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REVIEW

Aortic annulus sizing in stenotic bicommissural non-raphe-type bicuspid aortic valves: reconstructing a three-dimensional structure using only two hinge points

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Abstract

Bicuspid aortic valve (BAV) anatomy is becoming an increasingly frequently encountered challenge in transcatheter aortic valve implantation (TAVI). Bicommissural non-raphe-type BAV (Sievers and Schmidtke Type 0) is composed morphologically of two aortic cusps with no raphe and is less common than the tricommissural or bicommissural raphe-type configurations. Precise annular sizing is a key step for successful TAVI in BAV. The challenge in bicommissural non-raphe-type BAV is that a three-dimensional structure has to be reconstructed using only two anatomical hinge points. For this reason, available software are limited when it comes to bicommissural non-raphe-type BAV. We propose that manual assessment of the aortic root in bicommissural non-raphe-type BAV using multi-planar reconstruction (MPR) software can be performed successfully by aligning the two available hinge points and measuring the smallest identifiable annular dimensions in the transverse plane (Fig. 1). We identified 12 patients with bicommissural non-raphe-type BAV undergoing TAVI between January 2013 and December 2017 in our high-volume institution. Our novel sizing strategy was employed prospectively in three patients—with good clinical outcomes—and evaluated retrospectively in the remainder (Table 1). No patient suffered a central major vascular complication or required new permanent pacemaker implantation. Device success occurred in all patients except one (post-procedural echocardiographic transvalvular gradient of 23 mmHg). In the retrospectively assessed cases, the novel annulus measure was concordant with the implanted THV size in 7 out of 9 procedures and, importantly, did not overestimate the annulus dimensions in any case. Furthermore, in two balloon-expandable THV cases the new measure may, in retrospect, have prompted consideration of a smaller implant size. To be noted, balloon sizing of the aortic annulus has additional value when selecting the valve size in BAV anatomy. Further prospective validation of this novel MDCT sizing technique is required.

Keywords Transcatheter aortic valve implantation · TAVI · Aortic annulus sizing · Bicuspid aortic valve · Computed tomography

Antonio H. Frangieh and Jonathan Michel are equally contributed to this work.

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Introduction

Bicuspid aortic valve (BAV) is the most common congenital cardiac anomaly in the general population, with an estimated incidence of 0.4–2.25% [1], and a strong association with subsequent valvular dysfunction. In some reported series, almost 50% of stenotic aortic valves excised surgically have been found to be congenitally bicuspid [2].

Morphologically, BAV covers a spectrum of anatomical abnormalities; however, by definition there are less than three functional leaflets. Whilst several classification systems for BAV have been proposed [3] including a widely utilized anatomically-derived numerical grading system developed by Sievers and Schmidtke [4], a consensus amongst transcatheter aortic valve implantation (TAVI) operators has yet to be agreed upon. More recently, Jilaihawi et al. have published a simplified BAV classification developed specifically for TAVI based on the number of commissures, the presence or absence of raphe, and fusion pattern of the aortic cusps [5].

Bicommissural non-raphe-type BAV (Sievers and Schmidtke Type 0) is composed morphologically of two aortic cusps with no raphe and is less common than the tricommissural or bicommissural raphe-type configurations (Fig. 1).

In comparison with tricuspid aortic valves, BAV is associated with larger annulus dimensions, asymmetric calcification and dilatation of the ascending aorta [6–9]; factors traditionally deemed unsuitable—or of uncertain suitability—for TAVI. Patients with BAV have been excluded from all major trials evaluating outcomes of TAVI against surgical aortic valve replacement (SAVR) [4, 10–12].

However, data from registries and observational reports have shown generally favorable results in this population [13–19], particularly with newer generation devices [20].

Given the trend toward treatment of younger and lower risk patients with TAVI [12, 21, 22], BAV anatomy is becoming an increasingly frequently encountered challenge.

The aortic valve annulus typically represents the tightest part of the left ventricular outflow tract (LVOT) and is defined, in the case of tricuspid aortic valves—or functionally bicuspid valves with three identifiable cusps—as a virtual ring with three anatomical anchor points (hinge points) at the base of each of the attachments of the aortic leaflets [23].

In tricuspid aortic valves, the LVOT tissue at the height of the aortic hinge points provides the anchoring necessary for stable implantation of transcatheter heart valves (THV) [24]. Of note, it has been suggested by some operators that the narrowest anchoring point within stenotic BAVs may theoretically occur above the level of the annular plane. The upper LVOT is usually elliptical, and often irregular, in shape. As a result, proper imaging and accurate measurement of the aortic valve annulus is essential to avoid undersizing or oversizing of the THV [25].

