

Quadricuspid Aortic Valve: A Chance for Repair

Nuran Yener¹, Adem Resatoglu², EL Amin E. Elnur³, Ali Yener³

¹Hacettepe University, Department of Anatomy, ²Cankaya Hospital, Department of Cardiovascular Surgery, ³Gazi University, Gazi Hospital, Ankara, Turkey

Quadricuspid aortic valve is an uncommon congenital anomaly that often causes aortic regurgitation in adulthood, necessitating valve replacement. A 54-year-old woman presented with symptoms and signs of aortic regurgitation, but during surgery a quadricuspid aortic valve was identified. The abnormal

Quadricuspid aortic valve (QAV) is a rare congenital anomaly that has a reported incidence of between 0.008% and 1% (1-4). Although the QAV is a congenital anomaly, aortic regurgitation does not occur until adulthood. Valvular regurgitation usually develops due to fibrous thickening with incomplete coaptation, as a result of the unequal distribution of stress and abnormal leaflet movements (5).

The management of QAV is generally valve replacement (6). Here, a case of QAV is described, together with a modified method of repair.

Case report

A 54-year-old woman who presented with shortness of breath was referred to the authors' clinic for further assessment. On examination, she was in NYHA functional class II, and auscultation revealed a 2/6 diastolic murmur at the aortic area. Transthoracic echocardiography showed second-degree aortic regurgitation, but did not visualize the number of valve leaflets present. Angiography confirmed the presence of regurgitation and ruled out any coronary anomaly; the patients' left ventricular function was only mildly impaired.

The patient underwent elective open-heart surgery, during which it was noted that four aortic cusps were present (Fig. 1). Three of the cusps were of equal size, while the fourth was smaller and fenestrated, and

cusps were fibrotic, short, thick, and fenestrated. Excision of the fourth cusp was performed and valve repair carried out successfully, with only minimal regurgitation.

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located between the right coronary cusp and the non-coronary cusp (Fig. 1). A modified resection of the fourth cusp was carried out (Fig. 2), after which a repair with approximation of the right and non-coronary cusps was performed (Fig. 3). The postoperative period was uneventful, and early and late postoperative echocardiographies at follow up showed a well-functioning valve, with minimal regurgitation.

Discussion

Congenital QAV is a very uncommon lesion which causes significant aortic regurgitation, and often leads

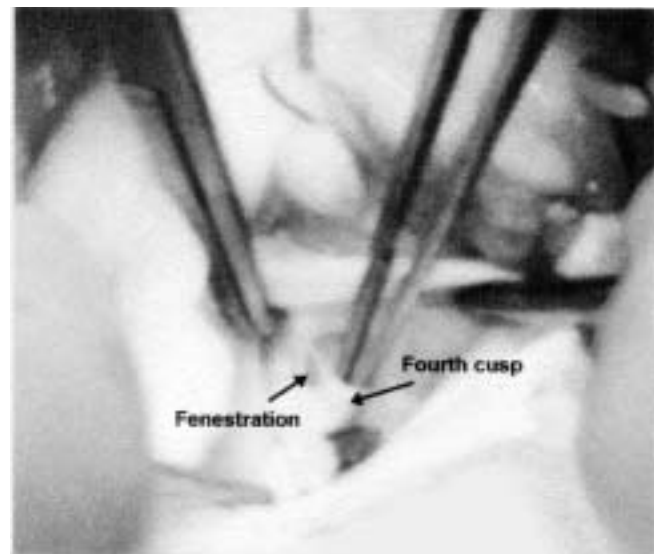


Figure 1: A cranial view of the aortic valve, showing the fourth cusp with a fenestration.

Address for correspondence:
EL Amin E. Elnur, Gazi University, Gazi Hospital, Department of Cardiovascular Surgery -KVC, Besevler, Ankara, Turkey
e-mail: aminnur1@yahoo.com

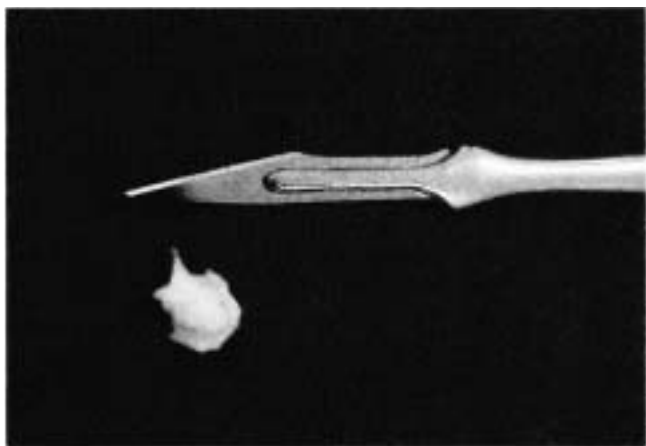


Figure 2: The resected leaflet, which was fibrotic and thickened.

