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Fetal Cardiac Tumors: Long Term Follow-up.

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Introduction
Cardiac tumors are rare in children with an incidence of 0.009% in prenatal evaluations and 0.08 - 0.2% in pediatric cardiology clinics. Rhabdomyoma is the most frequent, and although benign, serious conditions might be associated such as hemodynamic compromise, arrhythmias or Tuberous sclerosis and its neurologic compendium. The aim of this study is to describe prenatal and postnatal management and long-term follow-up of fetal cardiac tumors.

Methods
Retrospective study of fetal cardiac tumors diagnosed between 1998-2017 at our fetal cardiology unit. All Clinical records were reviewed: fetal echocardiography, prenatal and postnatal management, postnatal echocardiography and neurological status.

Results
A total of 36 fetuses with cardiac tumors were identified at a mean gestational age of 30 weeks. Seven legal pregnancy interruptions were performed (4 neurological involvement, 1 teratoma with hydrops, 2 intracardiac tumors). Rhabdomyoma represent 90%, 5% fibroma, 2.5% teratoma, 2.5% pericardium tumor. Electrocardiogram abnormalities were found in 17% (1 complete auriculoventricular block with remission at 9 months of age, first degree AV block, 3 supraventricular and ventricular premature beats). Surgery was required in 10%: 1 ventricular outflow obstruction, 1 progressive growth, 1 massive pericardial effusion (despite evacuating pericardiocentesis performed in utero). Median follow-up of the 29 newborns was 8,5 years (1-19 years) with an overall survival of 96,5%. One death was reported secondary to cardiogenic shock in a giant obstructive neonatal tumor prior starting ECMO program (1999). Although most patients are currently cardiologically asymptomatic with spontaneous regression of the tumors without medical treatment, 53% associate tuberous sclerosis of which 58% have serious neurologic manifestations such as West syndrome, autistic spectrum disorder and neurodevelopmental delay. A neonate with giant left ventricle rhabdomyoma with severe hemodynamic compromise had an outstanding response to everolimus.

Conclusions
Prenatal diagnosis of cardiac tumors is standard practice. Although most tumors are benign, they might associate serious neurologic comorbidity or a relevant need of cardiac surgery with extracorporeal circulation and a non-negligible mortality. Everolimus can be considered as a treatment alternative for multiple rhabdomyomas or those that cannot be surgically removed.