

Rare congenital abnormality of “acardiac amorphous”: Case report and review of literature

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ABSTRACT

The phenomenon of an acardiac twin is an extremely rare congenital abnormality. It occurs with monochorionic twins and has been recently explained by the twin reverse arterial perfusion (TRAP) syndrome. This latter is a consequence of abnormalities in placentation as well as a deformed vascular anastomosis between the donor and the recipient fetuses. Undoubtedly, the prenatal course has been proven essential in the diagnosis as well as in the prediction of prognosis in cases where interventions are needed.

Keywords: Acardiac amorphous, Intrafetal ablation, Monochorionic twin, Twin reverse arterial perfusion syndrome, Ultrasound

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INTRODUCTION

Acardiac twin is a rare complication of monozygotic twin pregnancies occurring in 1% of monochorionic twin gestations with an incidence of 1 in 35,000 births [1]. The early diagnosis of this pathology gives the chance for therapeutic options such as radiofrequency ablation or the new invasive technique of “tissue-sealing” material and hence increasing the prognosis of the normal fetus. Prenatal obstetrical ultrasound remains the corner stone in the diagnosis and paves the path of prognosis dictating interventions.

In this article, we present a case of a pregnant patient who initially presented to Sahel general hospital in Beirut for preterm labor and preterm premature rupture of membranes with a monochorionic twin gestation and was found to carry an acardiac twin.

CASE REPORT

Our patient was a 34-year-old Caucasian lady G6P3A2, previously healthy, nonsmoker, nonalcoholic, not known to have any drugs or food allergy, and having had three previous normal vaginal deliveries of healthy babies. The current pregnancy was a spontaneous monochorionic diamniotic pregnancy that did not benefit from an appropriate follow-up due to financial reasons. In fact, the patient had lost follow-up after the 2nd trimester ultrasound scan. To note that folic acid was taken in the first trimester with adequate dosage (400 mcg once daily).

The 2nd trimester obstetrical ultrasound report done in the second trimester showed a monochorionic, diamniotic twin gestation: fundal placenta, fetus A with fetal heart rate (FHR) = 156, cephalic presentation, BPD (bi-parietal diameter) = 4.58 cm, HC (head circumference) = 16.39 cm, AC (abdominal circumference) = 15.62 cm,

EFW (estimated fetal weight) = 340 g. Gestational age based on: AUA (actual ultrasound age) = 19+6 WD (week day), LMP (last menstrual period) = 21+1 WD, EDD (estimated date of delivery) = 02/07/2019 (Figures 1 and 2). Fetus B: Non-viable, fetal growth: limited growth, absent cardiac structure or activity, hydropic fetus with BPD 4.29 cm, correlates with 18 weeks of gestation, no parameters to estimate the fetal weight of fetus B are present. Normal quantity of amniotic fluid was noted in both amniotic sacs (Figure 3).

The patient was admitted at 35+5 weeks of gestation for preterm labor and preterm premature rupture of membranes with clear fluid 1 hour prior to presentation after referral from another obstetrical institution. Vaginal exam showed a 3–4 cm cervical dilation, 80% effacement, S-2 station, breech presentation with fluid leak.

Upon presentation, the fundal height of the patient was consistent with 36 weeks of gestation. Cardio-tocography (CTG) of baby 1 was category 1, reactive. Cardio-tocography of baby 2 showed no fetal heart tracing. Contractions were regular.

C-section was performed for twin malpresentation, the intraoperative course was smooth: under spinal anesthesia, with Pfannenstiel incision and transverse lower uterine incision. Baby A girl was extracted, breech presentation, APGAR 9/10 at 0 and 5 minutes, respectively, and with a weight of 2020 g transferred to nursery. While attempting to deliver baby B, a limb was felt and grasped in order to deliver the body, however, a large mass was perceived. The mass was almost the size of a regular newborn with what looked like a limb attached to it. The delivery process was difficult. The mass was oval with a face like appearance and a limb, weighing 1455 g, suggestive an “acardiac twin” (Figures 4 and 5).

