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VALVULAR HEART DISEASE

CLINICAL CASE

Long-Term and Multidisciplinary Treatment of Tetralogy of Fallot in Pediatrics

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ABSTRACT

BACKGROUND Right ventricular outflow tract (RVOT) dysfunction is a long-term postsurgical complication of tetralogy of Fallot (TOF) that needs long-term multidisciplinary treatment.

CASE SUMMARY We report a case of TOF patient who underwent radical surgical repair in infancy and who presented with pulmonary artery stenosis and pulmonary regurgitation during follow-up. Pulmonary stent placement and percutaneous pulmonary valve implantation (PPVI) were pursued successfully when he was aged 5 and 13 years, respectively.

DISCUSSION PPVI has been widely used as a minimally invasive treatment alternative to surgical pulmonary valve replacement. This technology has shown significant advantages in pediatric patients to improve RVOT dysfunction, relieve symptoms, optimize hemodynamic parameters, and avoid reintervention.

TAKE-HOME MESSAGE This case provides a long-term and multidisciplinary strategy for TOF in pediatrics, with a focus on the possibility and effect of PPVI in adolescent patients. (JACC Case Rep. 2025;30:103940)

HISTORY OF PRESENTATION

A 6-month-old male patient was presented to our cardiology department (Shanghai Children's Medical Center [SCMC], Shanghai, China) with cyanosis for 5 months that had been worsening for 2 weeks. On physical examination, the patient had sweating, pallor, and central cyanosis, and pulse oximetry indicated an oxygen saturation (Spo_2) of 90%. His heart rate and respiratory rate were 165 beats/min and 45 breaths/min, respectively. A grade 2 systolic murmur was heard between the third and fourth intercostal spaces on the left sternum edge, with reduced pulmonary valve closure (P_2).

TAKE-HOME MESSAGES

- This case highlights the importance of long-term and multi-disciplinary treatment of TOF.
- The case also provides possibility to treat PAS and PR by stent and PPVI in pediatric patients after TOF surgery.

PAST MEDICAL HISTORY

Fetal echocardiography had detected tetralogy of Fallot (TOF) in this patient at 20 weeks' gestation age.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

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ABBREVIATIONS AND ACRONYMS

CT = computed tomography

LPA = left branches of the pulmonary artery

MPA = main pulmonary artery

PAS = pulmonary artery stenosis

PPVI = percutaneous pulmonary valve implantation

PR = pulmonary regurgitation

RPA = right branches of the pulmonary artery

RV = right ventricular

RVEDVi = right ventricular end-diastolic volume index

RVOT = right ventricular outflow tract

TOF = tetralogy of Fallot

VSD = ventricular septal defect

His mother was G1P1; his birth weight was 3,200 g, and he had no birth complications. He had no chronic conditions, past surgical procedures and hospitalizations, allergies, medications, or family history.

DIFFERENTIAL DIAGNOSIS

According to his clinical presentation and echocardiography, the diagnosis of TOF could be confirmed, and we excluded other cyanotic congenital heart diseases, such as critical pulmonary stenosis, transposition of the great arteries, total anomalous pulmonary venous connection, or truncus arteriosus.

