Congenital Complete Atrioventricular Block in a Young Man: A Case Study

Connie S. Chronister, RN, MSN, CCRN

Congenital complete atrioventricular block (CCA VB) occurs from fetal age to young adulthood in a person who has had no other cardiac arrhythmia detected before the heart block and no other history or signs and symptoms of generalized disease. In this article, I present the etiology of CCA VB, risk factors, electrophysiology, pathophysiology, clinical features, diagnostic test findings, treatment, a case study, implications for nurses, and teaching topics for patients with pacemakers. Increased awareness of suspecting an atrioventricular heart block, in children and young adults undergoing a physical examination or who are hospitalized for unrelated problems and then found to have a slow heart rate, will ensure earlier recognition of this problem. Early recognition can help prevent cardiac decompensation and sudden cardiac death in these young persons.

Diagnostic criteria for CCA VB in children include occurrence of complete atrioventricular block (CA VB) in utero or at birth as indicated by echocardiographic or electrocardiographic (ECG) findings or CA VB with some evidence of a slow pulse at a fairly early age with no history of myocarditis that might have caused the condition after birth. The estimated incidence of CCA VB is 1 in 20,000 live births, but in a study by Siren et al, the incidence in Finland during the 1990s had increased to 1 in 11,000 births. This increase may be due to improved technological advances and increased prenatal care. CCA VB affects males and females equally and has been associated with familial clustering. The common occurrence of heart block in older patients lowers expectations of CA VB in younger patients. CA VB most often occurs in patients who are older (>70 years) who may have injury to or degeneration of the fibers in the conduction system.

PRIME POINTS

- Health care professionals performing physical assessments should suspect the possibility of an underlying heart condition in young adults and children with slow heart rates.

- Arrhythmias associated with a slow heart rate can easily be missed during a physical assessment if an ECG is not obtained.

- Patients with congenital complete atrioventricular heart block can be totally asymptomatic or can have subtle signs and symptoms that can easily be attributed to other causes.

CE Continuing Education

This article has been designated for CE credit. A closed-book, multiple-choice examination follows this article, which tests your knowledge of the following objectives:

1. Discuss the pathophysiology of complete congenital atrioventricular block
2. Identify the clinical features of complete congenital atrioventricular block
3. Describe 3 teaching points for a patient with a pacemaker

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The estimated incidence of CCA VB is 1 in 20,000 live births, but in a study by Siren et al, the incidence in Finland during the 1990s had increased to 1 in 11,000 births. This increase may be due to improved technological advances and increased prenatal care. CCA VB affects males and females equally and has been associated with familial clustering.

The common occurrence of heart block in older patients lowers expectations of CA VB in younger patients. CA VB most often occurs in patients who are older (>70 years) who may have injury to or degeneration of the fibers in the conduction system.
Etiology and Risk Factors

CCA VB is thought to be caused by destruction of the conduction system by myocarditis and is associated with maternal autoimmune disease, structural heart disease, and, when diagnosed in utero, neonatal lupus syndrome. CCA VB is strongly associated with maternal connective tissue disorders (CTDs), especially those involving autoantibodies to Ro/SS-A, which have been detected in maternal sera in up to 98% of cases of CCA VB. These autoantibodies are thought to cause damage to the cardiac conduction system. CCA VB may occur in association with destruction of the normal conduction system by maternal autoantibodies to the nuclear antigens Ro/SS-A and La in CTDs such as systemic lupus erythematosus (SLE), which triggers immune-mediated inflammation of the atrioventricular nodal and myocardial tissues in a susceptible fetus. Viral infections and long-QT syndrome might be responsible for cases in which these autoantibodies are not detected in the mother. CCA VB is thought to be primarily the result of maternal CTDs such as Sjögren syndrome, SLE, and rheumatoid arthritis. In SLE, small-vessel vasculitis and fibrous tissue infiltration are the major causes of dysfunction of the sinus or atrioventricular node. These conditions are thought to cause a placental transfer of antibodies to Ro/SS-A and La that damage the fetal conduction system. When tested for CTD, 75% of mothers who had a child with CCA VB had no signs and symptoms of CTD but had antibodies to Ro/SS-A.

Other causes of CCA VB (25%-33%), which may or may not have an inherited component, include structural cardiac abnormalities such as L-transposition of great arteries, ventricular inversion, and atrioventricular septal defect in which the atrioventricular node ends blindly. CCA VB patients with structural heart disease have higher mortality than do those without structural heart disease.

Neonatal lupus is rare and may lead to CCA VB due to transplacental transfer of autoantibodies from mothers who are positive for antibodies to Ro/SS-A and La, but CCA VB in patients with neonatal lupus is thought to occur in only 3% of infants with antibody-positive mothers. Neonatal lupus syndrome has a 12% to 41% first-year mortality rate and is rarely responsible for previously asymptomatic CCA VB that occurs in childhood or young adulthood. Jaeggi et al reported that the mechanism of heart block may differ between CCA VB diagnosed in utero or during the neonatal period (<28 days) and CCA VB diagnosed in childhood. This finding suggests that the mechanism of heart block is not due to autoimmune destruction of the conduction system in these patients because CCA VB diagnosed in childhood has a less dire prognosis.

