Of the benefits which graphic methods have conferred upon practical medicine, it is my desire to speak but briefly. These records have placed the entire question of irregular or disordered mechanism of the human heart upon a rational basis..., they have influenced prognosis..., they have potentially abolished the promiscuous administration of certain cardiac poisons, and have clearly shown the line which therapy should follow.1

Thomas Lewis, 1924

Electrocardiography, accurate physical examination, and radiology form the tripod on which rests the clinical diagnosis in pediatric cardiology. Omission of, unfamiliarity with, or misinterpretation of any of these three tools spells disaster.2

Alexander S. Nadas, 1957

The optimism of Sir Thomas Lewis and the cautions of Dr. Nadas regarding the electrocardiogram (ECG) remain valid even in this day of sophisticated echocardiography, Doppler flow analysis, and magnetic resonance imaging. Although it must be admitted that fine details of cardiac anatomy are now best evaluated with these modern techniques, the ECG is not (and never will be) obsolete. It is still the quickest, safest, and least expensive diagnostic tool in cardiology and is unparalleled in its ability to register arrhythmias and conduction defects. With proper interpretation, the ECG also offers a useful reflection of cardiac position, chamber enlargement, myocardial damage, and certain metabolic disorders. It has clearly proven its worth after more than a century of continuous clinical use.

This chapter is intended as a review of electrocardiography as it applies to the pediatric patient. Rather than simply catalogue a litany of rules for ECG interpretation, we have expanded the discussion here to encompass the basic cellular events underlying cardiac electrical activity, along with a survey of invasive and noninvasive techniques used for in-depth analysis of cardiac rhythm and conduction patterns. This information is intended not only to clarify the origin of ECG signals recorded from the body surface but also to serve as an introduction to the broader topic of cardiac arrhythmias that will be addressed further in Chapter 29 of this text.

BASIC ELECTROPHYSIOLOGY

Cellular Action Potential

The ECG is several steps removed from electrical activity at the cellular level, but the two are intimately related. Cellular events can be recorded directly using microelectrodes equipped with tips that are small enough to pierce individual cell membranes. When a microelectrode invades a normal cardiac cell, it encounters a field of net negative charge relative to the outside environment. This is the
diastolic resting potential of the cell, which is maintained by
the selective permeability of membrane channels to certain
ions, as well as the operation of membrane ion pumps. If the
cell interior becomes slightly depolarized (i.e., less nega-
tively charged), it may reach a critical value referred to as
the threshold potential. At this point, an abrupt change in
membrane channel properties allows a sudden flood of posi-
tive ions to enter the cell, and an action potential develops.

Two general types of cardiac action potentials can be
observed. The most common, known as the fast response or
sodium channel type, normally occurs in cells of atrial muscle,
ventricular muscle, His-Purkinje cells, and probably accessory
atrioventricular (AV) conduction tissue (e.g., Wolff-Parkinson-White syndrome). These cells generally register
a resting potential at about $-90$ mV and depend on sodium
ions as the positive charge carrier for their initial rapid
phase 0 depolarization (Fig. 12-1). Immediately after phase 0,
there is a complex sequence of activation and deactivation
for the various ion channels involved with potassium, sodium,
chloride, and calcium flux. This proceeds in an orderly
pattern that initially maintains the net intracellular charge in
balance with the outside environment for a period known as
the phase 2 plateau, but eventually progresses to phase 3
repolarization to restore the cell back to its phase 4 resting
state.

A second variety of action potential, referred to as the slow response or calcium channel type, occurs predomi-
nantly in cells of the sinoatrial (SA) node and the AV node.
It is distinguished by a resting potential of about $-60$ mV and
has a less acute upstroke for the initial phase 0 depolarization.
These cells utilize calcium along with some sodium to provide
the inward ionic current for depolarization (Fig. 12-2). An important feature of slow response cells is the property
of automaticity. Spontaneous upward drift of the diastolic
potential during phase 4 enables the cell to reach threshold
of its own accord and thereby act as a natural pacemaker for
the heart (Fig. 12-3). Some fast response cells are also capa-
ble of spontaneous automaticity, but at much slower rates.

Cell-to-Cell Conduction

When a cardiac cell depolarizes, it usually stimulates
neighboring tissue in such a fashion that the activation
sequence is transmitted from cell to cell throughout the heart.
For this process to repeat smoothly, cells must have sufficient time to repolarize and recover between stimuli. If the initiating impulse is premature, the cells may not be prepared, or may be only partially prepared, for reactivation. The time needed to recover from a prior stimulus is known as the **refractory period**, which usually lasts until cells have nearly completed their repolarization process (late phase 3 or early phase 4). During the **absolute refractory period**, a cell does not respond in any way to a new impulse, regardless of the stimulus strength. During the **relative refractory period**, a cell may respond occasionally, but only if the premature stimulus is sufficiently strong. During the early phase of the relative refractory period, a cell sometimes responds to a premature stimulus with an incomplete and low-amplitude action potential that is too weak to propagate any further to cells downstream. This is designated as the **effective refractory period** (ERP). The distinction between absolute and effective refractoriness is subtle but important because the ERP may be measured in the intact heart with clinical electrophysiologic techniques. For practical clinical purposes, the effective and absolute refractory periods can be considered similar (Fig. 12-4).

### Conduction through the Intact Heart

A normal heartbeat begins with the spontaneous depolarization of a cell in the SA node, located at the junction of the superior vena cava and right atrium in the area of the sulcus terminalis. This event then activates adjacent atrial muscle cells so that a wave of depolarization spreads out from high in the right atrium like ripples in a pond. The wavefront reaches the lower parts of the right atrium after about 30 msec and finishes at the lateral part of the left atrium after about 80 msec. The electrical activity from SA node depolarization is too small to be recorded from the body surface, but atrial muscle cell depolarization is clearly registered as the P wave on the ECG. The P wave corresponds to phase 0 of the action potentials from individual atrial myocytes and reflects the leading edge of the depolarization wavefront as it travels from cell to cell. Once all atrial cells have undergone their initial rapid depolarization and entered the phase 2 plateau, the P wave is complete. Phase 3 repolarization of atrial cells causes a very small deflection on the surface ECG, referred to as the Tₐ wave. This wave is rarely seen because it is usually obscured by the QRS complex, but under special conditions such as heart block, the atrial repolarization wave may be appreciated (Fig. 12-5).

As the atrial activation wavefront passes through the lower right atrium, depolarization of the AV node is initiated. This node is a complex interface consisting predominantly of slow response cells located in an anatomic region referred to as the **triangle of Koch** (Fig. 12-6). Conduction velocity within the AV node is relatively slow, and it varies according to the timing of atrial impulses. Premature beats or accelerated atrial rhythm exaggerate AV nodal delay in a gradual and progressive manner described as **decremental conduction**, which can ultimately produce the stereotypic sequence of conduction block that is easily recognized as Wenckebach periodicity (Fig. 12-7). This pattern is rather specific to slow response cells. Conduction through fast response cells, by contrast, tends to be all or none, with a fairly fixed conduction velocity.

Electrical activity within the AV node is not directly registered on the surface ECG. One must rely on upstream events (P wave) and downstream events (QRS complex) as indirect measures of the process. On the surface ECG, the

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**FIGURE 12–4** Diagrammatic fast response action potential showing the time course of refractoriness and excitability. During the **absolute refractory period** (ARP), the cell cannot be re-excited regardless of stimulus strength. During the **relative refractory period** (RRP), only large-amplitude stimuli can re-excite the cell. Some cells may display a supernormal period (SNP) at the end of phase 3, which permits excitability with low-amplitude stimuli. From Fyler DC. Nadas’ Pediatric Cardiology. Philadelphia: Hanley & Belfus, 1992.

PR interval provides a rough estimation of AV node conduction, but there is actually much more to this interval than AV node activity alone. To be precise, the PR interval includes conduction times: (1) from high to low in the right atrium, (2) in the AV node proper, and (3) in the His-Purkinje system. To dissect the PR interval into these individual components, an intracardiac electrode catheter can be positioned across the medial aspect of the tricuspid valve as part of an electrophysiologic study to straddle an area near the bundle of His. This specialized recording reveals localized low right atrial activation, followed by a small deflection from the common bundle of His, and finally the local right ventricular activation. An index of true AV node conduction time can be obtained by measuring the interval between low right atrial depolarization and the initial depolarization of the bundle of His, which is referred to as the AH interval (Fig. 12-8).

Beyond the AV node, the excitation process enters the common bundle of His. This bundle crosses the AV junction on the right ventricular side, just beneath the membranous septum (see Fig. 12-6). Cells of the His-Purkinje system have fast response action potentials, and generally rapid conduction velocity. The propagated impulse traverses the common bundle of His, splits into right and left bundle branches, and quickly exits from the terminal Purkinje fibers to begin activation of ventricular myocytes, all in about 40 msec. On the intracardiac His recording, the time from the initial His deflection to the beginning of ventricular activation (HV interval) is an accurate measure of His-Purkinje conduction time (see Fig. 12-8).

Conduction along the His-Purkinje network occurs as the cells are depolarizing (i.e., during phase 0). Their repolarization begins long after the excitation wavefront has passed on to ventricular muscle. Indeed, final repolarization of His-Purkinje cells tends to be one of the last electrical events during a cardiac cycle and probably contributes to some portion of the small terminal U wave seen occasionally on the normal ECG.

As the excitation wavefront leaves the His-Purkinje system, phase 0 of ventricular myocyte depolarization begins. The His-Purkinje system is highly arborized in its terminal portion, and these multiple exit sites promote depolarization of several different ventricular regions at one time, a process that is further complicated by the heart's threedimensional geometry. To better visualize ventricular activation, it is useful to divide events into small time segments and examine the order of regional depolarization. A simplification of the sequence is shown in Figure 12-9, beginning with left-to-right septal activation, followed by
activation of the left and right apex, the endocardium of the right and left ventricular free walls, the free wall epicardium, the base of the left ventricle, and finally, the right ventricular outflow tract. The advancing wavefront in each region generates a signal that can be recorded from the body surfaces as the QRS complex.

The QRS is complete once all ventricular cells have depolarized (usually within 90 msec). The ST segment is registered while the cells remain at the phase 2 plateau. At the onset of phase 3 repolarization, inscription of the T wave begins. Repolarization is less homogeneous than depolarization; hence, there is a relatively protracted duration for the T wave compared with the QRS. Additionally, the repolarization sequence is just the opposite of depolarization and appears to follow the reverse direction of epicardium toward endocardium. When all ventricular myocytes are back to phase 4, the T wave is complete.

