

Congenital Atrioventricular Block in an Adolescent Female: A Case Report

ABSTRACT

Introduction:

Congenital atrioventricular block (CAVB) is a rare cardiac condition characterized by impaired electrical conduction between the atria and ventricles. We present a case report of an 18-year-old female patient with no significant medical history admitted to the cardiology department with complete atrioventricular block.

Case Presentation:

The patient presented with a two-week history of syncope episodes without chest pain or other symptoms, alongside preserved general condition. Physical examination revealed bradycardia (40 bpm) and confirmed complete atrioventricular block on electrocardiogram. Transthoracic echocardiography showed preserved ejection fraction with no significant valvular disease. Laboratory investigations and serological tests were conducted to exclude underlying pathologies, with negative results. Subsequently, the patient underwent implantation of a double-chamber pacemaker.

Discussion:

The case highlights the diagnostic challenges and management strategies associated with congenital atrioventricular block. While congenital and acquired causes should be considered, the absence of identifiable risk factors and negative serological markers suggested a congenital etiology in this case. Management typically involves pacemaker implantation to restore normal heart rhythm and prevent complications. Regular follow-up is essential for monitoring device function and detecting potential complications.

Conclusion:

Congenital atrioventricular block is a rare but potentially serious condition that requires prompt diagnosis and management. Pacemaker implantation is the cornerstone of treatment to prevent complications and improve patient outcomes. Further research is needed to better understand the pathophysiology and optimize treatment strategies for this complex cardiac disorder.

Keywords: *Cardiac arrhythmia, Congenital atrioventricular block, Pacemaker implantation*

1. INTRODUCTION

Congenital atrioventricular block (CAVB) is a rare condition characterized by impaired electrical conduction between the atria and ventricles, leading to various degrees of heart block. This condition can result in significant morbidity and mortality, especially if not diagnosed and managed appropriately. The etiology of CAVB is often linked to maternal autoimmune diseases, such as lupus, which can result in transplacental passage of autoantibodies affecting the fetal cardiac conduction system.[1]

Despite advances in prenatal screening and diagnostic techniques, CAVB remains a diagnostic challenge due to its variable presentation and the potential for progression to complete heart block. Early identification and management are crucial to improving outcomes in affected individuals. Current management strategies may include medical therapy, pacemaker implantation, and close monitoring during pregnancy and after birth. [2-13]

In this case report, we present an 18-year-old female patient with a history of maternal lupus who was diagnosed with CAVB. This case underscores the importance of awareness and early diagnosis, and highlights the challenges and strategies in managing this condition

2. CASE PRESENTATION

An 18-year-old female with a history of maternal lupus presented to the cardiology department with a two-week history of syncopal episodes without chest pain or other signs, all developing in a context of preserved general condition; admitted to the cardiology department of CHU IBN ROCHD for high degree atrioventricular block.

Physical examination revealed bradycardia (40 bpm), with normal arterial tension at 130/60mmHg, cardiovascular auscultation was normal and second degree Mobitz II block was confirmed by electrocardiogram (ECG) (Figure 1).

Laboratory tests, including blood count, electrolytes, and renal function tests, were within normal limits:

- Hb was correct at 13.3 g/dl, leukocyte count at 13,520/mm³, with PNN at 9010; platelet count at 243,000, SV count at 27 mm/1st hour, and CRP at 7.8 g/l,

- Serum electrolyte count was correct with sodium at 142 mmol/l, potassium at 4.7 mmol/l, chloride at 100 mmol/l, calcium at 96 mg/l, magnesium at 20mg/l and phosphorus at 44 mg/l.

- Renal function was correct: urea 0.26 g/L, creatinine 7.8 mg/L, glomerular filtration rate estimated by MDRD at 102 ml/min/1.73m²,

- Troponin (HS) normal at 7.8 ng/L.

As part of an etiologic work-up, we performed the following:

- TSHus: normal at 2.57 mU/L.

