Case Report

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Therapeutic dilemma in twin reversed arterial perfusion sequence

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Abstract

The dissemination of minimally invasive in utero surgery reduced the mortality of twin reversed arterial perfusion sequence, but the mortality of expectantly treated surgical candidates remains high. A 26-year-old, non-parous, Japanese woman at 13 weeks of gestation had been diagnosed with twin reversed arterial perfusion sequence and was judged as a surgical candidate for radiofrequency ablation. However, she did not undergo surgery because of the anatomical location of the acardiac twin. At 18 weeks of gestation, the blood flow to the acardiac twin disappeared spontaneously. The pump twin began to demonstrate fetal growth retardation during the third trimester. The patient delivered a 1891 g female at term. We macroscopically identified the cause of the fetal growth retardation as velamentous insertion of the umbilical cord and microscopically diagnosed the acardiac twin with acardiac acephalus. We should give the same attention to the management of post–twin reversed arterial perfusion sequence as twin reversed arterial perfusion sequence itself.

Keywords

Twin reversed arterial perfusion sequence, acardiac twin, fetal growth retardation, radiofrequency ablation

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Introduction

Twin reversed arterial perfusion (TRAP) sequence is an abnormal fetal circulation of monochorionic twins in which a twin with an absent or nonfunctioning heart (i.e. the acardiac twin) is perfused by its co-twin (the pump twin) in a retrograde fashion via placental arterio-arterial and veno-venous anastomoses.[1] Consequently, the well-being of the pump twin may be threatened by congestive heart failure due to increased cardiovascular demands, a mass effect due to the continued growth of the acardiac twin and chronic hypoxia due to reperfusion of over-deoxygenated blood from the acardiac twin through the vein-vein anastomosis.[2]

Although in 1990 the reported overall peritoneal mortality of 49 acardiac twin pregnancies with TRAP sequence was 55%,[3] in the 2000s, the dissemination of minimally invasive in utero surgery reduced the mortality to 15%.[4] However, the mortality of expectantly treated surgical candidates remains as high as 57%.[5]

Case report

A 26-year-old, gravida 1, para 0 Japanese woman at 37 weeks of gestation was referred to Nakatsu Municipal Hospital with fetal growth retardation (FGR). Her past medical history included childhood asthma and cigarette smoking, and she had the comorbidity of rhinitis.

The patient had conceived naturally, visited Fujiyoshi Clinic, and was diagnosed with a single fetal demise of the co-twin in a monochorionic diamniotic twin pregnancy at 9 weeks of gestation. However, the ultrasound performed at 12 weeks' gestation indicated that one fetus appeared anatomically normal and the other lacked cardiac activity, but had a blood flow in the acardiac mass. TRAP sequence was suspected, and the patient was referred to Fukuoka Children's Hospital for a definite diagnosis and for an investigation of the possibility of in utero surgery.

At Fukuoka Children's Hospital, the Doppler ultrasound of the acardiac twin's umbilical cord performed at 13 weeks

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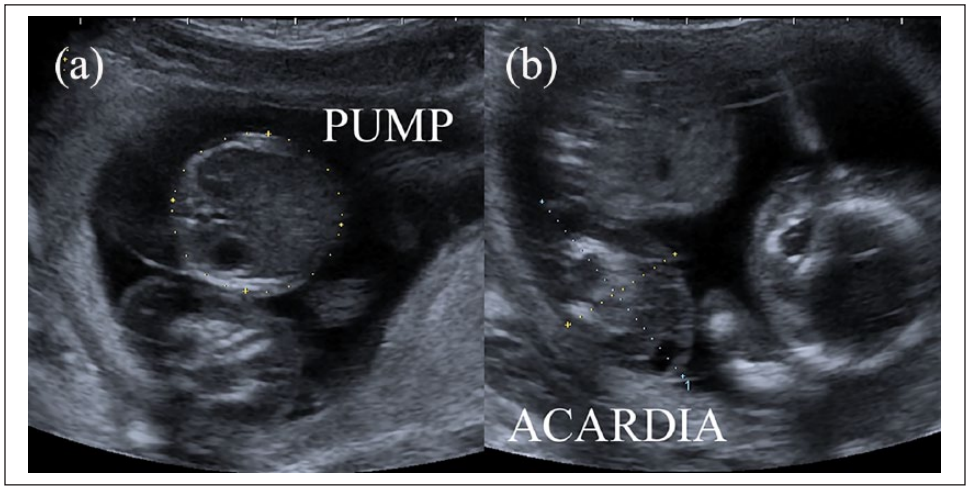

