Cardiol Young 2005; 15: 422–424

ISSN 1047-9511

Brief Report

A very unusual combination of straddling and overriding of the tricuspid valve associated with clefting of the mitral valve

Liane Eberhardt,[1] Philippe Chetaille,[1] Bernard Kreitmann[2]

[1]*Department of Cardiology, Pediatric Cardiology Unit,* [2]*Pediatric Thoracic and Cardiovascular Surgery, Hôpital d'Enfants de la Timone, Marseille, France*

Abstract We describe a patient in whom we found dual orifices in a straddling and overriding tricuspid valve, with two normally sized ventricles and a cleft in the mitral valve. The patient underwent successful surgical repair. We discuss the concept of "double-orifice right atrium", as well as the need to differentiate the isolated cleft of the morphologically mitral valve from the zone of apposition between the left ventricular components of the bridging leaflets seen in the setting of atrioventricular septal defect with common atrioventricular junction. We emphasise the unusual association of these abnormalities of the right and left atrioventricular valves in patients with separate atrioventricular junctions.

Keywords: Double-outlet right atrium; mitral insufficiency; congenital heart disease; endocardial cushions; surgical repair

We describe a patient with straddling and overriding of the tricuspid valve, but in the absence of any significant ventricular septal defect. The patient had two normally sized ventricles, and the morphologically mitral valve was cleft. The anomaly involving the morphologically tricuspid valve has been described as "double-orifice right atrium", but this can be morphologically and embryologically confusing. It is our intention to emphasise the unique elements seen in our patient. We discuss the distinction of clefting of the morphologically mitral valve from the zone of apposition between the left ventricular components of the bridging leaflets as seen in atrioventricular septal defect with common atrioventricular junction, the latter feature often described erroneously as a "cleft". We also discuss the uncommon association of the true cleft of a morphologically mitral valve with the right-sided anomalies encountered in our patient.

Case report

A 15-year-old girl was admitted for surgical treatment of mitral regurgitation. Echocardiography revealed that the regurgitation was due to a cleft in the aortic leaflet of the mitral valve being directed towards the aortic root without any features of atrioventricular septal defect with common atrioventricular junction (Figs 1 and 2). There was no sub-aortic obstruction. The membranous interventricular septum was seen to be long and thin, seemingly, formed mainly by apposition of the septal leaflet of the tricuspid valve to the muscular septum (Fig. 2a). A very small perimembranous ventricular septal defect was present.

Intraoperative examination confirmed clefting of the aortic leaflet of the mitral valve, the cleft components being unsupported by tendinous cords (Fig. 1b). Surgical interrogation also revealed the unusual aspect of the right atrium often described surgically as "double-outlet right atrium". There were two discrete orifices within the morphologically tricuspid valve. The first orifice, located anteriorly, superiorly, and to the right, opened to the morphologically right ventricle, and was made up in essence of a single large antero-superior leaflet, with a small fenestration in the leaflet. The second orifice, positioned postero-inferiorly and to the left, entered the left ventricle. It possessed two leaflets, which were attached to a papillary muscle in the left ventricle, close to the annulus, and posterior to the sub-aortic outflow tract, being unrelated to any of the tendinous cords supporting the leaflets of the mitral valve. The two orifices in the

Correspondence to: Dr Philippe Chetaille, Hôpital d'Enfants de la Timone, 264 rue Saint Pierre, 13385 Marseille Cedex 05, France. Tel: +33 4 91 38 6750; Fax: +33 4 91 38 5638; E-mail: philippe.chetaille@mail.ap-hm.fr

Accepted for publication 26 January 2005

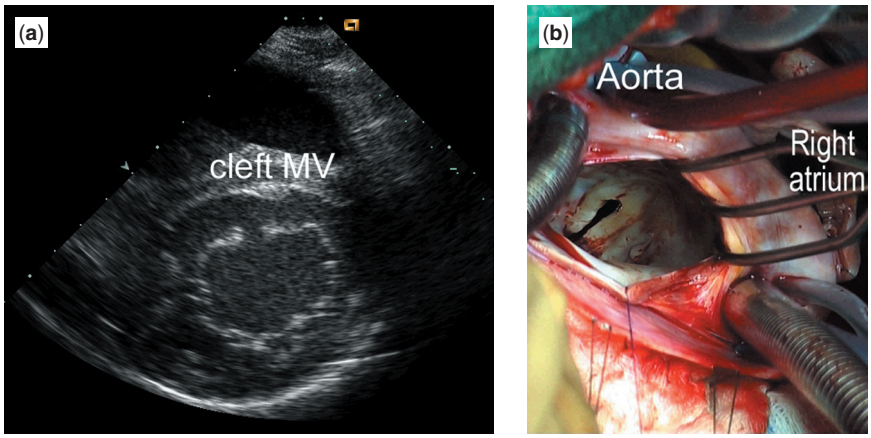

Figure 1.
A transthoracic echocardiographic short axis view of the mitral valve shows a cleft in the aortic leaflet. The surgical view of the valvar orifice confirms the presence of the cleft in the otherwise normal aortic leaflet of the mitral valve (MV). The edges of the cleft are thickened. The direction of the cleft is toward the aortic orifice.

