Fetal and Neonatal Arrhythmias

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Objectives

After completing this article, readers should be able to:

1. Delineate electrocardiographic parameters for the healthy newborn.
2. Distinguish benign variations in rhythm from pathologic arrhythmias.
3. Describe the acute management of supraventricular tachycardia.
4. Outline the acute management of complete heart block.
5. Recognize the most common fetal arrhythmias.

Abstract

Arrhythmias in fetuses and newborns are relatively common, occurring in up to 90% of newborns and in 1% to 3% of pregnancies. Although life-threatening arrhythmias are uncommon, practitioners should be aware of the more common disorders of rhythm and conduction that can affect fetuses and neonates. This article reviews the healthy newborn electrocardiogram (ECG) and discusses both benign and life-threatening disorders of cardiac rhythm in neonates, including premature atrial and ventricular contractions, atrioventricular block, congenital complete heart block, supraventricular tachycardias, ventricular tachycardias, and long QT syndrome. We also review the diagnosis and management of fetal arrhythmias.

Normal ECG Parameters

In the healthy newborn, the average heart rate ranges from 120 to 160 beats/min, but the normal range includes heart rates from 90 to 230 beats/min. Sinus arrhythmia describes a normal variation in heart rate with respiration. Normal sinus rhythm is characterized by upright P waves in leads I and aVF on the surface ECG, followed by narrow QRS complexes. The PR interval, from the onset of the P wave to the Q or R wave, is measured best in lead II and normally ranges from 70 to 140 msec. In a term neonate, the QRS axis ranges from 55 to 200 degrees, and the normal QRS duration, as measured in lead V_5, ranges from 20 to 80 msec. In the preterm infant, the QRS axis ranges from 65 to 174 degrees.

The QT interval, from the onset of the QRS complex to the end of the T wave, is measured best in leads II, V_5, and V_6. Because the QT interval duration changes with heart rate, Bazett's formula is used commonly to determine the corrected QT interval (QTc): QTc=QT (sec)/$\sqrt{}$RR (preceding RR interval in seconds). The mean QTc on the fourth postnatal day is 400±20 msec, with an upper limit of normal (2 standard deviations above the mean, 97.5%) of 440 msec. Therefore, in 2.5% of healthy newborns, the QTc is expected to be greater than 440 msec. The more prolonged the QTc, the greater the likelihood that it is clinically significant. In healthy infants, there is physiologic prolongation of the QTc by 2 months of age (mean QTc, 410 msec), with return to a mean of 400 msec by 6 months of age. Mild QTc prolongation (≤470 msec) may occur following stressful labor or delivery but usually resolves within 48 to 72 hours.

Abnormalities of the ST segment are uncommon in neonates. In the first postnatal week, T waves vary substantially but are usually positive (upright) in lead V_1. After the first postnatal week, the T wave typically is negative (downgoing) in lead V_1 and positive in V_5

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and V_6. Although the ECG is a key component of the assessment of a newborn in whom cardiac disease is suspected, normal findings do not rule out congenital heart disease.

Neonatal Arrhythmias

Arrhythmias are disorders of cardiac rhythm that can range from benign to life-threatening. Although life-threatening arrhythmias are uncommon in the neonatal period, practitioners should be aware of the more common disorders of rhythm and conduction that can affect neonates.

Premature Atrial and Ventricular Contractions

Premature atrial contractions (PACs) are atrial beats (P waves) that arrive earlier than the normal sinus beat. PACs usually have a different morphology than sinus P waves and may be conducted to the ventricles normally or "blocked." Atrial bigeminy, characterized by blocked PACs occurring in a bigeminal sequence, is a benign cause of neonatal bradycardia. In utero, frequent premature beats are associated with an approximately 1% risk of associated supraventricular tachycardia (SVT); postnatally, that risk may be even lower. In neonates, PACs may occur frequently but tend to decline in frequency within the first few postnatal weeks.

Less common than PACs are premature ventricular contractions (PVCs). Isolated PVCs can occur in otherwise healthy neonates, and progression to ventricular tachycardia is rare among otherwise well infants who have PVCs. Aberrantly conducted PACs, with a wider QRS, may mimic PVCs. Although follow-up for PACs can be minimal, PVCs require more careful attention.

Bradycardias

Some 20% to 90% of healthy newborns experience transient bradycardia, including sinus bradycardia, sinus pauses, and junctional escape beats. Transient bradycardias may occur following stressful labor or delivery but usually resolve within 48 to 72 hours.

SINUS BRADYCARDIA. Sinus bradycardia describes a sinus rhythm in which the heart rate is below the lower limit of normal for age. In the first week after birth, the lower limit of normal is 90 beats/min; in the first month after birth, the lower limit of normal is 107 beats/min. Causes of sinus bradycardia include oversedation, the effect of drugs passed through the placenta from mother to infant, hypothermia, central nervous system abnormalities, increased intracranial pressure, increased vagal tone, obstructive jaundice, and hypothyroidism.

Sinus pauses from 800 to 1,000 msec may occur in healthy newborns. Such pauses usually are followed by escape beats from the atria or the atrioventricular (AV) junction. Pauses of more than 2 seconds are considered abnormal.

WANDERING ATRIAL PACEMAKER. Wandering atrial pacemaker, characterized by a change in P wave axis and morphology most clearly noted on the rhythm strip, is due to a shift of the pacemaker from its usual location in the sinus node to other sites in the atrium and in the AV junction. Wandering atrial pacemaker is a benign arrhythmia but generally is associated with high vagal tone. Accordingly, it may accompany other bradyarrhythmias.

