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Case Study and Literature Review

Intrapartum Asymptomatic Congenital Complete Heart Block; Case Study and Literature Review

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Abstract
Congenital complete heart block (CCHB) might be unnoticed for a long time. Often, 30 percent are discovered incidentally during pregnancy without any symptoms; patients should be evaluated further to determine the current and long-term care plan.We describe a case of a 42-year-old pregnant woman who developed pre-eclampsia during her third trimester of pregnancy and was later discovered to have CCHB. Throughout her life, she enjoyed a regular life free of symptoms.
Asymptomatic complete heart block (CHB) should be thoroughly investigated to ascertain its type. Temporary or permanent pacemakers should be considered on an individual basis, depending on the clinical status, investigations, and long-term prognostic concerns.
Keywords; Congenital complete heart block, AV dissociation, complete heart block, pregnancy complete heart block, pacemaker

1 | 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 andis 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 the atrioventricular (AV) node of a structurally normal heart, impeding signal conduction[3].CHB may remain undetected and often 30%are discovered during pregnancy [5]. 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[4,14].

CHB requires a multidisciplinary approach, which involves the obstetrician, cardiologist, anesthesiologist, and neonatologist[2,5].A heart pacemaker should be used to manage symptomatic pregnant patients; however, asymptomatic pregnant women are safely managed during labor and delivery without pacing [6].

2 | CASE REPORT

A 42-year-old Gravida 6, Para 5, female in her third trimester (32 weeks) of pregnancy, was admitted from the emergency department to Obstetrics and Gynecology ward for preeclampsia workup. She had one unremarkable antenatal visit with apparently normal vitals and a registered heart rate of 52 bpm. Cardiology consultation was sought based on the telemonitor heart rate.She denied dizziness, fatigue, syncope, or pre-syncope. On physical examination, she was afebrile with a blood pressure of 165/68 mm Hg, heart rate of 46/minute, and oxygen saturation of 100% on ambient air. No cyanosis, jugular venous distension skin rash, or pedal edema.

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Auscultation of the heart revealed a slow regular heart rate with a diastolic murmur. Auscultation of the lungs revealed clear breath sounds. Admission electrocardiogram (ECG) in (Figure 1) showed complete heart block (CHB) characterized by atrioventricular (AV) dissociation with narrow QRS escape rhythm, atrial rate of 90/minute, and ventricular rate(VR) of 45/minute. Chest X-ray was unremarkable. Lab work included a complete blood count, chemistry panel, troponin, erythrocyte sedimentation rate, and thyroid panel all within normal limits. Urine toxicology was negative. Interestingly, serology workup for antinuclear antibody and rheumatoid factor, toxoplasmosis, and rubella were also negative. A transthoracic echocardiogram revealed normal left ventricular systolic and diastolic function with moderate diastolic mitral regurgitation (MR) tricuspid regurgitation (TR), and pulmonary hypertension.A further decision was made to proceed with an emergency caesarian section comprehending the case's complexity. She was anesthetized spinally with bupivacaine and fentanyl, and a temporary pacemaker was not deemed necessary since she maintained sound cardiac output. In the post-surgery follow up the patient remained asymptomatic with CHB and occasionally switched to an apparent 2:1 heart block on the telemonitor. Furthermore, an exercise stress test was performed to assess the patient's chronotropic competency. The patient exercised for 11 minutes, initially 6 minutes on regular Bruce then 5 minutes on a manually adjusted grade reduction, heart rate rose from 45 to 63 bpm only, she achieved a metabolic equivalent of 7.64%, and remained in junctional escape rhythm with CHB (Figure 2) with noPremature ventricular ectopics on exercise nor on recovery. Her resting ECG showed CHB with isorhythmic AV dissociation throughout the exercise and recovery mimicking 2:1 block. Although she was asymptomatic, looking back to her modest stress chronotropic competency and acquired MR/TR; a permanent pacemaker (PM) was advised,however, she declined and continued an activelifestyle, with irregular outpatient cardiology follow up.