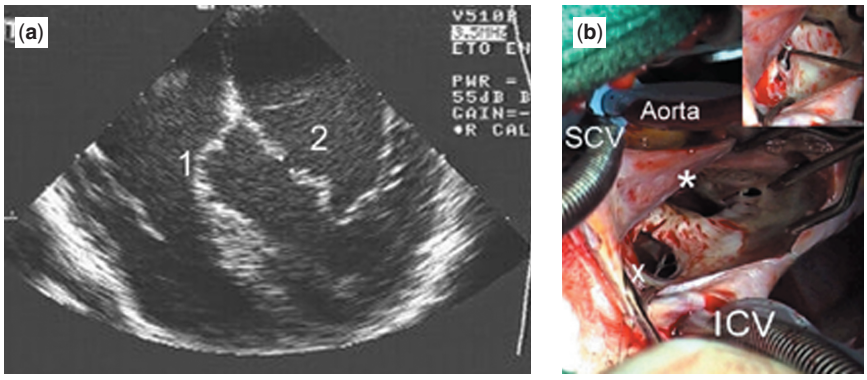

Figure 2.
*A transesophageal echocardiographic view of the tricuspid valve (**a**) shows a long and thin membranous septum formed by the septal leaflet of the valve (1). The cleft in the aortic leaflet of the mitral valve is well seen (2), as well as the malalignment of the atrial and ventricular septums. The surgical view of the right atrioventricular valve (**b**) reveals the presence of dual orifices. The right-sided orifice is close to the asterisk (*), where a small fenestration is seen, the valvar orifice communicating with the right ventricle. The other orifice, close to the "X", communicated with the left ventricle. A magnified view of the tip of the small papillary muscle inserted in the left ventricle is shown in the right upper corner. ICV: inferior caval vein; SCV: superior caval vein.*

tricuspid valve were separated by a tongue of fibrous tissue almost entirely adherent on the muscular crest of the ventricular septum. There was a small ventricular septal defect below the antero-superior part of this tongue (Fig. 2b). Although probably competent, because it connected in the left ventricle, we closed the left orifice of the tricuspid valve as well as the ventricular septal defect. The cleft in the aortic leaflet of the mitral valve was sutured. Recovery was uneventful.

Discussion

The atrioventriculars junctions constitute a complex region of the heart, being the site of many combinations of anomalies. In this respect, the term "double-outlet right atrium", often used to describe a malformation of the right atrioventricular junction, is imprecise, since it does not reflect all the possible combinations. It has been described as a right atrium opening in two different ventricles through a straddling tricuspid valve without any ventricular septal defect, on the basis that simple straddling of the tricuspid valve is always associated with a ventricular septal defect. Should the atrioventricular junctions be discordantly connected, however, double-outlet from the right atrium would be associated with straddling of the morphologically mitral valve.[1] Furthermore, in the setting of a common atrioventricular valve, with malalignment of the atrial and ventricular septums, "double-outlet right atrium" can occur through a common atrioventricular valve.[2] "Double-outlet right atrium" can also coexist in combination with absence of the left atrioventricular

https://doi.org/10.1017/S1047951105000879 Published online by Cambridge University Press

connection, then being one form of the rare uniatrial but biventricular connection.[3] In this setting, straddling and overriding of the morphologically tricuspid valve appears to offer the best way description.

In our patient, the orifice of the morphologically tricuspid valve was split by a tongue of fibrous tissue, thus creating dual orifices within the right atrioventricular junction. Uncommonly, however, each orifice opened to a separate ventricle. Futhermore, the bridge of valvar tissue dividing the orifices was partly attached to the crest of the muscular ventricular septum, incompletely closing an inlet ventricular septal defect. Isomatsu et al.[4] have previously reported a case of straddling tricuspid valve, in which a potential ventricular septal defect was probably closed by a tongue of valvar tissue connected to the crest of the ventricular septum. In our case, a very small ventricular septal defect still existed beneath this fibrous band, albeit that the other aspects of the tricuspid valve were very similar to those described by the Japanese workers.[4]

The essence of straddling of the morphologically tricuspid valve is ventriculo-atrial septal malalignment. In a postmortem study, Pessotto et al.[5] found a much wider ventriculo-atrial septal angle, with a median superior to 60 degrees, in the setting of the lesion they called "double-outlet right atrium" when compared to normal hearts, in which the median angle was 6 degrees. The malaligned ventricular septum is displaced under the normal tricuspid orifice, creating the condition for straddling of the valvar tension apparatus. Such displacement is well seen in our case (Fig. 2b), but with normal ventriculo-atrial angulation.

