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CONSENSUS STATEMENT

Summary: International Consensus Statement on Nomenclature and Classification of the Congenital Bicuspid Aortic Valve and Its Aortopathy, for Clinical, Surgical, Interventional and Research Purposes

Hector I. Michelena, MD, Alessandro Della Corte, MD, PhD, Arturo Evangelista, MD, Joseph J. Maleszewski, MD, William D. Edwards, MD, Mary J. Roman, MD, Richard B. Devereux, MD, Borja Fernández, PhD, Federico M. Asch, MD, Alex J. Barker, PhD, Lilia M. Sierra-Galan, MD, Laurent De Kerchove, MD, Susan M. Fernandes, LPD, PA-C, Paul W. M. Fedak, MD, PhD, Evaldas Girdauskas, MD, Victoria Delgado, MD, Suhny Abbara, MD, Emmanuel Lansac, MD, Siddharth K. Prakash, MD, PhD, Malenka M. Bissell, MD, Bogdan A. Popescu, MD, PhD, Michael D. Hope, MD, Marta Sitges, MD, PhD, Vinod H. Thourani, MD, Phillippe Pibarot, DVM, PhD, Krishnaswamy Chandrasekaran, MD, Patrizio Lancellotti, MD, Michael A. Borger, MD, John K. Forrest, MD, John Webb, MD, Dianna M. Milewicz, MD, PhD, Raj Makkar, MD, Martin B. Leon, MD, Stephen P. Sanders, MD, Michael Markl, PhD, Victor A. Ferrari, MD, William C. Roberts, MD, Jae-Kwan Song, MD, PhD, Philipp Blanke, MD, Charles S. White, MD, Samuel Siu, MD, SM, Lars G. Svensson, MD, PhD, Alan C. Braverman, MD, Joseph Bavaria, MD, Thoralf M. Sundt, MD, Gebrine El Khoury, MD, Ruggero De Paulis, MD, Maurice Enriquez-Sarano, MD, Jeroen J. Bax, MD, Catherine M. Otto, MD, and Hans-Joachim Schäfers, MD

Department of Cardiovascular Medicine, Mayo Clinic, Rochester, Minnesota; Department of Translational Medical Sciences, University of Campania "L. Vanvitelli", Naples, Italy; Department of Cardiology, Hospital Vall d'Hebron, Vall d'Hebron Research Institute (VHIR) Ciber-CV, Barcelona, Spain; Department of Laboratory Medicine and Pathology, Mayo Clinic, Rochester, Minnesota; Division of Cardiology, Weill Cornell Medicine, New York, New York; Departamento de Biología Animal, Facultad de Ciencias, Instituto de Investigación Biomédica de Málaga, Universidad de Málaga, Ciber-CV, Málaga, Spain; MedStar Health Research Institute, Washington, DC; Department of Radiology, Children's Hospital Colorado, University of Colorado, Anschutz Medical Campus, Aurora, Colorado; Cardiovascular Division, American British Cowdray Medical Center, Mexico City, Mexico; Division of Cardiothoracic and Vascular Surgery, Cliniques Universitaires Saint-Luc, Université Catholique de Louvain, Brussels, Belgium; Division of Pediatric Cardiology, Department of Pediatrics, Stanford University, Palo Alto, California; Division of Cardiovascular Medicine, Department of Medicine, Stanford University, Palo Alto, California; Department of Cardiac Sciences, Libin Cardiovascular Institute, Cumming School of Medicine, University of Calgary, Calgary, Alberta, Canada; Department of Cardiovascular Surgery, University Heart and Vascular Center Hamburg, Hamburg, Germany; Department of Cardiology, Leiden University Medical Center, Leiden, Netherlands; Cardiothoracic Imaging Division, Department of Radiology, UT Southwestern Medical Center, Dallas, Texas; Department of Cardiac Surgery, Institute Mutualiste Montsouris, Paris, France; Department of Internal Medicine, McGovern Medical School, The University of Texas Health Science Center at Houston, Houston, Texas; Department of Biomedical Imaging Science, Leeds Institute to Cardiovascular and Metabolic Medicine, University of Leeds, Leeds, United Kingdom; Department of Cardiology, University of Medicine and Pharmacy "Carol Davila"—Euroecolab, Emergency Institute for Cardiovascular Diseases "Prof. Dr. C. C. Iliescu", Bucharest, Romania; Department of Radiology and Biomedical Imaging, University of California San Francisco, San Francisco, California; Cardiovascular Institute, Hospital Clínic, Universitat de Barcelona, IDIBAPS, CIBERCV, ISCIII (CB16/11/00354), CERCA Programme, Barcelona, Spain; Department of Cardiovascular

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Address correspondence to Dr Michelena, Department of Cardiovascular Medicine, Mayo Clinic, 200 First St SW, Rochester, MN 55905; e-mail: michelena.hector@mayo.edu.

Surgery, Marcus Valve Center, Piedmont Heart Institute, Atlanta, Georgia; Department of Cardiology, Québec Heart & Lung Institute, Laval University Québec, Québec, Canada; Department of Cardiology, University of Liège Hospital, GIGA Cardiovascular Sciences, CHU Sart Tilman, Liège, Belgium; Gruppo Villa Maria Care and Research, Maria Cecilia Hospital, Cotignola, and Anthea Hospital, Bari, Italy; University Clinic of Cardiac Surgery, Leipzig Heart Center, Leipzig, Germany; Yale University School of Medicine & Yale New Haven Hospital, New Haven, Connecticut; St Paul's Hospital, University of British Columbia, Vancouver, British Columbia, Canada; Cedars Sinai Heart Institute, Los Angeles, California; Division of Cardiology, Columbia University Irving Medical Center/NY Presbyterian Hospital, New York, New York; Cardiac Registry, Departments of Cardiology, Pathology and Cardiac Surgery, Boston Children's Hospital, Boston, Massachusetts; Department of Pediatrics, Harvard Medical School, Boston, Massachusetts; Department of Radiology, Feinberg School of Medicine, Northwestern University, Chicago, Illinois; Cardiovascular Medicine Division, University of Pennsylvania Medical Center and Penn Cardiovascular Institute, Philadelphia, Pennsylvania; Baylor Heart and Vascular Institute, Baylor University Medical Center, Texas A & M School of Medicine, Dallas Campus, Dallas, Texas; University of Ulsan College of Medicine, Asan Medical Center, Seoul, South Korea; Department of Radiology, St. Paul's Hospital, Vancouver, British Columbia, Canada; Department of Radiology, University of Maryland School of Medicine, Baltimore, Maryland; Schulich School of Medicine and Dentistry, London, Ontario, Canada; Heart, Vascular and Thoracic Institute, Cleveland Clinic, Cleveland, Ohio; Cardiovascular Division, Department of Medicine, Washington University School of Medicine, St. Louis, Missouri; Division of Cardiac Surgery, University of Pennsylvania, Philadelphia, Pennsylvania; Division of Cardiac Surgery, Massachusetts General Hospital, Boston, Massachusetts; Department of Cardiac Surgery, European Hospital and Unicamillus University Rome, Rome, Italy; Division of Cardiology, University of Washington, Seattle, Washington; and Department of Thoracic and Cardiovascular Surgery, Saarland University Medical Center, Homburg/Saar, Germany

This International evidence-based nomenclature and classification consensus on the congenital bicuspid aortic valve and its aortopathy recognizes 3 types of bicuspid aortic valve: 1. Fused type, with 3 phenotypes: right-left cusp fusion, right-non cusp fusion and left-non cusp fusion; 2. 2-sinus type with 2 phenotypes: Latero-lateral and antero-posterior; and 3. Partial-fusion or forme fruste. This consensus recognizes 3 bicuspid-aortopathy types: 1. Ascending phenotype; root phenotype; and 3. extended phenotypes.

(Ann Thorac Surg 2021;112:1005-22)

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INTENDED AUDIENCE AND PURPOSE

This international evidence-based nomenclature and classification consensus on the congenital bicuspid aortic valve (BAV) and its aortopathy is intended to be universally used by clinicians (both pediatric and adult), echocardiography sonographers and physicians, cardiovascular advanced-imaging specialists, interventional cardiologists, cardiovascular surgeons, pathologists, geneticists and researchers encompassing these clinical and basic research areas. In addition, if and when new landmark research is available, this international consensus may be subject to change in accordance with evidence-based data.[1]

GENERAL NOSOLOGY OF THE CONGENITAL BAV CONDITION

The congenital BAV condition is a valvulo-aortopathy characterized by significant heterogeneity of its valvular and aortic phenotypic expressions, of its associated disorders, of its complications and of its prognosis.[2-6] To reconcile this clinical and prognostic heterogeneity, the BAV condition is broadly categorized into 3 clinical-prognostic subgroups (Figure 1): (i) Complex valvulo-aortopathy[1,5,6] is characterized by concomitant or associated disorders that may be clinically and prognostically worse than the BAV condition *per se* (ie, Turner syndrome, Loeys-Dietz syndrome, Shone complex and severe aortic coarctation) and/or by early/accelerated valve dysfunction and/or aortopathy, more commonly diagnosed earlier in the pediatric, adolescent and young adult population.[7,8] This presentation frequently requires early surgical/invasive treatment and close surveillance. (ii) Typical valvulo-aortopathy[1,2,6] is the most common type, with progressive BAV dysfunction and/or aortic dilatation without other major associated disorders, is more commonly diagnosed in the young adult and adult, requires long-term surveillance and commonly requires subsequent surgical/invasive treatment. Patients with complex-presentation and typical-presentation valvulo-aortopathies are at risk of developing infective endocarditis and aortic dissection (Figure 1), although aortic dissection is extremely rare in young children with BAV and rare in adults without aortic dilatation.[2,9] (iii) The undiagnosed or uncomplicated BAV subgroup[2] exhibits a lifelong silent condition with mild or non-progressing valvulo-aortopathy

Abbreviations and Acronyms

BAV = bicuspid aortic valve
CT = computed tomography
ECG = electrocardiographic
MR = magnetic resonance
STJ = sinotubular junction
TTE = transthoracic echocardiography

that does not manifest clinically but may come to light at autopsy or incidentally by imaging (Figure 1); therefore, it represents a retrospective definition. A critical difference between the typical and complex valvulo-aortopathies is the preserved long-term overall life expectancy that is similar to that of the age- and sex-matched general population in patients with the typical valvulo-aortopathy [10], whereas the life expectancy of the patient with complex valvulo-aortopathy may be reduced.[1]

COMMON COMPLICATIONS AND FUNDAMENTALS OF IMAGING ASSESSMENT OF THE CONGENITAL BAV CONDITION

In order of frequency, the most common major complications of the typical valvulo-aortopathy are (i) the need for aortic valve surgery due to aortic stenosis; (ii) ascending thoracic aortic dilatation that may require surgical repair; (iii) the need for aortic valve surgery due to aortic regurgitation; (iv) mitral valve prolapse including the anterior leaflet that may require surgery[1]; (v) infective endocarditis; and (vi) aortic dissection.[1,11]

At the center of the BAV condition is echocardiography, which serves as the first-line imaging modality in 6 major capacities[11]: (i) BAV diagnosis, (ii) valvular phenotyping, (iii) assessment of valvular function[11], (iv) measurement of the thoracic aorta (the expression of