- Viral serologies (CMV, EBV, HBV, HCV, VIH 1, 2 and syphilis serology): negative.

- Rheumatoid factor: negative

- Anti-nuclear antibodies, anti-DNA antibodies, ANCA: negative.

Transthoracic echocardiography showed preserved ejection fraction with ventricular dilatation (ventricular remodeling). There was no significant valvular disease. (Figure 2)

The patient was implanted with a dual chamber pacemaker (Figure 3). Short-term follow-up showed positive ventricular remodeling after pacemaker implantation, the cavities were no longer dilated and the patient remained asymptomatic. (Figure 4)

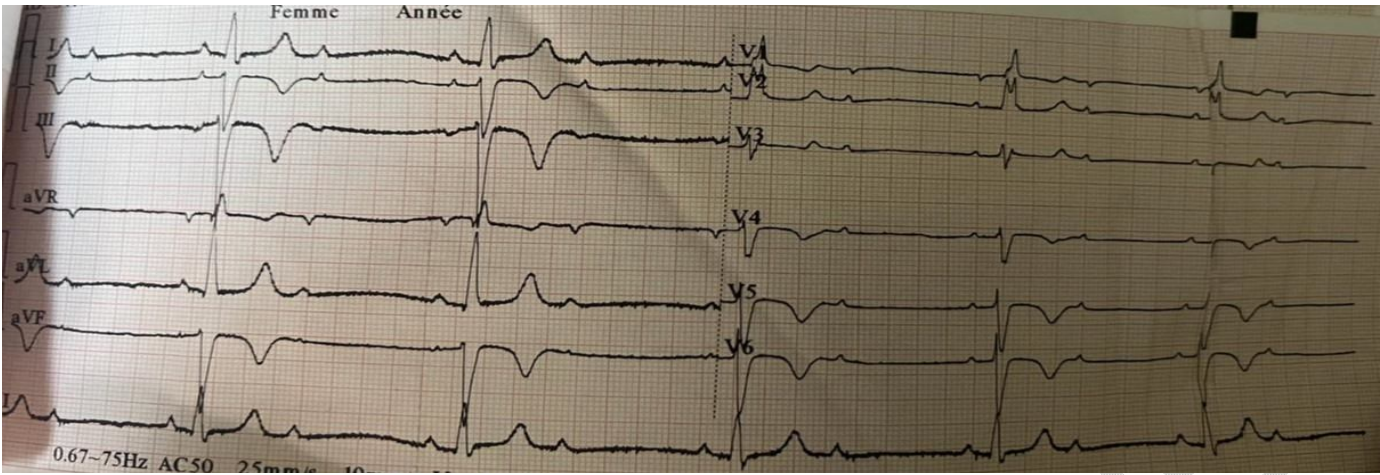


Figure 1: high degree atrioventricular block 2nd degree Mobitz II with 2/1 conduction