AV BLOCK. AV block occurs when conduction from the atria to the ventricles is delayed or interrupted. First-degree AV block is defined as prolongation of the PR interval for age and may be caused by increased vagal tone, a nonsinus atrial rhythm, and drugs that prolong the PR interval. First-degree AV block typically is not associated with disease of the conduction system. Mobitz type I second-degree AV block (Wenckebach) describes cycles of progressive PR prolongation prior to a single blocked atrial impulse. Wenckebach conduction occurs in healthy neonates and may represent changes in autonomic tone. Mobitz type II second-degree AV block describes abrupt failure of AV conduction of an atrial impulse without prior PR prolongation. This high-grade block is abnormal and may progress abruptly to third-degree (complete) AV block. Variation in the RR interval distinguishes second-degree from third-degree AV block. In third-degree AV block, no conduction occurs from the atria to the ventricles, and the resulting RR typically is fixed (Fig. 1). Modest beat-to-beat variation in the RR interval may occur due to "ventriculophasic variation" in the presence of third-degree AV block and should not be confused with intermittent conduction. High-grade AV block (type II second-degree and third-degree AV block) in neonates may be associated with certain forms of congenital heart disease, with congenital complete AV block, and as the result of pharmacologic therapy. Occasionally, infants who have congenital long QT syndrome (LQTS) present with bradycardia due to second-degree AV block. Symptomatic bradycardias due to AV block or sinus node dysfunction usually are responsive to atropine, epinephrine, or isoproterenol until temporary pacing is available.

CONGENITAL COMPLETE AV BLOCK. Congenital complete AV block occurs in 1 in 15,000 to 1 in 20,000 live births and is associated most commonly with maternal

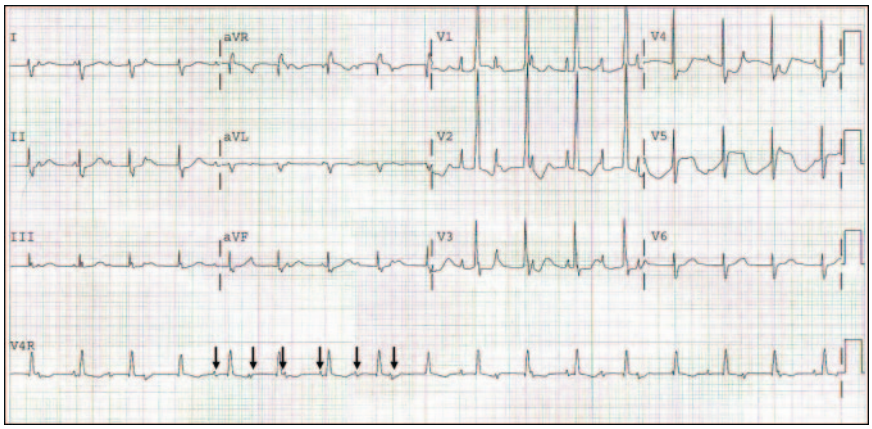

Figure 1. Complete AV block with narrow QRS escape rhythm in a neonate who has congenital AV block. The RR interval is fixed and independent of the atrial rate.

anti-Ro and anti-La antibodies transmitted across the placenta. By cross-reacting with fetal cardiac tissue during a critical stage of development, the anti-Ro and anti-La antibodies are believed to disrupt formation of the normal conduction system. Congenital AV block is being diagnosed increasingly with fetal echocardiography. Prenatal symptoms include hydrops fetalis, which necessitates prompt intervention with delivery and cardiac pacing to prevent fetal demise. Attempts to pace the fetus generally have resulted in fetal demise. Newborns who have complete AV block and no associated heart disease frequently are asymptomatic but may demonstrate congestive heart failure. This could be due to profoundly slow ventricular rates or associated myocardial injury from transplacental antibody exposure. Sudden death, although uncommon, may result from bradycardia-dependent polymorphic ventricular tachycardia. Most patients who have congenital complete AV block require a pacemaker at some point during childhood or adolescence, but a minority display symptoms that necessitate a pacemaker in the newborn period.

Tachycardias

Sinus tachycardia describes a sinus rhythm with a rate above the upper limit of normal for age. In the first week after birth, the upper limit of normal is 166 beats/min and increases to 179 beats/min during the ensuing month. Causes of sinus tachycardia in the neonate include fever, infection, dehydration, pain, and anemia; heart rates should normalize when such conditions are treated. More persistent sinus tachycardia warrants evaluation for neonatal hyperthyroidism or tachycardia due to medications such as beta-adrenergic agonists or theophylline. Careful assessment with a 12-lead ECG and monitoring for a nonsinus mechanism simulating sinus tachycardia also are warranted.

"TYPICAL" PAROXYSMAL SVT.

Paroxysmal SVT usually refers to a rapid, regular tachyarrhythmia (rates of 230 to 300 beats/min) classically characterized by abrupt onset and termination, narrow QRS complexes, a regular RR interval, and absence of clearly discernible P waves. This is the most common tachycardia seen in fetuses, neonates, and infants. Sustained SVT in the fetus may result in congestive heart failure or hydrops fetalis; intermittent tachycardia, even at relatively slow rates, may produce heart failure over days to weeks. In the neonate, SVT of 12 to 24 hours' duration usually results in heart failure. The most common mechanism of SVT in the newborn is orthodromic reciprocating tachycardia (ORT). Less common among neonates is AV nodal re-entry tachycardia (AVNRT). Because the clinical approach to both arrhythmias is similar, most centers do not attempt to distinguish between these mechanisms. However, in our experience, neonatal AVNRT more commonly is associated with other cardiorespiratory problems (eg, congenital diaphragmatic hernia, congenital heart disease, sepsis) and is uncommon in an otherwise healthy infant.

ORT and AVNRT usually display a 1:1 AV relationship, distinguishing them from atrial tachycardias, junctional ectopic tachycardia, and ventricular tachycardias. A premature atrial or ventricular contraction or a junctional escape beat usually initiates ORT or AVNRT. Although the QRS is typically narrow, a wide QRS can occur in the presence of bundle-branch block and in some pre-excitation syndromes.

