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Fetal Cardiomyopathy multicentre study: aethiology and clinical outcome

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Objectives: Although prenatal diagnosis of cardiomyopathy (CM) is rare, it has a high mortality and morbidity. The aim of our study is to describe the underlying causes, hemodynamic and echocardiographic findings and the outcome of these patients.
Methods: Multicentre retrospective study between January 2007 and November 2010 at 6 tertiary Spanish centres. We included 45 fetuses with dilated (DCM), hypertrofic (HCM) or Non Compactation (NCCM) cardiomiopathy. We excluded fetuses with congenital heart disease, arrhythmias, twin-twin transfusion and hypervolemic causes. Mean gestational age at diagnosis was 26 weeks (range 17-38 GW), with 2 twin pregnancies. Post-natal follow-up or necropsy was achieved in 92% of cases.
Results: DCM was diagnosed in 26 cases including 3 congenital infections, 4 endocardial fibroelastosis due to maternal anti-Ro/La and 19 idiopathic causes. During postnatal life 4 dilated cases turned out to be NCCM. 17 fetuses had HCM: 1 with familial hypertrophy, 4 associated with maternal diabetes, 3 with Noonan Sindrome, 1 Pompe, 1 with polichystic kidneys and 7 with idiopathic hypertrophy. Intrauterine diagnosis of NCCM was found in 2 cases. Fetal echocardiographic findings were: cardiomegaly 80%, abnormal Ductus Venosus /inferior vena cava flow 63%, tricuspid or mitral regurgitation 56% , systolic dysfunction 43%, diastolic dysfunction 33%, prolonged Isovolumetric relaxation time and Tei Index 71% (when measured). Hydrops was found in 38% of cases. Termination of pregnancy and intrauterine death occurred in 20% and 9% of patients. Transplacental medication was administered to 7% of fetuses. Fetuses with tricuspid/mitral regurgitation, prolonged Tei index and anomalous venous flow had a higher mortality. Out of the 23 pregnancies that continued, 16% died perinatally, 8% died before 2 years old, and 4% (2) were included in heart transplant program (1 died on Berlin Heart). Overall survival was 44% (56% in non-hydropic fetuses compared to 21% in hydropic fetuses), 47% of the survivors are under oral medication.
Conclusions: Fetal cardiomiopathy has a high prenatal and postnatal mortality. Hydrops, tricuspid/mitral regurgitation and abnormal venous flow pattern are strong predictors of poor outcome. Diagnosis of NCCM is feasible during fetal life.