Introduction

The practice of pediatric cardiac anesthesia has evolved significantly over the years, expanding beyond the operating room environment to many non-surgical locations. It is likely that anesthetic care will be provided for patients with cardiac rhythm disturbances in a variety of settings (operating rooms, intensive care units, treatment rooms, emergency facilities, cardiac catheterization/electrophysiology laboratories). Basic knowledge of dysrhythmia diagnosis and management is essential when caring for patients in any of these settings. This chapter provides a practical approach to pediatric cardiac dysrhythmias with discussions centered on diagnosis, mechanisms, and acute management strategies. A review of antiarrhythmic drug therapy in children is presented as well as the basic principles of cardiac pacing as applicable to the practice of this specialty.

Cardiac rhythm disturbances

Sinus bradycardia

Slow heart rates can be observed during sleep or at times of high vagal tone. When there is significant sinus bradycardia, a slow junctional escape rhythm or a slow atrial rhythm with an ectopic P wave focus may be present. Certain forms of congenital heart disease (CHD) may be more prone to slow heart rhythms. Patients with visceral heterotaxy (malpositioning of the abdominal organs) and polysplenia syndromes may be included in this category due to absence, displacement or hypoplasia of a true sinus node. The sinus node is a right-sided structure and this condition is characterized by bilateral left-sidedness.

In the intraoperative setting, particularly upon induction of anesthesia, with laryngoscopy, endotracheal intubation or tracheal suctioning, sinus bradycardia may occur. Sinus bradycardia may also be related to drug administration (i.e. opioids) or increased parasympathetic activity. This type of sinus bradycardia rarely poses a significant hemodynamic problem and, if so, can be easily treated with removal of the stimulus or chronotropic agents such as atropine or epinephrine (Table 15.1). In the postoperative setting, slow sinus rates may be associated with surgical interventions such as atrial septal defect repair (sinus venosus type) and cardiac transplantation. Sinus bradycardia can also be secondary to hypoxemia, hypothermia, drugs, acidosis, electrolyte abnormalities, or increased intracranial pressure. Bradycardia related to hypoxemia should be treated promptly with supplemental oxygen and appropriate airway management. The approach to other forms of secondary sinus bradycardia should focus on addressing the underlying cause. For worrisome low rates, particularly in small infants, or clinical evidence of low cardiac output, pharmacologic therapy (isoproterenol infusion) or temporary pacing should be considered.

Sinus node dysfunction

Sinus node dysfunction, sometimes termed sick sinus syndrome, encompasses a spectrum of disorders characterized by slow or irregular heart rates with a variety of escape rhythms alternating with periods of tachycardia. The tachycardia may be atrial tachycardia, atrial flutter, or atrial fibrillation. The term tachycardia–bradycardia syndrome is frequently used to characterize this association. Surgical interventions most likely to be associated with sinus node dysfunction include extensive atrial baffling procedures, such as Mustard or Senning operations, and the Fontan procedure. Management of symptomatic patients may include pacemaker implantation, drug therapy for tachydysrhythmias, atrial antitachycardia pacing, and in some cases radiofrequency ablation (transcatheter or surgical).

Sinus tachycardia

Sinus tachycardia is more commonly seen in the perioperative
the lack of normal atrial systolic contribution to ventricular filling.

Conduction disorders

Bundle branch block

In the unoperated patient, bundle branch block is an uncommon ECG finding, although incomplete right bundle branch may be seen in patients with right ventricular volume overload (atrial septal defects, anomalous pulmonary venous return, etc.). In the postoperative patient, a right bundle branch pattern is a frequent finding after surgical procedures for various congenital heart defects including tetralogy of Fallot, right ventricular outflow tract reconstructions, ventricular septal defect, and AV septal defect (AV canal or endocardial cushion defect). The bundle branch block pattern may be related to the ventriculotomy incision, damage to the moderator band, the ventricular septal defect repair, or resection of infundibular muscle. Left bundle branch block patterns are uncommon but can be seen in some patients following surgery involving the subaortic region.

Atrioventricular block

First-degree atrioventricular block

First-degree AV block is characterized by prolongation of the PR interval beyond the normal for age. Each P wave is followed by a conducted QRS complex. This can be a normal variant in healthy individuals but can also been seen in various disease states (e.g. rheumatic fever, structural heart...
Third-degree (complete) atrioventricular block

Third-degree AV block is characterized by total failure of atrial impulses to conduct to the ventricle. There is complete dissociation between the atria and ventricles and the ventricular rate is usually slow and regular. The diagnostic feature on ECG is the fact that atrial impulses that should be propagated to the ventricle fail to do so (Fig. 15.3). Complete heart block may be either congenital or acquired. Congenital AV block in infants with otherwise structurally normal hearts may be due to intrauterine exposure to maternal antibodies associated with collagen vascular diseases. Anatomic substrates at high risk of complete AV block include patients with l-transposition of the great arteries with ventricular inversion (congenitally corrected transposition) and those with polysplenia/left isomerism.1 Acquired postoperative AV block is thought to occur from damage to the compact AV node or bundle of His and may be of a transient or permanent nature. The surgical repairs most commonly associated with complete AV block are AV septal defects, ventricular septal defects, tetralogy of Fallot, resection of subaortic obstruction, and interventions in patients with l-transposition of the great arteries.2 The incidence of surgical AV block has been reported to be as high as 2–4% of pediatric patients.3 Eventual recovery of normal conduction occurs in over 60% of subjects and usually does so within the first 10 postoperative days.
Supraventricular tachycardia

Supraventricular tachycardia (SVT) is the most common significant dysrhythmia in infants and children. This rhythm disturbance is characterized by a narrow or “usual” complex QRS morphology and can occur in structurally normal hearts as well as in various forms of CHD. “Usual” complex implies that the QRS morphology in tachycardia is similar to that in normal sinus rhythm. This differentiation is made because patients with CHD often have abnormalities on their baseline ECG including a wide QRS complex. On occasion, widening of the QRS in SVT may be secondary to aberrancy in the right or left bundle branches or because of the tachycardia mechanism. When the QRS complex is wide the distinction of supraventricular from ventricular tachycardia may be difficult.

There are two general types of SVT: automatic and reentrant. These can be differentiated by evaluating characteristics of the tachycardia as listed in Table 15.2. The most common mechanisms of SVT and their electrographic features are listed in Table 15.3. Evaluation of a tachydysrhythmia typically includes a surface 12-lead ECG, a continuous six-lead rhythm strip to document onset and termination, and response to medication (i.e. adenosine) or pacing maneuvers. Bedside or transport monitor strips are helpful to determine tachycardia rate, but are not sufficient for diagnosis or to differentiate among tachycardia mechanisms. In the postoperative patient, temporary atrial pacing wires can be helpful in both diagnosis and management. These wires are typically placed on the epicardial surface of the heart at the conclusion of the surgical intervention. If it is difficult to discern P waves on the surface ECG, an atrial electrogram may be obtained by the use of these wires. This may assist in clearly defining atrial activity and the relationship between

Supraventricular dysrhythmias

Premature atrial contractions

Isolated premature atrial beats, often referred to as premature atrial contractions (PACs) are relatively common in the younger age group (infants and small children). The early P waves on the ECG frequently have an axis and morphology differing from those in normal sinus rhythm and are followed by a normal QRS. On occasion, these may blocked at the AV node or conduct aberrantly (abnormal QRS). Premature atrial contractions are usually benign in nature and require no therapy. Investigation may be initiated in cases of symptomatic, frequent or complex (arising from multiple foci) PACs. If a central venous catheter or any other type of intracardiac line is present, radiographic or echocardiographic assessment of the tip position should be considered and appropriate adjustments performed to eliminate direct atrial irritability as a potential etiology.

Supraventricular tachycardia

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tachydysrhythmias associated with evidence of low cardiac output, recognizing the fact this approach may not always result in restoration of normal sinus rhythm.

Management of SVT depends on the clinical status of the patient, type of tachycardia and precise electrophysiologic mechanism (Table 15.4). If the tachydysrhythmia is associated with moderate to severe hemodynamic compromise, emergent therapy is indicated. Synchronized direct-current cardioversion should be considered for essentially all acute tachydysrhythmias associated with evidence of low cardiac output, recognizing the fact this approach may not always result in restoration of normal sinus rhythm.

**Automatic supraventricular tachycardias**

Automaticity of atrial or AV junctional tissues account for this group of supraventricular tachydysrhythmias. In general these rhythm disorders are more resistant to standard pharmacological therapy than reentrant types.

