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Innocent Murmur since Childhood: Elderly Patient with Undiagnosed Interventricular Septal Defect

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ABSTRACT

Ventricular septal defects in adulthood only account for 10 per cent of congenital heart defects, In patients with evidence of LV volume overload and no Pulmonary Arterial Hypertension (no non-invasive signs of Pulmonary artery pressure elevation or invasive confirmation of Pulmonary Vascular Resistance <3 Woods Units in case of such signs), VSD closure is recommended regardless of symptoms. “Maladie de Rogers” is referred to as a VSD with a small left-to-right shunt, the pulmonary vascular resistance is not significantly elevated and the left-to-right shunt is small (Qp: Qs <1.5:1), in the absence of aortic valve prolapse and regurgitation or Infective endocarditis, may be managed by observation without need for operative intervention.

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INTRODUCTION

In the world, the estimated incidence of congenital heart disease (CHD) is 4-5 per 1000 births. In Mexico, there are no databases that systematically record its frequency, so the calculation is based on an estimated prevalence of 8-10 per 1000 newborns (NB), and extrapolated to the birth rate, 18,000 to 20,000 new cases per year are expected. (1)

Ventricular septal defect (VSD) is one of the most common congenital heart defects (second only to bicuspid aortic valve) at birth but accounts for only 10 per cent of congenital heart defects in adults because many close spontaneously. (2)

Adults with an isolated small restrictive VSD with the small left-to-right shunt (often referred to as "Maladie de Roger") generally remain asymptomatic and present with a systolic murmur, often with a palpable thrill from the VSD. There is a risk of endocarditis associated with this lesion, but the magnitude of risk is low.

Small restrictive defects of the muscular or membranous septum may be managed by observation without the need for operative intervention. However, 6% of patients with small supracristal (subaortic) or perimembranous defects may develop aortic valve prolapse and resultant AR that may be progressive. (4)

CASE REPORT

A 75-year-old patient with diabetes and long-standing arterial hypertension referred to the cardiology clinic because of cardiovascular risk factors. In the interrogation, the patient refers to an "innocent murmur in childhood", in her clinical evaluation the patient finds an holosystolic murmur of greater intensity in the 4th intercostal left parasternal space with bar irradiation to the 4th intercostal right parasternal space.

The chest X-ray shows a left heart profile with increased left atrial silhouette (Fig 1), the electrocardiogram shows a QRS of 120 msec, with aQRs of -30°, with an image suggestive of anterior bundle branch block of the left bundle, presents deep S in DIII and AFV together with inversion of the T wave as data of overload of the right ventricle (Fig 2), for this reason, an echocardiogram was performed where a non-dilated left ventricle was found, Normal LVEF, without mobility alterations, without increased LV filling pressures, with the slightly dilated left atrium, without valvular heart disease, with non-dilated right ventricle with preserved systolic function, a 2 mm gauche perimembranous interventricular septal defect was found (Fig 3), a Qp/Qs of 1.02:1 was calculated.

Because the patient is asymptomatic, without dilation or overloading of the left cavities, it was decided that the patient should continue to be closely monitored, until now she remains asymptomatic.

