Uncommon mitral valve anomalies associated with Ebstein anomaly

Alteraciones mitrales poco frecuentes asociadas con anomalía de Ebstein

To the Editor,

Ebstein anomaly (EA) is the most common congenital malformation of the tricuspid valve (TV). Its presentation varies from mild abnormalities with few clinical findings to severe forms that are incompatible with life. The most common associated defect is atrial septal defect, followed by pulmonary stenosis.[1] Association with left heart lesions is rare but includes bicuspid aortic valve, noncompacted myocardium, and mitral anomalies.

We present 2 cases of pediatric patients with EA associated with double-orifice mitral valve (DOM) in one and anterior mitral leaflet cleft (MC) in the other, assessed on 2-dimensional and 3-dimensional transthoracic echocardiography (2D-3DTTE). The parents gave informed consent via telephone to publish the cases in an article. To determine the volume of clinical cases of mitral lesions associated with EA in pediatric patients in our institution, we reviewed the electronic archive of examinations performed in the Pediatric Echocardiography Department of the National Institute of Cardiology of Mexico, from January 2012 to December 2019.

The first case was a 10-year-old boy with a history of palpitations and dyspnea. He was admitted to hospital with a supraventricular tachycardia that was chemically cardioverted. On admission, 2D-3DTTE showed a TV with 40% displacement of the septal leaflet, limited movement of the anterior leaflet, no compromise of the posterior leaflet, and moderate tricuspid regurgitation (figure 1A). Three-dimensional echo of the mitral valve showed a DOM, with a larger 16-mm orifice in a medial position with 2 leaflets: anterior and posterior, with adequate opening; the smaller lateral orifice, in a more apical position, measured 5.7 mm, and had failure of coaptation (figure 1B and video 1 of the supplementary data), with severe regurgitation (figure 1C), and subvalvular apparatus with accessory papillary muscles. Pulmonary systolic pressure was calculated at 38 mmHg and ejection fraction was 51%.

The second case was a 12-year-old girl referred for assessment of supraventricular tachycardia, with 2 previous ablations. 2D-3DTTE showed EA with 50% displacement of the septal leaflet, limited movement of the anterior leaflet, mild regurgitation, a hypertrophic, anatomically small right ventricle (figure 2A-C), patent foramen ovale, and severe mitral regurgitation (figure 2D) caused by the MC, which completely divided the anterior leaflet and ran toward the left ventricular outflow tract, as shown on the 3DTTE images (figure 2E,F and video 2 of the supplementary data). Significant dilatation of the left heart was observed, with

