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Feasibility and effectiveness of transcatheter aortic valve implantation in adults with congenital heart disease

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ABSTRACT

Aim: This study sought to evaluate the feasibility and effectiveness of TAVI for treatment of aortic stenosis in the ACHD population.

Methods and results: Review of 802 patients that underwent TAVI from January 2008 to November 2019. 13 ACHD patients with different underlying congenital anatomy were identified (isolated bicuspid aortic valve was excluded and not defined as ACHD). The main endpoints were: paravalvular aortic regurgitation> grade 2 post-procedure, and all-cause 30-day mortality. Periprocedural and post-procedural complications were defined according to the UK TAVI registry. Median age was 66.7 years (range 29–84 years). Eleven patients had severe aortic stenosis (AS), 1 had both severe AS and severe aortic regurgitation (AR) and 1 had severe AR. Median hospital stay was 9 days (range 6–28 days). One patient required a pacemaker post-TAVI. No mortality or paravalvular AR > grade 2 at 30-days. Twelve patients had a reduction in NYHA class on their first post TAVI follow-up.

Conclusion: TAVI is viable option as an alternative to surgery for ACHD patients. Further experience with the use of TAVI in the ACHD patients is required to assess long-term outcomes in this unique group of patients.

1. Introduction

The past 70 years has witnessed considerable development in the diagnosis and management of congenital heart disease, leading to an increased number living into adulthood [1]. Despite increased survival, adults with congenital heart disease (ACHD) often have complex anatomy, and have more cardiovascular complications and a higher mortality risk than the general population [2,3]. Many will develop significant valve lesions requiring multiple interventions in their lifetime.

Transcatheter aortic valve implantation (TAVI) was initially introduced to treat elderly patients with calcific aortic stenosis who were considered high risk or with contraindications to surgery [4]. The indications for TAVI have expanded to those at intermediate and low risk for conventional surgical valve replacement [5–7].

ACHD patients have frequently undergone sternotomies in childhood, have unusual anatomy of the aortic valve, or the consequences of previous surgery, and may have access problems to the heart, such as the presence of a coarctation repair (surgical or stent). Furthermore, as they are often young, the longevity of whatever valve is implanted surgically or percutaneously has to be taken into consideration.

TAVI has potential advantages for ACHD patients who have had previous operations or other relative contraindications to surgery, but TAVI for severe aortic valve (AoV) disease in the ACHD population is not well described.

This study reports all TAVI procedures undertaken in ACHD patients over an 11-year period at a tertiary level I centre for ACHD and TAVI.

2. Methods

2.1. Patients

We retrospectively reviewed the cardiac catheterization reports and medical notes of all TAVI patients from January 2008 until November

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Abbreviation list

ACHD	Adult Congenital Heart Disease
AR	Aortic regurgitation
AS	Aortic stenosis
AoV	Aortic valve
BAV	Bicuspid aortic valve
CoA	Coarctation of aorta
CT	Computed tomography
EuroSCORE	European System for Cardiac Operative Risk Evaluation
MDT	Multi-Disciplinary Team meeting
NYHA	New York Heart Association
PAR	Paravalvular aortic regurgitation
TAVI	Transcatheter Aortic Valve Implantation
TGA	Transposition of great arteries
ViV	Valve in Valve

2019, at the Bristol Heart Institute, Bristol, UK. Thirteen ACHD patients were identified from a total of 802 TAVI procedures during this period. A total of 293 AoV surgical procedures were performed on ACHD patients by comparison. Patients with native bicuspid aortic valve (BAV) alone as their only congenital diagnosis were not included in our study. Baseline demographic and clinical characteristics were recorded. All patients were discussed at ACHD and TAVI Multi-Disciplinary Team meetings (MDT), confirming an indication and mode of treatment (central illustration). Eleven of the TAVI ACHD patients were deemed very high risk by the surgical team. One patient refused blood transfusion and declined surgery. In one patient, surgery was attempted, but cross clamping was not feasible (because of significant scar tissue and risk of continued surgical dissection,) and the surgeon requested TAVI. This retrospective service evaluation was exempt from Research and Ethics Committee review in accordance with National Health Service Research Ethics Service guidelines [1].

2.2. Echocardiography

The diagnosis of severe AoV disease was confirmed by transthoracic echocardiography according to established guidelines [2]. Prior to discharge all patients received a detailed transthoracic echocardiogram including peak velocity, peak and mean gradient, and systemic ventricular ejection fraction.

Semi-quantitative grading of paravalvular AR (PAR) was performed using periprocedural transoesophageal echocardiography and aortography and post-procedure transthoracic echocardiography. Semi-quantitative grading of PAR was; 0 = absent, 1 = trace, 2 = mild, 3 = moderate, 4 = severe [3].

2.3. Computed tomography

Prior to the procedure, all patients underwent CT to assess: 1) valve morphology, 2) annulus size 3) coronary ostial height and sinus of Valsalva assessment, 4) aortic root, 5) evaluation of the peripheral access route.

CT scans were performed using ECG gating with either a Somatom AS+ (Siemens Healthineers, Erlangen, Germany) or an Aquilion One Genesis (Canon Medical Systems, Otawara, Japan) system. The Siemens system is a 64-detector row system with flying focal spot, 0.6 mm collimation, 300 ms rotation time, and the Canon system 320 detector row, 0.5 mm collimation, 270 ms rotation time. Non-contrast evaluation of the aortic root was not an essential component of the TAVI work-up as some members of our patient population were young and been recommended in recent guidelines as it is useful in the setting of uncertain AS severity. If non-contrast scans were acquired, AoV calcium score was performed at 120 kV. In patients who did not have a non-contrast scan, severity of calcium on contrast scans was based on visual assessment (supplementary data). Contrast enhanced CT scans were 100–120 kV retrospective ECG gated examination of the aortic root (maximum tube output in systole). Typically, 0.75 mm thick reconstructions were used for annular assessment at 250 ms and 75% phase (Canon) and 250 ms and 70% phase (Siemens). Assuming adequate renal function (eGFR >45) an iodine flux of ~15–20 mg Iodine/kg patient body weight/second was used for 20 s

Table 1
Baseline characteristics, medical history and indication of TAVR.

