CHAPTER 1

Indications for permanent and temporary cardiac pacing

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Anatomy

To understand the principles and concepts involved in cardiac pacing more completely, a brief review of the anatomy and physiology of the specialized conduction system is warranted (Fig. 1.1, Table 1.1).

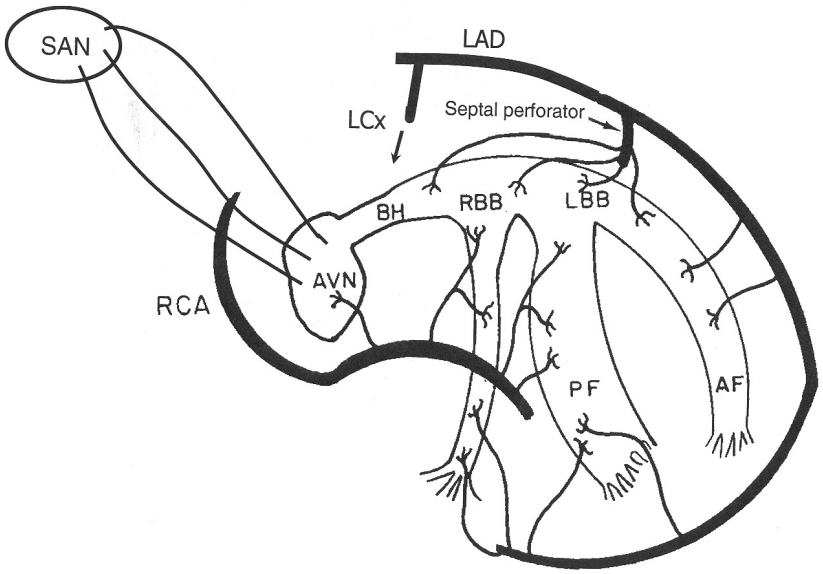

Fig. 1.1 A schematic diagram of the conduction system (SAN, sinus node; AVN, AV node; BH, bundle of His; LBB, left bundle branch; RBB, right bundle branch; AF, left anterior superior fascicle; PF, posterior inferior fascicle of the left bundle branch). The arterial supply is shown (RCA, right coronary artery; LAD, left anterior descending coronary artery; LCx, left circumflex coronary artery). For further description, see text.

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Table 1.1 The specialized conduction system

Structure	*Location*	*Histology*	*Arterial blood supply*	*Autonomic innervation*	*Physiology*
SA node	Subepicardial; junction of SVC and HRA	Abundant P cells	SA nodal artery from RCA 55% or LCx 45%	Abundant	Normal impulse generator
AV node	Subendocardial; interatrial septum	Fewer P cells, Purkinje cells, "working" myocardial cells	AV nodal artery from RCA 90%, LCx 10%	Abundant	Delays impulse, subsidiary pacemaker
His bundle	Membranous septum	Narrow tubular structure of Purkinje fibers in longitudinal compartments; few P cells	AV nodal artery, branches of LAD	Sparse	Conducts impulses from AV node to bundle branches
Bundle branches	Starts in muscular septum and branches out into ventricles	Purkinje fibers; highly variable anatomy	Branches of LAD, RCA	Sparse	Activates ventricles

AV, atrioventricular; HRA, high right atrium; LAD, left anterior descending coronary artery; LCx, left circumflex coronary artery; RCA, right coronary artery; SA, sinoatrial; SVC, superior vena cava.

Sinoatrial node

The sinoatrial (SA) node is a subepicardial structure located at the junction of the right atrium and superior vena cava. It has abundant autonomic innervation and a copious blood supply; it is often located within the adventitia of the large SA nodal artery, a proximal branch of the right coronary artery (55%), or the left circumflex coronary artery. Histologically, the SA node consists of a dense framework of collagen that contains a variety of cells, among them the large, centrally located P cells, which are thought to contain the principal pacemaker cells which initiate impulses and spontaneous electrical activity; transitional cells, intermediate in structure between P cells and regular atrial myocardial cells; and Purkinje-like fiber tracts, extending through the perinodal area and into the atrium. Once the impulse exits the sinus node and the perinodal tissue it traverses the atrium to the atrioventricular (AV) node.

Atrioventricular node

The AV node is a small subendocardial structure within the interatrial septum located at the convergence of the specialized conduction tracts that course through the atria. Like the SA node, the AV node has extensive autonomic innervation and an abundant blood supply from the large AV nodal artery, a branch of the right coronary artery, in 90% of patients, and from the left curcumflex artery in 10%. Histological examination of the AV node reveals a variety of cells embedded in a loose collagenous network including P cells (although not nearly as many as in the SA node), atrial transitional cells, ordinary myocardial cells and Purkinje cells.

His bundle

Purkinje fibers emerging from the area of the distal AV node converge gradually to form the His bundle, a narrow tubular structure that runs through the membranous septum to the crest of the muscular septum, where it divides into the bundle branches. The His bundle has relatively sparse autonomic innervation, although its blood supply is quite ample, emanating from both the AV nodal artery and septal branches of the left anterior descending artery. Longitudinal strands of Purkinje fibers, divided into separate parallel compartments by a collagenous skeleton, can be discerned by histological examination of the His bundle. Relatively sparse P cells can also be identified, embedded within the collagen. The rapid conduction of electrical impulses across the His–Purkinje system is responsible for the almost simultaneous activation of the right and left ventricles.

Bundle branches

The bundle branch system is an enormously complex network of interlacing Purkinje fibers that varies greatly among individuals. It generally starts as one or more large fiber bands that split and fan out across the ventricles until they finally terminate in a Purkinje network that interfaces with the myocardium. In some cases, the bundle branches clearly conform to a trifascicular or

quadrifascicular system. In other cases, however, detailed dissection of the conduction system has failed to delineate separate fascicles. The right bundle is usually a single, discrete structure that extends down the right side of the interventricular septum to the base of the anterior papillary muscle, where it divides into three or more branches. The left bundle more commonly originates as a very broad band of interlacing fibers that spread out over the left ventricle, sometimes in two or three distinct fiber tracts. There is relatively little autonomic innervation of the bundle branch system, but the blood supply is extensive, with most areas receiving branches from both the right and left coronary systems.

Physiology

The SA node has the highest rate of spontaneous depolarization (automaticity) in the specialized conduction system and, under ordinary circumstances, it is the major generator of cardiac impulses. Its unique location astride the large SA nodal artery provides an ideal milieu for continuous monitoring and instantaneous adjustment of heart rate to meet the body's changing metabolic needs. The SA node is connected to the AV node by several specialized fiber tracts, the function of which has not been fully elucidated. The AV node appears to have three major functions: it delays the passing impulse for approximately 0.04 s under normal circumstances, permitting complete atrial emptying with appropriate loading of the ventricles; it serves as a subsidiary impulse generator, as its concentration of P cells is second only to that of the SA node; and it acts as a type of filter, limiting ventricular rates in the event of an atrial tachyarrhythmia.

The His bundle arises from the convergence of Purkinje fibers from the AV node, although the exact point at which the AV node ends and the His bundle begins has not been delineated either anatomically or electrically. The separation of the His bundle into longitudinally distinct compartments by the collagenous framework allows for longitudinal dissociation of electrical impulses. Thus, a localized lesion below the bifurcation of the His bundle (into the bundle branches) may cause a specific conduction defect (e.g. left anterior fascicular block). The bundle branches arise as a direct continuation of the His bundle fibers. Disease within any aspect of the His bundle branch system may cause conduction defects that can affect AV synchrony or prevent synchronous right and left ventricular (LV) activation. The accompanying hemodynamic consequences have considerable clinical relevance. These consequences have provided the impetus for some of the advances in pacemaker technology that will be addressed in later chapters.

Although a detailed discussion of the histopathology of the conduction system is beyond the scope of the present chapter, it is worth noting that conduction system disease is often diffuse. For example, normal AV conduction cannot necessarily be assumed when a pacemaker is implanted for a disorder seemingly localized to the sinus node. Similarly, normal sinus node func-

tion cannot be assumed when a pacemaker is implanted in a patient with AV block.

Indications for permanent pacemakers

The decision to implant a permanent pacemaker is an important one and should be based on solid clinical evidence. A joint committee of the American College of Cardiology (ACC) and the American Heart Association (AHA) was formed in the 1980s to provide uniform criteria for pacemaker implantation. These guidelines were first published in 1984 and most recently revised in 2002.[1] It must be realized, however, that medicine is a constantly changing science, and absolute and relative indications for permanent pacing may change as a result of advances in the diagnosis and treatment of arrhythmias. Accordingly, it should be noted that the joint committee is again revising its recommendations for pacemaker implantation and the new guidelines should be available in the near future. It is useful to keep the ACC/AHA guidelines in mind when evaluating a patient for pacemaker implantation. When approaching a patient with a documented or suspected bradyarrhythmia, it is important to take the clinical setting into account. Thus, the patient's overall general medical condition must be considered as well as their occupation or desire to operate a motor vehicle or equipment, where the safety of other individuals may be at risk.

In the ACC/AHA classification, there are three classes of indications for permanent pacemaker implantation, defined as follows:

Class I
Conditions for which there is evidence and/or general agreement that a pacemaker implantation is beneficial, useful, and effective.

Class II
Conditions for which there is conflicting evidence and/or a divergence of opinion about the usefulness/efficacy of pacemaker implantation.

Class IIa
Weight of evidence/opinion in favor of efficacy.

Class IIb
Usefulness/efficacy less well established by evidence/opinion.

Class III
Conditions for which there is evidence and/or general agreement that a pacemaker is not useful/effective and in some cases may be harmful.

Level of evidence

Additionally, the ACC/AHA Committee ranked evidence supporting their recommendations by the following criteria.

• Level A: Data derived from multiple randomized trials involving a large number of patients.
• Level B: Data derived from a limited number of trials involving a relatively small number of patients or from well-designed analyses of non-randomized studies or data registries.
• Level C: Recommendations derived from the consensus of experts.

Acquired atrioventricular block

Acquired AV block with syncope (e.g. Stokes–Adams attacks) was historically the first indication for cardiac pacing. The site of AV block (e.g. AV node, His bundle, or distal conduction system) will to a great extent determine the adequacy and reliability of the underlying escape rhythm (Figs 1.2–1.4). It is worth noting that, in the presence of symptoms documented to be due to AV block, permanent pacing is indicated, regardless of the site of the block (e.g. above the His bundle as well as below the His bundle). Because of different indications for permanent pacing of heart block due to acute myocardial infarction (MI), congenital AV block and increased vagal tone, these indications are discussed in other sections.

The indications for permanent pacing with AV block follow.

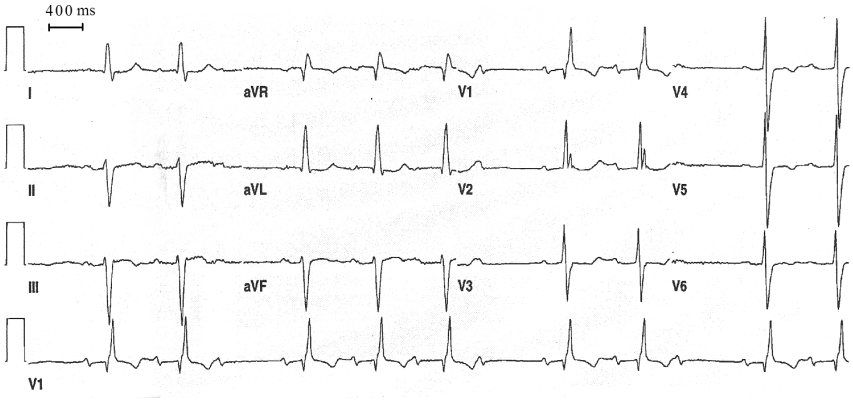

Fig. 1.2 A 70-year-old man with known right bundle branch block, left anterior fascicular block and first-degree atrioventricular (AV) block was seen in the emergency department with a complaint of recurrent syncope. Electrocardiogram revealed type I second-degree AV block. On electrophysiological study, the site of block was found to be infranodal and he was referred for a permanent pacemaker. It should be noted that approximately 65% of patients who develop complete heart block with a wide QRS escape complex have antecedent right bundle branch block with left anterior fascicular block. In this situation, even type I second-degree AV block is frequently infranodal and warrants permanent pacemaker implantation, especially when there is a history of syncope, since idioventricular escape rhythms are notoriously unreliable.

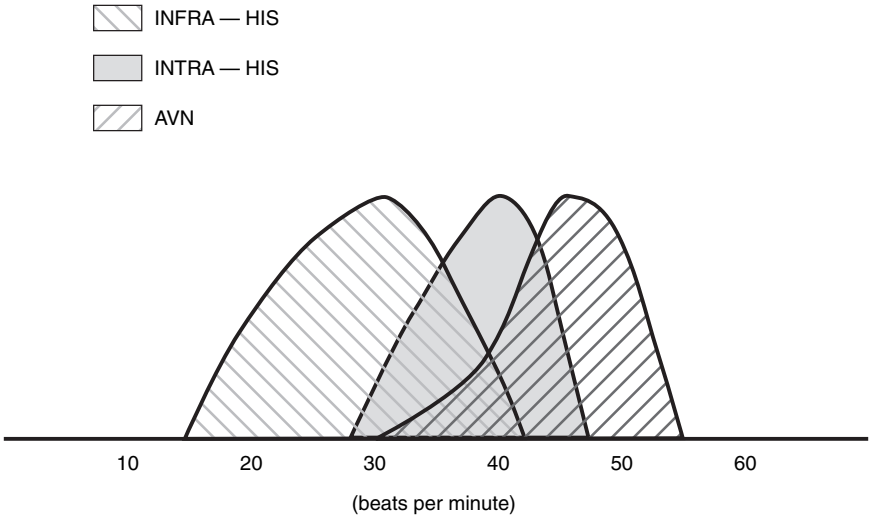

Fig. 1.3 A diagram outlining the rate of the escape rhythm in patients with high-grade atrioventricular (AV) block. As can be seen, the escape rate in a patient with block at the AV node (AVN) is usually considerably faster than in individuals with intra-Hisian or infra-Hisian block, although there is considerable overlap between groups.