**Graphic Recording of Cardiac Electrical Activity**

It should be apparent from the preceding discussion that the signal registered on the surface ECG occurs during abrupt changes in cellular conditions, with the P wave and QRS complex marking the acute phase 0 transition from resting potential to the fully depolarized state, and the T, A, T, and U waves marking phase 3 repolarization back to the diastolic resting potential. In fact, when all cells in a given cardiac chamber are either fully depolarized (phases 1 and 2) or fully repolarized (phase 4), the signal recorded from the body surface has the same isoelectric appearance even though intracellular conditions differ dramatically. Thus, what is measured with an ECG is not cellular voltage, but rather the current that arises at the boundary between depolarized and repolarized cells as activation and deactivation wavefronts move through a cardiac chamber. This boundary acts as a dipole, which generates current because of the presence of opposing charges in front and behind. Movement of this dipole relative to an ECG electrode produces the electrocardiographic signal (Fig. 12-10). During depolarization, the leading edge of the activation wavefront is charged positive, and the trailing edge negative. Movement toward a recording electrode results in an upward deflection on the ECG, and movement away results in a downward deflection. Repolarization wavefronts have a negative leading edge and generate signals with just the opposite deflections. In addition to direction, electrocardiographic signals...
can be further qualified by amplitude, which is largely proportional to the number of cells being stimulated at a given time by the wavefront. Thus, these electrical events can be described as a series of vectors, with directionality reflecting the path for the wavefront, and amplitude (voltage) reflecting the muscle mass involved.

The simplest example of body surface recording is the cycle of atrial activation, shown in Figure 12-11 with three hypothetical ECG leads on the left arm, right arm, and leg. The normal atrial depolarization wavefront advances from the upper right atrium, resulting in a mean vector that inscribes positive P waves in the left arm and leg leads, with a negative deflection in the right arm lead. Repolarization follows in the same path after a short isoelectric period, generating a Tₚ of opposite polarity.

The ventricular cycle results in a more complex series of vectors (Fig. 12-12). The process begins with septal activation, which generates a small negative deflection (Q wave) in the left arm lead and a small positive deflection in the leg lead. The right arm, being nearly at right angles to this vector, records little activity at this point. With left apex depolarization, the vector abruptly shifts leftward and inferior, beginning the inscription of an R wave in the left arm, further increasing the positive amplitude of the leg recording, with a negative deflection now appearing in the right arm lead. As events proceed to the ventricular free walls, there is competition between the simultaneous vectors of left- and right-sided depolarization. In a normal heart, the left ventricle wins out by virtue of its larger muscle mass, and the resultant net vector continues leftward. Final depolarization of the left base and right outflow tract generates superior and rightward vectors, causing inscription of a negative S wave in the left arm and leg leads, with a positive deflection in the right arm lead.

The repolarization process of the ventricle is even more involved. Unlike the atrium, the vectors of repolarization do not exactly retrace the same steps as depolarization because ventricular myocytes generally repolarize in an
epicardial to endocardial direction. For this reason, the
direction of the T wave is similar to that of the QRS, and
not reversed as in the case of atrial tissue or simple experi-
mented models of isolated cardiac muscle fibers (Fig. 12-13).
The basic principles of cardiac excitation and recording
are summarized in Figure 12-14. The interested reader is
referred to several comprehensive reviews3–8 for further
details of these topics.

THE ELECTROCARDIOGRAM

Lead Systems and Technique

The standard ECG evolved from a three-lead system
introduced by Einthoven to a 15-lead tracing in current
use for pediatric recording. The two major lead groupings
include the limb leads and the precordial leads. The limb
leads can be further divided into Einthoven’s standard
bipolar system (I, II, and III), and an “augmented” varia-
tion of Wilson’s unipolar lead system (aVR, aVL, and aVF).
Einthoven’s leads record potentials between electrode
pairs: left arm (positive) to right arm (negative) = lead I;
left leg (positive) to right arm (negative) = lead II; and left
leg (positive) to left arm (negative) = lead III. Wilson’s
leads record from a single limb in reference to a zero
potential central terminal: right arm (positive) = aVR; left
arm (positive) = aVL; and left leg (positive) = aVF. A wave-
front moving toward the positive terminal of one of these
leads registers a positive deflection on the ECG. These leads

form a compass around the frontal plane, which is divided
into 360 degrees, with lead positions and degree coordi-
nates as shown in Figure 12-15.
The precordial leads (V4R through V7) view the electrical
activity in the horizontal plane. They are all unipolar (posi-
tive) and are referenced to a zero potential central terminal,
but without augmentation. Electrode placement is slightly
modified in pediatric studies to obtain lead positions far
out on the right side of the chest and laterally on the left
side of the chest (Fig. 12-16).

Routine recordings are made with a chart paper speed
of 25 mm/sec and are usually standardized with an amplitude
response of 1 mV/10 mm. If a patient’s ventricular voltages
are exceptionally large, the amplitude response should be
reduced to 1 mV/5 mm or even 1 mV/2.5 mm to avoid QRS
overshoot and superimposed signals. However, whenever
nonstandard amplification is used for ECG display, the
appropriate calibration mark must be clearly highlighted on the recording.

**The Normal Electrocardiogram**

The normal values referred to in this discussion have been drawn from our experience at Children's Hospital Boston and the classic publications of Davignon and colleagues,8 Garson,9 and Fisch.10 Data are abstracted for quick reference in Table 12-1. The ECG should be read in a systemic fashion, beginning with measurements of axes and intervals, followed by waveform analysis, all of which must be synthesized into a final impression based on history and physical examination.

**Axis**

The electrical axis refers to the predominant direction (or mean vector) of a waveform in the frontal plane. By identifying the limb lead with the largest positive deflection for the waveform in question, and remembering the coordinates for this lead on the frontal compass face, one can assign a value in degrees for the mean axis. The easiest example involves the P wave. Because normal atrial activation begins at the SA node and spreads through the atrium high-to-low and right-to-left, the wavefront of depolarization flows toward the southeast quadrant of the frontal plane. Lead II (+60 degrees) best records this area and usually registers the largest positive P wave. A lead from the northwest quadrant (aVR) simultaneously registers a deep negative P wave. Leads that record at nearly right angles to the P-wave vector (aVL and III) are equiphasic or isoelectric, with relatively low amplitude. The P-wave axis for a normal heart in sinus rhythm should be between 0 and +90 degrees regardless of the patient’s age. An abnormal axis can be seen in ectopic atrial rhythms or atrial malpositions.

The mean QRS axis is calculated in a similar fashion by identifying the limb lead with the largest positive R wave and assigning the corresponding degree value. In contrast to the P-wave axis, the normal QRS axis has wide age-dependent variation. In the case of newborns, for whom the right ventricle is relatively hypertrophied by virtue of its intrauterine workload, the axis is directed rightward, usually about 120 degrees. As the left ventricle becomes relatively more dominant during the first 6 months of life, the axis gradually shifts toward +60 degrees and should remain between about 0 and +90 degrees thereafter. An abnormal QRS axis can be seen with ventricular hypertrophy, malpositions, intraventricular conduction disturbances, and infarction.

The T-wave axis is usually concordant with the QRS in the frontal plane. There may be some discrepancy in the early months of life, but by the time the child is 6 months old, the QRS and T axes should not differ by more than 60 degrees. An abnormal T-wave axis can be seen in marked
TABLE 12–1. Normal Range and Mean Value for Selected Electrocardiogram Measurements in Children*

<table>
<thead>
<tr>
<th>AGE</th>
<th>0-7 days</th>
<th>1 wk–1 mo</th>
<th>1 mo-6 mo</th>
<th>6 mo-1 yr</th>
<th>1 yr-5 yr</th>
<th>5-10 yr</th>
<th>10-15 yr</th>
<th>&gt;15 yr</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rate (beats/min)</td>
<td>90-160 (125)</td>
<td>100-175 (140)</td>
<td>110-180 (145)</td>
<td>100-180 (130)</td>
<td>70-160 (110)</td>
<td>65-140 (100)</td>
<td>60-130 (90)</td>
<td>60-100 (80)</td>
</tr>
<tr>
<td>QRS axis (degrees)</td>
<td>70-180 (120)</td>
<td>45-160 (100)</td>
<td>10-120 (80)</td>
<td>5-110 (60)</td>
<td>5-110 (60)</td>
<td>5-110 (60)</td>
<td>5-110 (60)</td>
<td>5-110 (60)</td>
</tr>
<tr>
<td>PR lead II (msec)</td>
<td>80-150 (100)</td>
<td>80-150 (100)</td>
<td>80-150 (100)</td>
<td>80-150 (100)</td>
<td>80-150 (120)</td>
<td>80-150 (120)</td>
<td>90-180 (140)</td>
<td>100-200 (160)</td>
</tr>
<tr>
<td>QRS duration (msec)</td>
<td>40-70 (50)</td>
<td>40-70 (50)</td>
<td>40-70 (50)</td>
<td>40-70 (50)</td>
<td>40-70 (50)</td>
<td>45-80 (65)</td>
<td>45-80 (65)</td>
<td>50-90 (70)</td>
</tr>
<tr>
<td>Maximum QTc† (msec)</td>
<td>450 max</td>
<td>450 max</td>
<td>450 max</td>
<td>450 max</td>
<td>440 max</td>
<td>440 max</td>
<td>440 max</td>
<td>430 max</td>
</tr>
<tr>
<td>QRS V₁ Q (mm)</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>R (mm)</td>
<td>5-25 (15)</td>
<td>3-22 (10)</td>
<td>3-20 (10)</td>
<td>2-20 (9)</td>
<td>2-18 (8)</td>
<td>1-15 (5)</td>
<td>1-12 (5)</td>
<td>1-6 (2)</td>
</tr>
<tr>
<td>S (mm)</td>
<td>0-22 (7)</td>
<td>0-16 (5)</td>
<td>0-15 (5)</td>
<td>1-20 (6)</td>
<td>1-20 (10)</td>
<td>3-21 (12)</td>
<td>3-22 (11)</td>
<td>3-13 (8)</td>
</tr>
<tr>
<td>QRS V₃ Q (mm)</td>
<td>0-1 (0.5)</td>
<td>0-3 (0.5)</td>
<td>0-3 (0.5)</td>
<td>0-3 (0.5)</td>
<td>0-5 (1)</td>
<td>0-5 (1)</td>
<td>0-3 (0.5)</td>
<td>0-2 (0.5)</td>
</tr>
<tr>
<td>R (mm)</td>
<td>2-20 (10)</td>
<td>3-25 (12)</td>
<td>5-30 (17)</td>
<td>10-30 (20)</td>
<td>10-35 (23)</td>
<td>13-38 (25)</td>
<td>10-35 (20)</td>
<td>7-21 (13)</td>
</tr>
<tr>
<td>S (mm)</td>
<td>2-19 (10)</td>
<td>2-16 (8)</td>
<td>1-16 (8)</td>
<td>1-14 (6)</td>
<td>1-13 (5)</td>
<td>1-11 (4)</td>
<td>1-10 (3)</td>
<td>0-5 (2)</td>
</tr>
<tr>
<td>QRS V₅ Q (mm)</td>
<td>0-2 (0.5)</td>
<td>0-2 (0.5)</td>
<td>0-2 (0.5)</td>
<td>0-3 (0.5)</td>
<td>0-4 (1)</td>
<td>0-4 (1)</td>
<td>0-3 (1)</td>
<td>0-2 (0.5)</td>
</tr>
<tr>
<td>R (mm)</td>
<td>1-12 (5)</td>
<td>1-17 (7)</td>
<td>3-20 (10)</td>
<td>5-22 (12)</td>
<td>6-22 (14)</td>
<td>8-25 (16)</td>
<td>8-24 (15)</td>
<td>5-18 (10)</td>
</tr>
<tr>
<td>S (mm)</td>
<td>0-9 (3)</td>
<td>0-9 (3)</td>
<td>0-9 (3)</td>
<td>0-7 (3)</td>
<td>0-6 (2)</td>
<td>0-4 (2)</td>
<td>0-4 (1)</td>
<td>0-2 (1)</td>
</tr>
<tr>
<td>T-wave V₁ (mm)</td>
<td>0-4 days =</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4-7 days =</td>
<td></td>
<td></td>
<td></td>
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<td></td>
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</tr>
</tbody>
</table>

Adapted from references 8-10.

