Pulmonary valve replacement in patients with tetralogy of Fallot and pulmonary regurgitation: early surgery similar to optimal timing of surgery?

Ernst E. van der Wall1* and Barbara J.M. Mulder2

1Department of Cardiology, C5-P28, Leiden University Medical Center, PO Box 9600, 2300 RC Leiden, The Netherlands; and 2Department of Cardiology, Academic Medical Center, Amsterdam, The Netherlands

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This editorial refers to "Remodelling of the right ventricle after early pulmonary valve replacement in children with repaired tetralogy of Fallot: assessment by cardiovascular magnetic resonance"7 by E.R. Valsangiaco Büchel et al., on page 2721

Tetralogy of Fallot is the most common form of cyanotic congenital heart disease, with a prevalence of 0.26–0.8 per 1000 live births.1 Total repair for tetralogy of Fallot has been available for 50 years with a favourable outcome in most patients. Today, one is faced with an increasing number of patients with residual pulmonary regurgitation. It was previously thought that pulmonary regurgitation in Fallot patients was rather harmless. However, accurate measurements of right ventricular volumes using cardiovascular magnetic resonance (CMR) imaging have visualized important enlargement of the right ventricle in patients with severe pulmonary regurgitation.2 Moreover, it has been recently demonstrated that pulmonary regurgitation leads to progressive right ventricular dilatation and, with time, to right ventricular dysfunction, exercise intolerance, ventricular arrhythmias, and sudden death.3

Pulmonary valve replacement can be performed electively with little risk and may improve symptoms of right ventricular failure and provides excellent mid-term survival. The surgical procedure has a peri-operative mortality of 1–4% and a 10-year survival of 86–95%.4 Previous echocardiographic evaluation of right ventricular dimensions in children and adolescents showed a decrease in end-diastolic volume and end-systolic volume after pulmonary valve replacement. However, in adults, radionuclide angiography measurements showed no effects of pulmonary valve replacement on right ventricular volumes and ejection fraction.

To date, CMR is the gold standard for evaluation of right ventricular volumes and quantification of the degree of pulmonary and tricuspid regurgitation.5 There are distinct advantages of CMR over other imaging modalities in that CMR is independent of geometrical assumptions for evaluation of right ventricular volumes, mass, and function. Moreover, the unlimited field of view of CMR permits extensive evaluation of other right ventricular abnormalities such as aneurysms of the right ventricular outflow tract and regional right ventricular wall motion abnormalities.

In a previous study by Vliegen et al.,6 CMR was used in 26 adult Fallot patients (mean age 29 years) with pulmonary regurgitation and right ventricular dilatation to assess right ventricular functional parameters. Patients underwent CMR imaging with a median of 5 months before and 7 months after pulmonary valve replacement, and the authors observed a remarkable haemodynamic improvement in right ventricular function; right ventricular end-systolic and end-diastolic volumes decreased and right ventricular ejection fraction increased from 25 to 43%. For the first time, improvement of right ventricular function after pulmonary valve replacement was observed using CMR data. On the basis of these findings, accurate assessment of right ventricular volumes assisted in better timing of pulmonary valve replacement. The authors recommended a more liberal policy in pulmonary valve replacement in Fallot patients with pulmonary regurgitation and right ventricular dilatation.

Valsangiaco Büchel et al.7 studied 20 children (mean age 14 years) with tetralogy of Fallot and severe pulmonary regurgitation with right ventricular dilatation using CMR, 6 months before and 6 months after pulmonary valve replacement. The authors clearly showed that early pulmonary valve replacement resulted in beneficial remodelling of the right ventricle. Following pulmonary valve replacement, right ventricular volumes decreased and right ventricular mass showed a reduction from a mean of 49 to 36 g/m². Careful analysis showed that normalization of right ventricular function and mass by pulmonary valve replacement was largely dependent on the pre-existing right ventricular end-diastolic volume. Prompt remodelling of the right ventricle occurred when the right ventricular end-diastolic volume exceeded 150 mL/m², and the authors concluded that a cut-off level of 150 mL/m² should be used as a practical guide to advocate valve surgery. In line with the findings by Vliegen et al.,6 CMR may therefore usefully assist in better timing of pulmonary valve replacement in Fallot.

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patients. In addition, Valsangiacomo Büchel et al.\textsuperscript{7} provided a more precise functional parameter and potentially new criterion for better surgical guiding of these patients. As a result, the authors suggested that early pulmonary valve replacement might prevent the detrimental complications of severe pulmonary regurgitation, thereby favourably influencing long-term prognosis of patients after previous repair of tetralogy of Fallot.

Optimal timing of pulmonary valve replacement is still a subject of debate. So far, clinical symptoms, exercise tolerance, ventricular arrhythmias, progressive right ventricular dilatation, and onset of tricuspid regurgitation have been used as indicators for re-intervention. In addition, in symptom-free patients with right ventricular dilatation, pulmonary valve replacement has been considered for prevention and reduction of right ventricular dilatation. Additional arguments for this strategy are the predisposition of severe pulmonary regurgitation to ventricular dysrhythmias, and the beneficial effect of pulmonary valve replacement on electrical instability and the incidence of tachyarrhythmias.

However, one should realize that early pulmonary valve replacement is not similar to optimal timing of pulmonary valve replacement. It remains highly important to know the proper time span that the right ventricle can endure pulmonary regurgitation before irreversible dysfunction develops. Optimal timing of pulmonary valve replacement is critical for preserving right ventricular function (not too late) and avoiding the need for early re-operation (not too early). Amelioration of right ventricular function following pulmonary valve replacement has to be weighed against the risk of subsequent re-operation for homograft failure. In this context, it is interesting to note that Valsangiacomo Büchel et al.\textsuperscript{7} observed that none of their patients with a right ventricular end-diastolic volume >200 mL/m\textsuperscript{2} showed normalization of right ventricular function. Consequently, this functional parameter may become a helpful indicator for defining both a lower limit (150 mL/m\textsuperscript{2}) and an upper limit for re-intervention (200 mL/m\textsuperscript{2}).

In the future, an increasing number of children and young adult patients with previous repair of tetralogy of Fallot will require elective pulmonary valve replacement. Consequently, younger patients will already receive a ‘ticket’ for re-operation at a relatively young age. In addition, the younger the patient, the sooner the new grafts will deteriorate. In a mixed population of children and adults undergoing pulmonary valve replacement, the rate of freedom from further valve replacement is 81, 58, and 41\% at 5, 10, and 15 years, respectively. In adults, the life span of a pulmonary valve prosthesis ranges between 15 and 30 years. Despite these caveats and considerations, the overall opinion of experts in congenital heart disease is presently changing, and there is an increasing number of publications describing the detrimental effects of this condition.\textsuperscript{6,8–10}

In these articles, a less restrictive strategy is advocated with respect to pulmonary valve replacement in patients with severe pulmonary regurgitation and right ventricular dilatation late after total repair for tetralogy of Fallot. Pulmonary valve replacement should be offered to those patients long before irreversible right ventricular dysfunction occurs. For a definite strategy, prospective follow-up studies are needed for better defining the optimal timing of pulmonary valve replacement. The suitable moment for re-intervention can be either early or late, pending the use of well-established criteria. The study by Valsangiacomo Büchel et al.\textsuperscript{7} may be an important step in an attempt to define the appropriate criteria, based on CMR, for optimal timing of pulmonary valve replacement in patients with tetralogy of Fallot and pulmonary regurgitation.

**Conflict of interest:** none declared.

**References**