Special Article

Acardiac Twin

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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. Without treatment, the perinatal mortality of the pump twin is about 50-70%. The optimal management of acardiac twin pregnancies is controversial. Management options include elective termination, observation with close antepartum surveillance (serial cardiotocography, ultrasonography and echocardiography) and surgical interventions

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Acardiac twin or twin reversed-arterial perfusion (TRAP) sequence is a rare complication of monozygotic twin pregnancies that was first described by Benedetti in 1533[1]. It occurs in 1% of monochorionic twin gestations with an incidence of 1 in 35,000 births[1-8]. It was previously thought to occur only in monochorionic pregnancies, but recently French et al described a diamnionic dichorionic gestation which was confirmed by gross and histological examination[9]. It is more common in nulliparous women[10] and three times higher among monozygotic triplets than among twins[11]. Sixty percent of acardiac twins are diamniotic[12]. There is a slight female excess among them[12]. The incidence of chromosomal abnormality is 9% with the recurrence rate of 1 in 10,000. There have been scattered reports of chromosomal anomalies in the donor fetus, including autosomal trisomies[13] and the Klinefelter's syndrome[14]. This syndrome is a consequence of primary or secondary cardiac development disruption and direct arterioarterial and venovenous placental anastomoses[3-5]. Theories of origin of acardius have changed throughout the centuries. Prior to the eighteenth century, the anatomic and physiologic relationships of acardiac twins were obscured by superstition and were thought to be due to the disturbed imaginings of expectant mothers. During the eighteenth century, the defective germ plasma theory proposing that acardia was determined at the time of conception was introduced. During the following century, this concept was refuted by the observations that acardius occurred in monochorionic twins of monozygotic origin and that the anastomosis between the vessels of these twins were found to be primarily arterio-arterial type so that the circulation in the acardiac twin was reserved making the second twin a parasite of the first. The primary etiology was thought to be due to an accidental relationship of the placental vessels. During the twentieth century reports of studies of the embryopathology and embryogenesis of the acardiac twins suggested that the primary deficiency is a failure of the heart to develop and that the acardiac twin survives in utero only where an anastomosis is formed between the vessels of the two umbilical cords. The low oxygen content of the blood perfusing the twin results in a number of characteristic anomalies including total or partial absence of cranial vault, holoprosencephaly, anencephaly, anophthalmia, absent facial structures, microphthalmia, cleft lip, cleft palate, diaphragmatic defects, absent or rudimentary limbs, absent heart and lungs, esophageal atresia, ventral wall defects, ascites, absent liver and gallbladder, skin edema and cystic hygroma[15,16]. The lower part of the body received better oxygenated venous blood though the hypogastic artery and consequently the most severe abnormalities are in the upper part of the body[11,17]. Recently, Petersen et al[18] reported a case

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of acardiac twins with a nearly normal external appearance and an almost normally developed brain. 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 to form is acardius amorphous and developed head but no body is called acardius acormus, this is the rarest type[4]. Morphologic categories of this syndrome have no effect on management or prognosis[19]. Typically, the pump twin is anatomically normal.

Without treatment, the perinatal mortality of the pump twin is about 50-70%[4,5,7,20]. The goal of the treatment of acardiac twin is to interrupt the blood flow to the acardiac twin without harming the pump twin. Unfortunately, as the fetus share a common placenta and have vascular communications between them, damage to the pump twin may occur. Thus, optimal management of acardiac twin pregnancies is controversial. Management options include elective termination, observation with close antepartum surveillance (serial cardiotocography, ultrasonography and echocardiography) and surgical interventions[3,5,6,20-26].

Perinatal mortality of the pump twin is the result of congestive heart failure, polyhydramnios and preterm delivery. The mortality rate of the pump twin is about 64% when the acardiac twin is greater than 50% of the size of the pump twin and about 90% when the acardiac twin is greater than 75% of the size of the pump twin by estimated weight[3]. Weight of the acardiac twin is estimated with 1.21 (length^2)-1.66 (length)[3].

Ultrasonography has assumed increasing importance in prenatal diagnosis of congenital anomalies. Using this technique, intrauterine diagnosis of acardiac twins has been made during the first trimester of pregnancy. Ultrasonography in acardiac twins reveals a normal pump twin with an acardiac twin. The pump twin may show signs of hydrops with hepatosplenomegaly, cardiomegaly, ascites and pleural effusion. 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 acardiac fetus. Polyhydramnios is common. If there are two sacs, polyhydramnios is found with the pump twin and oligohydramnios with the acardiac twin[2]. The acardiac fetus usually has a two-vessel umbilical cord[3].