Undersizing is associated with higher risk of significant paravalvular leak (PVL) [26] or valve embolization [27]. In contrast, oversizing may increase the risk of annular rupture, coronary obstruction, conduction disturbances requiring long-term pacing, or, when associated with underexpansion of the prosthesis, may lead to reduced valve durability [26, 28–31].

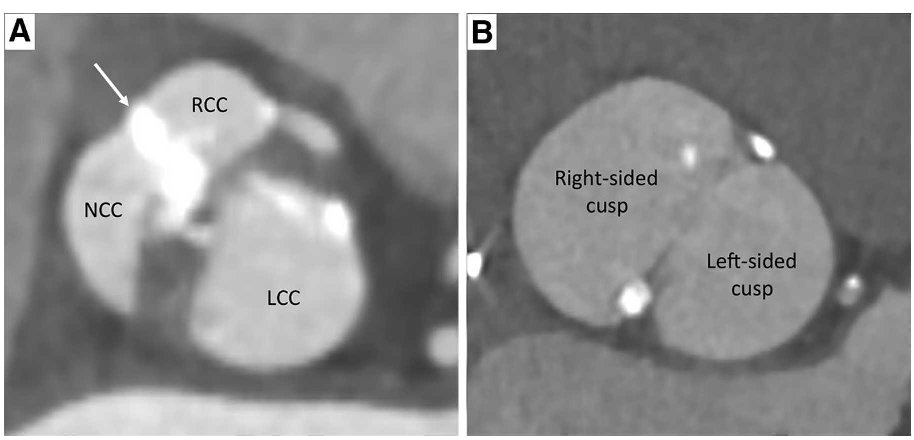


Fig. 1 Bicommissural bicuspid valves. **a** Bicommissural raphe-type BAVs are identified by a congenital fusion (raphe) of one of the commissures with three identifiable cusps. Fusion may occur between any of the three cusps and involves the lower third of the commissure nearest to the base of the cusps. In this example, a calcified raphe is present between the right- and non-coronary cusps (white arrow). **b** Bicommissural non-raphe-type BAVs have two identifiable cusps. A raphe is not present. *BAV* bicuspid aortic valve, *RCC* right coronary cusp, *LCC* left coronary cusp, *NCC* non-coronary cusp

Echocardiography, multidetector computed tomography (MDCT), and angiography represent the three cornerstones of aortic root assessment in TAVI [32]. However, annulus sizing in bicommissural non-raphe-type BAV can be especially challenging. In particular, the absence of a third aortic cusp hinders the use of most standard multi-planar reconstruction (MPR) software (with the 3 planes locked at a 90° angle) and manual annulus alignment techniques.

In this paper, we review the available modalities for aortic root assessment in bicommissural non-raphe-type BAV, describe a standardized and simplified manual MPR technique for annulus sizing using MDCT, and present an example step-by-step minimalist TAVI procedure in this particular aortic valve anatomy.

Echocardiography

Echocardiography plays an essential role during case selection for TAVI and long-term assessment of THV function. Aortic stenosis severity, aortic valve and root morphology, ascending aorta dimensions, left ventricular function, concomitant valvular disease, and pulmonary hypertension may all be assessed echocardiographically [33].

There is a lack of published data for the utility of echocardiography in bicommissural non-raphe-type BAV. In tricuspid valves, aortic annulus sizing is commonly performed using transthoracic echocardiography (TTE) and/or transesophageal echocardiography (TEE) [33, 34].

Echocardiographic measurement of the LVOT diameter can be performed using two-dimensional (2D) images acquired during systole in the 3-chamber long-axis view and measuring at the height of the hinge points of the aortic valve, with a trailing edge to leading edge convention [33, 34]. The annulus dimensions can be extrapolated from the LVOT diameter; however, due to the difficulty obtaining an ideal plane through the aortic root and the elliptical shape of the virtual ring, 2D echocardiography tends to underestimate the size of the aortic annulus [35–37].

The development of real-time 3D echocardiography enabled high spatial resolution volume acquisition of the aortic root and improved visualization of the virtual basal ring. Assessment of coronary height and anatomical landmarks associated with the aortic valve can be performed more accurately with 3D TEE [36]. Annulus measurements derived from 3D TEE have been reported to correlate well with MDCT [38] although other investigators have found that 3D TEE, whilst performing better than 2D, results in persistent underestimation of LVOT dimensions [39, 40]. More recently, the use of automatic algorithm-derived annulus diameter measurements from 3D TEE images have been shown to perform as well as those derived manually [41].

