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Acardius- acephalus Twin Delivery: A Case Report and Review of Literature

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Authors' contributions

This work was carried out in collaboration between all authors. Authors OCO and OAO designed the study, wrote the protocol, and wrote the first draft of the manuscript. Authors NAO and AEN managed the literature searches while author OMO analyzed the data. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

We present a case of a 27 year old primigravida at 36^{+6} weeks gestation with monochorionic diamniotic twin gestation who presented late in pregnancy with twin reversed arterial perfusion (TRAP) sequence and a diagnosis of severe preeclampsia. Twin reversed arterial perfusion syndrome is a very rare manifestation of twin to twin transfusion in a monochorionic gestation where the affected twin receives vascular supply in a retrograde manner from the healthy twin. The antenatal period was poorly supervised due to the temporary closure of our centre. She had her first ultrasound scan at 15 weeks but TRAP sequence was not noted. She was delivered via an emergency lower segment caesarean section. The donor twin is currently doing well. This case underscores the need for proper training and recertification of Fetal Medicine Specialists for accurate and early diagnosis so that the Obstetrician will be fully prepared for all eventualities.

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1. INTRODUCTION

Twin pregnancies continue to intrigue the human population. Nevertheless, this report showcases one of the rare complications of monochorionic twin gestation, namely; acardiac amorphous twin.

Ultrasound remains the mainstay for diagnosis. The use of assisted reproductive technology has remarkably increased the incidence of twins especially monochorionic diamniotic twin gestation globally. Monochorionic diamniotic twin gestations are associated with many complications which include preterm births, Twin to Twin Transfusion Syndrome (TTTS), TRAP etc. The incidence of acardiac twin is 1 in 35,000 deliveries1. In TRAP sequence, one twin is structurally normal and is referred to as 'pump twin' because it pumps blood to the other twin while one is structurally abnormal. It may consist of legs and lower body but no upper body, head or heart. The abnormal twin is referred to as an acardiac twin.

2. CASE REPORT

A 27- year old primigravida at 36^{+6} weeks gestation was admitted from the antenatal clinic for an emergency lower segment caesarean section on account of severe pre-eclampsia. She had mild to moderate occipital headaches but no reduction in urine output, abdominal pains or blurring of vision. There was bilateral lower extremity oedema noted by the patient in the past two weeks prior to presentation. She felt fetal movements. The index pregnancy was booked at a gestational age of 10 weeks in a private hospital in Owerri. At booking, her weight was 95 kg, Blood pressure was 130/80 mmHg and no proteinuria was noted on urinalysis.

Retroviral screening, Hepatitis B surface antigen and venereal disease research laboratory (VDRL) tests were all negative. She attended antenatal clinic in the private hospital and there was no rise in blood pressure or excessive weight gain. She took her routine antenatal medications regularly and did not take any local herbs or concoctions. The routine antenatal medications included tablets fersolate, tablets folic acid, tablets calcium lactate, tablets vitamin and tablets multivite. She had her first ultrasound (USS) on 4th June 2015 with a diagnosis of normal twin gestation at 15 weeks and 5 days. There were no details of chorionicity and TRAP sequence was not noted. Based on this result of twin gestation, she booked at this facility at 15 weeks and 5 days.

Between 15 and 36 weeks there was temporary closure of our hospital Federal Medical Centre, Owerri and we could not see her for subsequent antenatal care. At her second visit at this facility at 36^{+0} weeks, her weight was 104 kg, blood pressure was 152/95 mmHg and urinalysis was normal. She was counselled on the need for admission but she declined. She was placed on tablets methyldopa 250 mg 8 hourly. The next clinic visit was 6 days later. On her third visit on 36^{+6} weeks she had a blood pressure of 160/110mmHg and proteinuria of 2+. Her packed cell volume was 37%. An urgent USS was arranged.

Fig. 1. Shows the picture of Acardius acephalus twin

USS which was done at private ultrasound centre at 36^{+6} showed that a dividing membrane was present which was suggestive of at least a diamniotic gestation. There was one viable twin and the other twin was non-viable.

A repeat ultrasound scan done at Federal Medical Centre, Owerri at the same gestational age (36^{+6}) showed:

Multiple (twin) gestation with a viable fetus and a macerated second fetus at 36weeks, EDD= 01-12-2015 ± 2 weeks. Pelvic examination revealed a cervix that was <40 effaced, posterior, firm. Cervical Os was closed and station was at -3. A diagnosis of unfavorable cervix in a Primigravida with twin gestation, severe preeclampsia and non-viable second twin at term was made.

