

Case Report

Rapid progression of congenital heart block with negative maternal anti-Ro/SSA antibody :

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Abstract

Severe congenital heart block is strongly associated with maternal anti-Ro/SSA antibody. However, we experienced an infant with negative maternal anti-Ro/SSA antibody who had showed progression of severe congenital heart block.

A 2-month-old infant with 2 : 1 second-degree heart block progressed to 4 : 1 heart block within a month. At 6 months of age, he was administrated to our hospital because of frequent vomiting and loss of body weight. His ECG showed complete heart block. A VVI pacemaker was implanted under the diagnosis of severe heart failure due to congenital complete heart block. During a one-year follow-up, he has been free from cardiac events.

Frequent electrophysiologic examinations are mandatory for an infant with progressive heart block.

Key words (atrioventricular block, infant, a pacemaker implantation)

Introduction

A 2-month-old infant with second-degree heart block progressed to complete heart block, requiring pacemaker implantation 4 months after the diagnosis. Clinical features suggested congenital complete heart block (CCHB), however, maternal anti-Ro/SSA antibody, which has been strongly associated with severe CCHB, was negative.

Case

A 2-month-old infant was referred to our hospital due to bradycardia. A fetal echocardiogram showed normal heart rate. Previously, he had been active and asymptomatic with normal growth and development. His past medical history and family history were unremarkable. The heart rate was 96/min, respiratory rate was 48/min, and blood pressure was 84/41mmHg. Heart sounds were regular and no heart murmur was detected. Cardio-thoracic ratio (CTR) on chest X-ray film was 58%. Electrocardiogram (ECG) demonstrated 2 : 1 second-degree heart block

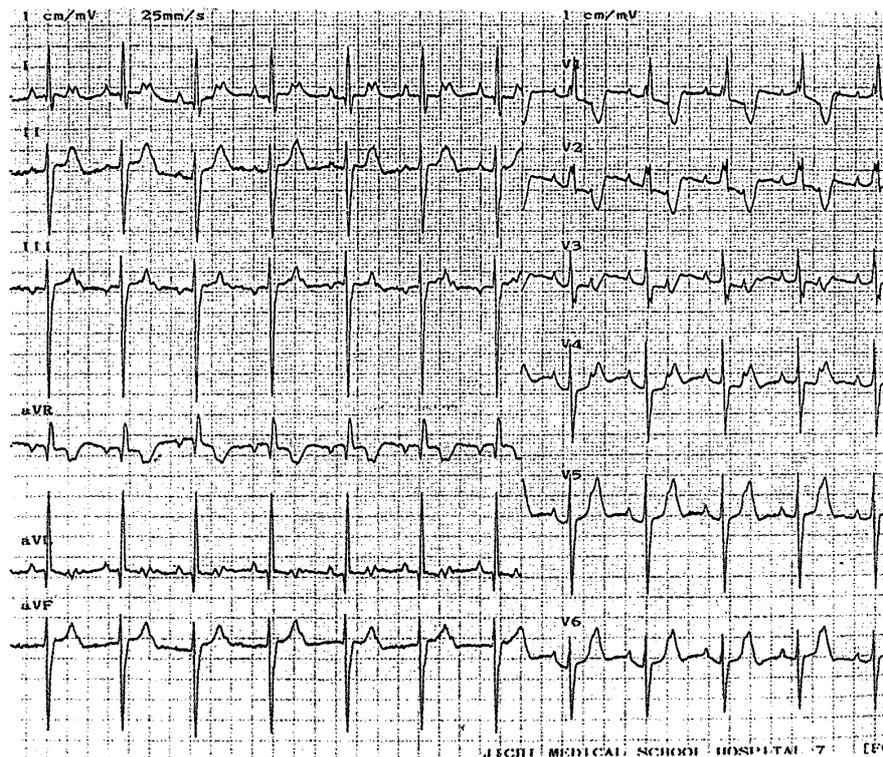


Figure 1. ECG at 2 months of age showing 2 : 1 second-degree heart block.

and incomplete right bundle branch block (Fig. 1). On 24-hour ambulatory ECG, the heart rate varied from 45 to 131/min with a 2 : 1 heart block. Two-dimensional echocardiogram showed normal cardiac anatomy without cardiac tumor. We assessed that neither medication nor pacemaker implantation was needed at that time.

At 3 months of age, his ECG showed 4 : 1 heart block (Fig. 2). At 4 months of age, 24-hour ambulatory ECG demonstrated that the heart rate varied from 29 to 51/min with a 4 : 1 heart block. Despite the decreased heart rate, he remained active.