Color Doppler velocimetry may help to determine the site of the vascular connection and assess circulatory health of the pump twin. The pulsatility index (PI) in the acardiac twin's umbilical artery is significantly lower than that of the pump twin[13]. The S/D ratio of the reversed flow in the acardiac twin umbilical artery has been reported to be elevated and provides a measure of the systemic vascular resistance of the acardiac fetus itself [13,27,28]. Small resistive index differences and a rapid growth rate of the mass are associated with poor outcome, including cardiac failure and central nervous system hypoperfusion[28,29]. The cardiothoracic ratio and presence of cysts or of a rudimentary heart don't correlate with outcome. Serial ultrasonography with color Doppler should be performed every 1-2 weeks to assess growth status, hydrops or signs of heart failure in the pump twin[3].

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 heart failure in the pump twin, and selective preterm delivery of the acardiac twin by hysterotomy. Ash et al reported a case of acardiac twins complicated by hydramnios managed by maternal administration of indomethacin with good outcome[30].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, which has been done with varying degrees of success by radiofrequency ablation, percutaneous injection of thrombogenic coils or sclerosing agents to occlude the umbilical cord of the acardiac twin; by umbilical cord ligation performed at hysterotomy or with endoscopic or ultrasound guidance; by thermocoagulation; and by fetoscopic laser coagulation. Thrombosis of one of the vessels of the umbilical cord has been achieved under ultrasound guidance. The thrombosis must be carried out in the artery, as blood in this vessel flows towards the acardiac twin. If the vein is thrombosed, there is a risk of emboli traveling back to the pump twin. The umbilical cord ligation is a reliable technique and can be performed with or without the need of an endoscope. Fetoscopic cord ligation was firstly reported in 1993[23]. The pump twin is not affected by the procedure. It is associated with 70-80% of success rate. Risks of the procedure include technical impossibility (7.6%), premature rupture of membranes (10%) and bleeding. Recently, Norwitz ER et al[31] described prenatal planning for neonatal surgery using three- dimensional modeling with magnetic resonance imaging. The best time to intervene and the best mode of intervention are not yet known[3,5,6,20-26].

Because of increased antenatal diagnosis, the outcome in expectantly managed cases may be better than previously reported[32]. Sullivan AE et al[32] reported the success rate of conservative treatment was 8 in 9 cases, the mean gestational age at delivery was 34.2 weeks and the mean weights of the pump and acardiac twin were 2,279 and 1,372 grams.

