**Supplemental Figure 1.** Panels A and B present standard two-dimensional echocardiographic images of the two common forms of the functionally bileaflet aortic valve with trisinuate aortic root, while Panels C and D represent the two common forms of the bileaflet aortic valve with bisinuate aortic root. Column 1 and 2 represent the diastolic and systolic short axis view of the same plane of interrogation, while Column 3 represents the systolic long axis view, with the red hashed line in Column 2 demonstrating the approximate corresponding plane obtained across the short axis view. In the functionally bileaflet aortic valve, it is only the systolic frame which allows delineation of the raphe and zone of fusion between two leaflets. In the functionally bileaflet aortic valve with fusion between the right and left coronary leaflets (Panels A1-3), the long axis plane (Panel A3) demonstrates doming of the anterior, or right coronary leaflet in systole (white star). The non-coronary leaflet, which is not involved in the leaflet fusion, reaches up near the sinutubular junction, without doming. The height this doming leaflet reaches superior to the plane of the virtual basal ring in systole is roughly analogous to the height of the corresponding hypoplastic interleaflet triangle. In the functionally bileaflet aortic valve with fusion between the right and non-coronary leaflets (Panels B1-3), the long axis plane (Panel B3) demonstrates doming of both the anterior, or right coronary leaflet, and posterior, or non-coronary leaflet (white stars). Similarly, the heights of these doming leaflets correspond to the hypoplastic interleaflet triangle between these two leaflets. In the bileaflet aortic valve with bisinuate aortic root (Panels C and D), there is no leaflet fusion, and each of the two interleaflet triangles are normal in height. As expected, there is no doming of either leaflet. In the bileaflet valve with antero-posterior configuration (Panels C1-3), the long axis image cuts across both leaflets (Panel C3), and both leaflets are seen to reach near the sinutubular junction in systole. In the form with latero-lateral configuration (Panels D1-3), the long axis now cuts obliquely across the right-sided leaflet, without clear delineation of this normal leaflet, without doming in systole. With this understanding, the long axis plane can be used in two-dimensional echocardiography when the clinician questions if an aortic valve represents a functionally bileaflet and trisinuate aortic root with indistinct raphe versus a bileaflet and bisinuate aortic root.

**Supplemental Figure 2.** These panels demonstrate two different patients, both with a functionally bileaflet and trisinuate aortic root with fusion between the coronary leaflets. Panels A-D demonstrate a patient in which the raphe is indistinct, while Panels E-H demonstrate a patient in which the raphe is more prominent. Three-dimensional volume-rendered computed tomographic reconstructions are demonstrated in Panels A-C and E-G. The aortic valves are viewed in systole (Panels A and E) and diastole (Panels B and F). The location of the apex of the hypoplastic interleaflet triangle (blue star) and the zone of fusion with variable degree of raphe (red arrow) are marked. There is a slightly longer length of fusion in the first case (Panels A and B) compared to the second case (Panels E and F), with more significant hypoplasia of the corresponding interleaflet triangle in the first case (Panel C) compared to the second (Panel G). In the first case, a clinician could easily mistake this to be a bileaflet valve with bisinuate root, however, close inspection of the aortic root with three-dimensional imaging demonstrates three sinuses with one of the three interleaflet triangles being hypoplastic (blue star in Panel C) corresponding with the zone of fusion between the coronary leaflets. For surgical planning, the height of this interleaflet triangle, or commissural height (CH), accurately can be assessed with multiplanar reformatting (Panel D).The blue dot in Panel D represents the nadir of the non-coronary leaflet, with the green line representing the plane of the virtual basal ring. These same techniques can be applied to three-dimensional echocardiography, including assessment of the zone of fusion and corresponding raphe as demonstrated in Panel H. LCA, left coronary artery; RCA, right coronary artery.

