

Prenatal diagnosis of idiopathic dilatation of the right atrium. A case report

Melina J. Saavedra M.D.^a, Sofía Grinenco M.D.^a and Natalia Nápoli M.D.^a

ABSTRACT

Idiopathic dilatation of the right atrium is a rare condition. The real incidence of the disease is difficult to estimate since most patients are asymptomatic, although a wide spectrum of symptoms have been reported in children and adults. In the literature, prenatal diagnosis has been reported in few cases. We report a case of prenatally diagnosed right atrial dilation with a 3 years and 8 months follow up. The patient is on acetylsalicylic acid since birth and remained asymptomatic without any atrial thrombosis. We performed regular transthoracic echocardiograms during follow up, however was a cardiac magnetic resonance imaging the method for achieving diagnostic confirmation.

The aim of this case report is to describe a rare, potentially serious disease and how prenatal diagnosis allows anticipated actions such as monitoring for cardiac arrhythmia detection and early starting of primary thrombosis prophylaxis.

Key words: right atrium, aneurysm, dilation, prenatal diagnosis, congenital heart defects.

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INTRODUCTION

Idiopathic dilatation of the right atrium is a rare disease and its pathogenesis is unknown.¹ Clinical presentation ranges from asymptomatic patients, to syncope, chest pain, arrhythmias, thromboembolism, congestive heart failure and even sudden death.²

In the literature, prenatal diagnosis has been reported in few cases without a long-term follow-up, being the management individualized on a case by case basis.¹⁻⁴

We report a case of idiopathic dilatation of the right atrium prenatally diagnosed, describing an uncommon and potentially serious disease and highlighting the relevance of antenatal diagnosis in order to prevent possible complications.

CASE REPORT

A 34-year-old woman, previously healthy, was referred to the fetal cardiology service during her first pregnancy due to dilatation of the right atrium found on obstetric ultrasound after 35 weeks of gestation (*Figure 1*). She had no family history of cardiovascular disease. A color Doppler fetal echocardiography was performed at 36 weeks of gestation, an extremely right atrium dilation and mild regurgitation of a normal tricuspid valve were observed. No arrhythmias and no other findings were detected during the study. Weekly appointments were scheduled with stable parameters.

A male infant was delivered at term by C-section with a birth weight of 3860 g and Apgar score of 8/9. On physical examination pink skin was observed and S1 and S2 were present, without S3 and any murmurs. Baseline electrocardiogram was performed exhibiting normal values as sinus rhythm, heart rate 120 beats per minute, normal voltage and duration of P wave, PR interval duration was 100 milliseconds, QRS complex was 70 milliseconds, right axis deviation (+130°) and right R wave progression without repolarization abnormalities. A transthoracic echocardiogram was performed revealing the following notable findings: small patent foramen ovale with a left-to-right shunt, right atrium dilation measuring 7 cm² (mean 2.6 cm², SD 1.8 cm²), normal tricuspid

a. Department of Pediatrics. Division of Pediatric Cardiology. Hospital Italiano de Buenos Aires. Argentina.

E-mail address: :

Melina J. Saavedra M. D.: melina.saavedra@hiba.org.ar

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valve with mild regurgitation, patent ductus arteriosus of 3 mm with a left to right shunt. According to the high thromboembolism risk of this disease, acetylsalicylic acid (5 mg/kg/day) was started as antiplatelet therapy. The patient remained in the Neonatal Intensive Care Unit during 24 hours and his telemetry monitoring were within normal limits, therefore we have decided to transfer him with his mother until the fourth day of life when both were discharged.

Subsequently, he started outpatient follow-up with pediatric cardiology. In the first visit, closure of the patent ductus arteriosus was verified. During the first two years of life, we perform physical examination, electrocardiogram and transthoracic echocardiogram every two

month and order a Holter monitor yearly. Based on World Health Organization's standards, the patient was at the 50th percentile for weight and 90th percentile for height. He did not show any arrhythmias and the right atrium area remained stable with values from 10 cm² (mean 4.2 cm², SD 2.2) to 20 cm² (mean 6.0 cm², SD 2.7) according to the child growth curves.

