Case Report

Transcatheter aortic valve implantation in a patient with type 0 bicuspid aortic valve with severe stenosis and calcium extending in to LVOT

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Abstract

Transcatheter aortic valve implantation is the most preferred treatment for aortic stenosis in elderly patients at high surgical risk. We present a case experience of Transcatheter aortic valve implantation using a balloon-expandable Myval transcatheter heart valve in a Type 0 bicuspid aortic stenosis.

Introduction

Transcatheter aortic valve implantation (TAVI) is well-established in patients with severe symptomatic aortic stenosis (AS) with high surgical risk for conventional aortic valve replacement. However, this recommendation was based on clinical trials that excluded patients with bicuspid aortic valve (BAV),[1,2] a common cardiac anomaly present in 0.5-2% of the general population and associated with the development of AS that requires intervention.[3] Though there are different classification of BAV, the most widely used classification is the surgical one, described by Sievers and Schmidtke, which defines three types of BAVs based on the presence, number and spatial orientation of the raphe: BAV type 0 (no raphe); type 1 (one raphe), and type 2 (two raphes).[4]

The most common type (88%) is Sievers 1 with raphe between left and right coronary cusps (71%). This type is particularly related to asymmetric annular geometry (oval shape of the annulus) and presents predominantly with stenosis (insufficiency in about 26–31% of specimens). On the other hand, Sievers types 0 and 2, which are quite rare (7% and 5% respectively), present in similar proportions with stenosis and insufficiency.[5] Regardless of the classification used, to date, no clear correlation has been demonstrated between BAV phenotypes and clinical outcomes after TAVI.[6]

TAVI procedure in BAV anatomy is challenging as the Bicuspid Aortic valve is associated with: larger dimensions of the aortic valve components (Aorta[7], Sinus of Valsalva[8], Annulus.[9]); aortopathy[7] and horizontal aorta;[6] less elliptical aortic annulus;[9] heavily calcified leaflets8 and raphe;[7] eccentric calcification;[9] valve asymmetries due to raphe and different calcification patterns;[10] higher take-off of the left main coronary ostium;[8] separate coronary ostia in the left system;[8] and left coronary artery dominance[8] compared to tricuspid aortic valve.

Because BAV has several anatomic features that can complicate the accurate device delivery and apposition of the prosthetic valve during TAVI,[11,12,13] and can also increase the risk of TAVI procedural complications or failure,[14] it is also associated with higher rates of paravalvular leak;[2] higher risk of permanent pacemaker;[7] higher risk of need for a second TAVI valve;[7] risk of annulus rupture or aortic dissection, stroke;[7] risk of aortic injury;[10] valve misalignment or under-expansion during valve deployment;[10] and early valve deterioration.[10]

Therefore, proper planning of the TAVI procedure is important for successful TAVI in the BAV case. Outcomes of TAVI in patients with BAV using new-generation TAVI valves have shown promising results.[15,16,17] The Meta-Analysis and Trial Sequential Analysis suggest that the outcome of TAVI in stenotic BAV patients is largely comparable to stenotic TAV patients.[18] Typically, the TAVI valve is selected based on the patient's clinical features.[19] BEAT Registry[20] is the only study that has attempted to directly compare the safety and efficacy of BEV (Sapien3) versus SEV (Evolut R/PRO) in the context of BAV stenosis, revealing similar rates of device success (BEV 85.7% vs. SEV 84.4%, p = 0.821) and clinical outcomes up to 1 year. Better hemodynamic performance was observed among patients treated with SEV, but also a higher rate of significant PVL (BEV 0% vs. SEV 9.3%, p = 0.043).

In this case report, we present a case of TAVI in a type 0 bicuspid aortic valve severe aortic stenosis using BEV Myval THV at Shahid Gangalal National Heart Center, Kathmandu, Nepal.

Case Report

A 78-year-old female was referred for TAVI. She complained of shortness of breath on exertion and a few episodes of syncope. ECG showed a normal sinus rhythm. The echocardiographic assessment revealed a type 0 bicuspid aortic valve with peak aortic velocity = 590cm/sec and gradient (peak pressure gradient 139 mmHg and mean pressure gradient of 91 mmHg) (Fig. 1) with a calculated aortic valve area of 0.4 cm^2, LV concentric hypertrophy with normal left ventricular ejection fraction.