Patient	Diagnosis	Age at TAVI (yr)	Original AoV morphology	Pre TAVI morphology	Indication for TAVI
1	Atrio-pulmonary Fontan, Tricuspid & pulmonary atresia, previous Brock procedure + right Blalock Taussig shunt	53.1	Bicuspid	Bicuspid	Severe AS
2	CoA - surgical repair; severe bicuspid AS + saccular calcified aneurysm at site of CoA repair.	80.2	Bicuspid	Bicuspid	Severe AS
3	Congenitally corrected TGA with systemic right ventricle dysfunction + moderate systemic right atrioventricular valve regurgitation	84.2	Tricuspid	Tricuspid	Severe AS
4	Bicuspid AS, pulmonary hypertension due to PDA	32.1	Bicuspid	Bicuspid	Severe AS
5	Aortic homograft, previous infective endocarditis.	29.2	Bicuspid	Homograft Tricuspid	Severe prosthetic AS
6	Double inlet left ventricle, TGA, S/P arterial switch + Glenn, S/P total cavopulmonary connection, Ehlers Danlos syndrome.	27.9	Tricuspid	Tricuspid	Severe AR
7	Native CoA, bicuspid AS, Jehovah's witness	51.3	Bicuspid	Bicuspid	Severe AS
8	CoA surgical repair, recoarctation, bicuspid AoV replacement, bioprosthetic valve degeneration-severe AS, previous atrial fibrillation and complete heart block, dual-chamber implantable cardioverter-defibrillator	62.1	Bicuspid	Homograft Tricuspid	Severe prosthetic AS
9	Congenital bicuspid AoV, previous homograft AVR with severe prosthetic AS	54.4	Bicuspid	Homograft Tricuspid	Severe prosthetic AS
10	Infective endocarditis and homograft AVR x2 childhood, presenting with cerebrovascular accident + severe AS and AR	27.8	Tricuspid	Homograft Tricuspid	Severe prosthetic AS and AR
11	Previous CoA surgical repair, tiny pseudoaneurysm, severe bicuspid AS, heavy calcification of ascending aorta, peripheral vascular disease, single functioning kidney	72.6	Bicuspid	Bicuspid	Severe AS
12	CoA, bicuspid AoV with root dilatation, previous bioprosthetic valve + ascending & aortic arch replacement), dilatation ascending aorta (52 mm) & aorta beyond aortic arch replacement	39.7	Bicuspid	Perimount trileaflet	Severe prosthetic AS
13	Moderate-severe residual bicuspid AS, post sub-aortic resection and valvotomy, AoV repair (3x sternotomy) & previous CoA repair (thoracotomy), residual AS and AR	47.0	Bicuspid	Repaired Bicuspid	Severe AS

