Atrioventricular Reentry Tachycardia

4

Vincent C. Thomas, Nicholas Von Bergen, and Ian H. Law

Accessory Pathways

An anomalous strand of working myocardium, called an accessory pathway, bridges the electrically isolated gap between the atria and ventricle (i.e., the AV groove) and supports atrioventricular reentrant tachycardia (AVRT, Fig. 4.1). These pathways are likely formed as a result of incomplete senescence (apoptosis) and differentiation during the first trimester of gestation. During embryologic development, the fetal muscle fibers are oriented longitudinally in the straight heart tube which loops and differentiates into the four chamber heart. Some of the fibers of the straight heart tube undergo senescence (apoptosis—programmed cell death) leaving only the atrioventricular node and His-Purkinje system (AVN-HPS) electrically connecting the atria and ventricles. However, in some cases there is failure of apoptosis and anomalous connections—the accessory pathways—may persist beyond birth (see Chap. 1).

V.C. Thomas, M.D., M.H.A. (✉)
Department of Pediatrics,
University of Nevada School of Medicine,
3006 S. Maryland Parkway, Suite 690,
Las Vegas, NV 89109, USA
e-mail: vthomas@childrensheartcenter.com

N. Von Bergen, M.D.
Department of Pediatrics, University of Wisconsin—Madison, Madison, WI, USA

I.H. Law, B.S.E., M.D.
Division of Cardiology, Department of Pediatrics,
University of Iowa Children's Hospital, University of Iowa Carver College of Medicine,
Iowa City, IA, USA

Manifest Accessory Pathways

Accessory pathways may be manifest or concealed. Manifest accessory pathways comprise antegrade activation of the ventricular myocardium through the accessory pathway and can "manifest" as preexcitation on an electrocardiogram during sinus rhythm (Figs. 4.1, 4.2, 4.3). This pattern is called the Wolff–Parkinson–White syndrome for the three physicians who described the syndrome in 1930—short PR interval, preexcitation (delta wave), paroxysmal tachycardia, usually first appearing in young persons. Even when ventricular preexcitation is present on the surface ECG during sinus rhythm, the mechanism of the tachycardia is usually orthodromic AVRT.

Concealed Accessory Pathways

Concealed accessory pathways are those that do not have antegrade conduction, but only

© Springer Science+Business Media New York 2015

M. Dick, II (ed.), *Clinical Cardiac Electrophysiology in the Young*,
DOI 10.1007/978-1-4939-2739-5_4

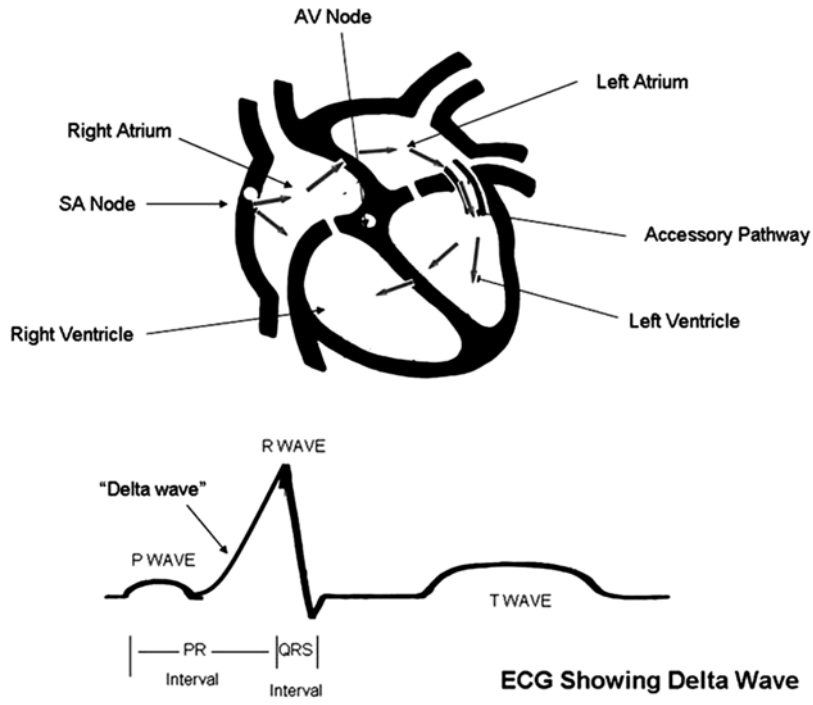

Fig. 4.1 Cartoon of the heart demonstrating the atrioventricular conduction system with a left accessory connection. The P-wave and QRS complex are show short the indicating the impulse PR interval bypassing the AV node; also shown is the "delta wave" transcribing the slow anomalous ventricular activation and resulting in the early arrival (preexcitation of the ventricle) of the impulse through the accessory pathway. [Reprinted Kuilig J, et al. Wolff-Parkinson White syndrome and accessory pathways. Circulation 2010;122(15): e480–e483. With permission from Wolters Kluwer Health]

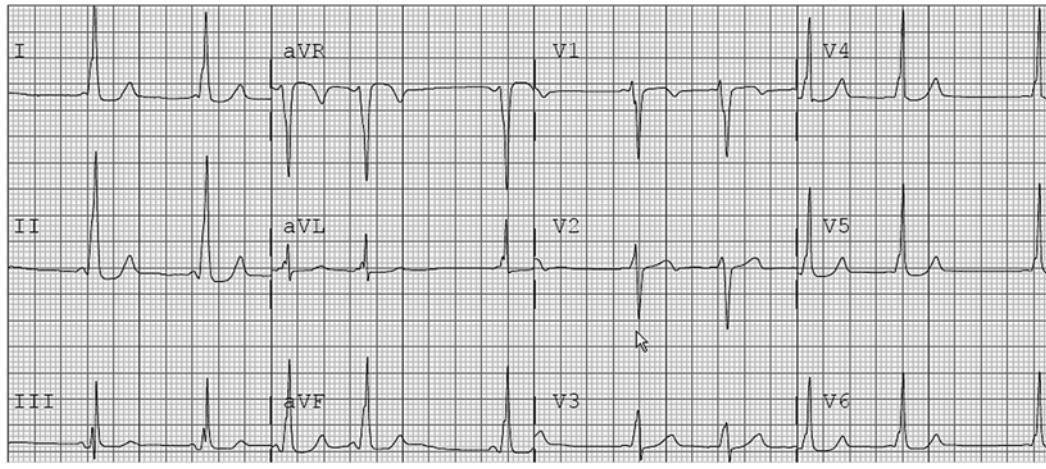

Fig. 4.2 12-Lead ECG demonstrating preexcitation with a right-sided pathway in an 8-year-old female. This was mapped to the right anterior septal area as suggested with the R/S transition between V2 and V3 (suggesting a possible septal pathway) and the positive preexcitation noted in II, III, aVF

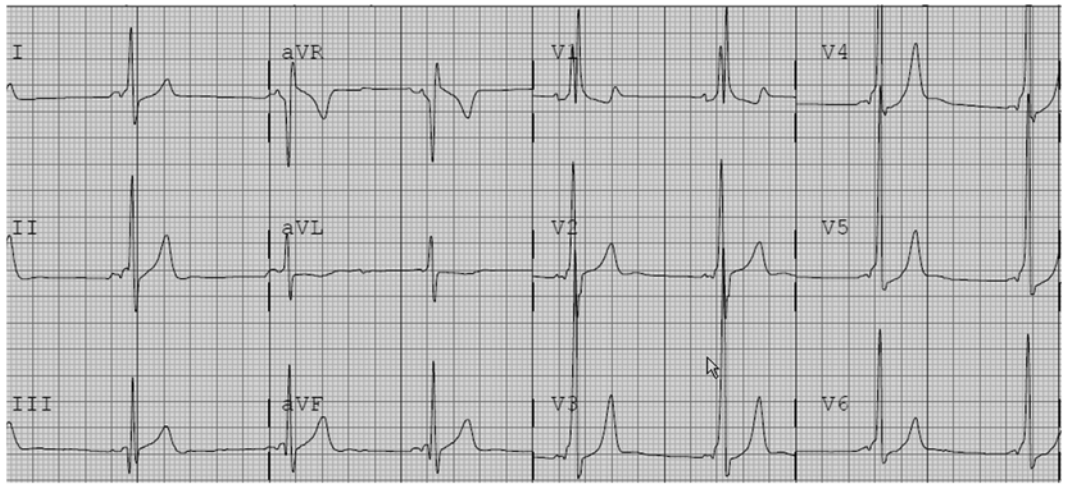

Fig. 4.3 12-Lead ECG of patient with Wolff–Parkinson–White syndrome with a left posterior accessory pathway. The PR interval is short and delta waves are easily visualized. The R/S transition is before V1 suggesting a left-sided pathway

Definitions:

Orthodromic is used to describe accessory pathway (AP)-mediated tachycardias in which there is normal conduction from the atria to the ventricle via the AVS-HPS.

Antidromic is used to describe AP-mediated tachycardias which traverse the AP from atrium to ventricle and proceed backwards up the HPS.

Antegrade indicates conduction from atria to ventricle.

Retrograde indicates conduction from ventricle to atrium.

Concealed pathways only conduct in the retrograde direction.

Manifest pathways conduct in the antegrade direction with or without retrograde conduction as well.

