Basics of Pediatric Electrocardiography and Invasive Electrophysiology: Principles of Cardiac Testing

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Abstract

The pediatric electrocardiogram is the cornerstone of evaluation for all arrhythmia conditions and acute arrhythmia analysis and treatment. In the presence of an arrhythmia, additional modalities are available for capturing a paroxysmal rhythm disorder. The interpretation of the electrocardiogram is dependent on understanding the evolution of the electrocardiogram as the child ages. Electrocardiographic abnormalities that occur in children include Wolff-Parkinson-White syndrome, supraventricular tachycardia, complete heart block, junctional ectopic tachycardia, ventricular tachycardia, atrial flutter, atrial fibrillation, long QT syndrome, Brugada syndrome, and myocardial ischemic changes in patients with anomalous origin of the left coronary artery from the pulmonary artery. The indications for invasive intracardiac electrophysiology studies include the risk assessment of patients with ventricular or supraventricular tachycardia, the assessment of anti-arrhythmia drug efficacy, and the evaluation of unexplained syncope.

Keywords

ALCAPA • Anomalous left coronary artery from the pulmonary artery • Arrhythmias • Atrial fibrillation • Atrial flutter • Atrial wire ECG • Brugada syndrome • Cardiac testing • Complete heart block • ECG • Electrocardiography • Event monitor • Holter monitor • Implantable loop monitor • Invasive electrophysiology • Junctional ectopic tachycardia • Long QT syndrome • Loop monitor • Myocardial ischemia • Paroxysmal rhythm disorder • Supraventricular tachycardia • Syncope • Telemetry • Ventricular tachycardia • Wolff-Parkinson-White syndrome

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Basics of Electrocardiography and Invasive Electrophysiology

Evaluation of the electrophysiology of pediatric cardiac patients is an integral part of the complete evaluation of any heart problem. Though supplanted by imaging modalities such as angiography, echocardiography, and cardiac magnetic resonance imaging for the assessment of cardiac anatomy in the pediatric patient, the basic 12- or 15-lead electrocardiogram (ECG) remains the cornerstone of evaluation for all arrhythmia conditions and acute arrhythmia analysis and treatment. Though stated over 50 years ago, Nadas' comments regarding the ECG's central role as part of the "tripod" upon which rests clinical diagnosis in pediatric cardiology remain as true today as then [1].

Based upon the work of many prior investigators, Einthoven described the first string galvanometer which is commonly viewed as the true predecessor of the modern day ECG machine [2]. Improvements in electrocardiographic recording technology have allowed for miniaturization of the recording device. This has allowed for ECG recording in various hospital and out-of-hospital settings. The various forms of electrocardiographic recording include the basic ECG, 24-h ambulatory ECG recording ("Holter" monitoring), "event" recorders of various types, and, finally, telemetry [3–5].

By allowing for multiple (limb and parasternal) leads of simultaneous electrocardiographic recording, the ECG allows the clinician the ability to view both electrical depolarization and repolarization of the heart from various angles. The multiple angles of "electrical visualization" allow for various determinations such as rhythm assessment, hypertrophy, and axis determination of both the atria and ventricles as well as allow for greater accuracy when diagnosing arrhythmia conditions such as Wolff-Parkinson-White syndrome (WPW), long QT syndrome, or Brugada syndrome, to name but a few. There are many conditions or clinical situations in which there is simply no noninvasive or invasive substitute for the basic ECG in assessing the heart.

However, because of the ephemeral or intermittent nature of various cardiovascular arrhythmias or symptoms, the basic 12- or 15-lead ECG is, at times, simply impractical for the capture of various different arrhythmia conditions. For this reason, electrocardiographic surrogates have arisen. These typically provide between one and three leads of the ECG with the thought that capturing even a single lead of ECG during an intermittent event is superior to no tracing. There are an increasing number of these devices and they will be briefly described below (Table 20.1).

Holter Monitor

The simplest such device is the 24- or 48-h ambulatory ECG, or "Holter monitor" [6]. This device can typically record up to 3 leads of the basic ECG for 1 or 2 days continuously. The device will also allow for heart rate variability analysis [7–11]. For patients in whom there are daily symptoms, this form of monitoring is useful for capturing frequent sensations or arrhythmias [12, 13]. Typically, patients are given a "diary" which is usually a single sheet of paper on which daily activities and symptoms can be recorded and then double checked with the timed electrocardiographic tracings. Because it is continuously recording the entire time period, the onset and offset of arrhythmias can often be documented which can, in certain situations, be useful in determining arrhythmia mechanism.

Event Monitor

When symptoms are less frequent than daily, there are a virtual plethora of options for the cardiologist to use to try capture the event electrocardiographically. [14–19] The first of these is the so-called event recorder. These devices, which can be as small as a typical credit card, allow for the recording of any arrhythmia or sensation by placing the device directly to the

Table 20.1 Types of noninvasive electrocardiographic recorders and their strengths and weaknesses

Type of ECG recorder	Advantages	Disadvantages
Holter	Single day, full disclosure, onset and offset of arrhythmias, heart rate variability data, quantification of ectopic beats	Improper for infrequent symptoms (only 1–2 days)
Event recorder	Infrequent symptom capture, ease of use	Requires patient coordination and compliance, poor for very brief symptoms, some versions must be uploaded manually
Loop recorder	Infrequent symptom capture, ease of use, can automatically upload	Must wear ECG electrodes daily for 3–4 weeks, no full disclosure
Ambulatory telemetry	Infrequent symptom capture, ease of use, can automatically upload, full disclosure	Must wear ECG electrodes daily for 3–4 weeks
Implantable loop recorder	No need for ECG electrodes, minimal patient involvement in recording, ease of downloading of information	Requires surgical procedure, no full disclosure

chest and then pressing a button to record. The devices typically can record a few episodes of 30–60 s duration. Once recorded, the device can then be taken to a telephone, and the recording downloaded to a "central station" where the ECG tracing is generated. The main advantage of this form of recording is ease of use. The device is not affixed to the patient continuously and only needs to be applied at the time of symptoms. There are, however, several disadvantages of such a device. It is often challenging for the patient to carry the device continuously for 3–4 weeks, and when symptoms are very brief, such a device may not be practical to capture an episode, even if the device is immediately adjacent to the patient. These devices can also only record a single ECG lead, thus limiting the interpretation of the tracing compared to a typical 12-lead or 15-lead ECG.

Loop Monitor

The "loop recorder" was designed to address some of the disadvantages of the event recorder [20, 21]. This device is similar to the event recorder, but the patient typically wears ECG electrodes that are attached via cables to the device that is worn 24 h per day for a 3–4-week period. The device continuously records a period of time (typically 1–3 min of "looping") and also continuously erases this period of recording with new recording. When the patient has the sensation for which the recording is performed, a button can be pressed which will typically command the device to record the prior 30 or 45 s as well as the subsequent 30 or 45 s. This recording can then be sent, in similar fashion to the event recorder, over the phone, to a "central station" for conversion to an ECG tracing. Two newer variations of the loop recorder have become more common. So-called "automated" recorders can be programmed to work similarly to the "loop recorder" with the primary difference being that the device will also automatically record anything that it believes is an arrhythmia, typically using algorithms that are based upon heart rate ranges [22]. Thus, this sort of recorder will automatically record any preprogrammed activity that may represent an arrhythmia while also allowing the patient to override the system and record any symptoms that they may be feeling. This feature is particularly useful in young or incapacitated patients that are unable to press the button or for complaints such as syncope where the patient may be unable to press the button during an episode. Finally, newer technology allows for the device to automatically send the data to a central station using cell phone technology without the need for uploading by the patient.

Ambulatory Telemetry

More recently, with the advent of cellular phone technology, a newer technology that is often referred to as "ambulatory telemetry" allows for complete and total recording of a patient's heart rhythm with near "full disclosure" [23–26]. This allows the cardiologist to view not only device- and patient-commanded arrhythmias but the entire daily rhythm as well as rhythm and heart rate trends in similar fashion to a 24-h ambulatory "Holter" device, though for multiple days. There are some data to suggest that this form of arrhythmia monitoring may be superior in rhythm sensitivity for various different arrhythmia conditions [25, 26].

Implantable Loop Monitor

The final form of ambulatory monitoring is the so-called implantable loop recorder/ILR [27–32]. This device, which is a few inches in size, can be implanted beneath the skin in the left parasternal area in a minor surgical procedure. The device allows for single ECG lead recordings and can record data that can then be downloaded either in the physician office or over the telephone/Internet. The device can automatically record either bradycardias or tachycardias as defined by the physician and can also be patient activated. The indications for their usage are largely the same as for any of the ambulatory systems previously discussed above with the main advantage being that they require essentially no patient cooperation. As a general rule, these are best reserved for patients in whom the above modes of monitoring have failed or in patients who are highly noncompliant [33].

Atrial Wire Postoperative Studies

Following open heart surgery, any sort of arrhythmia may be potentially hemodynamically embarrassing. As a result, rapid and accurate diagnosis followed by treatment is imperative in the postoperative setting. The most important "test" for assessing postoperative arrhythmias is undoubtedly the 12- or 15-lead ECG as described above. However, there are situations in which it is challenging to accurately assess the relationship of atrial to ventricular depolarizations on the surface ECG alone. For this reason, atrial wire studies are often conducted, in addition to the 12-lead ECG, to help the clinician better assess this relationship and therefore better understand the nature of the arrhythmia.

At the time of surgery, it is common for a congenital heart surgeon to place one or two wires on the surface of the atrium and ventricle [34]. As these are directly touching the atrial myocardium, intracardiac recordings can be made at the bedside. Typically, a precordial lead (e.g., V1–V6) is used in addition to the standard limb leads. The precordial lead is usually attached to the metal atrial lead using an "alligator clip," resulting in a large atrial electrogram deflection on the ECG. By running the standard surface leads simultaneously, the relationship of the atrial depolarization to ventricular depolarization (as inscribed by the surface QRS) can be easily determined. Examples of rhythms in which atrial wire tracings can be useful would be JET or ventricular tachycardia in which the ventricular rate often exceeds the atrial rate. Incorporation of an atrial electrogram tracing in concert with the surface ECG (plus the response to pacing of the atrium or ventricle) is often all that is necessary to make accurate postoperative rhythm determinations.

