Congenital Complete Heart Block in Young Women

Triwedya Indra Dewi, Giky Karwiky, Rekha Nova Iyos, Mega Febrianora

Department of Cardiology and Vascular Medicine, Faculty of Medicine Universitas Padjadjaran-Dr. Hasan Sadikin General Hospital Bandung, Indonesia

pISSN: 2302-1381;
eISSN: 2338-4506;
http://doi.org/10.15850/ijihs.v9n1.2142
IJIHS. 2021;9(1):42–48

Received:
September 21, 2020

Accepted:
March 30, 2021

Abstract

Objectives: To present a rare case of Congenital Complete Heart Block (CCHB) in the setting of post-cesarean delivery of an asymptomatic young patient.

Methods: A 30-year-old female patient complained of sudden weakness after C-section delivery with spinal anesthesia. She presented a slow heart rate and Complete Heart Block (CHB) on electrocardiogram (ECG). After one week of observation, the ECG still presented a CHB condition. A permanent pacemaker (PPM) with DDDR mode was then installed for this patient.

Result: The etiology of CHB, especially at a young age, is unclear, hence challenging. A patient with a CCHB is difficult to diagnose, especially without any previously related symptoms. This abnormality is usually detected during routine screening not related to cardiovascular disease. The patient in this case study presented an ECG of persistent CHB from the time this patient was admitted until one week after observation. The echocardiography showed normal results. Other modalities to confirm diagnosis and evaluate the prognosis of a CCHB should be done.

Conclusion: Establishing the etiology of CHB in young patients is challenging. The implantation of PPM is needed because the condition is permanent, regardless the etiology. However, implanting a permanent pacemaker is not always an easy decision, especially in young patients.

Keywords: Complete heart block, congenital, young

Introduction

Heart block is a disturbance of impulse conduction that can be permanent or transient, depending on the anatomic or functional impairment. It must be distinguished from interference, a normal phenomenon that is a disturbance of impulse conduction caused by physiologic refractoriness resulting from inexcitability secondary to a preceding impulse. Interference or block can occur at any site where impulses are conducted, but they are recognized most often between the sinus node and atrium (SA block), between the atria and ventricles (AV block), within the atria (intra-atrial block), or within the ventricles (intraventricular block).[1,2]

Complete heart block can be acquired or congenital. It can occur due to multiple pathological conditions. The etiology of complete heart block, which ensues in the young population, was only identified in approximately half the patients, in whom complications to cardiac surgery or congenital complete heart block were the most common aetiologies. The acquired CHB occurs due to various reasons: medicines, acute myocardial infarction, chronic ischemic heart disease, degenerative diseases, rheumatic diseases, infiltrative processes, neuromyopathy, infectious diseases, iatrogenic, and AV blocks

Correspondence:
Triwedya Indra Dewi,
Department of Cardiology and Vascular Medicine, Faculty of Medicine Universitas Padjadjaran-Dr. Hasan Sadikin General Hospital Bandung, Indonesia
e-mail: adeindradewi@gmail.com

mediated by vagal processes. In some rare cases, spinal anesthesia can cause CHB. Congenital heart block that occurs after spinal and epidural anesthesia may combine several processes, such as drugs, iatrogenic, and vagal.[1,3-7]

The incidence of CCHB varies from 1 in 15,000 to 1 in 22,000 live births.[1] The signs and symptoms of patients with CCHB depend on the baseline ventricular rate and underlying structural defects. Patients may be asymptomatic until cardiac decompensation begins to occur in adulthood.[8] In this case report, the author will discuss etiology of CCHB in a young post partum patient and diffrentiate it from other causes of CHB.

Case

A thirty-year-old female (P1, A0) was referred from a satellite hospital and consulted in the emergency room due to a complete heart block after Caesarean delivery with spinal anesthesia. She complained of progressive generalized fatigue a few hours after Caesarean delivery, so that she was unable to perform a regular activity. There were no accompanying neurological symptoms, heart failure, nor a history of hypertension.

