Transcatheter aortic valve implantation in patients with bicuspid aortic valve

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Introduction

Bicuspid aortic valve (BAV) is one of the most common congenital heart defects with a population prevalence of 0.5–1.3% [1,2]. BAV has been identified as the main cause of aortic valve disease leading to surgical treatment in children and young adults. A large pathological survey revealed that BAV could result in a stenotic lesion in 75% of patients and insufficiency in 15% [3].

In the aspects of anatomy, compared to a normal tricuspid aortic valve (TAV), a BAV is formed with only two as a result of the fusion of two leaflets into a larger one. Although BAV is often considered to be a benign lesion early in life, the complications associated with cardiovascular diseases, including aortic stenosis (AS), aortic insufficiency (AI), infective endocarditis (IE), and aortic dilation and dissection, can result in marked increases in morbidity and mortality later in life [3–5]. There have been many surgical techniques and different therapeutic options for patients presenting with BAV stenosis with or without regurgitation [6]. In clinical practice, at least 30% of patients with severe symptomatic aortic stenosis do not undergo surgery for replacement of the aortic valve, owing to advanced age, left ventricular dysfunction, or the presence of multiple coexisting conditions [7,8]. With the advent of transcatheter valves specifically for minimally invasive implantation procedures, AS patients at high risk with conventional surgery have benefited with transcatheter aortic valve implantation (TAVI). Nevertheless, BAV is generally considered to contraindicate TAVI in most randomized controlled trials [8,9], because of the poor stability of the prosthetic valve or paravalvular regurgitation due to non-ideal expansion of a valve in elliptical and calcified annulus. Not only may the nonstandard shape and geometry of bicuspid valves predispose backflow leak during ventricular diastole after TAVI, but also the asymmetric dilatation resulting with irregular distribution of calcium deposits on the annulus of the BAV increase the risk of incomplete
sealing, severe paravalvular leak and aortic regurgitation (AR), the complications which already exists as the major drawback of TAVI technology [10,11]. Here, we detailed a case of successful trans-femoral TAVI in a 77-year-old male with BAV.

Case report

We present a 77-year-old male with symptomatic BAV stenosis. The patient was admitted with progressive dyspnea (NYHA IV) for 1 year in December 16, 2014. The ECG findings included atrial fibrillation rhythm, heart rate of 150 beats/min, ST depression, and T inversion in leads I, AVL, and V1–V6. Echocardiography revealed BAV with severe AS and moderate thickening and calcification and trivial aortic insufficiency. Baseline transthoracic echocardiography reported an aortic valve area of 0.82 cm², mean gradient of 41.0 mmHg, mean velocity of 312 cm/s. Left ventricular ejection fraction was 48%. At the same time, aortic valve area index (AVAI) estimated approximately 0.51 cm²/m², which was an indicator of severe AS.

He was declined for surgery on account of high operative risk (logistic EuroSCORE 26.03%, STS estimated Morbidity or Mortality 25.84%) after consultations by the multidisciplinary heart team (consisting of interventional cardiologists, cardiac surgeons, anesthesiologists, and imaging specialists), thus he was evaluated for TAVI. But we underlined that BAV is currently considered a contraindication and the procedure could be considered only after a careful examination with transesophageal echocardiography (TEE), multidetector computed tomography (MDCT) and coronary and peripheral angiography. He accepted the risks and signed an informed consent. On his coronary angiography in December 19, 2014, significant luminal narrowing (80%) of distal LM to opening of LAD (90%), tight stenosis (95%) at D1 and intermediate disease (60%) at proximal RCA were observed. LM to proximal LAD was pre-dilated and 3.0 mm × 36 mm EXCEL Stent was successfully deployed at LM to LAD (Fig. 1). After post-stenting, high pressure balloon dilatation with a Voyager NC balloon 4.0 mm × 8 mm at LM to pLAD (Fig. 1).

The patient’s symptoms subsequently improved from NYHA class IV to class III. TEE and computed tomographic angiography (CTA) confirmed the bicuspidy with a severe horizontal angulation (Fig. 2) and with an annulus diameter ranging from 24.3 to 28.3 mm (26.3 mm [Mean]), an annulus perimeter of 82.5 mm (annulus perimeter derived diameter, 26.2 mm), an ascending aorta diameter of 37.1 mm. Aortoiliac CTA detected normal lumen and adequate sizes of the iliac and femoral arteries. There was no significant stenosis or calcification in the iliac or common femoral artery. A 29-mm CoreValve (Medtronic Inc., Minneapolis, Minnesota, USA) implantation via trans-femoral access was decided upon. General anesthesia, TEE, pre-implantation balloon aortic valvuloplasty, and rapid ventricular pacing were carried out as a routine standard protocol. Arteriotomy was performed to obtain

Fig. 1 – Coronary angiography images before (left), post-stenting high pressure balloon dilatation (middle) and after (right) stent deployment.