Mechanism of Atrioventricular Reentrant Tachycardia

Atrioventricular reentrant tachycardia can be either orthodromic or antidromic. Orthodromic AVRT (Fig. 4.4), comprising approximately 90 % of AVRT, denotes antegrade conduction through the AVN-HPS and retrograde conduction through the accessory pathway, producing a narrow QRS tachycardia. This pattern can be seen in all patients with concealed and the great majority of patients with manifest pathways. If a patient has a manifest accessory pathway (WPW), ventricular preexcitation is no longer present during orthodromic AVRT as the antegrade ventricular activation occurs through the His-Purkinje system, no longer passing from the atrium to the ventricle across the accessory pathway.

In contrast, antidromic AVRT is characterized by antegrade conduction through a manifest accessory pathway (and not a concealed one) and retrograde conduction through the His-Purkinje-AV nodal system, resulting in a wide QRS complex tachycardia, mimicking ventricular tachycardia (Figs. 4.5a and 4.6). Activation of the ventricular myocardium via the accessory pathway results in a wide QRS complex secondary to working myocardial cell-to-cell conduction rather than conduction through the His-Purkinje system.

ECG Diagnosis

For those patients with manifest pathways, the ventricular preexcitation pattern in sinus rhythm can be helpful in determining the location of the

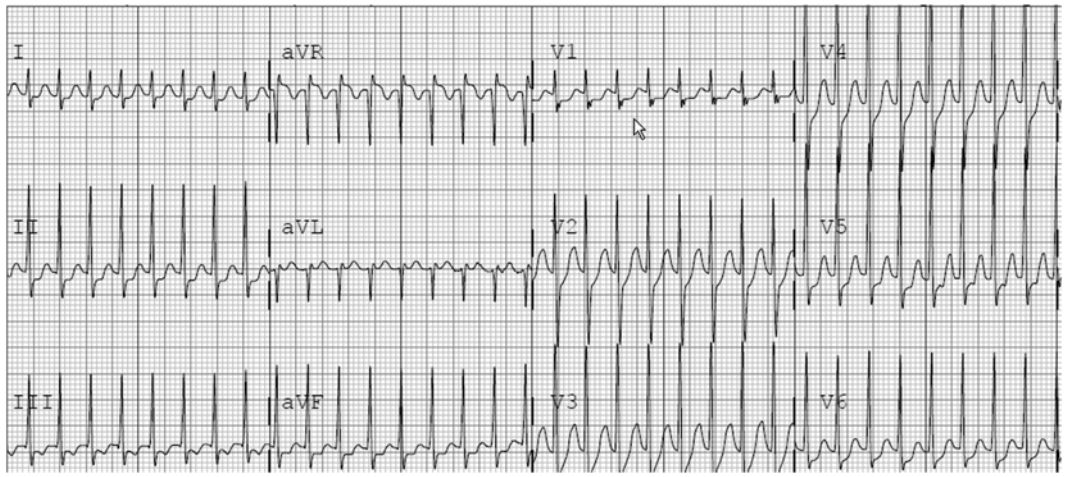

Fig. 4.4 12-Lead ECG with narrow QRS tachycardia in a 10-year-old female

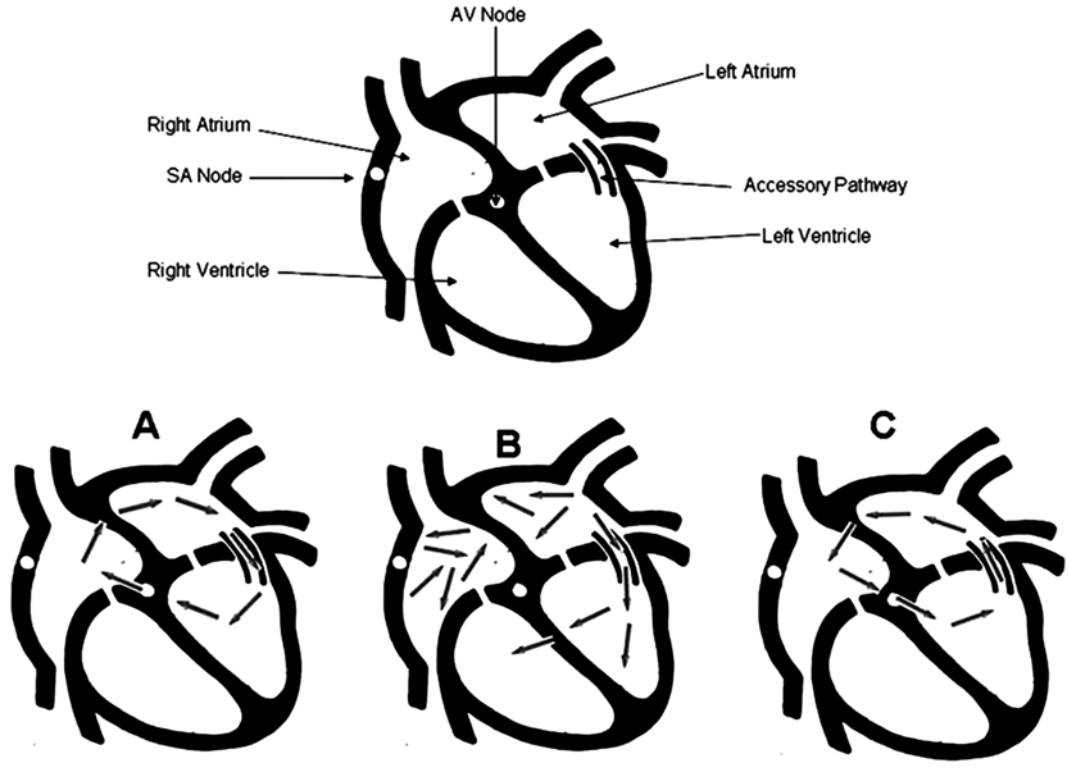

Fig. 4.5 Cartoon demonstrating typical orthodromic AVRT (*C*), rare antidromic AVRT (*A*), and antidromic AVRT during atrial fibrillation (*B*). (see text). [Reprinted Kuilig J, et al. Wolff–Parkinson–White syndrome and accessory pathways. Circulation 2010;122(15): e480–e483. With permission from Wolters Kluwer Health]

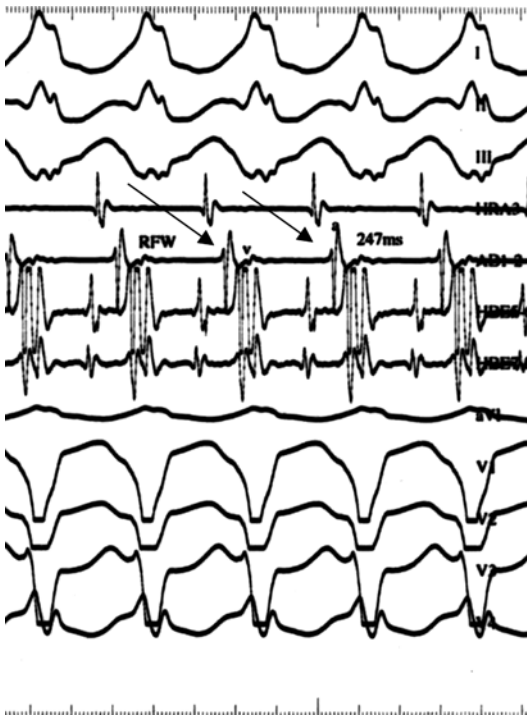

Fig. 4.6 An 8-Lead ECG demonstrating antidromic tachycardia in a 5-year-old boy. The *top* three tracings are recorded through the limb leads I, II, III; the *bottom* four tracings are aVL, V1, V2, V3, V4. The tracing fourth from the top is recorded through the high right atrial catheter. The tracing fifth from the top is recorded through the mapping catheter located in the right atrial free wall near the tricuspid valve. The sixth and seventh tracings are recorded through the His bundle catheter located at the low right atrial-septal area. Note the wide QRS tachycardia with maximal preexcitation of a left bundle branch configuration, compatible with a right-sided pathway. Note the tight AV relationship between the right atrial free wall electrogram (*arrows*) and the surface QRS complex (and intracardiac ventricular electrogram) indicating antidromic conduction down the right lateral accessory conduction to the ventricle. Retrograde conduction (not shown) supporting the tachycardia was through a left-sided concealed accessory pathway. Ablation of both pathways was successful

accessory pathway. A number of algorithms have been developed to aid in pathway location with variable accuracy depending on the accessory pathway location. Briefly, if the QRS transition (R greater than S-wave) is after V3 in the precordial leads, the pathway is most likely on the right side; if it is at or between V2 to V3, it is most likely a septal pathway. If the transition is before V2, it is most likely on the left side. Using published algorithms, further analysis of the delta wave polarity in the ECG leads can identify the location of the accessory pathway even more precisely. In patients with a concealed accessory pathway, the resting ECG is of no diagnostic value to determine accessory pathway location.

An ECG obtained during SVT can be very helpful to indicate AVRT (vs. AVNRT) and occasionally to evaluate the location of the accessory pathway, especially when compared to an ECG obtained in sinus rhythm. Retrograde P-waves are typically present (typically with an R-P retrograde interval greater than 65–70 ms) in the ST segment or T-wave during AVRT, especially in Lead I, and suggests the diagnosis. This contrasts with a retrograde P-wave less than 40–60 ms in typical AVNRT. Table 4.1 summarizes the findings for manifest and concealed pathways characteristically seen on the surface electrocardiogram during either sinus rhythm or SVT.

Manifest Accessory Pathways

Wolff–Parkinson–White Syndrome

The preexcitation is the result of a congenital accessory pathway that conducts antegrade reaching the ventricles in advance of activation through the AVN-HPS (Figs. 4.1, 4.2, 4.3). The incidence of Wolff–Parkinson–White syndrome in children has been estimated around one in every 1,000 individuals. Ventricular preexcitation can be quite subtle, occasionally apparent only as a lack of a q-wave in the lateral precordial leads due to slow conduction in the accessory pathway; accelerated conduction through the AV node (as is common in children); or in later accessory pathway activation of a left-sided pathway (the most frequent site of a pathway).