The overall mortality rate for CCA VB is 4% to 29% (Table 1). The general population potentially includes a small percentage of individuals who have grown to young adulthood with CCA VB that has not been diagnosed. The exact incidence of this problem in individuals who are now young adults is unknown because CCA VB may not have been recognized when they were younger and their bodies were able to compensate for the slow heart rate.

CCA VB is associated with many potential cardiac complications (Table 2). The main causes of early death due to CCA VB are cardiac failure associated with cardiomyopathy and sudden cardiac death. These cardiac complications account for

**Table 1** Mortality rates associated with congenital complete atrioventricular block (CCA VB)

<table>
<thead>
<tr>
<th>Population</th>
<th>Estimated mortality rate, %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Children with isolated CCA VB</td>
<td>6-8</td>
</tr>
<tr>
<td>Infants with isolated CCA VB</td>
<td>4-8</td>
</tr>
<tr>
<td>Infants with associated structural heart disease</td>
<td>29</td>
</tr>
<tr>
<td>Children with associated structural heart disease</td>
<td>10</td>
</tr>
<tr>
<td>Adults with isolated CCA VB who were asymptomatic during infancy and childhood</td>
<td>5</td>
</tr>
</tbody>
</table>

*a Based on data from Balmer et al and Michaelsson et al.

Author

Connie S. Chronister is a clinical instructor at the University of Akron in Akron, a doctoral student in nursing practice at Case Western Reserve University in Cleveland, and a staff nurse at Medina General Hospital in Medina, Ohio.

Corresponding author: Connie S. Chronister, RN, MSN, CCRN, 2824 Sharon Copley Rd, Medina, OH 44256 (e-mail: csc21@akron.edu).

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up to 14% of deaths in patients with CCAVB. Adams-Stokes attacks and heart failure may develop at any age in children with CCAVB. Patients with CCAVB can appear healthy, especially when they have no other cardiovascular problems, but it is clearly established that the first manifestation of CCAVB can be sudden death.

Electrophysiology of CAVB
CAVB is characterized by a complete absence of conduction of electrical impulses from the atria to the ventricles; the cardiac conduction system (Figure 1) is interrupted at the level of the atrioventricular node, bundle of His, or bundle branches. Because of the dissociation of the atria and ventricles, patients with CAVB have a ventricular heart rate that is slower than the atrial rate. The atrial and ventricular chambers beat independently of each other because no communication exists between the chambers. This lack of communication results in a conduction block at the level of the atrioventricular node. For example, the atrial rate may be 80/min and the ventricular rate 40/min. A regular P-P interval may be visible, or the interval may appear to change because of nonvisible P waves that may be hidden in the QRS complex or the T waves. Importantly, although the PR intervals vary widely, the PR measurement in complete heart block is meaningless because no relationship exists between atrial conduction and ventricular conduction; they are independent of each other. Table 3 indicates differences in types of heart block and provides examples of ECG findings.

The ventricular rate in CAVB is slow because the atrioventricular (junctional) or ventricular pacemaker takes over to pace the heart at a rate much slower than the intrinsic sinus node rate. The inherent rate of automaticity of the atrioventricular pacemaker is 40/min to 60/min, with a narrow QRS complex; the inherent rate of automaticity of the ventricular pacemaker is 20/min to 40/min, with a wide QRS complex. However, escape rhythms such as accelerated junctional or accelerated idioventricular rhythms are common. The morphology of the escape rhythm may help

Table 2 Cardiac complications associated with congenital complete atrioventricular blocka

<table>
<thead>
<tr>
<th>Complication</th>
<th>% of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dilated cardiomyopathy, heart failure (more common in patients &lt;4 years old)</td>
<td>3-4</td>
</tr>
<tr>
<td>Syncope or Adams-Stokes attacks (more common in patients &gt;4 years old)</td>
<td>40</td>
</tr>
<tr>
<td>Mitral valve insufficiency in adults</td>
<td>10</td>
</tr>
<tr>
<td>Sudden death</td>
<td></td>
</tr>
<tr>
<td>In newborns</td>
<td>20</td>
</tr>
<tr>
<td>In adults</td>
<td>10</td>
</tr>
</tbody>
</table>

a Based on data from Esscher, Kertesz et al, Gordon, Beaufort-Krol et al, and Michaelsson and Jonzon.

Figure 1 Cardiac conduction system. Reprinted with permission of SensorMedics.
distinguish its origin. A narrow complex (<120 ms) indicates a block in the atrioventricular node with the escape pacemaker at the atrioven-
tricular junction, a condition that is considered “stable” and usually
does not progress to asystole.20 A wide QRS (>120 ms), signifies a dis-
tal or infrahisian block (in the lower part of the bundle of His or in the
bundle branches) and is inherently less stable, with a risk of asystole or
polymorphic ventricular tachycar-
dia20 (Figure 2). A wide QRS com-
plex (>120 ms) may also occur if the

