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REVIEW ARTICLE

Diagnosis and management of common fetal arrhythmias

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Abstract Fetal arrhythmias are detected in at least 2% of unselected pregnancies during routine obstetrical scans. Most common are transient, brief episodes of a slow or fast heart rate or of an irregular heart rhythm. Less common are prolonged or persistent abnormalities such as supraventricular tachycardia and complete heart block which may lead to low cardiac output, fetal hydrops and demise. The objectives of this review are to update the reader on the diagnosis and management of the more common arrhythmias.

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1. Introduction

The cardiac conduction system is composed of highly specialized tissue that is able to rhythmically generate electrical impulses in the sinus node and to propagate the electrical current across the atriums, the AV-node and the His-Purkinje system throughout the ventricles. Synchronized depolarization and repolarization of atrial and ventricular myocardial tissue result in rhythmic contraction and relaxation of these cavities,

allowing the coordinated filling and emptying of the heart during each cardiac cycle. Depending on the level of fetal activity, the typical fetal heart rate ranges between 120 and 180 beats per minute (bpm) with little beat-to-beat variation. Fetal arrhythmia may be defined as an irregularity of the cardiac rhythm, as an abnormally slow (< 100 bpm) or fast (> 180 bpm) heart rate, or as a combination of irregular rhythm and abnormal heart rate. Fetal arrhythmias are detected in at least 2% of unselected pregnancies during routine obstetrical scans and are a common reason for referral to the fetal cardiologist. The goal of this review is to discuss the diagnosis, clinical manifestation and management of the most common fetal rhythm abnormalities.

2. Echocardiographic assessment of the fetal heart rhythm

After birth, the etiological classification of a newly detected cardiac arrhythmia relies primarily on characteristic electrocardiographic findings including the rate, morphology and chronology of atrial and ventricular electrical events. In the fetus, the assessment of arrhythmias becomes more challenging as conventional real-time electrocardiogram (ECG) is not obtained. Fetal ECG and fetal magnetocardiography are only available in a few centers world-wide and are mainly used as research tools. Alternatively, fetal M-mode and Doppler echocardiography can be used to study the rate and timing of atrial and ventricular mechanical events which occur briefly after their respective electrical depolarization (Jaeggi et al., 1998a; Fouron et al., 2000; Rein et al., 2002). Fetal echocardiography does not inform about the morphology of electrical complexes (e.g. P-wave axis) nor on QRS- and QT-interval durations. Despite these limitations, with some ultrasound experience and a basic understanding of electrophysiological principles, it is almost always possible to correctly identify the arrhythmia mechanism based on characteristic echocardiographic pattern (Jaeggi et al., 1998a; Fouron et al., 2003; Fouron, 2004).

Figs. 1a and 1b show examples of M-mode and pulsed-wave Doppler recordings of a normal fetal cardiac rhythm. To record sequences of atrial and ventricular events by myocardial M-mode imaging it is important to align the M-mode cursor simultaneously through the atrial and ventricular walls.

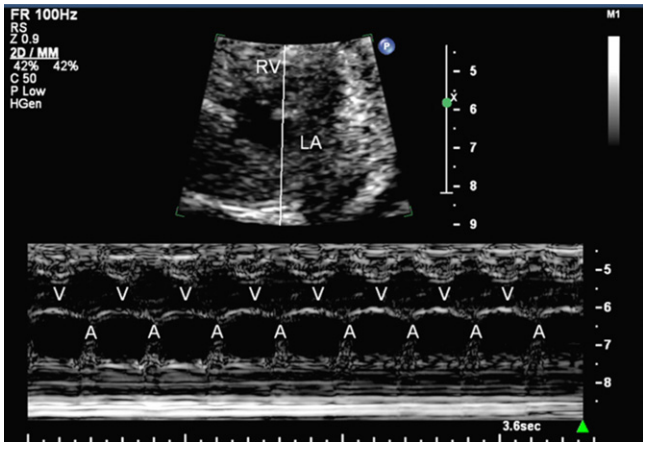

Figure 1a M-mode tracing using a four-chamber view to demonstrate atrial (A) and ventricular (V) wall motions. The M-mode beam is directed through the left atrium (LA) and right ventricle (RV).