INVESTIGATIONS

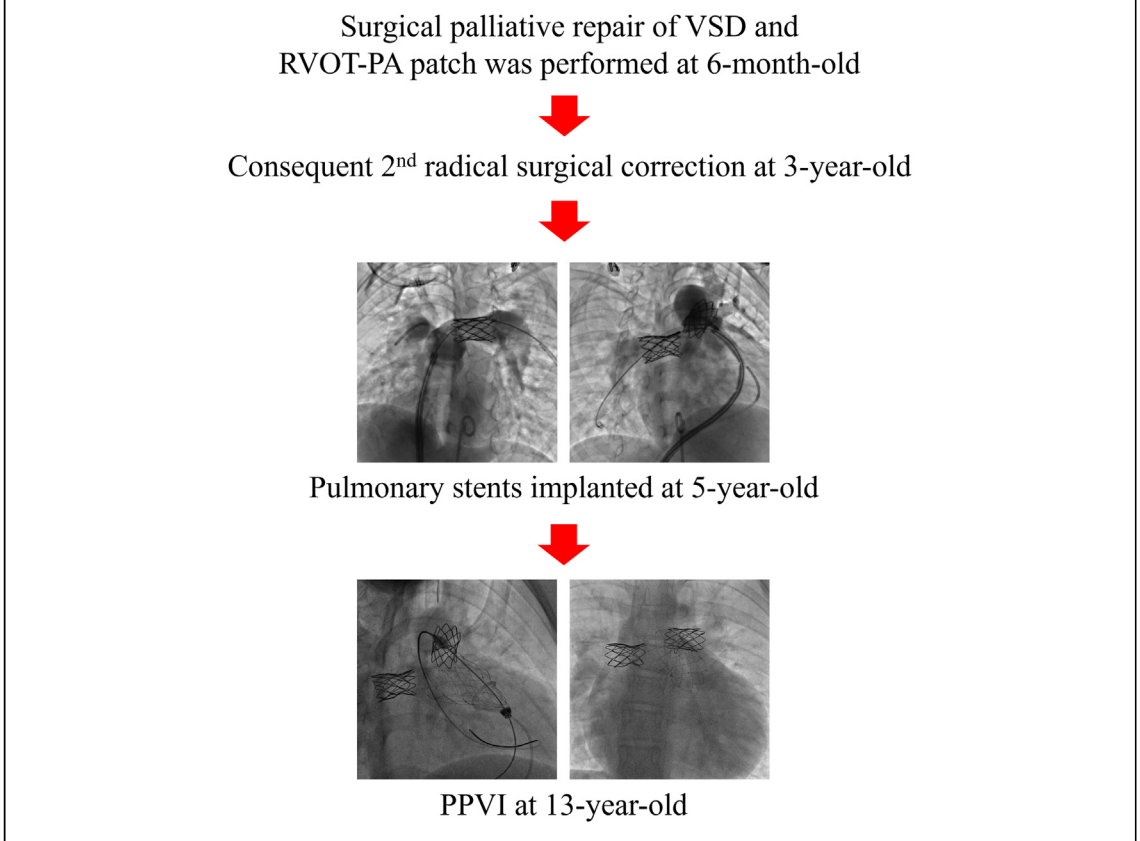
Electrocardiography indicated right-axis deviation and right ventricular (RV) hypertrophy. Echocardiography enabled the diagnosis of TOF. Further cardiac computed tomography (CT) angiography and cardiac catheterization confirmed the diagnosis and also detected the presence of minor aortopulmonary collateral vessels.

MANAGEMENT

Given that the patient had recurrent "tet spells," surgical palliative repair of the ventricular septal defect (VSD) and a RV outflow tract (RVOT)-pulmonary artery (PA) patch were performed when the patient was aged 6 months. The second radical surgical repair was finished after 3 years, including VSD repair and patch enlargement.

