Postoperative Arrhythmias in Patients with Congenital Heart Disease*

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Improvement in cardiac surgical techniques has resulted in increased longevity and improvement in the quality of life for most patients with congenital heart disease who underwent open-heart repair. At present, virtually all patients, even patients with the most complex congenital malformations, are subjected to physiologic or anatomic repair. In recent years, much attention has been focused on postoperative arrhythmias evident in this population of patients. Indeed, with increasing years of follow-up, it seems evident that ever-increasing numbers of patients who underwent surgery in infancy or childhood manifest different forms of cardiac electrical instability. In most patients the arrhythmias are a direct result of surgical injury to the various parts of the specialized conduction system. In other patients, cardiac arrhythmias result from marked myocardial hypertrophy and scarring, which in some patients may be associated with ischemia or cyanosis. Because open-heart repair of different forms of congenital heart disease is carried out in different parts of the heart and close to different sections of the specialized conduction system, the injury to the conduction system may vary with different operations.

This report will detail the mechanism, pathologic findings, clinical course, and management of postoperative cardiac arrhythmias which result from surgical injury in the pediatric population.

Sinus Node Injuries

Injuries to the sinus node occur most often following repair of transposition of the great arteries by the Mustard or Senning operations; however, it may occur following any type of open-heart surgery and has been reported after surgery for tetralogy of Fallot, atrial septal defect, and other forms of congenital heart disease. The injury may be transient, as seen following cannulation of the superior vena cava with the superior vena caval tape and applying excessive pressure to the sinus node. This type of injury may result in an inappropriate low sinus rhythm following surgery, with or without junctional escape, and usually resolves spontaneously within a few days following the operation. Permanent damage to the sinus node may result from incision of the sinus node or placement of sutures either through the sinus node or in its vicinity, such as often occurs following the Mustard procedure. Interruption of the blood supply to the sinus node may also lead to degeneration and fibrosis of the approaches to the sinus node or the node itself. In such patients the clinical manifestation may not be apparent for years following the operation.

After surgery in pediatric patients, the clinical manifestations usually include inappropriate sinus bradycardia, sinoatrial block, evidence of sinus standstill with or without atrioventricular junctional escape and, less frequently, the tachycardia-bradycardia syndrome and paroxysmal atrial fibrillation. These clinical manifestations may appear immediately following surgery or late (that is, years following the open-heart repair). Some patients may demonstrate these abnormalities intermittently, usually as excessive bradycardia during sleep, with normal sinus rhythm appearing during waking periods and exercise. Others will manifest complete sinus arrest, with junctional rhythms throughout their daily activities. Unless the cardiac rhythm is extremely slow or additional hemodynamic abnormalities exist, bradycardias are usually well tolerated, and the patients remain essentially asymptomatic from a cardiovascular standpoint. With maturation the cardiac rhythm decreases, and many patients with permanent low cardiac rates may eventually require treatment with a cardiac pacemaker (Fig 1).

The work-up of patients who are likely to manifest evidence of injury to the sinus node, particularly patients who have undergone surgery for transposition of the great arteries, should include electrocardiographic evaluation of cardiac rhythms during follow-up visits and 24-hour ambulatory electrocardiographic recordings in children with a higher rate of suspicion. The extent of malfunction of the sinus node may be tested by exercise or administration of atropine.

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Figure 1. Spectrum of cardiac rhythms seen at different times in patient with transposition of great arteries two years after Mustard's operation. A, Sinus rhythm; B, junctional rhythm at sleep; C, supraventricular tachycardia; and D, marked sinus arrhythmia.

(0.01 to 0.015 mg/kg of body weight, to a maximum of 0.4 mg) or isoproterenol. In patients with the sick sinus syndrome, the expected acceleration of sinus rhythm does not occur or may be rather sluggish. The function of the sinus node may also be studied during cardiac catheterization. Such studies include the response of the sinus node to atrial pacing, measurement of sinoatrial conduction time, and the recovery time of the sinus node. Patients with the sick sinus syndrome who manifest alternately tachycardia and bradycardia and who require medical management for control of the tachycardia may also require insertion of a cardiac pacemaker, because any measures to reduce the tachycardia may induce cardiac asystole.

 Interruption of the internodal pathways has been suggested to occur following the Mustard operation;6,11 such injury is likely to cause changes in P-wave morphology and possibly may result in supraventricular arrhythmias; however, no clear-cut clinical patterns related to injury or interruption of internodal pathways has so far been recognized.