**Supplemental Figure 3.** The panels show three-dimensional volume-rendered computed tomographic reconstructions of a functionally unileaflet valve in a trisinuate scaffold. Panels A and B visualize the short axis of the aortic root in diastole (Panel A) and systole (Panel B). The virtual basal ring is colored green (compare to the normal aortic root in Figure 1A). The single commissure is shown by the white star with red borders. The apices of the hypoplastic interleaflet triangles are marked with blue and yellow stars. Panel C demonstrates a long axis view, highlighting the angle of valvar opening (white hashed lines). Panels D, E and F demonstrate the blood-filled cavity of the aortic root, with variable degrees of hypoplasia of two of the three interleaflet triangles. Panels G, H and I then demonstrate corresponding virtual dissections during systole. L, left coronary leaflet or sinus; LCA, left coronary artery; N, non-coronary leaflet or sinus; R, right coronary leaflet or sinus; RCA, right coronary artery.

**Supplemental Figure 4.** The three commonly used methods for measuring the trisinuate aortic root by cross-sectional imaging are demonstrated, with measured values given for the thicker dashed line for comparison. As demonstrated in this example, these three methods can provide significant differences in measured dimensions. Of note, two of the methods are indistinctly referred to as “cusp-to-cusp” measurements. Normative adult data only exists for the widest sinus-to-sinus and center of sinus-to-opposite commissure measurements. The center of sinus-to-center of sinus measurement may better correlate with two-dimensional echocardiographic measurements. Currently, there is no robust normative data available for children. While there is no evidence to suggest the superiority of one method over the other, institutional consistency and clarity in the method used is paramount.

**Supplemental Table 1.** **Terms and definitions for the components of the normal aortic root and its evaluation.**

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
| **TERM** | **DEFINITION** | **COMMENTARY** | | **SYNONYMS** |
| **Normal Structures of the Aortic Root and Its Support** | | | | |
| **Aortic root (anatomical definition)** | The space and structures contained between the planes of the virtual basal ring and the sinutubular junction. | The virtual basal ring indicates the proximal junction of the aortic root. There is, however, ventricular myocardium incorporated into the bases of the coronary sinuses, forming a myocardial-arterial junction which becomes incomplete in the area of fibrous support of the root. | |  |
| **Aortic root (hemodynamic definition)** | The space and structures extending from the arterial surfaces of the aortic valvar leaflets in their closed positions to the plane of the sinutubular junction. As such, the interleaflet triangles are not part of the hemodynamic aortic root. | Defined during diastole, portion of the anatomic aortic root proximal to the ventricular aspect of the closed leaflets exposed to left ventricular pressure is hemodynamically part of the left ventricular outflow tract. | |  |
| **Aortic valvar complex** | The functional unit made up of the valvar leaflets, their supporting sinuses, and the fibrous interleaflet triangles. |  | |  |
| **Aortic valvar leaflets** | The semilunar moving components of the valvar complex, with each leaflet having nadirs at the level of the virtual basal ring and zeniths at the sinutubular junction. The leaflet is attached to the wall of the aorta in semilunar fashion from commissure to commissure. |  | | Aortic valvarcusp |
| **Nadir of the leaflet** | The lowest point of semilunar attachment of an arterial valvar leaflet. Joining together the nadirs defines the plane of the virtual basal ring. |  | | Leaflet nadir |
| **Zenith of the leaflet** | The highest point of attachment of the semilunar leaflet of an arterial valve at the sinutubular junction. |  | | Leaflet zenith, commissure |
| **Belly of the leaflet** | The middle portion of the leaflet between its attachment lines and free margin. |  | | Leaflet belly |
| **Zone of Apposition** | The surface areas where leaflets meet during ventricular diastole. In the normal root, three such areas extend from the centroid of the valvar orifice to the peripheral attachments at the sinutubular junction. | The peripheral attachments are the commissures. The areas of apposition extend to the nodules of Arantius at the free edge of the leaflets, as well as the lunules on either side of the nodules. | | Leaflet coaptation, coaptation area |
