Fetal and Neonatal Arrhythmias

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KEYWORDS

• Arrhythmia • Fetal • Pediatric • Tachycardia • Bradycardia • Diagnosis

KEY POINTS

- Arrhythmias may present as an irregularity of the cardiac rhythm, as slow or fast heart rate, or as a combination of irregular rhythm and abnormal rate.
- The identification of the underlying arrhythmia mechanism and hemodynamic impact is critical because the management and prognosis differ among the various disorders.
- The most common arrhythmia is an irregular heart rhythm caused by premature atrial contractions (PACs). Isolated PACs are usually benign and self-resolving.
- Dysrhythmias presenting with sustained slow or fast heart rates are uncommon but potentially life threatening because of the hemodynamic consequences and the underlying cause. Antiarrhythmic treatment to control the tachycardia is often key to ensuring a good outcome.

INTRODUCTION

Understanding the normal rhythm is essential for diagnosing any dysrhythmia. This article reviews the normal electrophysiology and then addresses the mechanisms, features, management, and outcomes of the common fetal and neonatal rhythm disorders.

NORMAL IMPULSE GENERATION AND PROPAGATION

The main function of the heart is to pump blood throughout the body to allow a sufficient supply of oxygen and nutrients to the tissues while removing toxic wastes. Cardiac output, the volume of ejected blood per minute, is equal to the stroke volume of each ventricle in a single heart beat times the heart rate. The normal heart rate ranges between 120 and 160 beats/min (bpm) in the mid to late gestational fetus and between

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100 and 150 bpm in the newborn. Heart rate is usually controlled by the sinoatrial (SA) nodal cells. These cells are capable of spontaneously depolarizing and thus acting as a pacemaker. The electrical impulse from the SA node is then propagated across the atria, the atrioventricular (AV) node, and the His-Purkinje system throughout the ventricles, allowing the sequential depolarization of the atrial and ventricular myocardium with each heartbeat. The cardiac mechanical actions, contraction of myocytes in systole, and relaxation in diastole are then orchestrated by rapid cyclic changes in their transmembrane action potentials and ion currents with each heartbeat. Following depolarization, the conducted impulse is prevented from immediately reactivating the conduction system and myocardium by refractoriness of the tissue that just has been activated. The heart must then await a new electrical impulse from the SA node to initiate the next heartbeat.

METHODS OF PERINATAL CARDIAC RHYTHM ASSESSMENT

The electrocardiogram (ECG) is the main diagnostic tool after birth to record the electrical activity of the heart. The normal ECG entails a sinus P wave with a P-wave axis between 0 and +90° (positive P wave in lead II) that precedes each QRS complex within a regular, normal PR interval. The neonatal cardiac rhythm is typically regular and the rate within the normal range for patient age. Because noninvasive fetal electrocardiography is available at a few centers only, the antenatal rhythm evaluation is primarily based on the chronology of atrial and ventricular systolic mechanical events that are recorded by echocardiography. M-mode imaging is useful to simultaneously record the atrial and ventricular systolic wall motions.[1] Similarly, simultaneous pulse wave Doppler evaluation of the mitral inflow and aortic outflow (mitral valve/aorta) or, preferably, the superior vena cava (SVC) and the ascending aorta (SVC/aorta Doppler) is used to examine the sequence and time relationship of blood flow events that are secondary to atrial and ventricular contractions.[2] The beginning of the mitral A wave and the retrograde SVC a wave reflect the onset of atrial systole, whereas the onset of aortic forward flow marks the beginning of ventricular systole. The diagnosis of a normal fetal cardiac rhythm is based on the documentation of a regular atrial and ventricular rhythm with a normal rate for gestational age (**Fig. 1**A).[3] Each atrial event is followed by a ventricular event within a normal AV time interval, which confirms normal 1:1 AV conduction.[4] Although echocardiography provides useful information on mechanical systolic events, it does not inform on the morphology, duration, and amplitude of electrical events. Hence, it is not possible to confirm repolarization abnormalities like long QT syndrome (LQTS) solely by echocardiography.

MECHANISMS OF ARRHYTHMIAS

Arrhythmias may present as an irregular cardiac rhythm, as a slow or fast heart rate, or as a combination of abnormal rhythm and rate. The contributing causes can be broadly divided into abnormalities in the generation and the propagation of electrical impulses. These disturbances result from critical alterations in electrical activity and may occur in every region of the heart.

Abnormal Impulse Generation

Cardiac cells in the atria, AV node, and His-Purkinje system can spontaneously depolarize and manifest automaticity outside the SA node. They are called latent pacemakers because they are physiologically suppressed by the faster sinus rate. Rhythm disorders whose origin is the SA node include a sinus node that fires at an unusually fast or slow rate. Ectopic cardiac rhythms occur when the dominant

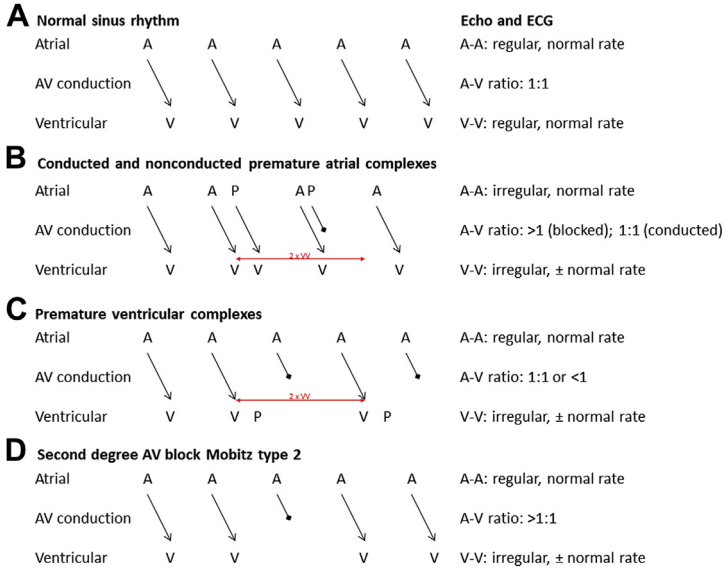

Fig. 1. (*A–D*) The sequence of electrical activation and impulse propagation of (*A*) the normal sinus rhythm compared with (*B–D*) the main disorders of an irregular heart rhythm. →, nonconducted atrial beat; A, normal atrial event; Echo, echocardiography; P, premature atrial or ventricular complex; V, normal ventricular event.

