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Surgical Pulmonary Valve Replacement Due to Failed Percutaneous Pulmonary Valve Intervention in a Patient After Correction of Fallot's Tetralogy: Surgery Remains the Standard

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
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Conflict of interest: None declared

Patient: Male, 26
Final Diagnosis: Pulmonary valve replacement
Symptoms: No symptom
Medication: —
Clinical Procedure: —
Specialty: Surgery

Objective: Congenital defects/diseases

Background: One of the most common complications after repair of Fallot's tetralogy is pulmonary valve regurgitation (PR). There are many concepts of treatment, such as surgical, percutaneous, or hybrid pulmonary valve repair. Surgical pulmonary valve replacement is associated with low operative morbidity and mortality and shows very good long-term results. For that reason, it remains the standard of treatment.

Case Report: We present a case of a 26-year-old male patient who underwent a successful emergent surgical pulmonary valve replacement after a failed percutaneous pulmonary valve intervention, which was performed due to pulmonary regurgitation.

Conclusions: Despite the modern (interventional and hybrid) procedures in the treatment of pulmonary valve regurgitation after repair of Fallot's tetralogy, surgical treatment for pulmonary regurgitation still remains the gold standard for young adult patients.

MeSH Keywords: Pulmonary Valve • Pulmonary Valve Insufficiency • Tetralogy of Fallot

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Background

Tetralogy of Fallot is a congenital cardiac malformation that occurs in 3 of every 10 000 live births, and accounts for 7–10% of all congenital cardiac malformations. It includes a ventricular septal defect, obstruction of the right ventricular outflow tract, override of the ventricular septum by the aortic root, and right ventricular hypertrophy. The standard therapy is the surgical correction [1]. Pulmonary regurgitation (PR) is a common complication after the repair of Fallot's tetralogy (TOF) [2]. If moderate, it is usually well tolerated in childhood. In the long term, chronic PR has negative influences on right ventricular function and exercise capacity and increases the risk of ventricular arrhythmias and sudden cardiac death [2,3].

We present a patient who had previously undergone a surgical repair of TOF and suffered from clinically-relevant PR. First, he underwent a percutaneous right ventricular outflow tract (RVOT) pre-stenting before the scheduled pulmonary valve replacement. The procedure was complicated with a dislocation of the stent. Our case report, like other cases from the literature, emphasizes the relevance of surgical therapy in PR after repair of TOF compared to other therapeutic modalities.

Case Report

A 26-year-old man was admitted to the Cardiology Department of our institution due to PR for pre-stenting of the RVOT before an elective percutaneous pulmonary valve replacement. He presented heart failure NYHA Class II–III°. At the age of 15 months, he had undergone a repair of TOF with patch implantation into the RVOT. The patient was in a sinus rhythm and in acceptable general condition before the procedure. The right ventricle was considerably dilated with slight obstruction of the RVOT. The right ventricle end-diastolic volume index (RVEDVI) was 161 ml/m^2. During the intervention of the pre-stenting, the stent graft should have been initially placed into the pulmonary valve ring, but during inflation it slipped out of the valve ring and moved into the left pulmonary artery. There were various unsuccessful rescue attempts. Under these conditions, the intervention had to be discontinued. The patient was referred to our clinic for emergent surgical treatment. On the following day, the surgery was performed without operative complications. After median re-sternotomy, an extensive preparation was necessary due to excessive adhesions. The severe adhesions were released without tissue injury. The extracorporeal circulation was connected with the selective drainage of the superior and inferior vena cava. The old patch material was removed and the RVOT was opened. The pulmonary valve presented a relevant insufficiency. The main pulmonary artery was opened with a longitudinal incision. In doing so, the stent graft was removed without any further injury to the left pulmonary artery (Figure 1A, 1B). The RVOT was measured. The native pulmonary valve was excised (Figure 1C). A stentless pulmonary valve conduit with a 25-mm diameter was sewn (BioPulmonic Conduit™, Company SyGan Medical) (Figure 1D, 1E). The postoperative course was uneventful. Six months after surgery, the echocardiographic examination showed no pulmonary regurgitation, as well as slight improvement in right ventricular function. Furthermore, the patient was in a good clinical condition with a remarkable improvement in exercise capacity.

