

Hammock Bridge on Fire: Complete AV Block in a Patient with Congenitally-corrected Transposition of the Great Arteries and Wolff-Parkinson-White Pre-excitation Pattern

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Authors' contributions

Both authors designed, analyzed and interpreted and prepared the manuscript.

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Case Study

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ABSTRACT

Aims: To recognize heart block as a complication associated with congenitally-corrected transposition of the great arteries (CCTGA).

Presentation of the Case: A healthy 36 year old male with CCTGA presented with syncope as a manifestation of heart block. A unique feature of this case was the presence of an accessory pathway that served as the atrio-ventricular conducting structure for more than 3 decades.

Discussion: rationale for a cardiac resynchronization device (CRT) as opposed to a simple pacemaker system is emphasized in this case.

Conclusion: clinicians must be aware of heart block a frequent complication of CCTGA. CRT appears as a more favorable option than a single systemic ventricular pacemaker in such patients.

Keywords: *Congenitally-corrected transposition; complete heart block; wolff-parkinson-white; cardiac resynchronization therapy.*

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1. INTRODUCTION

Congenital heart disease is the most common group of birth defects. Advances in detection, medical and surgical treatment have allowed infants and children with inborn cardiac structural defects to reach adulthood. As a consequence, physicians face the challenge of taking care of a growing number of survivors, and thus, must be familiar with the long-term complications of congenital heart disease, whether related or not to prior surgical interventions. We discuss the unusual case of a patient with congenitally-corrected transposition of the great arteries associated with an electrical accessory pathway, presenting with a relatively common complication of this congenital abnormality: complete heart block.

2. CASE PRESENTATION

A 36-year old active male with a history of congenitally-corrected transposition of the great arteries (CCTGA), situs solitus type (normal position of thoraco-abdominal structures) presented with complaints of weakness, fatigue and an episode of syncope. He was known to have a Wolff-Parkinson-White (WPW) pattern without evidence of spontaneous supra ventricular dysrhythmias (Fig. 1).

At an outside institution, he had undergone an EP study a decade prior which revealed no

antegrade conduction through the AV node, as impulses travelled through a left postero-septal accessory pathway. Additionally, he had a long-standing non-ischemic cardiomyopathy on maximum medical therapy, with an estimated EF of 25% for his systemic ventricle. Initial assessment revealed an athletic gentleman in no acute distress, with a blood pressure of 90/60 mmHg and a pulse of 40 beats per minute. Cardiovascular examination was significant for prominent, intermittent venous pulsations of the neck, variable S1 and a 2/6 systolic ejection murmur best heard at the left lower sternal border. A 12-lead ECG at the office is shown in Fig. 2.

3. INTERPRETATION OF ECG FINDINGS

His baseline ECG (Fig. 1) indicates sinus bradycardia at a rate of 55 bpm, a wide QRS complex with slurred-onset (delta wave) and a short PR interval consistent with a pre-excitation pattern of WPW. In contrast, his ECG on presentation (Fig. 2) shows atrio-ventricular dissociation, with an atrial rate of 68 bpm and a junctional escape rhythm at lower rate of 39 bpm, consistent with complete heart block. Occasional premature atrial complexes (third and last QRS complexes) are conducted through the failing accessory pathway. The junctional escape complexes utilize the His-Purkinje system and not the bypass tract, resulting in narrow complexes that lack the delta wave.