*Values reported as 2nd-98th percentile (mean), except for QTc (maximum value only) and >15 yr data, which report ± 1 SD.
†QTc as corrected by Bazett’s formula (QTc = QT / \sqrt{RR}).
hypertrophy with ventricular strain, ischemia, myopathy, and some intraventricular conduction disturbances.

Rhythm and Rate
Cardiac excitation arising from the SA node generates a P wave with a normal axis at a rate within the limits for age (see Table 12-1). Rate determination is usually a straightforward exercise (Fig. 12-17). Respiratory sinus arrhythmia is a normal finding in healthy children, as is the observation of a shifting atrial pacemaker, where a subsidiary P wave with a different axis than sinus rhythm takes over during episodic slowing of the SA node.

P Wave
The contour and amplitude of the P wave are an indirect measure of atrial size. The normal P wave should have a smooth dome shape in lead II and should never be taller than 0.3 mV or wider than 0.12 second in duration. Occasionally, there may be a small notch in the P wave of lead II, but this is acceptable if amplitude and duration fall within the normal range.

T_A Wave
The shallow wave of atrial repolarization is rarely seen on the normal ECG because of its low amplitude and superimposition of the QRS complex. It may be seen occasionally in patients with heart block when the QRS is delayed or dissociated. The normal T_A wave is directed opposite from the P wave and is usually less than 0.1 mV in depth. Atrial enlargement or inflammation distorts the T_A wave.

PR Interval
The PR interval is measured from the beginning of the P wave to the initial deflection of ventricular activation (it is more precisely a PQ interval). As noted earlier, the PR includes several electrical events, with AV node conduction accounting for the major portion. The normal PR interval is less than 0.16 second in young children or 0.18 second in adolescents and adults. A prolonged PR interval can be due to enhanced vagal tone, cardiac medications (digoxin and antiarrhythmic agents), or disease involving either the AV node or the His-Purkinje system. A short PR interval (less than 0.08 second) may be observed in Wolff-Parkinson-White syndrome.

QRS Complex
Registration of the QRS begins when cardiac excitation leaves the His-Purkinje system and the ventricular myocytes begin to depolarize. The QRS complex is evaluated for its morphology, amplitude, and duration.

QRS morphology is dictated by the sequence of regional ventricular activation, and the balance (right versus left) of ventricular muscle mass, as previously discussed. The normal heart leaves a characteristic QRS shape in each lead of the ECG, which may change because of distorted activation sequence or hypertrophy. Beyond infancy, the normal pattern is one of a small Q wave, followed by a large R and a small S wave in left-sided leads (I, II, or aVL, V_3–V_6), whereas right-sided leads (aVR, III, V_4R–V_2) typically register a small R followed by a deep S wave.

QRS amplitude is a more quantitative measure of ventricular mass and compliments QRS morphology when evaluating a trace for hypertrophy. Normal values are established for R-wave amplitude, as well as for Q- and S-wave amplitudes, in each individual lead. Measurements in the precordial leads are particularly sensitive indicators of an abnormality. Normal amplitude values vary widely with patient age, and tables of normal data should be on hand during review of all pediatric ECGs.

The duration of the QRS complex is related to the speed of conduction within the His-Purkinje system, as well as from myocyte to myocyte within the ventricles. Duration increases slightly with age. In normal infants, the QRS width should be less than 0.08 second, and in those older than 6 months, less than 0.10 second. Prolongation of the QRS may be seen with block of His-Purkinje conduction (bundle branch block), slow myocyte conduction (due to muscle injury, drugs, or electrolyte disturbances), severe ventricular hypertrophy, and some cases of preexcitation.

ST Segment
Ventricular muscle cells are in the plateau phase of their action potential (phase 2) during the ST segment. Because no electrical wavefronts are advancing or retreating through the heart, the body surface recording is normally isoelectric. The J point at the termination of the S wave marks the beginning of the ST segment and should not deviate more than about 1 mm from the baseline.
Deviations in ST level may be caused by ischemia, inflammation, severe hypertrophy, and some medications. One normal variant is the *early repolarization* pattern, seen occasionally in healthy adolescent patients, where the J point can be elevated 2 to 4 mm. Usually, this elevation is observed in the lateral (V4–V6) and inferior (II, III, aVF) leads and is accompanied by strikingly tall T waves in the same leads (Fig. 12-18). The diagnosis of benign early repolarization should not be made if the elevation is more than 4 mm or the T wave is of low amplitude.

**T Wave**

The T wave corresponds to phase 3 repolarization of ventricular myocytes. The normal T-wave amplitude is variable and is not routinely quantitated. On the other hand, the direction of the T wave deserves careful attention. As mentioned, the T-wave axis should follow the same net direction as the QRS in the frontal plane within about 60 degrees, and discordance of the axis in the limb leads may suggest pathology. The precordial leads follow somewhat different rules for T-wave direction. Over the left chest, the T wave is still normally concordant with the QRS, but there are important age-dependent variations in the rightward leads. From birth to 4 to 7 days of age, the T wave is upright in all precordial leads. After this, the T wave becomes negative over the right chest (V_{4R}-V_1), while remaining positive in the left chest leads. This pattern persists until adolescence, when the T waves tend to resume an upright direction in all chest leads. This sequence is critical to remember during analysis of ECGs in children because an upright T wave in the right precordial leads between the age of 7 days and early adolescence is a potential indicator of right ventricular hypertrophy.

**QT Interval**

The QT interval is a reflection of the total action potential duration for ventricular myocytes. It is measured from the onset of the QRS to the point of T-wave termination. Because the normal QT interval varies with heart rate (longer at slow rates, shorter at fast rates), the measurement is adjusted with the formula: QT (seconds)/square root R-R (seconds). This rate-corrected interval (QT\(_c\)) should be less than 0.45 second in infants, 0.44 second in children, and 0.43 second in adults. A prolonged QT\(_c\) interval can be of dramatic clinical significance. The hereditary long QT syndromes are potentially fatal disorders, and early detection on an ECG is imperative. The QT\(_c\) is also prolonged by many antiarrhythmic drugs and by some electrolyte imbalances.

**U Wave**

The U wave is thought to reflect the relatively late repolarization process of His-Purkinje cells and certain left ventricular myocytes. It is not always seen on the ECG of normal patients. When present, a normal U wave is of low amplitude (less than one fourth the height of the T wave) and has the same polarity as its T wave. When the U wave is abnormally prominent (more than half the height of the T wave), it should be included in the measurement of the QT interval. Occasionally, the amplitude of the U wave may become accentuated secondary to hypokalemia, antiarrhythmic drugs, and some forms of long QT syndrome.

**The Abnormal Electrocardiogram**

The ECG should never be interpreted in isolation, and there should always be specific questions to answer when the test is ordered regarding rhythm, hypertrophy, myocardial injury, and so forth. Additionally, one should constantly bear in mind that a child can have serious heart disease with a normal-appearing ECG, particularly in the first few days of life.

**Rate and Rhythm**

Cardiac rhythm should be the first item scrutinized on the ECG tracing. If the rhythm is abnormal, many of the assumptions regarding hypertrophy, malpositions, ischemia, and so forth will become invalid. To evaluate these issues accurately, the ECG needs to be repeated after sinus rhythm is restored. A detailed discussion of cardiac arrhythmias is presented in Chapter 29.

**Cardiac Malpositions**

Chamber orientation is reflected on the ECG by the axis and the morphology of the P wave and the QRS complex. Atrial situs is determined with considerable accuracy by deciding which side of the atrium contains the SA node. In situs solitus, the SA node activates high in the right atrium, and the resultant atrial depolarization wavefront generates a P-wave axis of about +60 degrees, whereas in situs inversus, the activation emanates from high in the left-sided atrium.
so that the P axis is about +120 degrees (Fig. 12-19). Variable patterns may be seen in the heterotaxy syndromes. For example, patients with asplenia may have bilateral SA nodes, and the P-wave axis may alternate or fuse between +60 and +120 degrees. In polysplenia, there may not be a true SA node, and such patients usually rely on a subsidiary atrial pacemaker focus that can have a variable location.

Gross ventricular orientation is best estimated from the precordial leads. Normal levocardia has a characteristic pattern of relatively low (or predominately negative) voltage in the right chest leads (V4R–V2) with positive forces of higher amplitude in the mid and left chest leads. In dextrocardia (Figs. 12-19 and 12-20), the pattern is classically reversed. Further insight into ventricular anatomy may be gained by determining the embryologic ventricular looping. The normal D-loop orientation has an anatomic right ventricle and tricuspid valve located on the right side of the heart. In an L-loop anomaly, the ventricular relationship is inverted; hence, the septal activation wavefront must travel right-to-left. This changes the QRS morphology to one of initial Q waves in right-sided limb or precordial leads, with small initial R waves on the left side (see Fig. 12-20).

**Atrial Enlargement**

The surface ECG is a fair indicator of atrial enlargement. Because the right atrium is the first to depolarize, indicators of right-sided enlargement are found in the early portions of the P wave. The diagnostic criterion for isolated right atrial enlargement is the presence in lead II of a peaked narrow P wave greater than 0.30 mV in amplitude, accompanied by either a tall P wave or a biphasic P wave with an early deep negative deflection in lead V1 (Fig. 12-21). Left atrial enlargement is reflected in the terminal portion of the P wave. The classic findings include a broad, notched P wave in lead II (duration greater than 0.10 to 0.12 second) or a deep slurred terminal portion of a biphasic P wave in V1 (Fig. 12-22). A combination of the above amplitude and duration criteria is indicative of biatrial enlargement (Fig. 12-23).