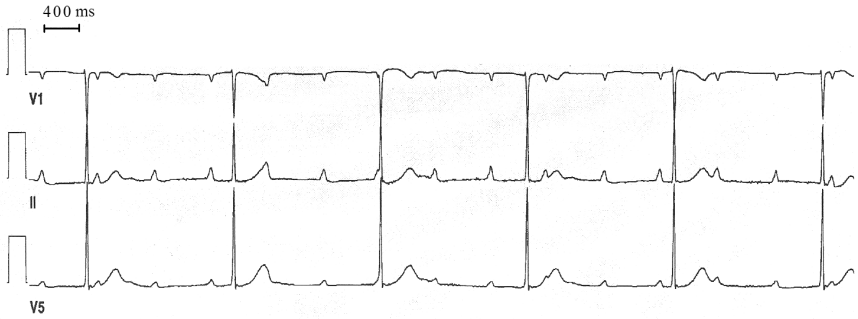

Fig. 1.4 A 70-year-old man was admitted to the hospital complaining of weakness and presyncopal episodes. Rhythm strips revealed complete atrioventricular block and a slow junctional escape rhythm with narrow QRS complexes. He received a permanent dual-chamber pacemaker, which completely relieved his symptoms.

Class I

1 Third-degree and advanced second-degree AV block at any anatomical level, associated with any one of the following conditions:

a Bradycardia with symptoms (including heart failure) presumed to be due to AV block. (Level of evidence: C.)

b Arrhythmias and other medical conditions requiring drugs that result in symptomatic bradycardia. (Level of evidence: C.)
c Documented periods of asystole ≥3.0 s or any escape rate <40 bpm in awake, symptom-free patients. (Levels of evidence: B, C.)
d After catheter ablation of the AV junction. (Levels of evidence: B, C.) There are no trials to assess outcome without pacing, and pacing is virtually always planned in this situation unless the operative procedure is AV junction modification.
e Postoperative AV block that is not expected to resolve after cardiac surgery. (Level of evidence: C.)
f Neuromuscular diseases with AV block, such as myotonic muscular dystrophy, Kearns–Sayre syndrome, Erb's dystrophy and peroneal muscular atrophy, with or without symptoms, because there may be unpredictable progression of AV conduction disease. (Level of evidence: B.)

2 Second-degree AV block regardless of type or site of block, with associated symptomatic bradycardia. (Level of evidence: B.)

Class IIa

1 Asymptomatic third-degree AV block at any anatomical site with average awake ventricular rates of ≥40 bpm, especially if cardiomegaly, ventricular arrhythmias or LV dysfunction are present. (Levels of evidence: B, C.)
2 Asymptomatic type II second-degree AV block with a narrow QRS. When type II second-degree AV block occurs with a wide QRS, pacing becomes a class I recommendation. (Level of evidence: B.)
3 Asymptomatic type I second-degree AV block at intra- or infra-His levels found at electrophysiology study performed for other indications. (Level of evidence: B.)
4 First- or second-degree AV block with symptoms similar to those of pacemaker syndrome. (Level of evidence: B.)

Class IIb

1 Marked first-degree AV block (>0.30 s) in patients with LV dysfunction and symptoms of congestive heart failure in whom a shorter AV interval results in hemodynamic improvement, presumably by decreasing left atrial filling pressure. (Level of evidence: C.)
2 Neuromuscular diseases such as myotonic muscular dystrophy, Kearns–Sayre syndrome, Erb's dystrophy, and peroneal muscular atrophy with any degree of AV block (including first-degree AV block), with or without symptoms, because there may be unpredictable progression of AV conduction disease. (Level of evidence: B.)

Class III

1 Asymptomatic first-degree AV block. (Level of evidence: B.)
2 Asymptomatic type I second-degree AV block at the AV nodal level or not known to be intra- or infra-Hisian. (Levels of evidence: B, C.)

3 AV block expected to resolve and/or unlikely to recur (e.g. drug toxicity, Lyme disease, or during hypoxia in sleep apnea syndrome in the absence of symptoms). (Level of evidence: B.)

The majority of these diagnoses can be made from the surface electrocardiogram (ECG). Invasive electrophysiology studies are only rarely necessary, but may be helpful or of interest in elucidating the site of AV block (Figs 1.5–1.7). Regarding the first two items in class II, it is likely that permanent pacemakers are more frequently implanted in patients with wide QRS complexes and/or documented infranodal block than in patients with narrow QRS complex escape rhythms.

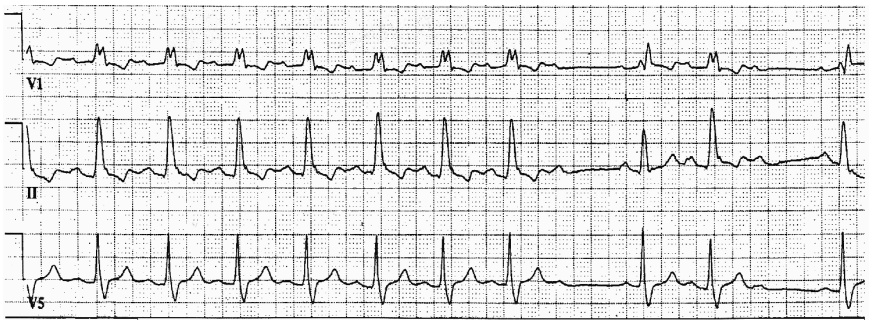

Fig. 1.5 A rhythm strip recorded from a patient with recurrent syncope showing right bundle branch block, left posterior fascicular block and type II second-degree atrioventricular (AV) block. Type II second-degree AV block is almost always infranodal (this was documented by intracardiac recordings). Symptomatic second-degree AV block is a class I indication for permanent pacing.

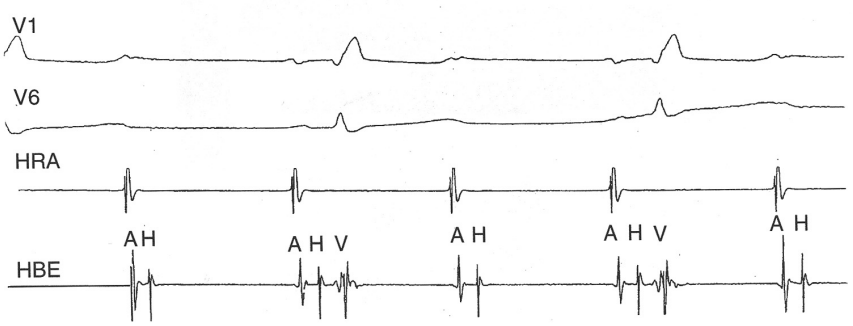

Fig. 1.6 An example of 2 : 1 atrioventricular (AV) block with the level of block occurring within the His–Purkinje system. In the presence of a narrow QRS complex, 2 : 1 AV block is usually situated at the AV node, whereas a wide QRS complex in the conducted beats often indicates infranodal block. Note that every other P wave is blocked below the His bundle. The paper speed is 100 mm s^{-1}. From top to bottom: V_1 and V_6 are standard ECG leads; HRA is the high right atrial recording and HBE is the intracardiac recording of the His bundle electrogram. A, atrial electrogram; H, His bundle electrogram; V, ventricular electrogram.

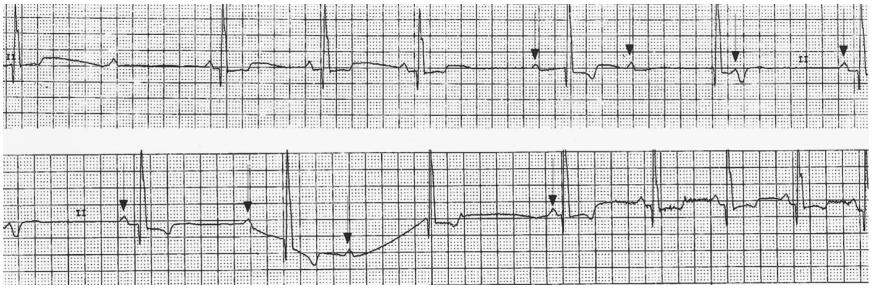

Fig. 1.7 An example of "vagotonic" block. P waves are indicated by the arrows. The simultaneous occurrence of atrioventricular (AV) block and slowing of the sinus rate is diagnostic of hypervagotonia. This type of block is located at the level of the AV node. It is generally considered benign and does not warrant a permanent pacemaker unless the patient is very symptomatic with medically refractory recurrences.

It is worth emphasizing that 2:1 AV block may be either type I or type II, but this cannot always be discerned from the surface ECG (Table 1.2). As a rough approximation, if the QRS complex is narrow, the block is probably localized to the AV node and considered type I. If the QRS complex is wide, the level of block may be in the AV node or His bundle, and the site of the block can best be determined from an invasive electrophysiological study (His bundle recording). The causes of acquired high-grade AV block are listed in Table 1.3.

Table 1.2 Differential diagnosis of 2:1 atrioventricular (AV) block

Condition	*Block above AV node*	*Block below AV node*
Exercise	+	+/– or –
Atropine	+	+/– or –
Carotid sinus massage	–	+ or +/–
Isoprenaline	–	+ or +/–

+Represents improved AV conduction, –represents worsened AV conduction.

Table 1.3 Causes of acquired high-grade atrioventricular (AV) block

Ischemic
- Acute myocardial infarction
- Chronic ischemic heart disease
- Prinzmetal's angina

Non-ischemic cardiomyopathy
- Hypertensive
- Idiopathic dilated

Fibrodegenerative
- Lev's disease
- Lenègre's disease

Table 1.3 (*Continued*.)

After cardiac surgery/cardiac catheterization laboratory
Coronary artery bypass grafting
Aortic valve replacement or aortic root replacement
Ventricular septal defect repair
Septal myomectomy or ethanol ablation of the interventricular septum
Other iatrogenic
After His bundle (AV junction) ablation
After ablation of septal accessory pathways, AV nodal re-entry
After radiation therapy (e.g. lung cancer, Hodgkin's lymphoma)
Infectious disease
Bacterial endocarditis
Chagas' disease
Lyme disease
Syphilis
Myocarditis
Rheumatic fever
Other (viral, rickettsial, fungal, etc.)
Neuromuscular disease
Myotonic dystrophy
Muscular dystrophies (fascioscapulohumeral)
Kearns–Sayre syndrome
Friedreich's ataxia
Infiltrative disease
Amyloid
Sarcoid
Hemochromatosis
Carcinoid
Malignant tumors
Connective tissue disease
Rheumatoid arthritis
Systemic lupus erythematosus
Systemic scleroderma
Ankylosing spondylitis
Cardiac tumors
Drug induced
Digitalis
β-Blockers
Calcium channel blockers
Lithium
Class I or III antiarrhythmic drugs
Congenital
Congenital heart disease
Maternal systemic lupus erythematosus

Chronic bifascicular or trifascicular block

Patients with chronic bifascicular block [right bundle branch block (RBBB) and left anterior hemiblock, RBBB and left posterior hemiblock, or complete left bundle branch block (LBBB)] and patients with trifascicular block (any of the above and first-degree AV block) are at an increased risk of progression to complete AV block.

In the 1980s, the results of several large prospective studies of the role of His bundle recordings in asymptomatic patients with chronic bifascicular block were published. In the combined analysis of these studies, more than 750 patients were observed for 3–5 years. The incidence of progression from bifascicular to complete heart block was low, varying from 2% to 5% per year. Most important, the total cardiovascular mortality was 19–25%, and the mortality from sudden cardiac death was 10–20%. In these patients, the presence of bifascicular block on the ECG can be taken as a sign that there is a high likelihood of coexisting structural heart disease. We can conclude from these studies that patients with chronic asymptomatic bifascicular block and a prolonged HV interval (HV interval represents the shortest conduction time from the His bundle to the ventricular endocardium over the specialized conduction system) have more extensive organic heart disease and an increased risk of sudden cardiac death. The risk of spontaneous progression to complete heart block is small, although it is probably slightly greater in patients who have a prolonged HV interval. Permanent pacing appears to prevent recurrent syncope in these patients, but does not reduce the incidence of sudden death, which may often be due to heart failure or ventricular arrhythmias. Routine His bundle recordings are therefore of little value in evaluating patients with chronic bifascicular block and no associated symptoms (e.g. no syncope or presyncope) (Fig. 1.8).