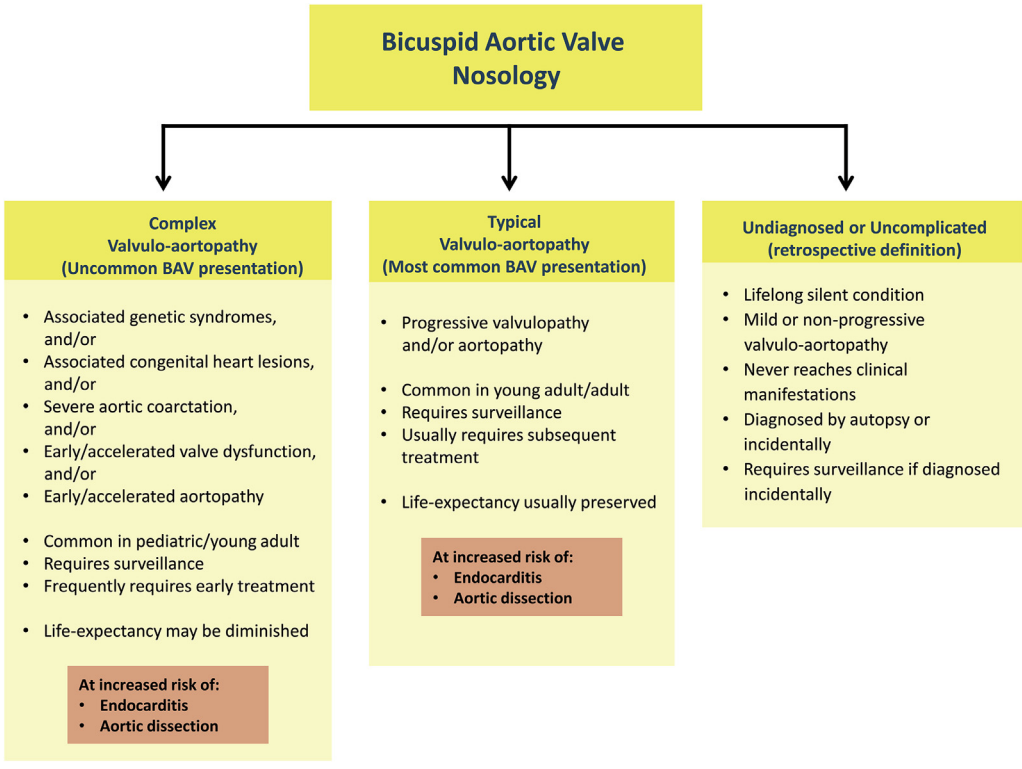

FIGURE 1 Nosology of the congenital bicuspid aortic valve condition. (Left) Anatomically and prognostically complex presentations of the bicuspid aortic valve valvulo-aortopathy are those associated with syndromes, left-sided obstructions, significant aortic coarctation, early/accelerated valve dysfunction (stenosis or regurgitation) and/or early aortopathy, manifested as thoracic aorta dilatation. These conditions are more commonly diagnosed in childhood, adolescence and young adulthood. (Middle) The anatomically and prognostically typical valvulo-aortopathy is usually diagnosed in young and middle-aged adults, although it may be diagnosed in children as well and comprises various degrees of progressive valvular dysfunction with a high cumulative incidence of aortopathy over the long run, manifested as thoracic aortic dilatation, without major associated conditions. Complex- and typical-presentation forms are susceptible to development of infective endocarditis and aortic dissection, although dissection is rare in the pediatric population and adults without aortic dilatation. (Right) The undiagnosed or uncomplicated form is not diagnosed in the patient's lifetime (without any bicuspid aortic valve-related complications, some are diagnosed post-mortem) or is diagnosed during the patient's lifetime but does not cause complications requiring treatment. Therefore, it is a retrospective definition. (Modified from Michelena et al[6] with permission from Elsevier.)

TABLE 1 Critical Limitations of the Sievers Classification Compared to the New International Consensus

Sievers and Schmidtke[16], Type of Limitation	Specific Sievers Limitation	International Consensus
Comprehension and retention	Not language-intuitive: types: 0, 1 and 2	Language-intuitive: types: fused, 2-sinus and partial fusion
Unable to define all BAV phenotypes	Type 0 does not differentiate between a fused BAV with no raphe and a 2-sinus BAV	Fused types may have raphe or not; 2-sinus types do not have raphe
Lack of prerepair assessment of symmetry	Nonexistent	Fused types require assessment of symmetry for surgical repair planning
Lack of recognition of BAV phenotypes	Does not recognize the partial-fusion (forme fruste); does not recognize fused BAV with no raphe	Recognizes partial-fusion (forme fruste); recognizes fused BAV with no raphe, which is different from 2-sinus BAV
Lack of recognition of aortopathy phenotypes	Nonexistent	Aorta phenotypes: root, ascending and extended
Includes a non-BAV congenital aortic valve abnormality	Type 2 is not BAV, is unicuspid aortic valve	Does not include unicuspid aortic valves
Evidence based	Anatomical pathology only	Imaging, anatomical surgical pathology, surgical-functional pathology, clinical associations

BAV, bicuspid aortic valve.

BAV aortopathy is dilatation of the thoracic aorta), (v) exclusion of aortic coarctation and other associated congenital lesions[2,7] and (vi) assessment of uncommon but serious complications such as infective endocarditis[12] and aortic dissection.[9] Transthoracic echocardiography (TTE) is the best modality for hemodynamic assessment of valvular dysfunction and the initial modality for assessment of thoracic aorta size, presence of aortic coarctation and other congenital lesions. Transesophageal echocardiography may aid in the diagnosis and phenotyping of BAV if it is not well visualized by TTE and has excellent accuracy for the diagnosis of aortic dissection and infective endocarditis.[1]

Advanced-imaging modalities are also at the center of the BAV condition: electrocardiographic (ECG)-gated cardiac computed tomography (CT) and ECG-gated cardiac magnetic resonance (MR). These imaging techniques improve the diagnostic accuracy and phenotyping of BAV[13,14] and represent the gold standard for measuring the thoracic aorta because they accurately assess aortic diameters that are truly perpendicular to the longitudinal axis of the aorta. After initial TTE imaging, if any aortic segment cannot be visualized or coarctation cannot be ruled out or any thoracic segment measures $\geq$45 mm by TTE, then ECG-gated CT angiography or MR angiography is recommended.[15]

WHY HAVE A STANDARD NOMENCLATURE AND CLASSIFICATION CONSENSUS FOR THE CONGENITAL BAV CONDITION?

Nomenclature refers to the choice of name that is given to a particular structure, abnormality or phenotype, whereas classification refers to the process of arranging or categorizing something according to shared features. The clinician evaluating the patient with BAV must be able to communicate all specific morphological, functional and prognostic aspects of the BAV condition to the patient, other clinicians, surgeons, interventionalists and researchers, in a common language.[1,6] In addition, there are multiple gaps in the knowledge and understanding of the BAV condition.[2] To advance the clinical, biological and genetic understanding of the BAV condition, a common language must be articulated among researchers in all clinical and laboratory research disciplines. Multiple nomenclatures and classifications exist for the BAV condition, and they are as heterogeneous or more than the BAV condition itself.[1,6] For example, the Sievers[16], Schaefer[17] and Kang[18] classifications use multiple letters and numbers to describe different aspects of the BAV and its aortopathy that are not intuitive and thus difficult to remember.[1,6]

The Sievers classification, based on the presence and number of raphes identified at surgery (ie, direct visualization), has several shortcomings (Table 1): (i) it is not based on imaging, which is the most common method of diagnosing, phenotyping and surveilling the BAV and its aortopathy; (ii) it is unable to define all known BAV phenotypes (ie, partial fusion/forme fruste); (iii) it lacks the recognition of aortopathy phenotypes; (iv) it lacks the assessment of BAV symmetry, which is critical for planning surgical regurgitant-BAV repair[19,20]; and (v) it is inclusive of the unicuspid aortic valve morphology as a subtype of BAV (Sievers type 2). Although the morphological spectrum of human congenital aortic valve abnormalities includes unicuspid, bicuspid and quadricuspid aortic valves, their embryological origins may not necessarily be closely linked, such that animal models of BAV have displayed all possible BAV phenotypes, quadricuspid valves and pulmonary valve abnormalities but not unicuspid anatomical forms.[21] In addition, the prevalence, age at presentation and prognosis of unicuspid and BAV are not equivalent.[22,23] Furthermore, the definition of unicuspid aortic valve (1 cusp with or

without a commissure: unicommissural or acommissural)[24,25] is very different from that of BAV (2 cusps, 2 commissures); therefore, the Sievers classification includes only 1 (unicommissural) of the 2 types of unicuspid aortic valves as if it were a subtype of BAV, which is confusing.

The use of one or another of the many classifications for research varies according to authors and institutions, and there are specific terminologies that lead to confusion such as the 'true' BAV[2]; does it mean that the others are false BAV? These numerous and heterogeneous classifications cause confusion in clinical practice; failure to identify phenotypes that may predict outcomes; the inability to analyse clinical outcome data in registries, systematic reviews and meta-analysis formats; failure to capture anatomical information critical for surgical aortic valve repair and TAVR; and hamper identification of phenotypic-genetic associations. Herein, we present an imaging-based, descriptive, simple-but-comprehensive nomenclature and classification system that is based on the English language rather than on numbers or letters and on important available anatomical, clinical, surgical and pathological scientific data.[1,6] This new nomenclature/classification system represents the combined efforts of international BAV experts including clinicians (both adult and pediatric), surgeons, interventionalists, pathologists, geneticists and imagers (echocardiography, CT and MR experts).[1]

DEFINITION OF CONGENITAL BAV AND AORTIC ROOT COMPLEX

CONGENITAL BAV. The congenital BAV is most commonly diagnosed by base-of-the-heart, short-axis aortic valve imaging with TTE and gated cardiac computed tomography or cardiac magnetic resonance, demonstrating the existence of only 2 commissures delimiting only 2 valve cusps (Figure 2).[2,26] On echocardiographic long-axis imaging, systolic doming of the conjoined cusp may be appreciated particularly

