ESC European Society of Cardiology

European Heart Journal - Case Reports (2020) **4**, 1–6
doi:10.1093/ehjcr/ytaa102

CASE REPORT
Congenital heart disease

Transcatheter aortic valve replacement for bicuspid aortic valve regurgitation in a 17-year-old patient with congenitally corrected transposition of great arteries: a case report

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Received 18 January 2019; first decision 20 February 2019; accepted 9 April 2020; online publish-ahead-of-print 8 May 2020

Background Limited research has been conducted on the surgical management of the aortic valve in congenitally corrected transposition of great arteries (ccTGA) and to our knowledge there have been no reports on the treatment of bicuspid aortic regurgitation (AR) in ccTGA. We report on a ccTGA patient with bicuspid AR and systemic right ventricule (SRV) dysfunction who underwent transcatheter aortic valve replacement (TAVR).

Case summary A 17-year-old male with a history of ccTGA and cerebral palsy diagnosed at birth presented with heart failure. During childhood, he did not experience any heart failure symptoms, however, secondary to progressive bicuspid AR he experienced worsening SRV dysfunction beginning at 15-year-old. Echocardiography showed reduced SRV ejection fraction and severe bicuspid AR. The heart team, including a cardiac surgeon and paediatric cardiologist, discussed the treatment strategies and decided to proceed with TAVR as surgical aortic valve replacement was deemed high risk. TAVR was performed with the 34 mm Evolut R (Medtronic, Minneapolis, MN, USA). Post-operative echocardiography showed severe paravalvular leak (PVL). Therefore, valve-in-valve TAVR using a 29 mm Edwards SAPIEN 3 (Edwards Lifesciences, Irvine, CA, USA) was performed on post-operative Day 2 for PVL reduction. Following second procedure, PVL was significantly improved. The patient was discharged in stable condition.

Discussion This is the first case wherein TAVR was performed for bicuspid AR in a patient with ccTGA. With appropriate preparation and planning and a collaborative multi-disciplinary team approach, TAVR can be a treatment option for severe AR in patients with ccTGA at high risk for surgery.

Keywords Case report • Transcatheter aortic valve replacement • Congenitally corrected transposition of great arteries • Bicuspid aortic valve • Aortic regurgitation

Learning points

- We report the case of a surgical high-risk congenitally corrected transposition of great arteries (ccTGA) patient who underwent transcatheter aortic valve replacement (TAVR) for bicuspid aortic regurgitation (AR).
- As post-operative echocardiography demonstrated severe paravalvular leak (PVL), valve-in-valve TAVR was performed on post-operative Day 2 for the purpose of PVL reduction. Following second procedure, PVL was improved.
- With appropriate preparation and planning and a collaborative multi-disciplinary team approach, TAVR can be a treatment option for severe AR in patients with ccTGA at high risk for surgery.

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Handling Editor: Georg Goliasch

Peer-reviewers: Maria Antonieta Albanez A de Medeiros Lopes and Dejan Milasinovic

Compliance Editor: Christian Fielder Camm

Supplementary Material Editor: Peregrine Green

Introduction

Congenitally corrected transposition of great arteries (ccTGA) is a rare disease. The frequency is reported to be 1/33 000 live births, accounting for ~0.05% of congenital heart disease.[1] It has been reported that 84% of cases of ccTGA are associated with other cardiac defects.[2] Common defects associated with ccTGA are ventricular septal defect (VSD), pulmonary stenosis, tricuspid regurgitation, and Ebstein's anomaly of the systemic atrioventricular valve. Additionally, although infrequent, aortic regurgitation (AR) has been associated in 7% of patients with ccTGA.[2] These associated cardiac malformations may require surgical treatment depending on their severity, and systemic right ventricle (SRV) dysfunction is often associated with increased operative risk.[3] Herein, the authors report the case of a 17-year-old patient with ccTGA and SRV dysfunction who developed severe AR and was treated with transcatheter aortic valve replacement (TAVR) rather than surgical aortic valve replacement because of presumed elevated surgical risk associated with SRV dysfunction. Little information is available regarding the treatment of bicuspid aortic stenosis (AS) in patients with ccTGA. Only two case reports on such patients exist,[4,5] and there are no other studies describing AR in patients with ccTGA and bicuspid aortic valve. The authors believe that a detailed description of this case provides valuable information.

