Cardiac Arrhythmias

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Introduction

Cardiac arrhythmias are disturbances in the rhythm of the heart, manifested by irregularity or by abnormally fast rates (tachycardias) or abnormally slow rates (bradycardias). Patients who perceive these abnormalities most frequently observe palpitations, which some describe as the sensation of 'my heart turning over in my chest', or awareness that their hearts are beating rapidly or slowly. Other symptoms include weakness, shortness of breath, lightheadedness, dizziness, fainting (syncope) and, occasionally, chest pain. The symptoms tend to be more severe when the rate is faster, the ventricular function is worse, or the arrhythmia is associated with abnormalities of autonomic tone. However, many patients with arrhythmias report no symptoms, and the condition may first be discovered during a routine examination. A tachyarrhythmia that is rapid enough and lasts long enough can produce cardiomyopathy and congestive heart failure. In these cases, treatment of the arrhythmia can often return normal function to the ventricles.

Although certain physical signs present during arrhythmias can help the physician make a correct diagnosis, electrocardiography is the standard method used for recognizing cardiac arrhythmias. A prolonged electrocardiographic recording, often called a 'Holter monitor', or an event recorder that the patient activates when sensing an abnormality, may assist in confirming the diagnosis when the arrhythmia occurs sporadically, as is often the case.

Sinus Rhythms

The normal rhythm of the heart originates in the sinus node, a collection of cells at the junction of the right atrium and the superior vena cava with the unique property of automaticity, shared with few other cardiac tissues. Automatic cells in the sinus node discharge at rates affected by autonomic influences through the parasympathetic nervous system, which regulates heart rates during most normal activities and at rest, and by sympathetic stimulation, which raises the heart rate during exercise. Atrial activation, which produces P-waves on the electrocardiogram during normal sinus rhythm, passes from the sinus node towards the atrioventricular node and from right to left atrium. This process produces an axis of the P waves in the frontal plane that ranges from approximately 0 to $+75°$. After atrial activation, the electrical activity traverses the atrioventricular node and depolarizes the ventricles, producing the QRS complex on the electrocardiogram.

For decades, sinus rhythm in adults has been defined as present when the heart rate is 60 to 100 beats per minute. Some authorities, however, suggest the lower rate should be 50 beats per minute, a rate often found in normal athletes whose resting parasympathetic tone is greater than in most people. Sinus tachycardia is diagnosed when the rate exceeds 100 beats per minute, and sinus bradycardia when the rate is less than 60 or 50 beats per minute. In children, normal sinus rhythm is faster than in adults.

Sinus rhythm tends to be slightly irregular. Inspiration briefly increases the rate, and expiration decreases it. The name 'sinus arrhythmia' is given to the cardiac rhythm when the variation is particularly marked. Sinus arrhythmia may frequently be found in young subjects with normal hearts.

In a few patients, more often women than men, the sinus pacemaker may continuously discharge faster than normal in the absence of any obvious structural heart disease, another atrial arrhythmia or diseases such as thyrotoxicosis. If this condition produces symptoms, the diagnosis of 'inappropriate sinus tachycardia' may be assigned. Those patients requiring treatment may be given beta-blocking or calcium-blocking drugs to decrease the heart rate. Rarely, ablative or surgical treatment has been applied.

Premature Beats

Premature beats, which may originate in the atria or the ventricles, are the most common cardiac arrhythmia. They occur in subjects with otherwise normal hearts and in patients with heart disease of lesser or greater severity. Palpitations are the principal symptoms produced by premature beats, whatever their origin. The sensation is produced by the early contraction of the ventricles

followed, after a pause, by a stronger than normal contraction. The name bigeminy is applied to those premature beats that alternate with sinus beats, trigeminy when one or two of three beats are premature, and quadrigeminy when one of four beats is premature.

Premature beats in patients whose hearts are otherwise normal do not require treatment. Some patients find that avoiding use of caffeinated beverages or smoking will reduce the number of premature beats that afflict them. If patients demand relief from the palpitations, β-adrenergic blocking drugs may be prescribed.

Atrial premature beats

Atrial premature beats are produced by abnormalities of atrial electrical activity that discharge the atria early in competition with the normal function of the sinus node. The form or morphology of the P waves of atrial premature beats is abnormal, reflecting their origin in locations other than in the sinus node. Atrial premature beats may indicate the presence of atrial pathology, which can produce sustained atrial arrhythmias such as atrial fibrillation or atrial flutter (see below.) In general, however, atrial premature beats tend to have less clinical significance than ventricular premature beats, particularly in patients with intrinsic heart disease. Most patients with atrial premature beats do not require specific treatment for the arrhythmia.

Ventricular premature beats

Ventricular premature beats are recognized by the presence on the electrocardiogram of early QRS complexes with abnormal forms indicating their origin in the ventricles. When present in normal subjects, they are seldom complex, a characteristic defined by their being repetitive, bigeminal, frequent or multiform. Ventricular premature beats occur in every patient during myocardial infarction and more frequently in those who have sustained greater amounts of myocardial damage. Successful thrombolysis is often associated with additional simple and complex beats soon after therapy. Many patients have premature beats after recovery from myocardial infarction. Patients with angina pectoris have more ventricular premature beats than those without the condition and both simple and complex beats frequently develop during episodes of coronary vasospasm.

Most patients with mitral valve prolapse have no more ventricular premature beats than patients without the lesion. Other conditions giving rise to ventricular premature beats include cardiomyopathy, hypertension, pulmonary disease, congenital heart disease, cardiac surgery, metabolic disturbances, alcohol and certain drugs. The time of day, whether one is awake or asleep, and stress also affect the frequency of ventricular premature beats.

Palpitations are the principal symptom produced by ventricular premature beats, and the patient is usually aware of the premature beat itself as well as the strong sinus beat afterwards. Principal physical findings include the premature beat in the pulse, the abnormality of the heart rhythm, abnormal waves forms in the neck veins and variations in the heart sounds.