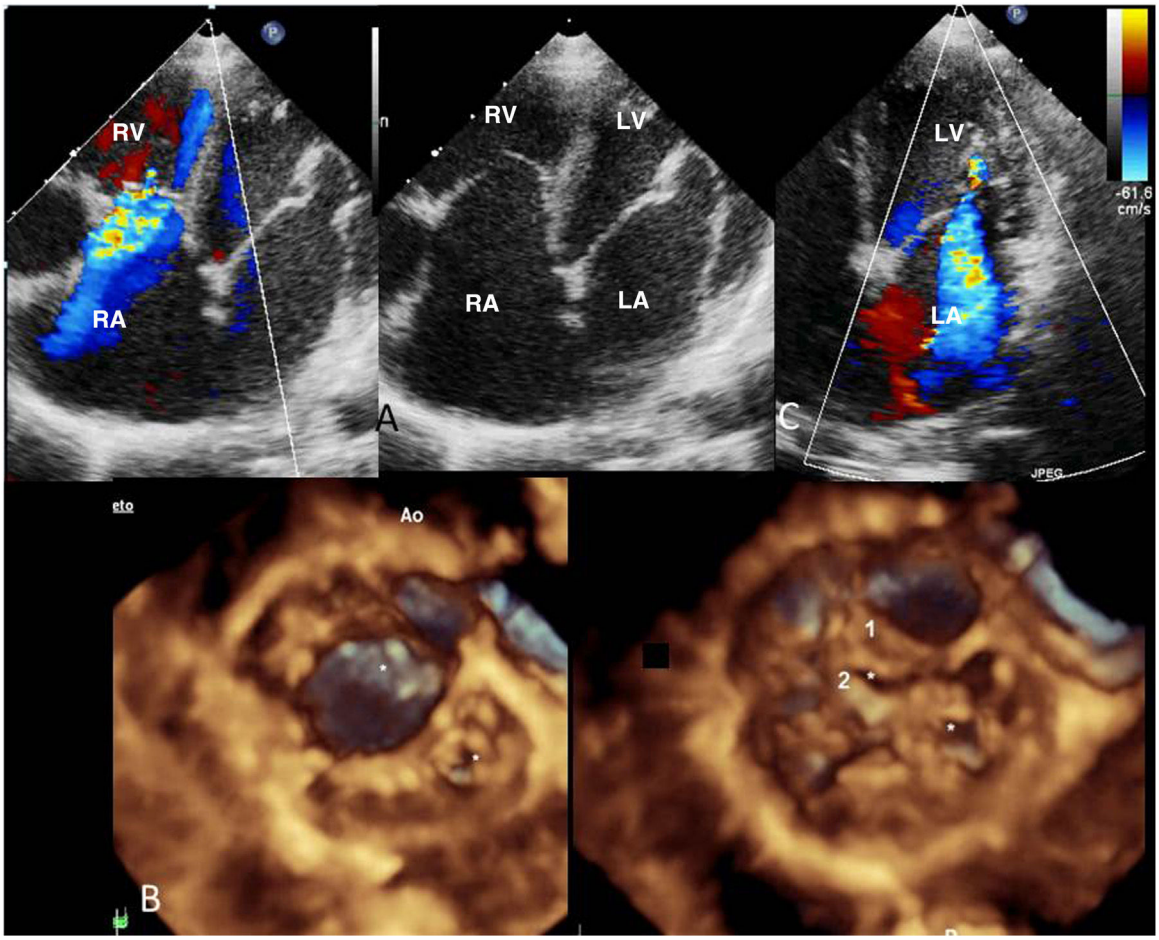

Figure 1. TTE images on apical 4-chamber view. A: displacement of the septal leaflet of the tricuspid valve and moderate tricuspid regurgitation on color Doppler. B: 3DTTE reconstruction from the ventricular view; both mitral orifices can be seen in diastole and systole, the larger, medial orifice has 2 leaflets: anterior (1) and posterior (2); the smaller, lateral and more apical leaflet has failure of coaptation. C: severe mitral regurgitation is observed originating at the smaller orifice (video 1 of supplementary data). Ao, aorta; LA, left atrium; LV; left ventricle; RA, right atrium; RV, right ventricle; TTE, transthoracic echocardiogram.

