The Management of Acardiac Twins at King Chulalongkorn Memorial Hospital: Case Series

Somchai Tanawattanacharoen MD*, Saknan Manotaya MD*,
Teera Wacharaprechanont MD*, Boonchai Uerpairojkit MD*,
Yuen Tannirandorn MD*, Dhirapongs Charoenvidhya MD*

** Department of Obstetrics and Gynecology, Faculty of Medicine, Chulalongkorn University*

Acardiac twin is a rare event with high perinatal mortality rate and the management strategies remain controversial. The authors report 4 cases of twin pregnancies associated with one acardiac twin diagnosed at King Chulalongkorn Memorial Hospital during the period 1993 to 2002. Two cases were expectantly managed and intrauterine interventions were performed in order to occlude umbilical artery of the acardiac twin in 2 cases, by using Guglielmi detachable coil (GDC) embolization and absolute alcohol injection. Overall, the perinatal mortality rate for the pump twin was 50% and the survival rate of expectant management in the present series was 100%. GDC embolization was judged to be technically successful since it completely occluded the circulation to the acardiac twin. However, the pump twin was dead which might have resulted from the compromised state due to cardiac failure. At present, it seems that conservative management with close antenatal surveillance is the treatment of choice when the twin-weight ratios are substantially less than 70%. Invasive techniques should be considered when there is ultrasound evidence of hydramnios or congestive heart failure of the pump twin at a previable gestational age.

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Acardiac twin is a unique complication of monochorionic twin gestations. It is a rare condition, occurring in 1 in 35,000 pregnancies, and in 1 % of monozygotic twins[1,2]. The condition involves a "pump" or donor twin perfusing a recipient or "acardiac" twin through vascular placental anastomoses. It is well accepted that the acardiac twin has no direct vascular connection to the placenta, so the lower part of the body receives better oxygenated blood through a hypogastric artery, whereas the upper part is more poorly perfused with less well oxygenated blood. This may cause secondary disruption of organogenesis and subsequent fetal acardia[2]. This condition is also known as twin reversed arterial perfusion (TRAP) sequence.

The acardiac twin obviously has no chance of survival. Perinatal mortality rate of the pump twin is extremely high (50-55%)[2,3] and can be due to increased cardiac demands in an effort to perfuse its acardiac sibling, hydramnios, and complications from prematurity. The management strategies for TRAP sequence remain controversial. The therapeutic efforts by invasive procedures were aimed to occlude the vascular communication between the two fetuses. Several techniques have been proposed with varying degrees of success, including injection of thrombogenic coils or sclerosing agents, to occlude the umbilical cord, cord ligation performed at hysterotomy or with endoscopic or ultrasound guidance and fetoscopic laser coagulation. In 2003, Tan and Sepulveda[4] have systematically reviewed minimally invasive treatment modalities in acardiac twinning and suggested the intrafetal ablation is the treatment of choice for acardiac twins. They also recommended that the treatment should be mandatory in those cases that show deterioration, even mild, on close surveillance scans or in cases of excessive growth of the acardiac twin.

Sullivan et al[5] have recently reported less neonatal mortality of pump twins that were expectantly managed than previously reported. Even the twin-weight ratio was not accurate in predicting outcome of the pump twins in their series. They suggested that expectant management with close antepartum surveillance deserves consideration when counseling patients with acardiac twins. The authors present the outcomes of 4 cases of acardiac twins with different management.

Correspondence to : Tanawattanacharoen S. Department of Obstetrics and Gynecology, Faculty of Medicine, Chulalongkorn University. Rama IV Road, Bangkok 10330, Thailand. Phone: 0-2256-4824, Fax: 0-2256-4825, E-mail: dr_coke98@hotmail.com

Method

During the period 1993 to 2002, four cases of twin pregnancies associated with one acardiac twin were diagnosed at King Chulalongkorn Memorial Hospital. All patients were evaluated and managed by maternal-fetal medicine specialists. The gestational age of the pregnancy was based on the patient's last menstrual period and/or the mid-trimester sonograms. Management of each case was different and was at the discretion of the clinician. Patients were counseled about the natural history and prognosis of this condition. Two patients who opted for intrauterine interventions were counseled at length before the procedure was carried out.

