Arrhythmias in the Newborn
Anne M. Dubin, MD*

OBJECTIVES

After completing this article, readers should be able to:

1. Describe the most common arrhythmia seen in neonates.
2. Delineate the patients for whom radiofrequency ablation of supraventricular tachycardia is reserved.
3. State the most common cause of congenital complete heart block in an infant who does not have structural heart disease.
4. Describe the indications for pacemaker placement in congenital complete heart block.

Introduction

Identifying and treating arrhythmias in neonates differs substantially from approaches used in an older child and can be challenging. Neonatal sinus tachycardia can be very rapid and may be difficult to distinguish from other arrhythmias. The natural history of arrhythmias in the neonatal age group also differs markedly from other ages and must be considered when planning a treatment strategy.

Incidence and Prevalence

Arrhythmias are found in 1% to 5% of newborns during the first 10 days of life. Most are premature supraventricular beats that will disappear over the first month of life. The most common symptomatic arrhythmia in the neonatal period is supraventricular tachycardia (SVT), which has an incidence of 1/25,000.

Bradycardias

FIRST-DEGREE HEART BLOCK

First-degree heart block is defined as a PR interval greater than the upper limit of normal for age (0.16 sec on day of life 1, 0.14 sec for the remainder of the newborn period). A prolonged PR interval can be caused by delayed conduction in the atrium, atrioventricular (AV) node, or His-Purkinje system. It usually is seen with disorders of the AV node and commonly is associated with congenital heart disease or inflammatory disorders of the heart. Medications that increase vagal tone also may increase the PR interval. Most patients are asymptomatic and need no further therapy.

SECOND-DEGREE HEART BLOCK

Second-degree heart block is characterized by intermittent failure of conduction to the ventricle. It is classified further into Mobitz type I or II block. Mobitz type I or Wenckebach block is due to blockage in the AV node. Its characteristic appearance on electrocardiography (ECG) is gradual prolongation of the PR interval with eventual failure of conduction and dropped beat. This type of block generally is related to medications or maternal connective tissue disease. Mobitz type II block, which is more ominous, is due to blockage in the distal conducting system. There is no lengthening of the PR interval prior to the blocked beat. Pacemaker placement is required.

LONG QT WITH 2:1 BLOCK

Long QT syndrome, an abnormality of repolarization of the ventricle, can present with an extremely long QT interval and associated 2:1 AV block due to ventricular refractoriness. Affected patients are at high risk of sudden death from torsades de pointes. They require high-dose beta blockade and pacemaker implantation.

COMPLETE HEART BLOCK

Complete heart block (CHB) is characterized by no conduction from the atrium to the ventricle. The atrial rate is higher than the ventricular rate, and P waves have no relationship to the QRS complexes (Fig. 1). This arrhythmia has been associated with maternal connective tissue disease or structural heart disease.

CHB in most patients who have structurally normal hearts is believed to be due to exposure to anti-Ro and anti-La antibodies in utero. These antibodies are common in women who have systemic lupus erythematosus or Sjögren syndrome. In animal models, these antibodies, which cross the placenta in the second trimester of pregnancy, attack the myocardium and conduction system, causing a myocarditis-like picture and conduction abnormalities. Preliminary work suggests that early maternal steroid administration may protect the fetus. Newborns who are symptomatic, have a wide-complex escape rhythm, or have ventricular rates less than 50 beats/min should undergo permanent pacemaker implantation.

The combination of congenital heart disease and congenital CHB is associated with a mortality rate of 29% in the neonatal period. Corrected transposition of the great arteries and left atrial isomerism have been related to CHB in the newborn.

Supraventricular Arrhythmias

PREMATURE ATRIAL CONTRACTIONS

Premature atrial contractions represent the most common arrhythmia seen in the neonatal period. They may be conducted normally or aberrantly or completely blocked. They are benign in the structurally normal heart. In patients who have central venous lines, mechanical irritation of the right atrium may cause premature atrial contractions. The association of atrial septal aneurysms and premature atrial contractions in the fetus and the newborn is considered controversial.
ATRIOVENTRICULAR RE-ENTRANT TACHYCARDIA

Atrioventricular re-entrant tachycardia is the most common cause of tachycardia in the newborn period. It accounts for 50% of all pediatric patients who have SVT. A bypass tract that either can conduct in the antegrade and retrograde direction (also known as Wolff-Parkinson-White [WPB] syndrome) or solely in the retrograde direction (concealed pathway) is mandatory for this type of tachycardia. Tachycardia usually is initiated with a premature atrial or ventricular contraction. This allows for unidirectional block (usually in the accessory pathway, but occasionally in the AV node) and the start of a re-entrant circuit. In orthodromic tachycardia, this is exhibited by antegrade conduction across the AV node and retrograde conduction up the bypass tract.