pacemaker shifts from the SA node to a latent pacemaker, either when the sinus rate decreases to less than the intrinsic rate of the secondary pacemaker (atrial or junctional escape rhythm) or the intrinsic rate of a secondary pacemaker increases to more than the normal sinus rate (atrial ectopic tachycardia, junctional ectopic tachycardia, ventricular tachycardia [VT]), or the sinus beat fails to conduct across the AV node, leaving a secondary junctional or ventricular pacemaker free to fire at its slower intrinsic rhythm.

Abnormal Impulse Propagation

Reentry is the propagation of an impulse through myocardial tissue already activated by the same impulse in a circular movement. Reentry is the underlying mechanism of most types of perinatal tachyarrhythmia, including atrial flutter (AF) and AV reentrant tachycardia (AVRT). AF is sustained by a macroreentrant circuit that is confined to the atria. AVRT, the most common mechanism of a fast heart rate in the young, is a reentrant circuit that uses the AV node to conduct from the atria to the ventricles and a fast-conducting accessory pathway to propagate the ventricular impulse back to the atria. On the other side, nonconduction of the impulse occurs when it arrives in nonexcitable tissue, either because it is still refractory after a recent depolarization (eg, blocked premature atrial contraction [PAC]) or because of abnormal tissue (eg, heart block).

CONSEQUENCES OF ARRHYTHMIAS

Compared with adults, the fetal and neonatal heart beats significantly faster, is structurally and functionally immature, and performs close to the maximum of the ventricular function curve. Because of the limited pump reserve of immature hearts, any

significant change in heart rate leads to a decline in cardiac output, impaired cardiac filling, and venous congestion, the severity of which depends on arrhythmia characteristics and myocardial properties. As a general rule, the more abnormal the heart rate and the younger the age, the less likely it is that a significant arrhythmia will be well tolerated by the fetus and infant. Rhythm disorders that manifest with enduring slow (complete heart block) or fast (AVRT) heart rates represent the main cardiac causes of fetal hydrops, prematurity, and perinatal death. To provide optimal care on any new arrhythmia diagnosis it is therefore essential to first discern the mechanism and the hemodynamic impact of the rhythm disorder and then to decide on the need of treatment, if this option is available.

ASSESSMENT OF ARRHYTHMIAS

Although arrhythmias have diverse causes, most abnormalities can be deducted by the experienced investigator. **Box 1** presents a stepwise approach that can be used to diagnose and differentiate most fetal arrhythmias and that may also be used for neonatal patients.

Box 1
Stepwise approach to the assessment of fetal rhythm abnormalities

Atrial (A) rate and rhythm (A-A):

Absent – slow – normal – fast

Regular – irregular – regular-irregular (eg, atrial bigeminy)

Ventricular (V) rate and rhythm (V-V):

Slow – normal – fast

Regular – irregular – regular-irregular (eg, ventricular bigeminy)

Ratio of atrial and ventricular events:

Normal 1:1 (equal A and V)

Greater than 1:1 (more A than V)

Less than 1:1 (more V and A)

Relationship and timing of atrial and ventricular events:

Normal AV time – prolonged AV time – A-V dissociation

Arrhythmia pattern:

Duration: brief (<10%) – intermittent (10%–50%) – sustained (>50%) – incessant (100%)

Onset/termination: sudden or gradual; triggered by other event (ie, PAC)

Health state:

Effusions, heart size and function, AV valve regurgitation; fetal movements; ↓ or ↑ amniotic fluid

Structural heart disease and other associations:

Heart block: anti-Ro antibodies; left isomerism; congenital corrected transposition

Supraventricular tachycardia: Ebstein anomaly

Sinus tachycardia: Graves disease; beta-mimetic therapy; myocarditis

Sinus bradycardia, 2:1 AV block, VT: LQTS; anti-Ro antibodies

ARRHYTHMIAS PRESENTING WITH AN IRREGULAR RHYTHM

Probably the most frequent presentation of an irregular rhythm disorder is coincidental during a routine assessment of an otherwise asymptomatic patient. The main differential diagnosis of the underlying mechanisms includes (Fig. 1B–D):

- PAC
- Premature ventricular contractions (PVCs)
- Second-degree AV block

Premature Atrial Contractions

PACs account for most patients with an irregular heartbeat at any age. ECG criteria of PACs include the documentation of premature P waves with abnormal P-wave axes and with an AV conduction that may be normal, aberrant (bundle branch block), or blocked. By echocardiography, a PAC is detected by a shorter than normal atrial (A-A) interval (see Fig. 1B). If the AV conduction is normal, the premature atrial event is followed by a timely related premature ventricular event. If the PAC is premature enough to fail conduction across the refractory AV node, no ventricular event is observed, which manifests as a skipped heartbeat. The true fetal incidence of PACs is unknown but the arrhythmia seems to be common. In healthy newborns, PACs were documented in 51% over a 24-hour ECG surveillance study.[5] At least after birth, isolated PACs may be considered a finding within the normal range unless they are associated with other conditions, such as electrolyte abnormalities, myocarditis, or tachyarrhythmia. Before birth, PACs have been associated with less than a 1% risk of fetal tachycardia, although a higher risk has been suggested for atrial bigeminy and couplets.[6,7] PACs are usually spontaneously resolving and no medical treatment is warranted.

