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The acardiac twin : a case report

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The acardiac twin or twin reversed arterial perfusion sequence (TRAPS) represents one of the most severe, but rare, congenital anomalies which occurs only in multiple gestations associated with vascular anastomoses between the affected fetus and its co-twin. We report an interesting case of a macrosomic acardiac fetus weighing 3.5 - fold of another donor fetus. A 20-year-old Thai woman, gravida 1, para 0 was referred to Chulalongkorn Hospital at 19 weeks' gestation for further management of twin with one fetal demise due to absence of identifiable cardiac pulsation. Ultrasound scan confirmed a diamniotic twin gestation. The anatomy of Twin A appeared normal, but the upper part of the body of Twin B was markedly distorted. Cystic hygroma, ascites and marked tissue edema were noted. No definable cardiac pulsation was demonstrated. A twin with one fetal hydrops, associated with acardiac fetus, was suspected. Bed rest was recommended, and follow-up sonographic examinations documented progressive discrepancy of BPD. At 31 weeks' gestation, hydramnios and preterm labor were presented despite labor inhibition with salbutamol. The infants were delivered by low transverse cesarean section because of transverse-oblique line. The male acardiac twin weighing 4,200 g., demonstrated no cardiac activity but showed a huge cephalic part with imperfect face and omphalocele. The normal male twin weighed 1,210 gm, with Apgar scores of 8 and 9 at 1 and 5 minutes, respectively, but died of pneumonia and sepsis at age 7 days. Examination of the placenta demonstrated interfetal vascular anastomosis. The mother's postoperative course was unremarkable.

Key words : *Acardiac twin, Twin reverse arterial perfusin sequence (TRAPS).*

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การตั้งครรภ์แฝดโดยทารกหนึ่งคนไม่มีหัวใจ จัดเป็นภาวะที่พบน้อยมาก แต่มีความรุนแรง เกิดในการตั้งครรภ์แฝดสองขึ้นไป โดยมีการเชื่อมต่อของหลอดเลือดจากทารกทั้งสอง ใต้รายงานทารกแฝดที่ไม่มีหัวใจ ซึ่งมีน้ำหนัก 3.5 เท่าของทารกคู่แฝดปกติ มารดาอายุ 20 ปี ตั้งครรภ์แรก ได้รับการส่งต่อเข้ารับการรักษาในโรงพยาบาลจุฬาลงกรณ์ขณะอายุครรภ์ 19 สัปดาห์ด้วยพบว่าเป็นครรภ์แฝดและสงสัยมีทารกเสียชีวิตคนหนึ่ง เนื่องการตรวจไม่พบการเต้นของหัวใจขณะตรวจคลื่นเสียงความถี่สูง ได้ทำการตรวจคลื่นเสียงความถี่สูง พบว่าเป็นการตั้งครรภ์แฝดชนิดมีถุงน้ำคร่ำ 2 ถุง ทารกแฝดคนแรกตรวจไม่พบความผิดปกติ แต่ทารกคู่แฝดอีกคนมีร่างกายส่วนบนพบความผิดปกติมาก โดยมีถุงน้ำขนาดใหญ่รอบศีรษะและคอ มีภาวะท้องมาน บวมทั่วตัว และไม่เห็นหัวใจเต้น ได้ให้การวินิจฉัยเป็นทารกแฝดไม่มีหัวใจและมีภาวะบวมน้ำ แนะนำให้มารดารักษาดูแลในโรงพยาบาล โดยให้ยาคลายกล้ามเนื้อ และตรวจติดตามด้วยคลื่นเสียงความถี่สูงพบขนาดศีรษะของทารกทั้งสองมีความแตกต่างกันมาก เมื่ออายุครรภ์ 31 สัปดาห์ ภาวะครรภ์แฝดน้ำมีความรุนแรงมากและทำให้เจ็บครรภ์ก่อนกำหนดร่วม แม้จะให้ยาชาลบลูตามอลอยู่ก็ตาม ทารกคู่แฝดได้รับการผ่าตัดคลอดทางหน้าท้องเนื่องจากเป็นท่าขวาง ทารกแฝดชายที่ไม่มีหัวใจมีน้ำหนัก 4,200 กรัม เพศชายมีศีรษะและร่างกายส่วนบนขนาดใหญ่ ผิดปกติรูปหน้าวิปริต และมี omphalocele ร่วมด้วย ทารกคู่แฝดชายอีกคนมีลักษณะปกติ น้ำหนัก 1,210 กรัม มีคะแนนแอปการ์ที่ 1 และ 5 นาทีเท่ากับ 8 และ 9 ตามลำดับ แต่เสียชีวิตในเวลาต่อมาเนื่องจากปอดบวม และติดเชื้อเมื่ออายุ 7 วันหลังคลอด การตรวจศพมีการเชื่อมต่อกันของหลอดเลือด หลังคลอดบุตรมารดาไม่มีภาวะแทรกซ้อน

Acardiac malformation, characterised by failure of heart development and severe dysmorphogenesis, is a rare consequence of monozygotic twinning.⁽¹⁾ The incidence of this phenomenon is 1 per 35,000 deliveries,⁽²⁾ with 1% of all monozygotic twins affected.⁽³⁾ The condition has also been reported to occur in non induced pregnant triplet pregnancies with a higher incidence.^(4,5) The acardiac monster represents one of the most severe congenital anomalies so prenatal diagnosis of an acardiac fetus must be suspected in any multiple gestation in which cardiac activity cannot be demonstrated sonographically in a growing fetus. We report an interesting case of a macrosomic acardiac fetus weighing 3.5-fold the other variable donor fetus.