ORT is a form of SVT using an accessory AV connection that functionally links atrial and ventricular tissue. During tachycardia, conduction occurs from atria to ventricles (antegrade) via the AV node and subsequently back up from ventricles to atria (retrograde) via the accessory pathway, completing a re-entrant circuit. During sinus rhythm, Wolff-Parkinson-White (WPW) syndrome (classically characterized by a short PR interval and a delta wave) may be observed, but ORT also may be associated with a concealed accessory pathway or a "URAP" (unidirectional retrograde accessory pathway), such that WPW is absent during sinus rhythm. Typically,

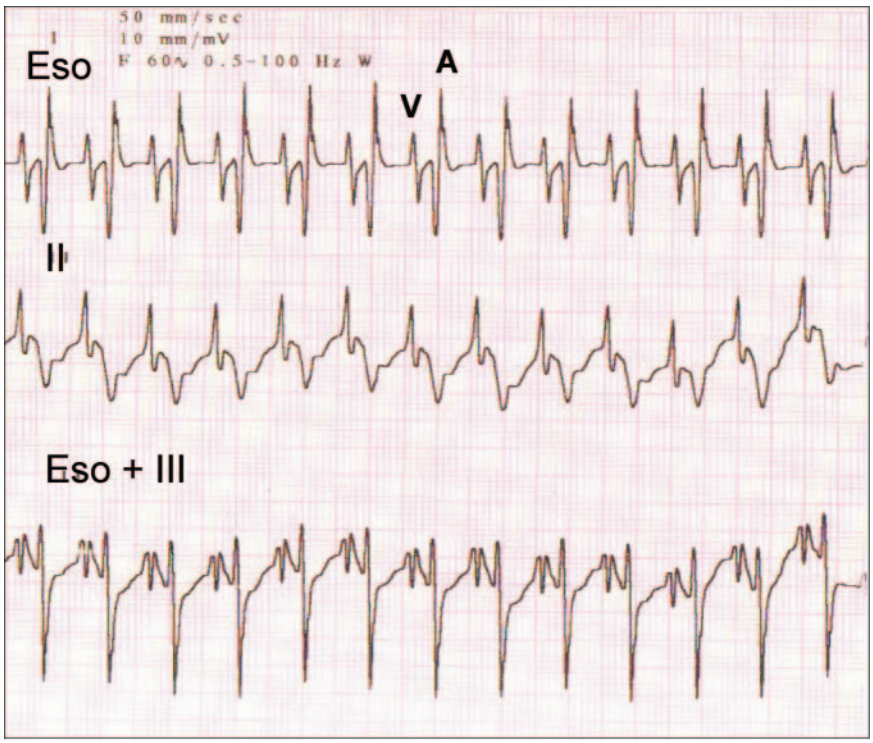

Figure 2. Neonatal supraventricular tachycardia due to orthodromic AV re-entry. The QRS is normal. The esophageal recording displays the atrial depolarization following the preceding QRS by more than 70 msec.

the RP (ventriculoatrial [VA]) interval in tachycardia is short during ORT but usually more than 70 msec, placing the P waves in the ST segment (Fig. 2).

AVNRT is a form of SVT involving the region around the AV node. Antegrade and retrograde conduction occur over anatomically discrete atrial inputs into the AV node. One pathway, the "fast" pathway, conducts more rapidly than the other pathway, the "slow" pathway. Tachycardia usually proceeds antegrade down the "slow" pathway and retrograde up the "fast" pathway. The RP (VA) interval in tachycardia is extremely short (<70 msec), such that P waves are essentially obscured by the QRS complex (Fig. 3).

Vagal stimulation, such as created by applying ice to the face to elicit a dive reflex, may terminate certain SVTs by producing transient block in the AV node. Adenosine, administered as a rapid intravenous bolus (initial dose of 0.1 mg/kg), also can terminate tachycardias that use the AV node by producing transient block in the AV node. Continuous ECG recording during such interventions allows diagnostic recognition of SVT. Temporary pacing via the esophagus is another effective means of terminating SVT and is preferred over repeated administrations of adenosine for SVT displaying incessant behavior as maintenance therapy is established (Fig. 4).

ATRIAL FLUTTER. Atrial flutter is a form of SVT characterized by regular, rapid (atrial rates of 240 to 360 beats/min), "saw-toothed flutter waves," often seen best in leads II, III, and aVF on the surface ECG. In neonates, 2:1 atrial conduction is common, resulting in a rate somewhat slower than typical paroxysmal SVT. Recognition of a secondary nonconducted P wave on ECG distinguishes atrial flutter from typical SVT. This is usually evident from the surface ECG but can be verified by esophageal ECG recording (Fig. 5A). Atrial flutter also may be recognized when transient changes in the A:V conduction ratio (1:1, 2:1, 3:1) result in an irregular ventricular rate. In infants, atrial flutter usually occurs in structurally normal hearts and may be associated with an accessory AV connection. Acute termination of atrial flutter with adenosine varies (Fig. 5B), but termination usually can be accomplished in the neonate with transesophageal pacing (Fig. 5C). DC cardioversion should be reserved for the unstable infant or when transesophageal pacing is unavailable. Atrial flutter generally is self-limited in infants who do not have heart disease, and therapy beyond initial conversion generally is not required.

UNCOMMON FORMS OF SVT. A variety of other forms of SVT are seen less commonly among neonates, and a thorough discussion of each is beyond the scope of this review. However, recognition of atypical SVT is important and warrants the early involvement of a pediatric arrhythmia specialist because acute treatments for typical ORT are of no lasting benefit and may prove deleterious. Notable examples include incessant SVTs due to several factors. Permanent junctional reciprocating tachycardia is an incessant form of ORT in which unusually slow conduction occurs over the accessory pathway, resulting in incessant behavior and a characteristically short PR interval. Ectopic atrial tachycardia often simulates an incessant, monotonous, and otherwise unexplained sinus tachycardia. Junctional ectopic tachycardia typically is