While both manifest and concealed pathways can mediate reentrant tachycardia, patients with Wolff–Parkinson–White syndrome have a greater rate of recurrent SVT than those without preexcitation. The clinical course of AVRT is age related. If diagnosed as an infant, the preexcitation may resolve to complete disappearance as a child

Table 4.1 Summary of the findings for manifest and concealed pathways

	Sinus			SVT				
							Mahaim	
	Concealed	Manifest WPW	Mahaim (Manifest)	Concealed	WPW Orthodromic	WPW Antidromic	Atriofascicular	Nodo-or fasciculoventricular
Rate—beats/min	Within normal limits for age	Within normal limits for age	Within normal limits for age	Infants: 200–300; children: 160–250	Infants: 200–300; children: 160–250	Infants: 200–300; children: 160–250	Infants: 200–300; children: 160–250	Infants :200–300; children: 160–250
Rhythm	Regular	Regular	Regular	Regular	Regular	Regular	Regular	Regular
P-wave	Normal	Normal	Normal	Absent or negative, within T-wave of lead II, aVF	Absent or negative, within T-wave of lead II, aVF	Absent	Absent	Absent or negative, buried within T-wave of lead II
PR interval and QRS complex	Normal	Pre-excited (delta-wave, short PR interval)	Pre-excited (delta-wave, short PR interval)	Normal or wide when aberrantly conducted	Normal or wide when aberrantly conducted	Wide with QRS pattern similar to or wider than sinus rhythm	Wide (ART-AVRT) with LBBB QRS pattern	Wide (ORT-AVRT) with QRS pattern

Comparison of concealed and manifest accessory pathways, and Mahaim fibers during sinus rhythm and SVT. *WPW* Wolff–Parkinson–White syndrome, *ART* antidromic reentrant tachycardia, *ORT* orthodromic reentrant tachycardia, *AVRT* atrioventricular reentrant tachycardia, *LBB* left bundle branch

grows, usually by 18–24 months of age (30 % recurrence if diagnosed less than 1 year of age). However, if preexcitation or documented SVT is seen after this age, AVRT is unlikely to self-resolve (94 % recurrence rate).

Furthermore, patients with WPW are noted to have a very small, albeit increased, risk for a sudden cardiac arrest. During atrial fibrillation, a conduction of the fibrillatory impulses may preferentially cross a fast conducting manifest accessory pathway, bypassing the more slowly conducting AV node resulting in a rapid life-threatening ventricular response (>250–300 bpm) (Fig. 4.5b; Chap. 20, Fig. 20.2). The incidence of sudden cardiac arrest in WPW patients has been estimated between one and 4.5 per 1,000 patient years, reflecting a higher incidence of atrial fibrillation in WPW patients. The highest prevalence of sudden cardiac arrest appears to be between the ages of 15 and 35 years. Patients with WPW syndrome who experience sudden cardiac death may have no history of a tachyarrhythmia. On the other hand, they may have had previous syncopal episodes. Potential risk factors for a sudden cardiac event with WPW include a younger age (<30 years), prior syncope or atrial fibrillation, male gender, familial WPW, or other heart disease.

The majority of accessory pathways are seen in patients with structurally normal hearts; however, approximately 10–15 % of patients with Wolff–Parkinson–White syndrome has congenital heart disease (Ebstein's anomaly, L-transposition of the great arteries, cardiomyopathy, and intracardiac tumors). Six to nine percent of patients who have WPW syndrome and congenital heart disease are more likely to have multiple pathways. Approximately a quarter of patients with Ebstein's anomaly also have preexcitation, the most common association. In addition, up to 43 % of patients with WPW and Ebstein's anomaly have multiple pathways. In patients with other forms of congenital heart disease, 25 % had multiple pathways. Pathway location is also associated with congenital heart disease: right-sided pathways are more common in congenital heart disease (63 %) whereas left-sided pathways are more common in the structurally normal heart (61 %). Interestingly, the appearance of preexcitation in patients with congenital heart disease may be misleading, as some malformations—tricuspid atresia and hypoplastic left heart syndrome—may be associated with "pseudo-preexcitation" or the appearance of preexcitation and not have an accessory pathway.

The management of the asymptomatic patient with WPW has been addressed by the Pediatric and Congenital Electrophysiology Society in a recent consensus statement (2012). Patients who are 8 years of age or older and are found to have manifest preexcitation on an ECG without symptoms are recommended to undergo risk stratification, initially with noninvasive studies (i.e., exercise treadmill test; Holter monitor tracings). If inconclusive, invasive studies can be considered to determine those at higher risk for sudden cardiac arrest. In asymptomatic individuals under 8 years of age, the low risk of sudden death due to atrial fibrillation, allow conservative management and continued follow-up.

Atrioventricular and Mahaim Fibers

Mahaim fibers, as first described in 1938, are direct histologic links between either the AV node and the right ventricle (nodoventricular fibers) or the His bundle and the right ventricle (fasciculoventricular fibers). These nodo or fascicular ventricular fibers are anatomically common (though often inactive, i.e., not resulting in preexcitation or tachycardia) When they do produce ventricular preexcitation, it may be only in a "by-stander" roles and not participate in an arrhythmia reentrant circuit. In patients with a nodoventricular fiber, the PR interval varies relative to the fiber's take-off from the node. In those with a fascicular-ventricular fiber arising from the His bundle and inserting into the ventricular myocardium the PR interval is normal. Rarely the nodoventricular fibers produce a wide QRS complex in the context of a different mechanism for SVT, such as AVNRT (Fig. 4.7), or even more rarely participate as a true antegrade limb of an atrioventricular circuit. Fasciculoventricular fibers have not been shown to participate in a reentrant circuit other than as a bystander. These bystander pathways are often best left alone and not ablated.

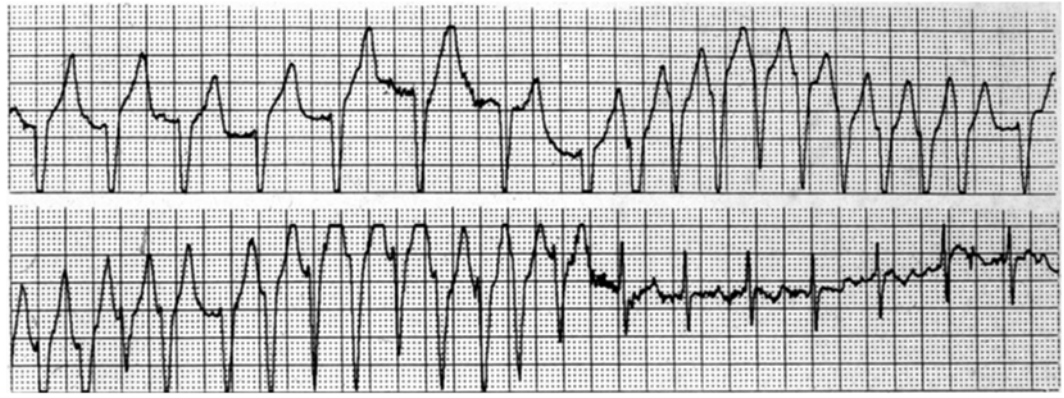

Fig. 4.7 Wide QRS rhythm and tachycardia followed by narrow QRS sinus rhythm in a 9-year-old girl. Electrophysiology study demonstrated a nodoventricular fiber (not involved in the reentrant circuit), which was ablated. Narrow QRS tachycardia was demonstrated post ablation of the Mahaim fiber and was found to be due to AVNRT. The slow pathway and arrhythmia were ablated

An uncommon right-sided atriofascicular fiber (somewhat confusingly also referred to as a Mahaim fiber) is located in the right posterior AV groove (Fig. 4.8). This accessory pathway has several features: (1) it typically conducts in only an antegrade direction producing preexcitation in a left bundle branch block configuration; (2) it inserts deep (not along the AV ring) in the right ventricle, often near the distal fascicle of the right bundle (moderator band); (3) the pathway demonstrates decremental conduction; (4) if present, the tachycardia is antidromic resulting in a wide QRS left bundle branch pattern SVT. These fibers were shown to be the cause of preexcitation characterized by left bundle branch block QRS morphology and wide QRS tachycardia. Intracardiac electrophysiologic studies demonstrate that the preexcitation and the wide QRS tachycardia are due to an atriofascicular accessory pathway coursing from the lateral right atrial tricuspid valve annular area to insert deeper in the right bundle rather than at the AV groove. As these fibers often have AV nodal properties, they may exhibit slow conduction velocity and "decremental conduction" with atrial extrastimulus and adenosine sensitive, resulting in AV block with adenosine administration. The hallmark of identifying the site for ablation is the identification of a fast action specialized conduction tissue electrogram distant in both location and timing from the His bundle electrogram, often near the moderator band. Ablation at that site terminates conduction within this pathway (eliminates the preexcitation), as well as the wide QRS tachycardia (Fig. 4.8). It can also be ablated on the right lateral annulus.

Diagnostic Evaluation of AVRT

Exercise Treadmill Testing and Holter Monitoring

Abrupt disappearance of ventricular preexcitation on the electrocardiogram during exercise suggests an accessory pathway effective refractory period perhaps as long as 360–390 ms, placing the patient in a lower risk category (Fig. 4.9). Exercise testing or Holter monitoring during activity has been used as a noninvasive method to evaluate the conduction properties of a manifest accessory pathway. Abrupt loss of preexcitation during exercise treadmill testing occurred in 15 % of a predominately s pediatric group of patients. However, in practice, disappearance of ventricular preexcitation exercise testing can be difficult to interpret due to movement artifact on the ECG and enhanced AV node conduction from increased adrenergic tone during exercise resulting in gradual (and less) not abrupt diminution of the ventricular preexcitation.

