



Twin Reversed Arterial Perfusion Sequence in a Triplet Natural Pregnancy: Case Report from Korle Bu Teaching Hospital, Accra, Ghana and a Review of the Literature

Séquence de perfusion artérielle inversée dans un triplet lors d'une grossesse naturelle. Cas clinique à l'Hôpital Universitaire de Korle Bu, Accra, Ghana et revue de la littérature

E. V. Badoe[†], A. Darko[‡], B. H. Atuguba[§]

ABSTRACT

BACKGROUND: Twin reversed arterial perfusion sequence is probably the most severe malformation found in humans. It is also called acardia, acardiac twinning or acardius acephalus and is a very rare condition that occurs in monozygotic twins and rarely in triplet pregnancy. It has not been described in the West African Medical literature. A case was recorded in a twin delivery at the labour ward four years ago in the same hospital but the underlying diagnosis was not made.

OBJECTIVE: To describe a rare case of twin reversed arterial perfusion sequence occurring in a triplet pregnancy and review the current literature.

METHODS: A case was evaluated at the delivery ward, Department of Obstetrics and the Neonatal unit of the Korle Bu Teaching Hospital upon information from the obstetric team that a monster had been born from a triplet set. Full documentation was done.

RESULTS: The case documented was a twin reversed arterial perfusion sequence in a triplet pregnancy.

CONCLUSION: This rare condition has been well characterized and Obstetricians and Paediatricians should now be able to make an accurate diagnosis. *WAJM* 2013; 32(1): 73–75.

Keywords: Twin reversed arterial perfusion (TRAP) sequence, Acardiac acephalus.

RÉSUMÉ

CONTEXTE: La séquence de perfusion artérielle inversée chez les jumeaux est probablement la plus sévère des malformations dans l'espèce humaine. On l'appelle, acardia, jumeaux acardiaques ou acardius acephalus et elle constitue une affection rare survenant chez des jumeaux monozygotes et rarement dans la grossesse trimellaire. Elle n'a pas été décrite dans la littérature Ouest Africaine. Un cas a été décrit dans un accouchement gémellaire dans une salle d'accouchement il y'a de cela 4 ans dans le même hôpital mais le diagnostic sous-jacent n'a pas été posé.

OBJECTIF: Décrire un cas rare de séquence de perfusion artérielle inversée survenant dans une grossesse trimellaire et faire une revue de la littérature actuelle.

MÉTHODES: Un cas a été évalué dans la salle d'accouchement du département d'Obstétric et de l'unité de Néonatalogie de l'Hôpital Universitaire de Korle Bu après information par l'équipe d'obstétrique qu'un monstre était né d'un groupe de triplet. Une documentation complète a été faite.

RÉSULTATS: Le cas documenté était une séquence de perfusion artérielle inversée dans une grossesse de triplet.

CONCLUSION: Cette affection rare a été bien caractérisée et à présent les Obstétriciens et Pédiatres doivent pouvoir faire un diagnostic juste. *WAJM* 2013; 32(1): 73–75.

Mots clés: Séquence de perfusion artérielle inversée chez des jumeaux, Acardiaque acéphalique

INTRODUCTION

The term twin reversed arterial perfusion syndrome (TRAP) was first defined by Gruenwald in 1942 as probably accounting for an acardiac foetus and a form of twin to twin transfusion syndrome.¹ This condition is very rare and occurs in 1 in 35,000 deliveries, 1 in 100 monozygotic twins, rarely in triplet pregnancy and even in quintuplet gestations.² It has not been described in the West African Literature to the best of our knowledge. We present a case of triplets where one represented the TRAP syndrome or acardiac acephalus with twice the weight of the pump twin who did not appear to have any health problems. A review of the literature is also presented.

