Fetal Arrhythmia and Related Fetal and Neonatal Outcome

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ABSTRACT

The evolution of fetal echocardiography (Motion and pulsed Doppler modes) has made possible the antenatal diagnosis of cardiac rhythm anomalies. Fetal cardiac arrhythmias are common anomalies, 20% of which are potentially dangerous. M-mode (motion-mode) echocardiography recording of the fetal heart cannot identify electrical events in the heart, but does depict the mechanical events that succeed them. Pulsed Doppler echocardiography identifies possible arrhythmias over a period of time, allowing for the measurement of these time intervals, a parameter absolutely necessary for the classification of various types of cardiac arrhythmias. Irregular cardiac rhythm (premature atrial contraction, premature ventricular contractions) is the most common disorder seen in practice in specialized fetal echography. These anomalies are usually benign and resolve spontaneously until delivery, thus do not require treatment. Fetal tachyarrhythmia (increased heart rate of 160–189 bpm) is detected by echocardiography, with a wide range of rhythm disorders. It is imperative to understand the mechanism of tachyarrhythmia, so that an accurate and rational management strategy can be formulated in each case. Fetal bradyarrhythmia is characterized by frequent irregular rhythm, low sustained heart rate (10 seconds–few minutes), below 110 bpm, or a combination of both. The diagnosis of the specific bradycardia depends on the echocardiogram findings and the atrioventricular conduction pattern. Once fetal bradycardia is noted, a quick fetal ultrasonographic examination should be performed to assess the normal fetal movements, fetal tone, thus contributing to the diagnosis of fetuses in distress, who require emergency delivery by cesarean section. This paper is an update of the diagnostic approaches in the current practice for different types of fetal heart rhythm disorders, the impact on the fetus and the newborn and their management.

Keywords: Doppler mode, echocardiography, Fetal cardiac arrhythmia, M mode.

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INTRODUCTION

The evolution of fetal echocardiography has made possible the antenatal diagnosis of cardiac rhythm anomalies—M and pulsed Doppler modes—playing an important role in identifying complex arrhythmias and monitoring the response in case of therapeutic intervention.

Fetal cardiac arrhythmias are common anomalies that occur with an incidence of 1–2% in the general unselected pregnant population¹ during routine ultrasounds, but only 20% are potentially dangerous.

Irregular fetal cardiac rhythm is the most common disorder seen in practice in specialized fetal echography and echocardiography centers, the vast majority being nonrepetitive atrial extrasystoles, not requiring therapy and re-evaluation. In 10% of cases, repetitive prolonged tachycardia or bradycardia is seen, requiring serial re-evaluations (in order to rule out structural anomalies) and therapeutic intervention, as they increase neonatal morbidity and mortality.²

This chapter is an update of the diagnostic approaches in the current practice for different types of fetal heart rhythm disorders, the impact on the fetus and the newborn and their management.

FETAL RHYTHM ASSESSMENT

M-mode Echocardiography

M-mode echocardiography (motion-mode) can be performed starting from the end of the 16th week of gestation until delivery (Fig. 1).

M-mode recording of the fetal heart cannot identify electrical events in the heart, but does depict the mechanical events that succeed them.

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Information obtained from real time M-mode echocardiography includes:

- Cardiac rate (both atrial and ventricular rates). Each atrial contraction is followed by a ventricular contraction (Fig. 2)
- Cardiac arrhythmias
- Motion of the atrioventricular and semilunar valves
- Ventricular cavity motion
- Cardiac wall thickness and motion
- Small pericardial effusions (separation of parietal and visceral pericardium).

M-mode echocardiogram can be obtained from many planes:³

- Plane through the left and right atrial chambers
 - Size and contraction of the atrial chambers only
 - Movement of foramen ovale.

