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Brief Report

Duplication of the tension apparatus of the tricuspid valve

Sivadasan R. Anil,[1] Suresh G. Rao,[2] Raman K. Kumar[1]

Department of [1]Pediatric Cardiology and [2]Cardiovascular and Thoracic Surgery, Amrita Institute of Medical Sciences and Research Centre, Kochi, Kerala, India

Abstract We report an infant with a large perimembranous ventricular septal defect, who had two separate orifices in the tricuspid valve, each supported by separate cordal apparatus, detected incidentally during surgery.

Keywords: Double orifice tricuspid valve; valvar duplication

Double orifices in the mitral valve, with duplication of the tension apparatus, is well known to occur in association with a number of congenital cardiac defects. Duplication of the tricuspid valvar apparatus, in contrast, is relatively rare.[1–4] We report an infant with a large perimembranous ventricular septal defect in whom two separate orifices were found in the right atrioventricular valve during surgical closure.

Case report

A 3-month-old male infant, weighing 4 kg, was admitted to our center with tachypnea and difficulties in feeding, which had continued from the end of the first month of life. Clinical examination revealed a tachypneic infant, with saturations of oxygen measured at 90%. Cardiac examination revealed 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 tachycardia, and preexcitation with features of an accessory pathway crossing to the free wall of the right ventricle.

The initial echocardiogram showed usual atrial arrangement, normal systemic and pulmonary venous connections, and patency of the oval foramen, with bi-directional flows. The tricuspid valve and right ventricle appeared somewhat small, the tricuspid annulus being measured at 11 mm diameter, with a Z Score of −0.3. There was a large perimembranous ventricular septal defect, with severe pulmonary arterial hypertension and predominant left-to-right shunting and significant left atrial and left ventricular dilation. The rest of the cardiac anatomy, including the mitral valve, was within normal limits. Cardiac catheterization was done in view of the presence of resting desaturation, and for angiographic assessment of right ventricle. The hemodynamic findings revealed systemic pulmonary arterial pressures, with a large left-to-right shunt and pulmonary vascular resistance of 0.85 Wood units m^2. The right atrial mean, and the right ventricular end diastolic, pressures were both 12 mmHg. The right ventriculogram in two orthogonal views showed a tripartite right ventricle, with a relatively small inlet portion. The decision was made to close the ventricular septal defect, leaving the oval foramen open to decompress the right atrium.

Operative findings

The right atrium was connected to the morphologically right ventricle through two atrioventricular orifices, of 4 and 10 mm diameter, respectively. On probing from the right atrium, the smaller orifice was seen anterior to the coronary sinus, and was inferiorly placed compared to the larger orifice. There were three well-developed leaflets for both the valves, with supporting cordal structures. The ventricular septal defect was adjacent to the larger tricuspid valve orifice, as for usual perimembranous defects. The tripartite right ventricle was of adequate size, so the ventricular septal defect was closed and the oval foramen was left

Correspondence to: R. Krishna Kumar, Consultant Pediatric Cardiologist, Amrita Institute of Medical Sciences and Research Centre, Kochi, 682 026, Kerala, India. Tel: 91 484 339080; Fax: 91 484 340801; E-mail: rkrishnakumar@aimshospital.org

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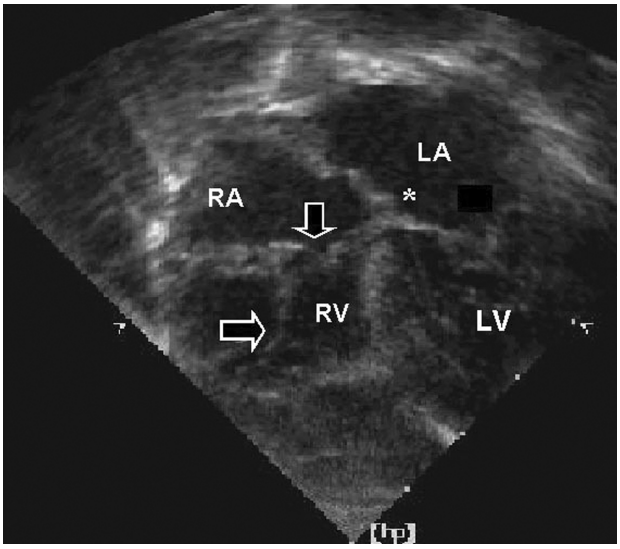

