Treatment of Aortic Homograft Dysfunction with Transcatheter Aortic Valve Implantation in a Young Patient

The introduction of transcatheter aortic valve implantation (TAVI) has transformed the treatment of aortic stenosis in patients at high risk of operative mortality, although it has been limited almost exclusively to elderly patients. (1, 2) This procedure has been performed mostly on native valves, and occasionally on heterologous bioprosthetic heart valves (valve-in-valve). In many patients, aortic root homograft (AH), used for surgical aortic valve replacement principally in infective endocarditis, is often at risk for calcification and structural deterioration, requiring replacement within 15 years of surgery. Valve dysfunction of AH, together with coronary ostium and aortic root calcification, represents a major challenge for the cardiovascular team.

We describe the clinical case of a 24-year-old male patient with history of congenital aortic stenosis, undergoing aortic valve repair with balloon at the age of one month, and aortic valve replacement (AVR) with mechanical prosthesis at 13 years of age. Five months after that procedure, he presented with early prosthetic endocarditis requiring further surgery with AH implantation. In 2015, the patient suffered new prosthetic endocarditis caused by Streptococcus spp., for which he received medical treatment. During the following year, the patient presented with progressive dyspnea in functional class (FC) III-IV and had several admissions due to heart failure in the context of aortic valve disease, with left ventricular enlargement and progressive deterioration of systolic function. Chest CT angiography and transesophageal echocardiography showed severe degeneration and calcification of the AH, severe stenosis and aortic valve regurgitation, and two images consistent with aseptic vegetations. The patient was considered inoperable in another medical center and cardiac transplantation was suggested, which was rejected by the patient and his family. The patient was evaluated by the interdisciplinary cardiovascular team of our institution, which ruled out surgical AVR due to the high risk of a third reoperation (Euroscore II 10.5%, STS 30.4%) and extended calcification of the homograft. A transfemoral TAVI was therefore decided using a repositionable self-expanding valve. Given the patient's small body structure and small femoral arteries, implantation with a 23 mm Lotus™ Valve System (Boston Scientific™) under general anesthesia and without predilatation was the preferred option. Echocardiographic parameters and symptoms improved immediately after the operation; the patient progressed with no postoperative complications, and was discharged 48 hours after the procedure. At the 6-month follow up, the patient remains asymptomatic, with good exercise tolerance (FC I), no signs of pump failure and echocardiographic improved left ventricular function, normal valve function, and absence of paravalvular leak.

Surgical aortic root and valve homograft implantation is used mainly in cases of extended endocarditis of the valve with root destruction. Structural deterioration of the AH is the main reason for reoperation. According to the different series, the rate of repeat intervention at 10 years in patients below 20 years of age is almost 20 times greater than in patients aged over 65 years. (3) Calcification is the major structural alteration causing AH dysfunction, both of the aortic root and leaflets. As a result, insufficiency and/or aortic valve stenosis often occur, and conventional surgery is the treatment of choice. Reoperation is a complex and atypical procedure with high technical difficulty, resulting in approximately 50% morbidity rate. However, surgical risk can be reduced with new technologies like TAVI or sutureless aortic valve replacement. (4) The latter option was not feasible for our patient because we did not have the Perceval Sorin™ System (sutureless valve with minimal surgical approach and short cardiopulmonary bypass times) available in our country.

Although the experience is limited, small series of patients with AH dysfunction and high surgical risk, which have been safely and efficiently treated with TAVI, have been reported to date. In most cases, a CoreValve was used. (5) The uniqueness of the present case lies in the indication of TAVI with a Lotus™ valve in a young patient who had AH dysfunction due to severe stenosis and insufficiency, with a history of two previous surgeries and porcelain aorta.

Some technical aspects of the procedure should be considered. A series of previous and intraprocedural assessments are necessary to adapt the TAVI technique used for aortic stenosis. CT angiography is

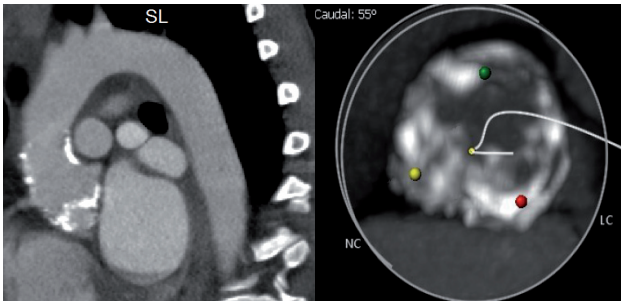

Fig. 1. *Left,* CT angiography showing severe homograft calcification, including annulus and valve. *Right,* cross sectional images of aortic valve calcifications.