The patient's heart rate was reported to be slow by her husband, with no significant symptoms before the procedure. Spinal anesthesia was performed using Bupivacaine 10 mg and Fentanyl 25 U. The procedure went smooth with 2900 gram healthy baby girl was delivered. Shortly after the delivery, the patient has hypotension and bradycardia. Resting ECG showed a complete heart block with QRS duration 0.08 s and prolonged QTc of 516 ms. Soon thereafter, she was referred to Hasan Sadikin General Hospital for Temporary Percutanues Pacemaker (TPM).

On arrival in the emergency room, the vital signs showed a slow heartbeat of 40 beats per minute and the blood pressure 120/80 mmHg. Her physical examination was unremarkable. The ECG showed CHB with QRS duration 0,08 s and prolonged QTc of 574 ms. Her chest X-ray examination revealed an enlarged heart. Her blood examination showed; hemoglobin 12.3 g/dL, leukocyte 7,200 /mm^3, thrombocyte 342,000, urea 52 mg/dL, creatinine 0.98 mg/dL, sodium 137 mg/dL, potassium 4.2 mg/dL, calcium 4.81 mg/dL, magnesium 2.1 mg/dL. The transthoracic echocardiogram showed normal heart chambers, normal valves, and normal systolic and diastolic functions. There was no LV or RV thickening, no "granular sparking" or "speckling," and aneurysm.

Based on the data above, the patient was diagnosed with a CHB, possibly due to spinal anesthesia. We still investigated the other cause of the CHB. We suspected a congenital involvement as a cause due to the previous history of slow heart rate but undocumented ECG. We then put her on a temporary pacemaker. Afterward, she was transferred to the High Care Cardiac Unit and was monitored intensively for one week. However, the ECG showed persistent CHB,