In patients with bifascicular or trifascicular block and associated symptoms of syncope or presyncope, electrophysiological testing is useful. A high incidence of sudden cardiac death and inducible ventricular arrhythmias is noted in this patient group. Electrophysiological testing may be helpful for identifying the disorder responsible for syncope and potentially avoiding implantation of a pacemaker (Fig. 1.9). In patients who have a markedly prolonged HV interval (>100 ms) and syncope not attributable to other causes, there is a high incidence of subsequent development of complete heart block, and permanent pacing is warranted. However, these patients comprise a relatively small percentage of patients undergoing electrophysiological testing with cardiac symptoms and bifascicular block. In the majority of patients, the HV interval is normal (HV 35–60 ms) or only mildly prolonged, and His bundle recording does not effectively separate out high-risk and low-risk subpopulations with bifascicular block who are likely to progress to complete heart block. Electrophysiological testing will often provoke sustained ventricular arrhythmias, which are the cause of syncope in many of these patients. In patients with LV systolic dysfunction, advanced heart failure and bundle branch block, espe-

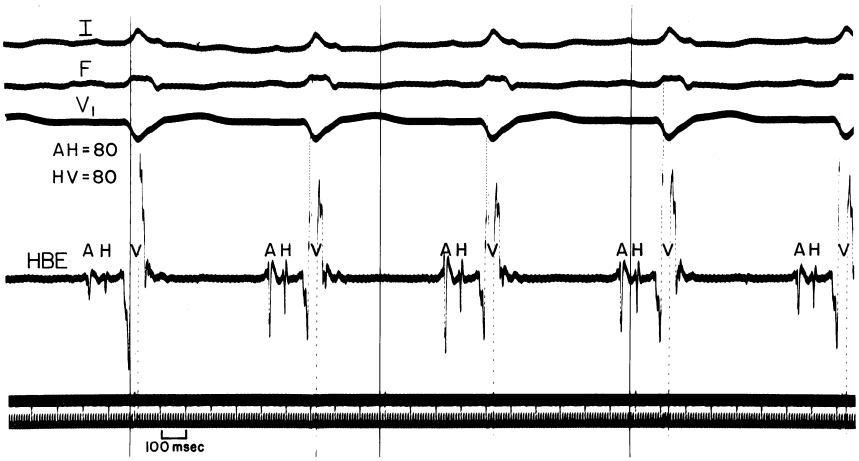

Fig. 1.8 An intracardiac recording in a patient with left bundle branch block. The prolonged HV interval (80 ms) is indicative of infranodal conduction disease, but in the absence of transient neurological symptoms (syncope, dizzy spells, etc.), no specific therapy is indicated. From top to bottom: I, F and V_1 are standard ECG leads; HBE is the intracardiac recording of the His bundle electrogram. A, atrial depolarization; H, His bundle depolarization; V, ventricular electrogram. Paper speed is 100 mm s^{-1}.

cially LBBB, and a QRS interval > 120–130 ms, defibrillators with biventricular pacing have been shown to improve symptoms from heart failure and reduce mortality.[2]

Barold has pointed out that the standard definition of trifascicular block is often too loosely applied.[3] Thus, in patients with RBBB and either left anterior or left posterior fascicular block or in patients with LBBB and first-degree AV block, the site of block could be located either in the His–Purkinje system or in the AV node. The term "trifascicular block" should be reserved for alternating RBBB and LBBB or for block of either bundle in the setting of a prolonged HV interval.

The indications for pacing in the setting of chronic bifascicular/trifascicular block are listed below.

Class I

1 Intermittent third-degree AV block. (Level of evidence: B.)
2 Type II second-degree AV block. (Level of evidence: B.)
3 Alternating bundle branch block. (Level of evidence: C.)

Class IIa

1 Syncope not demonstrated to be due to AV block when other likely causes have been excluded, specifically ventricular tachycardia (VT). (Level of evidence: B.)

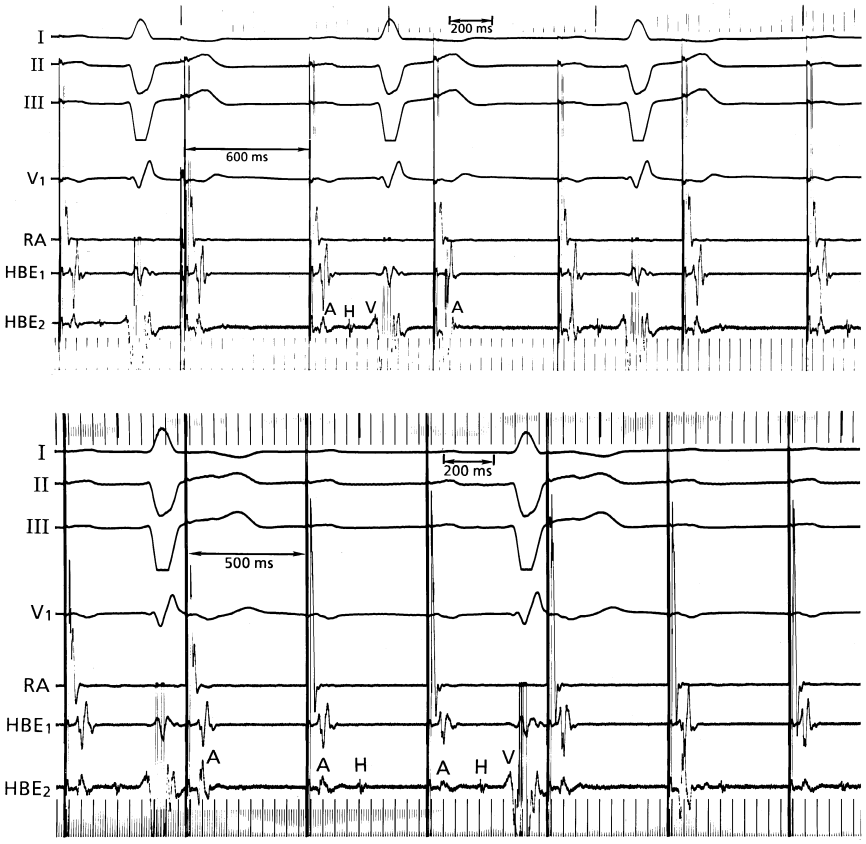

Fig. 1.9 A 68-year-old man was admitted complaining of recurrent dizziness and syncope. His baseline 12-lead ECG showed a PR interval of 0.20 s and a right bundle block QRS morphology. During the electrophysiological study, the patient's baseline HV interval was 90 ms. Top: During atrial pacing at a cycle length of 600 ms (100 ppm), there is block in the atrioventricular node. Bottom: During pacing at 500 ms (120 ppm), there is block below the His bundle. These findings are indicative of severe diffuse conduction system disease. A permanent dual-chamber pacemaker was implanted, and the patient's symptoms resolved. From top to bottom: I, II, III and V_1 are standard ECG leads; intracardiac recording from the right atrial appendage (RA) and His bundle (HBE_1 for the proximal His bundle and HBE_2 for the distal His bundle). A, atrial depolarization; H, His bundle depolarization; V, ventricular depolarization.

2 Incidental finding at electrophysiology study of markedly prolonged HV interval (≥100 ms) in asymptomatic patients. (Level of evidence: B.)
3 Incidental finding at electrophysiology study of pacing-induced infra-His block that is not physiological. (Level of evidence: B.)

Class IIb

1 Neuromuscular diseases such as myotonic muscular dystrophy, Kearns–Sayre syndrome, Erb's dystrophy and peroneal muscular atrophy with any

degree of fascicular block with or without symptoms, because there may be unpredictable progression of AV conduction disease. (Level of evidence: C.) Class IIa indication in European guidelines.

Class III

1 Fascicular block without AV block or symptoms. (Level of evidence: B.)
2 Fascicular block with first-degree AV block without symptoms.

Permanent pacing for chronic neuromuscular disorders

The frequent involvement of the cardiac conduction system in chronic neuromuscular disorders should not be surprising, considering the many similarities between cardiac and skeletal muscle. In some of these conditions, cardiac disease may be responsible for greater morbidity and mortality than the neuromuscular manifestations, highlighting the importance of evaluating the extent of cardiac involvement. Most often, bradyarrhythmias in neuromuscular disorders are due to direct involvement of the specialized AV conduction system, with symptomatic sinus node dysfunction being unusual. The relatively small numbers of patients involved and the absence of randomized, placebo-controlled clinical trials make it difficult to provide definitive guidelines for pacemaker implantation. In general, the recommendations for permanent pacing in these patients are similar to those in other groups with conduction system disease. However, since mortality and the incidence of sudden cardiac death are high in this group of disorders, and because conduction system disease tends to be unpredictable, the development of second- or third-degree AV block, even in the absence of symptoms, is considered a class I indication for permanent pacing. In addition, suggestive symptoms such as syncope should be promptly and aggressively investigated. Some authorities recommend yearly ECGs and 24-h ambulatory recordings for patients with one of these disorders to facilitate early recognition of AV block. It should also be realized, however, that life-threatening ventricular arrhythmias are also fairly common in this population, especially when LV function is impaired or complicated by hypertrophic cardiomyopathy, so use of a permanent pacemaker will not necessarily prevent sudden cardiac death. There is almost no definitive information available on determining whether pacemakers or defibrillators should be implanted in this population. The neuromuscular disorders most frequently associated with symptomatic conduction system disease are as follows:

1 Duchenne's muscular dystrophy—A progressive X-linked disease that usually becomes clinically apparent in the mid-teens and is fatal by the end of the third decade. The ECG typically shows prominent R waves in V_1 with deep narrow Q waves in the lateral precordial leads. Although cardiac involvement is almost universal, the incidence of arrhythmias is variable, with many patients dying from heart failure. In the absence of definitive data, it seems prudent to recommend permanent pacemaker implantation in patients who

develop second-degree or higher degrees of AV block, especially in the setting of a wide QRS complex.

2 Becker muscular dystrophy—An X-linked condition closely related to Duchenne's muscular dystrophy. It has similar electrocardiographic abnormalities, but progresses more slowly. The severity of cardiac involvement does not parallel the severity of neuromuscular disease. Although there is less experience with this disorder, the indications for permanent pacing appear similar to Duchenne's muscular dystrophy.

3 Myotonic muscular dystrophy—An autosomal dominant disorder that usually becomes clinically manifest in the third decade. Cardiac involvement is common and is mostly confined to the conduction system. Most adults have electrocardiographic abnormalities. Both bradyarrhythmias and tachycardias are common, and suggestive symptoms should be promptly evaluated. Permanent pacemakers are warranted for second- or third-degree AV block, even in the absence of symptoms. A recent large prospective study has suggested that HV $\geq$70 ms identifies a subgroup likely to benefit from prophylactic (no documented high-degree block) permanent pacing.[4]

4 Emery–Dreyfuss muscular dystrophy—A slowly progressive X-linked muscular dystrophy with a high incidence of conduction system disease and arrhythmias. Sudden cardiac death due to bradyarrhythmias has been well documented, and permanent pacemakers are often necessary.

5 Limb girdle muscular dystrophy—A heterogeneous group of disorders that usually begin with weakness in the upper legs and pelvic musculature. Cardiac involvement is variable, although there is a familial form with a high incidence of conduction disease. Patients with a family history of heart block or sudden death should be considered for permanent pacing relatively early in the course of their disease.

6 Kearns–Sayre syndrome—A multisystem mitochondrial disorder characterized by progressive external ophthalmoplegia, pigmentary retinal degeneration and AV block. Involvement of the distal conduction system is the rule and high-degree AV block is common. Although definitive data are lacking, it seems prudent to implant a permanent pacemaker prophylactically when marked first-degree AV block is appreciated.

Infiltrative disorders

The infiltrative disorders are a diverse group of conditions characterized by infiltration of the myocardium by a tissue or substance. These include hematological malignancies (leukemia, lymphoma, myeloma), primary tumors of the heart (primarily sarcomas), "solid" tumors that reach the heart by local extension (breast, lung) or metastases, and non-malignant conditions such as amyloidosis, sarcoidosis, and some of the collagen vascular diseases. The prognosis of these disorders is usually more closely related to the underlying disease, although the actual cause of death may be cardiac. For example, malignancies involving the heart, especially "solid" tumors, tend to have a uniformly poor prognosis. Nonetheless, infiltrative disorders may directly af-

fect the conduction system and cause life-threatening bradyarrhythmias and tachyarrhythmias. In these situations, permanent pacemakers or defibrillators can be life saving.

1 Amyloidosis—A group of disorders in which deposition of an insoluble protein within the myocardium can produce a restrictive cardiomyopathy, resulting in congestive heart failure and arrhythmias.[5] This protein can infiltrate the intramural vasculature, causing microvascular ischemia. The resulting perivascular fibrosis can affect the specialized conduction system, causing sinus node disease, intraventricular conduction defects or AV block. Permanent pacemakers may be helpful in alleviating symptoms, but have not been demonstrated to provide a survival benefit.

2 Sarcoidosis—A relatively common disorder of unknown etiology. Cardiac involvement, with the characteristic non-caseating granulomas, is frequent and, when extensive, portends a poor prognosis. The conduction system is often involved, and permanent pacemakers are often required for symptomatic sinus node disease or AV block. Implantable defibrillators are most often necessary because of recurrent malignant ventricular tachyarrhythmias. Unfortunately, death from progressive heart failure is not uncommon.

3 Collagen vascular diseases—Cardiac involvement is relatively frequent in individuals with a collagen vascular disease, especially polymyositis. Arrhythmias are not common, but fibrosis of the conduction system can cause AV block, necessitating a permanent pacemaker.

