
RESOLUTION OF HYDROPS FOETALIS CAUSED BY ATRIOVENTRICULAR BLOCK: GOOD POSTNATAL EVOLUTION WITH TERBUTALINE TREATMENT.

Type:

Recommendation, comment, report

Abstract:

Case of complete atrioventricular block with a poor prognosis (hydrops foetalis and foetal cardiac frequency < 55 beats/min) 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.

Keywords:

congenital atrioventricular block, terbutaline, foetal arrhythmias, foetal cardiology, hydrops foetalis

1 **RESOLUTION OF HYDROPS FOETALIS CAUSED BY**
2 **ATRIOVENTRICULAR BLOCK: GOOD POSTNATAL EVOLUTION WITH**
3 **TERBUTALINE TREATMENT.**

4 **Introduction:**

5 Complete atrioventricular block (CAVB) is rarely seen, as it occurs in only 1:11 000 to
6 1:20 000 newborns.⁹ It is produced by a primary alteration of the atrioventricular
7 conduction system if it is associated with a heart disease (asplenia/polysplenia
8 syndrome, left atrial isomerism, atrioventricular septal defect, discordant
9 atrioventricular connection).^{2,9,10} It can also be caused by an alteration in the conduction
10 system produced by an inflammatory reaction in cases of expectant mothers with anti-
11 Ro (SSA) and anti-La (SSB) antibodies.^{3,9,10} CAVB cases have been described in the
12 absence of any of these pathologies, and this suggests that they could be due to long QT
13 interval syndrome or a viral infection.^{1,5}

14 There is a serious risk of mortality in CAVB, mainly in those cases associated with
15 hydrops, foetal cardiac frequency ≤ 55 beats/min (bpm), and premature delivery.^{6,10}

16 The natural evolution of prenatally diagnosed CAVB is poor,^{6,10} but insertion of a
17 pacemaker in a newborn has been shown to have good results if the foetus is removed
18 during the viable stage and has an adequate weight. As a result, transplacental treatment
19 with anti-inflammatory drugs has been tried but shown little results.⁸ Currently,
20 immunosuppressive treatment with corticoids is being tried ⁷ when the block is still
21 partial, or they may be used in conjunction with positive inotropic drugs,
22 anticholinergics, or b-mimetics (ritodrine, salbutamol, terbutaline) in the attempt to

23 maintain adequate ventricular frequency and thus prevent hydrops foetalis from
24 occurring.^{4,7}

25 We discussed a CAVB case produced by anti-Ro (SSA) and anti-La (SSB) with hydrops
26 foetalis. After terbutaline treatment, the hydrops went into remission and the patient
27 progressed well with a postnatal pacemaker.

28 **Case report:**

29 Expectant mother 28 years of age whose husband has various close relatives with long
30 QT syndrome. Her first pregnancy resulted in a boy born at full term who progressed
31 well. In her second pregnancy, the 30-week old foetus was diagnosed with hydrops
32 foetalis caused by foetal heart block; it was born at 33 weeks and died during the first
33 24 hours. In the hydrops foetalis study protocol, maternal Sjögren's syndrome was
34 diagnosed with SSA/Ro52 and SSA/Ro50 antibodies and positive SSB/LA and anti-
35 cardiolipin antibodies in a low titre.

36 In the third pregnancy, a gestational exam was carried out with a combined low-risk test
37 (1:526, nasal bone present and normal ductus venosus). The foetal morphology scan
38 was within normal limits with a normal foetal echocardiogram and a foetal heart rate of
39 130bpm.

40 At 26 weeks, a partial atrioventricular block was observed with foetal heart rate at 75
41 bpm. We began treatment with dexamethasone (8mg/day), and at 28 weeks we observed
42 complete AV block with foetal heart rate at 56 bpm and hydrops foetalis (foetal ascites,
43 pericardial effusion and foetal edema) (image 1). In light of these findings, we decided
44 to treat with terbutaline (5mg every 8 hours). By week 30, the hydrops foetalis was in
45 remission and we observed a foetal heart rate of 68-70 bpm. The pregnancy was

46 monitored until week 34; it progressed well, with foetal heart rate between 65 and
47 70bpm. At week 35, a female infant weighing 2300g with an Apgar score of 8-10 and a
48 heart rate of 60 bpm was delivered by caesarean section.