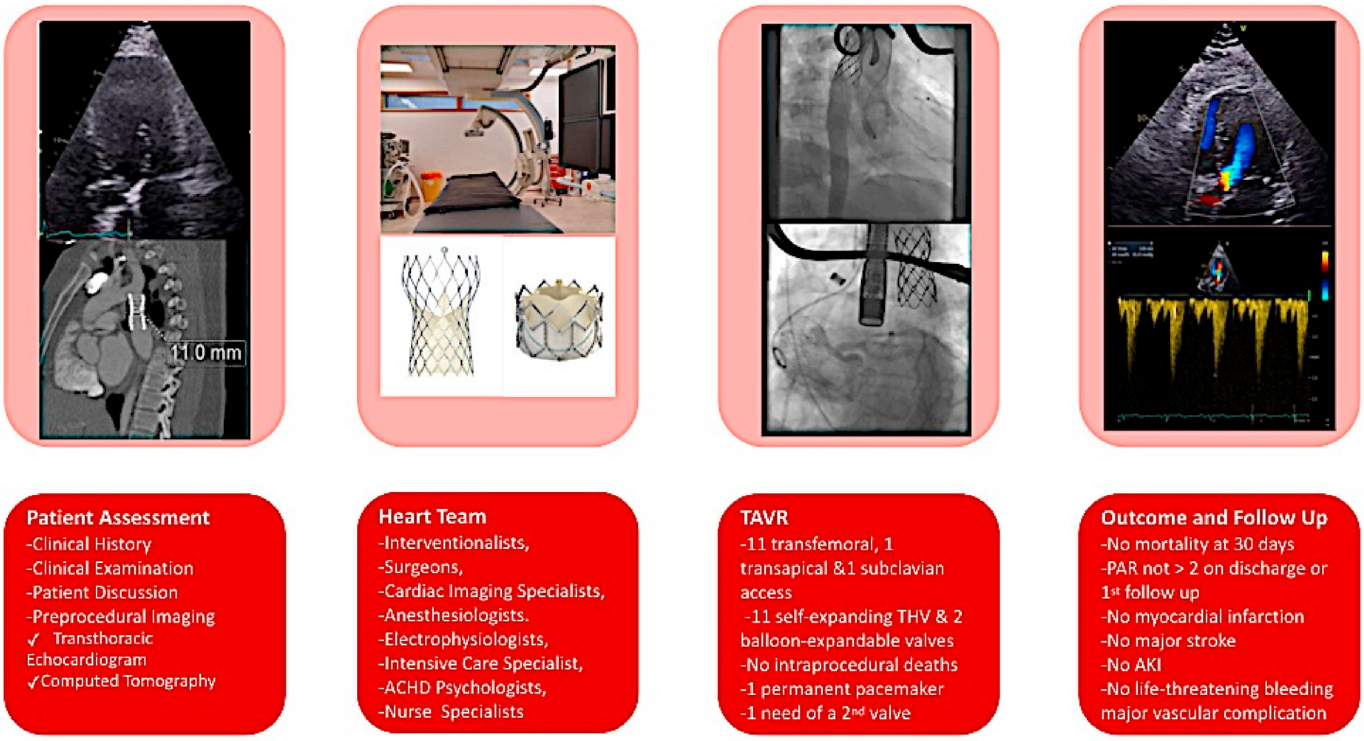

Fig. 1. Summary of TAVI in ACHD Feasibility Study. Evolut and Sapien XT images used with the courtesy and permission of Medtronic (© 2016 Medtronic) and Edwards Lifesciences Corporation.

(Canon) or 30 s (Siemens) - to also allow follow on body imaging. Either Iomeron 400 (Bracco, Milan, Italy) or Omnipaque 300 (General Electric, USA) iodinated contrast was used.

2.4. Vascular access and valve selection

The vascular approach was determined by the size, calcification, tortuosity and patency of ilio-femoral arteries. Valve type was determined by the operator based on anatomical and clinical factors. Valve size was based on pre-procedural imaging. If the patient had a prosthetic valve, the size of the valve was based on true internal diameters of the prothesis. From 2014, ViV (valve in valve) Aortic smart phone application was used for planning ViV procedure (https://www.pcronline.com/PCR-Publications/PCR-mobileapps/Valve-in-Valve-Aortic-app) [4].

2.5. Procedure

TAVI procedures were performed in a hybrid cardiac catheterization room under general anaesthesia or local anaesthesia with conscious sedation. Transoesophageal echocardiography was used if the TAVI was under general anaesthesia. Ultrasound guidance for vascular access was used for femoral access. Both self-expanding and balloon expandable valves were used, and pre-dilatation was performed at the operator's discretion. Intravenous heparin was administered to achieve an ACT of around 300s. The large vessel access was closed with a Prostar closure device and pacing was established with a transvenous temporary pacing wire. The result was assessed angiographically, hemodynamically and by echocardiography.

2.6. Postprocedural care

All patients were observed in the coronary care unit or cardiac intensive care unit for at least 24 h. For patients who did not have an indication for anti-coagulation, dual antiplatelet therapy was continued for 3 months and, thereafter, aspirin was continued indefinitely.

2.7. Study outcomes

The main endpoints were the frequency of PAR > grade 2 post-procedure and all-cause 30-day mortality. Periprocedural and post-procedural complications were defined according to the UK TAVI registry [5]. Adverse events were defined as myocardial infarction, major stroke, acute kidney injury, major vascular complication, life-threatening bleeding, annulus rupture and pacemaker implantation. Any other significant event identified during follow up was also reported.

2.8. Statistical analysis

Statistical analysis was performed using commercial spread sheet software. (Excel 2019 for Mac version 16.29, Microsoft, Redmond, Washington). Continuous data were reported as mean ± SD or median (range) depending on normality of their distribution. Categorical variables were presented as frequencies and percentages. Complete data were available in all patients. Inferential statistical analyses were not carried out, as the sample was too small and diverse.

3. Results

3.1. Baseline characteristics

Baseline characteristics, medical history and indication of TAVI are summarized in Table 1. All 13 patients (9 males) had different underlying congenital heart disease. All presented with decreased physical capacity (average NYHA class III), 3 of whom as urgent inpatients. Eleven patients had severe AS, 1 had both severe AS and severe aortic regurgitation (AR) and 1 severe AR. Ten of the 13 (77%) had undergone previous cardiac