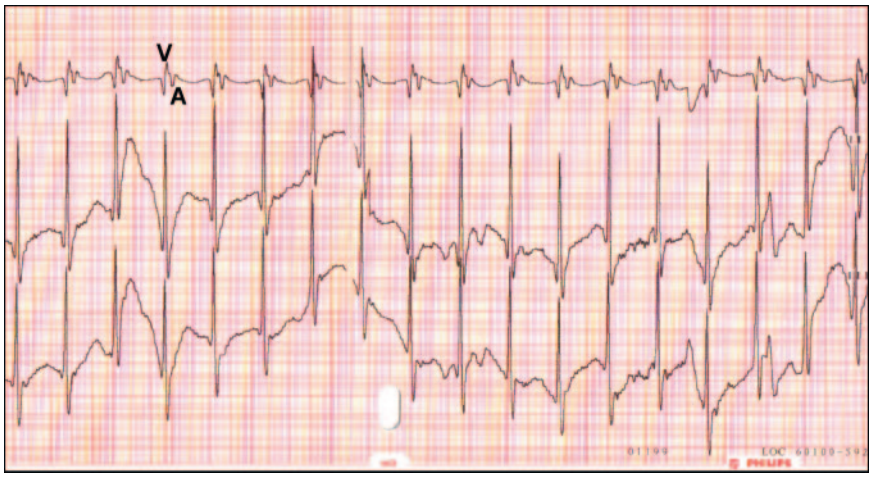

Figure 3. Neonatal supraventricular tachycardia due to AV nodal re-entry. The VA interval is nearly fused during tachycardia (<70 msec). The surface ECG tracing can be difficult to distinguish from orthodromic AV re-entry.

recognized as a narrow QRS tachycardia with VA dissociation, a slower atrial rate, and intermittent shortening of the RR interval due to "sinus capture." It is seen more commonly following surgery for congenital heart disease in the infant. Chaotic atrial tachycardia usually is seen in infants beyond the neonatal period, often in association with a respiratory illness such as that caused by respiratory syncytial virus. It is characterized by highly variable atrial rates, varying P wave morphologies, and periods of apparent atrial flutter and atrial fibrillation.

The ECG features of these arrhythmias may be unique, as in permanent junctional reciprocating tachycardia, which displays a characteristically short PR interval and inverted P wave in the inferior ECG leads II, III, and AVF during tachycardia (Fig. 6). In other instances, the ECG tracing may mimic more typical paroxysmal SVT. Thus, the diagnosis is established by a combination of ECG pattern, pattern of initiation and termination, and response to acute medical and pacing maneuvers. Typically, a cardiologist well-versed in neonatal arrhythmia evaluation and therapy should be involved in diagnosis and treatment.

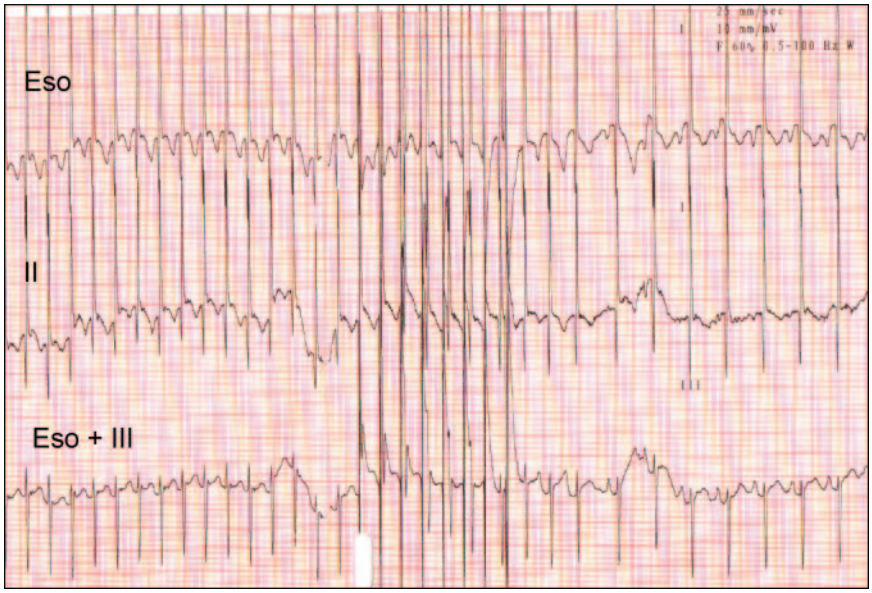

Figure 4. Pace termination of SVT due to AV re-entry with transesophageal pacing. The stimulus train "advances" atrial activation, disrupting the re-entrant circuit and restoring sinus rhythm.

TREATMENTS OF SVT. Treatment of SVT in the neonate often is accomplished in three phases: termination, initial therapy, and maintenance therapy (Table). An extensive discussion is beyond the scope of this article, but a few important principles warrant discussion. For most forms of SVT, adenosine administered as a rapid intravenous bolus either should terminate tachycardia (even if only transiently) or alter the AV relationship in a manner that assists with the diagnosis. Therefore, an ECG strip always should be recording when adenosine is administered. Once the diagnosis is established, it may be helpful to initiate intravenous therapy until enteral administration is deemed safe and feasible. When reinitiation results, repeated dosing with adenosine may perpetuate incessant behavior, so alternative termination and therapeutic strategies should be employed.

Virtually all intravenous antiarrhythmic medications have the potential to produce hypotension, either due to vasodilation or direct negative inotropic effects. Therefore, therapy should be initiated in the presence of a physician and con-

