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ADVANCED

CASE REPORT: CLINICAL CASE

Tetralogy of Fallot

Pushing Through Despite Long Odds

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ABSTRACT

The natural history of tetralogy of Fallot depends on whether a transannular pulmonary valve patch or shunt surgery was necessary in infancy. This case illustrates the feasibility of cardiac rest on extracorporeal membranous oxygenation for a very ill adult with conduit endocarditis who received a right ventricle-to-pulmonary artery valveless conduit for later transcatheter pulmonary valve replacement. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2019;1:523–5)

HISTORY OF PRESENTATION

We describe a 26-year-old male patient with tetralogy of Fallot, valvular pulmonary stenosis, and infundibular stenosis. His most recent surgical procedure consisted of a right ventricular (RV) outflow tract (RVOT) reconstruction, ventricular septal defect (VSD) patch repair, and a pulmonary homograft. His first surgery was a standard tetralogy of Fallot repair with VSD patch closure and infundibular resection. He later developed RVOT obstruction severe enough to require a right ventricle-to-pulmonary artery (RV-PA) conduit as his second surgical procedure. Afterward, he experienced multiple recurrences of RVOT obstruction, as well as pulmonary valve insufficiency. As a result, he required multiple palliative operations and transcatheter pulmonary valve replacements (PVRs). He also developed acute on chronic renal failure, with dialysis dependence, 1 year before the current presentation. The renal failure was thought to be related to a hypoperfusion injury related to his chronically depressed left ventricular (LV) systolic function because no intrarenal cause was found after consultation with nephrology. He also had an automated implantable cardioverter-defibrillator (AICD) placed 7 years before presentation, for sustained symptomatic ventricular tachycardia. This case describes the patient's most recent admission for endocarditis of his RV-PA conduit and AICD leads.

His physical examination on presentation was significant for fever and for systolic and diastolic murmurs indicative of pulmonary stenosis and severe pulmonary conduit insufficiency. He was admitted to the cardiology service for an endocarditis work-up and cardiac surgery, and he was treated with antibiotics throughout his hospitalization. Blood cultures grew methicillin-resistant *Staphylococcus epidermidis*.

LEARNING OBJECTIVES

- Recognize the most common surgical procedures required for tetralogy of Fallot.
- Generate a management plan for patients with tetralogy of Fallot who also have endocarditis.

DIFFERENTIAL DIAGNOSIS

Fever without a source on physical examination in a patient with cardiac hardware or devices should always raise the possibility of endocarditis. In this scenario, the endocarditis was localized to the pulmonary conduit and pulmonary valve, and it had

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Informed consent was obtained for this case.

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ABBREVIATIONS AND ACRONYMS

AICD = automated implantable cardioverter-defibrillator

ECMO = extracorporeal membranous oxygenation

ICU = intensive care unit

LV = left ventricular

PVR = pulmonary valve replacement

RVOT = right ventricular outflow tract

RV-PA = right ventricle-to-pulmonary artery

VSD = ventricular septal defect

spread towards the tricuspid valve and AICD leads. A thorough history of exposures may lead to the diagnosis of endocarditis with rare organisms.

INVESTIGATIONS

Echocardiography was significant for conduit vegetations. Blood culture results were persistently positive. The source was thought to be the hemodialysis catheter. Several echocardiograms also demonstrated depressed biventricular function during his hospitalization, which gradually improved. Pulmonary angiography showed free pulmonary valve insufficiency (**Figure 1**). The cause of the right-sided heart dysfunction was likely related to the pulmonary valve stenosis, pulmonary valve insufficiency, and previous RVOT reconstruction.

MANAGEMENT

He was referred for redo surgery involving the removal of his infected AICD leads, removal of his vegetations, excision of his entire pulmonary valve, and recovery on extracorporeal membranous oxygenation (ECMO) with free pulmonary insufficiency after difficulty separating from cardiac bypass. The surgery was technically challenging from a sternal re-entry standpoint because there was very dense scar. A new pulmonary valve was technically impossible at the time of surgery because the tissue was thought to be not sufficiently robust to hold new suture. There was also a risk of immediate endocarditis of a new prosthesis. He survived the operation and returned to the intensive care unit (ICU) with an open chest. The patient underwent multiple rounds of resuscitation from ventricular fibrillation and tachycardia both in the operating suite and in the ICU postoperatively, and he needed to recover from a state of stunned myocardium. These issues were thought to be related to his scar tissue and his depressed systolic function, after consultation with electrophysiology. Despite the volume loading of acute free pulmonary insufficiency, standard ECMO management was performed in the ICU with inotropic support. ECMO was chosen above the Impella device (Abiomed, Danvers, Massachusetts) because of the relatively new status of the RV Impella device at the time and the need to support right-sided heart dysfunction. He was hemodynamically stable enough to sustain traditional weaning from ECMO. He returned to the operating room 2 days later for chest closure, ECMO decannulation, and placement of a new subcutaneous AICD. Renal function was maintained with peritoneal hemodialysis, and later hemodialysis was performed through his fistula under the guidance of the infectious disease and nephrology staff. He awaited sterilization of blood cultures before later transcatheter PVR. Heart transplantation was not considered because of the need for simultaneous cardiac and renal transplantation and the possibility of being unable to clear the patient's endocarditis. He successfully recovered from this admission and was discharged for a later planned readmission for transcatheter PVR.

