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CASE REPORT

Monochorionic–diamniotic discordant growth in a twin pregnancy with one fetus affected by Ebstein's anomaly of tricuspid leaflets

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Key Clinical Message

Our patient was diagnosed as having discordant twin growth with Ebstein's anomaly in the larger fetus. Cardiac function was deteriorated in accordance with progression of gestational age. Our observation indicated cardiac failure of the larger fetus. The most important issue in this situation is management of the timing of delivery.

Keywords

Cardiac function, Ebstein's anomaly, twin pregnancy.

Introduction

Ebstein's anomaly is a congenital malformation of the heart that is characterized by apical displacement of the septal and posterior tricuspid valve leaflets. This condition leads to atrialization of the right ventricle, with a variable degree of malformation and displacement of the anterior leaflet. Ebstein's anomaly has a prevalence of 0.2–0.5% of all cardiac malformations [1, 2]. One-third of prenatally diagnosed patients with Ebstein's anomaly will die in utero and the 1-year survival rate is only 15% [3, 4]. These patients almost always have severe disease with tricuspid regurgitation and significant cardiomegaly. We present a case of fetal severe Ebstein's anomaly that was diagnosed by fetal echocardiography in monochorionic–diamniotic twins. We report the management of Ebstein's anomaly in one fetus of discordant twins.

Case Report

A 21-year-old gravida 1, para 0 woman at 10 weeks' gestational age was referred for prenatal management for a monochorionic–diamniotic twin pregnancy. Her medical history was unremarkable. She had no family history of congenital abnormalities or multiple pregnancies. The patient was suspected as having discordant twin growth with Ebstein's anomaly in the larger fetus at 20 weeks' gestation. In the smaller fetus, we observed oligohydramnios and biometry at the first percentile, with normal structural anatomy. In the larger fetus, a fetal structural survey showed no structural abnormalities. Fetal echocardiography was performed with two-dimensional, M-mode Doppler, and color flow imaging. The findings were consistent with Ebstein's anomaly with severely downward displaced tricuspid valve leaflets tethered at the muscular ventricular septum (including the trabecular portion of the right ventricle) (Fig. 1A), right ventricular atrialization, a small effective right ventricular cavity, and cardiomegaly (cardiothoracic area ratio: 40%). The echocardiogram also showed moderate to severe tricuspid valve regurgitation (Fig. 1B). These findings did not match the criteria of twin-to-twin transfusion syndrome [5]. The patient received periodic sonographic evaluation for changes in cardiac function. Both fetuses grew according to gestational age with 40–45% of estimated fetal body weight discordancy (Fig. 2A). The

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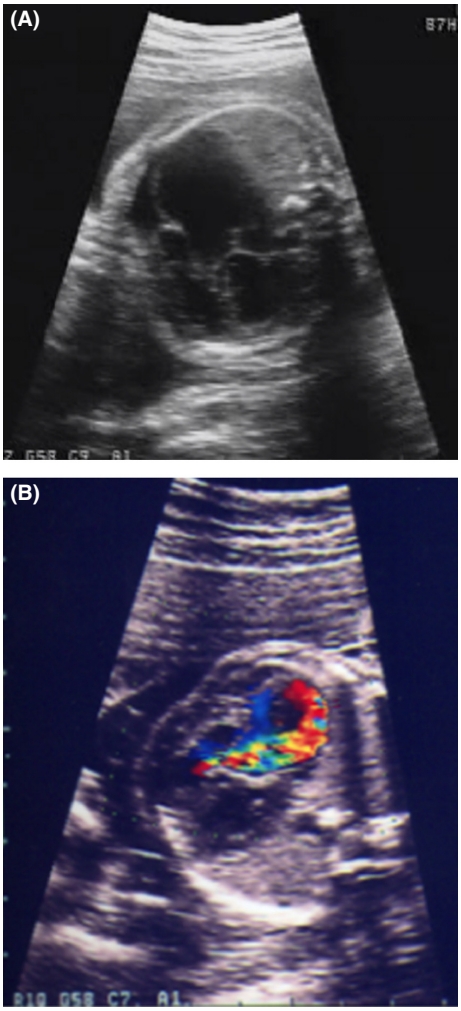