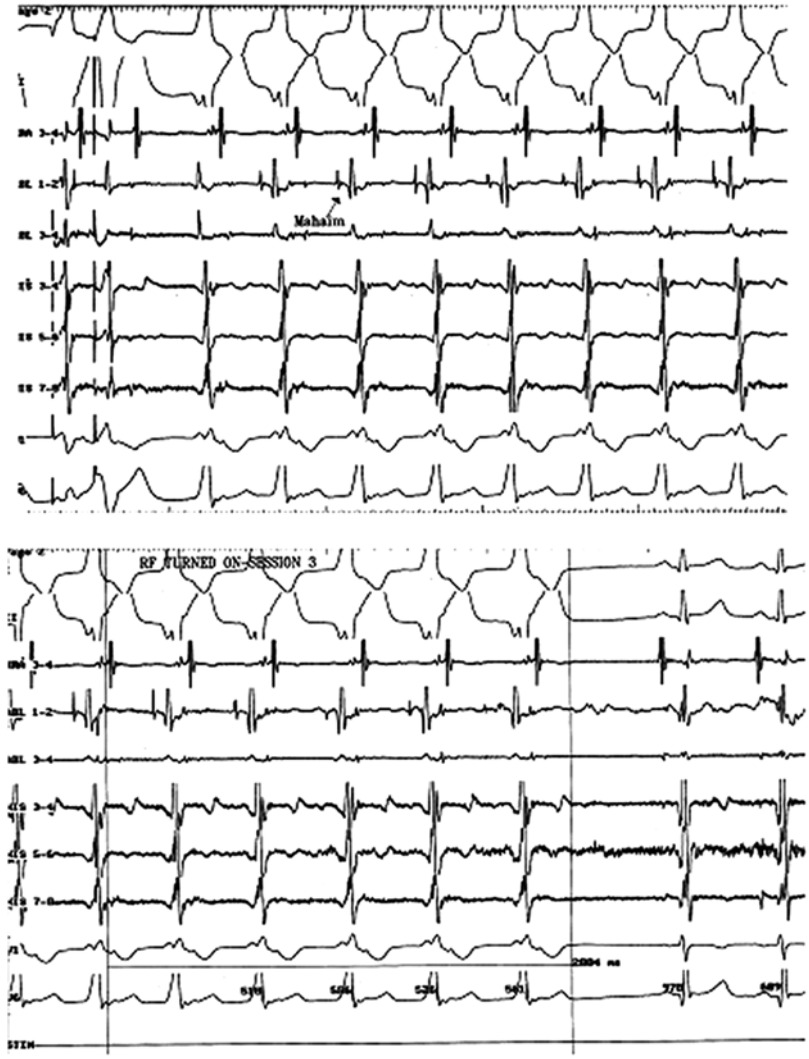

Fig. 4.8 Patient with an atrioventricular fiber causing wide QRS tachycardia. *Top panel*: Initiation of wide QRS tachycardia with antegrade conduction through the Mahaim fiber (note fast action deflection of atriofascicular fiber activation) and retrograde conduction through the normal His-Purkinje–AV node system. *Bottom panel*: Radiofrequency ablation of antegrade conduction in the atriofascicular fiber, terminating the tachycardia. Note the His bundle electrogram in the two sinus beats, as well as the absence of fast action deflection of the atrioventricular fiber in the mapping/ablation catheter post ablation

Holter monitoring of patients with WPW may demonstrate intermittent ventricular preexcitation. This intermittent conduction down the manifest accessory pathway is thought to confer low-risk attributes for the ability to conduct atrial fibrillation rapidly. This finding can be reassuring in the risk stratification of patients for sudden cardiac arrest, but does not exclude the possibility of SVT.

Electrophysiology Study

The electrophysiology study (see Chap. 3) is an essential tool in the assessment of the patient with documented SVT. Although atrial extrastimuli can be delivered through a transvenous catheter or transesophageal catheter (particularly useful for infants and small children), the transcatheter intracardiac technique is necessary for a

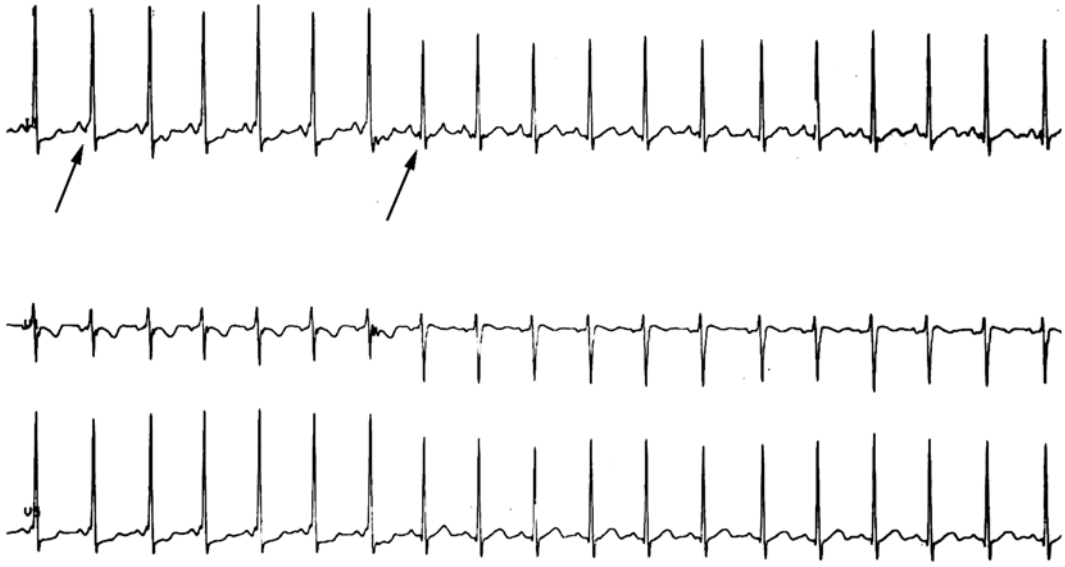

Fig. 4.9 Leads II, V1, and V5 during an exercise treadmill test. Note the delta wave in the onset of the QRS (*left arrow*) and abrupt loss over 2–3 beats (*right arrow* and next two beats) as the heart slightly increases, suggesting a slowly conducting accessory pathway

definitive examination of the properties and location of the accessory pathway as well as for transcatheter ablation.

Baseline Studies

The patient is usually placed under general anesthesia supervised by an anesthesiologist; conscious sedation is usually well tolerated in the adolescent but in many centers still requires the presence of the anesthesiology service. At the start of the electrophysiology study, baseline surface electrocardiogram and electrogram recordings are obtained. Baseline intracardiac intervals measured in patients with manifest accessory pathway conduction typically reveal an HV interval (Fig. 4.10) much less than normal (40 ms) or at times <0 ms (ventricular activation occurring before the His electrogram). When the patient has manifested accessory pathway conduction, the earliest area of early ventricular activation (prior to the onset of the surface QRS complex) during antegrade conduction through the accessory pathway can identify the location of ventricular insertion of the accessory pathway.

Antegrade and retrograde electrophysiologic properties of the AV node and accessory pathway can then be determined. Atrial pacing is performed at a cycle length slightly shorter than the sinus rate, and this is gradually decreased in 10–20 ms increments until 1:1 AV conduction is no longer seen (Wenckebach cycle length), or for a sudden loss of preexcitation. The loss of 1:1 AV conduction represents either the accessory pathway Wenckebach cycle length (if the preceding beats were preexcited), or the AV node Wenckebach cycle length (if the preceding beats were conducting through the AV node, typically narrow complex).

The antegrade effective refractory period of the AV node and accessory pathway can then be determined by delivering extra stimuli after an 8–10 beat drive train cycle length (e.g., at both 600 and 400 ms. The S2 stimulus is decremented by 10–20 ms observing for a loss of AV conduction reflecting the antegrade effective refractory period of the AV node or the loss of preexcitation representing the effective refractory period of the accessory pathway or both (Fig. 4.11). In rare cases, it may be possible to differentiate between two separate antegrade conducting pathways noted by alterations in the pre-excited QRS

