Clinical Electrophysiology of the Cardiac Conduction System

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Cardiac Electrophysiology in the Young

The electrophysiologic properties of the heart consist of impulse formation, conduction velocity, and refractoriness. These properties are evaluated in several components of the conduction system, including the sinoatrial (SA) node, atria, inter-nodal tracts, atrioventricular (AV) node, His bundle, bundle branches, Purkinje network, and ventricle by clinical electrophysiologic study.

The SA node, AV node, and His-Purkinje system exhibit automaticity—spontaneous depolarization dependent upon phase 4 depolarizing outward potassium current. Atrial and ventricular tissues typically develop automaticity only during pathological conditions such as ischemia. This activity is rate variable, relative to the site of origin; the SA node has the fastest spontaneous discharge rate (140–160 bpm at birth, decreasing with age), and thus is the dominant pacemaker in the normal heart. A progressive decrease in spontaneous impulse formation distinguishes different cardiac excitable tissues as the pacemaker moves from atrium to AV node to His-Purkinje system, and lastly to ventricular myocardium. When diastolic depolarization of the dominant pacemaker reaches threshold, it initiates cell-to-cell propagation (conduction), generating an excitation wave front through ordinary atrial and ventricular myocardium and the specialized conduction tissue of the AV node and His-Purkinje system. The rate of normal automaticity is a complex process modulated by circulating catecholamines, the autonomic nervous system, and pathophysiologic state of the myocardium. Autonomic tone varies with age. The newborn and infant are highly susceptible to excessive parasympathetic input that can be aggravated by analgesia, anesthetics, or manipulation of the airway, resulting in a deleterious slowing of the heart rate. Beyond the first several months of life, until adolescence, autonomic tone is fairly stable and well balanced.

The intra-atrial conduction time is a function of the size and state (fibrosis, scars, suture lines, electrical remodeling) of the atria. The AV node displays decremental conduction: conduction

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through the AV node prolongs as the input stimulus interval shortens. AV node conduction time is highly influenced by the basic metabolic state, vagal tone, circulating catecholamines, and drug administration. Conduction through the His-Purkinje system is age dependent.

The natural history of arrhythmias is, in large part, dependent on the underlying structural and functional changes in the developing child. Accessory pathway (AP)-mediated tachycardia is the most common tachyarrhythmia occurring in infants and young children. Atrial fibrillation (AFib) and atrial flutter (AFL) are rare in the infant and small child due to insufficient cardiac mass to support the microreentry or macroreentry circuits which sustain this tachyarrhythmia, respectively. On the other hand, when anatomic conditions exist, such as dilated atria providing a circuit of sufficient length, AFib or AFL may appear, even in infants. By approximately 5 years of age, increased heart size and longer refractory periods increase the potential for reentry tachyarrhythmias. During adolescence, the predominate tachyarrhythmia shifts to AV nodal reentry tachycardia, likely reflecting developmental changes in AV node physiology and the autonomic nervous system. Automatic and triggered arrhythmias are less clearly age dependent, though multifocal atrial tachycardia and junctional ectopic tachycardia are, in general, arrhythmias of infants. Ventricular tachycardias unrelated to surgery span all age groups.

The Electrophysiology Study

Myocardial electrical activity can be recorded using electrodes on catheters placed within the heart (Fig. 3.1), and displayed as unipolar or bipolar electrograms (Fig. 3.2) which provide information on the timing of local electrical activation.

Unipolar electrograms display the electrical activation relative to a single electrode using a distant "indifferent" electrode (Wilson's central terminal or a remote electrode) as the opposite pole. The unipolar electrogram provides information on the local activation based on signal polarity, but is disadvantaged because they contain additional far-field signal. Bipolar electrograms are more commonly used. They are recorded between two adjacent electrodes (interelectrode distance is 1–5 mm), thereby minimizing much of the far-field signal and providing more precise information on the timing of local activation. Thus, by virtue of this proximity

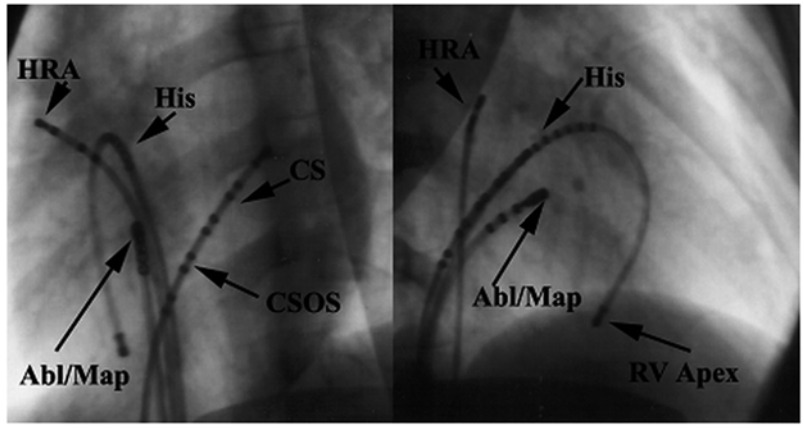

Fig. 3.1 Chest radiographs in two projections (*left panel*: left anterior oblique; *right panel*: right anterior oblique) demonstrating placement of four bipolar electrode pair electrodes; one each in the high right atrium (HRA), coronary sinus (CS), His bundle region (His), and right ventricular apex (RV apex); the last two sites are recorded through a single catheter. The mapping catheter (Abl/Map) is in the mid septal region in the triangle of Koch. The CS catheter in the right anterior oblique projection is superimposed on and obscured by the His catheter

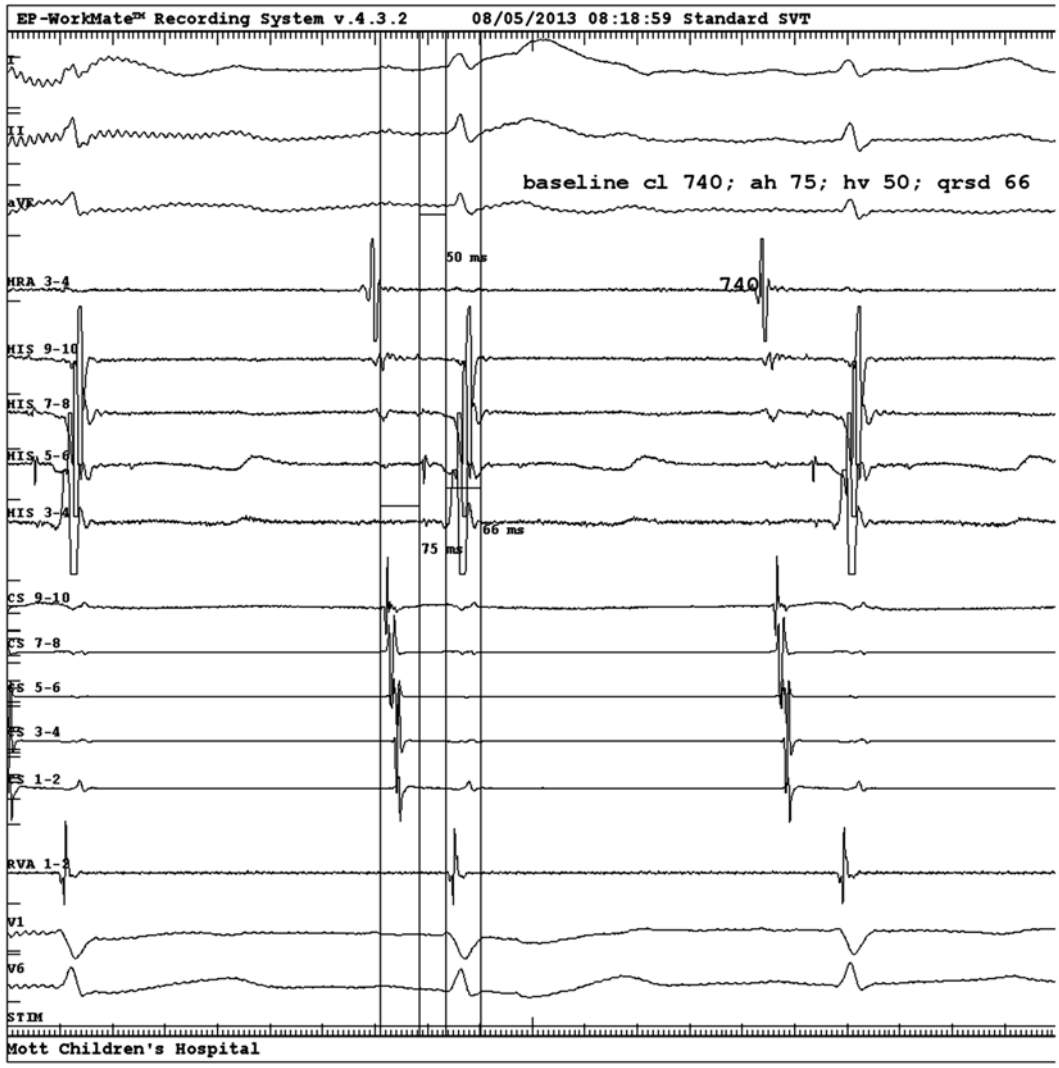

Fig. 3.2 Baseline electrograms from EPS. From *top* to *bottom*: surface ECG Leads I, II, aVF; HRA, His electrograms proximal (9–10) to distal (3, 4), CS proximal (9, 10) to distal (1, 2), RV apex, surface V1 and V6. Measured intervals as displayed are sinus cycle length 740 ms, AH interval 75 ms, HV interval 50 ms, QRS duration 66 ms. *HRA* high right atrium, *CS* coronary sinus, *RV* right ventricle

effect, the electrical activity of localized areas of the atrial and ventricular myocardium, and the His bundle potential can be recorded with intracardiac electrode pairs.

Sedation/Anesthesia

Pediatric patients and most adult patients will require sedation or general anesthesia for a typical catheter ablation procedure due to case duration. The preference for general anesthesia or moderate to deep sedation tends to be center or substrate dependent. We utilize general anesthesia almost exclusively except for substrates which may be repressed by general anesthesia. Modifications in the procedural regulations over the past decade have dictated at many centers that a dedicated physician be in charge of patient sedation. For our own center, this has been the

pediatric anesthesia service. General anesthesia allows for complete control of patient movements during ablation but requires intubation and may result in post-procedure nausea and longer recovery. General anesthesia may also act to suppress certain arrhythmia substrates—focal atrial tachycardia and atrioventricular reentrant tachycardia AVNRT due to effects of gas anesthesia on electrophysiology. For these reasons, moderate sedation may be preferable in some circumstances.

Access

The Electrophysiology Study (EPS) begins with access to the intravascular space. The chosen sites of access are operator dependent. Access is obtained using modified Seldinger technique to place valved sheaths into the vein allowing easy catheter exchange. In our laboratory, with rare exception, all access is via the femoral veins; typically, two sites on each side to accommodate three diagnostic and one ablation catheter. Many labs, however, routinely utilize jugular venous access typically for coronary sinus catheter placement. For those patients with limited femoral access, not uncommon in congenital heart disease patients, an alternative access point is a transhepatic sheath (Fig. 3.3). This typically will require interventional cardiologist involvement as coil closure of the transhepatic tract is recommended to decrease the risk of bleeding complications.