Discussion

PR is common in patients after repair of TOF [2]. It is well tolerated for many years following the primary repair of TOF and does not show symptoms. However, it can result in progressive exercise intolerance and heart failure, and can cause tachyarrhythmia and sudden death [4]. Nowadays, there are many concepts of treatment, such as surgical, percutaneous, or hybrid pulmonary valve repair.

Surgical pulmonary valve replacement is associated with low operative morbidity and mortality and shows very good long-term results [4]. Cavalacnti et al. reported a meta-analysis of 48 studies comprising 3118 patients who underwent surgical pulmonary valve replacement (PVR) after surgical repair of TOF [5]. The 30-day mortality rate was 0.87% and the 5-year mortality rate was only 2.2%. The rate of 5-year re-PVR was 4.9%. Furthermore, the results of this meta-analysis demonstrated improvement in the left and right ventricular functions, as well as reduction in symptoms [5]. Another study investigated the mid-term results of 131 patients after bioprosthetic pulmonary valve replacement in PR after TOF repair and showed that there was neither early nor late mortality. Moreover, it revealed that the RV function showed marked improvement within the first post-operative year, with acceptable rates of freedom from reoperation due to prosthetic valve failure [6]. The improvement of the RV function was also confirmed in our present case. Catheter-based interventions present an attractive alternative to surgery in patients with TOF and PR. Bonhoeffer et al. performed the first percutaneous pulmonary valve replacement (PPVR) in 2000 [7]. Percutaneous approaches are limited by patient size and size of the valve or the anatomy of the right ventricular outflow tract [3]. Furthermore, it does not offer the opportunity to repair additional defects such as pulmonary artery dilatation, which is common in severe pulmonary regurgitation [3]. Khambadkone et al. showed no mortality in 10 months within a collective of 59 patients; however, device complications occurred in 14 patients (25%) [8]. Furthermore, in a US study of 102 cases of PPVR, the authors reported a significant reduction of PR along with improved RV function. However, PPVR was associated with stent fractures in 5% of the patients [9].

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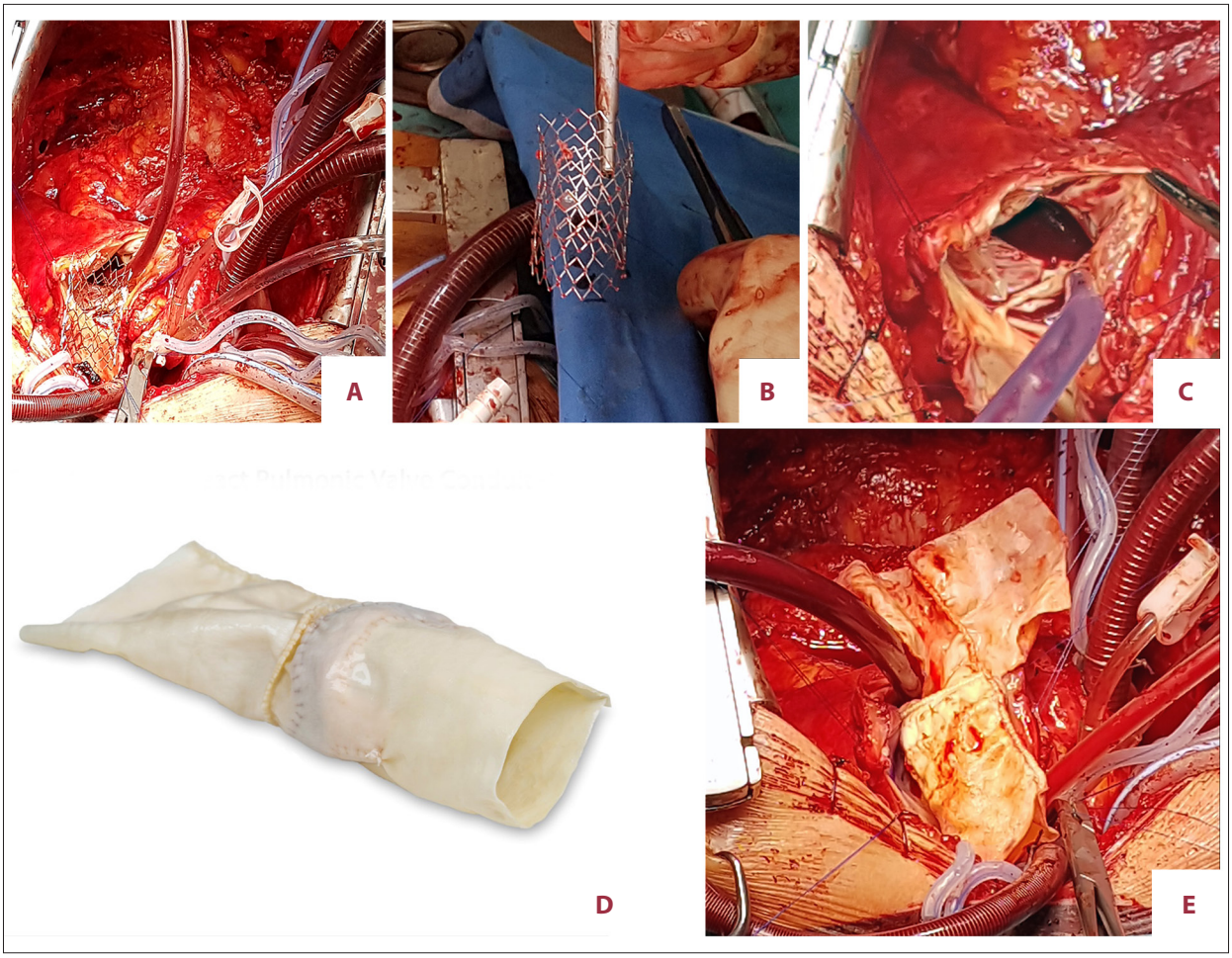