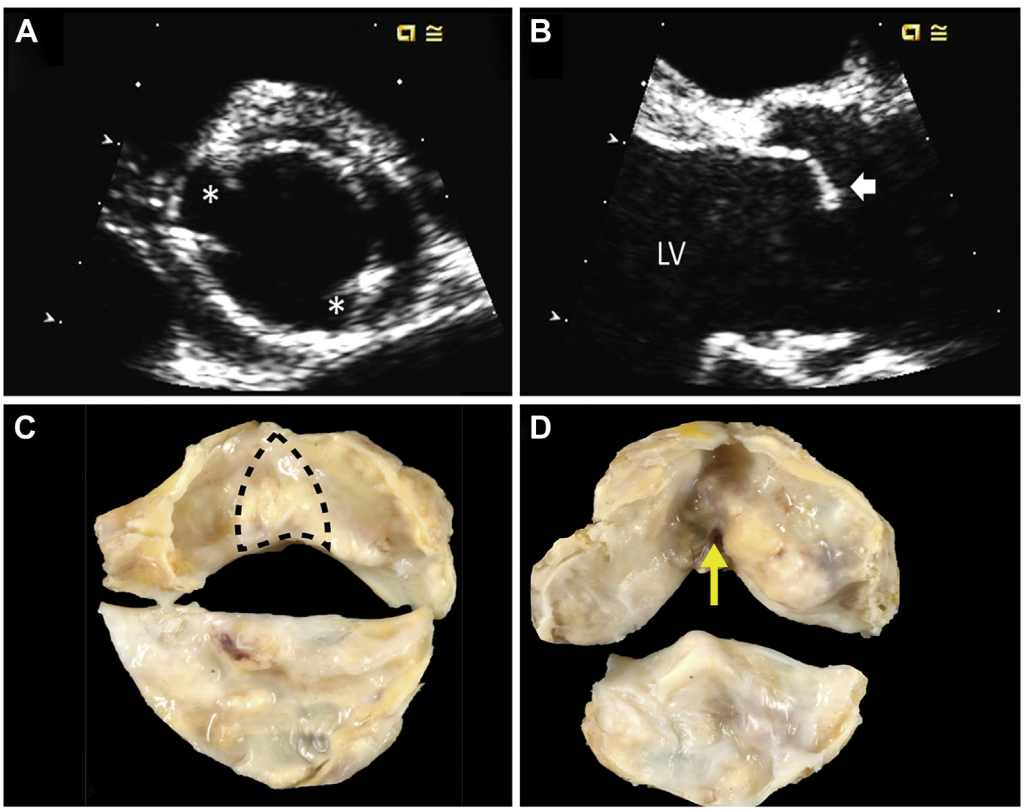

FIGURE 2 Diagnosis of congenital bicuspid aortic valve by transthoracic echocardiography and pathological manifestations. (A) Parasternal short-axis aortic valve systolic still image demonstrating the existence of only 2 commissures (asterisks) delimiting only 2 cusps. (B) Parasternal long-axis systolic still shows systolic doming of the fused (conjoined) cusp (arrow), common for right–left–coronary cusp fusion. (C) Pathological congenital bicuspid aortic valve specimen shows the area of the raphe (dashed line) from the left ventricular perspective, forming an obtuse angle between the fused cusps. (D) Ventricular side of a tricuspid aortic valve with acquired rheumatic fusion shows the cleavage plane with acute angle (yellow arrow). (LV, left ventricle.)

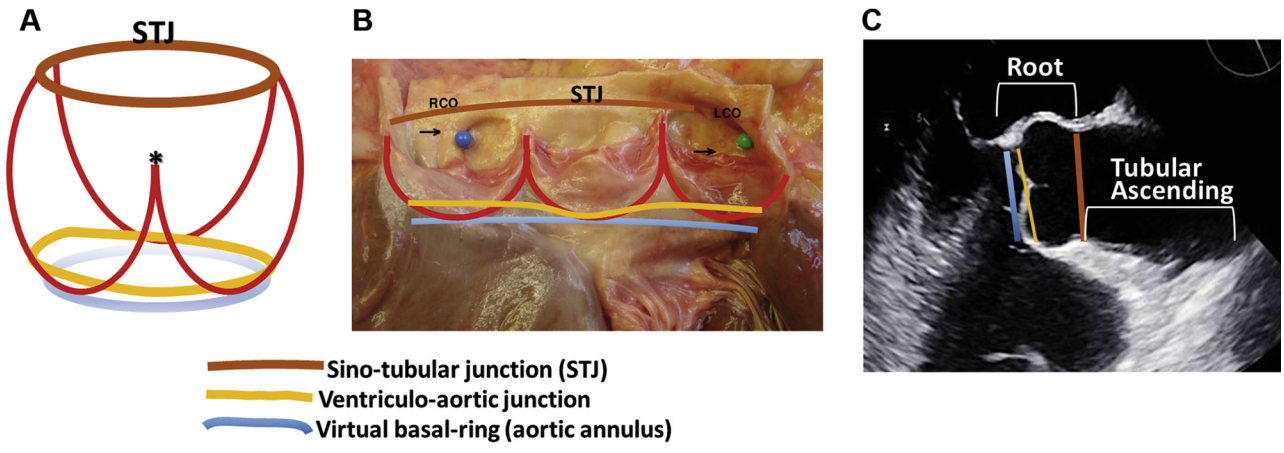

FIGURE 3 The aortic root complex. (A) Schematic drawing of the aortic root: the blue line indicates the virtual basal ring (aortic annulus); the yellow line depicts the ventriculo-aortic junction (whose non-planar nature is emphasized schematically)[30]; the red lines show the crown-shaped attachments of the cusps to the wall of the aortic sinuses (note the different height of the underdeveloped commissure [asterisk] under the raphe compared to the other 2 true commissures); and the brown line depicts the sinotubular junction. (B) All the above boundaries and structures are shown (same colors as above) in an anatomical specimen of a normal aortic root and tricuspid aortic valve. (C) Echocardiographic view of the aortic root: the levels of the aortic annulus, ventriculo-aortic junction and sinotubular junction are shown (same colors as above). It is important to recognize that it is the measurement of the virtual annulus, sinuses and sinotubular junction that have clinical and practical implications for the patient with bicuspid aortic valve. (LCO, left–coronary orifice [green pin and arrow]; RCO, right coronary orifice [blue pin and arrow]; STJ, sinotubular junction.)

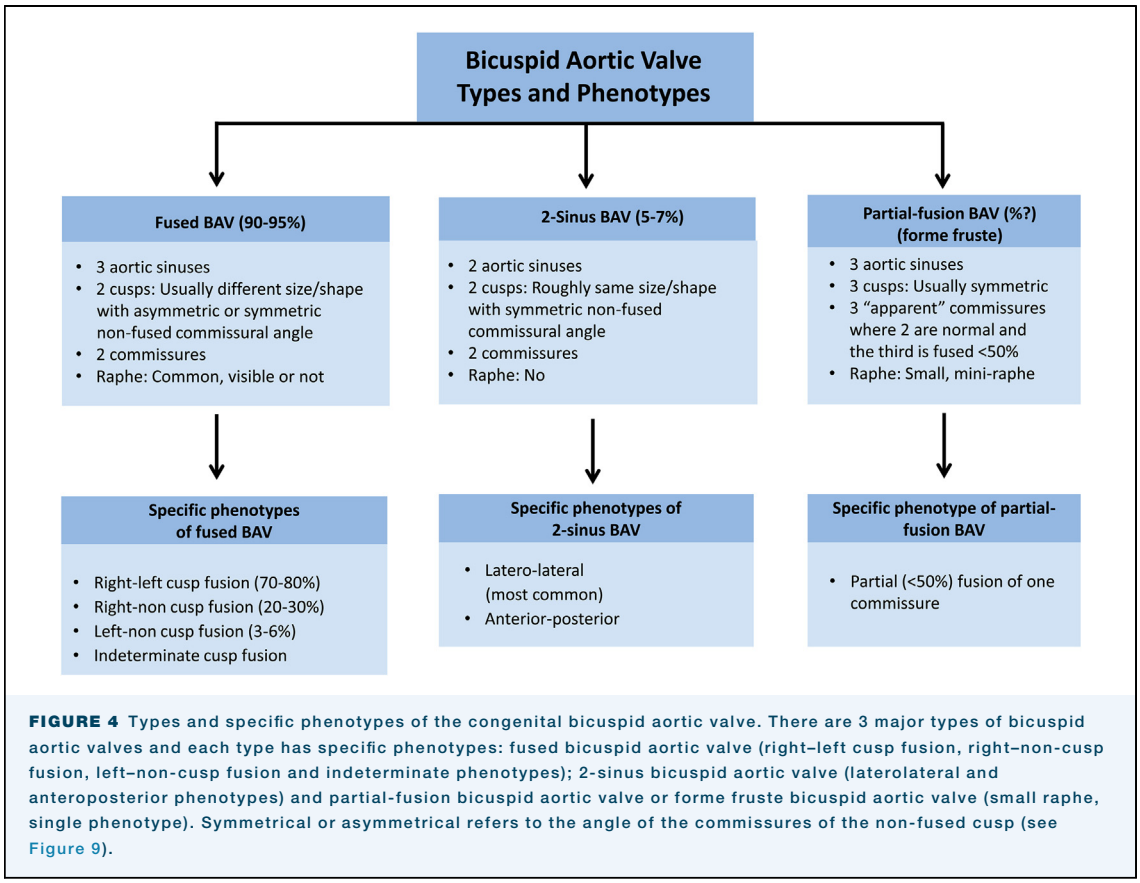

FIGURE 4 Types and specific phenotypes of the congenital bicuspid aortic valve. There are 3 major types of bicuspid aortic valves and each type has specific phenotypes: fused bicuspid aortic valve (right–left cusp fusion, right–non-cusp fusion, left–non-cusp fusion and indeterminate phenotypes); 2-sinus bicuspid aortic valve (laterolateral and anteroposterior phenotypes) and partial-fusion bicuspid aortic valve or forme fruste bicuspid aortic valve (small raphe, single phenotype). Symmetrical or asymmetrical refers to the angle of the commissures of the non-fused cusp (see Figure 9).

for right-left-coronary cusp fusion (Figure 2), but it is less reliable for identifying other BAV phenotypes. The diagnosis can also be made by direct surgical observation[20,27] and by a pathological examination.[28] It is important to recognize that a tricuspid aortic valve that is calcified or rheumatic may present a pattern of acquired (non-congenital) fusion of 2 cusps that may be difficult to differentiate from congenital BAV; in these cases, surgical inspection and/or pathological examination may identify whether the fusion is congenital or not. In the operating room, although it is not always possible, the surgeon can define the congenital bicuspid condition by comparing the height of the 'pseudocommissure' (the attachment of the raphe [pseudocommissure] at the aortic wall), which is lower within the root compared to the height of the true commissures, whose attachment is higher (Figure 3). Additional gross features can be used on surgical or pathological inspection, such as the angle formed between the fused cusps (obtuse = congenital fusion; acute = acquired fusion) and the cleavage plane on the ventricular aspect of the fused cusps (absent = congenital; present = acquired) (Figure 2).