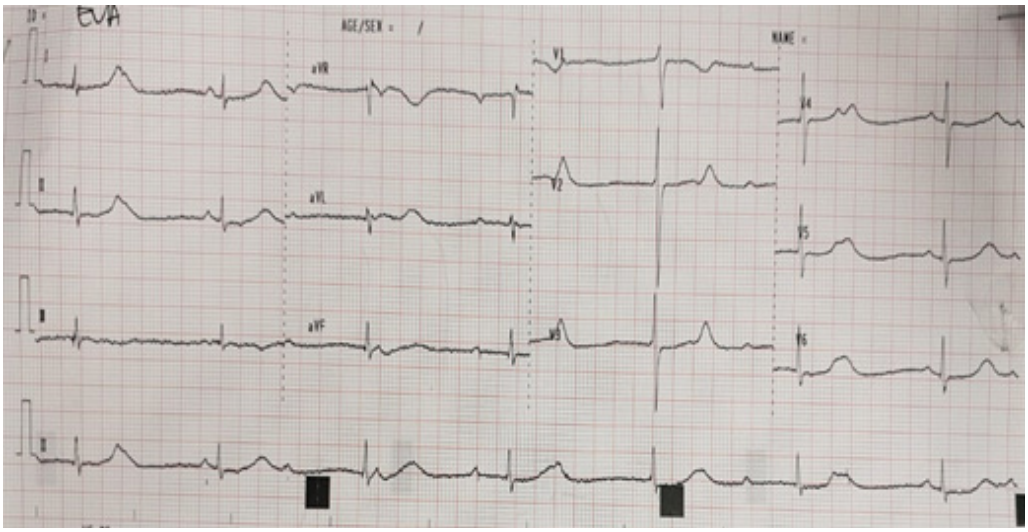

Fig. 1 ECG with Complete Heart Block

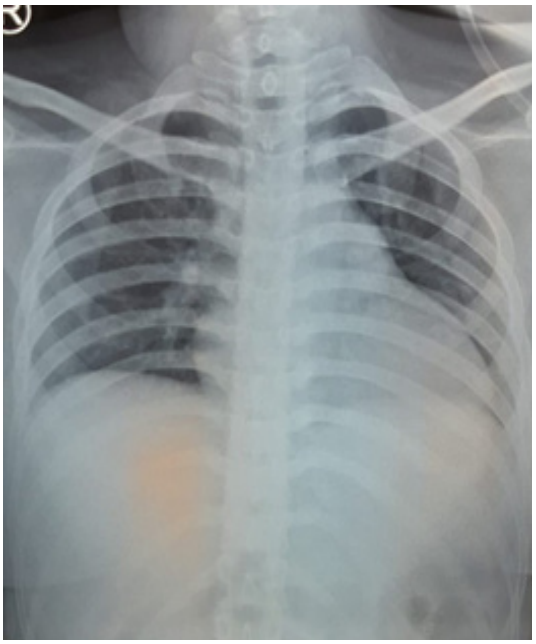

Fig. 2 Chest X-Ray

despite the already diminished effect of the anesthetic drug. Doctors have excluded spinal anesthesia as the etiology and perform workup for other etiology such as congenital or autoimmune. Unfortunately, the patient's mother was not present, so we could not examine immunoassays for SSA/Ro or SSB/La antibodies, but this patient did not have any signs and symptoms of autoimmune disease. Patient heart rate persists below 50 beats per minute and still with prolonged QTc of 515 ms. Based on the recommendation by guideline, we decided to perform a permanent pacemaker (PPM) installation on the patient and programmed with DDDR mode without any complications. The patient had provided informed consent for scientific publication and agreed to be published. This case report was conducted from January 2019 to February 2019.

Discussion

The etiology of CHB was unclear, hence challenging. A complete heart block in this patient was initially suspected of spinal anesthesia complication after the Caesarean delivery procedure. In a case report by Joseph et al.[4], they said that they recorded the systemic symptoms after spinal anesthesia injection after 4 minutes, and a CHB was reported several minutes afterward. The reported time needed for the heart's impulse to return to sinus rhythm after CHB was 4 minutes. Another case report by McHugh *et al*.[5] said that 5 minutes after subarachnoid injection, the non-specific symptoms were noted, and a CHB was revealed. It sustained first-degree heart block for 6 hours after subarachnoid injection, and afterward, the rhythm returned to sinus rhythm.

In this patient, we rejected the possibility of complications of spinal anesthesia as the cause of the heart block because the CHB persisted after one week. In contrast, most CHB caused by spinal anesthesia would return to sinus rhythm after several hours. This patient complained of symptoms of CHB after the Caesarean delivery had finished, where most of the spinal anesthesia complications would be seen several minutes after the spinal anesthesia drug was injected.

The etiologies of CCHB include the following: autoimmune antibodies, structural heart abnormalities due to congenital heart disease (e.g., congenitally corrected transposition of the great arteries, endocardial cushion defects), idiopathic familial CCHB.[6,7,9] CCHB due to the autoimmune disease usually begins in utero, though clinical detection may occasionally be postponed until after birth or during early childhood.[10] In many cases, the block is a complete heart block. The mechanism is due to damage to the development of specialized conduction tissue from passive transplacental passage of maternal autoantibodies to Ro/SSA and/or La/SSB intracellular ribonuclear proteins.[9]

The etiology possibility of CHB in this patient due to congenital cause is getting more likely because the etiology of CHB at a young age, such as cardiomyopathy, infection, infiltrative disease, and ischemic heart disease, had been excluded based on the clinical history and examinations that have been carried out. The patient reported no prior history of complaints. This study found several data that support this condition. Baruteau *et al*.[8] and Bordachar *et al*.[11], said that some people with CHB are asymptomatic with bradycardia and detected only during routine screening. Patients may be asymptomatic until cardiac decompensation begins to occur in adulthood. It raised suspicion of CCHB as the mechanism of the slow heartbeat in this patient.