At 5 months of age, he developed occasional vomiting after feeding, but, 24-hour ambulatory ECG did not show any interval changes since 4 months of age.

At 6 months of age, he still vomited repeatedly and lost weight, but looked well. Heart rate was 26/min, respiratory rate was 52/min, and blood pressure was 96/63mmHg. Cardiomegaly was noted on chest X-ray film (CTR was 63%). ECG demonstrated a complete heart block (Fig. 3). Two-dimensional echocardiogram showed mildly dilated ventricles with a normal ventricular systolic function. Laboratory tests demonstrated elevated urea nitrogen (45mg/dl), hyponatremia (Na 128mmol/l) and hyperpotassemia (K 5.5mmol/l). Serum CPK was within normal level and there was no evidence of infection and inflammation. The maternal and infant's serum anti-Ro/SSA antibodies at 6 months of age were negative. Serum HANP (974pg/ml, normal range <43) and BNP (1700pg/ml, normal range <18.4) were elevated. There was also no evidence of overdose of medications. Under a diagnosis of severe heart failure due to CCHB, a VVI pacemaker (Medtronic Kappa KSR701, programmed to a lower rate of 80/min) was implanted.

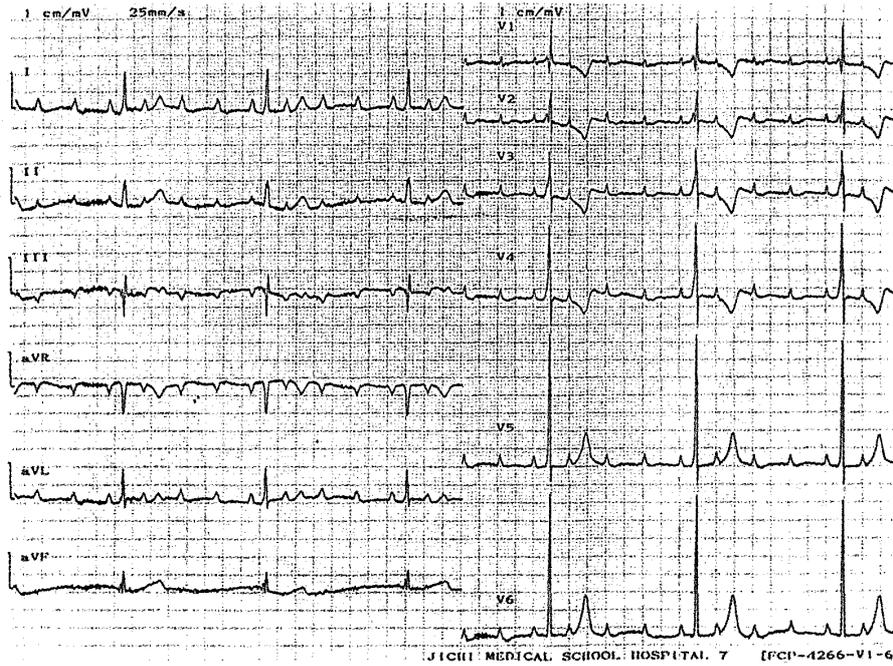


Figure 2. ECG at 3 months of age showing 4 : 1 heart block.

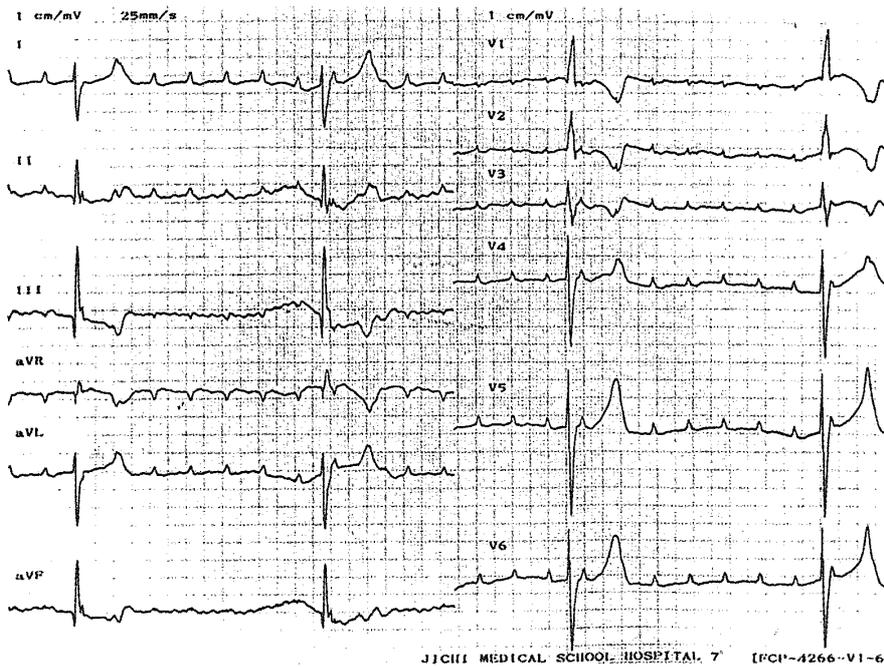


Figure 3. ECG at 6 months of age showing complete heart block with a heart rate of 21/min.