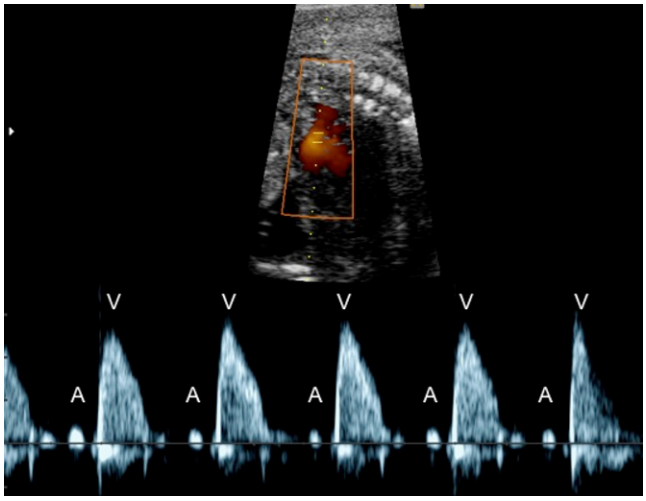

Figure 1b Example of a normal Doppler tracing obtained by simultaneous pulse wave Doppler of the aorta and the SVC. The Doppler sample volume is simultaneously placed within the aorta and the SVC.

The preferred M-mode beam direction is e.g. in the cardiac 4-chamber view simultaneously through the atrial free wall and the opposite ventricular free wall close to the AV junction because these parts show the most pronounced lateral excursion during the cardiac cycle. This approach is particularly relevant to assess hearts with reduced wall motion. A main limitation of M-mode is that mechanical events are not well-defined on M-mode recordings. Therefore, it may not be possible to achieve precise measurements of time intervals between mechanical events. Doppler flow signals are far better delineated which makes Doppler echocardiography a superior modality for the assessment of the fetal cardiac rhythm and AV-conduction. Simultaneous recording of the vena cava superior (SVC) and the ascending aorta Doppler flow is a particularly useful imaging tool for the detailed assessment of any arrhythmia. Anatomically the SVC and aorta are closely-related. This is best illustrated in the sagittal plane by 2-dimensional echocardiography. By placing the Doppler sample volume simultaneously within both vessels it becomes possible to concurrently record their blood flow patterns. The normal SVC flow consists of three flow waves which are the larger S and D waves that are directed towards the heart during ventricular systole (S-wave) and in early diastole (D-wave) followed by a brief retrograde a-wave. The a-wave is explained by intermittent flow reversal into the vena cava at the time the atrium contracts (atrial systole) which is immediately after atrial depolarization. The a-wave can therefore be considered as the mechanical equivalent of the electrocardiographic P-wave. The aortic ejection flow from the ascending aorta Doppler correlates with ventricular systolic contraction that follows electrical ventricular depolarization which corresponds to the electrocardiographic QRS complex. Using SVC-aorta Doppler, the timing between atrial and ventricular events can therefore be measured from the onset of the venous a-wave (A) to the onset of the ascending aorta flow (V) and then compared to published reference data. Despite differences in delay between electrical and mechanical activation of atriums and ventricles, there is a good correlation between echocardiographic AV-

durations and electrical PR-durations (Nii et al., 2006). AV-intervals physiologically prolong with advancing fetal age (Andelfinger et al., 2001; Nii et al., 2006).

As previously mentioned, the normal fetal heart beats regularly somewhere between 120 and 160 bpm. Each atrial event is followed by a ventricular event which occurs within a well-defined time-interval (normal 1:1 AV conduction). By means of echocardiography it is possible to accurately differentiate most fetal arrhythmias. To be able to do this it is crucial: (1) to obtain good quality M-mode or Doppler recordings to identify atrial and ventricular events; (2) to systematically analyze the regularity and rate of atrial–atrial (A–A) and ventricular–ventricular (V–V) events and to determine the relation between atrial and ventricular beats (AV-chronology; AV-conduction ratio); and, (3) to examine the impact of the arrhythmia on fetal well-being. Major arrhythmias that result in persistently faster or slower heart rates may be associated with poor ventricular function and fetal hydrops. Moreover, there is a strong association between some types of fetal arrhythmias and structural heart disease for which the fetus should also be examined by detailed echocardiography.