On the electrocardiogram, the form of the QRS complexes of the ventricular premature beats is abnormal. Their duration is prolonged, and their amplitude is frequently greater than normal. Ambulatory electrocardiographic monitoring best defines the frequency in character of premature beats in individual patients. Exercise can also expose premature beats. Clinical electrophysiological study can identify ventricular premature beats when the diagnosis is confused with aberrant conduction of supraventricular beats.

Few patients with ventricular premature beats require specific therapy. Although many antiarrhythmic drugs are effective, they can reduce ventricular function and endanger the patient's life because of their proarrhythmic effect. Beta-blocking drugs can be helpful for a few patients with intolerable symptoms. Antiarrhythmic therapy does not prolong survival in patients with ventricular premature beats and myocardial infarction and may increase mortality. Coronary artery bypass graft surgery does not reduce the incidence of ventricular premature beats. The site of origin of ventricular premature beats can be ablated.

The prognosis is worse for patients with established coronary heart disease and ventricular premature beats than for those without the arrhythmia. When ventricular premature beats are present and ventricular function is diminished, survival is reduced further. Ventricular premature beats also increase mortality in patients with other forms of chronic heart disease, including patients resuscitated from out-of-hospital ventricular fibrillation, cardiomyopathy and cardiac transplantation.

Supraventricular Tachyarrhythmias

Rapid, regular or irregular arrhythmias, characterized by QRS complexes of supraventricular form and of normal duration unless distorted by an intraventricular conduction defect can be called supraventricular tachyarrhythmias. The most common is atrial fibrillation. Other examples are atrial flutter, atrial tachycardia, multifocal atrial tachycardia, paroxysmal supraventricular tachycardia and junctional ectopic tachycardia.

Atrial fibrillation

The most common sustained cardiac arrhythmia, atrial fibrillation occurs infrequently in the general population but more often as people age. Most patients with atrial

fibrillation have structural heart disease such as mitral stenosis or regurgitation, acute myocardial infarction, Wolff–Parkinson–White syndrome, thyrotoxicosis, recent cardiothoracic surgery, cardiomyopathy, myocarditis or pulmonary disease. Patients with the arrhythmia but no organic heart disease are said to have 'lone atrial fibrillation'. Atrial fibrillation may be paroxysmal and convert to sinus rhythm spontaneously or easily after medical or electrical treatment, or may be chronic and defy successful, sustained conversion. Atrial fibrillation increases the risk of stroke and emboli and the mortality of adults with structural heart disease.

Patients usually recognize the onset of a paroxysm of atrial fibrillation by feeling palpitations and observing that their hearts are beating rapidly. Some patients, however, remain unaware of the presence of the arrhythmia, which is particularly the case when the arrhythmia is chronic. The examiner, and the informed patient, can usually detect the characteristic pattern in the pulse or the heart beat. In the neck veins, the A waves produced by contraction of the right atrium will be absent, and the intensity of the first heart sound will often vary.

The electrocardiogram of patients with untreated atrial fibrillation usually shows a rapid, irregular ventricular rate in which the P waves are replaced by an undulating baseline. The range of ventricular rates is wide and may exceed 200 per minute in patients with pre-excitation. The duration of the QRS complexes, although usually normal, may be prolonged (aberrant) and simulate ventricular beats and ventricular tachycardia. The left atrium is often enlarged and, after long periods of chronic atrial fibrillation, loses contractile ability when sinus rhythm is re-established. Cardiac function usually decreases when atrial fibrillation replaces sinus rhythm. Many patients develop polyuria during episodes of paroxysmal atrial fibrillation.

Electrophysiological studies show replacement of organized, regularly occurring atrial signals by rapid, irregular electrical activity. Abnormalities of atrial vulnerability, conduction, excitability and refractoriness characterize patients with atrial fibrillation. The likelihood of very rapid ventricular rates and ventricular fibrillation developing in patients with Wolff–Parkinson–White syndrome and atrial fibrillation can often be determined in the electrophysiology laboratory. These arrhythmias occur because of faster than normal conduction of the atrial signals to the ventricles over the accessory pathways present in patients with the Wolff–Parkinson–White syndrome. The duration of the QRS complexes in these cases will be abnormally prolonged and the form abnormal and changing depending upon the amount of conduction through the atrioventricular node or the accessory pathway.

Administration of a β-adrenergic or calcium channel blocking drug or digoxin will decrease the ventricular rate through their affect on conduction within the atrioventricular node. However, digoxin or verapamil should not be employed if pre-excitation (Wolff–Parkinson–White syndrome) is suspected because these drugs may increase the ventricular rate in these patients. If conversion to sinus rhythm is thought feasible, the presence of a clot in the left atrium should be ruled out with a transoesophageal echocardiogram and, if it is absent, pharmacological or electrical cardioversion may be conducted. Patients are usually anticoagulated with heparin before conversion and then with warfarin for several weeks afterwards. If a clot is found, three weeks of anticoagulation should be prescribed before elective conversion. All patients with chronic atrial fibrillation or with frequent episodes of paroxysmal atrial fibrillation should be anticoagulated, unless such treatment is strongly contraindicated, to reduce the occurrence of thromboembolic events.

To maintain sinus rhythm in patients prone to recurrence of atrial fibrillation, various antiarrhythmic drugs may be prescribed. Often used today are flecainide for patients with normal ventricular function and amiodarone for those with reduced left ventricular ejection fractions, since amiodarone in these patients is less likely to induce dangerous ventricular arrhythmias (proarrhythmia). Dofetilide, a recently released class III antiarrhythmic drug, assists in the conversion to, and maintenance of, sinus rhythm in patients with atrial fibrillation.

Patients who are symptomatic because of atrial fibrillation and in whom sinus rhythm cannot be sustained, may be treated with ablation of their atrioventricular junctions and insertion of a permanent ventricular pacemaker. The regular rhythm produced by a pacemaker, the rate of which may vary in accordance with the physiological needs of the patient, provides better ventricular function than the irregular rhythm of atrial fibrillation at the same average rate. All patients with the Wolff–Parkinson–White syndrome who present with atrial fibrillation should be evaluated in the electrophysiological laboratory for ablative treatment to prevent the occasional occurrence of cardiac arrest. Some cases of paroxysmal atrial fibrillation may originate from foci that can be ablated in the electrophysiology laboratory. The Maze surgical operation can restore sinus rhythm in some patients with atrial fibrillation that is otherwise difficult to treat successfully.