Interpretation of the Electrocardiogram in Children

The basic approach to reading a pediatric electrocardiogram (ECG) should be systematic and consistent. There is no one "right way," but a methodical approach should be used in order to avoid missing any abnormalities. We would advocate a "12-step" approach with attention to the following details and findings (Table 20.2). There are published tables on the normal pediatric ECG values for heart rate, PR intervals, QRS axis, etc. based on patient age (Fig. 20.1),

Table 20.2 "12-step" approach to ECG interpretation

1. Relationship between Ps and Qs (P waves and QRS complexes)
2. Atrial and ventricular rates
3. P wave axis
4. QRS axis
5. Right precordial lead (V1/V3R/V4R)
6. R to S transition in the precordial leads
7. Hypertrophy (atrial and ventricular hypertrophy)
8. ST segments
9. T waves
10. Q waves
11. Calculation of intervals (PR, QRS, QTc)
12. Assessment of other abnormalities

but perhaps the most important consideration in reading an ECG in a pediatric patient is the understanding of the evolution of the ECG from infancy to adulthood [35–38].

The Evolution of the Electrocardiogram Throughout Childhood

The 15-lead ECG changes significantly throughout childhood. [35–38] Caregivers therefore cannot interpret the electrocardiogram of a pediatric patient without knowing the age of the patient. Understanding the changes seen on ECG throughout childhood is best done by understanding the evolution of cardiac physiology from fetus to adulthood, as the changes in the ECG reflect the changing physiology from the fetus, to the newborn, to the young child, and to the adult. The ECG thus evolves from primarily prominent right ventricular (RV) forces in the newborn period to primarily left ventricular (LV) dominant forces seen in late adolescence and adulthood. In the fetus, the right ventricle is the primary output for systemic blood flow, pumping roughly 65 % of the cardiac output to the body as the pulmonary resistance is elevated and blood flows from right to left through the patent ductus arteriosus [39]. This is reflected in the electrocardiogram of the newborn with prominent RV forces and a rightward QRS axis. As pulmonary resistances drop over the first few weeks of life, the RV forces on the ECG gradually diminish and this continues over the first few years of life. Left ventricular dominance predominates in later childhood and adolescence, ultimately taking the form of the typical adult ECG. The changes on the ECG during childhood are thus reflected in the QRS axis, the T wave patterns, and the prominence of RV and LV forces as illustrated below. In addition, the "normal" values for heart rate, PR interval, QRS interval, QTc interval, and amplitudes of the R and S waves in the right and anterolateral precordial leads also change with age.

Newborn

The ECG in the newborn reflects the importance of the RV in fetal life and shows prominent RV forces (Fig. 20.2). The QRS axis is usually rightward, between 30 and 180°. The prominent RV forces are demonstrated by tall R waves noted in V1 and the right precordial leads, V3R and V4R, with a relatively small S wave. The T waves are usually upright in V1, V3R, and V4R. In V6 there is often a small R wave with a more prominent S wave. The notable exception to this typical newborn ECG is in the ECG of a premature infant. The pulmonary vascular resistance of the neonate is typical lower than a full-term neonate, and the premature infant does not demonstrate the prominence of RV forces that are usually seen in the typical full-term newborn. The ECG of the premature neonate is perhaps more similar to that of an adolescent or adult pattern, with a more leftward QRS axis, minimal RV forces, and prominent LV forces.

Two Weeks

By 2 weeks of age, pulmonary vascular resistance has usually diminished and the RV pressures have started to decrease. This is reflected on the ECG with a change in the T wave morphology in V1, V3R, and V4R (Fig. 20.3). While upright in the newborn, the

NORMAL PEDIATRIC ECG PARAMETERS

Age	Heart Rate (bpm)	QRS Axis*	PR Interval (sec)*	Lead V_1 QRS Duration (sec)†	Lead V_1 R Wave Amplitude (mm)†	Lead V_1 S Wave Amplitude (mm)†	Lead V_1 R/S Ratio	Lead V_6 R Wave Amplitude (mm)†	Lead V_6 S Wave Amplitude (mm)†	Lead V_6 R/S Ratio
0–7 days	95–160 (125)	+30 to 180 (110)	0.08–0.12 (0.10)	0.05 (0.07)	13.3 (25.5)	7.7 (18.8)	2.5	4.8 (11.8)	3.2 (9.6)	2.2
1–3 wk	105–180 (145)	+30 to 180 (110)	0.08–0.12 (0.10)	0.05 (0.07)	10.6 (20.8)	4.2 (10.8)	2.9	7.6 (16.4)	3.4 (9.8)	3.3
1–6 mo	110–180 (145)	+10 to +125 (+70)	0.08–0.13 (0.11)	0.05 (0.07)	9.7 (19)	5.4 (15)	2.3	12.4 (22)	2.8 (8.3)	5.6
6–12 mo	110–170 (135)	+10 to +125 (+60)	0.10–0.14 (0.12)	0.05 (0.07)	9.4 (20.3)	6.4 (18.1)	1.6	12.6 (22.7)	2.1 (7.2)	7.6
1–3 yr	90–150 (120)	+10 to +125 (+60)	0.10–0.14 (0.12)	0.06 (0.07)	8.5 (18)	9 (21)	1.2	14 (23.3)	1.7 (6)	10
4–5 yr	65–135 (110)	0 to +110 (+60)	0.11–0.15 (0.13)	0.07 (0.08)	7.6 (16)	11 (22.5)	0.8	15.6 (25)	1.4 (4.7)	11.2
6–8 yr	60–130 (100)	−15 to +110 (+60)	0.12–0.16 (0.14)	0.07 (0.08)	6 (13)	12 (24.5)	0.6	16.3 (26)	1.1 (3.9)	13
9–11 yr	60–110 (85)	−15 to +110 (+60)	0.12–0.17 (0.14)	0.07 (0.09)	5.4 (12.1)	11.9 (25.4)	0.5	16.3 (25.4)	1.0 (3.9)	14.3
12–16 yr	60–110 (85)	−15 to +110 (+60)	0.12–0.17 (0.15)	0.07 (0.10)	4.1 (9.9)	10.8 (21.2)	0.5	14.3 (23)	0.8 (3.7)	14.7
>16 yr	60–100 (80)	−15 to +110 (+60)	0.12–0.20 (0.15)	0.08 (0.10)	3 (9)	10 (20)	0.3	10 (20)	0.8 (3.7)	12

Fig. 20.1 Table of normal ECG parameters during childhood [38]

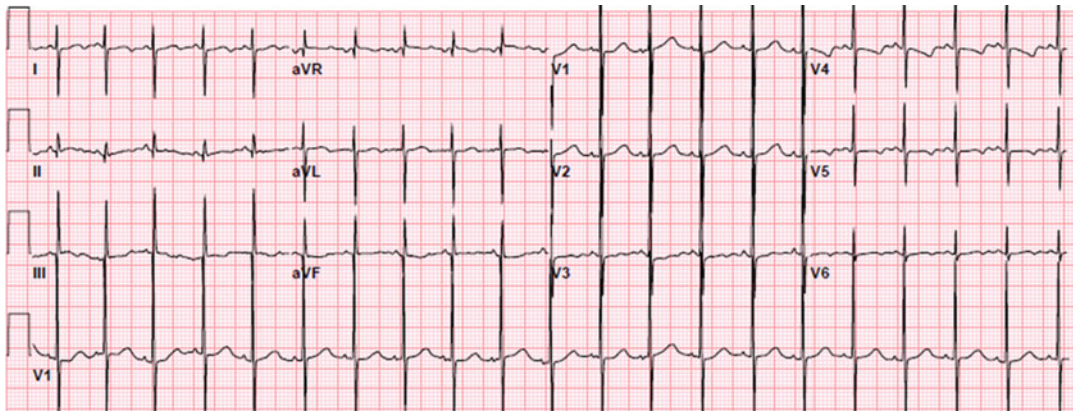

Fig. 20.2 Normal newborn ECG. Note the *right* axis deviation, prominent R wave in V1, and *upright* T wave in V1

T wave now becomes inverted in V1, V3R, and V4R. This T wave pattern, with inversion in V1/V3R/V4R, should persist throughout childhood into late adolescence. If the T wave were to remain upright in the newborn after 2 weeks of age, it would indicate possible right ventricular hypertrophy or RV hypertension.

One Year of Age

By 1 year of age, the RV forces are beginning to diminish with more prominent LV forces noted (Fig. 20.4). The QRS axis is usually slightly less rightward. The T waves remain inverted in V1/V3R/V4R. There is usually slightly more LV forces, with a slightly smaller R wave and slightly larger S wave in V1 and usually less S wave in V6.