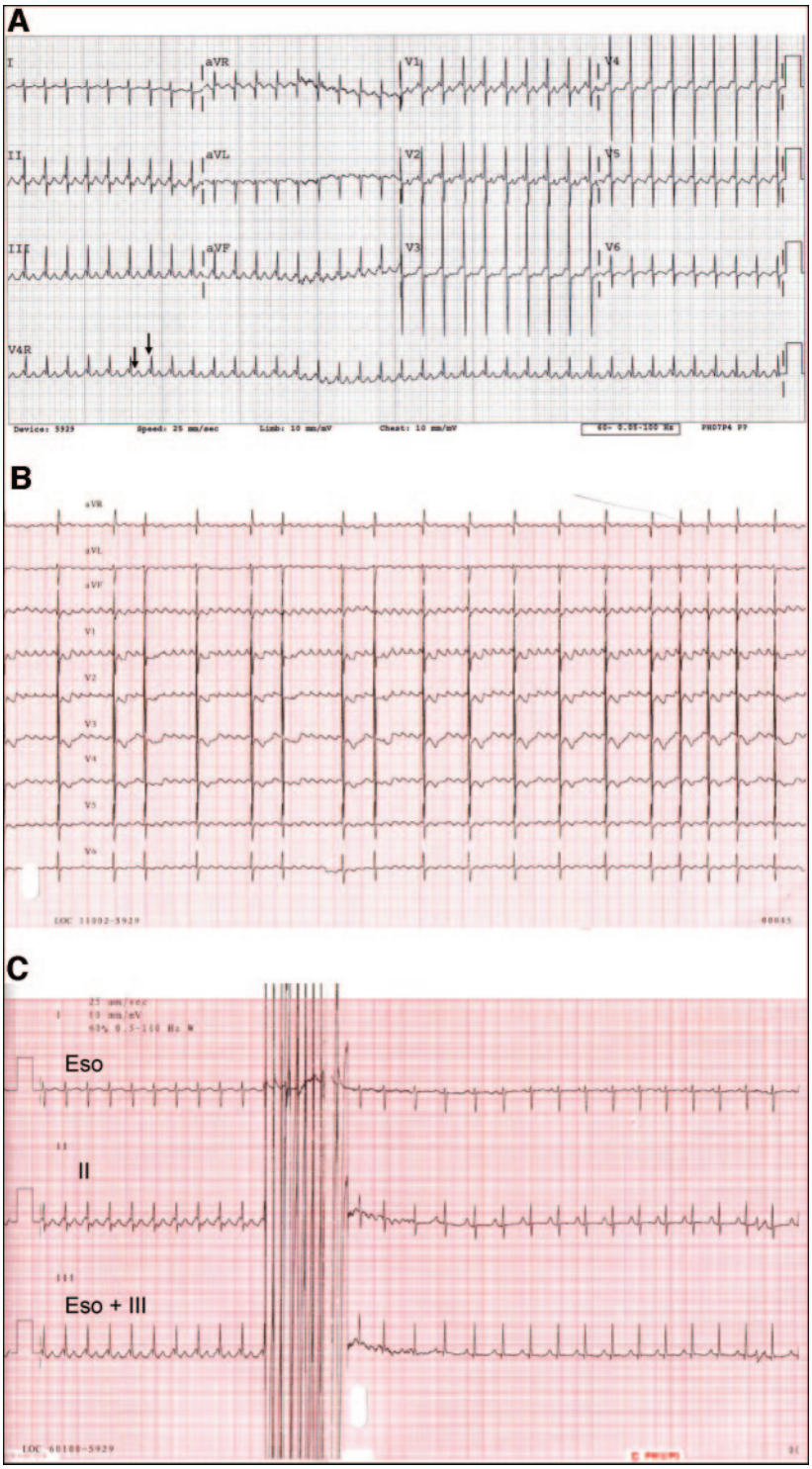

Figure 5. A. Atrial flutter in a neonate. Flutter waves are visible on the surface ECG and correspond with atrial depolarization at twice the ventricular rate (2:1 AV conduction). B. Atrial flutter unmasked during administration of adenosine. Characteristic "saw-tooth" flutter waves are clearly evident when adenosine blocks conduction through the AV node, slowing the ventricular rate. C. Termination of atrial flutter with transesophageal pacing. Entrainment from the atrium disrupts the atrial re-entrant pattern, and after a brief period of disorganized tachycardia, sinus rhythm is restored.

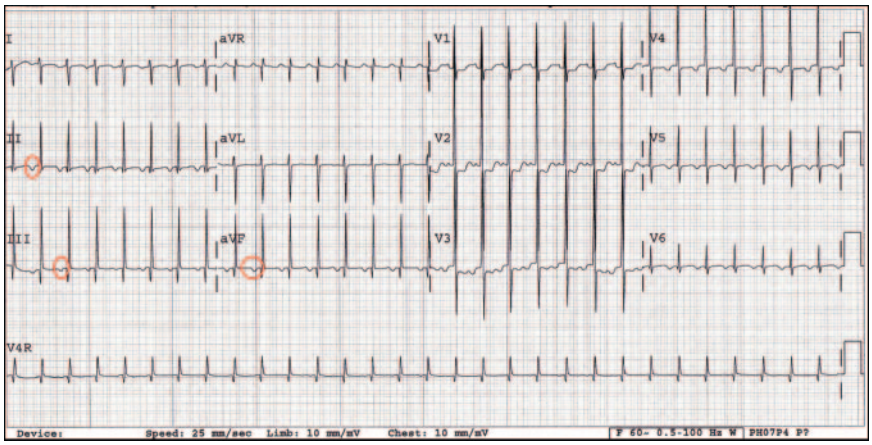

Figure 6. SVT due to permanent junctional reciprocating tachycardia, an uncommon form of orthodromic AV re-entry (ORT) that typically displays incessant rather than paroxysmal behavior. The P wave is inverted in leads II, III, and AVF, and the PR interval is short. This is the result of retrograde conduction over a slowly conducting accessory connection, allowing for recovery of the AV node and more normal PR interval than that seen in more common forms of ORT.

tinuous monitoring of blood pressure and heart rate to recognize and intercede in any hemodynamic deterioration.

In establishing maintenance (oral) therapy, the dosing may need to be adjusted according to multiple factors, including weight, body surface area, age, hormonal, and ECG effects. For example, the initial dose for sotalol employs a correction factor that varies through the first few postnatal weeks. Flecainide absorption may be affected by variations in dietary milk protein. Thyroid function is altered by amiodarone, possibly necessitating thyroid replacement therapy.

Table. Treatment of Supraventricular Tachycardias

Termination
• Ice to face
• Adenosine 0.1 to 0.2 mg/kg
• Transesophageal pacing
• DC cardioversion
Initial Infusion Therapy
• Esmolol 50 to 300 mcg/kg per minute
• Procainamide 30 to 50 mcg/kg per minute
• Amiodarone 0.2 to 0.4 mg/kg per hour
Maintenance Therapy (Oral)
• Propranolol 1 to 4 mg/kg per day
• Sotalol
• Propafenone
• Flecainide
• Amiodarone
• Digoxin
• Verapamil*

*Oral verapamil may be used with caution in neonates. Intravenous verapamil may result in profound hemodynamic collapse and should be avoided in neonates and young infants.

