# ACARDIAC ACEPHALUS TWIN: A CASE REPORT

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## ABSTRACT

Acardiac acephalus twin is a rare complication of monochorionic twinning. A case of a 26year old who presented to the emergency unit of the University of Maiduguri Teaching Hospital [UMTH] with ultrasound report of non viable but morphologically normal twin pregnancy is presented. She was delivered of a twin still born; one apparently normal and an acardiac twin.

**Key words:** Multiple pregnancy, Ultrasound scan, Acardiac twin

## INTRODUCTION

Multiple pregnancies, which account for 1.5% of all pregnancies, are responsible for 10% of perinatal morbidity and mortality. [1] Congenital malformations are twice as common in multiple than in singleton pregnancies. [1,2] Malformations, such as conjoined twins and acardiac twin (reversed-arterial-perfusion (TRAP) sequence) are unique to multiple pregnancies. Acardiac twinning is one of the most severe congenital malformations seen in humans and is unique to monochorionic placentation. The prevalence is 1% of monozygotic twins and one in every 35 000 pregnancies. [1,2] Acardiac twin results from abnormal placental vascular anastomoses through which the structurally normal 'pump' twin provides blood supply to the parasitic acardiac twin in a retrograde, paradoxical fashion.[3] As a result, the 'pump' twin perfuses deoxygenated blood into the 'recipient' twin and a spectrum of anomalies due to reduced formation of body tissues, as a consequence of severe hypoxemia, results in acardiac twins.[2,3] In many cases, the continuous growth of the acardiac twin and the associated "vascular steal" phenomenon may lead to cardiac insufficiency and polyhydramnios in the pump twin.[1,3] The morphology of the pump twin is usually normal but without treatment mortality is the rule in 50 – 75%, particularly if the recipient twin weighs more than half as much as the pump twin.[4,5]

Interruption of the vascular communication between the twins, although simple in concept has been challenging with varying degrees of success.[3,4]. These techniques include injection of sclerosing agents to occlude the umbilical cord of the acardiac twin, ligation of the umbilical cord of the acardiac twin (via hysterotomy or endoscopically), endoscopic laser coagulation of the acardiac cord and thermocoagulation of the main intra-abdominal vessel of the acardiac twin.[6-9] Endoscopic techniques, though more effective are more invasive and require surgical skills and equipment that are not widely available.[3,4]

Conventional fetocide by intracardiac injection of potassium chloride, fetectomy following maternal laparotomy and hysterotomy for removal of the abnormal fetus, intrafunicular insertion of stenting systems or coils, and injection of either fibrin glue into the umbilical cord or absolute alcohol into the systemic circulation of the affected fetus have fallen out of favour because of the associated adverse effects on the healthy twin.[4-6]

## CASE REPORT

A 26 year old $G_4P_2^{+1}$ 2 alive, presented to the emergency unit of the University of Maiduguri Teaching Hospital [UMTH]Maiduguri, with complaint of absent fetal movement for a week at 30 weeks of gestation. There was no history to suggest the cause of the failure to appreciate fetal movement. She had not booked for antenatal care. General examination was unremarkable. Examination of the gravid uterus revealed a fundal height compatible with 29 weeks gestation with multiple fetal parts.

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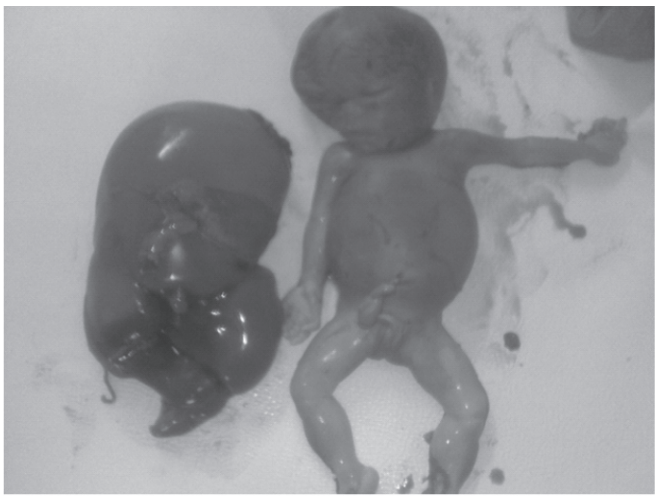
Fetal heart sounds were not heard with Pinard stethoscope. Investigations done included full blood count, urinalysis, syphilis screening, and blood glucose measurement. The results were all normal. Ultrasonography revealed twin pregnancy, both presenting cephalic with a single fundally placed placenta. Cardiac activity were absent in both twins. No malformation was noted. The patient consented to induction of labor after the findings of the ultrasound scan was explained to her. Labor was successfully induced after the 4th dose of 100 microgrammes of misoprostol leading to delivery of two still-born males with monochorionic diamniotic placentation. The placenta weighed 500 gm. And there was no demonstrable arterial and venous communications. Twin A, weighed 900g, apparently normal with polyhydramnios of about 2.5 litres. Twin B, the acardiac weighed 800g, the head and upper extremities were absent, and the external genitalia were poorly developed. The lower spinal column and lower leg bones were apparently normal [Figure 1]. The umbilical cord of the acardiac twin was thicker but both cords contained three vessels, one vein and two arteries. Neither autopsy nor radiographs were obtained as the bodies were taken for burial immediately after delivery.



