Sant Joan de Déu
HOSPITAL MATERNOINFANTIL - UNIVERSITAT DE BARCELONA

Idiopathic dilatation of the right atrium. REPORT OF FOUR FETAL CASES

C. Walter, J.M. Carretero, J. Bartrons, O. Gómez, J.M. Martínez, C. Pérez, J. Mayol, J.M. Caffarena, F. Prada.

Introduction

Idiopathic dilatation of the right atrium (IDRA) is a rare anomaly defined as an isolated enlargement of the right atrium (RA) in the absence of other cardiac lesions or a condition cause RA dilatation. The clinical presentation ranging from asymptomatic to cardiac failure or even sudden death. It can be associated with atrial arrhythmias, thrombus formation and congestive heart failure.

Antiplatelet prophylaxis is carried out in most described cases due to the risk of pulmonary thrombosis (and systemic if SIAD is present). The main indications for surgical reduction are rapid progression of growth, compression of adjacent structures and refractory arrhythmias.

We report 4 cases of IDRA observed during prenatal diagnosis and describe the intrauterine and the postnatal course and management.

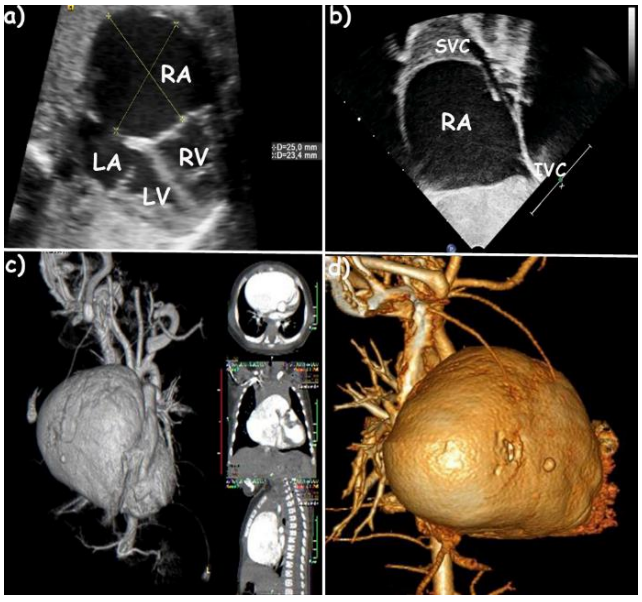
Methods

The 4 cases described were admitted to hospital in a period shorter than 13 months. 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. None had extracardiac malformations. (Table 1).

Case #	Age and size of RA at diagnosis	Date of birth	Growth of RA (age and size of RA)	Evolution	Additional tests	Therapy
1	39 w= 35x30 mm	08.12.2012	15 m = 40X37 mm	Asymptomatic, stable atrial enlargement	Blood analysis of thrombofilia, awaiting CT	AAS 5 mg/Kg/day
2	28w= 25x23 mm	13.08.2013	7 m= diam 67 mm	Asymptomatic, very fast atrial enlargement	Blood analysis of thrombofilia CT, MR	AAS 5 mg/Kg/day Surgical atrial reduction
3	32w= 32x22 mm	04.12.2013	unknown	Unknown	Blood analysis of thrombofilia. Other data unknown	AAS 5 mg/Kg/day. Other data unknown
4	20 w= 20x17 mm	Unborn	33 w= 42x35 mm	-	-	-

Postnatal echocardiography showed a dilated RA with spontaneous echo contrast (Figure1) but none had thrombus formation. In all patients thrombophilia study was normal but we initiated prophylaxis with salicylic acid due to the potential risk for pulmonary embolism. In one of the four cases we have found considerable progression of RA diameters with massively dilated (diameter of 67 mm). Cardiac resonance imaging showed global hypokinesia (Figure 1).

The patient was surgically corrected at the age of 7 months whereupon atrial diameter is 37 mm and the spontaneous contrast disappears. The other 3 cases have no sign of arrhythmias or symptoms.



Conclusion

Optimal management of IDRA in children is controversial, and depends on the individual case. This disease is probably underdiagnosed, this is reinforced by the fact that we have seen four cases, but all in the last 13 months.

Long-term follow-up is mandatory to monitor progression of RA size and the occurrence of arrhythmias or thrombus. 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 for evaluation and treatment decisions.