### Table 15.2 Characteristics of supraventricular tachycardia mechanisms.

<table>
<thead>
<tr>
<th>Mechanism</th>
<th>Automatic</th>
<th>Reentry</th>
</tr>
</thead>
<tbody>
<tr>
<td>Onset and termination</td>
<td>“Warm-up” at initiation</td>
<td>Abrupt</td>
</tr>
<tr>
<td></td>
<td>“Cool-down” at termination</td>
<td>Premature beats</td>
</tr>
<tr>
<td>Mode of initiation</td>
<td>Spontaneous</td>
<td>Narrow</td>
</tr>
<tr>
<td>Ability to initiate/terminate with timed premature beats</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Variation in tachycardia rate</td>
<td>Wide</td>
<td>Narrow</td>
</tr>
<tr>
<td>Response to catecholamines</td>
<td>Increased rate</td>
<td>None or slight rate increase</td>
</tr>
<tr>
<td>Response to adenosine</td>
<td>None</td>
<td>Termination</td>
</tr>
<tr>
<td>Response to drugs that increase refractoriness</td>
<td>Variable</td>
<td>Slowing or termination</td>
</tr>
<tr>
<td>Response to overdrive pacing</td>
<td>Transient suppression, quick resumption</td>
<td>Termination</td>
</tr>
<tr>
<td>Response to cardioversion</td>
<td>None</td>
<td>Termination</td>
</tr>
</tbody>
</table>

### Table 15.3 Mechanisms of most common types of supraventricular tachycardia and electrocardiographic features.

<table>
<thead>
<tr>
<th>Mechanism</th>
<th>Electrocardiographic features</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Automatic tachycardias</strong></td>
<td></td>
</tr>
<tr>
<td>Ectopic atrial tachycardia</td>
<td>Atrial rates of 90–330 beats/min</td>
</tr>
<tr>
<td></td>
<td>Incessant rhythm</td>
</tr>
<tr>
<td></td>
<td>From atrial focus distinct from sinus node</td>
</tr>
<tr>
<td></td>
<td>Abnormal P wave morphology and/or axis</td>
</tr>
<tr>
<td></td>
<td>Distinct P waves preceding QRS complexes</td>
</tr>
<tr>
<td></td>
<td>No influence of AV block on tachycardia</td>
</tr>
<tr>
<td>Junctional ectopic tachycardia</td>
<td>Narrow QRS tachycardia</td>
</tr>
<tr>
<td></td>
<td>Incessant rhythm</td>
</tr>
<tr>
<td></td>
<td>AV dissociation frequent feature</td>
</tr>
<tr>
<td></td>
<td>Atrial rate slower than ventricular rate</td>
</tr>
<tr>
<td></td>
<td>Capture beats frequently seen (QRS complexes slightly earlier than expected from antegrade conduction of normal sinus impulses)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Mechanism</th>
<th>Electrocardiographic features</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Reentrant tachycardias</strong></td>
<td></td>
</tr>
<tr>
<td>Atrial flutter</td>
<td>Sawtooth pattern or more discrete undulating P waves (leads II, II, AVF)</td>
</tr>
<tr>
<td></td>
<td>Various degrees of AV block may be seen</td>
</tr>
<tr>
<td>Atrioventricular reentrant tachycardia (accessory pathway-mediated)</td>
<td>P waves immediately following the QRS complex, on ST segment or T wave</td>
</tr>
<tr>
<td>Concealed bypass tract</td>
<td>AV block results in termination of tachycardia</td>
</tr>
<tr>
<td>Wolff–Parkinson–White syndrome</td>
<td></td>
</tr>
<tr>
<td>Atrioventricular nodal reentry tachycardia</td>
<td>P waves buried within QRS and not discernible</td>
</tr>
<tr>
<td></td>
<td>AV block results in termination of tachycardia</td>
</tr>
</tbody>
</table>

AV, atrioventricular.
<table>
<thead>
<tr>
<th>Rhythm disturbance</th>
<th>Treatment considerations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sinus bradycardia</td>
<td>See Table 15.1</td>
</tr>
<tr>
<td>Sinus tachycardia</td>
<td>Correct underlying cause</td>
</tr>
<tr>
<td>Premature atrial contractions</td>
<td>Evaluate position of central venous line or intracardiac catheter</td>
</tr>
<tr>
<td>Ectopic atrial tachycardia</td>
<td>Correct fever, electrolyte abnormalities Adequate sedation Consider possible role or inotropes/vagolytics Digoxin, usually first drug but rarely effective as single agent Beta-blockers, use with caution if depressed cardiac function Procainamide Amiodarone, sotalol Flecaïnide, propafenone</td>
</tr>
<tr>
<td>Multifocal atrial tachycardia</td>
<td>As in ectopic atrial tachycardia Goals are rate control and decreased automaticity</td>
</tr>
<tr>
<td>Accelerated junctional rhythm</td>
<td>Correct fever Consider possible role or inotropic agents Temporary atrial pacing</td>
</tr>
<tr>
<td>Junctional ectopic tachycardia</td>
<td>Correct fever, electrolyte abnormalities Consider possible role or inotropes/vagolytics Surface cooling to 34–35°C Temporary atrial pacing (for JET rates below 180 beats/min) Hypothermia plus procainamide Amiodarone</td>
</tr>
<tr>
<td>Atrial flutter</td>
<td>Adenosine to confirm diagnosis Atrial overdrive pacing Digoxin Procainamide Amiodarone, sotalol Propafenone</td>
</tr>
<tr>
<td>Atrial fibrillation</td>
<td>Digoxin (except in WPW) Beta-blockers Procainamide, quinidine Amiodarone, sotalol</td>
</tr>
<tr>
<td>Atioventricular reentrant tachycardia or atioventricular nodal reentry tachycardia</td>
<td>Consider vagal maneuvers Adenosine Atrial overdrive pacing Procainamide Amiodarone</td>
</tr>
<tr>
<td>Premature ventricular contractions</td>
<td>Consider and treat underlying cause Lidocaine</td>
</tr>
<tr>
<td>Ventricular tachycardia</td>
<td>Lidocaine Amiodarone Procainamide Magnesium (for torsade de pointes) Beta-blockers Phenytoïn (for digitalis toxicity) Bretylium? (no longer recommended in Pediatric Advanced Life Support Guidelines)</td>
</tr>
<tr>
<td>Ventricular fibrillation</td>
<td>Check for loose ECG electrode mimicking VF Lidocaine Amiodarone (to prevent recurrence)</td>
</tr>
</tbody>
</table>

ECG, electrocardiogram; JET, junctional ectopic tachycardia; VF, ventricular fibrillation, WPW, Wolff–Parkinson–White syndrome.
Institution of antiarrhythmic drugs is based on overall heart rates and the hemodynamic status of the patient. Digoxin has a minimal effect on the atrial focus, but can slow the overall heart rates by slowing AV conduction. After digoxin, the choice of therapy is based on clinical judgment and myocardial function. There are no large clinical series investigating antiarrhythmic drug efficacy in postoperative EAT. Intravenous medications such as β-blockers, procainamide, and amiodarone can be effective in slowing the tachycardia rate. Oral agents (class I, II, and III drugs) may also be of benefit.

In very rare cases in the postoperative patient, EAT may be incessant and life threatening and consideration should be given to a radiofrequency catheter ablation of the atrial focus. Atrial pacing and cardioversion are not likely to be effective.

**Multifocal atrial tachycardia**

Multifocal atrial tachycardia (MAT) also known as chaotic atrial rhythm, is an uncommon atrial dysrhythmia characterized by multiple (at least three) P wave morphologies. These different morphologies correspond to multiple foci of automatic atrial activity. Characteristic ECG features include variable P–P, R–R, and PR intervals. Multifocal atrial tachycardia may be seen in young infants without structural heart disease, in patients with cardiac defects after surgical intervention, and in children with non-cardiac medical conditions. Treatment focuses on ventricular rate control and/or decreasing automaticity. Drugs such as digoxin, procainamide, flecainide, amiodarone, and propafenone have been found to be successful in converting MAT to sinus rhythm in children. Adenosine, pacing and direct-current cardioversion are ineffective.
Numerous therapies have been advocated for JET. In the acute setting strategies include:

1. Control of fever to at least normothermia. Core temperature cooling (to 34 or 35°C) in the younger patient by the use of cooling blankets, fans, or cold compresses has been shown to be of benefit in reducing the tachycardia rate. Shivering, if significant, should be avoided by the use of muscle relaxants in view of potential detrimental increases in oxygen consumption.

2. Decreasing or withdrawing medications associated with catecholamine stimulation or vagolytic agents.

3. Correction of electrolyte abnormalities, especially potassium and calcium.

4. Temporary atrial pacing at heart rates 10–20 beats/minute above the JET rate. This establishes AV synchrony and often benefits the patient hemodynamically. However, if the JET rates are faster than 180 or 190 beats/minute, there is often little benefit with overdrive atrial pacing.