AORTIC ROOT AND ROOT COMPLEX. Although 'ascending aorta' and 'aortic root' are sometimes used interchangeably to indicate the entire vascular segment from the aortic valve to the brachiocephalic artery take-off (beginning of the arch), the term aortic root correctly refers only to the most proximal part of the ascending thoracic aorta, from the distal end of the left ventricular outflow tract to the sinotubular junction (STJ), formed by the sinuses of Valsalva and containing the aortic valve[29] (Figure 3). The anatomy and

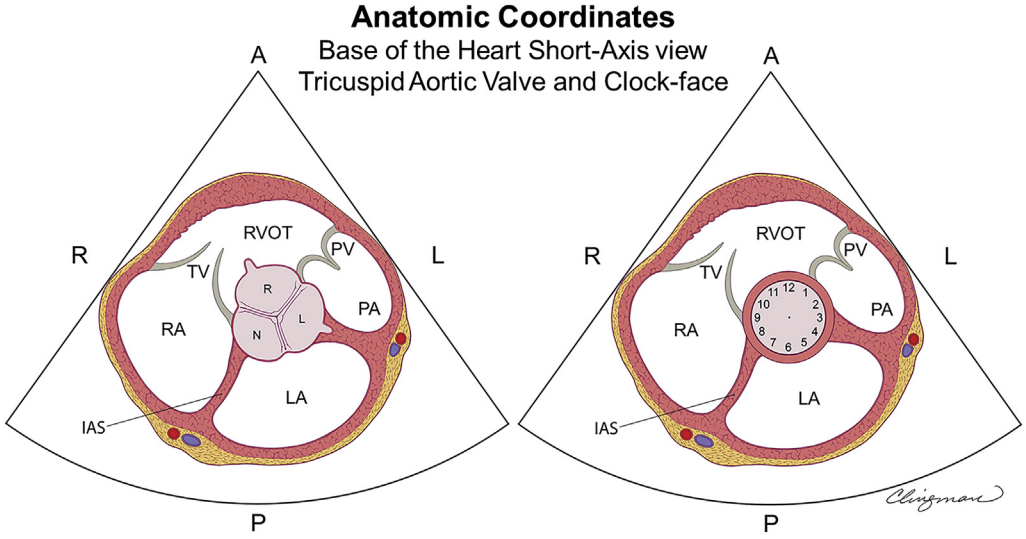

FIGURE 5 Schematic transthoracic echocardiography-based short-axis, base-of-the-heart anatomical landmarks and clock face for bicuspid aortic valve diagnosis and phenotyping. (Left panel) Schematic representation of the normal tricuspid aortic valve in the echocardiographic parasternal short-axis view, applicable to similar views obtained with cardiac computed tomography and cardiac magnetic resonance. The right coronary cusp (small R) is anterior and positioned between the tricuspid valve and pulmonary valve insertions. The left–coronary cusp (small L) is posterior-lateral and related to the left atrium, whereas the non-coronary cusp (small N) is the most posterior and related to the interatrial septum (IAS). Note the origin of the coronary arteries at the right and left cusps. These landmark anatomical relations of each cusp relative to adjacent structures are critical in determining which 2 cusps are fused. (Modified from Michelena et al[6] with permission from Elsevier.) (Right panel) The annular circumference of the aortic valve can be visualized like the face of a clock. Fused bicuspid valves with right–left cusp fusion usually have commissures at 4 and 10 or 5 and 11 o'clock (see Figures 6 and 7), and the anatomy relative to adjacent structures suggests right–left cusp fusion. In right–non-coronary cusp fusion, the commissures are usually at 1 and 7 or 12 and 6 o'clock (see Figures 6 and 7); the anatomy relative to adjacent structures suggests right–non-cusp fusion. In left– non-coronary cusp fusion, usually 2 and 8 or 9 and 3 o'clock (see Figures 6 and 7) and the anatomy relative to adjacent structures suggest left–non-fusion. It is important to note that there can be overlap between the clock positions; thus, it is critical to know the landmark anatomical relations of each cusp. Identification of the raphe can be invaluable in determining the conjoined cusp. Identification of the origin of the left and right coronary arteries (left panel) may also be invaluable. (LA, left atrium; large L, left side of the patient; large R, right side of the patient; P, posterior aspect of the heart; PA, pulmonary artery; PV, pulmonary valve; RA, right atrium; RVOT, right ventricular outflow tract; TV, tricuspid valve.) (Modified from Michelena et al.[6] with permission from Elsevier.)

physiology of the aortic root complex and its interaction with the valve have been thoroughly investigated as contemporary techniques for aortic valve repair have been introduced and more widely adopted.[30,31] Functionally, and particularly in relation to the competency of the BAV and surgical repair, 3 elements form the aortic root complex and cooperate in determining physiological valve dynamics[32]: (i) the STJ, (ii) the aortic sinuses with the crown-like attachment line of the aortic valve cusps to the aortic wall at the aortic sinuses (which, as mentioned, assumes a peculiar form in the fused BAV, with 1 of the 3 'crown tips' corresponding to the under-the-raphe pseudocommissure, reaching a lower height than the other 2) (Figure 3) and (iii) the aortic annulus, which is a virtual circular line inside the left ventricular outflow tract, running through the nadir of the aortic cusps and the bases of the respective inter-cusp triangles (Figure 3). The aortic annulus is a virtual surrogate for the ventriculo-aortic junction, which is the real boundary of the aortic root complex, identified anatomically as the transition from the ventricular muscle to the aortic media and located circumferentially slightly above the nadir of the aortic cusps, crossing the semilunar lines of each cusp's attachment (Figure 3). However, for both surgical and imaging purposes, the virtual aortic annulus is the practical and clinically used anatomical landmark that constitutes the 3rd component of the root complex, as described above. The aortic root complex, particularly

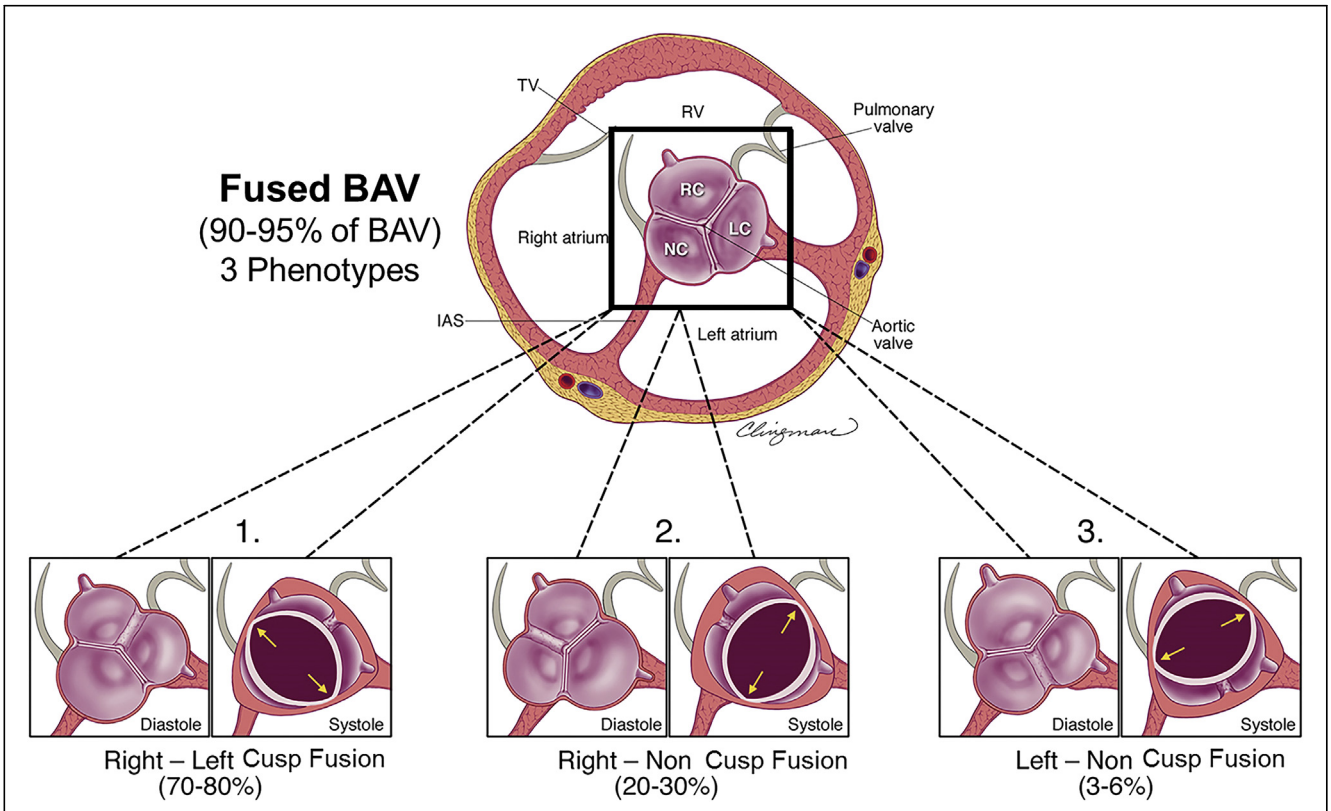

FIGURE 6 Schematic representation of fused bicuspid aortic valve phenotypes as seen by parasternal short-axis transthoracic echocardiography. Applicable to similar tomographic views by cardiac CT and cardiac MR, the figure demonstrates the 3 fused bicuspid aortic valve phenotypes as zoomed views of the base of the heart (black square) for anatomical landmark correlation. Note that all fused bicuspid aortic valves have 3 distinguishable aortic sinuses. Note the oval (American football shape) systolic opening of these 3 valves as opposed to the triangular opening of a tricuspid aortic valve. (1) Right–left cusp fusion (most common) with visible raphe, 2 different size/shape functional cusps (the non-fused cusp [non-coronary] is commonly of larger 'compensatory' size than the others). (2) Right–non-cusp fusion with visible raphe, 2 different size/shape functional cusps (the non-fused cusp [left] is larger than the others). (3) Left–non-cusp fusion with a visible raphe (least common), 2 different size/shape functional cusps (the non-fused cusp [right] is larger than the others). It is important to note that these short-axis imaging views do not correspond to the surgeon's intraoperative view. Note how, in diastole, the commissural angle of the non-fused cusp of these 3 asymmetrical bicuspid aortic valves is <170–180° (see Figure 9); in systole, the right–left commissures are at 10 and 4 o'clock (1: yellow arrows), right–non-commissures at 1 and 7 o'clock (2: yellow arrows), and left– non-commissures at 2 and 8 o'clock (3: yellow arrows) (see Figure 7). These 3 fused phenotypes may not have a visible raphe and may also have symmetrical non-fused cusp angle (see Figure 8). (IAS, interatrial septum; LC, left cusp; NC, non-coronary cusp; RC, right cusp; RV, right ventricle; TV, tricuspid valve.) (Modified from Michelena et al.[6] with permission from Elsevier.)

