Case report

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. We present a case of CAVB in an 18-year-old female

patient with no significant medical history, highlighting the diagnostic challenges and management strategies associated with this condition.

2. CASE PRESENTATION

An 18-year-old female with no notable medical history presented to the cardiology department with a two-week history of syncope episodes with no notion of chest pain or other sign, all evolving in a context of preserved general condition; admitted to the cardiology department of CHU IBN ROCHD for a complete atrioventricular block.

Physical examination revealed bradycardia (40 bpm) , with normal arterial tension at 130/60mmHg, cardiovascular auscultation was normal and a complete atrioventricular block confirmed by electrocardiogram (ECG) .**(Figure1)**

Transthoracic echocardiography showed preserved ejection fraction with ventricular dilatation (ventricular remodeling). There was no significant valvular disease. **(Figure2)**

Laboratory investigations, including blood count, electrolyte levels, and renal function tests, were within normal limits:

- Hb was correct at 13.3 gr/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 7.8g/l,

- The count of serum electrolytes was correct with sodium at 142 mmol/l, Potassium at 4.7 mmol/l, chlorides 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 7.8mg/L, estimating glomerular filtration rate according to MDRD at 102 ml/min/1.73m2, ,

- Troponin (HS) normal at 7.8 ng/l

As part of an etiological work-up, we completed the following:

- TSHus: normal at 2.57 mUI/L.

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

- Rheumatoid factor : negative

- Anti nuclear antibody, Anti DNA antibody, ANCA : negative.

The patient was implanted with a double-chamber pacemaker**(figure3).** short-term follow-up showed positive ventricular remodeling after implantation of the pace maker, the cavities were no longer dilated and the patient remained asymptomatic. **(Figure4)**

DISCUSSION

The etiology of CAVB can be congenital or acquired. Congenital cases are often associated with maternal autoimmune diseases or congenital heart defects. 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. 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.[1]

The pathophysiology of CAVB involves disruption of the normal electrical conduction system between the atria and ventricles. In congenital cases, this may result from abnormal development of the conduction tissue during fetal life. Alternatively, 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. [2,3]

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.[4]

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.

Management of CAVB focuses on restoring normal heart rhythm and preventing complications such as syncope, heart failure, and sudden cardiac death. 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. 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. [5,6]
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. Further research is needed to better understand the pathophysiology of CAVB and refine treatment strategies for this complex cardiac disorder.[7,8]

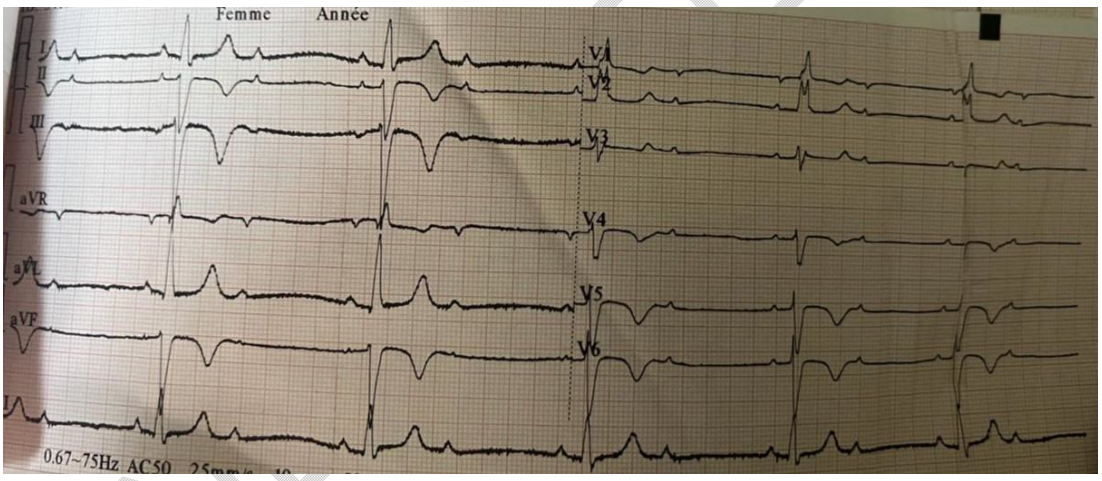

Figure 1: Complete atrio-ventricular block.

