JACC: CASE REPORTS VOL. 29, 2024

STRUCTURAL HEART DISEASE

CASE REPORT: CLINICAL CASE SERIES

Transcatheter Aortic Valve Replacement in Congenital Heart Disease

Betemariam Sharew, MS,[a,b] Beka Bakhtadze, MD,[a,b] Thomas Das, MD,[a,b] Kenneth Zahka, MD,[a,d] Hani Najm, MD,[a,c] Robert Stewart, MD,[a,c] Aaron Weiss, MD,[a,c] James Yun, MD,[a,c] Shinya Unai, MD,[a,c] Gosta Pettersson, MD, PHD,[a,c] Rishi Puri, MD, PHD,[a,b] Samir Kapadia, MD,[a,b] Tara Karamlou, MD, MS,[a,c] Joanna Ghobrial, MD, MS[a,b]

ABSTRACT

Transcatheter aortic valve replacement is not widely used in patients with congenital heart disease. We describe our single-center experience of transcatheter aortic valve replacement in congenital heart disease, demonstrating short-term feasibility and safety, role in lifetime management of congenital aortic valve disease, and use as a bridge to recovery, future surgery, or transplantation. (J Am Coll Cardiol Case Rep 2024;29:102199) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

The role of transcatheter aortic valve replacement (TAVR) has evolved immensely over the past decade, now spanning high- to low-surgical-risk patients.[1,2] Congenital heart disease (CHD) often involves the aortic valve (AV), in isolated lesions such as bicuspid aortic valves or part of a complex defect such as Shone syndrome, and can require multiple surgical interventions throughout a lifetime.

LEARNING OBJECTIVES

- To describe the unique characteristics of congenital TAVR, anatomical variety and complexity, and unique multidisciplinary evaluation.
- To introduce the feasibility and safety of TAVR in CHD and the critical role of TAVR in CHD as a bridge to recovery, future surgery, transplantation, or palliation.
- To emphasize the need for future larger clinical trials of TAVR in CHD.

In TAVR trials, CHD patients were excluded due to younger age and increased complexity. While TAVR use in the younger adult is increasing,[3] there is a dearth of outcomes data for it to be more widely adopted in the congenital population. Recently, case reports of TAVR use in the pediatric congenital population demonstrate its potential role.[4] We add importantly to this literature, describing our single-center experience of 6 diverse cases of adult CHD patients undergoing TAVR.

CASE SERIES

CASE 1. Case 1 was a 43-year-old man with tetralogy of Fallot who underwent 5 total surgeries, 4 of which were sternotomies, and 2 percutaneous interventions: right classic Blalock-Taussig-Thomas (RBTT) shunt; complete tetralogy of Fallot repair; right ventricular outflow tract conduit replacement; tricuspid valve replacement; a redo right ventricular outflow tract conduit replacement; redo tricuspid

From the [a]Department of Cardiovascular Medicine, Cleveland Clinic, Cleveland, Ohio, USA; [b]Cleveland Clinic Lerner College of Medicine, Case Western Reserve University, Cleveland, Ohio, USA; [c]Department of Cardiothoracic Surgery, Cleveland Clinic, Cleveland, Ohio, USA; and the [d]Department of Pediatric and Congenital Cardiology, Cleveland Clinic Children's, Cleveland, Ohio, USA.

The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received October 4, 2023; revised manuscript received December 4, 2023, accepted December 6, 2023.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2023.102199