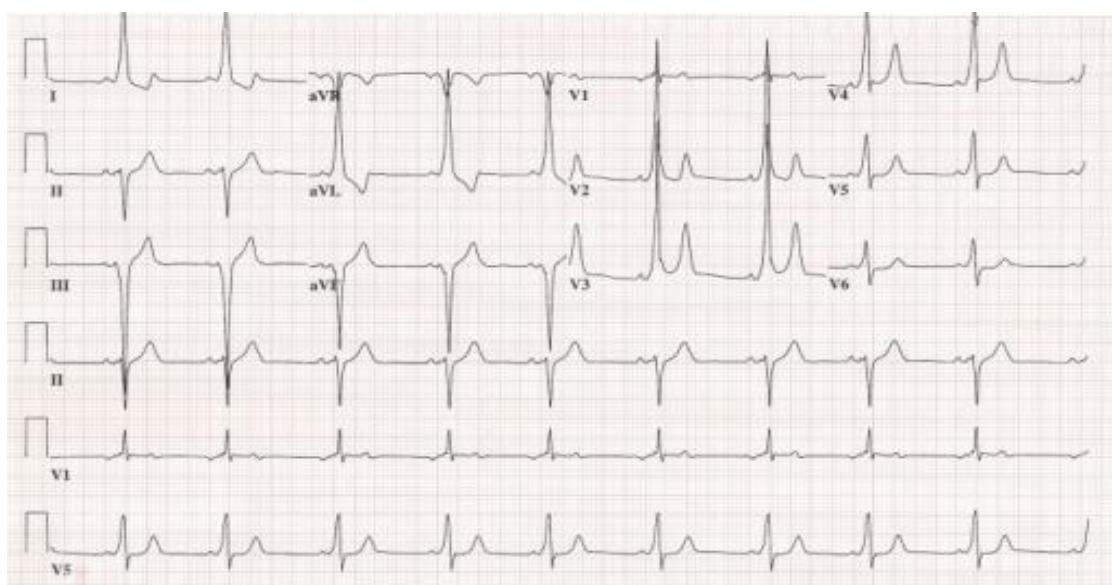


Fig. 1. Baseline electrocardiogram with normal sinus rhythm prior to presentation

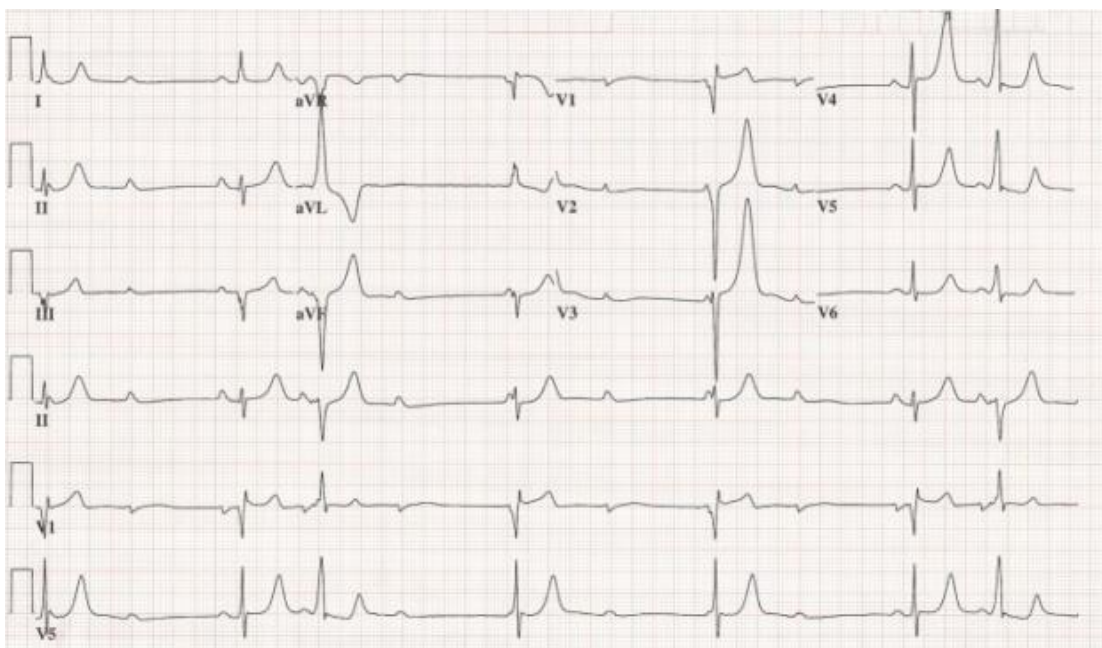


Fig. 2. Electrocardiogram on presentation, displaying complete AV dissociation, consistent with complete heart block

4. CLINICAL COURSE

Dual chamber pacemaker was clearly indicated, but decision making regarding other components of his device was more challenging. Given his depressed systemic ventricular function on adequate medical therapy, as well as impending high frequency of ventricular pacing, a cardiac resynchronization (CRT) system was highly recommended to the patient. A typical CRT system includes a left ventricular lead that is placed into a coronary sinus branch vein to pace the LV epicardially in addition to the standard right atrial and right ventricular leads. While use of CRT systems in CCTGA has been described, the diminutive venous system of the morphologic right ventricle can make LV systemic lead placement difficult [1]. Prophylactic defibrillator implantation was also offered to the patient, but he declined. He underwent placement of a standard CRT-P, with RA, RV and LV leads with no peri-procedural complications. During the procedure, there was evidence of intermittent retrograde conduction through the accessory pathway, but no pacemaker-induced tachycardia could be elicited, thus, ablation of the accessory pathway was deferred. Repeat echocardiogram at 3 months indicated an improvement in ejection fraction to 35%, and at 9-months of follow up, he remained asymptomatic, with normal device function and no dysrhythmias noted.

