Asian Journal of Cardiology Research

Volume 7, Issue 1, Page 104-110, 2024; Article no.AJCR.118491

Congenital Atrioventricular Block in an Adolescent Female: A Case Report

Zahri Soukaina [a*], Ghali Bennani [a], Lamiaa Tlohi [a], Samia Ejjebli [a], Imad Nouamou [a], Salim Arous [a], Abdennaser Drighil [a] and Rachida Habbal [a]

[a] Cardiology Department, UHC Ibn Rochd, Casablanca, Morocco.

Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Article Information

Open Peer Review History:
This journal follows the Advanced Open Peer Review policy. Identity of the Reviewers, Editor(s) and additional Reviewers, peer review comments, different versions of the manuscript, comments of the editors, etc are available here: https://www.sdiarticle5.com/review-history/118491

Case Report

Received: 02/04/2024
Accepted: 07/06/2024
Published: 12/06/2024

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.

**Corresponding author: Email: zahrisoukaina@gmail.com;*

Cite as: *Soukaina, Zahri, Ghali Bennani, Lamiaa Tlohi, Samia Ejjebli, Imad Nouamou, Salim Arous, Abdennaser Drighil, and Rachida Habbal. 2024. "Congenital Atrioventricular Block in an Adolescent Female: A Case Report". Asian Journal of Cardiology Research 7 (1):104-10. https://journalajcr.com/index.php/AJCR/article/view/207.*

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) (Fig. 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 (Fig. 2).

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

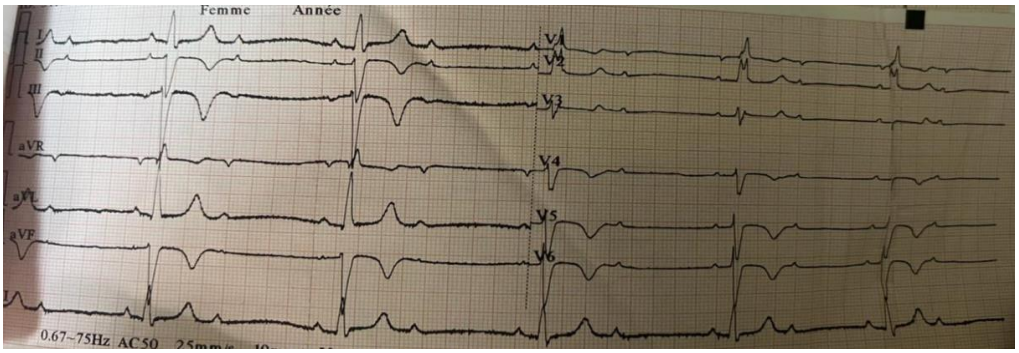

Fig. 1. High degree atrioventricular block 2nd degree Mobitz II with 2/1 conduction