Premature Ventricular Contractions

PVCs are uncommon observations during fetal life. In healthy newborns, PVCs were documented in 18% by 24-hour ECG.[5] The ECG diagnosis is based on a premature QRS complex that is not preceded by a P wave. Moreover, the QRS morphology of the PVC differs from normally conducted ventricular beats. By echocardiography, the PVC is not preceded by an atrial beat, whereas the atrial intervals are usually normal and regular (see Fig. 1C).

Isolated atrial and ventricular ectopy are typically benign and self-limited, and no treatment is required.[5] Fetal heart rate should be monitored weekly or every other week by an obstetrician or midwife until the PACs or PVCs have resolved. In addition, recently published American Heart Association (AHA) guidelines also recommend fetal echocardiography to assess the cardiac structure and function and to determine the mechanism of the arrhythmia if the fetus presents with frequent ectopic beats, if there is any question about the mechanism, or if the ectopic beats persist beyond 1 to 2 weeks.[6]

An irregular rhythm can also be caused by second-degree AV block, which is characterized by failure of AV conduction of some, but not all, atrial activity to the ventricles (see Fig. 1D). The atrial rate is normal and the ventricular rate depends on the number of conducted atrial impulses. In Mobitz type I or Wenckebach-type AV block, the nonconducted atrial event is preceded by progressive PR/AV lengthening. In Mobitz type II, the AV conduction is either normal or blocked. Type II is considered serious because the site of the conduction block is below the AV node. Second-degree fetal AV block has been associated with antibody-mediated conduction disease and may

benefit from antiinflammatory treatment to prevent progression to complete heart block (CHB).[8]

ARRHYTHMIAS PRESENTING WITH A SLOW HEART RATE

Fetal bradycardia is defined by a heart rate less than 110 bpm; in the newborn it is a resting heart rate less than 100 bpm on a surface ECG. Occasional, brief sinus bradycardia is a benign physiologic response whereby the rate of the SA node is slower than normal for age. Of more concern is bradycardia that is prolonged or persistent, which should trigger a more detailed assessment for the cause. The main mechanisms of perinatal bradycardia include (Fig. 2):

- Sinus bradycardia
- CHB
- Functional AV block: nonconducted atrial bigeminy and 2:1 AV block

Sinus Bradycardia

Sinus bradycardia is defined as a rhythm that originates from the SA node but in which the rate is slow for age (see Fig. 2A). A subsidiary pacemaker (ie, in the lower atrium) may become the dominant pacemaker if the rate of the SA node decreases to less than that of the secondary pacemaker. By echocardiography, fetal sinus or atrial bradycardia resembles that of a normal rhythm with the only difference that the atrial and ventricular rates are slow for gestational age, usually in the range between 80 and 110 bpm. Sinus bradycardia per se is well tolerated but may be secondary to fetal distress, sinus node dysfunction (anti-Ro antibody related, left isomerism), and LQTS (KCNQ1 mutations).[9–12]

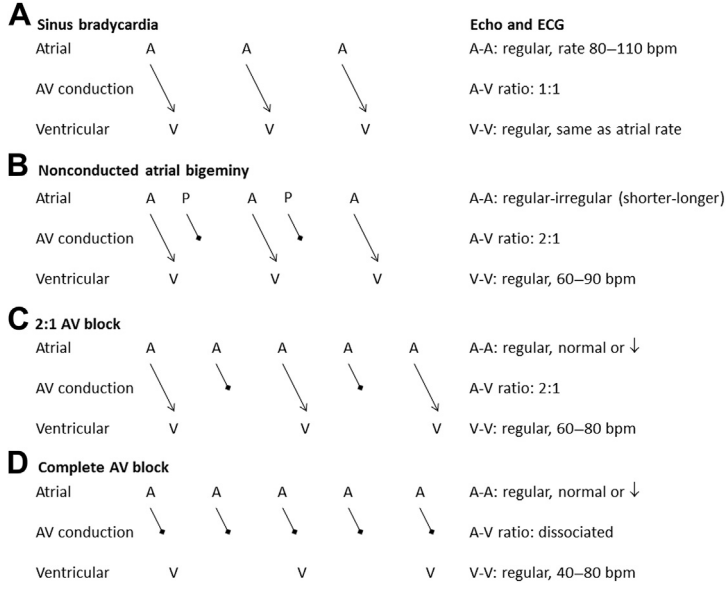

Fig. 2. (*A–D*) Electrical activation and impulse propagation of the main disorders of a slow heart rate. P, premature atrial complex.

The perinatal management of sinus bradycardia depends on the underlying cause and may include no treatment, antiinflammatory medication for myocarditis (anti-Ro antibodies, parvovirus), premature delivery (fetal distress), and postnatal therapy with β-blocker with or without pacing (LQTS).

