Supravalvular Arrhythmia
Identifying and Ablating the Substrate

Niloufar Tabatabaei, MD; Samuel J. Asirvatham, MD

Mapping and ablating arrhythmias in the great arteries were rarely in the realm of consideration to treat tachyarrhythmias for the first 2 decades of invasive electrophysiology. More recently, it has become clear that a variety of arrhythmias including ventricular tachycardia, atrial tachycardia, and accessory pathways can be ablated successfully in the great arteries and the cusps of the semilunar valve.

This set of arrhythmias is associated with a unique set of challenges that need to be overcome to obtain high degrees of success and avoid complications.

An understanding of substrate being mapped or ablated is important. Because of the complex relative anatomy of the cardiac valves and great arteries, the electrophysiologist must learn the exact relationship, which is not always intuitive.

Equally important is for the operator to correlate the mapped electrograms with fluoroscopy and ultrasound and to repeatedly establish in his or her mind the neighboring structures where arrhythmia may originate and avoid potentially catastrophic complications that may arise from ablation at these locations.

Important clues with regard to precise catheter location and the nature of the mapped substrate exist from the electrograms being seen at each site, as well as from details of activation mapping and pace mapping performed at each of these locations.

In this article, we will review the regional anatomy of the great arteries and semilunar valves with special emphasis on the arrhythmogenic substrate that may be present at these sites and with regard to avoiding complications as a result of collateral damage. We will then discuss the characteristic electrograms and electrocardiography of the various arrhythmias that are today mapped and ablated above the semilunar valve. Finally, we will provide information on technical details and pitfalls while manipulating catheters and when mapping and delivering energy for ablation at these locations.

Anatomy Relevance to Supravalvar Arrhythmia Ablation

There is perhaps no other set of arrhythmias that requires an exact knowledge of the relative anatomy of the various structures existing in close proximity as this region. The semilunar valve, the overlapping nature of the outflow tract, the valve cusps, proximal conduction system, proximal coronary arteries and their ostia, and coronary veins and the atrial appendages all relate to one another over this relatively small area.

The purpose of studying the anatomy of this region is for the invasive electrophysiologist to be able to clearly picture in his or her mind what structure the mapping catheter exactly makes contact with and, in three dimensions, the surrounding electrically active structures as well as structures that may be damaged by ablating from that site.

Relative Orientation of the Outflow Tracts and the Semilunar Valve

Right and left ventricular outflow tracts (LVOT) are in some ways misnomers. The right ventricle itself is rightward and anterior to the left ventricle. The right ventricular outflow tract (RVOT) courses anterior and leftward off the LVOT such that the pulmonic valve is placed to the left and anterior to the aortic valve. In addition, the pulmonic valve is typically placed 5 to 10 mm cephalad to the aortic valve such that the supravalvar portion of the aorta lies in immediate proximity to the portions of the pulmonary valve. This relationship has important consequences for the electrophysiologist. First, when mapping in the RVOT where the relatively early signals preceding the onset of the premature ventricular contraction (PVC) or ventricular tachycardia (VT) are far-field signals located rightward and posteriorly, then mapping of the LVOT and the supravalvar portion of the aorta should be considered. Second, the posterior wall of the RVOT and the anterior wall of the supravalvar portion of the aorta (right and left coronary cusps) are immediately adjacent to each other and are virtually indistinguishable by surface ECG patterns and require precise mapping to define origin before ablation (Figure 1).

The Central Location of the Aortic Valve

It is imperative to appreciate the important central location of the aortic valve and its relationships (Figure 2). The aortic...
valve comes into contact with the right atrium, left atrium, interatrial septum, RVOT, mitral valve (aorto-mitral continuity), pulmonary valve, tricuspid valve, and conduction system.

The right coronary cusp (RCC) lies immediately posterior to the relatively thick posterior wall of the RVOT. The left coronary cusp (LCC) is related to the posterior wall of the RVOT, which is close to the pulmonic valve but more posteriorly continuous with the anterior leaflet of the mitral valve as the aorto-mitral continuity. The posteriorly direct noncoronary cusp (NCC) lies immediately anterior to the interatrial septum and on either side to the right and left atria.

