56 Fetal arrhythmias

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Fetal rhythm abnormalities occur in 2% of pregnancies. They are usually identified by the obstetrician or midwives after 20 weeks.

In avian embryos, a pacemaker is first established in the inflow tract at the straight heart tube stage. When the embryonic heart first begins to contract, it has no specialized conduction system. It is thought that the cardiac conduction system is derived from independent parent cells linked together to establish the integrated conduction system of the heart. Terminal differentiation of conduction tissue occurs late in development (1,2). By 6 weeks postconception, atrioventricular (AV) synchrony can be demonstrated using standard Doppler techniques. The heart rate is 150 bpm by 14 weeks; by 20 weeks it is 140 bpm with a gradual decrease to 130 bpm by term (3).

There are different methods used to assess fetal arrhythmias: scalp electrodes attached to electrocardiographic recordings, fetal magnetocardiography (FMCG), fetal electrocardiographic recordings from the maternal abdomen, and fetal echocardiography (M-mode, pulsed Doppler, tissue Doppler).

The usefulness of the invasive method, scalp electrodes attached to electrocardiographic recordings, is limited because it is possible to use only at ruptured membranes. An FMCG provides a noninvasive means of analyzing complex tachyarrhythmia in utero, with efficacy approaching that of postnatal electrocardiographic rhythm monitoring (4–6). The disadvantage of FMCG is that it is expensive.

The fetal electrocardiographic recordings from the maternal abdomen are not performed routinely because of the limited quality of the surface fetal electrocardiographic signal. With newer equipment it was found that electrical PR interval was better than mechanical PR interval at differentiating between normal and prolonged PR in the fetus, and the reproducibility of the PR interval using noninvasive fetal electrocardiography was significantly better than that of the Doppler methods (7–9).

In the fetal heart, electrical activity can be inferred from the timing of the mechanical events. With *M-mode method*, the simultaneous recording of the movement of the right atrial wall or the opening and closing of the AV valves and of the ventricular wall or the opening and closing of the aortic valves make it possible to determine the relationship of the atrial and ventricular electric activity. The fetal M-mode echocardiogram has its limitations: inaccurate measurement of the fetal AV conduction time, the loss of clear markers of atrial and ventricular contractions in hydropic fetuses with hypocontractile myocardium, and the poor picture resolution related to factors such as maternal obesity, fetal position, or polyhydramnios.

The *Doppler method* was found to be superior compared with the M-mode approach when measuring fetal AV time intervals. There are different places where it is possible to

measure mechanical PR interval. The MV-Ao Doppler method is recorded in five-chamber view with the septum in a vertical position, velocities generated by both inflow of blood through the mitral valve (MV) during diastole and by the outflow through the left ventricular outlet (Ao) during systole. The PR interval is measured from the intersection of the mitral E and A waves to the beginning of the ventricular ejection wave (Fig. 1). With simultaneous recording of superior vena cava (SVC) and ascending aorta (AA) flow velocity waveforms (Fig. 2), pulsed-Doppler sample volume is adjusted to encompass both vessels. The AV time interval is measured from the beginning of the retrograde venous A wave to the beginning of the aortic ejection wave (9–13). A PR interval of >150 msec on Doppler, FMCG, or postnatal ECG has been determined to be prolonged. PR intervals can vary in the fetus as they do in neonates or that at birth; changes in AV conduction or autonomic nervous system activity alter the PR interval. In cases with AV time prolongation, the SVC-Ao method seemed superior. Higher heart rates have been demonstrated to result in increasing fusion of the E and A waves in the mitral valve, overlapping the beginning of the A wave and leading to an artifactual shortening of the AV time interval. As the PR interval gets longer with no associated decrease in heart rate, the A wave will have an earlier start in relation to the E wave, in turn causing increasing overlapping of the E wave and an artifactual shortening of the A wave (14).