Complete Heart Block

CHB is defined as a complete failure of the normal propagation of atrial impulses to the ventricles (see **Fig. 2**D). It is the most common congenital conduction abnormality and accounts for about 40% of all major arrhythmias before birth. The typical fetal echocardiogram shows a regular normal atrial rhythm and rate, whereas the ventricles beat independently at a much slower rate of between 40 and 80 bpm. On the ECG, the QRS morphology of congenital CHB is usually narrow complex, which means that the ventricular escape rhythm is junctional (**Fig. 3**.). In about half of fetal cases with CHB, it is associated with major structural heart disease, most importantly left atrial isomerism, which carries a very high risk of in-utero demise.[13,14] In the absence of structural heart disease, congenital CHB is strongly linked to the fetal transplacental passage of anti-Ro antibodies, which are prevalent in about 2% of pregnant women.[15] In 1% to 5% of exposed fetuses, the maternal antibodies lead to complications, including CHB, sinus bradycardia, myocarditis, endocardial fibroelastosis, and/or dilated cardiomyopathy. Although isolated fetal CHB is often tolerated, at the severe end of the disease spectrum it results in low cardiac output, fetal hydrops, and death. Risk factors associated with perinatal death include fetal hydrops, endocardial fibroelastosis, myocarditis, and bradycardia less than 50 to 55 bpm.[16,17]

There is currently no consensus about the indications of prenatal therapy for isolated CHB. There is no treatment available to reverse CHB.[18] However, dexamethasone, intravenous immune globulin (IVIG), β-adrenergic medication, and postnatal pacing have been used to prevent or treat more severe immune-mediated myocardial inflammation, to augment cardiac output and to improve the chances of survival.[19,20] In contrast, possible treatment-related adverse events that may preclude the routine use of high-dose steroids include fetal growth restriction, oligohydramnios, and maternal mood/behavioral changes.[17,19] Chronic prenatal steroid therapy for CHB

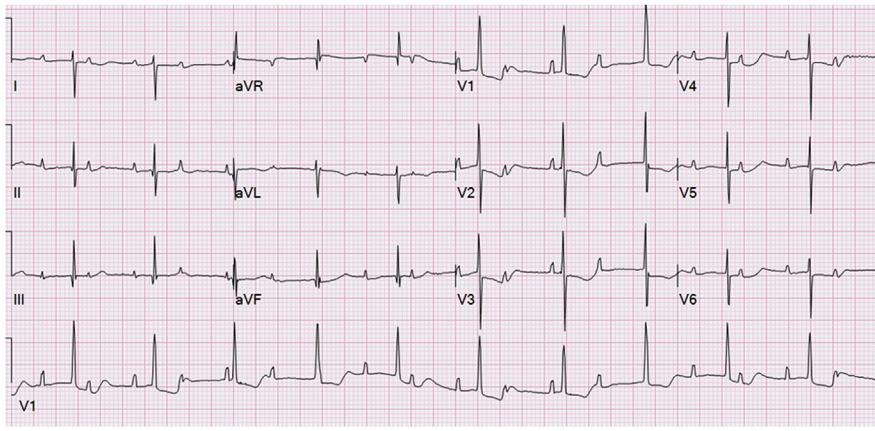

Fig. 3. ECG recording of a newborn with immune-mediated congenital CHB. The atrial rate is 135 bpm, the narrow complex ventricular rate is 105 bpm, and there is complete AV dissociation. The baby has no immediate indication for a permanent pacemaker implantation.

had no obvious impact on neurocognitive function at school age.[21] In our institution, maternal dexamethasone (8 mg/d for 2 weeks; 4 mg/d to 28 weeks; 2 mg/d to birth) is routinely used to treat immune-mediated cardiac disease from the time of diagnosis to birth.[22] Maternal IVIG (1 g/kg every 2–3 weeks) is added if we detect signs of endocardial fibroelastosis and ventricular dysfunction. In contrast, idiopathic isolated CHB (not associated with maternal anti-Ro antibodies) can be managed without antiinflammatory medication. If the average fetal heart rate is less than 50 bpm, we also use transplacental salbutamol (usually 10 mg 3 times a day orally) and postnatal isoprenaline infusion to maintain an adequate ventricular output until the neonatal implantation of a permanent pacemaker system. Current American College of Cardiology/AHA guidelines[23] recommend permanent pacing for CHB in children for the following class 1 indications: (1) symptomatic bradycardia, ventricular dysfunction, or low cardiac output; (2) wide-complex QRS escape rhythm; and (3) infants with ventricular rate less than 55 bpm or with congenital heart disease and a ventricular rate less than 70 bpm. During the past decade, the neonatal survival rate of isolated congenital CHB was 95% at our center, which is improved compared with predominantly untreated patient cohorts reported by us and others.[16,17,19,24] Most patients with isolated congenital CHB require permanent pacing during childhood and most commonly during the first month of life.[25]

Functional Atrioventricular Block

Functional AV block may occur when the AV node is refractory not excitable; that is, following recent depolarization or because of QT prolongation. In nonconducted atrial bigeminy (see **Fig. 2**B), every second atrial impulse occurs prematurely enough to fail conduction by the physiologically refractory AV junction. By echocardiography, the interatrial intervals are irregular but in a regular pattern alternating between a shorter (A-PAC [time interval between sinus beat to premature atrial beat]) and a longer (PAC-A [time interval between premature to normal beat]) atrial interval. If each PAC is nonconducted and each SA beat (A) is forwarded to the ventricles, the ventricular rate will be half of that of the averaged atrial rate, which is in the range between 60 and 90 bpm in fetuses. Nonconducted atrial bigeminy is a possible cause of fetal bradycardia and may last for days to weeks. Blocked PACs are benign, well tolerated, and resolve spontaneously. Weekly assessment by an obstetrician is recommended until resolution of the PACs is documented.