The Leftward and Anteriorly Located Pulmonic Valve

The pulmonary valve and supravalvar portion of the pulmonary arteries are well established locations of origin for ventricular arrhythmias. Because of its anterior and leftward location, only the posterior and rightward parts of the pulmonary artery have important relations with other cardiac structures. Of the three cusps of the pulmonary valve, the septal or right pulmonary cusp lies at variable distances from and sometimes adjacent to the distal portions of the right atrial appendage. The left pulmonary cusps, being the most superficial, lie immediately beneath the pericardium and have no other cardiac structures related to it. The posterior pulmonary cusps, however, have a very important anatomic relationship that must be understood when ablating in this region; externally it lies in the proximal portion of the left main coronary artery and the distal portions in the left atrial appendage. The supravalvar portion of the aorta lies close to and in some cases adjacent to the junction and surrounding parts of the septal and posterior pulmonary cusps.

Myocardial Extensions Above the Semilunar Valve

In the past, electrophysiologists pictured the semilunar valve to be an abrupt transition point between ventricular myocardium and the great arteries. Anatomists and cardiac surgeons have clearly documented sleeves of syncytial myocardium extending above the semilunar valve and to variable lengths into the great arteries (Figure 3). This situation is akin to what we now know to exist in the pulmonary vein.4,5

The myocardial sleeve typically extends circumferentially around the pulmonic valve between and above all 3 cusps.7 However, with the aortic valve, the myocardial extensions are asymmetrical, with sleeves routinely being found along the
RCC and anterior portions of the LCC.\textsuperscript{7–9} Posterior LCC and NCC, particularly in relation to the anterior leaflet of the mitral valve, are usually devoid of myocardium that is in continuity with the LVOT (Figure 4).

Electrograms Recorded Above the Semilunar Valve: Correlation With Catheter Position, Fluoroscopy, and Ultrasound Imaging

When catheters are placed above the semilunar valve, the exact location of where the recording electrode is located relative to other cardiac structures, coronary arteries, and so forth, is important to know. Understanding the typical recorded electrograms above the valve and correlation with the standard fluoroscopic images, as well as intracardiac ultrasound when used, maximizes success and safety with these procedures.

RCC

The recorded electrogram from the RCC typically shows a large ventricular electrogram. The amplitude of this electrogram is far in excess of what one would expect as a result of activation of the small myocardial sleeve extending above the cusp. The electrogram results from the overlying relatively thick posterior wall of the infundibular/RVOT. Because this tissue is immediately adjacent to the anterior portion of the RCC, the electrogram may appear near field, and the site of early activation can be difficult to distinguish between arrhythmia origination from the myocardium extending above this cusp. The atrial electrogram is typically small and often absent. When large, it signifies close proximity of the right atrial appendage. A variable amount of epicardial fat around the right coronary artery and autonomic tissue separate the appendage from the RCC.

On fluoroscopy, the catheter that has been advanced retrograde when seated in the RCC will point rightward in the left anterior oblique (LAO) projection and anterior in the right anterior oblique\textsuperscript{10} projection. When the fluoroscopic view appears to suggest RCC position but a large atrial electrogram is noted, the catheter should be manipulated with clockwise torque to see whether the atrial electrogram becomes larger. If so, the catheter is likely to be in the NCC and the fluoroscopic images are misleading, perhaps because of atrial enlargement or counterclockwise rotation of the heart as may occur with right ventricular enlargement and chronic obstructive pulmonary disease. Adjunctive intracardiac ultrasound imaging can be particularly useful in identifying mapping and ablation catheter location.\textsuperscript{11} (Figure 5) Longitudinal views are best to appreciate the relationship of the right and left ventricular outflow tracts and for identifying the ostia of the coronary arteries. A cross-sectional view is of most value in identifying the cusps themselves. Once the 3 cusps are visualized, the interatrial septum will be seen in immediate relationship to 1 of the cusps; this defines the noncoronary cusp rightward and anterior to the noncoronary cusp immediately behind the RVOT with the right coronary cusp. Another landmark for the RCC is that the septal and anterior leaflets of the tricuspid valve will meet at the junction between the NCC and RCC. The LCC is identified as being leftward and anterior to the NCC and in close relation to the anterior leaflet of the mitral valve.