The family refused to do X-rays or autopsy for the dysmorphic baby due to ethical and cultural beliefs.

The placenta was monochorionic with two umbilical cords weighing 600 g and was sent to pathology.



Figure 1: Morphoscan of the fetus A (normal fetus).



Figure 2: Baby B: acardiac amorphous (yellow arrow: umbilical cord, red arrow: deformed limb-femur like extremity, black arrow: eye, green arrow: skull).



Figure 3: Morphoscan of the fetus B—the acardiac amorphous with no defined structures (abdomen, umbilical cord, limb).



Figure 4: Morphoscan of the fetus A [FHR = 156, cephalic presentation, BPD (bi-parietal diameter) =4.58 cm, HC (head circumference) = 16.39 cm].



Figure 5: Blue arrow: femur-like extremity. Baby B: Possible deformed head with hair and eye).

Results showed no evidence of trophoblastic disease or chromosomal abnormalities.

Postoperative course in hospital was smooth and the patient discharged on day 2. Followed after 6 months, the baby girl had normal growth with no signs of developmental delay.

DISCUSSION

The first case of acardiac fetus with twin gestation was reported by Mery in 1720 and Winslow in 1740 [2]. Over years, and due to its rarity, a hypothesis about the pathophysiology behind acardiac fetuses was initially proposed in 1850. It described a process of reversed perfusion due to large arterio-arterial and veno-venous anastomoses after arrest of heart development which leads to an acardiac fetus being maintained by arterial perfusion from the pump twin [2]. A second hypothesis was then declared by Claudius in 1859 stating that the heart degenerates after the normal initial development when a reversed flow from the aorta leads to thrombosis [2]. Twin reverse arterial perfusion (TRAP) syndrome was also identified as the inter-placental anastomoses between the fetuses being arterio-arterial and veno-venous in contrast to the arterio-venous anastomoses found in the twin-twin transfusion syndrome [2]. Overall, the acardiac twin phenomenon is defined by the presence of both a normal formed twin and an amorphous twin with the absence of a head [3], its incidence is 1 over 35,000 of pregnancies and 1% of the monozygotic twin gestation [2, 3]. To note, a few cases were reported in the literature where TRAP syndrome was associated with di-chorionic twin, triplet gestation, and triploidy [2, 4].

The etiology remains unknown, and most acardiacs had shown a normal karyotype [4]. However, some chromosomal studies of acardiac fetuses have shown abnormalities like trisomy 21, 22 leading to the thinking that acardiacs could be merely a part of a generalized chromosomal defect [5] after having found that 50% of chromosomal abnormalities can be identified in acardiac fetuses [6]. Also, a difference in cytogenesis between the two twins was noted to be present [7]. On the other hand, the researchers found that several acardiacs have different

karyotypes compared to the normal co-twin. In a case report of triplet gestation after in vitro fertilization (IVF), one of the fetuses was an acardiac identified with trisomy 2 [8]. The hypotheses of error in embryonic development stages, primary endodermal defects, and errors in the morphogenesis of the heart were also proposed [4].

The acardiac twins are classified into four classes depending on their external appearance, the degree of cephalic and truncal maldevelopment [2, 3, 4, 9]:

