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Case report - Congenital

Repair of quadricuspid aortic valve by bicuspidization: a novel technique

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Abstract

Quadricuspid aortic valve (QAV) is a rare congenital lesion, generally manifesting with valve regurgitation. Standard treatment involves valve replacement, though anecdotal cases of successful repair by means of valve tricuspidization have been reported. Here, the successful application of a repair technique previously unreported in the setting of QAV is described.
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Keywords: Quadricuspid aortic valve; Bicuspid aortic valve; Aortic valve repair; Bicuspidization; Tricuspidization; Aortic insufficiency; Valve repair

1. Introduction

Quadricuspid aortic valve (QAV) is a congenital anomaly rarely seen in clinical practice with incidence rates ranging from 0.008% to 0.04% [1, 2]. Most cases have been discovered as incidental finding at surgery or autopsy. Valve insufficiency is the most frequent abnormality in patients with QAV. The management of regurgitant QAV commonly involves valve replacement. More recently, a few cases of valve repair have been reported [3–5]: in all these cases, tricuspidization of the aortic valve was used to restore competence. Here, we report is a case of QAV, which was repaired using an alternative technique.

2. Case report

A 68-year-old-man was referred to our center for severe aortic incompetence identified during his admission for heart failure at a nearby hospital. He complained of severe dyspnea and presented hypertension and diabetes as comorbidities. The chest film showed moderate cardiomegaly. Surface EKG demonstrated regular signs rhythm with signs of left heart hypertrophy. Trans-thoracic echocardiographic examination showed severe aortic regurgitation with undefined valve morphology, left ventricular dilatation with moderate systolic dysfunction (ejection fraction = 42%). The results were confirmed at angiography, which also showed mild ascending aortic ectasia (42 mm), while coronary lesions were excluded. Trans-esophageal echocardiography performed during surgery, revealed QAV anatomy (Fig. 1a) with severe regurgitation, due to a larger central jet caused by lack of coaptation and multiple finer paracommissural jets. Via median sternotomy, under normothermic cardiopulmonary bypass, using retrograde and selective antegrade blood cardioplegia, the aortic valve was inspected through a transverse aortotomy. The valve comprised of four unequal sized cusps: a larger-sized left coronary, a medium-sized right coronary and two smaller non-coronary leaflets, classified as type G according to Hurwitz and Roberts [2] (Fig. 2a,b). The latter two, in addition, were proplapsing and large fenestrations were present at the commissural level in all leaflets. Repair was performed by detachment of the inter-non-coronary commissure and plication of the new non-coronary leaflet using a locking 5-0 polypropylene suture (Fig. 2c). Due to persistence of prolapse and the presence of large fenestrations at the left-to-non-coronary commissure, the latter commissure was demolished (Fig. 2d) and the non-coronary leaflet was then anchored to the left coronary leaflet, thereby turning the valve from quadricuspid to bicuspid (Fig. 2e,f). Furthermore, all remaining fenestration were closed with running 7-0 polypropylene suture. Finally, sub-commissural anuloplasty with interrupted pledgeted 4-0 polypropylene sutures was carried out to allow insertion of a size 20 Hegar dilator. Post-repair trans-esophageal echocardiography showed trivial residual central regurgitation and minimal stenosis (mean pressure gradient = 16 mmHg), with normal orifice area ($=3.80\ cm^2$ or $1.73\ cm^2/m^2$) (Fig. 1b,c). Patient recovery was uneventful with discharge on postoperative day 5. Discharge trans-thoracic echocardiographic examination confirmed intraoperative findings. At 18-month follow-up, the patient is symptom-free and trans-thoracic echocardiographic examination showed mild aortic regurgitation, with effective height of coaptation of 11 mm (Fig. 1d), preserved leaflet mobility and improved left ventricular function (ejection fraction = 61%).

