PATHOGENESIS AND MANAGEMENT OF TACHYCARDIA ASSOCIATED WITH WOLFF-PARKINSON-WHITE SYNDROME

A Review Article

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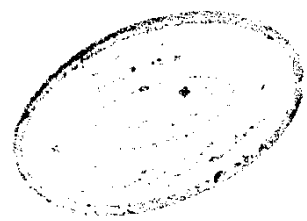

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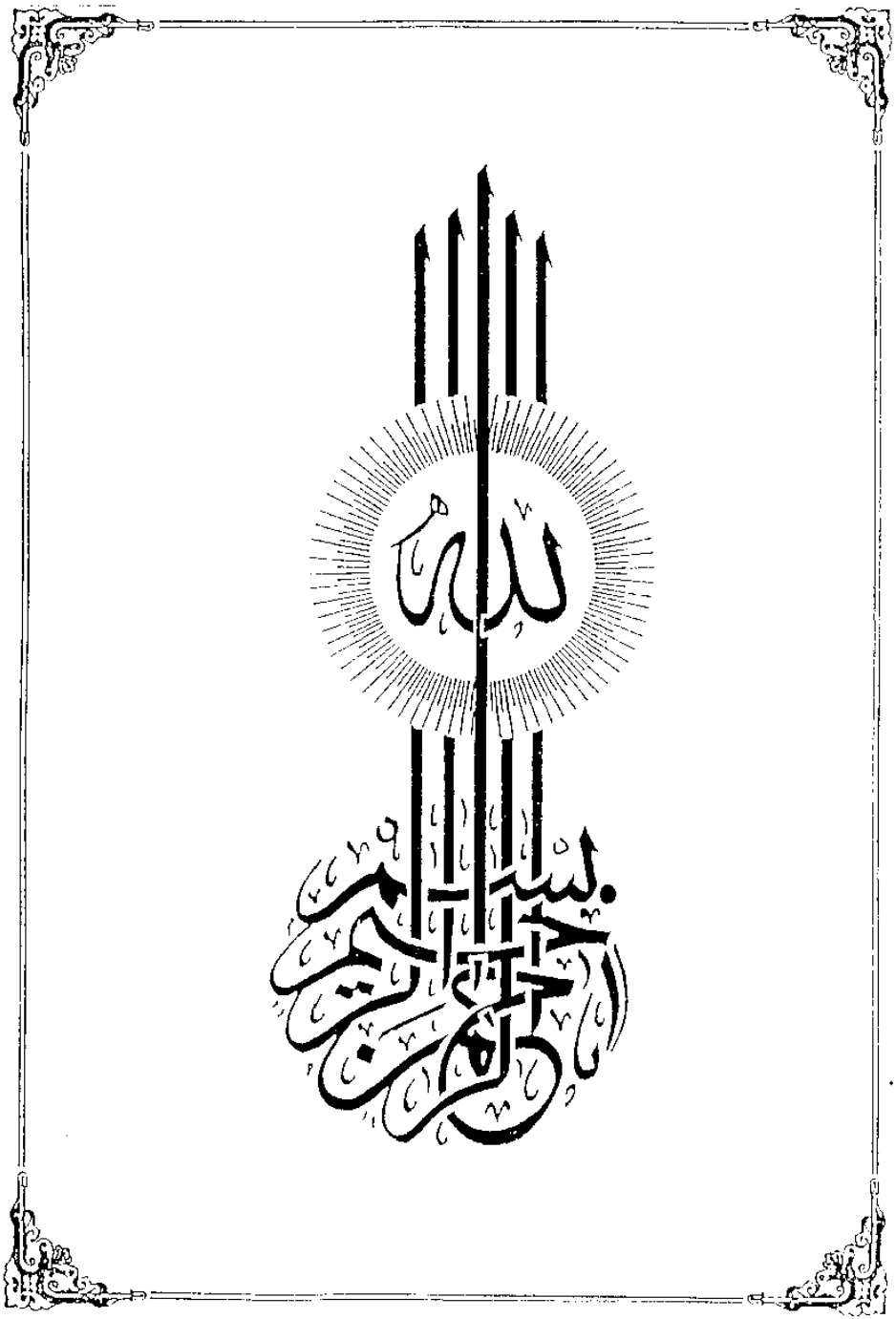

