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CASE REPORT

Congenital Complete Heart Block Complicated by Atrial Flutter Diagnosis and Management

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Abstract

Seventeen-month-old child was diagnosed in utero to have congenital complete heart block. The mother has Sjogren's syndrome with high Anti Ro antibodies. The baby was delivered at term with a heart rate of 55–60 beats per minute. Echocardiography revealed a structurally normal heart with a small atrial septal defect and moderate patent ductus arteriosus. At the age of 17 months, he developed atrial flutter which was aborted using electrical cardioversion in the Cath lab. No recurrence of the atrial flutter during a one-year follow-up.

Keywords: Atrial flutter, Congenital complete heart block, Cardioversion

Article summary

A description of a child with congenital complete heart block complicated by atrial flutter, managed with cardioversion.

1. Introduction

Congenital complete heart block (CCHB) is a rare complication of maternal connective tissue disorders and transplacental transfer of autoantibodies from the mother to the fetus [1]. The occurrence of atrial flutter on top of CCHB is rare and was not presented in the literature. We are presenting a child with CCHB complicated by atrial flutter managed successfully by electrical cardioversion. To our knowledge, this is the first case reporting postnatal atrial flutter in a patient with CCHB.

2. Case presentation

Seventeen months-old male infant known to have congenital complete heart block (CCHB). His Mother is positive for Anti Ro (SS-A) antibodies & possible Sjogren's syndrome. The mother (G8P3+5) was referred during pregnancy at *20 weeks' gestation* because of fetal irregular heart rhythm, and pericardial effusion. There is a history of one child with gastrointestinal obstruction (died in the neonatal period). They have two other healthy siblings. Mother had recurrent abortions with no symptoms suggestive of connective tissue disorders apart from itching and mild dryness of the eyes.

Fetal echocardiography was done and revealed a structurally normal fetal heart with congenital complete heart block. The atrial rate was 130–156 bpm and the ventricular rate was 53 bpm. There was mild to moderate pericardial effusion with mildly depressed ventricular systolic function. The UAC, UVC, and Ductus venosus Doppler were abnormal. No ascites or pleural effusion (Fig. 1).

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The baby was delivered at complete 37 weeks' gestation by CS with a birth weight of 2.6 kg. He was clinically stable with a HR of 55–60 beats per minute. ECG was done postnatally & showed a complete heart block with junctional escape rhythm with a ventricular rate of 53/min (Fig. 2).

Echocardiography revealed a PFO with a left to right shunt and moderate patent ductus arteriosus (PDA) with a bidirectional shunt. Ventricles were dilated with good biventricular systolic function. Mother investigations revealed a normal level of anti-LA (SS-B), anti-SM, anti-SCL-70, anti-JO-1, and anti-DNA antibodies. Anti Ro (SS-A) antibodies were strongly positive with a titer of 141.69 (normal <20 U/mL).

The patient was kept on regular follow-up. He was growing and developing normally. At the age of 6 months (weight 7 kg), he had PDA device closure.

Abbreviations

AF	atrial fibrillation
AFL	atrial flutter
ASD	Atrial septal defect
bpm	beat per minute
CCHB	Congenital complete heart block
CHD	Congenital heart disease
ECG	electrocardiogram
EFE	endocardial fibroelastosis
EP	electrophysiology
HCQ	hydroxychloroquine
PDA	Patent ductus arteriosus
PFO	patent foramen ovale
TEE	trans-esophageal echocardiography

Twenty-four hours Holter monitoring was done frequently and showed complete heart block, junctional escape rhythm with a ventricular rate of

