We report an extremely rare case of deformity of the pulmonary sinus of Valsalva with pulmonary valvular stenosis 42 years after a pulmonary annular-sparing operation for tetralogy of Fallot. Aortic regurgitation with deformity of the sinus is also noted. At the previous operation, the right ventricular outflow tract was augmented by a prosthetic subvalvular patch. Through the years, the pulmonary valve and sinus were distorted because the patch was pulled over toward the right ventricle.

Keywords: Tetralogy of Fallot, pulmonary valve, aortic valve, imaging, long term

Introduction

The long-term results of complete repair of tetralogy of Fallot (TOF) have been excellent. However, some patients require reoperations due to aortic regurgitation (AR) as well as pulmonary regurgitation (PR) and/or stenosis (PS). AR caused by aortic root dilatation has been increasingly recognized as a feature of TOF. Treating PR and/or PS is certainly the major aspect of TOF repair affecting long-term results. Preserving native pulmonary valves and avoiding the use of a transannular patch might be important to prevent PR. We report a case of PS and AR 42 years after the corrective repair of TOF. Our patient developed valvular PS in conjunction with deformity of the pulmonary sinus of Valsalva, although the right ventricular outflow tract (RVOT) repair was originally performed with a subvalvular patch technique.

Case Report

A 58-year-old man, who had TOF repair performed in 1967 when he was 16 years old, presented with severe congestive heart failure (New York Heart Association class III). He has not been followed by any hospitals for a long time. A transthoracic echocardiogram demonstrated severe AR due to a prolapsed right coronary cusp with a poor left ventricular ejection fraction of 23% (left ventricular diastolic and systolic dimension were 52 mm and 47 mm, respectively) as well as an immobile, thickened valve, causing stenosis and regurgitation from the anterior aspect of the pulmonary artery. The posterior pulmonary valve was functioning well, with normal thickness and size. The condition of the subaortic ventricular septum was compatible with patch closure of the ventricular septal defect (VSD). Cardiac catheterization demonstrated severe AR due to a prolapsed right coronary cusp with a poor left ventricular ejection fraction of 23% (left ventricular diastolic and systolic dimension were 52 mm and 47 mm, respectively) as well as an immobile, thickened valve, causing stenosis and regurgitation from the anterior aspect of the pulmonary artery. The posterior pulmonary valve was functioning well, with normal thickness and size.
magnetic resonance imaging did not show evidence of left ventricular fibrotic changes. Coronary angiography revealed no abnormal finding.

Upon operation, a thickened valve consistent with the bicuspid anterior pulmonary valve was found under the cranial edge of the severely calcified RVOT patch. The patch demonstrated no aneurysmal change. No patch was identified on the pulmonary artery. Pulmonary valves were dissected out, and a 27-mm Medtronic freestyle porcine stentless valve (Medtronic Inc, Minneapolis, MN) was implanted. Aortic valve was dissected out and the top of the VSD closure patch was adhered to the right coronary cusp as preoperatively detected from MDCT images (Fig. 2). Aortic valve replacement was then performed with a 27-mm St. Jude mechanical bileaflet valve (St. Jude Medical Inc., Minneapolis, MN), as well as tricuspid annuloplasty with a 28-mm MC" annuloplasty ring (Edwards Lifesciences, Irvine, CA). The thickened valve was histologically confirmed as a native pulmonary valve, and the patch was also confirmed as pericardium. There were no serious complications after the operation, and the patient was discharged from the hospital. A transthoracic
echocardiogram 6 months after surgery demonstrated an improved left ventricular ejection fraction of 58%.

**Discussion**

Long-term survival after corrective surgery for tetralogy of Fallot (TOF) has been excellent since the technique was first implemented.\(^1\) We report a case of valvular PS in conjunction with AR 42 years after the repair of TOF (performed in 1967). The patient demonstrated severe congestive heart failure and was found to have severe AR due to a prolapsed leaflet. AR after the repair of TOF is not a common problem but is serious when it does occur.\(^2\) Previous studies\(^2\) have found a correlation between AR and aortic root dilation or dilated aortic sinuses. Aortic valve-sparing root operations are probably the ideal operations for young patients. The aortic root was normal in size in our case, but MDCT images clearly demonstrated that the aortic sinus of Valsalva was crushed. This deformity of the sinus probably led to prolapse of the aortic valve. Another reason of the prolapse is the relationship to the patch closure of ventricular septal defect as the patch was adhered to the cusp.

Late PR with right ventricular dysfunction is also a major problem after the corrective repair of TOF; the optimal timing and indications for surgical intervention related to the pulmonary valve remain controversial.\(^4\) Late development of PR is closely related to the usage of a transannular patch.\(^3\) The transannular patch technique and PR are also related to the development of ventricular tachycardia and sudden death.\(^5\) Various materials have been used for RVOT reconstruction to achieve successful long-term results, i.e., avoiding PR and consequent right ventricular failure.\(^6\) We have designed a fan-shaped expanded polytetrafluoroethylene-valved conduit and patch with bulging sinuses for greater long-term durability and reliability.\(^6\) These valved conduits and patches have functioned well, retaining low-pressure gradients at the valves and minimum PR for up to 5 years. To prevent delayed pulmonary problems, many surgical modifications for preserving the pulmonary valve as well as pulmonary annulus have also been developed.\(^3\)\(^,\)\(^7\) Stewart and associates\(^3\) assert the importance of annular preservation; therefore, they use two patches above and below the annulus instead of a transannular patch for cases in which an infundibular patch is required. In our case, the infundibular patch enlargement was performed adjacent to the native pulmonary annulus, and many years later, the degeneration of a patch without growth potential pulled the pulmonary annulus toward the ventricular apex side. These factors deformed the anterior pulmonary sinus of Valsalva. Long-term follow-up for PS at the valvular position after the repair of TOF is rare and required for individuals with congenital bicuspid valves.\(^8\) The large bicuspid pulmonary anterior valve showed severe degenerative change and developed PS in our case.

Although the long-term results of complete repair of TOF have been excellent,\(^1\) many problems such as right ventricular failure and PR secondary to RVOT repair, AR secondary to aortic root native problems, and arrhythmias develop slowly with time. Regular long-term follow-up is required for patients who have undergone TOF repair.

**Acknowledgment**

The authors acknowledge Mr. Takaaki Kitai, Mr. Katsunori Sakurai and Mr. Yoshinori Obata for their technical contributions.

**References**