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Original Research Article

Is it possible to operate four heart valves in a patient with heart failure, congenital heart disease and pulmonary hypertension?☆

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ABSTRACT

We describe a 60-year old man with the history of radical correction of the Tetralogy of Fallot (TOF) in the year 1964. This patient has had a long lasting decompensation of a severe right heart failure with ascites and pulmonary hypertension. On echocardiography he had residual mild pulmonary stenosis (PS) and severe pulmonary and tricuspid regurgitation (TR), moderate aortic and mitral regurgitation. He also had residual ventricular septal defect (VSD) and severe pulmonary hypertension with the maximal gradient on TR 83 mmHg. He was considered unoperable by his cardiologist, however patient decided to undergo a high-risk operation. The operation comprised pulmonary and aortic valve replacement with bioprosthesis, mitral and tricuspid repair, closure of ventricular septal defect, bilateral MAZE and volume reduction of both atria. After a very complicated postoperative course with multiorgan failure he recovered and was discharged home 2 months after operation. The NYHA class improved from IV before operation to II. Twenty months after this operation he experienced infective endocarditis with a leak on the aortic bioprosthesis. He was reoperated with reimplantation of a new aortic bioprosthesis and with a very complicated postoperative course. Sildenafil was added to his therapy due to the persistent severe pulmonary hypertension. He survived and was discharged home.

The presented unique high-risk complex reoperation of a congenital heart disease with pulmonary hypertension can be performed only by a very experienced cardiosurgical and cardio-anesthesiological team with a high level of the early and late postoperative care.

Abbreviations: AR, aortic regurgitation; CI, cardiac index; CFM, colour flow mapping; CHD, congenital heart disease; LVEF, left ventricular ejection fraction; RVEF, right ventricular ejection fraction; LV, left ventricle; LA, left atrium; MR, mitral regurgitation; PAP, pulmonary arterial pressure; PAPm, mean pulmonary arterial pressure; PAR, pulmonary arterial resistance; PCW, pulmonary wedge pressure; PH, pulmonary hypertension; RV, right ventricle; RA, right atrium; Qp, pulmonary flow; Qs, systemic flow; TOF, tetralogy of Fallot; TR, tricuspid regurgitation; VCI, vena cava inferior; VSD, ventricular septal defect

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1. Background

At present, the survival of children born with a congenital heart disease (CHD) into adulthood exceeds 85% [1–3]. The reason is probably the improved intensive care and cardiac surgery in newborns and babies with critical CHD. However, even after successful operation in childhood there is a risk of residual findings. Some of the residual findings (sequelae) are obligatory, e.g. degeneration of homografts or pulmonary regurgitation after the excision or decision of pulmonary valve. Other residua (e.g. residual defects) are due to the insufficient surgical technique, especially in 60th or 70th. Besides that, we can find acquired valve disease in a patient originally treated for CHD. Even severe residual lesions may be clinically silent for a long time. At the time of limiting symptoms the risk of the operation may be already very high. According to our experience, the most important risk factors of mortality in reoperations of adults with CHD are NYHA class III and IV, cyanosis and dysfunction of the right or left ventricle [4].

Tetralogy of Fallot (TOF) is one of the most common cyanotic CHD, it represents 4–10% of all CHD [2,3]. The long-term survival after the corrective surgery in childhood is very good. Approximately 90% of those, who survived operation, are still alive after 20–30 years [5–7]. The most common residual finding in TOF is pulmonary regurgitation, less common is pulmonary stenosis or residual ventricular septal defect (VSD) [2,3,8].

We describe a unique case of two reoperations in an older patient with TOF, who was considered inoperable because of the severe state with long lasting severe right heart failure, pulmonary hypertension and because of the extent of the surgery. We did not find any similar case report in the literature.

1.1. *Description of the case*

Our patient was born in 1950 with Tetralogy of Fallot. He had radical correction in 14 years, in the year 1964, the operation was performed by professor Navrátil in Brno, Czechoslovakia. Residual VSD was found in 1970, but reoperation was not performed because of the high operative risk and small patient's complaints. The reoperation was offered to our patient again in 2002, but he refused. In 2006 he was repeatedly hospitalized because of severe right heart decompensation with repeated ascites evacuation. He had persistent atrial fibrillation and non-sustained ventricular tachycardias (VT). He was referred to Hospital Na Homolce for implantable cardioverter-defibrillator (ICD) implantation, which was not performed because of the hypocalemia as the possible reason of VT. We performed a thorough reevaluation of all the residual findings and proposed a possibility of a high-risk reoperation.

We performed echocardiography (Table 1) and catheterization (Table 2). We found a high Doppler CW gradient (83/40 mmHg) on a massive tricuspid regurgitation (TR) during the first examination. This finding could imply irreversible pulmonary hypertension and inoperable state. However, the gradient on TR was increased by the residual pulmonary stenosis with the systolic gradient assessed by Doppler 46 mmHg. We also found moderate pulmonary, aortic and mitral regurgitations. The aortic regurgitation was caused by degenerative changes of the leaflets, vena contracta was 5 mm, pressure half time 366 ms and width of the jet was 15 mm=65% of the left ventricular outflow tract. Moderate mitral regurgitation was caused by annular dilatation to 46 mm and a small prolaps of the anterior leaflet, vena

Table 2 – Catheterization before operation (right and left catheterization, oxymetry, selective coronarography).

Qp (pulmonary flow)	9.7 l/min
Qs (systemic flow)	4.8 l/min
PAP syst./diastol.	62/22 mmHg
PAPmean	34 mmHg
PCW	19 mmHg
PVR	1.5 WU
Qp/Qs	2:1
PS: peak gradient	25 mmHg
Coronarography	No coronary stenosis

Qp, pulmonary flow; Qs, systemic flow; WU, Wood units; PVR, pulmonary vascular resistance; PAP, pulmonary arterial pressure; PAPm, mean pulmonary arterial pressure; PCW, pulmonary wedge pressure; PS, pulmonary stenosis.

Table 1 – Comparison of the echocardiographic examinations before and after operation.

Echographic parameters	ECHO before operation	ECHO before operation	ECHO after operation
	3.12.2008	7.4.2010	10.1.2011
LV	56/42 mm	53/42 mm	60/44 mm
LVEF	45–50%	45%	55–60%
RV	45 mm (PLAX) 57 mm (A4CH) RV anterior wall 15 mm	45 mm (PLAX) 57 mm (A4CH) RV anterior wall 14 mm	46 mm (PLAX) 39 mm (A4CH)
RVEF	40%	40%	40–45%
LA	81 × 75 mm	100 × 60 mm	63 × 77 mm
RA	94 × 81 mm	93 × 91 mm	61 mm in the long axis
TR gradient (max/mean)	83/40 mmHg	55/26 mmHg	34/ 23 mmHg
VCI	41 mm	41 mm	26/14 mm

LVEF, left ventricular ejection fraction; RVEF, right ventricular ejection fraction; VCI, vena cava inferior; TR, tricuspid regurgitation; LA, left atrium; RA, right atrium; RV, right ventricle; LV, left ventricle; PLAX, parasternal long axis projection; A4CH, apical four-chamber view.