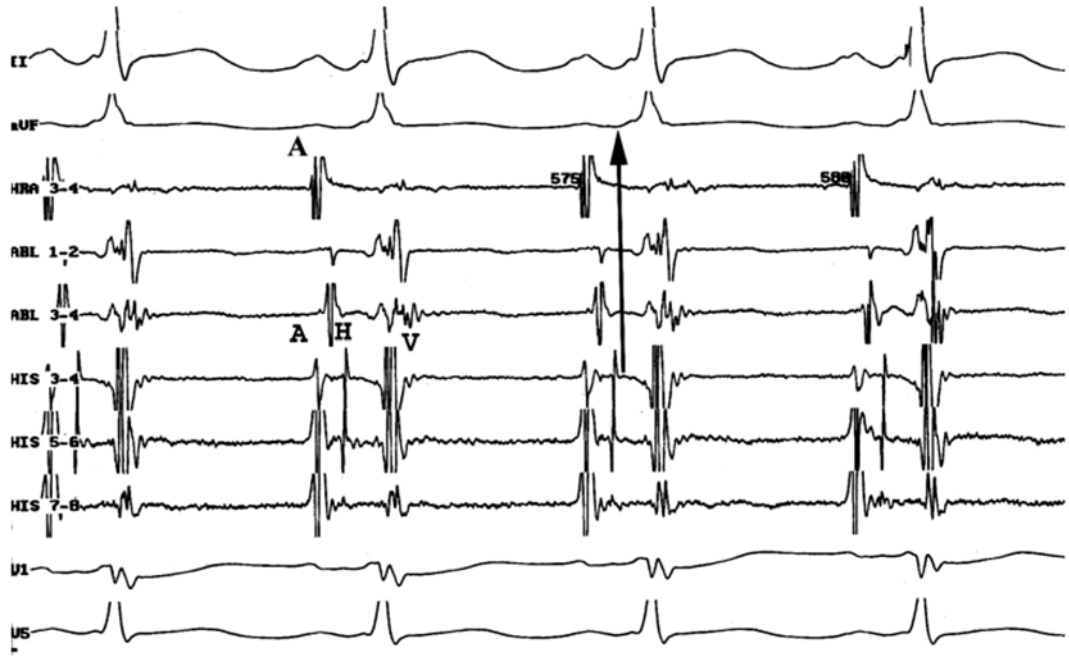

Fig. 4.10 Surface electrocardiograms (II, aVF, V1, V5) and intracardiac electrograms obtained during sinus rhythm in a patient with a left lateral accessory pathway. A very short HV interval (H to arrow) is seen in the His electrogram tracings indicating that ventricular activation is reaching the ventricles earlier than expected if the wave front from the sinus impulse were only traversing the atrioventricular nodal–His-Purkinje axis (i.e., the ventricles are pre-excited). *A* atrial electrogram, *HRA* high right atrium, *HIS = H* His, *Abl* ablation catheter in RA

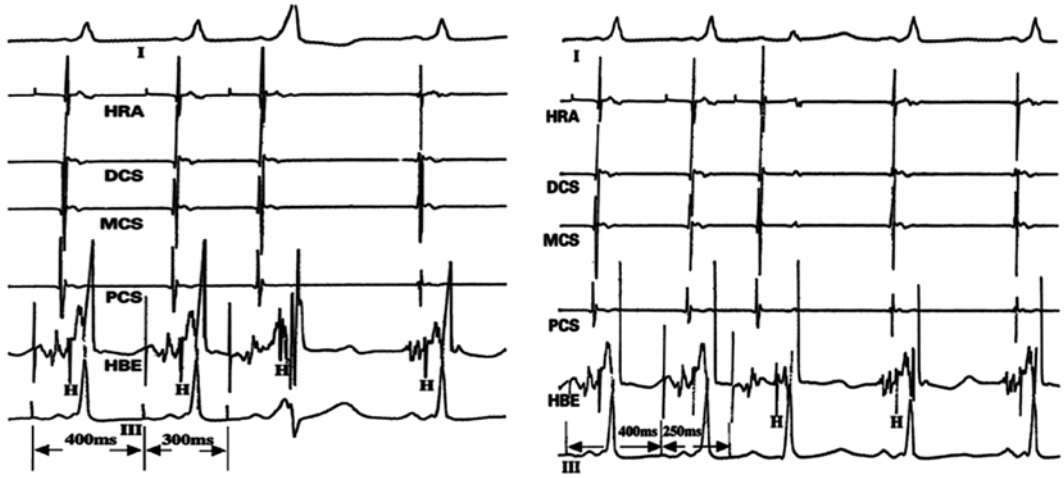

Fig. 4.11 Surface electrocardiograms and intracardiac electrograms obtained during an atrial extrastimulus in a patient with preexcitation. In the *left panel*, the premature atrial stimulus (300 ms coupling interval conducts antegrade through the accessory pathway resulting in more pronounced ventricular preexcitation and a short AV interval in the distal CS electrode pair. By shortening the atrial extrastimulus by 40 ms, the accessory pathway becomes refractory resulting in normal conduction through the AV node, a narrow QRS complex, and lengthening of the AV interval in the coronary sinus electrodes. *HRA* high right atrium, *HIS* His, *CS* coronary sinus, *RVA* right ventricular apex

morphology. In a similar manner, retrograde properties of the AV node and accessory pathway can be determined using the same techniques but pacing from the ventricle and monitoring for loss of VA conduction or eccentric accessory pathway conduction. If the manifest accessory pathway is noted to have decremental conduction, this may be indicative of an atriofascicular pathway.

Electrophysiologic Evaluation of the Manifest Accessory Pathway

As part of a risk stratification strategy for WPW syndrome, induction OD atrial fibrillation during the electrophysiology study provides useful information. Once atrial fibrillation is induced, by burst atrial pacing, the pre-excited R–R intervals are monitored. In a study of 60 children, a short, pre-excited R–R interval during rapid atrial fibrillation was the most sensitive predictor of sudden cardiac death (Chap. 20, Fig. 20.2), with all patients with a cardiac arrest having an interval <220 ms (though not all patients in this group had previously had an arrest). In this group, an interval <220 ms was potentially associated (the natural history and risk was eliminated by surgery in some subjects with <220 ms pre-excited R–R interval) with a positive predicted value for cardiac arrest of 68 %. In some patients, atrial fibrillation may not be inducible or sustained; rapid atrial pacing with or without isoproterenol, although used, is not a good surrogate for atrial fibrillation. The antegrade accessory pathway effective refractory period has also been used for risk stratification but has been less well correlated than atrial fibrillation. Multiple manifest accessory pathways have also been demonstrated to confer a higher risk for ventricular fibrillation. Conversely, intermittent loss of preexcitation during sinus rhythm suggests a low-risk pathway.

Electrophysiologic Evaluation of the Concealed Accessory Pathway

By definition, concealed accessory pathways cannot be evaluated during atrial pacing due to lack of antegrade conduction. However, both manifest and concealed accessory pathways can be evaluated by retrograde assessment using ventricular pacing. A straightforward method to determine the presence of a concealed accessory pathway is performed by ventricular pacing during a bolus infusion of IV adenosine (200 mcg/kg up to 12–18 mg) followed by a rapid flush (Fig. 4.12). Ventricular pacing at fast enough rates (<500 ms cycle length, 120 bpm) assures that the pacing rate is faster than the reflex sinus tachycardia that follows adenosine infusion in 15–20 s. As adenosine usually blocks retrograde conduction in the AV node a change in the atrial activation sequence suggests a shift of retrograde conduction from the AV node to the accessory pathway (Fig. 4.12), particularly evident with a left-sided accessory pathway. At times, a significant change in the retrograde atrial activation may not be seen due to either a paraHisian or anterior pathway accessory pathway or an adenosine-resistant AV node. Occasionally, a concealed accessory pathway conducting slowly retrograde will block with an adenosine bolus during ventricular pacing.

Similar to antegrade assessment, the retrograde Wenckebach cycle length and retrograde effective refractory periods of the AV node and accessory pathway are determined by rapid ventricular pacing and extrastimulus pacing. Though uncommon, the presence of multiple concealed pathways can be determined during extrastimulus pacing observing for multiple changes in the retrograde atrial activation sequence or after ablation of one pathway.

Induction of SVT and Diagnostic Pacing Maneuvers

During the atrial and ventricular pacing maneuvers, SVT is induced and usually well tolerated. However, the hemodynamic status is closely monitored for any compromise. If there is significant hemodynamic compromise, overdrive pacing maneuvers, adenosine, or rarely direct current cardioversion may be required. On occasion, an isoproterenol continuous infusion is required to initiate SVT. The starting dose of isoproterenol is 0.01–0.02 mcg/kg/min; rarely is more than

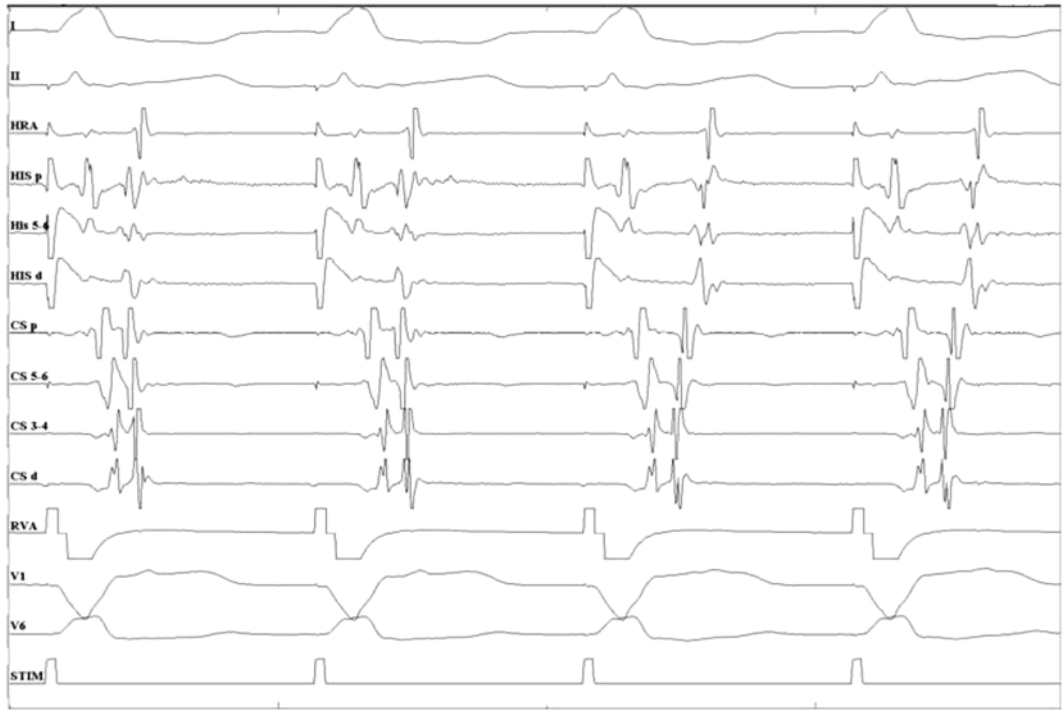

Fig. 4.12 Change in retrograde atrial activation sequence following adenosine infusion during ventricular pacing. At the far left the earliest atrial activation is seen in the proximal CS (CSp) and the His bipolar electrode pairs (His d, His 5–6, His p). As the adenosine blocks retrograde AV node conduction the earliest activation shifts (compare second and third complexes in CSp) to the distal coronary sinus bipolar electrode pairs (CS 5–6, CS 3–4). *HRA* high right atrium, *HIS* His, *CS* coronary sinus, *RVA* right ventricular apex

0.08 mcg/kg/min needed. When SVT cannot be induced with the previously described atrial and ventricular pacing maneuvers, two and three extrastimuli in the atrial or ventricle may be tried. Infrequently intravenous atropine, epinephrine, or caffeine may also be used in an attempt to induce arrhythmias.