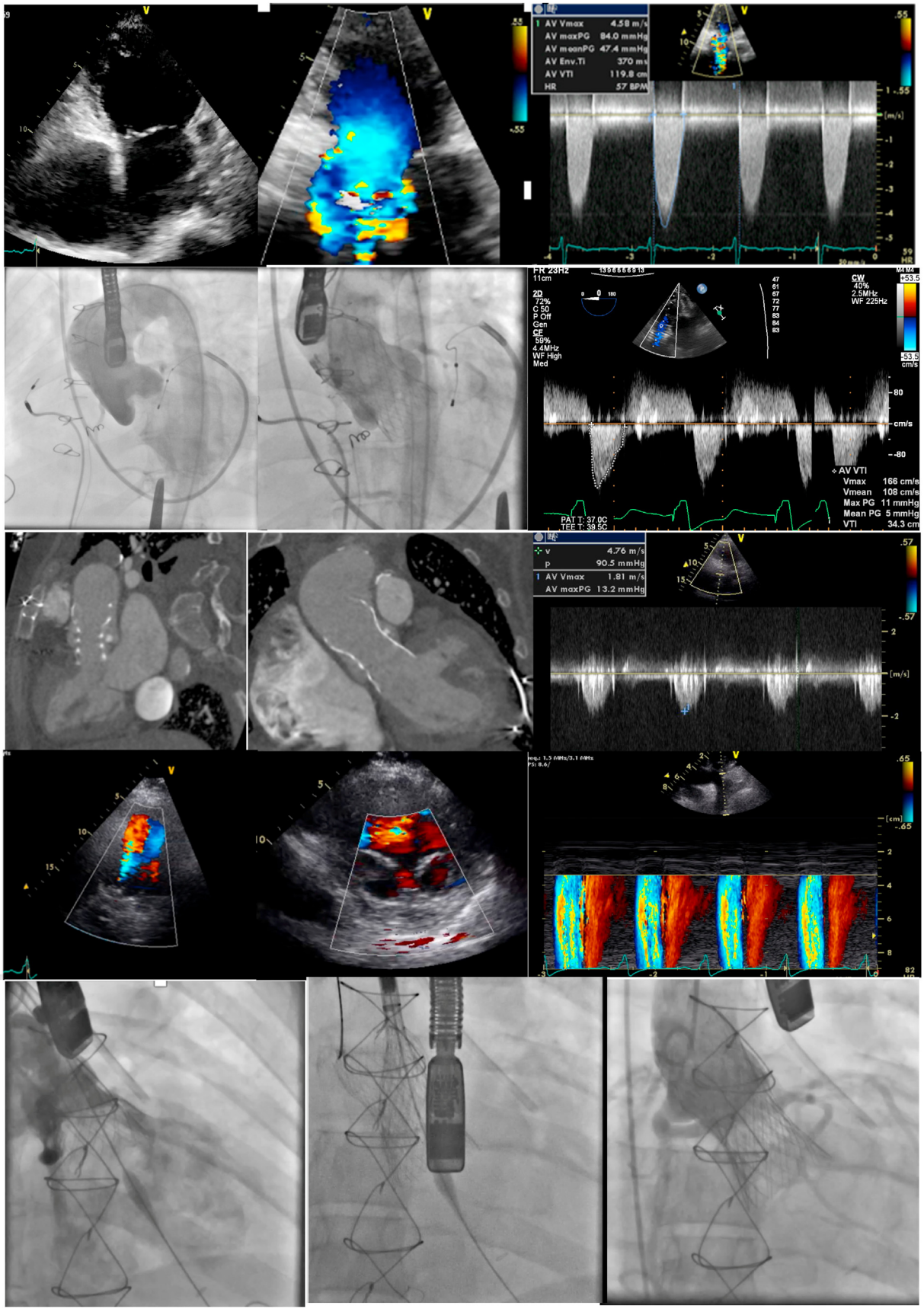

Fig. 2. TAVI in Fontan Circulation. Panel A–C preprocedural imaging, procedure and postprocedural imaging of Patient 1. Panel D–E preprocedural imaging and procedure of Patient 6.

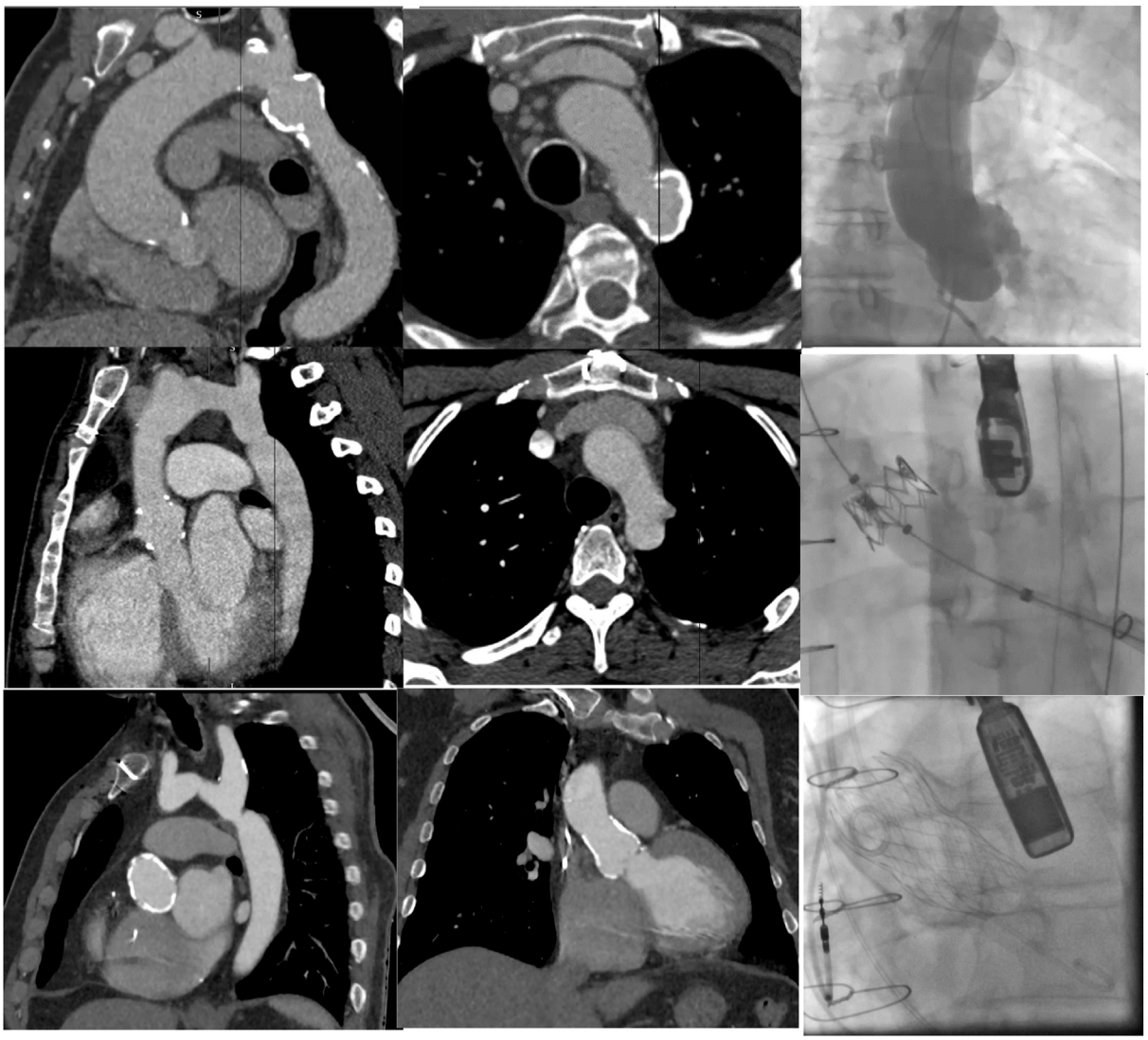

Fig. 3. Vascular access in patients with a background diagnosis of CoA.

surgery. Five patients had bioprosthetic AoV replacements (4 of 5 originally had BAV) and 6 native BAV in addition to their underlying condition. The aortic annulus size was 26.1 mm ± 1.96 (range 23 mm–31 mm). Patient 6 had an annulus of 33 mm, but the superior aspect had previously been resuspended in a 31 mm Goretex tube during his previous valve sparing aortic root operation. In 12 patients with severe AS, the pre-procedural mean gradient was 45 mmHg±12.8.