3. Irregular rhythm

Irregularities of the cardiac rhythm belong to the most common rhythm anomalies at any age. In the fetus this finding is almost always associated with isolated premature atrial contractions (PACs). PACs are explained by atrial cells that electrically fire before the normal atrial beat is to occur which resets the normal sinus beat. Depending on the degree of prematurity of the ectopic event, a PAC may be conducted to the ventricles or be blocked within the AV node and thus manifest either as extra beat or as missed beat on auscultation or ultrasound examination (Fig. 2). Frequently blocked PACs will result in a markedly irregular cardiac rhythm with slowing of the average heart rate. If every 2 beat is a non-conducted PACs (atrial bigeminy) this will result in a regular bradyarrhythmia between 60 and 80 beats/min and needs to be differentiated from more serious causes of fetal bradycardia such as complete heart block (see below). Irrespective of whether PACs are conducted or blocked and are frequent or rare, they are usually well-tolerated and resolve spontaneously with no need of treatment. In rare cases, nevertheless, progression to supraventricular tachycardia is possible. Therefore, intermittent heart rate monitoring (e.g. once per week for the next 2–3 weeks by the obstetrician) is recommended to exclude the development of a major tachyarrhythmia. PACs have also been associated with structural heart disease in up to 2% of cases, intracardiac tumors and redundancy of the atrial septum which are detectable by echocardiography. Premature ventricular contractions (PVCs) are an uncommon, usually benign cause of an intermittently irregular heart rhythm in the fetus. Still, serial echocardiographic assessment is advised as PVCs have been associated with myocarditis and long QT syndrome. Treatment for isolated PVCs is not required.

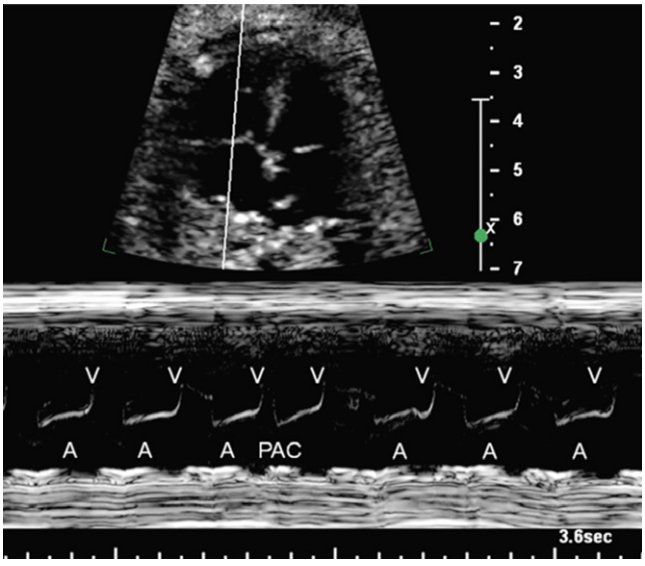

Figure 2 Conducted PAC. M-Mode tracing from a four chamber view with the transducer beam through the RA and the RV. There is a conducted PAC followed by an early ventricular contraction (V).

4. Fetal bradycardia

Fetal bradycardia is conventionally defined by a decrease in heart rate < 100 bpm although a heart rate persistently slower than 120 beats/min is not a normal finding. Brief heart rate deceleration of less than 1–2 min is common and, in general, without clinical relevance. More concerning is the observation of sustained bradycardia which typically relates to one of the following mechanisms: (1) sinus bradycardia; (2) atrial bigeminy; and, (3) complete heart block (CHB).

Sinus bradycardia secondary to advanced fetal hypoxia is an obstetrical emergency: survival depends on expedited delivery. Cardiac etiologies of sinus bradycardia are less common but include long QT-syndrome due to extremely prolonged repolarization and the congenital absence or dysfunction of the sinus node e.g. in left isomerism of the atrial appendage (Ho et al., 1995). The cardiac causes are distinguished based on tell-tale echocardiographic and postnatal electrocardiographic findings.

