Modified underlying cardiac disease severity in twin-twin transfusion syndrome

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ABSTRACT

Twin-twin transfusion syndrome or related conditions affect fetal loading. We report monochorionic-diamniotic twins. Twin 1 had Ebstein anomaly with mild tricuspid regurgitation (TR) and slightly thickened tricuspid valve leaflets with plastering. Twin 2 had tricuspid valve dysplasia (with abnormal thickening but without plastering) with moderate TR and mild right atrial dilatation. After birth, the severity of TR was greatly reduced in the recipient but increased in the donor. Therefore, intravascular volume change which was due to twin-twin transfusion syndrome seemed to affect the severity of the valvar disease in fetuses. This case suggests that the intrinsic severity of fetal tricuspid valvular disease may be overestimated in the recipient and underestimated in the donor twin. These factors need to be taken into consideration in clinical decision-making.

Keywords: Fetal echocardiography, tricuspid insufficiency, twin amniotic fluid discordance, twin-twin transfusion syndrome

INTRODUCTION

Pathology can be induced by vascular communication between monochorionic-diamniotic twins as occurs in twin-twin transfusion syndrome and related conditions. Twin amniotic fluid discordance (TAFD)[1] is one such condition. TAFD in monochorionic twins is characterized by one amniotic pocket measuring 3 cm or less and the other measuring 7 cm or more.[2] However, in severe twin-twin transfusion syndrome, one pocket measures 2 cm or less and the other measures 8 cm or more.[3] TAFD may have a circulatory impact on the fetus similar to that of severe twin-twin transfusion syndrome[1] and can even progress to severe twin-twin transfusion syndrome.[2] In twin-twin transfusion syndrome, large volume overloading is imposed on the recipient twin, with volume unloading in the donor twin. Large volume overloading in the recipient twin causes fetal heart failure, and volume unloading in the donor twin causes fetal dysfunction. The extent to which loading alterations induced by TAFD modify the intrauterine severity of tricuspid regurgitation (TR) in monochorionic-diamniotic twins with tricuspid valve disease remains unclear.

We recently experienced a case of monochorionic-diamniotic twins where TAFD modified the fetal severity of TR, making it difficult to precisely assess intrinsic valve disease before delivery.

CASE REPORT

A monochorionic-diamniotic twin case was referred to our department at 20 weeks of gestation. Ebstein anomaly was diagnosed in Twin 1, with mild TR

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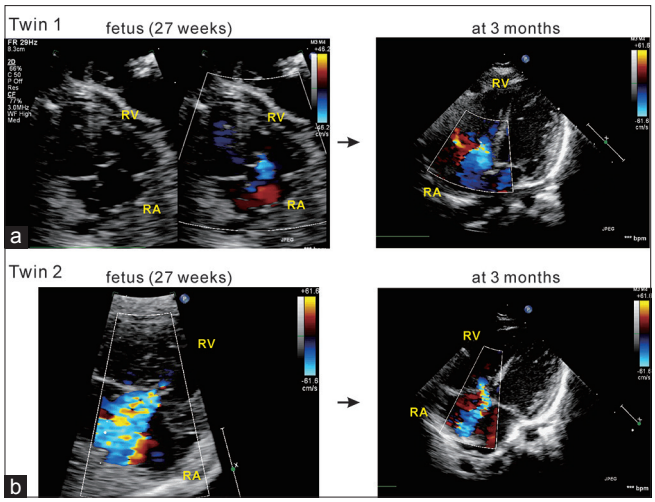

Figure 1: Changes in tricuspid regurgitation severity before and after birth. Left panels showing fetal echocardiograms at 27 weeks and 6 days, and right panels showing echocardiograms at 3 months after birth. (a) In Twin 1, the donor in twin amniotic fluid discordance circulation, severity of tricuspid regurgitation worsened after birth (from "mild" to "moderate"). (b) In Twin 2, the recipient of twin amniotic fluid discordance circulation, severity of tricuspid regurgitation was ameliorated after birth (from "moderate to severe" to "moderate")