Figure 1.
Sub-xiphpoid view in the coronal plane showing two right atrio-ventricular valves between right atrium and right ventricle. Arrows show the separate orifices between the right atrium and right ventricle. The atrioventricular junction at the crux of the heart is shown by (*). RA: right atrium; LA: left atrium; LV: left ventricle; RV: right ventricle.

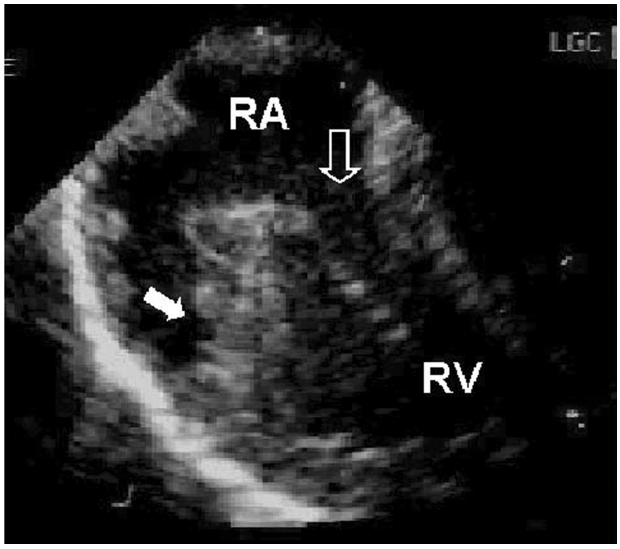

Figure 2.
Trans-esophageal echocardiogram (horizontal plane) done in the early post-operative period showing a pouch in the right atrium (RA) which communicates with the right ventricle (RV) via the smaller orifice. The larger orifice of the tricuspid valve is shown by the arrow.

open. The post-operative central venous pressure varied from 8 to 10 mmHg.

Postoperative echocardiography

Echocardiography was repeated because of the new operative findings. This revealed that the right atrium had two outlets, each guarded by a separate atrioventricular valve. The two valves were oriented in planes perpendicular to each other. The larger orifice was located in the usual position, and its annulus measured 1.1 cm, the leaflets being supported by cordal apparatus and papillary muscles (Figs 1 and 2). The smaller orifice was located inferiorly, lateral and anterior to larger orifice, and was seen at the lower end of a pouch-like extension from the right atrium (Fig. 2). Its leaflets had independent cordal apparatus and papillary muscles (Fig. 1). The child recovered well after surgery, and the postoperative period was uneventful. At follow up three months later, the child was asymptomatic and thriving, with no cardiac failure.

Discussion

The division of an atrioventricular valve into two similar and functioning units is described as duplication of the valvar apparatus, and also as double orifice valves.[4,5] Although this anomaly is well known in the mitral position, duplication of the tricuspid valve is rare.[1,3,4] In these situations, it is the presence of accessory subvalvar components that distinguishes true duplication from simple fenestration of a valvar leaflet.[4,5] In our patient, there was a bar of muscle separating the two valvar orifices from each other (Figs 1 and 2). In addition, well-developed tendinous cords and papillary muscles were supporting both the valvar components.

The diagnosis of duplication of the tricuspid valve was missed initially, being discovered incidentally during surgery. After the operation, the two orifices were easily demonstrable by cross-sectional echocardiography using sub-xiphoid sweeps in the coronal plane. Our experience suggests that it may be necessary to scan for accessory orifices in all patients in whom the atrio-ventricular valve appears to be small. The association of pre-excitation resulting from a possible right free wall accessory pathway in this case is also of interest, particularly since such arrhythmias are known to be associated with aneurysms of the coronary sinus.[6]

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