If a bundle branch block pattern is seen during the initiation of the SVT, the difference in the cycle length in the SVT between wide QRS (bundle branch) and narrow QRS tachycardia (or changes in the cycle length as the bundle block branch pattern resolves) can be helpful in determining the location of the accessory pathway (Chap. 12, Fig. 12.6). An initially wide bundle block branch pattern during the tachycardia that prolongs the tachycardia cycle length is indicative of an ipsilateral accessory pathway (e.g., right bundle branch block lengthens the cycle length of orthodromic AVRT using the right-sided accessory pathway). This finding is explained by the slower conduction from myocyte to myocyte through the reentry circuit as compared to through the rapidly conducting His-Purkinje system. Therefore, a bundle branch block prolongs the ventricular transit time to the ipsilateral accessory pathway, thus lengthening the cycle length (Figs. 4.13 and 4.14). The corollary to this finding is that if the bundle branch block pattern does not alter the SVT cycle length, the bundle branch block is contralateral to the accessory pathway (e.g., the right bundle branch block pattern with left-sided accessory pathway does not alter SVT cycle length). Occasionally, this observation is confounded by a compensating faster conduction through the AV node neutralizing the cycle length change. This confounder is corrected by examining the AV conduction (time) as the AV conduction time remains constant regardless of cycle length change in the presence of an ipsilateral accessory pathway.

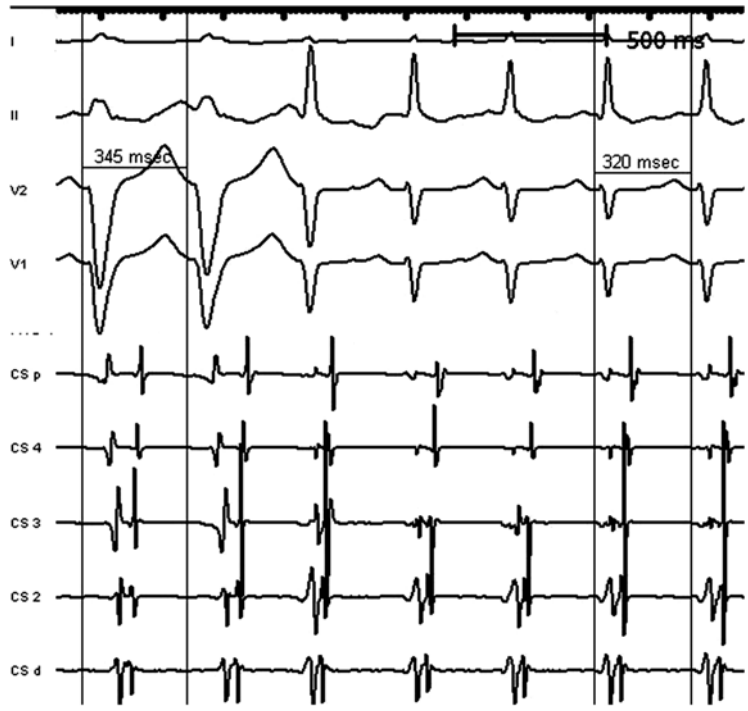

Fig. 4.13 A change in the cycle length of the tachycardia is noted concurrently to a normalization of the rate-related aberrancy (left bundle block configuration) to a narrow QRS complex on the surface and intracardiac electrograms. The electrograms in the CS catheters during narrow QRS tachycardia show left to right atrial activation from the distal CS to the proximal CS indicating a left-sided pathway with a slower tachycardia cycle length during the LBBB seen in the first two beats. This change suggests that the reentry circuit proceeds through an accessory pathway ipsilateral to the left bundle branch block

Various pacing maneuvers can be important diagnostic procedures. Premature ventricular beats introduced into the SVT can be helpful in distinguishing between AVRT and AVNRT and can help determine the location of the accessory pathway. The degree of preexcitation may help in deciphering between left- and right-sided accessory pathways as right-sided pathways typical pre-excite more prominently with right-sided pacing. Conversely, pacing from the left side (e.g., via the coronary sinus or left ventricle) may preferentially engage a left sided pathway. To ensure that retrograde conduction is not through the His bundle but rather the accessory pathway, premature beats may be given during His refractoriness (i.e., not earlier than 10–25 ms prior to the His deflection). If the ventricular stimulus is delivered during SVT while the His bundle is refractory in the presence of an accessory pathway, the retrograde conduction is through the accessory pathway shortening the atrial cycle length. The preexcitation index may be calculated to assist with localization of the accessory pathway and is determined by subtracting the longest premature ventricular pacing interval that shortened the atrial cycle length from the SVT cycle length (Fig. 4.14). The shorter the preexcitation index (longest PVC coupling interval), the more likely the pathway is to be on the right side. Posterior septal accessory pathways have a mean preexcitation index of 38 ms whereas anteroseptal septal accessory pathways have a mean preexcitation index of 17 ms. The longer the preexcitation index (shortest PVC coupling interval), the more likely the pathway is left-sided but the possibility of conduction through the His-Purkinje system/AV node should be considered, suggesting AVNRT. This can be usually distinguished by adenosine bolus infusion during ventricular pacing. Additionally, paraHisian pacing may be used to distinguish retrograde conduction through the AV node or an accessory pathway.

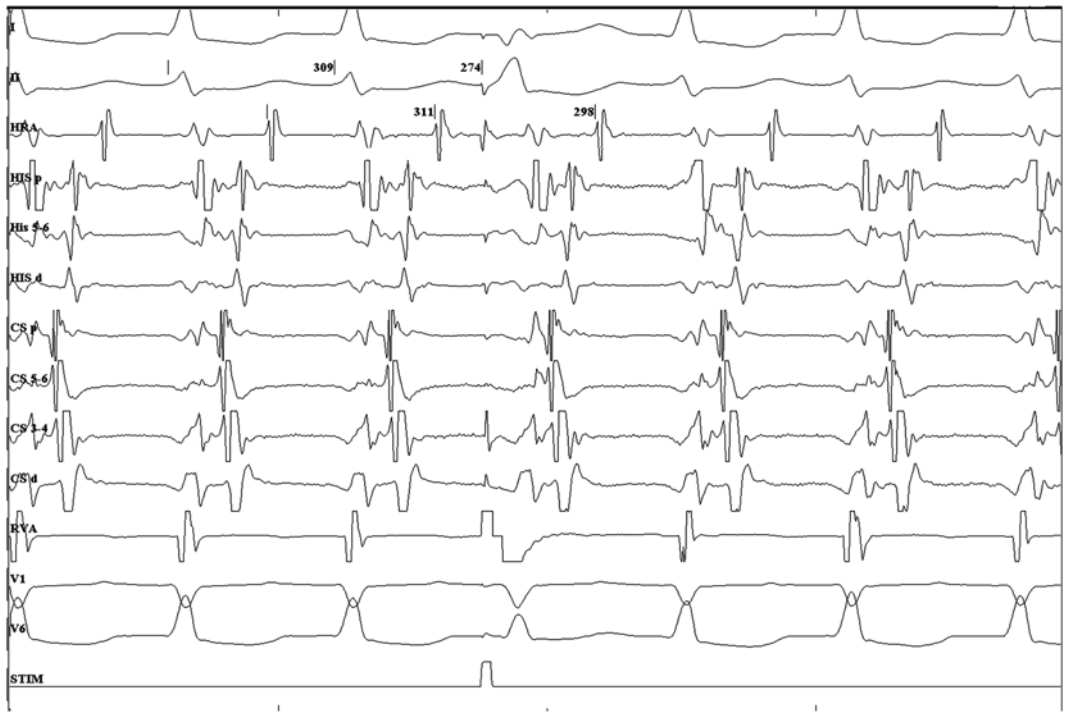

Fig. 4.14 Surface electrocardiograms and intracardiac electrograms obtained during orthodromic AVRT. A premature ventricular extrastimulus introduced during His refractoriness shortens the atrial cycle length by greater than 10 ms. The preexcitation index is calculated by subtracting the premature interval from the tachycardia cycle length (309–274 ms), resulting in a preexcitation index of 35 ms, consistent with a right-sided accessory pathway. *HRA* high right atrium, *HIS* His, *CS* coronary sinus, *RVA* right ventricular apex, *STIM* stimulation

The VA time can be measured after high output pacing at the His signal to capture both the His bundle (as noted with a more narrow QRS) and the ventricular myocardium. By slowly decreasing the pacing output until there is loss of His capture, with continued ventricular myocardial capture, the VA time is again measured. Accessory pathways continue to demonstrate consistent VA conduction time regardless of captured tissue (both or just heart muscle), while AV nodal retrograde conduction demonstrates an increase in the VA time consistent with the delay required for the ventricular myocardium to propagate into the His-Purkinje system.