Sinus node dysfunction

Sinus node dysfunction, or sick sinus syndrome and its variants, is a heterogeneous clinical syndrome of diverse etiologies. This disorder includes sinus bradycardia, sinus arrest, SA block, chronotropic incompetence, and various supraventricular tachycardias (atrial or junctional) alternating with periods of bradycardia or asystole (Table 1.4). Sinus node dysfunction is quite common, and its incidence increases with advancing age. In patients with sinus node dysfunction, the correlation of symptoms with bradyarrhythmias is critically important. This is because there is a great deal of disagreement about the absolute heart rate or length of pause required before pacing is indicated. If the symptoms of sinus node disease are dramatic (e.g. syncope, recurrent dizzy spells, seizures, or severe heart failure), then the diagnosis may be relatively easy. However, the symptoms are often extremely non-specific (e.g. easy fatigability, depression, listlessness, early signs of dementia) and in the elderly may be easily misinterpreted. Instead, many of these patients have symptoms as a result of an abrupt change in heart rate (e.g. termination of tachycardia with a sinus pause or sinus bradycardia) (Fig. 1.10). It is important to realize that the degree of bradycardia that may produce symptoms will vary depending on the patient's physiological status, age, and activity at the time of bradycardia (e.g. eating, sleeping or walking) (Fig. 1.11). In patients with sinus node dysfunction whose symptoms have not been shown to cor-

Table 1.4 Diagnosis of sinus node dysfunction

Sinus bradycardia—Sinus rates persistently <60 bpm and associated with symptoms. Prolonged sinus node recovery time (atrial pacing) may help in the diagnosis.

Chronotropic incompetence—Sinus rate does not increase with exertion. Diagnosis made with exercise test or continuous electrocardiographic monitoring.

Sinoatrial (SA) block—Sinus beats are "dropped" in a regular pattern (e.g. 2:1 SA block, 3:2 SA Wenckebach, etc.) due to blocking of impulses in the perinodal area between the sinus node and atrial muscle (by disease, medications, etc.). Diagnosis is made by continuous electrocardiographic monitoring. It may be facilitated by sinus node potential recordings.

Sinoatrial pause—Failure of impulse formation in the sinus node due to pathology, medications, etc. The diagnosis is made electrocardiographically by an absence of sinus P waves that occurs without any discernible pattern.

Bradycardia–tachycardia syndrome—The diagnosis is made electrocardiographically by alternating periods of sinus bradycardia and tachycardia (most commonly atrial fibrillation or flutter). The bradycardia is often manifested by periods of sinus node arrest which often occur when the tachycardia terminates.

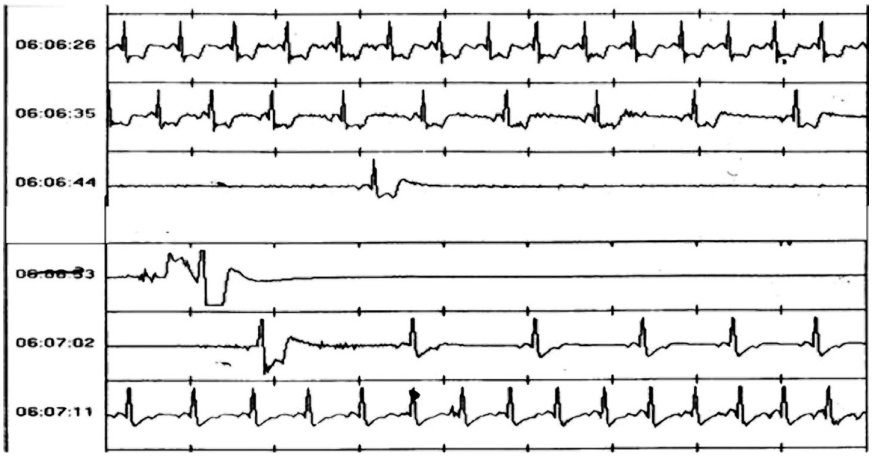

Fig. 1.10 A dramatic example of sinus node dysfunction manifested by 7 and 10 s of asystole as documented by an implantable loop monitor in a patient with recurrent undiagnosed syncope. He underwent permanent pacemaker implantation.

relate with electrocardiographic abnormalities, a simple exercise test may be helpful (to assess the degree of chronotropic incompetence, especially in the individual with vague symptoms) or an electrophysiological study may be considered.

More permanent pacemakers are implanted for sinus node disease than for any other indication in the USA. Patients with alternating periods of brady-

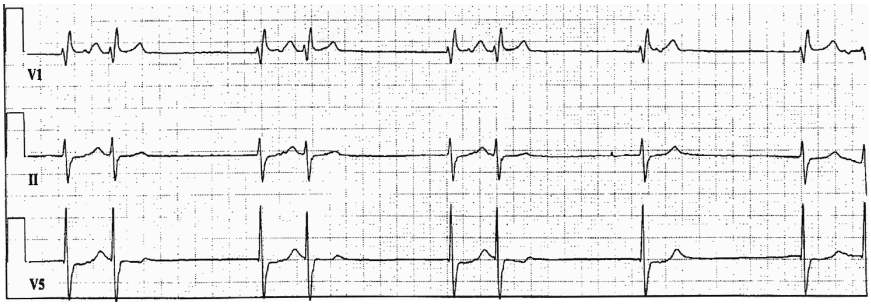

Fig. 1.11 A 69-year-old man had been started on atenolol 75 mg/day for treatment of hypertension approximately 2 weeks earlier. He was seen in the emergency room complaining of feeling weak and lightheaded. The ECG shows a slow junctional escape rhythm followed by a sinus beat in a pattern termed "escape capture bigeminy." Discontinuation of atenolol resulted in return of normal sinus rhythm within 36 h. Patients with sinus node dysfunction may be dependent upon sympathetic stimulation, and β-blockers, even in low doses, may result in profound bradycardia.

cardia and tachycardia (i.e. tachy–brady syndrome) are especially likely to require permanent pacing because medical treatment of the tachycardia often worsens the bradycardia and vice versa (Fig. 1.12). Up to 30% of patients with sinus node disease will also have AV nodal or distal His–Purkinje conduction system disease. Thus, atrial fibrillation, which is a common expression of sinus node disease, may be accompanied by a slow ventricular response, even in the absence of medications that depress AV conduction. Other important complications of sinus node disease include systemic emboli, especially in the setting of alternating periods of bradycardia and tachycardia, and congestive heart failure, usually related to the slow heart rate. In addition, many commonly used medications may exacerbate sinus node dysfunction (Table 1.5). For many patients, an acceptable alternative cannot be found, and pacing is necessary so that they can continue their medications. In some patients, the AV nodal conduction disturbance may not resolve even after discontinuation of drugs that may cause conduction system disturbance.

A group of patients has been identified who have a relatively fixed heart rate during exercise; this condition is referred to as chronotropic incompetence. These patients frequently have other symptoms of sinus node dysfunction. Some may have symptoms at rest (generally non-specific), but most of these patients will note symptoms such as fatigue or shortness of breath with exercise. In some cases, the diagnosis is straightforward; there is no or only a very slight increase in heart rate with exercise. In other cases, the diagnosis is difficult and will require comparison of the patient's exercise response with that of age-matched, gender-matched patients using specific exercise protocols.

Although the indications for permanent pacing for sinus node dysfunction are fairly well delineated, there is considerable debate as to which pacing mode is most appropriate. Because of the high incidence of chronotropic in-

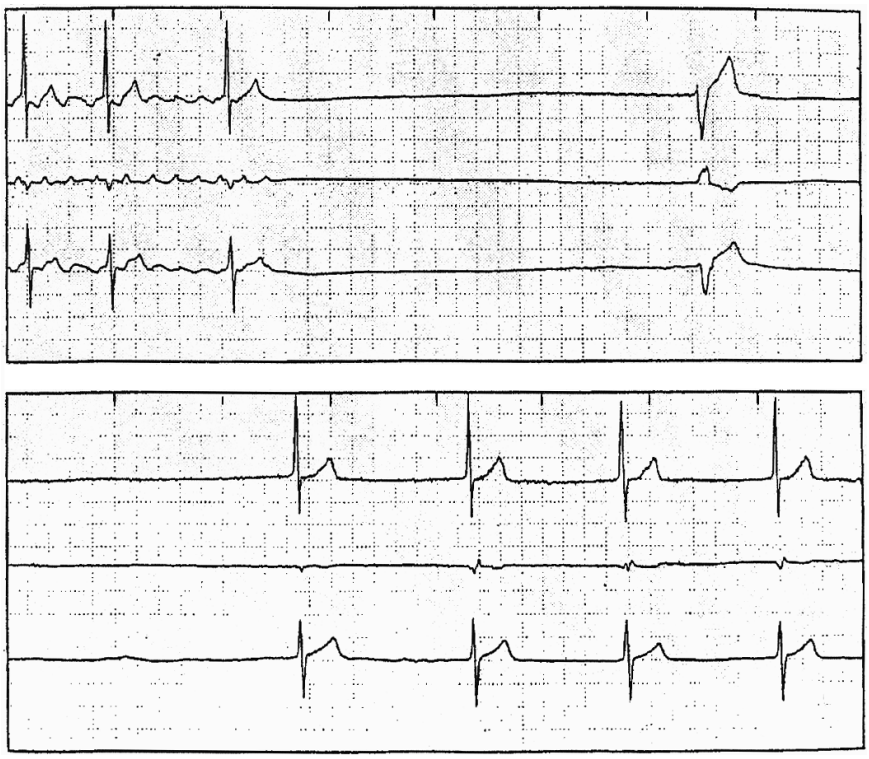

Fig. 1.12 A 53-year-old man with a history of atrial flutter complained of syncope preceded by palpitations. Twenty-four-hour ambulatory recordings (Holter) revealed prolonged periods of asystole following spontaneous termination of atrial flutter. Alternating periods of tachycardia and bradycardia ("bradycardia–tachycardia syndrome") are notoriously difficult to manage without a permanent pacemaker.

Table 1.5 Commonly used medications that may cause sinus node dysfunction or atrioventricular block

- Digitalis (especially in the setting of hypokalemia)
- Antihypertensive agents (clonidine, methyldopa, guanethidine)
- Beta-adrenergic blockers (inderal, metoprolol, nadolol, atenolol), and administration as eyedrops
- Calcium channel blockers (verapamil, diltiazem)
- Type 1A antiarrhythmic drugs (quinidine, procainamide, disopyramide)
- Type 1C antiarrhythmic drugs (flecainide, propafenone)
- Type III antiarrhythmic drugs (amiodarone, sotalol)
- Psychotropic medications
  Tricyclics
  Phenothiazines
  Lithium
  Phenytoin
  Cholinesterase inhibitors

competence, the need for rate-responsive pacing is generally accepted. However, whether dual-chamber (DDD/DDDR) pacing confers any advantage over the VVIR mode is less well established.[6] Pacing to maintain AV synchrony (AAI/DDD) has been shown to reduce the incidence of atrial fibrillation, but does not prevent strokes or prolong survival.[7] Similarly, there is debate about whether patients with intact AV conduction might benefit more from AAI/AAIR than from DDD/DDDR pacing. Single-chamber devices are less complicated and cheaper and allow for normal ventricular activation. These issues are discussed in a separate chapter.

The indications for pacemaker implantation in patients with sinus node dysfunction are listed below.

Class I
1 Sinus node dysfunction with documented symptomatic bradycardia or sinus pauses. Sinus node dysfunction as a result of essential long-term drug therapy of a type and dose for which there are no acceptable alternatives. (Level of evidence: C.)
2 Symptomatic chronotropic incompetence. (Level of evidence: C.)

Class IIa
1 Sinus node dysfunction occurring spontaneously or as a result of necessary drug therapy, with heart rates < 40 bpm, when a clear association between significant symptoms consistent with bradycardia and the actual presence of bradycardia has not been documented. (Level of evidence: C.)
2 Syncope of unexplained origin when major abnormalities of sinus node function are discovered or provoked in electrophysiological studies. (Level of evidence: C.)

Class IIb
1 In minimally symptomatic patients, chronic heart rates < 40 bpm while awake. (Level of evidence: C.)

Class III
1 Sinus node dysfunction in asymptomatic patients, including those in whom substantial sinus bradycardia (heart rate < 40 bpm) is a consequence of long-term drug treatment.
2 Sinus node dysfunction in patients with symptoms suggestive of bradycardia that are clearly documented as not associated with a slow heart rate.
3 Sinus node dysfunction with symptomatic bradycardia due to non-essential drug therapy.

Neurocardiogenic syncope/hypersensitive carotid sinus syndrome

Neurally mediated syncope is a form of abnormal autonomic control of the circulation. It may take one of three forms:

1 The cardioinhibitory type is characterized by ventricular asystole of ≥3 s due to sinus arrest or (occasionally) complete heart block.
2 The pure vasodepressor response is marked by a decrease in arterial pressure of at least 20–30 mmHg, but little or no change in heart rhythm.
3 The mixed type has features of both the cardioinhibitory and vasodepressor types.