Atrial bigeminy should not be confused with 2:1 AV block (see **Fig. 2**D), which may be related to a congenital QT prolongation. Unlike with atrial bigeminy, the atrial rhythm in 2:1 AV block seems fairly constant and 1:1 AV conduction recurs at slower atrial rates. Similar to other possible LQTS manifestations (unexplained sinus bradycardia, VT), patients with 2:1 AV block (and their families) should undergo a complete work-up for the possibility of an inherited ion channel disorder. Because of the predisposition of patients with LQTS for VT-related cardiac arrests and sudden death, postnatal treatment with a long-acting β-blocker (ie, nadolol) with or without a pacemaker or implantable cardioverter-defibrillator is usually required.[9,10]

ARRHYTHMIAS PRESENTING WITH A FAST HEART RATE

The detection of a fast heart rate greater than 180 bpm in a fetus or newborn constitutes a medical emergency because it carries a significant risk of hemodynamic compromise, heart failure, morbidity, and mortality.

Possible mechanisms include (**Fig. 4**):

- Supraventricular tachycardia (SVT), including AVRT, atrial ectopic tachycardia (AET), and permanent junctional reciprocating tachycardia (PJRT)

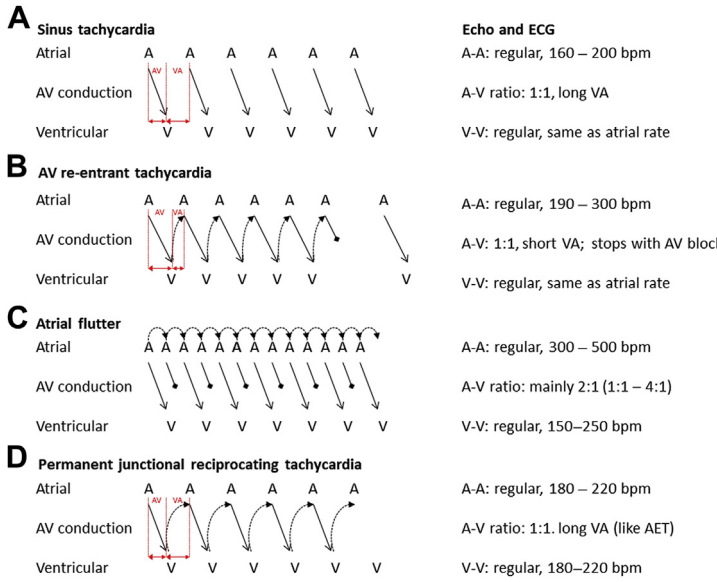

Fig. 4. (*A–D*) Electrical activation and impulse propagation of the main disorders of a fast heart rate. Long VA indicates that the tachycardia VA interval is longer than the AV interval, which is the case in sinus tachycardia, permanent junctional reciprocating tachycardia, and atrial ectopic tachycardia (not shown). The VA interval is shorter than the AV interval in AVRT (short VA or short RP tachycardia). ⇢, reentrant pathway; →, nonconducted atrial event; V, ventricular event.

- AF
- Sinus tachycardia
- Rare: VT

Atrioventricular Reentrant Tachycardia and Atrial Flutter

AVRT and AF account for 90% of the fetal and neonatal tachyarrhythmias.[26] Both arrhythmias are readily distinguished by echocardiography and electrocardiography.

Atrioventricular Reentrant Tachycardia

AVRT commonly manifests as an intermittent or persistent tachycardia between 190 and 300 bpm. It can occur any time after the first trimester. The usual reentrant circuit involves the AV node for anterograde conduction (AV) and a fast retrograde ventriculoatrial (VA) conducting accessory pathway. AVRT starts suddenly with a PAC and terminates with AV block. Most hearts are structurally normal, but Ebstein anomaly of the tricuspid valve is a known association with accessory pathways. By fetal echocardiography, the tachycardia has a short VA pattern because the atrial contraction occurs closely after the ventricular contraction (see Fig. 4B). Because of the almost simultaneous atrial and ventricular contraction the AV valves are closed during atrial systole and there is pronounced a-wave flow reversal in the precordial veins and the ductus venosus. On the tachycardia ECG (Fig. 5), a retrograde P wave is seen immediately following QRS (short RP tachycardia). The QRS morphology is typically regular and narrow complex unless there is rate-related bundle branch block. A delta wave is

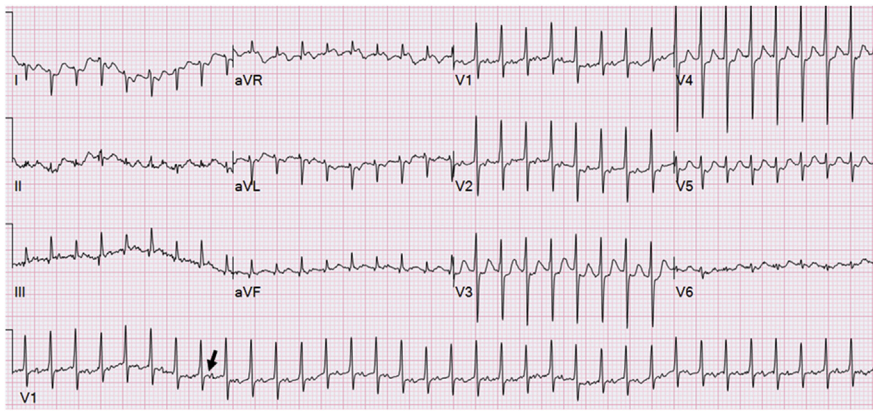

Fig. 5. Neonatal AVRT with a ventricular rate of 265 bpm. P waves are seen in the ST segment in leads III and V1 (*arrow*). The short RP interval is typical for this arrhythmia.

seen if there is anterograde pathway conduction during sinus rhythm (Wolff-Parkinson-White syndrome).