During the second admission, the technical aspects of transcatheter PVR were uneventful (**Figure 2**). However, he required resuscitation from multiple episodes of unstable ventricular arrhythmias. He had cardiogenic shock with ejection fractions as low as 5%, requiring high degrees of inotropic support. After consultation with electrophysiology, this response was again thought to be caused by his depressed ventricular function and multiple scars. He also needed continuous renal replacement therapy. He experienced shock liver and severe pulmonary edema. Because he has significant abdominal ascites, he was given a peritoneal dialysis catheter, but this, too, became infected.

After a 2-month hospitalization, the patient survived and was discharged with normal LV systolic function and a properly functioning pulmonary valve. Despite his multiple episodes of unstable ventricular arrhythmias, he survived with no neurological sequelae. Because he did not have any significant recurrences of ventricular arrhythmia after hospitalization, ablation was not performed.

DISCUSSION

Tetralogy of Fallot refers to 4 defects: a VSD, an overriding aorta, RV hypertrophy, and varying degrees and locations of pulmonary stenosis. If a shunt was the initial surgery, the patient will require complete repair later (1). If a pulmonary valve transannular patch was used to relieve valvular stenosis by augmenting the diameter of the valve, the patient will likely need a new pulmonary valve later on. In those patients who require minimal RVOT reconstruction and who retain a normally functioning native pulmonary valve, clinic follow-up is generally uneventful. A significant proportion of patients with tetralogy of Fallot will develop progressive RV dysfunction from residual pulmonary insufficiency or stenosis. By the time of his presentation, our patient already had RV dysfunction, which was likely multifactorial. He had at least 3 risk factors for persistent RV dysfunction, including pulmonary stenosis, pulmonary

FIGURE 1 Pulmonary Angiogram Demonstrating Free Pulmonary Valve Insufficiency

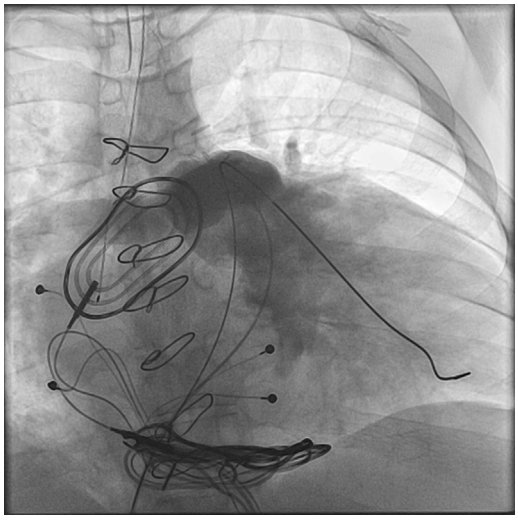
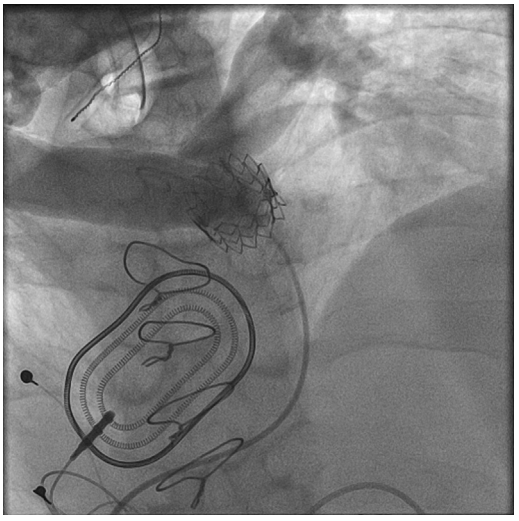


FIGURE 2 Successful Percutaneous Pulmonary Valve Placement Within the Surgical Conduit



insufficiency, and the need for an RVOT reconstruction (2). Although ECMO is rarely used in postoperative adult congenital heart surgery (3), it is currently unclear whether ECMO is helpful for the acute volume load in pulmonary insufficiency. This patient required multiple reinterventions for conduit stenosis and insufficiency. He also developed endocarditis. He was in a complicated situation because of his need for urgent cardiac surgery to correct endocarditis in a high-risk multiple redo sternotomy, as well as the need not to reinfect a valved conduit immediately. Hence, the decision was made to place a valveless conduit and allow him cardiac rest on ECMO before later admission for transcatheter PVR.

FOLLOW-UP. At his most recent clinic visit, he was recovering well from a cardiac standpoint and had been following along with his baseline dialysis regimen. He has regained much of his original body weight and can do regularly scheduled physical therapy. He sustained no significant neurological sequelae.

CONCLUSIONS

Patients with tetralogy of Fallot who have received pulmonary valve transannular patches to relieve valvar pulmonary stenosis are very likely to require future reintervention on their pulmonary valves. If pulmonary artery intervention was required, the patient could also require percutaneous or surgical intervention for restenosis of prior surgical repairs. Our patient experienced multiple adverse cardiac events, up to and including pulmonary conduit with AICD endocarditis. The difficulty in decision making came when the patient had active endocarditis but no means to confirm that the patient would not immediately reinfect a new conduit. The surgical team decided, in this case, to allow cardiac rest on ECMO and later planned readmission for percutaneous PVR. With the assistance of inotropic support, the patient demonstrated an improvement of both his LV systolic dysfunction and the ability to sustain some RV function in the presence of acute pulmonary insufficiency. This case overall serves to illustrate the feasibility of percutaneous PVR after valveless conduit placement with the assistance of ECMO for right-sided heart dysfunction in desperately ill patients with refractory conduit endocarditis.

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