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Images in Congenital Cardiac Disease

Fetal echocardiographic imaging of ventricular noncompaction

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A 25-YEAR-OLD WOMAN, PREGNANT FOR THE FOURTH time after 2 successful deliveries, was referred for fetal echocardiography at 30 weeks gestation to follow-up an abnormal screening examination. There was no family history of cardiac disease. This initial fetal echocardiogram (Fig. 1a and b), demonstrated highly trabeculated areas of myocardium lining the luminal surfaces of both left and right ventricles, with deep recesses within the trabeculated layer and communicating with the lumen as shown on colour flow Doppler. The cardiac apex was leftward, and there was significant cardiac enlargement, the cardiothoracic area ratio being 34% (Fig. 2). There was qualitative biventricular depression of systolic function with a moderately sized pericardial effusion, but no other evidence of fetal hydrops. The heart rate and rhythm were normal. These findings persisted unchanged on subsequent fetal echocardiograms.

The mother gave birth to a male infant weighing 2.4 kilograms at 37 weeks gestation, with Apgar scores of 7 and 8 at 1 and 5 minutes, respectively. The fetal echocardiographic diagnosis was confirmed after delivery (Fig. 3), with a small, muscular ventricular septal defect also being noted. During nine days of observation in the neonatal intensive care unit, the patient had mild tachypneoa, but remained breathing room air, and tolerated oral feedings. Investigations, including a karyotype, pyruvic acid, lactic acid, plasma amino acids, urine amino acids, acylcarnitine profile, and Barth syndrome sequence analysis, were all within normal limits. The urine organic acid testing suggested a possible mitochondrial disorder,

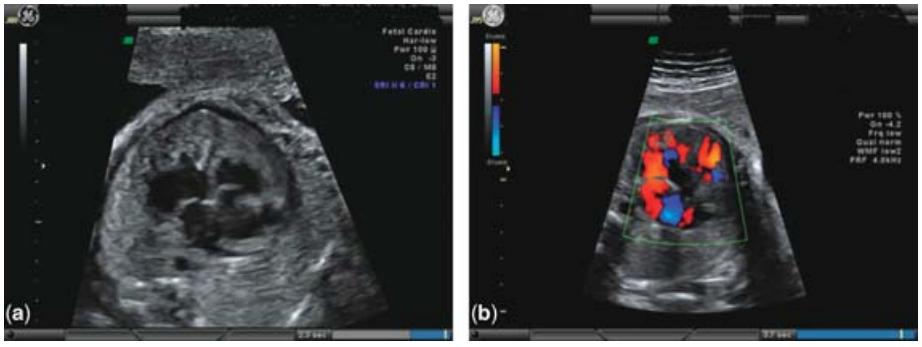

Figure 1.

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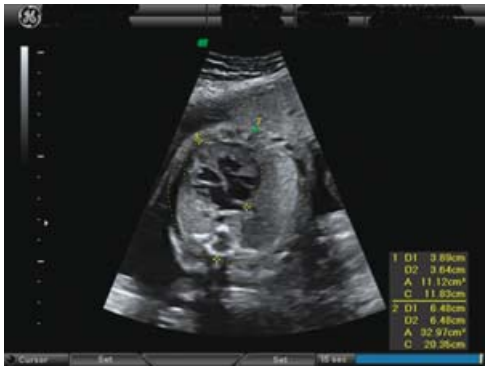

Figure 2.

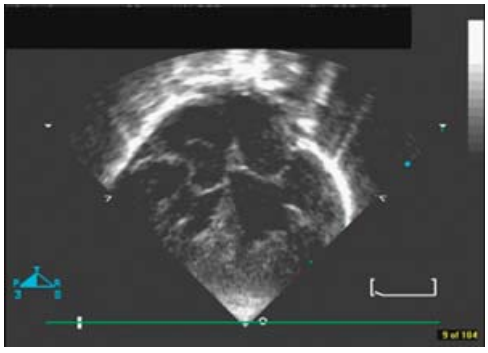

Figure 3.

but his parents declined muscle biopsy for further characterization. The patient was discharged from hospital on aspirin, digoxin, furosemide and aldactone. Currently, this 10 month old infant continues to be asymptomatic, with weight at the 10th centile. His recent echocardiograms continue to demonstrate severe biventricular noncompaction, with subjectively moderately decreased biventricular function. An inhibitor of angiotensin converting enzyme, and monthly injections of immunoglobulin as prophylaxis against the respiratory syncytial virus, has been added to his therapeutic regime. Continuous ambulatory monitoring over 24 hours on several occasions has documented normal sinus rhythm.

While the deep trabeculations and the spongy appearance of the myocardium, both before and after birth, are typical of ventricular non-compaction, our patient differs from others diagnosed during fetal life[1] in not having bradycardia, nor severe hydrops with enlarged atrial chambers due to restrictive cardiomyopathy, nor perinatal death. In this regard, he could represent either a less severe end of the spectrum of ventricular non-compaction, or an earlier stage in the process of the disease. More widespread identification of ventricular non-compaction on fetal echocardiography, and careful follow-up of such patients over the long term, is clearly needed further to characterize this malformation.

Reference

1. Guntheroth W, Komarniski C, Atkinson W, Flinger CL. Criterion for fetal primary spongiform cardiomyopathy: Restrictive pathophysiology. Obstet Gynacol 2002; 99: 882–885.

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