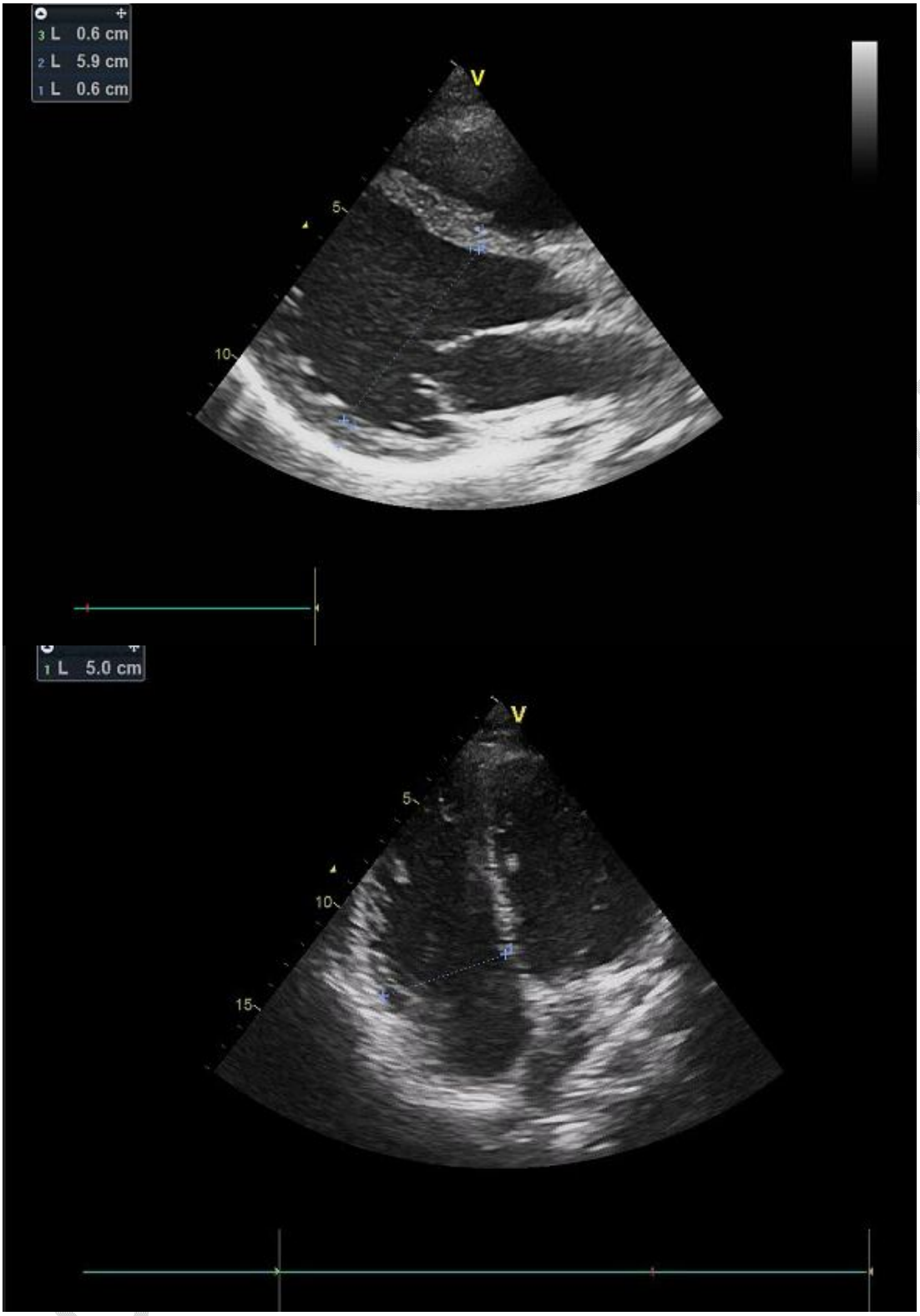

Figure2: right and left ventricular dilation secondary to CAVB

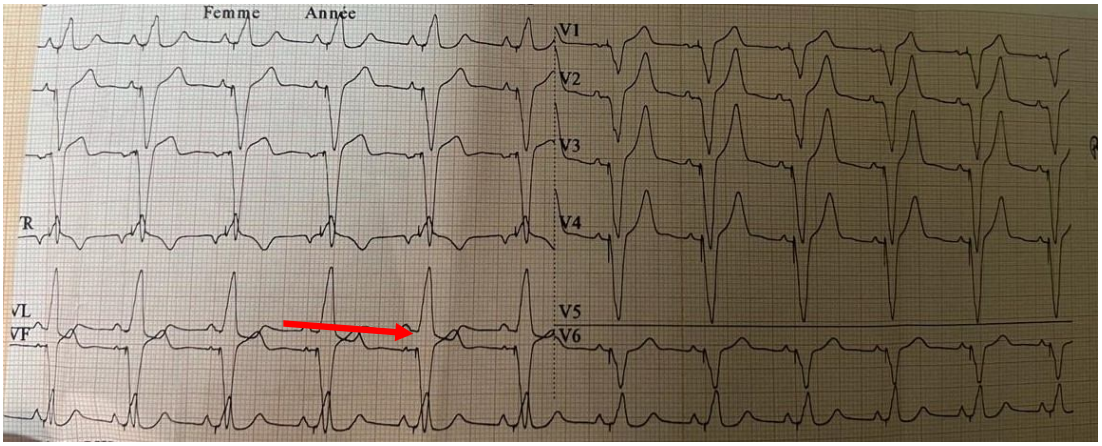

Figure 3: ECG after implantation of a double-chamber pacemaker shows an electro simulated rythm

