

Third Degree Atrioventricular Block in Children

Vinko Vrdoljak¹ , Matej Šapina², Suzana Bitanga¹, Matej Katavić¹

¹Department of Pediatrics, University Hospital Center Sestre Milosrdnice, Zagreb, Croatia

²Department of Pediatrics, University Hospital Center Osijek, Osijek, Croatia

ABSTRACT:

Atrioventricular (AV) block is defined as a delay or interruption in the transmission of an impulse from the atria to the ventricles due to an anatomical or functional impairment in the conduction system. The conduction disturbance can be transient or permanent. In third degree AV block, also referred to as complete heart block, there is complete dissociation of the atrial and ventricular activity. Atrioventricular block is considered to be “congenital” when it occurs spontaneously in a fetus or young child. In children, the most common cause of permanent acquired complete AV block is surgery for congenital heart disease. Injury to fetal conduction tissues caused by transplacental exposure to maternal autoantibodies related to systemic lupus erythematosus or Sjogren’s syndrome is responsible for 60 to 90 percent of cases of congenital CHB overall¹⁻³. As many as 40 percent of cases of congenital CHB do not present until later in childhood (mean age five to six years). Only rarely do these patients (5 percent) have proven autoimmune etiology. The increased risk of sudden death is associated with the onset of deep bradycardia or ventricular arrhythmia. A routine electrocardiogram is sufficient to diagnose the disease. A 15-year-old girl has been examined at the emergency pediatric outpatient clinic of the University Hospital Centre “Sestre Milosrdnice” for recurrent episodes of presyncope. Physical examination revealed no major deviations other than bradycardia. Her vital signs were within the reference range, with the exception of a pulse of about 44 beats per minute. The electrocardiogram showed atrioventricular dissociation consistent with third degree atrioventricular block. The echocardiogram showed a structurally normal heart except for sinus bradycardia. The girl underwent permanent epicardial pacemaker implantation after which there were no symptoms.

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Correspondence:

Vinko Vrdoljak MD
vinkov@yahoo.com
orcid.org/0000-0002-7321-8057

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KEYWORDS: complete heart block, CHB, complete atrioventricular block, third degree atrioventricular block, congenital atrioventricular block, acquired atrioventricular block, children, bradycardia

SAŽETAK:

ATRIOVENTRIKULARNI BLOK SRCA TREĆEG STUPNJA U DJECE

Atrioventrikularni (AV) blok je definiran kao kašnjenje ili prekid u prijenosu impulsa iz atrijske u ventrikularne komore zbog anatomskih ili funkcionalnih oštećenja u provodnom sustavu srca. Poremećaj provodljivosti može biti prolazan ili trajan. U AV bloku trećeg stupnja, koji se još naziva i kompletni srčani blok, dolazi do potpune disocijacije atrijske i ventrikularne aktivnosti. Atrioventrikularni blok se smatra “kongenitalnim” kada se pojavljuje spontano kod fetusa ili malog djeteta. Stečeni permanentni kompletni AV blok kod djece najčešće je posljedica kirurške korekcije prirodnih srčanih grešaka. Ozljeđa fetalnog provodnog srčanog tkiva uzrokovana transplacentalnom izloženošću majčinim autoantitijelama povezanim sa sistemskim eritematoznim lupusom ili Sjogrenovim sindromom odgovorna je za 60 do 90 posto slučajeva kongenitalnog srčanog bloka 1-3. Čak 40 posto slučajeva kongenitalnog srčanog bloka se javlja kasnije u djetinjstvu (prosječna dob od pet do šest godina). Od navedenih samo rijetki pacijenti (5 posto) imaju dokazanu autoimunu etiologiju. Povećan rizik iznenadne smrti se veže uz nastup duboke bradikardije ili ventrikulske aritmije. Za dijagnozu bolesti je dovoljan rutinski elektrokardiogram. Djevojčica od 15 godina je pregledana u hitnoj pedijatrijskoj ambulanti kliničkog bolničkog centra “Sestre Milosrdnice”, zbog ponavljajućih epizoda presinkopa. Tjelesnim pregledom se nije utvrdilo većih odstupanja, osim bradikardije. Njeni vitalni znakovi su bili unutar referentnog raspona, s izuzetkom pulsa koji je iznosio oko 44 otkucaja u minuti. Elektrokardiogram je pokazao atrioventrikularnu disocijaciju u skladu s atrioventrikularnim blokom trećeg stupnja. Ehokardiogram je izuzev sinusne bradikardije pokazao strukturno normalno srce. Djevojka je podvrgnuta ugradnji trajnog epikardijalnog pacmakera nakon čega više nije imala simptoma i tegoba u budućnosti.