Atrial flutter

Atrial flutter is a relatively uncommon supraventricular tachyarrhythmia that develops in adults with various types of heart disease or severe pulmonary disease, after cardiac surgery, and in some with no structural heart disease. Children develop flutter after operations to correct congenital defects involving the atria. Atrial flutter is more often paroxysmal than chronic, but flutter may persist continuously for years.

On physical examination, the pulse is rapid and more frequently regular than irregular. Normal atrial wave

forms in the neck veins are absent. Auscultation reveals rapid heart action and, if it is irregular, the signs of partial atrioventricular dissociation such as varying intensity of the first heart sound.

The electrocardiogram shows the characteristic atrial flutter waves at about 250–350 per minute. In most cases, these are negative in the inferior leads and upright in lead V1 and are due to counterclockwise rotation through a re-entrant circuit in the right atrium. Clockwise rotation produces flutter waves which are upright in the inferior leads and inverted in lead V1. In untreated patients, the atrioventricular ratio is usually 2:1, which produces a ventricular rate of about 150 per minute. The atrioventricular ratio is most frequently an even number, such as 2:1 or 4:1, or may vary between these two, which produces an irregular ventricular rhythm. Occasionally, the ventricles beat very rapidly with each atrial signal transmitted through the normal conducting system or via accessory pathways in patients with the Wolff–Parkinson–White syndrome.

Carotid sinus pressure and the Valsalva manoeuvre, by increasing vagal tone and atrioventricular block, decrease the ventricular rate in many patients with atrial flutter and, thereby, reveal the flutter waves in the electrocardiogram when they cannot be discerned at rapid ventricular rates.

Drugs are used in patients with flutter to decrease the ventricular rate, convert to sinus rhythm or prevent recurrences of the arrhythmia. Adenosine, amiodarone, β-adrenergic blocking agents, calcium channel antagonists and digitalis slow the ventricular rate by increasing atrioventricular block. Various antiarrhythmic agents convert some patients to sinus rhythm and help maintain normal rhythm afterwards. However, the most effective method is electrical cardioversion, which produces sinus rhythm in almost all patients with atrial flutter. Before elective cardioversion is performed, the left atrium should be searched for thrombi with transoesophageal echocardiography, even though the likelihood of finding intra-atrial clots in patients with atrial flutter is probably lower than in patients with atrial fibrillation. Rapid atrial or oesophageal pacing will also convert many cases of flutter, although pacing may produce transient or persistent atrial fibrillation.

Catheter ablation of the re-entrant circuit with radio-frequency current produces long-term relief from further paroxysms in many patients with atrial flutter. Recurrence of flutter or conversion to atrial fibrillation limits the results, however. A few will require ablation of the atrioventricular junction with subsequent ventricular pacing when symptom-producing flutter cannot be controlled or cured. Patients with persistent flutter should receive chronic anticoagulation.

Atrial tachycardia

Atrial tachycardia is a relatively uncommon supraventricular tachyarrhythmia produced either by re-entrant circuits or by automatic foci in the atria. Adults with this arrhythmia almost always have intrinsic heart disease. In most cases, each abnormal atrial beat is transmitted to the ventricles, but, occasionally, atrioventricular block may develop as in the arrhythmia previously known as 'PAT (paroxysmal atrial tachycardia) with block' which can be caused by digitalis intoxication. The rate of the atrium in patients with this arrhythmia ranges from less than 100 to more than 250 beats per minute, with the average rate of about 150 beats per minute. The rate in young children is faster. In patients with atrial tachycardia due to re-entry, the onset and offset of the arrhythmia is sudden, and the patient may report this phenomenon. In automatic atrial tachycardia, the arrhythmia may come and go without the typical features of a paroxysmal arrhythmia. Abnormally formed, identical P waves, indicating the non-sinus origin of atrial activation, are characteristic of the electrocardiogram of patients with atrial tachycardia. In a few patients, the form of the P waves during the arrhythmia may appear almost identical to that present during sinus rhythm. In these cases, some authorities assign the name 'sinus node reentry' to the arrhythmia.

Adenosine, amiodarone and beta and calcium channel blocking drugs, given intravenously, convert most patients with re-entrant atrial tachycardia. Combinations of these and other antiarrhythmic drugs often suppress recurrences. Amiodarone, flecainide and β-adrenergic blocking drugs seem to be the most effective agents for suppressing automatic atrial tachycardia. Cardioversion and atrial pacing convert re-entrant atrial tachycardia, but cardioversion is ineffective for reverting automatic atrial tachycardia and pacing only temporarily suppresses the arrhythmia.

Ablating the site of origin of atrial tachycardia with radiofrequency current delivered through electrode-catheters may suppress the focus of the arrhythmia whatever its mechanism. Unfortunately, the arrhythmia may return after ablative treatment, with the tachycardia now originating from another focus. In refractory cases, ablation of the atrioventricular node or bundle of His and insertion of a ventricular pacemaker may provide a satisfactory solution.

Multifocal atrial tachycardia

Multifocal atrial tachycardia is a rapid irregular supraventricular tachyarrhythmia produced by the discharging of several pathological foci. Patients with the arrhythmia are usually elderly, usually have severe pulmonary, cardiovascular and/or infectious diseases and are often acutely ill. Many have diabetes. Multifocal atrial tachycardia, or a similar rhythm that pediatricians often call 'chaotic atrial tachycardia', occurs occasionally in children, more often in boys than in girls.

Electrocardiographically, multifocal atrial tachycardia is characterized by premature P waves with at least three abnormal forms. Although some of the P waves may be blocked from transmission to the ventricles, most are conducted. The heart rates range from 100 to about 180 beats per minute. Atrial fibrillation is the arrhythmia with which multifocal atrial tachycardia is most frequently confused. In both cases, the ventricular rhythm is irregular, and the abnormal P waves of multifocal atrial tachycardia may be misinterpreted to be the undulations of atrial activation in atrial fibrillation.