Postnatally, neonates and young children present in a wide variety of different ways. Some are asymptomatic with bradycardia and detected only during routine screening. Others may present with nonspecific symptoms, such

as poor growth, abnormal tiredness, sleep disturbance, or frequent nightmares. Syncope, heart failure, or sudden death may be the first manifestation of congenital AV block in the absence of previous symptoms and signs of cardiovascular disease.[11]

Congenital CHB is related to congenital heart defects. In various series of fetal CCHB, 30 to 53% of cases have associated congenital heart disease. Various forms of congenital heart disease are related to abnormalities in the development of AV conduction tissue, such as L-looped transposition of the great arteries (L-TGA), Endocardial cushion defects, and syndromes with simple atrial septal defects. Anatomic disruption between the atrial musculature and peripheral parts of the conduction system and nodoventricular discontinuity are two common histologic findings.[2,12]

Idiopathic familial CCHB — Non-immune CHB in patients with a structurally normal heart has also been described as an idiopathic disorder with a strong familial tendency. In a retrospective cohort of 141 children with AV block diagnosed in utero or up to age 15 years (51% female, 84% asymptomatic, 71% with CHB on presentation, with an additional 21% progressing from incomplete to CHB), 112 patients (79%) received permanent pacemakers, most prophylactically in asymptomatic patients (70 of 112 patients [63%]).[13]

There were several modalities that can be utilized to diagnose CCHB. Electrocardiography can be used used to diagnose arrhythmia and the type of heart block. An electrophysiology study can be performed to locate the level block in the conduction system. The treadmill stress test was needed to evaluate functional capacity. Holter monitoring can be conducted to determine how heart rate and rhythm vary with activities of daily living. Immunoassays for SSA/Ro or SSB/La antibodies. Lastly, echocardiography can be used to determine structural problems.[8,11]

Patients with CCHB may undergo routine 12-lead ECG, echocardiography, holter monitoring, electrophysiology studies, and treadmill testing to determine the effects of the disease process. The results of diagnostic tests are not predictive of who will die from CCHB, but the results do indicate risk factors and can be used to determine which patients should receive a pacemaker. Resting heart rate decreases with age in patients with CCHB during infancy and childhood, and heart rates less than 50/min are associated with signs and symptoms and increasing mortality.[14]

In nearly all cases, the diagnosis of complete heart block can be made by obtaining a surface ECG, ideally a full 12-lead ECG, but sometimes a single-lead rhythm strip is adequate if a full 12-lead ECG cannot be obtained. The diagnosis is usually suspected when a slow pulse is detected, and heart block is confirmed by ECG or by ambulatory ECG monitoring.[14,15]

As mentioned in several previous studies, a CCHB could be initially recognized from routine ECG records. In our patient, we suspected having a complete heart block by ECG recorded after the Caesarean delivery procedure, even though there was no previous record of such findings. The recorded ECG of this patient revealed a complete heart block with prolonged QTc of 516 ms and 574 ms. This is in accordance with the conditions described in previous studies that CHB might have prolonged QTc. Seven percent of the patients were identified as having a prolonged QT interval (corrected QT interval 450 ms) at the time of presentation in the study by Michaelsson et al.[14]

In other ECG evaluations, our patient had a narrow QRS complex of 80 ms. The width of the QRS complex is used to infer whether hemodynamically unstable cardiac rhythms may develop. A ventricular rhythm with a wide complex (>120 ms) and a prolonged QTc (>450 ms) in patients with CCHB is an unfavorable prognostic sign because it may be related to underlying myocardial damage.[14] Prolonged QTc is associated with congenital CHB and occurs in approximately 15% to 22% of patients.[16] In multiple studies, a greater percentage of patients with congenital CHB and QTc prolongation greater than 450 ms had signs and symptoms related to the CCHB or suddenly died than did CCHB patients without QTc prolongation.[8] Michaelsson *et al.*[14], described the electrocardiographic characteristics in a large population of patients with congenital AV block; the mean heart rate was 41 beats/min, the heart rate usually decreased as the child grew older, and they observed a broad complex escape rhythm in 10% of the patients.

