The quadricuspid pulmonary valve: Its importance in the Ross procedure

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The quadricuspid pulmonary valve is a rare congenital anomaly that can appear as an isolated morphologic variation or in the presence of other cardiac anomalies. Approximately 200 cases of quadricuspid pulmonary valves have been reported to date. Despite the relatively large number of reported cases, this is the first report detailing the anatomy of this malformation.

With the present renaissance of the Ross operation, a great deal of attention has been given to every facet of this procedure. The quadricuspid pulmonary valve is an example of congenital malformations of the pulmonary valve, which are very difficult to diagnose preoperatively. This could have an effect on the planning and outcome of the Ross operation and can seriously affect not only the operating procedure but also the postoperative quality of life. Previously, nearly all cases of quadricuspid pulmonary valve were reported as coincidental postmortem findings, and only one case was diagnosed in a viable newborn child. Because its clinical symptoms are very modest, the diagnosis of a quadricuspid pulmonary valve could be overlooked during clinical investigations.¹

Pulmonary valve lesions are invariably congenital in origin. Because the aortic and pulmonary valves have a common morphogenesis, the quadricuspid pulmonary valve is frequently associated with anomalies of the aortic valve.² It is well known that the Ross procedure has been proved to be a useful intervention in cases of congenital aortic valve malformations when a tricuspid pulmonary valve is used.³ However, the questionable hemodynamics of a quadricuspid pulmonary valve suggest that it is an inadequate candidate for use as an autograft in the Ross procedure. Therefore it is necessary to investigate whether the quadricuspid pulmonary valve could be an acceptable autograft in the Ross procedure.

Clinical Summary

A necropsy was performed on a 23-year-old man who died of a heroin overdose. In the course of the postmortem investigation, the

heart appeared to be normal (350 g), except for the pulmonary valve, which was composed of 4 cusps. The basal ring of the pulmonary valve was slightly dilated, but the pulmonary artery was normal. Three cusps of the quadricuspid pulmonary valve were of equal size and shape. The intercommissural distances were 19 mm. The distances between the sinutubular junction and the deepest point of the sinuses were 18 mm. Although this was a case of a quadricuspid pulmonary valve, only 3 intervalvular triangles were found. The left and right ventral intervalvular triangles were composed of 2 portions. The upper two thirds of the triangles consisted of the wall of the pulmonary artery, and the lower third was composed of the musculature of the right ventricle. The dorsal intervalvular triangle, being positioned between the rudimentary and left cusp, had the same structural nature as ventral two. The fourth cusp was smaller than the other 3 cusps. The intercommissural distance was 0.7 mm, and the height of the sinus was 2.1 mm. The smallest cusp was fenestrated at its free edge and interposed between the left and right cusps. The border between the fourth cusp and the right leaflet was macroscopically indicated by a shallow groove (Figure 1). The rudimentary cusp was not separated from the right leaflet. The intervalvular triangle, which indicated the border between the rudimentary and the right leaflet, was observed to be reduced in size and depth and was only a slight incision marking the border between the 2 cusps.

Discussion

The quadricuspid pulmonary valve is a rare congenital heart anomaly. Comparing it with the bicuspid pulmonary valve, which is frequently associated with serious cardiac malformations, the quadricuspid pulmonary valve is not commonly associated with additional clinical manifestations.

Furthermore, even in combination with other cardiac anomalies, such as patent ductus arteriosus, atrioventricular defect, and aortic valve malformation, the clinical symptoms arising from a quadricuspid pulmonary valve are moderate.^{1,4} Davia and colleagues demonstrated 10 cases in which neonates were examined and given diagnoses of aortic valve anomaly during postmortem investigation. Five of the 10 newborns had quadricuspid pulmonary valve in addition to the aortic valve malformations.

The reported incidence of a quadricuspid pulmonary valve ranges from 1 in 400 to 1 in 1000 autopsies.² Because almost all the cases were reported as coincidental findings and registered as retrospective studies, the actual occurrence of the quadricuspid pulmonary valve is very difficult to estimate. Furthermore, because of the moderate clinical symptoms, the presence of a quadricuspid pulmonary valve might easily escape observation during the postmortem investigation.²

Only one case of a quadricuspid pulmonary valve has been diagnosed in a living patient. In that case the use of 2-dimensional echocardiography was critical in revealing the adequate function

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Figure 1. Quadricuspid pulmonary valve seen after the division of the anterior pulmonary valve: 1, rudimentary leaflet; 2, right pulmonary leaflet; 3, left pulmonary leaflet; 4, intervalvular triangle between the fourth and left leaflet; 5, groove indicating the border between the right and fourth leaflet; 6, sinutubular junction.

of the quadricuspid pulmonary valve, and no morphologic changes were found in the right ventricle.¹ This finding is confirmed by many reports describing the anatomy of the quadricuspid pulmonary valve. A review of the relevant literature reveals no evidence of valvular insufficiency or stenosis being associated with a quadricuspid pulmonary valve. Interestingly, in contrast to an incompetent pulmonary valve, in the case of the quadricuspid pulmonary valve, no right ventricular dilatation or wall hypertrophy has been reported.² Despite the fact that the coaptation of the 4 leaflets is poor, as well as in those cases in which the leaflets are asymmetric, there is no evidence of valvular stenosis or regurgitation.

Carr-White and coworkers⁵ analyzed the long-term results of right ventricular function after the Ross procedure. It was observed that the absence of the pulmonary valve does not result in long-term dysfunction of the right ventricle.

The phenomena that the quadricuspid valve can function as a competent valve without negative consequences seems to be true only in the pulmonary circulation. In contrast with this finding, in cases of a quadricuspid aortic valve, there is a clear occurrence of stenosis or insufficiency.^{4,5}

Aforementioned findings suggest that the use of a quadricuspid pulmonary valve as an autograft in the Ross operation might cause several complications that affect the competence of the graft. This assumption is supported by the fact that the implementation of a tricuspid pulmonary valve in the Ross procedure causes autograft insufficiencies.⁶ In conclusion, the quadricuspid pulmonary valve does not appear to be an adequate autograft for the Ross procedure.

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