The newer technique of tissue-velocity Doppler imaging may be helpful when evaluating the fetus with borderline findings and negative maternal anti-Ro/anti-La antibodies (15). With tissue Doppler method, the best correlation was found between postnatal PR time and prenatal PR time with measurement of right ventricular Aa-IV (atrial contraction–isovolumetric contraction) interval (16,17).

The most common fetal rhythm anomaly is the *irregular rhythm*. The causes of irregular rhythm could be most often premature atrial contractions (PACs), premature ventricular contractions, or tachyarrhythmias with AV block.

Extrasystoles are most often “PACs,” which are usually identified in third-trimester fetuses, and their frequency may be highly variable. They may be conducted (Fig. 3) or blocked (Fig. 4) at the AV node. These are usually benign, resolving just before or shortly after birth. The follow-up is necessary because some (1–3%) of the affected fetuses have intermittent runs of supraventricular tachycardia (SVT). PACs identified with cardiomegaly, evidence of ventricular systolic dysfunction, AV valve regurgitation, and/or hydrops could be signs of a temporary SVT. Control of the foetal heart rate on each third day, weekly or biweekly obstetric ultrasound assessment is warranted until they have resolved or until delivery. About 2.5% of affected fetuses have associated structural heart defect—lesions associated with significant mitral or tricuspid

Figure 1 MV-Ao Doppler method to measure mechanical PR interval from the intersection of the mitral E and A waves to the beginning of the ventricular ejection wave. Abbreviations: E, E wave; A, A wave; V, aortic ejection wave; RV, right ventricle; LV, left ventricle; LA, left atrium; RA, right atrium; MV, mitral valve; mPR: 0.11."

Figure 2 AA/SVC Doppler method. Abbreviations: A, A wave in the superior vena cava, retrograde wave from the right atrium; V, aortic flow; SVC, blood flow in the superior vena cava. mPR: 0.11 and mPR: 0.13.

insufficiency, atrial dilatation, intracardiac tumors (Fig. 5), and aneurysm like dilation of the foramen ovale flap (Fig. 6). In such fetuses, the minor hemodynamic alterations that occur with the extrasystoles may not be tolerated.

Premature ventricular contractions are rare (Fig. 7). They are usually benign. It is necessary to rule out myocardial disease or intracardiac tumors. Prenatal and postnatal follow-up is always indicated to confirm the nature of the ectopy and exclude intermittent runs.

Tachycardia is defined as a fetal heart rate that exceeds 180 bpm. The origin of the tachycardia could be ventricular or supraventricular.

Ventricular tachycardia is rare during fetal life. With echocardiography, in the setting of fetal tachycardia, the findings of AV dissociation with a ventricular rate faster than the atrial rate suggest ventricular tachycardia (18). If there is 1:1 retrograde conduction, it is impossible to distinguish between ventricular and supraventricular tachycardias.

Figure 3 Conducted premature atrial beat, Doppler method. Abbreviations: E, E wave; A, A wave; EA, fused E and A waves because of premature beat; V, aortic ejection wave.

Figure 4 Blocked premature atrial beat, Doppler method. Abbreviations: E, E wave; A, A wave; EA, fused E and A waves because of premature beat; V, aortic ejection wave.

A rare presentation of fetal long QT syndrome is second-degree AV block and ventricular tachycardia leading to congestive heart failure. The best method to diagnose QTc prolongation and torsades de pointes is the FMCG. As a sign of torsades de pointes, the peak-to-peak intervals of AA flow during ventricular tachycardia are markedly irregular (14,18,19).

The therapy of the fetal ventricular tachycardia is difficult. In cases where it is not possible to rule out the ventricular tachycardia, digoxin therapy is contraindicated; rather, amiodarone

is the drug of the choice. If the diagnosis of the ventricular tachycardia is sure, the pregnancy is before 34th week, and there is a sign of the decompensation, then therapy with lidocaine is reasonable with strong maternal control.

