

Tricuspidation of Quadricuspid Aortic Valve: Case Reports

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Quadricuspid aortic valve in adulthood is a rare pathology which often leads to aortic valve regurgitation that requires surgical treatment. Herein are described two patients with severe regurgitation on a quadricuspid aortic valve and with dilated left ventricle, who were successfully repaired using a technique of tricuspidation of the valve at the level of the abnormal commissure. In each patient, the repair was

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Case reports

Two male patients, aged 45 and 53 years, were referred, within a six-month period, to the authors' center for asymptomatic aortic insufficiency discovered during a routine medical evaluation. Preoperative cardiac echocardiography showed a dilated left ventricle (end-diastolic dimensions 70 and 56 mm, respectively) with preserved left ventricular function (shortening fractions 32% and 29%). In both patients, a quadricuspid aortic valve was seen with severe aortic regurgitation. No significant findings were identified during the remainder of the evaluation.

The patients were transferred to the operating room to undergo aortic valve repair. Intraoperative transesophageal echocardiography showed good mobility of the four leaflets, with no signs of leaflet prolapse or

stabilized and leaflet coaptation increased by sub-commissural annuloplasty stitches at the level of the three commissures. After one year and six months' follow up, respectively, both patients presented with trivial aortic regurgitation and good mobility of the three leaflets.

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calcification (Fig. 1). In both patients, the mechanisms of regurgitation included a central jet caused by a lack of central coaptation associated with a commissural jet from the extra and abnormal commissure. In one patient, the ascending aorta was slightly dilated, but with preservation of the sinotubular junction (STJ).

In both patients surgery was performed via a sternotomy, under normothermic cardiopulmonary bypass (CPB) and antegrade warm blood cardioplegia. A transverse aortotomy was made at the level of the STJ in order to evaluate possible valve repair. Virtually the same configuration of quadricuspid valve was seen in both cases. The aortic valve comprised two larger leaflets of equal size (corresponding to the right and non-coronary sinuses in the first patient, and to the left and right coronary sinuses in the second patient), and of two smaller leaflets (corresponding to the left sinus in one patient and to the non-coronary sinus in the



Figure 1: Peroperative transesophageal echocardiography, showing a quadricuspid aortic valve with good leaflet mobility.

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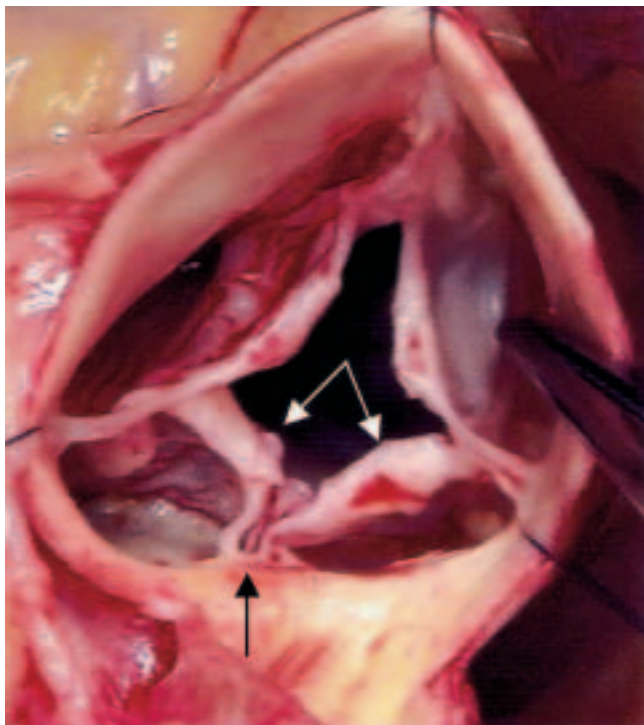


Figure 2: The two smaller leaflets (white arrows), with their incomplete commissure (black arrow), are visible on the posterior side.

other). The three commissures of the larger leaflets appeared normal; the fourth commissure between the smaller leaflets was either incomplete or thickened (Fig. 2). One patient presented with hypertrophy of the nodules of Arantius.

