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Brief Report

Ventricular tachycardia secondary to prolongation of the QT interval in a fetus with autoimmune mediated congenital complete heart block

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Abstract We report a case where fetal echocardiography identified both complete heart block and ventricular tachycardia. The mother tested positive for anti-Ro antibodies. Prenatal detection of this unusual combination of arrhythmias prompted early postnatal evaluation, which revealed prolongation of the QT interval. Autoimmune mediated congenitally complete heart block associated with such prolongation of the QT interval has a poor prognosis. The child was successfully treated with beta blockers and implantation of a pacemaker.

Keywords: Long QT syndrome; fetal heart; anti-Ro antibody; echocardiography; prenatal diagnosis; arrhythmia

PRENATAL ECHOCARDIOGRAPHY CAN SUCCESSFULLY identify fetal complete heart block, using M-mode to demonstrate atrioventricular dissociation with an atrial rate faster than the ventricular rate. Fetal echocardiography also allows the mechanism of tachyarrhythmias to be evaluated. Ventricular tachycardia can be distinguished from supraventicular tachycardia, using M-mode to demonstrate atrioventricular dissociation with a ventricular rate faster than the atrial rate. We report a case where prenatal echocardiography identified both complete heart block and ventricular tachycardia.

Case report

A 24-year-old lady was referred for fetal echocardiography at 20 weeks gestation, following an admission to hospital for a viral infection, during which an irregular fetal heart rate and pericardial effusion had been detected. M-mode echocardiography demonstrated complete heart block (Fig. 1). Although the ventricular rate was mostly 60 beats per minute, runs of faster rate were observed. The cardiac structure was normal, there was mild to moderate tricuspid regurgitation, and both ventricles were dilated with moderate function. There was a small pericardial effusion, but no other sign of hydrops. The mother was found to be positive for anti-Ro antibodies. Repeat echocardiography one week later demonstrated complete heart block, some periods of 1 to 1 atrioventricular conduction, and brief salvos of ventricular tachycardia (Fig. 2). Ventricular function had normalised, with left ventricular fractional shortening of 40 per cent. In view of the short-lived nature of the periods of ventricular tachycardia, no

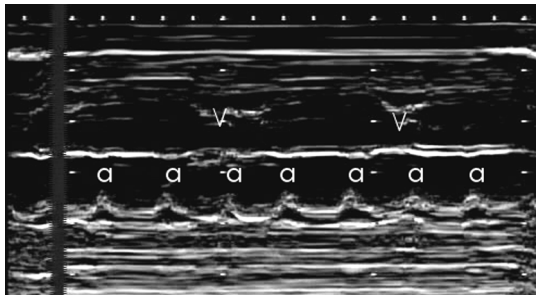

Figure 1.
M-mode fetal echocardiogram demonstrating complete heart block: there is atrioventricular dissociation with an atrial rate of 150 beats per minute (a) and a ventricular rate of 60 beats per minute (V).

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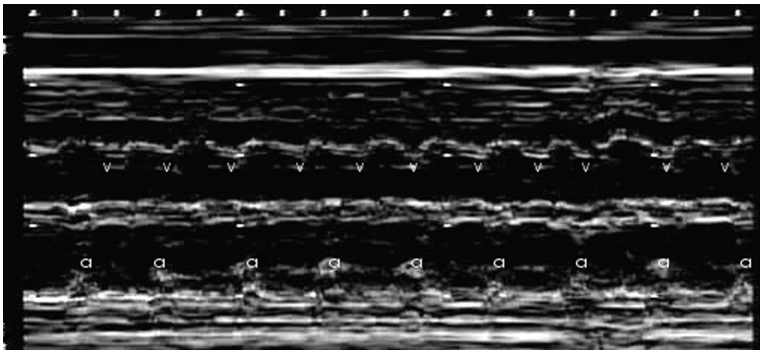

Figure 2.
M-mode fetal echocardiogram demonstrating ventricular tachycardia: there is atrioventricular dissociation with a ventricular rate of 220 beats per minute (v) and an atrial rate of 150 beats per minute (a).

antiarrhythmic drugs were administered. By 24 weeks gestation, complete heart block was consistently observed, with no episodes of ventricular tachycardia. By 37 weeks gestation, the atrial rate was 110 beats per minute, the ventricular rate was 45 beats per minute, and ventricular function remained good, with no atrioventricular valvar regurgitation. The fetus was delivered by elective Caesarean section at 39 weeks gestation, owing to the low ventricular rate and large fetal size with a previous history of failure to progress during labour.