| **Commissure** | The peripheral attachments of adjacent leaflets at the sinutubular junction, marking their zeniths of attachment. | The peripheral extents of the zones of apposition. | |  |
| **Aortic sinus** | The wall of the aortic root within the boundaries of the semilunar insertion lines of the leaflet, delimited superiorly by the sinutubular junction. | The normal arterial root has three such sinuses, as do the majority of the examples of “bicuspid” and “unicuspid” roots. | | Sinus of Valsalva |
| **Interleaflet triangle** | The fibrous tissue forming the walls of the anatomic root between the semilunar attachments of adjacent leaflets, delimited inferiorly by the virtual basal ring. | A variable portion of the membranous septum is included within the interleaflet triangle between the non- and right coronary sinuses. | | Intercusp triangle |
| **Virtual basal ring** | A geometric planar surface, limited by the confines of the aortic root, created by joining the nadirs of attachment of the semilunar valvar leaflets. | This marks the proximal boundary of the anatomic aortic root. | | Annulus, echocardiographic valvar annulus |
| **Myocardial-arterial junction** | The boundary between the myocardial and arterial walls within the aortic root. | Such a junction in the aortic root is found only within the valvar sinuses giving rise to the coronary arteries. This junction has historically been referred to as an anatomical ventriculo-arterial junction, however, incorrectly suggesting a continuous boundary extending across the area of aortic-mitral fibrous continuity where no anatomical boundary can be distinguished. This junction, therefore, is better referred to as the myocardial-arterial junction, and the proximal anatomical boundary of the aortic root is better inferred by the plane of the virtual basal ring. | | Ventriculo-arterial junction |
| **Hemodynamic ventriculo-arterial junction** | The anatomical boundaries responsible for producing the changes in pressure, during ventricular diastole, between the ventricular cavities and the intrapericardial arterial trunks. | The junctions are obvious only during ventricular diastole, since during systole the pressures are equalized between the ventricles and the arterial trunks. | | Hemodynamic ventriculo-aortic junction |
| **Sinutubular junction** | The ring at the junction between the distal extent of the valvar sinuses and the tubular aorta. | The peripheral ends of the zones of apposition between the leaflets, described as the commissures, are at the level of the sinutubular junction. | | Sinotubular junction |
| **Ascending aorta** | The portion of the thoracic aorta extending from the plane of the sinutubular junction to the origin of its first brachiocephalic branch. |  | | Tubular ascending aorta |
| **Nodule of Arantius** | A small fibrous tissue formation found normally at the center of the free margin of each arterial valvar leaflet. The nodules of all three aortic leaflets meet at the center of the valve during diastole. |  | | Nodes of Arantius, nodes of the semilunar cusps |
| **Central fibrous body** | The largest part of the fibrous skeleton of the heart, made up of the membranous septum, the roof of the inferoseptal recess, and the right fibrous trigone. |  | |  |
| **Membranous septum** | The fibrous component of the septal structures, divided by the insertion line of the septal leaflet of the tricuspid valve into atrioventricular and interventricular portions. |  | | Membranous interventricular septum |
| **Inferoseptal recess** | The space within the left ventricle extending below the plane of the virtual basal ring of the aortic root. Its roof, representing fibrous tissue between the atrioventricular valves, is bordered by the right fibrous trigone, the membranous septum and crest of the ventricular septum. | Its roof is now recognized as an integral part of the central fibrous body. | | Sibson’s vestibule |
| **Right fibrous trigone** | The rightward thickening of the area of fibrous continuity between the aortic, or anterior, leaflet of the mitral valve and the leaflets of the aortic valve. |  | |  |
| **Aortic-mitral fibrous curtain** | The area of fibrous continuity between the aortic, or anterior, leaflet of the mitral valve and the leaflets of the aortic valve. |  | | Aorto-mitral curtain, mitral-aortic intervalvar fibrosa |
