



Atrioventricular Discordance Associated to Double-Outlet Right Ventricle. A Rare Case in a 28-year old Patient with no Surgery

Discordancia Atrioventricular Asociada a ka Doble Vía de Salida del Ventrículo Derecho. Un Raro Caso con 28 años y Ninguna Cirugía

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SUMMARY

We report a rare case of a 28 years old patient with the diagnosis of atrioventricular discordance and double outlet right ventricle, obtained only after transesophageal echocardiogram and no surgery. Severe pulmonary stenosis was the natural banding allowing this outcome.

Descriptors: Congenital Heart Diseases; Double Outlet Right Ventricle; Echocardiography, Transesophageal; Pulmonary Valve Stenosis

RESUMEN

Relatamos el raro caso de un paciente de 28 años, con diagnóstico de discordancia atrioventricular y doble vía de salida del ventrículo derecho, obtenido solamente después del ecocardiograma transesofágico y sin cirugía previa en donde la estenosis pulmonar importante actuó como vendaje pulmonar natural, posibilitando esa evolución.

Descriptores: Cardiopatías Congénitas, Doble Vía de Salida del Ventrículo Derecho, Ecocardiografía Transesofágica, Estenosis

Introduction

Innumerable complex congenital cardiopathies may be associated to cardiac looping abnormalities and ventricular septation. In embryos with normally positioned viscera, the primitive cardiac tube revolves to the right and ventricles have usual orientation. The left morphologically ventricle is on the left and posterior to the right morphologically ventricle. In many complex lesions, the primitive cardiac tube revolves to the left, resulting in ventricle inversion or atrioventricular discordance.

Double outlet is a specific ventriculo-arterial connection, and more than half of the circumference of both semilunar valves must be connected to the right morphologically ventricle^{1,2}.

Case Report

A.S., 28 years old, male, 1.79 meter high, 50 kg, complaining of dyspnea, low weight gain, and cardiopathy diagnosed at early childhood (cardiac murmur). Cardiac surgery was proposed when he was 12 years old, but not authorized by his parents. Symptoms got worse sin-

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Received on: 23/09/2012 - **Accepted on:** 29/10/2012



ce then. He has fatigue, excessive tiredness, shortness of breath and cyanosis on moderate exertion.

At clinical examination: Systolic murmur in left sternal border, more audible at the tip, and pancardic murmur with B2 unfolding, blood pressure 110 x 70 mmHg, dyspnea +++/4, cyanosis ++/4, anicteric. No visceromegaly, no edema, lungs with good expansion and slight bilateral basal crackles.

Transthoracic echocardiogram did not define the type of ventriculo-arterial connection. Transesophageal echocardiogram showed situs solitus with atrioventricular discordance and ventriculo-arterial connection of the double outlet right ventricle type (located on the left). Broad interventricular communication (IVC) related to the pulmonary artery, approximately 1.7 cm with flow preferably from the LV to the RV, no significant gradient. Moderate dilation of the left ventricle (located on the right) and of the right atrium. Mild biventricular hypertrophy with preserved functions. Important mitral incompetence and moderate tricuspid. Slight pulmonary trunk hypoplasia, anterior and to the right of the aorta,