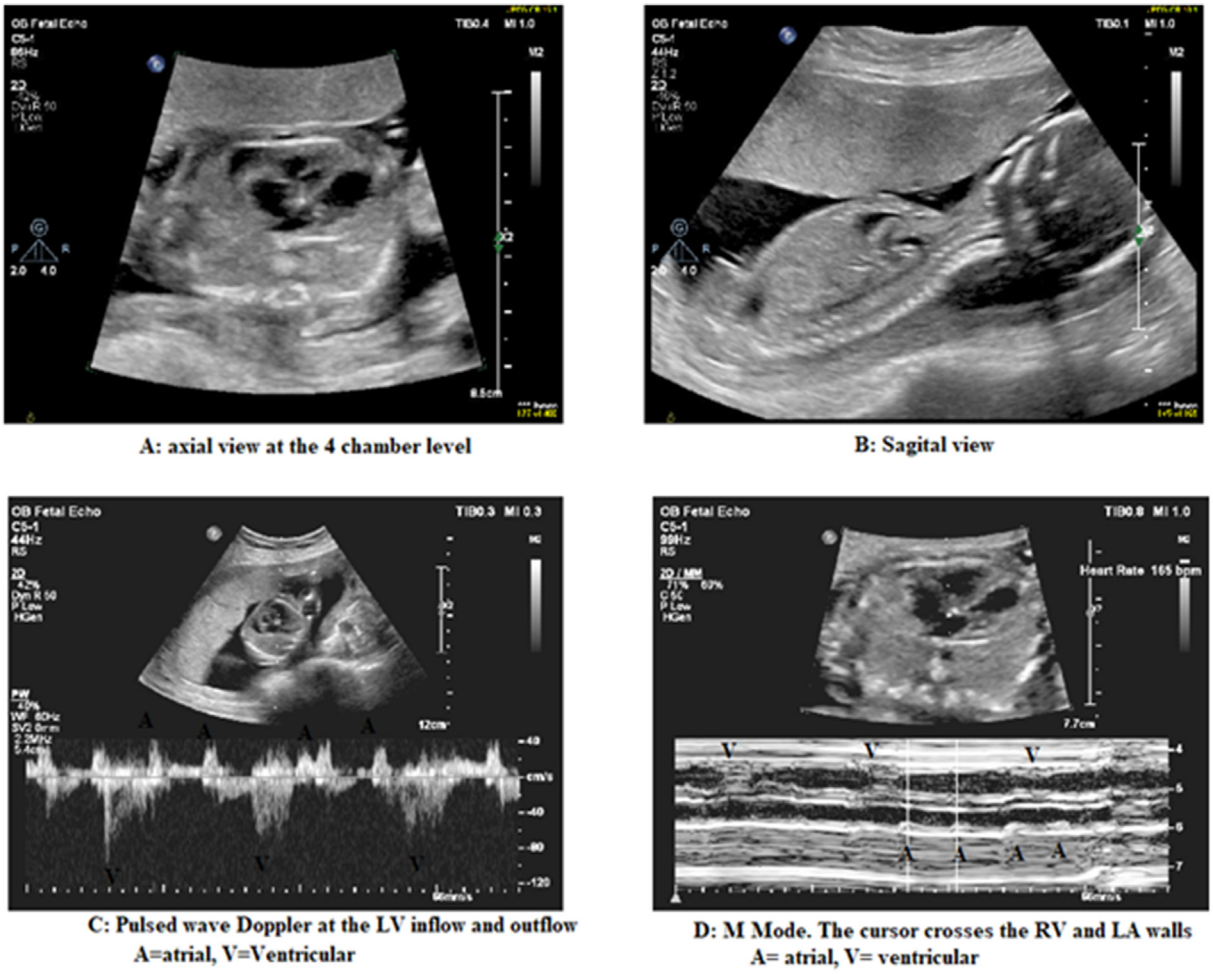

Fig. 1. Fetal echocardiography. A) axial view at the 4 chamber level showing a normal 4 chambers with mild pericardial effusion (Arrow). B) Sagittal view showing the chest and abdomen of the fetus, with mild pericardial effusion (Arrow). C) LV Inflow/outflow Pulse-Doppler showing mitral inflow atrial (A) more than ventricular (V) aortic outflow waves. C) M Mode: the cursor line cuts the right ventricular (RV) anterior wall and the left atrial (LA) wall (Atrial rate (A) is 153, and ventricular rate (V) is 55 bpm.