Fig. 1: M-mode—ventricular contraction (v)—superior wave, atrial contraction (a)—inferior wave, between them, the movements of the interventricular septum (arrow)

- Plane through the left and right ventricles
 - One can recognize the anterior wall of the near field ventricle, the atrioventricular valves, the interventricular septum, the distal ventricle and its posterior wall.
 - The ventricular rate can be determined but one would not know the atrial rate nor could one ascertain whether atrial contractions are transmitted to the ventricles.
- Plane through the left/right atrium, semilunar valve, left/right ventricle or through the right atrium, semilunar valve and left ventricle:
 - This is the most informative view and allows one to analyze both the rate of atrial and ventricular contractions as well as the transmission of an impulse from the atrium to the ventricle.
 - Different arrhythmias can be more easily differentiated.

Pulsed Doppler Echocardiography

Pulsed Doppler echocardiography provides valuable information in the assessment and diagnosis of fetal rhythm arrhythmias and is a classic method, preferably in conjunction with motion-mode echocardiography.

In order to obtain accurate Doppler indices in fetal echocardiography, the sample volume should be placed distal to the valves to be examined, the insonating angle should be between 15° and 20° of the direction of blood flow. Proper Doppler measurements should be obtained during fetal apnea.

Pulsed Doppler echocardiography³ has the ability to acquire simultaneous signals from atrial and ventricular contraction, which leads to the identification of possible arrhythmias over a period of time, allowing for the measurement of these time intervals, a parameter absolutely necessary for the classification of various types of cardiac arrhythmias. Measurements can be made by placing the Doppler gate across the aortic and mitral valves, pulmonary artery and vein, renal artery and vein, superior vena cava or aorta. When the assessment is performed synchronously on the aorta and superior vena cava, the

Fig. 2: M-mode with sinus rhythm: ventricular contraction (v)—superior wave, atrial contraction (a)—inferior wave, between them, the movements of the interventricular septum (arrow)

retrograde flow of the latter marks the beginning of the atrial systole and the onset of aortic forward flow marks the beginning of ventricular systole.

Figures 3 and 4 present Doppler indices frequently used in fetal echocardiography.

FETAL CARDIAC ARRHYTHMIA

General Considerations of Cardiac Automaticity

Cardiac automaticity represents heart's property to self-excite, to develop rhythmic stimuli. This automaticity is due to the existence of the embryonic myocardial tissue (nodal) that functions in a hierarchical order, successively as follows:

- Sinoatrial node (Keith-Flack) that develops sinus rhythm, normal heart rate
- Atrioventricular node (Aschoff-Tawara) generating nodal rhythm: When the sinus rhythm is suppressed, the Aschoff-Tawara node takes command, determining the nodal heart rate
- The bundle of His and Purkinje network, which generates idioventricular rhythm.

Synchronized depolarization and repolarization of atrial and ventricular myocardial tissue result in the contraction and relaxation of cardiac chambers, allowing the coordinated filling and emptying during a cardiac cycle. The normal fetal heart rhythm ranges between 120 bpm and 180 bpm.

Any derangement that occurs in these normal sequences or changes in heart rate causes changes in the filling and emptying of the cardiac chambers, which if persistent lead to myocardial dysfunction, low cardiac output and ultimately congestive heart failure manifested in the fetus by fetal hydrops.⁴

Classification of Fetal Cardiac Arrhythmia

Fetal arrhythmias are classified in three main groups:

- Irregular cardiac rhythm
- Fetal tachyarrhythmia
- Fetal bradyarrhythmia.

Fig. 3: Pulsed Doppler of tricuspid valve

Fig. 5: M-mode with sinus rhythm: premature blocked atrial contraction—the superior wave and the arrow, ventricular contraction which appears after a pause—the inferior wave and arrow, between them, the movements of the interventricular septum and valves

IRREGULAR CARDIAC RHYTHM

Irregular fetal heart rhythm is seen in 1–3% of pregnancies.⁴

Premature Atrial Contraction

The most common pathophysiology of irregular fetal cardiac rhythm disorders is premature atrial contractions (PACs). These PACs are explained by triggering an electrical stimulus in atrial cells before the normal atrial rhythm, thereby causing normal sinus beat reset.⁵ Depending on the degree of prematurity of the ectopic electrical discharge, the PACs may be conducted to the ventricles or may be blocked within the atrioventricular node. This manifests itself either as an extra heartbeat or as an incomplete missed beat on ultrasound examination (Figs 5 and 6).