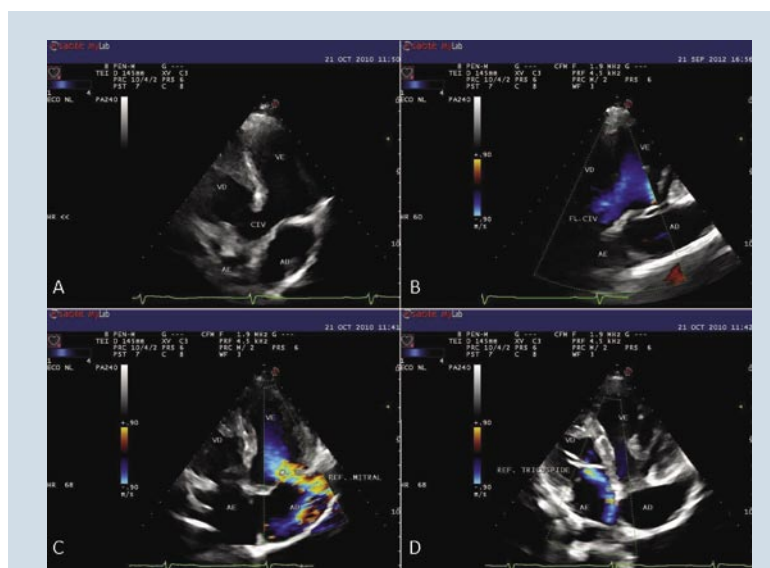


Figure 2: In A and B, we see aspects of ventricular septal defect (VSD) to two-dimensional echo and color flow mapping. In C and D the mitral and tricuspid valve regurgitation. RA = right atrium, LA = left atrium, RV = right ventricle, LV = left ventricle)

pulmonary stenosis with no incompetence. Right ventricle-pulmonary trunk systolic gradients: maximum of 75 mmHg and mean of 50 mmHg (Figures 1 and 2). The aorta measured 3.1 cm in its origin, slightly posterior and to the left of the pulmonary artery, no obstructions. The aortic arch was located to the left, no anatomical changes. Slight pericardium effusion.

Discussion

Lesions associated to atrioventricular discordance are common and IVC is the most common one (around 88%). It is of the perimembranous type most of the cases. Our patient had subpulmonary IVC, which represents only 5% of associated IVCs (Figure 3). Pulmonary stenosis, by valve or subvalve involvement, is found in around 80% of the cases.

In double outlet right ventricle ventriculo-arterial connections, intraventricular communication is classified by Zamora et al.³ according to its association to great heart basis vessels. The communication can be subaortic, subpulmonary, double related and not related.

Very rarely, there is no interventricular communication and blood from the left

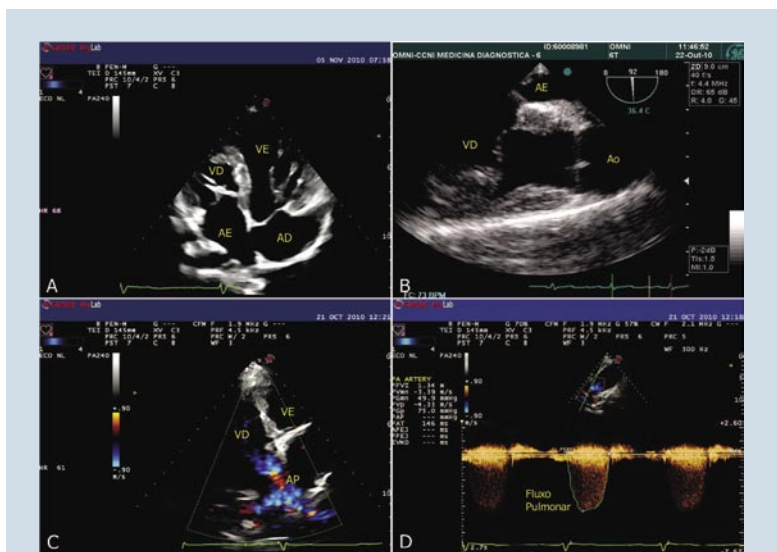


Figure 1: Patient with atrioventricular discordance and double-outlet right ventricle. In A, it is possible to see atrioventricular discordance. It was only possible to confirm the origin of the aorta by transesophageal echocardiogram (B). In C, it is possible to see the pulmonary artery originated in the right ventricle (RV). This confirms the double-outlet right ventricle diagnosis. The patient also had pulmonary stenosis with a maximum right ventricle-pulmonary trunk systolic gradient of 75 mmHg (D). RA = right atrium; LA = left atrium; LV = left ventricle; PV = pulmonary valve.