“SVT” is the most common cause of the fetal tachycardia. FMCG could help to clarify electrophysiologic patterns of initiation and termination of reentrant fetal SVT. Five distinct patterns of initiation and four patterns of termination were documented, with the most common patterns of initiation

Figure 5 Multiplex rhabdomyomas in the heart. Abbreviations: RH, rhabdomyoma; LA, left atrium; RA, right atrium; SP, spine; AO, descending aorta.

Figure 6 Premature atrial beat caused by aneurysm-like flap of the foramen ovale (M-mode). Each fourth beat is a premature atrial contraction. Abbreviations: A, atrial contraction; FL, aneurysm-like flap of the foramen ovale; RA, right atrium; RV, right ventricle.

involving reentrant PACs (Wolff–Parkinson–White syndrome, QRS aberrancy, and multiple reentrant pathways). FMCG actocardiography revealed an unexpectedly strong association between fetal trunk movement and the initiation and termination of SVT, suggesting that autonomic influences play a key role (6).

In everyday routine, it is the best to plan the management of the fetal tachyarrhythmia based on analysis of pulsed-Doppler recordings of fetal heart's blood flow. It is a short V-A (ventriculo-atrial) tachycardia, when V-A period is shorter than

A-V (atrioventricular) period (Fig. 8). Short V-A tachycardia with 1:1 AV conduction and with ventricular rates of 230 to 280 bpm suggests reentrant tachycardia, and digoxin is proposed as the first choice drug. Short V-A tachycardia with AV dissociation means that with high probability of junctional ectopic tachycardia, which is usually resistant to digoxin and sotalol, amiodarone could have success. The atrial ectopic tachycardia usually responds to sotalol.

Long V-A tachycardias are seen in sinus tachycardia, in permanent junctional reciprocating tachycardia, or in

Figure 7 Ventricular extrasystole, M-Mode method. Abbreviations: A, atrial contraction; V, ventricular contraction; RA, right atrium; LA, left atrium; RV, right ventricle; LV, left ventricle.

Figure 8 Long V-A tachycardia. The V-A interval is 150 msec, and the A-V interval is 90 msec. Abbreviations: EA, atrial inflow; AO, ventricular outflow; RA, right atrium; LA, left atrium; RV, right ventricle; LV, left ventricle.

atrial ectopic tachycardia. The first-choice drug is sotalol. In intra-atrial reentrant tachycardia, the first-choice drug is digoxin (10,20).

The sotalol was found to be useful in transplacental treatment of atrial flutter and SVT. The daily maternal sotalol dose is linearly related to the maternal plasma concentration. The mean fetal/maternal sotalol plasma concentration is 1.1 (range 0.67–2.87, SD 0.63), and the mean amniotic fluid/fetal blood ratio of sotalol is 3.2 (range 1.28–5.8, SD 1.4). The effectiveness of sotalol therapy could not be extrapolated from

maternal blood levels. A large difference in the sensitivity of sotalol was found. As proarrhythmia is known to be dose related, low initiation doses are preferable and dosage increases should be stepwise. Close monitoring, especially during the initiation phase, is recommended. In addition, maternal potassium levels and magnesium levels should be monitored regularly and supplemented if necessary. The maternal toxic level is 2.5mg/L at which marked QTc prolongation is noted. The F/M ratio of 1.1 shows that the fetus also stays below the “toxic” level (21).

SVT results in reduced diastolic filling time, which, when combined with the normal limitations of relaxation and reduced compliance of the fetal myocardium, leads to increasing atrial and central venous pressures in the evolution of placental edema and dysfunction, which leads to fetal hypoxemia. Treatment of the SVT is proposed for fetuses with the risk of signs of heart failure—incessant SVT and early onset of SVT (<32 weeks). Altered maternal gastric emptying and increased maternal renal clearance could necessitate the use of higher doses. It is important to control the mother, maternal heart rate, serial ECGs and telemetry, and monitoring drug concentration (22). The mortality of SVT was found to be 8% to 10% (23).