A cardiac magnetic resonance imaging (MRI) was requested when he was 14 months old, and idiopathic dilatation of the right atrium diagnosis was confirmed (*Figure 2*). The MRI showed the following findings: severe aneurysmal dilation of the right atrium (area 26 cm²) with reverberant flow inside; thrombus were not observed; normal tricuspid valve; right ventricle end-diastole and

FIGURE 1. Prenatal echocardiography at 35 weeks of gestation. A and B. Excessively dilated right atrium in comparison with the other chambers

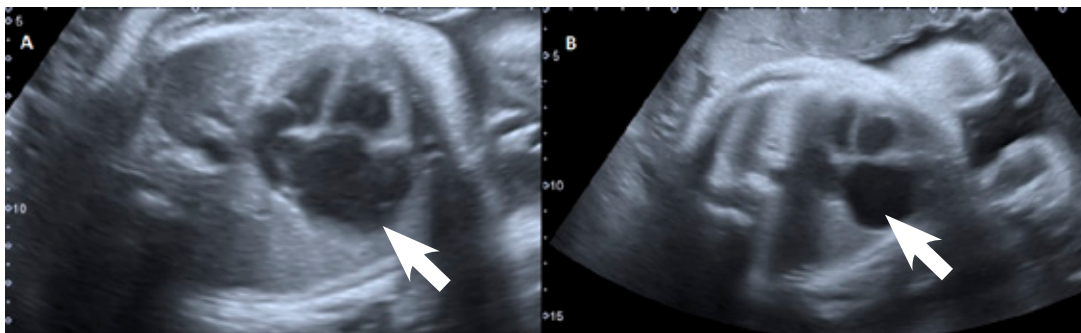
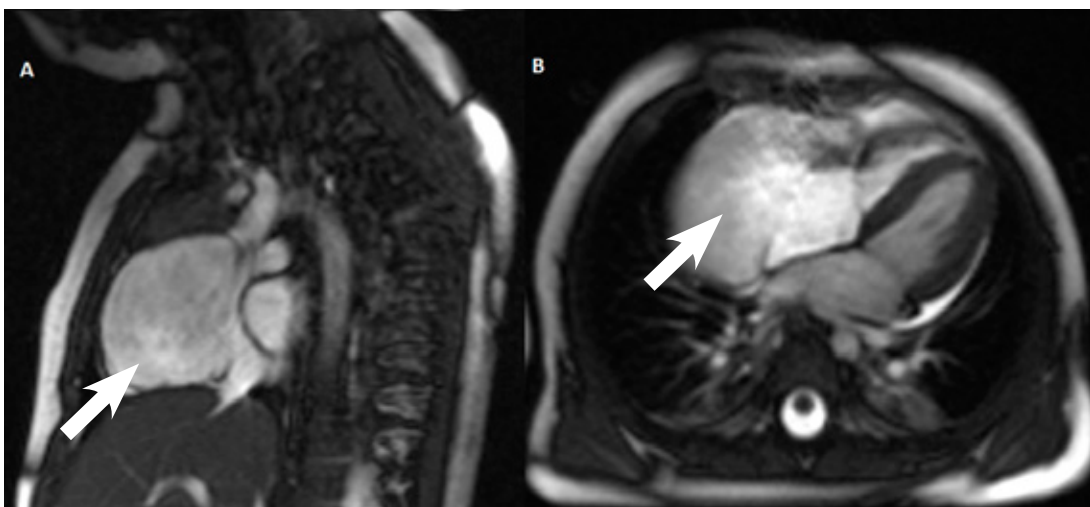


FIGURE 2. Cardiac magnetic resonance imaging (MRI). A and B. Disproportionately large right atrium in comparison with the other structures



end-systole volumes within normal range, with preserved regional motility, and global systolic function; intact pericardium. In the T1, T2 and fat saturation images, no fatty infiltration of the myocardium was observed. In the images after intravenous injection of gadolinium, no enhancement was observed in any region of the ventricles wall, ruling out the presence of fibrosis and myocardial necrosis.