Four Years of Age

By 4 years of age, the QRS axis has usually normalized (between 0 and 110°). There is again less RV prominence in the right precordial

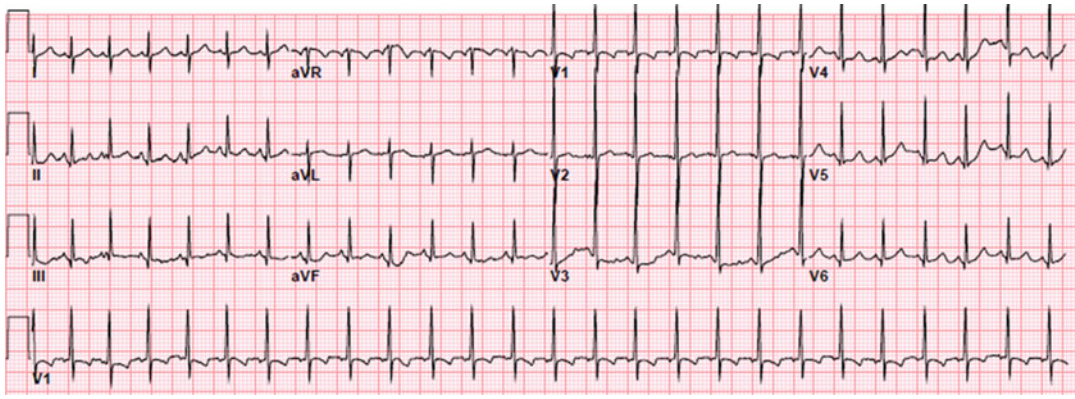

Fig. 20.3 Normal ECG of a 2-week-old infant. Note the *rightward* QRS axis, the prominent R wave in V1, and the inverted T wave in V1

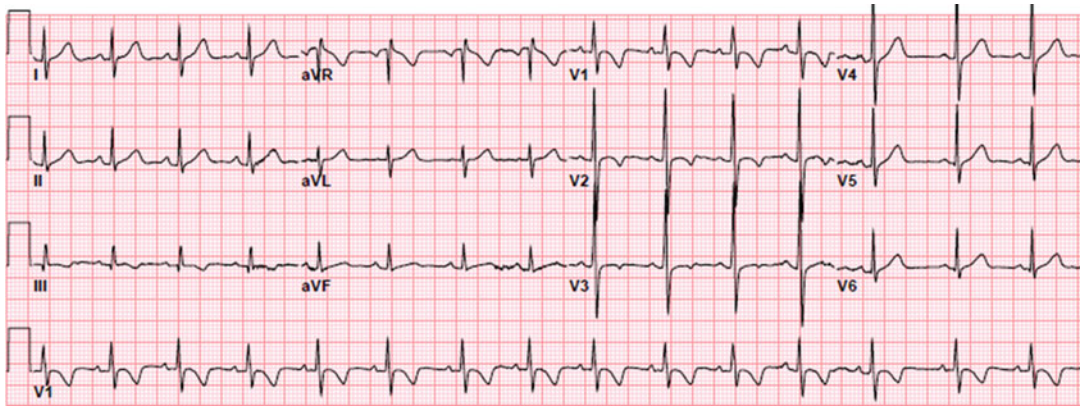

Fig. 20.4 Normal ECG in a 1-year-old. Note the more *leftward* QRS axis and more prominent LV forces in V6, though still a positive R/S ratio in V1 with an inverted T wave in V1

leads, with a more balanced R/S ratio in V1 (Fig. 20.5). The T wave in V1 has remained inverted. There should be little to no S wave in V6 with a predominant R wave in V6.

Note that the predominant change compared to the 1-year-old ECG is the more balanced R/S ratio in V1.

Twelve Years of Age

The ECG in a 12-year-old is similar to the adult, with the exception of the immature ("infantile") T wave pattern that is usually still present in V1, V3R, and V4R. The QRS axis should be predominantly leftward (between −15 and 110°). There is usually a small R wave and deep S wave with good R wave progression from a prominent S wave in V1 to a pure R wave in V6. The T waves are usually still inverted in V1 with upright T waves in V5/V6 (Fig. 20.6).

Late Adolescence: 18 Years of Age

By late adolescence the ECG takes the pattern of the normal adult (Fig. 20.7). The QRS axis is usually normal (−15 to 110°). The T wave now

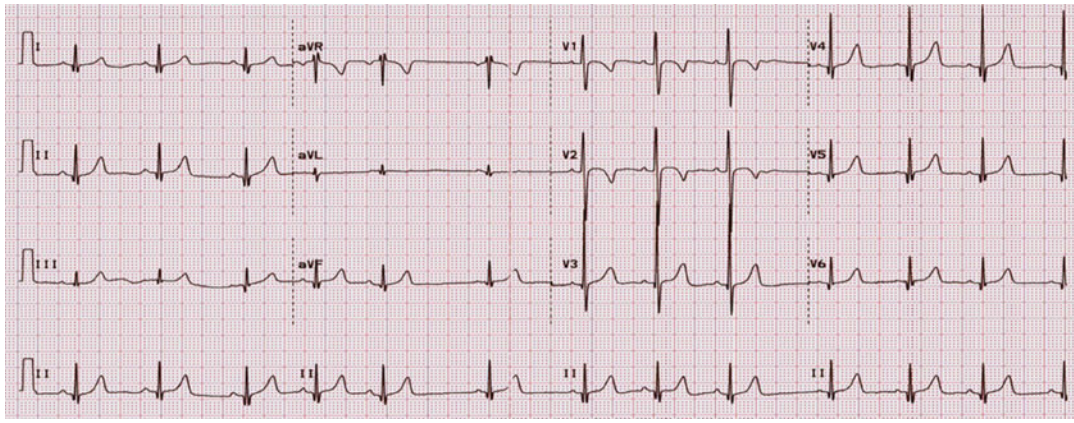

Fig. 20.5 Normal ECG in a 4-year-old

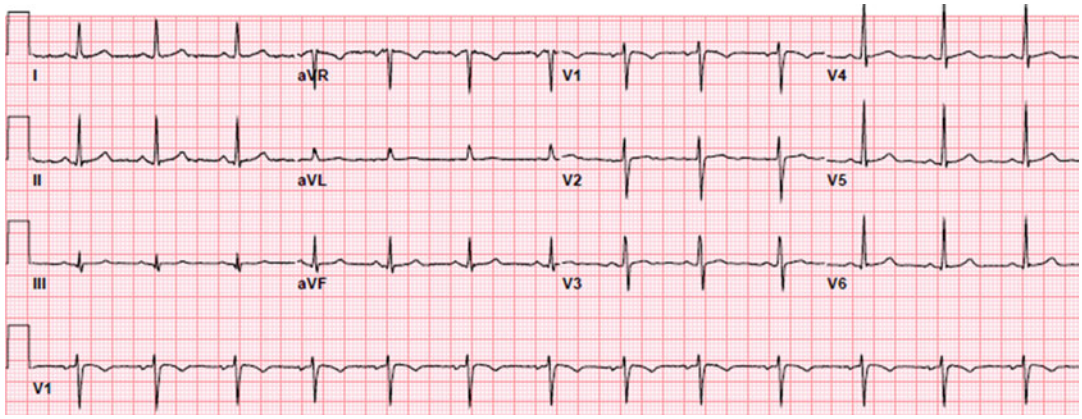

Fig. 20.6 Normal ECG in early adolescence. Note the similarity to the adult ECG, though with a juvenile T wave pattern in V1 with an inverted T wave

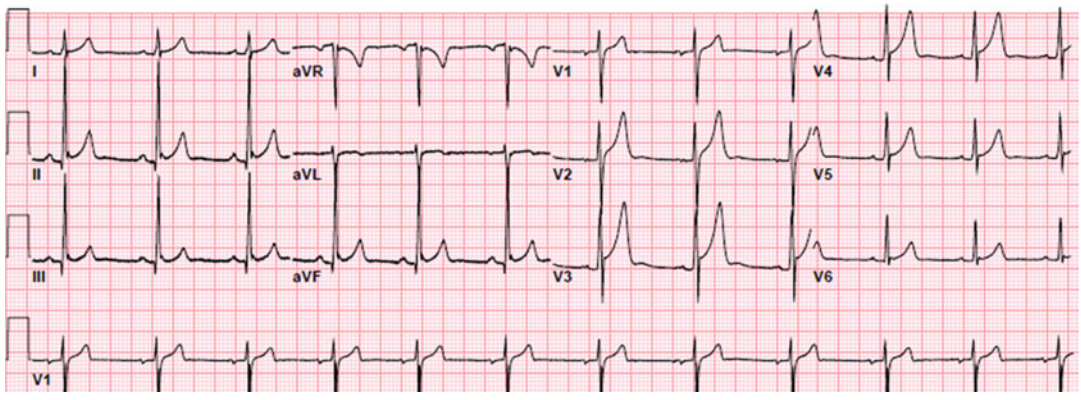

Fig. 20.7 Normal ECG in late adolescent and in young adults. Note the predominance of LV forces with an *upright* T wave in V1

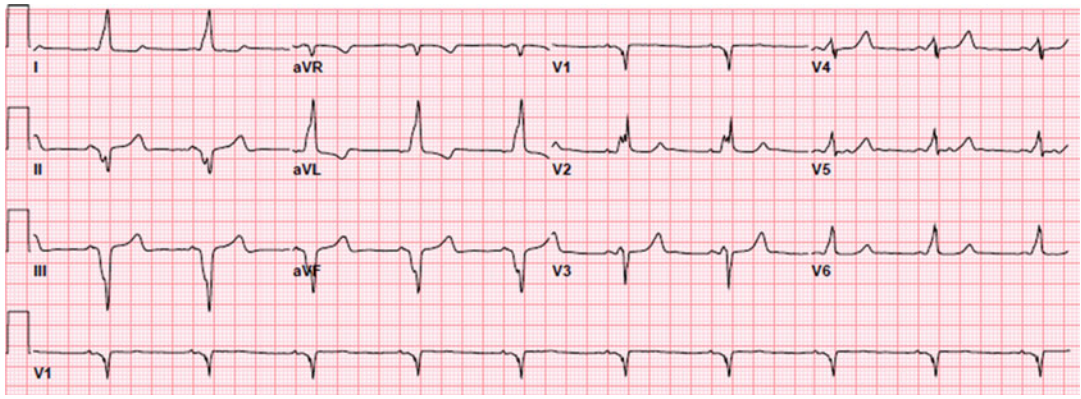

Fig. 20.8 ECG from a 17-year-old patient with palpitations and Wolff-Parkinson-White. Note the short PR interval, wide QRS complex, and delta wave (slurring of the QRS upstroke)

becomes upright in the right precordial leads though may remain inverted in V1. There is usually only a small R wave in V1 with a small S wave. Through the precordial leads there is progression from a deep S wave in V1 to a pure R wave in V6, so-called good R wave progression.

"Top 10" ECG Abnormalities

Wolff-Parkinson-White Syndrome

Wolff-Parkinson-White syndrome (WPW), or ventricular preexcitation, was first described in 1914 with the combined findings of a short PR interval, slurring of the initial upstroke of the QRS complex (delta wave), and a wide QRS associated with paroxysmal tachycardia (Fig. 20.8) [40]. The initial authors did not recognize that the etiology of WPW was the presence of an accessory pathway or extra electrical connection in the heart connecting the atrium and ventricle.

