The case of repeated reoperation of a patient with tetralogy of Fallot

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Intracardiac reparative correction of tetralogy of Fallot has been performed for more than 40 years: thus there are an increasing number of patients requiring reoperation during recent decades. The authors present an innovative surgical and cardiological approach in the case of a 42 year-old female patient who required a second reoperation (for residual left to right interventricular shunt, pulmonary and tricuspid valve regurgitation). The patient had undergone intracardiac repair of tetralogy of Fallot at 14 years of age, reoperation at 27 and had radiofrequency ablation for atrial arrhythmias with implantation of a pacemaker, at 35 years of age. At the time of the current operation, there was extensive calcification of the rim of the ventricular septal defect, and the residual defect could not be successfully repaired with sutures. Instead, the surgeon chose to close the ventricular septal defect by intraoperative deployment of an Amplatz occluder. The patient's postoperative clinical course was without complications, and the remaining shunt detected by transesophageal echocardiography was minimal. This offers an alternative to surgeons in the management of difficult reoperation for tetralogy of Fallot.

Key words: Tetralogy of Fallot – Reoperation – Amplatz occluder

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Intrakardiálna reparačná korekcia Fallotovej tetralógie sa realizuje viac ako 40 rokov, čo má za následok zvýšený počet pacientov v priebehu posledných desaťročí, ktorí sú indikovaní na reoperáciu. Autori predstavujú inováciu chirurgického a kardiologického prístupu pri opakovanej reoperácii na kazuistike 42-ročnej pacientky s reziduálnym defektom komorového septa, s pulmonálnou a trikuspidálnou regurgitáciou. Primárnu intrakardiálnu korekciu Fallotovej tetralógie mala pacientka vo veku 14 rokov, prvú reoperáciu v 27. roku života. Pre závažné poruchy rytmu podstúpila v 35. roku veku rádiofrekvenčnú abláciu a implantáciu kardiostimulátora. Pri druhej reoperácii (15 rokov po prvej a 28 rokov po primárnej operácii) sa peroperačne zistila výrazná kalcifikácia okrajov komorového defektu, ktorá nedovolila klasickú korekciu defektu (záplatou). Chirurg sa rozhodol perioperačne uzavrieť defekt Amplatzovým okluderom pri priamej kontrole zraku. Pooperačný priebeh pacientky bol bez závažných komplikácií. Pretrvával minimálny ľavo-pravý skrat, echokardiograficky dokumentovaný pri okraji okludera. Chirurgický postup pri komplikovanej reoperácii pacientky s Fallotovou tetralógiou bol alternatívnym (nevyhnutným) individuálnym riešením korekcie defektu.

Kľúčové slová: Fallotova tetralógia – reoperácia – Amplatzov okluder

Indications for reoperation in patients with tetralogy of Fallot (ToF) are: a residual ventricular septal defect (VSD) greater than 1.5 : 1, residual pulmonary stenosis with right ventricular pressure two thirds of systemic pressure or more, severe pulmonary and tricuspid regurgitation, a large RVOT aneurysm or evidence of infection, sustained clinical arrhythmias, significant aortic regurgitation, and aortic root enlargement 55 in diameter or more (1 – 3). A warranted intervention following reparative surgery is also the combination of residual VSD and/or residual pulmonic stenosis and regurgitation, all mild to moderate but leading to right ventricular enlargement (3). There is a rise of the number of patients who require reoperation following complete intracardiac repair during the last decades (4, 5).

The aim of our report is to demonstrate the importance of an individual approach to multiple repeat surgical corrections of the VSD in the case of a patient with ToF.