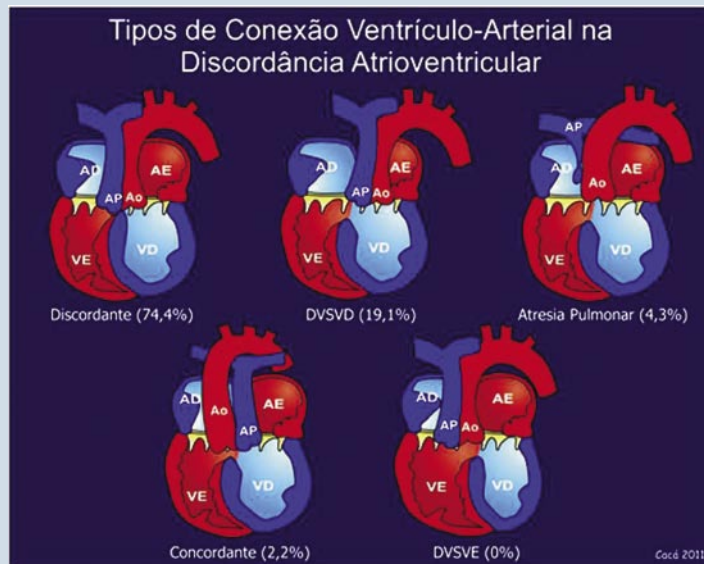


Figure 3: Scheme with types of ventriculo-arterial (VA) connection that may be present in cases of atrioventricular (AV) discordance. According to Tenório de Albuquerque et al.⁴, in a review of a series of patients with AV discordance, it was noted that 74.4% had VA discordance, 19.1% had double outlet of the right ventricle, 4.3% had pulmonary atresia, 2.2% had VA concordance and no case of double outlet of the left ventricle. RA = right atrium; LA = left atrium; Ao = aorta; PA = pulmonary artery; LV = left ventricle; RV = right ventricle.

chambers reaches the right atrium by interatrial communication, right ventricle and aorta. The double outlet of the right ventricle is called not related IVC when IVC is distant from both semilunar valves and is represented by localized defects in the entrance portion of the septum, type of defect of the atrioventricular septum, or in the trabecular portion of the muscle septum.

Its clinical manifestations are generally related to severe pulmonary stenosis, pulmonary atresia, with or without interventricular communication, or only broad interventricular communication. Cyanosis and dyspnea are the most common symptoms. We have observed in our case significant pulmonary stenosis with maximum right ventricle-pulmonary trunk systolic gradient of 75 mmHg, associated to dyspnea and cyanosis.

In atrioventricular discordance (AVD), as to the position of the heart in the thorax, around 70% are positioned to the left, with the aortic arch totally turned to the left. In relation to the types of ventriculo-arterial connections, ventriculo-arterial discordance is found in the great majority of the cases (around 75%). The AVD case with double outlet of the right ventricle re-

presents a bit over 19% of possible changes (Figure 2). In relation to the position of the ventricles, in most of the cases, they are side by side, or the morphologically right ventricle in anterior position. The aortic valve is predominantly to the left and anterior to the pulmonary artery or to the left side by side with the pulmonary artery.

In a study presented by Albuquerque et al.⁴, the most common electrocardiographic findings were sinus rhythm with few cases showing total atrioventricular blocking; SAQRS vector deviation (only 20% of the cases are between -20° and $+90^\circ$) and ventricular overload, the right overload is more common than the left overload. Thorax x-ray in almost half of the patients with this association is normal.

Echocardiography, especially transesophageal echocardiography, is one of the most important complementary exams in these situations. This exams allows the physician to see in a quick and simple way morphological and functional aspects of complex congenital cardiopathies, in addition to hemodynamic repercussion.

Palliative surgical corrections depend on initial clinical condition. In most of the cases, the objective is decreasing hyperflow or improving cyanosis. The most common procedures are pulmonary artery banding surgery and shunts (Blalock-Taussig and Waterston). In cases with significant pulmonary stenosis, the choice is pulmonary valvotomy.

Definite surgery includes ventricular septoplasty associated to broadening of the pulmonary trunk, with or without tube. A more complex surgery can be performed (Mustard surgery associated to Rastelli surgery)⁵.

In the case presented, we have noted important systolic gradient in the pulmonary trunk (maximum right ventricle-pulmonary trunk gradient of 75 mmHg). This allowed the 28-year old patient to follow without surgical intervention once pulmonary stenosis functionally acted like a natural pulmonary artery banding process, which would be the palliative proposal for this congenital cardiopathy.

We also highlight the importance of the transesophageal echocardiography which was crucial to provide adequate assessment of atrioventricular and ventriculo-arterial connections in this patient.



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