ABBREVIATIONS AND ACRONYMS

AR = aortic regurgitation

AS = aortic stenosis

AV = aortic valve

BE = balloon-expandable

CHD = congenital heart disease

LV = left ventricular

RFA = right femoral artery

SE = self-expanding

TAVR = transcatheter aortic valve replacement

TPVR = transcatheter pulmonary valve replacement

TTVR = transcatheter tricuspid valve replacement

ViV = valve-in-valve

valve replacement with a 33-mm bioprosthetic valve (Biocor, Abbott) and aortic valve replacement (AVR) with a 27-mm bioprosthetic valve (Biocor); transcatheter pulmonary valve replacement (TPVR) with a 22-mm bovine jugular valve; and implantable cardioverter-defibrillator for ventricular tachycardia. He had right ventricular failure, left ventricular (LV) diastolic dysfunction, liver cirrhosis with portal hypertension and ascites, gastrointestinal bleeding, severe restrictive lung disease, and cardio hepatorenal disease. Prior pulmonary valve prosthetic endocarditis was treated medically with redo palliative TPVR given surgical risk, on suppressive antibiotics. He presented with triscuspid valve and AV prosthetic valve dysfunction and biventricular failure. Transthoracic echocardiography and transesophageal echocardiography revealed severe prosthetic aortic regurgitation (AR) and aortic stenosis (AS) (peak gradient of 74 mm Hg, mean gradient of 34 mm Hg, dimensionless index of 0.24), as well as severe tricuspid regurgitation. After multidisciplinary discussion involving the transplant team, it was decided to proceed with heart and liver transplant evaluation after optimization using valve-in-valve (ViV) TAVR and transcatheter tricuspid valve replacement (TTVR) as a bridge to transplantation, recognizing his high recurrent endocarditis risk.

The case was performed under general anesthesia with sterile technique. Vascular access obtained using the micropuncture technique with ultrasound guidance. Preprocedural antibiotic was administered. Given right classic Blalock-Taussig-Thomas shunt history and right femoral artery (RFA) and bilateral femoral venous occlusions, a cerebral embolic protection device could not be used, and access was right internal jugular vein for ViV TTVR, left internal jugular vein for venous access, left femoral artery for ViV TAVR, and left ulnar artery for arterial monitoring. ViV TAVR with a 26-mm balloon-expandable (BE) bovine pericardium valve (SAPIEN S3, Edwards Lifesciences) was performed, and stiff curved wire in the LV was used for pacing using alligator clips. Postdilation with a 24-mm balloon was performed. The final angiogram showed trace AR. TTVR was subsequently performed. Transthoracic echocardiography showed no AR, mean gradient 5 mm Hg, no tricuspid regurgitation, and mean gradient 3 mm Hg (Video 1).

At 12 months, transthoracic echocardiography showed trace-to-mild AR with peak gradient/mean gradient 26/13 mm Hg. His clinical status improved significantly, and he was accepted for heart and liver transplantation.

CASE 2. Case 2 was a 40-year-old woman with atrioventricular septal defect (AVSD), cleft mitral valve with mitral regurgitation, and sub-AS for which she underwent 5 open heart surgeries: AVSD repair and mitral valve repair, subaortic resection, Konno procedure, bioprosthetic mitral valve replacement, and redo mechanical mitral valve replacement. Her medical history was complicated further by liver cirrhosis, prior bacteremia without valve vegetations treated medically, heart block with pacemaker dependence, and atrial arrhythmias. She presented with heart failure symptoms, and progressive decline in functional capacity. Transthoracic echocardiography and transesophageal echocardiography revealed mildly reduced biventricular systolic function and severe AR without stenosis that progressed since her Konno procedure. After multidisciplinary discussion, she was deemed high surgical risk, and TAVR was selected to improve her functional capacity and future surgical candidacy. Given pure native AR of a noncalcified valve, a self-expanding valve platform was selected, with a size of 26 mm based on a perimeter of 70 mm and annular diameter of 21 mm.

The case was performed in similar fashion to case 1 but with femoral access, using an 8-F in the RFA for valve implantation, 5-F in the RFA for arterial access, 8-F in the right femoral vein for venous access and pacing, and 6-F in the right radial sheath for cerebral embolic protection. A pigtail was placed in the noncoronary cusp and a pacer was positioned in the right ventricle. Hemodynamics showed LV end-diastolic pressure of 20 mm Hg with aortic diastolic pressure of 42 mm Hg. A 26-mm porcine self-expanding (SE) valve (Evolut, Medtronic) was deployed successfully, with an increase in aortic diastolic pressure to 60 mm Hg (Video 2).

At 3 months, transthoracic echocardiography showed no AR and no paravalvular leak. AV peak gradient/mean gradient was 17/9 mm Hg with a dimensionless index of 0.44. She was able to resume daily activities such as walking her dog.

CASE 3. Case 3 was a 29-year-old man with D-transposition of the great arteries and ventricular septal defect status after 3 open heart surgeries: arterial switch operation, redo neo-AV repair, and redo AVR with a 25-mm bovine pericardial valve (CE Perimount, Edwards Lifesciences). He was lost to follow-up and presented 10 years later with heart failure and critical prosthetic AS, peak gradient/mean gradient

125/90 mm Hg, and new onset LV dysfunction, with ejection fraction 20%.