Some patients were elderly, but all had congenital anatomical abnormalities that had a potential technical challenge, setting them apart from the usual TAVR population, including coarctation and an anterior aorta connected to a systemic right ventricle, with a muscular right ventricular outflow tract, below the aortic valve.

3.2. Procedural results

A summary of our approach and outcomes are shown in Fig. 1 and results of imaging and procedural details for the 13 patients are summarized in Table 2, some of which are illustrated in Figs. 2–5. Patient 1 has been published as a case report and only salient data is provided [6].

Eleven TAVI procedures were performed transfemorally. A transapical approach was performed in patient 5 due to residual coarctation and small peripheral vessels. Patient 5 also had an ejection fraction of ~10% with non-compaction cardiomyopathy and so peripheral bypass was established surgically with an apical vent. Patient 7 had subclavian artery access as there was right external iliac occlusion from previous native coarctation stenting.

Balloon-expandable valves were used in 2 patients (17%) (Sapien S3 and SapienXT, Edwards Lifesciences, Inc., Irvine, California and self-expanding THV in 11 patients (83%) (CoreValve, Evolut R and Evolut Pro Medtronic, Inc., Minneapolis, Minnesota).

Ten out of 13 procedures were uncomplicated. The complications seen in the other 3 patients were the need of a pacemaker and significant PAR (one valve repositioning and the other a second valve). Patient 1 experienced ventricular standstill after direct implantation of the 29 mm Medtronic Evolut R. She was the only patient who needed permanent pacing (epicardial) [6]. Patient 3 developed significant PAR with worsening systemic **right** ventricular function and renal impairment. He was returned to the lab and the 31 mm CoreValve was snared and pulled up into a higher position, with improved diastolic blood pressure and mild AR. Patient 6 required a second valve during the procedure. On multiple attempts, the first valve went too deep or popped up. After good positioning of the first valve was achieved, the valve dived into the ventricle after release. This resulted in severe AR and hemodynamic instability. Attempts to pull the valve up with a balloon failed. As hemodynamics

