Case Report

TRAP Sequence in an Acardiac Twin

Athita Chanthasenanont MD*,
Densak Pongrojpaw MD*

** Department of Obstetrics and Gynecology, Faculty of Medicine, Thammasat University, Pathumthani*

Case Report: *The authors present a case of an acardiac twin gestation diagnosed sonographically at 20 weeks with evidence of twin reversed-arterial perfusion (TRAP) sequence. The acardius appeared to be less than half the estimated weight of the pump twin. The acardius was identified as an amorphous mass, without extremities. The parents chose expectant management rather than surgical intervention. Observation by serial sonographic and color Doppler evaluation was performed. At 34 weeks, a healthy infant was delivered by cesarean section after showing signs of fetal congestive heart failure (CHF). The acardius was 2/3 the size of the infant at delivery.*

Conclusion: *Expectant management with close antepartum surveillance deserves consideration in cases of monozygotic twins with TRAP sequence. Neonatal mortality of the pump twin diagnosed antenatally may be considerably less than reported.*

Keywords: *Acardiac twin, TRAP sequence, Fetal heart failure*

J Med Assoc Thai 2007; 90 (4): 819-22
Full text. e-Journal: *http://www.medassocthai.org/journal*

Case Report

A 19-year old primagravida woman was first seen in the institution at 9 weeks' gestation. Her past history and family history were unremarkable for twins or perinatal issues. At 20 weeks' gestation, transabdominal ultrasonographic screening was performed. It revealed a twin gestation with a single placental mass and no dividing membrane. A normal size for dates twin was associated with an acardiac, acranial amorphous twin. The acardius had a two-vessel cord with twin reversed-arterial perfusion via the pump twin with anastomosis within the placenta. The size of the acardius appeared to be less than half of the size of the combined abdomen and thorax of the pump twin. The parents were counseled about the risks and benefits between expectant management and surgical intervention. Serial sonography and Doppler velocimetry was performed weekly.

At 34 weeks' gestation, the free loop of the umbilical artery of the pump twin revealed a marked abnormal S/D ratio (> 99th percentile), tachycardia, and an increased cardiothoracic circumference ratio (0.65). The acardius had also increased in size relative to the pump twin, who had maintained a normal weight for gestational age with an estimated fetal weight of 2,096 grams. There was no pericardial or pleural effusion, or ascites. A non-stress test was reactive. A course of dexamethasone was given. The patient was admitted and delivered by low transverse cesarean section following a normal shake test for fetal lung maturity (positive in 3 tubes). The pump twin was 2,085 grams with apgar scores 8 and 10 at 1 and 5 minutes. The acardiac twin was an oblong mass 19 x 14 cm. in diameter at 1,395 grams. It had a two-vessel cord, gastroschisis and a skin bud with the appearance of upper and lower lips and some hair. The pump twin developed transient heart failure at the first hour of life. He was treated with two doses of furosemide and clinically improved. The patient and her baby were discharged on the fourth day after delivery.

Discussion

Acardiac twin is a rare complication of monozygotic twin pregnancies occurring in 1% of monochorionic twin gestations with an incidence of 1 in

Correspondence to : Chanthasenanont A, Department of Obstetrics and Gynecology, Faculty of Medicine, Thammasat University, Pathumthani 12120, Thailand. E-mail: athitaka @gmail.com

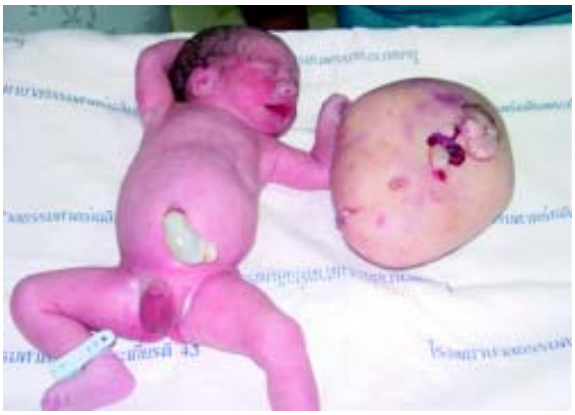

Fig. 1 The normal pump twin and acardiac twin

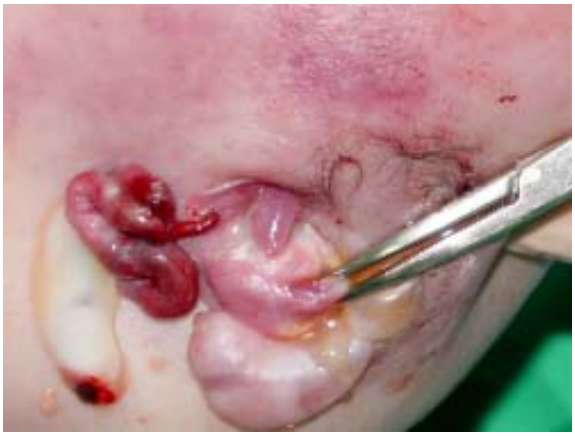

Fig. 2 Enlargement showing a small gastroschisis, some hair and the appearance of lips and a tongue