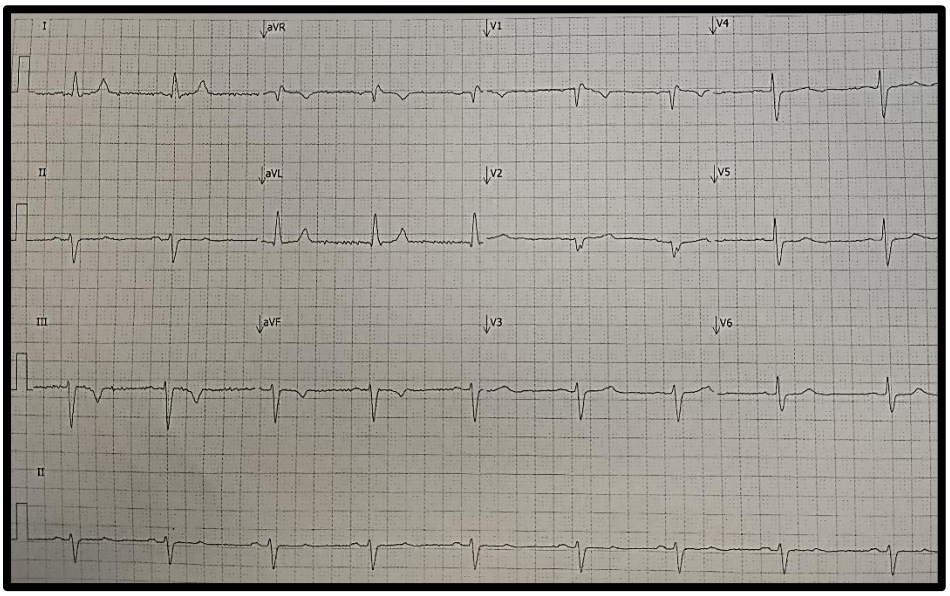

Fig 1)

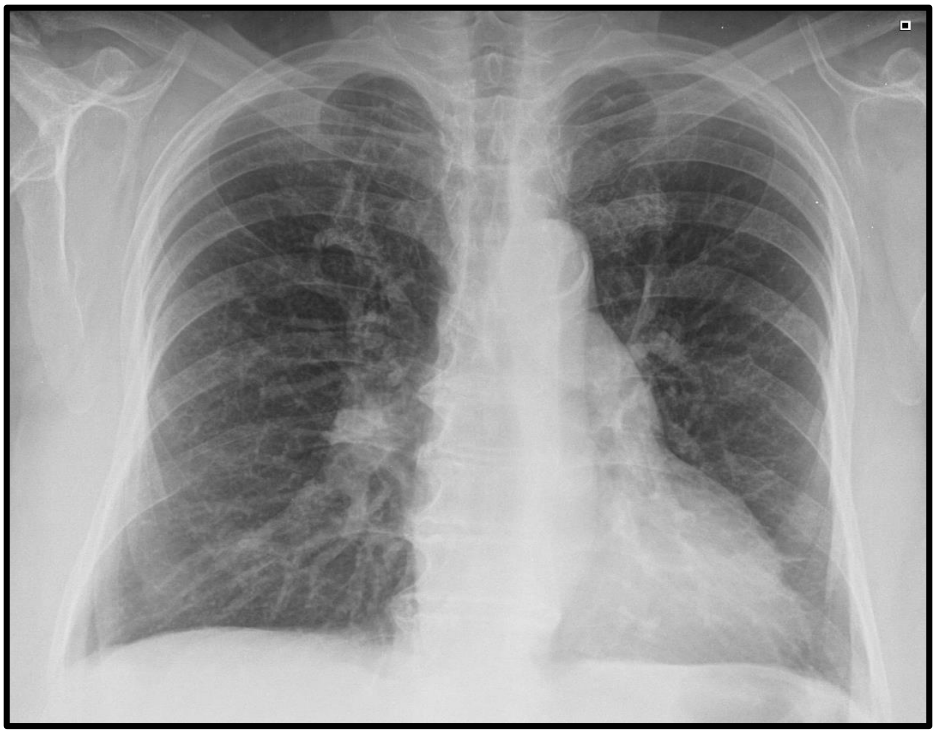

Fig 2)

