Case report

Antenatal and postnatal sonographic imaging findings of a single ventricle presenting as double outlet right ventricle with rudimentary left ventricle and single atrium

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Abstract: Congenital heart disease is a major cause of morbidity and mortality. Single ventricle is a rare finding and usually of left ventricular morphology. We present here interesting antenatal and postnatal echocardiographic findings of a baby having a rare single ventricle of right ventricular morphology with double outlet. Antenatally we saw a large ventricular septal defect indistinguishable from a single ventricle with left to right ventricular ratio of 1:1. Postnatally we saw a single ventricle having the outlets for both the main pulmonary artery and aortic root. The left ventricle is collapse with a rudimentary morphology possibly due to changes in hemodynamics after birth and absent of outlet from it.

Keywords: single ventricle, ventricular septal defect, single atrium, double outlet right ventricle, echocardiography

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Introduction
Congenital cardiac disease is seen in 2.0–6.5 of 1000 live births and is a major cause of morbidity and mortality. Transabdominal detection of cardiac anomalies is usually done at around 18 to 22 weeks of gestation. Transvaginal scan can detect anomalies even at 12–13 weeks [1].

Case Report
We present here interesting antenatal echocardiographic imaging findings of a baby at term with subsequent postnatal confirmation.

Antenatal echocardiography: Four chamber antenatal echocardiograms show the apex of the heart on the left side, a single atrium with a very large ventricular septal defect (VSD) indistinguishable from a single ventricle. The ratio of the left and right ventricle is approximately 1:1. Both tricuspid and mitral valve appear normal. Three vessel views cannot be carried out due to technical difficulty and foetal position (Figure 1).

Postnatal echocardiography: The baby was delivered by caesarean section under spinal anaesthesia and needed prolong resuscitation after birth. Postnatal echocardiography was done on day 2. Findings included a single atrium, opening and closing of the tricuspid valve (TV), prominent single ventricle having the morphology of the right ventricle (RV), and poor visualization of mitral valve movements. The left ventricle (LV) is collapse. Both the great vessels are seen arising from the right ventricle, oriented perpendicular to each other with the main pulmonary artery on the right and anterior to the aortic root (Figure 2).

Discussion
Single ventricle accounts for 2% of congenital heart defects and is caused by failure of development of the interventricular septum. The single ventricle has a LV configuration in 85% of cases. It may be associated with pulmonary stenosis, atrial septal defect (ASD), common atrium, asplenia and polysplenia. Differential diagnosis includes large VSD or hypoplastic RV or LV [2].

Demonstration of the origin of both the vessels from the same side of ventricular septum is essential to differentiate double outlet right ventricle (DORV) from transposition of great arteries [3].

Figure 1. Antenatal four chamber echocardiographic view of the heart, note the single atrium (A) and a very large VSD indistinguishable from the single ventricle (V), normal opening and closing of the mitral valve (MV) and tricuspid valves (TV) were seen.
Figure 2. Postnatal echocardiographic images of the heart, note the prominent right ventricle (RV) giving rise to both the aortic root (AO) and main pulmonary artery (MPA) with collapse left ventricle (LV) consistent with a single ventricle of RV configuration and rudimentary left ventricle (LV). Note the normal closing and opening of the tricuspid valve (TV).

Conclusion
Our case is a single ventricle with right ventricular morphology. The normal size left ventricle seen antenatally is small after birth and changes to rudimentary morphology due to absent of outflow tracts and resultant hemodynamics.

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References

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