49 At time of admission, she was in good general condition and presented grade II-IV
50 systolic murmur in the subclavian fossa with no signs of heart failure except for
51 moderate cardiomegaly. An ultrasound ruled out pericardial effusion and structural
52 heart disease; the permeable ductus and oval foramen were appropriate for the
53 newborn's age. The electrocardiogram showed signs of complete atrioventricular block
54 with an atrial frequency of 130 bpm and ventricular frequency (HR) of 60 bpm (image
55 2). At 24 hours, when the HR had dropped to 55 bpm, we placed a temporary
56 intravenous pacemaker through the axillary vein (following an attempt to catheterise the
57 internal jugular vein which failed due to the patient's small size); this stabilised the
58 heart rate at 120 bpm. Lastly, when the patient was 9 days old, we implanted a
59 permanent epicardial pacemaker (image 3), and the patient had progressed well by one
60 year.

61 **Discussion:**

62 In the case of maternal systemic lupus erythematosus with Anti-Ro or anti-La, the
63 probability of presenting with an AVB is between 2-5%,³ but in cases with a previous
64 episode of foetal AVB, probability increases to 15-20%. There is no prognostic factor to
65 predict whether or not pregnancy with anti-Ro or anti-La antibodies will develop into
66 AVB.⁴ Therefore, the foetus should be closely monitored, and if partial AVB does
67 appear, corticoid treatment could be established as early as possible in order to reduce
68 cardiac damage.⁷

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

References:

1. Batmaz G, Villain E, Bonnet D, et al. Therapy and prognosis of infectious complete atrioventricular block in children. *Arch Mal Coeur Vaiss* 2000;93:553–557.
2. Berg C, Geipel A, Kohl T, Breuer J, Germer U, Krapp M, Baschat AA, Hansmann M, Gembruch U. Atrioventricular block detected in fetal life: associated anomalies and potential prognostic markers. *Ultrasound Obstet Gynecol* 2005;26:4–15.
3. Buyon JP, Hiebert R, Copel J, et al. Autoimmune-associated congenital heart block: demographics, mortality and recurrence rates obtained from national neonatal lupus registry. *J Am Coll Cardiol* 1998;31:1658–66.
4. Groves AM, Allan LD, Rosenthal E. Therapeutic trial of sympathomimetics in three cases of complete heart block in the fetus. *Circulation* 1995;92: 3394–3396.
5. Hosono T, Shinto M, Chiba Y, et al. Prenatal diagnosis of fetal complete atrioventricular block with QT prolongation and alternating ventricular pacemakers using multi-channel magnetocardiography and current-arrow maps. *Fetal Diagn Ther* 2002;17:173–176.
6. Jaeggi ET, Hamilton RM, Silverman ED, et al. Outcome of children with fetal, neonatal or childhood diagnosis of isolated congenital atrioventricular block: a single institution's experience of 30 years. *J Am Coll Cardiol* 2002;39:130–137.
7. Jaeggi ET, Fouron JC, Silverman ED, Ryan G, Smallhorn J, Hornberger LK. Transplacental fetal treatment improves the outcome of prenatally diagnosed complete atrioventricular block without structural heart disease. *Circulation* 2004;110:1542–1548.
8. Lin MT, Hsieh FJ, Shyu MK, Lee CN, Wang JK, Wu MH. Postnatal outcome of fetal bradycardia without significant cardiac abnormalities. *Am Heart J* 2004;147:540–544.

101 9.Fetal heart block. In: Allan L, Hornberger LK, Sharland G (eds) Textbook of fetal
102 cardiology. London: Greenwich Medical Media;2000.pp 438–52.

