A rare case of contracted endocardial fibroelastosis
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Objective
Endocardial fibroelastosis (EFE) is characterized by diffuse endocardial thickening and myocardial dysfunction. Two pathologic forms of primary EFE include dilated and contracted. The less common contracted type of primary EFE is associated with a relatively hypoplastic or normal left ventricle (LV) size. The right and left atria and the right ventricle are markedly enlarged and hypertrophied, with minimal or no endocardial sclerosis. An early event in fetal life is believed to result in dilated endocardial fibroelastosis, which later morphs into a contracted type.

Methods
A 24-year-old healthy woman (gravida 1, para 0) without risk factors for congenital heart disease presented at 20 weeks gestation for routine early screening of fetal cardiac malformations.

Results
The fetal heart revealed a dilated left cardiomyopathy. The family did not accept the termination of pregnancy. Chromosomal analysis through amniotic puncture revealed a normal male karyotype: 46, XY. The reference range of anti-Ro and anti-La antibodies were negative; viral serology were negative and Secondary EFE was excluded. At 28 weeks of gestation fetal echocardiography showed a dilated and hypotonic left ventricle with a very bright echogenic line over the endocardium which suggested fibroelastosis. At 35 weeks of gestation echocardiography revealed LV hypoplasia, midly hypoplasia of aort and mitral anulus, increased left ventricle echogenicity and the patient was delivered by cesarean. First echocardiography in postnatal period showed markedly progression of contraction and hypoplasia in LV. Prostaglandin E1 infusion was started. The patient underwent the Norwood procedure at a median age of 14 days and after one week the patient died. The family did not consent to an autopsy.

Conclusion
This is a rare seen report about a case of prenatal diagnosis of contracted EFE without significant aortic stenosis.