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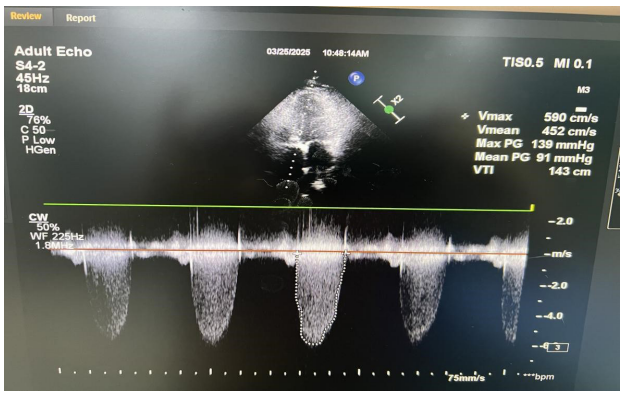

Figure 1: Echocardiogram

Multi-slice computed tomography (MSCT) revealed a Bicuspid Type 0 Aortic Valve. Severe Aortic Valve Calcification. Annular Calcification Extending into LVOT. The mean aortic annulus diameter was 21.4 mm, the annulus perimeter was 66.6 mm and the annulus area was 343.9 mm2 as shown in Fig. 2.

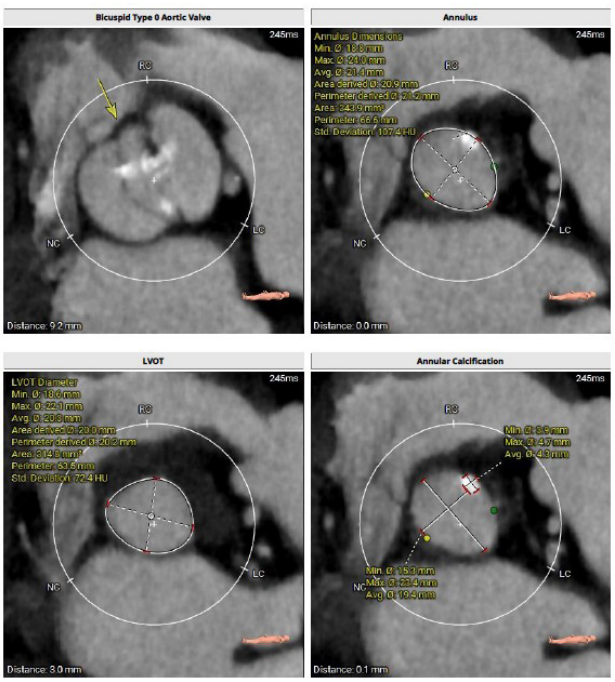

Figure 2: MSCT of Annuals and LVOT

The sinus of Valsalva's mean diameter was 31.6 mm. The heights of the right and left coronary ostia were 15.5 mm and 10.0 mm, respectively. The mean diameter of the sinotubular junction was 35.7 mm. The mean diameter of the ascending aorta was 41.2 mm, as shown in Fig. 4. Mild calcification was noted at the arch of the aorta and descending aorta, with an aortic angle of 64 degrees. The average diameters of the right common iliac (7.9 mm), external iliac (7.5 mm), and femoral (6.6 mm) arteries were found to be normal. The average diameters of the left common iliac (9.3 mm), external iliac (7.6 mm), and femoral (7.9 mm) arteries were also found to be normal.

The choice of treatment options and their benefits was discussed with the patient and her family, and they opted for TAVI. Written informed consent was obtained from both the patient and his family.