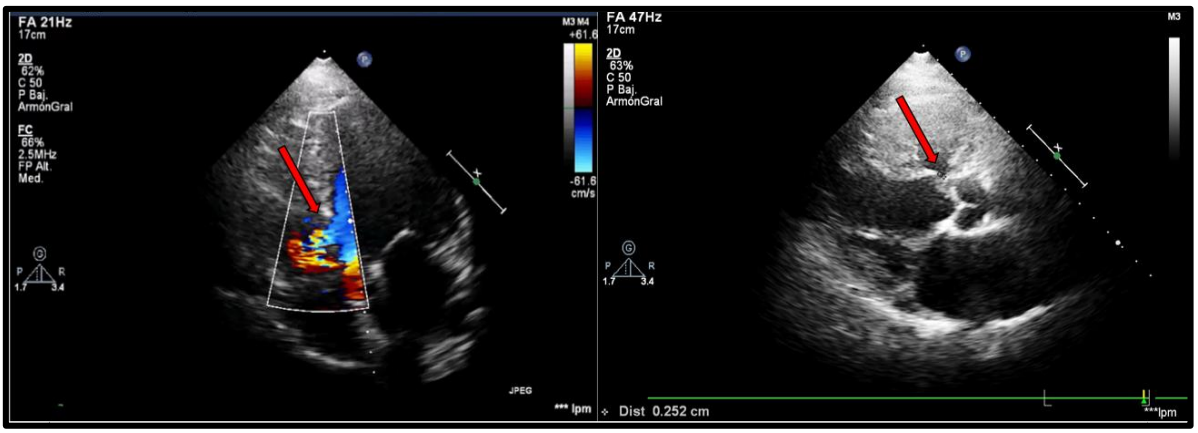

Fig 3)

DISCUSIÓN

Several locations of the defect within the interventricular septum are possible, and these can be divided into four groups according to their location within the RV (nomenclature varies and synonyms are added)[3]:

• Perimembranous/ perimembranous/ subaortic/ conoventricular (most common, 80% of Ventricular septal defects; located in the membranous septum with possible extension into the inlet, trabecular, or outlet septum; adjacent to tricuspid and aortic valve; so-called aneurysms of the membranous septum i.e. tissue from the septal leaflet of the TV are frequent and may result in partial or complete closure).

• Muscular/trabecular (up to 15-20%; surrounded by muscle; various locations; frequently multiple; spontaneous closure particularly frequent).

• Outlet (with or without malalignment of the outlet septum)/ supracristal/sub arterial/subpulmonary/infundibular/conal/doubly committed juxta-arterial [5%; located beneath the semilunar valves in the conal or outlet septum; may be associated with progressive aortic regurgitation (AR) due to prolapse of the right aortic cusp and aneurysm of the sinus of Valsalva].

• Inlet/AV canal/AVSD type (an inlet of the ventricular septum immediately inferior to the AV valve apparatus; associated with a common AV valve; may be associated with AV septal malalignment and straddling TV; typically occurring in Down syndrome).961

The direction and magnitude of the shunt are determined by PVR and systemic vascular resistance, the size of the defect, LV/RV systolic and diastolic function, and the presence of right ventricular outflow tract (RVOT) obstruction (RVOTO) and left ventricular outflow tract (LVOT) obstruction (LVOTO). [5]

Several possible problems may occur with advancing age:

• A double-chambered RV (DCRV) can develop over time, mostly in perimembranous defects, and maybe a result of the jet lesion of the RV endothelium caused by the high-velocity VSD jet.

• In the case of an outlet (supracristal) VSD (less commonly perimembranous), there is a risk for prolapse of the right coronary (or non-coronary) cusp of the aortic valve, resulting in progressive AR and formation of a sinus of Valsalva aneurysm.

• Arrhythmias can occur but are less frequent than in other forms of Congenital Heart Disease.

• Complete heart block rare nowadays was not uncommon in the earlier years of cardiac surgery, so can occur, especially in older patients. These patients usually require lifelong pacing.

• Late LV dysfunction and heart failure.

• Endocarditis.

In the absence of aortic valve prolapse and regurgitation or IE, small restrictive defects of the muscular or membranous septum may be watched conservatively without the need for operative intervention. [4]

For patients with small defects (Qp: Qs <1.5:1 and low PA pressure), the survival rate was 96%. (4)

CONCLUSION

VSD in adults accounts only for 10% of congenital heart disease.

Adults with small persistent VSDs with normal pulmonary artery pressure generally do not require intervention and have an excellent prognosis; however, these patients may remain at risk of development of endocarditis, DCRV, or aortic regurgitation (AR) and may be watched conservatively without the need for operative intervention. [6]

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