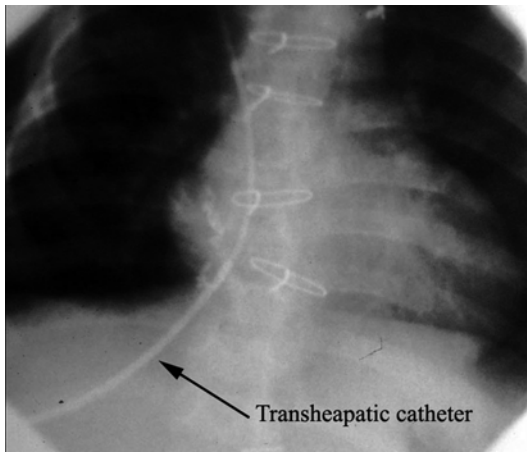

Fig. 3.3 Passage of transhepatic sheath and catheter through the liver and into the right atrium and superior vena cava in an 8-kg infant

Arterial cannulation for monitoring of systemic blood pressure can be used selectively. For most patients with supraventricular tachycardia, monitoring of the blood pressure by sphygmomanometer is sufficient, avoiding the need for an intra-arterial cannula. In patients with poor ventricular function, suspected ventricular arrhythmias, infants (≤15 kg), or patients with adult congenital heart disease, the use of direct intra-arterial blood pressure monitoring may be desirable.

Diagnostic Catheters and Placement

A variety of specially designed diagnostic catheters exist to record electrograms from varied or specific locations within the heart (Fig. 3.4). Positions are strategically chosen to allow for differentiation of the electrical activation in the heart in order to identify the origin and direction of electrical propagation. The routine sites for a typical EPS include the right atrium (RA), right ventricle (RV), coronary sinus (CS), and bundle of His (Fig. 3.2). A bipolar or quadripolar catheter suffices for the RA and RV sites. The CS catheter is usually an octapolar or decapolar catheter which allows for differentiation of the timing of left atrial activation from the CS os (proximal) to the distal CS along the lateral rim of the mitral valve annulus. The His bundle potential can be recorded by placing a catheter across the tricuspid valve annulus at the superior septum. Specially formed catheters (Josephson curve or St. Jude CRd2) may improve stability and quality of the recorded His potential. In our own lab, a combined His/RV catheter (Fig. 3.4) is preferred to decrease the number of access sites. This catheter has two distal electrodes placed in the RV and an array of proximal electrodes which sit at the His bundle. We primarily use deflectable His/RV catheters with a 30 mm His–RV separation or a 50 mm His–RV separation.

Additionally, a quadripolar or pentapolar esophageal catheter (Fig. 3.4) may be added

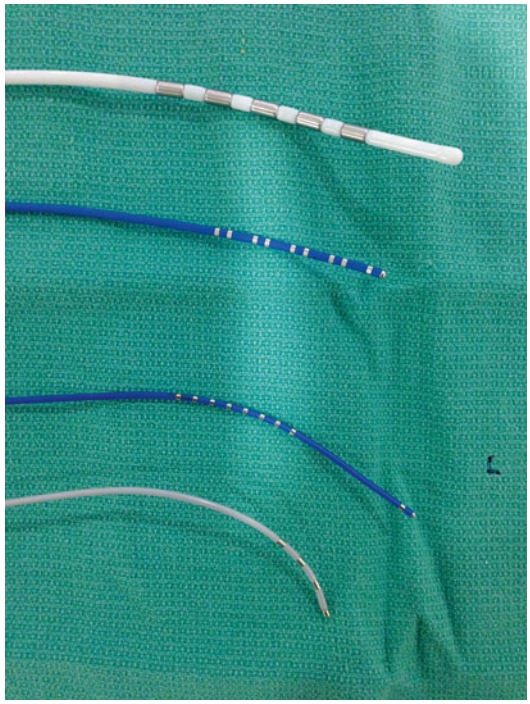

Fig. 3.4 Diagnostic catheters: Several types of diagnostic catheters used in an electrophysiology study. From *top to bottom*: Pentapolar esophageal catheter (*top catheter*). Decapolar deflectable catheter with 2–5–2 mm spacing (6Fr, *second catheter*). Deflectable decapolar His/RV combination catheter with distal bipolar pair of electrodes and a proximal array for His recording (*third catheter*). Josephson curve quadripolar catheter for multipurpose use (*bottom catheter*)

to provide an atrial pacing and recording site while potentially decreased the need for additional venous access. The esophageal catheter also functions well as a reference site for activation mapping as its position tends to be stable and easily confirmable with fluoroscopy if needed; in the infant, it can serve to deliver atrial extrastimulation.

Catheter Navigation

Visualization of the diagnostic and ablation catheters has traditionally been with fluoroscopy. Evolution of computerized 3D electro-anatomic mapping systems over the past decade has led to its nearly exclusive method for catheter navigation in our own and many pediatric laboratories across the country (also see Chap. 23).

The most commonly used systems are Ensite (St. Jude Inc., St. Paul, MN) (Fig. 3.5) and CARTO (Biosense-Webster, Diamond-Bar, CA) (Fig. 3.6). We currently use both Ensite Velocity and CARTO3 with approximately equal use between the two systems. Choice of system is primarily by attending preference and secondarily by arrhythmia substrate or patient characteristics. The Ensite system allows for visualization and placement of all diagnostic catheters prior to choosing an ablation catheter. Any radiofrequency or cryocatheter is compatible with the Ensite system. The CARTO system requires a proprietary catheter to create an endocardial geometry prior to additional catheter placement. Proprietary radiofrequency catheters must be used for ablation, though cryocatheters can be used with minor connection adjustments.

Specific Considerations for Pediatrics and Congenital Heart Disease

The EPS the infant or small child requires special considerations. First, the operator should be experienced in advancing and navigating an electrode catheter through the cardiac chambers of young patients. The operator should be fully informed regarding the clinical course and structural features of concomitant cardiac malformations. Structural congenital heart lesions affect the location of the SA and AV nodes and specialized conduction tissue. Operations for these congenital heart lesions can further alter the electrical properties of the atrial and ventricular myocardium, resulting in complex rhythm disturbances.

The risk of catheter ablation in pediatric patients was shown to carry increased, though not prohibitive, risk in children less than 5 years old or less than 15 kg. The enhanced risk includes difficulty of vascular and intracardiac access, alterations in hemodynamic state, potential perforation and pericardial effusion with tamponade, potential for excessive lesion size (and possible expansion), and injury to the AV node. Between

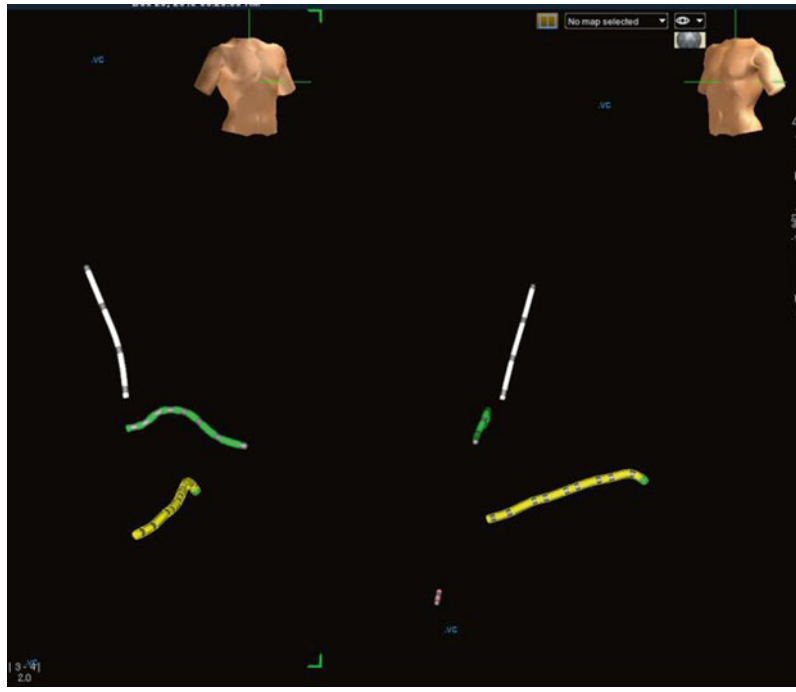

Fig. 3.5 Monitor image of Ensite Velocity electroanatomic mapping system as used for catheter navigation in routine electrophysiology study. *Left-side* is an RAO view as demonstrated by torso in upper screen. *Right side* is LAO view. *Yellow catheter* is a decapolar catheter placed in the coronary sinus. *Green catheter* is the His array of a His/RV combo catheter. RV apex electrodes are in *red* in the LAO view only. *White* catheter is a quadripolar high right atrial catheter

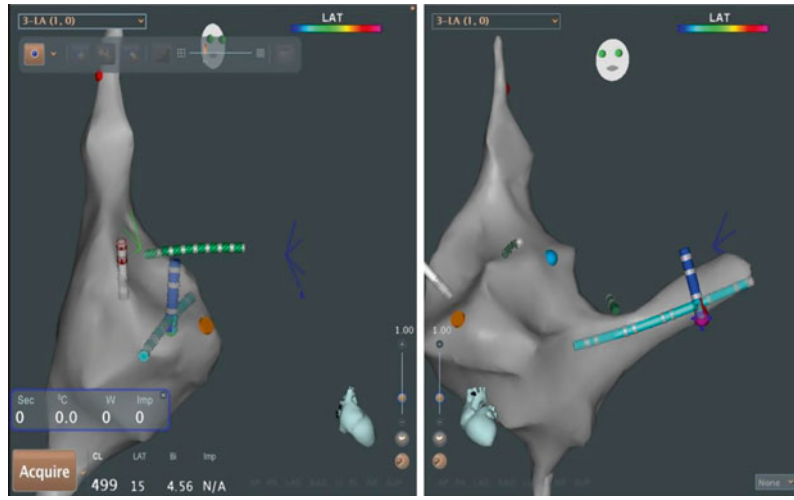

Fig. 3.6 Monitor image from CARTO3 electroanatomic mapping system as used for catheter navigation during electrophysiology study. *Left-hand panel* is RAO view as indicated by "face" position at top of screen. *Right-hand panel* is LAO view. *Aqua catheter* is decapolar catheter placed in the coronary sinus. *Green catheter* in the RAO view is the His array of a His/RV combo catheter. The *blue dot* in the LAO view was the location of a His potential. The *dark blue catheter* with *purple tip* in the LAO view is a radiofrequency catheter positioned to ablate a left lateral accessory pathway

15 and 35 kg, the risks are diminished and beyond 35–50 kg, the risk of electrophysiologic study is essentially equivalent to that in adult patients.

When considering electrophysiologic study in the small infant or child, the number of catheters, the size of the catheters, and the stiffness of the catheters should be minimized. Because of the variable dimension of the cardiac chambers in different size children, steerable catheters with different shapes and different sized curves are necessary to appropriately accommodate each patient. Adolescents are usually of sufficient size to allow use of catheters and sheaths of the length and caliber of those used in adults. However, smaller children require smaller catheters and sheaths, especially in the diameter of the operator-controlled curve. Typically, for a child 20 kg or less, diagnostic catheters should not exceed 6 French; though 7Fr ablation catheters may occasionally be necessary. For an infant under 10 kg, 4 and 5 French catheters are used including the mapping/ablation catheter; although, again 7Fr RF and cryocatheters may be required for effective ablation. Additionally, the esophageal catheter can be a useful addition to the EPS in a small patient.