A thorough assessment by a comprehensive heart team, consisting of a cardiac surgeon, anesthesiologist, and cardiologist, ensured that all necessary conditions for the intervention were met. The patient then underwent TAVI under local anesthesia and intraoperative transthoracic echocardiography (TTE) imaging guidance. The right and left femoral arteries and the right femoral vein were cannulated. A temporary pacemaker was inserted into the right ventricle through the right femoral vein. To guide valve placement and facilitate arteriography for positioning, a 6Fr pigtail catheter was introduced through a 6Fr sheath into the noncoronary sinus via the left femoral artery. The transcatheter heart valve (THV) was delivered through the right femoral artery. After achieving all necessary arterial and venous accesses, intravenous unfractionated heparin was administered to maintain an activated clotting time of over 250 seconds. Using the right femoral artery access, a J-tipped, soft 0.035 mm wire was advanced into the descending thoracic aorta. Following the procedure, two suture-mediated closure devices (Perclose ProGlide, Abbott Cardiovascular, Abbott Park, IL, USA) were deployed for pre-closure while maintaining arterial access through the J-tipped guidewire. Subsequently, a 14Fr Python expandable introducer sheath (Meril Life Sciences Pvt. Ltd., India) was advanced into the femoral artery. A 6Fr AL-1 catheter was threaded through the valve delivery sheath using a 145–150 cm 0.035-inch J-tipped guidewire. It was then switched to a straight-tip wire to navigate through the valve. After crossing the valve, the straight-tip wire was exchanged for a 300 cm J-tipped wire. The AL-1 catheter was replaced with a 6Fr angled pigtail catheter. A reshaped stiff guidewire, SAFARI 2™ (Boston Scientific, Marlborough, MA, USA), was advanced through the pigtail catheter into the left ventricle, positioning the guidewire's transition point above the apex, pointing away from the ventricular wall. The septum was pre-dilated with an 18X40mm Mammoth balloon. A 21.5 mm BE Myval THV was implanted under pacing at 180 beats/min. The placement was verified through a root aortogram using a pigtail catheter before and after deployment. The BE Myval THV was positioned in the native annulus Figure 3.

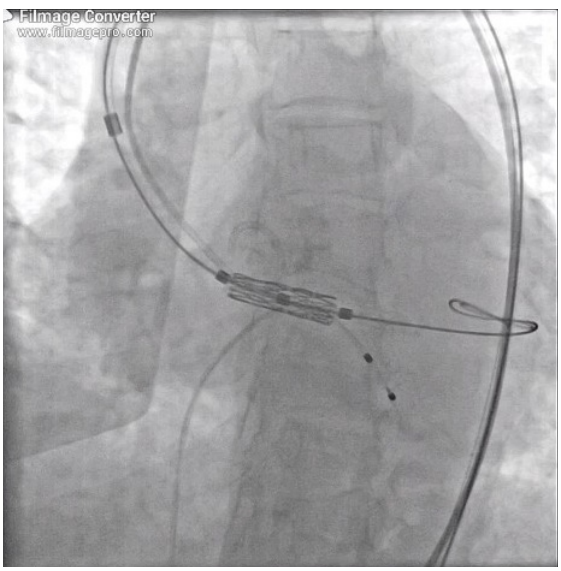

Figure 3: Positioning the Myval THV inside the aortic annulus

The root aortogram confirmed the optimal positioning of the TAVI valve, Figure 4.

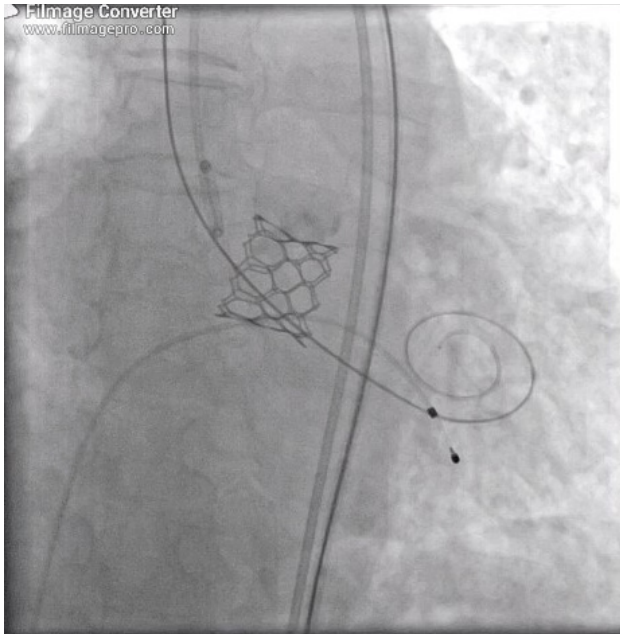

Figure 4: Fluoroscopic image shows the optimal deployment of the Myval THV

Immediate post-deployment Transthoracic echocardiography (TTE) demonstrated a well-placed valve with normal leaflet motion, no paravalvular leakage, and a mean pressure gradient of 7 mmHg. After successful valve implantation, the introducer sheath, catheter, and guidewires were removed, and the patient was transferred to the cardiac intensive care unit. The postoperative recovery period was uneventful, and echocardiography performed before discharge showed a normally functioning prosthetic valve with a mean aortic gradient of 7 mmHg and no paravalvular leak, as shown in Fig. 5. The patient was discharged on the third day in a hemodynamically stable condition.