Junctional Injuries

Injury to the atrioventricular node and the His bundle may result from surgical procedures in proximity to these structures. Injuries to the atrioventricular node may occur due to the placement of sutures to secure the lower part of the atrial baffle in the Mustard operation6,8 and following repair of various forms of endocardial cushion defects. Injury to the branching part of the His bundle may occur when sutures are required to repair membranous ventricular septal defects in patients with atrioventricular concordance. In such patients the His bundle is close to the postero-inferior aspect of the defect; however, such injury is more likely to occur in patients with atrioventricular discordance in whom the atrioventricular node and the His bundle are unusually located, with the His bundle traversing the superior anterior aspect of the ventricular septal defect. Similarly, injury to the His bundle may occur in patients with complex intracardiac malformations in whom the atrioventricular node and His bundle may be unusually located.6,11 The clinical manifestation of junctional injury may be either junctional tachycardia or evidence of increased atrioventricular block.

Atrioventricular Junctional Tachycardia

This form of supraventricular tachycardia is seen most often during the immediate postoperative period. Infrequently, atrioventricular junctional tachycardia occurs many years following repair of congenital heart disease. The hemodynamic effects result from the lack of atrial contribution to cardiac output, as well as from the inappropriately high ventricular rates which interfere with diastolic filling. The rate of junctional tachycardia may vary from 120 to 280 beats per minute; and when rapid ventricular rates are coupled with early postoperative borderline hemodynamics or with residual defects, the net result may be devastating.

The electrocardiogram usually shows a narrow QRS complex with absent P waves. Occasionally, the QRS complex is wide because of intraventricular conduction defects, and the differential diagnosis of ventricular tachycardia may be impossible without further electrophysiologic studies. Rate-related atrioventricular block in pediatric patients is rare because the atrioventricular conduction system in pediatric patients is capable of transmitting fast atrial or junctional rhythms.

Management of junctional tachycardia, particularly in the immediate postoperative period, may be extremely difficult. Junctional tachycardia responds poorly to vagal maneuvers and usually does not respond well to administration of digoxin; however, digoxin or propranolol may reduce the abnormal junctional rate or induce second-degree atrioventricular block, thus effectively halving the ventricular response. If unstable hemodynamic conditions exist, cardioversion should be attempted, but the abnormal rhythm may recur. If atrial wire electrodes or ventricular electrodes exist, they may be extremely helpful in the management of patients with junctional tachycardia in the immediate postoperative period.6,11 Such wires are helpful in determining precisely the origin of
the arrhythmia, as well as in its management by premature stimulation or overdrive pacing. After termination of the arrhythmia, many patients will require supplemental drug therapy, usually in the form of digoxin, to prevent recurrence. Continuous rapid atrial pacing for the infrequent patient whose condition does not respond well to electrocardioversion or entrainment of the arrhythmia has also been described.

**Atrioventricular Conduction Defects**

Injury to the atrioventricular node or His bundle or, on rare occasions, to both the proximal part of the right and the left bundle branch simultaneously may result in different forms of atrioventricular block. Surgical damage to this area results from the placement of sutures around the specialized conduction system at these sites or the interruption of the blood supply to these components of the specialized cardiac conduction system. Early damage to function results from ischemia, hemorrhage, and necrosis; and eventually, some further damage may occur due to fibrosis and healing. Similar to other injuries the clinical manifestation of such injury may be partial, with a low degree of atrioventricular block, or complete interruption of atrioventricular conduction. The clinical manifestations may also be transient (that is, only in the immediate postoperative period) or permanent. In some patients who have undergone open-heart surgery for congenital heart disease, evidence for junctional injury in the form of atrioventricular block has developed many years following their surgery.

Fortunately, in recent years, because of a better understanding of the location of the specialized conduction system, the incidence of complete heart block has decreased very significantly. In our institution, permanent complete heart block nowadays occurs very rarely, even in the more complex congenital heart diseases.

The decrease in surgical complete heart block results from improved knowledge of the precise location of the specialized conduction system from very meticulous histologic and electrophysiologic studies.

Since the early and late mortality from untreated high-degree atrioventricular block in patients after surgery is high, therapy with a pacemaker is currently indicated in all pediatric patients who manifest postoperative high-degree atrioventricular block. This is particularly true in patients with clinical manifestations such as dizziness or blackout spells or clear Stokes-Adams attacks. Because of the decreased incidence of atrioventricular block in pediatric patients, therapy with a pacemaker has been used less frequently in recent years. In patients treated with cardiac pacemakers, the rate of survival has been improved and so has been the care of such patients with improvement in pacemaker technology.

In most patients, transient atrioventricular block resolves within three weeks following surgery. Treatment with a pacemaker should be considered in all patients in whom normal atrioventricular conduction does not recur during this time. Patients with early postoperative complete heart block require either ventricular wires placed prior to closure of the chest or, if the atrioventricular block occurred after the chest had been closed, insertion of transvenous endocardial wires.

Patients with transient complete heart block have been shown to develop late complete heart block, particularly patients with transient complete heart block in whom right bundle-branch block and left anterior hemiblock are present when atrioventricular conduction resumes. This electrocardiographic pattern following transient complete heart block indicates an extensive residual damage to the branching portion of the His bundle and additional injury to the right bundle branch and to the anterior fibers of the left bundle branch.