5. Initiation of antiarrhythmic medications. The two most widely used drugs for JET are procainamide and amiodarone. The benefits of procainamide are that it has a faster onset of action and a shorter half-life. The concerns are that it appears to be efficacious only with the use of core cooling. It may also cause a decrease in peripheral vascular resistance and hypotension, especially during bolus infusions. Procainamide may also have negative inotropic properties. Usually, a saline bolus or other volume expander should be administered prior to or during procainamide therapy to maintain adequate hemodynamics.

Amiodarone has a longer onset of action and a longer half-life as compared to procainamide. It has been shown to reduce the heart rate in JET during the initial bolus infusion. Core cooling is often continued but is not needed for efficacy. This may avoid the challenge of having to evaluate clinical signs of adequate cardiac output (distal peripheral perfusion, skin temperature) in a hypothermic, tachycardic patient. Amiodarone does not influence
ventricular function and generally it causes less blood pressure changes during the initial bolus infusion. Both drugs have been shown to be effective in the treatment of JET in published retrospective studies; however, drug choice is significantly influenced by physician/center preference. Due to the fact that procainamide and amiodarone can each result in QT prolongation and proarrhythmic side effects, concomitant drug administration should be used with extreme caution and under guidance of a consultant. Anecdotally, digoxin loading may slow the JET rate, but this has not been well documented in the literature.21 Beta-blockers and calcium channel blockers can depress myocardial contractility, a feature that limits their application in the immediate postoperative period. The use of intravenous class IC agents such as propafenone and flecaainide has been reported, but these agents have not been studied extensively for JET and the intravenous form of propafenone is not yet available in the USA.28–30

6 Overdrive pacing and cardioversion are generally considered ineffective to terminate JET. The natural history of perioperative JET is that it resolves within 2–5 days from the surgical intervention. Long-term antiarrhythmic therapy is usually not necessary. In extreme cases where JET cannot be controlled medically, radiofrequency catheter ablation may be indicated.31,32

Reentrant supraventricular tachycardias

Reentry, also known as “circus” movement or reciprocation, implies that a single stimulus or excitation wave front returns and reactivates the same site or tissue where it came from. Reentrant forms of SVT may or may not involve accessory pathways.

Atrial flutter

Atrial flutter is a dysrhythmia confined to the atrial myocardium. The electrophysiologic basis for this rhythm disturbance involves reentry within the atrium. The typical or classic form of atrial flutter is characterized by a negative saw-tooth P wave pattern and atrial rates of 300 beats/minute (Fig. 15.6). This form of atrial flutter may occasionally be seen in an otherwise healthy neonate but is relatively uncommon in children. The diagnosis of atrial flutter is suggested by abrupt onset of a rapid atrial rhythm that remains relatively regular over time. Atrioventricular nodal conduction accounts for the ventricular response that may be 1:1 or variable. Rapid clinical deterioration is likely with fast ventricular rates and prompt intervention is frequently necessary. Atypical atrial flutter or intra-atrial reentrant tachycardia (IART) usually displays slower atrial rates than the classic form of atrial flutter and varying P wave morphologies. Intra-atrial reentrant tachycardia occurs predominantly in older children and young individuals in association with structural heart disease.33 This is one of the most common dysrhythmias in the postoperative patient and is considered the cause of significant morbidity following certain types of surgical interventions.34 Procedures that involve extensive atrial manipulations, such as atrial redirection procedures (Senning or Mustard operations) and those associated with atrial dilation (Fontan surgery) are particularly at high risk. It has been proposed that anatomic abnormalities related to suture lines, scars or fibrosis from previous surgical interventions in atrial tissue account for the variable atrial rates in these patients.

Management approaches for atrial flutter include:

1 Pharmacologic agents such as digoxin, procainamide, and amiodarone are recommended in acute situations. Drug therapy for controlling the ventricular response in atrial flutter may include in some cases β-blockers (esmolol) or calcium channel blockers (verapamil). Important considerations regarding drug selection are patient age, underlying ventricular function, and presence of sinus node dysfunction (a concomitant problem in patients with recurrent atrial flutter). Newer class III agents (e.g. ibutilide) are

Fig. 15.6 Atrial flutter. The typical features of the classic form of atrial flutter are shown on this surface electrocardiograph. The negative large sawtooth flutter waves in leads II, III, and aVF are seen in addition to the 2:1 atrioventricular block.
available for acute termination of atrial flutter in the adult age group, and may have applications in the postoperative adult with CHD.

2 Although adenosine will not terminate atrial flutter it may assist in confirmation of the diagnosis by enhancing AV block and uncovering flutter waves.

3 Atrial overdrive pacing (via esophagus, intracardiac pacing catheter, or epicardial wires) has been shown to be safe and effective in the acute termination of atrial flutter. After determination of the atrial cycle length, rapid atrial stimulation is performed in short bursts to attempt interruption of the reentry circuit.

4 Synchronized direct current cardioversion is the treatment of choice for any patient with unstable hemodynamics associated with atrial flutter (0.5–1.0 J/kg body weight).

5 Chronic drug therapy is frequently required in patients with CHD because of the potential for recurrence and associated fast AV conduction.

6 Pacemaker therapy, atrial antitachycardia pacing, and radiofrequency ablation are additional modalities more applicable to long-term management.

Atrial fibrillation

Atrial fibrillation is the result of many small reentrant circuits within the atrium. In the pediatric age group this tachydysrhythmia is less frequent than atrial flutter. The atrial rates are rapid and irregular ranging from 400 to 700 beats/minute. Ventricular response rates are variable but generally range between 80 and 150 beats/minute. Patients at potential risk for atrial fibrillation include those with pre-excitation syndromes, rheumatic heart disease, structural heart disease (Ebstein’s anomaly, tricuspid atresia, atrial septal defects), severe AV valve regurgitation and cardiomyopathies.

Management principles in atrial fibrillation are similar to those for atrial flutter except that atrial overdrive pacing is not effective in terminating the dysrhythmia. Cardioversion is more likely to be required. Generally a higher amount of energy is necessary for cardioversion in patients with atrial fibrillation as compared to those with atrial flutter. An orientation of the cardioversion pads over the front and back of the heart in order to shock the entire atrium may be necessary. Anticoagulation and consideration of transesophageal echocardiography for evaluation of intracardiac thrombi is recommended prior to cardioversion if atrial fibrillation has been present more than a few days.

Atrioventricular reentrant tachycardia and atrioventricular nodal reentrant tachycardia

Atrioventricular reentrant tachycardia (AVRT) mediated by an accessory pathway between the atrium and ventricle is the most common form of SVT in infancy and childhood. Typically, the tachycardia circuit consists of conduction from the atrium, down the AV node, through the His bundle and ventricles, up the accessory pathway, and back to the atrium. This form is called “orthodromic” SVT and occurs in patients with Wolff–Parkinson–White syndrome (WPW; short PR interval, delta wave, abnormal QRS morphology), concealed accessory pathways (bypass tract utilized only as retrograde limb of reentrant circuit in SVT), and permanent junctional reciprocating tachycardia (PJRT). In contrast, in “antidromic” SVT conduction travels from the atrium, down the accessory pathway, through the ventricles, up the AV node, and back to the atrium. The QRS complex in this form of SVT is wide. Antidromic tachycardia can occur in patients with WPW and other pre-excitation variants (Mahaim tachycardia).

Atrioventricular nodal reentrant tachycardia (AVNRT), or reentry within the AV node, is more likely in the adolescent or young adult. In AVNRT there are two physiologically distinct components of the AV node designated as “slow” and “fast” AV nodal pathways. The typical form of AVNRT consists of antegrade conduction (from the atrium to the ventricle) via the slow pathway followed by retrograde conduction (back to the atrium) via the fast pathway.

Both AVRT (Fig. 15.7) and AVNRT (Fig. 15.8) have clinical characteristics of the reentrant tachycardia mechanisms listed in Table 15.2. The two can often be distinguished by close evaluation of the surface ECG in tachycardia. Patients with structurally normal hearts as well as those with CHD can have either AVRT or AVNRT. Ebstein’s anomaly of the tricuspid valve is frequently associated with AVRT secondary to one or multiple accessory pathways. The accessory pathways in this condition are usually right-sided and of either the WPW or concealed varieties. Transposition of the great arteries can be associated with Ebstein-like features of the left-sided AV valve and left-sided accessory pathways can be identified in a subset of these patients.

