Review Article

Congenital Complete Heart Block in Adults

Abstract:

Complete heart block occurs when atrial and ventricular contractions are not communicating, each beating, at their own pace. Therefore, it should be thoroughly investigated to ascertain its type. Congenital complete heart block (CCHB) might be unnoticed for a long time. CHB patients are prone to decreased perfusion related to symptoms of bradycardia and low cardiac output, which can lead to serious arrhythmias such as ventricular tachycardia, syncope, and sudden death. We searched PubMed and Google Scholar for CCHB in adults, implications, and outcomes. Accordingly further management of CHB is achieved by the implantation of a cardiac pacemaker, but it might be a challenging decision, particularly in asymptomatic patients.

Keywords; Congenital complete heart block, AV dissociation, complete heart block, diastolic murmur

Introduction:

Complete heart block (CHB) and atrioventricular (AV) block is defined as an electrical disturbance in the transmission of impulses from the atria to the ventricles [(1)]. CHB occurs in 1 out of 15000 to 20000 live births and is either congenital or acquired [(2)]. Congenital CHB pathophysiology is associated with the transfer of maternal autoantibodies, anti-La/SSB (Sjögren syndrome-related antigen B), and anti-Ro/SSA (Sjögren syndrome-related antigen A) through the placenta. These autoantibodies bind to L-type calcium channels on the

cardiomyocytes and inhibit the currents upon entering the fetus's circulation. As a result, inflammation, calcification, and fibrosis occur in a structurally normal heart's atrioventricular (AV) node, impeding signal conduction [3]. Acquired CHB develops during the lifetime due to a specific primary cause, including infections, cardiac ischemia or myopathies, and electrolyte imbalance mainly due to hyponatremia [4,5]. Temporary or permanent pacemakers should be considered individually, depending on the clinical status, investigations, and long-term prognostic concerns.

Congenital complete heart block;

"CHB occurs when atrial and ventricular contractions are not communicating, each beating at their own Pace. CHB may be intra-Hisian or infra Hisian; intra-Hisian blocks mostly feature escape rhythms with narrow QRS complexes; meanwhile, infra-Hisian blocks often present with broad QRS complex escapes" [6,7]. "Patients with CHB are vulnerable to decreased perfusion related to symptomatic bradycardia and decreased cardiac output, resulting in serious arrhythmias like ventricular tachycardia, syncope, and sudden death. Patients are usually asymptomatic and respond to physical exertion or atropine" [9]. Congenital complete heart block (CCHB) was first recognized in 1846 [9] and documented into two categories: congenitally malformed and otherwise anatomically normal hearts [10]. "Secondary CHB might be caused by infections, cardiac ischemia or myopathies, autoimmune diseases, or endocrinological diseases that require extensive workup to be ruled out" [11]. The acquired is mainly seen after 50 years of age [2]. While CCHB may remain undetected and may be discovered during pregnancy [12]."One of the most typical complications of isolated CCHB is a progressive enlargement of the left ventricle leading to dilated cardiomyopathy even in asymptomatic patients. In a review of a multicenter retrospective study of 149 patients with CCHB, pacemaker (PM) therapy may result in decreased stress on the left ventricular over time and may benefit hemodynamically. In the same study, most patients who received PM had reduced their heart size during their follow-ups with echocardiography" [13] "Acquired diastolic mitral regurgitation (MR) is seen with AV dissociation. In patients with sinus rhythm and AV block, prolongation of the PR interval reverses the pressure gradient between the LV and left atrium, leading to an early partial closure of the mitral valve in diastole, then atrial contraction after a non-conducted P wave opens the Mitral valve/ resulting in MR during diastole" [5,14].

Table 1

Indications for PPM in Congenital CHB

Class I	Class II
1. In adults with a congenital complete atrioventricular block with any symptomatic bradycardia, a wide QRS escape rhythm, mean daytime heart rate below 50 bpm, complex ventricular ectopy, or ventricular dysfunction, permanent pacing is recommended. 2. In adults with adult congenital heart disease (ACHD) and symptomatic SND or chronotropic incompetence, atrial based permanent pacing is recommended.	1. In asymptomatic adults with congenital complete atrioventricular block, permanent pacing is reasonable. 2. In adults with repaired ACHD who require permanent pacing for bradycardic indications, a bradycardia device with atrial anti-tachycardia pacing capabilities is reasonable. 3. In adults with ACHD with preexisting sinus node and/or atrioventricular conduction disease who are undergoing cardiac surgery, intraoperative placement of epicardial permanent pacing leads is reasonable.

"It would be desirable to point out signs predicting an increased risk in an individual patient. Low ventricular rate (VR), less than 40 bpm in the young and less than 35 bpm in the elderly, prolongation of the QT time, the appearance of frequent ectopies, and low VR during heavy work have been reported as indicators of PM" [15,16,17]. The American Heart Association's latest guidelines for PM treatment include similar advice (Table 1) [18]. Temporary PM is necessary only for those whose heart rate does not increase during the exercise test [19,20]. Khardke et al. proposed "temporary pacing in patients with atropine-resistant bradycardia, first- and second-degree AV block, and atrial fibrillation with low VR" [21]. Most asymptomatic CCHB patients will eventually become symptomatic and require PM treatment [22,23]. The primary concern is when is the optimal time to implant a PM for patients who do not fit the criteria outlined above because PM implant has its own set of risks, including thrombosis, lead fractures, and other problems that can occur in up to 25% of instances [24]. The question has remained unanswered to this day. It's difficult to foresee a CCHB becoming a lower-degree block or a sinus rhythm. [16,17].