**Intraventricular Conduction Defects**

Postoperative intraventricular conduction defects lead to the development of different electrocardiographic patterns, such as the pattern of right bundle-branch block, the pattern of right bundle-branch block and left anterior hemiblock ("bifascicular block"), and the pattern of right bundle-branch block, left anterior hemiblock, and PR prolongation ("trifascicular block pattern").

**Right Bundle-Branch Block**

The pattern of right bundle-branch block occurs in almost all patients following repair of tetralogy of Fallot and in almost all patients following repair of complete atrioventricular canal. The pattern occurs less often following repair of ventricular septal defect or ostium primum defects. The pattern of right bundle-branch block occurs in almost all patients with complex congenital malformations in whom a membranous ventricular septal defect had to be closed during the intraoperative repair or who have pulmonary infundibular resection as part of their corrective surgery. The injury to the right bundle branch may occur along its entire course (that is, in its proximal part close to the ventricular septal defect), as the right bundle tranverses the moderator band, or to the right bundle-branch distal ramification. The injury occurs because of the placement of sutures, hemorrhage, necrosis, and inflammatory changes resulting in scarring and fibrosis of the right bundle branch.

Electrocardiographic evidence of injury to the right bundle branch is apparent immediately at the end of the intracardiac repair. There are no clinical or hemodynamic consequences to this injury. In patients with
proximal right bundle-branch block who may develop a left bundle-branch block due to the atherosclerotic process later in life, the risk of complete heart block may be real."

**Right Bundle-Branch Block and Left Anterior Hemiblock**

The electrocardiographic pattern of a right bundle-branch block associated with a counterclockwise and superiorly oriented frontal QRS complex reflects injury to the branching part of the His bundle or to the proximal part of the right bundle branch and additional injury to some of the left bundle-branch fibers oriented towards the anterior superior part of the left ventricle. The pattern occurs in 8 to 22 percent of the patients undergoing surgery for tetralogy of Fallot and results from a somewhat more extensive injury but quite similar to the injury which causes the appearance of a proximal right bundle-branch block.

It has been suggested that many patients with right bundle-branch block and left anterior-hemiblock may be prone to late development of complete heart block or sudden death. A review of the literature revealed that only one of 204 patients developed a late complete heart block, and four died suddenly, for a total occurrence of late complications of 2 percent. The varying reports regarding the long-term prognosis for patients with right bundle-branch block and left anterior hemiblock may stem in part from the different surgical techniques and possibly from the different sites of injury, resulting in the development of two populations of patients. The first, with injury to the branching part of the His bundle, may have a higher likelihood for late development of complete heart block and sudden death. The second population of patients, in whom the injury occurred to the proximal parts of the right bundle with injury to some fibers of the left bundle branch, has a decreased likelihood of developing late complete heart block or dying suddenly.

Electrocardiographic tracings are advised in the follow-up of such patients after surgery, but no other treatment is needed.

**Right Bundle-Branch Block, Left Anterior Hemiblock, and PR Prolongation**

In postoperative patients, this electrocardiographic pattern ("trifascicular block pattern") reflects extensive damage to the branching part of the His bundle and possibly the proximal part of the right bundle and left bundle branches (Fig 2). The PR prolongation may be due either to extensive injury to the His bundle or the atrioventricular node. Thus, this electrocardiographic pattern reflects injuries which may be located from the atrioventricular node to the proximal part of the right and left bundle branches (Fig 3). In most patients with congenital heart disease, surgery is carried out through the right ventricle; it is therefore very unlikely that injury to the posterior fibers of the left

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**Figure 2.** Six frontal electrocardiographic leads in patient following repair of tetralogy of Fallot. Note PR prolongation (PR-0.26 seconds), QRS prolongation, and superiorly oriented frontal QRS loop, consistent with pattern of "trifascicular block."

**Figure 3.** Schematic drawing of junctional area of specialized conduction system. PR prolongation may result from injuries to atrioventricular (A-V) node or penetrating part of His bundle (or both). Intraventricular conduction defects may result from injuries to branching part of His bundle or right and left bundle branches. Combined injuries or extensive injuries to atrioventricular node and penetrating part of His bundle and additional injuries to branching part of His bundle and bundle branches may result in complex electrocardiographic patterns such as "trifascicular block." Such a pattern in pediatric patients after surgery almost invariably indicates marked injury to junctional area of specialized conduction system, rather than injury to distal bundle branches.
bundle branch will occur, since they are remote from the surgical field. Therefore, the term, " trifascicular block," is a misnomer when applied to the electrocardiographic pattern of patients following open-heart repair for congenital heart disease.