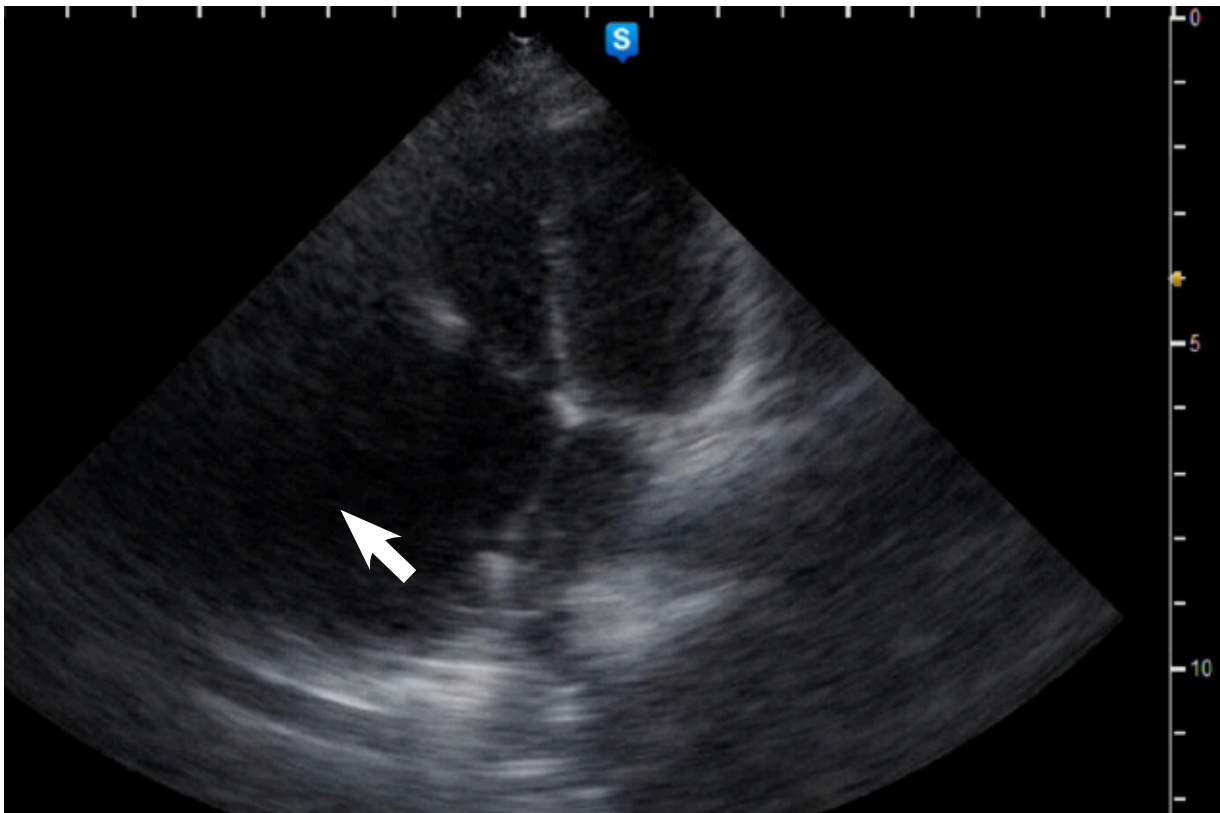
Currently, the patient attends to pediatric cardiology clinic every 6 months and he continues on acetylsalicylic acid treatment. Holter monitor is requested annually. After 3 years and 8 months follow-up in our division, the child remained asymptomatic, without any arrhythmias. The right atrium did not exhibit significant changes over time with an area of 18 cm² (mean 7.8 cm², SD 3) measured by transthoracic echocardiography (Figure 3). During follow-up, his younger brother was born without congenital heart disease.