Multiple non-conducted PACs may result in an irregular, slow ventricular rate below 100 bpm. The heart rate becomes regular if every second beat is a blocked PAC which defines

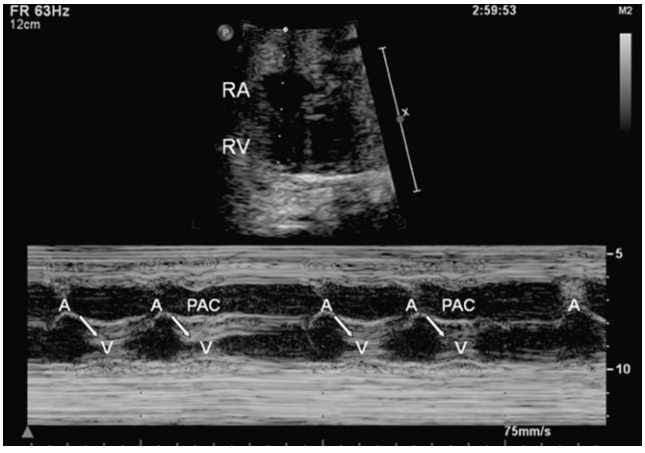

Figure 3 Blocked PAC. M-Mode tracing from a four chamber view with the transducer beam through the RA and the RV. There are blocked PAC's in a trigeminy pattern. Only the normal atrial contractions (A) are followed by a ventricular contraction (V).

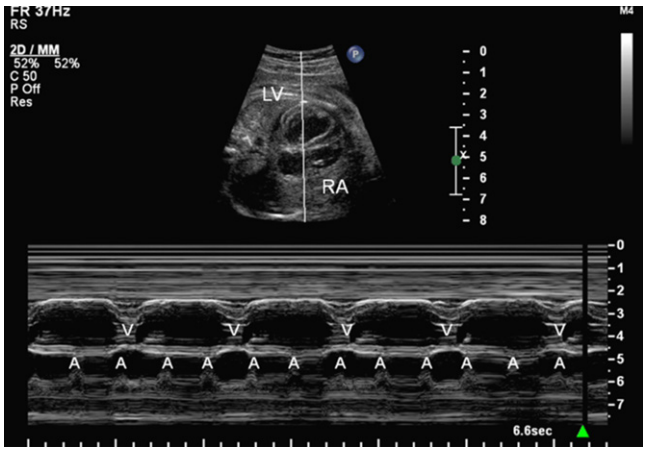

Figure 4a Complete heart block. M-Mode tracing from a four chamber view with the transducer beam through the RA and the LV. There are regular atrial contractions (A) with a normal heart rate of 125 bpm and independent ventricular contractions (V) with a heart rate of 50 bpm.

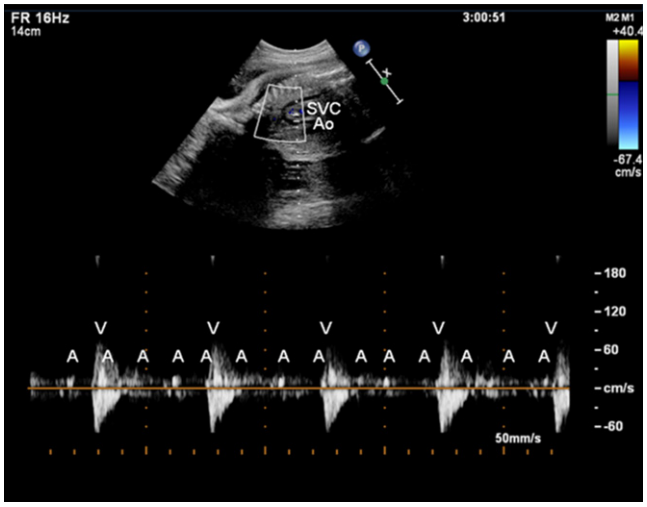

Figure 4b Simultaneous pulse wave Doppler of the aorta and vena cava superior of a fetus with complete heart block showing backflow following atrial contractions (A) in the superior vena cava with an atrial rate of 140/min (A) and in the same direction aortic outflow following ventricular contractions (V) which are independent with a regular rate of 60/min.

non-conducted atrial bigeminy (Fig. 3). On the M-mode or Doppler recording of atrial bigeminy, the atrial rate is irregular (alternating sinus and premature beats) while the ventricles beat at a regularly slow pace (60–80 bpm) that is half of the atrial rate. Atrial bigeminy may persist for hours but is clinically benign and ultimately will resolve without treatment.