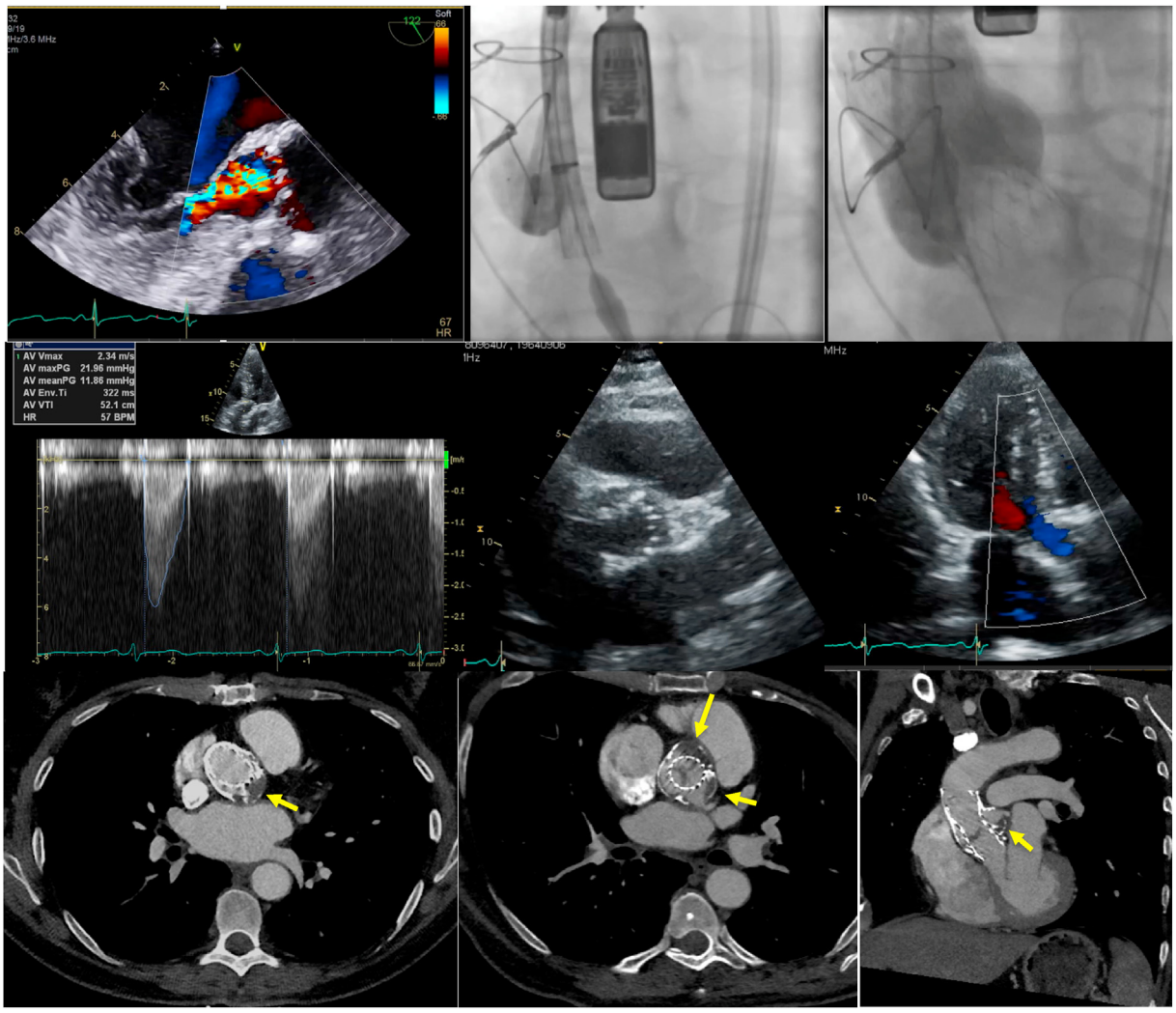

Fig. 4. TAVI procedure in a homograft (upper and middle panel). Lower panel shows thrombus (arrows) detected during routine follow up scan.

worsened, he was supported by percutaneous femoral bypass and a 28 mm Nucleus balloon was inflated in the valve to prevent retrograde flow and improve the pressure on bypass (as the aortic regurgitation was preventing effective perfusion), and a second valve was prepared. Successful implantation of the second valve was achieved at a much higher position with mild AR on transoesphageal echocardiogram and angiography. Coronary ostial obstruction was not a feature in any case.

3.3. Clinical outcome

3.3.1. Primary safety and effectiveness end point

There was no mortality at 30 days, nor any myocardial infarction, AKI, life-threatening bleeding or major vascular complications. The median stay in hospital was 8.0 days (range 4–35 days). Twelve patients were discharged directly home. Patient 6 who had an intraoperative cardiac arrest and woke up with quite significant visual loss, dysarthria and co-ordination difficulties. Ophthalmology review, CT and MRI head were unremarkable. This was likely ischemic posterior optic neuropathy and hypoxic brain injury. His vision gradually improved, and he was discharged to a rehabilitation facility. Twelve patients reported improvement in NYHA functional class at their first follow up. The mean gradient was reduced to 15 mmHg±7.2. There was no more than mild AR was reported on predischarge or first follow up transthoracic echocardiography.

3.3.2. Additional clinical outcomes

The median follow-up time was 2.9 years. Patient 1, with a background of atriopulmonary Fontan, experienced increased fatiguability and her mild PAR progressed to moderate at her 3.6 years follow up. Patient 5 died at 4 months post-TAVI from recurrent AoV endocarditis, patient 3 died aged 84 with congenitally corrected TGA at 7 months post-TAVI from sepsis and heart failure and patient 10 died from sepsis. Patient 4, who had severe pulmonary hypertension of 180 mmHg, is well, four years post TAVI. He had near normal systolic pulmonary artery pressures 2-years post-procedure on right heart catherization assessment and more recently had successful atrial flutter ablation. Incidentally at 4 months post TAVI, patient 9 (with a previous aortic homograft) was found to have extensive thrombus formation within the inferior aspects of all 3 aortic sinuses on CT which extended up to, but did not compromise the coronary ostia. There was a decrease in thrombus burden when scan was repeated at 1-year post TAVI, with no adverse or embolic events.

4. Discussion

The main finding of this study is that TAVI is feasible in ACHD patients, even though it is technically complex and challenging. Even in the very high-risk group described, where patients were not deemed good surgical candidates, there appears to be a favourable safety profile.