LCC

Electrograms obtained in the LCC are the most variable of the aortic cusps (Figure 6). More anteriorly close to the commissure with the RCC, a large ventricular electrogram, again originating in the posterior RVOT, can be recorded. More leftward and posteriorly, however, because of the aortic-mitral continuity, recorded ventricular electrograms are small and far field in nature. In the RAO fluoroscopic image, as the catheter is rotated counterclockwise in the LCC, a relatively larger but usually far-field atrial electrogram is located representing atrial activation close to the anteroseptal mitral annulus. Occasionally, mitral annular atrial tachycardias may be best mapped or ablated in this location (Figure 7).

\textbf{Figure 4.} Endocardial view of the junction between the LVOT and aorta. Note the RCC (R) inserts onto relatively thick ventricular myocardium and the myocardial sleeves extend above the valve and between this cusp and the NCC (P). Arrows point to the aortic mitral continuity between the mitral valve and the left/noncoronary cusps. Note the absence of myocardial extensions in this region. RC indicates right coronary artery; LC, left main coronary artery; N, nodes of Ranvier; L, left coronary cusp; LV, left ventricle; STJ, sino-tubular junction.

\textbf{Figure 5.} Intracardiac ultrasound image showing the cross-sectional view of the aortic valve with the ablation catheter in the LCC. His indicates His catheter; Abl, Ablation catheter.
NCC

The largest atrial electrograms are recorded in the NCC. Although it is unclear at present whether myocardium, either atrial or ventricular, actually extends above and into the NCC, the electrogram findings are consistent. The reason for this is the immediate proximity of the thick myocardium of the interatrial septum posterior to the NCC in its midportion. A ventricular signal may or may not be present. In the depths of the cusp, often a far-field ventricular electrogram will be seen probably representing far-field activation of the LVOT. In the RAO projection, the NCC, when entered, will be posterior. In the LAO projection, however, the NCC can be very difficult to distinguish from the LCC because both appear to be somewhat leftward (Figure 8). When counterclockwise torque is applied and a large near-field atrial electrogram is seen, the operator can be certain that the catheter is in the NCC. Intracardiac ultrasound may also help to clarify the situation. The probe should be placed close to the tricuspid annulus and directed leftward. If the catheter tip is located in a cusp and the interatrial septum can be visualized in the same plane, NCC location can be determined. If the tricuspid valve anterior leaflet is visualized along with the RVOT, then the catheter tip is probably in the RCC and simply directed posteriorly. Again, a simple maneuver of counterclockwise torque on the catheter while visualizing fluoroscopically and with ultrasound will result in a large ventricular electrogram being recorded and thus identifying catheter location in the RCC.

His Bundle Electrogram

The His bundle electrogram can be obtained at several supravalvar locations including the RCC posteriorly, the NCC anteriorly, and when the electrode has been prolapsed below the plane of the semilunar valve in between these 2 cusps. The intracardiac ultrasound location is characteristic
Significance of Near-Field and Far-Field Signals With Supravalvar Arrhythmia Mapping

Above the pulmonic valve and portions of the aortic valve, the ventricular electrogram itself may be of a multicomponent in nature. Both near-field and far-field signals are sometimes seen when mapping this region in patients with PVCs or VT. In sinus rhythm, a delay in activation into the great artery across the valve annulus results in an electrogram similar to that observed in the pulmonary veins. In sinus rhythm, the first component of this double potential is far field and represents ventricular outflow tract activation. The isoelectric period that follows is probably a result of delay across the annulus and then the sharp near-field potential activation of the myocardial supravalvar sleeve. During arrhythmia (PVCs or VTs), if a similar sequence of far-field to near-field activation is noted, this signifies that the arrhythmia is not supravalvar in origin, and there is passive activation of the supravalvar myocardium. On the other hand, if there is a reversal in this pattern of electrograms (far-field, near-field reversal) with a sharp near-field potential occurring first, then this is diagnostic of supravalvar arrhythmia origin. It should be noted, however, that this reversal does not signify that the catheter site is the origin of arrhythmia but only that the supravalvar myocardium (distal to the site of delay) is the substrate. Further mapping above the valve may be required to find the true earliest near-field electrogram representing the site of origin of an automatic tachycardia (see below).