Frequently blocked PACs will result in a markedly irregular cardiac rhythm with slowing of the average heart rate.

If every two beats is a non-conducted PAC-atrial bigeminy, this will result in a regular but bradycardic heart rate with a frequency between 70 bpm and 100 bpm, never below 65 bpm,⁴ requiring a mandatory differential diagnosis from other, more serious causes of fetal bradycardia such as complete heart block.

Fig. 4: Pulsed Doppler of pulmonary artery (PA) and superior vena cava (SVC). A represents the negative wave of SVC, the onset of the atrial contraction

Fig. 6: Premature atrial contractions

In 2% of cases,⁵ PACs were determined by structural cardiac defects, such as intracardiac tumors—rhabdomyomas⁴ or redundancy of the atrial septum, both detectable by echocardiography.

Premature Ventricular Contractions

Premature ventricular contractions (PVC) are unusual, usually benign. Serial echocardiographic assessment is recommended, as PVCs have been associated in some cases with myocarditis or long QT syndrome.^{4,5}

Pregnancy Management

In both situations of PACs, when conducted to the ventricles or blocked at the atrioventricular node, with increased or rare frequency, they are usually well-tolerated and resolve spontaneously with no need of treatment.⁵ Approximately 10–14%⁴ of fetuses may lead to supraventricular tachycardia (SVT), which is why such a case should be monitored once a week for the next 2–3 weeks to rule out the development of a major tachyarrhythmia.⁵

Isolated PVCs do not require treatment.⁵

In order to stop irregular heart rhythm disorders, the pregnant woman is advised to avoid excitatory factors such as nicotine, caffeine and stop β -mimetic administration for premature contractions when feasible.

When these rhythm disorders are common and do not disappear until delivery, it is recommended to perform a neonatal ECG.⁴

FETAL TACHYARRHYTHMIAS

Fetal tachycardia is a condition that occurs in approximately 0.4–0.6% of all pregnancies^{6,7} and presents with an increased heart rate of 160–180 bpm.⁴ Fetal tachyarrhythmia is detected by echocardiography, with a wide range of rhythm disorders, from sinus tachycardia⁴ to isolated extrasystoles, to ventricular and supraventricular tachycardia or atrial flutter.⁸

Sinus Tachycardia

In this situation, there is a 1:1 ratio of atrioventricular conduction, with a regular rate of 160–180 bpm and occurs most frequently in the context of fetal movements. Some maternal pathological conditions may cause this rhythm disorder, such as hypoxia, anemia, thyrotoxicosis, infections, and fever.⁹ In general, the onset and end of an episode of sinus tachycardia is not sudden and abrupt.

Isolated Extrasystoles

Isolated extrasystoles are the most common fetal arrhythmias (85%) and are often detected during a routine prenatal visit. They almost always have a supraventricular origin, only occasionally a junctional or ventricular origin. Most often they are self-limited, resolving completely pre or immediately after birth.

Ventricular Tachycardia

Ventricular tachycardia (VT) is a very rare rhythm disorder, representing less than 2% of fetal tachycardias¹⁰ and is correctly diagnosed by Doppler ultrasound and magnetocardiography.³ Ventricular rates are bigger than 180 bpm with atrioventricular dissociation, while the atrial rate is normal. The pathological conditions that determine these arrhythmias are: fetal myocarditis, long QT syndrome or ventricular tumors.⁴

- *Pregnancy management:* Conversion of fetal VT to a normal regular rate is achieved by intravenous maternal administration of lidocaine and magnesium^{3,10} and oral mexiletine, sotalol and amiodarone.¹¹
- *Ventricular tachycardia in newborns:* Ventricular tachyarrhythmias in the newborn are not only rare but also poorly understood. Several secondary ventricular tachyarrhythmias are described as metabolic abnormalities including hyperkalemia, hypoglycemia^{12–14} and hypoxia, so ruling out and correcting

these abnormalities are extremely important before using conventional anti-arrhythmic therapy.

