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SCIENTIFIC LETTER

Straddling of the mitral valve in the set of *situs solitus* in dextrocardia with atrio-ventricular connection discordance and double-outlet right-ventricle: a rare case report

Straddling *de la válvula mitral en el contexto de* situs solitus *en dextrocardia con conexión atrio-ventricular discordante y doble vía de salida del ventrículo derecho: un raro reporte de caso*

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A 3-year-old male with cyanosis and a heart murmur since birth. Class I of Ross. Physical examination: SaO_2 72%, cyanosis ++, ejective heart murmur on high parasternal right border III/VI, normal pulses, digital clubbing +++; chest plain: cardiothoracic ratio 0.51, *situs solitus* with dextrocardia, and decreased pulmonary flow (Fig. 1A); electrocardiogram: sinusal rhythm with biatrial growth and biventricular hypertrophy; transthoracic echocardiogram: *situs solitus*, dextrocardia with dextroapex, double superior vena cava (left superior vena cava drains into coronary sinus), ostium secundum 15 mm atrial septal defect with bidirectional shunt, pulmonary venous return to the left atrium, discordant atrioventricular (AV) connection, right straddling of AV valve (mitral) with light-moderate regurgitation, ventricular septal defect (VSD) of 17 × 16 mm (Figs. 1B and C), double outlet right ventricle (DORV) with right ventricle (RV) spatially at the left side (aorta anterior with pulmonary artery posterior with severe mixed pulmonary stenosis with maximum gradient of 62 mmHg, confluent pulmonary branches with normal size, left aortic arch without obstruction and normal biventricular function.

The catheterization procedure was in the same line as the echocardiogram with infundibular pulmonary stenosis (Fig. 2) and a peak-to-peak gradient of 68 mmHg. The end-diastolic pressure of both ventricles is 15 mmHg, pulmonary arteriolar vascular resistance is 2.62 UW, the Nakata Index is 475, and the McGoon Index is 2.52 and double superior vena cava without innominate vein.

As management, the cardiology team offered bidirectional cavo-pulmonary shunt; however, the procedure has not been carried out at the moment, because of sociocultural issues by the parents.

The straddling of AV valves has usually been described either in hearts in which they connect the morphologic right atrium to both the right and left ventricles, with the ventricles normally related, or in which they connect the left atrium to both normally related ventricles[1]. More rarely, the tricuspid valve may straddle a septum between a larger right-sided chamber of left ventricular morphology and a left-sided smaller chamber of right ventricular morphology[1]. However, a right-sided AV (mitral) valve straddling a septum in the setting of AV discordance is exceedingly rare[1].

Thus, straddling of the mitral valve (SMV) is a malformation that is invariably associated with a VSD and frequently with conotruncal anomalies[2-4]. The etiology

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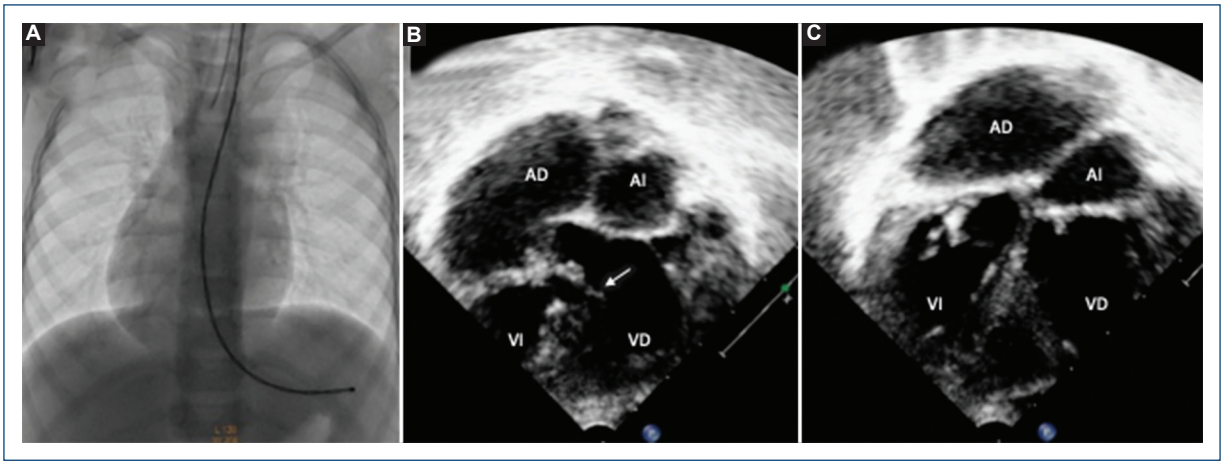

Figure 1. A: chest fluoroscopy: *situs solitus* in dextrocardia. **B** and **C:** transthoracic echocardiogram (apical 4-cambers view): straddling (arrow) of right AV valve (mitral) and anatomical right ventricle (RV) at the left side (dropped trabeculation and moderated band).

