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Brief Report

Unguarded tricuspid valvar orifice in the fetus

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Abstract The unguarded tricuspid valve is uncommon. We describe herein a fetus with a grossly dilated right ventricle and atrium, with severe tricuspid and pulmonary valvar regurgitation. The right ventricle was akinetic, and no tricuspid tissue or valvar apparatus was identified. Colour Doppler showed a highly unusual retrograde flow of blood through the right heart. The pregnancy was terminated, and necropsy examination confirmed the gross dilation of the right heart chambers, with severely dysplastic valvar tissue at the right atrioventricular junction effectively giving an unguarded orifice. There was no valvar displacement, and the left heart was normal. The fetus had a normal karyotype, albeit with absent kidneys.

Keywords: Dysplastic tricuspid valve; fetal echocardiography; right-to-left shunting

TRICUSPID VALVAR DISEASE ENCOMPASSES A spectrum from absent connection to complete absence of the leaflets resulting in an unguarded orifice. Initially described by Kanjuh et al.,[1] the unguarded tricuspid valve remains a rare malformation, usually associated with pulmonary atresia and intact ventricular septum.[2] It can be confused with Ebstein's malformation in the setting of a dilated right atrium and ventricle with significant tricuspid regurgitation.[2] We describe diagnosis of this rare lesion in a fetus, emphasizing the highly unusual pattern of flow of blood through the heart.

Case report

A fetal echocardiogram was performed on a fetus at 20 weeks gestation following the prenatal detection of multiple congenital abnormalities, including severe oligohydramnios, absent kidneys, and a positive triple marker screen for Trisomy 18. Cardiac ultrasound revealed normal arrangement of the abdominal organs and a left-sided heart. The striking feature seen in the off-axis four-chamber view was the grossly dilated, akinetic, and thin walled right ventricle, raising the suspicion for Uhl's anomaly (Fig. 1). There was no visible tricuspid valvar tissue movement, and it proved impossible to identify any tricuspid valvar mechanism. An echo density was seen separating the

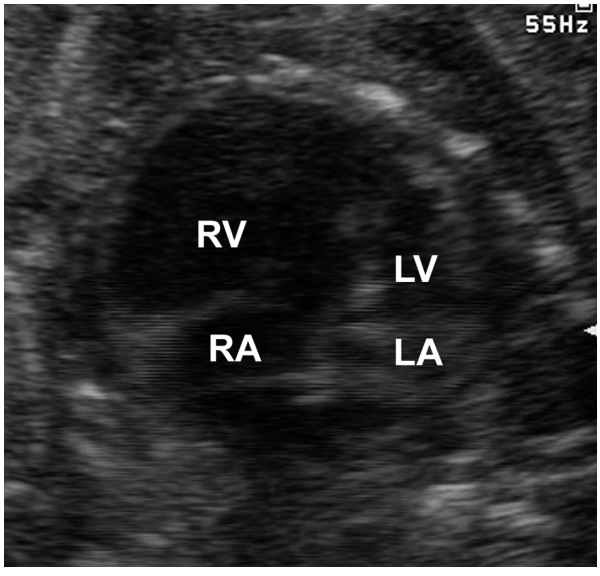

Figure 1.
Four-chamber view highlighting the dilated right ventricle (RV) and right atrium (RA). The left atrium (LA) and left ventricle (LV) are also shown.

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right atrium and ventricle in keeping with the right atrioventricular groove (Fig. 1). The left heart was normally developed, although there was regurgitation across the mitral valve. The pulmonary valve was present, but permitted severe regurgitation, such severe regurgitation also being seen across the unguarded tricuspid valvar orifice. In other views, retrograde flow of blood was seen across the arterial duct, which swirled into the grossly dilated right ventricle. Only retrograde flow was identified across the tricuspid valve. There was right-to-left shunting across the oval fossa, with no ventricular septal defects.