KLJUČNE RIJEČI: kompletni srčani blok, kompletni atrioventrikularni blok, atrioventrikularni blok 3. stupnja, kongenitalni atrioventrikularni blok, stečeni atrioventrikularni blok, djeca, bradikardija

CASE PRESENTATION

A 15-year-old girl was referred to pediatric emergency department outpatient clinic of the University Hospital Centre “Sestre Milosrdnice” due to a recurrence of an episode of presyncope back in 3 days. For the past 11 months, there has been a daily feeling of shortness of breath. Three days before admission, she felt weak, flushed before her eyes, faded. The day before her admission, her eyes turned black again at school and she collapsed. There were no convulsive elements and she did not lose consciousness. After the examination, her vital signs were within the reference range. The heart action was bradycardic, a systolic murmur 1/6 heard over Erb’s point was observed. The heart rate received was a bio variable and ranged from 42-60/min. The electrocardiogram showed atrioventricular dissociation consistent with third degree atrioventricular block. A 24-h holter was made, recording an average rhythm of 44/min (daily 51/min, night 36/min). The minimum rhythm was reduced by 28/min, the maximum by 109/min. RR breaks of up to 3.5 s with rare isolated VES were noted. An echocardiographically discontinuation of the continuous interatrial septum at the level of the oval fossa 2 mm with an L-R flow of 1.06 m/s, hemodynamically insignificant. Spirometry was neat, salbutamol test negative. She had normal electrolytes, renal and liver function, full blood count and random blood sugar test results. Anti-SSA/Ro and anti-SSB/La, anti-double-stranded deoxyribonucleic acid (dsDNA), antinuclear antibodies (ANA) and antineutrophil cytoplasmic antibodies (ANCA) autoantibodies were negative. There were no heart diseases in the family. The child underwent implantation of a permanent epicardial pacemaker after there were no symptoms.

DISCUSSION

The atrioventricular (AV) block is defined as the interruption or delay in the transfer of impulses from the atrium to the ventricles due to an anatomical or functional disorder in the conduction cardiac system. The atrioventricular block is considered “congenital” when it occurs spontaneously in a fetus or child. Congenital complete heart block (CHB) was first described in 1901 by Morquio, who also noted familial occurrence and association with Stokes-Adams attacks and death⁴. The presence of fetal bradycardia (40 to 80 beats per minute) as a manifestation of CHB was first noted in 1921 and is the initial sign of this disorder in many cases⁵. The incidence of congenital CHB is estimated from 1 in 15,000 to 1 in 22,000 newborns^{6,7}. In children, the most common cause of permanently acquired complete AV block is surgery for congenital heart disease. Fetal conduction injury caused by transplacental transfer of maternal antibodies to patients with systemic lupus erythematosus or Sjogren’s syndrome is thought to be responsible for 60-90 percent of the cause of congenital CHB.¹⁻³ Among women with anti-Ro / SSA and / or anti-La / SSB antibodies, fetal / neonatal CHB occurs in approximately 2 percent of pregnancies^{8,9}. 40 percent of all congenital heart block cases are not clinically presented until later in childhood (average between 5 and 7 years)³. Only rarely do these patients (5 percent) have proven autoimmune etiology

3. The etiology of complete heart block in children includes the following: autoimmune diseases, structural abnormalities due to congenital heart defects (eg congenitally corrected transposition of large blood vessels, defects of endocardial pads), idiopathic familial congenital complete heart block, or the consequence of overfeeding myocardial myocardial surgery¹⁰. Congenitally impaired AV nodal function may also be observed in some simple cases of atrial septal defect among patients with Holt-Oram syndrome, an autosomal dominant disorder causing cardiac and upper-limb abnormalities that causes a mutation on the TBX5 gene¹¹. The manifestation of congenital CHB varies depending on the age of presentation, causes, ventricular frequency, and ventricular function. Patients with autoimmune etiology present earlier than those with other causes of CHB¹². Clinically, CHB is presented differently at a certain age. Regarding neonatal age, the primary presentation of infants with congenital heart block is a slow heart rate below 100 beats per minute. Such newborns may appear pale due to decreased cardiac output. Signs of congestive heart failure (eg, crackles on lung examination, elevated jugular venous pulsations, peripheral edema, etc) may also occur. First or second degree AV block found in infants at birth can progress to CHB and should be followed carefully¹³⁻¹⁶. 40 percent of patients with congenital CHB present later in childhood. The primary finding in these children is a slow heart rate with or without symptoms, including poorer effort tolerance, presyncope or syncope (Stokes-Adams seizures). Sudden death has also been rarely described¹⁷⁻¹⁹.