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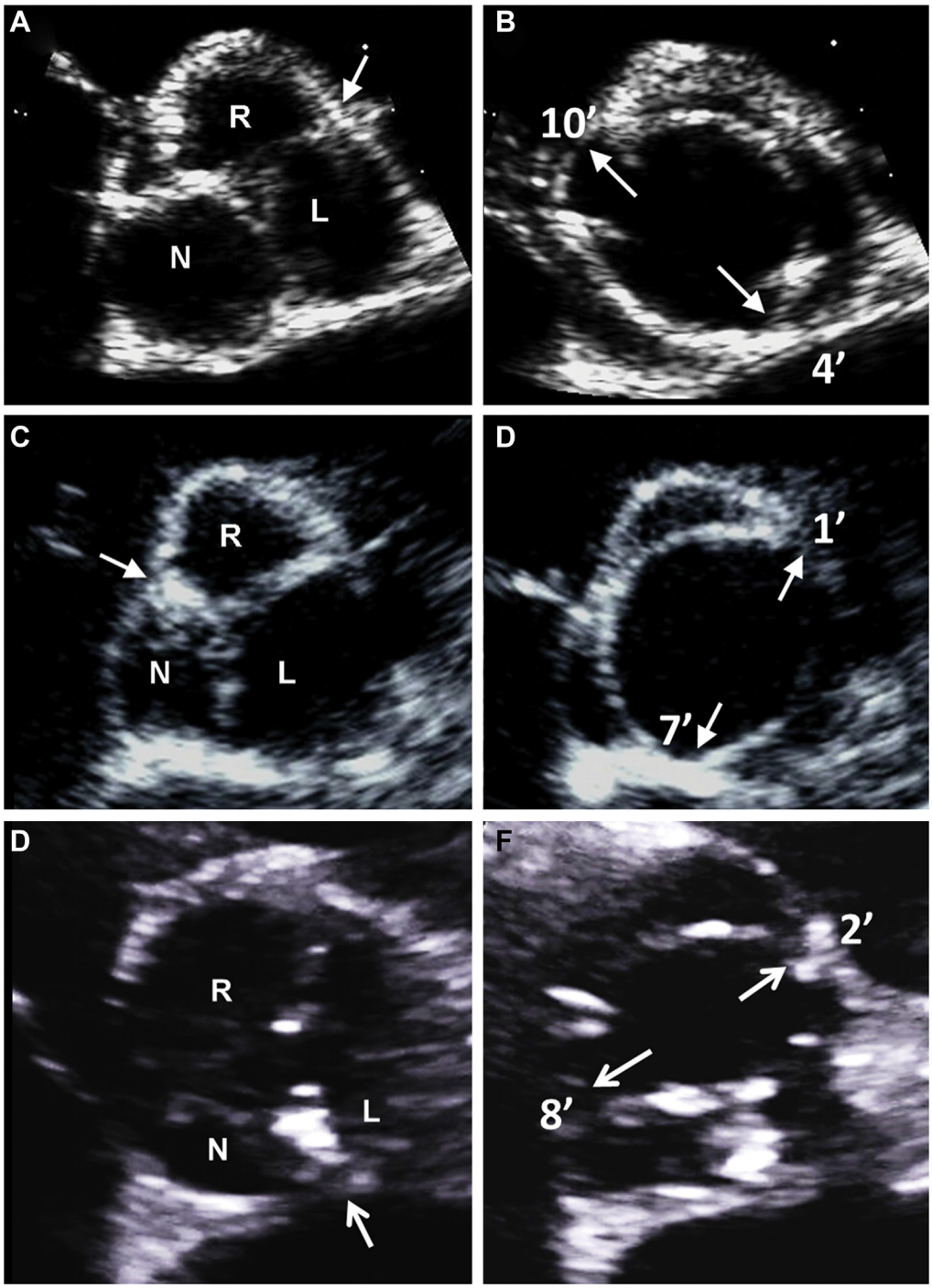

FIGURE 7 Diastolic and systolic transthoracic echocardiography parasternal short-axis still images of the 3 phenotypes of fused bicuspid aortic valve. Applicable to similar tomographic views obtained with cardiac CT and cardiac MR. (A) Right–left cusp fusion bicuspid aortic valve within 3 distinguishable aortic sinuses, with raphe (arrow) in diastole and (B) typical systolic opening with commissures marked as the clock face (arrows). (C) Right–non-cusp fusion bicuspid aortic valve within 3 distinguishable aortic sinuses, with raphe (arrow) in diastole and (D) typical systolic opening with commissures marked as the clock face (arrows). (E) Left–non-cusp fusion bicuspid aortic valve within 3 distinguishable aortic sinuses, with raphe (arrow) in diastole and (F) typical systolic opening with commissures marked as the clock face (arrows). (Modified from Michelena et al.[11] with permission from Elsevier.)

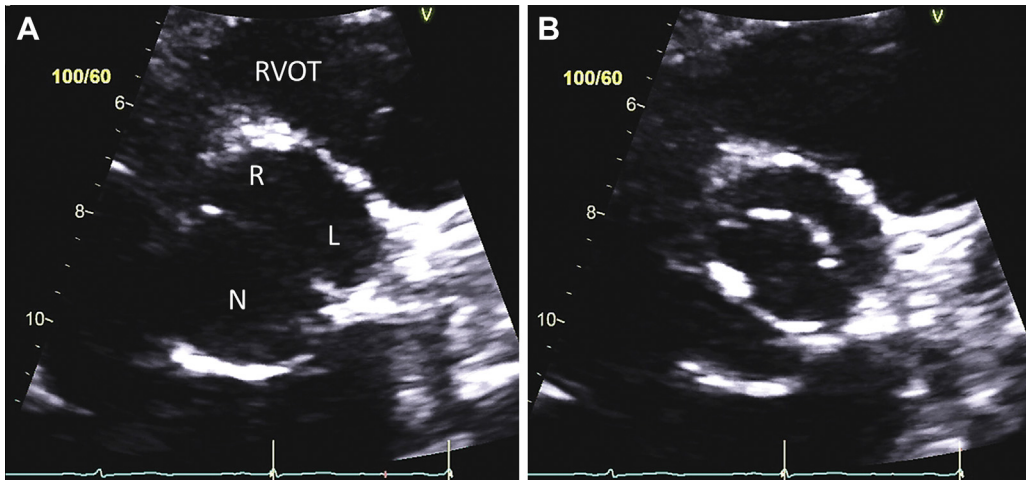

FIGURE 8 Fused-type right–left cusp fusion without visible raphe and symmetrical non-fused cusp commissural angle. (A) Diastolic transthoracic echocardiography short-axis still frame shows right–left cusp fusion without visible raphe (uncommon) and 180° angle of the non-fused cusp commissures (see Figure 9), yet the sizes and shapes of the 2 functional cusps are different, the conjoined cusp is smaller than the predominant non-fused non-coronary cusp (N) and there are 3 aortic sinuses. (B) Systolic transthoracic echocardiography short-axis still frame confirms the absence of a visible raphe and the 180° commissural angle. (L, left–coronary cusp; R, right coronary cusp; RVOT, right ventricular outflow tract.)

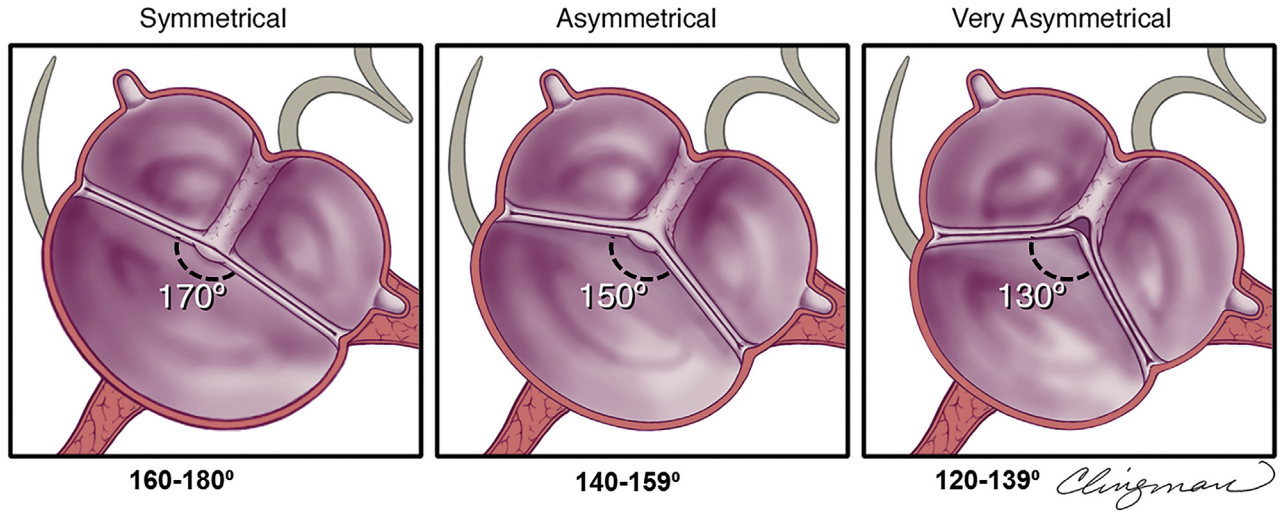

FIGURE 9 Schematic representation of the transthoracic echocardiographic evaluation of fused bicuspid aortic valve symmetry in the parasternal short axis. Applicable to similar tomographic views obtained from cardiac CT and cardiac MR, the figure demonstrates different commissural angles of the non-fused cusps (applicable to the 3 fused bicuspid aortic valve phenotypes, although only right–left cusp fusion is shown) that define symmetry. (Left panel) Symmetrical (angle 160–180°) right–left cusp fusion bicuspid aortic valve with raphe, where the 2 functional cusps are almost the same size/shape (the non-fused cusp is a little larger) and the commissural angle of the non-fused cusp is about 170°. (Middle panel) Asymmetrical (angle 140–159°) right–left fusion bicuspid aortic valve with a raphe, and the commissural angle of the non-fused cusp is about 150°. (Right panel) Very asymmetrical (angle 120–139°) right–left fusion bicuspid aortic valve shows retraction of the conjoined cusp at the raphe area and the commissural angle of the non-fused cusp is about 130°. Note that retraction is more prominent as the angle decreases and that this may cause aortic regurgitation. (Modified from Michelena et al.[6] with permission from Elsevier.)

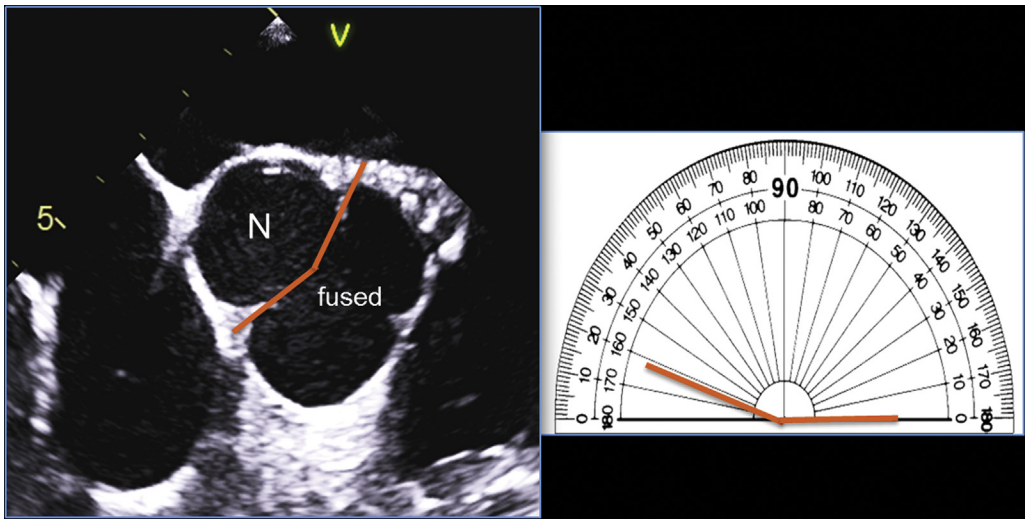

FIGURE 10 Transesophageal echocardiographic measurement of the commissural angle of the non-fused cusp prior to valve repair. Applicable to similar tomographic views obtained using cardiac CT and cardiac MR, after careful visualization of the systolic and diastolic motion of this regurgitant fused-type right–left cusp fusion bicuspid aortic valve, the non-fused commissures are identified, and a line is drawn from the position of the commissures to the center of the valve in diastole (left). The angle of the non-fused cusp (N) is then carefully measured at approximately 162° on the protractor to the right, suggesting a good chance for succesful repair. (Modified from Michelena et al.[11] with permission from Elsevier.)