<p>| Table 3 Differences between first-degree, second-degree, and complete heart blockα |
|-----------------------------------|-----------------------------------|-----------------------------------|</p>
<table>
<thead>
<tr>
<th>Feature</th>
<th>First degree</th>
<th>Second degree, Mobitz type 1 (Wenckebach)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Electrocardiographic appearanceb</td>
<td><a href="#">Image</a></td>
<td><a href="#">Image</a></td>
</tr>
<tr>
<td>Rate</td>
<td>Generally at intrinsic rate of 60/min to 100/min May be slower</td>
<td>Atrial rate generally at intrinsic rate of 60/min to 100/min Ventricular rate depends on number of impulses conducted through atrioventricular node; will be slower than atrial rate</td>
</tr>
<tr>
<td>Regularity of atrial rhythm</td>
<td>Regular</td>
<td>Regular</td>
</tr>
<tr>
<td>Regularity of ventricular rhythm</td>
<td>Regular</td>
<td>Regularly irregular</td>
</tr>
<tr>
<td>P wave</td>
<td>P to P wave is constant</td>
<td>P to P wave is constant</td>
</tr>
<tr>
<td>P:QRS ratio</td>
<td>1:1</td>
<td>1:1 until block occurs, then a P wave with a pause absent of a QRS complex, then cycle restarts with a P wave and associated QRS complex</td>
</tr>
<tr>
<td>PR interval (the primary clue to the degree of heart block)</td>
<td>Constant PR interval but prolonged &gt;0.20 s or 200 ms</td>
<td>Variable: PR interval progressively lengthens until a QRS complex is dropped A pause follows the dropped QRS complex, then PR interval resets to shorter interval and the cycle repeats, very cyclic</td>
</tr>
<tr>
<td>QRS width</td>
<td>Normal, &lt;0.12 s</td>
<td>Normal &lt;0.12 s</td>
</tr>
<tr>
<td>Dropped QRS complexes/pauses</td>
<td>None</td>
<td>Yes: pattern of progressive lengthening of the interval from QRS to QRS until a QRS is dropped after a P wave; then a pause occurs, and the interval resets</td>
</tr>
</tbody>
</table>

α Adapted from Huff17 and Garcia and Holtz.18
b Images of second-degree and complete heart block are reprinted from Huszar,19 with permission. Copyright Elsevier 2002.
atrioventricular junction pacemaker discharges before both the bundle branches have completely repolarized, thus causing the electrical impulse to be conducted down one bundle branch (usually the left one) and blocked in the other branch (usually the right one). CAVB most often occurs in patients who are older (>70 years) who may have degeneration of the fibers in the conduction system. CAVB is common in patients with ischemic heart disease, including patients with acute inferior or right ventricular myocardial infarctions, digitalis poisoning (because the sodium and potassium channels in the action potential are altered), or chronic degenerative changes in the conduction system. The common occurrence of heart blocks in older patients may lower a practitioner's awareness of the possibility of CAVB in younger patients.

### Pathophysiology

CCA VB is thought to be due to maternal autoantibodies associated with autoimmune disease that cross the placenta during fetal development, causing a myocarditis that damages the atrioventricular node and bundle of His region of the cardiac conduction system. The myocarditis leads to inflammation and scarring of the atrioventricular node and bundle of His areas, resulting in delayed and blocked conduction through these areas. ECG features of CCA VB are a consistently low ventricular rate, a prolonged QT interval, and a wide QRS complex. Because of incomplete ventricular emptying, mitral regurgitation and left ventricular dilatation are also associated with CCA VB. Cardiac output = stroke volume × heart rate. To maintain cardiac output, the stroke volume increases, and the increased force of contraction contributes to ventricular hypertrophy. This process combined with increased left ventricular end-diastolic volume can result in dilated cardiomyopathy. Cardiac dilatation as a compensatory mechanism may also predispose

<table>
<thead>
<tr>
<th>Type of heart block</th>
<th>Second degree, Mobitz type 2</th>
<th>Complete or third degree</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrial rate generally at intrinsic rate of 60/min to 100/min</td>
<td>Atrial rate and the ventricular rate are always different; atrial is faster than ventricular</td>
<td></td>
</tr>
<tr>
<td>Ventricular rate depends on number of impulses conducted through atrioventricular node: may intermittently be the same as atrial rate when impulses are being conducted</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ventricular rate will be slower than atrial rate when impulses are blocked</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Regular</td>
<td>Regular</td>
<td></td>
</tr>
<tr>
<td>Regularly irregular or irregularly irregular</td>
<td>Regular with ventricular rate slower than atrial rate</td>
<td></td>
</tr>
<tr>
<td>P to P wave is constant</td>
<td>P to P wave is constant but P waves may be difficult to see if buried in the QRS complex or T wave</td>
<td></td>
</tr>
<tr>
<td>1:1 until block occurs, then 2 or more P waves that are followed by a pause without QRS complexes</td>
<td>Variable, more P waves than QRS complexes</td>
<td></td>
</tr>
<tr>
<td>1:1 conduction may recur or may progress to ventricular asystole or complete heart block</td>
<td></td>
<td></td>
</tr>
<tr>
<td>PR interval remains constant until a block occurs and the QRS complex is dropped, resulting in a pause</td>
<td>PR interval is not consistent</td>
<td></td>
</tr>
<tr>
<td>When or if conduction resumes, the PR interval remains constant</td>
<td>Impulses are not conducted from the atria to the ventricles</td>
<td></td>
</tr>
<tr>
<td>Visually the PR interval is not constant and does not progressively lengthen, “no pattern”</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Normal if block is at level of bundle of His</td>
<td>Normal or wide</td>
<td></td>
</tr>
<tr>
<td>Wide (≥0.12 s) if block in bundle branches</td>
<td>Normal (&lt;0.12 s) if block at level of atrioventricular node or bundle of His</td>
<td></td>
</tr>
<tr>
<td>Wide (≥0.12 s) if block in bundle of His</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes: QRS-QRS rate is constant until a QRS complex is dropped and a pause occurs</td>
<td>None</td>
<td></td>
</tr>
</tbody>
</table>
patients to ventricular ectopy similar to the way atrial dilatation predisposes patients to atrial dysrhythmias. Risk of sudden cardiac death may increase in patients with a prolonged QT interval, which predisposes them to development of the ventricular arrhythmia torsades de pointes. Age and resting heart rate are also factors implicated in the risk for sudden cardiac death. The risk for sudden death is increased in infancy, decreases in childhood, and then increases again later in life.