Figure 1. Fetal echocardiography at 23 weeks of gestation. (A) Four-chamber view of the heart. Ebstein's anomaly with severely downward displaced tricuspid valve leaflets is shown. (B) Tricuspid valve regurgitation.

amniotic fluid pocket of the smaller fetus gradually increased and caught up to the larger fetus at 27 weeks of gestation (Fig. 2B). In the larger fetus, the cardiothoracic area ratio was enlarged to greater than 50% (Fig. 2C), the ejection fraction of the left ventricle was less than 80% (Fig. 2D), and the preload index was increased to greater than 50% (Fig. 2E) at 27 weeks of gestation.

Cardiac function was deteriorated in accordance with progression of gestational age, which indicated cardiac failure of the larger fetus (Table 1). Cardiac function of the smaller fetus was stable over time (Table 1). Cesarean section was performed at 31 + 2 weeks' gestation because of the high risk of intrauterine demise owing to cardiac failure of the larger fetus. This procedure was performed because of profound consequences for the outcome of the healthy twin. The larger neonate had a birth weight of 1301 g (25th percentile) and Apgar scores of 3 and 9 at 1 and 5 min, respectively, with cyanosis. The smaller neonate had a birth weight of 819 g (10th percentile) and Apgar scores of 5 and 7 at 1 and 5 min, respectively. The smaller neonate was transferred to the intensive care unit. There was no evidence of findings of twin-to-twin transfusion syndrome. The cardiothoracic ratio was 100% on a chest X-ray (Fig. 3). An echocardiogram showed severe Ebstein's anomaly with pulmonary stenosis, severe tricuspid valve regurgitation (grade 4), and atrial enlargement. We attempted to rescue the larger newborn for 7 h, but were unsuccessful. The cause of death was lung hypoplasia by cardiomegaly. The smaller infant was provided nasal continuous positive airway pressure for 2 days and supplemental oxygen for one further day. During the neonatal intensive care unit stay, the developing bronchopulmonary dysplasia was treated with synthetic surfactant therapy. The smaller newborn showed normal development at 2 years of age.

Discussion

Many cases of Ebstein's anomaly involve some degree of tricuspid regurgitation and stenosis. Increased right atrial pressure and volume enlarge the right atrium, resulting in a right-to-left shunt if a pathway exists [1, 2]. Generally, twin pregnancies are at higher risk for congenital heart disease than singletons. Singletons have a prevalence of congenital heart disease of 2% compared with 1.7–7.5% in all twin pregnancies. Monochorionic twins have a higher prevalence of congenital heart disease than dichorionic twins [6, 7]. Unequal division of the embryonic cell mass could be a risk factor for development of congenital heart disease in monochorionic twins. Monochorionic twins are associated with a high perinatal mortality and morbidity [8–10]. Abnormal vascular connections between infant twins and blood volume fluctuations or ischemia cause damage in multiple organs [8–10], especially in twin-to-twin transfusion syndrome. Doppler parameters were introduced to investigate blood flow in fetus to evaluate the fetal well-being and useful for decision making of the optimal time for termination the pregnancy. Umbilical artery and middle cerebral artery Doppler findings appear to be abnormal in the early stage of placental insufficiency. The preload index reflects ductus venous flow abnormalities and has been described as a late marker. This is because these abnormalities occur within the last week before delivery, reflecting an adverse perinatal outcome and neurological damage [11–14]. Fetal cardiac function analysis in twins shows their hemodynamic status and this finding may improve understanding of the development of growth discordance. When one