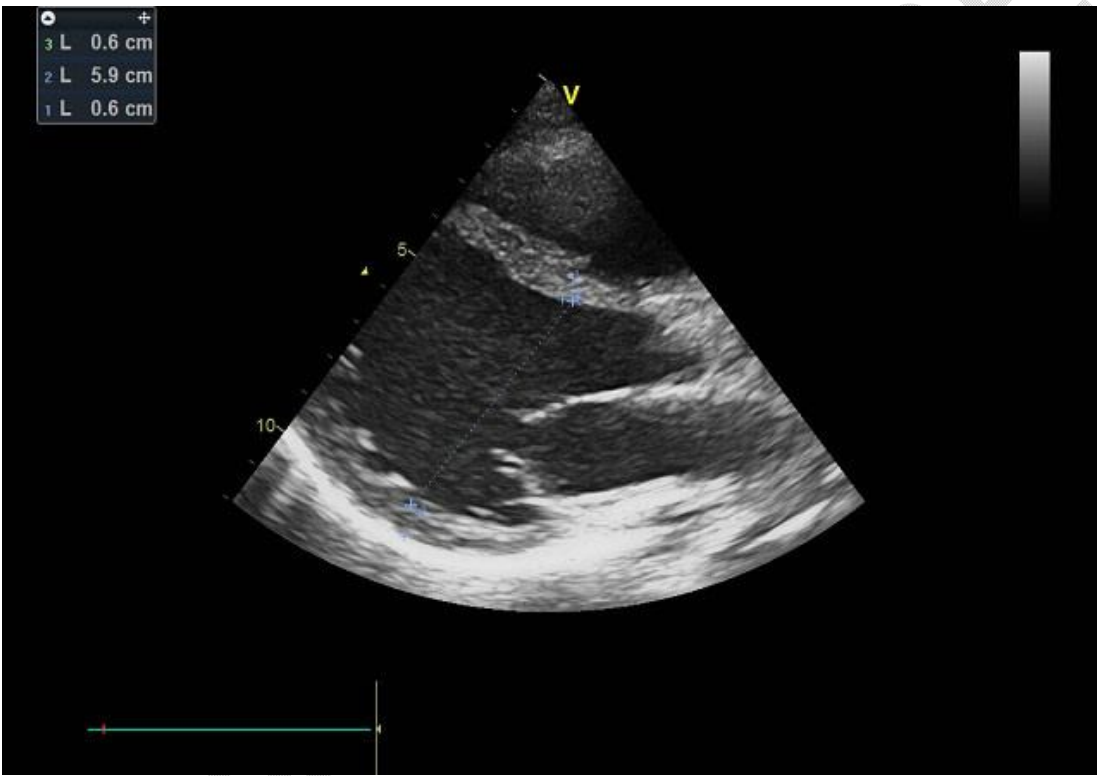




Figure2: right and left ventricular dilation secondary to CAAB

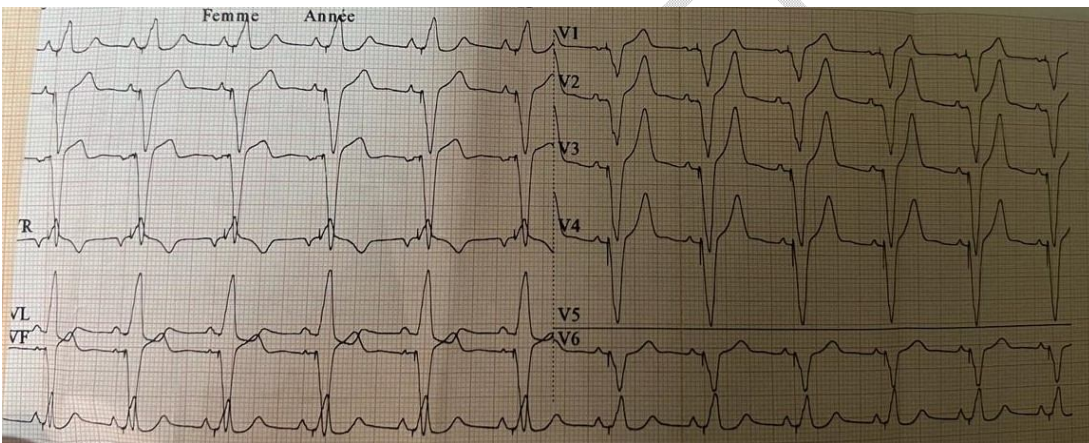


Figure 3: ECG after implantation of a double-chamber pacemaker shows an electro simulated rhythm

