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Ebstein's anomaly- The one and a half ventricle heart

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Objective: Ebstein's anomaly remains a relatively ignored disease. Lying in the 'No Man's land' between congenital and valve surgeons it largely remains, under studied. We report our short-term results of treating it as a 'one and a half ventricle heart' and to propose that the true tricuspid annulus (TTA) Z-score be used as an objective criterion for estimation of functional right ventricle (RV).

Method: 22 consecutive patients undergoing surgery for Ebstein's anomaly were studied. Echocardiography was performed to assess the type and severity of the disease, tricuspid annular dimension and its Z-score patients were operated by a modification of the cone repair, with addition of annuloplasty, BCPC and right reduction atrioplasty to give a comprehensive repair. TTA Z-score was correlated later with post-plication indexed residual RV volume.

Result: There was one (4.5%) early and no late post-operative mortality. There was a significant reduction in tricuspid regurgitation grade. $(3.40\pm0.65 \text{ to } 1.22\pm0.42, \text{ p}<0.001)$. Residual RV volume reduced to $71.96\pm3.8\%$ of the expected volume and there was a significant negative correlation (rho: -0.83) between TTA Z-score and indexed residual RV volume. During follow-up of 20.54 ± 7.62 months, the functional class improved from 2.59 ± 0.7 to 1.342 ± 0.52 (p<0.001).

Conclusion: In Ebstein's anomaly, a higher TTA Z-score, correlates with lower post-plication indexed residual RV volume. Hence, a complete trileaflet repair with offloading of right ventricle by BCPC shunt (when TTA Z-score is >2) is recommended. The short-term outcomes of our technique are promising.

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