In the setting of such complex congenital cardiac disease, coupled with other obvious major congenital abnormalities and probable Trisomy 18, the family chose to terminate the pregnancy. A necropsy was performed, and confirmed gross dilation of the right ventricle and atrium. The right ventricle was sparsely trabeculated, and the free wall measured 2–3 mm in thickness (Fig. 2). Dysplastic tissue representing the leaflets of the tricuspid valve was found at the right atrioventricular junction, but it was very nodular, and it proved impossible to identify the individual valvar leaflets. There was no displacement of the valvar remnants from the junction, however, and the rudimentary chordal apparatus was heavily muscularised. The leaflets of the pulmonary valve were also nodular, albeit not thickened. The left heart appeared normal. Cytogenetic analysis showed a normal male karyotype, and no 22q11 micro-deletion.

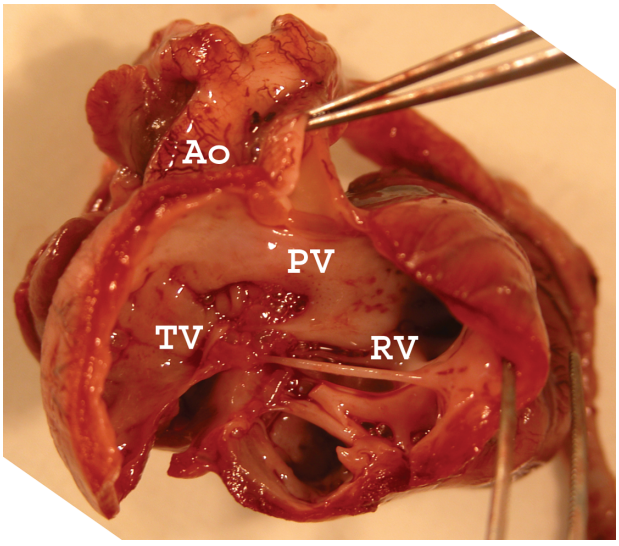

Figure 2.
View of the autopsied heart with the right ventricle opened, with the apex directed rightward and the great vessels superiorly. The right ventricle (RV) is dilated and sparsely trabeculated. Dysplastic and nodular tricuspid valvar (TV) tissue is attached at the level of the atrioventricular junction, and is supported by muscularized cords. The pulmonary valve (PV) is nodular but not thickened.

Discussion

The most striking aspect of our fetus was the retrograde flow of blood through the right heart. Along with the grossly dilated and akinetic right ventricle, and severe tricuspid regurgitation, we speculated on this being an example of Uhl's anomaly. This notion, however, was disproved by the necropsy examination, which revealed the presence of grossly dysplastic rudiments of the tricuspid valve attached at the level of the normal right atrioventricular junction. Anderson et al.[2] described the absence of the mural leaflet as an anatomic guide to diagnosing unguarding of the tricuspid valvar orifice, distinguishing this from the displaced mural leaflet found in Ebstein's malformation. The leaflets in our case were too dysplastic and clustered to identify accurately the mural component, but in the absence of displacement, and in the setting of severe regurgitation, we diagnosed our case as having effective unguarding of the tricuspid valvar orifice secondary to severe dysplasia of the leaflets. Freedom et al.[3] have described pulmonary regurgitation in association with florid tricuspid regurgitation in the neonate, and this combination may explain the unusual hemodynamics seen in our case.

Unguarding of the tricuspid valvar orifice is seen most frequently with pulmonary atresia and intact ventricular septum, usually resulting in a dilated, rather than an hypoplastic, right ventricle.[2] There are, however, case reports of the unguarded tricuspid valve orifice as an isolated anomaly presenting in childhood and late adulthood, progressing to severe right heart failure.[4–6] Other complications that have been described include atrial tachycardias and formation of thrombus secondary to the grossly dilated atrium.[7,8]

Hornberger et al.[9] reviewed 27 fetuses with tricuspid valvar disease and significant regurgitation. They identified 2 fetuses with an unguarded tricuspid valvar orifice, and 17 with Ebstein's anomaly. Overall, these lesions were associated with a high mortality. In those with only tricuspid valvar disease, a mechanical valve has been placed in the unguarded orifice.[10] Otherwise, surgery is dictated by the accompanying lesions, and often involves palliation by the functionally univentricular route. We hope to contribute to the understanding of this entity by adding our description of the retrograde flow of blood in the right heart in the setting of a severely dysplastic tricuspid valve effectively resulting in an unguarded valvar orifice.

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