cusps, a triangular resection followed by commisuroplasty was chosen. One patient had a conjoint large cusp, and the remaining 2 cusps were very small, so a plication of the conjoint cusp was performed; the commissure between the remaining cusps was closed, resulting in bicuspidization of the QAV. Misfield and colleagues⁹ described a case using the subcoronary Ross procedure, in 1 patient who had QAV and severe regurgitation.⁹ Although this technique was effective for treating this pathology, 42% of our patients had some evidence of aortopathy, which can predispose them to late failure.

In summary, QAV dysfunction includes both regurgitation and stenosis. Repair may be feasible in some patients with regurgitation, but most require valve replacement. Aortic root and ascending dilatation is frequent, and further studies of this population may clarify the etiology of congenital aortic valve disease.

Conflict of Interest Statement

Authors have nothing to disclose with regard to commercial support.

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EDITORIAL COMMENTARY

Quadricuspid aortic valve repair: Pushing the limits!

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See related article on pages 79-82.

Quadricuspid aortic valve (QAV) is a rare congenital abnormality that often leads to hemodynamically significant

valvular disease, typically in the fourth, fifth, or sixth decade of life. A number of associated cardiac abnormalities have been described, including proximal aortic dilatation, coronary artery anomalies, and ventricular and atrial septal defects. QAVs can have a variety of different configurations, which were originally classified by Hurwitz and Roberts¹ in 1973, largely on the basis of the sizes of the individual cusps. Interestingly, in their study of 11 autopsy specimens, only 5 valves were functionally abnormal, suggesting that many patients with this abnormality may never have development of significant valve disease and are seen for medical attention. Although the most common surgical

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treatment for this disease has been aortic valve replacement, a number of techniques to repair the valve have been described and successfully applied in selected cases.

The majority of available information on QAV is based on case reports or small case series numbering less than 10 patients. In contrast, Idrees and colleagues² from the Cleveland Clinic report in this issue of the *Journal* on a series of 31 patients with QAV undergoing surgical intervention. Analysis of this large experience provides some interesting insights into this rare disease. The authors observed a high incidence of associated aortic dilatation in 40% of patients, and 23% underwent associated aortic resection. Although aortic dissection and rupture have not been frequently reported with QAV, the high incidence of aortopathy suggests a pathophysiologic association, related to hemodynamic or molecular mechanisms. The incidence of coronary abnormalities was also high at 10%; however, only 1 patient underwent repair. Both these findings have implications for the workup and surgical treatment of patients with QAV.

Consistent with existing literature, most of the patients in this series (68%) were seen with valvular insufficiency as the predominant hemodynamic lesion, which opens the door to valve repair. Extension of established aortic valve repair techniques to this unusual anatomy requires a thorough understanding of aortic insufficiency mechanisms³ and further reinforces the general principles of aortic valve repair.⁴ The 2 important mechanisms related to cusp anatomy previously reported and also observed in this series are (1) cusp restriction (type 3) as a result of incomplete fusion of 2 small accessory cusps and (2) cusp prolapse (type 2). Cusp restriction can be treated by detachment of the pseudocommissure between the small accessory cusps (similar to the fused cups of a bicuspid aortic valve). After resection of the thick, fibrotic portion, the cusp edges can be reapproximated, rendering the valve tricuspid. A small pericardial patch may be required in some cases when cusp tissue is inadequate. Cusp prolapse can be treated with partial resection and plication of the free margin as described.

In addition to the abnormal cusp anatomy, it is important to identify and treat the changes in annular geometry that contribute to aortic insufficiency (type 1). Ascending aortic aneurysms frequently cause sinotubular junction dilatation (type 1a), which can be treated with downsizing and remodeling of the sinotubular junction, and chronic aortic

insufficiency is frequently associated with dilatation of the ventriculoaortic junction (type 1c), typically treated with subcommissural annuloplasty. It is important to address all lesions to achieve a durable repair, and in this series all repaired valves underwent some form of annuloplasty in addition to cusp repair.

Repair of QAVs, like that of any other valve, may ultimately be limited by the quality of cusp tissue available, which may be the reason that only a third of QAVs with pure aortic insufficiency were repaired in this series. Although an understanding and rigorous application of aortic valve repair principles may lead to a successful intraoperative outcome, there are few data on the progression of disease and long-term durability of QAV repair. Of the patients undergoing repair in this series, 1 required aortic valve replacement at 13 years and 1 each had development of moderate aortic stenosis and regurgitation. Repair should therefore be undertaken only in selected cases of patients with suitable anatomy who really stand to benefit from the avoidance of a prosthetic valve. Other small series of repaired QAVs in the literature, although initially successful, also report follow-up of less than 5 years.^{5,6}

As aortic valve repair matures as a discipline, these general principles will continue to be applied to increasingly challenging anatomy. The Cleveland Clinic group are to be congratulated for analyzing data on a large series of patients with QAV that provides further insights into this rare abnormality and for their successful application of aortic valve repair techniques in carefully selected cases.

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