WPW occurs in approximately 0.1 % of the population and is slightly more common in males [41]. The main clinical problem in patients with WPW is the occurrence of supraventricular tachycardia (SVT), primarily AV reciprocating tachycardia (AVRT). The most common form of SVT in patients with WPW is typically orthodromic reciprocating tachycardia (ORT), with antegrade conduction down the AV node and retrograde conduction via the accessory pathway leading to a narrow complex tachycardia. A rarer form of tachycardia in patients with WPW is antidromic reciprocating tachycardia (ART), with antegrade conduction down the accessory pathway and retrograde conduction through the AV node causing a wide complex tachycardia. Both types of SVT can be terminated with vagal maneuvers, administration of IV adenosine, or anti-arrhythmic medications. Current treatment options for patients with WPW include the use of vagal maneuvers to terminate paroxysmal tachycardia, use of chronic anti-arrhythmic medications to prevent episodes of tachycardia, or invasive electrophysiology (EP) testing and ablation of the accessory pathway to permanently destroy the accessory pathway and cure patients of their condition. Ablation is a catheterization procedure performed via access in the femoral and occasionally internal jugular veins with a low risk of complications (1 % of less) and high success rate (88–97 %) [42].

In addition to the risk of developing SVT, patients with WPW have a small risk of sudden cardiac death, estimated to be 0.015 per patient year, or 0.1 % annual risk [41, 43]. Sudden death is believed to be secondary to atrial fibrillation with rapid conduction via the accessory pathway

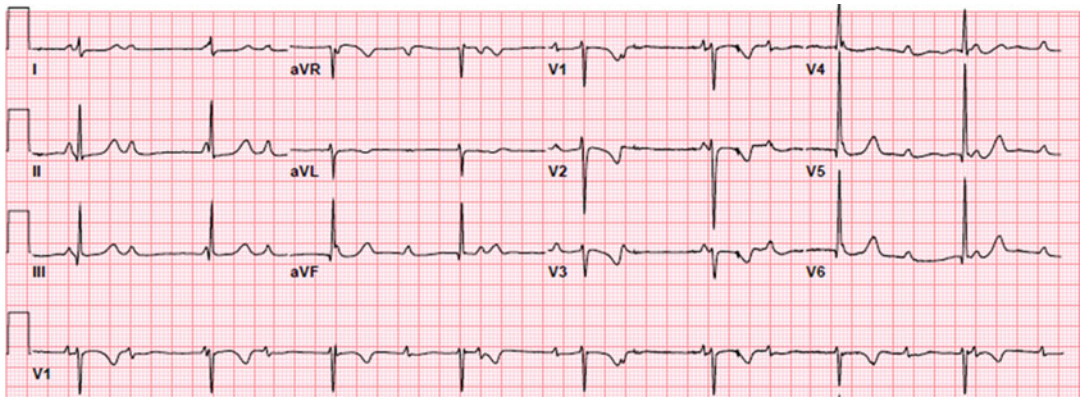

Fig. 20.9 ECG from a 4-year-old female with congenital complete heart block secondary to maternal lupus demonstrates an atrial rate of approximately 100 bpm with a narrow complex junctional escape rate of approximately 50 bpm. Note the regular RR interval with complete AV dissociation

leading to ventricular fibrillation [44, 45]. Invasive EP testing can help determine if a patient with WPW is at higher risk of sudden death. During testing atrial fibrillation is induced, and if the accessory pathway can conduct faster than 250 ms, this would place a patient in a higher-risk group [44]. Recent guidelines recommend exercise stress testing in patients over 8 years of age, and if there is no loss of ventricular preexcitation during the exercise test, invasive EP testing should be considered to determine if the accessory pathway was a "high-risk" pathway, with consideration of ablation in those high-risk patients or those with inducible SVT [12].

Complete Heart Block

Complete heart block (CHB), or third-degree AV block, is the failure of any atrial electrical impulses to be conducted to the ventricles (Fig. 20.9). While the SA node continues to fire regularly, there is no atrioventricular conduction with the result of an atrial rate that typically exceeds the ventricular rate. Patients may be able to maintain cardiac output and systemic blood flow via escape rhythms from within the His-Purkinje system, resulting in a narrow complex escape rhythm, or from within the ventricles, resulting in a wide complex escape rhythm [46].

In children, the most common etiology of CHB is secondary to damage to the AV node and conduction system during surgery for repair or palliation of congenital heart disease (postsurgical CHB) [47]. Other forms of congenital heart disease also have a risk of patients developing heart block, such as heterotaxy syndrome and congenitally corrected transposition of the great vessels (L-TGA), which carries a 1–2 % annual risk or roughly 30 % lifetime risk of developing CHB [48, 49]. Additional acquired causes include infectious diseases such as Lyme disease and other forms of myocarditis, infiltrative diseases such as Hunter's or Hurler's syndromes, thyroid dysfunction, and cardiomyopathies [50–52]. In neonates, the most common cause of CHB is congenital AV block secondary to maternal lupus. Maternal anti-Ro and anti-La antibodies cross the placenta and lead to damage of the conduction system in utero [53, 54].

Treatment of AV block depends on the heart rate, associated factors (such as presence of congenital heart disease), and escape rhythm. In older patients (teenagers and adults), treatment is placement of a permanent pacemaker. In those with postoperative CHB, a period of waiting of 7–10 days should be observed prior to placement

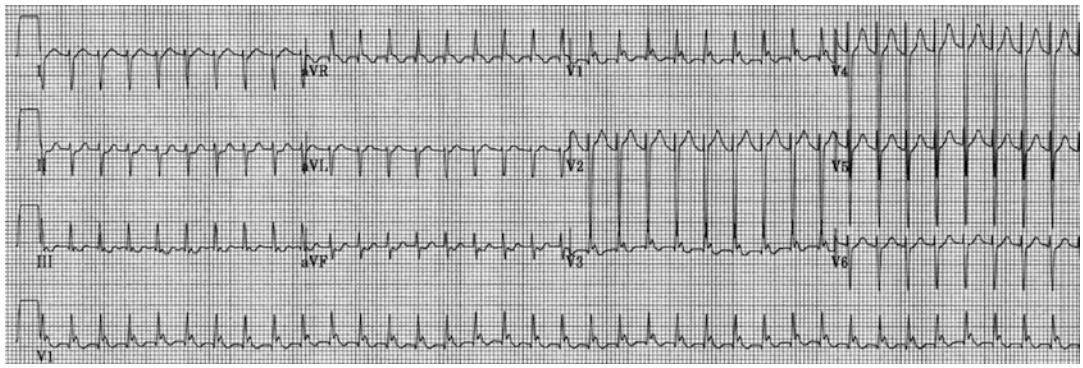

Fig. 20.10 ECG from a 4-month-old female following operative closure of a ventricular septal defect with junctional ectopic tachycardia (JET) at a rate of 215 bpm. The QRS complex was identical to that seen upon arrival to the intensive care unit when in sinus rhythm. Note that there is 1-to-1 ventriculoatrial conduction through the AV node retrograde as seen by the inverted retrograde P waves immediately following the QRS complex in lead III (and upright and more clearly seen in lead V1)

of a permanent pacemaker, as roughly 65 % of patients can recover atrioventricular conduction during this time [46, 55]. In neonates and young children, in the presence of a narrow complex escape rhythm and an adequate heart rate (>55 bpm), placement of a permanent pacemaker can often be delayed until the early teenage years, when the risk of syncope and sudden cardiac death increase.

Junctional Ectopic Tachycardia

Junctional ectopic tachycardia, commonly referred to as JET, is a tachycardia arising from the AV node or His-Purkinje system (Fig. 20.10) [56, 57]. The rhythm is characterized by a QRS complex that is identical to that seen in sinus rhythm, and there is a ventricular rate that is faster than the atrial rate (though in children with preserved AV nodal conduction properties there can be 1:1 retrograde ventriculoatrial conduction).

JET most commonly occurs in children following open heart surgery for congenital heart disease [56–58]. It can cause hemodynamic embarrassment with associated acidosis and poor urine output. Cardiac lesions more commonly associated with JET include ventricular septal defects (VSD), tetralogy of Fallot (TOF), atrioventricular canal repair (AVC), and other lesions that can lead to postsurgical trauma and edema to the His-Purkinje system [56–58].

JET is usually a self-limited disorder in the postoperative period and resolves after 48–72 h. Acute treatment usually involves sedation and pain control, careful cooling and temperature control, electrolyte repletion, minimizing exogenous catecholamines (including IV dopamine, epinephrine, and others), and atrial pacing at a rate faster than the JET rate to improve hemodynamics by providing AV synchrony [59, 60]. Failure of these techniques to resolve or control JET can then lead to treatment with an antiarrhythmic medication such as IV procainamide or IV amiodarone [61–63].