| **Left fibrous trigone** | The leftward thickening of the area of fibrous continuity between the aortic, or anterior leaflet of the mitral valve and the leaflets of the aortic valve. |  | |  |
| **Inferior pyramidal space** | The extracardiac fibro-adipose tissue wedging inferiorly between the four cardiac chambers from the diaphragmatic surface of the heart. | The atrioventricular nodal artery is commonly carried within this space to the atrioventricular node, which is commonly positioned at the apex of the inferior pyramidal space. | |  |
| **Measurement Planes and Metrics of the Aortic Root** | | | | |
| **Virtual basal ring dimensions** | Two-dimensional measurements of the length of lines drawn between the short axis view formed between the nadirs of the leaflets, across the aortic orifice. |  | |  |
| **Dimensions of the Sinuses of Valsalva** | A two-dimensional measurement of the plane extending between the maximum extents of the short axis of the sinuses. |  |  | |
| ***Center of sinus-to-center of sinus*** | A two-dimensional measurement in the short axis view of the aortic sinuses measuring from the center wall of one sinus to the center wall of an adjacent sinus. | There has been no distinction between this and the following measurement plane in the terminology used by those using the term ‘cusp-to-cusp’, despite significant differences on average between reported values.  It is also worth noting, that the common use of ‘cusp’ to denote this measurement, is using cusp synonymously with sinus in this circumstance, which we would discourage. | | Cusp-to-cusp |
| ***Largest sinus-to-sinus*** | A two-dimensional measurement in the short axis view of the aortic sinuses determining the longest length between the walls of two adjacent sinuses. | There has been no distinction between this and the preceding measurement plane in the terminology used by those using the term ‘cusp-to-cusp’, despite significant differences on average between reported values.  In contrast to the other two methods for measuring the dimensions of the sinuses of Valsalva, though this may achieve the off-center plane discussed below, there are no distinct points to guide this measurement. | | Cusp-to-cusp |
| ***Center of sinus-to-opposite commissure*** | A two-dimensional measurement in the short axis view of the aortic sinuses measuring from the center wall of one sinus and continuing along the zone of apposition between the adjacent two leaflets, ending at the midline of the opposite interleaflet triangle. | Similar to above, those using ‘cusp’ in this instance, are doing so when referring to the sinus. | | Cusp-to-commissure |
| **Long Axis Plane Cuts of the Aortic Root** |  |  |  | |
| ***Center bisecting plane*** | A long axis plane obtained by cutting the short axis of the sinuses from the center wall of one sinus, across the central point of coaptation and continuing along the zone of apposition between the adjacent two leaflets, ending at the midline of the opposite interleaflet triangle. | The only long axis plane which can accurately measure all the discussed aortic valvar leaflet metrics. | | Bisecting center plane |
| **Center non-specific plane** | A long axis plane obtained by cutting the short axis of the sinuses from an indistinct point along the wall of one sinus, across the central point of coaptation, and ending at an indistinct point along the wall of an adjacent sinus. |  | |  |
| ***Off-center plane*** | A long axis plane obtained by cutting the short axis of the sinuses from the nadir of one leaflet insertion line to the nadir of an adjacent one. |  | | Bi-nadir, largest sinus-to-sinus |
| **Metrics of the Aortic Valvar Leaflets** |  | All of these metrics can only accurately be measured using multiplanar reformatting. |  | |
| ***Effective Height*** | A linear measurement from a central point of the virtual basal ring to the cephalad end of the central coaptation segment of the aortic valvar leaflets. |  | |  |
| ***Coaptation Length*** | A linear measurement of the central segment of coaptation between two leaflets, from the proximal ventricular surface to central tip involved in coaptation. |  | |  |
| ***Geometric Height*** | A curvilinear measurement taken within the extent of the root along the midline of the leaflet from its nadir at the virtual basal ring to the center point of its free margin edge. |  | |  |