Data were collected from prenatal records, ultrasound reports, operative notes, labor and delivery charts, and neonatal records. The primary outcome variable was survival of the pump twin. Secondary outcome variables included gestational age at the time of diagnosis and delivery, the weight of the acardiac and pump twins.

Results

Pregnancy characteristics and outcomes are shown in Table 1. Three cases were diagnosed in the first half of pregnancy (at 19 weeks). Case 4 was misdiagnosed as single fetal demise in the first trimester and follow-up scan at 29 weeks revealed the continued growth of the "suspected dead" twin. This case was referred to our institute and TRAP sequence was diagnosed. Two of 4 (50%) cases that were managed expectantly resulted in the live birth of the pump twin. All cases had the twin-weight ratio greater than 70% (100.0-347.1%).

The first case was diagnosed in the year 1993 and no intervention was performed because of the limitation of our facility at that time. In case 2, the parents opted for conservative treatment at the beginning and decided to undergo the procedure after the ultrasound revealed mild pericardial effusion and hydramnios in the pump twin. The authors performed occlusion of umbilical artery of the acardiac twin using the Guglielmi detachable coil (GDC). The procedure went well and the cessation of blood flow to the acardiac twin was achieved immediately after detachment. Unexpectedly, follow-up scan on the first postoperative day indicated death of the pump twin. The authors injected absolute alcohol into the umbilical artery of the acardiac twin in case 3 on the same day of diagnosis. Unfortunately, the procedure failed and both twins died immediately after the procedure. The last case was referred to our institute at 30 weeks of gestation and showed no evidence of heart failure. Thus, the authors decided to expectantly manage until 34 weeks and do a cesarean section.

Discussion

Overall, the perinatal mortality rate for the pump twin was 50%. Like the study of Sullivan et al[5], the survival rate of expectant management in the present series (100%) was better than previous reports. This may be due to the small number of cases in the present study. Moreover, several studies have reported spontaneous cessation of blood flow to acardiac twin, resulting in good pregnancy outcomes [5-9] and this event might occur in the present series.

Moore et al[3] retrospectively described 49 cases of acardiac pregnancy and noted that prognosis was strongly associated with the weight ratio of the acardiac twin to the pump twin (the twin-weight ratio). In the present series, the twin-weight ratio was not a reliable prognostic factor of outcome. All cases had the twin-weight ratio greater than 70 % and 2 cases (50%) resulted in live births. However, the twin-weight ratio may be useful in counseling patients when using other prognostic factors.

The therapeutic efforts for this condition were aimed to treat congestive heart failure of the pump twin with digoxin[10] and palliation of hydramnios with amnioreduction[11] and, later, with indomethacin[12]. The first successful invasive intervention for acardiac

Table 1. Pregnancy characteristics and outcomes

Case	Year	Age	GA at diagnosis (wk)	Intervention	GA at delivery (wk)	Mode of delivery	Acardiac weight (g)	Pump weight (g)	Acardiac: pump (%)	Outcome
1	1993	20	19	no	31	C/S	4200	1210	347.1	LB
2	2000	39	19	Coil (23 wk)	23	NSVD	600	600	100.0	IUFD
3	2002	31	19	Alcohol (19 wk)	20	NSVD	300	240	125.0	IUFD
4	2002	30	30	no	34	C/S	3000	1700	176.5	LB

C/S, cesarean section; NSVD, normal spontaneous vaginal delivery; LB, live birth; IUFD, intrauterine fetal death

twinning was reported in 1989 by Robie et al[13], who selectively delivered a 710-g acardiac twin by hysterotomy (sectio parva) at 22 weeks, with subsequent delivery of a 2310-g healthy pump twin at 33 weeks. With this intervention, the survival rate was achieved, but maternal complications, such as pulmonary edema, abruptio placenta, were significantly increased. Platt et al[11] were the first to propose the management by cord occlusion because the real goal of the treatment was to occlude the vascular communication between the two fetuses. The first successful case of percutaneous cord occlusion by a thermogenic coil embolization was described in 1989 by Hamada et al[14]. In 1991, Porreco et al[15] reported another successful case by a similar approach. Other cord occlusion techniques were subsequently developed, both funicular and intrafetal approaches, with varying degrees of success. These techniques include percutaneous injection of sclerosing agents[16,17]; cord ligation performed at hysterotomy[18] or with fetoscopic[19-21] or ultrasound guidance; fetoscopic laser coagulation[22,23]; and radiofrequency[24].