**Clinical Features**

The signs and symptoms of patients with CCAVB depend on the baseline ventricular rate and underlying structural defects. Patients may be asymptomatic until cardiac decompensation begins to occur in adulthood. Slow heart rates and fainting or Adams-Stokes attacks, defined as abrupt loss of consciousness without warning, and near-fainting (dizzy) spells or presyncopal episodes are common overt indications. Other signs and symptoms of low cardiac output and exercise intolerance are sometimes subtle and often are attributed to other causes. Many children and young adults adopt a sedentary lifestyle as an adaptive mechanism. Table 4 lists overt and subtle signs and symptoms of low cardiac output.

Signs and symptoms, including the first Adams-Stokes attack, develop in adulthood in approximately 50% of adults with CCAVB who were asymptomatic in childhood and adolescence, and 10% die prematurely. Also, heart rate decreases progressively in adults with CCAVB as they become older, a change that does not occur in healthy adults. A physiological explanation for increased signs and symptoms of decompensation in older patients with CAVB is related to the deterioration of physiological compensatory mechanisms.

Signs and symptoms in CCAVB are due to a slow heart rate with resultant low cardiac output. Paroxysmal nocturnal dyspnea, a sudden attack of dyspnea that occurs during sleep, is related to CCAVB because the heart rate slows more severely during sleep. Cardiac output is then

### Table 4: Overt and subtle signs and symptoms of low cardiac output associated with slow heart rate in older children

<table>
<thead>
<tr>
<th>Overt</th>
<th>Subtle</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fainting or dizzy spells (Adams-Stokes attacks)</td>
<td>Sedentary lifestyle, avoidance of physical work or play</td>
</tr>
<tr>
<td>Syncope</td>
<td>Postural nocturnal dyspnea</td>
</tr>
<tr>
<td>Exercise intolerance (dyspnea with exertion, diaphoresis)</td>
<td>Shortness of breath with activity</td>
</tr>
<tr>
<td>Sudden death</td>
<td>Long naps or sleep intervals</td>
</tr>
<tr>
<td></td>
<td>Irritability</td>
</tr>
<tr>
<td></td>
<td>Insomnia (related to diaphoresis, postural nocturnal dyspnea)</td>
</tr>
<tr>
<td></td>
<td>Headaches</td>
</tr>
<tr>
<td></td>
<td>Intolerance to heat</td>
</tr>
</tbody>
</table>

^Based on data from Beaufort-Krol et al. 22
further compromised, decreasing oxygen delivery to the tissues, causing dyspnea. Paroxysmal nocturnal dyspnea disturbs sleep, and the person awakens with a feeling of extreme suffocation that resolves when he or she sits upright. Initially, the experience may be interpreted as awakening from a bad dream.\textsuperscript{24} Cardiac fatigue is manifested as tiredness that occurs later in the day, with a resultant need for naps. Exercise intolerance leads to exercise avoidance and adaptive mechanisms such as moving slowly and resting frequently. Irritability, fatigue, and headaches in adolescents can also be associated with heart failure; cardiac output is not sufficient to perfuse the brain and organs because the ventricular rate cannot increase.

**Diagnostic Test Findings**

Patients with CCAVB 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 CCAVB, 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 CCAVB during infancy and childhood, and heart rates less than 50/min are associated with signs and symptoms and increasing mortality.\textsuperscript{14}

In ECG evaluations, the width of the QRS complex is used to infer whether hemodynamically unstable cardiac rhythms may develop.\textsuperscript{14} A ventricular rhythm with a wide complex (>120 ms) and a prolonged QTc (>450 ms) in patients with CCAVB is an unfavorable prognostic sign because it may be related to underlying myocardial damage.\textsuperscript{14} A prolonged QTc is associated with CCAVB and occurs in approximately 15% to 22% of patients.\textsuperscript{1} In multiple studies,\textsuperscript{1,10,14} a greater percentage of patients with CCAVB and QTc prolongation greater than 450 ms had signs and symptoms related to the CCAVB or suddenly died than did CCAVB patients without QTc prolongation.