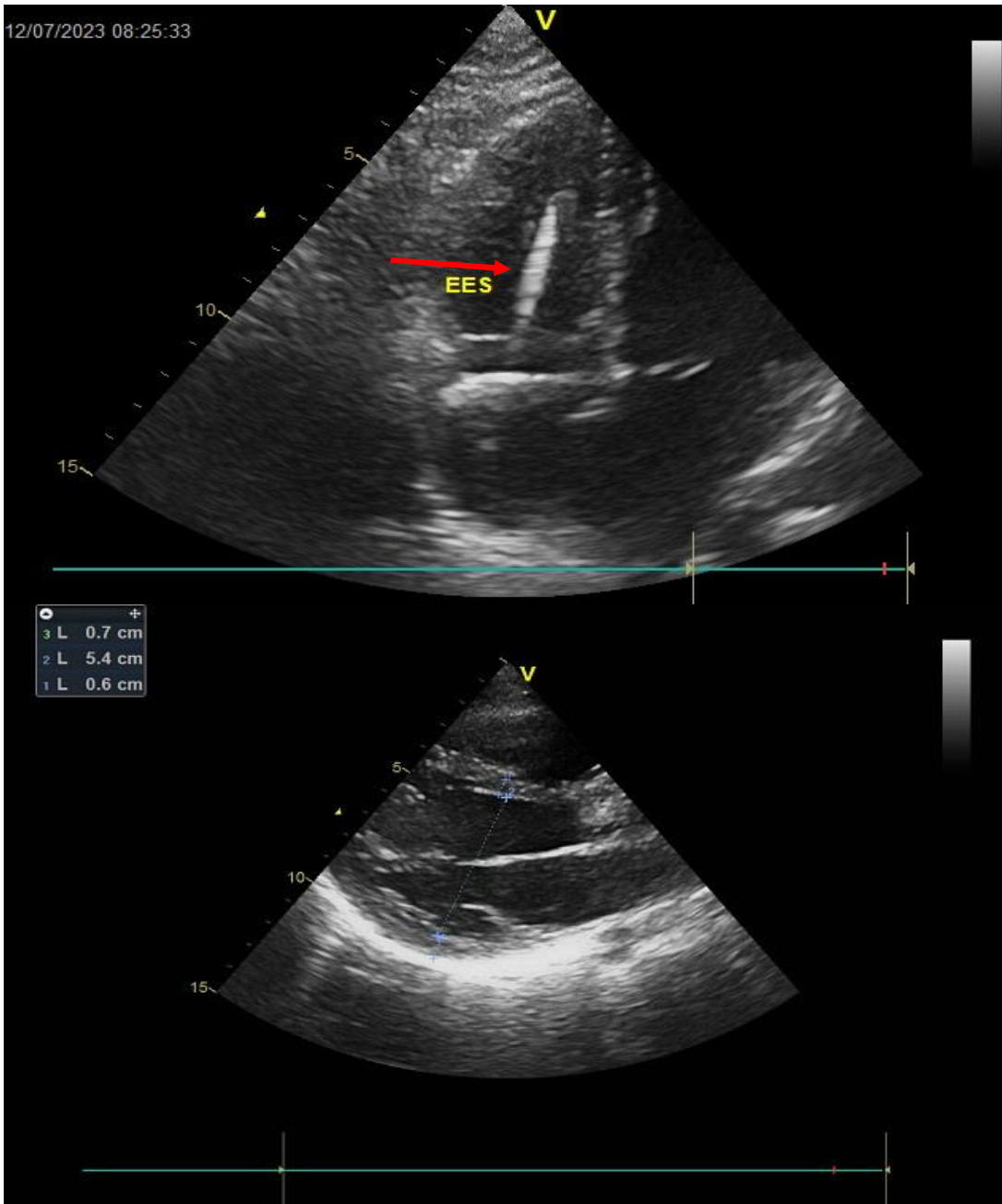


Figure4: electrocystolic entrainment lead in place with left ventricular remodeling

DISCUSSION

The etiology of congenital atrioventricular block (CAVB) can be congenital or acquired. Congenital cases are often associated with maternal autoimmune diseases or congenital heart defects, while acquired CAVB may result from infections, medications, or autoimmune conditions. In our case, the absence of risk factors and negative serological markers suggested a congenital etiology. This highlights the importance of considering congenital causes even in the absence of typical risk factors. [1,2]