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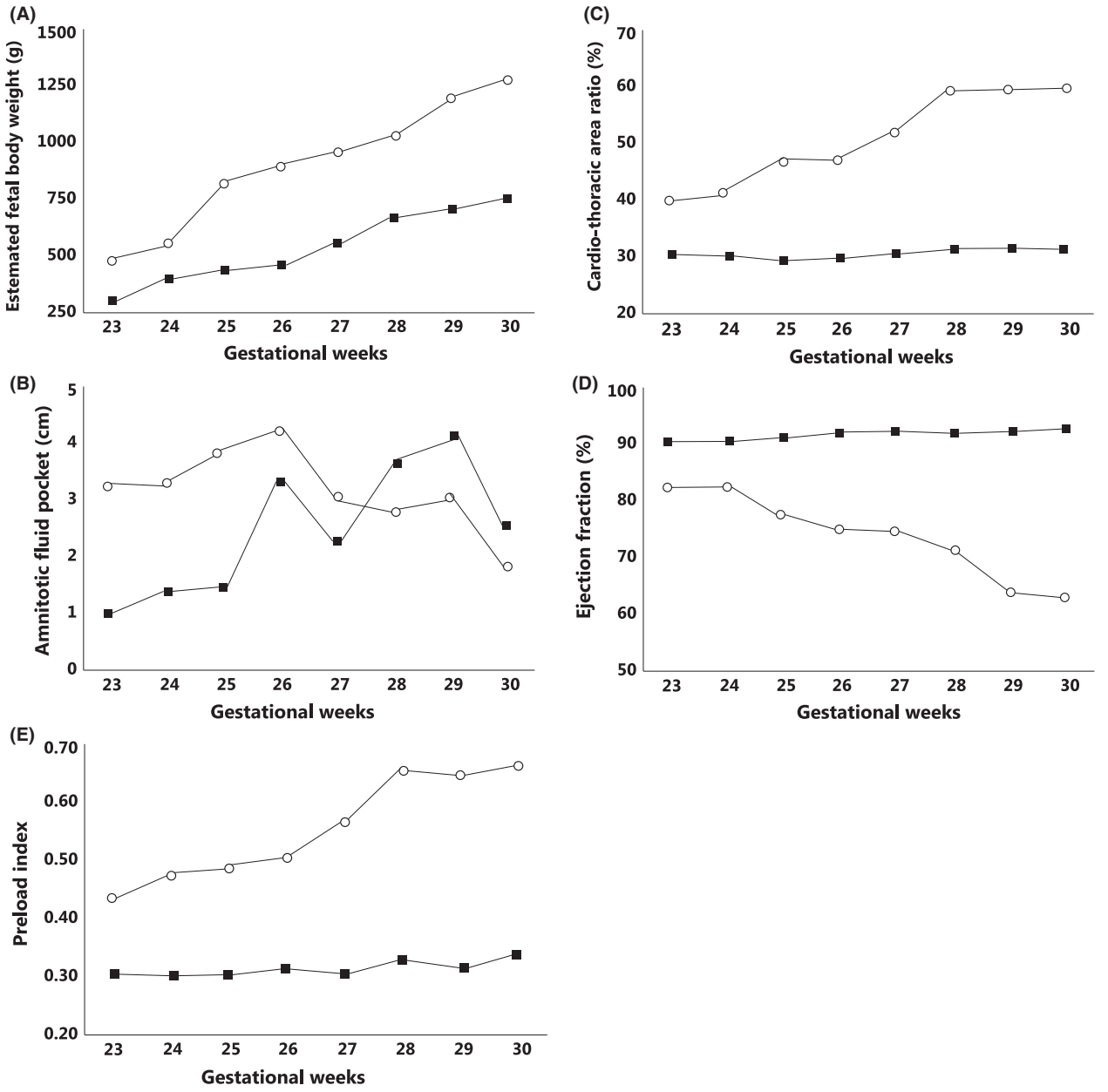

Figure 2. Changes in parameters with gestational age. (A) Estimated fetal body weight (g). (B) Amniotic fluid pocket (cm). (C) Cardiothoracic area ratio (%). (D) Ejection fraction (%). (E) Preload index.