**Ventricular Hypertrophy**

Identification of ventricular hypertrophy from the surface ECG is far from a perfect science. Although criteria are generally accurate for right ventricular hypertrophy (RVH), the diagnosis of left ventricular hypertrophy (LVH) is sometimes difficult until the process is far advanced.

**Right Ventricular Hypertrophy**

Screening for RVH is particularly important in children because the more common congenital defects impose an
increased work load on this chamber. Fortunately, the criteria that have evolved are fairly sensitive.

**R-Wave Amplitude in V1 Higher than the 98th Percentile for Age.** This finding is very specific outside the newborn period. The height of the R wave in this lead correlates well with right ventricular systolic pressure and is sufficiently quantitative to allow prediction of right ventricular pressure for isolated pulmonary valve stenosis using the formula: R-wave height (in millimeters) \times 5 = peak systolic pressure (mm Hg).

**FIGURE 12–22 ECG showing left atrial enlargement.**

**FIGURE 12–23 ECG showing biatrial enlargement.**

**Abnormal T-Wave Direction in V1.** As previously mentioned, the T-wave direction in lead V1 changes with time: it is upright in newborns, negative beyond the age of 7 days, and positive again in adolescents and adults. A persistently upright T wave after the seventh day of life is a sensitive indicator of elevated right ventricular pressure, and when combined with R-wave amplitude, even greater precision is possible. Mild degrees of RVH may show a normal R-wave amplitude but an upright T wave in V1. Moderate RVH is characterized by abnormal height of the R wave in conjunction with the upright T wave. In marked RVH, the R wave remains excessive, but the T wave may now be deeply inverted in what is referred to as a "strain" pattern (Fig. 12-24).

**S-Wave Depth in V6 Lower than the 98(th) Percentile for Age.** This measurement is useful in patients with increased right ventricular pressure secondary to chronic lung disease. Respiratory disorders such as cystic fibrosis can lower the voltage pattern recorded from the right chest because of heart rotation and hyperexpansion of the lungs. Despite low anterior forces, RVH can still be diagnosed when the lateral S wave is deep. This pattern of RVH, when associated with right atrial enlargement in a

**FIGURE 12–24 ECG patterns with varying degrees of right ventricular pressure load hypertrophy.** A, Mild RVH in a 9-month-old suggested by an upright T wave in V1, but without excess R-wave voltage. B, Moderate RVH in a 4-year-old with an upright T wave and excess R-wave voltage in V1. C, Marked RVH in a 7-year-old showing a very tall R wave with an inverted T wave ("strain" pattern) in V1.
patient with severe pulmonary disease, is characteristic of cor pulmonale (Fig. 12-25).

Right Axis Deviation. Isolated right axis deviation is not specific for RVH and may be observed in conduction disturbances such as left posterior hemiblock. When present in conjunction with other RVH criteria, it lends additional support for the diagnosis.

QR Pattern in V1. This criterion is likewise not absolute for RVH but is supportive evidence when associated with a tall R wave in the right chest leads. A QR pattern may also be seen with L-loop ventricles and anterior infarction.

RSR′ Pattern in V1. It is important to understand the significance and limitations of this finding in children. Increased right ventricular volume loads imposed by common lesions such as an atrial septal defect may create a pattern of R′ of a small initial R wave, followed by an S wave, terminating with a tall R′ wave (Fig. 12-26). A diagnosis of RVH should be made only when the secondary R′ wave is large in amplitude. Some normal children may have a similar pattern with a lower-amplitude R′ wave. It is also useful to examine the distribution of the RSR′ pattern in multiple precordial leads because large right ventricular volume loads may cause the RSR′ pattern to extend from V4R all the way across V3 or V4, whereas the pattern does not usually extend beyond V1 in normal children. An RSR′ pattern may also be caused by incomplete right bundle branch block.

Abnormal R/S Ratio in V1 or V6. Normal values for R/S ratios are well established, and these data can be drawn on when the decision regarding RVH is questionable. However, it is rare to see abnormal ratios as an isolated finding, and one should hesitate to make a firm diagnosis of RVH on the basis of this criterion alone.

Left Ventricular Hypertrophy

It is difficult to predict LVH with certainty from the ECG. The diagnosis is best entertained when multiple criteria are fulfilled.

R-Wave Amplitude in V5–V6 Greater than the 98th Percentile for Age. The voltage criteria for LVH are not very exact. Hypertrophy may be present with normal left precordial forces, and in some normal children (particularly athletic teenagers), the R-wave amplitude may exceed the 98th percentile. Attempts have been made to improve diagnostic accuracy by examining the reciprocal S-wave depth in lead V1 as an indicator of LVH using isolated S-wave measurement or a combination of S in V1 plus R in V6, but there remain limitations to voltage data alone.

Lateral T-Wave Inversion (Strain Pattern). In the Natural History Study of congenital aortic stenosis, T-wave abnormalities were identified as the most specific indicators of LVH. Left ventricular strain presents a pattern of inverted T waves in the inferior limb leads (II, III, aVF) and left precordial leads (V5 and V6), sometimes associated with depression of the ST segment. There may or may not be voltage indications of LVH (Fig. 12-27). Although the
presence of these T-wave changes usually suggests advanced
degrees of hypertrophy, the presence of ischemia or myocardial
inflammation must be excluded before this criterion
can be applied with certainty.

**Left Axis Deviation.** An abnormal leftward axis is
supportive evidence for LVH. The utility of this criterion is
best appreciated in the neonate, in whom the QRS axis
is normally directed rightward. The presence of a mature
axis in the 0 to +90 degree range or a superior axis in the 0
to −90 degree range in early infancy suggests a definite
cardiac abnormality. Left axis deviation may also be due to
conduction disturbances such as left anterior hemiblock.

**Abnormal Lateral Q Wave.** The Q wave in leads V5
and V6 (septal depolarization) may be distorted if the left
ventricle is very dilated or markedly thick. There can be
deviation and rotation of septal position, and increased
competition from vectors of left apex and left free-wall
depolarization. As a broad generalization, a dilated volume-
loaded left ventricle tends to have an abnormally deep
Q wave in the leftward leads in lesions such as aortic regurgi-
tation, patent ductus, or ventricular septal defect (Fig. 12-28).
Concentric hypertrophy from a pressure load such as aortic
stenosis is more likely to be associated with a small or
absent Q wave (Fig. 12-29).

**Single Ventricle Hypertrophy**

There are no firm criteria to apply for hypertrophy of a
single ventricle in complex congenital anomalies. What is
most surprising is that an ECG in such conditions can look deceptively normal at times, at least for the newborn. However, voltage criteria for single ventricles generally exceed those for either RVH or LVH in some precordial lead as the children age. The exact QRS morphology is variable, depending on the presence and location of the ventricular septum, which anatomic ventricle is indeed present, and the rotation of the abnormal heart in the thorax. One may be able to predict which ventricle is absent by noting which side of the precordium has deficient positive voltage (Fig. 12-30). Absent ventricle (or hypoplastic ventricle) may also be suspected if a septal Q wave is not seen in any precordial lead.

**Intraventricular Conduction Abnormalities**

From the common bundle of His, intraventricular conduction fibers divide into the right and left bundle branches. The left bundle actually fans out along the entire left ventricular septal surface, but may be considered to split into two major divisions: the left anterior and left posterior fascicles. Partial or complete block at any one of these sites creates delay in regional ventricular activation and a characteristic change in QRS pattern. Likewise, the presence of an accessory conduction pathway distorts regional activation.

**Incomplete Right Bundle Branch Block**

An RSR′ pattern in the right precordial leads with normal QRS duration may indicate an incomplete conduction disturbance in the right ventricle. However, an identical pattern may be seen in healthy normal individuals or in patients with right ventricular volume overload, as previously discussed. The diagnosis of incomplete block should be reserved for situations in which RSR′ is associated with a slightly prolonged QRS duration in the absence of a left-to-right shunt at the atrial level.

**Complete Right Bundle Branch Block**

When transmission is interrupted along the right bundle branch, the septum and left ventricle can activate normally, but the right ventricle must depend on slower cell-to-cell activation spreading left to right. The resultant QRS complex has prolonged duration (greater than 0.10 second for infants, greater than 0.12 second for older patients) and a characteristic morphology that reflects the late, slow activation wavefront spreading toward the right heart. The initial portion of the QRS is generated by the usual septal and initial left ventricular depolarization and thus is quite similar to normal (small R wave in V1, QR in V6). The subsequent slow wavefront traveling toward the right heart inscribes a tall shurred R′ wave in V1 and an equally sluggish S wave in V6 (Fig. 12-31). A pattern of complete right bundle branch block is a frequent observation after surgical repair of ventricular septal defects and tetralogy of Fallot. Of interest, the classic electrocardiographic picture of complete right bundle branch block can be seen with interruption of either the peripheral portions of the right His-Purkinje network or central right bundle branch itself. Although the surface electrocardiographic patterns are indistinguishable, by using an intracardiac electrode catheter, normal conduction times to the right ventricular apex can be measured in the former condition, whereas apex activation is delayed with a central injury.

The ability to diagnose ventricular hypertrophy on the ECG is lost in complete bundle branch block. Attempts to correlate right ventricular pressure with the height of the R′ wave in V1, or with the extent of RSR′ distribution across the precordium, have met with limited success. Additionally, bundle branch block results in diffuse changes in the S-T segment, the T wave, and QT interval, so that the usual electrocardiographic markers of ischemia, strain, and prolonged QTc are lost.

**Left Anterior Hemiblock**

Conduction block in the left anterior fascicle produces a shift in QRS axis to the range of −60 degrees, without prolongation of QRS duration. Whereas the anterior-superior and posterior-inferior portions of the normal left ventricle are usually depolarized simultaneously by their respective fascicles, block in the anterior limb changes the sequence.
The inferior regions activate normally, but the depolarization wavefront must then spread upward, producing a superiorly directed vector in the frontal plane (Fig. 12-32). Isolated block in the anterior fascicle is rare in children but may occur with myocardial inflammation, ischemia, and surgical or catheter trauma.

Certain congenital cardiac anomalies, notably endocardial cushion defects and tricuspid atresia, have electrocardiographic patterns with a leftward superior QRS axis that mimic left anterior hemiblock. The abnormal axis is not due to true conduction defects in these cases, but instead results from the abnormal anatomic location of the conduction fibers in cushion defects (Fig. 12-33) or the unusual left ventricular shape and orientation in tricuspid atresia (Fig. 12-34).

**Left Posterior Hemiblock**

The QRS duration remains normal, but the axis is shifted right and inferior to about +120 degrees when the posterior fascicle is interrupted. The activation pattern in the left ventricle is just the reverse of anterior hemiblock (Fig. 12-35). Because right axis deviation is seen commonly in infants and children with right ventricular hypertrophy, the label of posterior hemiblock should be reserved for instances when an abrupt and dramatic axis shift has occurred between serial ECGs.

