

Twin Reversed Arterial Perfusion: To Treat or Not?

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ABSTRACT

Acardiac twinning or Twin Reverse Arterial Perfusion (TRAP)-sequence is a rare complication of monochorionic twin pregnancy. Whether to start elective or therapeutic treatment in TRAP-sequence is still controversial. In the present case, acardiac twin was not diagnosed till her delivery at 39 weeks. A healthy baby weighing 2.45 kg was delivered along with another amorphous mass (acardiac twin) of about 150 g which was attached to the placenta with a short and separate cord. As outcome of normal twin vary according to the growth of acardiac twin, frequent follow-up of the normal twin is required to look for the features of heart failure. Hence, the diagnosis of acardiac twin is essential in early pregnancy.

Keywords: Acardiac twin, Monochorionic twin pregnancy, Radiofrequency ablation in TRAP sequence

CASE REPORT

A 26-year-old gravida 3, para 1 with one live baby and one abortion came to emergency labour room at 39 weeks of gestation with intrahepatic cholestasis of pregnancy. She had conceived spontaneously. Her 4D ultrasound at 18 weeks revealed single normal fetus without any congenital malformation. She delivered vaginally after induction of labour in view of intrauterine growth restriction with intrahepatic cholestasis of pregnancy at 39 weeks. Outcome of pregnancy was a 2.45 kg (small for gestational age) live born baby boy with apgar score of 9, 9 at one and five minutes respectively. Soon after delivery of normal twin, she expelled another amorphous mass (acardiac twin) which appeared as trunk like structure with one foot having three digits at caudal end and ocular placodes with hairs at cranial end [Table/Fig-1,2]. Weight of the acardiac twin was 150 g.

Grossly, placenta was single with two umbilical cords. The normal twin's cord was long, had three vessels while acardiac twin had a short cord with two vessels. Both twins shared the same placenta and had single amniotic sac. Placenta and this acardiac twin like mass were sent for autopsy after taking consent of parents. Patient was transferred to postnatal ward in satisfactory condition and was discharged from hospital on the 5th postpartum day.

X-ray of acardiac twin revealed multiple bones arranged vertically on dorsal aspect and metatarsals in the foot [Table/Fig-3]. Autopsy of the acardiac twin (10x5 cm) revealed an incompletely formed skeleton, with no head, upper limbs, heart, or thoracic structures. It had an abdominal stump without any intraabdominal organs; there was one incompletely developed foot with three toes. Histopathological examination of placenta revealed hypoxic changes and that of acardiac twin revealed disorganised architecture of mesenchymal elements such as cartilage and bone enclosing marrow elements. Also seen were, stratified squamous epidermis, adnexal structures, mature adipose tissue, fibrous connective tissue and skeletal muscle bundles in a disorganised array consistent with TRAP. Finally, a diagnosis of monochorionic monoamniotic twins with T2 acardiac twin acardius acephalus type was made.

DISCUSSION

TRAP refers to an unusual and exclusive complication of monochorionic twin pregnancy with an incidence of 2.6 percent [1]. Due to availability of highly developed ultrasonography, types of chorionicity and TRAP sequence can be diagnosed as early as 11 weeks of gestation [2,3]. As not all pregnant patients seek antenatal care in first trimester, some who visit may not have



[Table/Fig-1]: Acardiac twin with short umbilical cord attached with the placenta of normal twin.



[Table/Fig-2]: Acardiac twin. Hairs at the cranial end and partially developed foot at caudal end.



[Table/Fig-3]: X-ray of acardiac twin. Vertically arranged bone at dorsal aspects and metatarsals in the foot.

undergone antenatal ultrasonography. Due to same reason chorionicity was determined before 14 weeks only in 27% of twin pregnancies in a study done by T Dias [4]. So, there must be some guidelines to recommend first trimester ultrasonography for all antenatal patients to detect multiple pregnancies and to determine chorionicity. The NICE guidelines also suggest a second option if chorionicity determination is not possible by ultrasound at the time of detecting the multiple pregnancy. It is advised to manage them as monochorionic pregnancy if found doubtful on subsequent ultrasonography [5].