During a one-year follow-up, he has been free from frequent vomiting, regained body weight and begun growing normally.

24-hour ambulatory ECG showed complete cardiac pacing.

Discussion

Our patient was diagnosed as having severe CCHB, because of the severe outcome that required pacemaker implantation and the rapid progress of conductive disorder under His bundle.

A strong association between severe CCHB and maternal anti-Ro/SSA antibody is well established.^{1,2)} Maternal serum anti-Ro/SSA antibody is reported to be positive in 75 to 100% of cases involving severe CCHB infants.^{1,2)} However, this was rare case since maternal anti-Ro/SSA antibody was negative.

Our patient had another rare clinical feature that he was not diagnosed as CCHB in early infant period. According to the report by Hubscher³⁾, 9/12 of CCHB infants that were diagnosed in early infant period, which was before the age of 3 months, were associated with positive maternal anti-Ro/SSA antibody and the outcome was severe with all 9 CCHB infants requiring pacemaker implantation. However, 5/6 of CCHB infants diagnosed after the age of 3 months were associated with negative maternal anti-Ro/SSA antibody and none of 5 CCHB infants needed pacemaker implantation. Our patient was diagnosed as CCHB at the age of 6 month and with negative maternal anti-Ro/SSA antibody, which was likely with the latter case of Hubscher. However the prognosis of our patient was severe which was more likely the prognosis of CCHB infants with positive maternal anti-Ro/SSA antibody.

In our patient, heart block was accompanied by right bundle branch block on the first ECG recording, which suggested that conductive disorder had existed under His bundle. Slow heart rate and wide QRS in the ECG at 6 month of age also suggested that conductive disorder progressed into both left and right bundle branches, though we have not confirmed this with electrophysiologic study in the catheter laboratory.

The current hypothesis is that maternal anti-Ro/SSA antibody transmits through the placenta to a fetus and causing interactions that result in an inflammatory response with consequent calcification and apoptosis.^{4,5)} These changes are commonly seen in the area of the AV node and His bundle.⁴⁾ Some progressive conductive disorder after birth, which was seen in our patient, has been reported in CCHB^{6,7)}

Other mechanisms are considered that can cause CCHB after birth due to some injury, like the existence of autoantibody, anoxia, hypersensitivity or aging. Hackel proposed that genetic predisposition and a precipitating injury of the conducting fibers are related to the occurrence and progress of CCHB.⁸⁾ The hypothesis seems to be more acceptable because not all anti-Ro/SSA positive mothers have CCHB infants. However, we could not confirm any other factors which like autoantibody, anoxia, using of drugs or viral infections in our patient.

The conductive disorder in our patient rapidly progressed to complete heart block by the age of 6 months. Progressive CCHB was reported by Agarwala in 2000.⁹⁾ His case was diagnosed as having first-degree heart block at the age of 11 months and progressed to complete heart block at the age of 26 months, requiring pacemaker implantation. Maternal anti-Ro/SSA antibody was also negative in that case. To our knowledge, our patient showed most rapid progression to complete heart block that needed pacemaker implantation with negative maternal anti-Ro/SSA antibody.

In summary, even in infants with negative maternal anti-Ro/SSA antibody can develop

severe CCHB and the progression of CCHB can be rapid. Frequent electrophysiologic examinations are mandatory for an infant with progressive heart block.

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早期に進行した母体 Anti-Ro/SSA 抗体陰性の 先天性房室ブロックの1例

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要 約

4か月間に完全房室ブロックに至った先天性房室ブロックの児を経験した。一般に母体 anti-Ro/SSA 抗体陽性の先天性房室ブロックの児は、完全房室ブロックへ進行する率が高く、また完全房室ブロックへの進行が早いと報告されているが、本児では Anti-Ro/SSA 抗体は母児

とも陰性だった。

Anti-Ro/SSA 抗体陰性の先天性房室ブロックでも、早期に完全房室ブロックに進行する例があり、頻回の電気生理学的な評価が必要と考えられた。