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Idiopathic dilatation of the right atrium. Report of four fetal cases.

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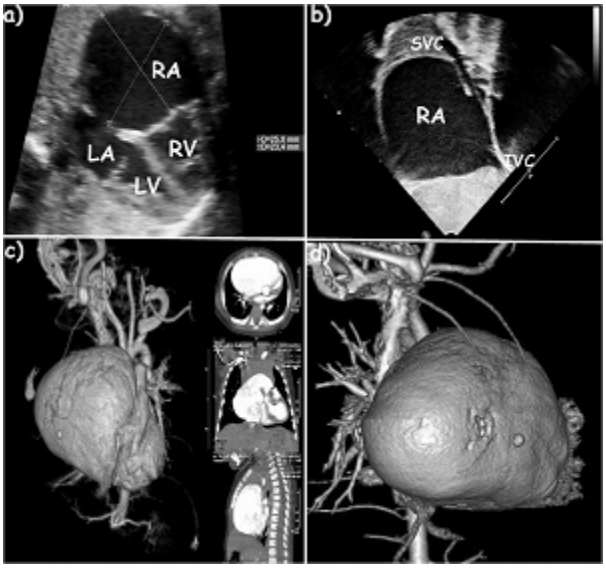
Introduction:
Idiopathic dilatation of the right atrium (IDRA) is a rare anomaly defined as isolated enlargement of the right atrium in the absence of other cardiac lesions or predisposing conditions to cause right atrial (RA) dilatation, especially tricuspid valve diseases. The clinical presentation is highly variable from asymptomatic to cardiac failure or even sudden death. It can be associated with atrial arrhythmias, thrombus formation and congestive heart failure.



Methods:
We report 4 cases of IDRA of prenatal diagnosis, three children and last one is still a prenatal case. We describe the intrauterine course, the postnatal management and its short-medium term follow-up. Echocardiography of the fetus showed an abnormal 4-chamber view with an increased cardiothoracic circumference ratio at the expense of enlarged RA without other anomalies. During the routine controls the dilatation of RA persisted. None had extracardiac malformations. Postnatal echocardiography demonstrated that the RA was dilated with spontaneous echo contrast (Figure1a-b). Although none had thrombus formation in the RA we decided initiate treatment due to a potential risk for pulmonary embolism. In all patients thrombophilia study was normal so we started thrombosis prophylaxis with salicylic acid. There has been no need for surgical intervention so far because of the lack of arrhythmias or symptoms, although one of our cases we have found an important progression of RA diameters. This patient is 4 months old and the RA is massively dilated (diameter of 65mmX71mm detected with angioCT). Cardiac resonance imaging showed global hypokinesia and he is awaiting surgical decision (Figure 1c-d).

Conclusions: Optimal management of IDRA is controversial and depends on the individual case. This disease probably is under diagnosed, reinforced by the fact we have seen four cases, but all in the last two years.
Long-term follow-up is necessary to monitor progression of RA size and occurrence of arrhythmias. Asymptomatic patients can be managed medically but symptomatic patients may require surgical reduction of the RA. Different imaging techniques including computed tomography and cardiac resonance imaging are useful to diagnose, evaluate and perhaps it may be helpful in treatment decisions. According to Paladini a Holt-Oram syndrome should be ruled out.