Management principles of AVRT or AVNRT include the following:

1 If the patient is hemodynamically unstable emergent direct current cardioversion (0.5–1.0 J/kg) should be carried out (Fig. 15.9). A lower energy setting is adequate for epicardial paddles. This should also be considered in the stable patient when potential rapid clinical deterioration is anticipated or after unsuccessful conventional therapy.

2 In the stable patient various modalities can be utilized to acutely terminate the tachycardia. Vagal maneuvers (ice application to face in infants, coughing, gag reflex stimulation, Trendelenburg position) enhance parasympathetic influences and may acutely terminate the tachycardia. Continuous electrocardiographic monitoring is recommended as well as the availability of atropine as transient bradycardia following tachycardia termination may be seen. Adenosine has become first line therapy for SVT. Other pharmacologic agents (digoxin, edrophonium, bi-blocking agents, calcium channel blockers, phenylephrine) have
In patients with high catecholamine states, termination of SVT can be successful but rapid recurrence may be seen. In this instance, it is helpful to sedate the patient and limit catecholamine stimulation if possible.

1. Once the tachycardia has terminated or if it terminates and then reinitiates, antiarrhythmic drug therapy can be instituted and the maneuvers again attempted. In patients with high catecholamine states, termination of SVT can be successful but rapid recurrence may be seen. In this instance, it is helpful to sedate the patient and limit catecholamine stimulation if possible.

3. Rapid atrial pacing may be conducted via a transesophageal electrode catheter or via temporary atrial pacing wires. One should first establish that at the pacing outputs utilized, the electrode catheter or the temporary wires do not capture the ventricle and cause ventricular contraction. Then, rapid atrial pacing is performed by pacing the atrium at 10–20% faster than the SVT rate for a period of up to 15 s, which typically terminates the tachydysrhythmia. If the tachycardia terminates but then reinitiates, antiarrhythmic drug therapy can be instituted and the maneuvers again attempted. In patients with high catecholamine states, termination of SVT can be successful but rapid recurrence may be seen. In this instance, it is helpful to sedate the patient and limit catecholamine stimulation if possible.

4. Once the tachycardia has terminated or if it terminates and then reinitiates, antiarrhythmic medication can be instituted. For perioperative patients not able to take oral medications, parenteral therapy includes digoxin, procainamide, and amiodarone. Beta-blockers and calcium channel blockers are much less desirable in view of their negative effects on myocardial contractility. Thus, their use is limited in the immediate perioperative period.
5 If the patient has incessant tachycardia and cannot be controlled with medications, radiofrequency catheter ablation may be warranted.

**Ventricular dysrhythmias**

Ventricular dysrhythmias are disorders that arise distal to the bifurcation of the common His bundle. These are relatively rare in young children and more commonly seen in the adolescent or young adult with a history of operated CHD. Patients with ventricular rhythm abnormalities may have minimal to no symptomatology or be gravely ill. Evaluation of ventricular dysrhythmias should include a review of the medical history for the presence of associated cardiovascular pathology or potential cause, analysis of the ECG and, very importantly, assessment of the hemodynamic state of the patient.

**Premature ventricular contractions**

Premature ventricular beats, often referred to as premature ventricular contractions (PVCs) are characterized by: (i) prematurity of the QRS complex; (ii) a QRS morphology that differs from that in sinus rhythm; (iii) prolongation of the QRS duration for age (this is a frequent finding but may not always be the case); (iv) abnormalities of the ST segment and T wave; and (v) a premature ventricular complex not preceded by premature atrial activity. Premature ventricular contractions of a single QRS morphology (uniform or morphic), without associated symptoms and in patients with structurally normal hearts are generally considered benign. Premature ventricular contractions that merit further investigation include those of multiple morphologies (multiform or polymorphic) on ECG, that occur with moderate frequency (runs) or in succession (couplets, triplets) and are associated with symptoms, or present in the context of an abnormal heart.

Ventricular ectopy in the perioperative period may be secondary to myocardial irritation from intracardiac catheters or direct surgical stimulation. Additional etiologies include respiratory (hypoxemia), electrolyte (hypokalemia) or metabolic (acidosis) derangements. Isolated PVCs may also be due to pharmacological agents (including recreational drugs), myocardial injury, poor hemodynamics, and prior complex surgical intervention. In some cases drug therapy (i.e. lidocaine) may be indicated to prevent degeneration of ventricular ectopy into a malignant rhythm.

**Ventricular tachycardia**

Ventricular tachycardia (VT) is defined as three or more consecutive ventricular beats occurring at a rate greater than 120 beats/minute. The QRS morphology in VT is different than that in sinus rhythm but not necessarily wide for age. ECG features that favor this diagnosis include: (i) AV dissociation; (ii) intermittent fusion (QRS complex of intermediate morphology between two other distinct QRS morphologies); (iii) QRS morphology of VT similar to that of single PVCs; and (iv) tachycardia rate in children usually below 250 beats/minute. A right bundle branch block QRS morphology is most common in infants with VT, whereas in older children a left bundle branch block QRS morphology is more frequent with likely widening of the QRS.

Various qualifiers have been proposed to further characterize VT. The classification as monomorphic or polymorphic is based on the evaluation of the QRS morphology in multiple ECG leads. Ventricular tachycardia is considered to be sustained or non-sustained if it lasts more or less than 10 seconds respectively.

Acute onset of VT in pediatric patients may be due to hypoxia, acidosis, electrolyte imbalance or metabolic problems. Ventricular tachycardia may also occur in the context of depressed myocardial function, poor hemodynamics, prior surgical interventions, cardiomyopathies (hypertrophic, dilated, arrhythmogenic right ventricular dysplasia), myocardial tumors, acute injury (inflammation, trauma), and prolonged QT syndromes. Among patients with CHD and ventricular dysrhythmias, those at higher risk include older patients following tetralogy of Fallot repair. The following potential causes of ventricular ectopy in patients with structural heart disease have been proposed: inadequate myocardial protection during the surgical procedure, chronic pressure or volume loads, residual or recurrent pathology, and scar formation at the ventriculotomy site.

**Monomorphic ventricular tachycardia**

Although occasionally seen in patients with presumably normal hearts, monomorphic (single dominant or constant morphology) VT is a more common phenomenon in patients with diseased hearts. In the abnormal heart the tachycardia is thought to originate from a reentry focus in scarred or damaged myocardial tissue. The electrographic findings are those of a wide regular QRS rhythm of uniform morphology (Fig. 15.10).

**Polymorphic ventricular tachycardia**

**Torsade de pointes**

Torsade de pointes (“twisting of the peaks”) refers to polymorphic VT. The characteristic ECG feature of this dysrhythmia is that of a varying QRS morphology manifested as positive and negative oscillations of the QRS direction that twist around an isoelectric baseline (Fig. 15.11). Polymorphic VT may occur in long QT syndromes, can be secondary to drug therapy or neurologic pathology or the result of
myocardial ischemia. Torsade may terminate spontaneously or degenerate into ventricular fibrillation (VF).

**Long QT syndromes**

The long QT syndrome can occur in the congenital (inherited) or acquired forms. This distinction is relevant to management strategies. The congenital varieties are considered to be the result of a genetic defect in the sodium or potassium channels responsible for maintaining electrical homeostasis in the heart. A diagnostic criteria has been suggested for the long QT syndrome. This scoring system includes ECG findings, clinical history (deafness, syncope), and family history. A frequent, but not essential, feature is prolongation of the corrected QT interval (QTc) on the resting ECG. The QTc is derived as follows:

\[
\text{Corrected QT} = \frac{\text{measured QT interval}}{\sqrt{\text{of preceding RR interval}}}.
\]

A QTc greater than 0.48 seconds is considered abnormal regardless of age. In addition to torsade de pointes, potential dysrhythmias in patients with long QT syndrome include VF and bradydysrhythmias. An important consideration in the care of these patients is ensuring adequate β-adrenergic blockade preoperatively and minimizing adrenergic stimulation. Conditions and drugs associated with QT interval prolongation should be avoided. Intraoperative dysrhythmias can be treated with additional doses of β-blockers. Others drugs to be considered include phenytoin and lidocaine. Bradydysrhythmias can be managed by pacing. Despite the fact that several anesthetics (intravenous medications and volatile agents) increase the QT interval, in most cases these drugs are used without untoward effects.

Acquired forms of long QT may result from electrolyte disturbances (hypokalemia, hypocalcemia, hypomagnesemia), drug therapy (antiarrhythmic agents, antipsychotic drugs, cisapride), and neurologic or endocrine abnormalities. Therapy in this setting should focus on correction of the underlying cause.