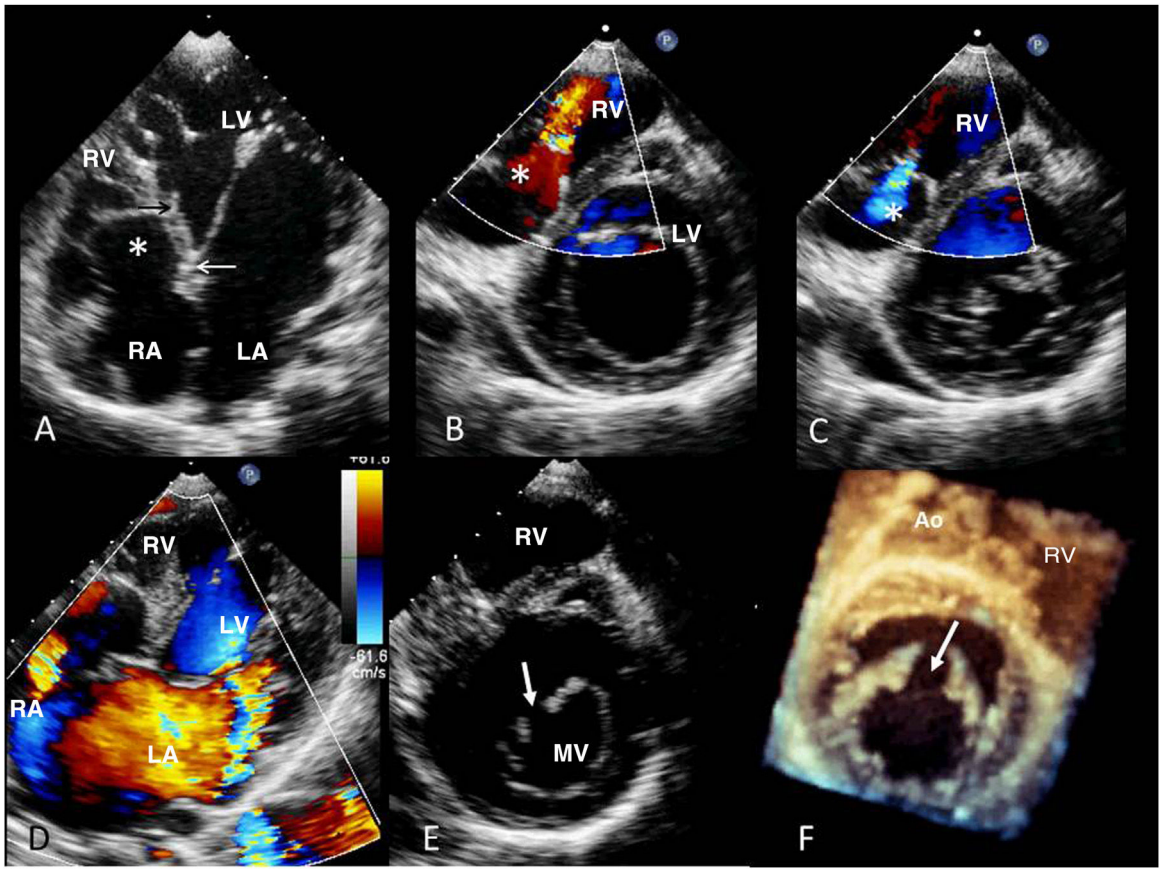

Figure 2. TTE on apical 4-chamber view showing displacement of the septal leaflet of the tricuspid valve (A) (black arrow) in relation to the mitral annulus (white arrow) and atrialization of the right ventricle (asterisk), and on parasternal short-axis view in diastole (B), limited movement of the anterior leaflet and displacement of the septal leaflet and mild regurgitation in systole (C). D: severe mitral regurgitation, dilatation of the left atrium, and patent foramen ovale. The 2 D parasternal short-axis (E) and the 3 D reconstruction from the ventricular surface (F) show the mitral cleft (arrow) that divides the anterior leaflet, running toward the left ventricular outflow tract. See video 2 of the supplementary data. Ao, aorta; LA, left atrium; LV, left ventricle; MV, mitral valve; RA, right atrium; RV, right ventricle; TTE, transthoracic echocardiography. This figure is shown in full color in the electronic version of the article.

ejection fraction of 56% and mean pulmonary artery pressure of 26 mmHg.

Our review of the archives found that, during this period, 25 842 echocardiograms were performed in pediatric patients, as outpatients and inpatients, of which 236 studies from 149 patients had a diagnosis of EA (0.91%). For the analysis of associated lesions, 4 patients with EA in the context of transposition of the great arteries were excluded. We observed a slight male predominance with a ratio of 1.1:1, and a mean age of 8.72 ± 5.3 years. Assessment of severity based on apical displacement of the septal TV leaflet measured on TTE[2] showed that 26% corresponded to mild EA, 25% to moderate, and 49% to severe. A total of 82.5% of the patients had at least 1 associated defect; the most common was atrial septal defect, in 117 patients, of whom 78 (54%) had an ostium secundum-type defect (2 of them had partial anomalous pulmonary venous connection) and 39 (27%) had patent foramen ovale, followed by ventricular septal defect in 13 (9%), persistent ductus arteriosus in 10 (7%), pulmonary stenosis in 6 (4%), and pulmonary atresia in 1 (0.7%), all of whom also had atrial septal defects, and 1 had tetralogy of Fallot. The left heart lesions found were bicuspid aortic valve in 1 (0.7%), mitral valve lesions in 11 (7.6%), and noncompacted myocardium in 3 (2%).

The mitral abnormalities found were prolapse with mild regurgitation in 7 patients (1 also had noncompacted myocardium), 2 with mitral dysplasia: 1 with moderate and the other with severe regurgitation, 1 patient with MC, and another with DOM, both with severe mitral regurgitation, which are the cases presented here. The severity of the septal TV leaflet displacement in this group was 1 patient with mild tethering, 5 with moderate, and 5 with severe.

Ebstein's original description mentioned a mild deformity of the mitral valve (MV); however, there are few publications reporting associated mitral abnormalities.[3] Gerlis et al.[4] described 14 patients with abnormal MV due to valve, chordae, or papillary muscle abnormalities. More recently, Attenhofer Jost et al.[5] published a series of 106 patients of whom 39% had left heart abnormalities affecting the myocardium (including systolic or diastolic ventricular dysfunction) and the mitral and aortic valves; the mitral lesions included prolapse in 16 patients and dysplasia in 4.