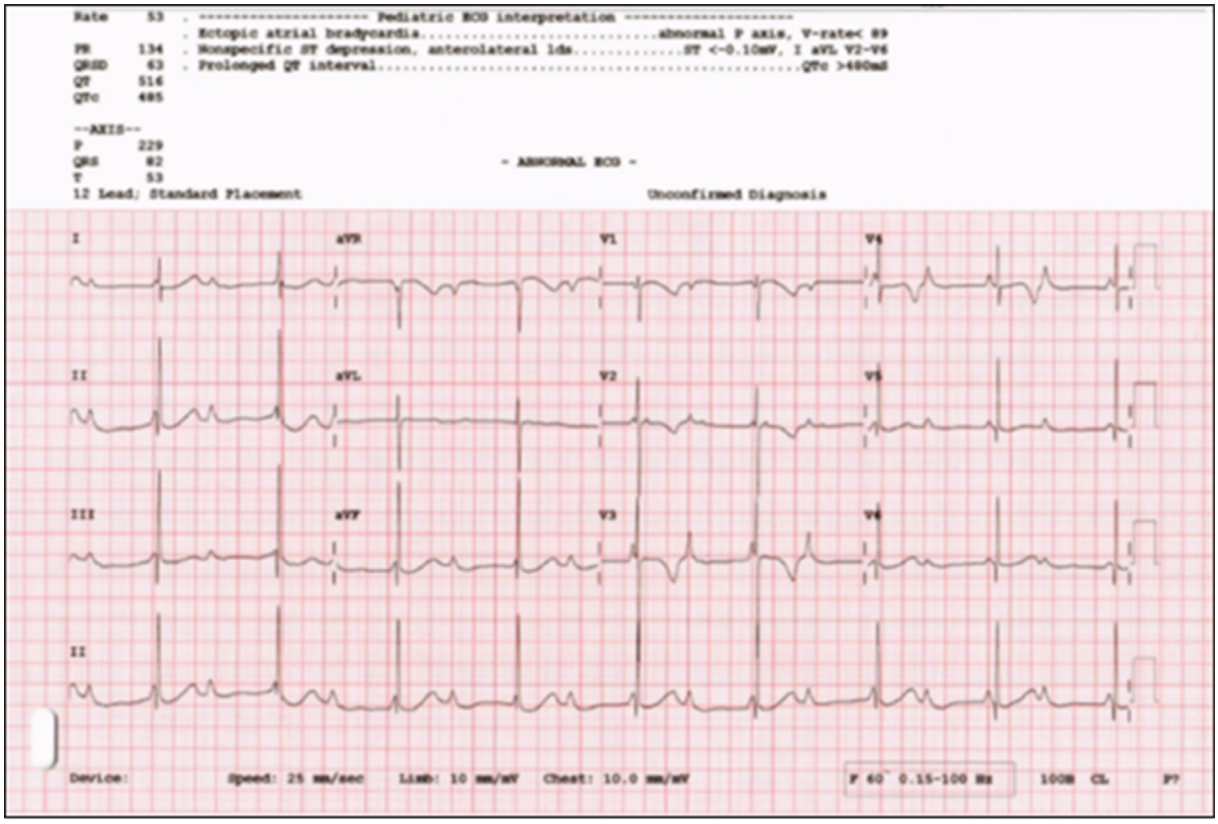

Fig. 2. 12 lead ECG showing complete heart block with atrioventricular dissociation. There is junctional escape rhythm, with narrow QRS complexes and a ventricular rate of 53 bpm.

53–57 and an average heart rate of 55 bpm. There are no long pauses and no PVCs.

At the age of 17 months, the patient was asymptomatic, but the ECG revealed atrial flutter (AF) with complete heart block and junctional escape rhythm, with a ventricular rate of 51 bpm, and an atrial rate of 250–300/minute (Fig. 3). Holter and repeated ECGs revealed the persistence of the Atrial Flutter (Fig. 4).

Clinical examination was unremarkable apart from the bradycardia and soft systolic murmur. His weight and height were 10.6 kg and 79 cm (both in the 50th percentile).

Echocardiography revealed the PDA device in position with no residual shunt. The ascending aorta is dilated (18 mm, Z score + 3.1), dilated main pulmonary artery, with normal systolic function. No pericardial effusion, no signs of pulmonary hypertension, and no evidence of atrial or mural thrombi.