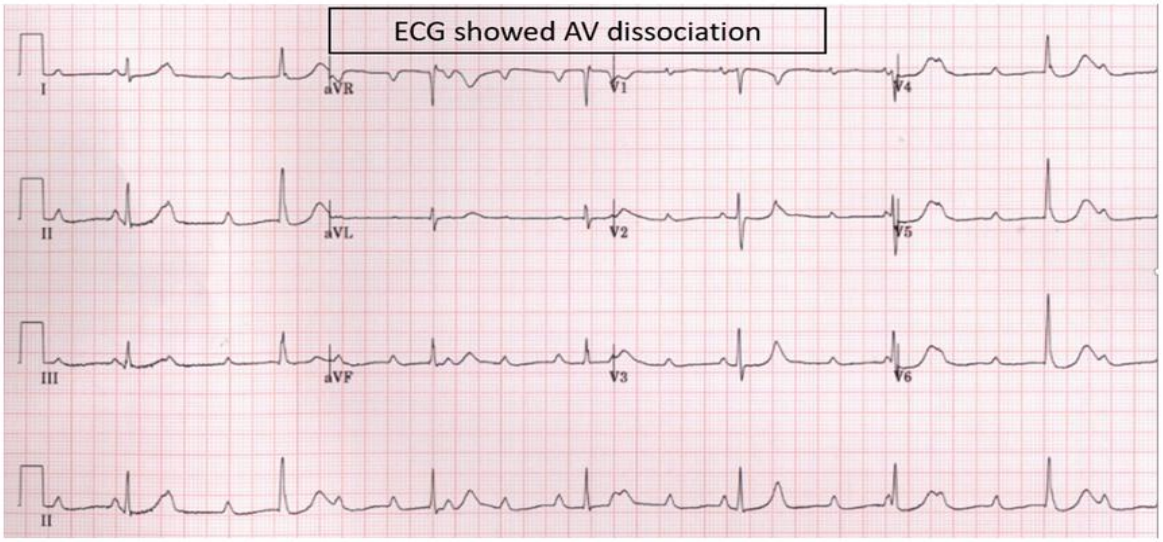

Figure 1. Twelve-lead electrocardiogram on admission showing complete heart block with junctional escape rhythm, atrial rate around 90/minute, and ventricular rate of 45/minute.

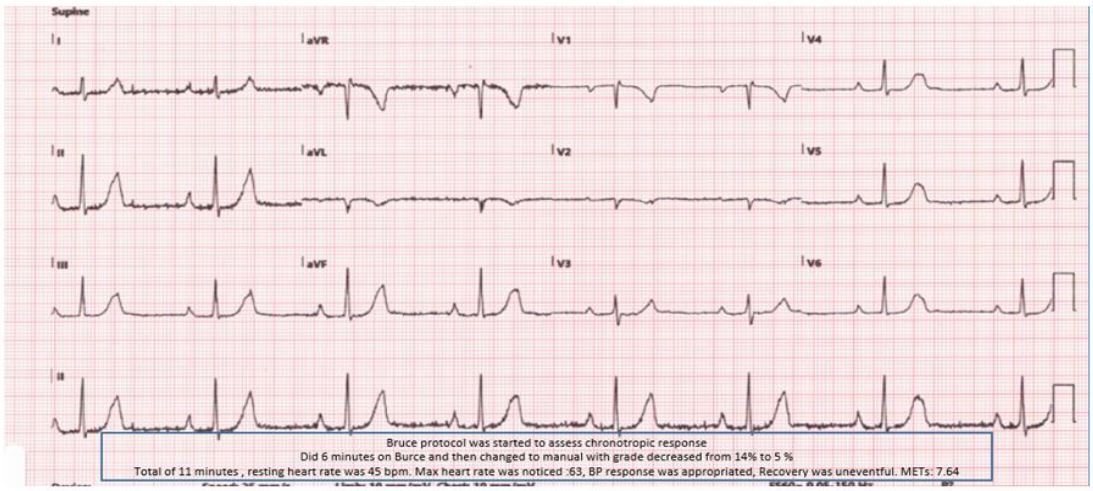

Figure 2. Twelve-lead electrocardiogram showing complete heart block with junctional escape rhythm, isorhythmic atrioventricular dissociation mimicking 2:1 block.

3 | DISCUSSION

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 [7,8]. 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.

Congenital complete heart block (CCHB) was first recognized in 1846 [9],anddocumented into two categories: congenitally malformed and otherwise anatomically normal hearts [10].Secondary CHB might be causedby infections, cardiac ischemia or myopathies, autoimmune diseases, or endocrinological diseases that require extensive workup to be ruled out [12].The acquired variety is rare during pregnancy as this type is mainly seen after 50 years of age [2], whilethe congenital variety is seen rarely during pregnancy.

