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Commentary: Cone repair for Ebstein anomaly: It's all in the details

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Since its introduction by da Silva and colleagues in 2004,^{1,2} the cone reconstruction has gained widespread recognition as the operation of choice for patients with Ebstein anomaly and dramatically changed the outlook for patients undergoing operation. In experienced hands, mortality rates are exceedingly low and valve performance excellent at midterm follow-up.³ Perhaps more importantly, the technique significantly expands the range anatomies amenable to repair, thus avoiding the once rather common necessity of valve replacement.⁴

The Mayo Clinic experience with surgical treatment of Ebstein anomaly is, of course, unparalleled, spanning several decades and involving more than 1400 patients. Such an expansive institutional experience affords a unique and powerful perspective on the role and transformative nature of the cone repair in the treatment of Ebstein anomaly. This clearly written and masterfully illustrated article by Dearani,⁵ who (Dr da Silva himself notwithstanding) is the world's foremost expert on Ebstein anomaly and the cone repair, will be an invaluable resource for any surgeon endeavoring to perform the operation.

The fundamental elements of design and execution of the cone repair are now widely understood. It is by adhering to certain details of the operation and perioperative management that consistently favorable outcomes will be achieved and the oft-mentioned learning curve mitigated. Several key details are worth emphasizing.

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CENTRAL MESSAGE

The emergence of the cone repair technique has dramatically changed surgical treatment of Ebstein anomaly. Familiarity with the technical details is crucial in achieving a successful outcome.

From a technical standpoint, surgical delamination of leaflet tissue, the platform upon which the cone is built, requires thorough inspection of the ventricular side of the leaflet to ensure hyphenated and secondary or tertiary fibrous or muscular attachments are divided with only leading edge attachments remaining. Maintenance of the anteroseptal commissure is desirable, but when leaflet to leaflet approximation is needed to address excessive deficiency there the surgeon must be alert to the creation of tricuspid stenosis. Patch material is preferably avoided, but its use to increase the circumference of the cone is an important modification if stenosis is a concern. Patch material is also used to augment a deficient anterior leaflet body when central coaptation is suboptimal. Primary closure of residual or surgically created fenestrations in the leaflet tissue is routinely performed.

What might be referred to as management decisions are no less important, representing lessons learned of which readers should make special note. They include management of the atrial septum as well as use of the Glenn shunt in moderate to severely impaired right ventricles and how that decision is made (utilized in 20% of patients in the series reported by the Mayo Clinic group 2 years ago).³ Although not explicitly stated by the author, anything worse than mild tricuspid regurgitation on postrepair transesophageal echocardiography should prompt return to bypass to correct identified mechanical deficiencies of the repair.

What these points of emphasis have in common is that they are details, some quite subtle. They are insights gained

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by doing hundreds of these repairs, and in them lies the difference between success and failure. If the principles so ably articulated here are learned and studiously applied, outcomes for these often difficult anatomies will be measurably improved.

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