Two mechanisms have been proposed for the development of QT prolongation in patients with CCHB. First, the conduction disorder and resultant bradycardia may be the initial phenomenon that subsequently leads to the development of altered repolarization. This is supported by data from animal models of chronic complete heart block. Second, patients with congenital AV block who develop QT

prolongation may manifest phenotypic latent congenital long QT syndrome.[11]

In prenatal conditions, echocardiography may be helpful. With fetal echocardiography, a high proportion of autoimmune-mediated complete heart block cases are now identified in utero (in populations where routine fetal echocardiography is performed). The finding may be an incidental finding. Fetal echocardiography remains the standard gold method for diagnosing congenital CHB. The diagnosis is made by establishing atrioventricular dissociation by using M mode or Doppler echocardiographic techniques. Reference values have been defined, and surveillance protocols have been designed to be implemented between 16 and 24 weeks of gestation, the period during which the fetus is at the highest risk of developing complete heart block.[11] The patient had normal echocardiography results without structural abnormalities.

The suspicions of congenital complete because there was no symptom nor chronic HF symptoms and no history of autoimmune disease. This condition might be due to idiopathic familial CCHB. Anti Ro/La negative cases constitute around 30% of all congenital CHB.[17] Fetal conduction tissue injury caused by transplacental exposure to maternal autoantibodies related to autoimmune disease is responsible for 70 to 90 percent of cases CCHB.[15] As stated by Bordachar et al.[11], he family history of autoimmune disease, especially in the patient's mother, was possible in a patient with congenital AV Block. Once heart block has been diagnosed, the mother should be screened for evidence of connective tissue disease. Immunoassays for SSA/Ro or SSB/La antibodies should be performed.[11] Unfortunately, we did not perform any examinations to prove it in this patient.

The incidence of CCHB is 2% in maternal anti-Ro/SSA antibody positivity cases, 3% when both anti-Ro/SSA and anti-La-SSB are positive. Isolated CCHB or CCHB with a structurally normal heart is frequently associated with Ro/SSA and La/SSB maternal autoantibodies. In this series of cases, all the mothers were positive for ANA, SS-A (Ro), antibodies, and SS-B (La) antibodies, except in the first case where SS-B (La) antibodies were negative. Pregnant women whose sera contain anti-Sjögren's syndrome A (SSA)/ Ro antibodies (in the presence or absence of anti-SSB/La antibodies) have a 1–7.5% risk of having a child with third-degree CHB.[18] Thus, in cases of children with CHB, the patient's mother should be examined for anti-Ro / SSA and anti-La-SSB.

Despite the conduction disorder, many patients have a normal exercise treadmill evaluation in terms of performance.[11] Treadmill exercise testing is done mainly to evaluate functional capacity.[8] In patients with CCHB without structural heart disease, up to 90% have normal results in exercise treadmill tests.[14,16] In this case treadmill test was not performed.

This patient has eventually implanted a PPM with DDDR mode. This was following the 2018 ACC/AHA/HRS Guideline on the Evaluation and Management of Patients With Bradycardia and Cardiac Conduction Delay:[19] 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. A similar statement was also mentioned in the 2021 ESC Guidelines on cardiac pacing and cardiac resynchronization therapy:[20] pacing is indicated in a high degree and complete heart block in symptomatic patients and in asymptomatic patients with any of the following risk conditions: ventricular dysfunction, prolonged QTc interval, complex ventricular ectopy, wide QRS escape rhythm, ventricular rate < 50 b.p.m., ventricular pauses > three-fold the cycle length of the underlying rhythm.

A patient with CCHB was hard to detect, especially without any previous related symptoms. This abnormality was usually detected from routine screening or when ECG was performed for other complaints related to the non-cardiovascular disease. Other modalities to confirmed diagnosis and evaluate the prognosis of CCHB could be done.