twin dies, a sufficient amount of thromboplastin from dead tissues may be released, causing generalized clotting problems in the survivor [15]. Neurological abnormalities of a surviving co-twin occur in 18% of monochorionic twins [16]. To avoid intrauterine fetal death in our case, we assessed fetal cardiac function. We diagnosed that the larger fetus had a poor prognosis. Our priority was to rescue the smaller fetus with intact survival. Fetal well-being was assessed by a biophysical profiling score, fetal blood flow analysis (middle cerebral artery resistance index, umbilical artery resistance index), and fetal cardiac functional evaluation, including the cardiothoracic area ratio, ejection fraction of the left ventricle, preload index of the inferior vena cava, and severity of tricuspid valve regurgitation. Cardiac function was deteriorated according to progression of gestational age in the larger fetus. This finding indicated cardiac failure of the larger fetus. The amniotic fluid pocket of the smaller fetus gradually increased and caught up to the larger fetus at 27 weeks of gestation.

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Table 1. Change in parameters with gestational age.

Gestational age (week)	23	24	25	26	27	28	29	30
Larger fetus								
EFBW (g)	494	532	779	811	939	1016	1206	1272
AF pocket (cm)	3.1	3.2	3.8	4.3	3	2.7	2.8	2
CTAR (%)	40	41.5	45	45	50	57	55	54
EF (%)	84	84	75	73	74	70	64	64
PLI	0.426	0.467	0.477	0.488	0.555	0.632	0.606	0.608
MCA RI	0.87	0.87	0.83	0.82	0.87	0.76	0.79	0.8
UmA RI	0.52	0.56	0.6	0.53	0.59	0.66	0.69	0.67
Smaller fetus								
EFBW (g)	278	348	346	422	540	629	688	801
AF pocket (cm)	1.2	1.5	1.6	3.2	2.3	3.5	3.9	2.6
CTAR (%)	30	28	27	30	31	28	28	28
EF (%)	90	90	91	91	89	91	90	92
PLI	0.300	0.312	0.352	0.332	0.282	0.333	0.287	0.340
MCA RI	0.69	0.7	0.72	0.74	0.77	0.86	0.84	0.85
UmA RI	0.77	0.61	0.59	0.71	0.73	0.67	0.71	0.68

EFBW, estimated fetal body weight; MCA, middle celebral artery; AF, amniotic fluid; UmA, umbilical artery; CTAR, cardiothoracic area ratio; RI, resistant index; EF, ejection fraction (of left ventricle); PLI, preload index.

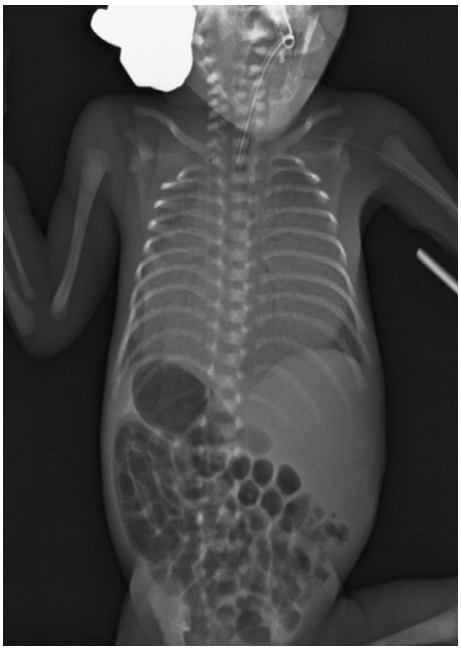

Figure 3. Neonatal chest X-ray of the larger fetus.

In conclusion, a smaller twin fetus of discordant growth increases the risk of an adverse pregnancy outcome by prematurity [17]. The timing of delivery is an important issue in the management of affected fetuses.

Consent for Publication

The patient's parents provided consent for the publication of this case report and any accompanying images.

Conflict of Interest

The authors declare that they have no conflict of interest.

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