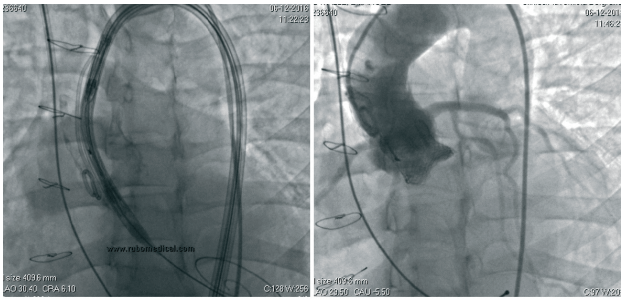

Fig. 2. Implantation through the left femoral artery showing the temporary pacemaker in the right ventricle and the 23 mm Lotus™ Valve System (Boston Scientific). No transvalvular gradient or aortic regurgitation were recorded in the angiogram.

very valuable for correct AH assessment, particularly its degree of calcification and the implantation area where the prosthesis will be fixed. Moreover, it allows establishing the anatomy and angulation of the aortic root, and its association with both coronary ostia, or the presence of bypass. (6) It is also important to assess the region of anastomosis with the ascending aorta, which can be stenotic or restrictive, hampering an appropriate expansion of the prosthetic valve. (3) Measuring valve annulus is of vital importance when choosing the size of the prosthesis to reduce the risk of significant paravalvular regurgitation (Figure 1). It should be taken into account that the degeneration of the AH may cause anatomical distortion and aortic root dilation. In case of aortic regurgitation, balloon dilation prior to implantation should not be performed because it may cause aortic insufficiency and acute hemodynamic deterioration. Correct positioning and deployment of the prosthetic valve are a challenge due to few anatomic landmarks in the fluoroscopy image and to severe valve regurgitation causing device instability. Transesophageal echocardiography is used to guide the positioning of the valve, and it is often supplemented by contrast injections in the aortic root. Valve stability during deployment may be improved with rapid ventricular pacing to reduce pressure and regurgitant anterograde flow in each beat may increase (Figure 2). The risk of coronary ostia obstruction should be evaluated in each patient, which increases in case of low-lying, rigid coronary ostia and less capacious sinuses of Valsalva. (5)

The choice of a repositionable self-expanding Lotus™ prosthesis facilitated adequate positioning, hemodynamic stability during deployment, correct functioning since implantation, and minimal paravalvular regurgitation. Prosthesis repositioning was unnecessary in our patient. TAVI results at one-year follow-up with this type of prosthesis have been published, and show excellent valvular hemodynamics, absence of moderate or severe paravalvular regurgitation, and significant and sustained improvement of functional capacity, with good clinical outcome. It has also been associated with permanent pacing in 33% of the patients, which was not necessary in our case.

Surgery is the treatment of choice in case of AH failure, but in patients with extensive calcification, the only option is to remove and replace it. This type of surgery is technically challenging, with a significant morbidity and mortality rate. In this context, TAVI can be an option in high risk patients. Although long-term durability of TAVI devices is not well-known, we believe it is an option even for young patients when the risk for surgery is very high.

Conflicts of interest

None declared.

(See authors' conflicts of interest forms on the web/Supplementary material).

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Retrospective Diagnosis of Takayasu's Arteritis from a ST-Segment Elevation Acute Coronary Syndrome

Takayasu's arteritis is a chronic idiopathic granulomatous vasculitis with a wide clinical variety, since it affects the aortic arch and its main branches. Its first description dates back to 1908 in Japan, where the highest incidence rates are reported today (1/3,000 inhabitants). In the West, its diagnosis is difficult because it is a rare condition, with an incidence of 1.2

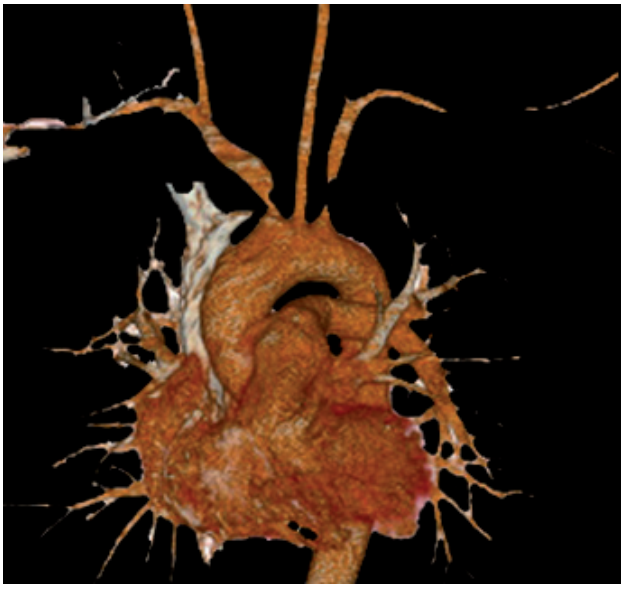

Fig. 1. CT angiography with 3D reconstruction showing stenosis of the great vessels. Image at the time of diagnosis.

cases/1,000,000 inhabitants and an 8:1 female/male ratio.

