CASE REPORT

PERMANENT PACEMAKER IMPLANTATION IN A PATIENT WITH CONGENITALLY CORRECTED TRANSPOSITION OF GREAT ARTERIES AND MESOCARDIA: AN INFREQUENT HAPPENSTANCE

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We present a compelling case of a 37-year-old male who, after three decades of asymptomatic living, was diagnosed with congenitally corrected transposition of great arteries (CCTGA). Initially presenting with troubling symptoms of dizziness and syncope, further evaluation revealed complete heart block, a significant finding necessitating intervention. Despite the rarity of symptomatic presentation in CCTGA, the severity of the heart block prompted the implantation of a DDD pacemaker to ensure adequate management. With meticulous care and precision, the implantation procedure was successfully executed, offering hope for improved cardiac function and symptom relief. Remarkably, the patient remained asymptomatic during subsequent follow-up visits, highlighting the effectiveness of the intervention in mitigating the risks associated with high atrioventricular block in CCTGA. This case serves as a testament to the importance of vigilant monitoring and timely intervention in managing complex cardiac anomalies, ultimately enhancing patient outcomes and quality of life.

Keywords: Congenitally corrected transposition of great arteries; pacemaker implantation; complete heart block; mesocardia; dizziness; syncope

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INTRODUCTION

Congenitally corrected transposition of the great arteries (CCTGA) presents a unique cardiac anomaly characterized by the transposition of the great arteries, ventricular inversion, and atrioventricular valve and conduction system inversion, while the atrial positioning remains normal. It is considered a rare congenital heart disease, with an incidence ranging from 0.5 percent to 1.4 percent among all congenital heart defects.[1,2] While approximately 90 percent of cases of CCTGA are associated with additional anomalies such as ventricular septal defects (VSD), pulmonary stenosis (PS), or pathological mitral and tricuspid valves, isolated cases do occur.[3]

Typically, individuals with isolated CCTGA remain asymptomatic for extended periods, often escaping clinical detection until later stages of life. Diagnosis may occur incidentally through routine physical examinations revealing murmurs on auscultation, abnormal findings on electrocardiography (ECG), or an enlarged cardiothoracic ratio on chest X-ray.[4] However, despite the asymptomatic nature of many cases, there exists a subset of patients who may experience life-threatening complications such as heart blocks, heart failure, and serious arrhythmias, contributing to heightened mortality rates.[5] Notably, the prognosis for individuals without associated anomalies largely hinges upon the presence or absence of ventricular dysfunction.[6]

In this report, we present a compelling case of a 37-year-old male diagnosed with CCTGA, who, contrary to the typical course, presented with troubling symptoms including dizziness and recurrent episodes of unconsciousness. Recognizing the gravity of the situation and the potential implications for systemic ventricular function, the patient underwent successful implantation of a DDD pacemaker. This intervention was crucial in managing the high atrioventricular block, ultimately aiming to preserve cardiac function and improve the patient's quality of life. Subsequent follow-up assessments have revealed the patient to remain asymptomatic, underscoring the effectiveness of the intervention in mitigating the risks associated with symptomatic CCTGA.

This case serves to illuminate the uncommon occurrence of symptomatic complete heart block in CCTGA patients, particularly in the presence of mesocardia. By sharing this case, we contribute to the growing body of literature on the management of

complex cardiac anomalies, emphasizing the importance of timely intervention and individualized treatment strategies in optimizing patient outcomes.

CASE REPORT

Patient Information: The subject of this case report is a 37-year-old male, professionally engaged as a heavy vehicle driver, who sought medical attention due to recurrent episodes of dizziness and syncope. Upon initial assessment, physical examination findings were unremarkable, with no audible murmurs detected, and laboratory investigations returned within normal parameters. However, electrocardiography (ECG) revealed a concerning finding of complete heart block, with a notably reduced heart rate of approximately 40 beats per minute.

Further diagnostic evaluation via echocardiography unveiled the presence of CCTGA, accompanied by mesocardia, a rare finding indicative of the heart being positioned more centrally within the chest cavity. Additionally, mild tricuspid regurgitation and mildly impaired right ventricular function were noted. This comprehensive assessment provided crucial insights into the patient's cardiac anatomy and function, guiding subsequent therapeutic interventions.

The patient's occupation as a heavy vehicle driver underscores the significance of prompt diagnosis and management of his cardiac condition, as episodes of dizziness and syncope pose a significant safety risk in his line of work. Through meticulous evaluation and diagnostic imaging, a clear understanding of the underlying cardiac pathology was established, informing tailored treatment strategies aimed at optimizing the patient's cardiovascular health and overall well-being.

Clinical Findings: The predominant clinical manifestations prompting medical evaluation in this case were recurrent episodes of dizziness and syncope, symptoms indicative of compromised cerebral perfusion secondary to cardiovascular dysfunction. These alarming symptoms were found to correlate with the presence of complete heart block, a critical electrocardiographic finding suggestive of impaired atrioventricular conduction.

