanalu – Substantial contributions to conception and design, acquisition of data, drafting the article, revising it critically for important intellectual content, final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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