Syncope is a common disorder that is estimated to account for up to 6% of all hospital admissions in the USA annually. Despite extensive evaluation, the cause of syncope may not be found in up to 50% of cases. It is believed that a substantial proportion of these cases may be due to neurally mediated syncope. The exact mechanism of neurally mediated syncope has not been fully elucidated, but appears to be initiated by an exaggerated response of the autonomic nervous system to a variety of stimuli. Although the syncope is sometimes an isolated event with an obvious precipitating cause such as severe fright or emotional upset, in many individuals these episodes are recurrent and without apparent triggers. A variety of other stimuli may give rise to cardioinhibitory or mixed cardioinhibitory responses. These conditions, when recurrent and refractory, may also be treated with permanent pacemakers. The conditions include pain, coughing, micturition, swallowing, defecation, and the relatively common vasovagal syndrome. In general, pacemakers may be considered in these patients only when symptoms are recurrent, severe, and cannot be controlled by more conservative measures [e.g. avoidance of stimuli, β-blockers, midodrine hydrochloride (ProAmatine), and/or fludrocortisone acetate (Florinef)]. Pacemaker therapy is thought more likely to be successful in patients who predominantly experience the cardioinhibitory type of response. The advent of head-upright tilt testing has had a major impact on the area of neurocardiogenic syncope. Vasodepressor and/or cardioinhibitory responses may be elicited, which appear to correlate only modestly with the clinical symptoms (Figs. 1.13–1.15). In some studies, the response to tilt table testing has been found to be different from that recorded with an implantable recorder during a spontaneous syncopal episode. The development of permanent pacemakers with the "rate-drop response," which initiates an interval of relatively rapid pacing when the heart rate suddenly drops below a pre-set limit, has stimulated renewed interest in the use of pacing for neurocardiogenic syncope and related disorders. Initial randomized but uncontrolled clinical trials had documented the ability of pacemakers with this feature to reduce syncopal recurrences compared with patients without pacemakers.[8] One double-blind, randomized controlled clinical trial (control group received pacemakers, but programmed to the ODO mode) has shown only a trend toward a reduction in frequency of syncope with active pacing without reaching statistical significance.[9] The final role of pacing in prevention of neurocardiogenic syncope is uncertain; at present, pacing is used only in truly refractory cases in which a significant bradycardic component has been well demonstrated.

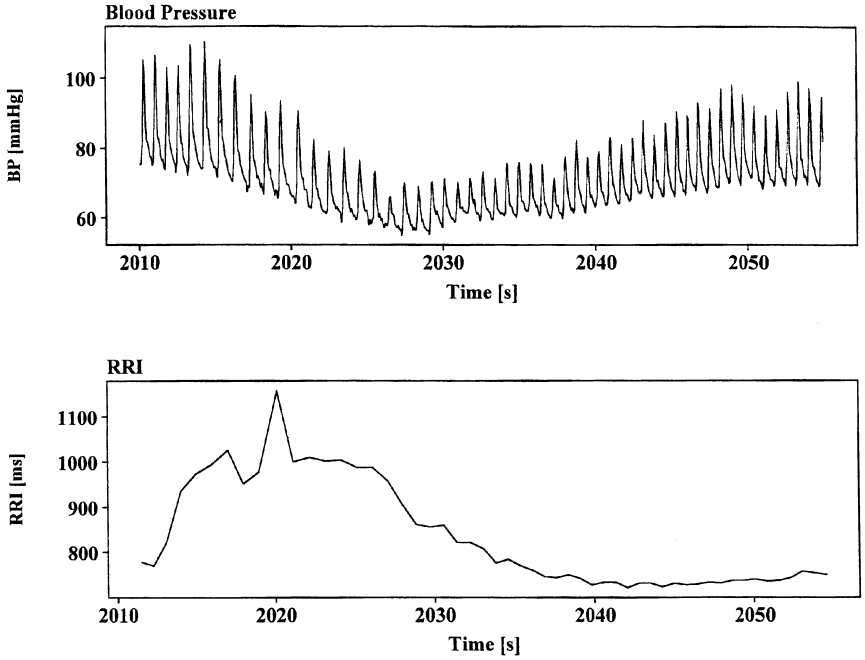

Fig. 1.13 An example of a pure vasodepressor response to tilt testing. The upper panel shows blood pressure and the lower panel shows heart rate (R–R intervals, expressed in milliseconds). Note the marked decrease in blood pressure at a time when the heart rate is actually increasing (the R–R interval is shortening). This type of individual is less likely to respond to permanent pacing.

One variant of neurally mediated syncope is the hypersensitive carotid sinus syndrome. A mildly abnormal response to vigorous carotid sinus massage may occur in up to 25% of patients, especially if coexisting vascular disease is present. Some patients with an abnormal response to carotid sinus massage may have no symptoms suggestive of carotid sinus syncope. On the other hand, the typical history of syncope—blurred vision and lightheadedness or confusion in the standing or sitting position, especially during movement of the head or neck—should be suggestive of this entity. Classic triggers of carotid sinus syncope are head turning, tight neckwear, shaving, and neck hyperextension. Syncopal episodes usually last only several minutes and are generally reproducible in a given patient. Symptoms associated with this syndrome may wax or wane over several years. Carotid sinus hypersensitivity is most often predominantly cardioinhibitory in nature, so that permanent pacing may be very helpful (Fig. 1.16). In contrast, other forms of neurocardiogenic syncope often have a significant vasodepressor component, so that permanent pacing has a more limited role.

The indications for pacemaker implantation in patients with neurally mediated syncope and hypersensitive carotid sinus syndrome are listed below.

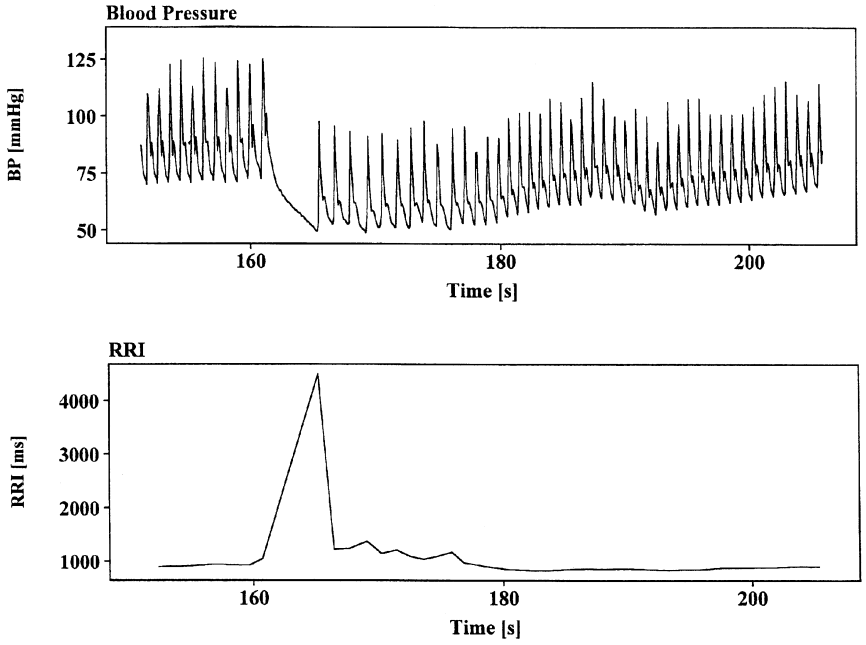

Fig. 1.14 Tracings from a tilt test showing a pure cardioinhibitory response. Note the abrupt increase in cardiac cycle length (R–R interval) reflecting marked bradycardia.

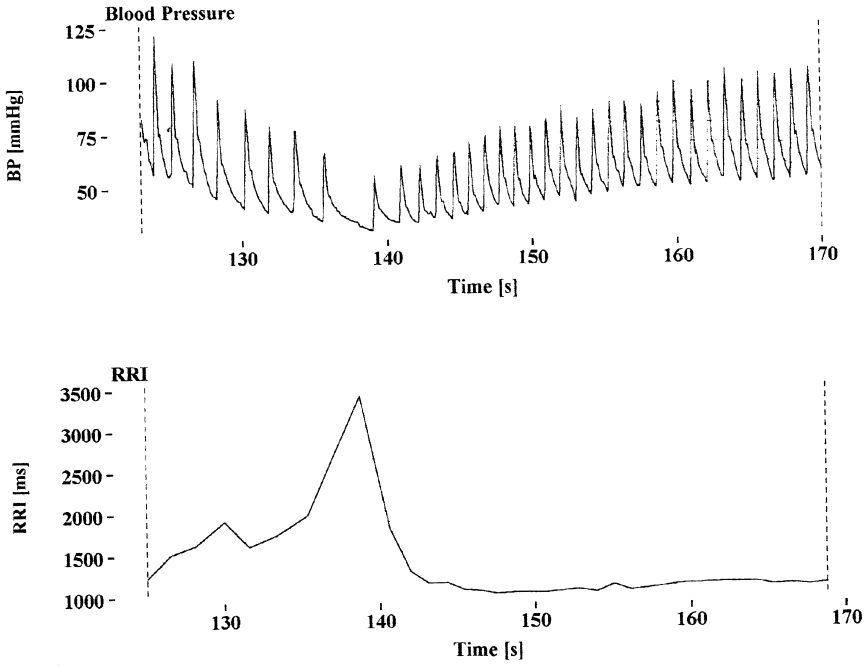

Fig. 1.15 An example of a "mixed" cardioinhibitory and vasodepressor response to tilt testing. An initial decrease in blood pressure is followed by a marked increase in cardiac cycle length.

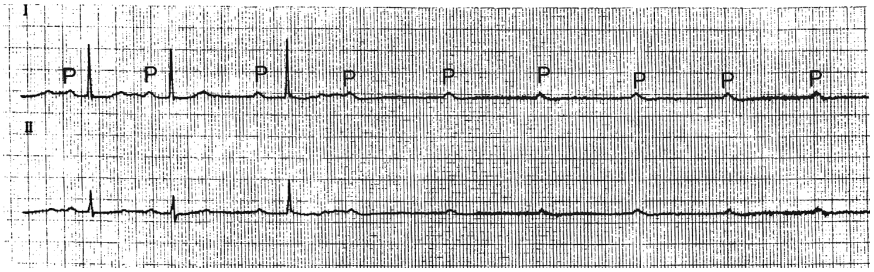

Fig. 1.16 A 49-year-old man complained of near-syncope, which typically occurred while shaving or turning his neck. Carotid sinus massage was performed shortly before the second QRS complex. Note that the sinus rate slows prior to the third QRS complex, followed by complete heart block with ventricular asystole.

Class I

1 Recurrent syncope caused by carotid sinus stimulation; minimal carotid sinus pressure induces ventricular asystole of >3 s duration in the absence of any medication that depresses the sinus node or AV conduction. (Level of evidence: C.)

Class IIa

1 Recurrent syncope without clear, provocative events and with a hypersensitive cardioinhibitory response. (Level of evidence: C.)

2 Significantly symptomatic and recurrent neurocardiogenic syncope associated with bradycardia documented spontaneously or at the time of tilt table testing. (Level of evidence: C.)

The new European guidelines state that recurrent severe vasovagal syncope with prolonged asystole during ECG recording or tilt table testing, after failure of medical therapy and informing patients of the conflicting results of clinical trials, is a class IIa indication in patients >40 years old and a class IIb indication in patients <40 years old.

Class III

1 A hyperactive cardioinhibitory response to carotid sinus stimulation in the absence of symptoms or in the presence of vague symptoms such as dizziness, lightheadedness, or both.

2 Recurrent syncope, lightheadedness or dizziness in the absence of a hyperactive cardioinhibitory response.

3 Situational vasovagal syncope in which avoidance behavior is effective.

Idiopathic orthostatic hypotension is a related neurocirculatory disorder that may respond to permanent pacing. Several reports have documented a beneficial response to atrial or AV sequential pacing in a small number of patients with idiopathic orthostatic hypotension refractory to salt and steroid therapy.[10] The rationale for pacing in this condition is that by increasing the paced rate (the lower rate in these series varies from 80 to 100 bpm), the cardiac

output increases and potentially leads to more vasoconstriction. This therapy usually results in some clinical improvement, but it varies considerably from patient to patient. There are currently no class I or class II indications for permanent pacing for idiopathic orthostatic hypotension.