**Complete Left Bundle Branch Block**

When the main left bundle branch is interrupted, ventricular activation begins solely through the right bundle. The septum must now depolarize right to left, and the left ventricle must rely on late transmission of the activation wavefront, which is directed leftward and posterior.
The QRS is prolonged, slurred, and directed away from the right chest leads (mostly negative in V₁) and toward the lateral precordial leads (positive in V₅–V₇) (Fig. 12-36). As with complete right bundle branch block, ventricular hypertrophy, ischemic changes, and QT prolongation cannot be interpreted easily from ECGs. Very advanced degrees of concentric left ventricular hypertrophy can produce an electrocardiographic pattern identical to that of complete left bundle branch block, and the two conditions may be impossible to distinguish by ECG recordings.

**Bifascicular Block**

The combination of complete right bundle branch block and left anterior hemiblock may occur after surgical correction of congenital heart defects. For example, following repair of tetralogy of Fallot, bifascicular block of this type is present in 10% of patients. The electrocardiographic pattern is essentially a combination of the findings for the two individual conduction defects. Recall that in right bundle branch block, the initial portion of the QRS reflects the normal pattern of septal and left ventricular activation. If a new shift to a superior axis is detected for this early portion of the QRS in conjunction with the terminal slurring characteristic of complete right bundle branch block, the coexistence of left anterior hemiblock should be considered (Fig. 12-37).

Combined right bundle and left posterior fascicular block is less common in children and is difficult to diagnose from the ECG. The initial portion of the QRS, representing septal and left ventricular activation, is shifted rightward in this case. Because children undergoing cardiac surgery often have preexistent right axis deviation from right ventricular hypertrophy, it may be impossible to appreciate this particular conduction change in the postoperative period.

**Preexcitation**

Preexcitation implies that a portion of ventricular tissue is being activated ahead of schedule relative to normal His-Purkinje conduction. In Wolff-Parkinson-White (WPW) syndrome, this early activation occurs over an accessory connection between atrial and ventricular muscle located anywhere along the right or left AV grooves. As an atrial depolarization wavefront approaches the ventricles, it may advance over the accessory pathway, the normal AV node, or both. Normal AV nodal conduction is relatively slow, but the accessory pathway transmits rapid activation to a focal ventricular segment. The region of early activation generates a delta wave on the ECG (Fig. 12-38) with a short PR (actually a P-delta) interval. Because some portion of ventricular activation still occurs over the normal His pathway, there is fusion between preexcitation and the normal depolarization sequence.
The electrocardiographic patterns in the WPW syndrome are variable, depending on the location of the accessory pathway and its conduction characteristics. At the most simplistic level, left-sided accessory pathways can be expected to produce negative delta waves in the left-sided ECG leads (I, aV₁, V₅, V₆) and positive delta waves in the right-sided leads (aV₂, aV₃, V₄R–V₁) because the early activation vector is traveling left to right. For the most part, the right ventricle is activated by the normal AV node, but only after the usual nodal time delay, thus generating a gross QRS morphology reminiscent of right bundle branch block (Fig. 12-39). Right-sided accessory pathways, by comparison, are usually associated with positive delta waves in the left-sided leads (Fig. 12-40) and a QRS pattern more closely resembling left bundle branch block. Electrophysiologists who deal with WPW syndrome on a regular basis have developed several ECG algorithms¹¹,¹² that provide far more exacting estimates of accessory pathway location based on fine details of delta wave polarity and QRS shape (see Chapter 29).

A less common form of preexcitation is associated with the presence of a Mahaim fiber. Until fairly recently, these fibers were thought to represent accessory connections running from the AV node into right ventricular muscle, but they are now better understood as long and slowly conducting accessory pathways running from the anterolateral tricuspid ring to the anterior surface of the right ventricle where they may join the terminal portions of the right bundle branch.¹³ Because conduction through the Mahaim fiber is rather slow, the PR interval remains fairly normal on the ECG. However, activation through the Mahaim pathway can still preexcite a small portion of the right ventricle to generate a delta wave (Fig. 12-41). Mahaim fibers are rare and may require intracardiac electrophysiologic study to confirm their presence or distinguish them from the WPW syndrome.

In both WPW and Mahaim preexcitation, the ability to use the ECG for evaluation of hypertrophy, changes in the ST segment and T wave, and QT measurement is lost (similar to bundle branch block).

Changes in the ST Segment and T Wave

No other aspect of ECG interpretation is as dependent on good clinical history as the evaluation of abnormalities of the ST segment and T wave. Unfortunately, pathologic changes are often nonspecific. Elevation or depression of the J point and changes in the T wave can be seen in almost any condition involving myocyte injury, pericardial inflammation, abnormal ion channel function, or certain electrolyte disturbances.


Pericarditis and Pericardial Effusion

Pericardial inflammation produces a sequence of changes in the ST segment and T wave that evolve as the disorder progresses. The earliest finding is elevation of the ST segment with preservation of normal T-wave amplitude and direction. Later, the ST segment returns to the baseline, but the T wave becomes flattened and, ultimately, inverted. As opposed to the focal changes in the ST segment and T wave that are seen in ischemic syndromes, the electrocardiographic findings in pericarditis are diffuse and usually involve all leads, except perhaps aVR and the right precordium. Additionally, pericarditis influences both atrial and ventricular surfaces, such that noticeable depression of the TA wave may sometimes be observed. Occasionally, the presence of a large effusion in the pericardial space results in diminished ventricular voltages and a pattern of QRS amplitude variation known as QRS alternans (Fig. 12-42).

Myocarditis

The electrocardiographic findings in myocarditis are variable but usually involved diminished ventricular voltages and T-wave inversion during the acute illness. Atrioventricular and intraventricular conduction disturbances, along with ventricular arrhythmias, are common (Fig. 12-43).

Hypertrophic Cardiomyopathy

The changes in the ST segment and T wave seen in hypertrophic cardiomyopathy are similar to the left ventricular strain pattern that occurs with advanced hypertrophy from any cause. The lateral T waves are inverted, and the J point may be depressed. Voltage criteria for LVH are usually present (Fig. 12-44). About 30% of patients also have prominent Q waves in the lateral and inferior leads and may display left axis deviation. A somewhat high percentage of patients with hypertrophic myopathy have also been found to have preexcitation from accessory AV pathways (WPW syndrome). The PR interval may appear short in some cases of hypertrophic myopathy even when such pathways are absent; hence, a formal electrophysiology study may sometimes be needed to distinguish pseudopreexcitation from true WPW.

Dilated Cardiomyopathy

There are no specific electrocardiographic findings to aid in the diagnosis of the dilated myopathy, although the ECG is rarely normal in such cases. Because the etiology
is so variable, the ECG can include almost any of the patterns seen with inflammatory disease or hypertrophy, although ST depression (rather than elevation) is most common.

Arrhythmogenic Right Ventricular Dysplasia

This familial disease involving right ventricular myopathy and recurrent ventricular tachycardia can be extremely difficult to diagnose by any testing modality. The ECG actually seems to be one of the most reliable tools for establishing its presence. The classic findings\(^\text{14}\) include variable degrees of right ventricular conduction delay, a pattern of inverted precordial T waves extending from V\(_4\)R all the way out beyond V\(_2\), and ventricular ectopy. Most importantly, a so-called epsilon wave (Fig. 12-45) can sometimes be detected in the right precordial leads as a small high-frequency spike during the early portion of ST segment. Epsilon waves are thought to be rather specific for this disease.

Long QT Syndrome

As mentioned, a QTc that exceeds normal limits may indicate a membrane channelopathy associated with one of the hereditary long QT syndromes. Because of the serious prognosis attached to this disorder, it is imperative that the QT interval be scrutinized carefully on all ECG recordings, particularly in leads II, V\(_6\), and V\(_8\) where prolongation seems to be most dramatic.\(^\text{15}\) In addition to prolonged duration, long QT syndrome can also produce abnormal contours for repolarization signals, such as notched T waves and gross alterations in T-wave direction known as T-wave alternans (Fig. 12-46).

Ischemia

Myocardial ischemia is a rare problem in pediatric practice, but it may occur with certain congenital anomalies or inflammation of the coronary arterial vasculature. Hypoxic insults result in an evolution of electrocardiographic findings, which tend to parallel cellular events. During the initial ischemic phase, the most dramatic changes occur in the T wave, which becomes tall and peaked in those leads that record near the affected myocardial segment. These changes are usually accompanied by some deviation of the

Brugada Syndrome

Brugada syndrome is a more recently recognized hereditary channelopathy causing ventricular tachycardia,\(^\text{16}\) which is usually associated with right bundle branch block and dramatic ST elevation in leads V\(_1\) through V\(_3\) (Fig. 12-47). The QTc tends to be normal. These findings can wax and wane in a given patient to the point that the ECG appears rather normal at times. Provocative challenges with certain antiarrhythmic drugs may be needed to uncover the abnormalities.

Short QT Syndrome

A new familial condition known as short QT syndrome has now been described.\(^\text{17}\) This rare disorder is associated with ventricular arrhythmias similar to long QT syndrome, but the patients demonstrate strikingly short QTc values of less than 0.30 second, along with very tall peaked T waves.

FIGURE 12–45 ECG from a teenage patient with familial arrhythmogenic RV dysplasia who had documented ventricular tachycardia as well as a dilated RV on echocardiogram. Note the deeply negative precordial T waves and the subtle epsilon wave in the right chest leads (arrows).

FIGURE 12–46 Dramatic T-wave alternans in a patient with a severe form of long QT syndrome.

FIGURE 12–47 ECG from a young boy with recurrent VT due to Brugada syndrome. Note unusual appearance of the ST segment and T waves in the right precordium.
ST segment (Fig. 12-48). If the pathologic process is promptly reversed, these changes can resolve. However, if the insult persists, the injury phase commences and may be seen on the ECG as a more dramatic shift in the ST segment. The ST deviation may be upward or downward, depending on whether the injured cells are epicardial or endocardial (Fig. 12-49). During the injury phase, correction of the underlying cause may still result in reversion to a normal ECG. When the injury persists, cell death (infarction phase) follows, reflected on the ECG as a diminution of R-wave voltage and the appearance of Q waves in those leads facing the infarcted segment (Fig. 12-50).

The most common cause of myocardial ischemia in pediatric practice involves anomalous origin of the left coronary from the pulmonary artery. At initial presentation, variable degrees of injury or infarction are apparent, typically involving ventricular muscle in the distribution of the left anterior descending artery (i.e., anterior and septal areas). The electrocardiographic findings in severely afflicted infants include deep Q waves in leftward and lateral leads (I, aVL, V3–V6) and loss of the mid-precordial R wave (V5), with a normal QRS axis (Fig. 12-51). Children who present beyond infancy are more likely to have left axis deviation, smaller Q waves in the leftward and lateral leads, and increased voltages suggestive of LVH (Fig. 12-52).