CASE REPORT

A 29-year old mother of one child was referred to Korle Bu Teaching Hospital on the 24th of May, 2012 from La Paz Community clinic at 35 weeks and 2 days gestation on account of imminent eclampsia. On arrival at Korle Bu Hospital, she was assessed and prepared for Emergency Caesarian section. The findings were that of eclampsia, unfavourable cervix and twin gestation. The history obtained showed that mother was a regular attendant at La Paz community clinic. She booked in at 28 weeks and had a late scan done at that time which showed a twin foetus. First twin was cephalic and the 2nd a breech presentation. Fetal heart beat for the twins was present and the average gestational age was 28 weeks. The expected date of delivery was recorded as 28th June, 2012. Her antenatal work up screen was negative for HIV, Hepatitis B and Sickling. She was a mother of a three year old son and parents lived in a suburb called Sowutuom. She was a graduate of a senior secondary school and worked as a seamstress. She did not smoke or take alcohol and the only medications taken in pregnancy were folic acid, fersolate, calcium supplements and Vitamin B complex. No fertility drugs were taken. There was no history of hypertension or diabetes in the family. The father of the infants was a driver at an Insurance firm. By the time of the emergency caesarean section the maturity of the pregnancy

was noted as 35 weeks and two days. The indications were imminent Eclampsia (BP was 190/110 mm Hg), urine protein with 3+ of urine, blurred vision, epigastric discomfort, headaches, with an unfavourable cervix and twin gestation. The findings were triplets. 1st triplet was in the cephalic position and weighed 1.8kg with an Apgar score of 7/10 and 7/10 at 1 and 5 minutes respectively. The second triplet was in the breech position and weighed 3.7 kg and was described as a monster. It was stillborn and severely malformed. It had male genitalia though not completely formed. The upper portion of the torso looked globular and swollen with a cystic feel. There was no heart, lungs, and upper limbs present. Rudimentary facial features of ears, eye and mouth were present. The lower limbs were both present with four digits in each including the big toes. The head looked poorly developed with a rudimentary face and ears. See Figure 1 and 2. The third triplet weighed 1.7kg and had good Apgar scores of 8 and 8 respectively at 1 and 5 minutes. The placenta was delivered whole and the lobes and membranes were complete weighing

1900g. It was a Dichorionic Triamniotic type of Placenta. The umbilicus appeared to have two veins and an artery. The surviving infants were sent to the Neonatal Intensive Care Unit of the Korle Bu Teaching Hospital where they made good progress. At the time of writing the two surviving triplets had been discharged home and were stable.

DISCUSSION

The acardiac malformation, characterised by failure of heart development and severe dysmorphogenesis was first described by Benedetti in a book published in 1533.³ The case presented are triplets. The chances of acardiac development in a multiple pregnancy of higher order are somewhat greater and this occurs in about 1 in 30 monozygotic triplets.⁴ With assisted reproduction on the increase in West Africa, Obstetricians and Paediatricians are likely to see this malformation described. There are no known maternal risk factors for acardiac development.⁵ There is a slight preponderance of female acardiac fetuses.⁶ Our case was male. The other two triplets survived though the perinatal mortality rate for the normal fetus may be as high as 50–75%. The cause of death is usually prematurity or congestive cardiac failure.⁷ The morphologic classification of the acardiac twin was described in 1925 as follows. Acardia amorphous, the least developed which is not recognisable as a human form. Acardia myelacephalus resembles the amorphous type except for the presence of rudimentary limb formation. See Figure 3 representing the first case recorded. Acardius acephalus,



Fig. 1



Fig. 2

Figs. 1 and 2 showing Acardia Acephalus: Legs Formed with a Cystic Trunk and Rudimentary Face.



Fig. 3: showing Acardia Myelacephalus – Rudimentary Legs and Poorly Developed Trunk

the commonest type which has a missing head, part of the thorax and upper extremities, acardiac acephalus in which there is an absence of the fetal head and thoracic organs. Acardius acormus is the rarest type and lacks a thorax with only a rudimentary head with the umbilicus inserting into the head and connecting directly to the placenta.⁸ The case presented was acardius acephalus. The anomaly is thought to result from an umbilical artery to artery anastomosis between twin fetuses and veno-venous placental anastomoses leading to circulatory problems in one twin. The acardiac twin loses direct vascular connection with the placental villi and receives its entire blood supply from the pump twin. The poorly oxygenated blood perfuses the lower part of the fetus first and the upper foetal structures undergo secondary atrophy and the resultant "twin" has been described as an acardiac monster.⁹ The hallmark for the diagnosis of an acardiac twin by ultrasound is the absence of identifiable cardiac pulsation. Other characteristic features are poor definition of the head, the trunk region and the upper extremities. The lower extremities are usually present and there is marked tissue oedema and abnormal cystic areas in the upper part of the body.¹⁰ The use of pulsed Doppler sonography has enabled the demonstration of reversed flow through the umbilical artery of the acardiac twin.¹¹ This was not done in this case. When twin reversal arterial perfusion is diagnosed venous Doppler ultrasound can be used to monitor the pump twin for signs of cardiac failure and facilitate delivery timing if a viable gestation has been reached.¹² Prior to 24 weeks gestation, cord ligation of the