5. DISCUSSION

In the 1950's, only 1 out of 5 children with congenital heart disease (CHD) was expected to survive through infancy; medical advances have been able to reverse those statistics, and nowadays only 1 out of those 5 children will not reach adulthood [2]. Familiarity of health-care providers with long-term cardiovascular complications of individuals with CHD becomes crucial in this context.

CCTGA is relatively rare, and represents less than 1% of all cases of congenital heart disease [3]. In the setting of situs solitus (normal position of thoraco-abdominal organs), the left ventricle lies to the right and the right ventricle to the left. The mitral valve then lies to the right and the tricuspid valve to the left. The left ventricle is usually slightly posterior or inferior to the right ventricle. In addition to the atrio-ventricular and ventriculo-arterial discordance (Fig. 3), patients may have other associated anomalies such as a ventricular septal defect, pulmonary stenosis or abnormalities of the systemic (tricuspid) valve, including the presence of a supra-valvular left atrial ring. Furthermore, the position of the AV node and His bundle in these patients is abnormal, with a more anterior orientation that makes it prone to fibrosis and degeneration [2].

It is estimated that at least 13% of individuals with CCTGA will have developed complete AV block at the time of diagnosis [4], and roughly 30% on long-term follow-up [5], at a rate of 2% per year [3]. Although complete heart block is thus a well-recognized long-term complication in patients with CCTGA, to the best of our knowledge this is the first case description of a concomitant accessory pathway, which served as the atrio-ventricular conducting structure for almost 4 decades, given the presence of a non-functional native AV node, but eventually this tenuous system collapsed.

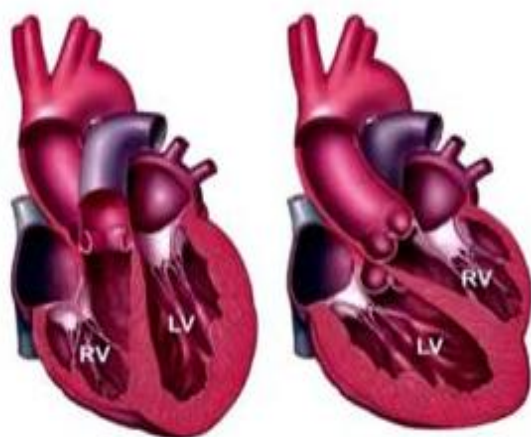


Fig. 3. Comparison of normal cardiac anatomy (left) to congenitally-corrected transposition of the great arteries (right). RV= right ventricle, LV= left ventricle

The clinical manifestations of complete heart block can be quite variable. In a classic clinical series of 251 patients [6], Samuel Levine and colleagues described that palpitations or self-perceived bradycardia were common complaints, as well as weakness, extreme fatigue and dizziness. Syncope was recognized as the gravest presentation in terms of morbidity and mortality. These authors described that the most useful physical finding, almost pathognomonic, is the changing intensity of the first heart sound in the setting of marked, regular bradycardia.

The noxious effects of isolated right ventricular pacing have been well established in structurally normal hearts with complete AV block: dyssynchronous contraction, increased oxygen consumption and adverse remodeling amongst others [7]. Patients with CHD and a systemic right ventricle have a limited ability to cope with dyssynchrony, due to inherent structural

abnormalities, lack of torsion and presence of fibrotic areas [8]. Therefore, a high burden of non-systemic ventricular pacing can have catastrophic consequences in these patients, including development of congestive heart failure [3]. Long-term pacing of the non-systemic ventricle in patients with atrial switch for TGA has been associated with significantly impaired functional status, exercise capacity and overall systemic ventricular performance [9].

Thus, we strongly recommended implantation of a cardiac resynchronization therapy (CRT) device instead of a standard pacemaker. From a pathophysiologic standpoint, this approach appears prudent, as ventricular dyssynchrony appears to be a major mechanism for systemic ventricular failure in patients with CHD; it is estimated that more than 25% of patients with CHD will eventually progress to symptomatic heart failure [10]. Based on the landmark CRT trials, about 10% of patients with a systemic RV would qualify for such a device according to commonly accepted criteria [11]. In a limited retrospective series of 7 patients with TGA, dyssynchrony and systemic ventricular dysfunction, CRT improved both NYHA functional class and peak oxygen consumption [12].