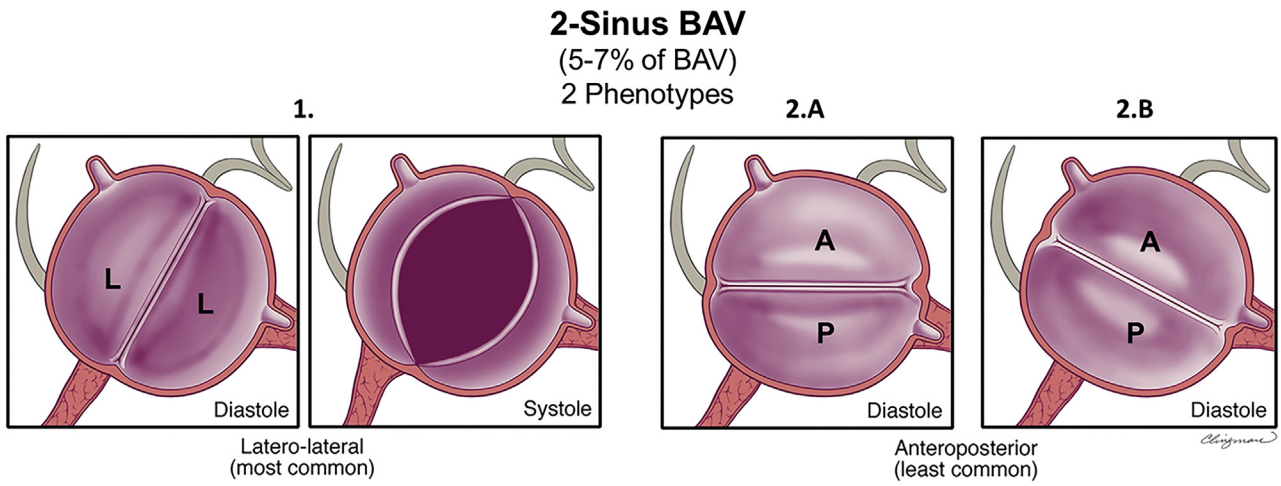

FIGURE 11 Schematic representation of the 2-sinus bicuspid aortic valve phenotypes as seen by the transthoracic echocardiogram parasternal short axis. Applicable to similar tomographic views obtained from cardiac CT and cardiac MR, the figure demonstrates 2-sinus bicuspid aortic valve phenotypes as zoomed views of the base of the heart for anatomical landmark correlation. (Left panels) (1) 2-sinus laterolateral bicuspid aortic valve with only 2 distinguishable aortic sinuses in diastole and 2 cusps of roughly same size and shape, each occupying 180° of the circumference, with a 180° angle of the commissures. Note that although it is possible to suspect right–non-fusion, the landmark anatomical relations are not clear because both the geographic 'left' and 'non-coronary' cusps occupy portions of the normal geographic location of the 'non-coronary' cusp, and the posterior commissural line is almost aligned with the interatrial septum, bisecting the geographical location of the normal non-coronary cusp (Figures 5 and 12). The 2-sinus bicuspid aortic valve laterolateral phenotype has 1 coronary artery arising from each sinus. (Right panels) (2A) a 2-sinus anteroposterior bicuspid aortic valve with only 2 distinguishable aortic sinuses in diastole and 2 cusps of roughly same size and shape each occupying 180° of the circumference, with a 180° angle of the commissures. Note that although it is possible to suspect right–left fusion, but the landmark anatomical relations are not clear because the commissural line actually bisects the normal geographical location of the left cusp, such that both anterior and posterior functional cusps appear to have a 'piece' of the left cusp (see Figures 5 and 12). (2B) A 2-sinus anteroposterior bicuspid aortic valve that resembles a fused right–left fusion but without a raphe, with only 2 distinguishable aortic sinuses in diastole and 2 same size/shape cusps each occupying 180° of the circumference. The 2-sinus anteroposterior bicuspid aortic valve may have coronary arteries arising from each cusp (2A) or from the anterior cusp (2B). (Modified from Michelena et al.[6] with permission from Elsevier.)

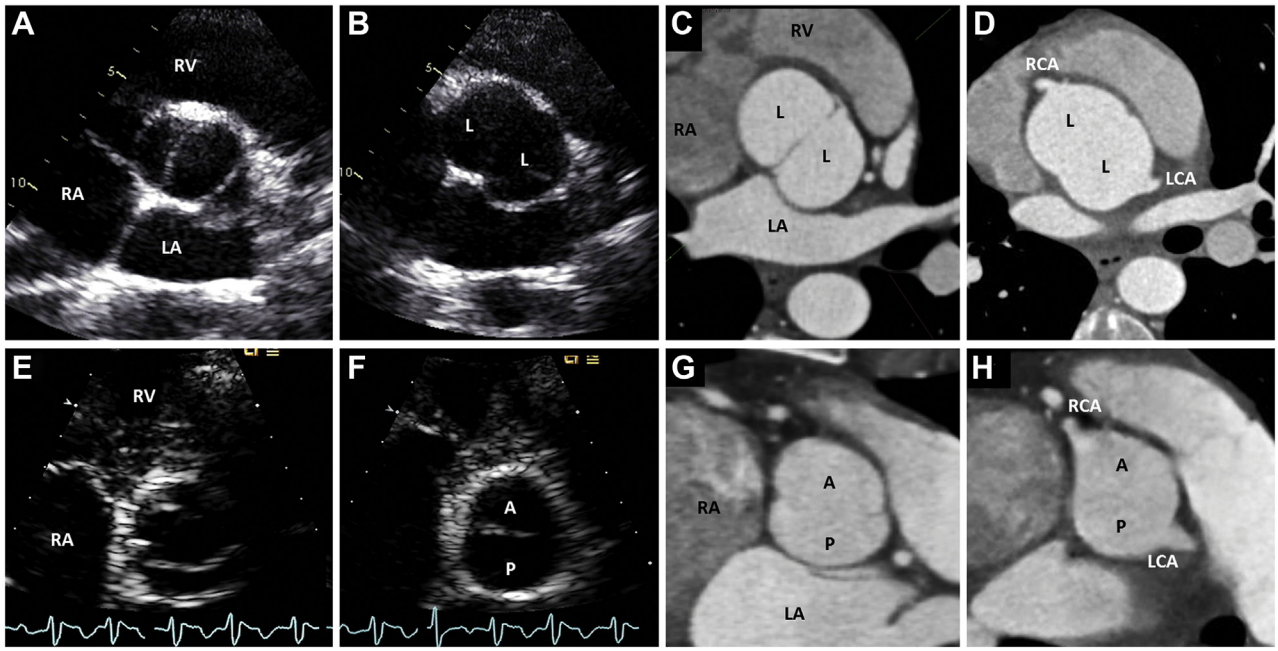

FIGURE 12 Diastolic and systolic short-axis still images of the 2-sinus bicuspid aortic valve phenotypes obtained from transthoracic echocardiographic and diastolic still images from electrocardiographic-gated cardiac CT. (A) A 2-sinus laterolateral bicuspid aortic valve in systole, with the commissural line bisecting the normal geographic position of the non-coronary cusp (B and C), with only 2 distinguishable aortic sinuses in diastole (B), and roughly equal size/shape cusps occupying 180° of the circumference, reproducible on an equivalent tomography cut as seen with cardiac CT (C). Note the coronary arteries arising, 1 from each cusp (D). (E) A 2-sinus anteroposterior bicuspid aortic valve in systole, with the commissural line bisecting the left–coronary cusp geographic position (F) (diastolic still frame), with only 2 distinguishable aortic sinuses and roughly equal size/shape cusps occupying 180° of the circumference, reproducible on an equivalent tomographic cut as seen with cardiac CT (G). Note the coronary arteries arising, 1 from each cusp in this particular example (H). (A, anterior cusp; L, lateral cusp; LA, left atrium; LCA, left–coronary artery; P, posterior cusp; RA, right atrium; RV, right ventricle; RCA, right coronary artery.)

the size of the aortic annulus and the STJ, is indispensable in the maintenance of sufficient diastolic cusp coaptation area to prevent the progression of aortic regurgitation[33] and its recurrence after surgery.[34] Therefore, the aortic root complex is the anatomical scaffold that maintains BAV competency, with the BAV cusps acting as a stentless valve and the root complex as its native stent.[32]

The tract of the proximal aorta, spanning from the STJ to the brachiocephalic artery take-off, should be referred to as the tubular ascending aorta or ascending aorta. The subsequent tract, from the brachiocephalic artery to the isthmus (the physiological narrowing just distal to the left subclavian artery origin), is called the aortic arch.

CONSENSUS ON BAV NOMENCLATURE AND CLASSIFICATION FOR CLINICAL, SURGICAL, INTERVENTIONAL AND RESEARCH PURPOSES

BAV TYPES AND SPECIFIC PHENOTYPES. There are 3 BAV types: the fused BAV, the 2-sinus BAV and the partial-fusion BAV, each with specific phenotypes[1,6] (Figure 4).

The fused BAV type. The fused BAV type is the most common (Figures 5 and 6), accounting for 90-95% of BAV cases[2,28]. The fused BAV is characterized by 2 of the 3 cusps appearing fused or joined within 3 distinguishable aortic sinuses, resulting in 2 functional cusps (1 fused or conjoined and the other non-fused) that are usually different in size and shape (Figures 6-8), with non-fused cusp commissural angles of varying degrees (Figure 9). Commonly, patients with a fused BAV demonstrate eccentric dominance of the non-fused aortic sinus and its cusp (compared to the other 2 sinuses and 2 fused cusps), irrespective of age[35] (Figures 6 and 7). Frequently (70%) but not always (Figure 8), a congenital fibrous ridge occurs between the fused cusps, termed 'raphe'[28,36]. The presence of a raphe has been associated with the progression of valvular dysfunction (particularly AS) and future valvular surgery.[26,36,37] A raphe may be present but not initially visible on the echocardiogram and may become visible years later.[38]

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There are 3 specific BAV phenotypes within the fused type: right-left cusp fusion, right-non (non-coronary)-cusp fusion and left-non-cusp fusion (Figures 4, 6 and 7). The right-left cusp fusion phenotype is the most common (70-80%).[2,28,39] The right- left cusp fusion phenotype is also the most common across all variations of aortic phenotypes (normal aorta, dilated ascending aorta, dilated root, dilated arch) and across valve dysfunction (regurgitation or stenosis). Although this right-left fusion phenotype statistically develops more AS[2], it has been associated in some patients[40,41] with aortic root dilatation, aortic regurgitation and male preponderance (these associations have been termed the 'root phenotype').[38,39] The right-left cusp fusion is also strongly associated with aortic coarctation in children.[42]

The right-non-cusp fusion phenotype is the next most common (20-30%). It is associated with a higher prevalence of AS in adults[37] and also independently predicts aortic regurgitation progression in adults.[33] Similarly, the right-non-cusp fusion phenotype is associated with a more rapid progression of AS and regurgitation in children and adolescents.[42,43] The left-non-cusp fusion phenotype is the least common phenotype (3-6%) across studies.