Due to the vigorous contractile state and the faster heart rates in small children compared to adults, both during sinus rhythm and tachycardia, catheter stability is at risk. There is always a trade-off between catheter size (and stiffness) and catheter stability. The combined His-RV catheter adds stability to the His catheter since it is, in part, secured by the placement of the catheter tip in the RV apex. Multiple catheters produce artifacts when the electrodes from different catheters strike one another, a confounder increased in the smaller volume of a child's heart ($\leq$20 kg).

Intracardiac Electrophysiology

The purpose of the electrophysiology study is to assess the basic electrophysiologic properties of the patient's cardiac conduction system and to determine the mechanisms of the suspected or clinically demonstrated cardiac arrhythmia. The study begins with the measurement of baseline intervals (Table 3.1). The P-A interval denotes the intra-atrial conduction time. The A-H interval assesses the conduction time through the AV node. The H-V interval marks the conduction time through the His-Purkinje system. This interval is fairly constant but does increase slightly from approximately 25 ± 10 to 45 ± 10 ms from infancy to adulthood. An H-V interval greater then 60 ms in the young is abnormal and suggests an abnormality in the His-Purkinje system. It is unusual in the child, even following surgery for congenital heart disease, to have a prolonged H-V interval. Top normal H-V intervals are not uncommon, however, in patients with AV septal defects (AV canal defects) and older postoperative tetralogy of Fallot and ventricular septal defect patients.

Table 3.1 Conduction intervals

Interval	Measurement (normal values)
P-A	Atrial conduction time—beginning from the earliest P-wave on the ECG to the earliest fast atrial deflection on the low septal right atrial electrogram
A-H	AV nodal conduction time—from earliest fast atrial deflection on the annular septal right atrial electrogram to the beginning of the His bundle potential recorded on the His bundle electrogram. Varies with heart rate and autonomic tone
H-V	His-Purkinje conduction time—from beginning of the His bundle potential recorded on the His bundle electrogram to the onset of the surface QRS or earlier ventricular electrogram

In the past, evaluation of the SA node was a routine part of the EPS, though is now seldom performed. SA node function is tested by measuring the SA node recovery time (SNRT) and corrected SA node recovery time (CSNRT). A fixed train (usually 30 s) of external stimuli is delivered to the atrium. When atrial pacing is terminated, the recovery interval of the SA node is measured (i.e., time from the last pacing stimulus to the first spontaneous signal emerging from the SA node recorded as an atrial electrogram in the catheter that delivered the pacing stimuli). This procedure is performed at a pacing rate of 90,

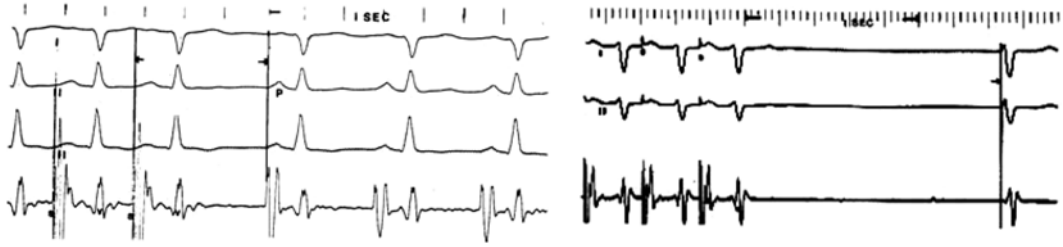

Fig. 3.7 Measurement of sinus node recovery time: *Left panel*: Atrial pacing at 400 ms (150 bpm). Upon termination of pacing, the normal recovery time of the SA node (P-wave) is 620 ms. Subtracting that interval from the basic sinus interval of this patient of 500 ms, yields a normal Maximal Corrected Sinus Node Recovery Time (MCSNRT) of 120 ms. *Right panel*: In contrast, cessation of pacing in a 15-year-old patient following the Mustard operation for transposition of the great arteries yields a junctional escape beat with an escape interval of 1,450 ms. Both the escape mechanism and the escape interval are abnormal

120, 150, 180, and 286 bpm. The maximal SA node recovery time (SNRT) is that maximal escape interval that follows termination of pacing at any pacing rate. The SNRT can be corrected for the normal spontaneous discharge rate by subtracting the basic cycle length (Fig. 3.7). To truly isolate the intrinsic automaticity of the SA node the autonomic nervous system must be attenuated using atropine and beta-blocker. Its clinical purpose was to evaluate an individual with suspected sick sinus syndrome (Chap. 13); this condition is effectively evaluated by surface ECG recordings such as the Holter 24 h dynamic ECG tracing.

Refractory periods are measured using programmed extrastimulation. A drive of 8–10 pacing stimuli are delivered at a constant interval (S1S1) followed by a premature stimulus delivered at a shorter interval (S1S2). This process is reiterated, sequentially shortening the premature beat (S2) until it fails to capture or propagate to the target tissue. This process yields the relative, functional, and effective refractory periods of the tissue in question (Tables 3.2 and 3.3). Because the AV node displays decremental conduction, several specific refractory periods can be measured (Table 3.3, displayed graphically in Fig. 3.8 and with intracardiac tracings in Fig. 3.9).

Following assessment of the electrophysiologic properties of the cardiac conduction system, induction of tachycardia and delineation of its mechanism(s) are performed. The arrhythmia classification is displayed in Fig. 3.10; all will be discussed in detail in the accompanying appropriate chapters. The great majority of tachyarrhythmias in children are due to a reentry mechanism (AVNRT, accessory pathway-mediated tachycardia). Induction of these arrhythmias typically requires a premature beat which is provided by the S1S2 programmed extrastimulation protocol described above. This produces unidirectional block in 1 limb of the circuit and delays conduction in the second limb, thereby initiating reentry. Tachycardia induction may not be as simple as a single premature atrial extrastimulus however; and other maneuvers may be required to induce tachycardia including burst atrial pacing, double or triple premature extrastimuli, alternate site for extrastimulus pacing and isoproterenol administration.

Diagnostic Electrophysiology

Whether or not the tachycardia can be initiated, diagnostic assessment and/or maneuvers are performed to identify the tachycardia mechanism. While there are numerous published techniques, the most commonly used by our laboratory will be outlined. The maneuvers are divided into those that are performed in the baseline state or during pacing and those that are performed during tachycardia. Because most pediatric arrhythmias have a reentry mechanism—AVNRT or AP mediated—most diagnostic pacing maneuvers are based on the concept of entrainment.

Table 3.2 Electrophysiologic data in children

	Group	<0.5 year	*N*	0.05–1 year	*N*	1–5 year	*N*	5–10 year	*N*	>10 year	*N*
Age		0.28	19	0.66	18	3.1	37	6.9	17	16.2	21
Range		0.01–0.18		0.52–0.98		1.0–4.9		5.1–9.9		10–25	
SCL ($x \pm SD$)	(ms)	456±50	19	471±66	18	547±37	37	610±83	17	814±132	21
Panel A: Conduction intervals ($x \pm SD$) (ms)											
	PA	24±7	10	22±14	12	29±15	30	29±12	11	29±14	13
	AH	72±14	11	75±14	14	71±20	30	75±18	12	91±21	14
	HV	33±9	11	35±4	14	36±8	30	34±6	13	41±8	17
	RVA	24±8	5	23±11	11	26±5	22	23±7	10	29±10	18
	RVO	31±5	4	41±11	8	41±10	14	52±7	5	47±14	7
	RVI	33±5	4	41±11	9	47±16	16	48±9	3	47±5	5
Panel B: Refractory periods ($x \pm SD$) (ms)											
	AT. ERP	166±25	17	163±29	12	203±26	22	239±28	7	269±39	4
	AT. FRP	205±35	17	212±18	12	243±29	22	264±24	7	301±45	4
	AVCERP	231±24	15	238±40	4	244±44	8	375	1	–	
	AVCFRP	284±30	15	305±39	11	329±35	18	381±32	6	454±85	4
	VENT ER			226	1	228	2	250	1	225	1
Panel C: Sinus node ($x \pm SD$) (ms)											
	TSACT	111±28	17	107±23	10	123±36	17	153±38	6	120±62	2
	MCSNRT	123±44	18	104±39	10	127±50	23	163±47	7	<250	

Notes: Adapted from Campbell RM, Dick M, Rosenthal A. Cardiac arrhythmias in children. Ann Rev Med 1984; 35:397–410

Abbreviations: *AT. ERP* atrial effective refractory period, *AT. FRP* atrial functional refractory period, *AH* atrial-His bundle interval, *AVCERP* atrioventricular conduction system effective refractory period, *AVCFRP* atrioventricular conduction functional refractory period, *HV* His bundle-ventricle interval, *MCSNRT* maximal corrected sinus node recovery time, *PA* high right atrial to low right atrial interval, *RVA* right ventricular apical activation time, *RVI* right ventricular inflow activation time, *RVO* right ventricular outflow activation time, *TSACT* total sinoatrial conduction time, *VERP* ventricular effective refractory period

Table 3.3 Refractory periods

Refractory period	Definition
Effective refractory period (ERP)	The longest input interval (S1-S2) that fails to depolarize (propagate to) the tissue in question
Functional refractory period (FRP)	The shortest output interval (e.g., H1-H2 or V1-V2) that propagates to the tissue in question
Relative refractory period (RRP)	The shortest output interval (e.g., H1-H2 or V1-V2) equal to the input interval (e.g., A1-A)

Diagnostic Evidence and Maneuvers Performed While Not in Tachycardia

The surface QRS morphology during sinus rhythm should be assessed for evidence of ventricular preexcitation confirmed by measurement of a short H-V interval (Fig. 3.11; also see Chap. 4, Figs. 3.2 and 3.3)).

Dual AV node physiology (DAVNP) is characterized by a "jump" in the AH interval when delivering incrementally shorter premature atrial extrastimuli (S2) (Fig. 3.12; Chap. 5). DAVNP is defined as an increase in the AH interval of at least 50 ms after a decrease in the S2 interval of 10 ms. Alternatively, a second method of diagnosing DAVNP has been identified in pediatric patients exclusively—rapid atrial pacing at progressively shorter pacing intervals which produces 1:1 conduction to the ventricle with the paced PR interval>paced RR interval. While DAVNP is typically associated with AVNRT, the finding itself is not particularly sensitive or specific. Approximately 30 % of pediatric patients

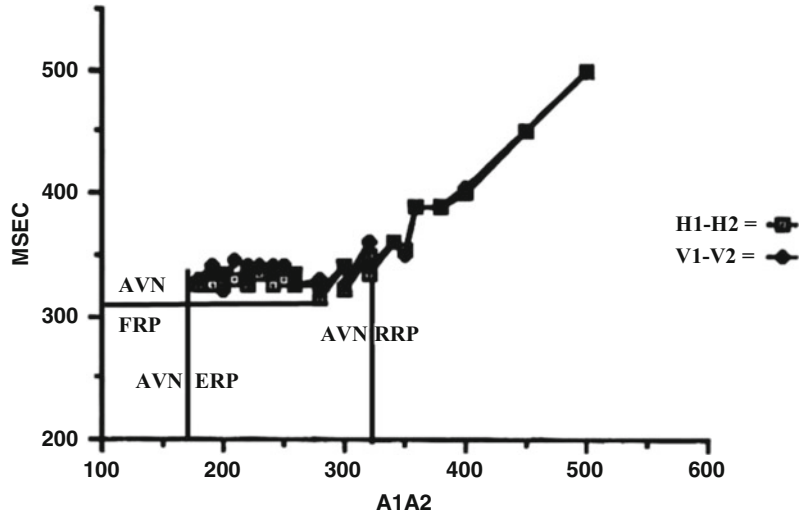

Fig. 3.8 Plot of the H1H2 and V1V2 intervals (*y*-axis) as the atrial premature interval (A1A2) is incrementally shortened (*x*-axis). Shown are the atrioventricular node relative refractory period AVRRP, the atrioventricular node effective refractory period (AVNERP) and the atrioventricular node functional refractory period (AVNFRP). The refractory periods are defined in Table 3.3

will have DAVNP without AVNRT. Retrograde DAVNP may also be demonstrated but appears to be much less specific for identifying patients with AVNRT.