General considerations in the management of VT are as follows:

1. A wide QRS tachycardia should always be considered to be of ventricular origin although some atypical forms of supraventricular dysrhythmias may mimic VT.
2 The primary approach in the care of a patient with an acute ventricular rhythm disturbance is consideration of possible causes and prompt evaluation of hemodynamics. In general, sustained ventricular dysrhythmias are poorly tolerated and require immediate attention. Cardiopulmonary resuscitation should be instituted in the unstable patient while preparing for cardioversion.

3 Pharmacological therapy may be indicated in the stable patient or for the prevention of recurrence in VT. Lidocaine is recommended as first line drug and procainamide as second choice therapy. Additional agents that may be of benefit in refractory dysrhythmias include β-blockers and amiodarone.

4 Electrical cardioversion in torsade de pointes should be performed only if the dysrhythmia is sustained. In patients with frequent but non-sustained runs of torsade de pointes cardioversion is of no benefit and may be detrimental. Magnesium sulfate is considered to be the first line drug and lidocaine may have a role in therapy. Procainamide is contraindicated due to QT prolongation.

5 In addition to β-blockade, congenital forms of the long QT syndrome may in some cases be treated with implanted defibrillators.

6 For polymorphic VT associated with acquired QT prolongation, such as that related to the proarrhythmic effects of certain drugs, pacing and isoproterenol may be more appropriate. In this setting it is thought that the electrophysiologic mechanism that initiates the tachycardia is the result of long pauses in the cardiac cycle.

**Ventricular fibrillation**

Ventricular fibrillation is an uncommon dysrhythmia in children characterized by chaotic, asynchronous ventricular depolarizations failing to generate an effective cardiac output. The ECG morphology in VF demonstrates low amplitude irregular deflections without identifiable QRS complexes. A loose ECG electrode may mimic these surface ECG features; therefore, immediate clinical assessment of cardiac output (checking for a pulse) and adequate pad contact should be performed when VF is suspected.

Considerations in the management of VF are as follows:

1 This is a lethal dysrhythmia if untreated; therefore immediate defibrillation (initial dose 2 J/kg for the transthoracic approach) is the definitive therapy. If this is unsuccessful, the energy dose should be doubled (4 J/kg) and repeated. Infant paddles are generally recommended for infants weighing less than 10 kg. Larger adult paddles are suggested for children weighing over approximately 10 kg in order to reduce impedance and maximize current flow.

2 Adequate airway control (oxygenation, ventilation) and chest compressions should be rapidly instituted while preparing for defibrillation or between shocks if several defibrillation attempts are needed. Resuscitative drugs such as epinephrine should strongly be considered without delaying defibrillation.

3 Adjunctive pharmacologic agents for VF include lidocaine and amiodarone. The intravenous preparation of sotalol may be an option in countries where available. Bretylium use in children with VF is not well documented and it is no longer considered an appropriate agent.

4 Additional therapies such as mechanical circulatory support may be an option in selected cases. The most generally accepted management strategies for acute therapy of perioperative rhythm disturbances without associated hemodynamic compromise are summarized in Table 15.4.

**Pharmacologic therapy of cardiac dysrhythmias**

Antiarrhythmic drugs exert blocking actions predominantly on sodium, potassium or calcium channels, or adrenergic receptors. These pharmacologic agents are generally classified according to their presumed mechanism of action and electrophysiologic properties (Table 15.5). The drug classification scheme described by Vaughan Williams in the 1960s and modified over the years is one frequently used and may be helpful in predicting the response to antiarrhythmic drug therapy. Appropriate selection of pharmacologic therapy by the anesthesiologist requires an understanding of dysrhythmia origin, presumed mechanism, and drug. This section discusses antiarrhythmic drug therapy focusing on the drugs most frequently used for acute management in the pediatric age group (Table 15.6). It should be emphasized that perioperative consultation with a cardiologist should be considered during the care of complex rhythm disturbances or if involving patients receiving chronic antiarrhythmic drug therapy.

**Class I agents**

The largest group of antiarrhythmic drugs is the sodium channel blockers. The relatively large size of this class has led to subclassification of these agents into IA, IB, and IC groups based on their cellular actions. Group IC is for oral therapy and will not be discussed in this chapter.

The class IA drugs include procainamide, quinidine, and disopyramide. The predominant electrophysiologic effects of these agents are prolongation of myocardial repolarization (QT interval) and duration of the action potential. Their mechanism of action is primarily related to inhibition of the fast sodium channels. The anticholinergic (vagolytic) properties of these drugs account for their more pronounced effects at fast heart rates.
**Table 15.5** Classification of antiarrhythmic agents.

<table>
<thead>
<tr>
<th>Class and action</th>
<th>Drugs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Class I: Sodium-channel blockers. Drugs may be subclassified into IA, IB, and IC categories</td>
<td></td>
</tr>
<tr>
<td>IA agents: Moderately depress phase zero upstroke of the action potential, slow conduction, and prolong repolarization. Effectively slow conduction in atria, ventricles, and accessory connections</td>
<td>Procainamide&lt;br&gt;Quinidine&lt;br&gt;Disopyramide</td>
</tr>
<tr>
<td>IB agents: Shorten action potential duration and result in minimal alteration of conduction. These agents are usually not effective in the treatment of supraventricular tachycardia</td>
<td>Lidocaine&lt;br&gt;Mexiletine&lt;br&gt;Phenytoin</td>
</tr>
<tr>
<td>IC agents: Significantly depress phase zero upstroke, with marked slowing of conduction but impart little change in refractoriness</td>
<td>Flecainide&lt;br&gt;Propafenone</td>
</tr>
<tr>
<td>Class II: Beta-adrenergic receptor blockers. Antiarrhythmic effects result from conduction and decreasing automaticity, particularly in the sinoatrial and atrioventricular nodes</td>
<td>Esmolol&lt;br&gt;Atenolol&lt;br&gt;Metoprolol&lt;br&gt;Propranolol</td>
</tr>
<tr>
<td>Class III: Potassium-channel blockers. Primarily prolong action potential duration with resultant prolongation of refractoriness</td>
<td>Amiodarone&lt;br&gt;Sotalol&lt;br&gt;Bretylium&lt;br&gt;Ibutilide</td>
</tr>
<tr>
<td>Class IV: Calcium channel blockers with predominant sites of action in the sinoatrial and atrioventricular nodes</td>
<td>Verapamil&lt;br&gt;Diltiazem</td>
</tr>
<tr>
<td>Others</td>
<td>Atropine&lt;br&gt;Digoxin&lt;br&gt;Adenosine&lt;br&gt;Magnesium sulfate</td>
</tr>
</tbody>
</table>

**Procainamide**

Procainamide is a potent sodium channel blocker, and to a lesser extent, a potassium channel blocker. This agent slows atrial conduction (prolongs PR interval) and lengthens the QRS duration and QTc interval. Procainamide is useful in the management of both atrial and ventricular dysrhythmias.\(^{21,46-48}\) The suppression of abnormal automaticity accounts for the role of this agent in the treatment of EAT, JET, and VT. This drug is more effective than lidocaine in acutely terminating sustained VT.

Procainamide may be administered via the oral, intravenous, or intramuscular routes. For the treatment of acute dysrhythmias, intravenous loading (doses of 10–15 mg/kg) is usually required. This is administered over a period of 30–45 minutes. The lower end of the loading dose spectrum is recommended for younger patients. Continuous ECG monitoring and frequent blood pressure assessments are recommended during the loading phase. The drug is rapidly distributed following intravenous injection. An infusion is frequently initiated at 40–50 µg/kg/minute. Monitoring of plasma levels is advisable during maintenance infusion. The infusion rate is adjusted accordingly to maintain therapeutic levels between 4 and 8 µg/mL. The drug is eliminated by the kidneys (50–60%) and via hepatic metabolism (10–30%). Hepatic acetylation accounts for the generation of N-acetylprocainamide (NAPA), a metabolite with antiarrhythmic (class III) properties.

Potential side effects include hypotension due to blocking of α-adrenergic receptors and decreased peripheral vascular resistance during rapid intravenous administration. Significant QTc prolongation and proarrhythmia is a known side effect. Additional non-therapeutic effects include negative inotropism and AV block. Gastrointestinal symptoms, a lupus-like syndrome, and blood dyscrasias may also occur.

**Class IB agents**

Class IB drugs include lidocaine, mexiletene, phenytoin, and tocainide. These inhibit fast sodium channels and shorten the action potential duration and refractory period.
Elevated plasma levels beyond the therapeutic range may cause gastrointestinal symptoms (nausea and vomiting), central nervous system pathology (paresthesias, tremor, confusion, seizures) and in rare instances hemodynamic perturbations may be seen.