- **Acardiusacephalus 62%:** The most common variety presenting no head but a rudimentary skull base can be present; the upper extremities are, in contrast to the lower ones, almost always absent. The diaphragm, thorax, and superior internal organs are absent. A subcutaneous necrosis due to hypoxia gives the fetus a thickened edematous skin.
- **Acardius amorphous 25%:** The least developed monster, not recognizable as a human form. It may have the appearance of a “blob” or ball of skin with or without hair and with no identifiable extremities. Bone, cartilage, fat, fibrous, and muscular tissue and blood vessels can be found.
- **Acardius anceps 8%:** The most developed form, part of the head is developed with remnant of cranial bone and brain tissue. The body and extremities are usually present.
- **Acardiusacormus 5%:** The rarest form with trunkless head, the head is present but not developed, the trunk can be present as cervical appendix. The main prenatal diagnostic tool is the obstetrical morpho-Doppler sonography done in the early second trimester. A reversal of flow in the pulsed Doppler of the umbilical artery of the recipient twin will confirm the diagnosis of TRAP [3]. The pump-to-acardiac umbilical venous diameter ratio of more than 0.5 also represents the excess of cardiac output for acardiac perfusion [10]. To note that in most cases of the acardiacs, approximately 75%, one single umbilical artery is present [9, 11]. Dashe et al. showed in their work that the interpretation of the Doppler of the umbilical artery of the acardiac fetus alone does not predict the evolution of the normal co-twin. So, the index of resistance between the Doppler of both twins is the most predictive value for the prognosis of pregnancy; if the index is more than 0.2 then the prognosis is expected to good while an index less than 0.05 predicts the occurrence of complications [3]. Abnormal flow directional properties of the TRAP sequence were also described by Bornstein et al. (2008), in addition to the volume mass ratio between the acardiac and the pump twin with a ratio of 0.95 predicting the prognosis [9]. The acardiac twin can appear as teratoma, heterogeneous mass, or intrauterine fetal demise. It can also present with a dorsal cystic hygroma in rare cases [12]. A twin weight ratio can also be considered to determine the prognosis despite the difficulty in measuring

the weight due to malformations. If the acardiac fetus weighs less than 25% of the normal co-twin, then favorable outcome is present. If the ratio is more than 70%, complications such as preterm labor, polyhydramnios, hydrops of the donor twin and congestive cardiac disease can occur [3, 13]. The more the acardiac is big, the more perfusion is demanded and the more the prognosis is severe. Perinatal mortality associated with TRAP syndrome is 50–70% mainly due to cardiac failure [2, 3, 4, 9, 11, 13].

The expectative management of the TRAP syndrome can be by serial ultrasound Doppler during the pregnancy if prognosis is suspected to be favorable [14]. Recently, pregnancy termination, intrafetal ablation, or interventional anastomosis sclerotization (alcohol ablation, thermal ablation, laser ablation, and bipolar coagulation) were introduced to reduce the risk of complications such as intrauterine fetal demise, preterm labor, cardiac failure, hydrops of pump twin and polyhydramnios [14–16]. The choice of therapy depends on the gestational age (in the first and second trimester as the Wharton's jelly becomes more edematous in the 3rd trimester with less success rates for ablation or coagulation [13]) and the degree of development of the acardiac fetus [14]. Recently, a new intrauterine invasive technique of "tissue-sealing" material, Histoacryl: a material that has appropriate qualities for vascular closure and even for amnion-defect closure was found to be safe, effective and possible in the early weeks of gestation to treat the acardiac fetus in patients who have high rates of fetal ablation failure and risk for preterm premature rupture of membrane [14]. This technique is difficult for its cost, availability and the need of an experienced operator. The choice of method to treat TRAP syndrome should be individualized to each patient conditions.

CONCLUSION

Twin reverse arterial perfusion syndrome is a rare congenital abnormality that occurs in monochorionic twinge stations. The definitive diagnosis is by Doppler morphoscan ultrasound. The perinatal mortality of the co-twin and complications like polyhydramnios, hydrops, cardiac failure, and preterm labor remain very high. The acardiac twin has a high index of resistance with reversal Doppler flow as well as a high volume mass compared to the pump twin. An early prenatal diagnosis allows possible therapeutic options and thus promising in preventing complications. Treatment is considered with either pregnancy termination or intrafetal ablation or coagulation. In addition, the invasive technique with tissue sealing of the umbilical artery of the acardiac fetus by Histoacryl represents a promising technique improving the chances to better decrease in morbidity and mortality.

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Zaynab Abed Kalach – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final

approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Mariam Zeineddine – Conception of the work, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Hala Hassan – Conception of the work, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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