A*trial activation pattern* is assessed during ventricular pacing and determined to be concentric (originating from the atrial septum) or eccentric (not originating from the septum) (Fig. 3.13). Normal retrograde conduction through the AV node produces concentric atrial activation. Left inferior or lateral APs will show eccentric activation early in the mid-distal CS electrograms. Right lateral APs will be early on the lateral tricuspid valve annulus provided the mapping catheter or the HRA catheter has been placed near this location; if the HRA catheter is high in the atrium in the absence of a mapping catheter, the earliest atrial activation for a right lateral AP will appear at the His or proximal CS and may mimic concentric activation. Concentric atrial activation should be further assessed with premature ventricular extrastimuli in order to identify decremental retrograde conduction which would suggest the conduction is via the AV node as opposed to a septal AP.

Base-apex pacing also can help in differentiating AP versus AVN conduction and is performed by delivering extrastimuli at a constant rate to the ventricle near the annulus (base) and measuring the stimulus-A interval, then pacing from the apex at the same rate and measuring the stimulus-A interval (Fig. 3.14). Because the His-Purkinje system terminates near the apex, an apically delivered extrastimuli is expected to enter the His-Purkinje system and conduct retrograde to the atria via the AV node. If there is not an AP, the stimulus-A time from the base should be longer than that from the apex, as the activation must travel through the ventricular muscle to the apex first and then enter the HPS. In the presence of a septal AP, the activation needs only to conduct through the AP to the atria, hence the stimulus-A time should be shorter. Cautious interpretation is necessary if a decremental AP is present or there is retrograde dual AV node pathway (DAVNP).

ParaHisian pacing is performed by pacing from the His catheter with the goal of capturing the His bundle—indicated by producing a relatively narrow QRS (Fig. 3.15). High output pacing is delivered from the His catheter (assuring there

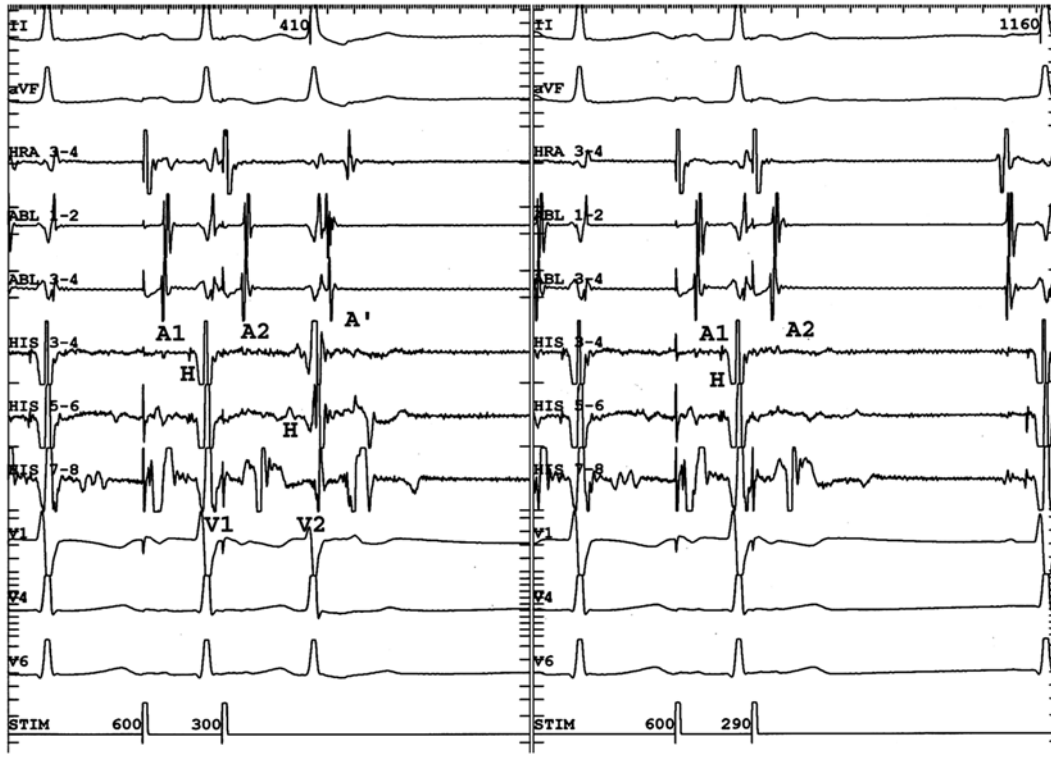

Fig. 3.9 AV node effective refractory period: Displayed from top to bottom: surface lead II, aVF, high right atrium, Ablation distal (1–2) to proximal (3–4), His distal (3–4) to proximal (7–8), surface leads V1, V4, and V6. Atrial extrastimulation yielding the effective refractory period of the AV node. *Left panel*: A premature atria stimulus (A2) is delivered at a coupling interval of 300 ms at a basic drive of 600 ms. Conduction through the AV node to the His bundle and the ventricle is intact. In addition, there is an echo beat (A′) earliest in the Abl 1–2 located in the coronary sinus. This last observation indicates a left-sided AP. *Right panel*: The coupling interval of the premature impulse (A2) was shortened to 290 ms with capture of atrial tissue and conduction block in the AV node (i.e., no His electrogram following the atrial electrogram), indicating the effective refractory period of the AV node

is no atrial capture) at a constant rate. The output is decreased gradually until a distinct increase in the surface QRS duration is noted—indicating that the His bundle is no longer being captured and ventricular activation is via local myocardial capture only. In the presence of a septal AP the stimulus-A interval is expected to be the same during both His capture and His non-capture. Without a septal AP the His captured stimulus-A interval will be shorter than the His non-captured stimulus-A as the latter must traverse the ventricle to the apex before entering the HPS and conducting retrograde via the AV node. Again, cautious interpretation is necessary in the presence of a decremental retrograde AP or retrograde DAVNP.

Adenosine may be delivered during sinus rhythm to produce AV node block and illicit ventricular activation down a manifest or latent AP. It may also be delivered during ventricular pacing for several purposes. Retrograde AV nodal conduction is expected to block and may uncover retrograde AP conduction (Fig. 3.16, also see Chap. 4, Fig. 3.12). Lack of retrograde AV block during adenosine administration is not absolutely indicative of the presence of an AP however, some patients with AVNRT may be resistant to the effects of adenosine on retrograde AV node conduction. For these patients, giving a higher dose of adenosine may produce retrograde AV node block. Additionally, rare APs

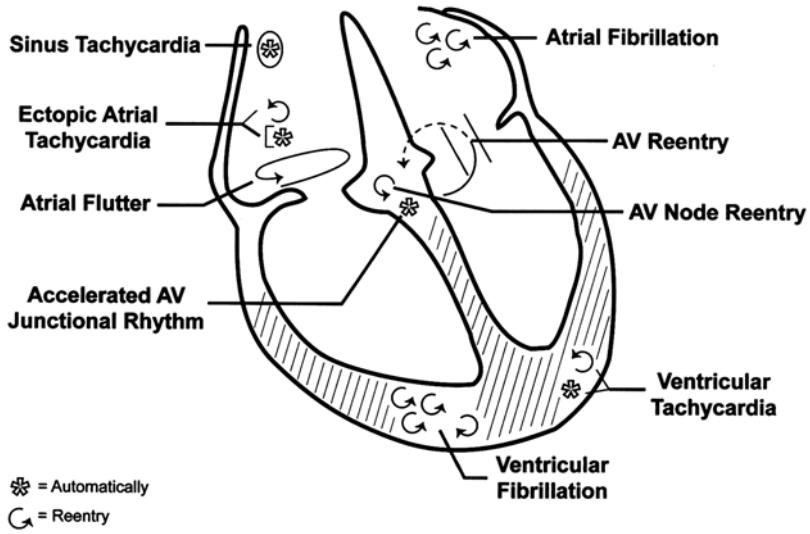

Fig. 3.10 Drawing of cardiac arrhythmias: their origin and their mechanism. Reentry arrhythmias include AP-mediated tachycardias (AV reentry) AV node reentry, atrial flutter, and some forms of ventricular tachycardia. Automatic arrhythmias consist of ectopic atrial tachycardia, accelerated AV junctional rhythm, and some forms of ventricular tachycardia (VT)

may be sensitive to adenosine, especially those displaying retrograde decremental or slow conduction. As a result of these caveats, the results of adenosine administration on retrograde AV node conduction should be interpreted along with other diagnostic evidence.

Diagnostic Evidence and Maneuvers During Tachyarrhythmia

The *surface QRS morphology* should be assessed for normality. Pediatric tachycardias tend to be narrow (or normal) complex or orthodromic tachycardias. However, preexcited tachycardias such as antidromic AP-mediated tachycardia or atrio-fascicular-mediated tachycardias are also possible and will demonstrate wide QRS morphology. Tachycardias may demonstrate rate-dependent bundle branch block (Ashman's phenomenon, aberrancy; see Chap. 13, Fig. 13.6).

The *atrial activation pattern* is assessed during 1:1 atrioventricular tachycardias. The site of earliest atrial activation should be noted and compared to atrial activation during ventricular pacing. Again, eccentric activation is differentiated from concentric activation to indicate AP-mediated tachycardia vs. AVNRT (see Chaps. 4 and 5).

A*tria to ventricular conduction* is 1:1 for all AP-mediated tachycardias and usually 1:1 for AVNRT (both block to the atrium and block to the ventricle is possible usually in a 1:2 or 2:1 pattern). Atrial tachycardia may be 1:1 or >1:1 if AV node block develops as in typical atrial flutter (Chap. 8); AV block essentially confirms atrial tachycardia except in the case of AVNRT with 2:1 block. Reciprocally, A-V conduction <1:1 should confirm ventricular tachycardia in the presence of wide complex tachycardia or junctional tachycardia if the QRS complex is narrow. Again, however, AVNRT with variable conduction to the atrium with or without bundle branch block may produce this pattern as well but is very rare.