Importantly, however, high spatial resolution imaging in 3D TEE involves an inevitable trade off with the size of the imaging sector and with the temporal resolution [42]. Furthermore, BAV show accelerated and asymmetric calcification compared to tricuspid aortic valves [43, 44] and bicommissural non-raphe-type BAVs tend to be more heavily calcified than other BAV types [5], often limiting the ability of both 2D and 3D echocardiography to adequately visualize the aortic annulus. Accordingly, MDCT has gained acceptance as the preferred modality for assessing aortic annulus dimensions in calcific aortic stenosis, including BAV [45].

Multidetector computed tomography

Modern MDCT scanners are well suited to assessment of the aortic root due to their high spatial and temporal resolution and ability to obtain a comprehensive 3D dataset. Thoracic evaluation with an electrocardiogram-gated multi-phase acquisition contrast-enhanced MDCT—with a minimum 64-slice capability and spatial resolution of 0.5–0.6 mm—is the recommended investigation of choice to assess the annulus and aortic root prior to TAVI [32, 45].

It should be noted that, in selected cases, magnetic resonance imaging provides accurate annulus sizing and may be used in patients in whom MDCT is contraindicated [39]; however, the extent and distribution of calcification cannot be determined using this modality.

Multi-planar MDCT assessment of the tricuspid aortic valve using the “turnaround” rule technique to identify the three aortic cusp hinge points and manually determine the annular plane is well established [23]. In addition, dedicated MDCT image reconstruction software for pre-TAVI sizing and planning have been developed that are capable of identifying three different hinge points and enable fully, or semi-automated, alignment of the virtual ring plane and sizing of the aortic annulus [46, 47].

The challenge in bicommissural non-raphe-type BAV is that to achieve proper identification and alignment of the annular plane, a three-dimensional structure has to be reconstructed using only two anatomical hinge points. For this reason, available software are limited when it comes to bicommissural non-raphe-type BAV, in which only two hinge points are present. MDCT has been used to compare aortic root characteristics in raphe-type BAV with tricuspid aortic valves from a large cohort of patients treated for severe aortic stenosis [44]. A semi-automated reconstruction approach was employed and the annulus plane was determined using three aortic cusp hinge points; however, patients with bicommissural non-raphe-type BAV were not described in this cohort.

Reconstruction of the aortic root also allows automated identification of a geometric centerline passing through the middle of the LVOT, aortic sinuses, and ascending aorta. Identifying a plane perpendicular to the centerline at the level of the aortic hinge points has been proposed as a means of measuring the virtual basal annulus in BAV [3]. However, this technique may not achieve optimal alignment of both hinge points given the variability in cusp symmetry and depth in BAV [44].

Operators may choose to rely on manual measurement to define the annular size in bicommissural non-raphe-type BAV using MPR software integrated into the majority of workstations used for cardiac imaging. There is no consensus regarding the optimal means of identifying the annular plane manually in this anatomy, however, in the manual MDCT sizing section below, we describe a novel technique that may simplify this task.

Angiography

Angiographic determination of the aortic annulus size has been employed since the advent of TAVI. The heterogeneity of the aortic annulus, unpredictable influence of heavy or asymmetric calcification, and the potentially compliant nature of the tissues adjacent to the aortic valve, annulus, and LVOT have prompted the use of inflated valvuloplasty balloons as a means of determining the annulus size and assessing the mechanical properties of the aortic root structures [32].

In this 'balloon sizing' technique, aortic valvuloplasty is performed in conjunction with rapid ventricular pacing using a balloon of pre-established size, with or without manometry, whilst a contrast volume of 10–15 ml at a flow rate of 10 ml/s is simultaneously injected into the aortic root [32, 48–50].

Balloon sizing during rapid pacing-induced ventricular standstill enables evaluation of the annulus dimensions independent of variability associated with the normal cardiac cycle. A balloon size close to the anticipated mean annulus diameter based on preprocedural imaging can mimic the mechanical properties of an implanted valve and enable angiographic assessment of the annulus dimensions, anticipated movement of bulky calcium deposits, displacement of aortic cusp leaflets, and confirmation of coronary artery patency [32].

For balloon-expandable THV, the American College of Cardiology recommends balloon sizing when the predicted annulus dimensions are borderline between the ranges of two available prosthesis sizes [45] and the same technique may also assist selection of self-expanding valve size.

In all BAV types, routine use of balloon sizing may be particularly important in view of the difficulty in accurately determining the aortic annulus dimensions preoperatively using MDCT or echocardiography, and the increased risk of post-implant paravalvular leak [17, 51]. Furthermore, BAV is generally associated with enlarged sinus of valsalva width, however, shallow sinus depth (effaced root) is seen in a minority of cases [44]. Shallow sinus width, in combination with longer leaflet length potentially increases the risk of coronary occlusion especially when the coronary ostia originate from a low position within the aortic root. In this situation, balloon sizing may also improve advanced identification of this serious complication.