Case Report

A 20-year-old Thai woman, gravida 1, para 0, had her last menstrual period on August 1st, 1992, and an estimated date of confinement of May 8th, 1993. A uterine size-date discrepancy became apparent at 16 weeks' gestation. The obstetric ultrasound scan on November 25th, 1992, revealed a twin gestation. An intrauterine fetal demise was suspected due to the absence of identifiable cardiac pulsation in the smaller twin with a biparietal diameter (BPD) of 30 mm. The viable twin had a BPD of 36 mm., which was compatible with the menstrual age.

The patient was referred to Chulalongkorn Hospital for further evaluation and management. The ultrasound scan repeated on December 17th, 1992, by Hitachi EUB40 at 19 weeks' gestation confirmed a diamniotic twin gestation.

The anatomy of Twin A appeared normal, despite a mild increase in the amniotic fluid

volume. However, there were marked tissue edema and abnormally large multi-cystic areas around the head and upper part of the body of Twin B compatible with cystic hygroma. The chest was represented by a large sonolucent area with no definable cardiac pulsation. Ascites were noted. Both kidneys were not demonstrated. The fetal bladder seen. The upper extremities were poorly identified. The lower extremities showed marked edema of the skin and subcutaneous tissues. (Figure 1) The umbilical cord of each twin was noted to contain three vessels. Bed rest was recommended, and follow-up sonographic examinations at 4 week intervals were performed at 24 weeks' gestation to assess the growth of the viable twin. The authors, the BPD of Twin A (viable twin) had grown from 47.0 to 60.2 mm., whereas Twin B had markedly grown from 43.0 to 68.2 mm. without cardiac pulsation. The diagnosis was twins pregnancy with an acardiac fetus. At 28 weeks' gestation, the patient was hospitalised because of a weight gain of 5 kg in a 2-week period, acute hydramnios, and uterine irritability. Her blood pressure was 130/80 mmHg. and regular uterine contractions were noted. Labor inhibition with oral salbutamol of 32 mg. per day was initiated; and with success in a few hours later. Continuation of oral salbutamol was recommended. At 31 weeks' gestation, the patient complained marked abdominal distention and was in labor despite the presence of tocolytics. Fundal height was 42 cm and pelvic examination revealed cervix was 6 cm dilated, and completely effaced. The infants were delivered by low transverse cesarean section because of transverse-oblique line of twin A and dystocia due to the huge upper part of the acardiac twin B.



Figure 1. Ultrasonography of Twin B shows cystic hygroma.

The normal male twin A, weighing 1,210 gm., was delivered with Apgar scores at one and five minutes of 9 and 9, respectively. He was admitted to the neonatal intensive care unit, but unfortunately twin A died from pneumonia and sepsis 7 days later.

The male acardiac twin, B weighing 4,200 gm, with huge cephalic part, imperfect face and omphalocele without cardiac activity (Figure 2) Examination of the placenta, weighing 1,200 gm, demonstrated a monochorionic diamniotic placenta with vascular anastomosis. The umbilical cords were noted to contain two arteries and one vein each.

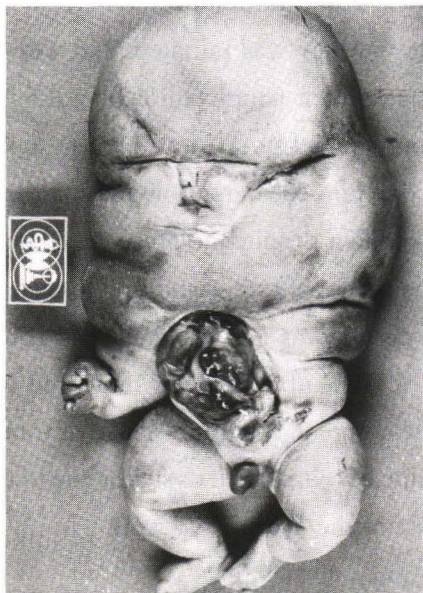


Figure 2. The acardiac twin.

The patient's postoperative course was uneventful.