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The Sinus Node:

The most significant feature of the morphology of the sinus node is its position. The node is a small spindle shaped structure, it is 10-20 mm long, 2-3 mm wide roughly flattened ellipse and it does not have a head or tail as commonly alleged. It is usually set around the prominent nodal artery. As *Treux, (1976)* has indicated, the precise shape of the node seen in sections varies depending on the plane of section employed, In section at right angles to the sulcus terminalis the node forms a wedge set into the junction of the wall of SVC with the crista terminalis. The sinus node lies less than 1 mm from the epicardial surface within the terminal sulcus lateral to the crest of the right atrial appendage, near the junction of the SVC and right atrium with its anterior margin very near the crest formed by the junction of atrial appendage with SVC. *(Hudson, 1960).* Since this feature is of most significance to surgeons, Fig. 1, is oriented so as to display the position of the node as it would be seen at operation. Although the node is most usually in the lateral position shown in Fig. 1, on occasions it may extend in a horse shoe fashion across the crest of the appendage so that its head becomes a medial limb which continues into the interatrial groove *(Anderson et al., 1979).*

The lateral position of the node has been substantiated by most subsequent investigators. *Anderson and Becker, (1980)* confirmed the opinion of those who observed the node in lateral position. Frequently the tail of the node is also extensive, running down the terminal sulcus towards the mouth of the inferior vena cava before burrowing into and merging with the musculature of the terminal crest. In most cases the sinus node is arranged around a prominent nodal artery, considerably larger than needed for mere nutrition of the node. This feature prompted *James, (1973)* to suggest a servomechanism function, i.e. drugs and procedures that accelerate the sinus node may decrease the calibre

of the artery while those which slow the node may be associated with increase calibre of the artery. The nodal artery is a branch of the right coronary in 55% of cases and left circumflex artery in 45%, the artery can then run clockwise or counter clockwise around the superior cavo atrial junction *(James, 1961)*. Certainly in animals such as the rat, ultrastructural studies have shown a particular specialization of the adventitial coat of the artery, which is composed of nodal cells, *(Taylor, 1980)*. However if it exists the servo mechanism function is not universal because there is considerable variation in the humans in the pattern the artery takes as it courses through the node. In a study done by *Anderson and Becker (1980)* it was found that in only 10 of 20 nodes did the artery run completely through the nodal substance in the other hearts it either ramified within the nodal substance (6 cases), run an eccentric course through the node (2 cases) or was not present as a prominent artery (2 cases). It is difficult to reconcile these findings with the contention that the sino atrial node and its artery function as a servomechanism. *James (1968)* has pointed to the significance of left main coronary arterial disease in obstructing the nodal artery and resulting in atrial arrhythmia following acute myocardial infarction. However others have denied any positive correlation between the main coronary atherosclerosis and impaired sinus node function *(Engel et al., 1975)*. Using sinoatrial conduction time as an indicator for sinus node function. *Jordan et al., (1977)* found that coronary artery disease does deleteriously affect sinus node performance. There are numerous nerve endings but no ganglia within the sinus node. At the anterior and posterior margins of the sinus node, however there are many ganglia. The autonomic neural input to the heart exhibits sidedness, with the right sympathetic and vagal nerves affecting the sinus node more than the AV node while the left sympathetic and vagal nerves affecting the AV node more than the sinus node *(Randall, 1977)*.

Sympathetic stimulation of the right side leads to sinus tachycardia, while stimulation of the left stellate ganglion produces a shift in the sinus pacemaker to ectopic site and shortens AV nodal conduction time and refractoriness, but inconsistently speeds

the sinus nodal discharge rate *(Randall, 1977, Levy, and Martin, 1979)*. Stimulation of the right vagus primarily slows the sinus nodal discharge rate and stimulation of the left vagus prolongs the AV nodal conduction time and refractoriness.

