Ultrasound Obstet Gynecol 2005; **25**: 514–516
Published online 9 February 2005 in Wiley InterScience (www.interscience.wiley.com). **DOI:** 10.1002/uog.1728

Color Doppler flow pattern in antenatal diagnosis of unguarded tricuspid valve

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KEYWORDS: Doppler; prenatal; unguarded tricuspid valve

ABSTRACT

We describe an antenatally diagnosed case of unguarded tricuspid valve in a 15-week fetus. Because of its rarity, the differentiation of this anomaly from other tricuspid valve dysplasias is difficult. Tricuspid regurgitation is the hallmark of Ebstein's anomaly and other tricuspid valve abnormalities, whereas in unguarded tricuspid orifice there is total absence of the valve, resulting in 'to and fro' flow across the right-sided chambers. The color Doppler flow pattern helps in differentiating this anomaly from others. The antenatal diagnosis was confirmed on autopsy.

CASE REPORT

A 27-year-old woman, gravida 4 para 2 abortion 1, with one 5-year-old normal child, was referred at 15 weeks of gestation to rule out congenital anomalies. Her third female child died on day 11 and had the following congenital anomalies: absent radius and ulna, hypoplastic palms and fingers in both upper limbs, absent tibia and fibula, deformed feet, hypoplastic toes in both lower limbs and hypoplastic tongue.

An ultrasound examination performed at 15–16 weeks of gestation revealed cardiomegaly, dysplastic tricuspid valve and dilated right ventricle (Figure 1). Retrospective analysis of the color Doppler images revealed 'to and fro' flow across the right ventricle and right atrium (Figure 2). There was difficulty in imaging the pulmonary artery. Amniocentesis revealed a normal karyotype.

A repeat scan was performed at 19 weeks of gestation. Cardiomegaly was present. The right atrium was enlarged. The tricuspid valve was markedly dysplastic. The septal cusp was rudimentary. The anterior leaflet showed redundant movement on real time. The right ventricle was dilated with thin myocardium. The subvalvular apparatus was not seen. The pulmonary artery was markedly hypoplastic and no flow was present across it. Minimal pericardial effusion was present. The final impression given was of severe tricuspid dysplasia with marked to and fro flow across the valve area with pulmonary atresia.

The patient opted for termination of pregnancy. A detailed autopsy revealed a dilated right atrium, atrial appendage and right ventricle. Marked thinning of the right ventricular myocardium was observed, which was confirmed on histopathology. The tricuspid valve leaflets were absent (Figure 3) and the pulmonary artery was narrow with an atretic pulmonary valve. These features are consistent with unguarded triscupid valve orifice with pulmonary atresia. The left ventricular myocardium appeared normal.

DISCUSSION

The common tricuspid valve abnormalities diagnosed *in utero* are Ebstein's anomaly and tricuspid valve dysplasia[1]. Unguarded tricuspid orifice is a rare abnormality of the tricuspid valve and its differentiation from Ebstein's anomaly may be difficult by echocardiography[2]. Although several cases of unguarded tricuspid orifice have been diagnosed in the pediatric age group[3–5], to our knowledge, this is the first case to diagnose it antenatally. Marked thinning of the right ventricular myocardium is present in this condition. Castellanos *et al*. postulated that a primary pathogenetic step occurs in the wall of the right ventricle and this prevents the morphogenesis of the tricuspid valve from the ventricle[6]. The differential diagnosis for this anomaly includes Ebstein's anomaly, Uhl's disease and arrhythmogenic right ventricular dysplasia.

In Ebstein's malformation there is annular attachment of the septal and mural leaflets within the right ventricle rather than at the atrioventricular junction. This results

