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## Case report

# Acardius acephalus with spontaneous umbilical cord occlusion: Reporting a rare case <sup>☆</sup>

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## ABSTRACT

Twin reversed arterial perfusion syndrome is a rare obstetric condition that occurs in mono-chorionic twin pregnancies, resulting in coexistence of a normal “pump” twin and an acardiac twin. The acardiac twin is dependent upon the normal twin to provide circulation by means of vascular anastomosis, thereby putting the pump fetus at risk of high output cardiac failure. Overall only 50% of pump twins survive. Mortality for acardiac twin is 100%. We present a case of 26-year-old primigravida female presenting with 8 months of amenorrhea with unsure LMP. Ultrasonography followed by fetal MRI was carried out which revealed acardius acephalus twin with absence of blood flow in umbilical vessels. Pump twin had multicystic dysplastic left kidney with single umbilical artery. Following delivery, the pump twin survived well and the deformed fetus showed features of twin reversed arterial perfusion syndrome.

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## Introduction

Acardiac twin is the most severe malformation known in humans. The peculiar appearance of acardiac twin is responsible for the term “Acardiac monsters” given to this malformation before the advent of ultrasonography. Other more acceptable terms for this malformation include Acardius, twin reversed

arterial perfusion (TRAP), and chorioangiopagus parasiticus [1].

The acardiac twin, is characterized by absence of heart and missing other upper body organ systems and is thus dependent upon the normal twin to provide circulation by means of vascular anastomosis, thereby putting the pump fetus at risk of high output cardiac failure [2]. Thus, treatment aims at occluding vascular supply to acardiac twin to ensure nor-

Abbreviations: TRAP, Twin Reversed Arterial Perfusion; MRI, Magnetic Resonance Imaging; HIFU, High Intensity Focussed Ultrasound; LMP, Last Menstrual Period; FOV, Field of View.

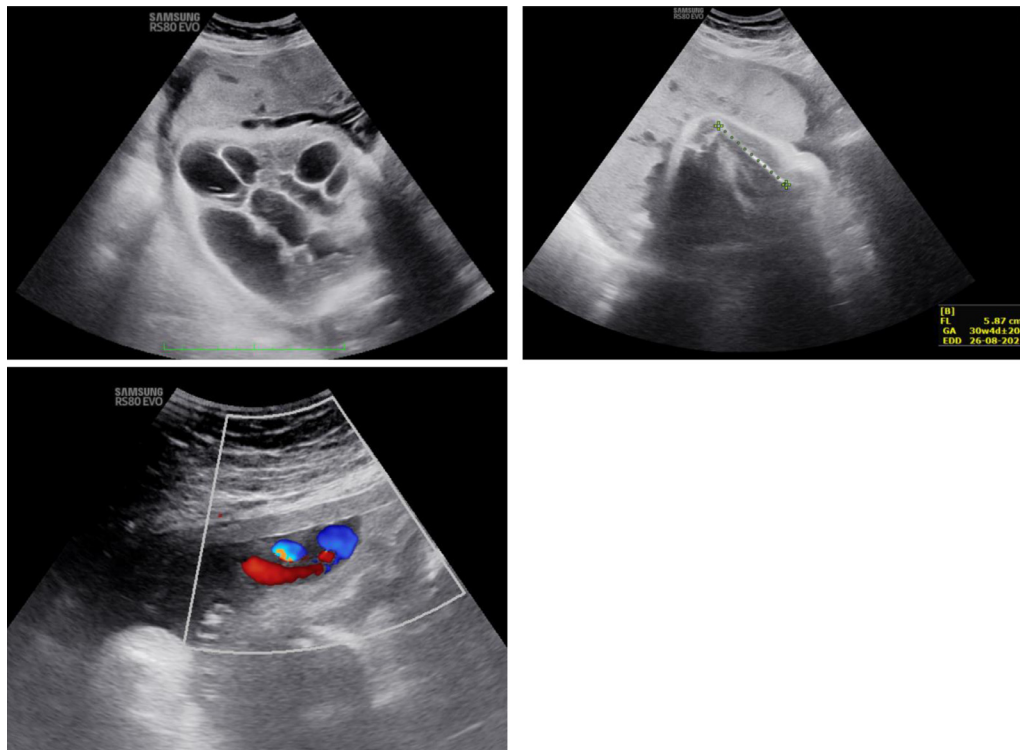
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**Fig. 1 – (A) Grayscale ultrasonography image reveals acardiac twin on right side of gestation sac with multiple cystic spaces noted in torso. (B) Grayscale ultrasonography image- relatively well-developed lower limb was noted in acardiac twin with femur length corresponding to 30 weeks of gestation. (C) Color doppler image- 2 vessel cord with single umbilical artery was noted supplying the normal twin, with no blood flow noted in umbilical cord supplying acardiac twin**

mal growth and development of pump fetus. We present a rare case of TRAP sequence with the pump twin having multicystic dysplastic kidney and single umbilical artery and the acardiac acephalus twin with spontaneous cord occlusion (Fig. 2). Such spontaneous cord occlusion has not been previously described in existing literature (Figs. 3 and 4).