Intrauterine interventions were performed in order to occlude the umbilical artery of the acardiac twin in 2 cases, by using GDC embolization and absolute alcohol injection. GDC embolization has been shown to be an effective non-invasive treatment of intracranial aneurysms. In case 2, the procedure was judged to be technically successful since it completely occluded the circulation to the acardiac twin. Death of the pump twin may have resulted from the compromised state due to fetal cardiac failure. However, the authors still proposed this intervention to be an effective and useful technique for the treatment of TRAP sequence[25].

It is unclear whether antenatal surveillance can improve the outcome in expectantly managed cases of TRAP sequence. Interventions were usually delayed until there was some evidence of cardiac dysfunction to avoid unnecessary interventions. However, Sullivan et al[5] reported that one case of pump twin death in their study occurred with a normal echocardiogram 1 week before demise. On the other hand, the death of the pump twin in case 2 of the present series was considered to possibly be due, in part, to too late intervention.

In summary, the rarity of TRAP sequence does not permit definitive conclusions to be reached concerning the prognostic factors and management strategies. At present, it seems that conservative management with close antenatal surveillance is the treatment of choice when the twin weight ratios are substantially less than 70%. Invasive techniques should be considered when there is ultrasound evidence of hydramnios or congestive heart failure of the pump twin at a previable gestational age. However, it remains to be established whether interventions would still be useful at that time.

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การดูแลรักษาการตั้งครรภ์แฝดโดยทารกคนหนึ่งไม่มีหัวใจในโรงพยาบาลจุฬาลงกรณ์

สมชาย ธนวัฒนาเจริญ, ศักนัน มะโนทัย, ธีระ วัชรปรีชานนท์, บุญชัย เอื้อไพโรจน์กิจ, เยื้อน ตันนิรันดร, ธีระพงศ์ เจริญวิทย์

การตั้งครรภ์แฝดโดยทารกคนหนึ่งไม่มีหัวใจเป็นภาวะที่พบได้น้อยแต่มีอัตราตายปริกำเนิดของทารกสูงและยังไม่มีแนวทางดูแลรักษาที่เหมาะสม คณะผู้วิจัยได้รายงานผู้ป่วยที่มีภาวะดังกล่าวที่ได้รับการวินิจฉัยและดูแลรักษาที่โรงพยาบาลจุฬาลงกรณ์ในช่วงปี พ.ศ. 2536 ถึง 2545 ผู้ป่วย 2 รายได้รับการดูแลรักษาแบบประคับประคอง ในขณะที่ผู้ป่วยอีก 2 รายได้รับการรักษาโดยการทำหัตถการต่อทารกในครรภ์เพื่อทำให้เส้นเลือดในสายสะดือของทารกที่ไม่มีหัวใจอุดตันไป ซึ่งได้แก่การใส่ Guglielmi detachable coil (GDC) รายหนึ่งและฉีดแอลกอฮอล์อีกรายหนึ่ง พบว่าอัตราตายปริกำเนิดของทารกของทารกคู่แฝดที่เป็นปกติเท่ากับร้อยละ 50 โดยอัตรารอดชีวิตจากการดูแลรักษาแบบประคับประคองเท่ากับร้อยละ 100 หัตถการใส่ GDC ประสบผลสำเร็จแต่ทารกคู่แฝดที่เป็นปกติเสียชีวิต ซึ่งน่าจะเกิดจากทารกดังกล่าวเริ่มมีภาวะหัวใจล้มเหลว ในปัจจุบันการดูแลรักษาแบบประคับประคองโดยมีการตรวจติดตามและเฝ้าระวังอย่างใกล้ชิดน่าจะเป็นวิธีดูแลรักษาที่เหมาะสมที่สุดหากสัดส่วนน้ำหนักของทารกทั้งสองยังน้อยกว่าร้อยละ 70 การพิจารณาทำหัตถการต่อทารกในครรภ์ควรทำเมื่อตรวจคลื่นเสียงความถี่สูงพบว่าทารกคู่แฝดที่เป็นปกติมีปริมาณน้ำคร่ำมากหรือมีภาวะหัวใจล้มเหลวเท่านั้น