**Electrolyte Abnormalities**

Significant changes may occur on the surface ECG with certain electrolyte disturbances, most notably potassium, calcium, and magnesium imbalance. With hyperkalemia, the electrocardiographic findings are quite specific for anything more than mild abnormalities. Moderate elevation of serum potassium concentration (greater than 6.0 mEq/L) causes tall, peaked T waves, along with some widening of the QRS complex (Fig. 12-53). Marked elevation (more than 8.0 mEq/L) causes profound widening of the P wave and QRS complex, resulting in a pattern for sinus rhythm...
that resembles a sine wave or mimics wide ventricular tachycardia (Fig. 12-54). Fibrillation and asystole can result from acute severe elevations of serum potassium. Hypokalemia (less than 3.0 mEq/L) results in a low-amplitude, flattened T wave, with the appearance of prominent U waves.

Calcium and magnesium predominantly influence speed of cellular repolarization. Low levels of either ion prolong the QT interval, whereas high serum levels may shorten the QT slightly.

Pathognomonic Electrocardiographic Patterns

Table 12-2 lists some cardiac conditions and syndromes in which the ECG findings are sufficiently specific to allow rapid diagnosis. These disorders are discussed individually elsewhere in this text but are abstracted here for quick reference.

**HOLTER MONITORING AND EVENT RECORDING**

Unless one is fortunate enough to have an ECG hooked up during a clinical arrhythmia episode, alternate techniques for long-term rhythm recording must be employed. The most familiar tool for this purpose is the Holter monitor, which provides 24-hour continuous rhythm recording from adhesive electrodes on the chest. Modern Holter equipment involves a lightweight battery-powered device that stores data to a digital file. Two or three simultaneous ECG channels are usually recorded and synchronized to a 24-hour clock. The patient and family maintain a written diary to correlate activity or symptoms with rhythm status. The recording is later played back on a high-speed analysis system by a technician using computerized arrhythmia detection templates. Paper copies of interesting events can then be printed for physician review (Fig. 12-55). Most analysis systems also provide data on heart rate trends (minimum, maximum, mean, and degree of variability) along with quantitation of supraventricular and ventricular ectopy.

The Holter monitor is suitable only if an arrhythmia occurs at a frequency greater than once in 24 hours. For patients who go many days or weeks between events, a long-term event recorder, which the patient keeps for a period of 30 to 60 days, is a far more appropriate tool.
Some event recording devices are intended to be worn on an episodic basis, whereas others are designed to be worn continuously to capture fleeting arrhythmic events (Fig. 12-56). When either device is activated, a rhythm strip is recorded for 1 minute or so and stored in memory. The recording is later played back as an oscillating audio signal that is easily decoded back into an electrocardiographic waveform with appropriate equipment. One major advantage of this technology is that the audio signals can be sent long distance over a standard telephone to a central decoding station, thus saving the patient travel time while also hastening detection of potentially serious rhythm disorders.

Event recorders are useful for the diagnosis of episodic palpitations of undetermined etiology, or for evaluation of vague symptoms (e.g., dizziness or chest pain) when an arrhythmia is part of the differential diagnosis. Implantable event recorders that are inserted subcutaneously in the pectoral area have also been developed in recent years and may be indicated in select cases.

EXERCISE TESTING FOR RHYTHM EVALUATION

Manipulation of autonomic tone can provide information regarding rhythm status that is not always available on a resting ECG. Enhancement of sympathetic drive during dynamic exercise permits analysis of sinus node function, intracardiac conduction, and certain tachyarrhythmias. The two primary exercise techniques used in pediatrics are...
the Bruce protocol on a treadmill, and the stationary bicycle protocol (see Chapter 16). The treadmill has an increased static component to its workload and generally produces higher peak heart rates, although bicycle exercise can provide higher fidelity electrocardiographic tracings because there is less body motion to generate baseline artifact. Both techniques are relatively straightforward to use for rhythm evaluation in patients as young as 6 years old. Although exercise testing in children is safe, the personnel need to be prepared for arrhythmias and other unexpected indications for test termination. Of 3120 consecutive studies analyzed from our center, 5% \((n = 156)\) were terminated prematurely for findings other than fatigue, including worrisome ventricular arrhythmias \((n = 59)\), syncope or marked presyncope \((n = 44)\), supraventricular tachycardia \((SVT; n = 27)\), or exercise heart rate approaching detection rates for an automatic implantable defibrillator shock \((n = 15)\).

Chronotropic incompetence from sinus node dysfunction can be assessed reasonably well with exercise testing by comparing the maximum achieved heart rate with norms for age. For patients with various degrees of AV block, exercise may be used to assess changes in conduction pattern, evaluate escape rates, or look for the development of ventricular ectopy at peak exercise.\(^{19}\) Such data may assist in determining timing for permanent pacemaker insertion. For patients with a pacemaker already in place, exercise studies can be used to adjust the degree of rate responsiveness and evaluate upper rate limit settings of the device.

Ventricular arrhythmias are a fairly common indication for exercise testing. It is generally believed that suppression of ventricular ectopy at peak exercise portends a benign prognosis, although there are certainly exceptions to this rule. More to the point, ventricular ectopy that either fails to extinguish at elevated heart rates or is exacerbated with exercise\(^{20,21}\) suggests clinically important disease (Fig. 12-57). Exercise testing may also be used to provide a spectrum of QT intervals for review at varied heart rates\(^{22}\) or to detect exercise-induced arrhythmias in suspected cases of long QT syndrome.

Supraventricular arrhythmias do not generally lend themselves to evaluation by exercise testing, with the notable exception of WPW syndrome, in which the anterograde conduction characteristics of the accessory pathway can be assessed. When patients with clear delta waves and

**FIGURE 12–56** An event recorder with a memory loop that records the preceding 30 sec of rhythm and a subsequent 30 sec when the device is activated. From Fyler DC. Nadas’ Pediatric Cardiology. Philadelphia: Hanley & Belfus, 1992.

**FIGURE 12–57** A. Abrupt onset of sustained monomorphic ventricular tachycardia with treadmill exercise in a 15-year-old with palpitations. B. Nearly sustained, somewhat polymorphic, ventricular tachycardia at peak exercise in an adolescent female with recurrent syncope.
short PR intervals develop sinus tachycardia in response to exercise, an atrial rate may be reached whereby the capacity of the accessory pathway to conduct anterograde is exceeded, so that all ventricular activation occurs over the AV node. At this point, the QRS complex and the PR interval normalize (Fig. 12-58). The sinus rate at which anterograde preexcitation is blocked correlates fairly well with conduction measurements for the accessory pathway obtained at electrophysiologic study and may help predict the potential risk for rapid anterograde conduction should the patient ever develop atrial fibrillation. The unequivocal loss of preexcitation in response to sinus tachycardia can usually be interpreted to indicate a patient is at relatively low risk for rapid preexcited atrial fibrillation.23

AUTONOMIC TESTING

Dynamic exercise testing permits observation of rhythm status during enhanced sympathetic state, but controlled manipulation of vagal tone is more difficult to achieve. Perhaps the best technique currently available for this purpose is the head-up tilt (HUT) procedure, during which the complex orthostatic reflexes involved with vasomotor regulation and heart rate control can be assessed.24–26

The basic technique for HUT involves positioning the patient supine on a motorized tilt table with a peripheral intravenous line in place, and monitoring the ECG along with noninvasive arterial blood pressure. After a 15-minute period of baseline observation in the supine position, the patient is tilted to a nearly upright posture (60 to 70 degrees), and the patient’s physiology is reassessed for an additional 15 minutes. If no symptoms or arrhythmias occur, the patient is returned to the supine position, and the entire sequence is repeated during an isoproterenol infusion (Fig. 12-59).

A positive HUT test may suggest a benign, neurally mediated mechanism as the etiology for recurrent syncope, or for episodic arrhythmias such as sinus bradycardia,

FIGURE 12–58 A. Treadmill exercise testing on a 16-year-old with palpitations and Wolf-Parkinson-White syndrome. At a heart rate of 189, there is abrupt loss of the delta wave. B. Another patient with Wolf-Parkinson-White syndrome who has progressive QRS fusion but maintains a subtle delta wave up to maximum sinus rates of 189.

FIGURE 12–59 Composite data from a head-up tilt test in a patient with intermittent syncope. Condition (A) is supine baseline before tilt. With initial head-up tilt (B), there is a minor increase in heart rate and narrowing of pulse pressure. The bottom panel shows preserved cerebral blood flow velocity. The condition shows preserved cerebral blood flow velocity. With longer-duration 60-degree tilt, there are increasing, cyclic swings in heart rate and blood pressure, eventually associated (C) with presyncope symptoms, decreased cerebral blood flow velocity, and then a marked decrease in both heart rate and blood pressure associated with syncope. In recovery, the heart rate remains relatively slow for several minutes before abruptly returning to a more normal pattern.
transient AV block, or exaggerated orthostatic sinus tachycardia. However, because of the relatively high incidence (up to 40%) of false-positive responses to HUT testing in asymptomatic children, the results need to be interpreted with caution. Inducing vasovagal syncope or a reflex bradycardia simply cannot be viewed as excluding other disease. Studies are only useful if the physiologic data correlate cleanly with symptoms, and the possibility of more serious cardiac disorders has been dismissed by other techniques.

**SIGNAL AVERAGING**

Signal averaging produces a high-resolution, high-amplitude electrical recording from the body surface that allows one to view subtle electrical events that are too small to be seen on a traditional ECG. This is accomplished by recording a hundred or more consecutive waveforms that are superimposed in the computer memory of the recording device, producing a composite or average signal from the multiple beats. Random electrical noise that is not part of the composite waveform is easily subtracted, and the resultant signal is amplified to permit a glimpse at low-voltage events that usually go undetected on the routine ECG.

Signal-averaging technology is used primarily as a risk-assessment tool for ventricular arrhythmias. The standard signal-averaged electrocardiogram (SAECG) involves a highly amplified QRS complex focused on the depolarization process. During normal ventricular depolarization, healthy myocytes activate quickly and inscribe the sharp QRS complex, but areas of diseased myocardium may have sluggish cell-to-cell conduction that continues to generate small-amplitude signals lasting beyond the normal QRS duration. The amplitude of these late potentials is typically too small to be appreciated through ambient electrical noise, but they can be uncovered by the SAECG during the terminal portion of the QRS (Fig. 12-60). Three orthogonal leads (X, Y, and Z) are typically used to record the SAECG, which are then summed and a vector magnitude obtained that transforms all net ventricular voltages into positive values. Commercial systems usually allow these data to be obtained in about 5 to 10 minutes. The transformed signal is then analyzed for three principle variables; total QRS duration, the root mean squared and mean voltages in the terminal 40 msec. Furthermore, both tests may have reduced value in the presence of specific amplitude alternans at heart rates between 100 and 130 beats/minute (Fig. 12-61).

