Foetal right ventricular outpouching associated with ventricular bigeminy

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Abstract

Introduction
Congenital cardiac right ventricular diverticula are transmural localised protrusions, within the free wall of the ventricles. These diverticula are infrequently diagnosed during the foetal period, and because of their rarity, their natural history remains unclear. We present a case of a prenatal diagnosis of right ventricular diverticulum at 26 weeks gestation associated with an irregular rhythm of ventricular bigeminy.

Case report
We present a case of a 32-year-old G1P1, who was referred at 26 weeks gestation on account of an abnormal rhythm and four-chamber view on a routine screening. Foetal echocardiography showed a large right ventricular outpouching (RVO) of 11 × 9 mm, with unrestricted flow, across a large orifice and situated along the lateral free wall, just beneath the tricuspid valve.

Conclusion
Foetal RVO are rare congenital malformations that need to be considered, when an extra chamber is visualised in the routine four-chamber view, in the assessment of foetal arrhythmia or presence of a pericardial effusion.

Introduction
Right ventricular (RV) aneurysms and diverticula are congenital heart defects that occur infrequently. The distinguishing feature between the two is made histologically within the wall of the outpouching, with absence

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Figure 1A: Four-chamber view. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle. White arrow indicates right ventricular outpouching.

Figure 1B: Short-axis view of the ventricles. LV, left ventricle; RV, right ventricle. White arrow indicates right ventricular outpouching.

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of myocardium in the aneurysm and presence of myocardium in the diverticulum. RV aneurysms tend to have a wide connection to the ventricle and move paradoxically in systole, whilst diverticula tend to have a narrow connection to the ventricle and contract during systole. Echocardiographic imaging to distinguish between the two, prenatal and postnatal diagnosis of right ventricular diverticulum, may be difficult even in the best setting. For this reason, the term right ventricular outpouching (RVO) has been coined to include both the entities.

We report a case of foetal RVO associated with irregular rhythm due to ventricular bigeminy.

Case report
We present a case of a 32-year-old G1P1, who was referred at 26 weeks gestation, on account of an abnormal rhythm and four-chamber view, on routine screening. Foetal echocardiography showed a large RVO of 11 × 9 mm (Figures 1A and 1B), with unrestricted flow, across a large orifice and situated along the lateral free wall, just beneath the tricuspid valve. Biventricular systolic function was preserved even though the rhythm was bigeminy. Doppler flow interrogation of the arteries, atrioventricular (AV) valve and superior vena cava (SVC), and motion (M)-mode echocardiography of atria and ventricles identified ventricular bigeminy (Figures 2A and 2B). There were no pericardial or pleural effusions observed. Chambers were not dilated. Doppler flow of the ductus venosus showed no retrograde flow. The normal foetal circulation was preserved with right to left shunting at atrial level and through the ductus arteriosus. The patient was followed with a serial foetal ultrasound scan every two weeks, with a stable, uneventful antenatal course. Foetal growth parameters remained on the appropriate centiles.

A female infant weighing 3,430 g with Apgar scores of 9, 9 and 10, was delivered normally at 39 weeks gestation. She remained persistent in ventricular bigeminy rhythm on monitoring. Postnatal echocardiogram confirmed the RVO and 12-lead electrocardiogram showed ventricular bigeminy originating in the RVO (left bundle branch block morphology with axis 5°), as depicted in Figure 3.
The infant remained haemodynamically stable, despite exhibiting persistent ventricular monomorphic ventricular extrasystoles (VES). Twenty-four hour Holter monitoring showed almost continuous ventricular bigeminy, no couplets and no runs of ventricular tachycardia. We postulated a mechanical-electrical trigger mechanism, within the pouch, as the aetiological focus of cardiac ectopy for the VES. Given the incessant nature of the VES bigeminy, we commenced beta-blocker therapy, in the form of atenolol 1 mg/kg, 12 hourly orally. Over the ensuing three weeks, the VES frequency regressed and completely disappeared. The medication was weaned for three months, after starting with no recurrence of VES. The patient was followed-up for six months, the RVO was observed to diminish in size and the infant was found to grow normally. Arrhythmia has not recurred.

**Discussion**

The aetiology of congenital cardiac ventricular aneurysm is largely unknown. There have been several theories, such as infection and redundant primordial tissue myocardial ischemia leading to infarct and damage. Focal weakening of the ventricular wall, caused by an intrinsic abnormality during embryogenesis has also been suggested. Aneurysms are not usually associated with other congenital malformations, whereas diverticula may be associated with thoracic and abdominal midline defects. Aneurysms and diverticula are now referred to as outpouchings.

Congenital RVO is a rare form of congenital heart disease, with unknown true incidence, as many are asymptomatic and undiagnosed. Advancement in foetal ultrasound has permitted the detection of this anomaly, with at least 17 cases of RVO, reported in the published literature. Apical RVO’s are more likely to be associated with a pericardial effusion and present much earlier. Two publications of non-apical RVO reported atrial extrasystoles and atrial ectopy (supraventricular tachycardia). Our case is the first reporting of the prenatal ventricular bigeminy.

The cases of non-apical RVO were published, which were presented later in gestation than apical RVO, perhaps because of the lack of pericardial effusion. Foetuses with non-apical RVO have been reported to have ventricular and valvular dysfunction. Close follow-up and prenatal and postnatal monitoring, is thus essential in these patients.

**Conclusion**

Foetal RVO are rare congenital malformations that are required to be considered, when an extra chamber is visualised in the routine four-chamber view, in the assessment of foetal arrhythmia or presence of pericardial effusion.

**Abbreviations list**

RV, right ventricular; RVO, right ventricular outpouching; VES, ventricular extrasystoles.

**Consent**

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

**References**