The Specialized Atrioventricular Junctional Area (Fig. 2)

The term atrioventricular junctional area describes the area of specialized conduction tissue forming the connection between the atrial and ventricular working myocardium *(Becker et al., 1976)*. The function of this area is to induce atrioventricular delay so that the ventricles fill with blood during diastole, systole then being synchronized by the rapid distribution of the cardiac impulse through the ventricular conduction system. It can be subdivided into: the transitional cell zone or nodal approaches *(Hecht, 1973)*, compact zone or the AV node itself and the penetrating part of the AV bundle (His bundle) which continues as a non branching portion of the AV bundle. Some investigators consider the branching portion of the AV bundle (i.e. the bundle branches) to be part of the AV junctional area anatomically *(Becker and Anderson, 1976)*, while others *(Hecht, 1973)*, relying more on electrophysiological function, separating the branching from non branching portion. Trauma to any part of the junctional area can produce atrioventricular dissociation. In terms of functional correlation, it is still not known with certainty which part of the specialized junctional tissue induces nodal delay. The most obvious is the compact zone, made up of interweaving fasciculi of small nodal cells, but in the rabbit node , correlative studies showed that most of the delay was induced in the transitional cell zone *(Anderson et al., 1974)*. The human heart has extensive transitional areas between the compact zone and the posterior right and left surfaces of the atrial septum, *(Becker and Anderson, 1976)*. It is highly probable therefore that some delay is induced within these zones.

Transitional Cell Zone or Nodal Approaches

In the rabbit AV node these are located in posterior superficial and deep group of cells. They differ histologically from atrial myocardium and connect the latter with the compact zone of the AV node. Some fibers passing from the posterior internodal tract to the distal portion of the AV node or His bundle that may provide anatomical substrate for a bypass tract. *(James, 1961)*. However the anatomical and functional importance of this structure is unclear *(Jackman et al., 1983)*.

A.V. Node

The compact portion of the AV node is a superficial structure lying beneath the right atrial endocardium anterior to the ostium of the coronary sinus and directly above the insertion of the septal leaflet of tricuspid valve. It is at the apex of a triangle formed by the tricuspid annulus and tendon of Todaro which originates in the central fibrous body and passes posteriorly through the atrial septum to continue with the Eustachian valve. The compact portion of the AV node is divided from and becomes the penetrating portion of the His bundle at the point where it enters the central fibrous body *(Zipes, 1984)*. In 85% to 90% of human hearts. the arterial supply to the AV node is a branch of the right coronary artery that originates at the posterior intersection of the AV and interventricular groove (Crux). A branch of the circumflex coronary artery provides the AV nodal artery in the remaining hearts *(Zipes, 1984)*. Internally the AV node exhibits both similarities and differences from the sinus node. Like those of the sinus node, slender fibers of the AV node interweave to form a meshwork but there is much less collagen between fibers. The fibers of the AV node are slightly thicker and shorter than those of the sinus node but not as thick as those of ordinary myocardium. Through the upper and middle portions of the AV node the fibers frequently connect with one another and interweave at random, but in the anterior and inferior portions of the node they

begin to orient into a longitudinal axis as they form the AV bundle. Behind the AV node is the very small area between it and the coronary sinus, there is a number of autonomic ganglia . These are vagal ganglia, having a receptor function *(James, 1968)* as well as being the probable route by which vagal stimuli of extracardiac origin arrive the AV node.

Internodal and Interatrial connecting Pathways

There are 3 potential pathways between the sinus node and the AV node connecting the impulse from sinus node to AV node more rapidly than that occur in ordinary myocardium. They correspond in part to areas previously described by *Bachmann (1916).* The pathways originally were described by *Bachmann* on the basis of both physiologic and anatomic studies concerned primarily on interatrial conduction and he made no mention of its function in internodal conduction *(Zipes, 1984).*

The anterior internodal pathways

Extends from the anterior margin of the sinus node curves around the superior vena cava and enters the anterior interatrial myocardial band *(Bachmann's bundle).* Fibers in Bachmann's bundle divide near the anterior margin of interatrial septum, some continuing into the left atrium and others descending obliquely and posteriorly within the interatrial septum behind the non coronary sinus of the aorta to enter the upper margin of the AV node. This pathway composed of both ordinary and Purkinje fibers.