A routine electrocardiogram is sufficient to diagnose the disease and echocardiography reveals an associated structural heart failure or cardiomyopathy. Holter monitoring and load tests assess the basal frequency of the downstream rhythm of the conductor and the possibility of its increase in various physiological requirements, tendency for deep bradycardia and arrhythmias. The therapeutic approach for prenatal diagnosed congenital CHB is largely expectant. If fetal hydrops or other signs of fetal distress develop, there is a possibility of early delivery or emergency pacing. If CHB diagnosed in utero is caused by transplacental transfer of maternal antibodies to patients with systemic lupus erythematosus or Sjogren’s syndrome, corticosteroid therapy should be considered depending on the time of diagnosis. If the heart block is already complete and has been present for more than three weeks, it is considered that an attempt at reversing this complete heart block is futile, and therefore serial echocardiographic and obstetrical follow-up, but no therapy is recommended. If, however, the third degree heart block has been recently diagnosed, a therapeutic course of dexamethasone 4 mg, orally once a day for a period of six weeks may be attempted. For infants and children who are presented later, the therapeutic approach depends on the presence or absence of symptoms. Patients with adequate ventricular frequency and no symptoms may be followed, while symptomatic patients (usually those with a slower ventricular rhythm) need a permanent pacemaker²⁰. Pacemaker implantation was recommended (class I) or felt to be reasonable (class IIa) for patients with congenital CHB and the following characteristics:

symptomatic bradycardia or low cardiac output (class I), wide QRS escape rhythm, complex ventricular ectopy, or ventricular dysfunction (class I), infants with normal anatomy and a ventricular rate of less than 55 beats per minute (class I), infants with other structural congenital heart disease and a ventricular rate of less than 70 beats per minute (class I), children beyond the first year of life with an average heart rate of less than 50 beats per minute, or abrupt pauses two to three times the basic RR cycle length (class IIa). CHB, which is presented prenatally, and is mainly due to maternal antibodies, is associated with a higher mortality rate^{10,21,22}. The outcome of patients diagnosed in the infant is better than the outcome of those diagnosed in utero. Children with CHB who are asymptomatic generally remain asymptomatic until later childhood, adolescence, or adulthood^{6,23}. Prognosis is generally excellent among infants and those diagnosed later in childhood¹⁰. However, exercise limitation and even mortality in childhood are not negligible²⁴⁻²⁶. Even patients who have been asymptomatic throughout childhood are at increased risk of sudden death. In a review of 102 patients who were without symptoms through age 15, 27 (26 percent) had a subsequent syncopal episode, eight of which were fatal¹⁸. The ventricular rate tends to fall slowly with age¹⁸. To compensate for the slow heart rate, the heart enlarges to produce a higher stroke volume; in some cases, this may lead to voltage criteria for left ventricular enlargement and nonspecific ST-T wave changes²⁷ as

well as to heart failure^{28,29}. In general, the prognosis of the following pacemaker implantation is excellent^{15,30,31}. However, a significant number of patients (5 to 11 percent) develop heart failure over the long term, even if a pacemaker is inserted^{28,29}.

CONCLUSION

The most common etiologies of CHB include the following: idiopathic familial congenital CHB, autoimmune antibodies, structural heart abnormalities due to congenital heart disease (eg, congenitally corrected transposition of the great arteries, endocardial cushion defects), consequence of myocarditis or mechanical trauma from surgical or transcatheter interventions. Symptoms of children with complete AV block and structurally normal heart range from unbearable physical exertion, syncope to complete absence of symptoms. Treatment of patients with low cardiac output syndrome or at risk of sudden death is performed by installing a permanent electrostimulator. Despite its infrequency and life-threatening potential, patients with congenital complete heart block have an excellent survival rate with timely diagnosis and intervention.

AUTHOR CONTRIBUTIONS:

All authors listed have made a substantial, direct and intellectual contribution to the work, and approved it for publication.

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