Optimal timing of pulmonary valve replacement in tetralogy of Fallot

Straten, A. van

1

General Introduction and Outline

Introduction

Tetralogy of Fallot (TOF) is a congenital heart defect consisting of four anatomical abnormalities (three of them are always present). It is the most common cyanotic congenital heart defect, representing 55-70% of all patients and it is the most common cause of cyanosis in the neonate [1]. The cardiac defects include a ventricular septal defect, overriding of the aorta and pulmonary valve stenosis. The fourth abnormality is a direct consequence of the pulmonary valve stenosis, namely hypertrophy of the right ventricle. Until the early 1940's TOF was thought untreatable. By that time surgeon dr. Alfred Blalock and cardiologist dr. Helen B. Taussig at Johns Hopkins University developed a palliative surgical procedure, which involved formation of an anastomosis between the subclavian artery and the pulmonary artery in order to bypass the pulmonary stenosis [2]. The Blalock-Taussig procedure was the only surgical treatment until the first total surgical repair was performed in 1954 [3]. Currently, Blalock-Taussig shunts are usually not performed on infants with TOF.

The first total repair was performed by dr. C. Walton Lillehei at the University of Minnesota in 1954. Total repair initially carried a high mortality risk which has consistently decreased over the years. Surgery is now often performed in infants in the first year of life. The operation generally involves incisions into the heart muscle, with relief of the right ventricular outflow tract stenosis and insertion of a (transannular) patch, and repair of the VSD using another patch.

A consequence of the insertion of a transannular of outflow tract patch is insufficiency of the pulmonary valve leading to pulmonary regurgitation.

For long, it was thought that mild to moderate pulmonary regurgitation following the repair of TOF was well tolerated. In recent years however, it has been shown that longstanding pulmonary regurgitation (PR) leads to right ventricular (RV) dilatation with decreased RV function and consequently, ventricular arrhythmias and the risk for sudden death [4].

Pulmonary valve replacement (PVR) is aimed at dissolving the PR in order to inverse the deterioration of right ventricular parameters. Several studies have now shown the

beneficial effects of PVR, including decrease of RV dilatation and improved systolic function [5, 6]. However, these effects have to be weighed against the inevitable risk for re-operations, since pulmonary homografts degenerate after 10-15 years [7].

Cardiac magnetic resonance imaging (MRI) is currently the gold standard for assessment of cardiac morphology and function in congenital heart disease due to its excellent spatial and temporal resolution. Cardiac MRI can be used to quantify cardiac volumes, ventricular function and the degree of valvular regurgitation and is therefore an important tool in the follow-up of TOF patients.

Repair of TOF is associated with an excellent prognosis. However, morbidity and mortality increase with age. PR is the most common lesion post repair and has been associated with exercise intolerance as well as atrial and ventricular arrhythmia [4]. PR relates to right ventricular outflow tract reconstruction and, in particular, the usage of a transannular patch during repair [8].

For the older patient with previously repaired TOF, PVR may lead to improvement of RV volumes and function, improved functional class, stabilization of QRS duration, and a reduction in atrial and ventricular arrhythmias. However, the optimal timing for late PVR remains unclear, and has been hampered by limitations in serial quantification of PR and RV function.

Outline of this thesis

The purpose of this thesis was to evaluate the time course of right ventricular function in repaired TOF before and after pulmonary valve replacement in order to establish criteria for optimal timing of pulmonary valve replacement.

Chapter 1 is a general introduction on magnetic resonance imaging in the follow-up of patients with TOF.

Chapter 2 contains a comprehensive review on right ventricular function late after total repair of TOF and the effects of PVR.

Chapter 3 describes the serial assessment of PR and right ventricular function in TOF. The time course of RV function was correlated to the surgical technique at initial repair.

Chapter 4 concerns the evaluation of possible preoperative thresholds for PVR in patients with corrected TOF.

Chapter 5 discusses the hemodynamic effects of PVR in adults late after repair of TOF.

Chapter 6 discusses the RV function following PVR in adult patients with TOF in correlation with the presence or absence of residual or recurrent PR.

Chapter 7 concerns the time course of diastolic and systolic function improvement after PVR in adult patients with TOF.

Chapter 8 describes the time course of QRS duration after PVR in adult TOF patients in relation to the reduction RV volume.

References

1 Ferencz C, Rubin JD, McCarter RJ et al. Congenital heart disease: prevalence at livebirth. The Baltimore-Washington Infant Study. Am J Epidemiol 1985;121:31-6.

2 Blalock A, Taussig HB. Landmark article May 19, 1945: The surgical treatment of malformations of the heart in which there is pulmonary stenosis or pulmonary atresia. By Alfred Blalock and Helen B. Taussig. JAMA 1984;251:2123-38.

3 Lillehei CW, Cohen M, Warden HE et al. Direct vision intracardiac surgical correction of the tetralogy of Fallot, pentalogy of Fallot, and pulmonary atresia defects; report of first ten cases. Ann Surg 1955;142:418-42.

4 Gatzoulis MA, Balaji S, Webber SA et al. Risk factors for arrhythmia and sudden cardiac death late after repair of tetralogy of Fallot: a multicentre study. Lancet 2000;356:975-81.

5 Graham TP, Jr., Bernard Y, Arbogast P et al. Outcome of pulmonary valve replacements in adults after tetralogy repair: a multi-institutional study. Congenit Heart Dis 2008;3:162-7.

6 Therrien J, Provost Y, Merchant N, Williams W, Colman J, Webb G. Optimal timing for pulmonary valve replacement in adults after tetralogy of Fallot repair. Am J Cardiol 2005;95:779-82.

7 Niwaya K, Knott-Craig CJ, Lane MM, Chandrasekaren K, Overholt ED, Elkins RC. Cryopreserved homograft valves in the pulmonary position: risk analysis for intermediate-term failure. J Thorac Cardiovasc Surg 1999;117:141-6.

8 d'Udekem Y, Ovaert C, Grandjean F et al. Tetralogy of Fallot: transannular and right ventricular patching equally affect late functional status. Circulation 2000;102:III116-III122.