Once an accessory pathway is identified a more precise method to determine the location of the pathway is by evaluating for the site of the shortest ventriculoatrial conduction time during ventricular pacing or orthodromic AVRT or the shortest AV conduction time during antidromic AVRT or in the presence of preexcitation. If the pathway is on the left side of the heart, a coronary sinus catheter advanced beyond the left-sided pathway may "bracket" the pathway (i.e., when there are bipolar electrograms from either side of the pathway that have longer VA or ventriculoatrial conduction times than the bipolar electrograms located near the accessory pathway during AVRT or ventricular pacing). If the accessory pathway is right-sided, a multipolar (e.g., duodecapolar) catheter or a roving catheter can be used to map around the tricuspid valve during SVT or ventricular pacing again observing for a short ventriculoatrial conduction time (or AV time with an antidromic SVT). 3-D mapping systems can enhance the ability to isolate the pathways using these techniques.

When mapping the accessory pathway during ventricular pacing (Chaps. 3 and 22), it is important to remember that when pacing the ventricle, retrograde conduction through both the accessory pathway and the AV node is likely. It is often

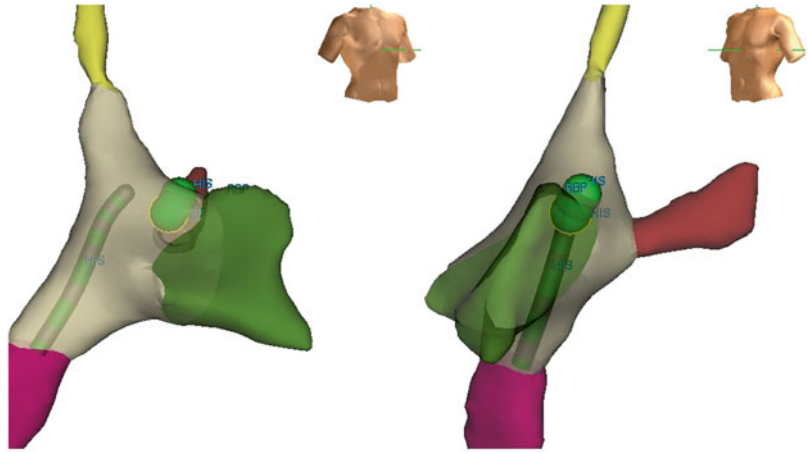

Fig. 4.15 RAO and LAO view of a paraHisian pathway mapped without radiation using 3-D mapping. The structures visualized include the SVC (*yellow*), IVC (*pink*), RA (*gray*), and RV (*green*). Note the location of the His labeled in close proximity to the ablation lesions (*green circles*). A small His signal was seen during the successful cryoablation of this accessory pathway. (Tie His catheter is seen in the RA and had been moved away from the His to facilitate cryoablation.) *HIS* His electrogram markings, *RBP* right bundle potential

possible to pace sufficiently fast to block conduction through the AV node and not the accessory pathway. If the pathway is located on the left side, pacing from the right ventricular outflow tract, LV apex or LV-free wall (retrograde through the aorta or transseptally across the atrial septum) may preferentially activate the left-sided accessory pathway prior to retrograde conduction through the AV node. Ventricular pacing at incrementally faster pacing rates may also be helpful in determining if there are multiple accessory pathways.

Complex anatomy and/or paraHisian pathways can make determination of the accessory pathway location more difficult. Three-dimensional mapping computer-based systems can prove to be extremely valuable in delineating the anatomy and precisely locating the area of His potentials (Fig. 4.15).

Treatment

Observation and Acute Treatment

As AVRT is rarely life threatening, and only rarely the cause of syncope (when orthodromic), several different options can be considered. For patients who have infrequent episodes that result in minimal or no symptoms, observation can be considered, especially in smaller children. In these patients, vagal maneuvers can be effective at terminating tachycardia (Chap. 23). If the tachycardia persists (>45–60 min) after vagal maneuvers, the patient should seek the assistance of a healthcare provider. In trained hands, esophageal overdrive pacing has been proven to be very effective, but is seldom used outside the infant age group. Medical cardioversion can also be very effective. Adenosine is 95–100 % effective in terminating AVRT. Typically when not effective, it is due to inadequate delivery of the medication to the coronary circulation and the AV node from too a slow flush, a distant delivery site that is too peripheral, or, in an infant, poor circulation. The advantage of adenosine is a very short half-life; its effects seldom last more than 10 s. However, once the adenosine effect is ended there is no further protection for recurrence of SVT and recurrences are not uncommon. When delivering adenosine, it is important to be prepared for atrial fibrillation that can occur as a result of adenosine infusion. Patients with Wolff–Parkinson–White syndrome and a fast conducting accessory pathway who develop atrial fibrillation after adenosine intervention are at risk for ventricular fibrillation. Although this combination is rare, an external

defibrillator should be readily available. Another option for medical cardioversion is verapamil, which is 80–95 % effective. Verapamil should not be used in infants (less than ~6 weeks old) and sparingly before 1 year of age, especially those in congestive heart failure as the calcium channel blocker effect can have a profound negative inotropic effect resulting in severe hypotension. Procainamide may have selective effect on the accessory pathway terminating the tachycardia but should be given slowly and with caution in the hypotensive patient.

For those patients who can tolerate their SVT but do not respond to vagal maneuvers, a "pill in the pocket" technique could be employed. These patients may take a beta-blocker or calcium channel blocker when the SVT begins, however if the SVT is not tolerated well, there may not be time for the antiarrhythmic medication to take effect.

Antiarrhythmic Medications

For those patients who have frequent events or do not tolerate their SVT, chronic antiarrhythmic medication is an option (Chap. 21). Digoxin for infants with SVT or a beta-blocker with or without digoxin have a long history and are safe when properly administered. However, as digoxin can shorten the accessory pathway effective refractory period, this should be avoided in patients who have Wolff–Parkinson–White syndrome. In older children digoxin has been largely replaced by others agents and beta-blockers are likely the most common antiarrhythmic medication used for daily prophylaxis. Sodium channel blockers and potassium channel blockers have are effective and there are liquid forms of all these antiarrhythmic medications. Amiodarone is an effective agent for especially resistant AVRT in infants and small children before they are candidates for ablation; however, duration of amiodarone should ideally not exceed 18–24 months. As noted previously, calcium channel blockers should not be used in children under ~6 weeks of age and caution in those less than 1 year. Some patients who have drug refractory SVT may require combination therapy such as sotalol (potassium with some beta blockade) and flecainide (sodium channel blockade) which has shown to be effective in a small group of children less than one year of age with refractory SVT.

Transcatheter Ablation

When antiarrhythmic therapy is ineffective or not desired, the ablation procedure offers the possibility of a definitive cure. Disruption of the accessory pathway was first performed surgically, followed by direct current ablation. Radiofrequency ablation was first introduced in 1987 and was reported in a series of pediatric patients in 1991. Today radiofrequency and cryotherapy are the most common energy sources used for transcatheter ablation and both have been shown to have excellent success and safety profiles.

Transcatheter ablation is discussed in Chap. 22. For patients who have paraHisian pathways or complex anatomy, 3-D mapping systems can be a useful adjunct to the electrophysiology study and the ablation procedure. Advances in mapping technology have resulted in 3-D spatial resolution of 1–2 mm, allowing for more precise localization of accessory pathways and reproducible targeting of an identical point within the heart. This technology cannot only increase the success rate and safety of the procedure but eliminate or decrease the fluoroscopic time and radiation dose. In patients with paraHisian pathways, accurate mapping of the AV node and accessory pathways using a 3-D system can allow for ablation with careful and consistent localization of the AV node and the accessory pathway (Fig. 4.15). For example, using the 3-D mapping system to map the AV node during ventricular pacing, then mapping the accessory pathway during adenosine infusion (blocking the retrograde conduction through the AV node) may facilitate a more precise location of the site of the accessory pathway, especially in those with initial failure. In patients who have congenital heart disease and AV tachycardia, 3-D mapping techniques can be used to better determine the complex anatomy and location of the His-Purkinje system, which can be

displaced secondary to the congenital defect or previous surgery.

Care should be taken when applying radiofrequency energy during SVT as the ablation catheter position on the AV ring can shift abruptly upon termination of the tachycardia. A technique to overcome this shift is to map the location of the accessory pathway during SVT, place the ablation catheter at that site, then entrain the tachycardia circuit by pacing the ventricle at a slightly faster rate than the SVT. This technique engages the pathway and because the ventricular pacing continues throughout the energy application, it prevents an abrupt shift of the catheter position away from the optimal site for ablation at the moment of successful ablation. Temperatures greater than 50 °C are desired. However, temperatures as low as 48 °C have been shown to cause irreversible damage. Seldom is it necessary to use temperatures greater than 65 °C. It is important to recognize that accessory pathways are in close proximity to other important structures (AV valve, coronary artery, coronary vein); therefore, excess temperature and time of energy radiofrequency application should be weighed against collateral injury. If success of radiofrequency does not occur within 10 s of the radiofrequency application, then the energy is turned off. Successful ablation within the 10 s frame usually leads to a full 30–60 s application (Chap. 22).