Timeline

Time	Events
Prior to presentation	17-year-old male Congenitally corrected transposition of great arteries Bicuspid aortic regurgitation Systemic right ventricle dysfunction Cerebral palsy
Day 0: transcatheter aortic valve replacement (TAVR)	TAVR using a 34 mm Medtronic Evolut R Post-operative echocardiography showed severe paravalvular leak (PVL)
Post-operative Day 2: valve-in-valve TAVR	Valve-in-valve TAVR using a 29 mm Edwards SAPIEN 3 for PVL reduction Following valve-in-valve procedure, PVL was significantly improved
Post-operative Day 4: patient discharge	The patient was discharged in stable condition

Case presentation

A 17-year-old male was diagnosed at birth with ccTGA, VSD, bicuspid aortic valve, and cerebral palsy. During childhood VSD closure occurred spontaneously and he was virtually asymptomatic until age 15. At that time, he was noted to have progressive bicuspid AR associated with SRV dysfunction. He developed worsening symptoms of heart failure despite the institution of medical therapy. He presented at our hospital for consultation on alternative treatment strategies for his AR. Physical examination revealed malaise and fatigue. His blood pressure was 102/49 mmHg. A grade 3/6 diastolic murmur was present. Water hammer wrist pulses were felt. Transthoracic echocardiography revealed an enlarged SRV with ejection fraction (EF) decreased to 28%. Aortic valve morphology was bicuspid, with severe AR. There was mild mitral regurgitation and mild-to-moderate systemic tricuspid regurgitation (TR). Transoesophageal echocardiography demonstrated severe eccentric AR (*Figure 1*) without an atrial- or ventricular-level shunt. Multidetector computed tomography showed an annulus mean diameter of 26.9 mm, annulus

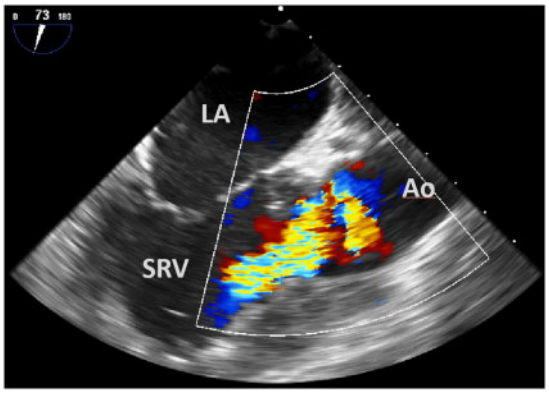

Figure 1 Baseline transoesophageal echocardiography, long-axis view. Severe eccentric aortic regurgitation. Ao, aorta; LA, left atrium; SRV, systemic right ventricle.

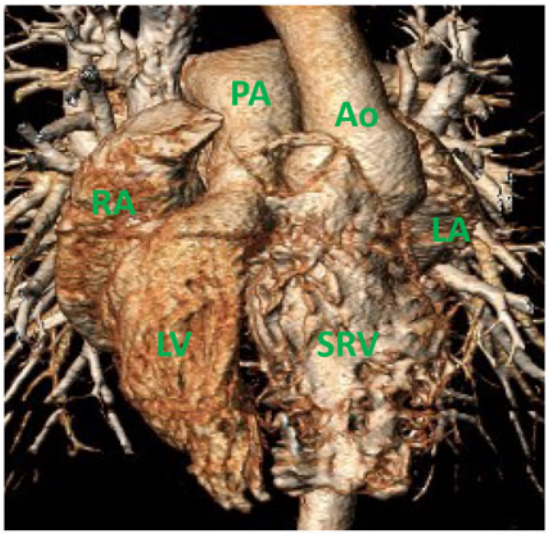

Figure 2 Multidetector computed tomography volume rendering image. Anatomical location relationship. Ao, aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; SRV, systemic right ventricle.