The condition was discussed for possible cardioversion with or without a temporary transvenous pacemaker.

He was electively admitted and given heparin infusion for 24 h. The next day he was taken to the electrophysiology (EP) lab. There was difficulty getting a femoral venous line. He was given 4 mcg of isoprenaline and transcutaneous pacing was attached to the chest as a backup. Then synchronized cardioversion was achieved using 10 J of direct current (1 J/kg). Atrial flutter was terminated immediately and the HR was in 40s bpm with narrow complex QRS (Fig. 5 A and B). He remained hemodynamically stable. Then he was shifted to the ward and kept on heparin infusion for another 24 h. Twelve lead ECGs showed complete heart block, with a junctional escape rhythm with a ventricular rate of 55 bpm. The patient was discharged home on aspirin 50 mg daily for 2 weeks. Over the one year of follow-up, there was no recurrence of the atrial flutter, and he maintained his baseline heart rate of 55 bpm.

3. Discussion

CCHB is a rare disorder that can be diagnosed antenatally. There is an increased risk for fetal hydrops and fetal demise, especially in fetuses with structurally abnormal hearts. The most common cause of isolated CCHB block is fetal exposure to autoimmune maternal antibodies that can occur as early as 11 weeks of gestation. Fetal heart block is generally not seen before 18 weeks' gestation and the onset is rare after 28 weeks' gestation. Half of these women are asymptomatic concerning their rheumatologic disorder and