In tachycardia with a fast conducting reentrant pathway, atrial contractions occur during ventricular ejection, thus against closed AV valves. Tall reverse “A” waves are present in the venous system of fetuses with reentrant SVT. This type of arrhythmia could induce backward venous congestion and hydrops, even in the absence of myocardial failure. The duration of the process also plays a significant role in the development of hydrops fetalis (24). Hydropic patients have higher ventricular rates (median: 240 bpm, range 240–300 bpm) than non-hydropic fetuses (220 bpm, range 200–310), and the atrial rates are not significantly different. In cases with fetal SVT and hydrops fetalis, flecainide could control the fetal arrhythmia. The therapeutic range is 200 to 700 µg/L. Strong maternal ECG and flecainide concentrations need to be checked out to term to avoid potential maternal and fetal toxicity. There is no evidence that flecainide is concentrated in the fetus (25). Another possibility of drug treatment for hydropic fetuses is digoxin and verapamil (26). In the absence of effective therapy, the mortality of the hydropic fetuses with supraventricular tachycardia approaches 50%, and with treatment, the mortality is 10%. It is recommended the maternal hyperglycemic state in the acute resuscitation of the hydropic fetus with SVT, which in providing metabolic support for the fetal myocardium, may ultimately be used coincident with initiation of antiarrhythmic treatment.

Fetal *atrial flutter* is usually identified late in gestation and is associated with atrial rates ranging from 300 to 550 bpm with variable AV conduction and ventricular rates. The atrial rate is confirmed through M-mode or systemic venous Doppler demonstration of the atrial wave rates.

Action potential prolongation in the absence of conduction slowing seems more effective in the termination of atrial flutter than depression of the excitability. Sotalol (class III drug) seemed to be more efficacious than other drugs in fetal atrial flutter. The proposed protocol in atrial flutter is as follows: 80mg of sotalol twice daily, increased stepwise with 80mg per 3 days to a maximum of 160mg three times daily. Sotalol has excellent transplacental passage. Monitoring of the maternal QT interval is essential in excluding maternal proarrhythmia. Amiodarone (class III drug) has many side effects and does not cross the placenta as readily as does sotalol. The treatment with sotalol as the first drug of choice is more reasonable because transplacental passage is slower for digoxin, relapse into atrial flutter occurs more often at birth, and subjective maternal complaints are prominent with sotalol (27).

Long-standing and/or fluctuating cerebral perfusion as a result of intrauterine atrial flutter is responsible for the

disturbance of the maintenance of adequate cerebral perfusion. It is exposing the brain to ischemia with increased risk of intracranial hemorrhage. It is opted to treat all fetuses with atrial flutter at time of presentation, unless immediate delivery is possible. The stress of the delivery and release of catecholamines may have an influence on retriggering the onset of atrial flutter, such seen in SVT. The early morbidity of atrial flutter is significant, but the long-term prognosis is excellent (28). The risk of neurologic morbidity must be taken into consideration. There are some case reports on probable inherited etiology of fetal atrial flutter (29,30).

IF THE FETAL HEART RATE IS BELOW 120/MIN, IT IS BRADYCARDIA INDEPENDENTLY FROM THE GESTATIONAL AGE

Persistent fetal *sinus bradycardia* is a rare condition and has been reported with central nervous system abnormalities, maternal treatment with beta blockers, excessive vagal tone, hydrops, neonatal long QT syndrome, and intrauterine growth retardation or as an atypical manifestation of congenital parvovirus B19 infection (31). Sinus bradycardia has also been associated with complex heart defects in which absence or hypoplasia of the sinus node is a typical finding (32). Sinus bradycardia has been reported recently as a possible sign of maternal anti-SSA/Ro antibodies. There are findings that in fetuses or newborns who died of congenital atrioventricular block (CAVB), the sinoatrial nodes were hypoplastic and, in some of these cases, surrounded by extensive fibrosis (33).