Supravalvar Ventricular Tachycardia

Anatomic Basis for the Substrate Ablated

Ventricular arrhythmias including VT and PVCs have been successfully ablated above the aorta and pulmonary valve.2,16,17 Once the possibility of these arrhythmias has been recognized and specific mapping above the valves undertaken, ablation is performed with a high degree of success and relatively low complication rate.17–19 Despite these advances, the nature of the actual substrate being ablated has not been clearly defined. For example, the ablation electrode placed in the depth of the RCC may be mapping and ablating a focus arising from the supravalvar extension into the cusp, LVOT myocardium, or a deeper posterior RVOT myocardium. When mapping, exact identification of the substrate is warranted because relatively lower energy ablation, thus limiting the chance of collateral damage, can be obtained when ablating at the true arrhythmogenic site. A combination of electrogram analysis (near field versus far field), early electrogram localization, and completely negative unipolar recordings along with integration of anatomic, fluoroscopic, and ultrasound images combine to identify the best-defined target.

ECG-Anatomy Correlation

Several algorithms and templates of electrocardiograms attempting to correlate location of origin of supravalvar ventricular tachycardia have been suggested.11,17 Because there is significant variation in the regional anatomy between individual patients with and those without structural heart disease, the electrophysiologist can have a broader picture of recorded electrocardiograms to consistently, albeit approximately, assess site of origin, based on the ECG.19

The ECG during RVOT tachycardia is characterized by a left bundle-branch block inferior axis pattern. Often noted, but unlike with infravalvar RVOT location for arrhythmia origin, is the presence of a small R wave in lead V1. Because lead V1 is a right anterior lead, a small R wave signifies either a left or posterior location for the focus. As described above, the pulmonary valve lies to the left of the body. Thus, a suprapulmonary valve tachycardia origin will have a small but definite vector toward the right and thus lead V1, resulting in a small initial R wave. In this instance, lead I will be negative and also reflective of the leftward origin and lead III will be more positive than lead II.

The second instance in which an R wave is produced in lead V1 is posterior origin for the arrhythmia. An accurate understanding of anteroposterior section of anatomy of the outflow tract will help the electrophysiologist correlate the height of the R wave with the site of the focus.16–20 (Figure 9) Ventricular tachycardia arising from the anterior RVOT will have no R waves in V1 (classic left bundle-branch block pattern). The next most posterior electrically active structure is the posterior wall of the RVOT, and immediately behind this the anterior LVOT and RCC, and both will produce a small R wave in lead V1. Distinguishing between these two further locations from the ECG alone is difficult, and simultaneous or sequential mapping of the posterior RVOT as well as the RCC must be done for exact diagnosis. As one proceeds more posteriorly, tachycardia origin in the posterior LCC, aortic mitral continuity, and so forth, will produce a significantly larger R wave in lead V1. When the focus is on the posterior mitral annulus, distinct right bundle-branch block morphology will be seen in lead V1. Leftward epicardial RVOT below the level of the pulmonary valve occurring within or in the region of the anterior interventricular vein will produce a pattern similar to origin in the neighboring supravalvular valve and LCC myocardium. Simultaneous mapping with careful analysis of the earliest near-field electrogram occurrence is required to find the true site of origin to define where ablation energy delivery should be delivered. These guidelines correlating anatomy and the ECG during arrhythmia have important limitations that should be considered for application in clinical scenarios. First, variation in body habitus and exact electrode placement can significantly alter the relative amplitude of the R and S waves in lead V1. Second, even within a cusp, for example, in the left coronary cusp, relatively anterior (junctional with RCC) foci may have a smaller R wave (similar to what Figure 8 shows), whereas more posteriorly located foci (junction of NCC or aortic mitral continuity) may have relatively larger R waves. These issues may explain the slight differences and published reports on correlating paced mapped R waves and cuspal origin.11,17

Activation Mapping

The majority of outflow tract arrhythmias including supravalvar arrhythmia result from either triggered or abnormal
Automaticity. Activation mapping is generally straightforward, with point-to-point mapping of the outflow tracts and supravalvar region. The situation, however, is it is not always straightforward, and a few causes of difficulty should be kept in mind (Figure 10).