The *ventricle to atria conduction time (VA time)* is an important clue to the tachycardia substrate and should be measured from the earliest onset of the surface QRS to the earliest intracardiac atrial electrogram. A VA interval <70 ms

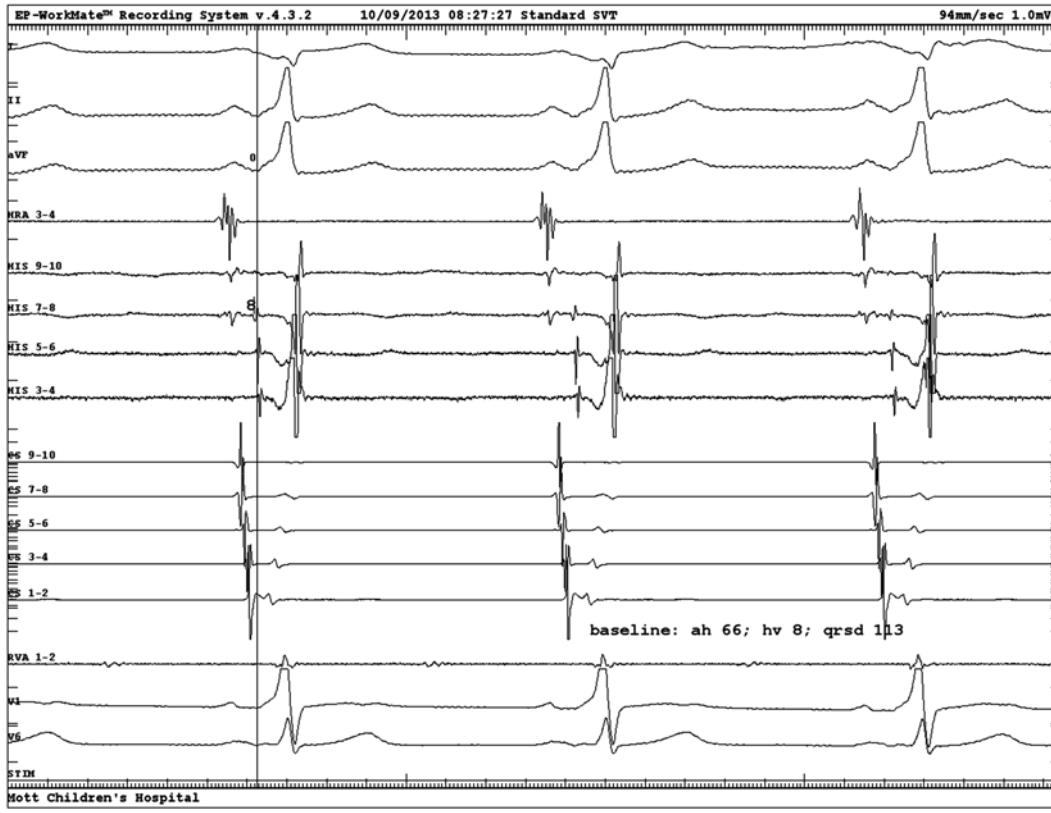

Fig. 3.11 Ventricular preexcitation: Intracardiac electrograms displayed from top to bottom as: surface leads I, II, aVF, HRA, His proximal (9–10) to distal (3–4), CS proximal (9–10) to distal (1–2), RV apex, surface leads V1 and V6. Baseline measures are shown with HV 8 ms. Horizontal line at first beat marks beginning of delta wave onset occurring simultaneous with His deflection. *HRA* high right atrium, *CS* coronary sinus, *RV* right ventricle

is diagnostic of typical AVNRT in adults and older adolescents (Fig. 3.17; Chap. 5). Younger patients, however, may have AP-mediated tachycardias with a VA time of 65 ms (Fig. 3.13, right panel). A VA time of less than 60 ms during inducible tachycardia is essentially diagnostic of typical AVNRT, though the less common junctional tachycardia may have a similar VA relationship. During AP-mediated tachycardia a change in the VA time associated with a rate-dependent bundle branch block may help localize the AP—a bundle branch block will increase the VA time through an ipsilateral AP (Coumel's phenomenon; Fig. 3.18, Chap. 4)

Entrainment is an important and useful concept and diagnostic maneuver that supports reentry as the mechanism of a tachycardia. Three criteria must be present to sustain a reentry: (1) a central obstacle for the circuit to navigate and (2) a zone of slow conduction which allows the rest of the circuit to repolarize on the return (reentry) of the impulse to the site of origin. A third criterion must occur in order to initiate a reentry tachycardia—unidirectional block in one limb of the circuit which allows for the circuit to progress in one direction. The central obstacle varies depending on the tachycardia type: the tricuspid valve annulus in typical atrial flutter, the fibrous annulus in AP-mediated tachycardias, scar in intra-atrial reentry tachycardia, and portions of the AV node and atrial septum in AVNRT. The slow zone varies as well: the cavo-tricuspid isthmus in typical atrial

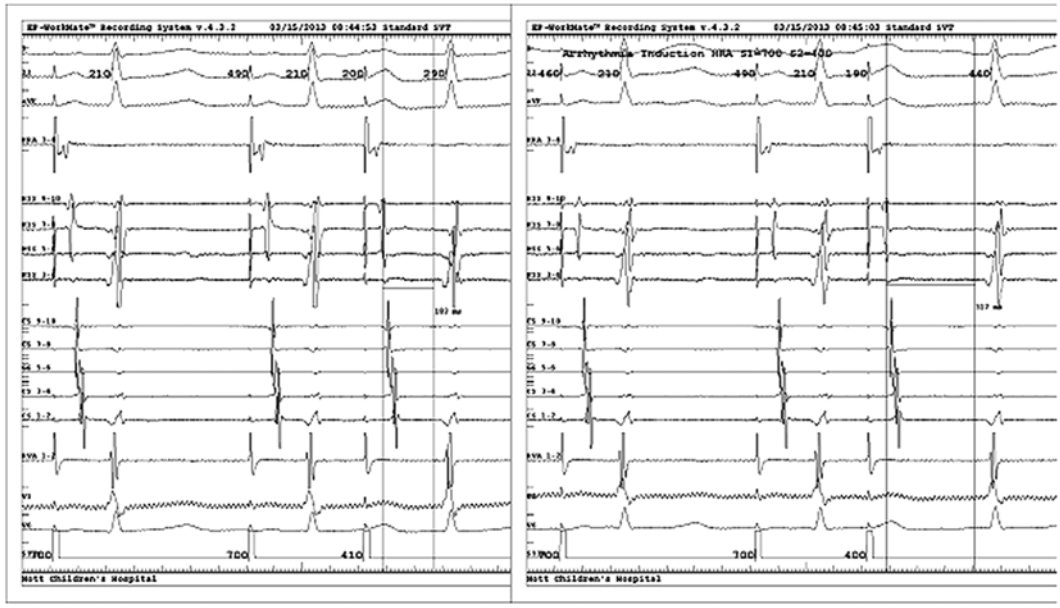

Fig. 3.12 Dual AV node physiology: Intracardiac electrograms displayed as: surface lead I, II, aVF, high right atrium, His proximal (9–10) to distal (3–4), CS proximal (9–10) to distal (1–2), RV apex, surface leads V1 and V6. *Left panel* shows premature atrial extrastimulus with S1S2 700/410 paced from the HRA. The AH interval following the premature extrastimulus is 183 ms. *Right panel* shows a premature atrial extrastimulus of S1S2 700/400 from the HRA. The AH interval has now "jumped" out to 317 ms. Important to the documentation of dual AV node physiology is that the AV node must continue to conduct with the longer AH as the premature extrastimulus shortens—700/390, 700/380, etc. *HRA* high right atrium, *CS* coronary sinus, *RV* right ventricle

flutter, the AV node its right and left extensions in AP-mediated tachycardias and AVNRT and scars in reentrant ventricular tachycardia.

Entrainment has been defined to mean: to adjust (an internal rhythm of an organism) so that it synchronizes with an external cycle, such as that of light and dark; in our case, the internal rhythm is the tachycardia and the external cycle is the pacing rate. Thus, entrainment is the delivery of programmed extrastimuli at a constant rate that is slightly faster than the tachycardia cycle length thereby transiently capturing the tachycardia mechanism by advancing the tachycardia to the paced cycle length Fig. 3.19) The delivered extrastimuli enter the tachycardia circuit in both the retrograde and antegrade directions—the retrograde activation collides with the prior wave front blocking it while the antegrade activation propagates and continues the tachycardia. As a practical example, an orthodromic AP-mediated tachycardia can be entrained from the RV. Assume a tachycardia cycle length of 300 ms. Pacing is delivered from the RV apical catheter at a cycle length of 280 ms. The paced wave front enters the HPS and propagates retrograde where it collides with the antegrade wave front of the tachycardia and blocks in the HPS. The paced wave front also activates the ventricle and traverses the AP thereby activating the atrium 20 ms earlier than the tachycardia (300 ms − 280 ms = 20 ms). With continued pacing at 280 ms, the atrial activation sequence would remain constant (IE: activated via retrograde conduction through the AP) but would occur every 280 ms instead of every 300 ms. Hence, the tachycardia would be entrained.

The post-pacing interval (PPI) is an important measure to determine whether or not the tip of the pacing catheter is positioned at a location

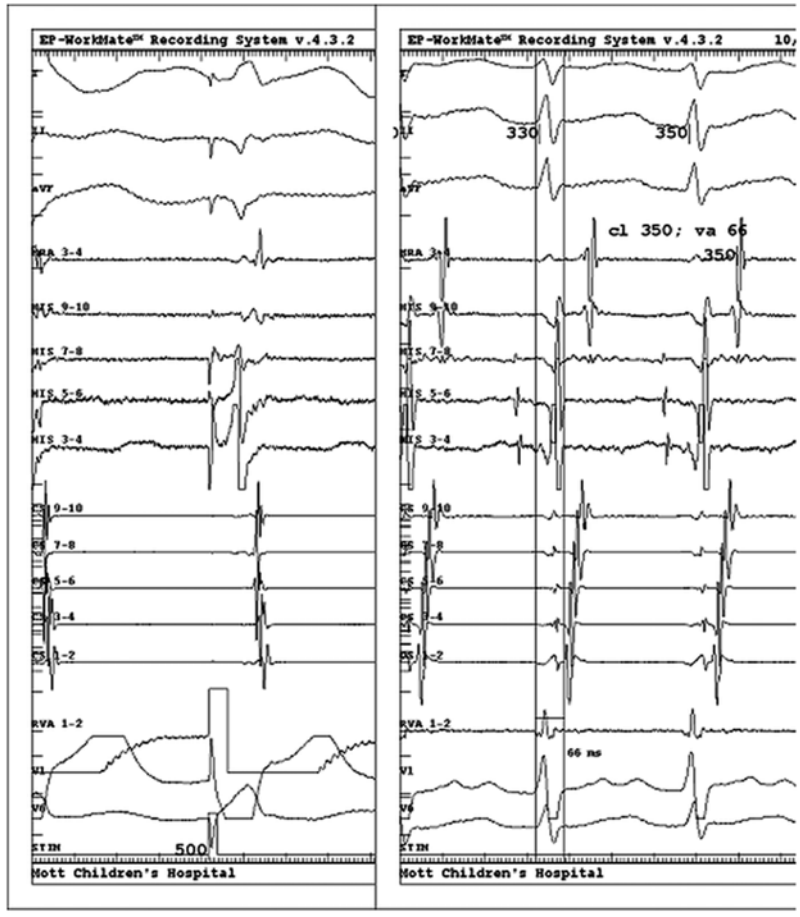

Fig. 3.13 Concentric vs. eccentric atrial activation: Intracardiac electrograms displayed as surface leads I, II, aVF, HRA, His proximal (9–10) to distal (3–4), CS proximal (9–10) to distal (1–2), RV apex, surface leads V1 and V6. *Left panel* shows ventricular extrastimulation from the RV apex. Concentric atrial activation is earliest in the His catheter and proceeds from proximal to distal in the CS catheter—consistent with retrograde atrial activation via a septal accessory pathway or the AV node. *Right panel* shows accessory pathway-mediated orthodromic tachycardia with VA time measured at 66 ms. There is eccentric atrial activation proceeding from distal CS to proximal CS consistent with retrograde activation via a left lateral accessory pathway. *HRA* high right atrium, *CS* coronary sinus, *RV* right ventricle

within the reentry circuit itself or is positioned outside the circuit in "passively" activated myocardium. The PPI is the interval from the last pacing stimulus during entrainment to the first sensed electrogram at the same pacing catheter. Using the above AP example, the final paced beat should enter the circuit in the antegrade direction and take 300 ms to traverse the entire circuit. If the pacing catheter is positioned in this 300 ms circuit, the PPI is expected to be around 300 ms. However, if the pacing catheter is outside the circuit, then the PPI will be the 300 ms circuit time plus the travel time from the catheter to the circuit and the travel time from circuit back to catheter. The tachycardia cycle length (TCL) is subtracted from the PPI to determine the difference. PPI-TCL of <|30ms| is typically considered within the circuit. These maneuvers are important when determining where to ablate an intra-atrial or ventricular reentry circuit.