Prenatal sinus bradycardia or recognition of second-degree AV block may lead to early detection and treatment of *long QT syndrome*. QTc prolongation was found to be more common in utero than postnatally, but it does not necessarily imply that fetal QTc prolongation is a benign condition. Several studies of newborns have suggested that QT prolongation and the concomitant increased risk of ventricular tachyarrhythmias may account for the significant mortality. QT prolongation was the highest in the fetuses with the poorest outcome and was often accompanied by T-wave alternant. Common genetic polymorphisms have been shown to alter the QT interval, and prenatal molecular genetic testing for long QT syndrome has been demonstrated. The recently reported environment–genetic interactions could result in arrhythmias or sudden infant death syndrome (4). Few cases of fetal ventricular tachycardia due to congenital long QT have been reported in the literature. Some cases presented fetal ventricular tachycardia with both bradycardia and AV block (14,18,19).

“CAVB” occurred in 2.6% of a large population of fetuses evaluated for an irregular rhythm. Congenital heart defects, like L transposition (Fig. 9), left isomerism (Fig. 10), cause the AV block at about 50% of cases. If there is no cardiac malformation, the fetal CAVB is the consequence of transplacental passage of maternal autoantibodies (Figs. 11 and 12). CAVB and the neonatal lupus are the model of passively acquired autoimmunity (33). The mean histologic lesion in the heart is fibrosis of the conducting tissue and, in some cases, the surrounding myocardium. The mechanism by which maternal anti-SSA/Ro-SSB/La antibodies initiate and perpetuate inflammation and eventuate in scarring of the AV node is not yet defined. In vitro and in vivo studies suggest that one pathologic cascade leading to scarring may be initiated via apoptosis,

Figure 9 Horizontal cross section of the chest, with univentricular heart, dextrocardia, common atrium, interruption of the inferior caval vein. Abbreviations: Ventr, ventricle; ATR, atrium; AO, aorta; VA, azygos continuation.

Figure 10 Horizontal cross section of the chest, with L transposition. The anatomical right ventricle (RV) is visible on the left side, below the left atrium (LA); the left ventricle (LV) on the right side, below the right atrium (RA). Abbreviations: SP, spine; R, right; L, left.

resulting in the translocation of SSA/Ro-SSB/La antigens and subsequent surface binding by maternal autoantibodies. In CAVB, the maternal anti-SSA/Ro-SSB/La antibodies result in opsonization and inflammatory/fibrotic sequel. These opsonized cardiocytes are phagocytosed by macrophages, which secrete factors those transdifferentiate fibroblasts into myofibroblasts, a scarring phenotype (34).

The development of CAVB is a two-stage process. In the first step, the maternal autoantibodies are transferred through

the placenta and bind structures in the developing fetal heart, leading to first-degree AV block. This state is spontaneously reversible with further maturation of the fetus. If the second phase is entered, a progression to complete heart block commences. Factors involved in this second phase may be mainly fetal and related to the immunogenetic profile of the individual. A predisposition to an intense inflammatory reaction to the antibody deposition might lead to the reaction to the subsequent mononuclear cell infiltration and fibrosis,

Figure 11 Complete atrioventricular dissociation. Abbreviations: EA, ventricular inflow wave; AO, ventricular ejection wave; RA, right atrium; LA, left atrium; RV, right ventricle; LV, left ventricle; SP, spine.

Figure 12 Cardiomegaly and focal myocardial fibrosis in a fetus with congenital atrioventricular block because of maternal autoantibodies. Abbreviations: RA, right atrium; LA, left atrium; RV, right ventricle; LV, left ventricle; < signs, fibrotic foci.

with a permanent injury as the outcome. The high prevalence of fetal first-degree AV block with normal postnatal PR intervals in a group of fetuses with maternal anti-Ro/La antibodies has been proposed due to a transient biologic response to the transplacental transfer of Ro/La antibodies (3).

