

Percutaneous closure of paravalvar leak after Cone repair for Ebstein's anomaly

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Ebstein's Anomaly is a rare congenital heart disease. The Cone-repair is used to repair the tricuspid valve. As in valve replacements, paravalvar leaks can affect the outcome.

A 5 year old boy with severe Ebstein's anomaly underwent a successful Cone-repair. At follow-up, his right ventricle function was reasonable, but he developed a paravalvar leak. The leak increased progressively and echocardiography at 1 year post repair showed right atrial dilatation, decreased RV function and important flow over the oval paravalvar defect of 8x5mm in postero-lateral position. Because of the hemodynamic importance of the leakage, percutaneous closure was scheduled.

Transoesophageal echocardiography confirmed the defect in infero-lateral position, but failed to provide accurate visualisation of the margins of the defect. As we failed to see the native valve under fluoroscopy, we monitored the procedure with transthoracic echocardiography which allowed perfect visualisation of the defect. An oval amplatzer vascular plug III type size 10/5 mm was positioned in the defect. The immediate limited residual leak was completely abolished after a few weeks, with recovery of RV function.

The "cone-repair" involves delamination and clockwise rotation of the anterosuperior leaflet, and use of the remnants of the septal and inferior leaflets to create a cone, subsequently the atrialized portion of the right ventricle is plicated. This is comparable to a valve replacement, re-inserting the own tricuspid valve instead of a (bio)prosthetic valve.

As in any other type of valve replacement, a paravalvar leak can compromise cardiac function, in which case closure of the leak is mandatory. Percutaneous closure is an option and is done under fluoroscopic guidance. If a bioprosthesis is inserted, fluoroscopic visualisation of the tricuspid annulus is often lacking and TEE is used in addition to guide the closure procedure. In our case, fluoroscopy and TEE failed to delineate the tricuspid annulus and the defect, but fortunately transthoracic echo provided accurate visualisation of the anatomic structures, which allowed us to close this paravalvar leak.

Conclusion: Paravalvar leak can compromise the results of Cone-repair for Ebstein's disease.

Percutaneous closure is possible, taking into account the limitations of guiding the procedure by fluoroscopy or even TEE.