Documentation of entrainment technically requires three criteria as described by Waldo.

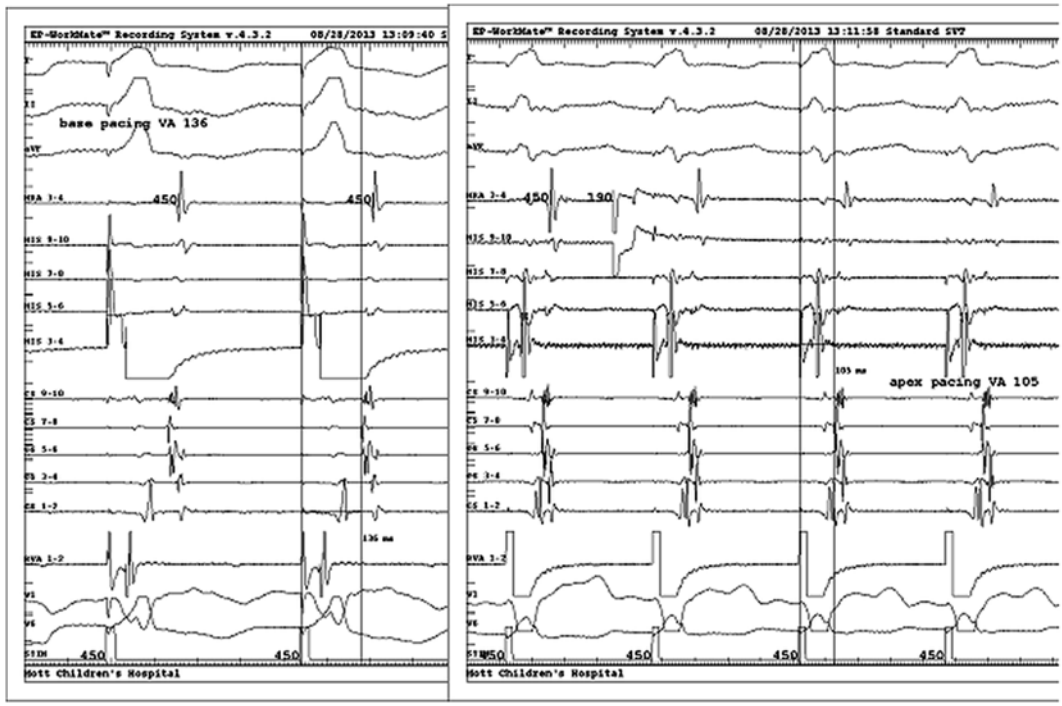

Fig. 3.14 Base/apex pacing: Intracardiac electrograms displayed as surface leads I, II, aVF, HRA, His proximal (9–10) to distal (3–4), CS proximal (9–10) to distal (1–2), RV apex, surface leads V1 and V6. *Left panel* shows RV pacing from the base with a measured stimulus to A interval of 136 ms. *Right panel* shows RV pacing from more distal toward the apex with a stimulus to A interval of 105 ms. Difference in base/apex pacing is consistent with retrograde activation via the AV node; see text. (Note: sweep speeds of tracings are slightly different.) *HRA* high right atrium, *CS* coronary sinus, *RV* right ventricle

(1) When pacing at a constant rate that is slightly faster than the rate of the tachycardia (but not terminating it), there is the demonstration of constant fusion beats in the ECG except for the last captured beat which is not fused. (2) When pacing at 2 or more constant rates faster than the tachycardia rate (but not terminating it), there is demonstration of different degrees of constant fusion on the ECG between the different pacing rates. (3) When pacing at a constant rate that is faster than and interrupts the tachycardia, local conduction block to a site can be demonstrated for 1 beat followed by activation of that site by the next paced beat with a shorter conduction time (Waldo 2004).

RV entrainment has been shown to distinguish septal AP-mediated tachycardias from AVNRT in children specifically when focusing on the post-pacing interval (PPI). Following tachycardia entrainment from the RV apex the corrected PPI minus the tachycardia cycle length (cPPI-TCL = (PPI-TCL) – (post-pacing AH – pre-pacing AH)) was measured and found to differentiate AVNRT (cPPI-TCL > 95 ms) from ORT (cPPI-TCL < 95 ms). Further, entrainment pacing from the RV during 1:1 narrow complex tachycardia by the same mechanism is diagnostic of an AP-mediated tachycardia if the first fully paced beat advances the atrial activation and consistent with AVNRT if the atrial activation is not advanced until after the third fully paced beat.

His-refractory Ventricular Extrastimulation (VES). During narrow complex 1:1 tachycardia, a single ventricular premature extrastimulus is

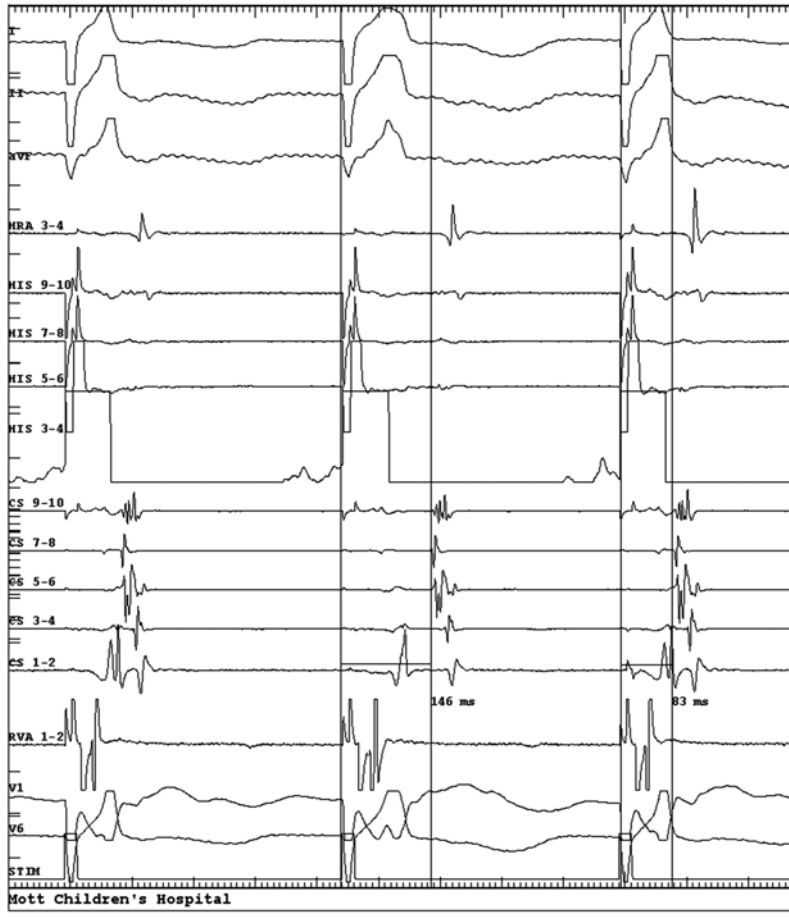

Fig. 3.15 ParaHisian pacing: Intracardiac electrograms displayed as surface leads I, II, aVF, HRA, His proximal (9–10) to distal (3–4), CS proximal (9–10) to distal (1–2), RV apex, surface leads V1 and V6. Pacing is from the distal His catheter. The second beat displays a wide QRS indicating the absence of His bundle capture—with a measured stimulus to A interval of 146 ms. The third beat displays a narrow QRS indicating capture of the His bundle with a stimulus to A interval of 83 ms. The shorter stimulus to A interval with His capture is consistent with retrograde conduction via the AV node and the absence of a septal accessory pathway. See text for details. *HRA* high right atrium, *CS* coronary sinus, *RV* right ventricle

delivered when the His is refractory (Fig. 3.20). If this *His-refractory VES* advances the subsequent atrial activation, it is 100 % diagnostic of the presence of an AP. Absence of the response, however, does not rule out an AP. Entrainment pacing from the RV during 1:1 narrow complex tachycardia by the same mechanism is diagnostic of an AP-mediated tachycardia if the first fully paced beat advances the atrial activation and consistent with AVNRT if the atrial activation is not advanced until after the third fully paced beat.

Ventricular pacing during narrow complex 1:1 tachycardias may also reveal the diagnosis of atrial tachycardia. If the ventricular activation, while pacing faster than the tachycardia rate, can be dissociated from the atrial activation without affecting the timing of atrial activation, then atrial tachycardia is diagnosed. Alternatively, if at the termination of ventricular pacing there is demonstrated a stimulus-V-A-A-V interval followed by continuation of the tachycardia, this too is diagnostic of an atrial tachycardia. In this

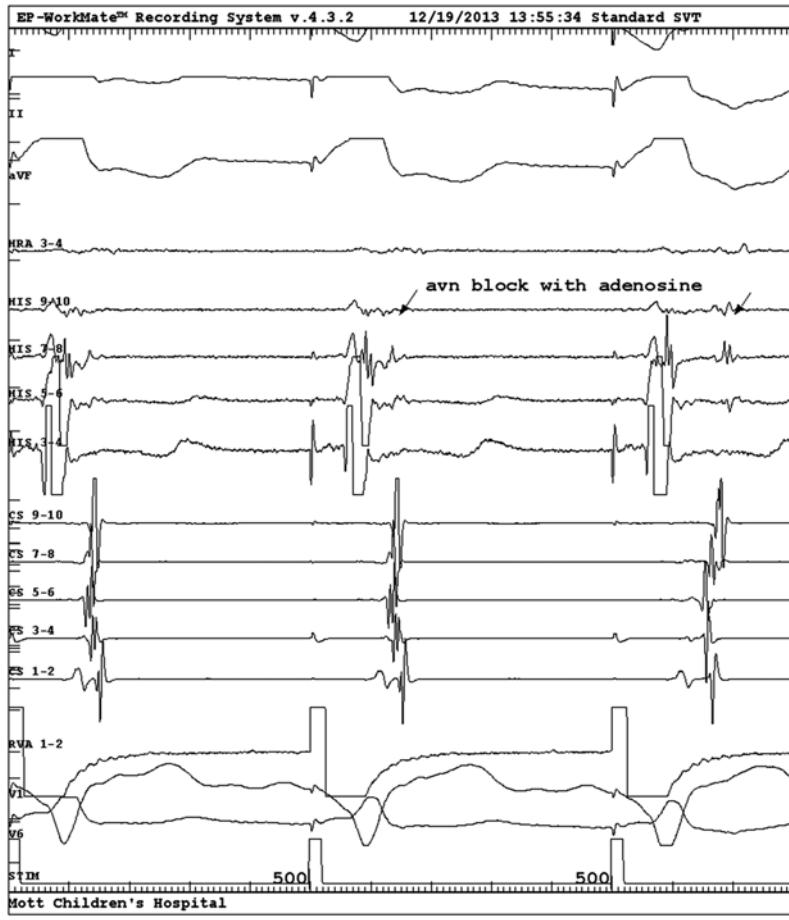

Fig. 3.16 Adenosine reveals AP conduction: Intracardiac electrograms displayed as surface leads I, II, aVF, HRA, His proximal (9–10) to distal (3–4), CS proximal (9–10) to distal (1–2), RV apex, surface leads V1 and V6. Pacing is from the RV catheter at 500 ms cycle length during adenosine administration. The first and second beats display retrograde activation via the AV node with earliest atrial activation at the His catheter. The third beat shows the effect of adenosine with a change in atrial activation. The atrial activation at the His is delayed and retrograde activation proceeds through a left inferior accessory pathway earliest in CS 5–6. *HRA* high right atrium, *CS* coronary sinus, *RV* right ventricle

circumstance, a pseudo V-A-A-V response must be ruled out which would occur during atypical AVNRT (Fig. 3.21).