Figure 1. Abdominal ultrasound images at 16 weeks' gestation. Calculated waist of the pump twin from the abdominal circumference (a), and that of the acardiac twin from the transverse trunk diameter and antero-postero trunk diameter (b).

of gestation demonstrated a reversed arterial blood flow from the placenta to the acardiac twin. We thus finally diagnosed TRAP sequence. At 16 weeks of gestation, the calculated waist of the pump twin was 9.63 cm (Figure 1(a)) and that of the acardiac twin was 10.93 cm (Figure 1(b)). A relatively large ratio of the size of the acardiac twin to the pump twin met one of the institutional criteria for in utero surgery. However, the acardiac twin's location was dorsal to the pump twin and thus the needle for radiofrequency ablation (RFA) could not access the acardiac twin's sac without disrupting the dividing membrane.

To avoid the occurrence of an iatrogenic monochorionic mono-amniotic twin, which causes unexpected fetal demise, we had no choice but to postpone the RFA until the acardiac twin re-positioned to become accessible to RFA without disruption of the dividing membrane. However, after 2 weeks, we confirmed the spontaneous cessation of blood flow in the umbilical artery of the acardiac twin. The patient thus returned to Fujiyoshi Clinic and restarted routine and regular perineal care. In the late preterm, the pump twin began to demonstrate FGR: the estimated fetal growth was 1985 g (−1.16 SD) at 35 weeks' gestation, 1995 g (−1.70 SD) at 36 weeks' gestation, and 1983 g (−1.91 SD) at 37 weeks' gestation.

At the patient's initial visit to Nakatsu Municipal Hospital at 37 weeks' gestation, ultrasound demonstrated a female, vertex-positioned pump twin weighing 2102 g (−2.16 SD). At the 1-week interval follow-up visit, the estimated fetal weight of the pump twin was 2185 g (−2.24 SD), and we planned an elective induction of labor. At 38 weeks and 6 days of gestation, induction of delivery was started using 0.5 mg of dinoprostone (Prostaglandin E_2, Kaken Pharmaceutical, Tokyo, Japan).

At 39 weeks and 1 day of gestation, the patient delivered a 1891 g female with the Apgar score of 8/9 (1 min/5 min). On gross examination, the acardiac twin was wrapped in an egg membrane and the placental end of the cord consisted of divergent umbilical vessels surrounded only by fetal membrane, demonstrating velamentous umbilical cord insertion.

A gross image (Figure 2(a)) and a loupe image (Figure 2(b)) showed that the upper body of the acardiac was deficient. On microscopic examination, a small umbilical cord-like structure was seen (Figure 2(c)). When we applied the methods proposed by Van Gemert et al.,[6] the pump/acardiac umbilical venous diameter (UVD) ratio was calculated as 4.1. Striated skeletal muscle tissue was identified in the sections (Figure 2(d)), but heart tissue composed of myocardium was not found in the section. The acardiac twin was pathologically classified as having acardiac acephalus, in which the fetal thoracic organs and head are absent. The maternal puerperal course was uneventful. We followed the infant until the age of 4 months, and she had no problem of development and height growth.

Discussion

We abandoned a plan for RFA at 16 weeks' gestation because of the anatomical location of the acardiac twin. This spontaneously resolved with the cessation of the blood flow to the acardiac twin by 18 weeks' gestation, and consequently, a full-term live birth of the pump twin occurred. Table 1 summarizes the perinatal outcome of expectantly treated surgical candidates in the English literature of the past decade. Although these reported cases vary regarding the gestational age at diagnosis, the gestational age at planned procedures, and the planned procedures, the perinatal outcomes of these expectantly treated surgical candidates were poor. Sullivan et al.[9] described 10 TRAP sequence cases treated by only a conservative approach. In their series, a live birth of the