Fetal Paroxysmal Supraventricular Tachycardia

Supraventricular tachycardia is considered the most common type of tachycardia and accounts for 60–90% of such cases.

Supraventricular tachycardia has a typical ventricular rate of ~230–280 bpm (Fig. 7), atrioventricular (AV) 1:1 conduction, and is often associated with an accessory AV conduction pathway leading to atrial contractions at a rate faster than the sinoatrial node,¹⁵ or a re-entry mechanism, in which there is a circular electrical current running between a fast-conduction accessory pathway, the ventricle, the AV node and the atrium in either direction.

Supraventricular tachycardia is rarely associated with intracardiac or extracardiac anomalies (as opposed to other tachyarrhythmias). Supraventricular tachycardia may be the clinical expression in the Ebstein anomaly,¹⁵ but in most cases, it is the sole pathology. In about 10% of cases, it can be associated with Wolfe-Parkinson-White syndrome.

Diagnosis is most commonly established incidentally during routine fetal ultrasound monitoring in the 2nd or 3rd trimesters and is established using M-mode echocardiography and fetal Doppler, thus being able to demonstrate the paroxysm of atrial tachycardia in the range of 230–280 bpm, often followed by an extrasystole.

When the heart rate remains elevated in the absence of treatment, diastole is shortened, thus decreasing atrial and ventricular filling time and increasing systemic venous volume load and central venous pressure,^{15,16} phenomena accompanied by reversible systolic dysfunction.^{17,18} Insufficient blood supply through the coronary arteries to the myocardium during heart diastole causes relative ischemia, a phenomenon leading to dilatation of the heart chambers, impairing the myocardial contractility according to Starling's law, eventually evolving toward massive AV valve regurgitation. These changes have been called "tachycardia-induced cardiomyopathy".^{17–19} The resulting heart failure causes fetal hydrops (Figs 8 and 9)—which rapidly leads to fetal death *in utero*.

Pregnancy management: Case management in fetal SVT is performed best within a multidisciplinary team, consisting of an obstetrician, a maternal-fetal medicine specialist, a neonatologist and, preferably, a pediatric cardiologist able to formulate a treatment plan.

Fig. 7: Supraventricular tachycardia—fetal heart rate 273 bpm—fetus 18 weeks of amenorrhea without hydrops associated

Fig. 8: Re-entry supraventricular tachycardia—fetal heart rate 273 bpm—fetus 22 weeks of amenorrhea without hydrops associated

Figs 9A and B: Supraventricular tachycardia—34 weeks of amenorrhea with hydrops associated

Following the diagnosis of a fetal SVT, it is recommended to perform a detailed fetal ultrasound to assess fetal growth according to gestational age to exclude any associated structural anomalies and the presence or absence of pericardial, pleural or ascites effusions. Echocardiographic assessment is recommended for atrial and ventricular rate, atrial and ventricular size, and the presence and extent of AV valve regurgitation.

The treatment plan depends on gestational age at the time of SVT diagnosis, the presence or absence of structural anomalies and the presence or absence of hydrops fetalis.

Pharmacological therapy is undertaken immediately if there is evidence of hemodynamic compromise or hydrops fetalis. Otherwise, the pregnant woman is monitored by continuous cardiotocography for assessing fetal heart rate in order to ascertain the proportion of time spent in tachycardia, with adoption of expectant management when arrhythmia is less than 50% of time and there is no evidence of fetal hemodynamic compromise.²⁰

In order to determine the mechanism of producing a general tachyarrhythmia and atrial flutter in particular, for the purpose of administering specific treatment according to its type—ventricular or junctional, whether or not associated with bradycardia or intermittent—it is recommended to perform fetal magnetocardiography (fMCG).