The patient was booked for an emergency lower segment caesarean section because that was the safest and fastest route of delivery. A vaginal delivery via induction of labour would have taken many hours to achieve since the cervix was not favourable and the patient had severe preeclampsia. Two units of blood were grouped and cross matched. She had the relevant laboratory investigations for preeclampsia which included full blood count and differentials, platelet count, liver function tests, serum urea creatinine with electrolytes, serum uric acid and bedside clotting time. All the results were within normal limits. She had Magnesium sulphate for prevention of convulsions using the Pritchard regimen. Maintenance dose was completed 24 hours later. The patient was also given intravenous labetalol for the prevention of hypertension. Outcome of the surgery was:

Twin 1: Cephalic presenting live male neonate with APGAR score of 9^1 10^5 and weight of 2.5 kg. No external congenital abnormalities were observed on gross examination.

Twin 2: Severely malformed demised male neonate without head or thorax. Abdomen was rudimentary. There was massive bilateral pitting oedema of the lower limbs. Only the first and second toes were formed. Imperforate anus was noted. The weight was 2.0 kg. A monochorionic twin placenta was noted. It weighed 0.8 kg. The cords were centrally inserted.

The umbilical cord of the first twin was 65 cm long but the acardiac twin had a short umbilical cord of about 25 cm. The uterus, tubes and ovaries were normal. The whole twin was sent for histopathology. The patient was transferred to the ward and was discharged on the 10^{th} day in good condition after managing the pre-eclampsia. The patient stayed for 10 days because the blood pressure was not well controlled. She was on oral methyldopa and nifedipine. The baby was seen at 6 weeks postnatal clinic and was doing very well.

2.1 Histopathology Report

An abnormal neonate with only a partially intact pelvis and lower extremities was noted on gross examination. A gastroschisis was also noted. Head, thorax and abdominal organs were all absent.

Waist-heel length = 21.5cm. Cord length = 3.5 cm. Bilateral genu valgus was noted. Both feet had two digits only.

3. DISCUSSION

This was a case of twin pregnancy with TRAP sequence and severe preeclampsia presenting late in pregnancy. TRAP Sequence is one of the manifestations of abnormal vascular anastomosis (twin to twin transfusion syndrome) that occur in monochorionic twins. The occurrence rate is about 1 in 35000 deliveries, and in 1% of monozygotic twin pregnancies [1]. Females are predominantly affected. This case was diagnosed late because the first USS at 15 weeks woefully missed it. This calls for more training and re-certification of Fetal Medicine Specialists.

It has been hypothesized that the TRAP is caused in the embryo by a large artery-to-artery placental shunt, often also accompanied by a vein-to-vein shunt [2].

In normal fetus, blood enters the fetus through the umbilical vein and exits through the umbilical artery, while in acardiac twin, blood enters via the umbilical artery and leaves via umbilical vein, hence the name Twin Reversed Arterial Perfusion (TRAP) Sequence. The perfusion pressure of the donor twin overpowers that in the recipient twin, who thus receives reverse blood flow from its twin sibling [3].

Acardiac twin does not send blood to the placenta but receives blood from the pump twin via the abnormal vascular anastomosis in the placenta. Blood supply from the pump twin enters the acardiac twin through umbilical artery which consists mainly of deoxygenated blood. The blood reaching the recipient twin preferentially goes to the iliac vessels and thus perfuses only the lower part of the body, leading to disruption or deterioration of growth and development of the upper body. The pump twin suffers from congestive cardiac failure and hydrops as a result of pumping blood to the acardiac twin. The pump twin has 50-70% mortality without an intervention while the acardiac twin has 100% mortality [4]. The importance of early identification, follow up and treatment would markedly improve the survival rate of the pump twin and cannot be over emphasized.

Acardiac twinning is classified according to the degree of cephalic and truncal maldevelopment [5-6].

The first type is Acardius acephalus, where no cephalic structures are present (head and upper

extremities are lacking). This is the most common variety of Acardia. Our case was a case of Acardiac acephalus.

The second type is Acardius anceps where some cranial structure and neural tissue or brain tissue is present. The body and extremities are also developed. It is a highly developed form of Acardia.

The third is Acardius acormus with cephalic structure but no truncal structures. It has a head without a body. The umbilical cord is attached to the head. This is the rarest form of Acardia.

The fourth type is Acardius amorphous with no distinguishable cephalic or truncal structure. It is least developed and not recognizable as human form with minimal development. This differs from teratomas only by its attachment to an umbilical cord.

The diagnosis of acardiac twin should be made early in pregnancy using ultrasound and by Doppler studies where reversal of flow in the umbilical arteries and absence of heart can be noted. Early diagnosis and diligent follow up is pertinent to reduce possible complications.