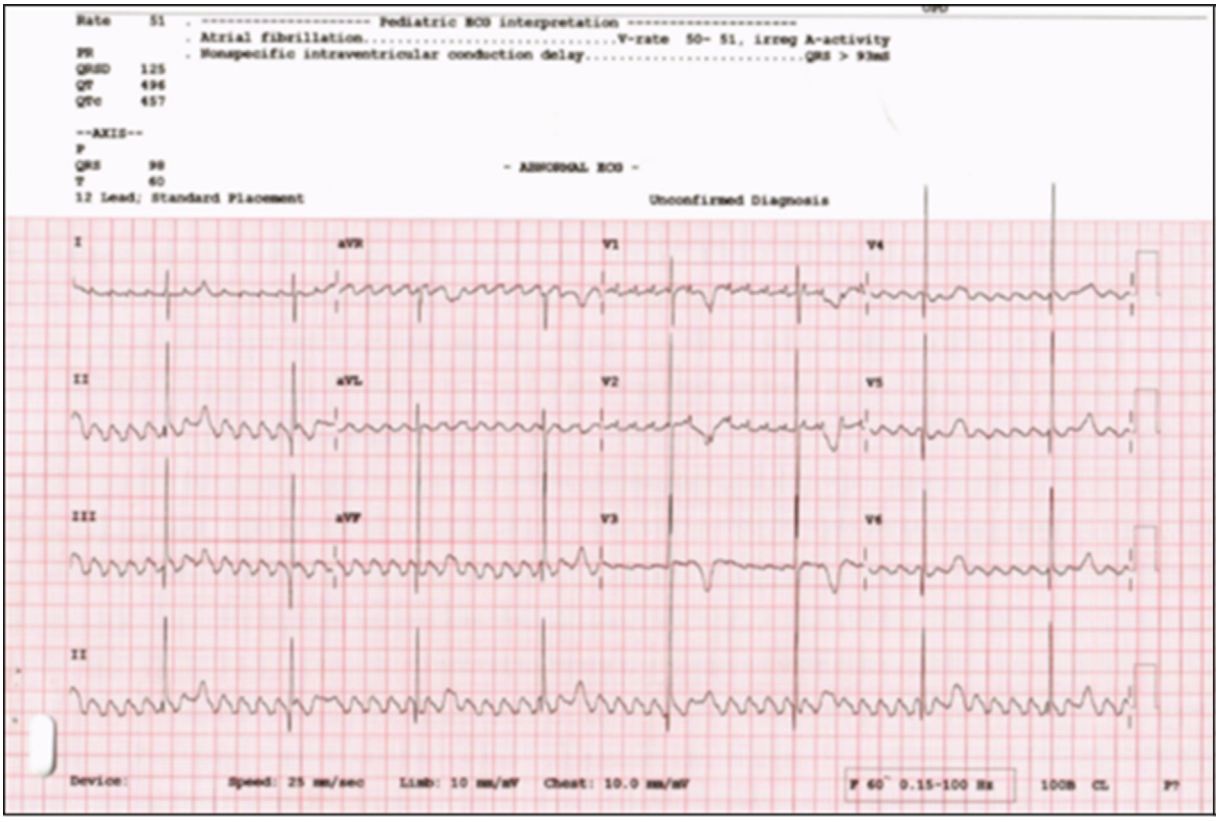

Fig. 3. 12 lead ECG showing atrial flutter with complete heart block with atrioventricular dissociation and almost 6 to 1 AV block. There is junctional escape rhythm, with narrow QRS complexes and a ventricular rate of 51 bpm.

are unaware that they carry the antibodies. This makes pre-emptive evaluation difficult [1,2].

The incidence of CCHB in patients with SSA/Ro and SSB/La antibodies is approximately 2–3%. The risk increases to 12–15% in fetuses of mothers with previous children with CCHB [3]. There is a risk for myocarditis, endocardial fibroelastosis (EFE), valve damage, and the development of cardiomyopathy [3,4]. In general, only a third of mothers of CCHB fetuses have an identified autoimmune disorder such as Sjogren's or Lupus at the time of CCHB diagnosis in their fetuses. Risk factors associated with *in-utero* mortality include fetal hydrops, diagnosis of CCHB at <20 weeks, ventricular escape rate <55 bpm, and impaired left ventricular function [3].

Fetal echocardiography is the standard tool for the diagnosis and management of fetal arrhythmias including CCHB [5].

Corticosteroids, plasmapheresis, and/or hydroxychloroquine (HCQ) are used for mothers with anti-

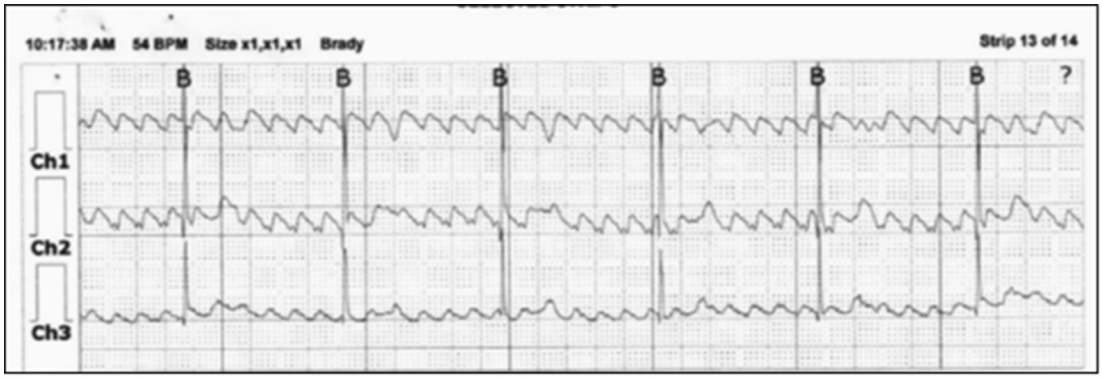

Fig. 4. Twenty-four-hour Holter monitoring, sample strip rhythm showing atrial flutter with an atrial rate of 300 bpm, narrow complex QRS complexes indicating a junctional escape rhythm, with a ventricular rate of 54 bpm.