The purpose of pharmacological treatment is to intervene in the vicious circle by slowing the heart rate and synchronizing atrial and ventricular contractions. This can be achieved by blocking automaticity of the ectopic rapid pace-dictating focus or by blocking fast conduction through the accessory pathway. An effective treatment will lead to sinus rhythm and will restore the normal cardiac function.

The first line of choice for restoring an acceptable fetal heart rate is Digoxin. The digitization is performed in the absence of hydrops fetalis and placental hydrops, since the transplacental transfer of antiarrhythmic medication in the case of the hydrophobic placenta is absent. Antiarrhythmic treatment is administered either orally to the pregnant, or directly via the fetoscopy. Successful *in utero* cardioversion of sustained fetal tachycardia complicated by hydrops fetalis has improved from 60% to 85%^{21,22} with fetal and neonatal mortality from 8% to 30%.^{23,24} Potential contributing factors^{24,25} for high fetal and neonatal mortality include failure to achieve cardioversion *in utero* with subsequent progression of hydrops

fetalis, premature delivery, and possible increases in arrhythmia during neonatal period (proarrhythmia).

The second treatment line is amiodarone in the fetus with refractory tachycardia that tends to evolve to hydrops fetalis or myocardial dysfunction. There is no consensus on the most effective and safest antiarrhythmic agent for *in utero* tachycardia conversion.

Two other antiarrhythmic agents currently considered to be of great success in fetal cardioversion are sotalol and flecainide, both of which have significant mortality rates. Sotalol, a class III anti-arrhythmia agent, managed to convert tachycardia in 40–60% of SVT cases complicated by hydrops, but with a mortality rate of 25–30%^{26,27} and flecainide has a 60–85% efficacy, while the mortality rate is of 18.5% in similar cases.^{23,28–31}

In case of SVT diagnosed late in gestation, cesarean section delivery is recommended, followed by newborn tachycardia conversion.⁴

- *Supraventricular tachycardia in newborns*: The most important clinical signs of SVT in newborns include hypotension, heart failure and signs of shock—pallor, or decreased level of consciousness. Other manifestations include irritability, malnutrition, and tachypnea.

The diagnosis is confirmed by ECG, with heart rate being sustained more than or equal to 220 bpm with a QRS less than 0.08 seconds^{32,33} (Fig. 10). Circulatory collapse caused by SVT requires chemical or electrical cardioversion.

In the neonatal period, echocardiography should be performed to exclude antenatal undiagnosed structural anomalies, considering that according to different studies, 15.3% of cases are identified postnatally.^{32,34,35}

Supraventricular tachycardia treatment for newborns is a challenge. If the newborn is not in critical condition, it is advisable to first try vagal maneuvers and adenosine intravenous injections, which should be given in a fast bolus, in a large vein, at an initial dose of 50–100 µg/kg, which is the most effective medicine.³⁶ In cases of critically ill newborns, intravenous amiodarone therapy is recommended, with little negative inotropic effect and which is relatively safe for those with compromised myocardial function³⁷ (Table 1).

After the neonatal acute phase of SVT, it is recommended to continue with the oral medication (Table 2).

Fig. 10: Postnatal ECG—SVT with large QRS

Table 1: Acute drug therapy for SVT—Intravenous administration

Group	Drug
Nucleoside	Adenosine
β-blocker	Propranolol, esmolol
Cardiac glycosides	Digoxine
Class III antiarrhythmic agent	Amiodarone

Table 2: Ongoing drug therapy for SVT, oral administration

Group	Drug
β-blocker	Atenolol, metoprolol, nadolol, propranolol
Calcium channel antagonist	Diltiazem, verapamil
Cardiac glycoside	Digoxine
Class Ic antiarrhythmic agent	Flecainide, propafenone
Class III antiarrhythmic agent	Amiodarone, sotalol

If intravenous adenosine conversion was easily achieved, the optimal first line follow-up treatment is by oral long-acting β-blockers, which are safe, effective and do not require special monitoring.³²

For a newborn in whom chemical cardioversion was difficult or requiring electrical cardioversion, it is recommended that oral therapy can be continued with a more powerful antiarrhythmic drug such as sotalol, flecainide, or amiodarone, each requiring intensive monitoring.