Referring to the fused phenotypes as BAV with right-left cusp fusion, right-non-cusp fusion or left-non-cusp fusion is appropriate. Occasionally, it is possible to recognize a fused BAV but not to be able to discern the fusion phenotype, in which case BAV with indeterminate cusp fusion is appropriate (Figure 4). It is important to recognize that some fused BAVs may not have a congenital raphe[28] or may have a raphe that is not visible via imaging[38], yet they have 3 distinguishable aortic sinuses, and the 2 fused cusps can usually be identified (Figure 8).

BAV symmetry for the fused BAV type is defined by the angle between the commissures of the non-fused cusp and has recently become a critical aspect in the planning and performance of BAV repair for pure aortic regurgitation.[6,20,44] From a regurgitation-treatment perspective, the BAV 'concept' offers a single-line coaptation surface (a tricuspid aortic valve has 3 coaptation lines; Figure 5, left); as long as that single coaptation line is straight or almost straight (Figures 8 and 9—symmetrical), the repair of the regurgitant BAV is reproducible. As the angle between the commissures of the non-fused cusp decreases <160° [44], the BAV becomes less symmetrical, resembling more a tricuspid (especially <140°) valve (Figure 9, very asymmetrical), which becomes technically more challenging for the surgeon to 'bicuspidize' during the repair, yet remains repairable in experienced hands. Asymmetrical valves may exhibit retraction of the free edge of the fused cusp at the raphe level, which is best appreciated by direct surgical visualization (Figures 2 and 9) or gross pathological inspection and not reliably by imaging. This retraction may contribute to valve regurgitation. Measuring the commissural angle of the non-fused cusp with transesophageal echocardiography before cardiopulmonary bypass aids the surgeon in planning the repair (Figure 10). Therefore, the symmetry of a fused-type BAV is defined by the angle between the commissures of the non-fused cusp.

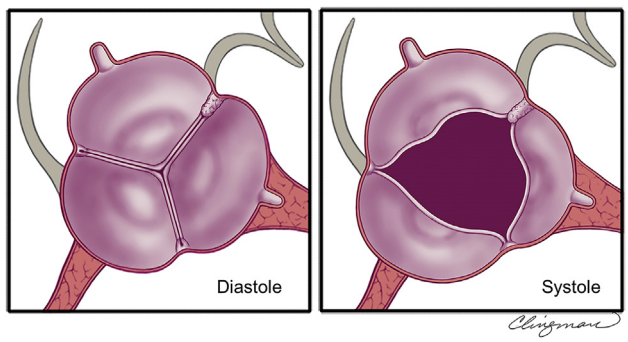

FIGURE 13 Schematic representation of the partial-fusion bicuspid aortic valve phenotype as seen from the transthoracic echocardiogram parasternal short axis view. (Left panel) The imaging appearance in diastole of the partial-fusion or forme fruste bicuspid aortic valve is that of a tricuspid aortic valve. (Right panel) The imaging diagnosis is usually made in systole. Although the opening appears triangular, there is a small fusion of the right and left cusps with a 'mini-raphe'. These can be suspected by transthoracic or transesophageal echocardiogram, and confirmed by a 3-dimensional transesophageal echocardiogram, cardiac MR or cardiac CT. Definitive confirmation is usually made by surgical inspection or pathological analysis. (Modified from Michelena et al.[6] with permission from Elsevier.)

The 2-sinus BAV type. The 2-sinus BAV type is uncommon, accounting for 5-7% of BAV cases.[2,6,28] In contrast to the fused type, the appearance of the 2-sinus BAV does not suggest that 2 of the 3 cusps have fused; instead, it suggests that 2 cusps of roughly equal size and shape, each cusp occupying 180° of the annular circumference, were 'formed' within only 2 aortic sinuses, resulting in a 2-sinus/2-cusp valve (Figures 11 and 12) with 180° commissural angles. It is difficult to determine which 2 cusps could have coalesced to form a 2-sinus BAV, but it is usually evident whether the cusps are laterolateral (side-to-side) or anteroposterior (front and back) within the short-axis base-of-the-heart plane (Figures 11 and 12); thus, these are 2 specific phenotypes of the 2-sinus BAV category. The 2-sinus laterolateral

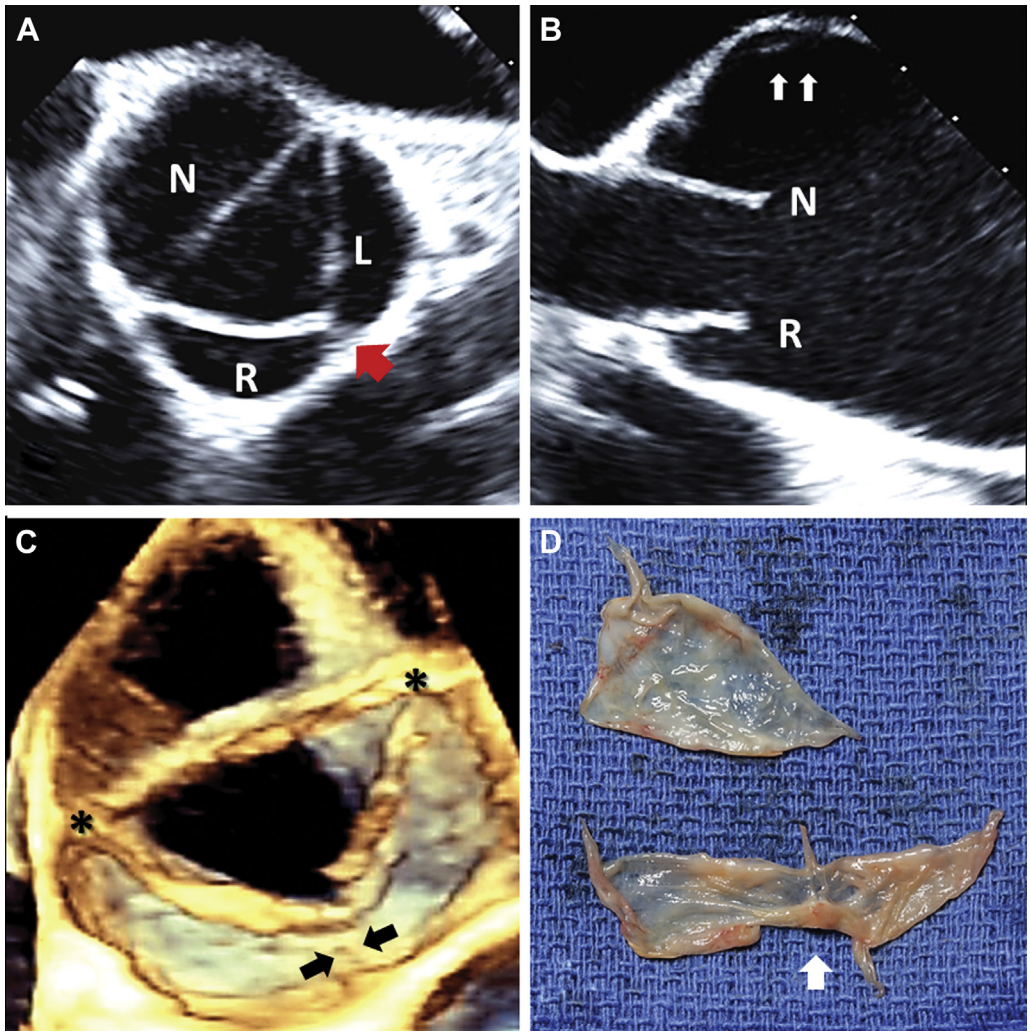

FIGURE 14 Systolic transesophageal echocardiogram still images and intraoperative photograph of a partial-fusion bicuspid aortic valve. (A) Intraoperative 2-dimensional transesophageal echocardiogram shows a triangular systolic opening with a suspected small fusion between the right and left cusps (red arrow). (B) The 2-dimensional transesophageal long axis demonstrates no evidence of systolic doming with asymmetrical dilatation of the non-coronary sinus (arrows), which was accompanied by significant dilatation of the ascending aorta in this patient. (C) 3-Dimensional transesophageal systolic short axis demonstrates a small raphe (arrows) between the right and left–coronary cusps with 2 other normal commissures (asterisks). (D) Explanted valve shows the small raphe between the right and left cusps (arrow). (N, non-coronary cusp.)

BAV has 1 coronary artery arising from each cusp, whereas the an-teroposterior BAV may have 1 coronary artery arising from each cusp or both coronary arteries arising from the anterior cusp (Figures 11 and 12). The 2-sinus BAV likely represents a more severe expression of the embryological mechanisms leading to the fused BAV. Referring to these phenotypes as 2-sinus latero-lateral BAV and 2-sinus anteroposterior BAV is appropriate. Occasionally, despite suspicion, it may be difficult to be certain whether there are only 2 sinuses, in which case, terms such as possible or probable 2-sinus BAV may be used. There is a lack of scientific data on the clinical/prognostic associations of the 2-sinus BAV, which represents a 'morphologically severe' form of BAV. Therefore, we hope that, through this nomenclature/classification system, the research community directs more attention towards this BAV type.

The partial-fusion BAV (or forme fruste BAV) type. The prevalence of this recently recognized partial-fusion BAV (or forme fruste BAV) is unknown[45] (Figure 13). The appearance of the partial-fusion BAV is that of a typical tricuspid aortic valve with 3 sinuses and 3 symmetrical cusps with a systolic triangular opening and commissural

angles of 120°, yet on surgical inspection or high-resolution imaging, <50% cusp fusion is noted at the base of a commissure, forming a small 'mini-raphe'.[6,45-47] It is important to recognize and further study the partial-fusion BAV, which has been described most frequently in the operating room in patients undergoing surgery for aortic dilatation[45] (Figure 14).[47] This forme fruste BAV results in alteration of aortic flow patterns, consisting of increased flow eccentricity and increased vortexes [46], perhaps partially explaining the apparent high prevalence of aortic dilatation in these patients. Referring to this phenotype as partial-fusion BAV or forme fruste BAV is appropriate, as is noting between which cusps the fusion occurs: right-left, right-non and so forth.