Ventricular Tachycardia

Ventricular tachycardia (VT) is a wide complex tachycardia arising from the ventricles (Fig. 20.11). Electrocardiographic clues to the diagnosis of VT include AV dissociation, a superior QRS axis, fusion beats, and a markedly wide QRS complex (>140 to 160 ms). Though the differential diagnosis for a wide complex tachycardia include ventricular tachycardia, supraventricular tachycardia with

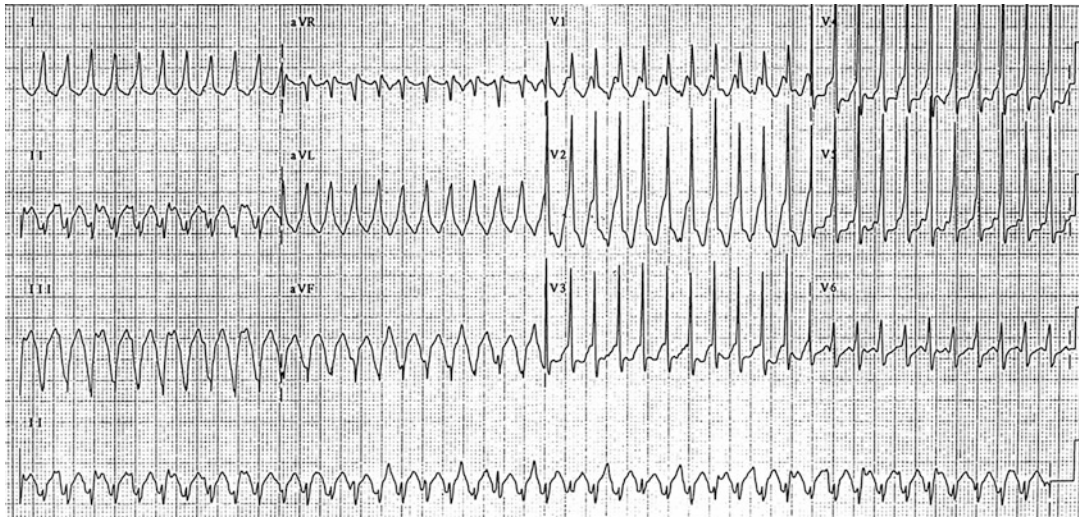

Fig. 20.11 ECG from a 6-month-old female after complete repair of tetralogy of Fallot with ventricular tachycardia at a rate of 230 bpm. The wide complex rhythm has a *left* bundle branch morphology with a superior axis and AV dissociation (as seen in lead II) and a change from a baseline postoperative *right* bundle branch block morphology

aberration, antidromic tachycardia (in a patient with WPW), a paced rhythm, and sinus tachycardia with electrolyte abnormalities or an underlying bundle branch block, it is generally recommended to presume that a wide complex rhythm is ventricular tachycardia until proven otherwise.

Treatment of VT depends in large part to the hemodynamic status of the patient. The hemodynamically stable patient can be given IV lidocaine, procainamide, or amiodarone. The VT could also be terminated with synchronized cardioversion using 2–4 J/kg. The hemodynamically embarrassed patient would require urgent cardioversion.

Supraventricular Tachycardia

Supraventricular tachycardia (SVT) is a regular, narrow complex tachycardia (Fig. 20.12) [64]. The most common cause of SVT in neonates and young children is AVRT mediated by an accessory pathway [64, 65]. In adolescence, the etiology begins to shift to the more common adult form of SVT, AV nodal reentry tachycardia (AVNRT) [65]. If the electrocardiogram is carefully scrutinized, and it can sometimes clue the reader into the diagnosis. In AVRT, there are often retrograde P waves, while in AVNRT the VA time in tachycardia is usually less than 70 ms and there are no discernible retrograde P waves [66]. SVT can often be distinguished from sinus tachycardia by the fixed rate, absence of discernible P waves or P waves with an abnormal axis, and rapid rate greater than 220 bpm. It is not uncommon for SVT rates in neonates to exceed 280–300 bpm.

Acute treatment of hemodynamically stable SVT involves the use of vagal maneuvers, such as ice to the face in neonates, exhalation against a closed glottis, or forcible exhalation. Rapid administration of IV adenosine (0.1 mg/kg) through a large bore IV that is as close to the heart as possible with a 3-way stopcock and a large flush will often terminate SVT [67]. Other anti-arrhythmic options include calcium channel blockers, beta-blockers, digoxin, procainamide, or Class III agents (amiodarone or sotalol) [68–70]. Anti-arrhythmic failure or hemodynamic instability should prompt synchronized cardioversion with 0.5–1 J per kilogram.

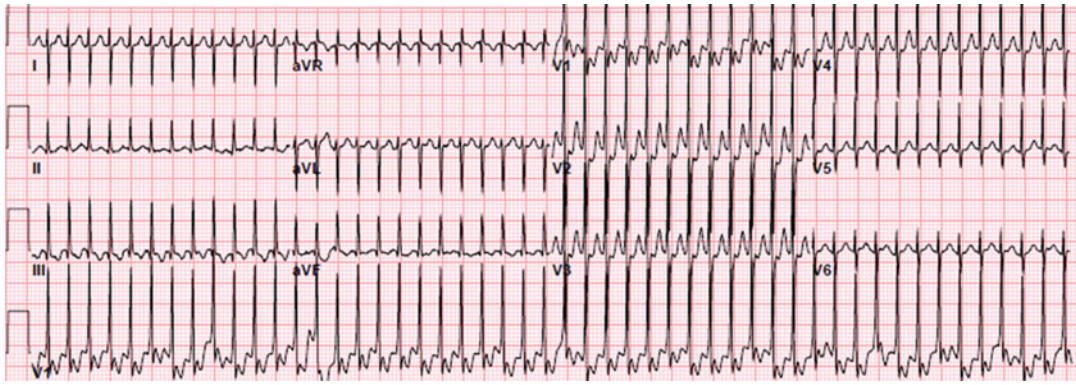

Fig. 20.12 ECG from a 2-day-old male with sudden onset tachycardia demonstrates SVT at a rate of 300 bpm. Note the retrograde P waves in V1 indicating the likely diagnosis of AVRT, the most common cause of SVT in a neonate

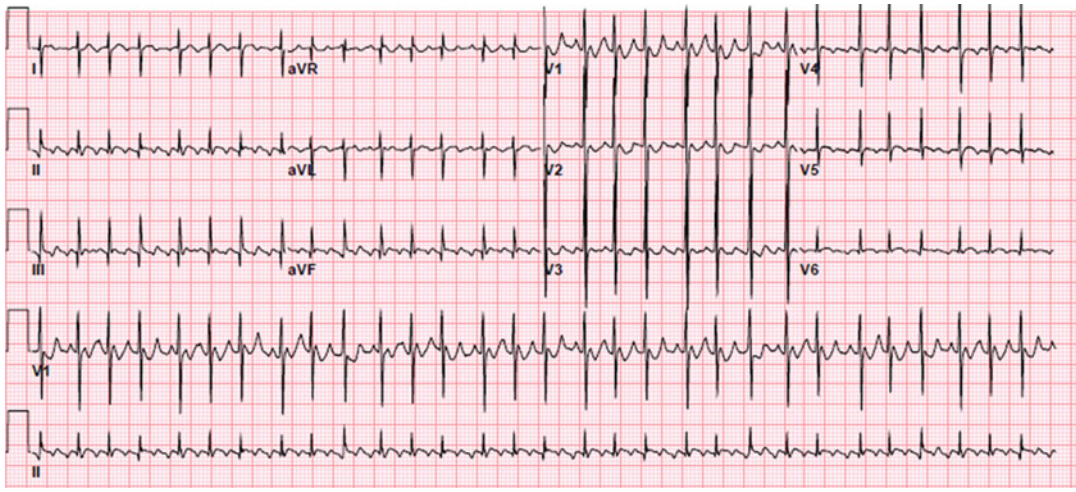

Fig. 20.13 ECG is from a 2-day-old female with tachycardia and atrial flutter at an atrial rate of roughly 450 bpm and a variable 2:1and 3:1 ventricular response rate. The rhythm is best seen in the rhythm strip of lead II. Note the classic sawtooth pattern of atrial activation that is commonly seen in atrial flutter

Long-term treatment can involve one of three different treatment strategies: (1) use of vagal maneuvers to terminate tachycardia, (2) use of medications (beta-blockers, calcium channel blockers, flecainide, or sotalol) to prevent episodes of tachycardia, or (3) EP study and ablation to prevent any future episodes of SVT [70].

Atrial Flutter

Atrial flutter, or intra-atrial reentry tachycardia (IART), is caused by areas of slowed conduction in the atria leading to a reentrant loop or propagation of atrial activation. The ECG typically demonstrates a sawtooth, regular pattern of atrial activation with an atrial rate of 200–400 beats per minute with constant or variable ventricular activation (Fig. 20.13). Atrial flutter is rare in children, though more common in those patients with repaired or palliated congenital heart disease, especially single-ventricle patients after Fontan palliation and patients who have undergone atrial switch procedures for transposition of the great vessels (e.g., Mustard or Senning procedures) [71–74].

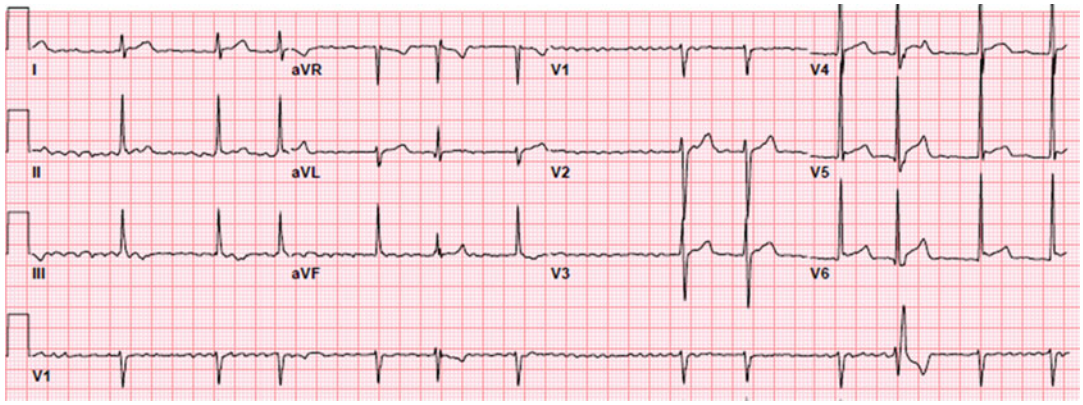

Fig. 20.14 ECG from a 17-year-old male with a 2-week history of palpitations. Note the irregularly irregular rhythm with disorganized atrial activity that is consistent with atrial fibrillation (AFIB). The rhythm strip in V1 also shows a common finding in AFIB, a wide complex beat that is often mistaken for a PVC. This represents "Ashman's phenomenon," or a wide complex beat that conducts with aberration after a long pause, and is not a PVC

Treatment of atrial flutter often depends on the patient presentation and the duration of the tachycardia. Atrial reentry for periods of greater than 24–48 h can be associated with the risk of development of an atrial thrombus and possible stroke. Patients with tachycardia that has clearly been present for less than 24–48 h have a low risk of thrombus formation and stroke and can undergo synchronized cardioversion with 0.5–1 J per kilogram [75]. Medical cardioversion with anti-arrhythmic agents such as Class IC agents (flecainide) or amiodarone could also be attempted [76]. With an unknown duration of tachycardia or tachycardia present for longer than 48 h, treatment options include performing a transesophageal echocardiogram (TEE) to rule out an atrial thrombus followed by immediate synchronized cardioversion or alternatively rate control with beta-blockers, digoxin, and/or calcium channel blockers, anticoagulation, and cardioversion in 6 weeks. Long-term treatment could involve anti-arrhythmic medications or possible curative treatment via catheter or surgical ablation.