Close observation without drug therapy may be a safe approach for fetuses with infrequent, brief SVT episodes, because heart failure rarely develops unless the arrhythmia is very fast and/or becomes more persistent. In contrast, fetuses with incessant tachyarrhythmia tend to develop heart failure with hydrops if left in tachycardia. Fetal hydrops, cardiovascular collapse, and death are strongly associated with incessant AVRT but even intermittent tachycardia may have serious consequences.[27] In retrospective studies, 40% of fetuses with AVRT presented with hydrops and this was associated with a perinatal mortality between 21% and 27%. In contrast, the rate of perinatal mortality was less than 5% for those cases without hydrops.[26,27] Rapid pharmacologic cardioversion to a normal sinus rhythm is therefore most pressing for hydropic fetuses with incessant tachyarrhythmia. Moreover, fetal hydrops resolves over time once the cardiac rhythm is normalized by antiarrhythmic therapy. Possible medications to treat fetal SVT until birth include maternal digoxin, flecainide, or sotalol either alone or in combination, whereas amiodarone and direct fetal therapy are usually reserved for treatment-resistant, poorly tolerated tachycardia. Newborns in sinus rhythm may not require postnatal antiarrhythmic treatment but close observation for 2- 3 days and thereafter for SVT recurrence is advised. Neonatal incessant AVRT warrants acute cardioversion by vagal stimulation (facial immersion in iced water or ice pack), rapid intravenous bolus injection over 1 to 2 seconds of the short-acting adenosine immediately followed by a rapid saline flush, transesophageal atrial overdrive pacing, and/or additional antiarrhythmic medication, whereas electrical cardioversion is rarely required. Prophylactic antiarrhythmic treatment (ie, with propranolol) is then often used to prevent SVT recurrence during the first 6 to 12 months of life or longer. In our experience, AVRT associated with manifest pathway conduction during sinus rhythm (Wolff-Parkinson-White syndrome) is more likely to persist and to require long-term postnatal antiarrhythmic treatment.[28]

Atrial Flutter

AF is sustained by a circular macroreentrant pathway within the atrial wall, whereas the AV node is not part of the reentry circuit (see **Fig. 4**C). Atrial rates range between 300 and 500 bpm, which is commonly associated with 2:1 AV conduction and

ventricular rates between 150 to 250 bpm.[29] Normal or near-normal ventricular rates are observed in AF with slower 3:1 or 4:1 AV conduction.

ECG diagnosis of AF is straightforward with saw-tooth flutter waves that are best seen in leads II, III, and aVF (Fig. 6). In the absence of structural heart disease, AF is almost exclusively observed in babies during the third trimester or at birth. AF is usually tolerated and fetal hydrops and death are uncommon. Sotalol or digoxin is the first-line medication to treat fetal AF.[26] The treatment aim is either to suppress the arrhythmia or, if this is not achieved, to slow the ventricular rate to a more normal heart rate. If AF persists to birth, sinus rhythm can be restored by transesophageal overdrive pacing or synchronized electrical cardioversion. Neonatal recurrence of AF is unusual and long-term treatment is rarely required.

Other

Other tachyarrhythmia mechanisms are less common and may be difficult to differentiate from each other without ECG. Before birth, sinus tachycardia, PJRT, and AET present similarly as long VA tachycardia with heart rates less than 220 bpm. Sinus tachycardia is usually 20 to 30 bpm slower than AET and PJRT and characterized by atrial rates of less than 200 bpm, normal 1:1 AV conduction, and some variability of the fetal heart rate (see Fig. 4A). Sinus tachycardia greater than 200 bpm is occasionally seen in critically ill babies. A variety of fetal and maternal conditions may be responsible for sustained sinus tachycardia, including distress, anemia, and infections. The importance of sinus tachycardia is recognizing and treating the underlying cause. PJRT is an AV reentry tachycardia with a fairly slow retrograde conducting accessory pathway, which explains the long VA pattern (see Fig. 4D). The typical ECG pattern is that of an incessant tachycardia with long RP intervals and inverted P waves in the inferior leads II, III, and aVF. Spontaneous resolution of PJRT is unusual. Definitive treatment of PJRT is possible with radiofrequency ablation of the pathway in later life.

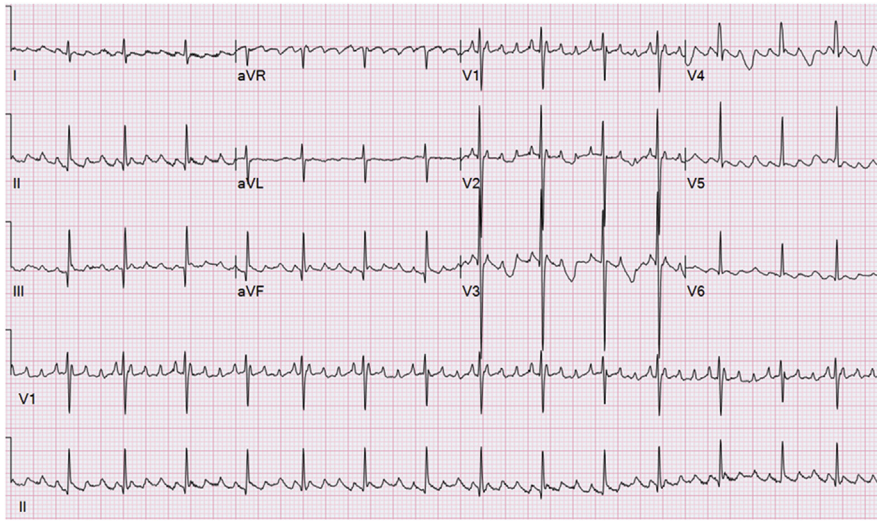

Fig. 6. Neonatal AF with an atrial rate of 460 bpm and a ventricular rate of 115 bpm caused by 4:1 AV conduction. Saw-tooth flutter waves are best seen in leads II, aVF, and V1.