The results of Holter monitoring in CCAVB have not been extensively studied, but patients who had a heart rate less than 50/min had an increased risk for syncope or sudden cardiac death.\textsuperscript{3,15} Treadmill exercise testing is done mainly to evaluate functional capacity.\textsuperscript{10} In patients with CCAVB without structural heart disease, up to 90% have normal results in exercise treadmill tests.\textsuperscript{1,14} During exercise testing, ventricular ectopy occurs in 50% to 70% of patients, but its importance in sudden cardiac death has not been determined. Patients whose peak exercise or target heart rates were less than 123/min had more cardiac deaths and/or pacemaker insertions than did patients with higher rates.\textsuperscript{10}

Before treadmill exercise testing, the patient’s target heart rate is calculated. The target rate is usually 85% of the maximum predicted rate. (In order to obtain the target rate,\textsuperscript{22} the patient’s age is subtracted from 220, and the new number is multiplied by 0.85.) Because of heart block, patients with CCAVB may not attain their target heart rate during exercise. In the electrophysiology laboratory, the location of the block in the bundle of His can be determined (Figure 1). Supraventricular block is the most common, but the location of the block is not predictive of syncope or sudden death.\textsuperscript{4} The terms supra (above) and infra (below) refer to the location of the block within the bundle of His; intrahisian (within) is a general term referring to the bundle of His. Table 5 lists types of diagnostic tests used to diagnose CCAVB.

**Treatment of CCAVB**

Insertion of a pacemaker is recommended for patients with symptomatic bradycardia,\textsuperscript{3} and treatment of CCAVB centers on temporary and permanent pacing.\textsuperscript{21} Prevention of sudden cardiac death and an increase in functional capacity are the primary goals of pacemaker insertion. Implantation is indicated in most

<table>
<thead>
<tr>
<th>Table 5 Diagnostic testing for congenital complete atrioventricular block\textsuperscript{a}</th>
</tr>
</thead>
<tbody>
<tr>
<td>Test</td>
</tr>
<tr>
<td>Electrocardiography</td>
</tr>
<tr>
<td>Electrophysiology studies</td>
</tr>
<tr>
<td>Treadmill stress testing</td>
</tr>
<tr>
<td>Holter monitoring</td>
</tr>
<tr>
<td>Echocardiography</td>
</tr>
</tbody>
</table>

\textsuperscript{a} Based on data from Vukomanovic et al\textsuperscript{16} and Dewey et al.\textsuperscript{25}
patients with isolated CCAVB\textsuperscript{26} (Table 6). Symptomatic patients or patients with congestive heart failure who are in complete heart block should be treated with an atrioventricular sequential (or dual chamber) pacemaker to maintain normal synchrony of the heart and physiological rate response. If sinus node function is normal in a child or young adult with CCAVB, then a single-lead ventricle dual-chamber pacemaker can be safely implanted. This type is preferred over dual-chamber pacing because of the long period of pacing ahead for young

\begin{figure}
\centering
\includegraphics[width=\textwidth]{table6.png}
\caption{Table 6: Indications for pacemaker implantation in children and adults with congenital complete atrioventricular block\textsuperscript{a}}
\end{figure}

<table>
<thead>
<tr>
<th>Children</th>
<th>Adults</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congestive heart failure</td>
<td>Congestive heart failure</td>
</tr>
<tr>
<td>Syncope</td>
<td>Repeated episodes of syncope</td>
</tr>
<tr>
<td>Dilatation of the left ventricle</td>
<td>Exertional dyspnea</td>
</tr>
<tr>
<td>Ventricular arrhythmia</td>
<td>Fatigue</td>
</tr>
<tr>
<td>Changing (widening) QRS morphology or abrupt pauses in ventricular rate that are 2-3 times longer than the preceding RR interval and bradycardia (heart rate, 50/min to 55/min) with QTc prolongation</td>
<td>Ventricular and supraventricular ectopic beats during exercise tests</td>
</tr>
<tr>
<td>Mitral regurgitation</td>
<td>Mitral regurgitation</td>
</tr>
<tr>
<td>Ventricular rate &lt;60/min</td>
<td>Ventricular rate &lt;50/min</td>
</tr>
<tr>
<td>Dizziness</td>
<td></td>
</tr>
</tbody>
</table>

\textsuperscript{a} Based on data from Balmer et al,\textsuperscript{2} Siren et al,\textsuperscript{5} Michaelsson and Jonzon,\textsuperscript{14} and Dewey et al.\textsuperscript{25}

\textbf{CASE STUDY}

A 30-year-old man came to the emergency department because of shortness of breath, dry persistent cough, and midsternal chest pain. His temperature was 40\textdegree{}C (103\textdegree{}F) orally, and his heart rate was 40/min. Cardiac monitoring indicated CAVB with narrow QRS complexes. The diagnosis was bronchitis and complete heart block, which was verified by findings on a 12-lead ECG (Figure 3). He left the emergency department against medical advice because he was “late to work.” He was told that he might need a pacemaker and that he should see a cardiologist.

At a follow-up appointment with a cardiologist, the patient said that he had been told since childhood, during his school physicals, that he had a “slow heart beat,” but he had not asked questions about it. No subsequent follow-up testing or ECG had been done. In retrospect, the patient had had signs and symptoms of low cardiac output throughout adolescence, but they were so subtle that the association with the slow heart beat was not recognized even by his mother, who was a nurse in an intensive care unit.