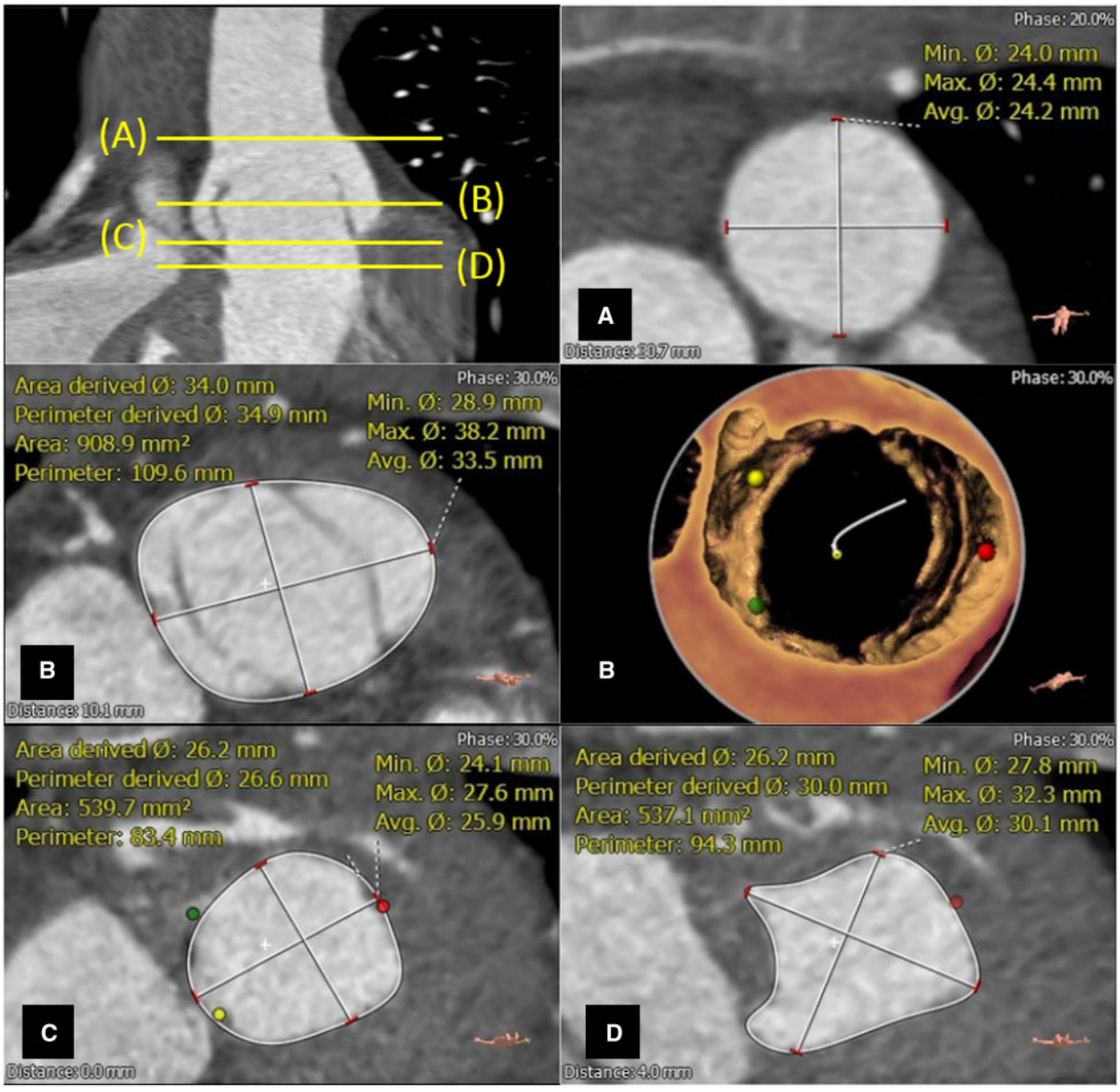

Figure 3 Multidetector computed tomography aortic valve complex analysis. (*A*) Sinotubular junction. (*B*) Sinuses of Valsalva. (*C*) Aortic annulus. (*D*) Systemic right ventricular outflow tract.

area of 506.3 mm^2, annulus perimeter of 80.9 mm, and sinus of valsalva area was 908.9 mm^2. The right coronary height was 18.8 mm, and the left coronary height was 27.5 mm. There was no calcification in the aortic valve complex. The SRV outflow tract had an irregular shape (*Figures 2 and 3*). The heart team, including an experienced cardiac surgeon and a congenital interventional cardiologist discussed the treatment strategy based on these results and ultimately decided to proceed with TAVR as surgical aortic valve replacement was deemed high risk. Under general anaesthesia, TAVR was performed using a transfemoral approach, and a 34 mm Medtronic Evolut R was implanted. Shortly after the procedure, post-operative echocardiography demonstrated severe paravalvular leak (PVL) necessitating a valve-in-valve TAVR using a 29 mm Edwards SAPIEN 3 performed on post-operative Day 2 for the purpose of PVL reduction (*Figure 4*, Supplementary materail online, *Video S1*). Following this second procedure, PVL was significantly improved (*Figure 5*). The final pre-discharge echocardiogram shows that the peak aortic valve jet velocity was 3.58 m/s, while the aortic valve mean pressure gradient was