WIDE QRS TACHYCARDIAS. Although SVT with bundle branch block is common in older children, particular with ORT, it is less common among neonates. Indeed, ventricular tachycardias may be attributed mistakenly to "wide complex SVT" because the QRS prolongation in the neonate who has ventricular tachycardia appears modest compared with that seen in older children and adults. Transient right bundle branch block or left bundle branch block can be seen with ORT, usually with the onset of tachycardia. Prolonged tachycardia cycle length (RR interval) during bundle branch block indicates the presence of an accessory pathway on the same side as the conduction delay.

Less common is SVT attributed to Mahaim fibers, slowly conducting accessory connections that originate from the right atrial free wall or the AV node and insert into the right ventricular His-Purkinje system. The accessory pathway serves as the antegrade limb of the re-entrant circuit and is characterized by a prolonged QRS with right bundle branch block pattern on ECG during tachycardia.

Distinguishing ventricular tachycardias from SVT with wide QRS complex has important therapeutic and prognostic implications. Response to maneuvers such as adenosine administration might provide some insight, but it is most helpful to obtain a direct recording of atrial activation using either an esophageal ECG or some other atrial recording. Neonates undergoing cardiac surgery have bundle branch block with prolonged QRS postoperatively. However, they also may be prone to ventricular tachycardias as the result of the operation, so the basis of a wide QRS tachycardia should be sought aggressively (Fig. 7).

Idiopathic Accelerated Ventricular Rhythm

Accelerated ventricular rhythm (AVR) is an uncommon, benign rhythm in otherwise healthy neonates character-

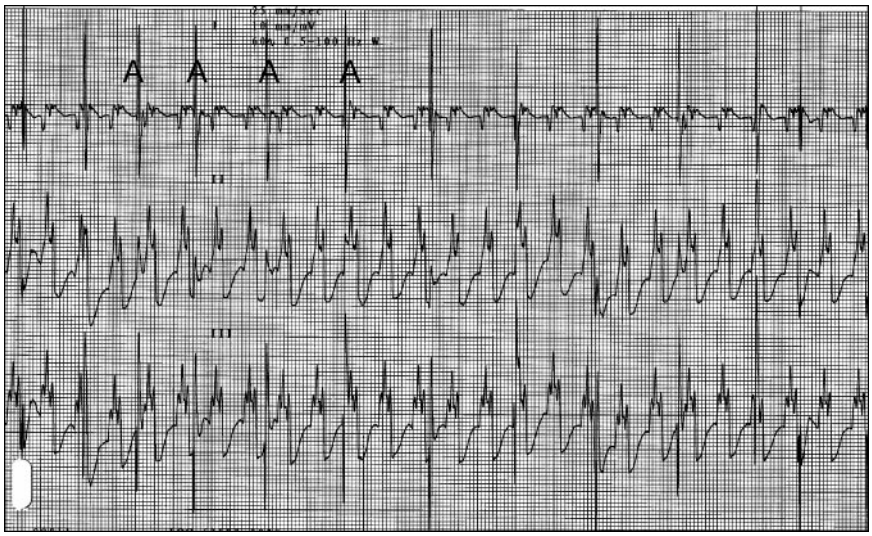

Figure 7. Assessment of wide QRS tachycardia in an infant following cardiac surgery. What might be presumed to be SVT due to bundle branch block is revealed to be ventricular tachycardia with ventriculoatrial block when a direct atrial recording is obtained using an esophageal ECG or (as in this case) a direct atrial recording from temporary epicardial pacing wires.

ized by a regular, wide QRS, with sustained or nonsustained rates 20% or less of the normal sinus rate. Often, periods of "fusion" between AVR and the slightly slow sinus rhythm are seen at onset and termination of AVR. This rhythm does not cause acute or progressive hemodynamic compromise, generally resolves with time, and rarely, if ever, warrants intervention.

Neonatal Ventricular Tachycardia

Ventricular tachycardia may occur in the seemingly healthy neonate and may display paroxysmal or incessant behavior. Any ventricular tachycardia in the newborn warrants prompt referral to a pediatric arrhythmia specialist for detailed characterization and therapy.

Paroxysmal ventricular tachycardia in the neonate often results from cardiac tumors, particularly rhabdomyomas (which also can cause atrial tachycardias and SVT) or cardiac fibromas. Such tumors usually can be seen by echocardiography, but magnetic resonance imaging also may be helpful. Rhabdomyomas, when multiple, usually indicate tuberous sclerosis; such tumors typically resolve. Medical therapy, perhaps with electrophysiology study-guided drug therapy, is warranted, with surgical resection or debulking reserved for extreme cases when the tumor mass also impairs normal cardiac function. Fibromas frequently are large, and unlike rhabdomyomas, generally do not regress. Surgical resection of small, discrete rhabdomyomas may be curative, but large and obstructive tumors may warrant consideration of heart transplantation.

Incessant ventricular tachycardia due to Purkinje cell tumors also is described. These tumors often are located in the epicardium, are relatively inconspicuous on echocardiography, and may prove difficult targets for catheter ablation or surgical excision. Medical therapy may require potent antiarrhythmic therapy with medications such as flecainide or amiodarone to prevent congestive heart failure, but typically these arrhythmias resolve over time.

Idiopathic tachycardias arising from the right ventricular or left ventricular outflow tracts or from the left posterior fascicular conduction system usually are seen in older children and teens. They are rare in infants.

Long QT Syndrome

Congenital LQTS encompasses a family of inherited conditions of abnormal cardiac excitability that affects approximately 1 in 2,500 people and is characterized by delayed ventricular repolarization, prolonged QTc on surface ECGs, and an increased risk of potentially fatal ventricular tachyarrhythmias. The arrhythmia associated with LQTS is a characteristic polymorphic ventricular tachycardia known as "torsades de pointes." This arrhythmia may be asymptomatic if brief and self-limited, but if sustained beyond several seconds, usually results in ventricular fibrillation. More than 400 different mutations have been identified in 10 LQTS-susceptibility genes, including genes that encode for subunits of Na^+, K^+, and Ca^{2+} voltage-gated ion channels. Children who have LQTS may come to clinical attention with syncope, seizures, aborted cardiac arrest, or sudden death; may be found to have a prolonged QTc on ECG performed for other reasons; or may present because of a family history of the symptoms. Symptomatic LQTS is uncommon inneonates, although it may be particularly lethal when associated with functional 2:1 AV block. LQTS is believed to be one cause of sudden infant death syndrome (SIDS); LQTS gene variants have been found in 9.5% of SIDS victims.