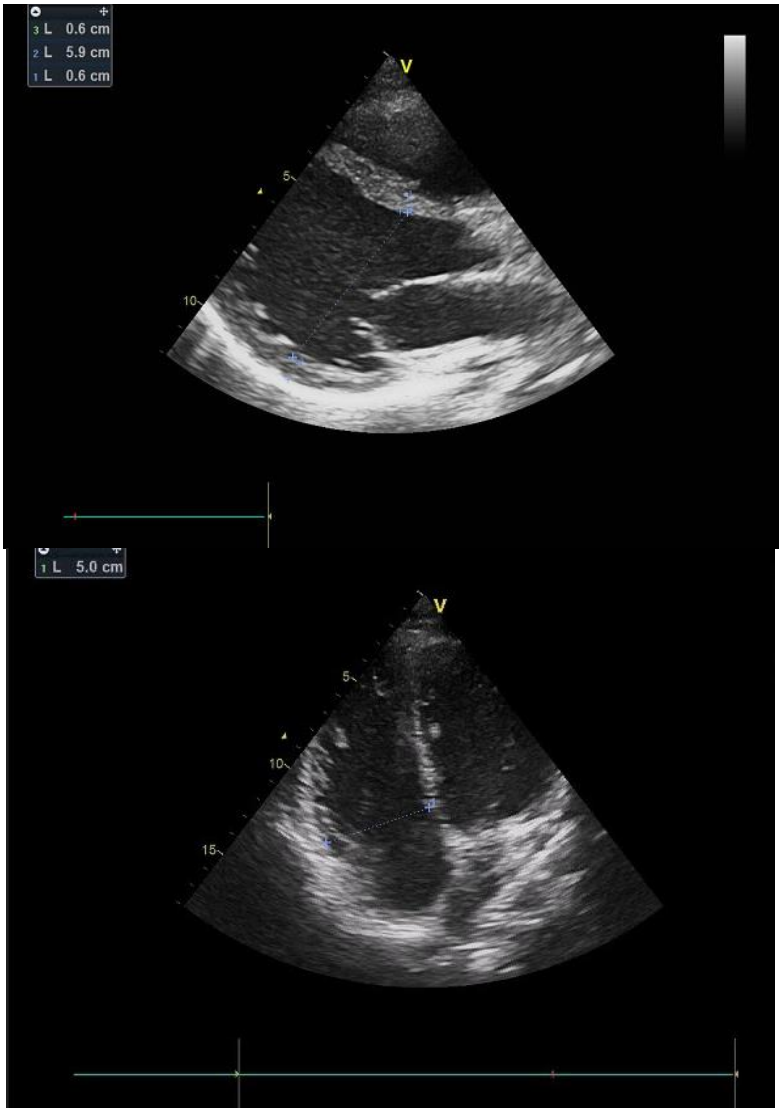

Fig. 2. Right and left ventricular dilation secondary to CAVB

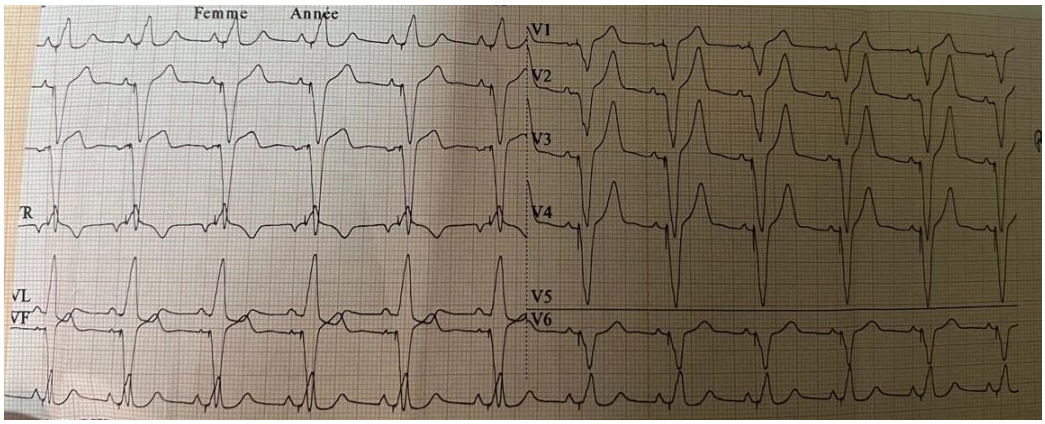

Fig. 3. ECG after implantation of a double-chamber pacemaker shows an electro simulated rythm