References

1. Issa ZF, Miller JM, Zipes DP. Clinical arrhythmology and electrophysiology : A companion to braunwald's heart disease. Philadelphia: Elsevier; 2019.
2. Zipes DP, Libby P, Bonow RO, Mann DL, Tomaselli GF, Braunwald E. Braunwald's heart disease : A textbook of cardiovascular medicine 11th ed. Philadelphia: Elsevier; 2018.
3. Jordi EM, Marsch SC, Strebel S. Third degree heart block and asystole associated with spinal anesthesia. Anesthesiology. 1998;89(1):257–60.
4. Joseph SE, Minehart RD. Third-degree heart block during spinal anesthesia for cesarean delivery. A A Case Rep. 2014;3(1):3–5.
5. McHugh SM, Ibinson JW. Development of third-degree heart block due to thoracic epidural anaesthesia. Injury Extra. 2012;43(2):18–20.
6. Rudbeck-Resdal J, Christiansen MK, Johansen JB, Nielsen JC, Bundgaard H, Jensen HK. Aetiologies and temporal trends of atrioventricular block in young patients: A 20-year nationwide study. Europace. 2019;21(11):1710–6.
7. Barra S, Providência R, Paiva L, Nascimento JW, Leitão-Marques A. Pacing Clin Electrophysiol. 2012;35(11):1395–405.
8. Chronister CS. Congenital complete atrioventricular block in a young man: A case study. Crit Care Nurse. 2009;29(5):45–56.
9. Brito-Zeron P, Izmirly PM, Ramos-Casals M, Buyon JP, Khamashta MA. Autoimmune congenital heart block: Complex and unusual situations. Lupus. 2016;25(2):116–28.
10. Eliasson H, Sonesson SE, Sharland G, Granath F, Simpson JM, Carvalho JS, et al. Isolated atrioventricular block in the fetus: A retrospective, multinational, multicenter study of 175 patients. Circulation. 2011;124(18):1919–26.
11. Bordachar P, Zachary W, Ploux S, Labrousse L, Haissaguerre M, Thambo JB. Pathophysiology, clinical course, and management of congenital complete atrioventricular block. Heart Rhythm. 2013;10(5):760–6.
12. Friedman D, Duncanson L, Glickstein J, Buyon J. A review of congenital heart block. Images Paediatr Cardiol. 2003; 5(3): 36–48.
13. Baruteau AE, Fouchard S, Behaghel A, Mabo P, Villain E, Thambo JB, 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(5):622–9.
14. Michaelsson M, Jonzon A, Riesenfeld T. Isolated congenital complete atrioventricular block in adult life. A prospective study. Circulation. 1995;92(3):442–9.
15. 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(1):130–7.
16. Kertesz NJ, Fenrich AL, Friedman RA. Congenital complete atrioventricular block. Texas Heart Institute journal. 1997;24(4):301–7.
17. Mitra S, Saha AK, Sardar SK, Singh AK. Remission of congenital complete heart block without anti-Ro/La antibodies: A case report. Ann Pediatr Cardiol. 2013;6(2):182–4.
18. Kapur A, Dey M, Tangri M, Bandhu HC. Maternal Anti-Ro/SSA and Anti-La/SSB Antibodies and Fetal Congenital Heart Block. J Obstet Gynaecol India. 2015;65(3):193–5. doi:10.1007/s13224-014-0608-2.
19. Kusumoto FM, Schoenfeld MH, Barrett C, Edgerton JR, Ellenbogen KA, Gold MR, 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):932–87.
20. Glikson M, Nielsen JC, Kronborg MB, Michowitz Y, Auricchio A, Barbash IM, et al. 2021 ESC guidelines on cardiac pacing and cardiac resynchronization therapy: Developed by the task force on cardiac pacing and cardiac resynchronization therapy of the european society of

cardiology (ESC) with the special contribution of the european heart rhythm association (EHRA). Eur Heart J. 2021;42(35):3427–520.