

References

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Commentary: Should the cone repair be the only option to consider for all patients with Ebstein's anomaly? Definitely not

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CENTRAL MESSAGE

The cone repair dramatically improved the short-term and mid-term outcomes for patients with Ebstein's anomaly but a lot still needs to be learned on long-term outcomes and patient selection.

In this issue of the *Journal*, the group from Munich, Germany, describe the outcomes of their surgical series in patients with Ebstein's anomaly.¹ Their aim was to investigate short-term and long-term outcomes (including tricuspid regurgitation, reoperation, and death) following conventional repair of Ebstein's anomaly and compare the results with the cone repair.

They demonstrate a clear advantage of the cone repair over conventional repair at a follow-up of 5 years. The 2 take-home messages are: (1) failed valve repair (that they define as in-hospital death, conversion to replacement, or in-hospital reoperation) is significantly less frequent after the cone repair (5%) than after other repair techniques (20%); and (2) the 5-year cumulative incidence of recurrent tricuspid regurgitation moderate or greater is significantly lower after the cone repair (8%)

than after other repair techniques (32%). These points are not arguable.

Nevertheless, this series has some significant limitations that the authors acknowledge quite well. The most important limitation is the significantly shorter follow-up of the cone group compared with the conventional group. This limitation prevents the authors from drawing any conclusions on the long-term assumed superiority of the cone over the conventional techniques. Moreover, the subjective and operator-dependent assessment of tricuspid regurgitation on echocardiography without objective measurement of the vena contracta in the setting of a retrospective single-center study brings more concerns about the reliability of the findings.

That being said, this nice study confirms the impression that almost all of us, cardiac surgeons taking care of this difficult and challenging population, share: the cone repair

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dramatically changed the short-term and mid-term outcomes in patients with Ebstein's disease, mainly by making this repair more reproducible.

The questions that our community should try to answer now are:

- (1) what are the long-term outcomes of the cone repair? Does the cone repair improve the right ventricular (RV) function, life expectancy, and quality of life of patients with Ebstein's disease long term?
- (2) Which patient subpopulation would benefit more from a quick and straightforward tricuspid valve replacement rather than a cone repair? Despite the outstanding results of the cone repair in the pediatric and young adult populations, the tricuspid valve replacement makes probably more sense than a cone in "older" patients

with a significant RV dysfunction and/or RV dilation to avoid postoperative RV failure, especially with the option of transcatheter valve-in-valve tricuspid valve replacement down the road. Nowadays, understanding when we should NOT perform a cone has become a more interesting question than knowing when we should do it.

Setting up an international registry on Ebstein's anomaly would be the next logical step to help our community to answer these questions. Who wants to take the job?

Reference

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