Atrial Fibrillation

Atrial fibrillation (AFib) is the most common arrhythmia seen in adults, but is rare in the pediatric population. AFib is characterized by rapid and disorganized atrial activation with no clear, regular discernible P waves and an irregularly irregular QRS response (Fig. 20.14). In children, AFib is more commonly seen in patients with underlying congenital heart disease or a cardiomyopathy, though can rarely occur in children with structurally normal hearts, a condition known as lone AFib [77]. The management and treatment is similar to that of atrial flutter.

Long QT Syndrome

Long QT syndrome is a cardiac channelopathy leading to abnormal ventricular repolarization and ECG findings of prolongation of the corrected QT interval (QTc). These patients are at increased risk of serious ventricular arrhythmias (e.g., torsades de pointes). In general a QTc greater than 450–460 ms is considered abnormal (Fig. 20.15). Additional ECG findings that can help in the diagnosis of long QT syndrome include T wave abnormalities (e.g., tall or peaked T waves, bifid T waves, long ST segments with late T wave development), bradycardia, and T wave alternans. A prolongation of the QTc on ECG can be caused by

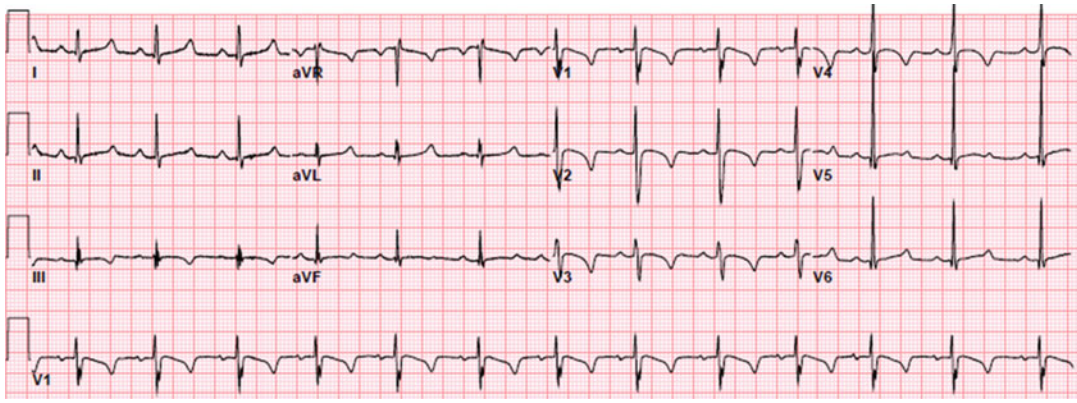

Fig. 20.15 ECG from a 5-year-old female obtained after the sudden cardiac deaths of her father and brother. The patient has a QTc of 520 ms with a long flat ST segment and was diagnosed with long QT syndrome

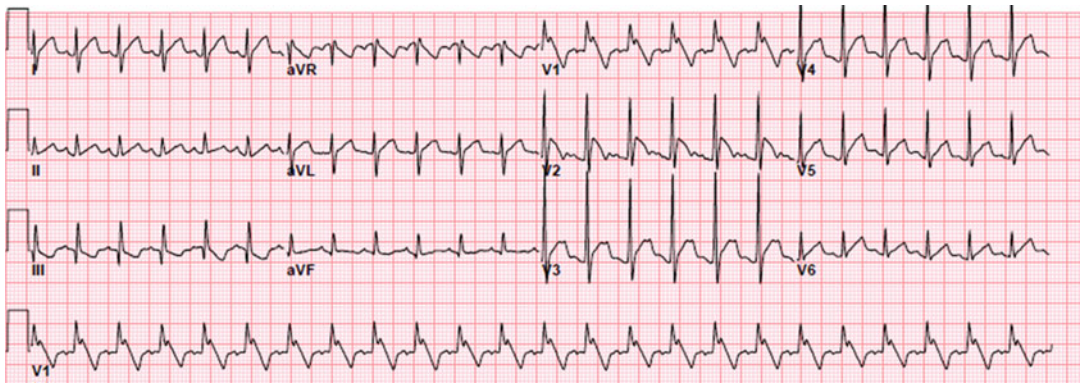

Fig. 20.16 ECG from a 4-year-old male obtained after a syncopal episode. Note the RSR' pattern in V1/V2 with a coved ST segment in V2 that is classic for Brugada syndrome

a true channelopathy or via an acquired cause, such as medications or electrolyte disturbance.

The clinical presentation in children and adolescents with long QT can vary from syncope and seizures to sudden cardiac death. Treatment usually entails use of a beta-blocker which has been demonstrated to decrease the risk of sudden cardiac death, avoidance of medications that prolong the QTc, and possible placement of an implantable cardioverter-defibrillator (ICD) [78, 79]. Specific genotype and long QT subtype therapy may include exercise restriction, avoidance of specific triggers, anti-arrhythmic therapy (Na channel blockers), and stellate ganglionectomy [80, 81].

Brugada Syndrome

Brugada syndrome was first described as a finding in patients surviving cardiac arrest in the late 1980s and early 1990s [82, 83]. The ECG findings in Brugada syndrome involve ST elevation in V1 through V3 with an RSR or RBBB pattern (Fig. 20.16). The syndrome is a heritable channelopathy that can lead to sudden death and most commonly occurring during sleep. The syndrome is one of the most common causes of sudden death in Southeast Asian populations.

Treatment options are limited in patients with Brugada syndrome. Avoidance of fever is a mainstay of treatment, as fever may precipitate

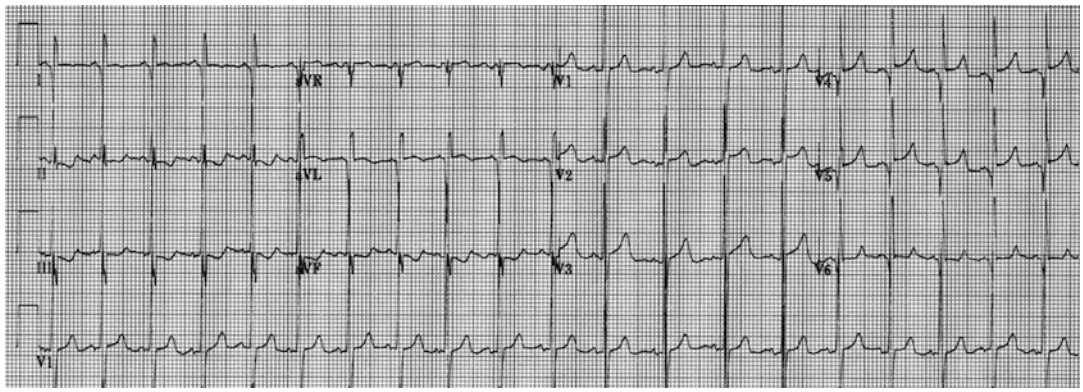

Fig. 20.17 ECG is from a 2-month-old female who presented with sweating, irritability, and feeding intolerance and was found to have an anomalous left coronary artery arising from the pulmonary artery (ALCAPA). Note the deep Q waves in leads I and AVL and also in V4/V5/V6 that are indicative are prior *left* ventricular myocardial infarction. Other notable findings include evidence of RV hypertension with an *upright* T wave in V1 and ST/T wave abnormalities in the inferior and lateral leads

malignant ventricular arrhythmias. ICDs may be indicated in certain patients to prevent sudden cardiac death from ventricular tachycardia and ventricular fibrillation (VF).

Anomalous Left Coronary Artery from the Pulmonary Artery

An anomalous left coronary artery arising from the pulmonary artery (ALCAPA) is a rare congenital anomaly [84]. As pulmonary resistance falls over the first several weeks of life, there is a steal phenomenon whereby coronary blood flow preferentially flows away from ventricular myocardium to the pulmonary artery, leading to myocardial ischemia and myocardial infarction [84]. Affected infants often present in the first 4–6 weeks of life with irritability, sweating or tachypnea, and with feeding and respiratory difficulties. The ECG in these infants often reflects a pattern consistent with an infarct of the left ventricle with deep Q waves in the lateral leads (I and AVL), diffuse ST/T wave changes, and LV enlargement (Fig. 20.17) [85]. Infants with ALCAPA require surgical repair, and the poor ventricular function, LV enlargement, and mitral regurgitation that frequently occur as a result of the LV infarct usually resolve over a period of months to years [86–89].

Basic Notions of Electrophysiology: Intracardiac Electrophysiology

Introduction

Though once viewed as an investigational pediatric procedure reserved either for research purposes or evaluation of arrhythmias that were not decipherable using noninvasive methods, the intracardiac electrophysiology study (EPS) has developed in the past two decades into a commonly performed study for both diagnostic and interventional purposes. [65, 90, 91] The rise of the EPS has been closely linked to the increasing ubiquity of catheter ablation, which has now become the treatment of choice for the management or cure of serious arrhythmias in children and adults [92–94]. When used in concert with noninvasive assessment techniques, it is rare that a patient's arrhythmia mechanism cannot be fully elucidated by the pediatric electrophysiologist, thus leading to more properly tailored arrhythmia therapies.