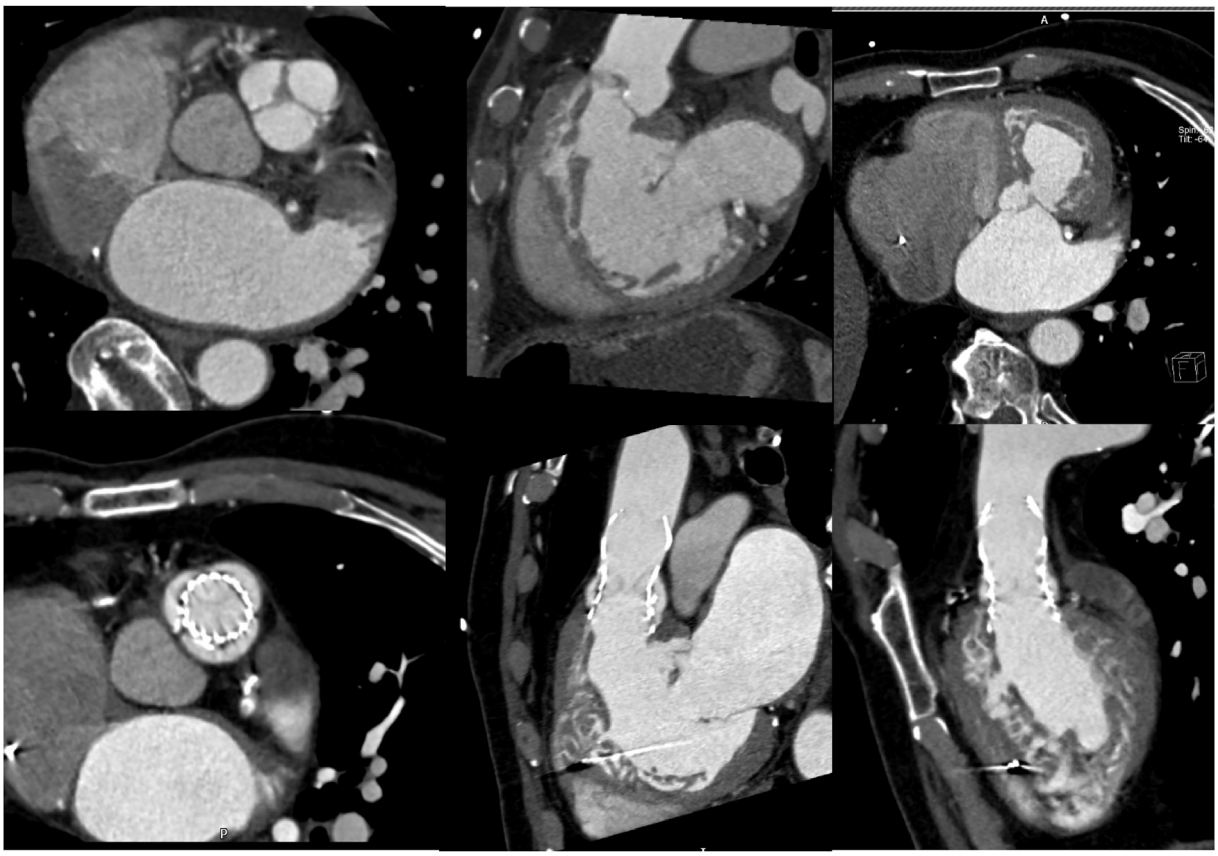

Fig. 5. CT images pre and post-procedure in a patient with corrected TGA.

4.1. Patient selection and pre-procedural planning

In this patient population, the use of the Society of Thoracic Surgeons score and EuroSCORE may not reflect the risk of AoV surgery in the presence of complex congenital anatomy [7–9]. At our centre decision-making was truly multi-disciplinary; the team involved in the pre-procedure planning consisted of structural and congenital heart disease interventionalists, congenital cardiothoracic surgeons, cardiac imaging specialists with expertise in structural and congenital imaging, and cardiac anaesthetists.

4.2. Choice of valves

The majority of our patients were young and did not have the classic degenerative calcific trileaflet AS that TAVI valves were designed for. Significant AoV lesions in the ACHD population can be due to either primary dysfunction as the result of an isolated AoV defect or part of a more complex condition or secondary dysfunction due to aortic root dilation, AoV leaflet prolapse or as a consequence of leaflet damage. Significant AoV deterioration may occur following valve repair or replacement with bioprostheses, homografts or autografts. Six patients had native BAV, 5 ViV-TAVI in failed bioprostheses (2 ViV were in homografts) and 2 severe AR.

A theoretical concern for TAVI is durability in younger patients. A recent study of 1403 patients reported long-term durability of TAVI [10]. 49 patients showed bioprosthetic valve failure with a 7-year cumulative incidence of moderate and severe structural valve deterioration of 7.0% (95% CI, 5.6%–8.4%) and 4.2% (95% CI, 2.9%–5.5%), respectively. However, the long-term assessment of durability is limited by the poor survival of the population beyond 5 years. More studies are needed in a younger and lower-risk patient population who have had undergone TAVI to determine long-term durability. Surgical and transcatheter bioprostheses share similar pericardial leaflet material and are both prone to Ref. [1] structural valve deterioration in the form of calcification, leaflet fibrosis, tear or flail [2], nonstructural valve deterioration in the form of PAR, size mismatch [3], thrombosis and [4] endocarditis [11]. To avoid the use of bioprosthetic valves, congenital surgeons tend to use Ross, valve sparing repair and more recently the Ozaki procedure for ACHD patients, but in the patients we describe, these options were not considered possible.

4.3. Procedural results

Passing the delivery systems through stented or surgically repaired coarctation did not cause significant problems, requiring only minor wire manipulation.

Valve positioning was problematic in two patients. Patient 3 with corrected TGA had a systemic right ventricle, with a muscular systemic right ventricular outflow tract and a large annulus requiring a 31 mm CoreValve. As this was not a recapturable and redeployable valve, we initially accepted a low position. Pulling the valve up with a snare was later successful. Patient 6 with a Fontan, single ventricle circulation and pure AR with a thin and mobile resuspended valve in a 31 mm tube graft presented a particular problem. The 34 mm Evolut R valve was being used outside of its design envelope, and moved down into the ventricle on release, probably because of the non-expandable nature of the prosthetic tube in which the valve had been resuspended – known as “melon-seeding”, and exacerbated by the absence of calcification. Whilst this was predictable (complication), the patient was young and deemed inoperable in another centre. The initial difficulty maintaining a pressure on

Table 2
Procedural data and outcome.

Paravaluvlar leak	3	2	2	1	0	2	0	0	2	0	2	0	1
Postgradient (peak/meanmmHg	21/11	22/11	23/14	39/23	30/18	28/13	31/18	22/12	22/12	55/34	25/7	28/10	24/10
Pregradient (peak/mean) mmHg	85/50	83/47	49/22	58/26	47/24	23/11	78/43	97/52	85/51	64/41	63/39	129/76	61/39
Post TAVI peak velocity m/s	2.3	2.4	2.4	3.0	2.8	2.7	2.8	2.4	2.3	3.7	2.5	2.6	2.4
Pre TAVI peak velocity m/s	4.6	4.6	5.6	3.8	3.4	2.4	4.4	5.0	4.6	4.0	4.0	5.7	3.9
Annulus* (mm)	30	24	29	19	24	33	22	25	28	29	26	28	26
TAVI Valve Size (mm)	29	23	31	29	23	34	26	26	29	29	29	26	29
TAVI Valve type	CoreValve	Sapien S3	CoreValve	CoreValve	Edwards Sapien XT	CoreValve	Evolut R	Evolut Pro	Evolut Pro	Evolut Pro	Evolut Pro	Evolut R	Evolut R
Pre TAVI morphology	Bicuspid	Bicuspid	Tricuspid	Bicuspid	Homograft tricuspid	Tricuspid	Bicuspid	Homograft tricuspid	Homograft tricuspid	Homograft tricuspid	Bicuspid	Perimount trileaflet	Repaired bicuspid
Original morphology	Bicuspid	Bicuspid	Tricuspid	Bicuspid	Bicuspid	Tricuspid	Bicuspid	Bicuspid	Bicuspid	Tricuspid	Bicuspid	Bicuspid	Bicuspid
NYHA (post)	1	1	1	1	2	1	1	1	1	1	2	1	1
NYHA (pre)	3	3	3	2	4	2	2	2	2	4	3	3	3
Hospital Stay days	10.0	6.0	16.0	7.0	28.0	35.0	8.0	2.0	2.0	11.0	4.0	2.0	3.0
Patient	1	2	3	4	5	6	7	8	9	10	11	12	13