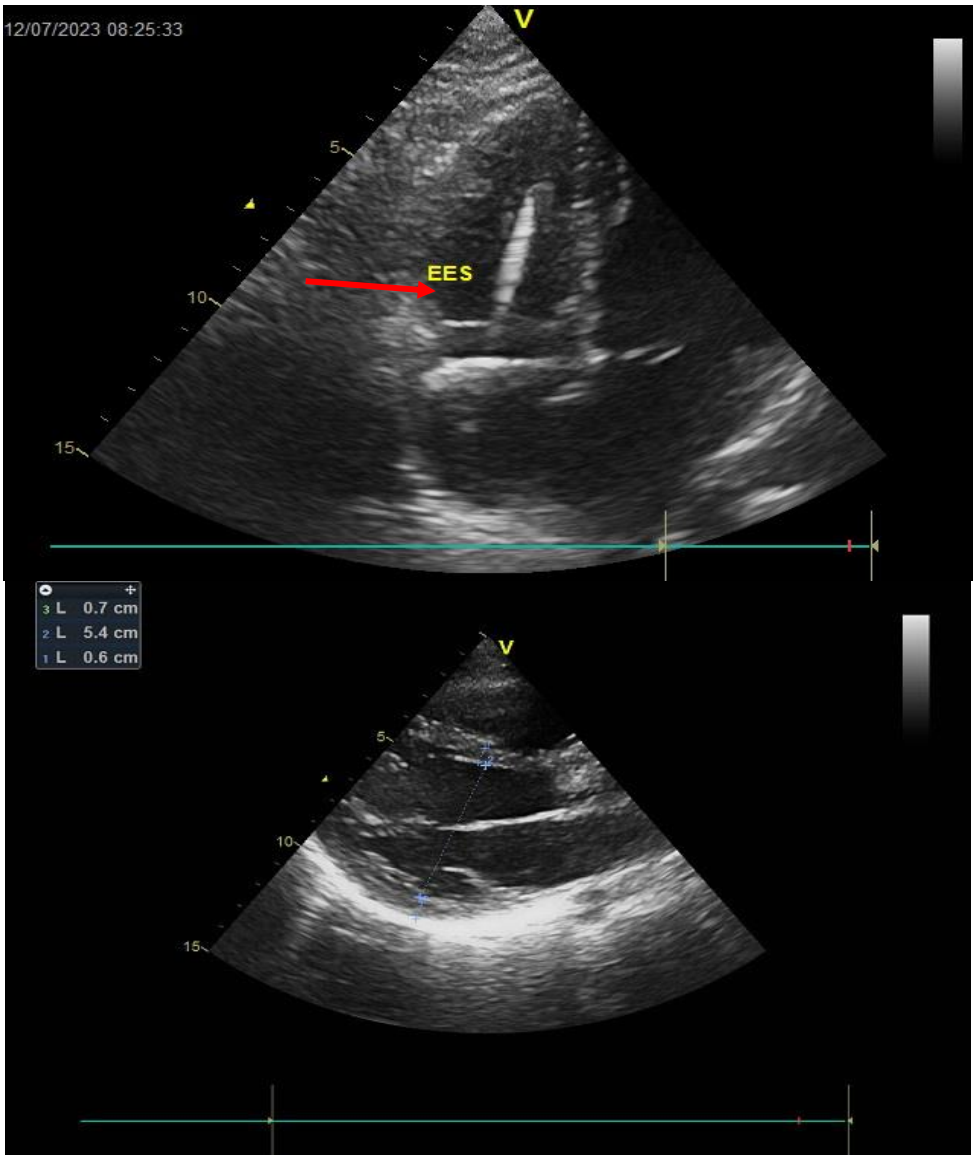

Fig.4. Electrosystolic entrainment lead in place with left ventricular remodeling

3. 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, patient written 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)

DISCLAIMER (ARTIFICIAL INTELLIGENCE)

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc) and text-to-image generators have been used during writing or editing of manuscripts.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Epstein AE, DiMarco JP, Ellenbogen KA, et al. 2012 ACCF/AHA/HRS focused update incorporated into the ACCF/AHA/HRS 2008 guidelines for device-based therapy of cardiac rhythm abnormalities: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines and the Heart Rhythm Society. J Am Coll Cardiol. 2013;61(3).
2. Baruteau AE, Pass RH, Thambo JB, et al. Congenital and childhood atrioventricular blocks: Pathophysiology and contemporary management. Eur J Pediatr. 2016;175(9): 1235-1248.
3. Jaeggi ET, Hornberger LK, Smallhorn JF, et al. Prenatal diagnosis of complete atrioventricular block associated with structural heart disease: Combined experience of two tertiary care centers and review of the literature. Ultrasound Obstet Gynecol. 2005;26(1):16-21.
4. Melim C, Pimenta J, Areias JC. Congenital atrioventricular heart block: From diagnosis to treatment. Servic¸o de Cardiologia Pediátrica do Centro Hospitalar de São João, Departamento de Ginecologia Obstetrícia e Pediatria da FMUP, Porto, Portugal; 24 February 2022.
5. Balmer C, Bauersfeld U. Do all children with congenital complete atrioventricular block require permanent pacing? Indian Pacing Electrophysiol J. 2003;3:178-83.
6. Baruteau AE, Fouchard S, Behaghel A, et al. Characteristics and long-term outcome of non-immune isolated atrioventricular block diagnosed in utero or early childhood: A multicentre study. Eur Heart J. 2012;33:622-9.
7. Jalal Z, Bordachar P, Labrousse L, Mondoly P, Ritter P, Thambo J.B. Pacing in the paediatric population: Indications, implantation strategies, evolution and new stimulation techniques. Archives of Cardiovascular Diseases Supplements. 2012;4:138-147.
8. Beaufort- Krol G, Schasfoort- Van Leeuwen MJ, Stienstra Y, et al. Longitudinal echocardiographic follow- up in children with congenital complete atrioventricular block. Pacing ClinElectrophysiol. 2007;30:1339- 43.
9. Thambo JB, Bordachar P, Garrigue S, et al. Detrimental ventricular remodelling in patients with congenital complete heart block and chronic right ventricular apical pacing. Circulation. 2004;110: 3766- 72.
10. Moak JP, Hasbani K, Ramwell C, et al. Dilated cardiomyopathy following right ventricular pacing for AV block in young patients: Resolution after upgrading to biventricular pacing systems. J Cardiovasc Electrophysiol. 2006;17:1068- 71.
11. Martin TA. Congenital heart block: Current thoughts on management, morphologic spectrum, and role of

intervention. Cardiol Young. 2014;24 Suppl. 2:41-6.

12. Anderson RH, Wenick AC, Losekoot TG, et al. Congenitally complete heart block. Developmental aspects. Circulation. 1977;56:90-101.

13. Barra sn, providencia r, paiva l, nascimento j, marques al. A review on advanced atrioventricular block in young or middle-aged adults. Pacing and Clinical Electrophysiology. 2012 nov;35(11):1395-405.

Peer-review history:
The peer review history for this paper can be accessed here:
https://www.sdiarticle5.com/review-history/118491