## Case report

Twenty-six-year-old primigravida female presented at 8 months gestation with unsure LMP with complaints of pain abdomen and decreased fetal movements. No previous antenatal checkup or ultrasonography had been carried out. Clinical examination revealed presence of fetal cardiac activity on auscultation.

Ultrasonography showed evidence of monochorionic twin pregnancy with single placenta along fundus and right anterolateral wall Fig. 1.

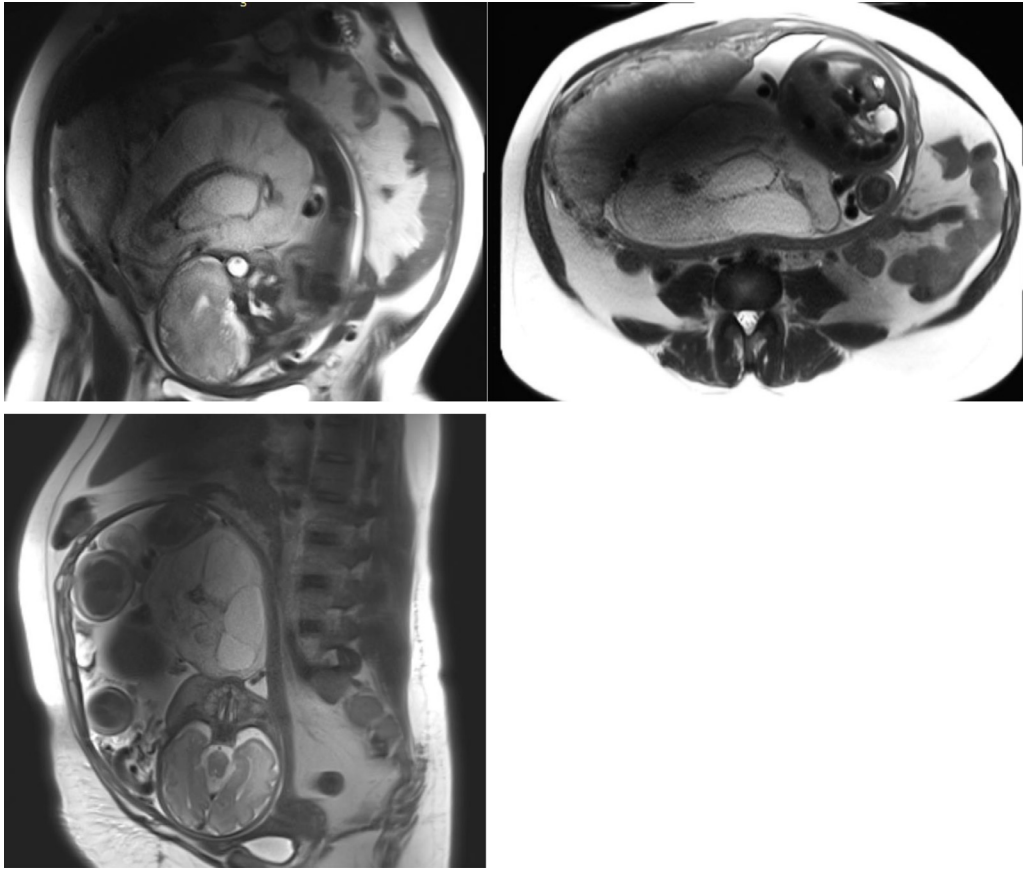
First twin (twin A) was noted in vertex presentation, with grossly normal fetal morphology. Biometry revealed ultrasonography gestation age as 33 weeks. Left kidney of twin A showed multiple cystic areas within, with normal right kidney. Two vessel umbilical cord was noted with single umbilical artery. Doppler study showed normal waveform in umbilical artery.

Second twin (twin B) had grossly abnormal morphology with nonvisualization of heart, head, and bilateral upper limbs. Multiple variable sized cystic areas along with rudimentary spine was noted in the visualized torso. Both lower limbs were visualized with femur length corresponding to 30 weeks gestation.