Hypertrophic cardiomyopathy

Hypertrophic cardiomyopathy is a disorder of the myocardium characterized by excessive myocardial hypertrophy, with a predilection for the interventricular septum. Although there may be obstructive (i.e. a demonstrable gradient across the LV outflow tract) and non-obstructive forms, there might be little difference between them because the gradient is dynamic and affected by preload, afterload, and other factors. Difficulty with diastolic relaxation (and ventricular filling) of the thickened and non-compliant ventricular musculature is present in both forms of this disorder and may be an important determinant of the clinical presentation. Pacing is thought to exert a beneficial effect by inducing paradoxical septal motion and ventricular dyssynchrony and dilation, thereby improving ventricular filling and reducing the outflow tract gradient. This is generally achieved with dual-chamber pacing with a short PR interval (i.e. usually 50–125 ms) to produce maximal ventricular preexcitation. The acute hemodynamic effects of dual-chamber pacing may be quite dramatic, with a major reduction in LV cavity obliteration and a concomitant decrease in LV outflow tract gradient (Fig. 1.17). More intriguing is the suggestion that the beneficial effects of dual-chamber pacing in this condition do not dissipate immediately once the pacing has been terminated.[11]

The mechanism of the beneficial effects of pacing is incompletely understood and the population who would most reliably benefit has not been fully elucidated. In a multicenter trial (the M-PATHY study) using a randomized, double-blind crossover design, Maron and colleagues have found that symptomatic improvement (quality of life and functional class) was not necessarily accompanied by improvement in objective indices such as treadmill exercise time and peak oxygen consumption.[12] Similarly, in the Pacing in Cardiomyopathy (PIC) study, Linde and colleagues have found significant improvement in both the active pacing and inactive pacing (placebo) group, although the improvement was greater in those assigned to active pacing.[13] These two studies suggest that some of the improvement seen in earlier studies may be partly due to placebo effect or the known variability in clinical course of the disorder. These studies, together, suggest a benefit from DDD pacing with a short AV delay in some patients with hypertrophic cardiomyopathy, but there is no way to predict which patients will respond to this therapy. Because prolongation of life has not been documented with this therapy, the current role of permanent pacemakers in hypertrophic cardiomyopathy is unclear. Accordingly, it should be remembered that surgical myotomy–myectomy is still considered the gold standard for treatment of this condition. Septal ethanol ablation is an emerging therapy also. The clinician managing these patients must determine

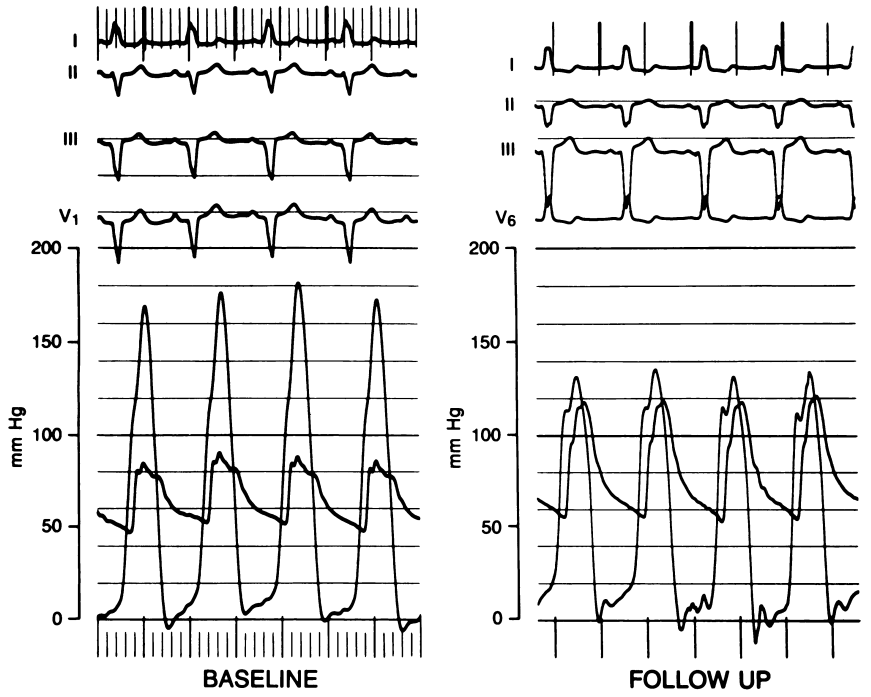

Fig. 1.17 Tracings show reduction of left ventricular outflow tract obstruction after chronic dual-chamber pacing. Left panel: At baseline, the left ventricular systolic pressure and left ventricular outflow gradient were 180 mmHg and 90 mmHg, respectively. Right panel: On follow-up assessment, the left ventricular systolic pressure and left ventricular outflow tract gradient, also measured in sinus rhythm, were reduced to 135 mmHg and 15 mmHg, respectively, despite the temporary inhibition of ventricular pacing. This finding suggests remodeling of ventricular function or anatomy by chronic pacing. From top to bottom: I, II, III, V_1 and V_6 are standard ECG leads.

if the device to be implanted will be a dual-chamber pacemaker or a dual-chamber defibrillator. Defibrillators are generally implanted when patients have one risk factor for sudden cardiac death (see Chapter 8).

The indications for permanent pacing for hypertrophic cardiomyopathy are as follows.

Class I

1 Class I indications for sinus node dysfunction or AV block as previously described. (Level of evidence: C.)

Class IIb

1 Medically refractory, symptomatic hypertrophic cardiomyopathy with significant resting or provoked LV outflow tract obstruction. (Level of evidence: A.)

Class III
1 Patients who are asymptomatic or medically controlled.
2 Symptomatic patients without evidence of LV outflow tract obstruction.

Dilated cardiomyopathy (left ventricular systolic dysfunction)

A related area in which permanent pacing may be of benefit is dilated cardiomyopathy. Early studies have suggested that dual-chamber pacing, especially with a short AV delay, may have important hemodynamic benefit in patients with severe congestive heart failure. Although the exact mechanism has not been determined, it was postulated that the improvement in hemodynamics may be related to optimization of ventricular filling or reduction of diastolic mitral regurgitation. However, controlled studies from several groups have failed to confirm these beneficial effects.[14] Studies of right ventricular (RV) outflow tract pacing for LV systolic dysfunction have been negative or mixed.[15]

In contrast, there is considerable evidence that the use of LV or biventricular permanent pacing improves hemodynamics in some patients with congestive heart failure. Because LV contraction is a key determinant of cardiac output, in theory the properly synchronized contraction of the left ventricle or both ventricles should enhance cardiac performance in patients with prolongation of the QRS duration. Randomized, double-blind, controlled clinical trials have clearly established the beneficial role of biventricular pacing therapy in advanced heart failure patients with prolonged QRS duration.[16]

The indications for pacing in patients with heart failure and impaired LV systolic function are:

Class I
1 Class I indications for sinus node dysfunction or AV block as previously described. (Level of evidence: C.)
2 Biventricular pacing in medically refractory, symptomatic New York Heart Association (NYHA) class III or IV patients with idiopathic dilated or ischemic cardiomyopathy, prolonged QRS interval (≥120 ms), LV end-diastolic diameter ≥55 mm and ejection fraction ≤35%. (Level of evidence: A.) Most of these patients will qualify for implantable cardioverter defibrillator (ICD) therapy, as listed in Chapter 8. The choice between a biventricular pacemaker and a biventricular ICD should be made based upon the patient's age and a variety of other clinical factors.

Class IIa
1 Biventricular pacing for heart failure patients with NYHA class III or IV symptoms, left ventricular ejection fraction (LVEF) ≤35%, LV dilation and a concomitant indication for permanent pacing (first implant or upgrading of conventional pacemaker). (Level of evidence: C.)
2 Biventricular pacing for patients with permanent atrial fibrillation and NYHA class III–IV heart failure on optimal medical therapy with an LVEF

≤35%, LV dilation, and indication for AV junction ablation. (Level of evidence: C.)

Class III
1 Asymptomatic dilated cardiomyopathy.
2 Symptomatic dilated cardiomyopathy when patients are rendered asymptomatic by drug therapy.
3 Symptomatic ischemic cardiomyopathy when the ischemia is amenable to intervention.

Prevention and termination of tachyarrhythmias including the prolonged QT syndrome

Permanent pacing can be used in some situations to prevent or terminate supraventricular [supraventricular tachycardia (SVT)] and ventricular arrhythmias. Individuals with prolongation of the QT or QT–U interval may be prone to a type of polymorphic ventricular tachycardia known as torsades de pointes (Fig. 1.18). Tachycardia is often preceded by a short–long–short series of changes in cycle length. Episodes tend to be paroxysmal, recurrent, and may become life-threatening. Therefore, it is critical that the clinical syndrome be recognized, any offending drugs be stopped, and any electrolyte deficiencies be corrected. A summary of the various conditions associated with torsades de pointes is provided in Table 1.6.

Permanent pacing may also be of help in patients with the long QT syndrome, especially for bradycardic patients who have a history of ventricular arrhythmias or syncope. It provides more uniform repolarization and an increased heart rate, which will shorten the QT interval. Permanent pacing may also permit the use of β-blockers, known to be of benefit in this syndrome, without worsening the resting bradycardia. Currently, many of these patients undergo implantation of defibrillators instead of pacemakers, depending mostly on whether the episode was caused solely by bradycardia or whether underlying structural or repolarization abnormalities are present.

Because radiofrequency ablation successfully treats most types of SVT, antitachycardia pacing is now rarely used for the treatment of these arrhythmias.

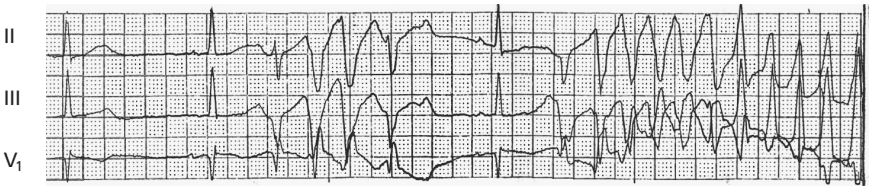

Fig. 1.18 A rhythm strip of ECG leads II, III and V_1 shows paroxysms of polymorphic ventricular tachycardia in an individual with QT interval prolongation and recurrent syncope. Note the short–long–short cycle length sequence that initiates the arrhythmia. In the absence of an identifiable cause, the patient received an implantable cardioverter defibrillator.

Table 1.6 Causes of torsades de pointes

Electrolyte abnormalities
Hypokalemia
Hypomagnesemia
Hypocalcemia
Antiarrhythmic agents
Quinidine
Procainamide
Disopyramide
Amiodarone
Sotalol
Dofetilide
Ibutilide
Hereditary long QT syndrome(s)
Bradyarrhythmias
Liquid protein diets
Myocardial ischemia/infarction
Neurological events
Subarachnoid hemorrhage
Head trauma
Non-cardiac drugs
Antihistamines (astemizole, terfenadine)
Tricyclic and tetracyclic antidepressants
Phenothiazines
Cisapride
Erythromycin
Trimethoprim sulfamethoxazole
Chloroquine
Amantadine
Pentamidine
Toxins
Organophosphates
Arsenic

See www.torsades.org or www.longqt.org/medications for updated list.

There is, however, growing interest in permanent pacing therapies for atrial fibrillation. In patients with concomitant sinus bradycardia, dual-site atrial pacing combined with drug therapy may reduce the recurrence rates of atrial fibrillation. In addition, preliminary data suggest that antitachycardia pacing may terminate atrial flutter and other atrial tachycardias that frequently coexist in some patients with atrial fibrillation. Specialized algorithms have been

developed that increase the frequency of atrial pacing based on the hypothesis that suppression of premature atrial contractions and decreasing atrial heterogeneity may reduce the frequency and duration of episodes of atrial fibrillation. Ventricular antitachycardia pacing without back-up defibrillation is contraindicated due to the risk of tachycardia acceleration.

The indications for permanent pacing to prevent or terminate tachycardias are:

Class I
1 Sustained pause-dependent VT, with or without prolonged QT, in which the efficacy of pacing is thoroughly documented. (Level of evidence: C.)

Class IIa
1 High-risk patients with congenital long-QT syndrome. (Level of evidence: C.)
2 Symptomatic recurrent SVT that is reproducibly terminated by pacing in the unlikely event that catheter ablation and/or drugs fail to control the arrhythmia or produce intolerable side effects. (Level of evidence: C.)

Class IIb
1 Recurrent SVT or atrial flutter that is reproducibly terminated by pacing as an alternative to drug therapy or ablation. (Level of evidence: C.)
2 AV re-entrant or AV node re-entrant SVT not responsive to medical or ablative therapy. (Level of evidence: C.)
3 Prevention of symptomatic, drug-refractory recurrent atrial fibrillation in patients with coexisting sinus node dysfunction. (Level of evidence: B.)

Class III
1 Tachycardias frequently accelerated or converted to fibrillation by pacing.
2 The presence of accessory pathways with the capacity for rapid anterograde conduction whether or not the pathway(s) participate in the mechanism of the tachycardia.
3 Frequent or complex ventricular ectopic activity without sustained VT in the absence of the long-QT syndrome.
4 Torsades de pointes VT due to reversible causes.

Pacing for children and adolescents, including congenital heart block

The general indications for pacing in children and adolescents are similar to those for adults with several additional considerations. The diagnosis of significant bradycardia in children depends on age, presence and type of congenital heart disease, and cardiac physiology. Following surgery for congenital heart disease, patients may have postoperative AV block that, if untreated by pacing, will worsen their prognosis.[17] Congenital heart disease patients may also have tachycardia–bradycardia syndrome, but the benefits of pacing for this indica-

tion are less clear. Congenital heart diseases such as corrected transposition of great arteries, ostium primum atrial septal defects and ventricular septal defects may be associated with complete heart block.

Congenital complete AV block is a rare anomaly that results from abnormal embryonic development of the AV node and is not associated with structural heart disease in 50% of cases. Congenital complete heart block is also associated with maternal lupus erythematosus. Most children with isolated congenital complete AV block have a stable escape rhythm with a narrow complex. Pacing is generally indicated in children with complete heart block if the heart rate in the awake child is < 50 bpm or if associated with LV systolic dysfunction or ventricular arrhythmias. The indications for pacing in congenital complete AV block have been clarified by a prospective study demonstrating improved survival and reduced syncope, myocardial dysfunction and mitral regurgitation even among asymptomatic patients.[18] Exercise testing does not predict future cardiac events in this population.

The indications for permanent pacing in children and adolescents are:

Class I

1 Advanced second- or third-degree AV block associated with symptomatic bradycardia, ventricular dysfunction, or low cardiac output. (Level of evidence: C.)

2 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. (Level of evidence: B.)

3 Postoperative advanced second- or third-degree AV block that is not expected to resolve or persists at least 7 days after cardiac surgery. (Levels of evidence: B, C.)

4 Congenital third-degree AV block with a wide QRS escape rhythm, complex ventricular ectopy, or ventricular dysfunction. (Level of evidence: B.)

5 Congenital third-degree AV block in the infant with ventricular rate < 50–55 bpm or with congenital heart disease and a ventricular rate < 70 bpm. (Levels of evidence: B, C.)

6 Sustained pause-dependent VT, with or without prolonged QT, in which the efficacy of pacing is thoroughly documented. (Level of evidence: B.)

Class IIa

1 Bradycardia–tachycardia syndrome with the need for long-term antiarrhythmic treatment other than digitalis. (Level of evidence: C.)