Atrial Ectopic Tachycardia

AET (Fig. 7) arises from an ectopic focus within the atria and is most commonly sustained. During AET, intermittent changes in tachycardia rate with warming up and cooling down may be observed. Although AET is usually 1:1, conduction delay with AV block may be seen. Short-term antiarrhythmic treatment is often required but AET usually resolves before 6 months of life.

Ventricular Tachycardia

VT is a rare arrhythmia in babies. The fetal echocardiogram shows a tachycardia less than 200 bpm that is often incessant on presentation. The ventricular rate is higher than the atrial rate and there is no clear relation between ventricular and atrial events (AV dissociation). The postnatal definition of VT is that of a ventricular rate that is greater than or equal to 120 bpm or 25% faster than the normal sinus rate. The term accelerated ventricular rate is used if the ventricular rate is less than 120 bpm. The characteristic ECG of VT shows QRS widening with a bundle branch block pattern as well as AV dissociation. VT needs to be differentiated from other rare mechanisms in infants that also produce wide QRS tachycardia, including SVT/AF with bundle branch block and SVT with anterograde conduction across an accessory pathway. If the QRS in the tachycardia is wide, then the diagnosis of VT is more likely.

When evaluating a fetus or newborn for VT, possible causes include viral and anti-Ro antibody-mediated myocarditis; cardiac tumors; structural heart disease; hereditary cardiomyopathy, including LQTS; and electrolyte imbalance. In the absence of an identifiable predisposing condition, perinatal VT is often a benign finding (idiopathic VT). Treatment and prognosis depend on the VT mechanism and pattern, the hemodynamic impact, and associated conditions. Before birth, short-term maternal intravenous magnesium has been recommended as first-line medication for VT greater than 200 bpm.[6] Other treatments to acutely control VT may include intravenous lidocaine, oral β-blocker, and mexiletine. In the absence of LQTS, amiodarone, flecainide, or sotalol may also be useful.

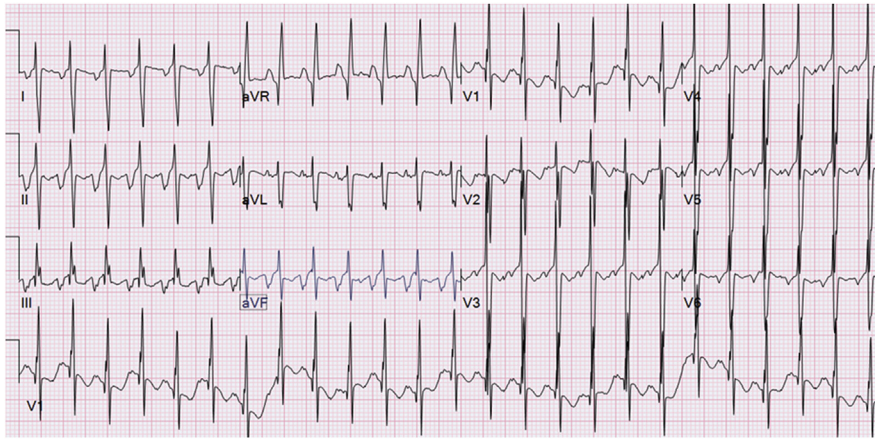

Fig. 7. Incessant AET with a ventricular rate of 190 bpm. The RP interval is prolonged and there are deeply inverted P waves in leads I, II, and III.

SUMMARY

Clinically relevant rhythm disorders are not common but may be associated with significant morbidity and mortality. Precise diagnosis is required before treatment is considered. AVRT is the most common type of SVT in fetuses and neonates. Antiarrhythmic treatment up to 12 months may be needed to prevent recurrence. CHB is the most common cause of persistent slow heart rate. Most survivors to birth require postnatal ventricular pacing.