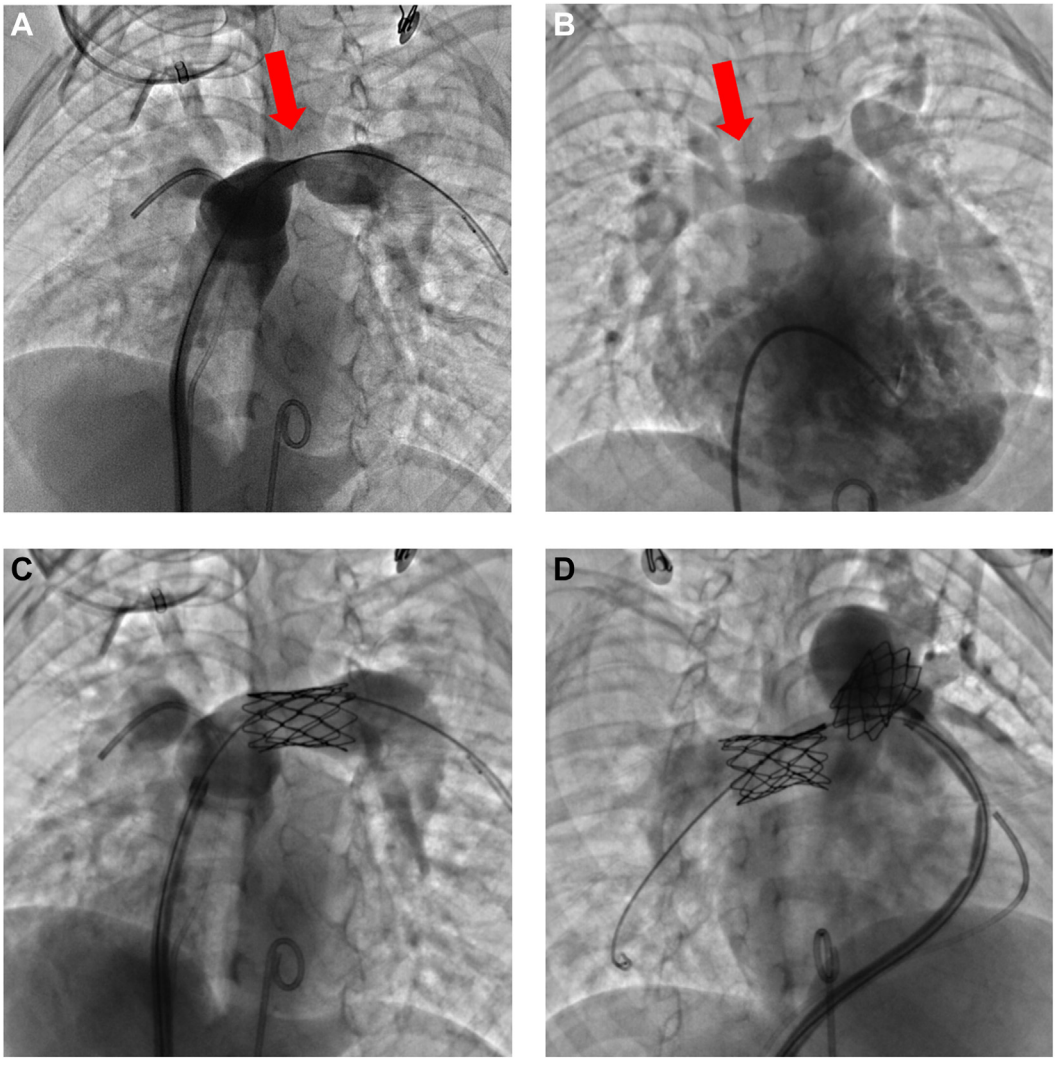
At the age of 5 years, the patient presented with exercise intolerance. Echocardiography and CT revealed stenosis of left and right branches of the PA (LPA and RPA, respectively). According to the velocity of pulmonary blood flow shown in Doppler cardiography (LPA, 3.8 m/s; RPA, 3.3 m/s), the pressure gradients had reached approximately 57.7mm Hg and 43.7mm Hg, respectively. Interventional angiography further confirmed the location of significant stenosis of the LPA and RPA (Figures 1A and 1B), and both

VISUAL SUMMARY Long-Term and Multidisciplinary Treatment of TOF in Pediatrics



PA = pulmonary artery; PPVI = percutaneous pulmonary valve implantation; ROVT = right ventricular outflow tract; VSD = ventricular septal defect.

