Caution: There is no “all or none” with Ebstein anomaly

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Hetzer and colleagues¹ describe their repertoire of repairs for Ebstein anomaly (EA) that they individualized depending on the specific anatomy in a series of 68 patients. They conclude that all morphologic types of EA are amenable to repair (no tricuspid replacements at initial operation) and that satisfactory long-term ventricular function and functional outcome can be obtained, even in the most severe cases.

The authors have a track record with the treatment of EA and are to be commended for the excellent results reported here. Strengths include solid intermediate-term follow-up (mean, 13 years), pre- and postoperative exercise testing that demonstrated significant improvement in maximal oxygen uptake, low early mortality (3%), and excellent freedom from reoperation and late survival at 20 years (93% and 91%, respectively). Weaknesses include no detailed late echocardiographic data, a lack of magnetic resonance imaging to complement echo data on ventricular size/function, and a belief that all patients can be categorized into the Carpentier classification system for EA.²

The authors acknowledge that EA morphology is “highly variable,” and indicate that previous reports fall short because they lack correlation between repair techniques and the Carpentier classification system. The fact is that the anatomic abnormalities encountered with EA are truly infinite and no 2 hearts are alike. Consequently, any classification system is completely flawed. The Carpentier classification system focuses on right ventricular atrialization... making it too simple, and our classification system³ that also factors in valve anatomy... makes it too complex. It is true that there are common features in EA—failure of leaflet delamination, apical displacement of leaflet(s), and some degree of right ventricular atrialization. But the infinite anatomic variability makes the creation of any classification system challenging. In our practice, we simply delineate the anatomic features in each individual case.

Although the authors are to be commended for their efforts at repairing every valve, the issue is whether this

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Central Message

Infinite anatomic variability in Ebstein anomaly precludes valve repair in all cases and makes classification systems flawed.

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strategy is appropriate for a surgeon who encounters this anomaly once or twice a year—the average frequency noted in the Society of Thoracic Surgeons database. The learning curve for EA repairs is steep and reproducible results are more predictable in centers with a larger experience. In general, the operation requires aortic occlusion and cardioplegic arrest for precise suture placement, avoidance of coronary or conduction tissue injury, and minimizing motion trauma during suture securement. The time required to perform EA repair can be lengthy, even in experienced hands, and the consequences of prolonged operation in a patient with depressed ventricular function may not be prudent.

The authors depict multiple techniques that deserve comment, particularly for surgeons who are interested in incorporating them into their armamentarium. First, significant annular reduction is often necessary. The authors nicely illustrate the use of pledgets; this reinforcement is important (particularly in older patients) because significant tension at the annulus is often present. Importantly, the right coronary artery is running in the atrioventricular groove and is vulnerable to kinking or occlusion with this maneuver, so external inspection of the heart and atrioventricular groove for coronary distortion with suture adjustment as needed is essential. The Sebens stitch is also applied liberally. In our experience, the anterior free wall papillary muscle should be mobilized (but maintaining its base intact) so

that it can be approximated to the ventricular septum without tension once the right ventricle is filled. When this is done properly, there should be no dimple in the right ventricular free wall after separation from bypass. When excessive tension is inevitable despite papillary muscle mobilization, patch augmentation of the anterior leaflet is an option to reduce tension and minimize dehiscence. When plication of the atrialized right ventricle is desired, it is important to confine it to the smooth, nontrabeculated inferior wall and avoid the suture line getting close to the ventricular septum because the posterior descending coronary artery is in close proximity. Again, careful inspection of the external surface of the heart is essential after all internal suture placements to ensure no inadvertent coronary injury. Finally, the authors describe their double-orifice technique that involves tissue approximation at the annulus level, in contrast to the more common double-orifice technique (described for the mitral) that focuses on approximation at the leaflet's leading edge inside the ventricle. Whereas the authors were able to obtain satisfactory results with their techniques, we suggest caution because substantial tension may be present and result in dehiscence, particularly when there is severe annular dilatation. In addition, the right coronary artery may also be compromised.

Although the authors believe that all EA valves can be repaired at the initial operation, the question is whether this is appropriate for all patients. It is not controversial that valve repair is preferred, particularly in children and young adults. However, the initial operation in EA may not be until adulthood and by this time significant right ventricular enlargement and dysfunction may be present. In addition, the infinite anatomic variability sometimes demonstrates inadequate leaflet tissue for successful valve

repair. The mistake made too often, particularly by less-experienced surgeons, is that every valve should or must be repaired and if it is not, then it is a failure on the part of the surgeon. The decision as to whether a good, durable repair can be accomplished must be made early in the course of an operation to achieve a good result. A long crossclamp time with or without multiple bypass runs and repeated attempts at valve repair is more detrimental than recognition up front that successful repair is unlikely and proceeding to valve replacement with shorter bypass and crossclamp times.

EA has the widest range of clinical presentation—it is seen in symptomatic neonates as well as asymptomatic adults. Surgeons have been humbled and haunted by this defect for more than 5 decades. As a result, there are more repair techniques described for EA than any other defect in cardiac surgery. The Mayo surgical experience now exceeds 1000 patients. Our repair rate in the era of cone reconstruction approaches 100% in children and > 80% in adults and we continue to learn with every new patient. The review by Hertzner and colleagues provides a few more methods for our armamentarium. But not all patients with EA can be categorized into a classification system and not all patients can or should have the valve repaired.

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