- Mapping of the posterior portion of the RVOT can be difficult. This is because a catheter placed via the femoral vein into the right ventricle is torqued clockwise to enter the RVOT. This natural maneuver tends to take the mapping electrodes toward the anterior wall of the RVOT. To map the posterior wall, either a guiding sheath must be placed in the RV or the catheter must be advanced while it is curved and torqued clockwise to obtain contact on the posterior RVOT without falling back into the right ventricle.

- Because of the above-mentioned difficulty, often the earliest site of activation for the posterior RVOT is found when mapping the anterior LVOT, specifically the supravalvar region of the RCC.

- When mapping above the valve, in addition to noting the actual timing of activation, whether reversal of near- and far-field potentials occurs should be watched for (see above).

- When the LCC site appears early but ablation is unsuccessful, the posterior supravalvar pulmonary artery region and the anterior interventricular ring and branches should be mapped.

- Sometimes the left atrial appendage, which lies close to the posterior pulmonary artery laterally, can be mapped and in rare instances ablated for supravalvar tachycardia. Similarly, the superior vena cava medially lies close to the supravalvar portions of the ascending aorta above the RCC.

- In performing electroanatomic mapping, because of the overlapping nature of the outflow tract and supravalvar region, when 2 potentials are seen, only the near-field potential should be used for timing, otherwise the same timing will be noted at disparate sites: first where the near-field potential was used and the second where the corresponding far-field potential was used (Figure 11). This will inappropriately give the appearance of a large area of simultaneous early activation.

Pace Mapping

Pace mapping where relatively low output pacing above capture threshold is performed at various outflow tract locations defined the best 12-lead ECG “match” with the clinical arrhythmia can be applied for supravalvar arrhythmia as well. Some potential causes of difficulty largely based on anatomy should, however, be kept in mind.

- Once again, because of the close anatomic proximity of the various structures in the supravalvar outflow tract region, high output pacing, for example, from the RCC, may reproduce the ECG or clinical arrhythmia even when the origin is in the posterior RVOT or subvalvar anterior LVOT.

- Similarly, high-output pace mapping from the anterior intraventricular vein proximally, LCC, and the aortic mitral continuity region will be similar.

- Remnants of the primitive atrioventricular conduction system are sometimes found in the outflow tract. These structures may or may not connect to the remaining infra-Hisian conduction system. Their presence, however, may produce a difference in pace-mapped ECG morphology with low or high output pacing. High output pacing from a location close to one of these tracts may give rise to both myocardial and “fascicular” capture, whereas low output pacing may capture only the fascicle exiting the myocardium elsewhere. Thus, pacing at high and low output may be required before making a judgment on the match with clinical arrhythmia.
Supravalvar arrhythmia origin may exit to the ventricular myocardium below the valve. Thus, pace mapping above the valve and at the exit site may be similar.

Pace mapping can be difficult to perform above the semilunar valves. Deep in the cusp where there is a clear sleeve of myocardium, particularly the RCC or the pulmonary valve cusps capture can usually be obtained at typical outlets. In the NCC, simultaneous atrial capture is the rule, and at lower outputs atrial capture without ventricular capture is often seen even when the focus of origin is above the NCC. Keeping these caveats based on anatomy in mind and carefully correlating with activation mapping will avoid ablation at an incorrect site.

Safety
Understanding the anatomic complexity of the region does help maximize ablation success but perhaps more importantly, allows the electrophysiologist to be cognizant of the potential for complication with inadvertent damage to the neighboring structures from mapping and ablation.18,25,26

- Damage to the coronary arteries. Because the left main coronary artery arises cephalad to the LCC and the pulmonary valve is also cephalad and leftward of the aortic valve, immediate posterior relation of the supravalvar pulmonary artery would be the proximal left main coronary artery. Thus, when ablating above the pulmonary valve posteriorly or just below the pulmonary valve in the posterior RVOT, either intracardiac ultrasound imaging or coronary angiography should be performed before ablation energy delivery.
- Conduction tissue damage. As noted above, a constant location of the penetrating bundle of His is at the membranous septum formed in part by the commissure between the RCC and NCC of the aortic valve. Ablation at this site could potentially damage the bundle of His. Ablation below the plane of the aortic valve but with the catheter brought between these 2 cusps will also record a His bundle electrogram, and this structure may be ablated during VT, in which the His bundle electrogram may be occurring within the ventricular electrogram itself (retrograde activation to the His).