The middle internodal pathway

These fibers leave the superior and posterior margins of the sinus node to curve behind the superior vena cava and cross the sinus intercavarum to the crest of the interatrial septum. There, they divide into a few sparse fibers continuing into the left atrium and a much larger number which descends within the interatrial septum to enter

the top of the AV node. The connections with the left atrium by this route are inconstant and suggesting that interatrial conduction by this route is important only in exceptional hearts.

The posterior internodal pathway

It was emphasized by *Soderstrom, (1948)* as a major route for internodal conduction. These fibers leave the posterior margin of the sinus node and continue along the crista terminalis to the eustachian ridge in which they sweep into the interatrial septum directly above the posterior margin of the AV node. There, they curve down to enter the node. Like the anterior internodal pathway this pathway is fairly constant especially in hearts with prominent eustachian ridge. The route of conduction in this posterior internodal pathways is similar in humans and the dogs. As fibers of all three internodal pathways approach the AV node, some merge with one another while others enter the node directly, which is at the anterior margin of fossa ovalis. Some fibers of all three pathways bypass the superior margin of the AV node (its crest) and enter the node at various points along its convex surface. The interatrial conduction may occur along any one of the three internodal pathways, but under normal circumstances in most hearts it is probably through the Bachmann's bundle, beginning along the anterior internodal pathways. The middle internodal pathway fibers do not carry an importance in interatrial conduction and the posterior internodal pathway fibers participate little if all in normal interatrial conduction. During ectopic rhythm originating in the AV node, however its septal connections to the left atrium and the posterior internodal pathway become more important routes of spread to both atria. Conduction to the right atrium from the sinus node is normally direct, radiating from the inferior margin of the node into the adjacent trabeculae of the free wall of the right atrium and extending into the crista terminalis, from which more distal radiation may occur. Many fibers in both atria exhibit Purkinje characteristics, particularly in the regions of the internodal and interatrial pathways.

Although it may reasonably be presumed that normal spread of excitation is roughly radial from the sinus node into the right atrium and through Bachmann's bundle to the left atrium, the normal (usual) pathway for internodal conduction is not so apparent, for example one internodal pathway may function selectively under normal conditions with the others providing alternate routes in the case of disease. On basis of anatomic length alone, the anterior internodal pathway is the shortest internodal pathway and under normal circumstances of sinus rhythm is probably the preferential one *(James and Sherf, 1971, Sherf and James, 1972).*

Anderson and Becker, (1980) reported that there are no histologically discrete tracts of specialized conduction tissue extending between the sinus and AV nodes. This tissue is best referred to as the internodal atrial myocardium. Nonetheless, it must be emphasized that although *Anderson and Becker, (1980)* were unable to identify histologically "specialized internodal tracts" this does not in itself rule out either preferential conduction of the cardiac impulse through the atrial myocardium or the possibility that cells with different electrophysiologic properties exist within the myocardium. The existence of preferential conduction is well established, but its pattern can be accounted for simply by the geometric arrangement of the muscle bundles of the right atrium.

The Bundle of His or Penetrating Portion of the AV Bundle

It is formed by convergence of fibers at the anterior and inferior margin of the AV node, the parallel fibers of the AV (His) bundle veerfrom the right atrial endocardial location of the AV node into the middle of the central fibrous body, continuing through the annulus fibrosis where it is called the non branching portion as it penetrates the membranous septum. The fibers of the AV bundle are separated by fine collagen septa *(James and Sherf., 1971).* Proximal cells of the penetrating portion are heterogenous resembling those of the compact AV node, while distal cells are similar to cells in the

proximal bundle branches. Connective tissue of the central fibrous body and membranous septum encloses the penetrating portion of the AV bundle which may send extensions into the central fibrous body *(James et al., 1982)*. The AV bundle descends along the posterior margin of the membranous interventricular septum to the crest of the muscular septum. The AV bundle is often triangular in cross section and the two lower corners of the triangle give rise to the right and left bundle branches.