A resting ECG showed CAVB with a junctional escape rhythm of 41/min. The QRS duration was 98 ms (normal, 80-120 ms), and the QT interval and the QTc were 524 and 432 ms, respectively (normal range, QTc >450 ms).\textsuperscript{19} An echocardiogram completed during this workup showed normal left ventricular systolic function and an ejection fraction of 72\% (normal range, 50\%-70\%), indicating strong heart muscle function. The echocardiogram showed no left ventricular hypertrophy, normal right ventricle, and normal right and left atria. All valves appeared normal except the pulmonary valve, which was not visualized.

On a stress test, the baseline ECG finding was complete atrioventricular dissociation with a resting heart rate of 38/min. Total exercise time was 10 minutes and 33 seconds with a heart rate response of only 58\%. Blood pressure response was somewhat abnormal, with a systolic pressure of 180 mm Hg. No ST-segment abnormalities occurred during the stress testing. The patient remained in second-degree heart block Mobitz type II during the test and at peak exercise appeared to be in sinus tachycardia, with a heart rate of 111/min. Then, less than 1 minute after exercise, the rhythm changed to complete heart block, with a resting ventricular rate of 38/min. The stress test was inconclusive because the patient did not achieve his calculated target heart rate.

A 24-hour recording from Holter monitoring indicated predominantly complete block with intermittent second-degree heart block Mobitz type II at a mean heart rate of 42/min. The ventricular rate varied from a low of 29/min to a high of 78/min. A single sinus pause of 3.7 seconds was evident, with rare asymptomatic ventricular and supraventricular ectopy.

The cardiologist diagnosed CCAVB on the basis of a history of slow heart rates, which were noted during
patients with CCAVB. Early implanta-
tion of a pacemaker may reduce or eliminate morbidity by prevent-
ing complications such as mitral valve insufficiency.2

**Implications for Nurses**

Advanced practice and school nurses who perform physical assess-
ments on children or young adults and discover a heart rate less than
55/min should use a cardiac moni-
tor to determine the patient’s heart rhythm. Health care professionals
should suspect a heart block until proven otherwise when the heart
rate is slow. Of note, patients born before the advent of routine fetal
ultrasound testing may grow to
adulthood without this cardiac prob-
lem being detected. They may begin
to manifest signs and symptoms of
cardiac decompensation later in life.

In this case study, the patient
had been told during routine physi-
cals that he had a slow heart rate
but did not know to ask how slow or to suspect that his fatigue, insomnia, and other signs and symptoms
might have been related to a slow
heart rate, and he never mentioned
the information to anyone. A parent
should accompany adolescents to
their physical examinations and
sports physicals so that he or she
can be available to ask questions
and obtain information from the
practitioner. Although routine sports
physical forms include questions
about a history of long QT syndrome
and abnormal heart rhythms, these
problems may not have been diag-
nosed or a family history of these
problems may not be known. Patients
with slow heart rates should have an
ECG, and the PR interval, QRS
complex, and QT interval should
be measured and documented.

![Figure 3](image)

**Figure 3** 12-Lead electrocardiogram shows complete heart block.

sports physicals in adolescence but attributed to an
“athletic heart.” One cannot be completely certain that
a complete heart block in a young adult is congenital
unless the block is noted at birth.3 In this case, the diag-
nosis was attributed to the previous lack of diagnostic
capabilities when the patient was an infant and the fact
that his slow heart rate was overlooked because he
appeared healthy. Also supporting the diagnosis was a
family history of great-uncles and uncles on the patern-
al side who died unexpectedly in their 40s of “heart
problems.” The patient’s mother had had tests for SLE;
the results indicated no SLE, although no assays for
Medical/surgical, emergency, and pediatric nurses should also consider the possibility of cardiac problems in patients who have heart rates less than 55/min even though the patients may have no symptoms of bradycardia. A high fever in a patient with a slow heart rate should be a red flag because fever usually causes tachycardia. Health care providers should not assume that a patient with a slow heart rate is athletic and physically fit; the patient should be questioned about his or her involvement in and tolerance of physical activities.

**Patient Teaching**

Patients who are receiving pacemakers to treat CCAVB or some type of heart block and the patients’ families should be taught how to measure a radial or carotid pulse. The patients should carry a card with them at all times that has their pacemaker information. Commonly, patients with CCAVB are pacemaker dependent because of their inherently slow heart rate. Family members of any patient who is pacemaker dependent or has CCAVB should learn cardiopulmonary resuscitation. Patients who are pacemaker dependent or at risk for syncope or cardiac arrest should be encouraged to obtain a medical identification bracelet.

Nurses should also be aware of the psychosocial issues involved when a young adult needs a pacemaker. Patients may deny the need for a pacemaker because it is associated with something old people need and may resist the surgery for pacemaker implantation because of a fear of scars and visibility of the implanted device. In addition, contact sports must be avoided, a situation that may influence the child or adolescent’s fear of social isolation or of “being different.” A comprehensive team approach with nurses, physicians, patients’ family members, and sometimes referral to professional counseling may be beneficial.