Class II agents

The class II drugs (esmolol, atenolol, metoprolol, and propranolol) block β-adrenergic receptors to variable extents (receptor selectivity and intrinsic sympathomimetic activity) depending on the specific agent. Antiarrhythmic effects result from slowing conduction and decreasing automaticity, particularly in the sinoatrial and AV nodes. These agents universally decrease sympathetic activity through β-receptor blockade.

Esmolol

Esmolol is a predominant β₁-selective (cardioselective) adrenergic receptor-blocking agent with a rapid onset and...
very short duration of action. The primary electrophysiologic drug property is inhibition of sinoatrial and AV conduction. The brief elimination half-life of this drug after intravenous injection (approximately 9 minutes) is a feature that has made it desirable in the perioperative and intensive care settings. Esmolol is commonly used for heart rate and blood pressure control and management of a wide number of tachydysrhythmias (supraventricular and ventricular). Following an intravenous loading dose of 500 µg/kg (over 1–2 minutes), an infusion is initiated at 50 µg/kg/minute and titrated to effect by increasing the infusion rate. Because of its short half-life, blood levels of esmolol can be rapidly altered by increasing or decreasing the infusion rate and rapidly eliminated by discontinuing the infusion.

Most adverse effects related to esmolol therapy have been mild and transient. Reported cardiovascular side effects include bradycardia, sinus pauses, AV block, hypotension, and negative inotropism. These are more likely to be seen during bolus therapy.

Class III agents

The class III drugs (amiodarone, sotalol, bretylium, ibutilide) block potassium channels and increase action potential duration and refractoriness in atrial and ventricular muscle and in Purkinje fibers.

Amiodarone

Amiodarone has a wide spectrum of actions with multiple and complex electrophysiologic effects which encompass all four antiarrhythmic drug classes. Class I actions include inhibition of fast sodium channels. Class II and IV effects result in depression of sinus node automaticity and function, and slowing of AV and His–Purkinje system conduction. As a class III agent, amiodarone delays repolarization and increases action potential duration resulting in prolongation of refractoriness in all cardiac tissues and accessory connections if present. In addition to blocking potassium channels, amiodarone exhibits vagolytic properties, weakly blocks calcium channels and non-competitively blocks α- and β-adrenergic receptors. The efficacy of this agent has been documented against many supraventricular (EAT, atrial flutter and fibrillation) and ventricular dysrhythmias (VT and VF). The usefulness of this drug in the treatment of life-threatening tachydysrhythmias accounts for its increasing role in emergency cardiovascular management.

Intravenous therapy requires a loading dose because of its rapid plasma disappearance during the distribution phase. In children the suggested dose is 5 mg/kg over 1 hour. The same dose is then infused over 12 hours, and repeated if necessary. Amiodarone binds extensively to most tissues, accounting for its extremely prolonged elimination. The slow elimination rate of amiodarone leads to an unusually long half-life (25–110 days).

Amiodarone administration may result in sinus bradycardia and AV block. Hypotension is an unlikely complication of intravenous therapy. Electrocardiogram effects include PR, QRS, and QTc prolongation. There are significant drug interactions with amiodarone that merit attention. Coadministration with other antiarrhythmic agents (digoxin, procainamide, flecainide, quinidine, phenytoin) may result in increased levels of these drugs. The concomitant use of amiodarone with β-blockers or calcium channel antagonists should raise concerns of potential synergistic effects on conduction tissue. A number of adverse effects have been reported with long-term oral therapy in children. These include skin discoloration, corneal microdeposits, alterations in hepatic and thyroid function, pulmonary fibrosis, and neurologic disturbances.

Bretylium

The clinical experience with this drug is limited in the pediatric age group. Indications have included VT or VF unresponsive to standard therapy. In the latest guidelines of the American Heart Association for pediatric resuscitation, bretylium is no longer considered an appropriate agent because of the risk of hypotension, the lack of demonstrable effectiveness in VT, and the absence of published studies of its use in children.

Ibutilide

Ibutilide is an intravenous class III agent approved in the adult population for the acute conversion of atrial flutter and fibrillation of recent onset (< 90 days) to sinus rhythm. Like other drugs that prolong ventricular repolarization, this agent may be associated with excessive QT prolongation and polymorphic VT requiring careful patient selection and monitoring during drug administration. The clinical experience with ibutilide in the pediatric age group is extremely limited.

Class IV agents

The class IV drugs, also known as calcium channel blockers (verapamil, diltiazem, nifedipine) inhibit the slow inward calcium current.

Verapamil

The actions of this drug are mediated through prolongation of conduction time and refractory period in nodal tissue. Verapamil has been shown to be efficacious in the management of SVT, certain types of VTs, and hypertrophic cardiomyopathy in children.
Verapamil should not be used in young children (< 1 year of age) in view of the potential for severe hemodynamic compromise (refractory hypotension, myocardial depression, asystole and cardiovascular collapse) following its administration. The detrimental effects are related to calcium channel blockade and uncoupling of excitation–contraction in myocardial cells. In older children (beyond 1 year of age) verapamil is infused in a dose of 0.1 mg/kg. The concomitant use of verapamil and β-blocking agents may result in serious cardiovascular side effects and is therefore not recommended. In the setting of WPW syndrome, verapamil may enhance the ventricular response rate of atrial fibrillation leading to hemodynamic compromise.

Other agents

Atropine

Atropine sulfate, an antimuscarinic, parasympatholytic drug, accelerates sinus or atrial pacemakers, and enhances AV conduction. Atropine is recommended in the treatment of symptomatic bradycardia caused by increased vagal activity or AV block, such as vagally mediated bradycardia during intubation. Atropine may be used to treat bradycardia accompanied by poor perfusion or hypotension; however, epinephrine may be a more effective therapy in this setting. Efforts to ensure adequate oxygenation and ventilation and exclude hypothermia should precede pharmacologic therapy of bradycardia.

The recommended dose is 0.02 mg/kg, with a minimum dose of 0.1 mg. The maximum single dose is 0.5 mg in a child and 1.0 mg in an adolescent or young adult. The dose may be repeated in 5 minutes, to a maximum total dose of 1.0 mg in a child and 2.0 mg in an adolescent. In the absence of intravenous access, atropine (0.02 mg/kg) may be administered tracheally or intramuscularly, although with less reliable absorption than through the intravenous route. Small doses of atropine may be associated with transient heart rate slowing. Atropine may rarely cause cardiac dysrhythmias.

Digoxin

Digitalis glycosides have been used for many years as frontline pharmacologic agents in the management of certain dysrhythmias. The electrophysiologic properties of digoxin are the result of direct effects on cardiac tissues (through inhibition of the sarcolemmic sodium pump) and indirect effects via the autonomic (parasympathetic) nervous system. Digoxin is known to increase the refractory period and decrease the conduction velocity of the specialized cardiac conduction system, slow the sinus rate (primarily by enhancing vagal discharge), and shorten the refractory period in atrial and ventricular muscle.

Digoxin is effective in the treatment of a wide spectrum of supraventricular dysrhythmias such as SVT, atrial flutter, atrial fibrillation, and chaotic atrial tachycardia. In patients with WPW and ECG evidence of antegrade conduction via the accessory pathway in tachycardia (wide QRS complexes), digoxin is not recommended. This is related to the fact that digoxin may alter the conduction properties of the accessory pathway and lead to malignant dysrhythmias (VT and VF) during episodes of atrial flutter or fibrillation and 1:1 AV conduction.

Digoxin can be administered orally or parenterally. In view of the fact that the onset of the digitalis effect may be delayed (up to 5 h), this drug may be less than ideal in the treatment of acute symptomatic tachycardias. Despite this limitation, digitalis glycosides remain useful in controlling the ventricular response in atrial tachydysrhythmias, particularly during atrial flutter or fibrillation. A common loading algorithm utilizes a total digitalizing dose of 30–50 µg/kg. Half of this amount is given initially followed by two doses at 6-hour intervals of 25% of the total digitalizing dose each. For intravenous use, the total digitalizing dose is reduced to 65–75% of the total oral dose given following a similar scheme. Inappropriate dose calculations may result in drug overdose emphasizing the fact that drug calculations should be carefully performed and corroborated prior to drug administration. Maintenance doses of digoxin are 7–10 µg/kg/day.

Digoxin is tightly bound to peripheral tissue proteins. Drug excretion is via the kidneys. Dose adjustments are indicated in the case of renal impairment or congestive heart failure.

The concomitant use of digoxin with other antiarrhythmic agents (amiodarone, quinidine, verapamil) requires an adjustment (reduction) in the digoxin dose and monitoring of plasma levels. Toxic manifestations of digitalis therapy may be classified as cardiac and non-cardiac. Digoxin toxicity can cause virtually any type of cardiac rhythm disturbance. Non-cardiac manifestations of digitalis toxicity include gastrointestinal (nausea, vomiting, anorexia) and neurologic symptoms (headache, lethargy, weakness, confusion, seizure), and visual disturbances. Although non-specific, non-cardiac symptoms are the earliest manifestations of digitalis toxicity.