Table 20.3 Common indications for electrophysiology study

1. Ventricular stimulation study to determine the ease of inducibility of potentially life-threatening ventricular arrhythmias in patients thought to be at possible risk of sudden cardiac death
2. Risk assessment in the setting of WPW, performed either via an intracardiac catheter or via an esophageal pacing catheter
3. Assessment of anti-arrhythmic drug efficacy
4. Evaluation of unexplained syncope

Table 20.4 Indications for ablation in children

1. Curative treatment of accessory AV connections (e.g., WPW, concealed accessory pathways)
2. Curative treatment of AV nodal reentrant tachycardia (AVNRT)
3. Curative treatment of «automatic arrhythmias (e.g., ectopic atrial tachycardia, ventricular tachycardia, some forms of junctional tachycardia)
4. Palliative or curative treatment of intra-atrial reentrant tachycardia in the postoperative patient (e.g., atrial flutter in a Fontan patient)

Indications

The intracardiac EPS has evolved from a procedure that was initially exclusively diagnostic in nature to one that is more commonly used for therapeutic purposes. With the advent of catheter ablation and the comprehensive array of noninvasive electrophysiologic monitors, it is increasingly uncommon for EPS to be performed for exclusively diagnostic purposes. However, despite this trend, the EPS is perhaps still the best means of providing certain diagnostic electrophysiologic data to the clinician. The most common diagnostic indications for EPS are shown in Table 20.3.

As the diagnostic indications for EPS have receded over the recent two decades, the therapeutic indications have widened and broadened. The reason for this is that the types of arrhythmias amenable to transcatheter ablation have grown and the general safety of ablation for the pediatric patient has improved for children of increasingly younger age. [95, 96] Though once viewed as a procedure for only the most recalcitrant or dangerous of arrhythmias, ablation has now become, for many common pediatric arrhythmia substrates, the treatment of choice. Indications for ablation in children are listed in Table 20.4.

Technical Aspects

Invasive EP procedures are typically performed in a cardiac catheterization laboratory and, in children, are usually performed with sedation or general anesthesia. Patients must be properly assessed prior to catheterization in order to evaluate suitability for an invasive procedure including anesthesia. This assessment must include a complete history and physical examination to rule out noncardiac conditions that might increase the risk of any invasive procedure. Unless an EP study is planned to test anti-arrhythmic drug efficacy, anti-arrhythmic drugs should be discontinued at least 5 half-lives prior to the procedure. Those patients with life-threatening arrhythmias may require hospitalization prior to ablation when anti-arrhythmic therapies are discontinued temporarily.

Assessment of the patient undergoing electrophysiologic study must be made by the anesthesiologist providing sedation prior to the procedure. Careful choice of anesthetic technique is imperative as certain agents are either pro-arrhythmic or may potentially reduce arrhythmia inducibility [92, 97, 98]. Additionally, it is often useful to have copies of a patient's resting surface ECG as well as tachycardia tracings in the laboratory in order to have them as a means of comparison during the procedure.

Once in the catheterization laboratory, extensive monitoring and safety precautions are taken. Radiolucent defibrillation pads are affixed prior to catheterization to allow for rapid cardioversion or defibrillation. Blood pressure monitoring, oxygen saturation monitoring, and full 12-lead ECG monitoring are standard for these procedures. Some laboratories will monitor blood pressure continuously with placement of

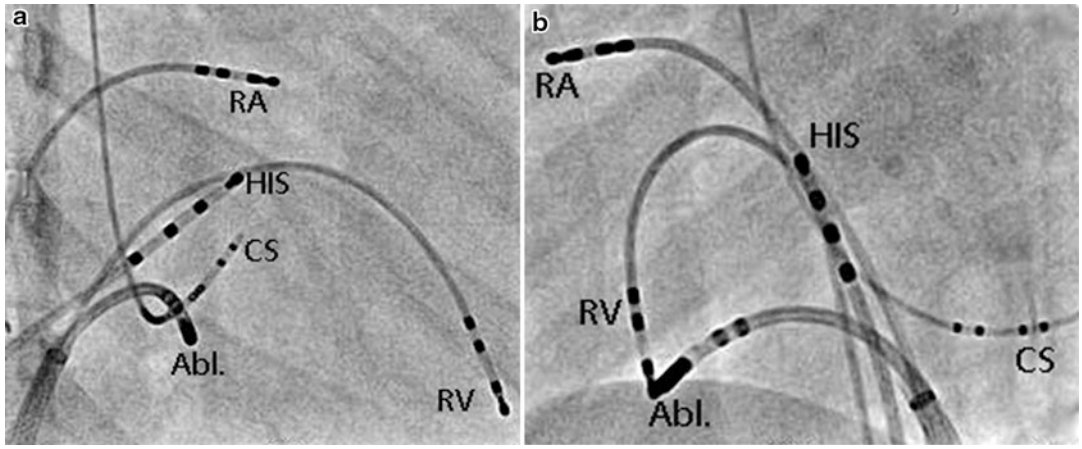

Fig. 20.18 RAO (**a**) and LAO (**b**) views of standard EP catheters in position with the ablation in *right* posterolateral location (*RA* right atrium catheter, *RV* right ventricle catheter, *His* His catheter, *Abl*. ablation catheter, *CS* coronary sinus catheter)

a small catheter either centrally (e.g., femoral artery) or peripherally (e.g., radial artery).

Proper laboratory staffing is essential to ensure patient safety during EP testing and ablation. Typically, most laboratories will have a trained electrophysiology nurse as well as laboratory technologist in addition to one or two electrophysiologists. Many laboratories prefer having 2 electrophysiologists with one operator at the table moving catheters and the other analyzing signals as well as conducting pacing maneuvers. As already noted, proper anesthesiology personnel who are knowledgeable about the management of cardiovascular arrhythmias are preferred.

After induction of anesthesia, catheters are typically placed using modified Seldinger technique in the femoral veins as well as the internal jugular or subclavian vein. The number of diagnostic catheters used can vary depending upon the type of invasive electrophysiologic study planned. For diagnostic studies, the operator typically uses one or two catheters which can be positioned in different locales in the heart during the procedure. For ablation procedures, it is routine to place between 4 and 5 multipole electrical catheters in the heart. The catheters are typically advanced to the heart and placed in standard locations. These include the right atrium, right ventricular apex, HIS position, and coronary sinus. The final catheter is usually the ablation catheter (Fig. 20.18). Fluoroscopy is most commonly used to position catheters in the heart during EP study and ablation. Recently, use of adjuncts such as three-dimensional mapping systems and intravascular echocardiography has been advocated by some groups to the reduce radiation dose associated with these procedures [99]. Three-dimensional electroanatomical mapping systems (e.g., CARTO Biosense Webster) are also useful when mapping complex arrhythmias such as atrial reentry or ventricular arrhythmias of any sort.

Central to all electrophysiologic testing is a programmable stimulator and electrophysiologic recording system. Programmable stimulators allow for provocative pacing maneuvers which can be used to induce arrhythmias as well as assess conduction characteristics of the atrium, ventricle, AV node, and accessory pathways. Most such devices should allow for programmable characteristics such as pulse width, current, and cycle length. By convention, most pacing in the heart is performed at two times the diastolic pacing threshold. The electrophysiologic recording system should be capable of recording multiple channels of information simultaneously. These would include the surface ECG as well as electrograms recorded from all of the intracardiac catheters. Optimally, the computer should allow for recording and display of all signals at varying

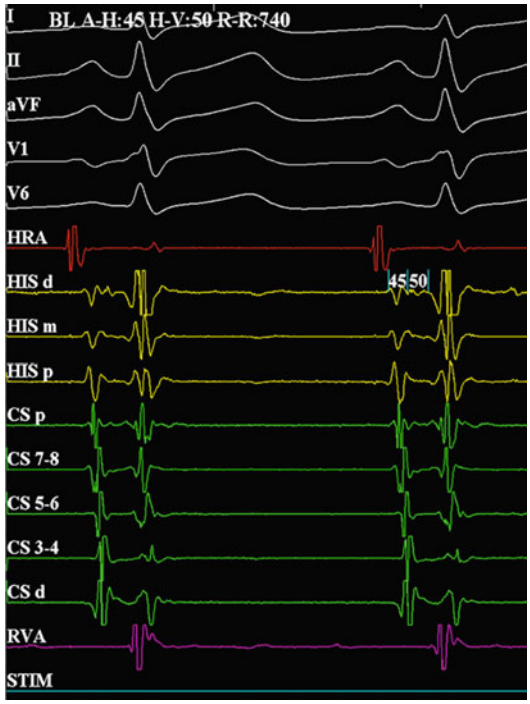

Fig. 20.19 Example of AH (45 ms) and HV (50 ms) intervals

paper speeds from 25 mm/s to 400 mm/s. Standard filtering should allow for filtering of all waveforms below 30 Hz and above 500 Hz in order to properly remove electrical "noise" from recordings.

The final critical device for invasive EP procedures is an ablation generator for either radiofrequency (RF) energy or cryoenergy. Most laboratories have the ability to do either, as RF current and cryoenergy have distinct and separate indications, and these are typically based upon specific patient and arrhythmia characteristics.

Interpretation

Interpretation of complex intracardiac tracings is beyond the scope of this chapter. However, basic concepts are important and relevant. After placing catheters as noted above, baseline resting intracardiac conduction time intervals are recorded. This includes recording of the surface ECG as well. These are often useful in helping make an initial diagnosis of preexcitation as well as providing a baseline as a means of comparison following ablation and catheter manipulation within the heart. The most important two intracardiac measurements are the AH and HV intervals (Fig. 20.19).

The AH interval allows the electrophysiologist to assess AV nodal function and is the measurement from the low right atrial electrogram to the earliest onset of the rapid His deflection in that same electrical pair. As the AV node is electrically silent and as the AV node conducts the electrical impulse from the low right atrium to the bundle of His, the AH interval is used as a surrogate of AV nodal function. A normal AH interval in children is between 50 and 100 ms. It is typically lengthened in patients with AV nodal injury ("first-degree heart block") and also as a result of certain drugs (e.g., digoxin). The second important intracardiac measurement is the HV interval which measures conduction from the proximal His to ventricular myocardium via the Purkinje system. It is measured from the rapid His deflection to the earliest ventricular activation in any lead on the recording system, including surface or intracardiac. A normal HV interval in children is typically between 30 and 50 ms. HV intervals can be prolonged in patients with conduction disorders such as infiltrative myopathies or in patients who have undergone congenital heart surgery and sustained injury. The interval can be shortened in preexcitation conditions such as WPW.