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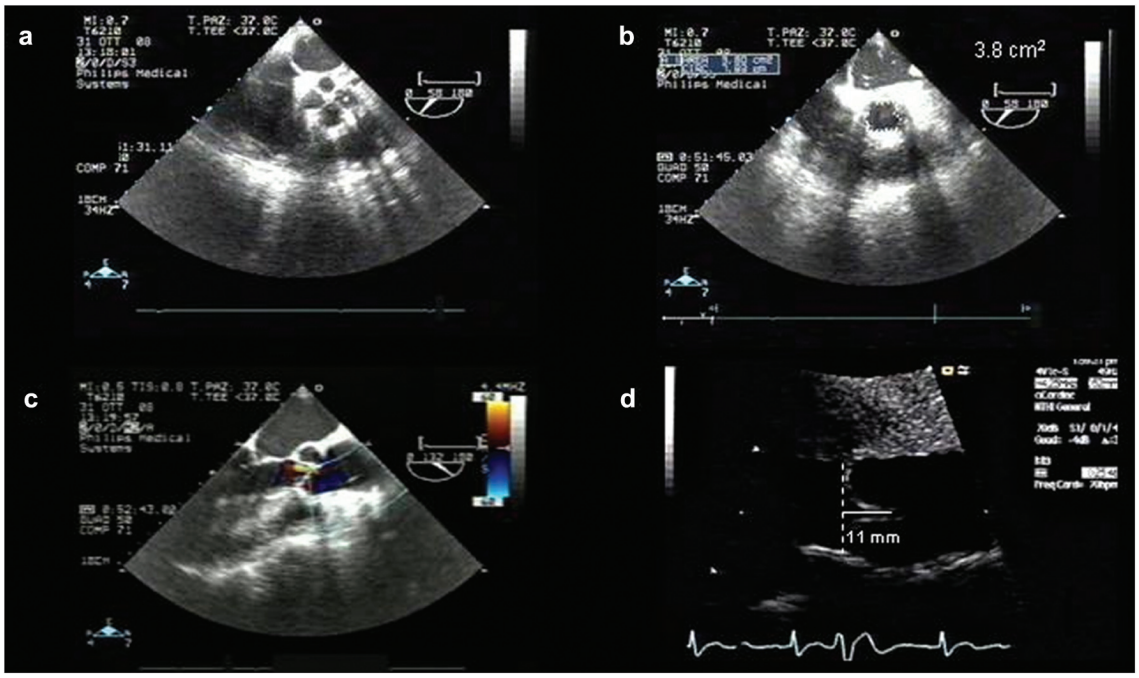

Fig. 1. (a) Pre-repair intraoperative trans-esophageal short-axis view of the aortic valve during diastole. Four unequal size cusps are present; (b) post-repair intraoperative trans-esophageal short-axis view of the aortic valve during systole showing bicuspid morphology and normal aortic orifice area (measured 3.80 cm²); (c) post-repair intraoperative trans-esophageal long-axis diastolic view showing trivial central jet regurgitation at Doppler interrogation; (d) follow-up trans-thoracic para-sternal long-axis view during diastole documenting effective height of leaflet coaptation (measured 11 m).