Sometimes, infants are screened for LQTS with an ECG due to a family history. Such screening should be delayed until the infant is 6 to 8 weeks of age because modest QT interval prolongation is relatively common (2.5%) among healthy infants and may cause undue family concern. Increasingly, genetic characterization

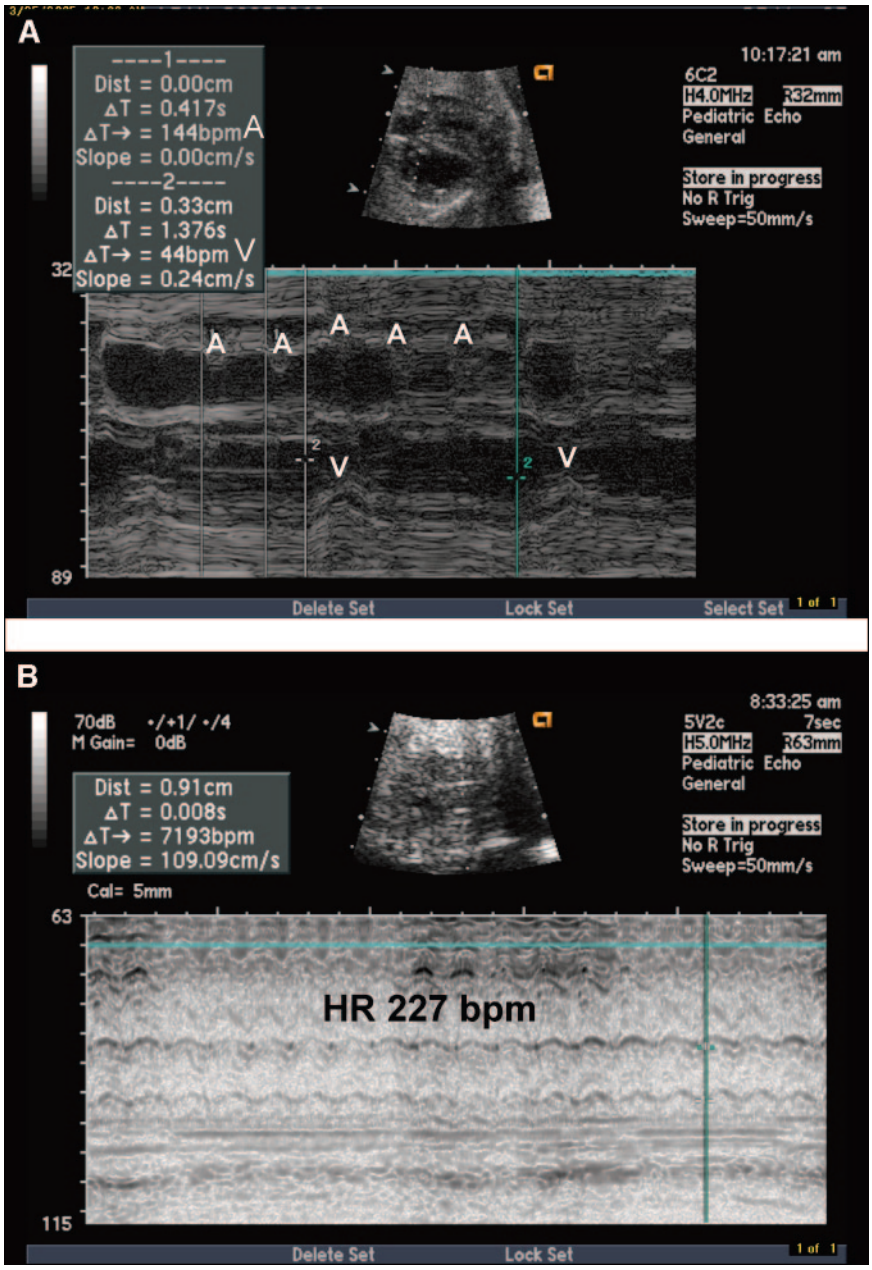

Figure 8. A. Fetal M-mode tracing of complete atrioventricular (AV) block, demonstrating atrial wall motion occurring out of phase with the much slower ventricular wall motion. B. Fetal supraventricular tachycardia (SVT). The atria and ventricles are contracting rapidly, with a 1:1 atrioventricular relationship. Irregularity of fetal SVT could be due to transient interruption and reinitiation or could reflect transient alterations in AV conduction during ongoing atrial tachycardia. This distinction could prove important when establishing medical therapy.

may prove superior to ECG in identifying asymptomatic neonates who carry LQTS-causing gene defects. However, initial genetic analysis should begin with the index patient, with family genetic screening following characterization of the specific defect responsible in the symptomatic individual.

Fetal Arrhythmias

Fetal arrhythmias are relatively common in otherwise uncomplicated pregnancies; isolated ectopic beats may be seen in 1% to 3% of pregnancies. However, the presence of any abnormality or irregularity of the fetal heart rate warrants further evaluation of the fetal rhythm and cardiac structure and function. Irregularity due to ectopic beats may be difficult to discern from incomplete AV block, and ectopic beats may serve as initiating triggers for fetal tachycardias.

Diagnostic Methods

Fetal arrhythmias usually are detected by direct fetal auscultation, Doppler monitoring, or ultrasonography. Abnormalities should prompt fetal echocardiography, which remains the primary diagnostic tool for assessing fetal arrhythmias. Wall motion of the atrial and ventricular walls by M-mode tracings may serve as a surrogate for atrial and ventricular electrical systole. The relative rate and timing of atria and ventricles can be used to infer AV and VA conduction patterns (Fig. 8). However, differences in mechanical contraction relative to electrical systole may limit direct interpolation of such events to standard electrocardiographic diagnosis. Two-dimensional imaging provides important information about fetal cardiac function and the development of hydrops fetalis. Fetal Doppler studies, particularly hepatic vein flow and tissue Doppler studies, may provide additional prognostic information in the fetus that has arrhythmias. Recently, fetal magnetocardiography has been used to obtain strikingly clear tracings of fetal arrhythmias, but

the dedicated facilities and personnel necessary to obtain such tracings are not widely available.