A novel technique for manual MDCT sizing of the bicommissural non-raphe-type BAV virtual annulus

Reliable identification of the virtual basal plane is key to accurate annulus sizing in bicommissural non-raphe-type BAV. In the following text we propose a simplified technique to perform manual annulus sizing using electrocardiogram-gated contrast-enhanced MDCT with a minimum 1 mm slice thickness (Fig. 2 and Supplementary Movie 1).

First, the longitudinal axis in the coronal and sagittal plane should be aligned with the axis of the aortic root (Fig. 2a) achieving a double oblique plane view in the transverse plane window. Identify the hinge point of one of the two aortic cusps in the transverse plane window by moving the crosshairs up and down through the valve (Fig. 2b). Place the transverse plane crosshairs on the identified hinge point and rotate the axis so that either the coronal or sagittal plane axis intersects the second cusp (Fig. 2b). Tilt the corresponding plane in the coronal or sagittal window to identify the second hinge point in the transverse plane window (Fig. 2c). Adjust the rotation of the crosshairs in the transverse plane window, if necessary, so that both hinge points are aligned on one of the axis lines (Fig. 2c). In contrast with tricuspid aortic valves, there is no third hinge point with which to align the final plane in bicommissural non-raphe-type BAV. In this case, to achieve accurate annulus measurement with no oversizing, tilt the sagittal or coronal plane (only manipulate the plane that maintains the previous hinge point alignment in the transverse window) and visually identify the smallest annulus area/diameter in the transverse plane window (Figs. 2d, 3). When this adjustment is achieved, the measurement can be performed in this plane (Fig. 2d) or 1 slice lower in the upper outflow tract. If it is difficult to determine the smallest area/diameter visually then make multiple measurements at different angles and use the smallest values obtained (Fig. 2d).

Depending on the software employed, the implantation angle corresponding to the two aligned hinge points can be

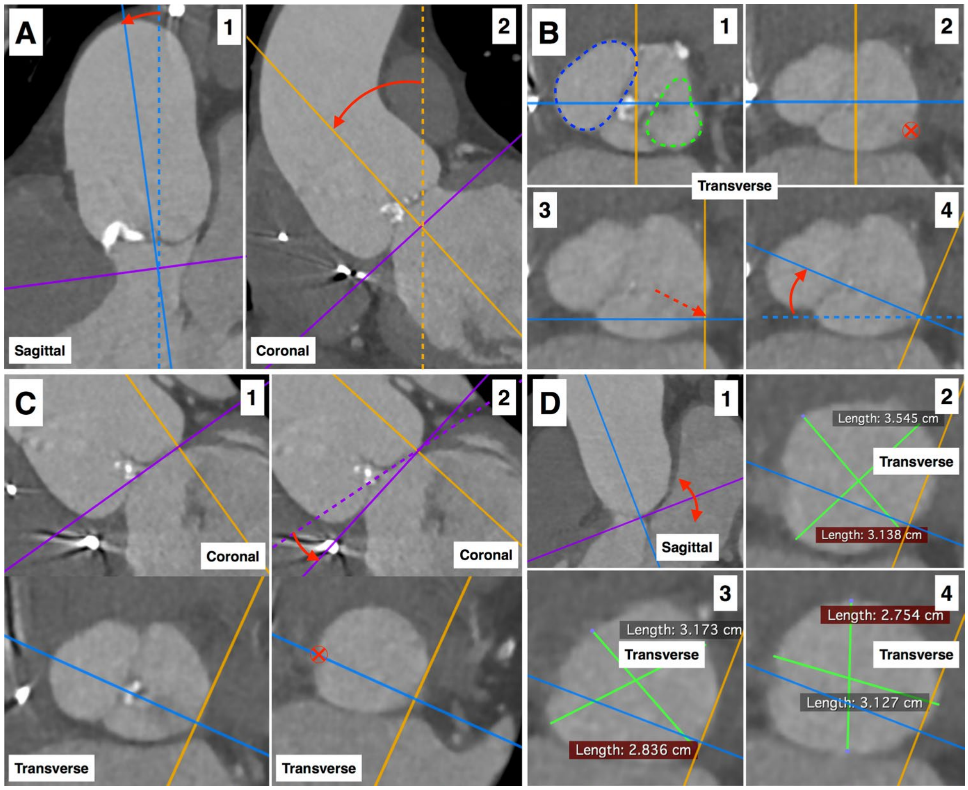


Fig. 2 MDCT annular sizing in bicommissural non-raphe-type BAV. **A.1 + 2** Align the coronal and sagittal planes with the aortic root and place the cursor in the center of the valve. **B.1** Scroll through the valve to identify the cusps (dotted green line = left-sided cusp, dotted blue line = right-sided cusp) and find one of the hinge points (red cross **B.2**). Position the crosshairs on the hinge point (**B.3**) and align the sagittal plane line (blue) to intersect the other cusp (**B.4**). **C.1** Identify the plane showing a transection of the two cusps (coronal plane in this example) and tilt the sagittal plane line (blue) in this window to locate and intersect the second hinge point (red cross **C.2**). **D.1** Finally, tilt the transverse plane line (purple) in the sagittal window and identify the smallest annulus dimensions. Measure the annulus size on the plane where the annulus has the smallest area/diameter (**D.4**) or perform repeated measures at different angles to find the smallest dimensions (**D.2-4**)

