REVIEW ARTICLE

The Significance of the Interleaflet Triangles in Determining the Morphology of Congenitally Abnormal Aortic Valves: Implications for Noninvasive Imaging and Surgical Management

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A comprehensive understanding of the normal and abnormal aortic root is paramount if we are to improve not only our assessment of the aortic root and its components but also the surgical approach to reconstructing this complex structure when congenitally malformed. Most anatomic and imaging-based classifications of the normal root recognize and describe the basic components, which include the shape and size of the three aortic sinuses and their three valvar leaflets, as well as the sinutubular junction and proximal ascending aorta. However, the three interposing fibrous interleaflet triangles, which share an intimate relationship with all elements of the root, are often ignored. In consequence, the important role the interleaflet triangles play in determining the function of the normal and congenitally malformed aortic root is underappreciated. Additionally, the subtle asymmetries found in the normal aortic root, such as differences between the sizes of the described components, underlie its hemodynamic efficiency. In this review the authors describe the complex structure of the normal aortic root, contrasting these normal characteristics with those found in the unicuspid and bicuspid variants of congenitally malformed aortic valves. Many of these features are readily recognizable using current imaging modalities and so should become a standard part of the description of aortic valvar disease. The authors believe that this thorough morphologic approach will provide a framework for the re-creation of a more normal aortic root at the time of repair or replacement, thereby improving current outcomes. (J Am Soc Echocardiogr 2016;■:■-■.)

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The bicuspid aortic valve is the most common form of congenital heart disease, with a prevalence of 0.5% to 2% in the normal population.[1] Its clinical presentation is highly variable. In its most benign form, the function of the valve can be near normal, and it may be manifest at routine examination only by the presence of an ejection click, with or without an associated systolic ejection murmur. Some degree of stenosis is more common, may range from mild to severe, and is often progressive during childhood, when associated with somatic growth, and/or during adult life when associated with degenerative changes. Its less frequent counterpart, the unicuspid unicommissural aortic valve, has been estimated to have a prevalence of approximately 0.02%. It is encountered in up to one in 20 of those undergoing surgery for isolated aortic stenosis[2,3] but is the most common pattern observed in neonatal critical aortic stenosis. Consequently, it has a marked bimodal age of presentation, either presenting with severe left ventricular outflow obstruction in fetal life or in early infancy[4] or else presenting in the third to fifth decades of life,[5] most commonly with degenerative aortic stenosis or mixed aortic valvar disease.[4,6] Its primary feature is a solitary zone of apposition within the persisting skirt of leaflet tissue. This opening extends leftward and posterior, appearing as an eccentric "teardrop," usually pointing toward the aortic leaflet of the mitral valve. It differs from the solitary zone of apposition found in the bicuspid valve, which spans the aortic root in various directions depending on the site of fusion between the developing leaflets.[4,6-8] In this review, we describe the anatomy of the normal aortic valve and root, taking this as the basis for understanding the distinct anatomic variations that characterize the unicuspid and unicommissural and bicuspid variants. We highlight the important role played by the fibrous interleaflet triangles, long underappreciated by those describing valvar architecture.[9,10] We then emphasize how the morphology of stenotic valves is best ascertained using contemporary imaging techniques and, on the basis of the dimensions and presence of the interleaflet triangles, provide the basis for accurate distinction of the bicuspid and

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unicuspid variants. Finally, we discuss the implications of these findings relative to the available surgical strategies, with the hope that this more detailed approach will improve their outcomes.

EARLY DEVELOPMENTAL CONSIDERATIONS

We still do not know the processes that are responsible for the formation of the arterial valvar sinuses as opposed to the valvar leaflets. We do know, however, that at the stage of formation and sculpting of the leaflets, the developing arterial roots are encased in their entirety within a turret of outflow tract myocardium, which extends distally to the level of the developing sinutubular junction.[11] Given that the valvar leaflets are developed from the outflow cushions, the spaces between them on their ventricular aspects are initially confined by the myocardial wall of the outflow tract. It is also the case that as the subaortic root begins its development, the myocardium of the inner heart curvature interposes between the developing leaflets of the aortic and mitral valves. There is, therefore, continuous regression of the distal level of the myocardial walls of the arterial roots as the valvar sinuses develop to support the valvar leaflets. It is the regression of this myocardial border, with details as yet undetermined, that permits the apexes of the forming fibrous interleaflet triangles to separate the cavity of the left ventricle from the pericardial space. In similar fashion, it is the transformation of the initial myocardial inner heart curvature to fibrous tissue that produces the extensive aortomitral curtain in the roof of the left ventricle. The details of this process also remain fully to be determined.

