Quadricuspid aortic valve in transposition of the great arteries

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Quadricuspid semilunar valves are rare and far less common than bicuspid or even unicuspid valves.¹ The most common anomaly associated with quadricuspid semilunar valve is truncus arteriosus, in which the frequency of the truncal valve being quadricuspid is reported to be around 20% to 25%.² In nontruncus hearts, quadricuspid semilunar valves are extremely uncommon. Quadricuspid semilunar valves are more often pulmonary than aortic valves.³ All reported quadricuspid aortic valves have been found in hearts with ventriculoarterial concordance. To the best of our knowledge, this is the first report of a quadricuspid aortic valve in a heart with ventriculoarterial discordance.

Description of Specimen

In the Leiden collection of heart specimens, we encountered a specimen with a quadricuspid aortic valve in the presence of ventriculoarterial discordance and ventricular septal defect. The anatomy of this heart is as follows:

- Situs solitus of the atria, atrioventricular concordance, and ventriculoarterial discordance
- Balanced ventricles
- Normal tricuspid and mitral valves
- Quadricuspid aortic valve with a small fourth leaflet between the right and left coronary cusps allowing for an X-shaped pattern (Figure 1)
- Unicuspid hypoplastic and stenotic dome-shaped pulmonary valve causing left ventricular outflow tract obstruction
- Great vessels in the anteroposterior position with the aorta anterior and slightly to the left of the pulmonary trunk and continuing with a right aortic arch
- Infundibular nonrestrictive ventricular septal defect

According to the Leiden classification, the coronary pattern can be considered 1 L,Cx-2 R. Apparently, a Blalock-Hanlon procedure had been performed, creating a generous atrial septal defect to allow mixing of pulmonary and systemic venous blood.

Discussion

Semilunar valves with an abnormal number of leaflets may cause clinical symptoms because they may be or may become stenotic or incompetent.^{4,5} Furthermore, abnormal semilunar valve anatomy leads to earlier and accelerated calcific leaflet degeneration. In the era of surgical techniques such as correction of truncus arteriosus or the Ross and the Nikaidoh procedures for complex congenital heart disease (involving translocation of semilunar valves), a correct diagnosis of semilunar valve anatomy is of great importance. Insufficient quadricuspid valves in truncus arteriosus must be repaired, and a quadricuspid pulmonary valve is a contraindication to the Ross procedure. Repair of a quadricuspid valve may involve tailoring or removal of the smaller accessory cusp.

Quadricuspid valves are very uncommon and have been reported to occur 9 times more frequently in the pulmonary than in the aortic position.³ Necropsy series showed an incidence of quadricuspid valves of around 0.003% to 0.008%.⁶ Aortic regurgitation was the most prevalent hemodynamic abnormality (56%) according to the review of Janssens, Klues, and Hanrath¹ with 70 hearts revised from 1923 to 1994.

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Figure 1. Cranial view of the specimen with quadricuspid aortic (Ao) valve and unicuspid pulmonary (P) valve. The asterisk indicates the additional (fourth) leaflet of the aortic valve. The left coronary artery (LCA) originates from sinus 1. The right coronary artery (RCA) originates from sinus 2. LAD, Left anterior descending artery; Cx, circumflex artery; LA, left atrium; RA, right atrium.

The question may arise whether we are dealing with a true quadricuspid aortic valve. Knowledge of the developmental background of valve formation, such as factors that regulate commissure and leaflet formation, is insufficient to answer this question. Moreover, a coronary artery may arise from a pulmonary sinus. On the basis of the embryonic background of the great arteries, the aorta being defined by its connection to the fourth pharyngeal arch arteries and the pulmonary trunk by its connection to the sixth arch arteries, we have to conclude that we are dealing with a true quadricuspid aortic valve.

Although a quadricuspid valve is usually an incidental autopsy or intraoperative finding, it may be diagnosed by means of noninvasive techniques such as 2-dimensional echocardiography or magnetic resonance imaging. Echocardiographic identification is easier in the presence of 4 cusps of equal size (X-shaped semilunar valve pattern on short axis) and is more difficult in cases with a small accessory cusp between the right and noncoronary cusps, which is the most frequent morphologic pattern.⁶ This pattern is more likely to result in valvular insufficiency or endocarditis because of unequal distribution of stress and abnormal leaflet coaptation, according to Feldman and associates.⁵

Only a few reports have been published concerning the association of quadricuspid valves with other cardiac defects. Of these, anomalies of the coronary ostia and coronary arteries are the most frequent. In the review by Janssens, Klues, and Hanrath,¹ only 6 of 70 hearts were associated with abnormalities other than coronary anomalies: 2 with

ventricular septal defect, 1 with persistent ductus arteriosus, 1 with fibromuscular subaortic stenosis, 1 with anterior mitral leaflet anomaly, and 1 with hypertrophic nonobstructive cardiomyopathy.

In conclusion, we want to add one more major congenital defect—transposition of the great arteries—to those associated with quadricuspid valves to emphasize the rarity of this condition. Even more unusual, in this case the quadricuspid aortic valve is associated with a unicuspid pulmonary valve in the setting of polyvalvular disease.

References

1. Janssens U, Klues HG, Hanrath P. Congenital quadricuspid aortic valve anomaly associated with hypertrophic non-obstructive cardiomyopathy: a case report and review of the literature. *Heart*. 1997;78:83-7.
2. Bogers AJJC, Bartelings MM, Bökenkamp R, Stijnen T, van Suylen RJ, Poelmann RE, et al. Common arterial trunk, uncommon coronary arterial anatomy. *J Thorac Cardiovasc Surg*. 1993;106:1133-7.
3. Davia JE, Fenoglio JJ, De Castro CM, McAllister HA, Cheitlin MD. Quadricuspid semilunar valves. *Chest*. 1977;72:186-94.
4. Bortolotti U, Sciotti G, Levantino M, Milano A, Nardi C, Tartarini G. Aortic valve replacement for quadricuspid aortic valve incompetence. *J Heart Valve Dis*. 1998;7:515-7.
5. Feldman BJ, Khanderia BK, Warnes CA, Seward JB, Taylor CL, Tajik AJ. Incidence, description and functional assessment of isolated quadricuspid aortic valves. *Am J Cardiol*. 1990;65:937-8.
6. Hurwitz LE, Roberts WC. Quadricuspid semilunar valve. *Am J Cardiol*. 1973;31:623-6.