A female infant was born in good condition with a birth weight of 3.4 kilograms. Her electrocardiogram confirmed complete heart block with a ventricular rate of 45 beats per minute, and an atrial rate of 100 beats per minute. There was a prolonged corrected QT interval of 530 milliseconds, with prominent U waves. Beyond the neonatal period, the QT interval has remained prolonged. The electrocardiograms of the parents demonstrated normal QT intervals, and there was no family history suggestive of congenital long QT syndrome.

An endocardial VVI pacemaker was inserted on the second day of life, but this became infected and was replaced with an epicardial pacing system in the neonatal period. Propranolol at doses of 1 milligram per kilogram twice daily was given because of the prolonged QT interval. A tiny patent arterial duct was identified on follow-up, which was occluded with a coil at the age of 4 years. A dual chamber pacing system was inserted at the age of 4.5 years, and propranolol has been maintained at a dose of 1milligram per kilogram three times daily because the corrected QT interval remains prolonged at 460 milliseconds. Ventricular function remains good, and the child remains asymptomatic. No episodes of ventricular tachycardia have been recorded on regular 24 hour tape recordings of the cardiac rhythm.

Discussion

Fetal ventricular tachycardia is very rare. It may therefore be misdiagnosed as supraventricular tachycardia if the M-mode is not critically evaluated.[1] Further diagnostic difficulties arise when atrioventricular dissociation cannot be clearly demonstrated, either because the atrial and ventricular rates are similar in a fetus with cardiac failure and reactive sinus tachycardia, or because there is 1 to 1 retrograde conduction across the atrioventricular node.

There have been reports of ventricular tachycardia and heart block occurring in the same fetus, but each case has subsequently been shown to have congenital long QT syndrome.[2] In such cases, complete heart block is not present. So-called "Pseudo 2 to 1 heart block" is observed, resulting from extreme prolongation of the ventricular refractory period.[3] This was clearly a diagnosis that needed to be excluded in our case. Features that were different in our case were the presence of true atrioventricular dissociation on the M-mode recordings, the absence of a family history of long QT syndrome, and the presence of maternal anti-Ro antibodies.

Fetal ventricular tachycardia occurred as a result of abnormal ventricular repolarisation, manifested postnatally as prolongation of the QT interval. The association of congenital complete heart block with such prolongation is well recognised, and defines a subgroup of patients with a particularly bad prognosis.[4] The reported incidence of prolongation, defined as corrected QT interval greater than 440 to 450 milliseconds, in patients with isolated congenital complete heart block is between 10% and 19%.[4,5] The corrected QT interval prior to pacing in our case was 530 milliseconds, which is very prolonged even for this subset of infants.[4] Bradycardia alone increases the tendency to develop ventricular arrhythmias owing to inhomogeneous ventricular refractoriness. It has been demonstrated that polymorphic ventricular tachycardia can be easily induced when bradycardia caused by congenital complete heart block is coupled with QT prolongation.[6] The increased mortality with this combination of findings is therefore almost certainly due to polymorphic ventricular tachycardia degenerating to ventricular fibrillation.[7] Treatment with beta-receptor blockade and implantation of a pacemaker, with pacing at relatively high rates, is aimed at shortening the QT interval and

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reducing QT dispersion. Although data is very limited, this treatment seems to be effective in preventing ventricular arrhythmias.[6,7] It is reassuring that our child has remained asymptomatic for more than four years.

It is unclear why a proportion of patients with isolated congenital complete heart block exhibit prolongation of the QT interval. More than nine-tenths of cases of isolated congenital complete heart block are associated with maternal anti-Ro antibodies that cross the placenta.[8] Congenital long QT syndrome results from a genetic defect in ion channels responsible for ventricular repolarisation. It is therefore tempting to speculate that anti-Ro antibodies, or other antibodies as yet unidentified that occur in mothers who are anti-Ro positive, are responsible for an autoimmune attack on both the atrioventricular node and myocardial ion channels. In support of this hypothesis, one study found that a proportion of infants with intact atrioventricular conduction born to anti-Ro positive mothers showed prolongation of the QT interval greater than 440 milliseconds.[9] A further study involving only children without congenital complete heart block showed that the corrected QT interval was significantly longer in children with anti-Ro positive mothers.[10] The effect of anti-Ro antibodies on the Ikr and Iks potassium currents involved in repolarisation, however, has yet to be tested.

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