The advent of cryothermal ablation has added another energy source for ablation (Chap. 22). Cryothermal ablation affords catheter stability during ablation as the catheter adheres (frozen) to the tissue, even during SVT. Additionally, damage to surrounding structures has been demonstrated to be minimal during cryothermal ablation, including the AV node and coronary arteries. Although the surrounding regions of the site of ablation also cool, tissue injury has been found to be reversible. During ablation of accessory pathways, careful monitoring of the PR interval and ST segments can prevent permanent injury if the cryo-application is terminated promptly. However, cryothermal ablation has also been felt to have a slightly higher recurrence compared to radiofrequency ablation. Several recommendations have been made to reduce recurrence. Use of a larger tip catheter (i.e., 6 mm vs. 8 mm) has may reduce recurrence. Elimination of pathway conduction early during the cooling process (less than 20–30 s after initiation and at warmer temperatures) suggests probable success. Lesion placement such as a freeze–thaw–freeze technique can also help in permanent pathway elimination. Finally, extensive testing post ablation to monitor for recurrence (at least 45 min post first successful lesion) may reduce the risk of recurrence.

The success rate for accessory pathway ablation is now between 90 and 95 % with the highest success rates found in patients having left-sided accessory pathways. Patients with paraHisian pathways have the lowest success rate, understandably due to more cautious ablation therapy applications in an effort to avoid AV block. Data from the pediatric radiofrequency ablation registry prior to adoption of cryotherapy by many centers recorded a risk of heart block at 1.2 % with a risk as high as 10.4 % for patients with mid-septal ablation site.

The overall risk of complications in patients undergoing an ablation is reported to be between 3 and 4 %, with the most common complication being a hematoma. The risk of early death has been reported at 0.05 % with the risk of late death between 0.07 and 0.18 %. However, it should be noted that these data were primarily obtained in the earlier era (1991–2004) of conventional mapping using fluoroscopy and only radiofrequency energy.

Summary

AVRT is a common form of reentrant arrhythmias, commonly diagnosed in the newborn period. Spontaneous resolution of SVT after 1 year of age in patients with preexcitation or who have AVRT is uncommon. Observation and medical management play a role in the care of very young patients. However, transcatheter ablation offers the advantage of a lifelong cure in older patients, or in younger patients with poorly tolerated, medication refractory tachycardia. Three-dimensional mapping systems have been a major

advance in the management of children with AVRT, providing more precise pathway location data and reducing the exposure to ionizing radiation. Continued advances in technology will allow for continued improvement in outcome for young patients with AVRT.

Suggested Reading

Anderson RH, Becker AE, Arnold R, Wilkinson JL. The conducting tissues in congenitally corrected transposition. Circulation. 1974;50:911–23.

Bink-Boelkens MT. Pharmacologic management of arrhythmias. Pediatr Cardiol. 2000;21:508–15.

Bokenkamp R, Wibbelt G, Sturm M, et al. Effects of intracardiac radiofrequency current application on coronary artery vessels in young pigs. J Cardiovasc Electrophysiol. 2000;11:565–71.

Bricker JT, Porter CJ, Garson A, et al. Exercise testing in children with Wolff-Parkinson-White syndrome. Am J Cardiol. 1985;55:1001–4.

Bromberg BI, Lindsay BD, Cain ME, Cox JL. Impact of clinical history and electrophysiologic characterization of accessory pathways on management strategies to reduce sudden death among children with Wolff-Parkinson-White syndrome. J Am Coll Cardiol. 1996;27:690–5.

Chiang CE, Chen SA, Teo WS, et al. An accurate stepwise electrocardiographic algorithm for localization of accessory pathways in patients with Wolff-Parkinson-White syndrome from a comprehensive analysis of delta waves and R/S ratio during sinus rhythm. Am J Cardiol. 1995;76:40–6.

Erickson CC, Walsh EP, Triedman JK, Saul JP. Efficacy and safety of radiofrequency ablation in infants and young children <18 months of age. Am J Cardiol. 1994;74:944–7.

Fitzpatrick AP, Gonzales RP, Lesh MD, et al. New algorithm for the localization of accessory atrioventricular connections using a baseline electrocardiogram. J Am Coll Cardiol. 1994;23:107–16 [Erratum appears in J Am Coll Cardiol. 1994;23(5):1272].

Giardina AC, Ehlers KH, Engle MA. Wolff-Parkinson-White syndrome in infants and children. A long-term follow-up study. Br Heart J. 1972;34:839–46.

Hahurij ND, Gittenberger-De Groot AC, Kolditz DP, Bökenkamp R, Schalij MJ, Poelmann RE, Blom NA. Accessory atrioventricular myocardial connections in the developing human heart: relevance for perinatal supraventricular tachycardias. Circulation. 2008;117(22):2850–8.

Haissaguerre M, Cauchemez B, Marcus F, et al. Characteristics of the ventricular insertion sites of accessory pathways with anterograde decremental conduction properties. Circulation. 1995;91:1077–85.

Iturralde P, Araya-Gomez V, Colin L, Kershenovich S, de Micheli A, Gonzalez-Hermosillo JA. A new ECG algorithm for the localization of accessory pathways using only the polarity of the QRS complex. J Electrocardiol. 1996;29(4):289–99.

Ko JK, Deal BJ, Strasburger JF, Benson Jr DW. Supraventricular tachycardia mechanisms and their age distribution in pediatric patients. Am J Cardiol. 1992;69:1028–32.

Kugler JD, Danford DA, Deal BJ, The Pediatric Electrophysiology Society, et al. Radiofrequency catheter ablation for tachyarrhythmias in children and adolescents. N Engl J Med. 1994;330:1481–7.

Kugler JD, Danford DA, Houston K, Felix G. Radiofrequency catheter ablation for paroxysmal supraventricular tachycardia in children and adolescents without structural heart disease. Pediatric EP Society, Radiofrequency Catheter Ablation Registry. Am J Cardiol. 1997;80:1438–43.

Law IH, Fischbach PS, LeRoy S, Dick M. Access to the left atrium for delivery of radiofrequency ablation in young patients: retrograde aortic vs. transseptal approach. Pediatr Cardiol. 2001;22:204–9.

Lundberg A. Paroxysmal atrial tachycardia in infancy: long-term follow-up study of 49 subjects. Pediatrics. 1982;70:638–42.

Mahaim I, Benatt A. Nouvelles recherches sur les connexions superieures de la branche gauche du faisceau de His-Tawara avec la cloison interventriculaire. Cardiologia. 1938;1:61–73.

Mandapati R, Berul CI, Triedman KJ, et al. Radiofrequency catheter ablation of septal accessory pathways in the pediatric age group. Am J Cardiol. 2003;92(8):947–50.

Miles WM, Yee R, Klein GJ, et al. The preexcitation index: an aid in determining the mechanism of supraventricular tachycardia and localizing accessory pathways. Circulation. 1986;74:493–500.

Munger TM, Packer DL, Hammill SC, et al. A population study of the natural history of Wolff-Parkinson-White syndrome in Olmsted County, Minnesota, 1953–1989. Circulation. 1993;87:866–73.

O'Connor BK, Dick M. What every pediatrician should know about supraventricular tachycardia. Pediatr Ann. 1991;20(368):371–6.

Pappone C, Santinelli V, Manguso F, et al. A randomized study of prophylactic catheter ablation in asymptomatic patients with the Wolff-Parkinson-White syndrome. N Engl J Med. 2003a;349(19):1803–11.

Pappone C, Santinelli V, Rosanio S, Vicedomini G, et al. Usefulness of invasive electrophysiologic testing to stratify the risk of arrhythmic events in asymptomatic patients with Wolff-Parkinson-White pattern: results from a large prospective long-term follow-up study. J Am Coll Cardiol. 2003b;41:239–44.

Pappone C, Manguso F, Santinelli R, et al. Radiofrequency ablation in children with asymptomatic Wolff-Parkinson-White syndrome. N Engl J Med. 2004;351(12):1197–205.

Pediatric and Congenital Electrophysiology Society (PACES), Heart Rhythm Society (HRS), American College of Cardiology Foundation (ACCF), American Heart Association (AHA), American Academy of Pediatrics (AAP), Canadian Heart Rhythm Society (CHRS), Cohen MI, Triedman JK, Cannon BC, Davis AM, et al. PACES/HRS expert consensus statement on the management of the asymptomatic young patient with a Wolff-Parkinson-White (WPW, ventricular preexcitation) electrocardiographic pattern: Heart Rhythm. 2012;9(6):1006–24.

Riggs TW, Byrd JA, Weinhouse E. Recurrence risk of supraventricular tachycardia in pediatric patients. Cardiology. 1999;91:25–30.

Schaffer MS, Silka MJ, Ross BA, Kugler JD. Inadvertent atrioventricular block during radiofrequency catheter ablation. Results of the Pediatric Radiofrequency Ablation Registry. Pediatric Electrophysiology Society. Circulation. 1996;94:3214–20.

Scheinman MM, Wang YS, Van Hare GF, Lesh MD. Electrocardiographic and electrophysiologic characteristics of anterior, midseptal and right anterior free wall accessory pathways. J Am Coll Cardiol. 1992;20:1220–9.

Sternick EB, Timmermans C, Rodriguez LM, Wellens HJJ. Mahaim fiber: an atriofascicualr or long ventricular atrioventricular pathway. Heart Rhythm. 2004;1(6):724–7.

Weng KP, Wolff GS, Young ML. Multiple accessory pathways in pediatric patients with Wolff-Parkinson-White syndrome. Am J Cardiol. 2003;91:1178–83.

Wolff L, Parkinson J, White PD. Bundle-branch block with short P-R interval in healthy young people prone to paroxysmal tachycardia. Am Heart J. 1930; 10:685–704.