FIGURE 1 **Angiography**

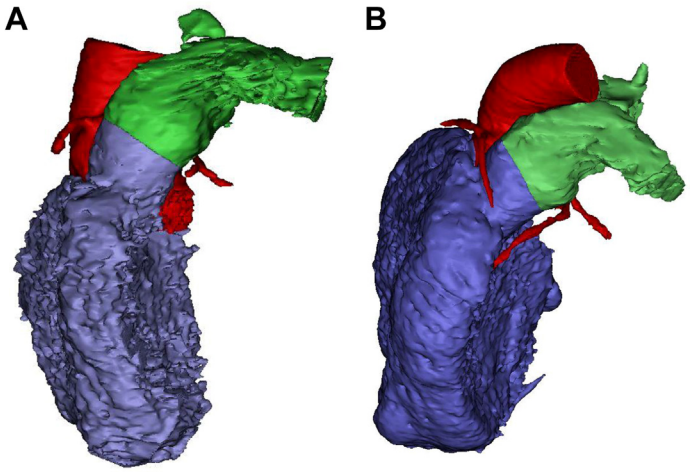


Imaging showed (A) left pulmonary artery stenosis (red arrow) and (B) right pulmonary artery stenosis (red arrow). Pulmonary stents were sequentially implanted in the (C) left and (D) right branches of the pulmonary artery.

pressure gradients of the LPA and RPA were >40 mm Hg. Two Cheatham-Platinum (CP, NuMed) stents (28-mm length stent with a 14 × 30 mm balloon in balloon catheter [BIB, B. Braun Interventional Systems]) were selected to cover the stenosed locations of the LPA and RPA sequentially (**Figures 1C and 1D**). The pressure gradient improved to only 3 mm Hg after the intervention, and the hemodynamics of pulmonary blood flow and symptoms were all improved during follow-up.

When this patient was aged 13 years, he presented with exercise intolerance again. Echocardiography detected severe pulmonary regurgitation (PR), and cardiac magnetic resonance demonstrated RV dysfunction with an RV end-diastolic volume index (RVEDVi) of 209.96 mL/m^2. Considering the patient's age (13 years) and weight (51 kg), to avoid surgical reintervention, and after a multidisciplinary treatment discussion, we decided to perform a percutaneous pulmonary valve implantation (PPVI) procedure. At that time, only the PT-Valve (MedZenith) was available in China. All protocols and procedures performed in this study were approved by the Ethics Committee of the SCMC, and informed

FIGURE 2 Cardiac Computed Tomography Reconstruction



Imaging demonstrated type II (cylindrical with small changes in caliper throughout) morphology of the right ventricular outflow tract and a low risk of coronary artery compression in the (A) systolic and (B) diastolic phases.

consent was obtained from the patient's guardians. All procedures were performed in accordance with the Declaration of Helsinki. CT reconstruction demonstrated a type II RVOT with a constant diameter and low risk of coronary compression (**Figures 2A and 2B**). Angiography showed an enlarged right ventricle and previously implanted stents in a suitable location without obstruction. Severe diastolic PR was noted (Videos 1 and 2). The inner diameters of the RVOT and the pulmonary valve ring were 22.38 mm and 21.18 mm, respectively, whereas the diameter of the main PA (MPA) and the length of the MPA were 23.45 mm and 41.63 mm, respectively. Next, 18-F and 22-F dilation sheaths were advanced sequentially through the right femoral vein. A PT-Valve (TPV3626) was delivered through an LPA pathway (**Figures 3A and 3B**). Finally, the pulmonary valve was released in the intended position with proper morphology, without blocking LPA and RPA blood flow (**Figures 3C and 3D**). PR disappeared soon post-implantation of the valve, as shown on angiography (Videos 3 and 4).