If second-degree AV block is present, transplacental steroid treatment theoretically may limit progression. Prophylactic steroid therapy in the fetus without documented AV conduction disease is not warranted due to maternal and fetal side effects. After a high-dose steroid therapy for fetal heart

block, several maternal complications could be found including diabetes, hypertension, infections, bone density loss, and cataract. Fetal risks are oligohydramnios and growth restriction, including the head and possibly central nervous system injury without clear evidence of benefit to fetal cardiac conduction (7).

Early detection of incomplete AV block successfully identifies a group at highest risk developing permanent AV block. Treating this group may incur greater benefit than risk. The anti-inflammatory effects of dexamethasone might have

interrupted ongoing damage to the conduction system secondary to maternal autoantibodies.

The findings of a PR interval of >150 msec at any gestational age should prompt a search for maternal anti-Ro/anti-La antibodies and close follow-up for other complex arrhythmias (11,12,15). The Doppler mechanical PR interval does not distinguish between electrical PR interval prolongation due to AV conduction delay and abnormalities of interatrial conduction. These limitations in Doppler mechanical PR interval measurements can be overcome if FMCG is available. The newer technique of tissue-velocity Doppler imaging may be helpful when evaluating the fetus with borderline findings and negative maternal anti-Ro/anti-La antibodies. May be that PR prolongation represents a variant of a normal, and only in retrospect does it have clinical significance if it is either sustained after birth or progresses to more advanced block. The prolongation of the fetal Doppler mechanical PR interval is not a definitive tool to detect early signs of anti-SSA/Ro-associated CAVB (34,35).

The risk of the third-degree heart block in pregnancies of mothers with a previous child with congenital heart block is 19% and in pregnancies in mothers without previous child with congenital heart block or rash is 4%, suggesting that there must be additional maternal and/or fetal factors that influence disease development and manifestation. The maternal factors may be related to the antibody profile. The fetal factor could be viral infection or genetic predisposition (36).

Advanced block and cardiomyopathy can occur within 1 week of normal echocardiogram without initial first-degree block. This more extensive injury can occur in utero or postnatally, even as late as 9 years of age (35).

In a large study on infants whose mothers had anti-Ro antibodies, the prevalence of hydrocephalus was 8%, which is significantly higher than the prevalence in the general population of 0.048% to 0.081%. It seems to be possible that hydrocephalus and macrocephaly are manifestations of neonatal lupus syndrome and infants born to mothers with anti-Ro antibodies should be carefully monitored for hydrocephalus as part of their routine physical examination (37).

In patients with fetal hydrops, cholestasis and severe and irreversible liver failure could be observed. Histologically, extensive collapse of the stroma and the absence of hepatocytes (foie vide) were observed. Cholestasis has been described in patients with neonatal lupus erythematosus without AV block (25).

When CAVB is associated with *endocardial fibroelastosis* (EFE), there is a very high mortality rate. The etiology of EFE remains unclear, but it may be a result of an autoimmune process. In fetuses or infants born from mothers positive for anti-Ro/La antibodies, EFE and cardiomyopathy with clinically normal conduction systems could conceive. The EFE and CAVB are two separate disease manifestations of neonatal lupus. In the histologic examinations, an active fetal B-cell response was found in addition to the passive deposition of transplacentally passed maternal immunoglobulin. The initial myocardial damage in utero is likely secondary to the passive deposition of maternal anti-Ro and anti-La antibodies, and the progression of EFE may be the result of an ongoing fetal and postnatal autoimmune reaction. The EFE accounted for over 80% of the deaths in these patients. Some cases may go

undiagnosed where there is a fetal demise. Fetal echocardiographic assessment is very subjective and often identifies only the most severe degree of EFE. Idiopathic dilated cardiomyopathy represents unrecognized maternal autoantibody-induced EFE.