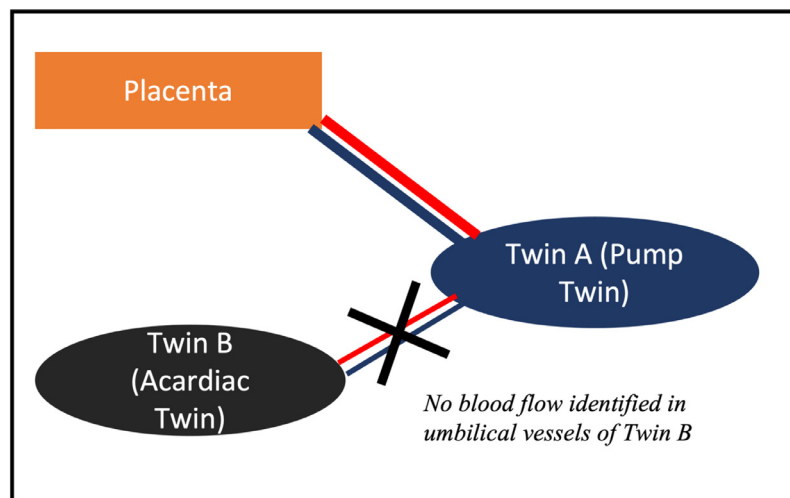
Fetal magnetic resonance imaging was carried out with large FOV to further characterize the anomaly. MR images revealed twin B (acardiac twin) with small sized umbilical vessels located on right side in gravid uterus, while twin A was noted on left side. MRI confirmed the findings elicited by ultrasonography, and a diagnosis of TRAP syndrome was confirmed Fig. 2.

Absence of blood flow in umbilical vessels supplying acardiac twin was indicative of spontaneous cord occlusion Fig. 3. Thus, relatively normal growth was observed in the twin A.

The patient was taken up for lower section caesarean section, and both twins were delivered. Normal twin cried immediately after birth, and birth weight was 2240 g. APGAR score was 8 at 1 minute and 9 at 5 minutes. Echocardiography performed on day 2 of life revealed no abnormality. Ultrasonography confirmed the finding of multicystic dysplastic left kidney in twin A. Gross anatomy of acardiac twin was consistent with imaging findings. Head and neck were not identified with a tuft of hair at cranial aspect of torso. Upper limbs were hypoplastic with relatively well-developed lower limbs Fig. 4. The weight of acardiac twin was 1115 g.



**Fig. 2 – Fetal MRI- T2 HASTE images in (A) coronal, (B) axial, (C) sagittal plane shows- grossly malformed acardiac twin on right side with multiple cystic spaces and rudimentary spine in torso with absence of heart, head and neck and upper limbs. Pump twin is noted on left side, with normally developed heart and brain. Multiple cystic spaces are noted in left kidney. Markedly different size of umbilical vessels is noted supplying the pump twin and acardiac twin**



**Fig. 3 – Pictorial representation of circulation of twin A and twin B**



**Fig. 4 – Gross appearance of acardiac acephalus twin with absent head and neck, and rudimentary upper limbs.**

## Discussion

The incidence of acardiac twinning has been widely accepted as approximately 1% of monozygotic twin pregnancies and approximately 1:35,000 pregnancies [3]. In twin pregnancy, approximately 20% are monozygotic; of these 75% share a placenta. Most monozygotic twin pregnancies are characterized by varying degrees of anastomoses. Artery-artery anastomosis predominate in 75% of monozygotic placentas in comparison to artery-vein or vein-vein anastomosis [4].

## Pathogenesis

The pathogenesis of acardiac twin can be summarized as TRAP which indicates that acardiac twin is dependent upon normal twin by means of anomalous reversal of umbilical arterial flow, resulting from vascular anastomosis between 2 circulations of twins [5,6].

Whether umbilical vascular anastomosis precedes cardiac embryogenesis defect in cases of acardiac twin is not yet certain. One theory postulates that vascular anastomosis in early embryogenesis leads to early tissue hypoxia and atrophy of heart and other organs while other theory states that due to failure in cardiac development in one twin, anastomosis between umbilical vessels gets established, allowing survival of affected twin [7].

In one case report, there is association of TRAP with VACTERL which indicates defect in early embryogenesis [8].

Chromosomal disorder has been reported in 50% of cases of TRAP syndrome. Monochorionicity always highlights risk of TRAP [9]. In our case it was monozygotic and diamniotic twins.

Eventually the acardiac twin becomes dependent on the perfusion of the “Pump” twin. The upper half of the body of an acardiac twin is extremely poorly developed and sometimes not developed at all. Head, cervical spine and upper limbs are usually absent. Oedema in the upper body consistent with cystic hygroma is common. In contrast the lower half of the body although malformed is better developed. This is because of mechanism of perfusion of the acardiac twin, where blood enters via abdominal aorta which is deoxygenated blood that left the normal twin. So the oxygen and nutrition available is extracted allowing some development of caudal aspect. Once blood enters upper torso in retrograde fashion oxygen saturation is extremely low, halting development of heart, head and upper torso giving rise to severe deficits [10]. The pump twin suffers from high output cardiac failure which is directly proportional to size/weight of the acardiac twin A weight ratio of more than 70% between the pump and acardiac twin is associated with increased incidence of preterm delivery and congestive heart failure [11].

