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Optimal timing of pulmonary valve replacement in tetralogy of Fallot

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Summary and conclusions

Summary

The goal of the present study was to establish parameters for optimal timing of pulmonary valve replacement (PVR) in patients after repair of tetralogy of Fallot (TOF). Until recently there were no absolute criteria for the triage of patient for PVR.

Chapter 1 gives an overview of the long term effects of pulmonary regurgitation (PR) after total repair of TOF and the application of cardiac magnetic resonance imaging (MRI) in the assessment of RV functional parameters.

The natural time course of RV function and PR in repaired TOF patients is discussed in chapter 2. It was demonstrated that the development of RV dilatation depends on the surgical technique at initial repair. Patients who underwent transannular patch (TAP) repair had significantly more PR and showed deterioration of RV function over time. Patients that were repaired without the use of a patch also showed enlarged RV dimensions and decreased RV function, however, these parameters remained stable over time. The degree of PR did not change over time in neither of the groups.

Chapter 3 discusses the preoperative parameters that may influence the effect of PVR on functional parameters. There was no threshold above which RV volumes did not decrease after surgery. Preoperative RV volumes were independently associated with RV remodeling, also when corrected for a surgical reduction of the RV outflow tract. However, normalization could be achieved when preoperative RV end-diastolic volume was $<160 \text{ mL/m}^2$ or RV end-systolic volume was $<82 \text{ mL/m}^2$.

In chapter 4, the effect of PVR on right ventricular (RV) function was assessed by cardiac MRI. A group of 26 TOF patients underwent PVR for moderate to severe pulmonary regurgitation (PR) and diminished RV function. It was shown that all patients had an improved RV systolic function and an average decrease of RV dimensions of around 40%.

The effects of residual and recurrent PR after PVR were assessed in chapter 5. Patients with residual, defined as PR present at the first postoperative cardiac MRI, or recurrent

PR, defined as PR only present at the second and/or third postoperative cardiac MRI, was associated with a less favorable outcome of RV function. Although all 26 patients showed marked improvement of RV function and a significant decrease of RV dimensions, the degree of improvement was hampered by the presence or recurrence of PR, even though the degree of PR in these patients was mild (<20%) in most patients.

Chapter 6 concerns the improvement of diastolic function following PVR. A delayed normalization of diastolic function parameters was observed, as compared to the rapid improvement of systolic function. The possible different mechanisms that play a role in the improvement of diastolic and systolic function after valve replacement are discussed.

The effects of PVR on QRS duration are discussed in chapter 7. Long QRS durations are associated with an increased risk for arrhythmias and sudden death. QRS duration decreased in 18 of 26 patients and the degree of reduction of QRS duration was correlated with the reduction of RV dimensions.

Conclusions

The overall conclusions of this study are:

1. The type of initial repair in TOF greatly determines the time course of late RV failure as TAP repair leads to more severe PR than repair without a patch. Higher degrees of PR are associated with progressive RV dilatation and RV failure.
2. PVR leads to rapid improvement of RV systolic function, rapid decrease of RV dimensions and QRS duration and late improvement of RV diastolic function in most patients.
3. Residual PR or the recurrence of PR following PVR hampers the improvement of RV function and the reduction of RV dimensions.

Recommendations for timing of PVR

From the present study it can be concluded that timing of PVR should focus on the degree of RV dilatation. If RV dimensions become too large, no normalization can be expected after PVR. The degree of PR plays a key role in the timing of PVR as the time course of

increase of RV dimensions is directly correlated with the degree of PR. Special attention is needed in patients who underwent a TAP procedure in initial repair as TAP repair is associated with the highest degrees of PR compared to patients with a patch confined to the RV outflow tract or no patch at all.