Gradual resumption of activities should be encouraged after implantation of a permanent pacemaker. Activities that a child or adolescent may perform should not overstretch or overextend the arm or shoulder on the side of the pacemaker, because doing so can cause lead dislodgement. For example, if a patient has a pacemaker on the left side, he or she may play tennis with the right arm but should avoid playing with the left arm. Noncontact sports should be encouraged to promote self-esteem and improve or maintain circulatory status. Patients and their family members should be

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antibodies to Ro were done. The patient had a paternal great-grandmother who had severe rheumatic arthritis.

The patient reluctantly had placement of an atrioventricular sequential permanent pacemaker with the low rate limit set at 60/min. He was initially resistant to his need for a pacemaker, feeling that the device was something only needed by “old people.” A larger than normal generator was inserted because he needed the pacemaker 100% of the time for atrial and ventricular pacing. Immediately after receiving the pacemaker, he began to feel better. He described having “more energy” and stated days after the pacemaker insertion, “The way I feel now compared to before, I don’t think I would have been alive in five years.”

This patient was a full-term and healthy infant but had a cesarean birth because of a slow heart rate, which was attributed to fetal distress. His childhood medical history was notable for a poor sleep pattern that included night terrors, heat intolerance, and activity intolerance. The activity intolerance was attributed to “laziness” by his parents. According to his mother, his childhood was basically normal except that he had one speed: “slow.” When he was about 13 years old, he became moody and irritable and complained of insomnia and nightmares. These problems were attributed by his parents to poor eating habits, not enough exercise, and just being a teenager who listened to heavy metal music. He also complained of headaches, which were attributed to stress at school and not sleeping. Unfortunately, he dropped out of high school in his junior year, partly because he had missed so much school because of his fatigue. Again these symptoms were attributed to “teenage rebellion.” The patient had actually received a detention at school for “walking too slow” when getting off the school bus. This patient also reported near-fainting spells that occurred occasionally when he would change from a sitting position to a standing position and profuse diaphoresis with light work. Despite having no overt signs or symptoms of cardiac problems in childhood, at 30 years old, the patient was beginning to have signs and symptoms of cardiac decompensation. He would quickly become short of breath and profusely diaphoretic with minimal exertion.
instructed to call 911 if syncope or cardiac arrest occurs.

According to the American Heart Association, patients with pacemakers should be taught to avoid devices that have strong magnetic fields because the magnetic fields can interfere with the pacemaker. Most electromagnetic fields in the home environment rarely affect pacemaker function. Most household and workplace appliances, tools, and equipment can be used without precautions. As a general rule, patients with pacemakers should avoid activities that cause vibration because such movement can dislodge the leads. Any patient with a pacemaker who begins to feel faint or dizzy or begins to have palpitations when around any equipment should move farther away from the equipment because it may be interfering with the pacemaker.

Examples of devices with strong magnetic fields that should be avoided because they cause electromagnetic interference and can interfere with pacemaker function are airport scanners and handheld metal detectors, magnetic resonance imaging machines (although some centers can reprogram the pacemaker and then perform low-energy imaging), arc-welding equipment, and electrocautery sources commonly used in surgery. Items containing magnets should be kept at least 15 cm (6 in) from a pacemaker because most pacemakers have a magnetically activated switch built into the electronics that closes, altering function and causing the pacemaker to change to a preset rate. A magnet may be used to monitor and test the pacemaker during follow-up visits to a physician’s office or during telephone monitoring. Removing the magnet returns the pacemaker back to the previous settings and functioning.

In the United States, patients with pacemakers who use cell phones with a power less than 3 W need take no special precautions because the phones do not appear to damage pulse generators or affect how the pacemakers work. Electromagnetic shielding has been incorporated into the design of modern pacemakers to prevent radiofrequency signals from interfering with the electronic circuitry in the pacemaker. The National Institutes of Health states that cell phones in the United States do not interfere with pacemakers, but patients with pacemakers should pay close attention to their surroundings to make sure no devices are present that might interfere with the pacemakers. The American Heart Association, the Texas Heart Institute, and the Federal Communication Commission all recommend keeping cordless or handheld cellular phones 15 cm away from the pacemaker just to ensure safety. If a cell phone transmits at greater than 3 W of power, the antenna should be kept 30 cm (12 in) away from all models of pacemakers. As an extra safety measure, to avoid any possible disruption in pacemaker function, patients with pacemakers should hold cell phones to the ear opposite the side of the pacemaker and should not place the cell phone in a shirt pocket on the same side of the body as the pacemaker generator. Table 7 provides general information for patients with pacemakers.