Treatment of multifocal atrial tachycardia should begin by improving cardiac, pulmonary, metabolic and infectious conditions that have given rise to the arrhythmia. Frequently, multifocal atrial tachycardia will resolve after such measures without specific antiarrhythmic therapy. However; multifocal atrial tachycardia frequently recurs during exacerbations of the underlying diseases.

β-Adrenergic and calcium channel blocking drugs will usually decrease the heart rate in multifocal atrial tachycardia and improve patients' cardiovascular function. These drugs, however, should be given with caution to patients with pulmonary spasm or severely decreased ventricular function. Amiodarone, flecainide and magnesium have proved useful in some cases by either suppressing the abnormal atrial activity or increasing atrioventricular block, thereby slowing the ventricular rate.

Digitalis is not particularly effective in suppressing multifocal atrial tachycardia, but neither does the drug produce the arrhythmia. Digitalis intoxication, however, can develop in patients with multifocal atrial tachycardia since they frequently have severe myocardial disease and may be hypokalaemic, hypomagnesaemic, hypoxic or uraemic. Atrioventricular block, ventricular arrhythmias and death have been reported when too much digitalis has been administered in a misguided attempt to slow the ventricular rate when atrial fibrillation has been incorrectly diagnosed. However, in the absence of toxicity, digitalis need not be withheld if the drug is otherwise indicated.

The prognosis of adults with multifocal atrial tachycardia depends more upon the severity of the underlying disease than on the arrhythmia.

Paroxysmal supraventricular tachycardia (PSVT) and Wolff–Parkinson–White syndrome

PSVT, a relatively common arrhythmia, arises from re-entry in dual pathways within the atrioventricular node or through accessory pathways in patients with the Wolff–Parkinson–White syndrome. It is the most frequent regular tachyarrhythmia in adults, and atrioventricular nodal re-entry is the more common mechanism. When PSVT first appears in relatively young subjects, the re-entry is more often sustained in accessory pathways. Re-entry in accessory pathways is more common in men and intranodal re-entry in women.

PSVT, as the name implies, is paroxysmal, but occasionally re-entrant supraventricular tachycardia may be persistent or permanent, a pattern that occurs less often in adults than in children, in whom the persistent arrhythmia may produce reversible cardiomyopathy. Most patients with PSVT have no structural heart disease. Palpitations are the most frequent symptom, and many patients can recognize that the paroxysms characteristically start and stop suddenly. Pounding in the neck, due to large venous waves produced by the right atrium contracting when the tricuspid valve is closed, occurs more often in patients with atrioventricular nodal re-entry than in those whose re-entry is sustained through accessory pathways. PSVT decreases the systemic blood pressure, cardiac output, stroke volume and ejection fraction, and raises pulmonary artery, right atrial and left atrial pressures. Fainting seems to depend more on vasomotor instability than on the haemodynamic effects of the tachycardia alone.

Few adults have dual atrioventricular nodal pathways, and not all that do have PSVT. Most tachycardias sustained within dual atrioventricular nodal pathways course in the antegrade direction through the slower conducting pathway and conduct retrogradely over the faster conducting pathway ('slow–fast' sequence). In a few cases, the reverse course is followed ('fast–slow' sequence) and many of these have persistent tachycardia. The arrhythmia usually starts after an atrial premature beat that dissociates the pathways and allows re-entry to begin. In the Lown–Ganong–Levine syndrome, the P–R intervals are short, but no delta waves are present. Some of these patients have PSVT sustained in dual pathways.

During sinus rhythm, the Wolff–Parkinson–White electrocardiogram of short P–R intervals and delta waves identifies the presence of accessory pathways that pre-excite the ventricles before normal conduction through the atrioventricular node can do so. When PSVT occurs in patients with the Wolff–Parkinson–White syndrome, the atrioventricular node is usually the antegrade limb supporting the tachycardia and the accessory pathway is the retrograde pathway. The circuit sustaining the tachycardia includes the ventricles and the atria. The electrocardiogram during the arrhythmia in patients with the Wolff–Parkinson–White syndrome shows a rapid regular rhythm usually with normal QRS complexes. Rate-related bundle branch aberrancy or antegrade conduction through accessory pathways, however, can produce abnormal QRS complexes. Incessant or persistent PSVT in these patients courses in the opposite direction to the usual route. Concealed accessory pathways cannot conduct in the antegrade direction and never produce ventricular pre-excitation, the electrocardiographic hallmark of Wolff–Parkinson–White syndrome. Ventricular

premature beats more easily start PSVT sustained within accessory pathways than within dual atrioventricular nodal pathways. Arrhythmias are also sustained through pathways that connect structures other than the atria and ventricles, and some patients have multiple pathways.

The P waves are inverted in the inferior leads when they can be found. In most cases of dual pathway re-entry, they are invisible, hidden within the QRS complexes; in re-entry through accessory pathways, the P waves often follow the QRS complexes. Parts of the P waves may produce what appear to be small Q waves, small S waves, or incomplete right bundle branch block.

Conversion is usually produced by slowing conduction in the atrioventricular node. This is the effect of carotid sinus pressure or the Valsalva manoeuvre that patients can administer themselves to produce sinus rhythm. Electrophysiological study can predict which drugs are most likely to convert or suppress re-entrant PSVT. Drugs that slow conduction include adenosine and calcium channel blockers, which, given intravenously, convert most paroxysms. β-Adrenergic blocking drugs are also, but somewhat less often, effective. The same drugs are frequently prescribed to prevent recurrences, but chronic suppression is usually less successful than acute conversion. Consequently, many patients are now treated by catheter ablation, which permanently cures most patients of the arrhythmia whether sustained in dual atrioventricular nodal or accessory pathways.

PSVT rarely affects prognosis. A few patients with accessory pathways are at risk of potentially fatal ventricular arrhythmias, but they can usually be identified and cured with catheter ablation.

