European Journal of Cardio-Thoracic Surgery Advance Access published January 20, 2012

European Journal of Cardio-Thoracic Surgery 0 (2012) 1–3
doi:10.1093/ejcts/ezr159

CASE REPORT

CONGENITAL

Double-orifice tricuspid valve: a rare entity

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Received 27 July 2011; received in revised form 9 September 2011; accepted 16 September 2011

Abstract

Duplication of the atrioventricular valves is a rare anomaly and is more commonly seen in the mitral position. We report the case of a 14-year old child with a large inlet muscular ventricular septal defect, who had a double-orifice tricuspid valve, which was detected incidentally during surgery.

Keywords: Double-orifice tricuspid valve • Valvar duplication

INTRODUCTION

Duplication of an atrioventricular valve is an extremely uncommon congenital anomaly that generally affects the mitral rather than the tricuspid valve [1–7]. Since Pisenti described the first case of tricuspid duplication in 1888, only 25 additional cases have been reported in the literature [2 –4]. Hartmann [1] first classified the types of double-orifice atrioventricular valves in 1937, and more recently Sanchez-Cascos and associates reviewed the literature and reported a new case of their own [1–7]. We present here a case of a duplicated right atrioventricular orifice in a child with a large inlet muscular ventricular septal defect (VSD), which was detected incidentally during surgery.

CASE REPORT

A 14-year old female was admitted to our institute with a history of recurrent respiratory tract infection and symptoms of breathlessness on exertion NYHA class II. On clinical examination, there was a loud systolic murmur and a prominent pulmonary component of the second heart sound. The chest X-ray revealed cardiomegaly and pulmonary plethora. The electrocardiogram revealed sinus rhythm and biventricular hypertrophy. The transthoracic echocardiogram showed situs solitus, levocardia, normal systemic and pulmonary venous connections, and a large inlet muscular VSD with a bidirectional shunt predominantly left to right, mild tricuspid and pulmonary regurgitation and trivial mitral regurgitation. The pulmonary artery end-diastolic pressure was 50 mmHg and right ventricular systolic pressure (RVSP) was120 mmHg. Cardiac catheterization was done in view of severe pulmonary hypertension and the pre-oxygen haemodynamic data showed systemic pulmonary arterial pressures with a left-to-right shunt of 1.7:1 and the pulmonary vascular resistance index (PVRI) of 6.6 Wood units (WU). The post-oxygen data showed a left-to-right shunt of 3.2:1 and PVRI of 4.0 WU. The right atrial mean and the right ventricular end-diastolic pressures were 3 mmHg. The left ventriculogram showed a posteriorly placed mid muscular VSD and the left-arm injection showed left superior vena cava draining into coronary sinus. On these findings, the decision was made to close the VSD.

OPERATIVE FINDINGS

There was a double-orifice tricuspid valve (DOTV) with equal-size orifices in the side-to-side position and the well-formed leaflets with a dividing leaflet having their own chordal apparatus. There was no regurgitation or stenosis observed (Fig. 1). There was a large inlet muscular VSD adjacent to the rightward opening of the tricuspid valve. The VSD was closed with a Dacron patch using 5-0 partially interrupted and partially continuous pledgeted polypropylene sutures.

POST-OPERATIVE ECHOCARDIOGRAPHY

Echocardiography was repeated because of the new operative finding and the subxiphoid sweeps in the coronal plane and a modified parasternal short-axis view revealed a double-orifice tricuspid valve and dilated annulus with mild tricuspid regurgitation (Fig. 2), no residual VSD and RVSP of 50 mmHg. The child recovered well after surgery and the post-operative course was uneventful.

DISCUSSION

The division of an atrioventricular valve into two similar and functioning units is described as a duplication of the valvar

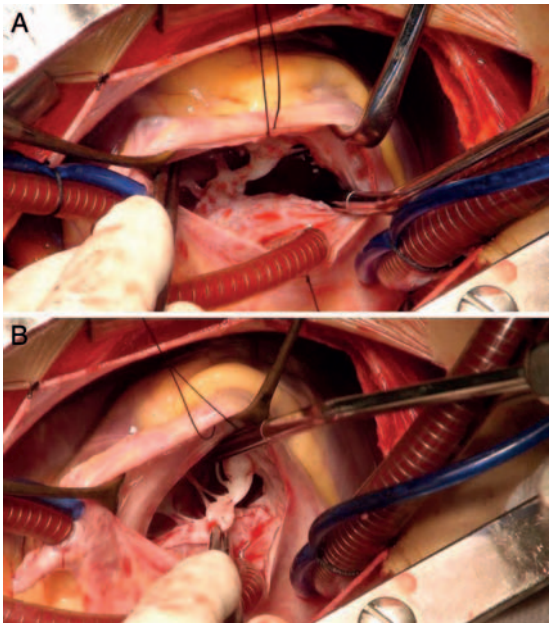

Figure 1: (**A** and **B**) The intraoperative view of a DOTV with equal sized orifices in the side-to-side position and well-formed leaflets with the dividing leaflet having their own chordal apparatus.