easily identified from the angles shown in the coronal plane window (Fig. 2 and Supplementary Movie 1).

In Fig. 4 and Supplementary Movie 2, we present a TAVI procedure for a patient with stenotic bicommissural non-raphe-type BAV. MDCT measurements are shown in Fig. 4d and Supplementary Movie 1. The annulus is nearly circular with minimal and maximal diameters of 27.5 and 31.3 mm respectively, perimeter of 93 mm and annular area of 682 mm^2. Figure 4a clearly shows the bicommissural non-raphe-type BAV anatomy on the root-shot aortic angiography. The implantation angle was determined based on the MDCT predicted angle as described above. In this case, retrograde crossing of the stenotic aortic valve was challenging and was performed using a straight tip 0.035″ glide-wire through a 5 Fr AL-2 catheter (Fig. 4b). MDCT annular sizing suggested suitability of an overexpanded 29 mm SAPIEN 3 valve (Edwards Lifesciences, Irvine, California), and balloon sizing using a 25 mm balloon was performed for more accuracy and safety (Fig. 4c). It showed that the balloon was much smaller than the root anatomy, away from hinge points, and confirmed the requirement for a SAPIEN 3 valve larger than 26 mm. With the crimped 29 mm valve in position, the coaxiality of the prosthesis stent frame should be checked and the coplanar view should be adjusted if needed, before valve deployment [51] (Fig. 4d). The proximal end of the stent frame should cover the cusps and the distal one should cover the annulus. In other terms, the balloon middle marker should be located slightly above a line in between the two hinge points of the cusps. After implantation of a 29 mm SAPIEN 3 valve (overexpanded with an additional 2 ml of volume in the balloon), the coaxial control showed that the valve was well-expanded and in a good position (Fig. 4e), along with excellent functional result with trivial PVL (Fig. 4f). Pre-discharge transthoracic echocardiography confirmed trivial PVL and a mean transvalvular gradient of 12 mmHg. Post-procedural non-contrast MDCT showed circular deployment of the prosthesis despite heavy calcification, with complete coverage of the virtual annulus (Fig. 5).