| ***Free Margin Length*** | A curvilinear measurement along the leaflet edge between its commissures. |  | |  |
| ***Commissural height*** | A two-dimensional measurement from the plane of the virtual basal ring, extending along the long axis of an interleaflet triangle to its apex at the sinutubular junction. | The common use of this term is applied when measuring the height of both a commissure, but also a pseudocommissure, above the plane of the virtual basal ring. | |  |

**Supplemental Table 2.** **Classification of the congenitally malformed aortic root with abnormal arrangement of the leaflets, sinuses and interleaflet triangles.**

|  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **TERM** | **IPCCC #** | **DEFINITION** | | | | | **COMMENTARY** | **SYNONYMS** |
| **Functionally Bileaflet Aortic Valve with Trisinuate Root** | 09.15.46 | A congenital cardiovascular malformation where there is a zone of fusion between two leaflets of the aortic valve so that the valve functions as two leaflets within an aortic root with three sinuses. | | | | | A raphe, defined as a ridge of tissue or seem located at the zone of fusion between two leaflets, is present to a variable extent, whether visibly obvious or indistinct. This variability should be described. | Functionally bicuspid valve with trisinuate root, Functionally bicuspid aortic valve with three leaflets and a three-sinus root |
|  | **PHENOTYPES** | | | | **IPCCC #** |  |  |  |
| **Fusion right and left coronary leaflets** | | | | 09.15.35 |  | Cusps can be substituted for leaflets. |
| **Fusion right and non-coronary leaflets** | | | | 09.15.36 |
| **Fusion left and non-coronary leaflets** | | | | 09.15.52 |
| **ADDITIONAL QUALIFIERS** | | | | **IPCCC #** |  |  |  |
|  | **Degree of fusion:** | **Partial fusion** | | | Q1.16.01 | Fusion of less than half of the zone of apposition. |  |  |
| **Complete fusion** | | | Q1.16.02 | Fusion of greater than half of the zone of apposition. | When present, this can be inferred and does not need to be specified. |
| **Relationship of commissures:** | **Symmetric** | | | Q1.16.03 | The two commissures and corresponding zones of apposition are positioned 160 degrees or greater related to each other. | Increasing asymmetry, or a decreasing angle, between the commissures and their zones of apposition directly correlates with increasing asymmetry between the fused leaflets and their sinuses, if considered as a single functional unit, when compared to the non-fused leaflet and its sinus. |  |
| **Asymmetric** | | | Q1.16.04 | The two commissures and corresponding zones of apposition are positioned 140 to 159 degrees related to each other. |
| **Very asymmetric** | | | Q1.16.05 | The two commissures and corresponding zones of apposition are positioned 139 degrees or less related to each other. |
| **Bileaflet Aortic Valve with Bisinuate Root** | 0.9.15.56 | A congenital cardiovascular malformation where there are two leaflets of the aortic valve within an aortic root with two sinuses. | | | | |  | Bicuspid aortic valve with bisinuate root, Anatomically bicuspid aortic valve with two leaflets and two-sinus root, Anatomically bicuspid aortic valve |
|  | **PHENOTYPES** | | | | **IPCCC #** |  |  |  |
|  | **Relationship of leaflets:** | | **Latero-lateral** | | 09.15.53 | There is no zone of fusion between leaflets and no raphe. The coronary arteries commonly originate from separate sinuses in the latero-lateral variant, but can originate from the same in the antero-posterior variant. |  |
|  | **Antero-Posterior** | | 09.15.54 |
| **Functionally Unileaflet Aortic Valve with Trisinuate Root** |  | A congenital cardiovascular malformation where there are two zones of fusion between three leaflets of the aortic valve so that the valve functions as one leaflet within an aortic root with three sinuses. | | | | | Two raphes, defined as a ridge of tissue or seem located at the zone of fusion between two leaflets, are present to a variable extent, whether visibly obvious or indistinct. This variability should be described.  Similar to the functionally bileaflet valve and trisinuate aortic root, asymmetry between the anatomical leaflets and sinuses can be present, and should be described. | Functionally unicuspid and trisinuate aortic root, Unicuspid aortic valve, Functionally unicuspid aortic valve with three-sinus root |
|  | **PHENOTYPES** | | | | **IPCCC #** |  |  |  |