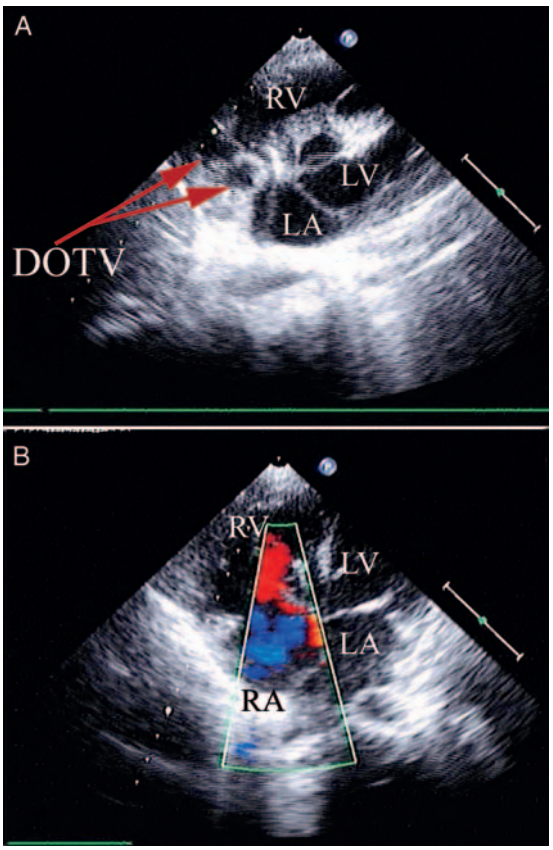

Figure 2: Transthoracic echocardiography views. (**A**) The modified parasternal short-axis view showing DOTV. (**B**) The modified subcostal view with the coronal sweep showing colour flow across the DOTV. DOTV: double-orifice tricuspid valve; RA: right atrium; LA: left atrium; RV: right ventricle; LV: left ventricle.

apparatus or a double-orifice valve. Although this anomaly is well known in the mitral position, duplication of the tricuspid valve is rare [1–7]. In these situations, it is the presence of an accessory subvalvar component that distinguishes true duplication from a simple fenestration of the valvar leaflet [1–7]. The first classification of DOTV was given by Hartmann in 1937 [1–7]. The 'L' type of defect was characterized by two ostia of unequal sizes, the 'B' type of defect had two equal sized ostia without an independent set of chordae and a papillary muscle for each ostia and 'S'-type anomalies had two similar sized ostia and each orifice had an independent set of chordae and a papillary muscle [1]. Cascos drew upon this classification and revised it to describe three types of tricuspid valve duplications [2–7]. The commissural type was similar to Hartmann's 'L' variant, in which the accessory ostia lay within the valve commissure. The central type encompassed Hartmann's 'B' and 'S' variants and was attributed to a fibrous fusion or band that divided the atrioventricular orifice into two. The third variant, or hole-type defect, was described as a hole within the cusp or valve leaflet. As previously mentioned by Hartmann, a subvalvular apparatus was necessary for true duplication. Our case was similar to Hartmann's type 'S' and Cascos's type 2 with equal sized orifices and independent set of chordae and papillary muscle sets. In most cases, these valves have been incompetent or stenotic and have been associated with additional congenital anomalies [1–7]. The occurrence of the DOTV is extremely rare and it is difficult to diagnose by echocardiography [3–5]. The modification of the subcostal long-axis view, apical four-chamber and parasternal views were helpful [3, 5]. The use of magnetic resonance imaging is helpful in diagnosis as well as its functional significance [6]. In our patient, the diagnosis of the DOTV was missed initially and discovered incidentally during surgery. After the operation, the two orifices were easily demonstrated by the cross-sectional echocardiography using subxiphoid sweeps in the coronal plane and modified parasternal short-axis views.

CONCLUSION

The DOTV is a very rare congenital heart defect. The isolated occurrence of this condition seems extremely rare and, in most cases, it is associated with other congenital cardiac malformations that determine patient's outcome. Even though the DOTV is a rare anomaly and can be easily missed if unaware of it, we recommend that it is necessary to scan for an accessory orifice in all patients in whom the atrioventricular valve appears to be small or excessively large.

Conflict of interest: none declared.

REFERENCES

[1] Hartmann B. Zur Lehre der Verdoppelung des linken Atrioventrikularostiums. Arch Kreisl-Forsch 1937;1:286.
[2] Sanchez Carlos A, Rabago P, Sokolowski M. Duplication of the tricuspid valve. Br Heart J 1967;29:943–6.
[3] Tutarel O, Westhoff BM. The double-orifice tricuspid valve: a review. J Heart Valve Dis 2007;16:508–10.

[4] Zhang J, Ma WG, Pan SW. Surgical management of double-orifice tricuspid valve. Journal of Cardiac Surgery 2011;26:425–8.
[5] Miyamura H, Matsukawa T, Maruyama Y, Nakazawa S, Eguchi S. Duplication of the tricuspid valve with Ebstein anomaly. Jpn Circ J 1984; 48:336–8.
[6] Quiroga JLS, Alvarez JR, Siebert MF, Carrewio CI, Duran D, Bengochea JBG. Duplication of the right atrioventricular orifice in a patient with common atrium and mitral cleft. Texas Heart Inst J 1988;15:68–71.
[7] Yoo SJ, Houde C, Moes CA, Perrin DG, Freedom RM, Burrows PE. A case report of double-orifice tricuspid valve. Int J Cardiol 1993;39:85–7.