Associated double-orifice mitral and tricuspid valves without ostium primum defect

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Abstract
The simultaneous existence of double orifice right and left atrioventricular valves in the absence of ostium primum defects is rare and scarcely reported [1]. In the following, we present the case of a 20-month old boy diagnosed with tetralogy of Fallot with pulmonary atresia who was found to have associated double-orifice mitral and tricuspid valves.

Keywords: Double-orifice valve • Atrio-ventricular canal • Cleft • Ostium primum

INTRODUCTION
The simultaneous existence of double-orifice right and left atrioventricular valves in the absence of ostium primum defects is rare and scarcely reported [1]. In the following, we present the case of a 20-month old boy diagnosed with tetralogy of Fallot with pulmonary atresia who was found to have associated double-orifice mitral and tricuspid valves.

The child was admitted for complete surgical correction of tetralogy of Fallot with pulmonary atresia. His previous history included two modified Blalock-Taussig shunts performed in the neonatal period and at 6 months of age.

The echographic examination showed an intact interatrial septum, a single malalignment-type ventricular septal defect (VSD) and moderately hypoplastic pulmonary arteries. The examination also found a double-orifice mitral valve and mitral cleft (Fig. 1a). There was trace mitral insufficiency. Moderate tricuspid regurgitation was found; however, the tricuspid anatomy seemed normal.

At operation, the mitral valve was inspected and found to have the expected appearance. There was an antero-lateral orifice whose anterior mitral leaflet presented a cleft. There were no secondary chordal attachments on the two sides of the cleft. The postero-medial margin of the cleft continued with a fibrous bridge that connected the two leaflets in the central portion of the mitral valve (Fig. 1b). Primary chordae supported the free margin of the cleft and extended onto the bridging tissue. Thus, two orifices were present: one that was antero-lateral and whose anterior leaflet presented the cleft, and a postero-medial one that was smaller. The papillary muscles inserted normally. No repair was deemed necessary, as there was only trace mitral insufficiency. The inspection of the tricuspid valve revealed a double-orifice valve that was not suspected before the procedure. There was a tissue band that bridged the anterior and septal leaflets (Fig. 2). Tendinous chordae inserted on all valvular elements, including the bridging tissue. The tricuspid annulus was dilated without chordal prolapse or rupture. The ventricular inlet septum was normally developed and there was no ostium primum defect. The patient underwent a complete correction of his cardiac malformation, with the closure of his VSD by a Dacron patch and pulmonary transannular patch enlargement. A small foramen ovale was surgically created.

His intensive care stay was marked by severe right ventricular dysfunction requiring extracardiac membrane oxygenation. An echographic examination revealed severe pulmonary insufficiency, a hypertrophic, non-compliant right ventricle and right-to-left shunt across the foramen ovale. The tricuspid insufficiency that was judged to be mild before the procedure had progressed to moderate. A decision to reintervene was taken. The right ventricular outflow tract was repaired with a 14 mm pulmonary homograft. The mechanism of tricuspid regurgitation was judged to be by annular dilatation. An associated tricuspid annuloplasty was performed, extending from the antero-septal commissure anteriorly to the postero-septal commissure posteriorly with a heterologous pericardial strip (CardioFix®, Medtronic).

His subsequent evolution was uneventful. The latest echographic follow-up at 7 months shows trace mitral and tricuspid incompetence, good homograft function and normal ejection fraction.

COMMENT
Double-orifice atrioventricular valves (DOAVV) are incidental findings. While a left DOAVV is found with non-negligible frequency, there are to date few reported cases of a right DOAVV. Furthermore, most reported cases of right DOAVV appeared in
the setting of atrio-ventricular canal defects and there is to date, to the best of our knowledge, only one report of their association with left-sided defects [1].

The development of a DOAVV has been explained in several ways. As most cases reported to date have occurred in the setting of a total or partial atrio-ventricular canal, some authors consider the anomaly to occur after the formation of the leaflets is complete, as a result of partial fusion of some of the five atrio-ventricular leaflets along their zones of apposition [2]. A second theory suggests an incomplete delamination during valve formation, which would explain the ‘hole’-type DOAVV, where an accessory orifice is found in the mid-portion of a leaflet [3]. However, the fact that left DOAVV appear to be much more frequent may suggest different embryologic mechanisms for right- and left-sided defects. Indeed, other authors consider a right DOAVV to appear as a result of the persistence of the inferior orifice in the floor of the tricuspid gully, developed in the 7th week of intrauterine development, with subsequent delamination producing a double-orificial valve [4].

The mitral cleft should be surgically addressed if it results in significant alteration of valve function. In our case, the repair was not deemed necessary, as there was trace mitral regurgitation.

Another option, as the position of the papillary muscles was normal, could have been to close the defect. The postoperative right-heart failure was most probably the result of the loss of pulmonary valve function, as demonstrated by the uneventful recovery of the patient once the right outflow tract was repaired by a homograft.

Surgical treatment of a DOAVV depends on the degree of chordal involvement, provided that there is no alteration of the valve cusps. In our case, a simple restrictive tricuspid annuloplasty was performed, as there was no prolapse or restriction of leaflet motion. The bridging tissue should be respected, as dividing it can result in severe incompetence [5].

Our case presented the rare occurrence of a conotruncal defect and associated DOAVV’s in the absence of ostium primum defect.

Conflict of interest: none declared.

REFERENCES