| **Unicommissural** | | | | 09.15.28 | There are two zones of fusion between leaflets resulting in a single commissure. | Most commonly there is fusion between the right and non-coronary leaflets and the right and left coronary leaflets, with a zone of apposition between the left and non-coronary leaflets. |  |
| **Acommissural** | | | | 09.15.29 | There are three zones of fusion between leaflets resulting in a central orifice. | This entity theoretically involves some degree of fusion between each of the three leaflets. This rare variant may occur if ventricular myocardium were to support each of the three zones of fusion, as is seen in the more common variant of the functionally unileaflet pulmonary valve. |  |
|  | **ADDITIONAL QUALIFIERS** | | | | **IPCCC #** |  |  |  |
| **Degree of fusion of the zones of fusion:** | | | **Partial fusion** | Q1.16.01 | Fusion of less than half of the zone of apposition. | Each zone of fusion needs to be described. |  |
| **Complete fusion** | Q1.16.02 | Fusion of greater than half of the zone of apposition. | Each zone of fusion needs to be described. When complete fusion is present, this can be inferred and does not need to be specified. |
| **Quadrileaflet Aortic Valve\* with Quadrisinuate Root**  ***\*****the valvar arrangement can be* ***‘Functionally Unileaflet’****,* ***‘Functionally Bileaflet’****,* ***‘Functionally Trileaflet’****,**or* ***‘Quadrileaflet’****,**and should be specified* | 09.15.24 | A congenital cardiovascular malformation where there are four anatomical leaflets within an aortic root with four sinuses. There can be the presence or absence of a zone of fusion between each of the four leaflets, which will dictate how the valve functions, and this should be specified in the term. | | | | | The valve can function as a quadrileaflet, trileaflet, bileaflet, or unileaflet valve, depending on the presence or absence of a zone of fusion between each of the four leaflets. This should be described. For example, when one zone of fusion is present, the valve should be described as a ‘Functionally Trileaflet Aortic Valve with Quadrisinuate Aortic Root’. A descriptive approach towards the quadrileaflet aortic root then becomes paramount.[55]  Similar to the functionally bileaflet or unileaflet valves within a trisinuate aortic root, asymmetry between the anatomical leaflets and sinuses can be present, and should be described. | Cusp can be substituted for leaflet to describe how the valve functions in the Quadrisinuate Aortic Root (i.e. Quadricuspid aortic valve with quadrisinuate root, or Quadricuspid aortic valve with four-sinus root) |
|  | **Phenotypes** | | | | **IPCCC #** |  |  |  |
| An approach using attitudinal description of the aortic leaflets and sinuses is necessary, including description of which leaflets have a zone of fusion present between them, asymmetry or hypoplasia of a leaflet and its sinus, as well as in describing the coronary artery origins.[33] | | | |  |  |  |  |
|  |  | | | | **IPCCC #** |  |  |  |
| **Degree of fusion of the zones of fusion:** | | | **Partial fusion** | Q1.16.01 | Fusion of less than half of the zone of apposition. | Each zone of fusion needs to be described, using an attitudinal approach in describing the leaflets and sinuses involved. |  |
| **Complete fusion** | Q1.16.02 | Fusion of greater than half of the zone of apposition. | Each zone of fusion needs to be described, using an attitudinal approach in describing the leaflets and sinuses involved. When complete fusion is present, this can be inferred and does not need to be specified. |  |

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**Supplemental Table 3.** **Terms and definitions for congenital and acquired abnormalities of the components of the aortic root.**

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
| **TERM** | **IPCCC #** | **DEFINITION** | **COMMENTARY** | **SYNONYMS** |
| **Normal Structures of the Aortic Root and Its Support** | | | | |
| **Congenital and Acquired Aortic Leaflet and Root Abnormalities\*** | | | | |
| **Hypoplasia** |  | Indicating a structure is below the normal size. | Hypoplasia is defined by Z-scores in children, and absolute values or values indexed to body surface area or height in adults. |  |
| **Dilation** |  | Indicating a structure is above the normal size. | Dilation is defined by Z-scores in children, and absolute values or values indexed to body surface area or height in adults. | Dilatation  Effacement |