To differentiate atrioventricular re-entrant tachycardia from other forms of SVT, it is important to identify the P-wave axis and location during tachycardia. In atrioventricular re-entrant tachycardia, a retrograde P wave (negative in lead II and avF) usually can be identified following the QRS complex. Transesophageal recordings may be helpful in elucidating a P wave buried in T waves. Atrioventricular re-entrant tachycardia requires both atrial and ventricular tissue to support the re-entrant circuit. Therefore, AV block or dissociation cannot occur during this tachycardia. Adenosine, which causes transient AV block, can help to differentiate SVTs. WPW syndrome may be identified on a baseline ECG by the characteristic short PR interval and slurred upstroke of the QRS complex, which is also known as the delta wave (Fig. 2).

Atrioventricular re-entrant tachycardia is seen most commonly in neonates who have structurally normal hearts. However, WPW syndrome has been associated with Ebstein malformation of the tricuspid valve as well as corrected transposition and hypertrophic cardiomyopathy.

Atrioventricular re-entrant tachycardia may be terminated acutely by eliciting a dive reflex with ice over the face for 30 seconds. Other vagal maneuvers, including gagging and rectal probe, also have been effective. Eyeball pressure should not be used. Rapid intravenous infusion of adenosine also will terminate atrioventricular re-entrant tachycardia. If the patient appears unstable, synchronized cardioversion should be performed. Esophageal overdrive pacing also has been effective in the neonate. Pacing is instituted at a rate 10% faster than the tachycardia rate and continued for 10 to 15 seconds. Cardioversion should be available during this procedure.

Because the previously mentioned maneuvers are only useful acutely, the clinician must plan chronic therapy to keep the patient out of tachycardia. Digoxin and beta blockers are considered first-line agents to prevent recurrence of atrioventricular re-entrant tachycardia. Class IA, IC, and III antiarrhythmics also have been used if the first-line agents fail. Most patients may be weaned from therapy within 1 year. Calcium channel blockers should be avoided in the neonate because there have been several reports of cardiovascular collapse following their administration in these infants. It has been hypothesized that the immaturity of the sarcoplasmic reticulum in the infant myocardium makes the neonate particularly dependent on calcium channels.

It is well established that patients who present with WPW syndrome...
or concealed accessory connections in infancy have a high probability of spontaneous resolution of SVT during the first year of life. Studies of children who have rapidly conducting accessory connections and present in infancy estimate the chance of resolution at 60% to 90%. However, approximately one third of patients who present in infancy with WPW syndrome and have spontaneous resolution by 1 year of age will experience re-onset of symptoms later in life, typically at approximately 4 to 6 years of life.

Radiofrequency ablation has been used in infants under rare circumstances. Several studies have examined the effect of radiofrequency energy on immature myocardium. Lesions created by radiofrequency in infant lambs were much deeper than those in studies of adult animals. Further, the lesions increased in size as the animals grew. Sudden death also has been reported in an infant after radiofrequency ablation of an incessant tachycardia. It was hypothesized that the ablation lesions were a focus for proarrhythmia, and the infant died of ventricular tachycardia. For these reasons, radiofrequency ablation is reserved for the infant who has failed all possible drug combinations or who has extremely poor cardiac function due to arrhythmia.

The permanent form of junctional reciprocating tachycardia is a relatively rare form of re-entrant tachycardia. It can present in utero or during the neonatal period and is characterized by a long R-P interval that represents a slowly conducting retrograde pathway. It is extremely difficult to control; affected infants commonly develop ventricular dysfunction and heart failure. They are a management challenge and frequently require amiodarone therapy to control the arrhythmia.

**ATRIAL FLUTTER**

Atrial flutter is a relatively uncommon rhythm of the fetus and neonate. As mentioned previously, it often is associated with high morbidity and mortality in utero. It is characterized by a saw-tooth pattern of P waves at rates of 300 to 600 beats/min. The ventricular response rate may be regular or irregular. It has been associated with both normal cardiac structure and congenital heart disease. It is important to rule out mechanical causes of flutter due to central venous lines.

Mortality is increased when atrial flutter does not respond to medications and when it is associated with congenital heart disease. Therapy consists of esophageal atrial overdrive pacing or direct synchronized cardioversion. Digoxin is usually the first-line drug for chronic therapy.

**CHAOTIC ATRIAL TACHYCARDIA**

Chaotic atrial tachycardia is an uncommon arrhythmia in the neonatal period. It is characterized by an irregular narrow-complex tachycardia in which at least three distinct P wave morphologies can be seen. This tachycardia appears to resolve as the conduction system matures.