Pediatric data remain limited regarding the significance of late potentials or microvolt T-wave alternans. Although some patients with severe forms of long QT syndrome may occasionally demonstrate gross T-wave alternans on a conventional ECG recording, far more subtle T-wave alterations can occur in other forms of heart disease. These small changes involve measurements in the range of microvolts, which would be far beneath the usual millivolt resolution available on the standard ECG. Microvolt TWA is detected by using specialized skin electrodes, collecting 128 consecutive beats that are averaged to allow spectral analysis of oscillations in T-wave amplitude. The patient’s heart rate is also raised with exercise, atrial pacing, or medication, and the measurements are repeated. Results are considered abnormal in the presence of TWA greater than 1.9 μV at rest, or the appearance of specific amplitude alternans at heart rates between 100 and 130 beats/minute (Fig. 12-61).

Pediatric data remain limited regarding the significance of late potentials or microvolt T-wave alternans. Furthermore, both tests may have reduced value in the presence of bundle branch block. At present, these tests are used primarily as confirmatory or supportive data for young patients with a normal QRS duration who already have other concrete risk factors for ventricular arrhythmias.
TRANSESOPHAGEAL RECORDING AND PACING

The proximity of the esophagus to the left atrium allows a recording and pacing electrode to be placed immediately adjacent to atrial muscle without requiring vascular entry. A properly positioned esophageal lead will record a sharp, high-amplitude atrial electrogram that is far superior to a surface ECG signal for identifying atrial timing during complex arrhythmias. Flexible bipolar pacing wires specifically designed for esophageal use are easily inserted with the same technique used for nasogastric tube placement.

Proper lead positioning is important to obtain a high-quality electrogram and achieve an acceptable threshold for pacing. The position may be checked by moving the lead up and down the middle portion of the esophagus and looking for the site that records the largest atrial electrogram (Fig. 12-62), or by consulting published charts that correlate ideal insertion depth with body size. Any device capable of registering electrophysiologic signals can be used to display the electrogram from an esophageal lead (Fig. 12-63), including conventional ECG equipment. An esophageal electrogram is exceedingly helpful for recording atrial activity during arrhythmias when the mechanism is
unclear on a surface ECG. A classic example is atrial flutter with 2:1 conduction to the ventricles, in which the esophageal electrogram can uncover a hidden flutter wave (Fig. 12-64) that might otherwise have been obscured by the QRS on a standard ECG.

Transesophageal pacing requires a higher pacemaker output than is used for an electrode in direct contact with atrial tissue. Whereas intracardiac pacing is routinely done at a pulse width of about 2.0 msec, the capacity for wider pulses of up to 10.0 msec is needed for transesophageal pacing. The expanded pulse width permits atrial capture at amplitude settings of about 10 to 15 mA. Except for very rare instances, ventricular pacing is not possible with an esophageal electrode. Atrial pacing through the esophageal lead can be used for temporary correction of sinus bradycardia, but it is more frequently employed for short bursts of rapid overdrive pacing of the atrium to interrupt reentry SVT. The overdrive technique is extremely successful in converting reentry circuits within the AV node or those involving an accessory pathway (Fig. 12-65), and it may be successful in as many as 80% of cases involving atrial flutter.

**INTRACARDIAC ELECTROPHYSIOLOGIC STUDIES**

Intracardiac electrical recording has evolved rapidly during the past 35 years from simple analysis of His bundle signals to complex three-dimensional maps of endocardial propagation. The systematic use of these intracardiac recordings, coupled with programmed stimulation to induce arrhythmias under controlled conditions, has expanded our understanding of the mechanism and risk for just about
every type of arrhythmia in man. Electrophysiologic study (EPS) has also laid the groundwork for the use of transcatheter ablative techniques that have revolutionized tachycardia management in patients of all ages. Although diagnostic EPS and catheter ablation are usually combined in a single procedure in the current era (Fig. 12-66), this section focuses exclusively on diagnostic techniques, with discussion of ablation reserved for Chapter 29.

**Equipment**

The basic equipment needed to perform EPS includes an imaging system for catheter localization, an electrophysiologic recording system, and a stimulator for cardiac pacing.35 Intracardiac electrograms are filtered and displayed on a multichannel recorder along with simultaneous surface ECG leads (most often I, aV\textsubscript{F}, V\textsubscript{1}, and V\textsubscript{6}). The recording system is usually computerized with digital processing that allows for real-time analysis at various sweep speeds (anywhere from 25 to 400 mm/sec), along with freeze and playback, annotation, and digital storage of the recorded signals. Studies are performed in a cardiac catheterization laboratory using fluoroscopic imaging to guide catheter navigation. Nonfluoroscopic catheter navigation techniques are now being introduced to many laboratories that can decrease radiation exposure and create virtual electrical activation maps of the heart. The other essential requirements for EPS are standard resuscitation equipment, reliable defibrillation equipment, and most importantly, an experienced staff. The type of sedation utilized for EPS is based on the type of study, patient’s age, and arrhythmia mechanism. General anesthesia is frequently used for children who are undergoing EPS combined with ablation, but conscious sedation is usually sufficient for purely diagnostic procedures in children.

Studies are performed with catheters ranging in size from 2 to 9 French. These are equipped with variable arrays of electrodes at the distal tip (Fig. 12-67) through which one can pace and record from various cardiac sites. Standard percutaneous techniques are used for vascular access. The number and types of catheters employed depend on the specific goals of the study, ranging from a single atrial catheter to induce supraventricular arrhythmias, to four or five catheters for a complete electrophysiologic survey of the heart. The standard pacing and recording sites are shown in Figure 12-68 and include the high right atrium, the bundle of His, the coronary sinus (CS), and the right ventricular apex.


**FIGURE 12–66** The yearly numbers of invasive diagnostic and therapeutic intracardiac electrophysiology studies done at Children’s Hospital Boston from 1985 to 2004. The percentage of purely diagnostic studies dropped from nearly 100% in the late 1980s to only 12% in 2004.

**FIGURE 12–67** Example of electrode catheters for intracardiac electrophysiologic study. Shown here (from top to bottom) are a 4 Fr quadripolar catheter, 5 Fr HBE catheter, 7 Fr deflectable quadripolar catheter with a 4-mm tip (used for ablation), and a 7 Fr deflectable octapolar catheter (used for coronary sinus recording).
**High Right Atrium**

The high right atrium (HRA) site is located at the superior vena cava–right atrial junction, in close proximity to the SA node. Sometimes this catheter is placed in the right atrial appendage instead, to improve stability during long procedures and ablations. Right atrial pacing is used for assessing SA and AV node function, inducing supraventricular arrhythmias, and on rare occasions, inducing idiopathic ventricular tachycardias.

**His Bundle Electrogram**

The His bundle electrogram (HBE) is obtained by positioning the catheter just across the tricuspid valve at its medial and superior rim, near the area of the common bundle of His. Because the catheter straddles the valve ring, it records local right atrial depolarization and local right ventricular depolarization, along with a distinct His deflection. The HBE dissects AV conduction into AV node and His-Purkinje components. It is a crucial recording site for analysis of AV conduction disturbances and for determining the supraventricular origin of an arrhythmia.

**Coronary Sinus**

A catheter in the CS records both atrial and ventricular signals from the left heart. This electrode is usually positioned from a femoral vein approach using a tip-deflecting mechanism built into the catheter shaft. If CS entry proves difficult, a CS angiogram may be performed to guide electrophysiologic catheter placement. Occasionally, a large Eustachian ridge guards the CS ostium, and a superior approach is needed. This can be accomplished through the right internal jugular vein, subclavian vein, or brachial vein. The CS catheter is mandatory for evaluation of SVT, particularly if an accessory pathway is suspected.

**Right Ventricular Apex**

The right ventricular apex (RVA) provides a stable site for ventricular pacing and recording. It is used during evaluation of SVT to examine retrograde conduction, as well as for the study of ventricular arrhythmias. An additional ventricular pacing site in the right ventricular outflow tract (RVOT) may be included in some cases.

**DIAGNOSTIC MANEUVERS**

Diagnostic EPS involves three general categories of data: (1) baseline rhythm, (2) pacing maneuvers to evaluate functional characteristics, and (3) programmed stimulation with a series of premature beats to initiate tachycardias. A comprehensive EPS will involve systematic collection of all these items, as outlined below.

**Baseline Recording**

Resting rhythm is examined to determine conduction times and activation sequence throughout the heart (Fig. 12-69). In normal sinus rhythm, the HRA catheter is
the first to register electrical activity by virtue of its close proximity to the SA node, followed by the low right atrium (first signal on HBE catheter) 20 to 25 msec later, and finally the lateral left atrium (first signal on distal CS catheter) after about 60 msec. Ectopic foci or other supraventricular arrhythmias will shift the atrial activation sequence in favor of the site of origin of the abnormal rhythm.

Conduction time within the AV node may be determined by measuring the AH interval of the HBE recording. The normal AH interval varies between 40 and 100 msec in children. A prolonged AH interval can be observed in patients taking certain antiarrhythmic drugs, those with high vagal tone, and those with disease of the AV node. It is shortened under conditions of high circulating catecholamines.

The HV interval, representing conduction time in the His-Purkinje system, is also recorded by the HBE catheter and should be between 35 and 55 msec in normal children. Prolonged resting HV intervals usually suggest His-Purkinje pathology. A short HV may be observed in the preexcitation syndromes, including WPW syndrome and Mahaim fibers.

The final baseline measurements come from regional ventricular activation times. The standard catheter positions provide ventricular activation from three locations, including the apex of the right ventricle (RVA catheter), right ventricular inflow (last signal on HBE catheter), and left ventricular base (last signal on CS catheter). Measurements are indexed against a V line, which is marked at the earliest evidence of ventricular activation in any recording lead (including the surface QRS). Normally, the RVA displays the earliest signals among these recording sites, about 10 to 35 msec after onset of the QRS. Block in the central right bundle branch can cause delayed RVA activation (more than 50 msec). In WPW syndrome, preexcitation through a right-sided accessory pathway may promote early activation of the right ventricular inflow area, or conversely, a left-sided pathway may cause the left ventricular base along the CS to activate ahead of schedule.