Figure 1. (**A–E**) Operative steps and the BioPulmonic Conduit, details in text.

The optimal timing of PR treatment is still under debate and can be very challenging because the majority of patients tolerate the right ventricular volume overload as a consequence of PR for many years without the development of symptoms [10].

The compensatory mechanisms fail and might lead to irreversible dysfunction of the right ventricle in the long term and a significant PR can result in ventricular arrhythmias and sudden death [6,10]. Harrild et al. and Jang et al. found that late pulmonary valve replacement for symptomatic PR did not reduce the incidence of ventricular tachycardia or death and did not show clinical improvement [6,11]. Therefore, several centers recommend early pulmonary valve replacement before the development of symptomatic heart failure [10]. This fact was considered in the present case, but the unsuccessful intervention necessitated urgent surgery. The recommendations for early treatment are based on the facts that pulmonary valve replacement has low mortality and re-intervention rates, and the improvement of symptoms, functional status, and right ventricular size are to be expected after treatment [6,12,13].

In a retrospective analysis of 278 cases over a 40-year period, Sabate Rotes et al. showed early survival rates of 98.6% and 80% at 15 years, with 97% freedom from re-intervention at 5 years [14]. Other studies have confirmed these results [13,15].

At present, there are various conduits and valve models. We decided to implant the BioPulmonic conduit because of its various advantages. The conduit is made of porcine pulmonary valve and pericardium. It is suitable as a replacement for diseased or damaged pulmonic valve or artery, such as for the Ross procedure or as an alternative to a pulmonic homograft, and in small children who need a replacement of their pulmonic valve or the total right ventricular outflow tract. With regard to adverse postoperative events such as thromboembolism, valve thrombosis, bleeding, endocarditis and valve dysfunction, the BioPulmonic conduit has shown significantly better results compared to the other valve models [16,17]. The most common failure of this conduit is distal stenosis. Nevertheless, compared to the other pulmonary conduits, the BioPulmonic conduit shows a lower frequency of distal stenosis [16,17].

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Conclusions

The surgical treatment for pulmonary regurgitation after the repair of TOF still remains the gold standard for young adult patients. On the basis of our case report, we were able to confirm this fact as already described in the literature. Despite the surgical challenges in this special patient group, this surgical treatment is a safe and effective technique with low procedural risk and is associated with low early and late mortality rates, as well as high freedom from re-intervention.

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Conflict of interest

None.

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