## Anomalies

Anomalies reported in acardiac twin are partial/total absence of cranial vault, holoprosencephaly, absent facial structures, anophthalmia, micro-ophthalmia, cleft lip, cleft palate, absent or rudimentary limbs, diaphragmatic defects, absent lungs & heart, esophageal atresia, ventral wall defects, ascites, absent liver & gall bladder, oedema of the skin, single umbilical artery (75% cases), markedly different sized umbilical artery, inconsistent membrane development between the twins and occasionally umbilical artery drawn directly into superior mesenteric artery [12].

## Classification

Depending on anomalies the acardiac twin are classified as Hemiacardius (if the heart is incompletely formed) and Holocardius (if the heart is absent). Other classification is Acephalus (if head is absent- 60-75% of cases), Aniceps (if head is poorly formed- 10% of cases), Acormus (presence of head only- 5% of cases) and Amorphous (unrecognizable amorphous masses- 20% of cases). In Acardius Acephalus, the cranial and thoracic structures are missing but there is preservation of the lower limbs, as seen in this case [5,9,13].

The classifications have little significance as acardiac fetus is invariably fatal and the importance of survival of pump twin is the priority. Congenital anomalies are present in about 9% of pump twins. More commonly, pump twin presents with features of congestive heart failure including ascites, pleural effusion, polyhydramnios and skin oedema [12]. Polyhydramnios in cases of acardiac twin is due to hyperperfusion of kidneys, leading to excessive urine production [5].

In our case, the cardiac twin showed absence of heart and head with poorly formed upper limbs and multiple cystic spaces in torso, and was thus classified as Acardiac Acephalus. In the pump twin, there was presence of multicystic dysplastic left kidney. However, no sign of congestive heart failure was noted either in antenatal or postnatal imaging. This can be attributed to the spontaneous cord occlusion noted in our case.

Treatment aims at increasing the probability of survival of the pump twin by reducing the risk for developing hydrops, cardiac failure, and preterm labor. Management aims at interrupting the blood flow from the pump twin to the acardiac twin without harming the normal twin [14]. Amnioreduction or medical therapy can be used to limit polyhydramnios and reduce the risk for preterm delivery. Other treatment options include surgical occlusion of the umbilical cord of the acardiac twin. More recently, intrafetal radiofrequency ablation and laser therapy have been performed [15]. The first successful case of treatment with high-intensity focused US was recently described [16]. The survival rate of the pump twin after treatment with laser coagulation and radiofrequency ablation ranges from 75% to 80% [17].

In our case, no intervention was required as there was no flow in the vessels supplying the acardiac twin. Spontaneous cord occlusion has not been described previously in existing literature in cases of TRAP.

In our case, no sign of congestive heart failure was noted in pump twin. An echocardiography of pump twin obtained at 2 days of age showed normal parameters.

## Conclusion

Fetus acardius is a dreadful outcome occurring in monozygotic multiple pregnancy resulting from TRAP sequence with the presence of a parasitic acardiac twin that thrives by getting blood supply from the donor hydropic twin. Prognosis is lethal for a the acardiac twin. It is associated with hydramnios, cardiac hypertrophy, congestive failure, hydrops fetalis, premature birth or mortality of normal fetus. Diagnosis is confirmed by transvaginal Doppler USG and is managed by interrupting blood flow from the normal fetus to the acardiac fetus if needed. We reported a rare case of spontaneous cord occlusion of parasitic twin in a case of acardiac acephalus, which improved the outcome of pump twin.

The obstetrician and radiologist should be aware of this anomaly especially in twin/multiple pregnancies so that timely proper measures can be taken for ensuring survival of the pump twin.

The pathologist should be aware of this entity so that proper autopsy report can be given to parents which can be helpful in planning future pregnancies.

## Patient consent

Patient's written consent was taken for images, photos, and clinical details to be published.

## Availability of data and material

The masterchart is not being shared to safeguard patient's privacy. However, we agree to share to the same when specifically asked for.

## Authors' contributions

All have contributed to conceptualise the design, analyze, interpret, and drafted the work. All have approved the submitted version and agree both to be personally accountable for the contributions and ensure that questions related to the accuracy or integrity of any part of the work, are appropriately investigated, resolved, and the resolution documented in the literature. All the authors have read and approved the manuscript.

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