In the first 12 months, spontaneous disappearance of tachycardia is expected in most cases, with the recommendation that prophylactic drug therapy can be continued until the end of the first year of life.³⁸ In medically refractory cases of oral antiarrhythmic medication, radiofrequency ablation can be employed successfully, but after 1 year of age.³⁹

Atrial Flutter

Atrial flutter is an uncommon condition that occurs in 0.4–0.6%⁴⁰ of all pregnancies, accounting for 26–29% of all fetal rhythm disorders.⁴¹

Fig. 11: Atrial flutter—fetal heart rate 484 bpm—fetus 28 weeks of amenorrhea

It is defined as a rapid regular tachycardia of 300–600 bpm (Fig. 11), originating in the body of the atrium, accompanied by variable AV conduction, with different degrees of AV block.⁴¹

It may occur in fetuses without structural heart abnormalities (20% of cases) such as AV septal defect, Ebstein's anomaly, hypoplastic left heart syndrome, and pulmonary atresia.^{42,43}

Atrial flutter has been associated with a poor prognosis with a higher mortality rate of approximately 10%, which may be due to its association with structural heart disease in 20% of cases^{20,41} and the increased incidence of developing hydrops fetalis, similar to the one in SVT.

- **Pregnancy management:** Prenatal management and prognosis of fetal atrial flutter (AF) depend on the gestational age at onset, the development of heart failure or hydrops, and the associated heart abnormalities. Unlike postnatal atrial flutter therapy, which aims at decreasing ventricular rate by increasing the degree of AV block and which may lead to a decrease in heart failure, control of the ventricular response rate in fetuses with severe hydrops

secondary to atrial flutter has not resulted in an improvement in fetal hemodynamics,²⁰ as the time to conversion to sinus rhythm is longer, and the transplacental passage of most antiarrhythmic agents is affected by the presence of hydrops fetalis.^{44–46} It takes an average treatment time of 10 days to obtain the conversion to sinus rhythm.^{47,48}

- *Atrial flutter in newborns:* Atrial flutter is a type of arrhythmia uncommon in newborns and infants. It is a type of SVT, characterized by rapid heartbeat (240–360 bpm), best evidenced in ECG, leads II, III, and aVF.

The management of this type of tachyarrhythmia can be achieved by administering sotalol at the dose of 160 mg, twice daily,⁴¹ amiodarone or flecainide,⁴⁹ after an optimal ventricular response with digoxin.

With the acquisition of sinus rhythm in newborns, the recurrence of atrial flutter is rare,⁴² therefore the expectative strategy may be an option, but considering the fact that there have been reported cases of late recurrence,⁵⁰ the follow-up period should be of at least 6 months. As a therapeutic safety alternative, prophylactic antiarrhythmic treatment should be administered for the first 6–12 months.^{42,51}

Lack of improvement or insufficient improvement after the antiarrhythmic treatment in newborn's atrial flutter^{20,52} is probably due to unique cardiovascular dynamics, the relatively restrictive ventricular myocardium, relatively volume-loaded right heart. Cardiac decompensation due to cardiac failure in newborns with atrial flutter reflects the diastolic rather than the systolic dysfunction. It is ideal to control the atrial flutter with resumption of a 1:1 AV contraction sequence. If this is not achieved, there will continue to be atrial contractions against a closed or partially closed AV valve, resulting in atrial pressure waves that keep an increased systolic venous pressure, edema and effusions.

Atrial Fibrillation

It is a rare type of fetal tachycardia, with a rapid and irregular rate, associated with AV block, which often cannot be distinguished from atrial flutter.

In conclusion, it is imperative to understand the mechanism of tachyarrhythmia, so that an accurate and rational management strategy can be formulated in each case.