Assessment of the pattern of conduction (both antegrade and retrograde) can be made in sinus rhythm and with ventricular pacing. When conduction is deemed "concentric," the conduction proceeds both antegrade and retrograde earliest in the His catheter pairs, suggesting that the conduction is either via the AV node or an accessory pathway located near to the AV node (Fig. 20.20). If, however, the conduction is "eccentric," the earliest signals can be seen in a location other than the "central" His catheter. If earliest in the coronary sinus pairs, the possibility of a left-sided accessory pathway would be raised. If earliest in the proximal coronary sinus

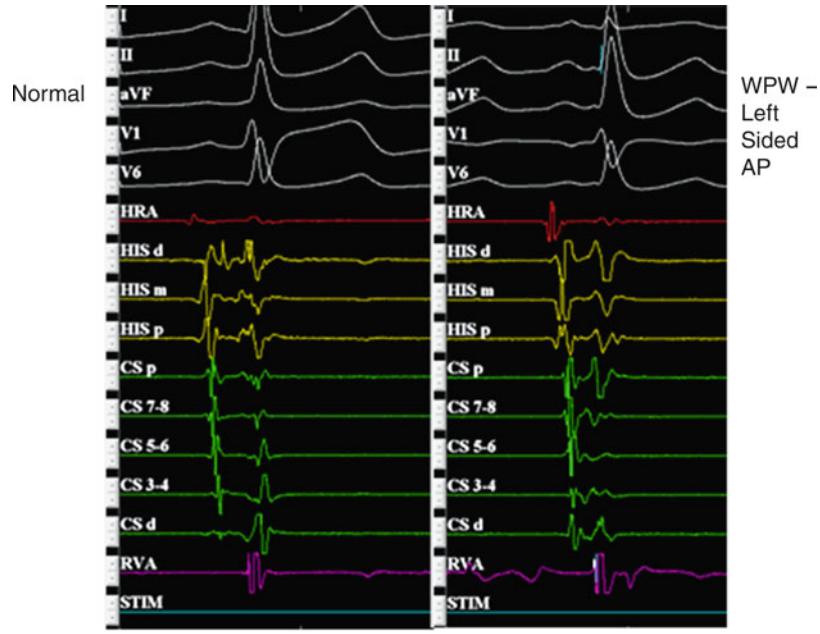

Fig. 20.20 Example of a patient with WPW and a *left*-sided accessory pathway. The tracings on the *left* show a patient in sinus rhythm with no preexcitation and a normal AH and HV intervals. The tracings on the *right* in a patient with WPW demonstrate an HV interval of 0 ms with earliest ventricular activation in the CS5–6 pair indicative of a *left*-sided accessory pathway

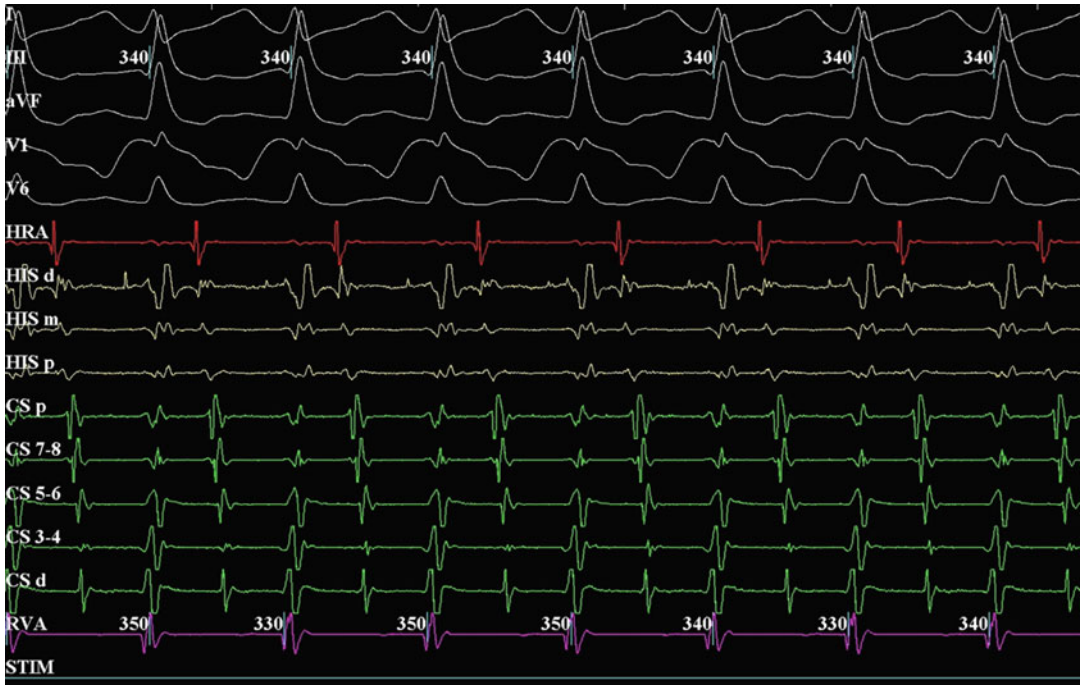

Fig. 20.21 Orthodromic reentrant tachycardia with a *right*-sided pathway. Note that the earliest atrial depolarization in tachycardia is the high right atrial pairs suggesting a *right*-sided pathway location

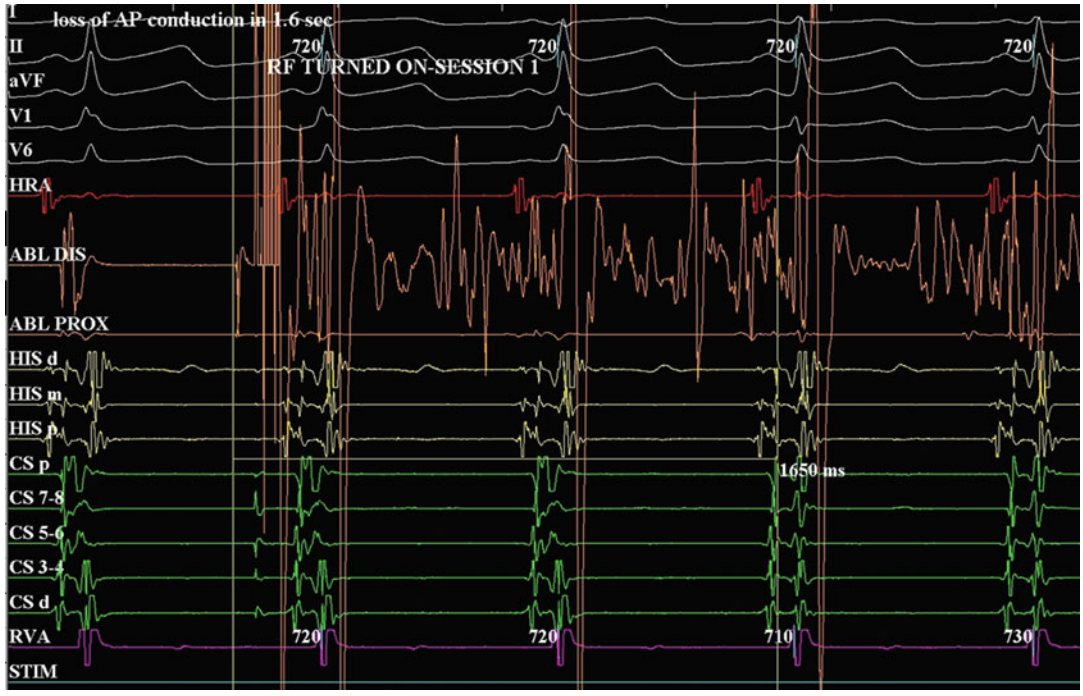

Fig. 20.22 Loss of AP conduction in a patient with WPW and a *left*-sided pathway. In this example radiofrequency energy is turned on producing a notable artifact on the ablation catheter (ABL DIS) and loss of accessory pathway conduction in less than 1.650 s. This is best seen by the change in the QRS morphology in surface lead V1 and the change in intracardiac conduction seen in the coronary sinus lead CS5–6

or in a catheter placed on the tricuspid annulus away from the atrial septum, the possibility of a right-sided accessory pathway would be raised (Fig. 20.21).

Once the above measurements and observations have been made, provocative testing is typically conducted. This includes determination of refractory periods of the atrium, AV node, accessory pathways (if present), and ventricle. Additionally, refractory periods of the ventricle, AV node, and accessory pathways (if present) are also routinely measured and recorded.

Following the above maneuvers, mapping of the arrhythmia substrate is performed (Fig. 20.22). Following mapping and ablation, it is routine to test for at least 20–60 min to confirm that the ablation was successful and there are no other mechanisms for tachycardia. If these two criteria have been established, catheters and vascular sheaths are removed and the patient awakened from anesthesia/sedation.

Overview of Therapies

Over the past 25 years, pediatric cardiac electrophysiology has markedly evolved from a predominantly diagnostic specialty to an interventional one. With the advent of catheter ablation, it is now possible to permanently cure most non-channelopathy-induced arrhythmias that previously required lifelong drug therapy. Ablation has become the treatment of choice for most supraventricular arrhythmias in children above the age of 5–8 years, and the technology has even been applied safely and effectively to younger and smaller patients [93, 94, 100–102]. The understanding of channelopathies has blossomed in recent decades, and the possibility for

personalized, gene-directed therapies is no longer the stuff of fiction [103, 104]. It is anticipated that continued work in the molecular genetics of channelopathies will continue to yield new therapies aimed at the source of arrhythmias in these patients.

From the perspective of device-based therapy, the technological advances in pacemakers and implantable defibrillators have allowed for expansion of their use in even the smallest patients [105, 106]. Additionally, newer data are helping refine which patients are most appropriate to receive these devices [107, 108]. Newer, less invasive technology is allowing further expansion of device therapy to patients who were not previously considered candidates, but further studies will be needed to demonstrate if this anticipated expansion in therapies will be justified or appropriate [109–111].

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