**CONGENITAL JUNCTIONAL ECTOPIC TACHYCARDIA**

Congenital junctional ectopic tachycardia (JET) is characterized by a narrow QRS tachycardia with AV dissociation and an atrial rate that is slower than the ventricular rate. (Fig. 3) It is usually incessant and has been associated with tachycardia-induced cardiomyopathy. Villian and colleagues documented a mortality rate of 35% in affected patients. Approximately 50% of patients will have an affected family member. These tachycardias do not respond to cardioversion or over-
drive pacing. Class III antiarrhythmics have been found to be the most efficacious therapies.

**Ventricular Arrhythmias**

**PREMATURE VENTRICULAR CONTRACTIONS**

Premature ventricular beats are relatively common in the neonate. One investigation documented premature ventricular contractions (PVCs) in 18% of newborns who had 24-hour Holter monitoring on the first day of life. The incidence of PVCs ranged from 4 to 50 beats/24 h. The following criteria may be used to recognize PVCs on ECG: 1) early onset of the electrical complex, 2) QRS duration greater than 80 msec, 3) abnormal QRS morphology with ST and T wave changes, and 4) absence of a preceding P wave. Most infants who have PVCs have structurally normal hearts. These infants do not require pharmacologic therapy unless complex ectopy (eg, multiform PVCs or couplets) is seen. Babies who have heart disease (myocarditis, tumors, or structural heart disease) require a further evaluation of frequent PVCs.

**ACCELERATED IDIOPATHIC VENTRICULAR RHYTHM**

Accelerated ventricular rhythm occurs at rates similar to the sinus rates recorded just before the onset of the arrhythmia. The majority of these rhythms are less than 200 beats/min. The QRS duration is prolonged (>80 msec), and the rhythm generally has a left bundle branch morphology and fuses in and out of sinus rhythm. It appears to be a phenomenon of the neonatal period that disappears early in postnatal development. It is a benign arrhythmia, infants experience no cardiovascular compromise, and they require no further evaluation or therapy.

**VENTRICULAR TACHYCARDIA**

Ventricular tachycardia (VT) in the neonate is a relatively uncommon event, but if it does occur, this arrhythmia must be investigated further and usually requires therapy. VT is diagnosed when three or more ventricular beats occur in sequence. The differential diagnosis of a wide-QRS tachycardia includes VT, rate-related aberration, and persistence of an underlying conduction defect (eg, WPW syndrome or underlying bundle branch block). Several factors can help distinguish VT: 1) the presence of fusion and capture beats (sinus beats that fuse with the ventricular beat or sinus beats in the midst of a VT), 2) AV dissociation, and 3) morphology similar to PVCs. Unfortunately, the absence of AV dissociation in the neonate does not rule out VT because infants tend to have 1:1 retrograde conduction. VTs are characterized as monomorphic or polymorphic and nonsustained (from 3 beats to 30 sec) or sustained. Incessant VT has been defined as an arrhythmia that lasts for more than 10% of the day.

Several etiologies of VT must be considered, including myocarditis, tumors, VT after myocardial infarction, electrolyte abnormalities, metabolic abnormalities, drug toxicity, and long QT syndrome. Myocarditis may have an initial presentation as VT. Affected infants are seriously ill, with poorly tolerated incessant arrhythmias, congestive heart failure, and even cardiac arrest. They usually have polymorphic incessant VTs. Myocardial function generally is severely depressed. These infants have an extremely poor prognosis. Intravenous amiodarone has been effective in only 50% of affected children.
Extracorporeal membrane oxygenation has been used if the child can be stabilized sufficiently to allow cannulation.

Harmatomas have been associated with ventricular arrhythmias. Garson and colleagues described the presentation and therapy of 21 children who had these tumors. All had incessant VT at an average rate of 260 beats/min, and the majority were symptomatic. Most had a right bundle branch pattern VT. All had normal echocardiographic and angiographic findings. A total of 19 children survived, and a definitive pathologic lesion was documented in 15. Twelve had myocardial harmatomas, two had rhabdomyomas, and one had myocarditis.

Rhabdomyomas rarely can cause VT or SVT in infancy. The natural history of these tumors is slow regression, with disappearance of the arrhythmia by the age of 2. Therefore, therapy is considered a temporizing measure.

Myocardial infarctions are rare in infancy. They are seen in patients who have anomalous origin of the left coronary artery from the pulmonary artery, thromboembolism, or a history of maternal cocaine or heroin use. This VT is polymorphic, unstable, and associated with a high mortality rate.

Electrolyte abnormalities also may be associated with VT. Hypokalemia, especially in newborns receiving digoxin, can cause VT. Hypocalcemia and hypomagnesemia have been associated with ventricular arrhythmias.