and slightly thickened tricuspid valve leaflets with plastering [Figure 1a, left]. Twin 2 had moderate TR and mild right atrial dilatation (Celermajer index = 0.43). Twin 2 had abnormal thickening without plastering of the tricuspid valve [Figure 1b, left] and was diagnosed with tricuspid valve dysplasia. There was almost no difference in the growth of the twins. While Twin 1 remained stable, Twin 2 gradually developed worsening and ultimately severe TR, with a cardiothoracic area ratio of 48% and a Celermajer index of 0.82 at 33 weeks. However, there were no abnormalities in umbilical flow patterns or pooling of effusion or hydrops. A TR flow rate of 3.5 m/s indicated that right ventricular pressure was sufficient for antegrade ejection into the pulmonary artery. At 35 weeks, maximum amniotic fluid depth met the criteria for TAFD[2] [Table 1]: 3 cm in Twin 1 and 8.9 cm in Twin 2. In addition, the middle cerebral artery flow rate[4] in Twin 1 increased to 89 cm/s, suggesting twin-twin transfusion syndrome circulation. The cardiothoracic area ratio further increased to 56% in Twin 2, with severe TR. However, the TR flow rate increased to 4 m/s in Twin 2, and the velocity time integral increased in both the aorta and pulmonary artery, suggesting favorable adaptation to the increased circulating blood volume caused by twin-twin transfusion syndrome circulation. This case did not meet the criteria of twin-twin transfusion syndrome before 26 weeks. Moreover, when maximum amniotic fluid depth met the criteria of twin-twin transfusion syndrome, the case became out of indication of the laser treatment because of over gestational weeks.

Table 1: Characteristics of the twins

	Twin 1	Twin 2
Disease	Ebstein anomaly	TVD
Fetal echocardiography at 35 weeks 2 days		
Maximum amniotic fluid depth (cm)	3	8.9
TR	Mild	Severe
TR pressure gradient	N/A	64
Celermajer index	0.43	0.82
Neonatal information at 35 weeks 3 days		
Gender	Female	Female
Birth weight (g)	1598	1594
Birth length (cm)	42.5	40.3
Apgar score (1, 5 min)	8–9	8–9
Hemoglobin (g/dL)	16.3	17.7
Albumin (g/dL)	3.3	3.2
CTR (%) at 0 day	55.5	70.2

TVD: Triple-vessel disease, TR: Tricuspid regurgitation CTR: Cardio-thoracic ratio

The twins were delivered by semi-emergent cesarean section at 35 weeks and 3 days because of rapidly progressive TAFD. Both twins were stable after birth. Twin 2 required oral diuretic therapy and temporary oxygen administration to stabilize the pulmonary circulation. At 3 months after birth, both infants had a cardiothoracic area ratio of about 50% on chest X-ray and showed well-balanced, cross-sectional four-chamber view on echocardiography [Figure 1, right]. The severity of TR worsened in Twin 1 but was markedly ameliorated in Twin 2 after birth [Figure 1]. The placenta showed an extremely thick anastomotic vessel between the twins.

DISCUSSION

To our knowledge, this is the first description that loading differences caused by twin-twin transfusion syndrome circulation result in altered fetal TR severity in reciprocal directions in congenital tricuspid valve disease. We think that the severity of TR was overestimated in Twin 2 because of volume overload by twin-twin transfusion syndrome and that the severity of TR was underestimated in Twin 1 because of volume unload by twin-twin transfusion syndrome.

Twin 2 (recipient) had progressively worsening TR with tricuspid valve dysplasia accompanied by marked right heart dilatation during gestation. Since there was rapid worsening during the third trimester, there was concern about increasing disease severity and even intrauterine fetal death.[1] However, right heart blood flow was maintained without obvious signs of fetal heart failure. TR markedly decreased after birth when the vascular communication between the twins was terminated [Figure 1b] and did not require aggressive treatment. Given the severity of TR and marked enlargement of the right atrium in Twin 2, such improvement after birth was beyond the expected

range of TR amelioration after birth,[5] due to reduction of pulmonary vascular resistance.[6-8] In contrast, TR worsened in Twin 1 (recipient), progressing from mild severity in the fetus to moderate severity after birth [Figure 1a]. Therefore, intravascular volume change which was due to twin-twin transfusion syndrome seemed to affect the severity of the valvular disease in fetuses.

This case indicates that twin-twin transfusion syndrome circulation may have exerted significant influence on fetal pathology; the intrinsic severity of fetal tricuspid valve disease may be overestimated in the recipient and underestimated in the donor twin.

As in our case, it is extremely difficult to determine to what extent twin-twin transfusion syndrome circulation modifies circulatory dynamics in monochorionic twins when they have shunts or valve lesions. Thus, caution should be exercised in predicting the prognosis and deciding the treatment strategy when in discussion with the family. Accumulation of future cases with such lesions and twin-twin transfusion syndrome circulation is needed to further clarify the relationship between fetal and postnatal hemodynamics.

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Conflicts of interest

There are no conflicts of interest.

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