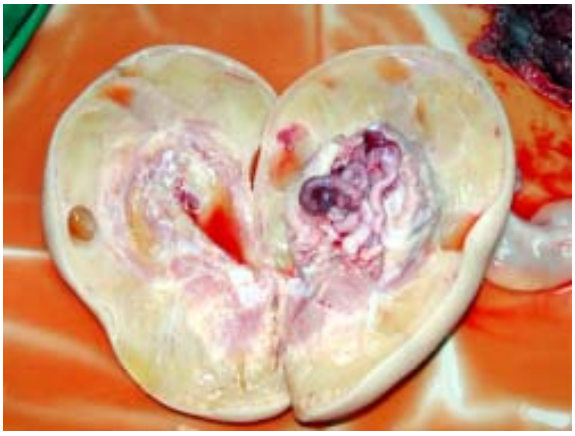

Fig. 3 Dissected acardius showing ribs, cartilaginous vertebrae, small bowel and edematous tissue

35,000 births. Without treatment, the reported perinatal mortality of the normal or pump twin is 50-70%[1-5].

The TRAP sequence is a consequence of arrested development during the formation of the endocardial tubes at approximately three weeks after fertilization. Profusion to the amorphous twin results from cord or and direct arterio-arterial and veno-venous placental anastomoses[2,6]. It is more common in nulliparous women and in monoamniotic monochorionic pregnancy[6]. The acardiac twin is typically grossly abnormal with gross edema and limb reduction anomalies, particularly of the upper part of the body and with an absent or rudimentary heart. Failure or disrupted growth of the head is called acardius acephalus; a partially developed head with identifiable limbs is called acardius myelacephalus. Failure of any recognizable structure in the recipient twin is called acardius amorphous[1]. Perinatal mortality of the pump twin is the result of CHF, polyhydramnios and preterm delivery. This is especially true when the acardiac twin is greater than 50% of the size of the pump twin by estimated weight[6].

The pump twin is typically a structurally normal, but may develop CHF over gestation with cardiomegaly, hepatosplenomegaly, pericardial and pleural effusions, ascites, and polyhydramnios. If there are two amnions, polyhydramnios is seen with the pump twin and oligohydramnios with the acardiac twin[5]. The recipient twin has severe anomalies and may have an anencephaly. Its upper trunk and neck area will be thickened. Upper limbs are sometimes absent. Club feet and absent toes are often seen. Limb movements are sometimes visible in the acardiac fetus, but may be a result of movement by the pump twin. The acardiac fetus has a two-vessel umbilical cord[6].

The optimal management of acardiac twin pregnancies is controversial. The surgical goal of the treatment is to interrupt the blood flow to the acardiac twin without harming the pump twin. Unfortunately, as the fetuses share a common placenta and have vascular communications between them, damage to the pump twin may occur. Management options include elective termination, observation with close antepartum surveillance and surgical interventions[2,5-13].

Various therapeutic options have been proposed, including expectant management, control of amniotic fluid volume by repeated amniocentesis or indomethacin therapy in the mother, administration of digoxin to the mother to treat fetal CHF in the pump twin, and selective preterm delivery of the acardiac twin by hysterotomy[12]. These interventions are usually

feasible only after 24 weeks of gestation, and they are hazardous for both the mother and the normal fetus. Another approach is to stop the perfusion of the acardiac twin by the pump twin. This has been done with varying degrees of success by percutaneous injection of thrombogenic coils or sclerosing agents to occlude the umbilical cord of the acardiac twin; by cord ligation performed at hysterotomy or with endoscopic or ultrasound guidance; by thermocoagulation; and by fetoscopic laser coagulation[9].

Color Doppler velocimetry may help to determine site of vascular connection and assess circulatory health of the pump twin. Small resistive index differences are associated with poor outcome, including CHF and central nervous system morbidity[14]. Serial sonography with color Doppler should be performed every 1-2 weeks to assess growth status in each twin as well as for indicators of CHF in the pump twin[6].

The best time to intervene and the best mode of intervention are not yet known[2,5-13]. With an increase in antenatal diagnosis, outcomes in expectantly managed cases may be better than reported[15]. Expectant management of the pregnancy is maintained with serial ultrasonographic and color Doppler examination.

In the presented case, the pregnancy was terminated at 34 weeks of gestation after signs of CHF and abnormal Doppler studies appeared, and amniocentesis for lung maturity test was performed. The outcome in this case was excellent.

Acknowledgment

The authors wish to thank Dennis C. Wood, RDMS, RCPT, and the fetal cardiology group of Thomas Jefferson University, Pennsylvania, USA, for editorial assistance.