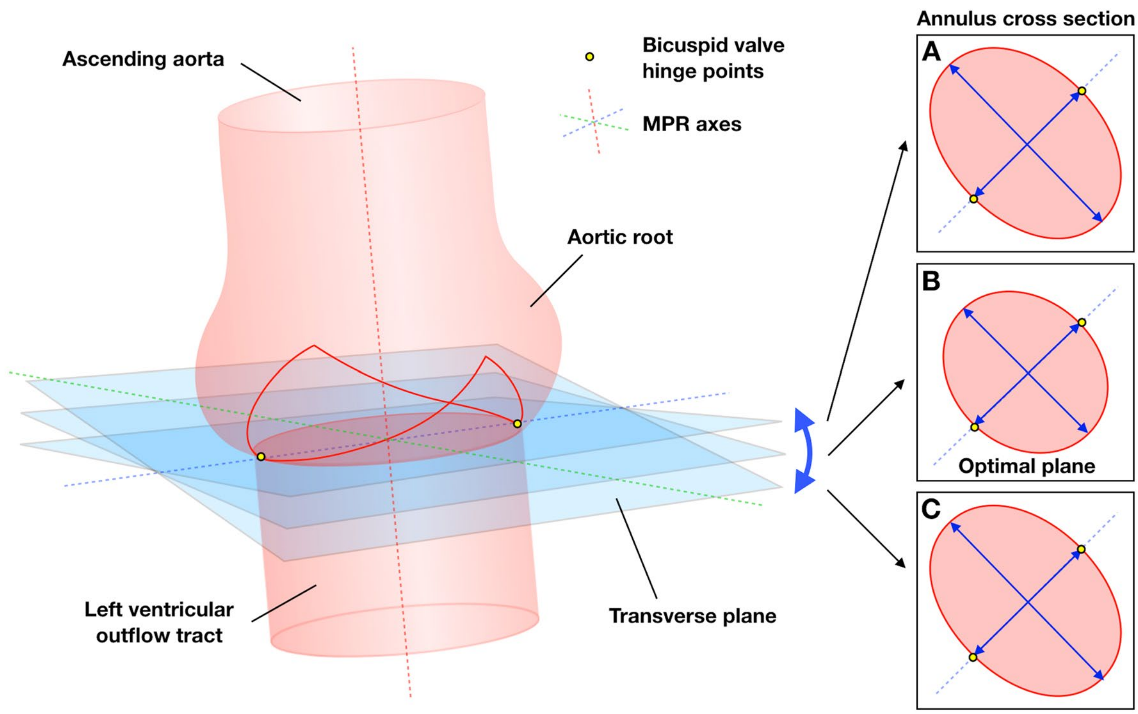


Fig. 3 Schematic representation of the bicuspid aortic root and annulus demonstrating the principal of using multi-planar reconstruction to determine the annular plane. After aligning the transverse plane as described in Fig. 2a–c, tilt the transverse plane whilst maintaining axis alignment through both hinge points. Oblique planes through the virtual annulus (**a, c**) have larger dimensions than the optimal annular plane (**b**)

Sizing evaluation in a bicommissural non-raphe-type cohort

A total of 12 patients with bicommissural non-raphe-type BAV undergoing TAVI between January 2013 and December 2017 were identified in our center. Our novel sizing strategy was employed prospectively in three patients—with good clinical outcomes—and evaluated retrospectively in the remainder (Table 1). No patient suffered a central major vascular complication or required new permanent pacemaker implantation. Device success occurred in all patients except one (post-procedural echocardiographic transvalvular mean gradient of 23 mmHg). In the retrospectively assessed cases, the novel annulus measure was concordant with the implanted THV size in 7 out of 9 procedures and, importantly, did not overestimate the annulus dimensions in any case. Furthermore, in two balloon-expandable THV cases (Table 1: Case ID 4 and 10) the new measure may, in retrospect, have prompted consideration of a smaller implant size. Certainly, the implanted THV in case 4 appears angiographically excessively oversized, although annulus rupture did not occur. It should be noted that we recommend the routine use of balloon sizing in all BAV cases, as described above, to confirm accurate sizing in these particular anatomical subgroups. We acknowledge that the small number of patients included in this cohort, due to the rarity of this anatomical configuration, represents a limitation of the current study.

Although we do not routinely perform TEE during TAVI, including BAV cases, further consideration should be given to validation of this technique in 3D TEE.

Conclusion

Precise annular sizing is a key step for successful TAVI in BAV. Manual assessment of the aortic root in bicommissural non-raphe-type BAV by means of MDCT and using MPR software can be performed by aligning the two available hinge points and measuring the smallest identifiable annular dimensions in the transverse plane. This measurement strategy showed good concordance with implanted THV size in a small prospective and retrospective sample of patients with bicommissural non-raphe-type BAV treated with TAVI and, in combination with intraprocedural balloon sizing, may reduce the risk of annulus size overestimation in these patients. Further prospective validation of this novel MDCT sizing technique is required.