### Functional Characteristics of the SA Node

The principal maneuver used to evaluate SA node status during EPS is the sinus node recovery time (SNRT). This involves pacing the HRA for 30 to 60 seconds at rates above resting rhythm. When pacing is abruptly terminated, there is usually a brief pause before the resumption of sinus beats in a normal individual, but if the pause is protracted, SA node dysfunction is suggested. The recovery time is usually “corrected” by subtracting the resting cycle length (i.e., the time between two normal sinus beats) from the recovery cycle length (i.e., the time from the last paced beat to the first spontaneous sinus beat) to yield a corrected sinus node recovery time (CSNRT). Normal values for CSNRT should be less than about 275 msec for children. When there is advanced disease of the SA node, the abnormal SNRT is usually quite blatant (Fig. 12-70).

### Functional Characteristics of the AV Node

The most straightforward technique for evaluation of AV conduction involves short bursts of pacing in the HRA

![Image](image-url)

**Figure 12-69** Baseline signals during sinus rhythm, including three surface ECG leads, along with multiple intracardiac recordings. Atrial activation is indexed against a P line and shows earliest intracardiac activation in HRA near the sinus node. Ventricular activation is indexed against the V line, and normally shows the earliest activity in RVA. HRA, high right atrium; CSd, distal coronary sinus; CSP, proximal coronary sinus; HBE, His bundle electrogram; RVA, right ventricular apex; Ao, arterial blood pressure.


![Image](image-url)

**Figure 12-70** An example of prolonged sinus node recovery. After 60 sec of pacing (S₁) in HRA at a rate of 150 beats/min (cycle length, 150 msec), pacing abruptly terminated. There is a pause of nearly 3.0 sec before a junctional escape beat restores the rhythm. HRA, high right atrium.

while observing the ventricular response. The pacing cycle length is decreased in 10-msec steps until a rate is encountered at which the Wenckebach phenomenon occurs. For normal children, Wenckebach is not usually observed until the pacing cycle length has been decreased to less than about 350 msec (i.e., rate faster than 171 beats/min).

More precise evaluation of the AV node is performed with the introduction of single premature beats at the HRA, delivered at an interval that is progressively shortened in 10-msec steps (Fig. 12-71). As the stimulus is moved earlier, a gradual and progressive increase in the A-H interval is seen related to the normal decremental properties of the slow response cells in the AV node. Eventually, a stimulus can be delivered early enough to block at the AV node. This registers on the HBE electrogram as an atrial signal without a His or ventricular deflection. The premature interval at which this conduction fails to occur is defined as the ERP for the AV node and normally ranges from 240 to 320 msec in children. As the stimulus timing is adjusted to a shorter coupling interval, a point will eventually be reached at which the atrial tissue no longer depolarizes. This is designated the atrial muscle ERP and is encountered between 170 and 240 msec in normal children.

**Functional Characteristics of His-Purkinje Conduction**

Function of the His-Purkinje system may likewise be evaluated with the single extrastimulus technique. Normally, the HV interval tends to remain constant, and the QRS complex tends to remain narrow during premature atrial stimulation. Some patients may develop QRS aberration due to rate-related bundle branch block in response to early beats, but this is usually a benign finding. More significant is the observation of block in the bundle of His in response to an early stimulus, which registers on the HBE as an atrial signal followed by a His deflection, but without conduction to the ventricles. When the ERP of the common bundle of His is encountered ahead of the AV node ERP in a young patient, His-Purkinje disease may be suggested.

**Functional Characteristics of Ventricular Tissue**

The ERP can also be measured for ventricular muscle, using the single extrastimulus technique. This information is most useful during the study of ventricular arrhythmias, when one might wish to compare baseline conditions to the alteration in ERP related to antiarrhythmic drugs. The longest premature interval at which a stimulus fails to capture the ventricular arrhythmia is defined as the ventricular muscle ERP, and normally occurs at 180 to 260 msec. Ventricular stimulation is also used to evaluate the pattern of retrograde conduction to the atrium.

**Evaluation of Supraventricular Tachycardia**

Many patients with SVT can be managed without a formal EPS. However, if the arrhythmia has been refractory to first-line management or is associated with serious

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**FIGURE 12–71** The effective refractory periods of the AV node (A) and atrial muscle (B). In panel A, the premature beat captures the atrium but blocks at the AV node such that no His bundle signal is seen. In B, a slightly earlier beat fails to capture the atrium entirely. HRA, high right atrium; CS, coronary sinus; HBE, His bundle electrogram; RVA, right ventricular apex.

symptoms, an intracardiac EPS is usually indicated to determine the mechanism and to map the substrate with a view toward catheter ablation.

Studies for SVT require catheters at the four standard recording sites. Resting rhythm is first examined for the atrial activation pattern and any evidence of preexcitation. A routine evaluation of the functional status of the entire conducting system is then performed as described previously. During these maneuvers, or with more aggressive stimulation, SVT may be induced.

There is a long list of mechanisms for pediatric SVT (see Chapter 29), but as a starting point, one may divide them into the broad categories of automatic foci or reentry circuits. The automatic variety tend to occur spontaneously or in response to increased catecholamine levels but generally are not induced or terminated with pacing techniques. Reentry, by far the more common mechanism in children, is easily induced by carefully timed premature beats during EPS and can usually be interrupted by overdrive pacing.

Mapping of SVT begins by determining whether the AV node and ventricle are critical components of the disorder. Those forms of SVT that arise from within atrial muscle (primary atrial SVT) usually display intermittent block at the AV node that does not modify the atrial rate. By comparison, tachycardias with a fixed 1:1 AV relationship (AV reciprocating SVT) terminate promptly whenever this ratio is disturbed. The latter is typical behavior for SVT owing to accessory pathways, as well as for most cases of AV node reentry.

Precision mapping of an arrhythmia location is possible with attention to the atrial activation sequence during SVT. Localization of both automatic foci (Fig. 12-72) and accessory pathways (Fig. 12-73) is accomplished by carefully noting the site of the earliest atrial signal.

**FIGURE 12–72** An example of atrial activation sequence mapping in a patient with ectopic atrial tachycardia. The earliest atrial activity was found mapping near the left lower pulmonary vein (LLPV). HBE, His bundle electrogram; HRA, high right atrium. From Fyler DC. Nadas’ Pediatric Cardiology. Philadelphia: Hanley & Belfus, 1992.

**FIGURE 12–73** Mapping during orthodromic reentrant tachycardia in Wolff-Parkinson-White syndrome. Tachycardia is induced with a single premature beat (S1) that conducts to the ventricle over the AV node and returns to the atrium through the accessory pathway. The shortest VA time (i.e., the earliest retrograde signal) is seen in the proximal coronary sinus, corresponding to a left-sided accessory pathway. HRA, high right atrium; CSd, distal coronary sinus; CSp, proximal coronary sinus; HBE, His bundle electrogram; RVA, right ventricular apex; Ao, arterial blood pressure. From Fyler DC. Nadas’ Pediatric Cardiology. Philadelphia: Hanley & Belfus, 1992.

### Evaluation of Ventricular Arrhythmias

Ventricular stimulation studies are performed with three major goals in mind. First, some tachyarrhythmias that appear to be ventricular tachycardia (VT) because of a wide QRS on surface ECG are actually SVT with aberration or preexcitation. Because the prognosis and treatment differ so drastically for SVT and VT, EPS is usually indicated to establish the exact mechanism whenever uncertainty exists. Second, EPS can be used to help elucidate the mechanism and location of a patient’s VT, which can assist the clinician in choosing among drug therapy, ablation, or an implantable defibrillator for long-term management. Finally, programmed stimulation can sometimes be used as a tool for stratifying a patient’s risk for spontaneous VT by observing the patient’s response to a standard ventricular pacing protocol.37

A complete VT study begins with baseline recording from at least three sites, including the HRA, HBE, and RVA. Atrial stimulation is then performed to evaluate functional characteristics of the conducting system and to rule out atypical SVT as the cause of the clinical arrhythmia. A ventricular stimulation protocol is then carried out in an attempt to induce VT. Protocols for ventricular stimulation vary, and it must be remembered that a point can be...
reached at which the stimulation becomes so provocative that an induced arrhythmia is nonspecific and bears little resemblance to the clinical tachycardia. Additionally, some forms of VT may involve a mechanism other than classic reentry, and normal stimulation techniques may be unsuccessful in reproducing the disorder. At most centers, ventricular stimulation studies usually involve the following protocol:

1. Stimulation at the RVA with an 8-beat drive train (S1 × 8) followed by one or more extrastimuli (Sn) delivered at progressively premature intervals (shortened in 10-msec decrements) until local ventricular muscle is refractory
   a. Single premature beats following 8 beats of ventricular pacing (S1 × 8, S2) using at least two different cycle lengths for S1 (usually 600, 500, or 400 msec)
   b. Double premature beats following 8 beats of pacing (S1 × 8, S2, S3) using at least two different cycle lengths for S1
   c. Triple premature beats following 8 beats of ventricular pacing (S1 × 8, S2, S3, S4) using at least two different cycle lengths for S1
2. Stimulation at a second site, usually the RVOT, with an identical sequence
3. Repetition of RVA and RVOT stimulation during isoproterenol infusion
4. Optional additions to the protocol:
   a. The standard sequence at a third site such as the left ventricular apex
   b. Stimulation with four premature beats (S1 × 8, S2, S3, S4, S5)
   c. Burst ventricular pacing at progressively rapid rates

The possible responses to ventricular stimulation are shown in Figure 12-74. Whenever sustained arrhythmias are induced and result in hemodynamic compromise, the team must react quickly to terminate the condition. For monomorphic VT, this can often be accomplished with a short burst of overdrive pacing in the ventricle (Fig. 12-75), but rapid polymorphic VT or ventricular fibrillation needs to be terminated promptly with a DC shock.

**Ancillary Tests**

During investigation of select ventricular arrhythmias, the EPS procedure may be coupled with hemodynamic assessment, angiography, endomyocardial biopsy, voltage mapping, and drug challenges. Hemodynamic data may reveal abnormalities not apparent on echocardiography, such as pulmonary hypertension or restrictive cardiomyopathy. Angiography may be indicated to delineate coronary anomalies, ventricular morphologic changes, and chamber aneurysms. When new-onset cardiomyopathy or myocarditis is suspected, an endomyocardial biopsy is useful to confirm the diagnosis. Voltage mapping is a relatively new procedure during which automated measurements of local electrogram voltage from one or both ventricular chambers is displayed on a virtual three-dimensional cardiac chamber map. Areas of very low voltage may correspond to scar or diseased myocardium. This technique may help in the diagnosis of certain cardiomyopathies, including arrhythmogenic right ventricular dysplasia. Finally, provocative drug challenges with adenosine, catecholamines,
or procainamide can uncover certain underlying electrical abnormalities. Adenosine is useful to expose cryptic premature excitation during sinus rhythm or to reveal the presence of a unidirectional retrograde accessory pathway during ventricular pacing. An isoproterenol infusion is frequently used to induce various types of catecholamine-sensitive VT. Procainamide infusion can be used to uncover evidence of certain membrane channelopathies, such as Brugada syndrome.

REFERENCES