Fetal AV Block

Fetal AV block is observed most commonly in mothers who have anti-Ro and anti-La antibodies associated with systemic lupus erythematosus or Sjögren syndrome. Less commonly, AV block is associated with congenital heart defects and associated abnormalities in AV concordance, such as congenitally corrected transposition of the great arteries or heterotaxy syndromes.

AV block may be observed as early as the 16th week of gestation. Most affected infants who have no structural heart disease do well and can be observed expectantly in utero to term delivery. However, congestive heart failure and the development of hydrops fetalis are ominous findings, and without delivery, fetal demise is likely. The fetus that has AV block associated with congenital heart disease is likely to be at particularly high risk, not only during gestation, but even with appropriate postnatal therapies.

Fetal therapies for symptomatic AV block have been largely unsatisfactory. Some evidence suggests that maternal steroid therapy may reverse the condition, if initiated early in gestation and in the presence of second-degree AV block. However, third-degree (complete) AV block is not reversible. Maternal administration of beta-agonists, such as terbutaline, may augment the fetal heart rate, but a benefit on fetal outcome is unproven. Several case reports have described "successful" in utero pacing techniques, although the ultimate outcome in all published cases has been fetal demise within hours of initiating therapies.

In general, the only potentially useful therapy for the fetus that has AV block and congestive heart failure or hydrops fetalis is prompt delivery and initiation of pacing. The size of the newborn at delivery determines whether a permanent pacemaker may be implanted or whether a secure temporary pacing technique can be employed until the neonate reaches a size suitable for permanent pacing.

Fetal Tachycardias

Fetal SVT, predominately due to AV re-entry, is the most common form of fetal tachycardia, followed closely by fetal atrial flutter. Junctional and ventricular tachycardias also may be observed in the fetus. If tachycardia episodes are self-limited and not associated with impaired cardiac function or hydrops, close observation and frequent follow-up without initiation of antiarrhythmic therapy may be sufficient. However, sustained, uninterrupted tachycardia warrants therapy, especially when cardiac dysfunction becomes apparent.

Therapy for fetal tachycardias is confounded by somewhat unpredictable transplacental transport and delivery to the fetus, potential adverse effects to the mother to whom the drugs are administered, and potential uncertainties in tachycardia mechanism. For fetal SVT, digoxin remains the usual first-line therapy, primarily due to its relative safety for mother and fetus. Some cardiologists advocate intravenous loading, with conversion to oral therapy once fetal cardioversion has been successful. Direct intramuscular administration to the fetal thigh sometimes is employed. When digoxin is unsuccessful, a number of other antiarrhythmic agents have been employed with varying success, including flecainide, sotalol, and amiodarone. The latter appears particularly effective and may achieve cardioversion in more than 90% of infants who have AV re-entry. Conversion of atrial flutter in utero is less successful, but this arrhythmia is better tolerated in the fetus. Maternal adverse effects are not uncommon, and fetal hypothyroidism is a concern. Although there is less experience with fetal therapy for ventricular and junctional tachycardias, they appear to respond favorably to amiodarone.

Proarrhythmia (aggravation of the existing arrhythmia or provocation of a new arrhythmia) may be a greater concern for both the mother and the fetus receiving flecainide or sotalol. Fetal risk is particularly difficult to gauge, except for effects on tachycardia rate, because usual electrocardiographic markers for excessive drug effect are unavailable in the fetus. Verapamil and procainamide have been used with varying success, but should be administered with caution when ventricular function is compromised. Although direct intravenous administration via the umbilical vein has been used, it is unclear whether the risk of such an invasive technique is warranted.

Not all fetal tachycardias controlled in utero become manifest following delivery. However, careful observation with cardiac monitoring is warranted as fetally administered medications are allowed to clear. When available, a provocative transesophageal pacing study to assess tachycardia susceptibility may help determine ongoing tachycardia susceptibility.

Suggested Reading

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NeoReviews Quiz

1. You are asked to evaluate a 4-day-old term newborn for a cardiac arrhythmia. You review with medical students the normal findings on neonatal electrocardiography (ECG). Of the following, the *most* accurate statement regarding normal ECG findings in such a neonate is that:

   A. Corrected QT interval averages 470 msec.
   B. PR interval is measured best in lead I.
   C. QRS axis ranges from 55 to 200 degrees.
   D. QRS duration ranges from 70 to 140 msec.
   E. T wave is usually negative in lead V_1.

2. Wandering atrial pacemaker, characterized by a change in P wave axis and morphology, is caused by a shift of the pacemaker from its normal location in the sinus node to other sites in the atrium and in the atrioventricular junction. Of the following, the wandering atrial pacemaker is *most* associated with:

   A. Excessive sedation.
   B. High vagal tone.
   C. Hypothermia.
   D. Hypothyroidism.
   E. Increased intracranial pressure.

3. Supraventricular tachycardia (SVT) refers to a rapid and regular tachyarrhythmia, which is characterized by abrupt onset and termination, narrow QRS complexes, fixed RR intervals, and absence of discernible P waves. Of the following, the *most* common mechanism of SVT in the newborn is:

   A. Atrioventricular nodal reentry tachycardia.
   B. Chaotic atrial tachycardia.
   C. Ectopic atrial tachycardia.
   D. Junctional ectopic tachycardia.
   E. Orthodromic reciprocating tachycardia.

4. Treatment for fetal tachycardia may be warranted if the tachycardia is sustained and the cardiac function is compromised. Although digoxin remains the first-line drug, other antiarrhythmia drugs may be needed if digoxin fails to induce a normal heart rhythm. Of the following, the antiarrhythmia drug *most* effective in inducing a normal heart rhythm in the fetus that has resistant atrioventricular nodal reentry tachycardia is:

   A. Amiodarone.
   B. Flecainide.
   C. Procainamide.
   D. Sotalol.
   E. Verapamil.

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