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Accepted: 29 November 2004

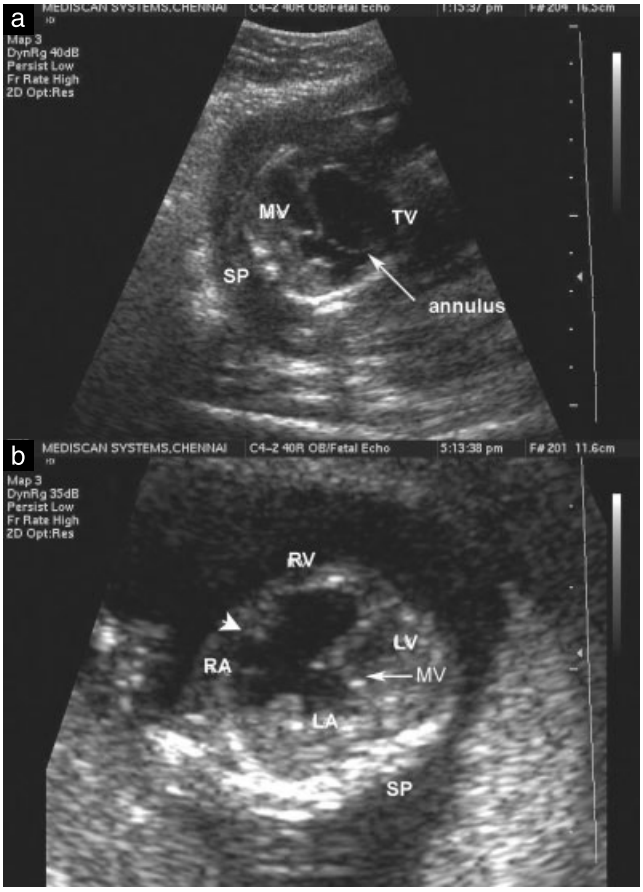

Figure 1 Ultrasound images of: (a) apical four-chamber view of the heart showing grossly dilated right ventricle with thin myocardium; (b) right ventricle (RV) with thin myocardium and absence of valve during systole (arrowhead indicates atrioventricular junction; mitral valve (MV) seen in closed position (arrow)). LA, left atrium; LV, left ventricle; RA, right atrium; SP, spine; TV, tricuspid valve.

developmentally from failure of liberation of these leaflets from the ventricular wall. As a result of this, the septal leaflet is placed low and is dysplastic but the anterosuperior leaflet is placed normally[7]. Ebstein's malformation has an extremely variable natural history depending on the degree of abnormality of the tricuspid valvular apparatus, which may range from mild to severe[8]. Severe tricuspid valve abnormality leads to profound congestive heart failure, whereas mild displacement of the hinge of the valve away from the atrioventricular junction may remain asymptomatic until late in adult life, or the person may remain symptomless throughout life[9]. The routine four-chamber view of the heart at 18–20 weeks of gestation reveals a dilated right atrium with a rudimentary mural leaflet of the tricuspid valve. On color Doppler, marked tricuspid regurgitation is present during systole due to abnormal closure of the mural leaflet. The right ventricular chamber appears small. Less severe forms of Ebstein's anomaly can appear normal in the early weeks of gestation and can present with dilated right atrium and tricuspid regurgitation in the third trimester or postnatally.

In Uhl's anomaly, there is total absence of myocardium of the right ventricle with normal septal trabeculations and normal structure of tricuspid and pulmonary valves[8].

Arrhythmogenic right ventricular dysplasia presents in infancy and childhood. There is degeneration of the right ventricular myocardium with replacement of fibrofatty tissue[10]. The tricuspid valve appears normal.

In unguarded tricuspid orifice, the septal cusps, chordae tendinae and papillary muscles are absent[3]. Due to the absence of the tricuspid valve and associated pulmonary atresia, there is to and fro flow across the tricuspid orifice. Enlargement of the right ventricular cavity is secondary to pulmonary narrowing. The annulus normally appears echogenic and should not be mistaken for the valve leaflet. In our case, the annular ring was seen flapping in the right atrial cavity on real time, and was mistaken as the valve leaflet. The right-sided chambers appear as an elongated single chamber due to the absence of the valves. The left atrium and ventricle appear relatively small due to reduced filling of the chambers.