Further investigation through echocardiography unveiled the complex cardiac anatomy characteristic of CCTGA, a rare congenital anomaly. Notably, in addition to the primary diagnosis of CCTGA, echocardiographic assessment revealed mild tricuspid regurgitation, indicative of valvular insufficiency, and mild right ventricular dysfunction, highlighting the multifaceted nature of the patient's cardiac pathology.

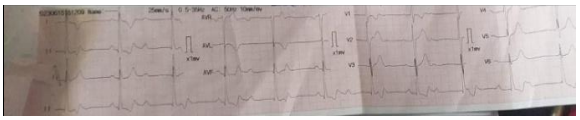

Figure 1: ECG showing complete heart block

Upon echocardiographic examination, CCTGA with mesocardia, mild tricuspid regurgitation, and mildly impaired right ventricular function was established (Figure 2).

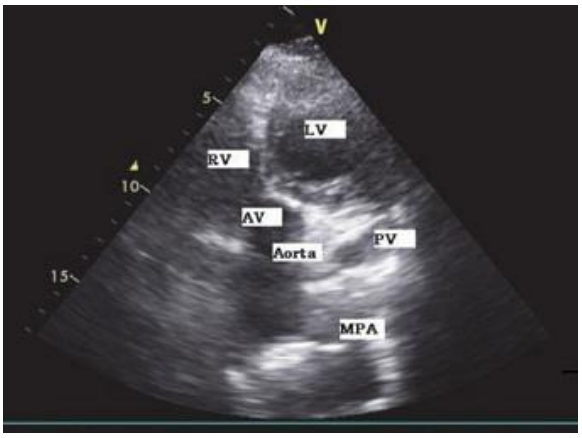

Figure 2: CCTGA on echocardiography depicting the left ventricle (LV), right ventricle (RV), aortic valve (AV), pulmonary valve (PV), and main pulmonary artery (MPA)

Given the presence of high atrioventricular (AV) block, permanent pacemaker (PPM) implantation was deemed necessary. Utilizing the DDD mode was selected to mitigate the risk of systemic ventricular dysfunction. The procedure was conducted via the left subclavian vein under local anesthesia and fluoroscopic guidance. A 58 cm screw-in lead was meticulously positioned in the morphological left ventricular (LV) apex, considering the smooth wall of the morphological LV and the heightened risk of dislodgement. Subsequently, a 52 cm screw-in lead was appropriately placed in the right atrium. Both leads underwent impedance and threshold checks with an analyzer before being connected to the pulse generator. Post-lead positioning verification, the incision was meticulously sutured in layers (Figure 3a, 3b).

The decision to avoid continuous pacing was made to preemptively prevent potential heart failure, given the patient's asymptomatic status with a heart rate of 45 beats per minute. Post-implantation electrocardiography revealed normal pacing in DDD mode. Favorable outcomes were observed at the 6-month follow-up, with no discernible deterioration in ventricular function.

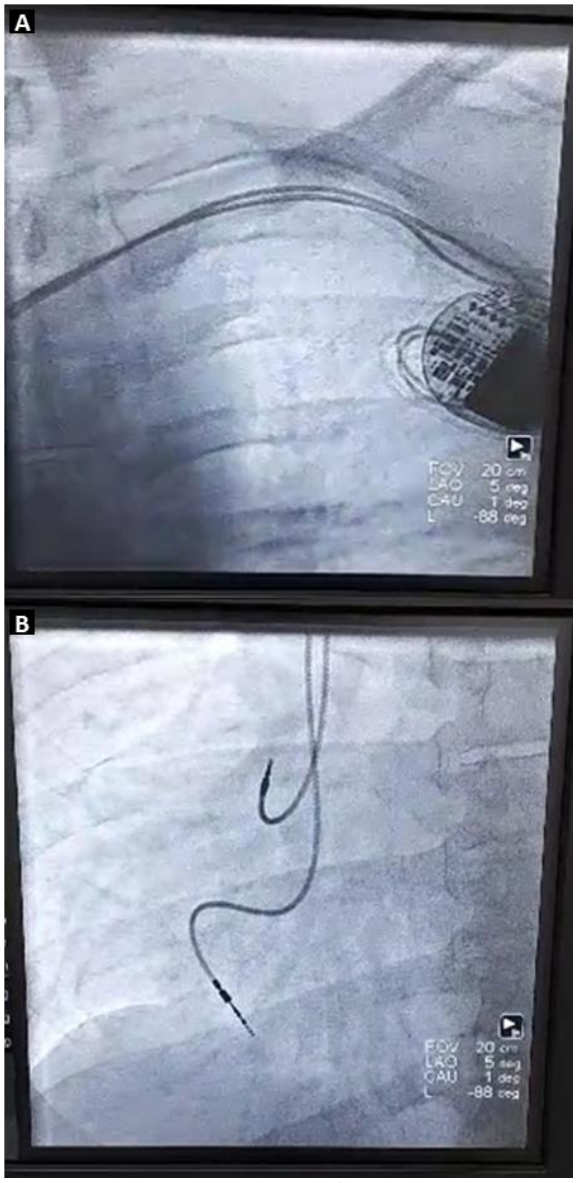

Figure 3: Pulse generator successfully implanted (A) and pacemaker leads in the right atrium (RA) and right ventricle (RV) (B)

