

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. Managing 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.

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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.

Ethical Considerations

Ethical approval: The study was approved by King Abdullah International Medical Research Center (KAIMRC)

Consent: Written informed consent was obtained from the patient for publication.

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