Late coupled premature atrial extrastimulus: A narrow complex 1:1 tachycardia with a very short VA time <70 ms may be consistent with either typical AVNRT or a junctional tachycardia. While AVNRT should be able to be pace induced and pace terminated, sometimes additional evidence is necessary to fully determine the tachycardia mechanism. A *late coupled premature atrial extrastimulus* during the tachycardia can serve to aid this differentiation (Fig. 3.22). The AES delivered during junctional tachycardia is expected to advance the immediately following ventricular activation. On the other hand, in typical AVNRT the AES would enter the antegrade slow pathway and alter the timing of not the immediate V, but the subsequent V activation (Padanilam et al. 2008).

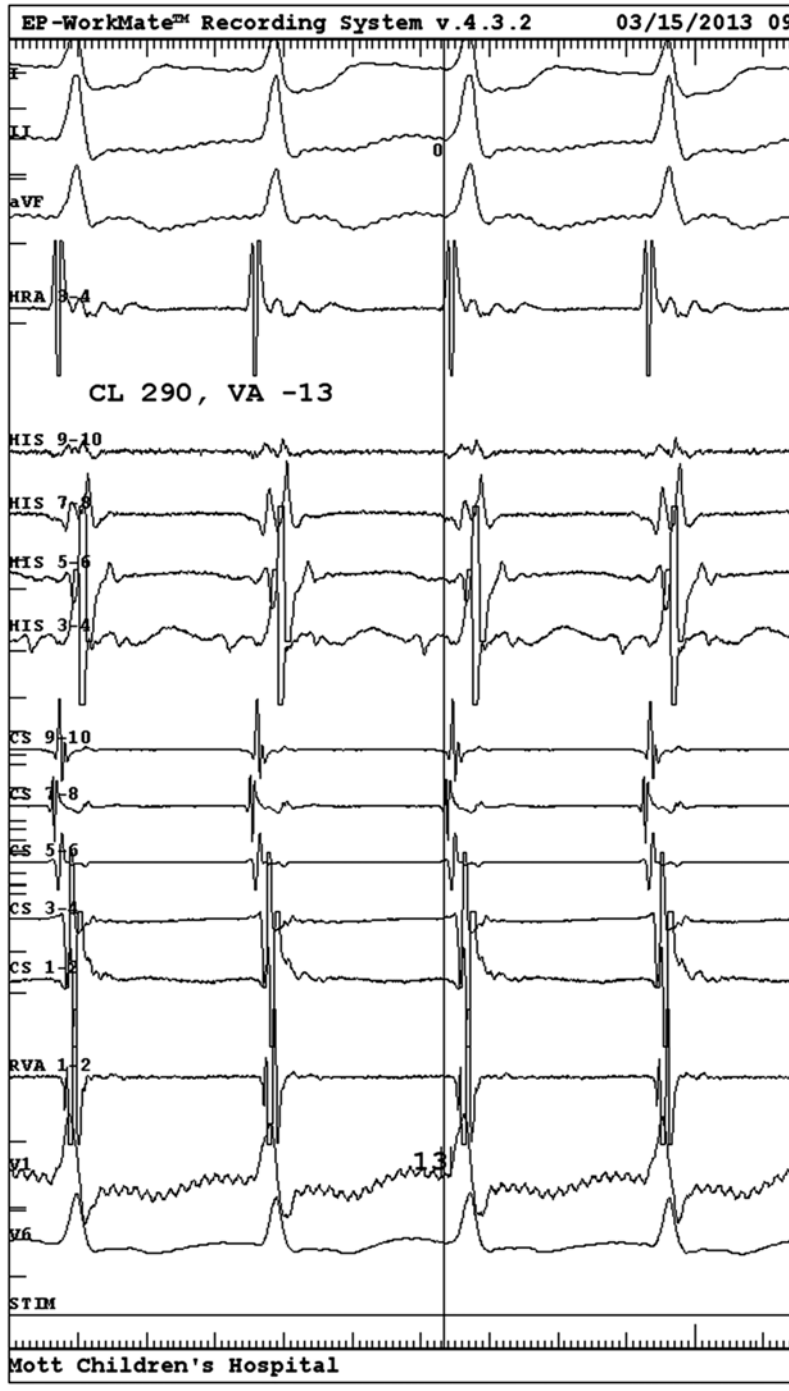

Fig. 3.17 AVNRT: Intracardiac electrograms displayed as surface leads I, II, aVF, HRA, His proximal (9–10) to distal (3–4), CS proximal (9–10) to distal (1–2), RV apex, surface leads V1 and V6. The rhythm is AVNRT. The atrial and ventricular activation is simultaneous. Measured cycle length is 290 with VA interval—13 ms. *HRA* high right atrium, *CS* coronary sinus, *RV* right ventricle

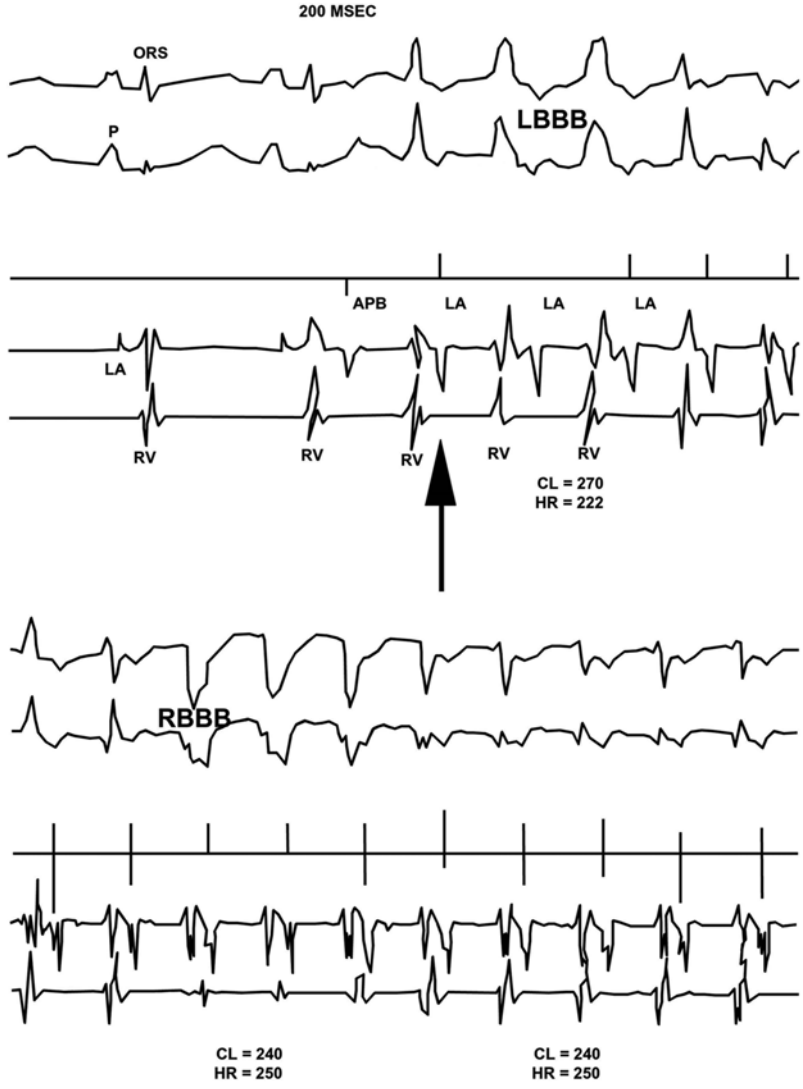

Fig. 3.18 Electrocardiographic tracings (Lead I and II, *top* 2 tracings) and cardiac electrograms form the left atrium (LA) and the right ventricular (RV) during supraventricular tachycardia. *Arrow* indicates spontaneous premature atrial beat initiating tachycardia. Note left bundle branch block configuration (3–5 beats) with increased retrograde conduction interval between the RV and the LA. In contrast as the QRS normalizes the RV-LA interval shortens; then, (*bottom tracings*) right bundle branch block develops, but the RV to LA interval stay short and persists even when the QRS totally normalizes, indicating a left-sided accessory pathway supporting the tachycardia

A multitude of alternative diagnostic maneuvers are documented in the literature. These generally serve to differentiate AVNRT from accessory pathway-mediated tachycardia or atrial tachycardia. Long RP tachycardias especially tend to require diagnostic maneuvers to prove the diagnosis. The majority of diagnostic maneuvers are variations on the concept of entrainment.