There are studies on infants with CAVB who developed late-onset *dilated congestive cardiomyopathy* despite early and adequate institution of cardiac pacing. At the time of clinical presentation, many infants have symptoms or signs of an upper respiratory tract infection, otitis media, or gastrointestinal infection. Myocardial biopsy or histologic examination of the explanted heart revealed the following: myocyte hypertrophy, interstitial fibrosis, and myocyte degradation in two subjects. An active inflammatory infiltrate was not detected. An upper respiratory/gastrointestinal infection might have reactivated a previously dormant inflammatory process. The increased rate that followed pacemaker implantation may have provoked a form of tachycardia-induced cardiomyopathy (38). The mortality rate is as high as 25%, and many patients require cardiac transplantation.

THERAPY OF THE CAVB

Although the prenatal use of corticosteroids, plasmapheresis, sympathomimetics, and either prenatal pacing or early delivery with postnatal pacing has been proposed, the true efficacy of *therapy of the CAVB* is not clear.

Current approach to affected pregnancies is to initiate dexamethasone therapy at diagnosis of fetal AV block (4 mg/day) and to add maternal sympathomimetics for ventricular rates of <55 beats/min. Weekly–biweekly fetal echocardiograms are used to follow the progress, and an elective delivery by caesarean section at 36 to 37 weeks is planned. If there are signs of fetal compromise, an earlier delivery with aggressive postnatal management is proposed, including early pacemaker placement. When there is oligohydramnios, maternal dexamethasone is tapered off; however, persistence of reduced amniotic fluid may prompt earlier delivery (39). (Prednisone and prednisolone are inactivated by placenta hydroxylases and only less than 10% of the mother blood level can reach fetus; dexamethasone and betamethasone cannot be inactivated.) Some data suggest that prolonged fetal exposure to dexamethasone may impair cerebral development, but there are reports on children treated with dexamethasone prenatally (cumulative dosage 20–260 mg), who were tested postnatally with different scales according to their age and they were found with normal intelligence quotient (40).

Intravenous immunoglobulin (IVIG) is efficient in various immune-mediated conditions. It seems conceivable to employ IVIG in cases of CAVB or neonatal lupus, either administered to the mother or directly through cordocentesis in the treatment of the fetus and neonate with reduced myocardial function suggesting more diffuse disease (41).

PROGNOSIS AND FOLLOW-UP

In an isolated AV block, diagnosed in utero, in neonatal age and in children, anti-Ro and/or anti-La are present in 95% of fetal, 90% of neonatal, but only 5% of children's mothers tested. Patients with CAVB having fetal, neonatal, and childhood diagnosis have a mortality of 43%, 6%, and 0%, respectively, in the first two decades of life. Increased mortality

is associated with a fetal diagnosis of CAVB, fetal hydrops (100%), EFE (100%), and delivery at or before 32 weeks of gestation. Timing of pacemaker implantation differ significantly among fetuses versus neonates and neonates versus children cases. At 20 years of age, only 11% and 12% of CAVB patients with neonatal or children diagnosis, respectively, are not paced. CAVB detected in utero represents a more severe spectrum of the disease than encountered postnatally (39).

The mortality decreased from 45% to 15% with close surveillance, use of maternally administered corticosteroids, delivery at 35 to 37 weeks or earlier with any evidence of fetal distress or cardiovascular compromise, neonatal cardiac intensive care management, and early permanent pacemaker therapy. Among fetuses with AV block only, the survival rate is 82%, with a heart defect; the prognosis is poor, and 50% of the patients survive. Sinus bradycardia is associated with survival rates of 75% (33,42).

All fetal arrhythmias are associated with a moderately high risk for fetal distress. In cases of compromise, fetal and neonatal prognosis is poor and is an indication for perinatal medication. The risk of neurologic morbidity must be taken into consideration.

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