2 Congenital third-degree AV block beyond the first year of life with an average heart rate < 50 bpm, abrupt pauses in ventricular rate that are two or three times the basic cycle length, or associated with symptoms due to chronotropic incompetence. (Level of evidence: B.)

3 Long-QT syndrome with 2 : 1 AV or third-degree AV block. (Level of evidence: B.)

4 Asymptomatic sinus bradycardia in the child with complex congenital heart disease with resting heart rate <40 bpm or pauses in ventricular rate >3 s. (Level of evidence: C.)
5 Patients with congenital heart disease and impaired hemodynamics due to sinus bradycardia or loss of AV synchrony. (Level of evidence: C.)

Class IIb
1 Transient postoperative third-degree AV block that reverts to sinus rhythm with residual bifascicular block. (Level of evidence: C.)
2 Congenital third-degree AV block in the asymptomatic infant, child, adolescent or young adult with an acceptable rate, narrow QRS complex, and normal ventricular function. (Level of evidence: B.)
3 Asymptomatic sinus bradycardia in the adolescent with congenital heart disease with resting heart rate <40 bpm or pauses in ventricular rate >3 s. (Level of evidence: C.)
4 Neuromuscular diseases with any degree of AV block (including first-degree AV block), with or without symptoms, because there may be unpredictable progression of AV conduction disease.

Class III
1 Transient postoperative AV block with return of normal AV conduction. (Level of evidence: B.)
2 Asymptomatic postoperative bifascicular block with or without first-degree AV block. (Level of evidence: C.)
3 Asymptomatic type I second-degree AV block. (Level of evidence: C.)
4 Asymptomatic sinus bradycardia in the adolescent with longest RR interval <3 s and minimum heart rate >40 bpm. (Level of evidence: C.)

Permanent pacing after the acute phase of acute myocardial infarction

Bradyarrhythmias and conduction defects are relatively common after acute MI. In patients who have these problems, a decision about permanent pacing must be made prior to the patient's discharge from the hospital. It is important to realize that the indications for temporary pacing in the setting of acute MI are different from those for permanent pacing following infarction. Unfortunately, there is some uncertainty regarding permanent pacing for these patients, because large prospective controlled trials have not been performed. In addition, the criteria for permanent pacing in patients after an MI do not necessarily require the presence of symptoms, and the need for temporary pacing in the acute stages of infarction is not by itself an indication for permanent pacing.

The prognosis for these patients is strongly influenced by the amount of underlying myocardial damage.[19] In general, sinus node dysfunction tends to be benign and reversible, and permanent pacemakers are rarely required. Similarly, second-degree and even third-degree AV block after inferior wall

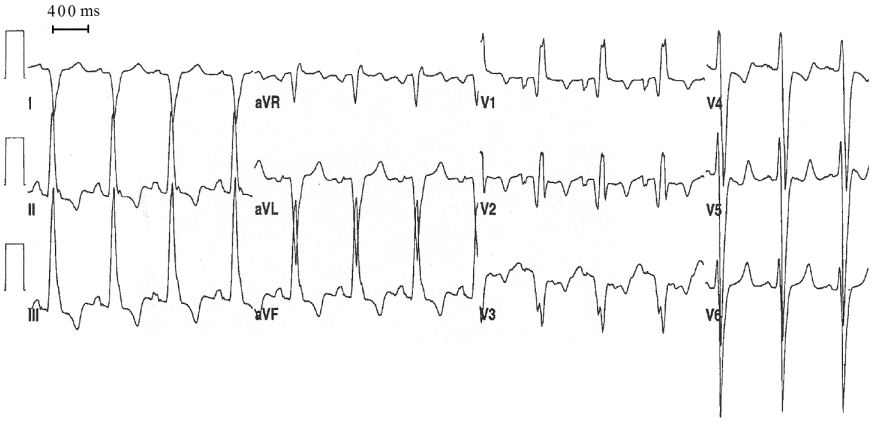

Fig. 1.19 A standard 12-lead ECG from an individual with a large anteroseptal myocardial infarction complicated by congestive heart failure and right bundle branch block with right axis deviation, presumably due to left posterior fascicular block. The patient developed transient high-degree atrioventricular block 72 h after admission (a class I indication) and subsequently underwent permanent pacemaker implantation.

MI is usually reversible and rarely requires permanent pacing. In contrast, conduction defects after an anterior wall MI usually warrant a permanent pacemaker or an ICD with appropriate pacing function insertion, although mortality remains extremely high because of pump failure (Fig. 1.19).

A class I recommendation (level of evidence: C) is that all patients who have an indication for permanent pacing after ST elevation MI should be evaluated for ICD indication. Likewise, class IIa recommendations (level of evidence: C) suggest that biventricular pacing should be evaluated if permanent pacing is indicated and that permanent dual-chamber pacing should be considered if the patient is in sinus rhythm.

The indications for permanent pacing following acute MI are:

Class I

1 Persistent second-degree AV block in the His–Purkinje system with bundle branch block or third-degree AV block within or below the His–Purkinje system after acute MI. (Level of evidence: B.)

2 Transient advanced (second- or third-degree) infranodal AV block and associated bundle branch block. If the site of block is uncertain, an electrophysiology study may be necessary. (Level of evidence: B.)

3 Persistent and symptomatic second- or third-degree AV block. (Level of evidence: C.)

Class IIb

1 Persistent second- or third-degree AV block at the AV node level. (Level of evidence: B.)

Class III
1 Transient AV block in the absence of intraventricular conduction defects. (Level of evidence: B.)
2 Transient AV block in the presence of isolated left anterior fascicular block. (Level of evidence: B.)
3 Acquired left anterior fascicular block in the absence of AV block. (Level of evidence: B.)
4 Persistent first-degree AV block in the presence of bundle branch block that is old or age indeterminate. (Level of evidence: B.)

European guidelines
Class I
Persistent third-degree heart block preceded or not by intraventricular conduction delay.
Persistent Mobitz II second-degree heart block associated with bundle branch block, with or without PR prolongation.
Transient Mobitz II second- or third-degree heart block associated with new-onset bundle branch block.

Class IIa
None.

Class IIb
None.

Class III
Similar to above.

Indications for temporary cardiac pacing

The following section reviews the clinical settings in which temporary cardiac pacing is indicated. Chapter 4 presents a review of the techniques and complications of temporary cardiac pacing. A summary of the general indications for temporary pacing is given in Table 1.7.

Acute myocardial infarction

In the setting of an acute MI, several different types of conduction disturbance may become manifest. They include abnormalities of sinus impulse formation or conduction, disorders of AV conduction, and disorders of intraventricular conduction. In general, any patient with bradyarrhythmias that are associated with symptoms or cause hemodynamic compromise must be treated. The ways of identifying the patient populations at greatest risk for the development of a significant bradyarrhythmia during acute MI, and in whom temporary pacing should be performed prophylactically, are discussed below. It is important to

Table 1.7 Indications for temporary pacing in the ABSENCE of acute myocardial infarction

In acute myocardial infarction
See Tables 1.8 and 1.9
In absence of acute myocardial infarction
Medically refractory symptomatic bradycardia
• sinus node dysfunction
• second- or third-degree AV block
Third-degree AV block with wide QRS escape or ventricular rate < 50 bpm
Prophylactic
Swan–Ganz catheterization or endocardial biopsy in patient with left bundle branch block
Cardioversion in setting of sick sinus syndrome
New AV or bundle branch block with acute endocarditis (especially aortic valve endocarditis)
Perioperatively in patient with bifascicular block and history of syncope
To allow pharmacological treatment with drugs that worsen bradycardia
Treatment of tachyarrhythmias
Termination of recurrent ventricular or supraventricular tachycardia
Suppression of bradycardia-dependent ventricular tachyarrhythmias including torsades de pointes

realize that the indications for temporary pacing in the setting of acute MI are different from those for permanent pacing following infarction.

Sinus node abnormalities

Sinus node dysfunction may include sinus bradycardia, sinus arrest and/or sinoatrial exit block. The incidence of these electrocardiographic abnormalities is quite variable, ranging from 5% to 30% in different series. Abnormalities of sinus rhythm are more common, with inferoposterior infarction because either the right or left circumflex coronary artery is occluded—these arteries most commonly supply the sinus node. Another potential reason is chemically mediated activation of receptors on the posterior left ventricular wall—these receptors are supplied by vagal afferent fibers. Treatment of sinus bradycardia is not necessary, unless symptoms such as worsening myocardial ischemia, heart failure or hypotension are documented. Atropine may be administered for vagally mediated bradycardia. If bradycardia is prolonged and severe, or is not responsive to atropine, temporary cardiac pacing is indicated.

Disorders of atrioventricular conduction

AV block occurs without associated intraventricular conduction system abnormalities in 12–25% of patients with acute MI. The incidence of this finding depends largely on the patient population and the site of infarction. First-degree AV block occurs in 2–12% of patients, second-degree AV block in 3–10%, and third-degree AV block in 3–7% of patients. The majority of patients with abnormalities of AV conduction without bundle branch block have evidence

of an inferoposterior infarction (approximately 70%). The reasons for the increased incidence of AV conduction abnormalities are related to the coronary blood supply to the AV node. The coronary artery supplying the inferoposterior wall of the left ventricle is typically the right or left circumflex coronary, which is occluded during an inferior infarction. In addition, activation of cardiac reflexes with augmentation of parasympathetic tone during inferior ischemia (infarction) may also be responsible. In some cases, AV block may be due to release of adenosine caused by inferior ischemia or during inferior infarction.

The risk of progression from first-degree AV block to high-grade AV block (during inferior infarction) varies from 10% to 30%, and that of second-degree AV block to complete heart block is approximately 35%. As would be expected, the development of high-grade AV block in the setting of acute inferoposterior infarction is usually associated with narrow QRS complex escape rhythms (Figs 1.20 and 1.21). The junctional escape rhythm usually remains stable at 50–60 pulses per minute and can be increased by intravenous atropine, so even complete AV block may not require temporary pacing in this situation.

Type I second-degree AV block with a narrow QRS almost always represents a conduction block in the AV node, and temporary cardiac pacing is rarely required unless the patient has concomitant symptoms. Type I second-degree AV block with a wide QRS complex may represent a conduction block in the AV node or His bundle or contralateral bundle branch block. In these

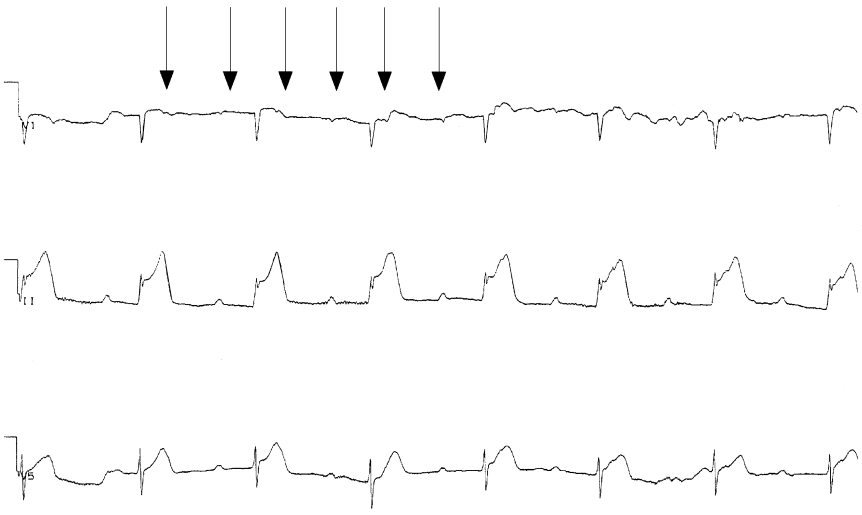

Fig. 1.20 A three-lead (standard leads V_1, II and V_5) rhythm strip in a 58-year-old man with an acute inferior wall myocardial infarction and complete atrioventricular block (the arrows in lead V_1 identify the P waves) with an escape rate of 45 ppm. Despite the slow rate, the patient did not exhibit signs or symptoms of hemodynamic compromise, so no therapy was required. Normal conduction resumed approximately 24 h later.

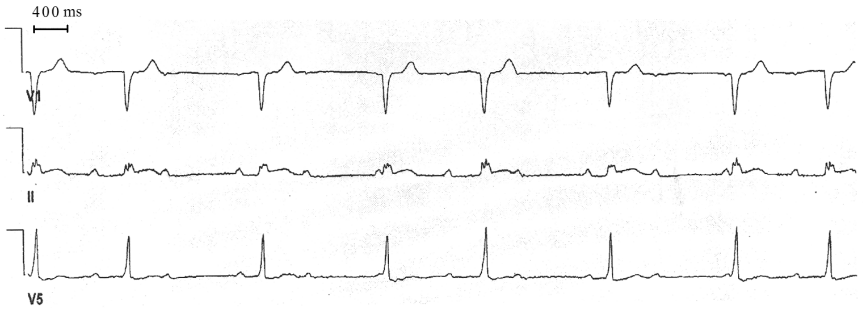

Fig. 1.21 Rhythm strips recorded from a 63-year-old woman with an acute inferior wall myocardial infarction showing high-grade atrioventricular (AV) block with junctional escape beats. The second, fourth and sixth QRS complexes are conducted with a prolonged PR interval (note shortening of the R–R interval with the conducted beats). The presence of junctional escape beats precludes typical Wenckebach conduction. Because the patient was asymptomatic, no therapy was administered. Normal AV conduction resumed spontaneously by the next morning.

patients, especially in the setting of anterior MI, temporary prophylactic pacing must be considered. In patients with type II second-degree AV block and a wide QRS complex in the setting of inferior infarction, or with a wide or narrow QRS complex during an anterior MI, a temporary pacemaker should be inserted. Patients with a narrow QRS complex and type II second-degree AV block in the setting of inferior infarction rarely progress to complete heart block.