Conclusion:

Adults with a congenital complete atrioventricular block (CCHB) typically have a favorable prognosis. Monitoring ectopics, mitral insufficiency, a long QTc interval, and widened QRS complexes all are sound reasons to consider Pacemaker treatment. If pacemaker implantation is declined, an annual evaluation with Holter monitoring, exercise testing, and echocardiogram is encouraged.

Competing interest.

Authors have declared that no competing interests exist.

Reference:

1. Kiblawi M, Naeem A, Al Attrash E, Kar S, Goud BK. Complete congenital heart block in a newborn associated with maternal systemic lupus erythematosus: a case report. *Int J Med Students*. 2013;1(3):128–31.
2. Perloff JK. The clinical recognition of congenital heart disease. 6. Philadelphia: Elsevier; 2003.
3. 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. doi:10.1007/s00431-016-2748-0
4. Badrinath AK, Suresh K, Ragunathan R, Babu SS. A case report of complete atrioventricular heart block due to hyponatremia. *Heart India*. 2017;5:105-7.
5. Levy MN, Edelstein J. The mechanism of synchronization in isorhythmic A-V dissociation. II. Clinical studies. *Circulation*. 1970;42:689-699.
6. Narula OS, Scherlag BJ, Javier RP, Hildner FJ, Samet P. Analysis of the A-V conduction defect incomplete heart block utilizing His bundle electrograms. Circulation. 1970;41(3):437-448.
7. Narula OS, Scherlag BJ, Samet P, Javier RP. Atrioventricular block. Localization and classification by His bundle recordings. *Am J Med.* 1971;50(2):146-165. doi:10.1016/0002-9343(71)90144-6
8. Mark EJ. Clinical Cardiac Electrophysiology: Techniques and Interpretations. Philadelphia, PA: Wolters Kluwer/Lippincott Williams & Wilkins; 2008:114-144.
9. Stokes W. Observations on some cases of permanently slow pulse. *Dublin Quarterly Journal of Medical Science*. 2014;2(1):73-85.
10. Carter JB, Blieden LC, Edwards JE. Congenital heart block: anatomic correlations and review of the literature. Arch Pathol 1974; 97:51-5
11. Buyon JP, Hiebert R, Copel J, et al. Autoimmune-associated congenital heart block: demographics, mortality, morbidity and recurrence rates obtained from a national neonatal lupus registry. *J Am Coll Cardiol*. 1998;31:1658-1666.
12. AlTaweel M, AlMukhaylid S, Alsultan N, et al. Intrapartum Asymptomatic Congenital Complete Heart Block; Case Study and Literature Review. *Innovative Journal of Medical and Health Sciences.* 2022;12(06):1935-1940. doi:10.15520/ijmhs.v12i06.3450

13. Michaëlsson M, Jonzon A, Riesenfeld T. Isolated congenital complete atrioventricular block in adult life. A prospective study. *Circulation*. 1995;92:442-449.
14. Kocabay G, Peluso D, Muraru D, Iliceto S, Badano LP. Diastolic mitral regurgitation in 2:1 atrioventricular block: insight of the diastolic pressure. Echocardiography 2013;30(2):E51-2
15. Smith RT. Pacemakers for children. In: Gilette PC, Garson A Jr, eds. Pediatric Arrhythmias: Electrophysiology and Pacing. Philadelphia, Pa: Saunders; 1990:532-558."
16. Levy AM, Camm AJ, Keane JF. Multiple arrhythmias detected during nocturnal monitoring in patients with congenital complete heart block. *Circulation*. 1977;55(2):247-253. doi:10.1161/01.cir.55.2.247
17. Esscher E, Michaëlsson M. Q-T interval in congenital complete heart block. *Pediatr Cardiol*. 1983;4(2):121-124. doi:10.1007/BF02076336
18. Kusumoto FM, Schoenfeld MH, Barrett C, et al. 2018 ACC/AHA/HRS Guideline on the Evaluation and Management of Patients With Bradycardia and Cardiac Conduction Delay: Executive Summary: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines, and the Heart Rhythm Society [published correction appears in J Am Coll Cardiol. 2019 Aug 20;74(7):1014-1016]. *J Am Coll Cardiol*. 2019;74(7):932-987. doi:10.1016/j.jacc.2018.10.043
19. Keepanasseril A, Maurya DK, Suriya Y, Selvaraj R. Complete atrioventricular block in pregnancy: report of seven pregnancies in a patient without pacemaker. *BMJ Case Rep*. 2015;2015:bcr2014208618. Published 2015 Mar 9. doi:10.1136/bcr-2014-208618
20. Kivrak T, Kivrak V, Kivrak YY, Karaca I. Presenting of pregnant woman with atrioventricular block. *SM J Case Rep.* 2017;3(5):1058.
21. Kharde VV, Patil VV, Dhulkhed VK, Divekar DS. A parturient with complete heart block for cesarean section. *J Anaesth Clin Pharmacol*. 2010 Jul 1;26(3):401-2.
22. Jaeggi ET, Hamilton RM, Silverman ED, Zamora SA, Hornberger LK. Outcome of children with fetal, neonatal or childhood diagnosis of isolated congenital atrioventricular block. A single institution's experience of 30 years. *J Am Coll Cardiol*. 2002;39:130-137.
23. Sholler GF, Walsh EP. Congenital complete heart block in patients without anatomic cardiac defects. Am Hear J. 1989;118:1193-1198

24. Tsuji A, Yanai J, Komai T, Sato M, Saishi T, Fukuda T. Recovery from congenital complete atrioventricular block. *Pediatr Cardiol*. 1988;9:163-166.