References

1. Loughead JR Jr, Halbert DR. An acardiac amorphous twin presenting soft tissue dystocia. South Med J 1969; 62: 1140-2.
2. 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.
3. Smith A P M. Abnormalities of twin pregnancies. In: Peter T, Josephine M, David WP,editors. Textbook of Fetal Abnormalities. 1st ed. London: Churchill Livingstone, 2000: 404-5.
4. Cunningham FG, Grant NF, Leveno KJ, Gilstrap LC III, Hauth JC, Wenstrom KD. Williams Obstetrics. 21st ed. New York: McGraw-Hill, 2001: 783-4.
5. Boris M. Fetal disorders: Diagnosis and Management. 1st ed. New York: Wiley-Liss,1999: 233-6.
6. Zwahr C, Petri E, Kirsch M, Pawlowski P. Monozygotic multiple pregnancies with acardiac anomalies. Z Geburtshilfe Neonatol 1998; 202: 80-5.
7. Sepulveda WH, Quiroz VH, Giuliano A, Henriquez R. Prenatal ultrasonographic diagnosis of acardiac twin. J Perinat Med 1993; 21: 241-6.
8. Moorthy NLN, Agarwal C, Gupta M,Chandrakala A, Mohan G. Case report: acardiac parabiotic twin. Indian J Rad 1998; 8:91-2.
9. French CA, Bieber FR, Bing DH, Genest DR. Twins, placentas, and genetics: acardiac twinning in a dichorionic, diamniotic, monozygotic twin gestation. Hum Pathol 1998; 29: 1028-31.
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.
11. Hanafy A, Peterson CM. Twin-reversed arterial perfusion (TRAP) sequence: case reports and review of literature. Aust N Z J Obstet Gynaecol 1997; 37: 187-91
12. James WH. A note on the epidemiology of acardiac monsters. Teratology 1977; 16: 211-6.
13. Al-Malt A, Ashmead G, Judge N. Color- flow and Doppler velocimetry in prenatal diagnosis of acardiac triplet. J Ultrasound Med 1991; 10: 341-5.
14. Moore CA, Buehler BA, McManus BM. Brief clinical report: acephalus-acardia in twins with aneuploidy. Am J Med Genet (Suppl) 1987; 3: 139-43.
15. Izquierdo L, Smith J, Gilson G. Twin, acardiac, acephalus. Fetus 1991; 1: 1-3.
16. Nevils BG, Maciulla JE, Izquierdo L. Twin, acardiac, anceps. Fetus 1993; 3: 15-7.
17. Langlotz H, Sauerbrei E, Murray S. Transvaginal Doppler sonographic diagnosis of an acardiac twin at 12 weeks gestation. J Ultrasound Med 1991; 10: 175-9.
18. Petersen BL, Broholm H, Skibsted L, Graem N. Acardiac twin with preserved brain. Fetal Diagn Ther 2001; 16: 231-3.
19. Cardwell MS. The acardiac twin: a case report. J Reprod Med 1988; 33: 320-2.
20. Pinet C, Colau JC, Delezoide AL, Menez F. Acardiac twins. J Gynecol Obstet Biol Reprod 1994; 23: 85-92.
21. Robie GF, Payne CG, Morgan MA. Selective delivery of an acardiac, acephalic twin. N Engl J Med 1989; 320: 512-3.
22. Porreco RP,Barton SM, Haverkamp AD. Occlusion of umbilical artery in acardiac, acephalic twin. Lancet 1991; 337: 326-7.
23. McCurdy CM, Childer JM, Seeds JW. Ligation of the umbilical cord of an acardiac- acephalus twin with an endoscopic intrauterine technique. Obstet Gynecol 1993; 82: 708-11.
24. Muhlhaus K, Beisler G. Acardius-prenatal diagnosis and pregnancy in 2 twin pregnancies. Z Geburtshilfe Perinatol 1986; 190: 98-103.
25. Ginsberg NA, Applebaum M, Rabin SA, Caffarelli MA, Daskal JL, Verlinsky Y, et al. Term birth after midtrimester hysterotomy and selective delivery of an acardiac twin. Am J Obstet Gynecol 1992; 167: 33-7.
26. Donnenfeld AE, van de Woestijne J, Craparo F, Smith CS, Ludomirsky A, Weiner S. The normal fetus of an acardiac twin pregnancy: perinatal management base on echocardiographic sonographic evaluation. Prenat Diagn 1991; 11: 235-44.
27. Sherer DM, Armstrong B, Shah Y. Prenatal sonographic diagnosis, Doppler velocimetric umbilical cord studies, and subsequent management of an acardiac twin pregnancy. Obstet Gynecol 1989; 74: 472-5.
28. Healey MG. Acardia: predictive risk factors for the co-twin's survival. Teratology 1994; 50: 205-13.
29. 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.
30. Ash K, Harman CR, Gritter H. TRAP sequence - successful outcome with indomethacin treatment. Obstet Gynecol 1990; 76: 960-2.
31. Norwitz ER, Hoyte LPJ, Jenkins KJ, van der Velde, Ratiu P, Rodriguez-Thompson D, et al. Seperation of conjoined twins with the twin reversed-arterial-perfusion sequence after prenatal planning with three-dimensional modeling. N Engl J Med 2000; 343: 399-402.
32. 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.

เด็กแฝดที่ไม่มีหัวใจ

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Acardiac twin เป็นภาวะแทรกซ้อนของการตั้งครรภ์แฝดที่เกิดจากไข่ฟองเดียวที่พบได้น้อย โดยพบได้ประมาณ ร้อยละ 1 ของครรภ์แฝดชนิด monochorion และพบประมาณ 1 ต่อ 35,000 ของการคลอด หากไม่ได้รับการรักษา จะพบอัตราการตายของทารกแฝดคนที่ปกติได้ถึงร้อยละ 50-70 การรักษาที่เหมาะสมในภาวะดังกล่าวนี้ยังคงไม่เป็นที่ตกลงกันอย่างแน่ชัด ซึ่งการรักษามีได้หลายวิธี ทั้ง การสิ้นสุดการตั้งครรภ์, การเฝ้าสังเกตอย่างใกล้ชิดโดยมีการตรวจติดตามด้วยเครื่องตรวจคลื่นเสียงความถี่สูงเป็นระยะ และการทำหัตถการเพื่อขัดขวางการเชื่อมต่อของเส้นเลือดที่ไปเลี้ยงแฝดคนที่ผิดปกติ