103 10. Simpson JM. Fetal arrhythmias. Ultrasound Obstet Gynecol 2006;27:599-606.

Accepted article

Figure 1

[Download source file \(65.88 kB\)](#)



Images 1. Complete atrioventricular block

Figure 2

[Download source file \(1.54 MB\)](#)

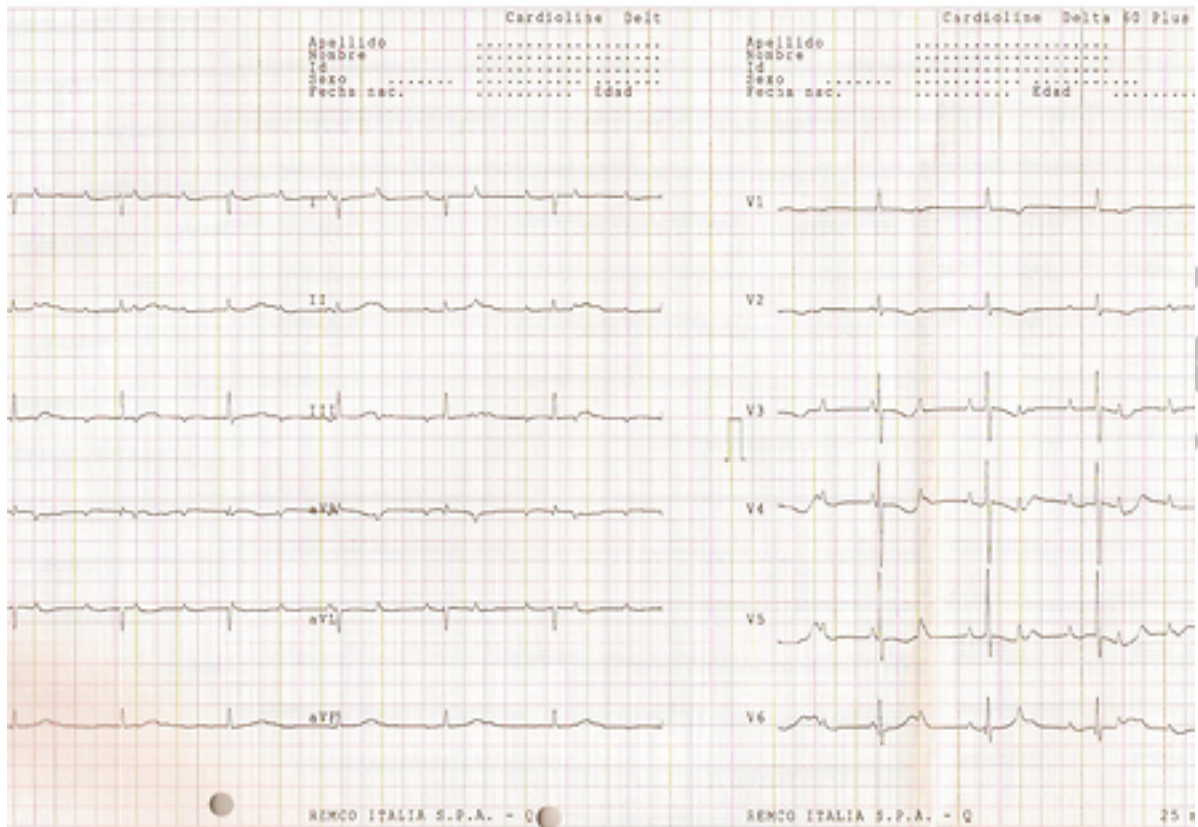


Imagen 2. Electrocardiogram of the newborn with complete atrioventricular block

Figure 3

[Download source file \(1.63 MB\)](#)