One of the common 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[11].

In the presenting ECG of our patient (figure 1), the escape rhythm is intra-nodal with narrow QRS complexes at a rate of 40 to 60 beats per minute, often these patients are asymptomaticand respond to physical exertion or atropine [13], Our patient's exercise-induced chronotropic response was mild.

There are a few essential factors worth addressing in our scenario; the first is(Figure 2) the ECG appears to be a heart block with a 2:1 pattern, which is seen chiefly in either Morbitz type I or type II second-degree AV block. Given that the patient probably has CCHB, the most likely diagnosis for (Figure 2) is CHB with isorhythmic AV dissociation. It occurs when the rates of independent atrial and ventricular PMs are in an integral ratio. However, there

is no actual synchronization between atrial and ventricular contractions [14].

The second factor; Cardiac output is the sum of the stroke volume and heart rate. However, when no increment in heart rate occurs with exercise stress, the cardiac output is primarily stroke volume dependent [6].

The third factor is acquired diastolic MR /TRwith AV dissociation. In patients with sinus rhythm and AV block, prolongation the PR interval reversesthe 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/TR during diastole[15,16].

Our patient had no evidence of structural heart disease and generally functioned without symptoms. Comprehensive laboratory testing excluded electrolyte imbalance, thyroid abnormalities, autoimmune diseases, rubella, toxoplasmosis, and syphilis. Therefore, CHB is most probably congenital in her case. This case illustrates the excellent prognosis of asymptomatic complete heart block but raises concern with advancing age. Complete heart block in pregnancy and labor is asymptomatic, except Mandal et al. reported that 21 pregnant women with complete heart block had syncope and palpitations [17]. Others reported complications such as preterm labor, IUGR, or preeclampsia like our patient [18,23]. Therefore, labetalol (in case of preeclampsia) and nifedipine (in preterm labor), which exacerbate the heart block, are contraindicated.

Appropriate anesthesia for the CHB is controversial. Spinal anesthesia is not recommended because of the possibility of blockage at high sympathetic levels, which may cause dangerous bradycardia [30]. General anesthesia-related complications may include bradycardia, hypertension, arrhythmia, and cardiac arrest, bupivacaine, fentanyl, and ketamine, are less likely to affect heart rates [12].

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.

In cases of asymptomatic CHB in pregnancy or labor, there are no precise guidelines. There have been numerous reports of cases, but no clear conclusions have been reached [17,18]. It would be desirable to point out signs predicting an increased risk in the 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 [19,20,21]. The American Heart Association's latest guidelines for PM treatment include similar advice. [22].

A PM may be indicated in asymptomatic patients before labor or postpartum, according to Mandal et al.[17]Modi et al., on the other hand, were against routine pacemakers for asymptomatic women, with emergency pacing arrangements in place if required. [26]. In patients with AV block, Hidaka et al. argued that a pacemaker isn't always needed during labor in absence of hemodynamic alterations. [23], and only for those whose heart rate does not increase during the exercise test a temporary PM is necessary [24,25].While Khardke et al. proposed temporary pacingin patients with atropine-resistant bradycardia, first- and second-degree AV block, and atrial fibrillation with low VR[27]. Thus, according to Thaman et al., not all women with complete AV block exhibit hemodynamic instability and accordinglydo not require pacing. They also suggested that various underlying heart structural disorders influenced and affected the course and outcome of pregnancy. [18]

Similarly, there is no clear consensus for long-term pacing; some authors have urged pacing early in pregnancy since syncopal attacks could be fatal. [13].Kivark and Modi, reportedcesarian sectionin asymptomatic complete heart block without an active pacemaker [25,26] Need for pacing, as well as the optimum timing or rate setting of temporary pacing, has not been objectively appraised, particularly for labor. The danger of consequences such as irradiation, bleeding, infection, or embolism must be weighed.

The majority of asymptomatic CCHB patients will eventually become symptomatic and require PM treatment [30,31]. The primary concernis 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 [34]. The question has remained unanswered to this day. It's difficult to foresee a CCHB becoming a lower-degree block or a sinus rhythm.[20,21].

4 | 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.

Competing interest: The authors declare no conflicts of interest.

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