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Resolution of hydrops fetalis caused by atrioventricular block: good postnatal evolution with terbutaline treatment

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Summary

Introduction: Complete atrioventricular block (CAVB) is rarely seen, as it occurs in only 1:11 000 to 1:20 000 newborns. There is a serious risk of mortality in CAVB, mainly in those cases associated with hydrops, fetal cardiac frequency ≤ 55 beats/minute, and premature delivery. *Case Report*: Case of complete atrioventricular block with a poor prognosis (hydrops fetalis and foetal cardiac frequency < 5 beats/minute) caused by anti-La and anti-Ro antibodies. Intrauterine symptoms improved after treatment with terbutaline, permitting foetal viability and successful postnatal treatment with a cardiac pacemaker. *Discussion:* In case of complete atrioventricular block of cause autoimmune with poor prognosis should be treated with positive inotropic drugs, anticholinergics or b-mimetic in the attempt to maintain adequate ventricular frequency, and thus prevent hydrops fetalis from occurring.

Key words: Congenital atrioventricular block; Terbutaline; Fetal arrhythmias; Fetal cardiology; Hydrops fetalis.

Introduction

Complete atrioventricular block (CAVB) is rarely seen, as it occurs in only 1:11 000 to 1:20 000 newborns [1]. It is produced by a primary alteration of the atrioventricular conduction system if it is associated with a heart disease (asplenia/polysplenia syndrome, left atrial isomerism, atrioventricular septal defect, and discordant atrioventricular connection) [1-3]. It can also be caused by an alteration in the conduction system produced by an inflammatory reaction in cases of expectant mothers with anti-Ro (SSA) and anti-La (SSB) antibodies [1, 3, 4]. CAVB cases have been described in the absence of any of these pathologies, and this suggests that they could be due to long QT interval syndrome or a viral infection [5, 6].

There is a serious risk of mortality in CAVB, mainly in those cases associated with hydrops, fetal cardiac frequency ≤ 55 beats/minute (bpm), and premature delivery [3, 7].

The natural evolution of prenatally diagnosed CAVB is poor [3, 7], but insertion of a pacemaker in a newborn has been shown to have good results if the fetus is removed during the viable stage and has an adequate weight. As a result, transplacental treatment with anti-inflammatory drugs has been attempted but has shown little results [8]. Currently, immunosuppressive treatment with corticoids is being attempted [9] when the block is still partial, or they may be used in conjunction with positive inotropic drugs, anticholinergics, or b-mimetics (ritodrine, salbutamol, terbutaline) in the aim to maintain adequate ventricular frequency and thus prevent hydrops fetalis from occurring [9,10].

The authors discussed a CAVB case produced by anti-Ro (SSA) and anti-La (SSB) with hydrops fetalis. After terbutaline treatment, the hydrops went into remission and the patient progressed well with a postnatal pacemaker.

Case Report

Expectant mother 28 years of age whose husband has various close relatives with long QT syndrome. Her first pregnancy resulted in a boy born at full term who progressed well. In her second pregnancy, the 30-week old fetus was diagnosed with hydrops fetalis caused by fetal heart block; it was born at 33 weeks and died during the first 24 hours. In the hydrops fetalis study protocol, maternal Sjögren's syndrome was diagnosed with SSA/Ro52 and SSA/Ro50 antibodies and positive SSB/LA and anti-cardiolipin antibodies in a low titre. In the third pregnancy, a gestational exam was carried out with a combined low-risk test (1:526, nasal bone present and normal ductus venosus). The fetal morphology scan was within normal limits with a normal fetal echocardiogram and a fetal heart rate of 130 bmp. At 26 weeks, a partial atrioventricular block was observed with fetal heart rate at 75 bpm. The authors began treatment with dexamethasone (eight mg/day), and at 28 weeks they observed complete AV block with fetal heart rate at 56 bpm and hydrops fetalis (fetal ascites, pericardial effusion, and fetal edema) (Figure 1). In light of these findings, they decided to treat with terbutaline (five mg every eight hours). By week 30, the hydrops fetalis was in remission and they observed a fetal heart rate of 68-70 bpm. The pregnancy was monitored until week 34; it progressed well, with fetal heart rate between 65 and 70 bpm. At week 35, a female infant weighing 2,300 grams with an Apgar score of 8-10 and a heart rate of 60 bpm was delivered by cesarean section.