acardiac twin has been used with success (up to 80% survival of the pump twin) in specialised centres.¹³ In the present case the baby was saved spontaneously. It is important to exclude a chromosomal abnormality before offering any procedure by amniocentesis in the TRAP sequence as about 10% of pump twins have documented abnormalities. Regarding the recurrence risk regarding genetic counseling the overall recurrence risk to a patient's sibling; in this case one of the triplets is about 1 in 10000. The patient's offspring is not applicable because it is a lethal condition.¹⁴ It would have been desirable to do a post mortem study on the acardiac monster as well as chromosomal studies. Unfortunately the specimen got discarded before that procedure could take place.

Conclusion

This bizarre malformation has been explained and it is hoped that Obstetricians in particular would report more on this condition in the sub region. Twinning rates are high in West Africa especially in Nigeria. The "monsters" should not be discarded but every effort must be made to characterise the malformation seen. Acardia is a rare complication of monochorionic twinning and multiple pregnancy.

REFERENCES

1. Ewa C, Maria R L, Stawerski P, Piotr K, Andrzej. Acardiac foetus N presentation of 3 cases. *Med Sci Monit* 1999; **5**: 953–7.
2. Aggarwal N, Suri V, Saxena SV, Malhotra S, Vashita K, Saxena Ak. Acardiac acephalus twins: A case report and review of the literature. *Acta Obstet Gynecol Scand.* 2002; **81**: 983–984.
3. Benedetti A. De Singulis corpori humani morbis a capite a pedis. Venice. 1533
4. James W.H. A note on the epidemiology of acardiac monsters. *Teratology.* 1977; **16**: 211–216.
5. Sato T, Kaneko K, Konuma S, Sato I, Tamada T. Acardiac anomalies: review of 88 cases in Japan. *Asia Oceania Journal of Obstetrics and Gynaecology.* 1984; **10**: 45–52.
6. Kaplan C, Benirschke K. The acardiac anomaly: new case reports and current status. *Acta Genetica Medica Gemellologica.* 1979; **28**: 51–59.
7. Lewi L, Valencia C, Gonzalez E, et al. The outcome of twin reversed arterial perfusion sequence diagnosed in the first trimester. *American Journal of Obstetrics and Gynecology.* 2010; **203**: 13. e1-e4.
8. Simonds JP, Gower GA. Fetus amorphous. *Surgery, Gynecology and Obstetrics.* 1925; **41**: 171–175.
9. Van Allen MI, Smith DW, Shephard TH. Twin reversed arterial perfusion (TRAP) sequence. A study of 14 twin pregnancies with acardius. *Semin Perinatol.* 1983; **7**: 285–93.
10. Nicolaidis P, Nasrat H, Tannirandom Y. Fetal acardia: aetiology, pathology and management. *J Obstet Gynaecol* 1990; **10**: 518–525.
11. Pretorius DH, Manchester D, Barkin S, Parker S, Nelson TR. Doppler ultrasound of twin transfusion syndrome. *J Ultrasound Med* 1998; **7**: 117–24.
12. Gibson J, Cameron A. Complications of monochorionic twins. *Current Paediatrics.* 2008; **18**: 12 568–573.
13. Quintero RA, Chmait RH, Murakoshi T, Barigyne O, Chappel L, Fisk N. Surgical management of twin reversed arterial perfusion sequence. *Am J Obstet Gynecol;* 2006; **194**: 982–991.
14. Chen H. Atlas of Genetic Diagnosis and Counseling. 1st Ed. Totowa (New Jersey). Humana Press Inc.; 2006: P 1–6.