ANATOMY OF THE NORMAL AORTIC ROOT

The anatomy of the congenitally malformed aortic valve cannot be understood in the absence of detailed knowledge of the structure of the normal aortic root.[9,10] The normal root is composed of the three aortic sinuses, which support the valvar leaflets in semilunar fashion (Figure 1). When these semilunar hinge lines are reconstructed, the valvar leaflets are seen to take the form of a crown or coronet, with a virtual plane, the virtual basal ring, drawn through their basal attachments to represent the enigmatic echocardiographic "annulus" (Figures 2 and 3).[10,12] The fibrous interleaflet triangles have as their base the true ventriculoarterial junction, or the plane of mitral-aortic continuity. They extend distally to the level of the sinutubular junction, are seen on the ventricular aspect of the root, and are bordered by the aortic components of the hinge lines of the aortic valvar leaflets. The free edges of the leaflets form three zones of apposition during diastole, with the level of coaptation formed at no more than half the overall height of the root (Figure 4).[13] The problems in defining the valvar "annulus" have previously been emphasized,[14] with the different approaches used by surgeons, interventional cardiologists, and echocardiographers underscoring potential problems when quantitating the dimensions of the root.[15] The only truly annular, or circular, anatomic structure found within the aortic root is the sinutubular junction. The anatomic ventriculoarterial junction is incomplete in the aortic root, because its posterior part is within the area of fibrous continuity between the leaflets of the aortic and mitral valves. The anterior portion of the junction is crossed by the semilunar hinges of the valvar leaflets and hence is not readily visible when the valve is intact.[9,10,16] The junction, however, is readily revealed by both virtual dissection using, for example, high-resolution computed tomography (Figure 2) or by anatomic dissection (Figure 5).

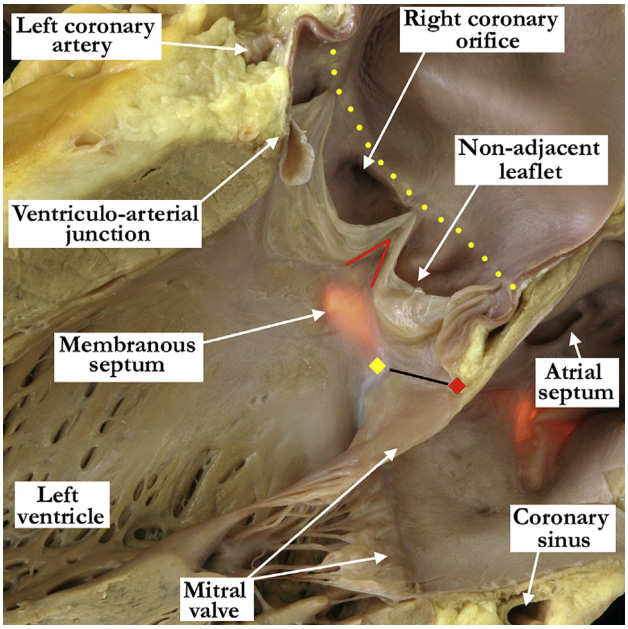

Figure 1 The left atrium, left ventricle, and aortic root are opened in a sagittal plane, with focus on the aortic root anatomy, demonstrating the semilunar lines of attachment of the aortic valve leaflets, extending distally to the sinutubular junction (*yellow dotted line*). The specimen has been transilluminated from the right side to show the location of the membranous septum. The interleaflet triangle between the right and noncoronary leaflets is shown (*red caret*), extending from the space between the ventricular aspect of the semilunar hinges of the valvar leaflets to the sinutubular junction. This interleaflet triangle is in fibrous continuity proximally with the transilluminated membranous septum, which itself is in fibrous continuity with the right fibrous trigone (*yellow diamond*), with these two latter structures together creating the central fibrous body. The area of fibrous continuity between the anterior leaflet of the mitral valve and the entirety of the noncoronary leaflet and portion of the transected left coronary leaflet of the aortic valve is demonstrated (*black line*), resting between the right (*yellow diamond*) and left fibrous trigones (*red diamond*). The right coronary sinus is largely supported by underlying ventricular muscle.

AORTIC ROOT GEOMETRY: ASYMMETRY AND DYNAMIC CHANGES WITH THE CARDIAC CYCLE ARE KEY TO NORMAL FUNCTION

Appreciation of the asymmetry found in the normal aortic root and its supporting structures can provide insight into what produces a competent aortic valve and what surgeons should be aiming to recreate when faced with the challenge of surgery involving the aortic valve and its supporting structures. The most obvious asymmetry is found with regard to the origin of the coronary arteries, with only two of the three sinuses giving rise to a coronary artery (Figures 1, 2, 4 and 5).[9] This feature permits the sinuses to be distinguished as being right and left coronary, as opposed to being noncoronary. The third sinus is usually named "noncoronary" because hardly ever does it give rise to a coronary artery. In the rare circumstances