| **Stenosis** |  | A narrowing or constriction of the diameter of a chamber, valve or artery. | This can be used to describe obstruction as any level: subvalvar, valvar or at the plane of the sinutubular junction, commonly referred to as being supravalvar. | Obstruction |
| **Regurgitation** |  | The backward flow of blood across a closed valve. |  | Incompetency |
| **Raphe** |  | A ridge of tissue or seam located at the zone of fusion between two leaflets. | The ridge of tissue is variably present at the zone of fusion between two leaflets. |  |
| **Zone of Fusion** |  | An abnormal permanent attachment along a variable portion of the ventricular surface of coaptation between two leaflets. | This abnormal fusion can be seen on a spectrum involving 1-100% of the zone of apposition. |  |
| **Leaflet perforation** |  | An abnormal tear or hole within a leaflet. | This commonly results in regurgitation during diastole. |  |
| **Leaflet fenestration** |  | An abnormal aperture or communication within the leaflet commonly located within the surface area of the zone of apposition. | Leaflet fenestration and perforation may be considered synonyms, however, the former is commonly used when located within the lunule of the leaflet. |  |
| **Leaflet elongation** |  | An abnormal increase in length to the leaflet from insertion to free edge of the leaflet. |  |  |
| **Leaflet retraction** |  | An abnormal decrease in length to the leaflet from insertion to free edge of the leaflet. | This is commonly associated with leaflet thickening. |  |
| **Leaflet bending** |  | An abnormal deformation in the leaflet, often its midportion, causing a more acute angle within the body of the leaflet. | This commonly is associated with prolapse of the leaflet. |  |
| **Leaflet prolapse** |  | An abnormal position of the leaflet in diastole where a portion of the leaflet, often its midportion, extends below the plane of the virtual basal ring. | Leaflet billowing is commonly used to describe normal to more marked inferior displacement of the belly of the leaflet, but which does not extend below the plane of the virtual basal ring. |  |
| **Leaflet flail** |  | An abnormal position of the leaflet in diastole where the leaflet midportion and tip extend below the plane of the virtual basal ring. There commonly is abnormal systolic opening as well. | This abnormality is commonly associated with loss of the normal support of a leaflet and sinus. |  |
| **Absent leaflet** |  | An abnormality where a leaflet is not present. |  |  |
| **Accessory leaflet tissue** |  | An abnormal amount of leaflet tissue is present resulting in leaflet elongation. |  |  |
| **Leaflet dysplasia** |  | A general description of leaflet abnormality. | This is a vague term commonly used to describe leaflet thickening. |  |
| **Aortic valvar atresia** | 09.15.06 | Absence of patency of the aortic valvar leaflets with complete fusion between all leaflets and no communication between the left ventricle and aortic root. |  |  |
| **Aortic atresia** | 09.15.03 | Congenital absence or closure of the orifice of the aortic root. |  |  |
| **Subcommissural fusion** |  | An area of tissue fusion within the left ventricular outflow tract on the ventricular surface underneath the commissure, or lateral zone of apposition, between two leaflets. | This subcommissural fusion invariably inhibits the mobility of the involved leaflets. |  |
| **Aorto-ventricular tunnel** | 0.9.17.01 | A congenital cardiovascular malformation of the aortic root in which a channel by-passes the leaflet insertion line of the aortic valve, continuing as an extracavitary passage, and opening into the left or right ventricle. |  | Aortoventricular tunnel; ventriculo-aortic tunnel’ aorto-ventricular tunnel |
| **Aneurysm of aortic sinus of Valsalva** | 0.9.18.01 | A cardiovascular malformation in which there is dilation of one or more aortic sinus of Valsalva. |  | Aortic sinus of Valsalva aneurysm |
| **Ruptured aneurysm of aortic sinus of Valsalva** | 09.18.03 | An abnormal communication between the aortic sinus and another adjacent cardiac chamber related to a tear within the wall of an aneurysm of an aortic sinus of Valsalva. |  | Ruptured aortic sinus of Valsalva aneurysm |

\*terminology for the various types of congenitally malformed aortic roots with abnormal arrangement of the leaflets, sinuses and interleaflet triangles are discussed in Table 2.

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