The Bundle Branches or Branching Portion of the AV Bundle

These structures begin at the superior margin of the muscular interventricular septum (from the two lower corners of the triangle representing the AV bundle) immediately beneath the membranous septum, with the cells of the left bundle branch cascading downwards as a continuous sheet on to the septum beneath the noncoronary aortic cusp. The AV bundle then may give off other left bundle branches, sometimes constituting a true bifascicular system with an anterosuperior branch, in other hearts giving rise to a group of central fibers, and in still others appearing more as a network without a clear division into a fascicular system *(Kulbertus et al., 1976)*. The right bundle branch continues intramyocardially as an unbranched extension of the AV bundle down the right side of the interventricular septum. In some hearts the right bundle branch forms an obtuse angle with the His bundle *(Massing et al., 1976)*. It generally remains unbranched to the apex of the right ventricle and base of the anterior papillary muscle. In some human hearts the His bundle traverses the right interventricular crest giving rise to a right sided narrow stem origin of the left bundle branch *(Massing et al., 1976)*.

Clearly the anatomy of the left bundle branch system maybe variable and may not conform to a constant bifascicular division represented as anterosuperior (thin) and posteroinferior (broad) fascicles. These two fascicles originate from the left bundle branch directly after an initial undivided course of about 20 to 30 mm, to the anterior and

posterior papillary muscles. This permits delivery of a slightly earlier signals to these two parts, since their effective contraction should slightly precede contraction of the ventricular free wall in order to prevent mitral regurgitation. The right bundle branch peripheral branches are fairly distributed. Terminal branches of the AV node artery supply the AV bundle and few millimeters to the right and left bundle branches. Distal to the inferior margin of the membranous septum, the bundle branches on both sides are supplied by septal arteries arising from anterior descending artery *(James et al., 1982)*. Most of the fibers in bundle branches resemble the classic Purkinje fibers. In human hearts the bundle branches are composed exclusively of Purkinje fibers and there are many other fibers which are directly continuous with Purkinje fibers but which are morphologically indistinguishable from ordinary myocardium. This occurs not only at the points of transition from the bundle branch area to definite myocardium but also, within the main course of the bundle branch area itself. *(James et al., 1982)*.

Terminal Purkinje Fibers

These fibers connect with the ends of the bundle branches to form interweaving networks on the endocardial surface of both ventricles that transmit the cardiac impulse almost simultaneously to the entire right and left ventricular endocardial surfaces. Purkinje fibers tend to be less concentrated at the base of the ventricle and at the papillary muscles tips. They penetrate the myocardium for varying distances depending on the animal spices, in man they apparently penetrate only the inner third of the endocardium, while in pig they almost reach the epicardium *(Zipes, 1984)*. Much confusion has resulted from the suggestion that rapid conduction is an exclusive property of Purkinje fibers. The presence of fibers that have ordinary appearance within the bundle branches suggest that some fibers also conduct rapidly. As discussed previously it is quite probable that rapidly conducting fibers in the human heart often exhibit Purkinje

characteristics similar to those in ungulates but some fibers which we now consider morphologically as ordinary myocardium must possess also the ability to conduct rapidly *(James et al., 1982)*.

Anatomical Substrates for Preexcitation

Ventricular preexcitation is defined as the situation where the ventricles are activated more rapidly than would be anticipated had the impulse been conducted vial the normal atrioventricular junctional area *(Durrer, 1970)*. Although some authorities have suggested that the disorder can result from a functional malformation in a normal conduction system *(Sherf et al., 1969)*. It is more usual to search for discrete anatomic short circuits of the junctional area as its explanation. The function of the specialized atrioventricular junctional area is to produce delay. There are several theoretical possibilities whereby anatomic connections could short circuit all or part of the delay produced within the specialized atrioventricular area. The morphology of various connections which indeed short circuit the junctional area has been previously described by various investigators notably *(Lev et al., 1966)*. However it is not always easy to correlate anatomic and clinical studies because of the eponymous names used to describe the various connections. For example accessory atrioventricular connections which exist outside the specialized junctional area are widely termed "bundles of Kent". Unfortunately the connections thus described bear no resemblance to the structure observed by *Kent* himself. The fibers described by *Mahaim (1947)* certainly do short circuit the junctional area. For present day needs it seems desirable to differentiate Mahaim fibers which arise from the node from those which arise from the penetrating bundle and branching bundle. Further confusion exists with regard to the nodal bypass fibers described by *James (1961)*. These tracts is no way resemble the fiber tract described by *Brechenmachen (1975)* in hearts with short P-R normal QRS syndrome. Yet such syndromes are