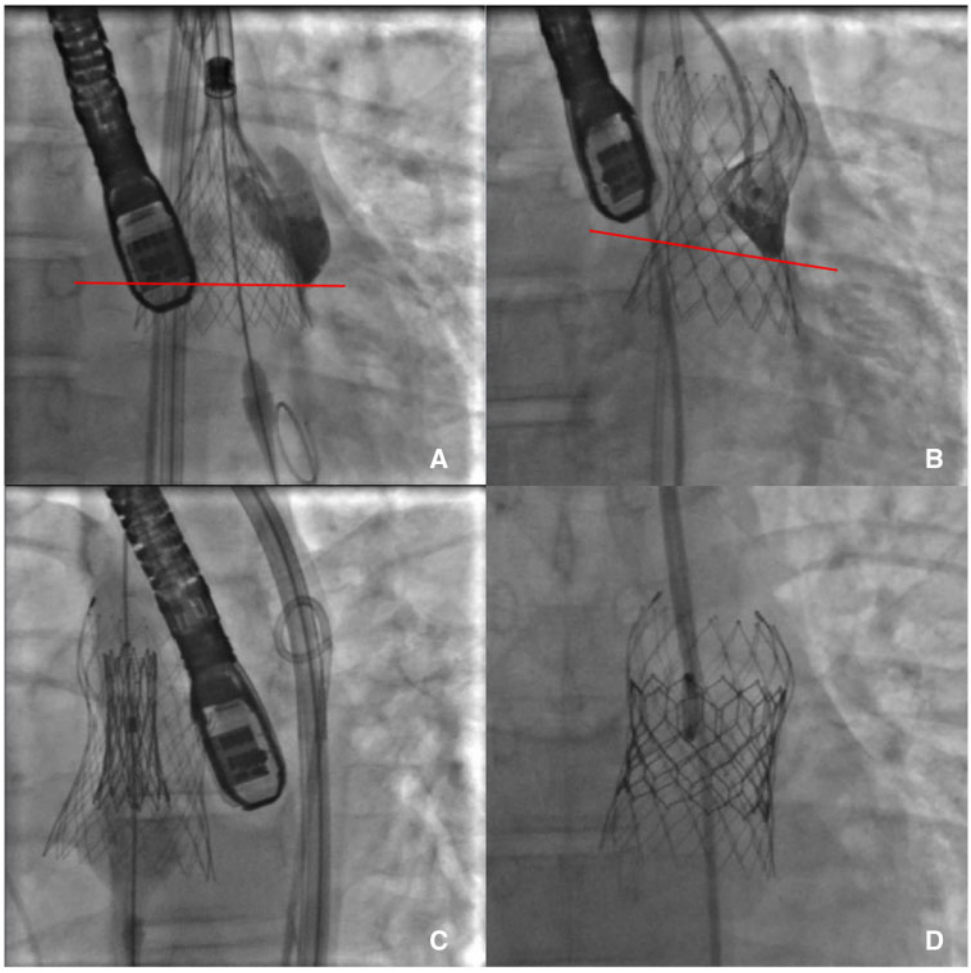

Figure 4 Fluoroscopic image. (*A*) Positioning of first valve. (*B*) Post-operative contrast of first valve. (*C*) Deployment of second valve. (D) Post-second valve deployment. The red lines in (*A*) and (*B*) are imaginary lines of aortic valve annulus height. When compared to (*A*), (*B*) shows that valve dislocation too deep for the height of one stent strut occurred. In (*C*) and (*D*), the second valve fully extends to the waist part of first valve. First valve: 34 mm Medtronic Evolut R. Second valve: 29 mm Edwards SAPIEN 3.

23.84 mmHg, and there was no description of the right ventricular ejection fraction (RVEF). The patient was discharged in a stable condition on post-operative Day 4. No further follow-up is available because of the patient transferred their care to another hospital.

Discussion

To the best of our knowledge, this is the first published case of TAVR for bicuspid AR in the setting of ccTGA with SRV dysfunction. While asymptomatic throughout early childhood, at 15 years of age this patient presented with heart failure symptoms related to the progression of AR and SRV. Previous studies have suggested that SRV dysfunction associated with systemic atrioventricular valve regurgitation (tricuspid regurgitation) in cases of unoperated adult ccTGA is problematic.[6] A preoperative SRV EF of <40% has been shown to be a predictor of poor prognosis in cases of unoperated adult ccTGA with SRV dysfunction requiring surgical tricuspid valve replacement.[3] In the present case, although the target valve was the aortic valve, the heart team believed that this patient's severe SRV dysfunction placed him at high operative risk prompting a decision to proceed with TAVR in this unique setting.