FETAL BRADYARRHYTHMIAS

Fetal bradycardia is a cardiac rhythm disorder that affects over 2% of pregnancies, characterized by frequent irregular rhythm, low sustained heart rate (10 seconds–few minutes), below 110 bpm, or a combination of both. Once fetal bradycardia is noted, a quick fetal ultrasonographic examination should be performed to assess the normal fetal movements, fetal tone, thus contributing to the diagnosis of fetuses in distress, who require emergency delivery by cesarean section.

There are various types of fetal bradycardia—sinus bradycardia, blocked atrial ectopic beats and AV conduction disorders. The diagnosis of the specific bradycardia depends on the echocardiogram findings and the AV conduction pattern.

Sinus Bradycardia

Atria and ventricles beat at the same rate with electrical impulses originating in the sinus node.⁵³

- *Transient sinus bradycardia:* It occurs in brief episodes of 1–2 minutes of bradycardia, most commonly during the second trimester of pregnancy and is usually physiological, generated by the immaturity of the parasympathetic nervous system, which is unable to adequately respond to increased vagal activity.
- *New-onset sinus bradycardia:* It is a sign of severe fetal hypoxia caused by obstetric emergencies such as premature separation of a normally placed placenta, uterine rupture or prolapsed umbilical cord and is an indication for emergency delivery.
- *Sinus node dysfunction* is seen as a bradycardia of 80–120 bpm, caused by an abnormal positioned sinus node in the context of structural heart anomalies such as stereotaxic syndrome, interrupted inferior vena cava, and single ventricle heart physiology.
- *Long QT syndrome:* LQTS includes a diverse group of myocardial repolarization disorders that can cause fetal death *in utero*. When sustained fetal bradycardia episodes are noted and congenital heart block is ruled out, LQTS diagnosis should be considered and can usually be ruled out only by a postnatal ECG, with a prolonged QT interval. In LQTS etiology, multiple genetic mutations (congenital LQTS) have been described, so a detailed family history is vital in attempting to assess this bradycardia with a genetic predisposition.

Two clinical phenotypes of LQTS have been described:

- *Jervell syndrome and Lange-Nielsen* is an autosomal recessive disorder. Mutations in the *KCNE1* and *KCNQ1* genes have been identified that normally code the structure and normal function of potassium channels found in the heart muscle and internal ear, which under pathological conditions result in serious sensory hearing loss and LQTS syndrome.
- *Romano—Ward syndrome* is autosomal dominant and includes LQTS without extracardiac abnormalities. *KCNE1*, *KCNE2*, *KCNH2*, *KCNQ1* and *SCN5A* genes mutations have been identified that normally code the structure and function of sodium channels in the cardiac tissue.
- *Pregnancy management:* No fetal therapy is required unless bradycardia is the result of fetal distress and acute hypoxia, and emergency fetal extraction being absolutely necessary.
- *Sinus bradycardia in newborns:* Fetal sinus bradycardia, which persists postnatally, requires careful and complex assessment in a pediatric cardiology department. Newborns and infants with persistent sinus bradycardia may require a pacemaker.

For LQTS cases, β -blockers, cardiac pacemakers and/or defibrillators are possible treatments.

Persistent Atrial Bigeminy and Trigeminy

Blocked atrial ectopic beats are a form of arrhythmia caused by premature ectopic beats arising from a site other than the sinus node manifested as fetal bradycardia, bigeminy and persistent atrial trigeminy being included in this entity.^{3,53} The diagnosis is challenging if the ectopic beats occur with regularity and regularly generate an irregular rhythm. Atrial bigeminy results from ectopic beats that alternate with sinus beats, but with the lack of conduction of the ectopic beats.

It is essential to differentiate blocked premature beats from AV block because the prognostic and therapeutic strategies are

different. The time intervals between consecutive atrial impulses remain relatively constant in the AV block compared with shortened atrial impulses on every second or third beat in bigeminy or trigeminy.