The diagnosis of unguarded tricuspid valve orifice with pulmonary atresia can be made early in gestation, due to the total absence of identifiable valve tissue and the dilated right ventricle. Very rarely, unguarded tricuspid valve can be partial, in which case it becomes difficult

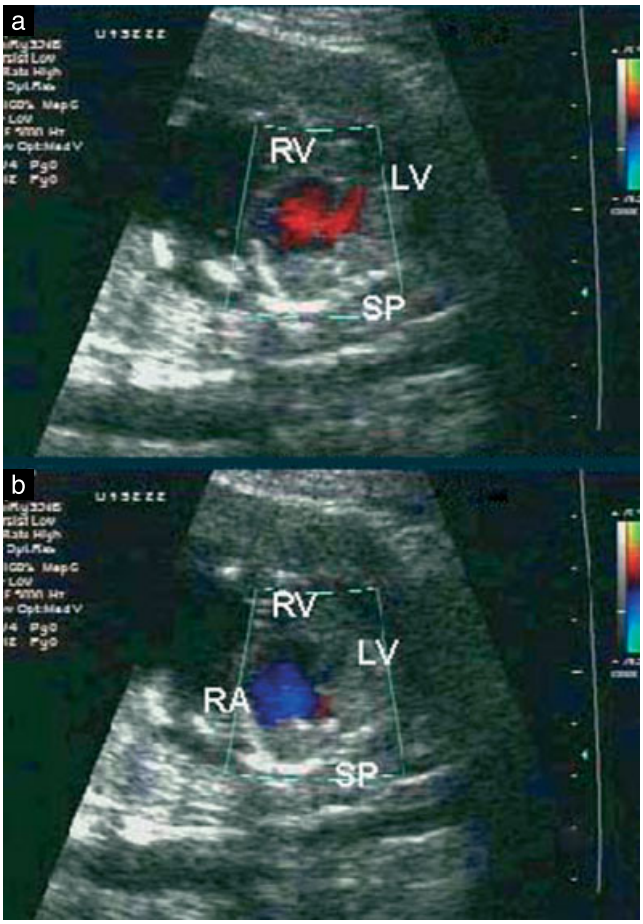

Figure 2 Color Doppler ultrasound images showing (a) ventricular filling (red) during diastole and (b) reversal of flow (blue) in the right atrium (RA) during systole. LV, left ventricle; RV, right ventricle; SP, spine.

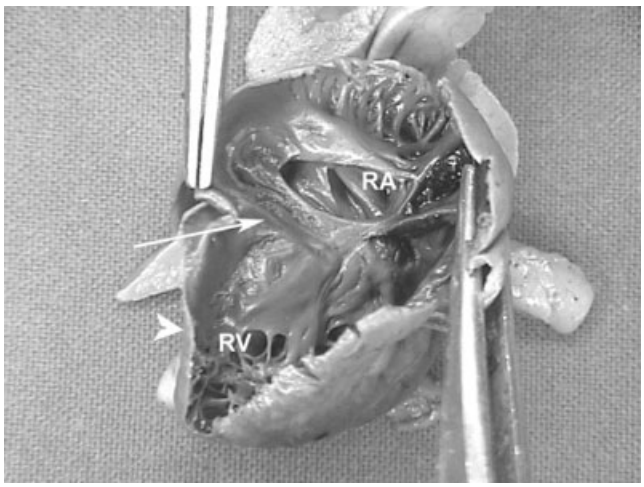

Figure 3 Autopsy photograph of the dissected heart showing absent tricuspid valve (long arrow represents atrioventricular junction). Note the thinning of the right ventricle (RV) myocardium (arrow head). RA, right atrium.

to differentiate from Ebstein's anomaly[2]. Unguarded tricuspid valve orifice, both partial and complete agenesis of valvular tissue with normal right ventricular outflow tract, can manifest late in childhood or rarely in adulthood[5,11–13].

Severe forms of Ebstein's anomaly and tricuspid dysplasias diagnosed *in utero* carry a poor prognosis[14] due to the presence of severe regurgitation, leading into right atrial enlargement and cardiac failure. Lung hypoplasia is due mainly to the space-occupying effect of the massive cardiomegaly[15]. The manifestation of the problem in patients with unguarded tricuspid valve orifice depends on the patency of the right ventricular outflow tract, whereas in Ebstein's anomaly the manifestation depends on the degree of displacement of the mural leaflet of the tricuspid valve into the right ventricular cavity.

To conclude, differentiation of unguarded tricuspid orifice from Ebstein's anomaly can be made by identifying the mural leaflet on B-mode. However, it may be difficult to differentiate severe forms (when the mural leaflet is rudimentary) of Ebstein's anomaly from partial agenesis of unguarded tricuspid valve orifice. Color Doppler helps in differentiating between the two conditions.

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