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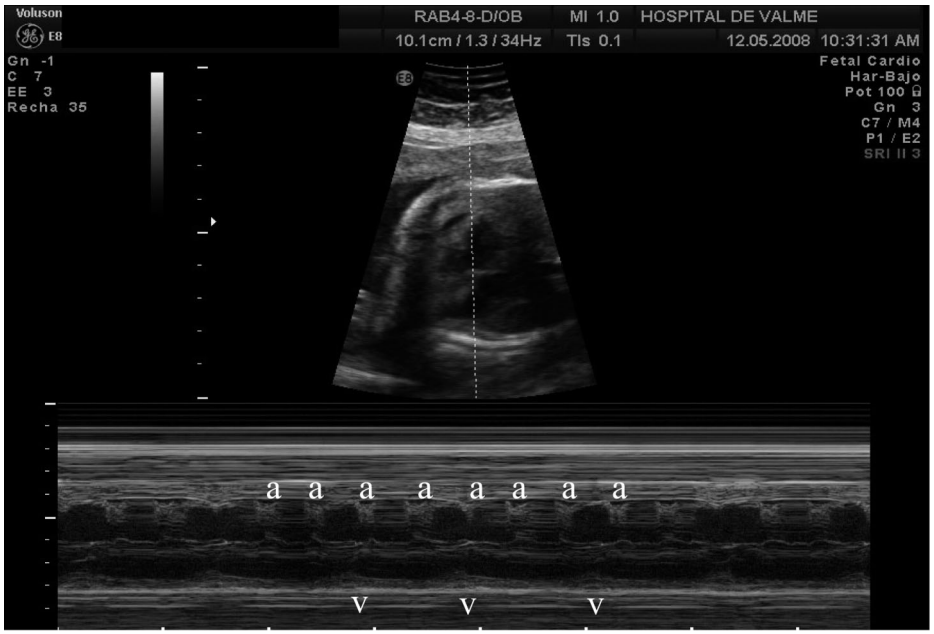

Figure 1. — Complete atrioventricular block (A: atrial contraction; V: ventricular contraction).

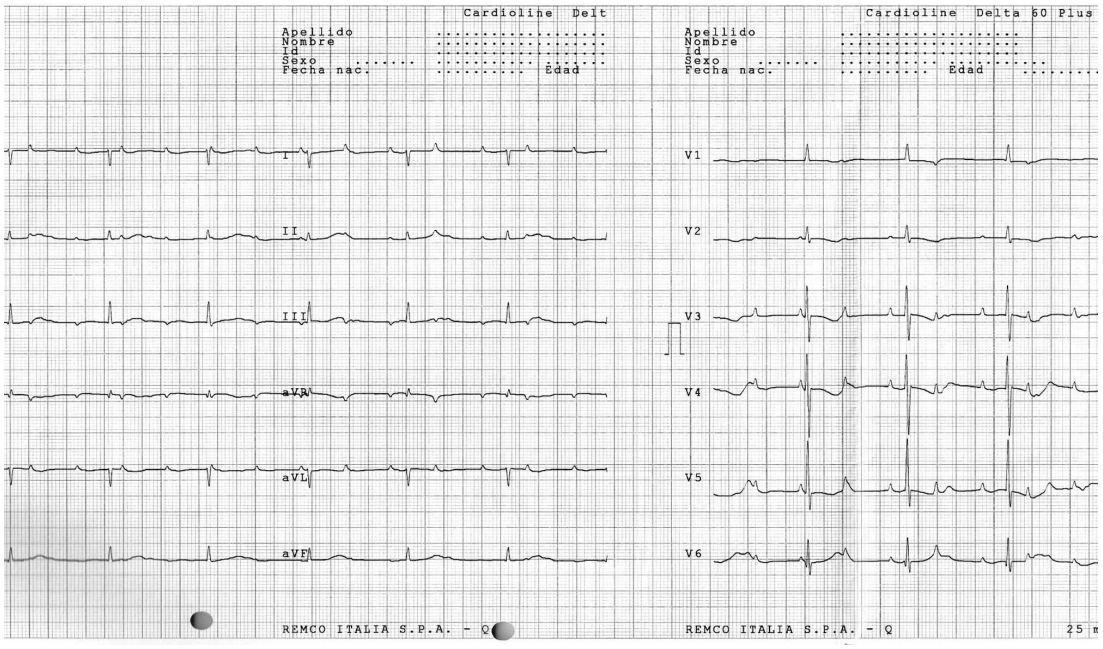

Figure 2. — Electrocardiogram of the newborn with complete atrioventricular block.