Adenosine

Adenosine is a purine agonist, with effects mediated via the activation of the A1-adenosine receptor (leading to activation of adenylate cyclase and intracellular cyclic AMP production). The electrophysiologic effects are secondary to an increase in potassium conductance and depression of the slow inward calcium current resulting in transient sinus slowing or AV nodal block. This accounts for its therapeutic value in terminating dysrhythmias that involve the AV node. Adenosine is the drug of choice for acute treatment of SVT. Reentrant supraventricular tachydysrhythmias that involve the AV node as part of the circuit are particularly sensitive to
Magnesium sulfate

Magnesium is a major intracellular cation, cofactor in multiple enzymatic reactions, and important regulator of numerous cardiovascular processes. Magnesium sulfate therapy is indicated as adjunct management for dysrhythmias in patients with documented hypomagnesemia or torsade de pointes. Magnesium deficiency is frequently seen in the context of other electrolyte abnormalities (hypokalemia and hypocalcemia). Rhythm disturbances associated with hypomagnesemia resemble those with hypokalemia or digitalis toxicity. In the setting of torsade de pointes, VT intravenous infusion (over several minutes) of 25–50 mg/kg (up to 2 g) is recommended. Approximately 70% of plasma Mg²⁺ is ultrafiltered by the kidney and the remainder is bound to protein. Side effects associated with magnesium administration include flushing, diaphoresis, muscle weakness, and central nervous system depression. Magnesium levels well above the therapeutic range can lead to serious morbidity such as cardiac conduction defects, respiratory depression, and circulatory collapse.

Pacemaker therapy in children

Pacemaker nomenclature

Pacemaker nomenclature as established by the North American Society of Pacing and Electrophysiology and the British Pacing and Electrophysiology Group is detailed in Table 15.7. The generic pacemaker (NBG) code has five positions. The first position or letter of the code refers to the chamber(s) paced, the second to the chamber(s) sensed, the third to the pacemaker’s response to sensing, and the fourth to programmability and rate modulation. The fifth position is restricted to antitachycardia function and is used infrequently.

Permanent cardiac pacing

Advances in pacemaker technology, enhancements in programmability, and miniaturization of units have resulted in the increasing use of these devices in the pediatric age group. The American Heart Association/American College of Cardiology published recent guidelines for permanent pacing in children and adolescents in 1998. Table 15.8 lists indications for which there is general agreement that the device should be implanted (class I) and for which pacemakers are used frequently but diverging opinions exist regarding benefits (class II). In general terms these indications can be summarized as follows:

<table>
<thead>
<tr>
<th>Position I</th>
<th>Position II</th>
<th>Position III</th>
<th>Position IV</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chambers paced</td>
<td>Chambers sensed</td>
<td>Response to sensing</td>
<td>Programmability, rate modulation</td>
</tr>
<tr>
<td>O, none</td>
<td>O, none</td>
<td>O, none</td>
<td>O, none</td>
</tr>
<tr>
<td>A, atrium</td>
<td>A, atrium</td>
<td>I, inhibited</td>
<td>P, simple programmable</td>
</tr>
<tr>
<td>V, ventricle</td>
<td>V, ventricle</td>
<td>T, triggered</td>
<td>M, multiprogrammable</td>
</tr>
<tr>
<td>D, dual (A + V)</td>
<td>D, dual (A + V)</td>
<td>D, dual (I + T)</td>
<td>C, communicating</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>R, rate modulation</td>
</tr>
</tbody>
</table>

Table 15.7 Generic pacemaker code.

Table 15.8 Indications for permanent pacing in children and adolescents.

<table>
<thead>
<tr>
<th>Class</th>
<th>Indications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Class I</td>
<td>• Advanced second- or third-degree atrioventricular (AV) block associated with symptomatic bradycardia, congestive heart failure, or low cardiac output&lt;br&gt;• Sinus node dysfunction with correlation of symptoms during age-inappropriate bradycardia. The definition of bradycardia varies with the patient’s age and expected heart rate&lt;br&gt;• Postoperative advanced second- or third-degree AV block that is not expected to resolve or persists at least 7 days after cardiac surgery&lt;br&gt;• Congenital third-degree AV block with a wide QRS escape rhythm or ventricular dysfunction&lt;br&gt;• Congenital third-degree AV block in the infant with a ventricular rate &lt; 50–55 beats/min or with congenital heart disease and a ventricular rate &lt; 70 beats/min&lt;br&gt;• Sustained pause-dependent VT, with or without prolonged QT, in which the efficacy of pacing is thoroughly documented</td>
</tr>
<tr>
<td>Class IIa</td>
<td>• Bradycardia–tachycardia syndrome with the need for long-term antiarrhythmic treatment other than digitalis&lt;br&gt;• Congenital third-degree AV block beyond the first year of life with an average heart rate &lt; 50 beats/min or abrupt pauses in ventricular rate that are two or three times the basic cycle length&lt;br&gt;• Long QT syndrome with 2 : 1 AV or third-degree AV block&lt;br&gt;• Asymptomatic sinus bradycardia in the child with complex congenital heart disease with resting heart rate &lt; 35 beats/min or pauses in ventricular rate &gt; 3 s</td>
</tr>
<tr>
<td>Class IIb</td>
<td>• Transient postoperative third-degree AV block that reverts to sinus rhythm with residual bifascicular block&lt;br&gt;• Congenital third-degree AV block in the asymptomatic neonate, child, or adolescent with an acceptable rate, narrow QRS complex, and normal ventricular function&lt;br&gt;• Asymptomatic sinus bradycardia in the adolescent with congenital heart disease with resting heart rate &lt; 35 beats/min or pauses in ventricular rate &gt; 3 s</td>
</tr>
</tbody>
</table>


1 Symptomatic sinus bradycardia.<br>2 Recurrent bradycardia–tachycardia syndromes.<br>3 Congenital AV block.<br>4 Advanced second- or third-degree AV block.<br>5 Congenital atrioventricular (AV) block. An important consideration in the setting of CHD is correlation of symptoms with recommended criteria for pacemaker placement in view of the physiologic alterations associated with structural heart disease or that may arise or persist following surgical intervention.

Implantation techniques

Permanent pacemaker implantation is accomplished via the transvenous or epicardial approach. These procedures are typically performed under sterile conditions in the cardiac catheterization laboratory, electrophysiology suite, or operating room. Local anesthesia with supplemental intravenous sedation may be used in the older age group; however, most infants and small children require a general anesthetic.

The transvenous technique uses the subclavian, cephalic, and axillary vein for access. Under fluoroscopic guidance pacing leads are advanced into the right atrium and/or ventricle and fixed to the endocardium according to the specific lead mechanism. After adequate sensing, capture thresholds and lead impedances are documented, and the leads are attached to a generator positioned in the pectoral region. The following are considered contraindications to transvenous pacing: right-to-left intracardiac shunts, prosthetic tricuspid valves, certain types of structural heart disease, anatomy not suitable for transvenous access to cardiac chambers, recurrent lead dislodgment, and small patient size (< 10 kg). Advantages of the transvenous route include longer generator longevity because of lower pacing thresholds and lower incidences of lead fractures. Disadvantages are potential narrowing or thrombosis of venous pathways, lead dislocation, risk of systemic embolization in the presence of an intracardiac shunt, and possible endocarditis.

For epicardial implantation the leads are attached to the epimyocardial surface of the heart and after appropriate testing, these are tunneled to the generator pocket. This approach requires a subcostal, subxiphoid, thoracotomy, or sternotomy incision. Advantages of epicardial implantation include ability for placement independent of intracardiac pathology and avoidance of considerations regarding venous thrombosis. Disadvantages include the invasiveness of the approach, higher incidence of lead failure, and early generator battery depletion.

Need for pacing, programmed settings, and device capabilities play a significant role in the longevity of pacemaker generators. These units are powered by lithium–iodide batteries with an expected service life of between 5 and 15 years.
Hardware selection and programming of devices

A variety of hardware options are available for cardiac pacing in infants and children. The selection of the particular generator system, mode for pacing, and pacing leads is dependent upon a number of factors. In general terms considerations include patient size, indications for pacing, requirements for specific programmability options, cardiac status/underlying cardiac pathology, and anticipated need for generator longevity.