DEFINITION OF AORTIC DILATATION AND BAV AORTOPATHY. Definition of aortic dilatation. The definition of 'aortic aneurysm' [48] is rarely applied in clinical practice, and the term aneurysm carries a sombre or dismal connotation for patients. Therefore, we propose a simple, universal term: aortic dilatation. Qualitative descriptive terms, such as saccular or fusiform dilatation or STJ effacement, may be important for aorta specialists and surgeons. A full discussion on aortic dilatation in patients with BAV is presented in the full document.[1]

BAV aortopathic phenotypes. The importance of recognizing BAV aortopathic phenotypes is that their presence and association with specific valvular phenotypes and dysfunction patterns may imply different clinical histories for a patient with BAV.[49] There are 2 major forms of aortic dilatation BAV phenotypes: (i) the ascending phenotype (dilatation preferentially located at the tubular ascending tract beyond the STJ) (Figure 15), accounting for ~70% of BAV aortopathy cases, and (ii) the root phenotype (dilatation preferentially located at the root [sinuses of Valsalva]) accounting for ~20% of BAV aortopathy cases (Figure 15).[6,40,41,50] Importantly, the root phenotype may have mild ascending dilation but significantly prevails at the root, and the ascending phenotype may have mild root dilatation but significantly prevails at the ascending portion. In addition, these 2 categories often correspond to 2 clearly distinct overall patient phenotypes: roughly, the older patient with BAV, either male or female, presenting more often with aortic valve sclerosis/stenosis (ascending phenotype) with fused-type right-non fusion, and the younger patient with BAV, usually male, presenting with aortic regurgitation of degrees ranging from mild to severe (root phenotype) with fused-type left-right fusion.[40,51,52] However, those associations are not unequivocal, and the right- left cusp fusion BAV can be associated with either aortic phenotype.[52] The root phenotype has been associated with greater rates of acute aortic dissection in the postoperative follow-up of patients with BAV who had undergone simple aortic valve replacement compared to the ascending phenotype.[53]

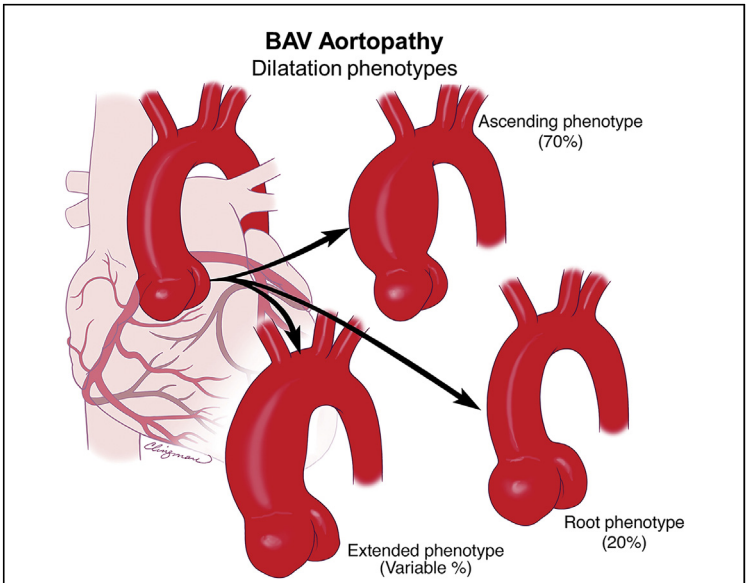

FIGURE 15 Bicuspid aortic valve aortopathy phenotypes. On the left is a normal aorta. (Top) The most common phenotype (70%), the ascending phenotype, is preferential dilatation of the tubular ascending aorta. (Middle) The root phenotype involves preferential dilatation of the root, seen in approximately 20% of patients with bicuspid aortic valve with aortopathy. (Bottom) The extended phenotype shows dilatation of the root, the ascending aorta and the arch. The most common extended phenotypes are root plus ascending aorta and ascending aorta plus arch.

Notably, in some cases, the dilation of the aorta does not significantly prevail at 1 segment. In a proportion of these cases, a localized dilatation at first observation can evolve during follow-up, with possible dilatation of previously normal adjacent segments of the aorta. In this scenario, the ascending phenotype can present, especially if a right-non-cusp fusion valve is present, with associated dilatation of the aortic arch; it is appropriate to refer to this condition as ascending phenotype extended. Similarly, the root phenotype has been demonstrated to be independently associated with faster growth of the ascending tubular tract, so that cases of 'cross-over' from an initial root phenotype configuration to significant dilatation of both tracts have been observed (Figure 15); root phenotype extended would be the appropriate definition of this form. In the context of a root phenotype, the presence and progression of effacement of the STJ may be an initial sign of this kind of evolution.

SUMMARY. The BAV phenotypic expression represents an anatomical continuum that is likely related to the severity of its embryological mechanisms. Therefore,

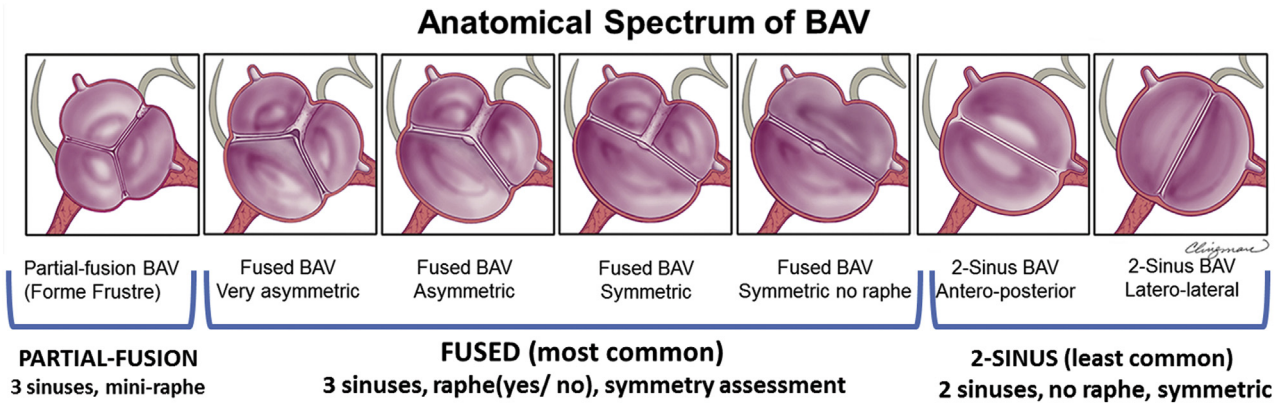

FIGURE 16 Schematic representation of the bicuspid aortic valve anatomical spectrum using the most common right–left cusp fusion as an example. From left to right, note the partial-fusion bicuspid aortic valve resembling a tricuspid aortic valve likely associated with a mild embryological defect, then spanning a continuum of increasing non-fused cusp commissural angles and increasing cusp size/shape similarity, ending with the 2-sinus bicuspid aortic valve phenotypes that represent almost perfect 'bicuspidity' and are likely associated with the most severe embryological defects. (Modified from Michelena et al.[6] with permission from Elsevier.)

we propose a general BAV anatomical spectrum (Figure 16) of BAV phenotypes in order of 'bicuspidity', defined as resemblance to a 2-sinus BAV. This spectrum represents a continuum of increasing non-fused cusp commissural angles and increasing similarity of cusp size and shape.

Based on the new nomenclature and classification consensus, Figure 17 presents a simple algorithm of the critical imaging evaluation for BAV valvulo-aortopathy. Three critical anatomic aspects to be described in all patients with BAVs are (i) the type and specific phenotype of the BAV and valve function; (ii) the presence and characteristics of the raphe and the cusp size/shape and symmetry of the BAV; and (iii) the presence and phenotype of aortopathy (aortic dilatation) and whether or not coarctation is present.

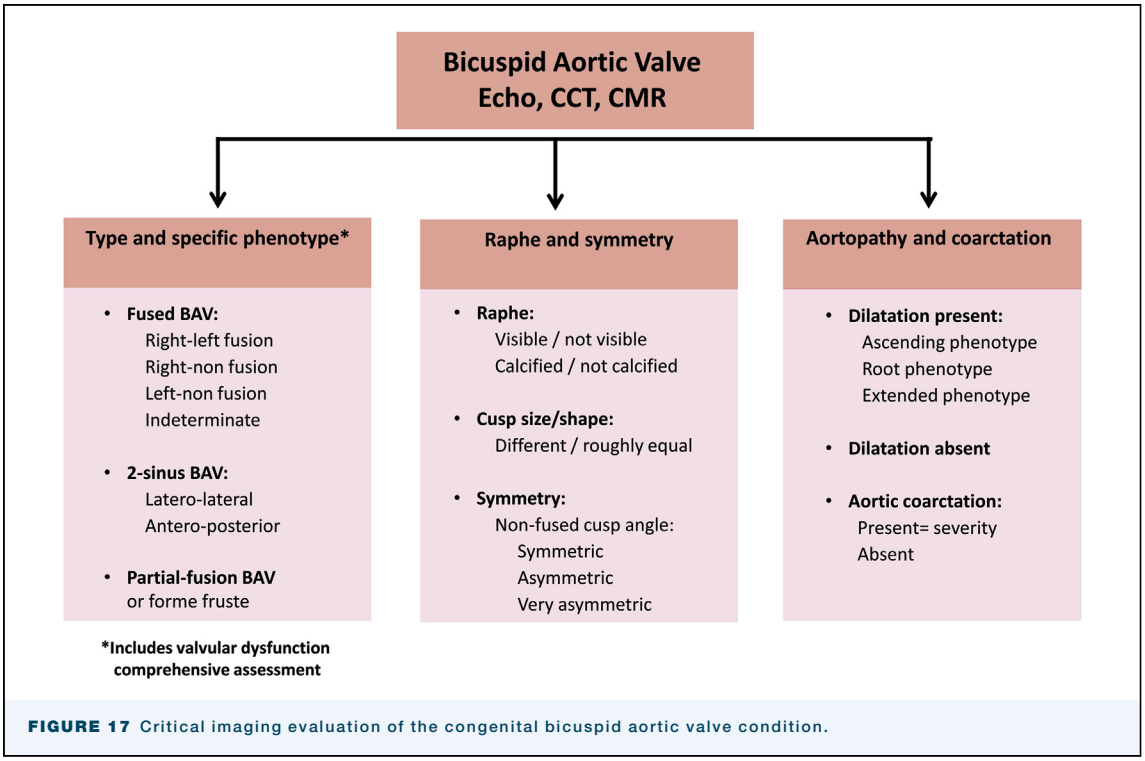

FIGURE 17 Critical imaging evaluation of the congenital bicuspid aortic valve condition.

REPORT

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Conflict of interest: Victoria Delgado: discloses a financial relationship with Abbott Vascular, Edwards Lifesciences, GE Healthcare, MSD, Medtronic, and Novartis. **Emmanuel Lansac:** patent Extra Aortic ring annuloplasty device with Coroneo Inc. **Phillippe Pibarot:** funding from Edwards Lifesciences and Medtronic for echocardiography core laboratory services with no personal compensation. **Michael A. Borger:** discloses a financial relationship with Edwards Lifesciences, Medtronic, Abbott, and CryoLife. **John K. Forrest:** grant support and consultant to: Edwards Lifesciences, Medtronic Inc. **John Webb:** consultant to: Edwards Lifesciences, Abbott, Boston Scientific. **Martin B. Leon:** institutional clinical research grants from Abbott, BSC, Edwards and Medtronic. **Michael Markl:** research support—Siemens Healthineers, Research Grant—Circle Cardiovascular Imaging, Consulting—Circle Cardiovascular Imaging, Research Grant—CryoLife Inc. **Victor A. Ferrari:** Senior Advisory Board, *Journal of Cardiovascular Magnetic Resonance*. **Philipp Blanke:** consultant for Edwards Lifesciences and Circle Cardiovascular Imaging; and provides CT core lab services for Edwards Lifesciences, Medtronic, Neovasc, and Tendyne Holdings, for which he receives no direct compensation. **Ruggero De Paulis:** patent on aortic root graft with Terumo Aortic. Consultant for Edwards Lifesciences, Medtronic, and Terumo Aortic. **Maurice Enriquez-Sarano:** consulting fees from Edwards LLC, CryoLife, and ChemImage. The Department of Cardiology of the LUMC received unrestricted research grants from Abbott Vascular, Bayer, Biotronik, Bioventrix, Boston Scientific, Edwards Lifesciences, GE Healthcare, and Medtronic. The other authors report no conflict of interest.

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