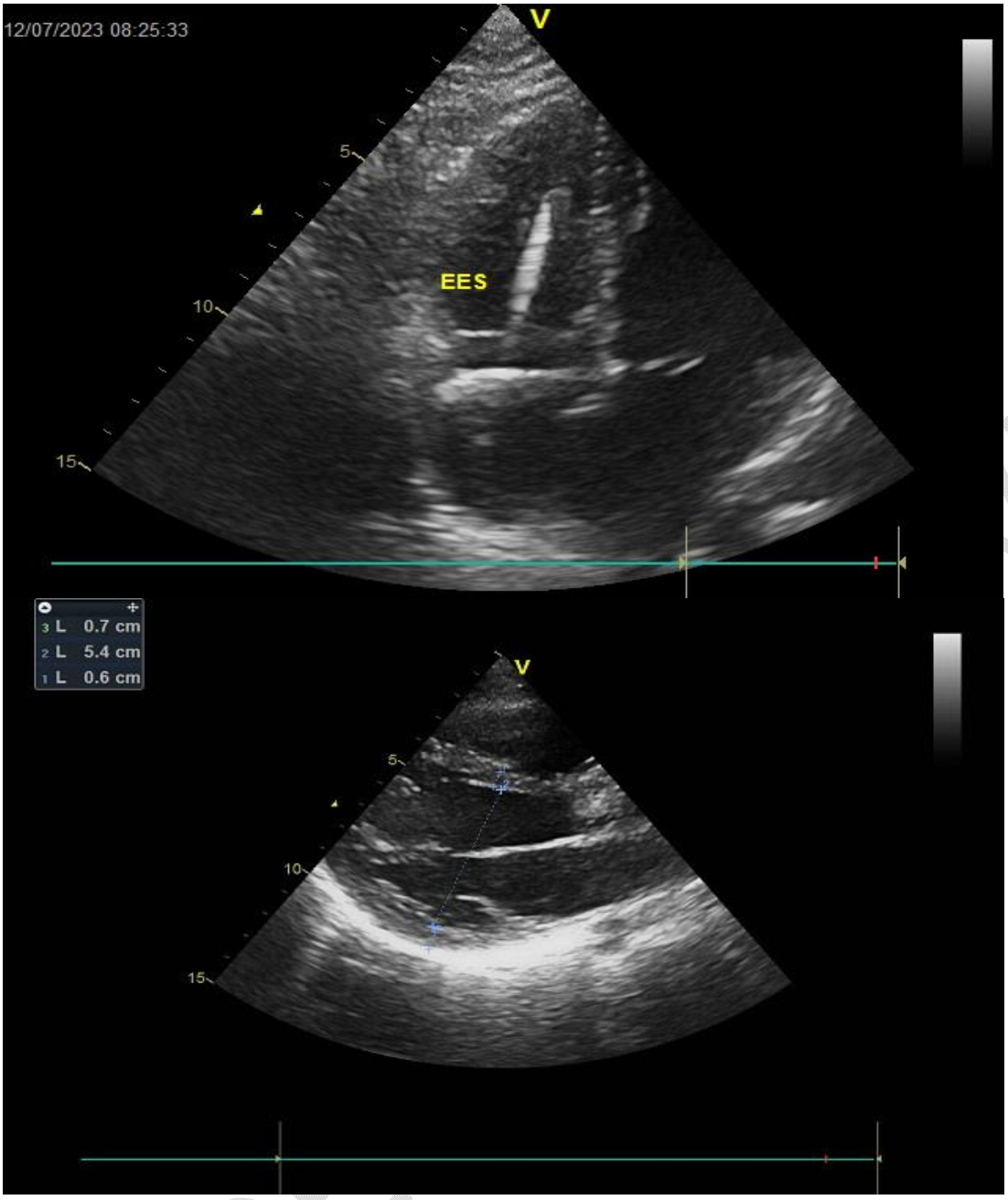

Figure4: electrosystolic entrainment lead in place with left ventricular remodeling

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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