Patient

Our 42 year-old patient had first been diagnosed with ToF at six months of age: her parents declined surgical intervention at that time. The first intracardiac repair was carried out when she was fourteen (unfortunately

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the details of this operation are missing). Her recovery was complicated by hepatitis during hospitalisation. The central cyanosis disappeared and her clinical condition improved to the point that she could engage in routine excercise; nevertheless, there remained evidence, by auscultation, of persistent VSD and residual right ventricular outflow tract obstruction. Within several years her exercise tolerance declined and signs of right ventricular failure appeared. A cardiac catheterisation at 21 years of age confirmed the persistent VSD and residual pulmonary stenosis (combined valvular and subvalvular 57 mmHg gradient). The patient at that time refused reoperation, despite clinical necessityot digoxin and diuretics in therapy. Six years later, after many respiratory tract infections and worsening of heart failure (NYHA III-IV) she finally agreed to have reoperation. The surgeon closed the VSD with a dacron patch, partially enlarged the calcified outflow tract and then implanted a pericardial bioprosthesis in the pulmonary position. The postoperative course of the patient had been complicated by bleeding and pleuropericarditis. The ausculatory picture of recurrent VSD and pulmonary valve regurgitation gradually reappeared over the next few years, and the patient continued to have frequent respiratory infections. She had atrial tachyarrhythmias which were difficult to control and at the age of 35 she underwent radiofrequency ablation of the atrioventricular node and implantation of a VVI pacemaker. For the five years prior to the current intervention heart failure had continued at NYHA Class III.. Another cardiac catheterization at the age of 42 had confirmed a significant VSD (Qp : Qs = 2.4 : 1), pulmonary regurgitation grade III and tricuspid regurgitation grade II. The end-diastolic pressure in the right ventricle was 4 mmHg and mean pulmonary artery pressure was 20 mmHg. The patient was offered a second reoperation sixteen years after the first one.

Operative procedure – second reoperation

The patient underwent operation with endotracheal anesthesis with mild hypothermia. Extrathoracic arterial cannulation for cardiopulmonary bypass was achieved by use of the iliac artery; the venous cannula was inserted into the right atrium. The pulmonary artery was calcified and was opened cranially to the bifurcation, caudally to the left ventricular outflow tract. The bioprosthesis in the pulmonary valve position was excised and an allograft valve was implanted using continuous suture. The VSD (dehistentio of the last sutures) was examined before suture of the patch to the pulmonary artery, and the anatomy was best appreciated after opening the aorta. The diameter of the defect was 1.2 cm and its rims were calcified. Because of this, it was not possible to close the defect by sutures. Thus, it was chosen to close the VSD with an Amplatz-ASD-017 occluder device (Code 2677) under direct vision. The occluder extended as far as the cleft of the right coronary cusp. It was necessary to fix the device in the RVOT by prolene sutures. The aorta was sutured in two layers by continuous suture. The duration of cardiopulmonary bypass was 200 minutes and aortic cross-clamp time was 17 minutes. Intra-operative transoesophageal echocardiography did not demonstrate a significant left to right interventricular shunt and the allograft in pulmonary valve position appeared satisfactory.

Results

The early postoperative period of the patient was without any severe complications. Extubation was provided on the first day after operation. Circulation was supported by small doses of catecholamines during the first days, diuresis was forced by diuretics and the intensive respiratory rehabilitation was provided during all the hospitalisation time. The patient was discharged on the tenth postoperative day from the cardiosurgery department, with the anticoagulation therapy and with small doses of diuretics and digitalis. Her clinical condition has improved from NYHA III class to NYHA II. Echocardiography recorded diminution of the right ventricle size. There was a minimal residual left to right shunt at the upper rim of an Amplatz occluder. The patient has good function of the allograft in the pulmonary orifice and left ventricular ejection fraction is 55% (two years follow-up).

Discussion

Reparative correction of ToF has been known for more than 40 years. The overall survival of patients who have had operative repair is excellent, provided that the VSD has been closed and the right ventricular outflow tract obstruction has been relieved. A 36-year survival rate of 83% has been reported (2). Death may occur from reoperation, endocarditis or congestive heart failure. The cumulative risk of sudden death following repair of ToF seems to be about 1.2% at 10 years, 2.2% at 20 years, 4% at 25 years and 6% at 35 years (accounting for approximately one third of late deaths) (2, 6). Reoperation is

necessary in approximately 10% to 15% of patients following reparative surgery over a 20 year follow-up. Patients can be free of symptoms after repair but still have residual or recurrent hemodynamic abnormalities, such as an intracardiac shunt, pulmonary valve regurgitation or peripheral pulmonary obstruction (6 – 9). The correction of a residual VSD is undergone usually by pericardial or dacron patch.