Our patient declined implantation of an ICD lead as part of the CRT system. Indeed, our ability to predict sudden cardiac death in patients with TGA is limited [13], and the evidence to support implantation of an ICD in patients with CHD is rather scarce. In a single-center experience from the Mayo Clinic [14], which included 73 patients with complex CHD, more than one-quarter of the devices were placed in patients with TGA; for all patients, the rate of appropriate shocks was 19%. However, ICD implantation in patients with CHD can be associated with considerable morbidity, including high rates of inappropriate shocks (30%) and procedure-related complications [15].

6. CONCLUSIONS

Physicians should be aware of the long-term complications of CHD in adult survivors. A significant proportion of patients with CCTGA will develop complete heart block at some stage in their lives. Syncope (Stokes-Adams syndrome) is one of the various manifestations of complete heart block, but weakness, fatigue and palpitations are also described by patients. Isolated pacing of the non-systemic ventricle is

associated with adverse long-term consequences; CRT appears to be a more attractive option than mono-ventricular non-systemic pacing in patients with CCTGA and heart block, particularly those with ventricular dysfunction at baseline. There is conflicting and limited data at the present time about the risks and benefits of ICDs in patients with CHD.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Bottega NA, Kapa S, Edwards WD, Conolly HM, Munger TM, Warnes CA, et al. The cardiac veins in congenitally-corrected transposition of the great arteries: delivery options for cardiac devices. *Heart Rhythm*. 2009;6(10):1450-1456.
2. Reid GJ, Webb GD, Barzel M, McCrindle BW, Irvine MJ, Siu SC. Estimates and life expectancy by adolescents and young adults with congenital heart disease. *J Am Coll Cardiol*. 2006;48(2):349-355.
3. Warnes CA. Transposition of the great arteries. *Circulation*. 2006;114:2699-2709.
4. Kafali G, Elsharshari H, Ozer S, Celiker A, Ozme S, Demircin M. Incidence of dysrhythmias in congenitally corrected transposition of the great arteries. *Turk J Pediatr*. 2002;44(3):219-223.
5. Daliento L, Corrado D, Buja G, John N, Nava A, Thiene G. Rhythm and conduction disturbances in isolated, congenitally corrected transposition of the great arteries. *Am J Cardiol*. 1986;58(3):314-318.
6. Penton GB, Miller H, Levine SA. Some clinical features of complete heart block. *Circulation*. 1956;13(6):801-824.
7. Tse HF, Lau CP. Long-term effect of right ventricular pacing on myocardial perfusion and function. *J Am Coll Cardiol*. 1997;29(4):744-749.
8. Giardini A, Lovato L, Donti A, Formigari R, Oppido G, Gargiulo G, Picchio FM, Fattori R. Relation between right ventricular structural alterations and markers of adverse clinical outcome in adults with systemic right ventricle and either congenitally complete or congenitally corrected transposition of the great arteries. *Am J Cardiol*. 2006;98:1277-1282.
9. Horovitz A, De Guillebon M, Van Geldorp IE, Bordachar P, Roubertie F, Iriart X, et al. Effects of Nonsystemic ventricular pacing in patients with transposition of the great arteries and atrial redirection. *J Cardiovasc -Electrophys*. 2012;23(7):766-770.
10. Piran S, Veldtman G, Siu S, Webb GD, Liu P. Heart failure and ventricular dysfunction in patients with single or systemic ventricles. *Circulation*. 2002;105:1189-1194.
11. Diller GP, Okonko D, Uebing A, Ho SY, Gatzoulis MA. Cardiac resynchronization for adult congenital heart disease patients with a systemic right ventricle: analysis of feasibility and review of early experience. *Europace*. 2006;8:267-272.
12. Jauvert G, Rousseau-Paziaud J, Villain E, Iserin L, Hidden-Lucet F, Ladouceur M, Sidi D. Effects of cardiac resynchronization therapy on echocardiographic indices, functional capacity and clinical outcomes of patients with a systemic right ventricle. *Europace*. 2009;11:184-190.
13. Wheeler M, Grigg L, Zentner D. Can we predict sudden cardiac death in long-term survivors of atrial switch surgery for transposition of the great arteries? *Congenit Heart Dis*. 2013;9(4):326-332.
14. Khanna AD, Warnes CA, Phillips SD, Lin G, Brady P. Single-center experience with implantable cardioverter-defibrillators in adults with complex congenital heart disease. *Am J Cardiol*. 2011;108:729-734.

15. Koyak Z, De Groot JR, Van Gelder IC, Bouma BJ, Van Dessel P, Budts W, et al. Implantable cardioverter defibrillator therapy in adults with congenital heart disease: Who is at risk of shocks? *Circ Arrhythm Electrophysiol.* 2012;5:101-110.

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