## DISCUSSION

Management of a twin pregnancy complicated by an acardiac twin is a challenging exercise. On the one hand, uninterrupted growth of the acardiac twin could lead to cardiac insufficiency, polyhydramnios, prematurity, and even perinatal death of the structurally normal twin as typified by the case presented, while on the other hand prenatal intervention to safe the pump twin could adversely affect the pump twin and the mother.[3,6]

Options of management of TRAP sequence includes elective termination of pregnancy, medical therapy for cardiac failure of the pump twin and interventions that target the blood supply of the acardiac fetus.[3,4] Conservative management and follow-up using ultrasound and intervening when there is evidence of cardiac failure in the pump twin, severe polyhydramnios or overgrowth of the acardiac twin has been suggested.[3,7] Others recommend prenatal intervention in early stages of pregnancies to prevent, rather than treat, these complications.[3,8] The index case presented only when she could not perceive fetal movements at 30wks of gestation. With severe polyhydramnious of the pump twin and overgrowth of the acardiac [pump:acardiac of 900gm:800gm] detected after delivery, the perinatal death was almost inevitable.[10] Perinatal mortality in the pump twin is reported to be up to 55%, as a consequence of polyhydramnios and cardiac failure.[10] Polyhydramnious of 2.5litres in the pump twin in our case would have contributed to the demise of the pump twin but the cardiac failure though probable is difficult to establish following fetal demise without an autopsy. The only option available to us at presentation was elective termination of pregnancy. Unfortunately, even if she had presented earlier with viable pump twin, the option of management offered would not have changed significantly as the other modalities of management are not available. Probably because of the rarity of acardiac twin coupled with lack of proficiency of the sonologist and/or poor resolution of the ultrasound scan, the diagnosis of acardiac twin was missed. But the gross polyhydramnious of the pump twin in a monochorionic placentation should have raised suspicion of a major congenital malformation.

## REFERENCES


1. Minakshi R, Seema C, Vanita S, Neelam A, Nittin V. **Acardiac-Acephalus Twins: A Report of 2 Cases and Review of Literature.** Medscape J Med. 2008; 10(8): 200-202
2. Sepulveda W, Sebire NJ. **Acardiac twin: too many invasive treatment options—the problem and not the solution. Ultrasound in Obstetrics and Gynecology 2004;24(4):387-389**
3. Waldo S, Jorge H, Victor D, Juan C. D, Juan LA. **Successful Sonographically Guided Laser Ablation of a Large Acardiac Twin at 26 Weeks'**

**Gestation. J Ultrasound Med 2004; 23:1663-1666**

4. Ruben AQ, Harry R, Karoline SP, Mordechai B, Mark IE, David BC et al Umbilical-Cord Ligation of an Acardiac Twin by Fetoscopy at 19 Weeks of Gestation. N Engl J Med 1994; 330:469-471
5. Ozeren S, Caliskan E, Corakci A, Ozka S Unsuccessful management of acardiac fetus with intrafetal alcohol injection. Ultrasound in Obstetrics and Gynecology2004;24(4):473-474
6. Ville Y. Selective feticide in monochorionic pregnancies: toys for the boys or standard of care? BJOG 2003;22(5):448-450
7. Weisz B, Peltz R, Chayen B. Tailored management of twin reversed arterial perfusion (TRAP) sequence. Ultrasound Obstet Gynecol. 2004;23:451–455
8. Jolly M, Taylor M, Rose G, Govender L, Fisk NM. Interstitial laser: a new surgical technique for twin reversed arterial perfusion sequence in early pregnancy. BJOG 2001; 108:1098–1102
9. Peter S, Karen S, Stephen C, Phillipa K Ultrasound-guided, intra-abdominal laser to treat acardiac pregnancies BJOG 2002;109(3):352-354
10. Moore TR, Gale S, Benirschke K. Perinatal outcome of forty-nine pregnancies complicated by acardiac twinning. Am J Obstet Gynecol 1990;163:907-12.