REFERENCES

1. Jaeggi E, Fouron JC, Fournier A, et al. Ventriculo-atrial time interval measured on M mode echocardiography: a determining element in diagnosis, treatment, and prognosis of fetal supraventricular tachycardia. Heart 1998;79(6):582–7.
2. Fouron JC. Fetal arrhythmias: the Saint-Justine Hospital experience. Prenat Diagn 2004;24(13):1068–80.
3. Mitchell JL, Cuneo BF, Etheridge SP, et al. Fetal heart rate predictors of long QT syndrome. Circulation 2012;126(23):2688–95.
4. Nii M, Hamilton RM, Fenwick L, et al. Assessment of fetal atrioventricular time intervals by tissue Doppler and pulse Doppler echocardiography: normal values and correlation with fetal electrocardiography. Heart 2006;92(12):1831–7.
5. Nagashima M, Matsushima M, Ogawa A, et al. Cardiac arrhythmias in healthy children revealed by 24-hour ambulatory ECG monitoring. Pediatr Cardiol 1987;8(2):103–8.
6. Donofrio MT, Moon-Grady AJ, Hornberger LK, et al. Diagnosis and treatment of fetal cardiac disease: a scientific statement from the American Heart Association. Circulation 2014;129(21):2183–242.
7. Sonesson SE, Eliasson H, Conner P, et al. Doppler echocardiographic isovolumetric time intervals in diagnosis of fetal blocked atrial bigeminy and 2:1 atrioventricular block. Ultrasound Obstet Gynecol 2014;44(2):171–5.
8. Raboisson MJ, Fouron JC, Sonesson SE, et al. Fetal Doppler echocardiographic diagnosis and successful steroid therapy of Luciani-Wenckebach phenomenon and endocardial fibroelastosis related to maternal anti-Ro and anti-La antibodies. J Am Soc Echocardiogr 2005;18(4):375–80.
9. Horigome H, Nagashima M, Sumitomo N, et al. Clinical characteristics and genetic background of congenital long-QT syndrome diagnosed in fetal, neonatal, and infantile life: a nationwide questionnaire survey in Japan. Circ Arrhythm Electrophysiol 2010;3(1):10–7.
10. Cuneo BF, Etheridge SP, Horigome H, et al. Arrhythmia phenotype during fetal life suggests long-QT syndrome genotype: risk stratification of perinatal long-QT syndrome. Circ Arrhythm Electrophysiol 2013;6(5):946–51.
11. Chockalingam P, Jaeggi ET, Rammeloo LA, et al. Persistent fetal sinus bradycardia associated with maternal anti-SSA/Ro and anti-SSB/La antibodies. J Rheumatol 2011;38(12):2682–5.
12. Lin JH, Chang CI, Wang JK, et al. Intrauterine diagnosis of heterotaxy syndrome. Am Heart J 2002;143(6):1002–8.
13. Berg C, Geipel A, Kohl T, et al. Atrioventricular block detected in fetal life: associated anomalies and potential prognostic markers. Ultrasound Obstet Gynecol 2005;26(1):4–15.
14. Jaeggi ET, Hornberger LK, Smallhorn JF, et al. Prenatal diagnosis of complete atrioventricular block associated with structural heart disease: combined

experience of two tertiary care centers and review of the literature. Ultrasound Obstet Gynecol 2005;26(1):16–21.
15. Jaeggi E, Laskin C, Hamilton R, et al. The importance of the level of maternal anti-Ro/SSA antibodies as a prognostic marker of the development of cardiac neonatal lupus erythematosus a prospective study of 186 antibody-exposed fetuses and infants. J Am Coll Cardiol 2010;55(24):2778–84.
16. Izmirly PM, Saxena A, Kim MY, et al. Maternal and fetal factors associated with mortality and morbidity in a multi-racial/ethnic registry of anti-SSA/Ro-associated cardiac neonatal lupus. Circulation 2011;124(18):1927–35.
17. Eliasson H, Sonesson SE, Sharland G, et al. Isolated atrioventricular block in the fetus: a retrospective, multinational, multicenter study of 175 patients. Circulation 2011;124(18):1919–26.
18. Saleeb S, Copel J, Friedman D, et al. Comparison of treatment with fluorinated glucocorticoids to the natural history of autoantibody-associated congenital heart block: retrospective review of the research registry for neonatal lupus. Arthritis Rheum 1999;42(11):2335–45.
19. Jaeggi ET, Fouron JC, Silverman ED, et al. Transplacental fetal treatment improves the outcome of prenatally diagnosed complete atrioventricular block without structural heart disease. Circulation 2004;110(12):1542–8.
20. Trucco SM, Jaeggi E, Cuneo B, et al. Use of intravenous gamma globulin and corticosteroids in the treatment of maternal autoantibody-mediated cardiomyopathy. J Am Coll Cardiol 2011;57(6):715–23.
21. Kelly EN, Sananes R, Chiu-Man C, et al. Prenatal anti-Ro antibody exposure, congenital complete atrioventricular heart block, and high-dose steroid therapy: impact on neurocognitive outcome in school-age children. Arthritis Rheumatol 2014;66(8):2290–6.
22. Hutter D, Silverman ED, Jaeggi ET. The benefits of transplacental treatment of isolated congenital complete heart block associated with maternal anti-Ro/SSA antibodies: a review. Scand J Immunol 2010;72(3):235–41.
23. Epstein AE, DiMarco JP, Ellenbogen KA, et al. ACC/AHA/HRS 2008 guidelines for device-based therapy of cardiac rhythm abnormalities: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Revise the ACC/AHA/NASPE 2002 Guideline Update for Implantation of Cardiac Pacemakers and Antiarrhythmia Devices) developed in collaboration with the American Association for Thoracic Surgery and Society of Thoracic Surgeons. J Am Coll Cardiol 2008;51(21):e1–62.
24. Lopes LM, Tavares GM, Damiano AP, et al. Perinatal outcome of fetal atrioventricular block: one-hundred-sixteen cases from a single institution. Circulation 2008;118(12):1268–75.
25. Jaeggi ET, Hamilton RM, Silverman ED, et al. Outcome of children with fetal, neonatal or childhood diagnosis of isolated congenital atrioventricular block. A single institution's experience of 30 years. J Am Coll Cardiol 2002;39(1):130–7.
26. Jaeggi ET, Carvalho JS, De Groot E, et al. Comparison of transplacental treatment of fetal supraventricular tachyarrhythmias with digoxin, flecainide, and sotalol: results of a nonrandomized multicenter study. Circulation 2011;124(16):1747–54.
27. Simpson JM, Sharland GK. Fetal tachycardias: management and outcome of 127 consecutive cases. Heart 1998;79(6):576–81.
28. Gilljam T, Jaeggi E, Gow RM. Neonatal supraventricular tachycardia: outcomes over a 27-year period at a single institution. Acta Paediatr 2008;97(8):1035–9.
29. Jaeggi E, Fouron JC, Drblik SP. Fetal atrial flutter: diagnosis, clinical features, treatment, and outcome. J Pediatr 1998;132(2):335–9.