bypass (subsequently improved with balloon occlusion) is likely to have contributed to the brain hypoperfusion injury. TAVI in native severe AR has been associated with increased embolization, migration and significant PAR due to the absence of annular and leaflet calcification, dilatation of the ascending aorta and increased stroke volume.

In homografts and stented surgical tissue valves, TAVI expansion can be limited. Some stented surgical valves can be post dilated with a Kevlar balloon to high pressure safely rupturing the surgical stent [12]. Heavily calcified degenerative homograft valve replacements do not always allow full expansion of the TAVI valve. ViV-TAVI is associated with coronary obstruction [13, 14] so careful pre-procedural planning is essential to identify high-risk features, such as low coronary heights, short virtual TAVI to coronary ostial distance, shallow sinuses of Valsalva and low sinotubular junction [15].

Patients with BAV morphology have increased risk of malpositioning, elliptical shape of the aortic annulus, eccentric valve calcifications and dilated ascending aorta [16–18]. Patient 2 had a highly eccentric BAV, as well as a calcified coarctation, (where the balloon expandable valve was a little high on one leaflet and a little low on the other), but crossing the coarctation was uncomplicated and there was mild PAR at 3 year follow up. (Our practice is to usually use self-expanding Medtronic valves for BAV as it provides a longer sealing zone, although this patient had a horizontal aorta, which led us to choose an Edwards valve.) New generation devices are being developed to improve outcomes by the development of external sealing cuffs, better deliverability, retrievability and repositioning capabilities or unique anchoring mechanisms [19–21].

4.4. Expansion of the TAVI population

To the best of our knowledge, this is the first study reporting the feasibility and effectiveness of TAVI in an ACHD population. Only three case reports have been published: 1)Turner's syndrome with severe symptomatic unicuspid AS, surgical correction of CoA with multiple comorbidities, 2) multiple surgical procedures for atrioventricular septal defect with severe AR and severe left ventricular function as a bridge to heart transplant and 3)surgically repaired double outlet right ventricle with severe AS and liver cirrhosis due hepatitis C in need of liver transplant [22–24].

The use of TAVI continues to grow for BAV, ViV treatment of degenerated bioprostheses and native AR. As the lifespan of ACHD patients increase, the need for procedures later in life will also increase, fuelling the rise of transcatheter interventions. This series includes young patients where valve longevity is important, as well as older patients with specific technical considerations, such as traversing coarctation repairs or stents, and complex aortic root anatomy, such as an anterior aorta attached to a muscular outflow tract, homografts and single ventricle physiology. TAVI is an appealing alternative to conventional open-heart surgery for those with severe AoV lesions, reducing sternotomies and blood transfusion. (Avoiding transfusion is particularly important as some young patients may be future candidates for heart transplantation.) A recent study with encouraging results showed ViV-TAVI had better short-term outcomes than redo SAVR (0.44–0.88; $p = 0.03$) and no significant difference of combined endpoints on long-term follow-up (18.6%/year vs. 21.9%/year; $p = 0.34$) [25].

However, careful evaluation and assessment of clinical factors and complex anatomy must be considered during the decision-making process in centres with expertise in both ACHD and TAVI. In the latest ACHD guidelines, TAVI treatment is a therapeutic option in “very rare cases with high surgical risk, when technically feasible” [26].

4.5. Study limitations

The aim of this study was to report initial outcomes of patients undergoing TAVI for complex congenital heart disease at a single institution and appears to be the first report of a series of TAVI procedures in this patient group. The major limitation is the relatively small number of

patients, and its retrospective nature.

5. Conclusions

TAVI appears to be feasible and effective in a group of ACHD patients with heterogeneous anatomical situations and age, deemed very high risk for conventional surgery by the MDT. Due to the variable nature of ACHD patients, large randomized trials are unlikely to be performed. Nevertheless, these patients have frequently had multiple cardiac operations at a young age and have much to gain by deferring or avoiding further sternotomies. Furthermore, many have a small annulus where a simple stented surgical AoV replacement decreases the effective orifice area. ACHD patients should be considered as potential candidates for TAVI if AoV intervention is needed.

5.1. Impact on daily practice

TAVI for ACHD patients requires multidisciplinary team discussion, meticulous imaging and adequate back up during the procedure. TAVI appears to be a viable alternative treatment option when surgery is not a good option. A larger number of patients and long-term follow-up studies are needed to confirm the safety and durability of this method and guide patient selection.

Declaration of competing interest

The authors declare the following financial interests/personal relationships which may be considered as potential competing interests: Dr Turner has served as a consultant and proctor for Abbott Vascular, Edwards Lifesciences Corporation and Medtronic; is a consultant for Occlutech and Medtentia and received educational grants from Gore. Dr Dorman has received honoraria for advisory boards and conference attendance fees from Abbott Vascular, Boston, Medtronic and Edwards Lifesciences Corporation. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ijcchd.2021.100116.

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