For the diagnosis of blocked premature atrial contractions, fetal therapy is not required.

Congenital Atrioventricular Heart Block

Congenital heart block^{3,53} is the delay or interruption in electrical conduction within the AV node, bundle of His or bundle branches, and occurs in approximately 1 out of 15,000 live births.

- First-degree block refers to a prolonged AV interval with a 1:1 lead pattern between the atria and ventricles, with a normal fetal heart rate. First-degree block may be a precursor for 2nd- and 3rd-degree blocks.
- Second-degree block allows some, but not all atrial impulses to penetrate the AV node, bundle of His and its branches, the atrial contraction rate being regular.
 - Second-degree block Type I (Wenckebach) involves a progressive lengthening of the AV interval, followed by a blocked impulse, resulting in an irregular ventricular rate. This is usually due to an abnormal conduction of the AV node.
 - Second-degree block Type II (Mobitz) refers to a normal AV interval with intermittently nonconducted impulses resulting in a slow irregular ventricular contraction rate.³ The intermittent lack of conduction represents a disease within the bundle of His and its branches and may progress to third-degree heart block.
- Third-degree block occurs when there is a complete disruption of the AV conduction, and the atria and ventricles beat independently, this phenomenon is usually due to dysfunction of the AV node and/or bundle of His or one of the branches.

The most significant type⁵⁴ is the congenital complete heart block (CHB). 50% of cases with this fetal rhythm disorder⁵⁵ come from pregnancies with associated maternal pathology, the most common of which is connective tissue disease such as systemic lupus erythematosus^{56,57} and Sjögren™ syndrome with transplacental passage of two specific antibodies (Anti-Ro and anti-La), which cause AV lesions. In the other 50% of CHB cases, congenital heart defects, including atrial isomerism, ventricular inversion and heterotaxy syndromes^{56,58} are involved. The diagnosis is established by echocardiography, showing a normal atrial rate with slower ventricular rate and AV dissociation^{54,59} (Fig. 12). In most cases, fetal heart rate is 45 bpm.

The prognosis depends on the presence or on the absence of structural heart disease and the secondary development of hydrops fetalis due to the very slow heart rate.^{59,60}

- *Pregnancy management:* In CHB cases caused by diseases of the maternal conjunctive tissue, the treatment consists of corticosteroids, in order to counteract the associated myocarditis and to improve the degree of AV block but with poor, limited results.^{61,62} It is used in cases with prolonged PR intervals in an attempt to prevent progression to the complete heart block.^{63,64}

In complicated cases with hydrops fetalis, sympathomimetic medications such as terbutaline, isoprenaline, ritodrine and salbutamol are recommended, which result in increase in heart rate and variable hydrops improvement.⁶⁵ The delivery of fetuses

Fig. 12: Fetal bradycardia by AV block—pregnant woman with systemic lupus erythematosus and anti-Ro antibodies present

diagnosed with CHB should be carried out in a tertiary care center that facilitates access to emergency heart stimulation techniques.

- *Complete heart block or third-degree atrioventricular block in newborns:* Neonatal first-degree CHB is most commonly associated with AV conduction disorders and is usually determined by congenital heart disease or inflammatory disorders of the myocardium. Most newborns are asymptomatic and do not require treatment.^{54,66}

Neonatal second-degree CHB is generally determined by maternal connective tissue disease, and in some cases, it is necessary to place a permanent pacemaker.^{54,66}

The prognosis for neonatal third-degree CHB depends on etiology, clinical situation, gestational age and associated structural heart defects. Specifically, the prognosis of the complete heart block is extremely poor when there are major structural heart anomalies and/or hydrops associated. In these situations, the risk of neonatal death is increased. Negative predictive factors are slow ventricular rate, dilated cardiomyopathy, tricuspid regurgitation and pericardial effusions.

The prognosis of the heart block caused by maternal conjunctive tissue disease without associated hydrops is better, but newborns and infants may need a pacemaker after birth or later in life.

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