Fig. 2 – TEE (left) and CTA (middle) showing the congenital bicuspid aortic valve with a severe horizontal angulation (right). Middle and right pictures from Medtronic company.
TAVI in patients with bicuspid aortic valve

an appropriate femoral access. Due to the patient had a relative large, congenital bicuspid aorta root anatomy with a severe horizontal angulation, we draw assistance from snare (in the left common femoral artery) to perform TAVI (Fig. 3). The snare has grabbed the CoreValve to ensure valve is aligned within the annulus and perpendicular to the basal plane. Pre-implantation balloon valvuloplasty was done with a 22 mm × 40 mm sizing balloon. Full balloon expansion could be achieved. The patient did not develop significant aortic incompetence after balloon valvuloplasty. The angiographic view, in which the three cusps are aligned, was obtained and the valve was deployed. The above-mentioned valve was thereafter successfully implanted (implant depth was 4 mm) without misplacement nor paravalvular leak, and with complete circular valve expansion confirmed by intraoperative TEE and fluoroscopy (Figs. 4 and 5). Post-TAVI mean gradient decreased strikingly to 9 mmHg, and peak velocity to 2.1 m/s. The patient was monitored in an intensive care unit for two days. No conduction abnormalities were observed on ECG. The patient’s symptoms subsequently improved from NYHA class III to class I. On day 5 post-procedure, the patient was discharged without any significant complications.

Discussion

Patients with an estimated mortality risk >20% by logistic EuroSCORE or >10% by the Society of Thoracic Surgeons score system are generally considered candidates for the TAVI procedure. BAV is generally considered to contraindicate transcatheter aortic valve implantation (TAVI) because of the poor stability of the prosthetic valve or paravalvular regurgitation partly due to non-ideal expansion of a valve in elliptical and calcified annulus [12].

Extensive calcium deposition in the body of BAV leaflets and asymmetrical nature of the bicuspid aortic root could impair TAVI outcomes. Many studies and some case reports have demonstrated a positive effect of TAVI when used on BAV patients whose condition is inoperable or has a high surgical risk [13–15]. By far, paravalvular leak is the major concern for TAVI in patients with BAV, owing to incomplete prosthesis apposition resultant from increased calcium or annular eccentricity [15]. Yousef et al. noted a 9.6% incidence of ≥3+ AR with incidence rising as high as 30.8% when including all patients with ≥2+ AR [15]. Such higher rates of paravalvular leak in this study could be related to the inherent aortopathy as-
associated with BAV, but also can be attributed to lower rates of MSCT performed (62.1%) pre-TAVI for sizing of the valve [15]. Close preoperative and intraoperative analyses of the aortic valve anatomy are mandatory for successful TAVI, especially in BAV cases. Since MSCT-based tricuspid aortic valve (TAV) sizing was clearly associated with reduced para-valvular regurgitation, MSCT should be considered a mandatory element of patient screening for TAV-in-BAV, certainly in view of the suboptimal echocardiographic results. Consequently, it was necessary to consider the characteristics of the current artificial aortic device. We should avoid the situation such as the artificial valve extension into the left ventricular outflow tract, artificial valve blocking the opening of the coronary artery and so on. In order to make the device better anchored in the aortic annulus, we performed MSCT to reconstruct the measurement of aortic root and combine with TEE, make evaluation of left ventricular outflow tract, aortic annulus, aortic sinus, the distance of coronary artery opening and ascending aorta. In addition, it is difficult to dilate the bicuspid valve into a completely circular shape due to the morphology of BAV itself. Therefore, the Medtronic Core Valve system (MCS), which incorporates a self-expanding valve, has been suggested due to sufficient radial strength for a bicuspid valve.

In this case, a 29-mm CoreValve was used and significant clinical improvement was achieved, and MPg decreased from 41 mmHg to 9 mmHg. In our patient, there was no obvious paravalvular leakage on intraoperative TEE and on TTE in follow-up. The recent study by Mylotte et al. [14] is the first large multicentre analysis of TAV implantation in patients with significant BAV stenosis or regurgitation. TAV-in-BAV proved to be feasible with encouraging short- and intermediate-term clinical outcomes, but the relatively high incidence (28%) of post-implantation aortic regurgitation is of serious concern. Therefore, longer-term follow-up of a larger cohort of patients is required to more completely assess the efficacy and durability of TAV implantation in patients with bicuspid disease.

Our case demonstrates that TAVI using a CoreValve can be performed safely in patients with symptomatic BAV stenosis.

Conflict of interest
None declared.

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Ethical statement
Authors state that the research was conducted according to ethical standards.

Informed consent
Informed consent was obtained from the patient participating in this study.

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