Junctional and Accelerated Idioventricular Arrhythmias

Arrhythmias that arise in automatic tissues of the atrioventricular node or bundle of His are called junctional arrhythmias because the tissues involved lie in the region where the atria and ventricles join. The pacemaking properties of these tissues are normally suppressed and only appear to protect the heart from asystole when the primary pacemaker, usually the sinus node, or atrioventricular conduction fails. Because of their similarities clinically and electrophysiologically, the accelerated arrhythmias of the atrioventricular junction and ventricles are discussed together in this section.

Junctional ectopic tachycardia

Junctional ectopic tachycardia is an abnormal automatic tachyarrhythmia that arises in the atrioventricular conduction system above the bifurcation of the bundle of His. Children develop junctional ectopic tachycardia as a

congenital disease without associated structural defects and from cardiac surgery. The arrhythmia is rare in adults.

Many children with congenital junctional ectopic tachycardia have a family history of the arrhythmia. Associated congenital cardiac defects are uncommon. Congenital junctional ectopic tachycardia usually presents early in life. When the arrhythmia follows cardiac surgery, the children have often had operations involving the atrioventricular junction. Congestive heart failure and hypotension accompany rapid heart rates.

The characteristic electrocardiographic finding is atrioventricular dissociation. The arrhythmia has the electrophysiological features of an automatic, not a re-entrant, mechanism.

Congenital junctional ectopic tachycardia is usually treated with β-adrenergic blocking drugs and amiodarone, which will decrease the ventricular rate. Digitalis is administered to improve cardiac function. Atrial pacing can temporarily re-establish atrioventricular synchrony and increase cardiac output. Ablation of the atrioventricular junction often obliterates the arrhythmia, but a pacemaker is then required.

Since junctional ectopic tachycardia can severely compromise cardiac function when it develops after cardiac surgery, usually in children with congenital heart disease, acute treatment is essential. Digitalis should be given to raise cardiac output. Drugs that increase catecholamine stimulation and may accelerate automatic foci should be avoided, and agents with vagotonic effects should be administered. Amiodarone, β-adrenergic blocking drugs and propafenone decrease the rate of the tachycardia, but other antiarrhythmic drugs are usually ineffective. Ablation of the atrioventricular junctional and ventricular pacing may occasionally be needed. The arrhythmia can be expected to resolve spontaneously in most cases if the patient survives the early postoperative period.

Accelerated junctional and idioventricular rhythms

Adults develop accelerated junctional and idioventricular rhythms from digitalis intoxication, acute myocardial infarction, myocarditis and cardiac surgery. Ablation of the atrioventricular node can produce accelerated junctional rhythm, and successful coronary thrombolysis often induces transient accelerated idioventricular rhythm. Most adults who develop these arrhythmias have significant myocardial disease, but occasionally they can occur in patients without structural heart disease. Some children with acute rheumatic fever develop both accelerated arrhythmias. Accelerated junctional and idioventricular rhythm are not paroxysmal, and patients are often unaware of their presence. The physical examination may reveal the signs of atrioventricular dissociation.

The rate of accelerated junctional rhythm is about 70–130 beats per minute and accelerated idioventricular rhythm is between 60 and 100 beats per minute, both faster than the normal escape rate of junctional or ventricular pacemakers. The ventricular rate is characteristically greater than the sinus rate, and partial atrioventricular dissociation with intermittent atrial capture of the ventricles occurs frequently. During isorhythmic dissociation in patients with accelerated junctional rhythm, the discharges of atria and of ventricles may appear to relate to one another. In accelerated junctional rhythm, the QRS complexes are usually of normal width unless concurrent bundle branch block is present. Prolonged, abnormal QRS complexes characterize accelerated idioventricular rhythm. Both arrhythmias have the characteristics of automatic rather than re-entrant disturbances.

Specific treatment of accelerated junctional or idioventricular rhythms is seldom required. Increasing the atrial rate with atropine, catecholamines or atrial pacing can suppress both. When digitalis causes the arrhythmias, the drug must be discontinued. Cardioversion will not restore normal rhythm. The prognosis depends primarily upon the severity of the underlying cardiac disease.

Ventricular Tachyarrhythmias

The tachyarrhythmias that originate in the ventricles, monomorphic and polymorphic ventricular tachycardia and ventricular fibrillation, threaten the lives of adults more often than do any other tachyarrhythmias.

Monomorphic ventricular tachycardia

Ventricular tachycardia in which the QRS complexes have a uniform morphology is an uncommon arrhythmia that occurs more frequently in men than women. The arrhythmia may appear briefly as 'transient' or 'unsustained' ventricular tachycardia within a range from three consecutive beats to a paroxysm lasting no longer than 30 s. When 'sustained' or 'incessant', the arrhythmia lasts longer than 30 s or requires conversion because of haemodynamic deterioration within that period.

The most common cause of monomorphic ventricular tachycardia is chronic coronary heart disease. The arrhythmia also occurs in patients with cardiomyopathy, rheumatic heart disease, or no evidence of structural heart disease, and occasionally in association with a variety of other cardiac conditions.

Patients aware of an episode of the arrhythmia usually complain of rapid heart action. Chest pain, dyspnoea, weakness and neurological symptoms, including fainting, can also develop. The most characteristic physical sign is a rapid heart rate of an apparently regular tachyarrhythmia. Varying systolic blood pressure, irregular, large A waves in the neck veins and changing intensity of the first heart sound and systolic murmurs identify atrioventricular dissociation, which is characteristic of about half the cases of sustained ventricular tachycardia.

The electrocardiogram shows a rapid, regular or slightly irregular, tachycardia with QRS complexes of greater than normal width and having the appearance of either right or left bundle branch block. The complexes may have constant (monomorphic) or changing (polymorphic, see below) form. P waves identify atrioventricular dissociation when present. The arrhythmia can be differentiated from atrial or supraventricular tachycardia with aberrant conduction in most cases through the clinical history and physical examination and careful review of the electrocardiogram. Ventricular tachycardia is the diagnosis when the width of the QRS complexes exceeds 0.14 s, or when atrioventricular dissociation, concordance or fusion beats are present. Late potentials are frequently seen during sinus rhythm in the signal-averaged electrocardiogram of patients with sustained ventricular tachycardia.