Several special situations are worthy of consideration. Patients with high-grade AV block occurring in the setting of right ventricular infarction tend to be less responsive to intravenous atropine and may demonstrate markedly improved hemodynamics during AV sequential pacing. The mechanism for this hemodynamic improvement is probably a reflection of the restrictive physiology that the infarcted right ventricle demonstrates. Another group of patients who may benefit from prophylactic temporary pacing are those with acute inferior wall infarction with alternating Wenckebach periods. This electrocardiographic finding is rare (2%), but without temporary pacing it frequently leads to hemodynamic embarrassment.

In contrast to inferior wall infarction, high-grade AV block complicating an anterior wall infarction is usually located within the His–Purkinje system. The transition from the first non-conducted P wave to high-grade AV block is often abrupt, and the resulting escape rhythm is typically slow and unreliable. Conducted beats usually have a wide QRS complex. In general, an interruption of the blood supply to the anterior wall and the interventricular septum severe enough to cause AV block usually causes severe LV dysfunction and results in high mortality. Emergency temporary pacing and prophylactic pacing are indicated, although survival may not be significantly improved because of the extent of myocardial damage.

Disorders of the intraventricular conduction system

A number of studies have examined the incidence of development of new bundle branch block in the setting of acute MI and have determined that it varies between 5% and 15%, depending on the site of infarction. New bundle branch block is three times more likely during anterior infarction than during inferior infarction, because the left anterior descending coronary artery provides the major blood supply to the His bundle and the bundle branches. Not surprisingly, there is a high incidence of heart failure in this setting, and the associated high cardiac mortality leads to controversy as to whether temporary or permanent pacing improves the poor prognosis in these patients. As with anterior MI and complete heart block, new bundle branch block reflects extensive myocardial damage.

Multiple studies have shown that patients with acute infarction and bundle branch block have a fourfold to fivefold increased risk of progression to high-grade AV block (e.g. an increase from 4% to 18%).[20,21] Both in-hospital and out-of-hospital mortality are higher for patients presenting with bundle branch block during acute infarction. The basis of this increased mortality may be a variety of causes, including heart failure, infarct extension, ventricular tachycardia and heart block. The mortality of patients with bundle branch block and acute infarction is 30–40%, compared with 10–15% in patients without bundle branch block. Most of the increase in cardiac mortality appears to be related to the degree of heart failure.

Several small retrospective studies have attempted to identify groups of patients who may be at increased risk of progression to high-grade heart block. Unfortunately, many of these studies are limited by their retrospective nature, their small sample size, or their ascertainment bias. On the basis of the results of several studies, patients with conduction system abnormalities who should have temporary pacemakers inserted prophylactically are listed in Table 1.8.

Patients with a new bundle branch block and first-degree AV block or old bifascicular block and first-degree AV block are at intermediate risk of progression (19–29%) to high-grade AV block; they may undergo prophylactic pacing depending on the availability of facilities for emergency placement of a temporary pacemaker. Because the greatest risk of progression to complete heart block occurs in the first 5 days following infarction, these decisions should be made promptly so that temporary pacing may be instituted.

The Multicenter Investigation of the Limitation of Infarct Size (MILIS) study has suggested a simpler method of risk stratification.[22] A "risk score" for the development of complete heart block was devised. Patients with any of the following conduction disturbances were given one point: first-degree AV block, type I second-degree AV block, type II second-degree AV block, left anterior fascicular block, left posterior fascicular block, RBBB, and LBBB. The presence of no risk factors was associated with a 1.2% risk of third-degree AV block, one risk factor with a 7.8% risk, two risk factors with a 25% risk, and three risk factors with a 36.4% risk of complete heart block. These findings were validated by testing the risk score in over 3000 patients from previously published

Table 1.8 Risk of high-grade atrioventricular (AV) block during acute myocardial infarction

Patient group	*Risk of high-grade AV block*
First-degree AV block and new bifascicular BBB	38–43%
First-degree AV block and old bifascicular BBB	20–50%
New bifascicular BBB	15–31%
Alternating BBB	44%
MILIS risk score[a]	
0	1.2%
1	7.8%
2	25%
3	36.4%

[a]One point each for first-degree AV block, Mobitz I second-degree AV block, Mobitz II second-degree AV block, left anterior fascicular block, left posterior fascicular block, right bundle branch block, left bundle branch block.
BBB, bundle branch block; MILIS, Multicenter Investigation of the Limitation of Infarct Size.

studies. The risk score appears to be an alternative to risk stratification using combinations of conduction disorders.

The effect of thrombolytic and early interventional therapies on the subsequent development of high-grade AV block in patients presenting with acute infarction and intraventricular conduction system disease has been poorly studied. Although the incidence of complete AV block in acute MI has decreased following thrombolytic therapy, mortality remains high.[23]

The most recent AHA guidelines for temporary transvenous pacing in the setting of acute MI are as shown in Table 1.9.[24]

Pacing during cardiac catheterization

During catheterization of the right side of the heart, manipulation of the catheter may induce a transient RBBB in up to 10% of patients. This block generally lasts for seconds or minutes, but can occasionally last for hours or days. Trauma induced by right ventricular endomyocardial biopsy also may result in temporary, or rarely long-lasting, RBBB. This is a problem only in patients with preexisting LBBB, in whom complete heart block may result. We therefore recommend consideration of placement of a temporary transvenous pacing wire in patients who are undergoing right heart catheterization or biopsy in the presence of previously known LBBB. Catheterization of the left side of the heart in patients with known preexisting RBBB only rarely gives rise to complete heart block because of the short length of the left bundle branch. Significant bradycardia and asystole can occur during injection of the right coronary artery. This complication is extremely rare, and the placement of a temporary pacing catheter does not alter the morbidity or mortality of cath-

Table 1.9 Indications for temporary pacing in acute myocardial infarction

Class I
1. Asystole
2. Symptomatic bradycardia (includes sinus bradycardia with hypotension and type I second-degree AV block with hypotension not responsive to atropine)
3. Bilateral bundle branch block [BBB; alternating BBB or right BBB (RBBB) with alternating left anterior fascicular block (LAFB)/left posterior fascicular block (LPFB)] (any age)
4. New bundle branch block with Mobitz II second-degree AV block
5. RBBB plus fascicular block with Mobitz II second-degree AV block
Class IIa
1. Narrow QRS plus Mobitz II second-degree AV block
2. Old or new fascicular block with Mobitz II second-degree AV block and anterior MI
3. Old bundle branch block and Mobitz II second-degree AV block
4. New bundle branch block plus first-degree AV block
5. New bundle branch block plus Mobitz I second-degree AV block
6. RBBB plus LAFB or LPFB (new or indeterminate) with first-degree AV block
7. RBBB plus LAFB or LPFB (new or indeterminate) with Mobitz I second-degree AV block
Class IIb
1. Bifascicular block of indeterminate age and Mobitz II second-degree AV block
2. Old bundle branch block and first-degree AV block or second-degree AV block
Class III
1. First-degree heart block
2. Type I second-degree AV block
3. Accelerated idioventricular rhythm
4. BBB or fascicular block known to exist before acute myocardial infarction without conduction system disease

eterization. The bradycardia usually resolves after several seconds. The same comments apply in general to placement of a temporary pacing wire during angioplasty.

Preoperative pacing

One of the questions most frequently asked of a consulting cardiologist by both surgeons and anesthesiologists is whether it is necessary to insert a temporary pacing catheter in patients with bifascicular block undergoing general anesthesia. The results of several studies have shown that the incidence of intraoperative and perioperative complete heart block is quite low. There does not appear to be any benefit from preoperative prophylactic pacemaker insertion. Even in patients with first-degree AV block and bifascicular block, there is a very low incidence of perioperative high-grade heart block.

However, in patients who have bifascicular block and also type II second-degree AV block or a history of unexplained syncope or presyncope, the risk of development of high-grade AV block is higher, and a temporary pacemaker

should be inserted. The appearance of new bifascicular block in the immediate postoperative period should also lead to insertion of a temporary pacemaker and should raise suspicion of an intraoperative MI. The general availability of transcutaneous pacing may make it an acceptable alternative to temporary transvenous pacing in lower risk individuals, although poor patient tolerance is often a limitation.

Open-heart surgery tends to be associated with a somewhat higher incidence of postoperative bradyarrhythmias than does non-cardiac surgery, due to the direct trauma to the conduction system and interference with blood supply. Cardiac surgeons generally implant temporary epicardial pacing wires at the time of surgery to facilitate temporary pacing. The major problem then becomes determining how long to wait for resumption of AV conduction and normalization of sinus node function before implanting a permanent pacemaker. Conventionally, a permanent pacemaker is recommended if the problem persists longer than 5–7 days after the operation. Although normal conduction may resume after this period of time, in the absence of definitive information about the natural history of these disorders, permanent pacing seems to be a prudent choice.

Other temporary pacing

Temporary pacing is indicated in patients with new AV or BBB in the setting of acute bacterial endocarditis. The development of a new conduction system abnormality generally suggests that there is a perivalvular (ring) abscess that has extended to involve the conduction system near the AV node and/or the His bundle. The endocarditis generally involves the non-coronary cusp of the aortic valve. In one study, high-grade or complete heart block developed in 22% of patients with aortic valve endocarditis and new first-degree AV block. Although these studies are retrospective, the patient with development of new AV block or BBB, especially in the setting of aortic valve endocarditis, should probably undergo temporary pacing while cardiac evaluation continues.

Treatment of tumors of the head and/or neck or around the carotid sinus may in some circumstances give rise to high-grade AV block. Temporary pacing may be required during surgical treatment, radiation therapy or chemotherapy. If the tumor responds poorly, permanent pacing may be necessary in some cases. The long-term risk for subsequent heart block due to tumor recurrence is difficult to predict in some cases.

Lyme disease, a tick-borne spirochete infection, causes a systemic infection with arthritis, skin lesions, myalgias, meningoencephalitis and cardiac involvement in 5–10% of patients. Lyme disease is epidemic in the summer months in the northeastern USA. Carditis typically occurs relatively late in the course of the illness, usually 4–8 weeks after the onset of symptoms. AV block is the most common manifestation of carditis and tends to be transient. Block is most common at the level of the AV node, and fluctuation between first-degree and higher degrees of AV block is frequent. Temporary cardiac pacing

may be required, but the conduction disturbances usually resolve spontaneously, especially with antibiotic treatment, so permanent cardiac pacing is rarely necessary. Similar conduction disturbances can occasionally be seen in patients with viral myocarditis, as well as with other tick-borne infections.

A number of medications may produce transient bradycardia that may require temporary pacing until the drug has been stopped (see Table 1.5). These drugs may cause sinus node dysfunction and/or AV block; if used in combination, their effects may become more potent and exacerbate mild or latent conduction system disease. If long-term therapy with these agents is necessary for an underlying disorder and a substitute cannot be found, permanent pacing may be required. Recent studies have suggested that at least some patients with transient drug-related bradycardia may experience recurrent bradycardia requiring a pacemaker, even after the drug related to the bradycardia is stopped.

Treatment of tachycardias with temporary pacing

Temporary cardiac pacing has been used for the termination and/or prevention of a variety of arrhythmias. Pacing-termination of ventricular tachycardia is discussed in detail in Chapter 8 on the ICD and will not be dealt with here. Type I atrial flutter can be successfully pace-terminated approximately 65% of the time in an unselected population and in >90% of patients in whom atrial flutter develops after surgery. Due to the development of radiofrequency catheter ablation techniques, there is currently less interest in pace-termination of atrial flutter. Similarly, the most common varieties of paroxysmal supraventricular tachycardia are usually pace-terminable, but tend to be equally amenable to radiofrequency ablation.

Torsades de pointes is a polymorphic ventricular tachycardia with a sinusoidal electrocardiographic appearance, due to the QRS complex undulating about the baseline. It results from prolongation and dispersion (inequality in different parts of the ventricle) of myocardial repolarization, and is often reflected on the surface ECG by a prolonged QT or QT–U interval. Episodes of tachycardia are often preceded by a short–long–short series of changes in cycle length. Importantly, episodes tend to be recurrent, paroxysmal, and non-sustained initially, but may become sustained later unless the underlying condition is identified and corrected. Therefore, it is critical that the clinical syndrome be recognized, any offending toxins or drugs be stopped, and any electrolyte deficiencies be corrected. (A summary of the causes of torsades de pointes is provided in Table 1.6.) In some patients these treatments will be adequate; in some patients, other forms of therapy must be considered. Overdrive atrial and/or ventricular pacing may be effective in suppressing torsades de pointes because it provides more uniform repolarization and an increased heart rate, which will shorten the QT interval. Intravenous magnesium and isoproterenol (which also increases heart rate and shortens repolarization) may also be effective in suppressing torsades de pointes, although the latter is associated with troubling side effects. For individuals who have recurrent

symptoms refractory to conventional management, implantation of an ICD should be strongly considered.

Summary

Updated AHA/ACC guidelines for pacemaker and defibrillator implantation are currently in progress. Recently, the European Society of Cardiology has published their updated guidelines for cardiac pacing and cardiac resynchronization therapy.[25]

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