Late Dynamic Right Ventricular Outflow Obstruction After the Ross Procedure for Bicuspid Aortic Valve Disease

Akhlaque Uddin, MBChB; John D.R. Thomson, MD; Sven Plein, PhD; John P. Greenwood, PhD

An asymptomatic 26-year-old man was under routine surveillance following a Ross procedure at 17 years of age for bicuspid aortic valve disease. During transthoracic echocardiography there was a suspicion of homograft stenosis following the finding of a high-velocity turbulent jet in the main pulmonary artery (MPA) (Figure 1A and 1B; online-only Data Supplement Movie IA and IB). The pressure gradient by ultrasound Doppler was calculated to be 60 mm Hg, but the cause could not be clearly elucidated. The ECG (Figure 2) showed sinus rhythm with a normal axis, normal P-wave morphology, and nonspecific interventricular conduction delay in V1-V2. Chest radiograph (Figure 3) did not reveal any distortion of the great vessels or show signs of heart failure.

A cardiovascular magnetic resonance (CMR) scan was organized to clarify the echocardiographic findings. CMR was performed on a Philips 1.5T scanner by use of standard T1-weighted black blood imaging, cines (steady-state free precession imaging), and phase-contrast velocity mapping. CMR revealed evidence of moderate right ventricular hypertrophy without evidence of chamber dilatation or impairment in function (Figure 4A; online-only Data Supplement Movie IIA). Turbulent high-velocity flow (4 m/s) was confirmed in the MPA, but there was no evidence of homograft stenosis or size mismatch. There was mild to moderate homograft incompetence with a calculated regurgitant fraction of 27% (Figure 4E). The turbulent high-velocity flow in the MPA was due to external compression by a large pulsatile mass measuring 5×3 cm (Figure 4B; online-only Data Supplement Movie IIB). Cine imaging in multiple planes identified a large false aneurysm arising from the left ventricular outflow tract (Figure 4C and 4D; online-only Data Supplement Movie IIC). The false aneurysm did not contain thrombus and appeared to originate from the suture line below the neoaortic valve. Of greater concern was the fact that the left main stem coronary artery occupied the potential space between the pulsatile false aneurysm and the MPA (Figure 4E). The neoaortic valve was functioning well with only a mild central jet of aortic incompetence (regurgitant fraction 10%) (Figure 4B).

In this case CMR was able to define exactly the late surgical complication of dehiscence at the proximal suture line of the autograft leading to false aneurysm formation. This was causing dynamic right ventricular outflow obstruction and moderate right ventricular hypertrophy. Because of the concern relating to compression of the left main stem, surgical correction was mandated. The patient underwent successful aneurysmectomy with repair of the left ventricular outflow tract and made an excellent recovery. Subsequent follow-up by CMR confirmed an excellent surgical result (Figure 5A and 5B; online-only Data Supplement Movie IIIA and IIIB).

The Ross procedure involves replacing the diseased aortic valve with the patient’s own pulmonary valve (autograft) to...
form the neoaortic valve, reimplanting the coronary arteries, and inserting a cadaveric homograft into the pulmonary position. This is a well-accepted strategy to manage aortic valve disease in children and young adults and is cost effective. The advantage of this technique is that, unlike mechanical aortic valve replacement, anticoagulation is not required. The disadvantage is that it converts single left-sided valvular disease into left and right heart valvular disease. Complications include neoaortic valve incompetence, aortic root dilatation, homograft stenosis, suture line aneurysms, and endocarditis. Late complications are rare, and aneurysm formation has been described but not to the extent of causing right ventricular outflow obstruction. Dilatation of the aortic autograft may be due to exposure to higher systemic arterial pressures in comparison with the lower right heart pressures. Alternatively, this dilatation may be due to an underlying preexisting aortopathy as seen in bicuspid aortic valve disease.

Transesophageal echocardiography may provide only limited information in complex right heart pathology and can be limited by patient tolerance during longer complex studies unless under general anesthesia. In this case, it would have been unlikely that transesophageal echocardiography could have determined the cause of external MPA compression. Thoracic computed tomography is an alternative modality for determining anatomic abnormalities, but would not have provided the functional information in relation to the dynamic nature of the right ventricular outflow tract obstruction or potential left main stem compression. Thus, we highlight the utility of CMR to noninvasively delineate complex anatomy and provide detailed functional information without exposure to ionizing radiation, which is important in young patients with congenital heart disease who will require life-long surveillance.

Disclosures
None.

References
Figure 4. A, CMR 4-chamber view showing a nondilated right ventricle with mild hypertrophy. B, Left ventricular outflow tract view showing the aortic root (Ao), the main pulmonary artery (p) being compressed from below by a false aneurysm (a); there is also a central jet of mild aortic regurgitation. C and D, Diastolic and systolic images (sagittal plane) showing the false aneurysm (a) compressing the main pulmonary artery (arrow) causing blood flow acceleration. E, This transverse plane shows the false aneurysm (a) wrapping around the left main coronary artery (arrow) which is being compressed onto the main pulmonary artery (p). F, Pulmonary flow analysis showing a forward volume of 95 mL and regurgitant volume of 26 mL yielding a regurgitant fraction of 27%. CMR indicates cardiovascular magnetic resonance.
Figure 5. A. Postoperative imaging showing the right ventricular outflow tract view in diastole with the obliterated cavity of the aneurysm and no further obstruction to the main pulmonary artery. B. The left ventricular outflow tract view shows the repaired site of the aneurysm and a normal pulmonary artery but the mild central aortic regurgitation persists.
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