Table 7  General information for patients with permanent pacemakers

- Learn how to take radial or carotid pulse, when to call health care provider
- Carry pacemaker identification information card with you at all times
- Encourage family members to learn cardiopulmonary resuscitation
- Wear medical identification bracelet if you are pacemaker dependent
- Follow physician’s guidelines for routine pacemaker checks
- Avoid strong magnetic fields (eg, arc-welding equipment, magnetic resonance imaging machines, high-voltage commercial transformers)
- Be aware that the average pacemaker battery lasts about 5 to 8 years
- Avoid full-contact sports
- Extracorporeal shock-wave lithotripsy to dissolve kidney stones may alter pacemaker function (reprogramming will be required) and should be avoided if the pacemaker generator is implanted in the abdomen
- Therapeutic radiation treatments to treat cancer may damage the pacemaker circuitry
- If you are scanned with a handheld metal detector, tell security personnel that you have a pacemaker and that the scanner should not be held near the device for any longer than absolutely necessary; you can ask for a personal search in place of the metal detector to avoid any risk of pacemaker malfunction
- Cell phones less than 3 W of power do not interfere with pacemakers

^ Based on data from the American Heart Association and the Federal Communications Commission.

Conclusions

Health care professionals performing physical assessments should not assume that young adults and children with slow heart rates (<50/min) are free of cardiac problems. Cardiac monitoring should be used to investigate the slow rhythm. Patients with CCAVB can have subtle signs and symptoms that can easily be attributed to other causes or can be totally asymptomatic until sudden cardiac death occurs. Health professionals should not be fooled into complacency because a patient “looks healthy.”

A thorough physical examination of patients with slow heart rates should include an ECG. Detection of CCAVB by health care professionals during routine health maintenance examinations with subsequent investigation and treatment may be a life-saving event for patients with this heart condition. 

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None reported.

References


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1. What is one of the diagnostic criteria for complete congenital atrioventricular block (CCAVB)?
   a. Complete heart block at birth as indicated by electrocardiographic (ECG) findings
   b. Endocarditis in fetus at 30 weeks
   c. Rapid heart rate at birth as indicated by echocardiographic findings
   d. Incomplete atrial septal defect closure at 5 years old

2. Which of the following disorders is CCAV associated with when diagnosed in utero?
   a. Congestive heart failure  c. Fetal antibody syndrome
   b. Neonatal lupus syndrome  d. Fetal alcohol syndrome

3. Which of the following maternal problems is associated with CCAVB?
   a. Gestational diabetes  c. Myocarditis
   b. Congestive heart failure  d. Connective tissue disorders

4. What are the main causes of early death due to in CCAV?
   a. Myocardial infarction and sudden cardiac death
   b. Cardiac failure associated with cardiomyopathy and sudden cardiac death
   c. Adam-Stokes attack with heart failure and myocarditis
   d. Connective tissue disorders and myocardial infarction

5. What is the ECG characterization of CCAVB?
   a. No P waves only QRS
   b. Absent conduction of impulse from atria to ventricle
   c. Rapid atrial and ventricular heart rates
   d. Irregular PR interval

6. What is the inherent rate of automaticity of the ventricular pacemaker?
   a. 100-120 minutes  c. 40-60 minutes
   b. 60-100 minutes  d. 20-40 minutes

7. What are the 2 components of cardiac output?
   a. Heart rate and stroke volume
   b. Systolic blood pressure and heart rate
   c. Stroke volume and systolic blood pressure
   d. Stroke volume and diastolic blood pressure

8. Which of the following statements about CCAVB is false?
   a. The signs and symptoms are not related to the baseline ventricular rate and underlying structural defects.
   b. Patients may be asymptomatic until cardiac decompensation begins to occur in adulthood.
   c. Common overt indications include slow heart rate, fainting, and presyncopal spells.
   d. Signs and symptoms of low cardiac output and exercise intolerance are sometimes subtle and are often contributed to other causes.

9. Which of the following could be a subtle sign or symptom of cardiac fatigue?
   a. Severe chest pain with exercise
   b. Syncope
   c. Tiredness and frequent naps
   d. Acute dyspnea

10. Which of the following ECG characteristics is an unfavorable prognostic sign because it may be related to potential myocardial damage in patients with CCAVB?
    a. Ventricular rhythm with a QRS complex >120 ms and a QTc >450 ms
    b. Atrial rhythm with a QRS complex <80 ms and a PR interval of 0.16 ms
    c. Ventricular rhythm with a QRS complex <80 ms and a QTc <400 ms
    d. Atrial rhythm with flutter waves and intermittent QRS at 3:1 rate

11. What are the 2 primary goals of pacemaker insertion for a patient with CCAVB?
    a. Decrease in functional capacity and prevention of paroxysmal nocturnal dyspnea
    b. Increase in functional capacity and prevention of sudden cardiac death
    c. Correction of heart rhythm and stabilization of thrombocytopenia
    d. Reversal of connective tissue disorder and prevention of pancytopenia

12. Patients with pacemakers should do which of the following?
    a. Not use cell phones because they damage the pulse generator
    b. Avoid participating in all types of sports
    c. Have pacemaker checks done only when they feel palpitations or dizzy
    d. Avoid strong magnet fields because pacemaker functioning may be altered

Test answers: Mark only one box for your answer to each question. You may photocopy this form.

1. 1a  2. a  3. a  4. a  5. a  6. a  7. a  8. a  9. a  10. a  11. a  12. a
2b  3b  4b  5b  6b  7b  8b  9b  10b  11b  12b
3c  4c  5c  6c  7c  8c  9c  10c  11c  12c
4d  5d  6d  7d  8d  9d  10d  11d  12d

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Congenital Complete Atrioventricular Block in a Young Man: A Case Study
Connie S. Chronister

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