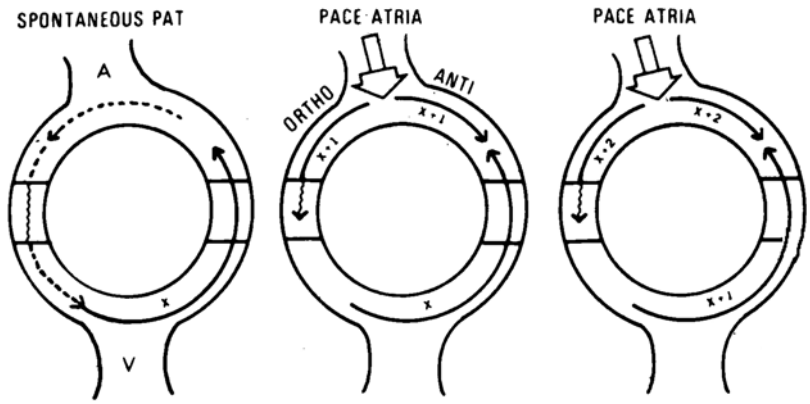

Fig. 3.19 Cartoon displaying *Entrainment*. See text. [Reprinted from Waldo AL, et al., Transient entrainment and interruption of the atrioventricular bypass tract type of paroxysmal atrial tachycardia. A model for understanding and identifying reentrant arrhythmias. Circulation 1983; 67(1): 73–83. With permission from Wolters Kluwer Health]

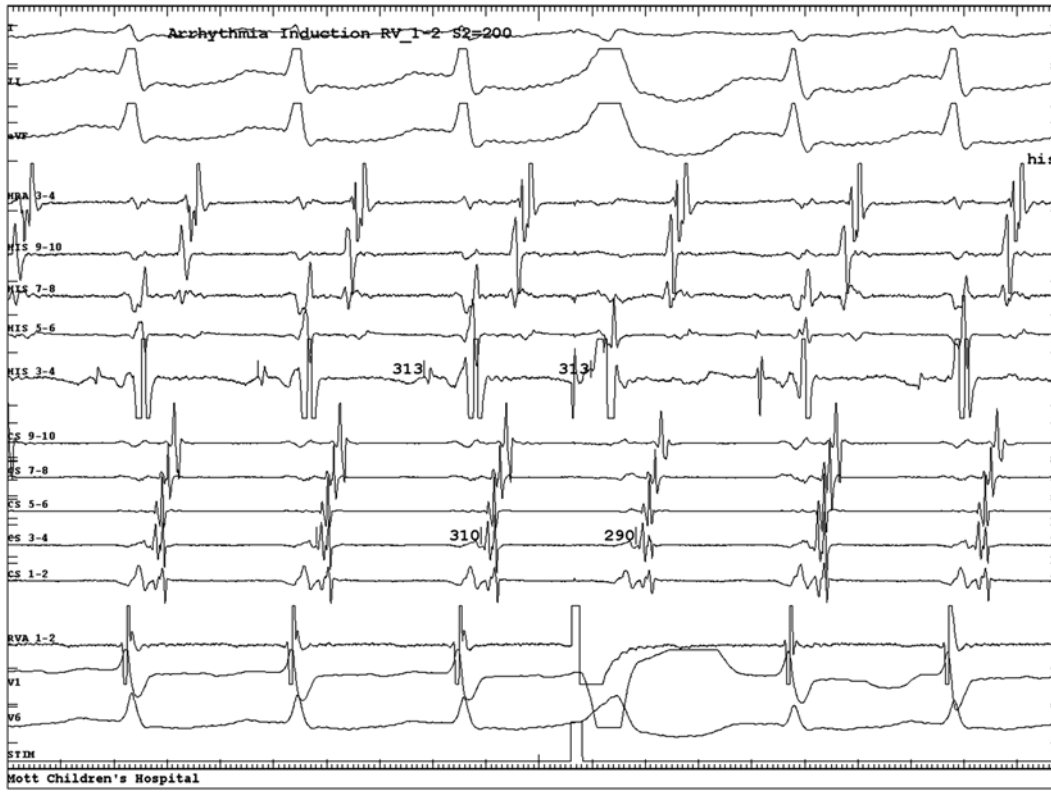

Fig. 3.20 His refractory PVC: Intracardiac electrograms displayed as surface leads I, II, aVF, HRA, His proximal (9–10) to distal (3–4), CS proximal (9–10) to distal (1–2), RV apex, surface leads V1 and V6. The rhythm is an orthodromic reentry tachycardia via a left lateral accessory pathway. The His bundle interval and tachycardia CL is measured in His 3–4. A premature ventricular extrastimulus delivered to time with His refractoriness. The subsequent atrial activation is early—measured at 290 ms confirming the presence of a retrograde accessory pathway. *HRA* high right atrium, *CS* coronary sinus, *RV* right ventricle

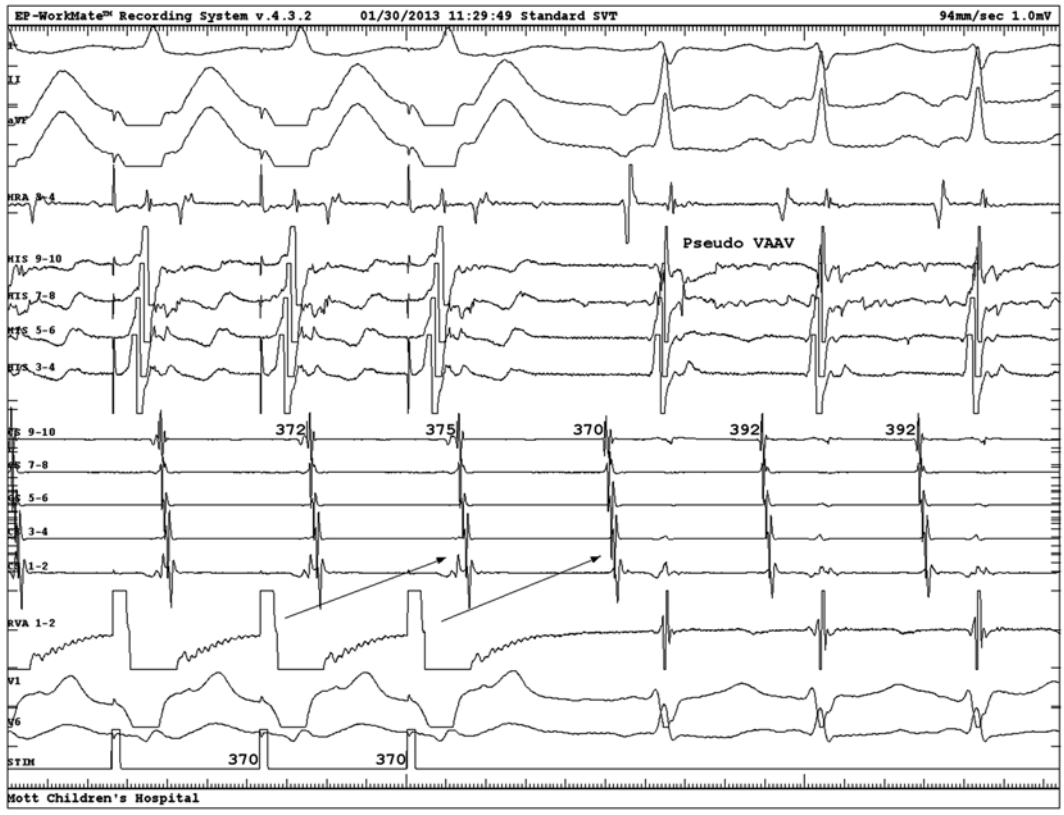

Fig. 3.21 Pseudo VAAV response during atypical AVNRT: Intracardiac electrograms displayed as surface leads I, II, aVF, HRA, His proximal (9–10) to distal (3–4), CS proximal (9–10) to distal (1–2), RV apex, surface leads V1 and V6. The rhythm is a long RP tachycardia with negative P-waves in the inferior leads. The tachycardia cycle length is 392 ms. Ventricular pacing at 370 ms conducts to the atrium at the same rate and with the same activation pattern. The response after termination of pacing is V-A-A-V which would typically indicate atrial tachycardia as the mechanism. However, this is a pseudo V-A-A-V response. The second A of the V-A-A-V response is entrained to the pacing rate of 370 ms indicating that this A is conducted from the last pacing beat. The ventricular pacing is conducting to the atrium via the slow pathway of the AV node. *HRA* high right atrium, *CS* coronary sinus, *RV* right ventricle

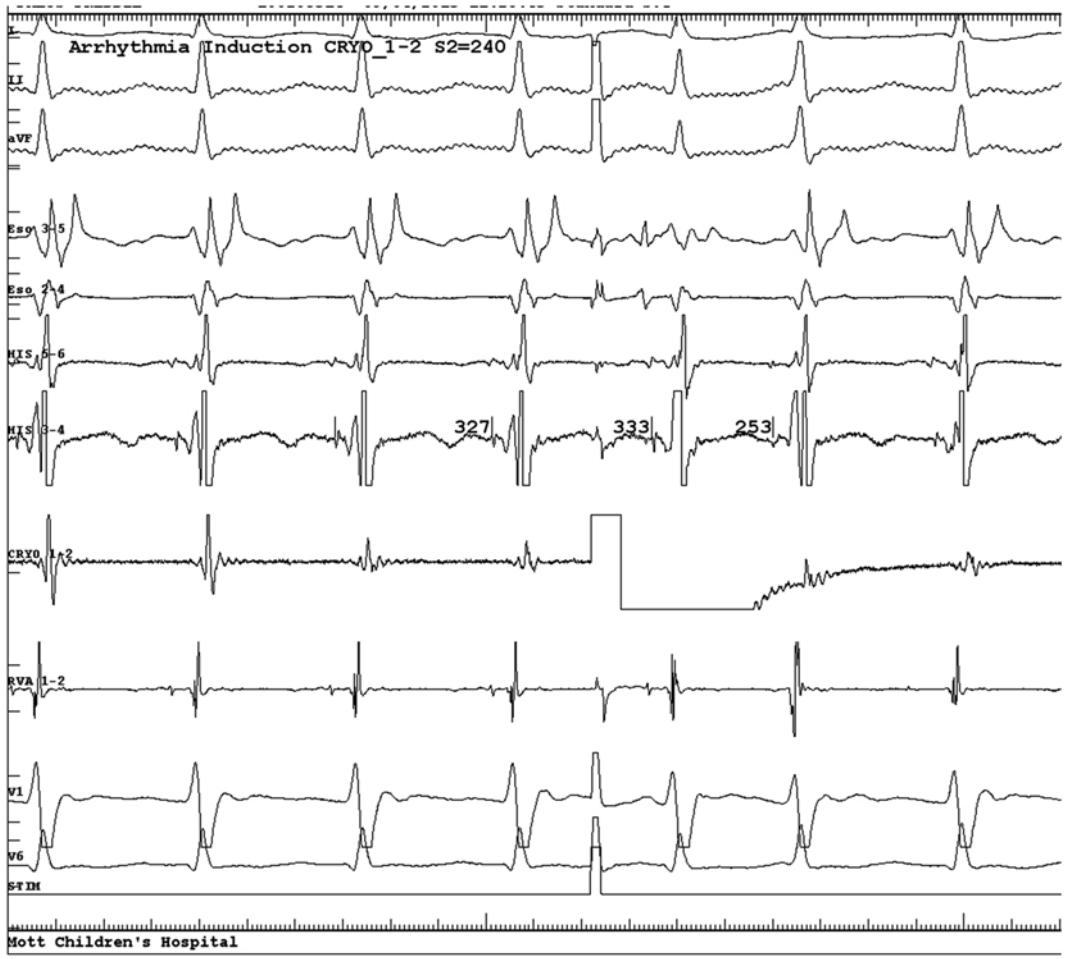

Fig. 3.22 Premature AES confirming AVNRT: Intracardiac electrograms displayed as surface leads I, II, aVF, esophageal, His proximal to distal, cryoablation distal, RV apex, surface leads V1 and V6. The rhythm is a narrow complex 1:1 tachycardia with near simultaneous VA activation at 330 ms, consistent with either AVNRT or junctional tachycardia. A premature atrial extrastimulus is delivered. The subsequent His signal is on time (333 ms), but the next His is early at 253 ms. This is consistent with AVNRT. The premature atrial extrastimulus entered the slow pathway of the AV node and conducted down to the ventricle bringing in the $n+1$ His. The His immediately following the premature atrial extrastimulus was activated from the beat prior to the premature extrastimulus. If this had been a junctional tachycardia, the first His after the premature extrastimulus would have been early. *HRA* high right atrium, *CS* coronary sinus, *RV* right ventricle

Conclusion

The EPS is the first step toward successful catheter ablation of tachyarrhythmias in pediatric and congenital heart disease patients. In-depth knowledge of clinical electrophysiology and a diagnostic armamentarium are important for procedure success. The ultimate goal of the EPS is to identify the target for ablation which will be addressed in the subsequent chapter.

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