TRAP should be suspected in monochorionic twin pregnancies when one fetus appears anatomically normal and the other lacks apparent cardiac structures and/or activity. Doppler study reveals presence of blood flow in it and flow of blood will be towards the acardiac twin. The acardiac phenotype ranges from well developed lower extremities, pelvis, and abdomen to a tissue mass like teratoma that is not readily recognizable as fetal parts. Acardiac twin with multiple anomalies may simulate single intrauterine death of one grossly abnormal monochorionic twin and with placental teratoma. Healey reported polyhydramnios in 70% of acardiac fetus sac who had a functional urinary tract and also reported two vessels in the umbilical cord in up to 70% of acardiac fetuses [6]. After diagnosis of TRAP sequence, ratio of weight of the acardiac twin to pump twin should be considered to predict fetal prognosis. The weight of acardiac twin can be calculated from its longest length by using following formula: weight (grams) = $(-1.66 \times \text{longest length}) + 1.21 \times (\text{longest length})^2$ [7]. Otherwise, ratio of abdominal circumference of acardiac twin to normal twin can be measured to predict fetal outcome and its ratio of ≥ 1.0 is considered significant [7]. According to Moore et al., when the ratio of the weight of the acardiac twin to that of the pump twin exceeded 0.70, the risks of preterm delivery and hydramnios were 90 and 40%, respectively [7]. Fetal surveillance from 16 weeks onwards with doppler studies of middle cerebral artery help in early diagnosis of anemia in pump twin. Further, reversed diastolic blood flow in the umbilical artery and umbilical vein's pulsatile blood flow, or abnormal ductus venosus blood flow in the pump twin are indicators of impending intrauterine fetal demise [8]. Other poor prognostic factors for normal twin are risk of cord entanglement and hydramnios in monoamniotic twin pregnancy.

Pregnancies with TRAP Sequence have the options of expectant management, antenatal intervention and delivery. Karyotype of the pump twin should be determined in all patients before doing

any intervention. In several acardiac twins, Blaicher W et al., found karyotypes that were different from those of the co-twin [9]. Laser YAG or diode laser (20 to 40 W) is one of the intervention modalities of TRAP sequence, which is done by a 2 to 3 mm fetoscope. Under direct visualization, either the arterial-arterial anastomoses or the umbilical cord itself are targeted. Bipolar cautery forceps (3 mm) can be used in case of large umbilical cord diameter or in stained amniotic fluid.

Radiofrequency Ablation (RFA) is used to coagulate the abdominal wall at the base of the umbilical cord, rather than direct coagulation of the umbilical cord itself. With all these techniques, cessation of flow is confirmed by Doppler ultrasound interrogation as the last step of the procedure. Maternal complications like bleeding, need for laparotomy to complete the procedure, thermal injury, chorioamnionitis leading to maternal sepsis, and disseminated intravascular coagulation have been reported with these procedure [10]. RFA is the preferred method of intrafetal ablation as compared to cord occlusion because it appears to have the lowest risk of membrane rupture and associated with a higher gestational age at delivery [11].

At present there is a controversy regarding elective versus therapeutic treatment of reversed arterial perfusion sequence. Lewi et al., found spontaneous flow arrest in 21% of cases during 16-18 weeks, which gives the hope of conservative management in these cases [12]. In one series of TRAP sequence, by Moore TR et al., and another series by Pagani G et al the perinatal mortality rate for conservatively managed pump twins was very high due to development of heart failure and preterm delivery [7,13].

Pagani G et al., reported a case series in which treatment was given after onset of poor prognostic factors in normal twin [13]. Outcome of treatment was good as compared to those who were kept under conservative management; hence they supported the elective intervention due to low sensitivity of prognostic factors to predict intrauterine fetal death.

Minakshi et al., reported a case that determines conservative treatment for salvation of the pump twin when the APTW ratio was less than 25 percent [14]. Wong and Sepulveda also suggest expectant management in these cases and treatment was indicated only in presence of adverse factors [15].

To improve the outcome for the pump twin, fetal monitoring should be done with weekly ultrasound and doppler flow to look for signs of fetal hydrops. Frequency of ultrasound should be increased to twice a week if there is evidence of pre-hydrops. Antenatal corticosteroids should be administered between 24 and 34 weeks of gestation to all patients with TRAP Sequence as there is risk of preterm labour and impending compromise of pump twin.

CONCLUSION

First trimester ultrasound should be done to detect all monochorionic pregnancies and follow up of vanishing twin must be done with Doppler to look for acardiac twin. Antenatal ultrasound should be critically analysed by radiologist and presence of any abnormal mass along with the normal fetus should not go unnoticed. Fetuses with signs indicative of poor prognosis are candidates for intervention and should be referred to higher institute.

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