References

1. Cunningham FG, Grant NF, Leveno KJ, Gilstrap LC III, Hauth JC, Wenstrom KD. Williams obstetrics. 21st ed. New York: McGraw-Hill; 2001.
2. Petrikovsky BM. Fetal disorders: diagnosis and management. New York: Wiley-Liss; 1999.
3. Sepulveda WH, Quiroz VH, Giuliano A, Henriquez R. Prenatal ultrasonographic diagnosis of acardiac twin. J Perinat Med 1993; 21: 241-6.
4. Moore TR, Gale S, Benirschke K. Perinatal outcome of forty-nine pregnancies complicated by acardiac twinning. Am J Obstet Gynecol 1990; 163: 907-12.
5. Pinet C, Colau JC, Delezoide AL, Menez F. Acardiac twins. J Gynecol Obstet Biol Reprod (Paris) 1994; 23: 85-92.
6. Smith AM. Abnormalities of twin pregnancies. In: Twining P, McHugo JM, Pilling DW, editors. Textbook of fetal abnormalities. London: Churchill Livingstone; 2000: 404-5.
7. Sogaard K, Skibsted L, Brocks V. Acardiac twins: pathophysiology, diagnosis, outcome and treatment. Six cases and review of the literature. Fetal Diagn Ther 1999; 14: 53-9.
8. Robie GF, Payne GG Jr, Morgan MA. Selective delivery of an acardiac, acephalic twin. N Engl J Med 1989; 320: 512-3.
9. Porreco RP, Barton SM, Haverkamp AD. Occlusion of umbilical artery in acardiac, acephalic twin. Lancet 1991; 337: 326-7.
10. McCurdy CM Jr, Childers JM, Seeds JW. Ligation of the umbilical cord of an acardiac-acephalus twin with an endoscopic intrauterine technique. Obstet Gynecol 1993; 82: 708-11.
11. Muhlhaus K, Beisler G. Acardius - prenatal diagnosis and pregnancy in 2 twin pregnancies. Z Geburtshilfe Perinatol 1986; 190: 98-103.
12. Ginsberg NA, Applebaum M, Rabin SA, Caffarelli MA, Kuuspalu M, Daskal JL, et al. Term birth after midtrimester hysterotomy and selective delivery of an acardiac twin. Am J Obstet Gynecol 1992; 167: 33-7.
13. Donnenfeld AE, van de WJ, Craparo F, Smith CS, Ludomirsky A, Weiner S. The normal fetus of an acardiac twin pregnancy: perinatal management based on echocardiographic and sonographic evaluation. Prenat Diagn 1991; 11: 235-44.
14. Dashe JS, Fernandez CO, Twickler DM. Utility of Doppler velocimetry in predicting outcome in twin reversed-arterial perfusion sequence. Am J Obstet Gynecol 2001; 185: 135-9.
15. Sullivan AE, Varner MW, Ball RH, Jackson M, Silver RM. The management of acardiac twins: a conservative approach. Am J Obstet Gynecol 2003; 189: 1310-3.

รายงานสตรีตั้งครรภ์เด็กแฝดไม่มีหัวใจ

อธิตา จันทเสนานนท์, เด่นศักดิ์ พงศ์โรจน์เผ่า

รายงานสตรีตั้งครรภ์เด็กแฝดที่ไม่มีหัวใจรายนี้ ได้รับการวินิจฉัยด้วยคลื่นเสียงความถี่สูงขณะอายุครรภ์ 20 สัปดาห์ ทารกรายที่ปกติมีขนาดใหญ่กว่าทารกแฝดรายที่ไม่มีหัวใจเท่าตัว โดยทารกแฝดรายที่ไม่มีหัวใจ มีลักษณะเหมือนก้อนเนื้อ ไม่มีแขนขา สตรีตั้งครรภ์รายนี้ได้รับการรักษาแบบประคับประคองและเฝ้าระวังภาวะแทรกซ้อนโดยการตรวจด้วยคลื่นเสียงความถี่สูงเป็นระยะ ๆ เพื่อประเมินทารกในครรภ์และการไหลเวียนเลือดของทารก สตรีตั้งครรภ์รายนี้ได้รับการผ่าตัดคลอดบุตรเมื่ออายุครรภ์ 34 สัปดาห์ เนื่องจากทารกแฝดคนปกติมีลักษณะภาวะหัวใจวายจากการตรวจด้วยคลื่นเสียงความถี่สูง ทารกเพศชาย น้ำหนักแรกเกิด 2,085 กรัม แข็งแรงดี โดยการรักษาแบบประคับประคองและเฝ้าระวังนี้ เป็นอีกหนึ่งทางเลือกในการดูแลรักษาสตรีตั้งครรภ์ที่มีภาวะดังกล่าว ซึ่งได้ผลเป็นที่น่าพอใจ