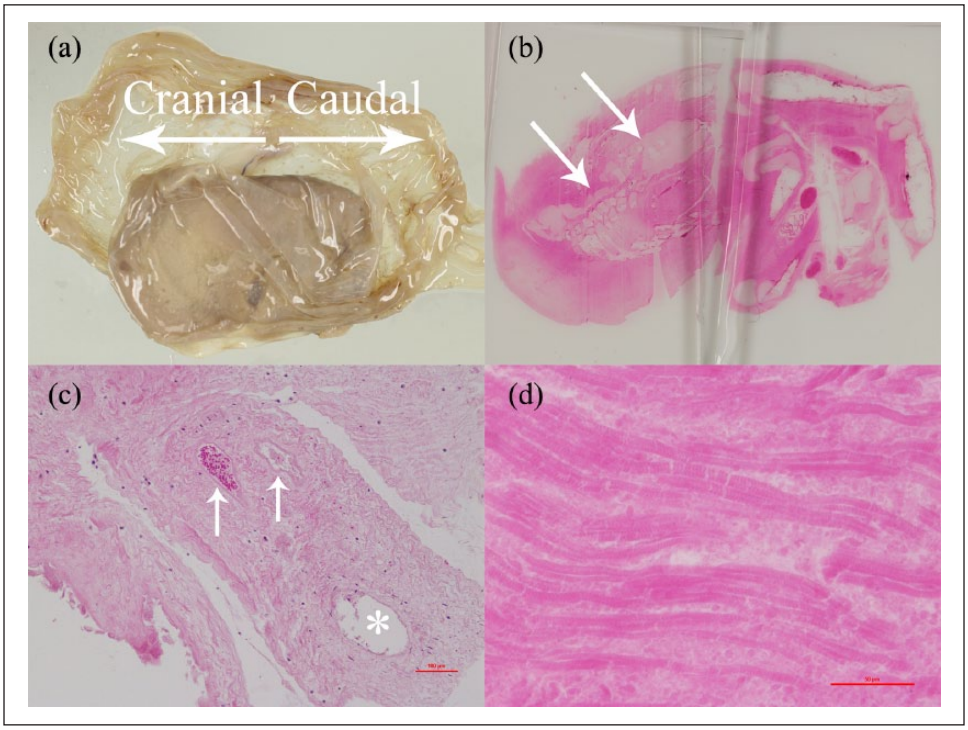

Figure 2. A gross (a) view of the acardiac twin suggested a deficit of upper body, which was confirmed by a loupe view (b), demonstrating the disruption of vertebrae (white arrows). (c) Umbilical cord of the acardiac twin consisted of two arteries and one vein with degenerative or necrotic change. (d) Striated skeletal muscle tissue was identified in the sections.

Table 1. Perinatal outcomes of expectantly treated surgical candidates: studies from 2006 to present.

Reference	Year	GA at diagnosis (weeks)	Planned GA at procedure (weeks)	Planned procedure	Patients (n)	Survivors (n)
Quintero et al.[5]	2006	20.4 ± 2.9[a]	21.2 ± 2.5	Extrafetal umbilical cord occlusion	14	6
Lewi et al.[7]	2009	11–13[b]	16–18	Extrafetal laser coagulation of cord/placental anastomoses	5	2
Chaveeva et al.[8]	2013	11–14	16–18	Intrafetal laser ablation	18	7
Present case	2018	13	16	Intrafetal radiofrequency ablation	1	1

GA: gestational age.
[a]Data are mean and standard deviation.
[b]Range of GA.

pump twin occurred in 9 of the 10 women, and 4 of the 10 cases resolved with the cessation of the blood flow to the acardiac twin after 19 weeks of gestation. Although it is difficult to compare different patients' characteristics, the gestational ages of the spontaneous cessation of blood flow in those four cases are almost identical to that in our patient's case. Based on the predicted pump-to-acardiac UVD curve,[10] the UVD ratio of this case was classified in the uncomplicated group, matching the perinatal outcome of our patient's case.

FGR was also confirmed in our patient's third trimester after a natural remission of TRAP sequence, and we identified the cause of FGR as the velamentous insertion of the umbilical cord. Velamentous insertion occurs in as many as 15% of monochorionic twin gestations, and this prevalence is 15-fold that in singleton gestations.[11] With the now widespread use of assisted reproductive technology and the use of first trimester obstetric ultrasound examinations, the incidence of TRAP sequence has increased from 1% in 1953[12] to 2.6% in 2015.[13] In addition, in utero therapy has improved the overall prognosis for TRAP sequence,[4] and as a result, complications other than TRAP sequence in the third trimester have become clinically evident. Clinicians should not only monitor the prognosis of patients with TRAP sequence but also pay sufficient attention to the complications of monochorionic twins.

Conclusion

Although in utero therapy has improved the prenatal outcome of TRAP sequence, the indications for in utero therapy remain controversial. We should use predictive indicators for prognosis from early pregnancy by ultrasonic measurement. Clinicians should give the same attention to the management of post-TRAP sequence as TRAP sequence itself, with the coordination of local clinics and hospitals.

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Ethical approval

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Informed consent

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