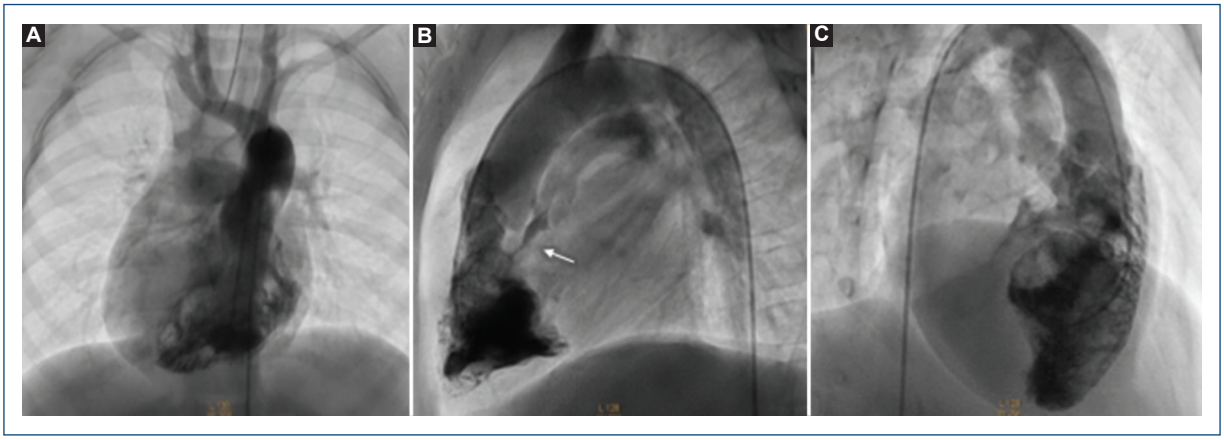

Figure 2. Anatomical right ventriculography: from the right ventricle both vases emerged (aorta anterior and pulmonary artery posterior with subpulmonary stenosis [arrow]). **A:** postero-anterior projection, **B:** left lateral projection, **C:** right anterior oblique (RAO) 45° + cranial 25° (4-cambers projection for dextrocardia).

and morphogenesis of SMV remain incompletely understood, but its anatomic features have been well described based on autopsy material[1-5]. These reports indicate that the mitral valve (MV) straddles the anterior aspect of the septum through a conoventricular-type VSD malalignment. In addition, they show that the most common associated conotruncal anomaly is DORV, followed by ventriculo-arterial discordance or transposition of the great arteries. They also highlighted that left and right ventricular size varies considerably[1,5]. However, because of its rare occurrence, most published reports on SMV are based on a relatively small number of cases, and no consistent anatomic classification has been proposed.

The morphologic and functional features of SMV in living patients have not been described in detail, and few reports have addressed the echocardiographic features of SMV[6,7]. Moreover, information on midterm outcomes and pre-operative echocardiographic features predictive of mortality is scarce. Only a few surgical reports have included patients with SMV[8,9]. Thus, the spectrum of echocardiographic findings and the optimal surgical management of patients with SMV are still unclear. Fraisse et al.[9] proposed SMV classification based on echocardiographic findings in a study of 46 patients, who reported their midterm clinical outcome, and identified pre-operative variables predictive of mortality.

Patients in *situs solitus* with dextrocardia may have concordance AV connection in 48.3% and discordance AV connection in 43.3%. However, only 13% of the patients in the latter group have been reported to have DORV[7-9].

In isolation, SMV is a rare malformation and much rarer in association with complex congenital heart disease, such as AV connection discordance; but has never been reported in association with DORV in the set of *situs solitus* in dextrocardia.

We report a rare case of SMV in a patient where the MV does not override the interventricular septum; rather the mitral subvalvular apparatus is clearly straddled. This case demonstrates that overriding and straddling are independent conditions that may or may not be associated.

Thus, it is very important to make the anatomical differential diagnosis principally with double-inlet RV; in this last situation, the AV annulus is overriding more of the 50% over the ventricular septum. Since in both pathologies the mitral and tricuspid subvalvular apparatus are located within the anatomical RV.

In the study of Becker et al.,[1] four pathological specimens were presented. Two of them presented similar morphological characteristics to our case. However, in their description, all cases were reported apparently in levocardia, but they did not commenting something about this situation. In the paper, an angiography of the first case was shown, where the heart in levocardia can be appreciated.

In another study of 125 patients with dextrocardia (Garg et al.[10]), *situs solitus* was found in only 43 patients. Of these, only 18 cases had AV connection discordance, out of which only 2 had DORV. However, they did not describe any case of straddling of the AV valve in the two patients.

In the present case, the most viable option of treatment is the univentricular physiology, and this is the management option preferable for our patient. Unfortunately, however, the parents have not accepted any type of surgery at the moment. In our country, it is common for parents to refuse to subject their children to heart surgery because of the risks of the surgery.

Finally, we report a very rare case of SMV in the set of *situs solitus* in dextrocardia with AV connection discordance and DORV with pulmonary stenosis. To our knowledge, a similar case has not been reported, thus making this report a very interesting one.

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Conflicts of interest

The authors declare no conflicts of interest.

Ethical considerations

Protection of humans and animals. The authors declare that no experiments involving humans or animals were conducted for this research.

Confidentiality, informed consent, and ethical approval. The authors have followed their institution's confidentiality protocols, obtained informed consent from all patients, and secured approval from the Ethics Committee. SAGER guidelines have been followed as applicable to the nature of the study.

Declaration on the use of artificial intelligence. The authors declare that no generative artificial intelligence was used in the writing or creation of the content of this manuscript.

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