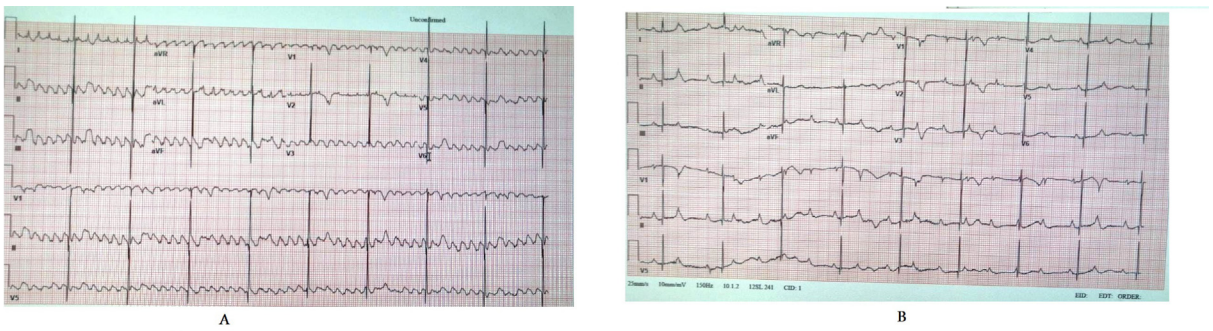

Fig. 5. (A and B)12 lead ECG pre-cardioversion (5A) as well as post cardioversion (5 B). Pre-cardioversion the atrial rate is above 300 and the ventricular rate is around 55 bpm. Post-cardioversion there is a return of organized atrial rhythm with complete heart block and junctional escape rhythm (narrow QRS complexes) with a ventricular rate of 53 bpm.

SSA/SSB antibody-positive pregnant women. Some are used before pregnancy to prevent the recurrence of CCHB in the coming fetus [3,6].

To our knowledge, this is the first case reporting postnatal atrial flutter in a patient with CCHB.

Atrial flutter on a background of CCHB was reported in a fetus who was delivered at 36 + 1 week's gestation with mild respiratory distress, CCHB, and a ventricular rate of 50 bpm. A permanent epicardial pacemaker was implanted at 7 days of age. In this case, no episodes of atrial flutter were noted during the postnatal course [7].

The management of isolated fetal atrial flutter depends upon the effect of the arrhythmia on the fetus. If the fetus is near-term, delivery is the best. Observation and monitoring without medications is an option if there are no hydrops and the cardiac function is maintained, otherwise, material administration of antiarrhythmic medications might be required. The presence of heart block would protect against fast ventricular rates resulting from atrial flutter giving time for conservative management in the absence of fetal compromise [5].

Postnatally, the development of atrial flutter in the background of CCHB is challenging. In atrial flutter, the rate is regular with an atrial rate of 300–600 bpm, accompanied by variable degrees of atrioventricular (AV) conduction block, resulting in slower ventricular rates.

Treatment of children with atrial flutter depends on the age of presentation and baseline cardiac anatomy. In general, treatment may involve medications, cardiac pacing, cardioversion, radiofrequency catheter ablation, or surgical procedures. The guidelines recommend trans-esophageal echocardiography (TEE) for patients with atrial fibrillation (AF) or atrial flutter (AFL) for >48 h, due to the risk of intracardiac thrombus formation [8].

Sinus rhythm can be restored by electrical cardioversion, transesophageal pacing, or antiarrhythmic drugs. In stable newborns, antiarrhythmic drugs can be tried. However, it takes some time to restore sinus rhythm. The recommended drugs are digoxin with the addition of flecainide or amiodarone in case of no therapeutic effect [9]. Direct-current cardioversion is reported to be the most effective treatment of atrial flutter [10].

Our patient had an atrial flutter on top of a baseline CCHB. He was successfully managed with direct current cardioversion using 1 J/kg, with no recurrence of the atrial flutter during the follow-up period.

4. Conclusion

Atrial flutter can be seen on top of CCHB. It can be managed as usual with synchronized cardioversion.

Author contribution

Conception and design of Study: AAA, AA, AA, HDA, WA. Literature review: AAA, AA, AA, HDA, WA. Acquisition of data: AAA, AA, AA, HDA, WA. Analysis and interpretation of data: AAA, AA, AA, HDA, WA. Research investigation and analysis: AAA, AA, AA, HDA, WA. Data collection: AAA, AA, AA, HDA, WA. Drafting of manuscript: AAA. Revising and editing the manuscript critically for important intellectual contents: AAA, AA, AA, HDA, WA. Data preparation and presentation: AAA. Supervision of the research: WA. Research coordination and management: AA.

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Conflict of interest

All authors have no conflicts of interest to disclose.

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