The case was performed in similar fashion as described in case 2. The patient underwent a ViV TAVR with a 26-mm BE bovine pericardial valve (SAPIEN S3). Transaortic gradients decreased from 90 mm Hg to 0 mm Hg (Video 3).

At 3 months' follow-up, transthoracic echocardiography showed an AV peak gradient/mean gradient of 23/13 mm Hg and LV ejection fraction improvement to 50% with medical management.

CASE 4. Case 4 was a 47-year-old man with tetralogy of Fallot for which he underwent a total of 5 open heart surgeries: classic right classic Blalock-Taussig-Thomas shunt; complete tetralogy of Fallot repair; AV repair and pulmonary homograft replacement; redo AVR and pulmonary valve replacement; redo 27-mm AVR (Biocor), 24-mm pulmonary valve replacement and mitral valve repair; and subsequent TPVR. He presented with symptomatic moderate-to-severe aortic bioprosthetic stenosis, AV peak gradient/mean gradient 51/30 mm Hg, and dimensionless index of 0.24, with no regurgitation and mildly reduced LV function (LV ejection fraction 50%) on transthoracic echocardiography. The prosthetic pulmonary valve was well functioning. After multidisciplinary discussion, the decision was made to proceed with ViV TAVR as a bridge to a future surgery, in which both aortic and pulmonary prostheses can be addressed.

The case was performed in similar fashion as described in case 2. A 26-mm BE bovine pericardium valve (SAPIEN S3) was successfully deployed at nominal volume and postdilated with a 24-mm balloon at 14 atm (Video 4).

Postoperative echocardiography demonstrated improved peak gradient/mean gradient of 27/16 mm Hg, a dimensionless index of 0.52, with no AR. At 3 and 6 months' transthoracic echocardiography with stable valve function, peak gradient/mean gradient was 25/12 mm Hg, with a DI of 0.6. He reported resolution of his symptoms and had resumed his work as a personal trainer.

CASE 5. Case 5 was a 25-year-old woman with bicuspid aortic valve, chronic immune thrombocytopenic purpura, severe idiopathic primary pulmonary arterial hypertension, and liver failure requiring transplantation. She presented with dyspnea and presyncope. Transthoracic echocardiography demonstrated moderate-to-severe AS with peak gradient/mean gradient 62/38 mm Hg and dimensionless index of 0.27. Catheterization with transaortic gradient of 43 mm Hg. Balloon valvuloplasty with gradient reduction from 40 to 22 provided symptom improvement, with subsequent severe AS recurrence over 9 months. After multidisciplinary discussion, to avoid a sternotomy in a young patient who will likely require future lung transplantation, TAVR was elected to manage her bicuspid aortic valve stenosis. Planning for a future SAVR with her lung transplantation, the heart team raised the concern that the taller SE platform can adhere to the ascending aortic wall and require further dissection with repair or replacement, so the shorter BE platform was chosen. The annulus was not markedly elliptical ($\sim 19 \times 23$ mm), and risk of perivalvular leak was not a major concern.