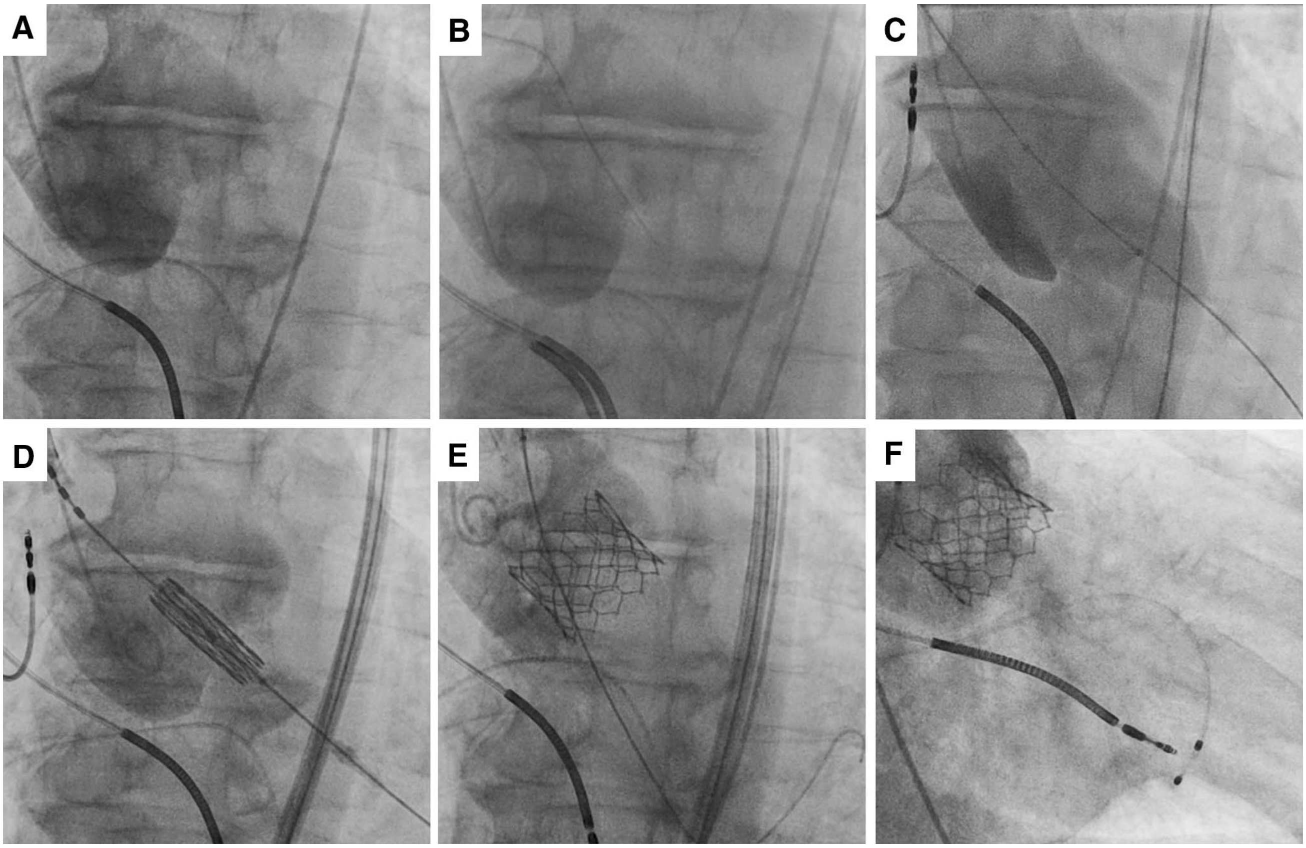


Fig. 4 stepwise description of a TAVI procedure in bicommissural non-raphe-type BAV. **a** Root-shot aortic angiography: Alignment of the two hinge points based on MDCT predicted angle. **b** Crossing with an AL 2 catheter because of its wider radius. **c** 25 mm Balloon Sizing: The balloon is much smaller than the root anatomy, away from hinge points. **d** Valve placement: coplanar alignment of the Stent frame, proximal stent frame should cover the cusps, distal stent frame should cover the annulus. **e** Coaxial control showing good valve position and expansion. **f** Functional result in RAO-30° view with 30 ml contrast and 15 ml/s flow showing none/trivial paravalvular leak

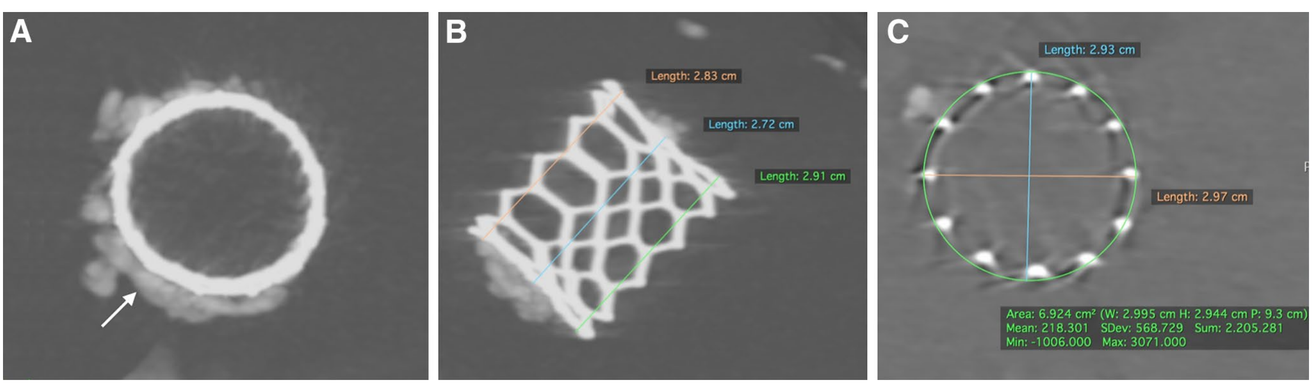
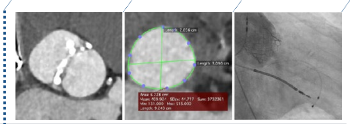
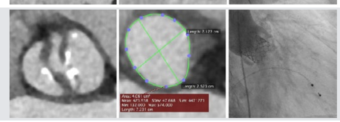
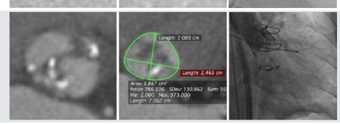
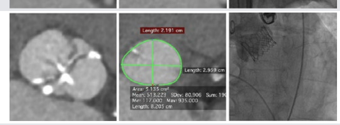
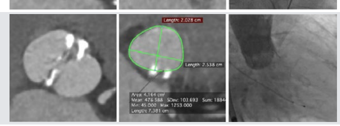
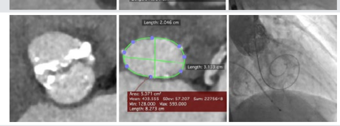
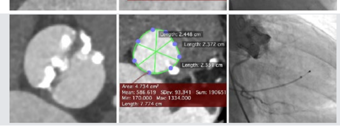
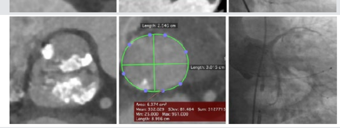
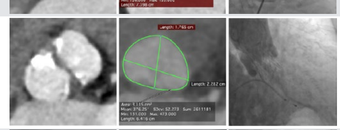
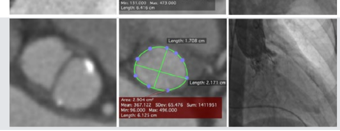