Our patient underwent three intracardiac repairs (14, 27 and 42 years of age). The first intracardiac repair was carried out when she was fourteen (closure of the VSD and enlargement of the right ventricular outflow tract by pericardial patch). The central cyanosis disappeared and her clinical condition improved to the point where she could engage in routine excercise. There remained evidence of persistent VSD and residual right ventricular outflow tract obstruction. The second intracardiac repair (the first reoperation) was carried out 13 years after the first one. The surgeon closed the VSD with a dacron patch, partially enlarged the calcified outflow tract and then implanted a pericardial bioprosthesis in the pulmonary position. The postoperative course of patient was complicated by bleeding and pleuropericarditis. Echocardiography did not show VSD in the first month after the first reoperation. The picture of recurrent VSD and pulmonary valve regurgitation gradually reappeared over the next few years, and the patient continued to have frequent respiratory infections. She underwent radiofrequency ablation of the atrioventricular node and implantation of a VVI pacemaker. The heart failure of the patient continued at NYHA Class III. Another cardiac catheterization at the age of 42 confirmed a significant VSD. We could not exclude infective endocarditis as a cause of severe calcifications at the ventricular septum with the repeated VSD.

The second reoperation was sixteen years after the first one. The last correction of VSD was complicated because its rims were calcified and it was not possible to close the defect by sutures. The using of an Amplatz occluder for closure of VSD under direct vision had been chosen after the careful consideration of another technique for the correction of this defect (**Figure 1**). An Amplatz occluder is usually used for transcatheter occlusion of atrial septal defect (10 – 12). Transcatheter occlusion of VSD was first reported in 1988 (10), and there are many other reports with percutaneous closure of VSD with an Amplatz occluder (11 – 13). Most defects are unsuitable for this procedure. Perimembranous VSD is usually too close to the aortic and atrioventricular valves to permit safe transcatheter closure (12, 13). We do not see in clinical practice the use of the Amplatz occluder

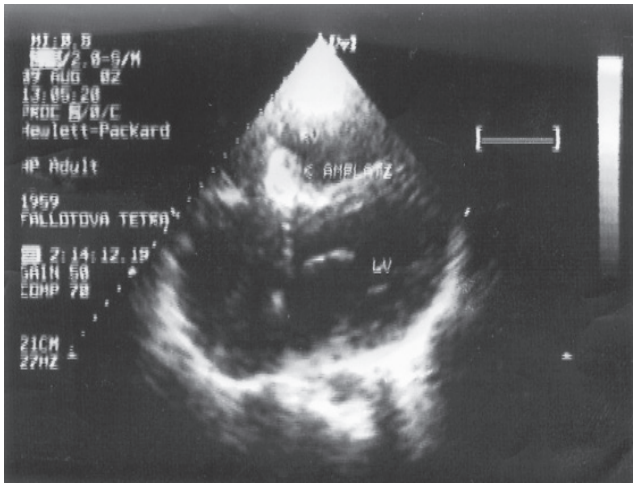

Figure 1 Postoperative echocardiography revealed occlusion of ventricular septal defect by the Amplatz occluder
LV – Left ventricle, LA – Left atrium, RA – Right atrium, Amplatz – device

for closure of VSD under direct vision in open-heart surgery – reoperation for repeated residual conotruncal VSD. The result of the operation confirmed the rightness of our decision. Pulmonary valve replacement after the correction of ToF had low operative risk (9, 14, 15). Allotransplant implantation at the pulmonary position during reoperation of ToF is indicated for the patients who have had transannular enlargement of the pulmonary artery and if leads to the postoperative regression of right ventricle (16).

Conclusion

Reoperations of ToF require an individual surgical approach. The implantation of an Amplatz occluder to the VSD (whose rims were calcified and where it was not possible to close the defect by sutures) under direct vision and allograft implantation at the pulmonary position during reoperation of ToF is a suitable surgical procedure. It provided for the success of the operation in our patient.

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