OUTCOME AND FOLLOW-UP

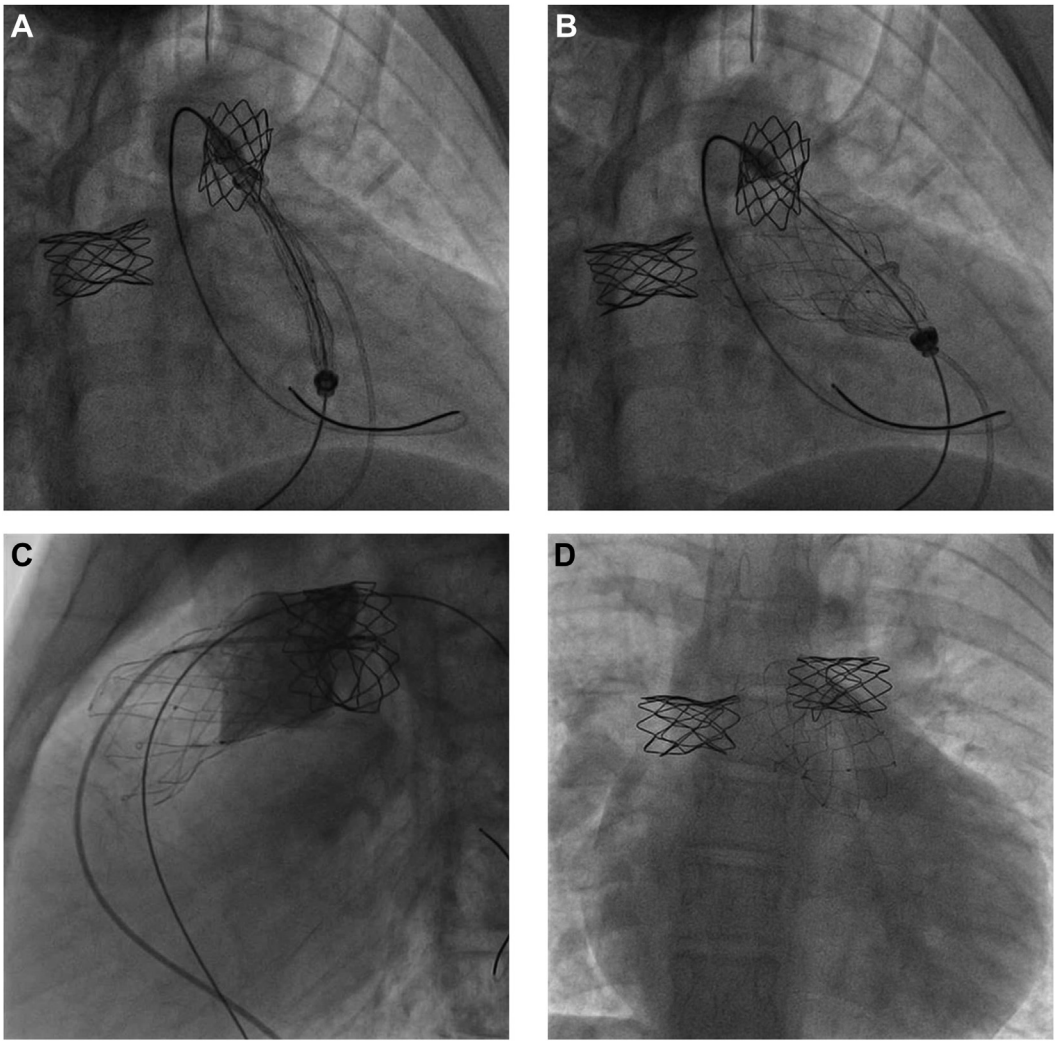
The patient continues to be followed up on an outpatient basis by cardiology and has been without symptoms, arrythmia, or other complications. Of note, the RVEDVi decreased to 142 mL/m^2 1 month post-implantation of the pulmonary valve. Recent follow-up has shown hemodynamic stability with normal a RVEDVi and RV function without stents or valve malposition. He is showing normal physical growth and development.

DISCUSSION

TOF is the most common cyanotic congenital heart disease, accounting for 5% to 7% of all congenital heart defects. Clinical presentation varies according to the severity of the RVOT obstruction, most commonly manifesting in a cyanotic neonate. With the advent of fetal echocardiography, the diagnosis can be made at the perinatal stage.[1] Although the current surgical mortality rate can be as low as 2%, with a 20-year survival rate in excess of 90%, most develop residual hemodynamic and/or electrophysiologic abnormalities.[2] However, a consensus on the optimal management of TOF in symptomatic neonates and infants, as well as management of long-term complications after surgery, has still not been reached. PA stenosis (PAS) and PR are common RVOT dysfunction complications, but treatment in pediatric patients remains controversial. Here we presented a TOF case in a patient treated by a surgical and catheterization team in a long-term and multidisciplinary manner, thereby providing a possible safe and effective strategy using pulmonary stent and valve implantation techniques.