In both patients, the repair involved detachment of

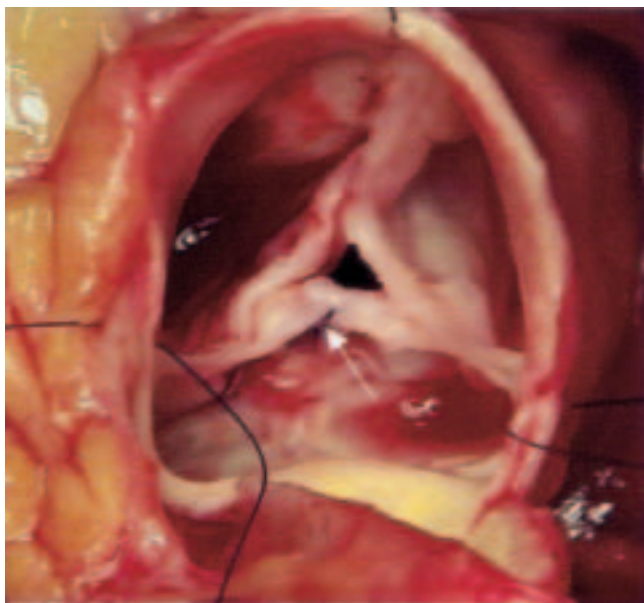


Figure 3: The incomplete commissure was resected, and the remnant adjacent leaflets were primarily sutured (arrow).

the abnormal commissure (between the smaller leaflets) from the aortic wall, with small resection of the thickened commissural tissues; the area of resection was closed with a running suture of Prolene 7-0, in a locking fashion. Additional simple Prolene 7-0 sutures were used to reinforce the line of suture. The end result was a tricuspid valve with a good coaptation (Fig. 3). In order to stabilize the repair and to increase leaflet coaptation, a subcommissural annuloplasty with sutures of Ethibond 2-0 reinforced with Teflon felt on both sides was performed on the three commissures. In the patient with a slightly dilated STJ, in order to increase leaflet mobility and stabilize the junction, a plication of the aortic wall was performed at this level with three sutures of Prolene 4-0 reinforced with a pericardial patch. In the patient with hypertrophy of the nodules of Arantius, the nodules were shaved to ensure a better central coaptation. The aorta was closed with a running suture of Prolene 4-0.

Both patients were weaned from CPB without difficulty, and post-bypass echocardiography demonstrated only trivial central regurgitation. The postoperative course was uneventful for both patients, who were discharged on postoperative days 6 and 8, respectively. At their 12- and six-month follow ups, both patients had recovered fully, with their most recent echocardiogram showing trivial aortic regurgitation and good mobility of all leaflets.

Discussion

Congenital quadricuspid aortic valves are rare, with reported incidences in two autopsy studies being reported as 0.008% (two in 25,666) (1) and 0.03% (two in 6,000) (2). Abnormal cusp formation results from either aberrant fusion of the aorticopulmonary septum, or from abnormal mesenchymal proliferation in the common trunk (1,2). Although congenital quadricuspid aortic valve is usually an isolated lesion, several concomitant congenital lesions have been described, especially coronary artery anomaly, as septation of the arterial trunk and development of the aortic valve leaflets occur just after development of the coronary artery origin from the sinus of Valsalva; hence, developmental errors might result in abnormalities of both structures. Hurwitz and Roberts (2) classified the quadricuspid valve as being of seven types, based on the relative size of the four cusps. The most common type consists of three equally sized cusps and one smaller cusp (type B).

Unlike quadricuspid pulmonary valve, the quadricuspid aortic valve often has an abnormal function, mainly regurgitation and much less frequently stenosis (3). Valvular regurgitation usually develops as a result of fibrous thickening with incomplete coaptation. With

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unequal distribution of stress and abnormal leaflet coaptation, aortic regurgitation may occur later in life, even if the valve was competent during childhood.

In most cases, the type of surgery performed for this pathology is aortic valve replacement, with the type of replacement depending upon the surgeon's experience (4,5). However, very few repairs of this type of valve have been reported (6).

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