

Echocardiographic diagnosis of distal aortopulmonary window

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ARTICLE INFORMATION

Received: November 27, 2012

Accepted: December 21, 2012

Competing interests

Authors have no competing interests

Acronyms

APW: aortopulmonary window

On-Line versions:

Spanish - English

ABSTRACT

The aortopulmonary window is a communication between the ascending aorta and the pulmonary artery, with well differentiated semilunar valves. Distal defects are less common and occur in approximately 25% of all patients with aortopulmonary window. Although two-dimensional transthoracic echocardiography can provide an accurate diagnosis in most cases, some authors ask for further studies. This is the case of a 40-day-old infant with echocardiographic diagnosis of distal aortopulmonary window that was confirmed in the operating room without further studies. At 9 months of clinical and echocardiographic follow-up, the patient remains asymptomatic.

Key words: Congenital heart disease, Aortopulmonary window, Echocardiography

Diagnóstico ecocardiográfico de ventana aortopulmonar distal

RESUMEN

La ventana aortopulmonar es una comunicación entre la aorta ascendente y la arteria pulmonar, con válvulas sigmoideas bien diferenciadas. Los defectos distales son menos frecuentes y se presentan aproximadamente en 25 % de todos los pacientes con ventana aortopulmonar. Aunque la ecocardiografía transtorácica bidimensional puede proveer un diagnóstico certero en la mayoría de los casos, algunos autores solicitan la realización de otros estudios. Se presenta una lactante de 40 días de edad con diagnóstico ecocardiográfico de ventana aortopulmonar distal, que fue confirmado en el quirófano, sin necesidad de realizar otros estudios. A los 9 meses de seguimiento clínico y ecocardiográfico, la paciente se mantiene asintomática.

Palabras clave: Cardiopatía congénita, Ventana aortopulmonar, Ecocardiografía

INTRODUCTION

The aortopulmonary window (APW) is a communication between the ascending aorta and the pulmonary artery with well differentiated semilunar valves¹. This rare disease represents 0.2 to 0.6% of all congenital heart diseases².

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According to the classification of Mori *et al.*³, type II APW occurs in the distal portion of aortopulmonary septum and extends to the right pulmonary artery. Distal defects are less common and occur in approximately 25% of all patients with APW⁴.

CASE REPORT

This is a 40-day-old female patient, with a history of heart failure at birth and probable diagnosis of ventricular septal defect.

During inspection a hyperdynamic precordium is detected with moderate bilateral subcostal retractions and respiratory rate of 64 per minute. The beat of the tip was visible and palpable in the fifth left intercostal space. On auscultation a continuous II / VI murmur at the base of the heart and a heart rate of 162 beats per minute were detected. The arterial oxygen saturation was 99% and bounding peripheral arterial pulses were noted. The electrocardiogram showed left ventricular hypertrophy and telecardiogram confirmed cardiomegaly and pulmonary congestion.

In the 2D transthoracic echocardiography, *situs solitus* with atrioventricular and ventricular-arterial concordance was detected. The left cavities were dilated with mild mitral regurgitation. There were no echoes in the distal portion of the aortopulmonary septum that corresponded with a type II APW, with 10 mm in length. A well formed edge toward the semilunar valves that measured 11 mm was visualized (Fi-

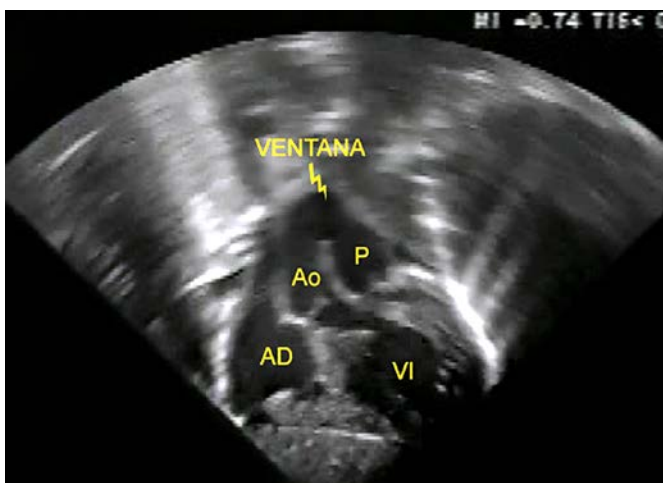


Figure 1. Coronal subcostal view of both outflow tracts where distal aortopulmonary window is visualized. AD: right atrium, VI: left ventricle, Ao: aorta, P: pulmonary artery.



Figure 2. Longitudinal suprasternal view of aortopulmonary window. Ao: aorta, P: pulmonary artery, RD: right pulmonary branch.

gures 1 and 2). Through the defect a slight shunt from left to right was visualized. A perimembranous ventricular septal defect (VSD) of 3 mm was diagnosed.

Surgical treatment by median sternotomy using cardiopulmonary bypass at normothermia was undertaken. The diagnosis was confirmed and by transaortic approach a polytetrafluoroethylene patch was placed in the defect, while with quilted stitches the VSD was closed.

There were no complications in the immediate postoperative period. At 9 months of clinical and echocardiographic follow-up the patient remains asymptomatic. In evolutionary echocardiograms neither residual shunt nor stenosis in the ascending aorta or pulmonary arteries are observed.

COMENTARIO

At the William Soler Pediatric Cardiology Hospital the APW shows a prevalence of about 1 patient per year, with a higher frequency in distal defects⁵.

The clinical characteristics are determined by the size of the defect and the kind of associated lesion. It is imperative to have a high index of suspicion in patients with congestive heart failure in the first days or months of life, with significant manifestations of left to right shunt, such as left chamber dilatation and functional mitral regurgitation, associated with severe pulmonary hypertension².

Electrocardiographic and radiological findings are nonspecific, so echocardiography is crucial for positive

diagnosis. Short axis and high parasternal views are used, as well as coronal subcostal of both outflow tracts and longitudinal suprasternal (Figures 1 and 2).

2D transthoracic echocardiography is a non-invasive study, useful for diagnosing distal APW, whereas cardiac catheterization is useful for those patients with complex anatomies or those over 6 months of age to evaluate the presence or absence of irreversible pulmonary hypertension. Some authors ask for further studies such as cineangiography or computed tomography to confirm diagnosis^{6,7}. It is considered that 2D transthoracic echocardiography is enough to make an accurate diagnosis without using other diagnostic means, and allows the follow-up of operated patients without exposing them to risk.

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