Many patients have severe haemodynamic distress during sustained and incessant ventricular tachycardia that produces many of the symptoms they observe. Hypotension and decreased ventricular function are frequent. Pathological ventricular endocardial tissue provides the substrate for the arrhythmia in those cases with structural heart disease.

Electrophysiological study shows that ventricular tachycardia is sustained most frequently by re-entry and occasionally by triggered activity or automaticity. Ventricular pacing by programmed stimulation induces the arrhythmia in most patients with spontaneous sustained ventricular tachycardia. The efficacy of many antiarrhythmic drugs can be assessed by their ability to prevent induction. Sustained ventricular tachycardia can be induced in some symptomatic patients with transient ventricular tachycardia. Programmed stimulation of the ventricles often terminates ventricular tachycardia.

Intravenous antiarrhythmic drugs and electric shock convert most episodes of spontaneous sustained or incessant ventricular tachycardia. Chronic treatment should be developed through electrophysiological study, which usually leads to implantation of cardioverter-defibrillators in patients with sustained ventricular tachycardia. Most patients with transient ventricular tachycardia do not need specific treatment. However, an implantable cardioverter defibrillator should be considered for those with coronary heart disease whose ventricular function is reduced and in whom a potentially fatal arrhythmia can be induced by electrophysiological study.

The prognosis of patients with recurrent ventricular tachycardia depends primarily upon their ventricular function. Consequently, survival is excellent in those with transient or sustained ventricular tachycardia and no structural heart disease but worsens as ventricular function

deteriorates from myocardial infarctions, cardiomyopathy or other diseases.

Polymorphic ventricular tachycardia, torsades de pointes, long-QT syndrome and bidirectional tachycardia

Polymorphic ventricular tachycardia is an uncommon tachyarrhythmia distinguished by the changing morphology of its QRS complexes. Most patients have organic heart disease and are taking drugs, often antiarrhythmic drugs, many of which prolong the QT intervals during sinus rhythm.

The polymorphic ventricular tachycardia that develops in patients with long-QT intervals is known as torsades de pointes and also occurs in patients with the long-QT syndrome, a genetically determined, uncommon condition. Genetic analysis can define, to some extent, their risk of sudden cardiac death due to a potentially fatal ventricular arrhythmia. Polymorphic ventricular tachycardia also develops in patients whose QT intervals are normal during sinus rhythm. Most of these patients have coronary heart disease or cardiomyopathy, but a few have no structural heart disease. The bradycardia produced by atrioventricular block or sick sinus syndrome can give rise to polymorphic ventricular tachycardia. Syncope is the most dramatic symptom that brings patients with polymorphic ventricular tachycardia to medical attention.

The most characteristic electrocardiographic feature of polymorphic ventricular tachycardia is the changing form of the QRS complexes. Their morphology and axis change beat-to-beat or within groups of beats, and their duration is prolonged and varies.

A special variety of polymorphic ventricular tachycardia is known as bidirectional tachycardia. In this arrhythmia, which is rapid and regular, the morphology of the abnormally wide QRS complexes alternates. The form in lead V1 is that of right bundle branch block, and the axes in the frontal plane alternate between left and right.

Abnormally prolonged corrected QT intervals during sinus rhythm are the fundamental electrocardiographic feature of the long-QT syndrome. Exercise shortens the QT intervals less than normal in patients with drug-induced torsades de pointes or the long-QT syndrome. Echocardiograms confirm the clinical observation that torsades de pointes occurs more often in patients whose left ventricular function is poor.

Polymorphic ventricular tachycardia usually begins in one of two patterns. A ventricular premature beat starts bradycardia-or pause-dependent torsades de pointes after supraventricular beats with particularly long or abnormally formed QT intervals. A pause follows the premature beat before the final supraventricular beat ('long–short' interval) that precedes the first beat of the tachycardia. The QT intervals during sinus rhythm are normal in patients

with the second pattern. In these, the arrhythmia begins immediately after a ventricular premature beat that occurs relatively soon after the last supraventricular beat.

Programmed stimulation in the electrophysiology laboratory seldom starts torsades de pointes or another sustained ventricular arrhythmia in patients with long-QT intervals, even when they have had spontaneous episodes of polymorphic ventricular tachycardia. However, the arrhythmia can be induced with electrocardiographic form closely resembling spontaneous episodes in most patients with a history of the arrhythmia and no electrolyte disturbances, antiarrhythmic drug therapy or ischaemia. Induced polymorphic ventricular tachycardia may progress to monomorphic ventricular tachycardia or ventricular fibrillation.

Acute treatment of torsades de pointes and polymorphic ventricular tachycardia is seldom necessary. On the occasions when the arrhythmia persists and acutely endangers the patient, cardioversion will usually produce sinus rhythm at least temporarily. Temporary pacing is the most reliable method for suppressing recurrent episodes until more definitive treatment can be applied. Isoproterenol and magnesium are also often effective. When the problem is due to proarrhythmia, the drug causing the arrhythmia must be discontinued. When the arrhythmia persists despite these methods, an implantable cardioverter defibrillator is indicated.

β-Adrenergic blocking drugs and/or left cardiac sympathetic denervation are the preferred methods of preventing torsades de pointes, syncope and ventricular fibrillation in patients with the inherited long-QT syndrome. Occasionally, implantable cardioverter-defibrillators may be needed.

Ventricular fibrillation

Patients who die suddenly in cardiac arrest usually succumb to ventricular fibrillation, the arrhythmia that most frequently causes cardiac arrest and the setting for at least 80% of those who die suddenly outside the hospital. Men die suddenly much more often than women and blacks slightly more frequently than whites. Few who are young die suddenly, the incidence rising rapidly over the age of 45. Cardiac arrest occurs more frequently in patients who are awake, active or highly active, cigarette smokers, obese, less educated, or have survived previous cardiac arrests. Cardiac arrest occurs more often on Mondays than on other days of the week and in the morning rather than at other times of the day or night.