Timeline and Diagnostic Assessment: Over the course of the past two years, the patient experienced recurring episodes of dizziness and syncope, prompting medical evaluation. A critical electrocardiographic finding of complete heart block emerged during diagnostic assessment, shedding light on the underlying cardiac conduction abnormality contributing to the patient's symptoms. Subsequent echocardiography played a pivotal role in confirming the diagnosis of CCTGA, providing comprehensive visualization of the complex cardiac anatomy characteristic of this rare congenital anomaly. Together, these diagnostic modalities facilitated a thorough understanding of the patient's clinical presentation, guiding tailored management strategies aimed at addressing both the structural and functional aspects of the underlying cardiac pathology.

Therapeutic Intervention: In response to the diagnosed complete heart block, the patient underwent a critical therapeutic intervention involving the implantation of a dual-chamber (DDD) pacemaker. This procedure was meticulously executed to address the impaired atrioventricular conduction observed in the setting of CCTGA. Careful consideration was given to the unique anatomical variations inherent in CCTGA, ensuring precise placement of the pacing leads to optimize therapeutic efficacy and minimize the risk of complications.

Follow-up and Outcomes: Subsequent to the pacemaker implantation, the patient underwent regular follow-up assessments to monitor therapeutic response and overall cardiovascular health. Encouragingly, at the 6-month follow-up milestone, the patient demonstrated sustained clinical improvement, remaining asymptomatic without the recurrence of dizziness or syncope. Furthermore, comprehensive evaluation revealed no evidence of deterioration in ventricular function, affirming the effectiveness of the therapeutic intervention in mitigating the risks associated with complete heart block and preserving cardiac performance.

DISCUSSION

This case highlights the imperative of maintaining a high index of suspicion for rare cardiac anomalies such as CCTGA when confronted with patients presenting with seemingly unrelated symptoms like dizziness and syncope. Despite the typical asymptomatic nature of CCTGA, this case elucidates the potential for incidental discovery of significant cardiac pathology underlying seemingly disparate clinical presentations.

In the context of CCTGA, the absence of associated anomalies often confers a relatively benign clinical course. However, as evidenced in this case, unforeseen complications may arise, necessitating vigilant monitoring and timely intervention. While the patient's initial symptoms were not directly attributed to CCTGA, further diagnostic workup revealed a constellation of findings including mesocardia, mild tricuspid valve regurgitation, mild right ventricular dysfunction, reduced heart rate, and third-degree atrioventricular (AV) block.

Notably, the patient's symptoms of dizziness and syncope were ultimately attributed to the identified AV block rather than tricuspid regurgitation, which is

commonly associated with CCTGA.[7] This underscores the importance of a comprehensive diagnostic approach to elucidate the underlying etiology of presenting symptoms, thereby guiding appropriate therapeutic interventions.

Furthermore, the development of AV block in CCTGA is a recognized complication, attributed to factors such as AV node dislocation and aberrant conduction tissue anatomy. This highlights the inherent complexity of managing cardiac arrhythmias in the setting of congenital heart disease, necessitating tailored therapeutic strategies to address the unique anatomical and physiological challenges posed by CCTGA.

While cardiac complications in CCTGA can be dire, ranging from systolic dysfunction to acute myocardial infarction, timely intervention offers the potential for improved patient outcomes. In this case, the successful implantation of a dual-chamber pacemaker effectively managed the AV block, mitigating the risk of adverse cardiac events and preserving ventricular function.[8] Notably, the absence of complications such as worsening systemic ventricular function or AV valve regurgitation underscores the importance of meticulous procedural planning and execution in optimizing patient outcomes.[9]

Informed Consent: Informed consent was obtained from the patient for the diagnostic workup and therapeutic intervention.

CONCLUSION

This case presents a unique clinical scenario wherein a patient with CCTGA remained asymptomatic for over three decades before being incidentally diagnosed with AV block.[10] The fortuitous identification of AV block underscores the importance of thorough cardiac evaluation in patients presenting with seemingly unrelated symptoms. One of the key takeaways from this case is the importance of meticulous attention to detail during therapeutic interventions, such as the placement of screw-in leads in CCTGA patients. Given the anatomical complexities inherent in CCTGA, particularly the reversal of ventricular positioning, careful consideration must be given to lead placement to optimize procedural success and minimize the risk of complications.

AUTHORS' CONTRIBUTION

RZ and ZAK: Concept and design, data acquisition, interpretation, drafting, final approval, and agree to be accountable for all aspects of the work. RK, ZAK, Hu, TM, AL, and MS: Data acquisition, interpretation, drafting, final approval and agree to be accountable for all aspects of the work.

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