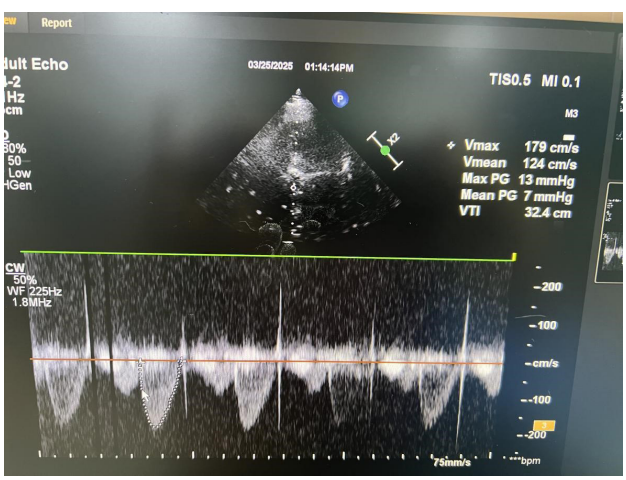

Figure 5: Aortic Valve gradient after TAVI.

Discussion

Though the latest guidelines did not fully support the use of TAVI in BAV patients,[21,22,18,14] it has not been fully explored in patients with BAV. In Meta-Analysis and Trial Sequential Analysis by Jeffrey Shi Kai Chan et. al[18] evaluated the evidence for the safety of TAVI in BAV severe aortic stenosis compared to that in TAV. A total of eight articles with 917 s BAV severe AS and 3079 TAV severe Aortic stenosis patients were meta-analyzed. There was no significant difference in primary and secondary outcomes, including AKI, PPM requirement, stroke, and 30-day mortality. It also confirmed that 30-day and 1-year mortality, as well as PPM requirements, were not significantly different between the two groups. However, BAV was associated with a considerably higher rate of significant aortic regurgitation postoperatively (P=0.002).[18]

Gentle predilatation with a semi-compliant balloon sized according to the smaller annular diameter is highly recommended in BAV patients, especially when using SEV or in patients with heavy valve calcifications. It can facilitate THV crossing through the stenotic native valve, offer useful information for valve sizing and positioning and allow the risk of coronary occlusion to be anticipated. In "gray zone" cases, valvuloplasty with simultaneous aortography could be used to confirm the most appropriate valve size and identify the best implantation depth according to the sealing point (waist of the balloon). Oversizing must be avoided, especially in severe calcified anatomies, to prevent intraprocedural complications.[6]

In the tricuspid anatomy, the aortic annulus is the narrowest part of the aortic the prosthetic valve will be sealed; hence, it is recommended that the THV should be sized based on annular dimensions, the so-called virtual basal ring (VBR). On the contrary, in a non-negligible proportion of BAV patients (approximately 13.8%), the tightest part of the aortic root may be situated above the annulus.[23] Tchétché et al. first developed the concept of the BAV landing zone. According to the BAVARD Registry,[23] the evaluation of the inter commissural distance (ICD) at 4 mm above the annulus, as compared to annular dimensions, enables identifying three different landing zone configurations (flared, tubular and tapered). TAVI sizing is done based on the different landing zone configurations. The Level of Implantation at the Raphe (LIRA) method,[24] CASPER (Calcium Algorithm Sizing for bicusPid Evaluation with Raphe)[25] and the circle method[26] are other TAVI sizing methods to avoid complications in BAV TAVI. The circle method is specific to BAV patients treated with BEV.[26]

THV Positioning is also an important procedural aspect in BAV TAVI. In BAV patients, a higher implantation depth should be attempted, while balancing the risk of THV malposition and embolization.[27] It is suggested that SEV should be placed at 0-2mm below the annulus and BEV at 90:10 (Stent frame height 90% aortic and 10% ventricular). This strategy reduces the risk of THV migration toward the left ventricle and conduction disturbances. To achieve this position with BEV, the center marker (the 3 mm radio-opaque marker placed in the middle of the deployment balloon) should be placed slightly above the line between the two or three bases of the cusps.[6]

In our case, we predilate the valve to help the crossing of the delivery system and ensure the appropriate expansion of the THV, as well as to assess the response of the supra-annular anatomy. The balloon valvuloplasty balloon was sized according to the minor axis of the AV complex.