Clinical examination, family history, ECG, and echocardiography usually are adequate for evaluating the infant who has VT. Magnetic resonance imaging has been used to help visualize tumors, but often they can be seen only at surgery. Electrophysiologic study should be considered only in those in whom the diagnosis is unclear or who have failed medical therapy.

Patients who have hemodynamic compromise should undergo immediate synchronized cardioversion or defibrillation. Nonsustained VT that is well-tolerated can be treated with lidocaine or amiodarone. As stated previously, neonates who have structurally normal hearts usually experience spontaneous resolution of ventricular arrhythmias. Therefore, therapy should be reserved for incessant or sustained VT or for rapidly conducting nonsustained tachycardia.

Chronic therapy is based on the underlying etiology of the arrhythmia. Beta blockade and class IA, IB, IC, and III antiarrhythmics all have been used in VT of the newborn with varying success.

LONG QT SYNDROME

Long QT syndrome in infancy is a known cause of sudden death and is associated both with bradycardia and torsade de pointes VT. The diagnosis is based on a family history and a prolonged QT interval on the resting ECG (>0.45 sec) (Fig. 4). The molecular genetics of this disease only recently have been elucidated. Long QT syndrome has been associated with at least three identifiable molecular abnormalities, two potassium channels (LQT1 and LQT2) and one sodium channel (LQT3). Recently, Zareba and colleagues have shown a relationship between the molecular abnormality and the phenotypic expression of the disease. Although cardiac events were more frequent with LQT1 or LQT2 mutations, there was greater lethality among patients who had LQT3. The authors suggested that these data indicated that patients who have LQT3 may warrant more aggressive therapy. This research represents the first step in relating
were significantly higher in survivors of SIDS babies and the total number of family members who have the mutation, was calculated at 25%. The implications of this study are very unsettling because they suggest that it is not possible to screen patients for this life-threatening disease based on their QT interval and that affected patients have a risk of transmitting the disease to 50% of their offspring. As molecular testing enters clinical practice, we should be able to offer much more information to these families, but in the interim, they should be alerted to avoid all drugs that block potassium currents. Further, they should be made aware of the possibility that their offspring may have long QT syndrome and, therefore, should undergo screening by ECG and, as it becomes available, by genetic testing.

One recent study has attempted to link prolongation of the QT interval with sudden infant death syndrome (SIDS). In this study, researchers obtained ECGs on babies at 3 to 4 days of life. ECGs were analyzed for QT interval, and the children were followed for 1 year. More than 33,000 children were included in the study, and 24 deaths were attributed to SIDS. The QT intervals among SIDS babies were significantly higher in survivors. Fifty percent of SIDS children had a prolonged QT (>440 msec). The odds ratio for SIDS in infants who had a prolonged QT was 41.3. This study obviously raises the question of routine screening for long QT syndrome in the neonate, but no consensus has emerged on this vexing issue. However, it does appear to be reasonable to screen high-risk infants (those who have a family history of SIDS or long QT syndrome or those who have survived a life-threatening event).

Therapy in patients diagnosed with long QT syndrome should be initiated with propranolol. If the infant continues to exhibit bradycardia or 2:1 block, epicardial pacing may be used. Mexilitene may be of use in infants who have the LQT3 form of long QT syndrome.

Conclusion

The natural history and prevalence of arrhythmias in the neonate are in striking contrast to that seen in the older child and adult. It is important for the physician caring for these children to have a good understanding of the developmental aspects of arrhythmia as well as the diagnostic and therapeutic options available.

SUGGESTED READING


NEOREVIEWS QUIZ

3. You are discussing cardiac arrhythmias in the newborn period with a medical student. Of the following, the most accurate statement regarding atrioventricular re-entrant tachycardia is that:
A. A retrograde P wave following the QRS complex is characteristic.
B. Atrioventricular block or dissociation can occur during tachycardia.
C. Calcium channel blockers are useful for preventing tachycardia.
D. Maternal history often is significant for anti-Ro and anti-La antibodies.
E. Placing pressure on the eyeball is a useful vagal maneuver for terminating tachycardia.

4. A term newborn has a constant heart rate of 60 beats/min, with no evidence of perinatal asphyxia, apnea, or respiratory distress. Blood pressure and perfusion are normal. Electrocardiography shows no conduction from the atrium to the ventricle, an atrial rate that is higher than the ventricular rate, and no relationship between P waves and QRS complexes. Echocardiography shows a structurally normal heart. You suspect complete heart block. Of the following, the most accurate statement regarding complete heart block is that:
A. It commonly is associated with truncus arteriosus.
B. It requires pacemaker implantation.
C. Neonatal mortality rate is in excess of 50%.
D. Postnatal steroid treatment can be beneficial.
E. There usually is a positive maternal history of connective tissue disease.

CROSS-REFERENCES

- Cardiology
- Arrhythmias

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