Irreversible *complete heart block,* the most common fetal presentation of a conduction disorder, accounts for almost half of all major fetal arrhythmias seen by Fetal Cardiology. On echocardiography, the atrial rate is normal and regular but the ventricles beat independently at a much slower rate (40–80 bpm) due to the failure of electrical AV conduction (Fig. 4). Heart block is most commonly associated either with structural heart disease or with maternal anti-Ro autoantibodies. The condition carries a significant mortality risk as the fetus needs to overcome the slow ventricular rate, the loss of coordinated atrial contribution to ventricular filling, and perhaps concomitant heart disease or carditis. The detection of a major structural heart disease, fetal hydrops, poor contractility, and ventricular rates below 50 bpm are all associated with a poor pregnancy outcome.

The most common association of fetal CHB with structural heart disease is unbalanced atrioventricular septal defect associated with left isomerism, which is almost universally lethal, independent of the choice of perinatal care. Fetal CHB without structural heart disease has a better prognosis and is mainly related to the transplacental passage of maternal autoantibodies directed to fetal Ro/SSA ribonucleoproteins. Anti-Ro antibodies are present in about 2% of pregnant women. In a similar percentage (1–2%) of fetuses these antibodies will trigger the inflammation of the AV node and the myocardium. The inflamed tissues may then heal with fibrosis which may cause heart block, endocardial fibroelastosis, and dilated cardiomyopathy. Heart block, the most common antibody-related fetal cardiac complication, typically appears between 18 and 24 weeks in fetuses exposed to high anti-Ro antibody levels (Jaeggi et al., 2010). The rationale to treat isolated fetal CHB in-utero is primarily to temper the antibody-mediated myocardial inflammation, to augment fetal cardiac output and to improve survival. Maternal dexamethasone administration has been shown to improve incomplete fetal AV block, myocardial dysfunction, and cavity effusions. Beta-sympathomimetics such as salbutamol and terbutaline may be used to increase the fetal heart rate and myocardial contractility. Published data from the Hospital for Sick Children in Toronto (Jaeggi et al., 2004) suggest an improved survival above 90% for antibody-related CHB if maternal high-dose dexamethasone was initiated at the time of anomaly diagnosis and was maintained during the pregnancy and if a β-adrenergic drug was added at fetal heart rates below 50–55 bpm.

Despite the improvement in outcome, the use of fluorinated steroids remains debated because a significant proportion of fetuses with immune-mediated CHB will also survive without anti-inflammatory steroid treatment and because the steroids increases the risk of oligohydramnios, fetal growth restriction, maternal hypertension and insulin-dependent diabetes. To minimize the risk of adverse events the treatment guidelines for immune-mediated heart block at the co-authors center has been modified in recent years. Dexamethasone, which is started at 8 mg/day, is now reduced to 4 mg/day after two weeks and then to 2 mg/day after 28 gestational weeks in uncomplicated cases. On the other hand, we do not recommend transplacental treatment for late diagnosis after 32 weeks with asymptomatic isolated fetal CHB and ventricular rate above 50 bpm. Delivery, usually by caesarian section, is recommended in a tertiary care facility with expertise in the postnatal management of cardiac patients.

5. Fetal tachycardia

Sinus tachycardia (ST), supraventricular tachycardia (SVT) and atrial flutter (AF) are the main causes of fetal tachycardia. Echocardiography is the diagnostic cornerstone in differentiating these mechanisms and in evaluating the hemodynamic impact. All forms of persistent tachyarrhythmia may be

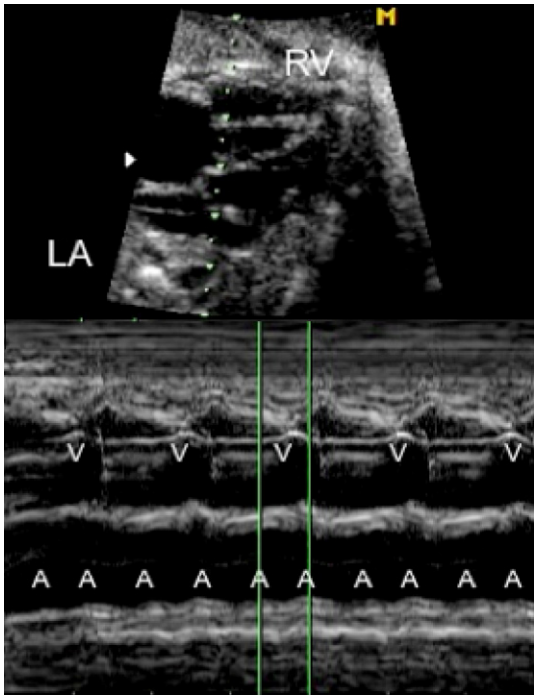

Figure 5 Atrial flutter. Simultaneous M-Mode tracing to record the RV and LA freewall movements during the cardiac cycles. The atrial rate is very fast and regular at 462 bpm while the ventricular rate is 231 bpm suggesting 2:1 AV conduction.

hemodynamically well-tolerated but on the more severe side of the spectrum may lead to heart failure, neurological complications and death.