THV under expansion with a residual high gradient and significant PVL can be quite a common complication in BAV patients, especially when using SEV.[28,29] This problem can usually be addressed with cautious balloon post-dilatation, while aggressive post-dilatation should be avoided as it exposes the patient to a higher risk of annular rupture, coronary obstruction and damage to the THV leaflets.[6] Post-dilation should also be more frequently considered in BAV than

in TAV because raphe and calcifications burden may lead to the valve under expansion and insufficient sealing in the pericommissural zones, particularly with SE valves that have a lower radial force.[14]

Balloon valvuloplasty should be sized according to the minor axis of the AV complex, and the appearance of a waist may indicate a calcified or resistant raphe and a risk of THV under expansion or annulus injury, thus influencing the THV type (SE valve preferred) and size selection. In cases of BE valve implantation, a low degree of oversizing is recommended (<10%) for the risk of annulus rupture.[14]

Conclusion

The case report demonstrates the successful completion of a TAVI procedure with balloon-expandable Myval THV in a patient with type 0 bicuspid aortic valve severe aortic stenosis in our setting.

References

1. Leon MB, Smith CR, Mack M, Miller DC, Moses JW, Svensson LG, et al. Transcatheter aortic-valve implantation for aortic stenosis in patients who cannot undergo surgery. N Engl J Med. 2010;363(17):1597-607. doi:10.1056/NEJMoa1008232.

2. Smith CR, Leon MB, Mack MJ, Miller DC, Moses JW, Svensson LG, et al. Transcatheter versus surgical aortic-valve replacement in high-risk patients. N Engl J Med. 2011;364(23):2187-98. doi:10.1056/ NEJMoa1103510.

3. Michelena HI, Desjardins VA, Avierinos JF, Russo A, Nkomo VT, Sundt TM, et al. Natural history of asymptomatic patients with normally functioning or minimally dysfunctional bicuspid aortic valve in the community. Circulation. 2008;117(21):2776-84. doi:10.1161/ CIRCULATIONAHA.107.740878.

4. Sievers, H.-H, Schmidtke, C. A classification system for the bicuspid aortic valve from 304 surgical specimens. J. Thorac. Cardiovasc. Surg. 2007, 133, 1226–1233. 10.1016/j.jtcvs.2007.01.039

5. Mikołaj Kosek a, Jan Jastrzębski, Krzysztof Kuśmierski, Maciej Dąbrowski, Piotr Szymański, Ilona Michałowska, Tomasz Hryniewiecki, Marcin Demkow, Janina Stępińska, Piotr Michałek, Zbigniew Chmielak, Adam Witkowski. Special considerations on TAVI implanted in bicuspid aortic valves Experience of Institute of Cardiology in Warsaw, Poland. coretvasa59(2017)e29–e34. http://dx.doi.org/10.1016/j.crvasa.2017.01.022

6. Gutierrez, L.,Boiago, M.,De Biase, C., Oliva, O., Laforgia, P.,Feliachi, S.,Beneduce, A., Dumonteil, N.,Tchetche, D. Transcatheter Aortic Valve Implantation for Bicuspid Aortic Valve Disease: Procedural Planning and Clinical Outcomes. J. Clin. Med. 2023, 12, 7074. https://doi.org/10.3390/jcm12227074

7. Giulia Costa,Marco Angelillis,Anna Sonia Petronio.Bicuspid Valve Sizing for Transcatheter Aortic Valve Implantation: The Missing Link. Front. Cardiovasc. Med. 2022; 27:8:770924. 10.3389/fcvm.2021.770924

8. Dayawansa, N.H., Noaman, S., Teng, L.E., Htun, N.M. Transcatheter Aortic Valve Therapy for Bicuspid Aortic Valve Stenosis. J. Cardiovasc. Dev. Dis. 2023, 10, 421. https://doi.org/10.3390/jcdd10100421 .