DISCUSSION

As a result of fetal cardiac medicine evolution, the role of the pediatric cardiologist has expanded and prenatal diagnosis became essential in order to plan medical strategies to improve congenital heart disease outcomes. In our case, antenatal diagnosis allowed an optimal delivery planification, in a third-level hospital with a neonatal intensive care unit as well as pediatric cardiology and electrophysiology services. We were able to perform an accurate heart rate monitoring since birth and antiplatelet medical treatment was started in order to reduce atrial thrombosis risk.

Idiopathic dilatation of the right atrium was first described in 1676 by Borrachin.⁵ However, in 1955, Charles Bailey, a cardiovascular surgeon, was who performed the first excision of a dilated right atrium in a 29-year-old woman who developed dyspnea, paroxysmal supraventricular tachycardia, and cardiomegaly.⁶ Since then, in

FIGURE 3. Transthoracic echocardiography. Apical four chambers view. Extremely right atrium dilation associated with normal tricuspid valve



the literature there have been isolated reports of this disease with a wide clinical presentation in children and adults.

Antenatal diagnosis occurs in a few cases and none of them reported long-term follow-up.¹⁴ The majority of those patients remained asymptomatic. Symptomatic children more frequently developed arrhythmias such as atrial tachycardia, most of them were controlled by antiarrhythmics and in some cases radiofrequency ablation was required.² Between the second and fifth week of birth, two patients were readmitted for respiratory distress. Fluoroscopy showed dynamic airway obstruction. Both infants underwent cardiovascular surgery for right atrial resection with good postoperative results.^{2,3} Children without antenatal diagnosis were mostly symptomatic and showed atrial thrombosis and arrhythmias, including an 8 years old child who developed refractory atrial tachycardia and died.^{7,8}

Idiopathic dilatation of the right atrium or congenital aneurysm of the right atrium requires for its diagnostic confirmation a disproportionately large right atrium in comparison with the other chambers with absence of abnormalities that could cause right atrial enlargement, such as Ebstein's anomaly, tricuspid valve dysplasia, pericardial agenesis, partially anomalous pulmonary venous connections, Uhl's anomaly, restrictive cardiomyopathy and right ventricular failure.⁹ Transthoracic echocardiogram has become the first-line imaging modality for the assessment of cardiac structure and function, however, two dimensional methods can be limited to rule out entities that could cause atrial enlargement. Imaging method development allow physicians to achieve a precise diagnosis. Computed tomography and cardiac resonance imaging are more accurate non-invasive diagnostic methods with 3D representation.^{1,8,10,11} In our experience, magnetic resonance imaging was essential to confirm the diagnosis of idiopathic dilatation of the right atrium, however, the echocardiogram allowed antenatal detection of right atrium enlargement and was the election method for monitoring right atrium area during the patient follow-up.¹²

As a result of a wide range of clinical presentations, management is individualized based on each case. Given the increased risk of atrial thrombosis and consequently thromboembolism, it is recommended to start prophylactic antiplatelet

therapy with acetylsalicylic acid.^{2,11} Cardiovascular surgery for right atrial resection in children, was proposed in those who presented with atrial thrombosis, progressive dilation of the right atrium, right ventricle or airway compression, and is still being controversial in asymptomatic children.^{2,11} Our patient started pharmacological treatment with acetylsalicylic acid as a newborn and we did not indicate surgical treatment, given his asymptomatic clinical course at the time of this report.

We presented a case of prenatally diagnosed idiopathic dilatation of the right atrium. Prenatal diagnosis allowed us to make anticipated medical planning in order to prevent potential complications. Those strategies were early referring to a third-level hospital for obstetrical follow-up and delivery, as well as heart rate monitoring and indication of starting antiplatelet primary prophylaxis in the newborn. ■

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