Brief *sinus tachycardia* is a common observation and usually related to fetal activity. A variety of pregnancy conditions causes *persistent ST* such as anemia, fetal hypoxia, infections, and thyrotoxicosis (Jaeggi et al., 2006). Echocardiographically, ST resembles the normal cardiac rhythm with the exception that the atrial and ventricular rates are increased above 180 bpm. The importance of ST is recognizing and treating the underlying cause.

Atrial flutter (Fig. 5) is sustained by a fast rotating macro-reentrant circuit that is confined to the atrium. The AV node is not part of the reentrant circuit. The atrial rate typically ranges between 300 and 500 bpm which is sufficiently fast that only every second (or third) atrial beat is conducted across the AV node, resulting in ventricular response rates between 150 and 250 bpm. AF is typically observed in the last trimester as the propagation of a sustained reentrant circuit depends on a critical atrial size (Jaeggi et al., 1998b). Management of AF aims to reverse the arrhythmia to sinus rhythm or otherwise to slow AV conduction rates thus reducing the ventricular response rate to a near normal rate. This allows better ventricular filling and improved cardiac output. Maternal digoxin, sotalol, flecainide, and amiodarone have all been used to restore sinus rhythm (Jaeggi and Tulzer, 2009). After 37 weeks of gestation, we prefer delivery by caesarian section followed by postnatal conversion to a normal rhythm e.g. by esophageal overdrive pacing. Once sinus rhythm has been established after birth, recurrence of AF is uncommon and no antiarrhythmic long-term prophylaxis is usually required.

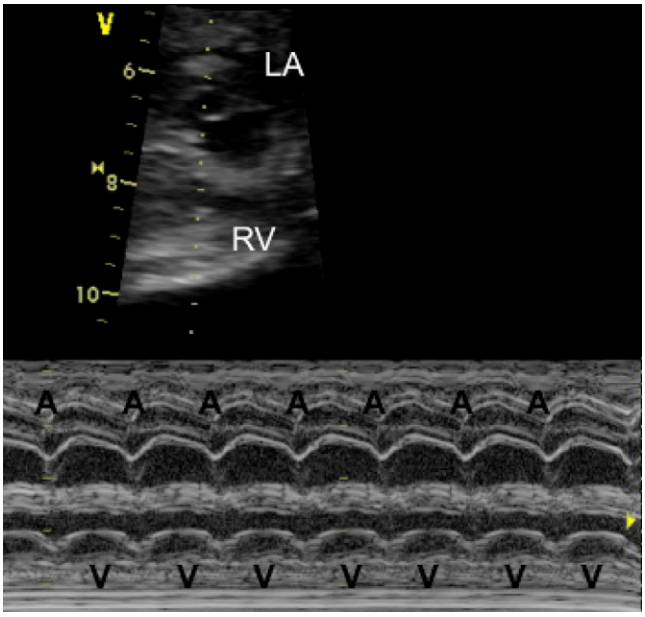

Figure 6 SVT with a heart rate of 240 bpm. M-Mode tracing through the right atrium (bottom) and the right ventricle (top). There is 1:1 AV-conduction with a short AV-interval.

Supraventricular tachycardia (*SVT*) (Fig. 6) is the most common cause of intermittent and persistent tachyarrhythmia with AV reentry related to a fast conducting accessory pathway being by far the most common mechanism. In reentrant SVT, the electrical circuit typically uses the AV node for the antegrade (atrio–ventricular) electrical conduction and the accessory pathway for the retrograde (ventriculo–atrial) conduction. As the retrograde pathway conduction is fast, the atrium is excited shortly after the ventricle, which yields a *short VA time interval* on Doppler or M-mode echocardiography (so-called short VA SVT). Other features of AV-reentry include the regular tachycardia rate that may range between 180 and 300 bpm, 1:1 AV relationship and, in intermittent SVT, the sudden onset and termination of tachycardia.