9. Femi Philip MD, Nadine Nadar Faza MD, Paul Schoenhagen MD, Milind Y. Desai MD, E. Murat Tuzcu MD, Lars G. Svensson MD, Samir R. Kapadia .Aortic annulus and root characteristics in severe aortic stenosis due to bicuspid aortic valve and tricuspid aortic valves: Implications for transcatheter aortic valve therapies Catheter Cardiovasc Interv. 2015;86:E88-98. doi: 10.1002/ccd.25948.

10. Marta Bargagna, Guido Ascione, Edoardo Zancanaro, Francesco Fioravanti, Alessandra Sala, Cinzia Trumello, Guohao Chang, Alessandro Verzini, Alessandro Castiglioni, Francesco Maisano.Rev. Cardiovasc. Med. 2024; 25(8): 310. 10.31083/j.rcm2508310

11. Zegdi R, Lecuyer L, Achouh P, Didier B, Lafont A, Latrémouille C, et al. Increased radial force improves stent deployment in tricuspid but not in bicuspid stenotic native aortic valves. Ann Thorac Surg. 2010;89(3):768- 72. doi:10.1016/j.athoracsur.2009.12.022.

12. Zhao ZG, Jilaihawi H, Feng Y, Chen M. Transcatheter aortic valve implantation in bicuspid anatomy. Nat Rev Cardiol. 2015;12(2):123-8. doi:10.1038/nrcardio.2014.161.

13. Himbert D, Pontnau F, Messika-Zeitoun D, Descoutures F, Détaint D, Cueff C, et al. Feasibility and outcomes of transcatheter aortic valve implantation in high-risk patients with stenotic bicuspid aortic valves. Am J Cardiol. 2012;110(6):877-83. doi:10.1016/j.amjcard.2012.04.064.

14. Flavien Vincent ,Julien Ternacle, Tom Denimal, Mylène Shen,Bjorn Redfors,Cédric Delhaye,Matheus Simonato,Nicolas Debry, Basile Verdier, Bahira Shahim, Thibault Pamart, Hugues Spillemaeker, Guillaume Schurtz, François Pontana , Vinod H. Thourani, Philippe Pibarot ,Eric Van Belle.Circulation. 2021;143:1043–1061. DOI: 10.1161/CIRCULATIONAHA.120.048048

15. Yoon SH, Lefèvre T, Ahn JM, Perlman GY, Dvir D, Latib A, et al. Transcatheter aortic valve replacement with early- and new-generation devices in bicuspid aortic valve stenosis. J Am Coll Cardiol. 2016;68(11):1195-205. doi:10.1016/j.jacc.2016.06.041.

16. Husser O, Pellegrini C, Kessler T, Burgdorf C, Thaller H, Mayr NP, et al. Outcomes after transcatheter aortic valve replacement using a novel balloon-expandable transcatheter heart valve: a single-center experience. JACC Cardiovasc Interv. 2015;8(14):1809-16. doi:10.1016/j. jcin.2015.08.014.

17. Yoon SH, Sharma R, Chakravarty T, Kawamori H, Maeno Y, Miyasaka M, et al. Clinical outcomes and prognostic factors of transcatheter aortic valve implantation in bicuspid aortic valve patients. Ann Cardiothorac Surg. 2017;6(5):463-72. doi:10.21037/acs.2017.09.03.

18. Jeffrey Shi Kai Chan, Sukhdeep Singh, Peter Eriksen, Lok Him Tsui, Amer Harky, for the Cardiovascular Evidence Review Collaboration (CvERC). Transcatheter Aortic Valve Implantation in Bicuspid Aortic Valve with Aortic Stenosis: a Meta-Analysis and Trial Sequential Analysis.Braz J Cardiovasc Surg 2022;37(1)88-98. DOI: 10.21470/1678-9741-2020-0146

19. Renker M, Kim WK. Choice of transcatheter heart valve: should we select the device according to each patient's characteristics or should it be "one valve fits all"?. Ann Transl Med.2020;8(15): 961.10.21037/atm.2020.04.13

20. Antonio Mangieri, Didier Tchetchè, Won Keun Kim,Matteo Pagnesi, Jean-Malte Sinning, Uri Landes, Ran Kornowski,Ole DeBacker, Georg Nickenig, Alfonso Ielasi, Chiara

DeBiase,Lars Søndergaard, Federico DeMarco, Matteo Montorfano,Mauro Chiarito,DamianoRegazzoli,Giulio Stefanini,-Patrizia Presbitero,Stefan Toggweiler,Corrado Tamburino, Sebastiano Immè, Giuseppe Tarantini, Horst Sievert, Ulrich Schäfer,Jörg Kempfert, Jochen Wöehrle, Francesco Gallo, Alessandra Laricchia, Azeem Latib,Francesco Giannini, AntonioColombo. Balloon Versus Self-Expandable Valve for the Treatment of Bicuspid Aortic Valve Stenosis: Insights From the BEAT International Collaborative Registrys. Circ Cardiovasc Interv. 2020 Jul;13(7):e008714. doi: 10.1161/CIRCINTERVENTIONS.119.008714.