At time of admission, the newborn girl was in good general condition and presented grade II-IV systolic murmur in the subclavian fossa with no signs of heart failure except for moderate cardiomegaly. An ultrasound ruled out pericardial effusion and structural heart disease; the permeable ductus and oval foramen were appropriate for the newborn's age. The electrocardiogram showed signs of complete atrioventricular block with an atrial frequency of 130 bpm and ventricular frequency (HR) of 60 bpm (Figure 2). At 24 hours, when the HR had dropped to 55 bpm, the authors placed a temporary intravenous pacemaker through the axillary vein (following an attempt to catheterise the internal jugular vein which failed due to the patient's small size); this stabilised the heart rate at 120 bpm. Lastly, when the patient was nine days old, the authors implanted a permanent epicardial pacemaker (Figure 3), and the patient had progressed well by one year.

Discussion

In the case of maternal systemic lupus erythematosus with Anti-Ro or anti-La, the probability of presenting with an AVB is between 2-5% [4], but in cases with a previous episode of fetal AVB, the probability increases to 15-20%.

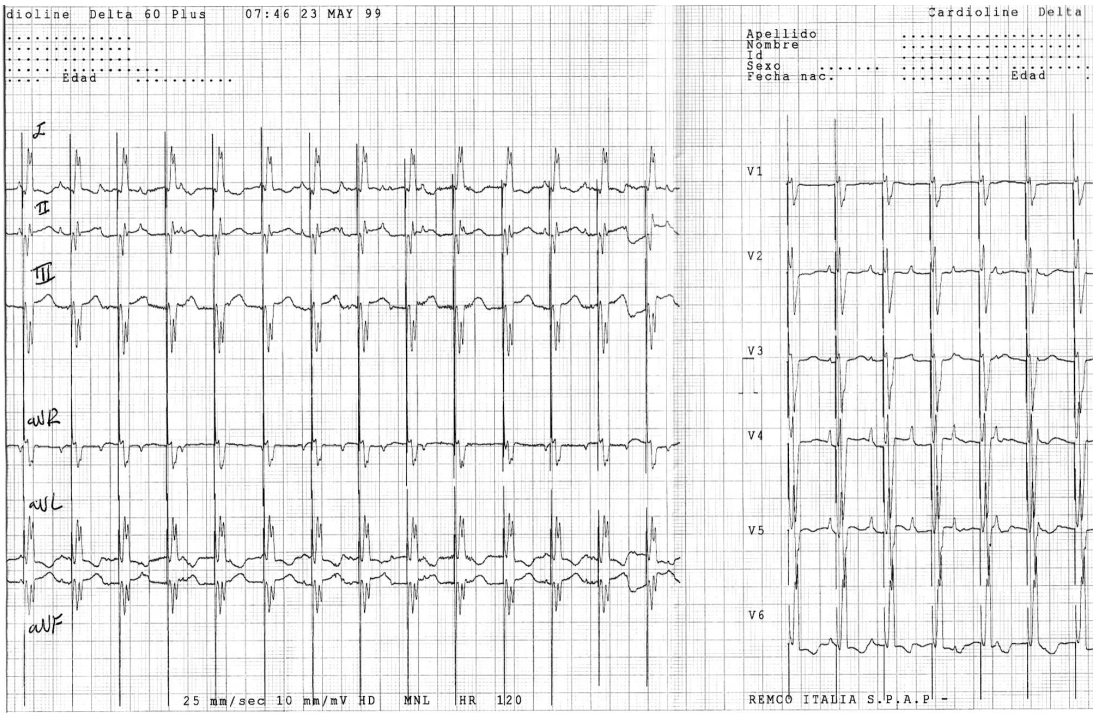

Figure 3. — Electrocardiogram of the newborn after the placement of the pacemaker.

There is no prognostic factor to predict whether or not pregnancy with anti-Ro or anti-La antibodies will develop into AVB [10]. Therefore, the fetus should be closely monitored, and if partial AVB does appear, corticoid treatment could be established as early as possible in order to reduce cardiac damage [9].

In the present case, this close monitoring was carried out, and dexamethasone treatment was established in an attempt to prevent it from developing into hydrops as it had in the previous pregnancy. When the treatment was no longer effective and the fetus became hydropic due to CAVB with a heart rate below 55 bpm, terbutaline treatment was administered to increase fetal heart rate and achieve remission of hydrops symptoms. This objective was accomplished, and pregnancy continued up to 34 weeks. At this time a cesarean section was performed, and permanent treatment in the form of a cardiac pacemaker was provided for the newborn.

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