TAVR for symptomatic severe AS is widely perceived as an established procedure, however, there are only a few reports of TAVR for bicuspid AR and the safety of this treatment, even in a so-called normal anatomic setting is not well established. In this case, preoperative multiplanar imaging suggested that the 34 mm Medtronic Evolut R would be best suited to the unusual configuration of the left ventricular outflow tract in the setting of ccTGA. However, post-operative echocardiography showed severe PVL. It has been reported that significant PVL after TAVR is strongly associated with poor prognosis.[7] For the purpose of PVL reduction, valve-in-valve TAVR was performed with a 29 mm Edwards SAPIEN 3 on post-operative Day 2, and PVL improvement was subsequently observed. After the first TAVR procedure, the authors thought deep implantation of transcatheter heart valve (THV)[8] was the principal cause of the severe

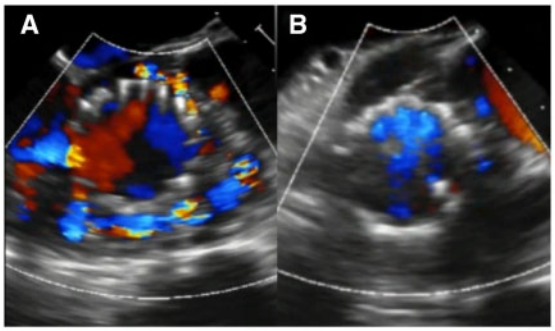

Figure 5 Pre- and post-transoesophageal echocardiography short-axis view for valve-in-valve transcatheter aortic valve replacement. (*A*) Pre-transoesophageal echocardiography short-axis view. (*B*) Post-transoesophageal echocardiography short-axis view. Following valve-in-valve transcatheter aortic valve replacement, paravalvular leak was significantly improved.

PVL. However, a simultaneous potential cause via the under-expansion of the THV could not be denied. Therefore, the authors waited 2 days for the decrement of PVL due to THV's self-expansion. However, AR did not decrease on post-operative Day 2. To remove the two potential causes, the primary operator decided to implant the SAPIEN3 valve, which has a stronger radial force than Evolut R, as the second valve.

Although a vascular plug would be another treatment option for PVL, we presumed that the use of a vascular plug was not effective because of its inability to seal the full circumferential jet of PVL. Unlike aortic valve stenosis, little or no calcification is observed in the aortic valve of patients with pure AR. In the absence of aortic valve calcification that might help anchor the THV, TAVR for pure native AR has been associated with valve dislocation.[9] In this case, no calcification of the aortic valve leaflet was observed placing this patient at risk for this complication. Additionally, the unusual orientation and anatomic structure of left ventricular outflow tract in ccTGA (i.e. presence of a subaortic conus) may have contributed to the observed valve dislocation (*Figure 4*). Ultimately, the performance of valve-in-valve TAVR utilizing a balloon-expandable valve served to markedly improve PVL as the SAPIEN 3 valve served to fully expand Evolut R through application of radial force. Perhaps not surprisingly, during the second procedure, deployment of the SAPIEN 3 valve to a precise position was simplified as the previously placed Evolut R served as an easily identifiable anchoring landing zone.

The long-term durability of THV over 10 years remains unknown, although recent investigations have revealed similar or better durability of THV in haemodynamic performance based on echocardiography compared to a surgical valve over 5–10 years.[10,11] In this study, the patient who underwent TAVR was young. Therefore, the long-term management strategy for this patient requires regular follow-up by the cardiologist and periodic examinations, including transthoracic echocardiography. This case had no other treatment option because the patient was at high risk for surgery. However, the durability of THV should be taken into account when considering TAVR as a treatment option for a young patient.

TAVR is a minimally invasive procedure compared to surgical aortic valve replacement and has been shown to serve an important role in high-risk patients with complex anatomy.[12,13] In this case, although a second valve deployment was required, an optimal result was ultimately achieved without the need for high-risk open-heart surgery. This result suggests that TAVR should be considered a treatment option for severe AR in patients with ccTGA at high risk for surgery.

Conclusion

To the best of our knowledge, this is the first case performed with TAVR for bicuspid AR in the context of ccTGA. With appropriate preparation and planning and a collaborative multi-disciplinary team approach, TAVR can be a treatment option for severe AR in patients with ccTGA at high risk for surgery.

Lead author biography

Takahiro Nomura is a Cardiologist. Specialty is ischaemic heart disease and structure heart disease.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: E.M.Z. has been a proctor for Edwards Lifesciences and Medtronic; and a consultant and national principal investigator for Edwards Lifesciences. R.R.M. received grant support from the Edwards Lifesciences Corporation and St. Jude Medical, and is a consultant for Abbott Vascular, Cordis, and Medtronic; he holds equity in Entourage Medical. All other authors declare no potential conflicts of interest.

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