21. A.Baumgartner H, Falk V, Bax JJ, De Bonis M, Hamm C, Holm PJ, et al. 2017 ESC/EACTS guidelines for the management of valvular heart disease. Eur Heart J. 2017;38(36):2739-91. doi:10.1093/eurheartj/ehx391.

22. B. Nishimura RA, Otto CM, Bonow RO, Carabello BA, Erwin JP 3rd, Fleisher LA, et al. 2017 AHA/ACC focused update of the 2014 AHA/ACC guideline for the management of patients with valvular heart disease: a report of the American college of cardiology/American heart association task force on clinical practice guidelines. Circulation. 2017;135(25):e1159-95. doi:10.1161/CIR.0000000000000503.

23. Tchetche, D, de Biase, C, van Gils, L,Parma, R, Ochala, A, Lefevre, T,Hovasse, T,De Backer, O. Sondergaard, L,Bleiziffer S, et al. Bicuspid Aortic Valve Anatomy and Relationship with Devices: The BAVARD Multicenter Registry. Circ. Cardiovasc. Interv. 2019;12: e007107.
doi: 10.1161/CIRCINTERVENTIONS.118.007107.

24. Iannopollo, G.; Romano, V.; Buzzatti, N.; Ancona, M.; Ferri, L.; Russo, F.; Bellini, B.; Granada, J.F.; Chieffo, A.; Montorfano, M. Supra-annular sizing of transcatheter aortic valve prostheses in raphe-type bicuspid aortic valve disease: The LIRA method. Int.J. Cardiol. 2020, 15: 317:144–151. doi: 10.1016/j.ijcard.2020.05.076

25. Petronio, A.S.; Angelillis, M.; De Backer, O.; Giannini, C.; Costa, G.; Fiorina, C.; Castriota, F.; Bedogni, F.; Laborde, J.C.; Søndergaard, L. Bicuspid aortic valve sizing for transcatheter aortic valve implantation: Development and validation of an algorithm based on multi-slice computed tomography. J. Cardiovasc. Comput. Tomogr. 2020, 14, 452-461. 10.1016/j.jcct.2020.01.007.

26. Blackman, D.; Gabbieri, D.; Del Blanco, B.G.; Kempfert, J.; Laine, M.; Mascherbauer, J.; Parma, R.; Tchétché, D. Expert Consensus on Sizing and Positioning of SAPIEN 3/Ultra in Bicuspid Aortic Valves. Cardiol. Ther. 2021, 10, 277–288. 10.1007/s40119-021-00223-9.

27. Xiong, T.-Y.; Ben Ali, W.; Feng, Y.; Hayashida, K.; Jilaihawi, H.; Latib, A.; Lee, M.K.-Y.; Leon, M.B.; Makkar, R.R.; Modine, T.; et al. Transcatheter aortic valve implantation in patients with bicuspid valve morphology: A roadmap towards standardization. Nat. Rev. Cardiol. 2022, 20, 52–67. DOI: 10.1038/s41569-022-00734-5

28. Chen, M.; Michel, J.; Kasel, A.M. Application of Balloon-Expandable Transcatheter Heart Valve in Bicuspid Aortic Valve. JACC Asia 2021, 1, 147–161. https://doi.org/10.1016/j.jacasi.2021.07.012

29. Yoon, S.-H.; Kim,W.-K.; Dhoble, A,Pio, S.M.; Babaliaros, V.; Jilaihawi, H.; Pilgrim, T.; De Backer, O.; Bleiziffer, S.; Vincent, F.; et al. Bicuspid Aortic Valve Morphology and Outcomes After Transcatheter Aortic Valve Replacement. J. Am. Coll. Cardiol. 2020, 76,1018–1030. doi: 10.1016/j.jacc.2020.07.005.