Patients with cardiac arrest over 30 years of age most frequently have coronary heart disease, often involving three vessels, previous myocardial infarction, and reduced ventricular function. About half of survivors of out-of-hospital cardiac arrest have a myocardial infarction. However, ventricular fibrillation develops in relatively

few patients with myocardial infarction once they come under medical care. Consequently, prophylactic administration of lidocaine is no longer recommended for patients with acute myocardial infarction in the absence of specific arrhythmic indications. The incidence of ventricular fibrillation is highest early after the onset of the infarction and occurs most often in those with anterior or large infarctions, congestive heart failure, more diseased coronary arteries, atrial fibrillation, atrioventricular block or bundle branch block. Opening the coronary obstruction that is producing the infarction with thrombolysis or angioplasty reduces the probability that ventricular fibrillation will develop during the hospitalization. Complex ventricular premature beats and ventricular tachycardia during and after myocardial infarction identify those patients most likely to sustain cardiac arrest after discharge.

About one-quarter of patients with dilated cardiomyopathy die suddenly, and cardiac arrest accounts for the majority of deaths in patients with hypertrophic cardiomyopathy. Valvular heart disease produces fewer cardiac arrests in developed countries than in former years; mitral valve prolapse seldom kills suddenly. The death of up to half of patients with congestive heart failure is sudden. Aortic stenosis, congenital anomalies of the coronary arteries, congenital complete atrioventricular block, Ebstein disease, Eisenmenger syndrome and tetralogy of Fallot are the principal congenital lesions producing cardiac arrest and ventricular fibrillation. Certain drugs, including some used to treat arrhythmias, Wolff–Parkinson–White syndrome, strenuous physical activity in healthy-appearing athletes and other young people, exercise testing and pulmonary disease have been associated with sudden death. Idiopathic ventricular fibrillation occurs in some subjects without any objective evidence for cardiac disease. The combination of right bundle branch block, ST segment elevation in leads V1–V3 and cardiac arrest due to ventricular fibrillation constitute an entity known as 'Brugada syndrome' in patients without structural heart disease.

About half of patients resuscitated from cardiac arrest recall no prodromal symptoms. When severe symptoms such as syncope or marked dizziness and weakness are remembered, the arrhythmia is more likely to be ventricular fibrillation; when palpitations or slight dizziness are perceived, ventricular tachycardia. The physical examination reveals an unconscious patient without palpable pulse or audible heart sounds.

A continuous undulating pattern characterizes the electrocardiographic pattern of ventricular fibrillation. The warning arrhythmias that may predict ventricular fibrillation are ventricular premature beats that are multiform or coupled or occur more frequently than 5 per minute or in the early portion of the preceding T wave ('vulnerable phase'). Ventricular tachycardia during myocardial infarction more frequently changes into ventricular fibrillation when it persists for more than 100 beats, or is faster than 180 beats per minute, polymorphic or initiated by an R-on-T beat. Most patients with ventricular fibrillation have abnormal signal-averaged electrocardiograms and heart rate variability. About half of patients with ventricular fibrillation and coronary heart disease have positive exercise tests. Spontaneous conversion occurs in many episodes of ventricular tachyarrhythmias including ventricular fibrillation.

In the electrophysiology laboratory, sustained ventricular tachyarrhythmias, but infrequently ventricular fibrillation, can be induced in the majority of patients successfully resuscitated from cardiac arrest, but less often than in those with stable sustained ventricular tachycardia and more often in those with organic heart disease compared with patients who have idiopathic ventricular fibrillation. Ventricular fibrillation can seldom be induced in survivors of cardiac arrest with dilated cardiomyopathy.

Ventricular fibrillation must be converted quickly if the patient is to recover. The operator should apply thumpversion and, if a pulse does not immediately return, chest compression and artificial respiration. Early electrical defibrillation and electrocardiographic monitoring are vital. If the arrhythmia recurs, intravenous lidocaine, procainamide or amiodarone should be given.

Each patient resuscitated from cardiac arrest should have an electrophysiological evaluation. If the history or the study suggests that ventricular fibrillation was the cause of the cardiac arrest and no reversible cause such as acute ischaemia or taking a pro-arrhythmic drug can be found, a cardioverter-defibrillator should be implanted. Antiarrhythmic drugs alone are no longer considered adequate treatment. Half of those resuscitated from cardiac arrest die in the hospital, and about one-third of those who are discharged die within three years. The implantable cardioverter-defibrillator dramatically reduces mortality in discharged survivors.

Bradyarrhythmias

Other than for sinus bradycardia, often a physiological finding, slow heart rates may indicate that patients have developed atrioventricular block or sick sinus syndrome.

Atrioventricular block

Atrioventricular block of any degree seldom occurs in patients with normal hearts. The P–R interval, the electrocardiographic indicator of conduction from atria to ventricles, lengthens as people age, and children and well-trained athletes may have asymptomatic P–R prolongation. Complete heart block develops most commonly in older patients, more of whom are male than female. Patients with congenital block are more frequently female.

Idiopathic fibrosis of the conducting tissue accounts for most cases of acquired permanent atrioventricular block in adults. Acute myocardial infarction produces complete block in about 7% of patients, most of whom have inferior infarctions often producing the syndrome of right ventricular infarction. Chronic coronary heart disease seldom produces heart block with the notable exception of variant angina due to coronary vasospasm. Atrioventricular block also develops in association with mitral and aortic valve disease, Lyme disease, infective endocarditis, some forms of myocarditis, acute rheumatic fever and the infiltrative cardiomyopathies. Surgical correction of valve disease and of certain congenital anomalies may produce atrioventricular block, but the arrhythmia seldom develops after coronary artery bypass grafting. Many cardioactive drugs as well as hyperkalaemia and hypermagnesaemia may induce block.

Congenital complete atrioventricular block is an uncommon anomaly. Most children who survive have no other structural heart lesions. Mothers of babies with congenital complete heart block have antibodies directed against SSA/Ro or SSB/La antigens.