Because a PAS lesion is difficult to treat surgically, and given the risks of repeated surgical operations, interventional therapy, especially stent implantation, has become the main treatment method. With the advent of expandable balloons in the late 1980s and the emergence of novel types, materials, and models of stents used for PAS treatment, stent implantation has become the first-line treatment for PAS.[3,4] Several studies have found that proximal and distal PA perfusion significantly improved after initial stent implantation in children with PAS.[5,6] In the present case, considering the patient's young age of 5 years, we implanted LPA and RPA stents sequentially, instead of using a "kissing stent technique," which involves simultaneously deploying 2 stents and could lead to total obstruction of pulmonary blood flow. Compared with the traditional Palmaz, the CP stent has a wider range of expandable diameters and length, the stent edge causes less damage to the balloon and blood vessels, and the CP stent has better plasticity. Recently, novel Pul-Stent (Med-Zenith) has shown post-expansion capabilities and a semiopen loop design that can adapt to the needs of children's growth and development while avoiding occlusion of branch vessels.[5,7]

FIGURE 3 **Cardiac Catheterization**



Imaging demonstrated proper morphology of pulmonary stents previously implanted. A PT-Valve (Med-Zenith) was then delivered through (A) a left pulmonary artery branch (LPA) pathway and (B) released. After implantation of the pulmonary valve, (C) the patient had no pulmonary regurgitation, and (D) the stents were in their correct positions.

In the past 2 decades, PPVI has become widely accepted as a minimally invasive treatment alternative to surgical pulmonary valve replacement that results in faster recovery and shorter hospital stays. The indications for PPVI have been extended to native and larger RVOTs, and PPVI is now an option for a growing number of people who need surgical valve replacement. At the same time, a life span revalving strategy should be taken into consideration.[8] PPVI technology has shown significant advantages and success in improving RVOT dysfunction, relieving patient symptoms, optimizing hemodynamic parameters, and avoiding the need for reinterventional surgery.[8,9] However, its widespread clinical application is still in early stages, especially in adolescents. The application of PPVI in pediatric patients has certain challenges, such as anatomical diversity of the RVOT and MPA, conveying system profile selection, and vascular access.

The Med-Zenith PT-Valve is a novel type of transcatheter self-expandable pulmonary valve. Different from other balloon-expandable or self-expanding valves, it is fully covered by pericardial tissue mounted on a nitinol frame. For most Chinese TOF patients who undergo RVOT with patch repair, the frame diameter is designed to be 28 to 44 mm at both

ends, with wide length range (38-54 mm). The stent is designed straight in the middle, with symmetrical trumpet-shaped expansion structures at both ends. The movement of the PA during diastole and systole is a compound process with diameter change and conduit twisting. The stent ring at the distal end of the Med-Zenith pulmonary valve stent is an open-loop structure with good compliance to adapt to the huge deformation of the PA in different cardiac cycles, minimize stent tension, and thus reduce the incidence of stent fracture and coronary artery compression.[10] Positive results from a clinical trial demonstrated the Med-Zenith PT-Valve's safety, effectiveness, and viability in treating severe PR in patients with noticeably enlarged RVOTs.[10]

This patient is becoming an adult with hemodynamic stability and normal physical growth and development. Longer follow-up in our children's hospital is still needed.

CONCLUSIONS

TOF is a complicated congenital heart disease with long-term complications, and long-term follow-up and multidisciplinary treatment should be performed. A cardiac catheterization strategy, especially including PPVI, is feasible in adolescent patients with suitable anatomy and appropriate device selection.

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KEY WORDS congenital heart disease, percutaneous pulmonary valve implantation, pulmonary stenosis, stent, tetralogy of Fallot

APPENDIX For supplemental videos, please see the online version of this paper.