Biventricular noncompaction: A rare cause of fetal distress and tricuspid regurgitation

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Abstract
Isolated noncompaction of the ventricular myocardium involving both ventricles is a rare entity. Here we report a rare case of biventricular noncompaction presenting with features of fetal distress and moderate tricuspid regurgitation. Noncompaction of both ventricles was diagnosed at birth.

MeSH: Biventricular noncompaction, Fetal distress, Tricuspid regurgitation

Case report
A 27 year old female, primigravida, 32 weeks of gestation, was referred for fetal echocardiography for intrauterine growth retardation (IUGR), cardiomegaly with suspicion of Ebstein's anomaly of tricuspid valve and moderate tricuspid regurgitation (TR) on routine ultrasound. Doppler imaging of the umbilical veins also showed moderate to severe increase in impedance in umbilical venous flow with no obstruction suggestive of high right atrial pressure. First and second trimester ultrasounds were within normal limits. Fetal echocardiography was done with a Phillips iE33 system, and showed normal situs, levocardia, concordant atrioventricular and ventriculoarterial connections, normal morphology of both tricuspid and mitral valves, normal right and left ventricular outflow tracts, widely stretched foramen ovale and a patent ductus arteriosus. Both right atrium and right ventricle were dilated with
the interatrial septum bowing toward the left (figure 1a). Color flow mapping showed severe TR (figure 1b).

Figure 1 Fetal echocardiogram. 1a: Four chamber view in fetus at 32 weeks of gestation showing dilated RA and RV, bowing of interatrial septum towards left (arrow). 1b: Same view with color flow mapping showing significant tricuspid regurgitation (arrow)

There was normal antegrade flow across the pulmonary valve. Systolic function of both ventricles was normal. Fetal heart rate was 150/minute with 1:1 atrioventricular conduction and a normal PR interval. The baby (male) was delivered by elective caesarian section at 38 weeks of gestation with a birth weight of 1.8 kg, and with normal Apgar scores. There was no suggestion of dysmorphism. Cardiac evaluation done at birth revealed tachycardia, tachypnoea and mild hepatomegaly. He was started on intravenous diuretic (frusemide) once daily and oxygen by hood. The baby was stabilized over 1 week. Echocardiography done at 1 week confirmed the finding of fetal echocardiography. There was moderate TR with a peak pressure gradient of 28 mmHg. In addition, there were features of noncompaction of ventricles (numerous trabeculations with deep intertrabecular recesses of both ventricles - figure 2a-c).

Figure 2 Echocardiography at 1 week of post natal life. 2a: Apical four chamber view showing dilated RA and RV with noncompaction of LV (arrows points to deep recess). 2b: Same view with color flow mapping showing moderate TR, deep recess of RV are marked by arrows. 2c: Parasternal short axis view of both ventricles showing noncompaction involving both RV and LV

There was biventricular diastolic dysfunction with mild systolic dysfunction of both ventricles. Recent follow up at 48 days of age showed a stable neonate with good weight gain (present weight 3.25 kg), no features of congestive cardiac failure (CHF). Echocardiography revealed regression of the features of noncompaction in the right ventricle (RV) with normal systolic function. There was persistence of noncompaction in the left ventricle (LV) with mild LV systolic dysfunction (LV ejection fraction 40-45%), moderacy (PA) pressure (figure (3a,b)).

Figure 3 Echocardiography at 48 days of life. 3a: Parasternal short axis view showing smooth interventricular septum and RV. 3b: Inverted four chamber view showing apical part of LV showing deep recess (arrows). RA-right atrium, RV-right ventricle, LA-left atrium, LV-left ventricle, TR-tricuspid regurgitation.

Discussion

Isolated ventricular noncompaction (IVNC), also known as spongy cardiomyopathy, is a rare congenital cardiomyopathy. It is characterized by the presence of numerous excessively prominent ventricular trabeculations and deep intertrabecular recesses. The main cause of this disease is due to an intrauterine arrest of normal myocardial development with lack of the loose myocardial meshwork. Although noncompaction of the ventricular myocardium is a congenital disorder, there are only a few case reports of its diagnosis in fetal life or in the immediate neonatal period. Diagnosis is made by echocardiography. Jenni et al defined 4 criteria for the echocardiographic diagnosis of IVNC and these are:

1. Coexisting cardiac anomalies caused by high-pressure exposure of the ventricle during intrauterine development are absent, and various forms of semilunar valve obstruction are absent.
2. The left ventricular wall is thickened and consists of a 2-layered structure: a compacted epicardial band of uniform tissue and a much thicker, noncompacted endocardial layer of prominent trabeculations.
and deep intertrabecular recesses. A maximal end-systolic ratio of noncompacted to compacted layers of greater than 2 is diagnostic.
3. The features are found predominantly in the apical and mid ventricular segments of the left ventricle.
4. Direct blood flow from the ventricular cavity into the spaces between the prominent trabeculations is visualized by color Doppler sonography throughout the cardiac cycle.

Although the features of IVNC can be detected during fetal life, these hypertrabeculations may not be initially apparent or easily recognized on fetal echocardiography due to difficulties inherent with resolution of very fine structures in the small fetus. In our case, there was IUGR, increased impedance in umbilical venous flow and severe tricuspid regurgitation. The diagnosis of noncompaction was not suspected during fetal echocardiography. For the same reasons, on retrospective review of the fetal echocardiogram we were not able to note any features of noncompaction.

In contrast to the predominant LV involvement seen in the adults with IVNC, the fetal and neonatal cases frequently display biventricular involvement. The high incidence of RV involvement in fetal IVNC may be related to the RV dominant fetal circulation. Six weeks post natal follow up in our case showed that features of noncompaction of the RV had decreased and RV function had improved while the LV still had features of noncompaction with persistent LV dysfunction. This could be due to the fact that after birth, RV dominance decreases with a fall in pulmonary vascular resistance and normalization of PA pressures leading to regression of features of noncompaction in the RV.

Clinical presentation of fetal and neonatal IVNC may vary from a hydropic fetus with severe ventricular dysfunction to minimal or less dramatic ventricular involvement. On reviewing the literature, we found case reports of fetal and neonatal diagnosis of IVNC. The majority of reports had a poor outcome, although Menon et al reported 6 cases (5 fetal and one neonate) with improvement in the ejection fraction (mean ejection fraction 36% which improved to 57% during an average follow up of 2 years).

**Conclusion**

If echocardiographic examination shows cardiomegaly, ventricular hypertrophy, unexplained tricuspid or mitral regurgitation and/or poor ventricular function, IVNC should be considered and in order to see recesses, the ventricle walls should be investigated by zooming in and comparing frame by frame.

**References**