Lesions of the MV associated with EA such as MC or DOM are uncommon: there is only 1 case report with associated DOM,[6] and none with MC. The severity of mitral regurgitation can affect the clinical presentation and patient outcomes in a heart disease that is generally well tolerated in childhood. 3DTTE provides detailed morphology of the TV and associated lesions, and in these cases of the MV, due to the advantage of unconventional views from the left atrium or ventricle.

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AUTHORS' CONTRIBUTIONS

C.A. Vázquez Antona contributed to the study conception and design, data analysis and interpretation, writing the article, and approval of the final article for publication. M.R. Álvarez Macedo contributed to obtaining and interpreting data and review and approval of the final version of the article.

CONFLICTS OF INTEREST

None.

APPENDIX. SUPPLEMENTARY DATA

Supplementary data associated with this article can be found in the online version available at https://doi.org/10.1016/j.rec.2021.01.013

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First experience of percutaneous Bi-Pella in Spain

Primera experiencia con Bi-Pella percutáneo en España

To the Editor,

Refractory cardiogenic shock in patients with severe biventricular dysfunction is a therapeutic challenge, as it often requires short-term mechanical circulatory support (ST-MCS) devices as a bridge to heart transplant (HT). Choosing the type of ST-MCS to use is a complex process. Univentricular ST-MCS devices enhance antegrade flow, leading to increased contralateral venous return, which can cause dysfunction of the ventricle due to lack of contractile reserve. Therefore, ideally these patients benefit from biventricular ST-MCS or extracorporeal life support systems such as the venoarterial extracorporeal membrane oxygenator (VA-ECMO), which performs the function of both ventricles simultaneously. However, these devices complicate transplant surgery and clinical course. Until now, central biventricular ST-MCS has been used most commonly. The cannulas are placed directly into the heart chambers or great vessels, which means that further surgery for heart transplant can prolong graft ischemia time and encourage bleeding. Moreover, VA-ECMO is usually inserted percutaneously into central vessels and, therefore, is less invasive but entails risks, such as increased afterload of the left ventricle (LV), complications related to vascular accesses (eg, limb ischemia), and high red blood cell and platelet destruction, which must be replaced with transfusions, thereby promoting cytotoxic antibody production. The recent development of percutaneous ST-MCS for the right ventricle (RV) combined with percutaneous ST-MCS for the LV offers an alternative. We describe the first case in Spain of percutaneous implantation of the biventricular Impella (Bi-Pella) as a bridge to HT, using the Impella CP and Impella RP catheters (Abiomed Inc, United States) to partially carry out LV and RV function, respectively.

A 47-year-old man with familial dilated cardiomyopathy, severe biventricular dysfunction, and severe mitral and tricuspid regurgitation listed for HT was admitted to the Acute Cardiology Care Unit for cardiogenic shock, systemic congestion, and stage 3 acute kidney injury (AKIN). Due to poor tolerance of dobutamine therapy (sustained ventricular tachycardia) and progressive impairment of kidney function (INTERMACS 2), ST-MCS was considered necessary. In the preliminary assessment, echocardiographic predictors (tricuspid annular plane systolic excursion [TAPSE], 10 mm; S', 5 cm/s; RV/LV ratio, 0.9; shortening fraction, 25%) and hemodynamic predictors (filling pressure ratio, 1.6; pulmonary artery pulsatility index, 0.8; RV stroke work index, 0.3 mmHg/L/m^2) indicated a high risk of RV dysfunction and, therefore, percutaneous biventricular ST-MCS was performed as a bridge to HT. The devices were implanted in the catheterization lab under conscious sedation and analgesia. The Impella CP device was inserted through the left femoral artery (14 Fr) and the Impella RP, through the right femoral vein (23 Fr) (figure 1). Assist was initially set to levels P8 and P6, which provided flow at 3.4 and 3.1 L/min, respectively. The patient gave written consent for publication of his case and the respective images in a scientific journal, with a commitment to avoid disclosing identifying information.

Following implantation, the patient's clinical progress was favorable (table 1), with immediate hemodynamic improvement, reduced filling volumes and pressures of both ventricles, and improved tissue perfusion and kidney function. This allowed inotropic support to be reduced with no new arrhythmic events. ST-MCS was maintained for 5 days until HT, which was successful, with early extubation and withdrawal of vasoactive support (24 hours) and a subsequent 11-day stay in the postoperative cardiology care unit and 30-day hospital stay. Several complications emerged during the ST-MCS period. First, initial bleeding at