We describe the case of a 37-year-old male patient with a history of hypertension since adolescence, smoking, dyslipidemia, sedentarism and prior hospitalization due to amaurosis fugax and right facial paresthesia interpreted as complex migraine.

The patient was admitted to our Department of Cardiology with a diagnosis of inferior ST-segment elevation acute coronary syndrome with electrical involvement of the right ventricle and Killip & Kimball A classification. Physical examination revealed right carotid and left subclavian murmur, asymmetric pulse in upper limbs, claudication and unrecordable blood pressure (BP) in the left upper limb, and differential BP of 20 mm Hg between upper and lower limbs.

Coronary angiography revealed a severe lesion in the proximal third of the anterior descending artery (ADA) and in its diagonal branch, moderate lesion in the proximal third of the circumflex artery (Cx) with significant lesion in its second lateral branch, and occlusive lesion in the mid-third of the right coronary artery (RCA), in which primary angioplasty with two stents was performed because it was considered the responsible artery.

Doppler echocardiography showed severe hypokinesis of the inferior left ventricular segments.

Due to the peculiar physical examination in a young patient, a CT angiography was performed, showing 52% stenosis in the brachiocephalic trunk (BCT) ostium, 77% in the left subclavian artery (LSA) and 38% in the superior mesenteric artery ostium (Figure 1).

As a result of these findings and of clinical suspicion, the diagnosis of Takayasu's arteritis was made, based on the American College of Rheumatology (1) and Ishikawa's (2) criteria. (2) It was therefore decided, together with the Department of Rheumatology, to start treatment with intravenous methylprednisolone and then with cyclophosphamide. (3)

The patient had an asymptomatic course and continued on outpatient treatment.

A reduction in the percentage of the involved artery obstruction was observed during follow-up. Follow-up CT angiography revealed 47% stenosis in the BCT ostium, and 62.5% in the LSA (Figure 2). Coronary angiography after 6 months of treatment also showed reduction in the following lesions: moderate lesion in the proximal third of the ADA, non-significant lesion in the lateral branch of the Cx, and RCA without stenosis in mid-third stents.

At present, the patient remains asymptomatic and continues under follow-up.

The association between early atherosclerosis and common rheumatic diseases such as systemic lupus erythematosus and rheumatoid arthritis is well known; therefore, cardiac manifestations may occur first and the cardiologist may be the first specialist to evaluate these patients. However, since Takayasu's arteritis is a rare disease in Argentina, it is difficult to be diagnosed, and hence there is scarce local literature. According to the American College of Rheumatology, (1) the presence of at least three criteria has a sensitivity of 92.1% and a specificity of 97% to diagnose the disease. Our patient met 6/6 criteria for the diagnosis of Takayasu's arteritis. The relevance of our case is based on the fact that it is extremely rare in our ethnicity, and even rarer in men. Moreover, initial presentation of Takayasu's arteritis with acute myocardial infarction is absolutely unusual. (5)

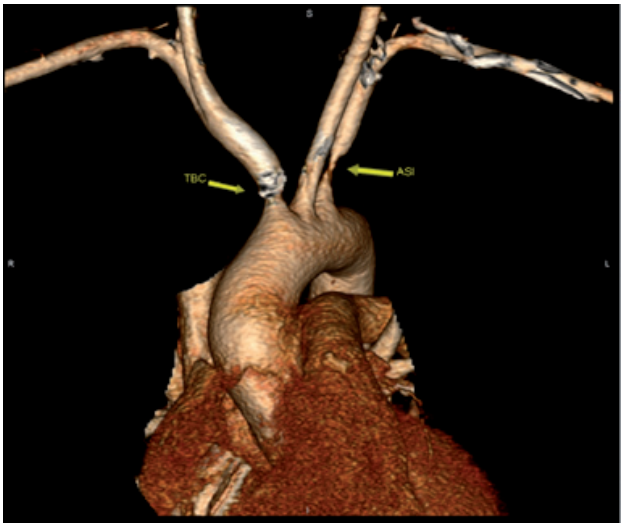

Fig. 2. Follow-up CT angiography at 6 months of treatment.

Conflicts of interest

None declared.
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