Supravalvar Atrial Tachycardias
Atrial tachycardias have been successfully ablated above the semilunar valve.27–29 Given the proximity of the semilunar valve to the ventricular outflow tract myocardium, it is easy to understand why ventricular tachycardia can be ablated above the valve. To appreciate the reason why atrial tachycardia can also be ablated above the valve, a careful review of the anatomy of the aortic annulus and valve cusps described above is required. Although it is presently not known why atrial myocardium may exist in the coronary cusps, the intimate anatomic relationship of the NCC with the interatrial septum and atria is indisputable.

Tachycardias that arise on the septal tricuspid annulus within the interatrial septum and the anteroseptal mitral annulus all occur very close to the location of the NCC.30 Because of this, the best vantage point to map and ablate these arrhythmias with stable catheter position may be in the NCC.26,28

Atrial tachycardias arising above the pulmonary valve have not been described; however, the left atrial appendage may be a vantage point for mapping or in rare instances ablating epicardial/leftward tachycardial origin in the RVOT in the region of the pulmonary valve.

Occasionally, atrial tachycardia or tachycardia beats initiating atrial fibrillation may be mapped when the rightward portion of the ascending aorta is above the RCC. Simultaneous mapping of
the adjacent septal superior vena cava above the right atrial junction is required to determine the true site of origin.

**Activation Mapping**

A supravalvar origin for atrial tachycardia should be suspected when near simultaneous but not necessarily very early activation is noted at several anatomically separate locations (Figure 12). For example, when mapping the atrial tachycardia sites either simultaneous with P-wave onset or 5 to 10 ms earlier may be noted in the fast pathway region, Bachmann bundle region, His bundle region, roof of the coronary sinus, and in the anteroseptal mitral annulus. As described previously, all of these locations skirt around the NCC of the aortic valve. When such simultaneous activation is noted, direct mapping of the NCC is required. Even when tachycardia origin is above the valve, not all sites in the NCC may be early because the NCC served as a vantage point to specific sites on the interatrial septum and right and left atria. One location may be distinctly earlier than the other. Thus, the entire NCC must be systematically mapped in much the same way that the right and left atria are mapped for nonvalvar atrial tachyarrhythmias.31

**Safety**

In addition to the precautions described previously, to avoid perforation of the valve cusps, care should be exercised to avoid conduction system damage. Mapping at the junction of the NCC and RCC may reveal a His bundle electrogram, and ablation at the site may damage the structure. Because the depth of the NCC is more caudal and posterior to the His bundle location, the fast pathway input to the AV node may be damaged when ablating in the NCC with the catheter electrode pointing rightward. To avoid this, either the catheter tip should point straight posterior or the catheter moved slightly more cephalad. In moving cephalad, particularly if counterclockwise torque is applied, care must be taken to avoid entering the right coronary artery. The compact AV node is generally not accessible from the NCC, but with cardiac pathology or if perforation of the valve occurs during ablation AV nodal, damage is possible.

**Supravalvar Accessory Pathway Mapping and Ablation**

Accessory pathways are usually myocardial “bypasses” connecting atrial and ventricular myocardium across the tricuspid or mitral annulus. Although such bypass tracts can occur.

---

Figure 11. Electroanatomic map images of patient whose ECG and intracardiac electrograms are shown in Figure 9. Note the earliest site of activation as seen in an LAO orientation (A) is leftward but importantly very posterior in the PA view (B). This simultaneously relative, leftward, and posterior origin correlates anatomically with the significant initial R wave seen in lead V1 (see text for details).
anywhere along the annulus, the anterior and anteroseptal region, the mitral annulus is typically thought devoid of such connections. This is because of the location of the aortic valve and the part of the cardiac skeleton that forms the aortic mitral continuity.32

There are very rare types of bypass tract previously referred to as trigone pathways.33 These were putative connections that traverse the central fibrous body and the trigone connecting either atria to either ventricle but usually the anterior annular left ventricle.