Syncope is the most characteristic symptom produced by complete atrioventricular block, but complaints arising from congestive failure and decreased cardiac output are frequent. Many patients with congenital heart block have no symptoms.

On physical examination, the dissociation between atrial and ventricular contraction in complete heart block produces large A waves in the venous pulse and variable intensity of the arterial pulse, the first heart sound and systolic murmurs.

The electrocardiogram reveals first-degree atrioventricular block as prolonged P–R intervals. Second-degree block is recognized by occasional nonconduction of P waves in the Wenckebach pattern or, in the Mobitz type II pattern, with fixed P–R intervals in the conducted beats. In complete heart block, the P waves and QRS complexes are dissociated from each other. The ventricular rate is characteristically slow and regular. Narrow QRS complexes suggest that the block is located in the atrioventricular node or within the bundle of His. Wide complexes suggest that the block has been caused by pathology below the atrioventricular junction. The rate of progression to higher degrees of block is unpredictable. Bifascicular block usually precedes the appearance of high degrees of atrioventricular block in patients with chronic acquired conduction system disease. However, most patients with bifascicular block do not develop atrioventricular block. Patients with anterior myocardial infarctions characteristically demonstrate bundle branch or bifascicular block before they develop complete block.

The clinical electrophysiological study can define the level of atrioventricular block, which has important clinical implications. Block within the atrioventricular node may be transient and is unlikely to progress to high degrees of atrioventricular block and syncope. Patients with partial block within or below the bundle of His often develop complete block and potentially fatal symptoms.

Drugs are seldom used for chronic treatment of atrioventricular block although atropine and adrenergic agonists may temporarily decrease the degree of block. Cardiac pacemakers, many of which change their rates in response to physiological requirements, are the preferred method for treating block that produces symptoms or threatens the patient's survival. Once pacemaking has been established, the survival of patients with acquired atrioventricular block depends primarily upon the severity of the accompanying cardiac and noncardiac disease. The mortality when high degrees of atrioventricular block complicate inferior myocardial infarction is at least three times greater than in the absence of this arrhythmia. However, heart block adds no risk to survivors of inferior myocardial infarction. At least three quarters of those with anterior infarction and acute atrioventricular block die. The prognosis is excellent for those children with congenital complete heart block who survive infancy and have no structural heart disease.

Sick sinus syndrome

Sick sinus syndrome encompasses those bradyarrhythmias due to malfunction of the sinus node that produce troubling or disabling symptoms. Many patients with sick sinus syndrome also have supraventricular tachyarrhythmias (bradycardia–tachycardia syndrome) and dysfunction of the normal escape mechanisms.

Although sick sinus syndrome affects a few children and young adults, the greatest frequency occurs in the sixth and seventh decades of life. Men and women are equally affected. Sick sinus syndrome is usually an acquired condition, but a few cases have a genetic basis. Patients with sick sinus syndrome most often have coronary heart disease; a few have cardiomyopathy. Many, however, have no structural heart disease. Among children, cardiac surgery to repair congenital lesions, most frequently the Mustard procedure for transposition of the great arteries, produces most of the cases of sick sinus syndrome. Some adults develop sick sinus syndrome from surgical closure of atrial septal defect. Sick sinus syndrome also appears in the donor heart of some patients after cardiac transplantation and occasionally after the Maze operation for atrial fibrillation. The function of the sinus node may be so affected by a long period of chronic atrial fibrillation that an adequate sinus rhythm may not appear following cardioversion. Vagotonia can transiently produce the bradyarrhythmias of sick sinus syndrome. The effects of drugs the patient is taking that slow the heart rate, such as β-adrenergic and calcium channel blocking drugs, must always be considered before making the diagnosis of sick sinus syndrome. Stroke and embolism are important

complications of the bradycardia–tachycardia syndrome but not of sick sinus syndrome without tachyarrhythmias.

Bradyarrhythmias cause lightheadedness, dizziness and fainting, the principal complaints of patients with sick sinus syndrome. Abnormal neural responses rather than bradyarrhythmias alone, however, contribute to much of the syncope that affects many patients with sick sinus syndrome. Palpitations suggest that the patient has the bradycardia–tachycardia syndrome.

The standard or extended ambulatory electrocardiograms of patients with sick sinus syndrome characteristically show sinus bradycardia, sinoatrial block, sinus pauses, sinus arrest or atrial standstill. Atrioventricular block may appear at the onset of sick sinus syndrome or develop later in the course of the disease. The most common sustained tachyarrhythmia of patients with the bradycardia–tachycardia syndrome is atrial fibrillation. Sick sinus syndrome limits the heart rate that patients can achieve when exercising.

On electrophysiological evaluation, sinoatrial conduction time, sinus node recovery time and refractoriness of the sinus node and atria are characteristically prolonged. The duration of the atrial electrograms are prolonged and their appearance is fractionated in many patients with sick sinus syndrome, particularly in those who also have atrial fibrillation. Electrophysiological studies show nonclinical atrioventricular conduction disease in many patients with sick sinus syndrome. However, only those with severe abnormalities are likely to develop atrioventricular block in the future.

Pacing is the most successful treatment for the symptoms produced by the bradyarrhythmias of sick sinus syndrome. However, one must be certain that the arrhythmias produce the symptoms before prescribing a pacemaker. When compared with ventricular pacing, atrial or atrioventricular pacing seems to reduce the incidence of atrial fibrillation, stroke and possibly congestive heart failure. Atrioventricular pacing also protects against the development of atrioventricular block and, consequently, has become the preferred pacing mode for most patients.

Use of appropriate pacemakers makes the prognosis of patients with sick sinus syndrome and no structural heart disease indistinguishable from that of the otherwise healthy general population. Coexisting disease accounts for any excess mortality. Patients who receive atrial or atrioventricular rather than ventricular pacers survive longer.

Further Reading

Kastor JA (2000) *Arrhythmias*, 2nd edn. Philadelphia: WB Saunders.

Zipes DP and Jalife J (2000) *Cardiac Electrophysiology from Cell to Bedside*, 3rd edn. Philadelphia: WB Saunders.