Discussion

Based on the associated anomalies, four types of acardiac twins are described in the literature: acardius anceps, acardius acephalus, acardius acornus, and acardius amorphus.^(4,6,7) The clinician must be aware that classification may be difficult in certain cases. The most highly developed type, acardius anceps, has a partly developed head and a deformed face, trunk, and extremities, and is exortic.⁽⁶⁾

The etiology of the acardiac twin remains unknown. Two main theoretical etiologies have been proposed.^(1,8) The first explanation is based on a chromosomal defect causing primary cardiac agenesis or dysgenesis in the acardiac twin, which is then perfused by the healthy twin through the anastomosis. The second theory (vascular reversal theory), abnormal placental vascular communications between the twins as the primary factor, resulting in secondary atrophy or arrest of cardiac and other tissue development in one twin.^(1,9) In the acardiac twin the anastomoses have classically been described as artery to artery and vein to vein, which differs from the twin-to-twin transfusion syndrome, where the anastomosis is artery to vein.⁽¹⁰⁾ The pressure of blood from the heart of the first (normal) twin forces the current backwards through the arteries of the second (acardiac) twin. Since normal morphogenesis of the cardiac tube depends on the magnitude and direction of hemodynamic forces, the heart of the second twin atrophies. The hemodynamically disadvantaged twin, known as the perfused twin, will depend on "used" blood from the normal twin, known as the pump twin.⁽¹⁰⁾ Such inadequate

perfusion with poorly oxygenated blood is the main cause of the various forms of tissue disruption and incomplete morphogenesis in the perfused twin. There are strong evidence in favour

of the vascular reversal perfusion theory. Recently, Doppler blood flow studies have further substantiated the vascular theory.^(8,10)

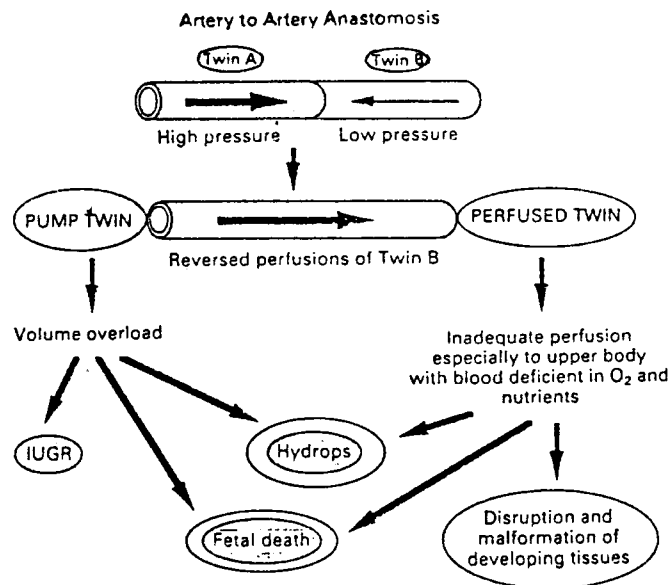


Diagram 1 Pathophysiological development of the twin reversed arterial perfusion sequence.

Obstetric ultrasonography has made pre-natal diagnosis of the acardiac twin possible. The hallmark for the diagnosis of an acardiac twin by ultrasound is the absence of identifiable cardiac pulsation. Other characteristic features are ill defined head, trunk and upper extremities. The lower extremities are usually present but often deformed. There are usually marked tissue edema and multiple cystic areas in the upper part of the body. Other associated features are two-vessel cord and hydramnios in the acardiac twin sac. Recently the use of Doppler sonography has enabled the demonstration of circulation reversal with arterial blood flow towards the acardiac twin^(8,10,11) but this case doppler sonography was not done because of limitation of instrument used.

Once the diagnosis of the acardiac twin is made, the therapeutic interest is focused on the normal twin since the monster has no chance of survival. Mortality rates for the normal twin are as high as 50 percent⁽⁹⁾ with the cause of death for congestive heart failure and extreme prematurity due to hydramnios and preterm labor. The aims of management are to prevent, detect and possibly treat congestive heart failure in the normal twin and continue pregnancy to term. When twin reversal arterial perfusion (TRAP) is diagnosed, close fetal surveillance should be employed. Hydrops due to congestive heart failure in the normal twin can be treated with digoxin given to the mother. If there is a threat of preterm labor and the normal fetus is not

compromised, tocolytic agents should be administered, with steroids. Serial amniocenteses are needed for symptomatic hydramnios. During labour, the delivery of the acardiac twin is usually associated with significant soft tissue dystocia of the upper part of abnormal twin due to multiple structural anomalies or hydrops.

Platt et al⁽¹²⁾ proposed severing the anastomotic circulation by clamping the umbilical cord of the acardiac fetus. Robie et al⁽¹³⁾ described hysterotomy with selective delivery of the acardiac fetus at 22 weeks' gestation and subsequent delivery of the healthy surviving fetus at 33 weeks. Porreco et al⁽¹⁴⁾ used a thrombogenic coil to interrupt vascular flow to the acardiac fetus with delivery of the healthy twin at 39 weeks.

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