Sullivan et al. advocated expectant management with close fetal surveillance as a management option [7]. Survival was reported for 9 of 10 donor twins managed.

Minimal invasive procedures like percutaneous insertion of helical metal coil to induce thrombogenesis in the single umbilical artery of the acardiac twin has also been advocated [8-9]. Quintero and colleagues have also reviewed methods of in utero treatment of acardiac twinning in which the goal is interruption of the vascular communication between the donor and recipient twins [10]. They also have described successful use of transabdominal fetoscopy to ligate the umbilical cord of 11 acardiac twins at approximately 21 weeks Tsao and colleagues reported the survival of 12 of 13 donor twins when, under ultrasonographic guidance, a 14-gauge radioablation needle was used to cauterize the umbilical vessels and terminate blood flow to the recipient twin at the site of insertion into the umbilicus [11]. Ablation of the abnormal vessels can be performed between 18 to 24 weeks. Without treatment, the donor or "pump" twin has been reported to die in 50 to 70 percent of cases [4]. Cerebral pathology in the survivor most likely results from acute hypotension at the time of the death of the co-twin [12-13]. A less likely cause is from emboli of thromboplastic material originating from the dead fetus. Blocking the vessels by coagulation using Nd: Yag laser and radiofrequency ablation [14] under ultrasound guidance have been considered now as the first line of treatment.

4. CONCLUSION

Early diagnosis helps in the determination of chronicity of twin pregnancy. Doppler and ultrasound scan can be used to make diagnosis of acardiac twin as early as the first trimester. Treatment at the appropriate time improves the survival of the pump twin. Our case is therefore an exception in terms of the mortality of the donor, however, a long time follow up is required.

CONSENT

All authors declare that 'written informed consent was obtained from the patient for publication of this paper and accompanying image.

ETHICAL APPROVAL

All authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Saritha S, Anjankar SS. Twin reversed arterial perfusion sequence (trap sequence). Int J Anat Res. 2013;03:140-4.
2. Nath A, Nair S, Pal R, Chakraborty A. Acardiac twin- A case report. IAIM. 2016; 3(3):158-162.
3. Jones KL. Smith's Recognizable Pattern's of Human Malformation, 5th ed. Philadelphia, Saunders. 1997;568.
4. Prameela RC, Ranganath P, Niveda S, Pooja GY, Prajwal S. A Rare Case of Acardiac Twin: A Case Report. Int J Sci Stud. 2014;2(8):254-257.
5. Mohanty C, Mishra OP, Singh CP Das BK, Singla PN. Acardiac Anomaly Spectrum. Tetralogy. 2000;62(5):356-9.

6. Napolitano FD. Schreiber I. The acardiac moster: A review of the word literature and presentation of two cases. Am J Obstet Gynecol. 1960;80:582–589.
7. Sullivan AE, Varner MW, Ball RH, Jackson M, Silver RM. The management of acardiac twins: A conservative approach. Am J Obstet Gynecol. 2003;189:1310-1313.
8. Tan TY, Sepulveda W. Acardiac twin: A systematic review of minimally invasive treatment modalities. Ultrasound Obstet Gynecol. 2003;22:409-19.
9. Weisz B, Peltz R, Chayen B, Oren M, Zalel Y, Achiron R, Lipitz S. Tailored management of twin reversed arterial perfusion (TRAP) sequence. Ultrasound Obstet Gynecol. 2004;23 (5):451-5.
10. Quintero RA, Reich H, Puder KS, Bardicef M, Evans MI, Cotton DB, Romero R. Umbilical-cord ligation of an acardiac twin by fetoscopy at 19 weeks gestation. N Engl J Med. 1994;330(7):469–71.
11. Tsao K, Feldstein VA, Albanese CT, Sandberg PL, Lee H, Harrison MR, Farmer DL: Selective reduction of acardiac twin by radiofrequency ablation. Am J Obstet Gynecol. 2002;187(3):635-40.
12. Benirschke K, Kim CK. Multiple pregnancy, 1. N. Engl. J. Med. 1973;288: 1276-1284.
13. Fusi L, Fisk N, Talbert D, Gau G, Rodeck C . When does death occur in an *acardiac twin*? Ultrasound diagnostic difficulties. J Perinat Med. 1990;18(3):223-227.
14. Stamation F, Muresan D, Caracosteae G, Kovacs T. Advances in ultrasonic assessment of acardiac. Donald Sch J Ultrasound Obstet Gynecol. 2011; 5:213-8.

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