More recently, successful ablation of accessory pathways in the aortic cusps has been demonstrated.10,34 The exact substrate being ablated in these cases is not clear. In some instances, a distinct pathway potential has been demonstrated suggesting that myocardium above the valve constitutes the accessory pathway. As mentioned previously, myocardium that continues with the ventricle has clearly been demonstrated anatomically,7 whereas the successful ablation of the atrial tachycardia from the aortic cusps suggests the possibility of atrial myocardium being present as well. It is therefore conceivable that bridges or connections between this atrial and ventricular myocardium could occur in some patients, constituting an accessory pathway. Also possible is that the pathways do in fact cross the fibrous trigone, but the best anatomic vantage point for catheter positioning and delivery of ablative energy is from the cusps.

Mapping of Supravalvar Accessory Pathways

Because of the central portion of the aortic valve, supravalvar accessory pathways could involve atrial myocardium of either the right or left atria and the ventricular myocardium of either in the region of the aortic mitral continuity or the LVOT. As such, suspicion for these pathways should arise when:

- a right anteroseptal pathway is suspected but ablation is unsuccessful;
- multiple sites of activation along the septum of either the right or left atria and in the case of antegrade preexcitation the right or left ventricular outflow tracts appearing to be activated early;
- a left anterior or left anteroseptal bypass tract is being considered;
- findings are consistent with a right or mid septal or anteroseptal bypass tract but a distinct pathway potential cannot be demonstrated; or
- a long pathway course in the septal region is suggested by either decremental conduction properties or slow conduction.

In addition to mapping along the annular, specific retrograde mapping of the cusp should be undertaken.10 Pacing mane-
vers to dissociate the ventricle and the atrium sequentially can be performed to determine whether the electrograms found in the aortic cusps represent sites of early atrial or ventricular activation, respectively, or the pathway potential itself.

Safety
In addition to the general issue of safety when ablating above the valve including coronary arterial damage and valvular damage mentioned above, specific attention to the conduction system must be made when ablating supravalvar accessory pathways.

Because multiple early sites of atrial activation may be found when mapping retrograde conduction and the operator may have already mapped 1 early site on the right anterosep-toral region near the fast pathway or compact AV node before specific supravalvar mapping, meticulous attention to which near-field electrogram truly represents the earliest site of activation must be made. Cryoablation can be considered when concern exists.34

Supravalvar ablation would not be expected to damage the compact AV node because of its midseptal location and the relatively superior location of the aortic annulus to the tricuspid valve (except for inadvertent cuff perforation during ablation). However, modulation of the fast pathway is probably from an anatomic perspective when ablating the depth of the NCC at its junction with an RCC. Such fast pathway modulation or ablation could potentially eliminate high septal atrial tachycardia as well as AV node reentry, but, as with other attempts at fast pathway ablation, would be less safe and more likely to result in AV nodal conduction abnormality than ablating the slow pathway in the region of the coronary sinus floor. Whether a role for supravalvar ablation exists for unusual variants of AV node reentry is not presently known.

Summary
Supravalvar ablation has now been well documented to be the ideal mode for ablating specific forms of ventricular tachycardia, atrial tachycardia, and accessory pathways. A studied appreciation of the anatomy of the supravalvar region is a prerequisite for electrophysiologists to safely and effect-ively approach these arrhythmias. In addition, the consistent ability to correlate the recorded electrograms with fluoroscopic anatomy and intracardiac ultrasound images enhances the chance of successful elimination of supravalvar arrhythmias.

Disclosures
None.

References


Key Words: aortic valve ■ outflow tract ■ pulmonary arteries ■ pulmonary valve ■ radiofrequency ablation
Supravalvular Arrhythmia: Identifying and Ablating the Substrate
Niloufar Tabatabaei and Samuel J. Asirvatham

Circ Arrhythm Electrophysiol. 2009;2:316-326
doi: 10.1161/CIRCEP.108.847962
Circulation: Arrhythmia and Electrophysiology is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2009 American Heart Association, Inc. All rights reserved.
Print ISSN: 1941-3149. Online ISSN: 1941-3084

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circep.ahajournals.org/content/2/3/316

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation: Arrhythmia and Electrophysiology can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation: Arrhythmia and Electrophysiology is online at:
http://circep.ahajournals.org/subscriptions/