to aortic valve replacement during the fifth and sixth decades of life (5). Seven anatomic variations of the quadricuspid semilunar valves have been described by Hurwitz and Roberts (7), depending on the size of the cusp. In the present patient, there were three equal cusps, while the fourth, smaller, cusp was of the type B classification detailed by Hurwitz and Roberts (7), and seen to be fibrotic and fenestrated. Fenestration is not considered by some to be a congenital abnormality, as its frequency increases with advancing age (5). It is possible that in the present patient the fenestration was so small in early life that its increase in size with age, combined with the turbulent flow through it, con-

tributed to the development of fibrous thickening of the affected cusp. The degree of risk for endocarditis is not clear, but it may be lower with the type A variation (four equal cusps) (3). Although the present patient was type B, endocarditis was not identified at any time. In addition, there were no coronary artery or other valvular anomalies identified, it having been reported that anomalies of coronary artery origin and distribution represents less than 1% of all heart lesions (8).

When managing these patients, the type of abnormality present affects to some extent the choice of procedure used. In general, valve replacement is the treatment of choice, with an average reported age for valve replacement of 54 years. However, the youngest patient who underwent replacement for quadricuspid valve and significant regurgitation was aged only five years (6). In the present patient, the other aortic cusps appeared normal, with no other abnormalities. Hence, excision of the fourth cusp was carried out (Fig. 3), and the right and non-coronary cusps were approximated with a two 4-0 Prolene sutures running from outside the aortic wall through the commissure to the opposite side. Support was provided with a Teflon pledget, and the repair was completed successfully, without any complications.

In conclusion, the repair of a QAV in a selected type of patient can be successfully carried out, notably in those patients who are young or who may develop complications due to long-term anticoagulant use associated with prosthetic valves.

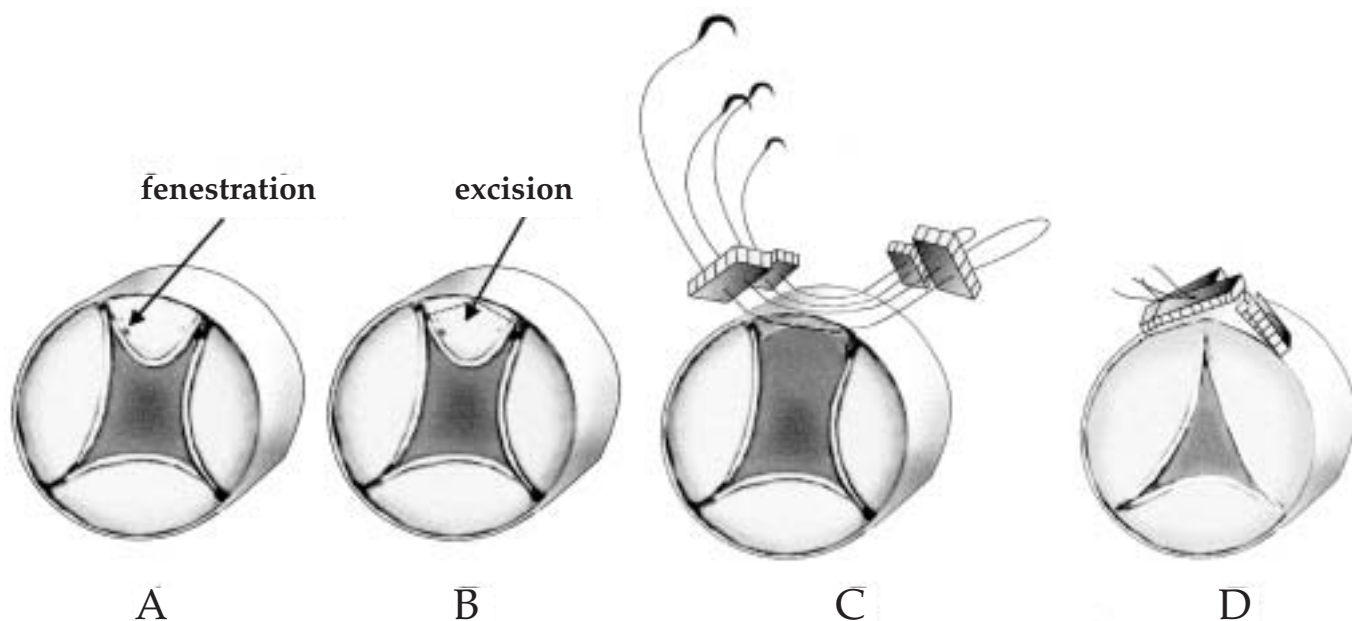


Figure 3: Schematic illustration of the repair procedure. A) Identification of the quadricuspid valve. B) Excision of the fourth cusp. C) Two sutures supported with a Teflon pledget were inserted from outside the aorta through the commissure and aortic wall. D) The suture was tightened against an opposite pledget and the right and non-coronary cusps approximated.

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