Both single- and dual-chamber units are commercially available for permanent pacing in the pediatric age group. The dual-chamber devices provide the benefit of AV synchrony and optimization of hemodynamics. The modes frequently used in single-chamber pacing are AAI or AAIR, VVI or VVIR, depending on whether atrial or ventricular leads are present. In the case of dual-chamber pacemakers, typical modes are DDD or DDDR. Under specific circumstances such as during surgical interventions, the pacing mode may be modified to asynchronous (AOO, VOO, or DOO) to prevent erratic pacemaker behavior and allow for an increased margin of safety. If active, the rate-responsiveness feature of the unit should be disabled prior to anesthetic induction.

Pacemaker malfunction

The problems most frequently accounting for pacemaker malfunction include complications related to lead placement and function, failure to pace, failure to capture, under or oversensing, phrenic nerve stimulation, and pacemaker-mediated tachycardia. Pacemaker troubleshooting may require a combination of chest radiograph, 12-lead ECG, rhythm strip, and device interrogation to determine pacing and sensing thresholds, lead impedances, battery status, and magnet rate.

Children are considered to be at higher risk for lead failure and fracture than their adult counterparts. These problems result in inappropriate pacemaker sensing or capture (underpacing or overpacing) and potential need for pacemaker revision.

With a single-chamber unit failure to pace implies no pacing artifacts or spikes on the ECG. A malfunctioning dual-chamber pacemaker may exhibit either no pacing deflections or pacing in only one chamber. The inability to pace may be intermittent or continuous. Failure to capture implies pacemaker stimuli without associated cardiac chamber response. Increasing the energy output of the pacing device may alleviate this type of malfunction. In the acute postoperative setting this problem requires immediate attention. Normally pacing thresholds increase with time emphasizing the fact that frequent testing of pacemaker function should be performed in the postoperative patient with temporary pacing wires. Steroid therapy may limit edema, inflammation and fibrosis that accounts for increases in pacing thresholds.

Undersensing (inability to sense) leads to intercalation of pacing stimuli during normal atrial (P waves) or ventricular (QRS complexes) activity. Oversensing, or failure of the pacing stimulus to be delivered at the pre-programmed time, results in pacemaker pauses. Diaphragmatic contraction related to pacing stimulation can be avoided by reducing the pacemaker output to a lower value, or adjusting pulse amplitude and duration. Pacemaker mediated tachycardia is an infrequent problem in the pediatric age group. Adjustments in pacemaker settings may remedy this potential issue.

Perioperative considerations

Device interrogation should be part of the complete preoperative evaluation in all patients with implanted pacemakers scheduled for surgical interventions (cardiac or non-cardiac). Consultation with a pediatric cardiologist/electrophysiologist to obtain details of unit type, settings, date of and indications for implantation, and underlying rhythm is highly recommended. Results of a recent 12-lead ECG should be reviewed. Reprogramming may be required prior to the planned procedure to avoid potential problems with pacemaker malfunction related to electromagnetic interference (electrocautery). Unipolar electrocautery may interfere with pacemaker function, thus bipolar electrocautery is preferred. Chronotropic agents and backup pacing modalities (transvenous, epicardial, transcatheter) should be readily available and carefully considered in the event of pacemaker malfunction and inadequate underlying rate. Capture thresholds can be affected by pharmacological agents (i.e. amiodarone can increase thresholds) and this should be considered if pacing is required in the patient receiving antiarrhythmic drug therapy. A magnet should also be accessible to allow for asynchronous pacing if required. Most generators respond to generator magnet application by pacing at a fixed rate asynchronously (AOO, VOO, or DOO). A potential concern is that the specific magnet rate, as determined by the manufacturer for the particular device, may not be in agreement with the desired or optimal pacing rate. Thus the use of a magnet should not be considered a substitute for preoperative pacemaker interrogation/programming. At pacemaker generator end of life the pacing rate upon magnet application to the generator may differ (slower) than the pre-specified magnet rate. In addition to perioperative ECG monitoring, additional modalities that confirm pulse generation during pacing (esophageal stethoscope for assessment of heart sounds, pulse oximetry, invasive arterial blood pressure monitoring) should be strongly considered. After completion of the procedure the device should be tested and reprogrammed.

Temporary cardiac pacing

The transvenous and epicardial routes are commonly used for temporary pacing, although the transthoracic
(transcutaneous) and transesophageal approaches are also suitable in some circumstances. Indications for temporary cardiac pacing are not clearly defined as in the case of permanent pacing. Temporary pacing is provided for during most cardiothoracic procedures by placement of wires in the atrial and/or ventricular epimyocardium near the completion of the intervention. Basic settings to be adjusted in the external temporary pulse generator (single or dual chamber device) include: (i) pacing rate; (ii) atrial and/or ventricular output amplitude (milliamperes); (iii) atrial and/or ventricular sensitivity (millivolts) or asynchronous mode; and (iv) A–V interval (milliseconds). Temporary pacing may be necessary for maintenance of adequate cardiac output in the context of bradydysrhythmias, abnormal AV conduction, AV asynchrony, and heart rates inadequate for physiologic state.80 Temporary pacing may also be helpful in individuals at risk of high degree AV block and can be used to suppress, overdrive, or terminate tachydysrhythmias. Atrial recordings (atrial electrograms) obtained through temporary pacing wires may provide diagnostic information in certain types of rhythm disorders. Temporary pacing is discontinued with resolution of the indication for pacing or transitioned to permanent pacing. In the care of patients who depend on temporary pacing for maintenance of hemodynamics, it is extremely important to be attentive to pacemaker settings, capture thresholds, and prepare to provide alternate means of pacing in the event of lead/pacemaker failure or malfunction.

External transcutaneous pacing

A transcutaneous external pacing unit with features superior to earlier transcutaneous systems was patented and introduced by Dr Paul Zoll in the early 1980s. This lead to renewed interest in the field and enhancements of this technology. Several of the devices currently available for commercial use combine defibrillation/cardioversion capabilities and external pacing features. In pediatric patients emergency transthoracic pacing may be considered as a temporizing measure in those with symptomatic bradycardia (secondary to abnormal sinus node function or to complete AV block).81 It is important to understand that transcutaneous pacing allows for simultaneous atrial and ventricular activation, thus optimal hemodynamics may not be feasible. Transcutaneous pacing has not been found to be effective in the treatment of asystole in children. PACing Electrode size should be selected according to patient size (usually patients under 15 kg require smaller adhesive pads). Device settings typically include heart rate and current output (milliamperes). Most current models provide the option for fixed rate (asynchronous) and demand (synchronous) pacing. After selection of a desired heart rate and pacing modality the current is increased as tolerated until capture is achieved. If the patient is not anesthetized, sedation may be necessary to improve tolerance to transcutaneous pacing. Prolonged periods of transcutaneous pacing may result in serious burns or skin trauma in infants and young children. In addition to monitoring for pacemaker capture by ECG, ongoing clinical assessment of the adequacy of cardiac output should be undertaken.

Esophageal overdrive pacing

A transesophageal catheter may be used for atrial sensing allowing for diagnostic information and discrimination of supraventricular tachydysrhythmias. The esophageal route also provides a minimally invasive approach for overdrive pacing a variety of supraventricular rhythm disorders (atrial flutter, SVT). For this purpose an electrode catheter is placed into the esophagus, advanced to a location that corresponds roughly to the region behind the atrial mass and an atrial electrogram is obtained to refine the catheter position. Local anesthesia to the nasopharynx or oropharynx and/or sedation is generally required in order to introduce the catheter and to prevent discomfort during atrial pacing. Standard cardiorespiratory monitoring should be undertaken during the procedure, in addition to airway support as necessary. Emergency drugs and cardioversion/defibrillation equipment should be readily available.

Implantable cardioverter–defibrillators

The primary goal of the internal cardioverter–defibrillator is the reduction of sudden death in patients at high risk. Although sudden cardiac death is an uncommon occurrence in pediatrics, certain patients groups may have a definitive risk deriving potential benefits from these devices. Individuals with arrhythmogenic right ventricular dysplasia (a specific type of cardiomyopathy), long QT syndrome, hypertrophic cardiomyopathy, and those with a history of near death events may be considered suitable candidates for pacemaker/defibrillator implantation. Additional patients are those with operated CHD and a history of malignant arrhythmias. At the present, the experience in the pediatric age group with these devices has been limited and reported mostly in retrospective fashion.82–84 Prospective trials are required to establish guidelines for use, address safety concerns, and evaluate long-term issues specific to children. The anesthetic considerations in patients with implanted units relate primarily to potential surgical electromagnetic interference (electrocautery). Perioperative consultation with a cardiologist/electrophysiologist is therefore essential in the care of these patients. In many cases the devices may need to be adjusted or deactivated prior to surgery. Careful evaluation and device reprogramming is advisable at the conclusion of the surgical intervention.
References

PART 4 Management