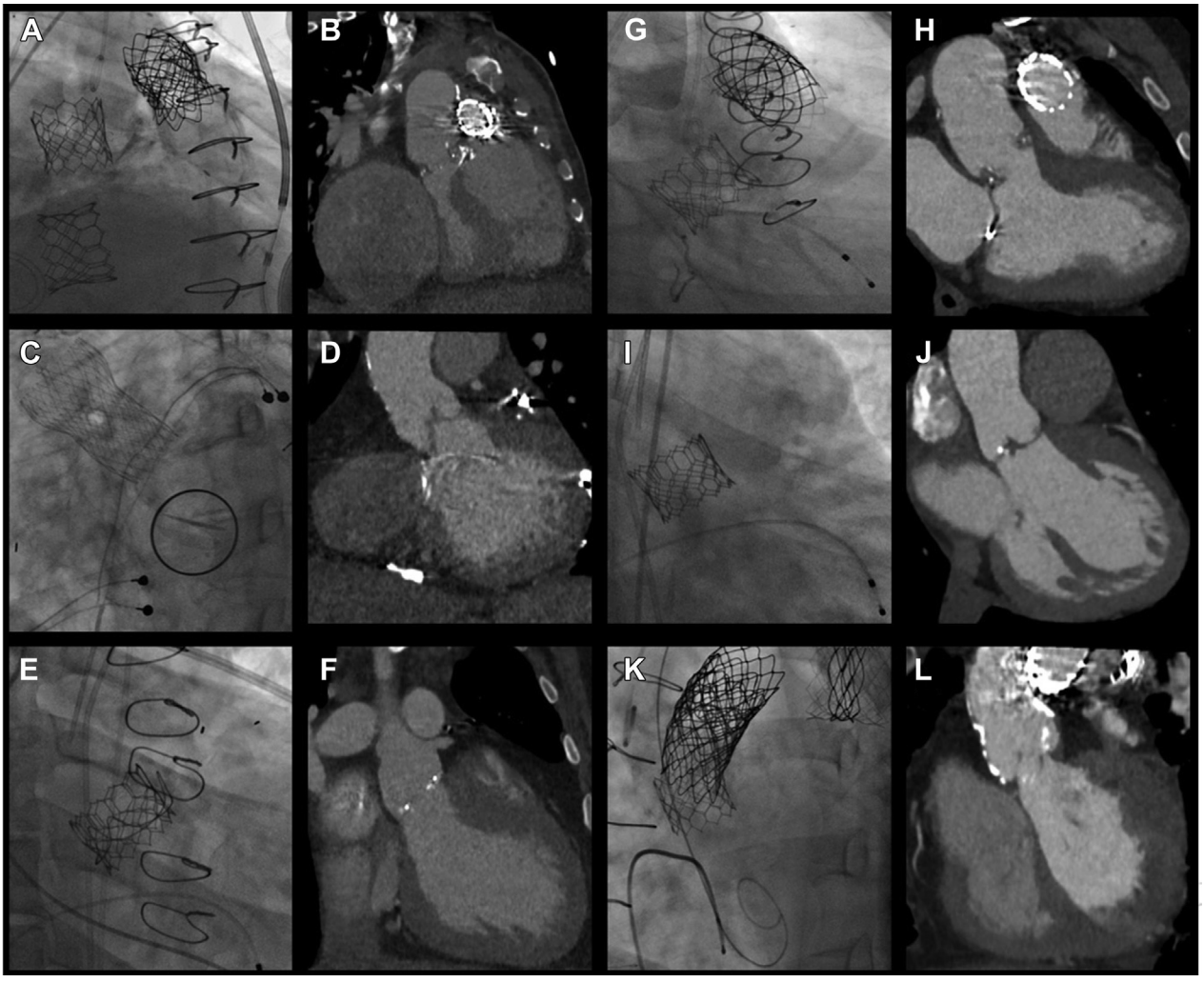
The case was performed in similar fashion as previously described in case 2. A 23/mm BE bovine pericardial valve (SAPIEN S3) was deployed at nominal volume and postdilated at nominal +1 cm^3 added fill volume (Videos 5a and 5b).

At 9 months' follow-up, transthoracic echocardiography showed peak gradient/mean gradient 23/11 mm Hg and a dimensionless index of 0.45, without AR and with resolution of symptoms.

CASE 6. Case 6 was a 49-year-old man with Shone syndrome with bicuspid aortic valve, subaortic membrane, and aortic coarctation. He underwent 5 open heart surgeries: AV repair, aortic coarctation repair with end-to-end anastomosis, redo AV repair, aortic and pulmonary homograft placement, and redo aortic (Freestyle 25 mm, Medtronic) and pulmonary homograft replacement. He underwent pulmonary artery covered stenting and TPVR to address a fistulous connection between his ascending aorta and pulmonary artery causing severe pulmonary arterial hypertension, with subsequent resolution of pulmonary hypertension (mean pulmonary arterial pressure of 62 to 23 mm Hg). Two years later, he presented with abrupt-onset dyspnea and volume overload. Transesophageal echocardiography showed severe AR of his heavily calcified homograft, with a ruptured cusp without AS (peak gradient/mean gradient 27/16 mm Hg), with mildly reduced LV function. After multidisciplinary discussion, TAVR was elected to relieve his symptoms as a bridge to future surgery that would likely tackle both his AV and pulmonary bioprosthetic valves. Despite pure AR as the underlying pathology, given that the homograft was heavily calcified, both SE and BE valve platforms were options, and a 23-mm BE valve was selected based on a diameter of 22.5 mm and area of 396 mm^2.