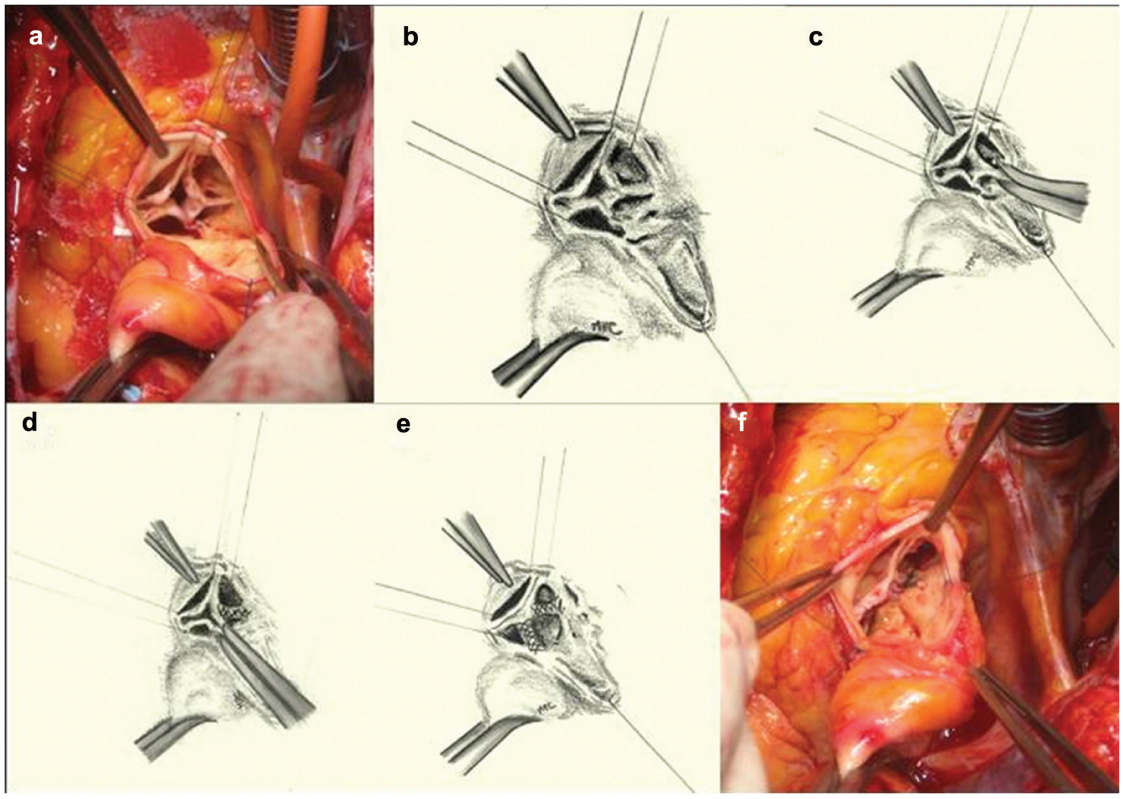

Fig. 2. Operative anatomy and surgical steps. (a) Quadricuspid aortic valve is present, with four unequal-sized leaflets (type G): the two smaller non-coronary leaflets also present multiple large fenestrations; (b) schematic of operative anatomy; (c) schematic of operative steps: detachment of inter-non-coronary commissure; (d) after non-coronary leaflet reconstruction, detachment of left-non-coronary commissure; (e) completed repair: the right coronary leaflet has been preserved, while the non-coronary leaflets have been joined and then anchored to the left coronary leaflet. The former now represents the smaller anterior leaflet, the latter the larger posterior leaflet; (f) operative view of completed bicuspidization.

3. Discussion

Seven anatomical variants of QAV have been described by Hurwitz and Roberts [2], based on the relative size of the four cusps. Over 85% of the reported cases belong to type A, B, or C. Most patients with this anomaly have required aortic valve surgery, which has traditionally consisted of prosthetic valve replacement. Valve repair has seldom been reported, mostly applied to type A, B or C [3–5] and has thus far entailed tricuspidization. Tricuspidization has originally been proposed for the repair of regurgitant tricuspid as well as bicuspid aortic valves in children and young adults [6]. However, when compared to bicuspidization or preservation of native bicuspid valve, tricuspidization has been found to be a risk factor for late repair failure [6]. On the contrary, bicuspidization has been applied successfully to tricuspid, unicuspid and bicuspid aortic valve anatomy [6, 7]. Similar to the high success rate observed with mitral valve repair, the experience of Schäfers and colleagues suggests that valve competence and stability are better achieved in bicuspid rather than tricuspid valves due to the presence of fewer coaptation lines [6, 7]. The case reported here supports this hypothesis by showing that bicuspidization may successfully be applied even to far more complex and rare morphologies, such as type G QAV, where the presence of four unequal cusps makes tricupidization geometrically difficult to apply. In agreement with prior clinical evidence on bicuspidization of unicusp aortic valves [7], this technique allows restoration of valve competence without significant residual trans-valvar pressure gradients, as is confirmed by mid-term follow-up in the present case. Although long-term observation after repair of QAV, particularly for the more uncommon variants, is unavailable, incidental findings of normally functioning QAV in the elderly would suggest this aortic valve anatomy is not necessarily pathological. Identical considerations apply for native (and, possibly, surgically created) bicuspid aortic valves. Therefore, theoretical grounds exist for a durable valve function after bicuspidization of QAV.

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