Management of CAVB involves pacemaker implantation to restore normal heart rhythm and prevent symptoms such as syncope and heart failure. Regular follow-up is essential to monitor device function and assess for complications. Our case emphasizes the critical role of early intervention and continuous monitoring in improving patient outcomes. [3,4]

The pathophysiology of CAVB involves disruption of the normal electrical conduction system between the atria and ventricles. In congenital cases, this disruption may result from abnormal development of the conduction tissue during fetal life. Conversely, acquired CAVB can occur due to inflammatory processes, ischemic events, or exposure to certain medications or toxins. Understanding the underlying cause of CAVB is essential for guiding appropriate management and prognosis.

Diagnostic evaluation of CAVB typically involves electrocardiography (ECG) to confirm the presence and severity of heart block. Transthoracic echocardiography is useful for assessing cardiac structure and function, ruling out associated congenital anomalies, and determining the need for additional imaging modalities. Laboratory investigations, including blood tests for autoimmune markers, viral serologies, and thyroid function, help identify potential underlying causes and guide further management. [5,6]

In our case, the patient underwent thorough serological and autoimmune screening, which yielded negative results, suggesting a congenital rather than acquired etiology. This highlights the importance of considering both congenital and acquired causes in the diagnostic work-up of CAVB, particularly in cases with atypical presentations or unclear etiology. The negative serological markers in our patient contrast with existing literature where maternal autoimmune disease is a common cause, suggesting that congenital CAVB can occur even in the absence of maternal autoimmune conditions. Management of CAVB focuses on restoring normal heart rhythm and preventing complications such as syncope, heart failure, and sudden cardiac death. [7]

Pacemaker implantation is the cornerstone of treatment, providing continuous electrical stimulation to ensure adequate ventricular pacing. The decision to implant a pacemaker depends on the severity of symptoms, degree of heart block, and overall clinical status of the patient. Cardiac stimulation permanent is indicated in symptomatic patients (syncope, congestive heart failure, heart failure chronotropic) or in asymptomatic patients with at least one of the following: dysfunction ventricular, prolonged QTc interval, ventricular extrasystole complexes, wide QRS escape rhythm, heart rate < 55 bpm, before age 1 year, breaks cores > 3 times the base cycle.[8]

However, retained patients should not be considered cured. Also, regular assessment of left ventricular size, volumes and function is required after implantation because these patients may develop dilated heart disease several years later the procedure, consequence of autoimmune myocarditis or asynchrony induced by ventricular stimulation prolonged. The stimulation site also plays an important role in these patients who will be stimulated throughout their life. The harmful effects of ventricular stimulation prolonged right are now well known and the biventricular resynchronization that the implantation of a left ventricular lead could be beneficial. Close monitoring and regular follow-up are essential to assess pacemaker function, adjust settings as needed, and detect any complications such as lead dislodgement, infection, or device malfunction. [9,10]

CAVB is a rare but potentially serious condition that requires a multidisciplinary approach for accurate diagnosis and optimal management. While congenital cases often present in childhood or adolescence, acquired forms can occur at any age and may be associated with various underlying etiologies. Early recognition, appropriate intervention, and long-term follow-up are essential for improving outcomes and quality of life in patients with CAVB. Our case underscores the importance of considering CAVB in differential diagnosis, even in the absence of common risk factors, and highlights the need for further research to better understand the pathophysiology and refine treatment strategies for this complex cardiac disorder[11,12]

4. CONCLUSION

Congenital atrioventricular block is a rare condition that can present with syncope and bradycardia in otherwise healthy adolescents. Prompt diagnosis and management, including pacemaker implantation, are crucial to prevent complications and improve quality of life in affected individuals. Further research is needed to better understand the underlying mechanisms and optimize treatment strategies for CAVB.

CONSENT

As per international standard or university standard, patientwritten consent has been collected and preserved by the author(s)

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s)

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- 1.
- 2.
- 3.

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