The case was performed in similar fashion as previously described in case 2. A 23-mm BE bovine pericardium valve (SAPIEN S3) was implanted via the

FIGURE 1 Computed Tomography and Fluoroscopy of Congenital TAVR



(A) Case 1 fluoroscopy of valve-in-valve (ViV) transcatheter aortic valve replacement (TAVR), ViV transcatheter tricuspid valve replacement, and transcatheter pulmonary valve replacement (TPVR). (B) Case 1 computed tomography angiography (CTA) preprocedure with dilated right atrium/right ventricle. (C) Case 2 fluoroscopy of TAVR and mechanical mitral valve replacement. (D) Case 2 CTA preprocedure demonstrating noncalcified native tricuspid aortic valve. (E) Case 3 fluoroscopy of ViV TAVR. (F) Case 3 CTA preprocedure demonstrating surgical aortic valve replacement, branch pulmonary artery around ascending aorta post-LeCompte. (G) Case 4 fluoroscopy of ViV TAVR and TPVR. (H) Case 4 CTA preprocedure. (I) Case 5 fluoroscopy of TAVR in bicuspid aortic valve. (J) Case 5 CTA preprocedure demonstrating thickened bicuspid aortic valve leaflets with minimal calcification and dilated pulmonary artery. (K) Case 6 fluoroscopy of valve-in-homograft TAVR, TPVR, and left pulmonary artery stent. (L) Case 6 CTA preprocedure demonstrating heavily calcified aortic homograft and covered-stented pulmonary artery.

transfemoral approach. Postoperatively, AV peak gradient/mean gradient was 10/6 mm Hg, without AR (Videos 6a and 6b).

At 6 months' follow-up, transthoracic echocardiography showed stable mildly reduced LV ejection fraction at 45%, AV peak gradient/mean gradient of 24/13 mm Hg, and dimensionless index of 0.42, and he reported resolution of symptoms (**Figure 1**, **Table 1**).

DISCUSSION

In our single-center 6-case series of TAVR in CHD, the average age was 38 years, and the median number of sternotomies was 4. All patients were at high or prohibitive surgical risk. All underwent TAVR safely and without acute complications despite anatomical and access challenges. Importantly, all had significantly improved and acceptable

TABLE 1 Demographic and Procedural Data

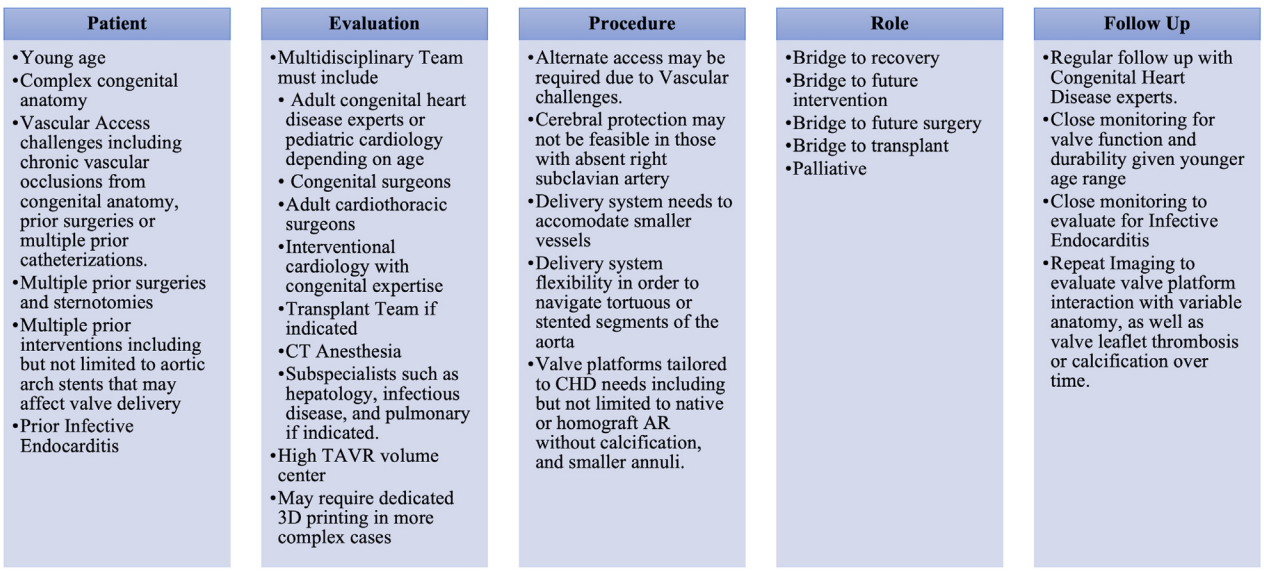
Patient	Age, y/Sex	CHD	Prior Surgeries	Aortic Valve Pathology	Reason for TAVR	TAVR Type/Size (mm)	Follow-Up (mo)	PG/MG (mm Hg)
1	43/M	TOF	5	Prosthetic AR	Bridge to transplantation	BE/26	12	26/13
2	40/F	AVSD	5	Native AR	Bridge to surgery	SE/26	3	17/9
3	29/M	D-TGA	3	Prosthetic AS	Bridge to recovery or surgery	BE/26	3	23/13
4	47/M	TOF	4	Prosthetic AS	Bridge to recovery or surgery	BE/26	6	27/16
5	25/F	bicuspid aortic valve	0	Native AS	Bridge to surgery and lung transplantation	BE/23	9	23/11
6	49/M	Shone complex	5	Homograft AR	Bridge to recovery or surgery	BE/23	6	24/13