The following management options are available for fetal SVT: (a) no treatment; (b) antiarrhythmic drug therapy; and (c) delivery. Abstention of treatment with close pregnancy monitoring is a valid option if the fetus presents with intermittent brief runs of tachycardia in the absence of hemodynamic impairment. More sustained tachyarrhythmia may significantly affect the fetal cardiovascular function due to impaired ventricular filling, reduced cardiac contractility and venous congestion. In this situation, rapid and permanent conversion to sinus rhythm to prevent or resolve congestive heart failure is a primary task. Experience in fetal SVT treatment exists with a number of antiarrhythmic agents, including maternally administered digoxin, flecainide, sotalol and amiodarone and, in life-threatening situations, with the direct fetal administration of adenosine, digoxin and/or amiodarone e.g. into the umbilical vein (Jaeggi and Tulzer, 2009). If a fetus presents with significant SVT after 35–37 weeks of gestation, expedited delivery followed by postnatal conversion becomes a first choice option. At an earlier gestational age, the risks associated with premature delivery probably outweigh the potential hazards of fetal–maternal pharmacological treatment.

In summary, by means of echocardiography, it is possible to accurately diagnose fetal arrhythmia mechanisms, to assess the hemodynamic consequences and to conclude on the need of prenatal care. If indicated, efficient treatment is available to control SVT and AF, the most common causes for intermittent and persistent tachyarrhythmias above 200 bpm. The outcome of isolated fetal CHB may be improved by transplacental treatment with dexamethasone and β-stimulation at heart rates of 50–55 bpm.

References

Andelfinger, G., Fouron, J.C., Sonesson, S.E., et al., 2001. Reference values for time intervals between atrial and ventricular contractions of the fetal heart measured by two Doppler techniques. Am. J. Cardiol. 88, 1433–1436.

Fouron, J.C., Proulx, F., Miro, J., et al., 2000. Doppler and M-mode ultrasonography to time fetal atrial and ventricular contractions. Obstet. Gynecol. 96, 732–736.

Fouron, J.C., Fournier, A., Proulx, F., et al., 2003. Management of fetal tachyarrhythmia based on superior vena cava/aorta Doppler flow recordings. Heart 89, 1211–1216.

Fouron, J.C., 2004. Fetal arrhythmias. The Sainte–Justine hospital experience. Prenat. Diagn. 24, 1068–1080.

Ho, S.Y., Seo, J.W., Brown, N.A., et al., 1995. Morphology of the sinus node in human and mouse hearts with isomerism of the atrial appendages. Br. Heart J. 74, 437–442.

Jaeggi, E.T., Fouron, J.C., Fournier, A., et al., 1998a. Ventriculo–atrial time interval measured on M-mode echocardiography: a determining element in the diagnosis, treatment and prognosis of fetal supraventricular tachycardia. Heart 79, 287–582.

Jaeggi, E., Fouron, J.-C., Drblik, S.P., 1998b. Atrial flutter in the fetal period: diagnosis, clinical features, treatment and outcome. J. Pediatr. 132, 335–339.

Jaeggi, E.T., Fouron, J.C., Silverman, E.D., et al., 2004. Transplacental treatment improves the outcome of prenatally diagnosed complete atrioventricular block without structural heart disease. Circulation 110, 1542–1548.

Jaeggi, E.T., Roman, K.S., 2006. Maternal autoimmune disease and its impact on the fetal heart: diagnosis and management. Prog. Pediatr. Cardiol. 22, 85–93.

Jaeggi, E., Tulzer, G., 2009. Pharmacological and interventional fetal cardiovascular treatment. In: Anderson, R.H., Baker, E.J., Redington, A., Rigby, M.L., Penny, D., Wernovsky, G. (Eds.), Paediatric Cardiology, 3rd ed. Elsevier, pp. 199–218.

Jaeggi, E., Laskin, C., Hamilton, R., et al., 2010. 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. 55 (24), 2778–2784.

Nii, M., Hamilton, R.M., Fenwick, L., et al., 2006. Assessment of fetal atrioventricular time intervals by tissue Doppler and pulse Doppler echocardiography: normal values and correlation with fetal electrocardiography. Heart 92, 1831–1837.

Rein, A.J.J.T., O'Donnell, C.O., Geva, T., et al., 2002. Use of tissue velocity imaging in the diagnosis of fetal cardiac arrhythmias. Circulation 106, 1827–1833.