Another embryological hypothesis to explain "double-outlet right atrium" with hypoplasia of the right ventricle can be incomplete rightward displacement of the developing atrioventricular junctions. Prior to the completion of ventricular septation, the embryonic atrioventricular canal is positioned so as to open exclusively into the developing left ventricle. Normal development includes successive steps leading to a rightward displacement of the right atrioventricular orifice. Straddling of the tricuspid valve may be an intermediate pattern between double-inlet left ventricle and a normal heart.[6] A hypoplastic apical part of the right ventricle is typically found in this setting of failure of normal transfer of the right atrioventricular junction,[5,6] but this feature was not noted in our case.

A so-called "cleft" of the left-sided atrioventricular valve is often considered to be part of atrioventricular septal defect with common atrioventricular junction. This morphological entity, in reality the zone of apposition between the left ventricular components of the leaflets that bridge the ventricular septum in the atrioventricular canal defect, needs to be distinguished from a cleft in an otherwise normal mitral valve. The morphology of these two different features is well described.[7,8] In hearts with a common atrioventricular junction, the so-called "cleft", in reality the space between the bridging leaflets, is directed towards the muscular ventricular septum, and associated with an underdeveloped left mural leaflet of the common valve and papillary muscles rotated in counter-clockwise direction. In the "isolated" cleft of the morphologically mitral valve, the arrangement of the papillary muscles is normal, the mural leaflet of the mitral valve is normally developed, and the cleft is directed towards the aortic root.

The combination of anomalies of the tricuspid valve seen in our patient, specifically straddling and overriding with dual orifices, each orifice opening in a ventricle of normal size with a residual inlet ventricular septal defect, and its association with an "isolated" cleft of the mitral valve, is unusual. It raises many questions, not least about the embryological origin of the valvar leaflets.[9,10] We are anxious to hear of others who may have had similar experiences.

Acknowledgement

The authors thank Dr Joël Errera for patient referral and Dr Alain Fraisse for his review.

References

1. Milo S, Ho SY, Macartney FJ, et al. Straddling and overriding atrioventricular valves: morphology and classification. Am J Cardiol 1979; 44: 1122–1134.
2. Alivizatos P, Anderson RH, Macartney FJ, Zuberbuhler JR, Stark J. Atrioventricular septal defect with balanced ventricles and malaligned atrial septum: double-outlet right atrium. Report of two cases. J Thorac Cardiovasc Surg 1985; 89: 295–297.
3. Ho SY, Milo S, Anderson RH, et al. Straddling atrioventricular valve with absent atrioventricular connection. Report of 10 cases. Br Heart J 1982; 47: 344–352.
4. Isomatsu Y, Kurosawa H, Imai Y. Straddling tricuspid valve without an atrioventricular septal defect. Br Heart J 1989; 62: 222–224.
5. Pessotto R, Padalino M, Rubino M, Kadoba K, Büchler JR, Van Praagh R. Straddling tricuspid valve as a sign of ventriculoatrial malalignment: a morphometric study of 19 postmortem cases. Am Heart J 1999; 138: 1184–1195.
6. Lamers WH, Wessels A, Verbeek FJ, et al. New findings concerning ventricular septation in the human heart. Implications for maldevelopment. Circulation 1992; 86: 1194–1205.
7. Sigfusson G, Ettedgui JA, Silvermann NH, Anderson RH. Is a cleft in the anterior leaflet of an otherwise normal mitral valve an atrioventricular canal malformation? J Am Coll Cardiol 1995; 26: 508–515.
8. Fraisse A, Massih TA, Kreitmann B, et al. Characteristics and management of cleft mitral valve. J Am Coll Cardiol 2003; 42: 1988–1993.
9. Wenink ACG, Gittenberger de Groot AC, Brom AG. Developmental considerations of mitral valve anomalies. Int J Cardiol 1986; 11: 85–101.
10. Lamers WH, Virágh S, Wessels A, Moorman AF, Anderson RH. Formation of the tricuspid valve in the human heart. Circulation 1995; 91: 111–121.