In pediatrics, this electrocardiographic pattern occurs infrequently following open-heart surgery. The limited available follow-up data suggest that such patients may be at a higher risk for late development of complete heart block and therefore may require more careful follow-up care. 40

**Ventricular Irritability and Arrhythmia**

Ventricular arrhythmias have recently been recognized with increasing frequency in patients following repair of congenital heart disease. 41-45 Many patients with complex congenital heart disease who undergo palliative surgery also show evidence of premature ventricular contractions in late adolescence and young adulthood. It has been reported that ventricular arrhythmias have accounted for sudden death in 30 to 38 percent of the patients who underwent open-heart surgery with "complete" repair and had evidence of premature ventricular contractions on ECGs, ambulatory electrocardiographic testing, or stress ECGs. 46-48 Premature ventricular contractions are seen in 5 to 18 percent of the patients with tetralogy of Fallot following open-heart surgery. If ambulatory electrocardiographic studies or stress ECGs are done, more patients will be found to have ventricular arrhythmias. The risk of sudden death is particularly of concern in patients with evidence of ventricular irritability and right ventricular hypertension or right ventricular dysfunction (or both). Currently, ventricular irritability, rather than conduction defects, is considered to be the major mechanism for late morbidity and mortality following repair of tetralogy of Fallot.

The likelihood of developing ventricular arrhythmias is increased in patients with a right ventriculotomy, older age at the time of surgery, and residual intracardiac defects.

It appears that postoperative premature ventricular contractions and ventricular tachycardia result from a reentrant mechanism in most patients, since the condition can be initiated and terminated with programmed electrical stimulation. 49 Electrograms recorded during electrophysiologic studies in patients with ventricular arrhythmias close to the site of the right ventriculotomy show prolongation and fragmentation of the electrograms. This is in contrast to electrograms from patients without evidence of ventricular irritability, in whom the electrograms are shorter and without evidence of fragmentation.

The work-up of patients who are likely to manifest ventricular arrhythmias should include a 12-lead ECG, ambulatory ECG, and stress ECG. A chest roentgenogram may occasionally show evidence of a right ventricular outflow aneurysm and calcification of the right ventricular outflow tract. A two-dimensional echocardiogram is useful to document right ventricular dysfunction and to rule out associated cardiac defects. Hemodynamic and electrophysiologic studies are important in all symptomatic patients in whom the symptoms may be due to ventricular arrhythmias. Such studies will identify the extent of residual defects, the ease with which ventricular arrhythmias may be initiated, the precise location and origin of such arrhythmias, and the efficacy of the antiarrhythmic treatment. In some patients, such electrophysiologic studies may reproduce the patient's symptoms. In contrast to the adult population with atherosclerotic heart disease, electrophysiologic induction of sustained ventricular tachycardia is difficult in the pediatric population, and most patients demonstrate premature ventricular contraction or, rarely, unsustained ventricular tachycardia.

Management of pediatric patients with ventricular arrhythmias after surgery depends on the severity of the ventricular arrhythmia, associated hemodynamic defects, the clinical symptoms, and the presence of additional cardiac malformations. Isolated uniform premature ventricular contractions in asymptomatic children with normal hemodynamics in whom higher degrees of ventricular arrhythmias have been ruled out do not require antiarrhythmic treatment. Patients with multifocal premature ventricular contractions and couplets and certainly patients with evidence of higher degrees of ventricular arrhythmias with or without associated malformations should be treated with antiarrhythmic medications.

Ventricular tachycardia should be treated promptly and can be controlled either with medical management by intravenously administering lidocaine or, in the presence of borderline hemodynamics, with electrical cardioversion (1 to 2 W/sec/kg). Occasionally, patients with ventricular arrhythmia associated with sinus bradycardia may benefit from administration of atropine or ventricular pacing to increase the cardiac rate. Long-term suppression of ventricular arrhythmias after surgery in pediatric patients may be achieved with drugs such as quinidine, propranolol, or phenytoin. Indeed, phenytoin has been shown to be very effective in the treatment of pediatric patients with ventricular arrhythmias and residual hemodynamic abnormalities after surgery. 50,51 In such patients, phenytoin may be superior to other antiarrhythmic medications because of its relative lack of negatively inotropic effects. Rarely, repeat surgery for excision of an arrhythmogenic focus located at the right ventricular outflow tract may be necessary; however, a surgical trial to abolish the arrhythmogenic circuit should be attempted in all patients with residual
postoperative cardiac arrhythmias. The recognition of these anomalies is critical, as they can lead to 10 reversible rhythm disturbances or premature ventricular contractions.

The prognosis of patients with mixed patterns of cardiac arrhythmias depends on the specific arrhythmia severity and the arrhythmias responsible for clinical symptoms. Management of patients with mixed arrhythmias is therefore targeted to each singular significant pattern of concern.

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