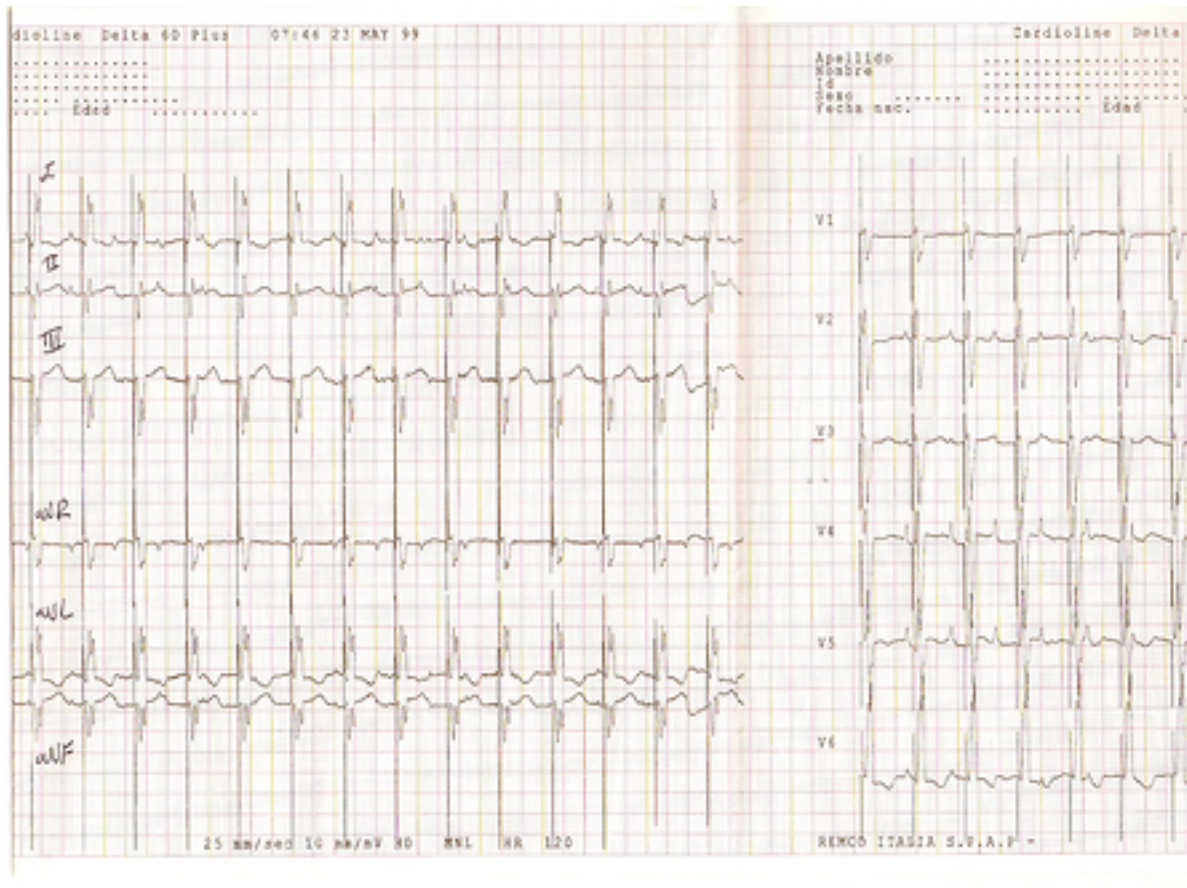


Imagen 3. Electrocardiogram of the newborn after the placement of the pacemaker

Manuscript body

Manuscript body 1 - [Download source file \(45 kB\)](#)

Figures

Figure 1 - [Download source file \(65.88 kB\)](#)

Images 1. Complete atrioventricular block

Figure 2 - [Download source file \(1.54 MB\)](#)

Imagen 2. Electrocardiogram of the newborn with complete atrioventricular block

Figure 3 - [Download source file \(1.63 MB\)](#)

Imagen 3. Electrocardiogram of the newborn after the placement of the pacemaker

Accepted article