Successful Repair of a Bicuspid Aortic Valve With Anomalous Chordal Attachment to the Aortic Wall

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The congenitally bicuspid aortic valve (BAV) occurs in an estimated 1% to 2% of the population [1]. Although aortic stenosis is the most frequent complication of BAV [2], pure aortic insufficiency develops in some patients in the absence of valve infection or other clear etiology. In a recently published article, Vowels and colleagues [3] called attention to a rare form of the BAV phenotype consisting of anomalous chordal attachment of the fused cusp to the aortic wall, leading to pure aortic insufficiency. The authors reported their experience with 5 patients who underwent aortic valve replacement (AVR) for this anomaly and provided a complete review of the literature on the subject. A total of 38 cases of this rare and underappreciated variant of BAV have been described, all from pathology reports or patients undergoing AVR. We have recently encountered a similar case. The aim of this report is to describe the technique for surgical repair of the BAV in this context. To our knowledge, this is the first report to describe the successful surgical repair of this rare variant of bicuspid aortic valve.

A 43-year-old male patient with no previous medical history presented with severe BAV insufficiency and left ventricular dilatation. A preoperative echocardiographic evaluation failed to identify the chordal anomaly, and the valve was thought to be tricuspid with prolapse of the right coronary cusp and an eccentric jet directed toward the mitral valve (Fig 1A).

An intraoperative evaluation revealed a BAV with fusion of the noncoronary and right coronary leaflets and an anomalous cord extending from the free margin of the fused cusp, vis-a-vis the raphe, to the ascending aortic wall cephalad to the sinotubular junction. This caused restricted movement of the fused cusp and asymmetrical coaptation height. In addition, the raphe was split, adding an additional cause of aortic insufficiency (Fig 2A).

Surgical repair of the valve was successful. The anomalous cord was fully resected. The raphe was shaved and reconstructed using interrupted 5-0 Prolene (Ethicon, Somerville, NJ) sutures, thus recreating an intact fused cusp. Resecting the anomalous cord induces a significant prolapse of the fused cusp. This is corrected using several...
interrupted central plication sutures to attain adequate cusp effective height (8 to 9 mm) and to ensure symmetry of the length of the free margins of both cusps [5]. In addition, 2 central plication sutures were placed on the nonfused cusp (Fig 2B). Postoperative echocardiography showed an excellent hemodynamic result, with no residual aortic insufficiency and no gradient (Fig 1B).

Comment

BAV is a common finding in pure aortic insufficiency in the absence of connective tissue disease, especially in younger patients [2]. The choice of a valve substitute is difficult in this population because biologic prostheses have limited durability, whereas mechanical valves carry the risk of thromboembolic complications and anticoagulation-related hemorrhage [4]. The Ross procedure is our favored approach for valve replacement; however, preservation of the native aortic valve remains our primary intention in these patients whenever feasible. Indeed, Schafers and colleagues [5] demonstrated excellent results of BAV reconstruction in durability, quality of life, and freedom from valve-related complications in the absence of associated aortic pathology.

The BAV containing an anomalous cord attaching the raphe of the conjoined cusp to the wall of the ascending aorta is a rare and unappreciated cause of aortic insufficiency. This report highlights the importance of recognizing this pathologic entity, as described by Vowels and colleagues [3], and emphasizes the possibility and surgical technique to restore adequate valve function using a tailored approach.

References