AR = aortic regurgitation; AS = aortic stenosis; AVSD = atrioventricular septal defect; BE = balloon expandable; CHD = congenital heart disease; D-TGA = D-transposition of the great arteries; F = female; M = male; MG = mean gradient; PG = peak gradient; SE = self-expanding; TOF = tetralogy of Fallot.

hemodynamics with no to mild AR and an average MG of 11 mm Hg. Multidisciplinary discussion with CHD experts and the heart team is essential to address the varied unique characteristics of each patient (Figure 2).

CHD patients have complex anatomies, multiple prior sternotomies, associated scars, and adhesions, making surgical access challenging. This increases their operative and postoperative risk of cardiovascular, pulmonary, and neurologic complications. TAVR is a minimally invasive option, reducing the risk of redo sternotomies, and can be an important tool in the lifetime management of AV disease in CHD. Importantly, in CHD patients awaiting transplantation, TAVR can improve functional capacity and extend survival while on the waitlist, without additional risks of surgery. There is an important need for innovation in the field, specifically to develop platforms tailored to the needs of CHD patients such as native or homograft pure AR (Figure 2). There are relevant concerns of the long-term durability in the younger congenital patient. To further understand the intermediate and long-term outcomes and durability of TAVR in CHD patients, prospective trials with larger cohorts are needed.

FIGURE 2 Congenital TAVR Unique Characteristic



Unique characteristics in congenital TAVR include anatomical, pre-procedural, intra-procedural, post-procedural challenges, and the different uses of TAVR in CHD. AR = aortic regurgitation; CHD = congenital heart disease; CT = computed tomography; TAVR = transcatheter aortic valve replacement; 3D = 3-dimensional.

CONCLUSIONS

TAVR can be an effective intervention in CHD in select clinical scenarios, proving both feasible and safe with good short-term outcomes in this small case series. In high surgical risk, it can be used as a bridge to recovery, future surgery, transplantation, or palliation. There is an important need for large prospective clinical trials of TAVR in CHD.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr Joanna Ghobrial, Department of Cardiovascular Medicine, Cleveland Clinic, 9500 Euclid Avenue, Cleveland Ohio 99175, USA. E-mail: ghobrij@ccf.org. @JGhobrialMD.

REFERENCES

1. Mack MJ, Leon MB, Thourani VH, et al. Transcatheter aortic-valve replacement with a balloon-expandable valve in low-risk patients. *N Engl J Med*. 2019;380:1695-1705.

2. Otto CM, Nishimura RA, Bonow RO, et al. 2020 ACC/AHA guideline for the management of patients with valvular heart disease: executive summary: a report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *J Am Coll Cardiol*. 2021;77(4):450-500.

3. Nelson JS, Maul TM, Wearden PD, et al. Aortic valve replacement in young and middle-aged adults: current and potential roles of TAVR. *Ann Thorac Surg*. 2021;112:132-138.

4. Sinha SA, Khan AA, Qureshi AA, et al. Application of transcatheter valves for aortic valve replacement in pediatric patients: a case series. *Catheter Cardiovasc Interv*. 2020;95:253-261.

KEY WORDS aortic regurgitation, aortic stenosis, congenital heart disease, transcatheter aortic valve replacement, valve-in-valve implantation

APPENDIX For supplemental videos, please see the online version of this paper.