Fig. 5 Post-procedural non-contrast MDCT of the implanted 29 mm Sapien 3 THV. **a** Circular deployment of the prosthesis is achieved despite heavy calcification (white arrow). **b** Measurement of the inflow [green], middle [blue], and outflow [orange] portions of the valve demonstrate complete deployment. **c** Area and diameter measurements show complete coverage of the virtual annulus

Table 1 Baseline characteristics, annular dimensions, and procedural data for 12 patients with bicommissural non-raphe-type BAV undergoing TAVI

Case ID	Age	Gender	STS score	LVEF	Cusp fusion type	Ann. min. diameter	Ann. max. diameter	Ann. mean diameter	Ann. area (mm^2)	Implanted THV	THV size (mm)	Postdilatation	Residual gradient	PVL grade	Annular rupture	New PPM	CT valve morphology	CT annulus measurements	Post-implant aortography
1	71	M	1.0	39	M	27.5	31.3	29.4	682	Sapien 3	29	No	12	0	0	N/A			
2	71	F	1.0	60	C	21.2	24.7	23.0	422	Sapien 3	23	+2 ml	19	0	0	0			
3	72	F	4.9	22	C	18.1	24.3	21.2	342	Sapien 3	23	No	16	0	0	0			
4	76	F	5.1	60	M	20.9	24.6	22.8	387	Sapien XT	26	No	7	0	0	0			
5	64	M	1.4	28	M	21.9	29.6	25.8	514	Sapien 3	26	+2 ml	6	0	0	0			
6	87	F	5.0	50	M	20.3	25.4	22.9	416	Evolut R	26	No	6	1	0	0			
7	76	M	1.8	65	M	20.5	31.3	25.9	507	Sapien 3	26	No	23	1	0	0			
8	80	F	2.5	43	M	23.7	25.5	24.6	473	Lotus	25	No	15	0	0	0			
9	66	M	0.8	60	M	25.5	30.2	27.9	637	Sapien 3	29	No	11	1	0	0			
10	54	F	0.9	65	M	19.7	26.1	22.9	401	Sapien 3	26	No	13	0	0	0			
11	73	F	1.6	60	C	17.7	22.8	20.3	312	Evolut R	26	No	8	0	0	0			
12	78	F	3.1	60	M	17.1	21.7	19.4	290	Core Valve	23	No	10	0	0	0			

All CTs were evaluated using the novel annulus measurement technique described above. In cases 1–3 the annulus dimensions were used to guide selection of THV size, whereas in cases 4–12 the annulus size and subsequent implanted THV were assessed in retrospect. No central major vascular complications or new PPM implant occurred. Valve academic research consortium device success was achieved in all but one patient, due to a persistent transvalvular mean gradient of 23 mmHg (Case 7). Annulus dimensions are shown in mm

Table 1 (continued)

Cusp fusion: *C* fusion of the left and right coronary cusps, *M* mixed fusion of either the left or right cusp or the non-coronary cusp

STS Society of Thoracic Surgeons, *LVEF* left ventricular ejection fraction, *THV* transcatheter heart valve, *PVL* paravalvular leak, *PPM* permanent pacemaker

Compliance with ethical standards

Conflict of interest AHF is a proctor and medical consultant for Edwards Lifesciences. AMK is a proctor and medical consultant for and receives research support from Edwards Lifesciences. SB is a proctor and medical consultant for Medtronic. The other authors have no conflict of interest to disclose.

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