Congenital Complete Atrioventricular Block

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Congenital complete atrioventricular block is found in 1 of 22,000 live births. Over time, it has become apparent that these patients represent not a single distinct disease process, but several processes with the common manifestation of atrioventricular block. The evaluation of these patients to determine their risk of sudden death and need for pacing is not well defined. ***(Tex Heart Inst J 1997;24:301-7)***

Morquio[1] 1st described congenital complete atrioventricular block (CCAVB) in 1901, but there was no electrocardiographic documentation until 1908.[2] The 1st large collaborative study of patients with CCAVB was performed in 1972, and from it and other studies the incidence of CCAVB was determined to be approximately 1 in 22,000 live births.[3-5] As technology has progressed, CCAVB can now be diagnosed as early as the 16th week of gestation, but the true incidence of the disease remains unknown.

Etiology

There are several theories about the etiology of CCAVB. It may be secondary to destruction of the normal conduction system or due to abnormal development. Lev[6] studied the conduction system of patients who had been diagnosed with CCAVB and found 3 pathologic processes: 1) lack of communication between the atrial musculature and the more peripheral part of the conduction system; 2) interruption of the atrioventricular (AV) bundle; and 3) pathologic changes in an aberrant conduction system.

The association of CCAVB with maternal connective tissue disease is a well-described phenomenon. The 1st series demonstrating the association between maternal systemic lupus erythematosus (SLE) and CCAVB was reported by McCue[7] in 1977 and further supported by Chameides.[8] Scott and associates[9] then demonstrated a high correlation between isolated CCAVB and anti-Ro (SSA) antibody in maternal serum, regardless of whether or not the mother had overt symptoms of connective tissue disease. In his series, all mothers with connective tissue disease who had a child with CCAVB were positive for the anti-Ro antibody, and 75% of asymptomatic mothers with a child with CCAVB were also anti-Ro antibody positive. This is 600 times the expected incidence, thus demonstrating the strong association between anti-Ro antibody and CCAVB. Further evidence in support of this association was provided by the work of Ramsey-Goldman,[10] who showed that the risk of a mother with SLE having a child with CCAVB increased from 1:60 to 1:20 if she was anti-Ro antibody positive. Women with SLE were then studied prospectively,[11] and the risk of their children developing neonatal lupus was found to be as low as 3% but as high as 32% if the mother was anti-Ro antibody positive; however, the risk of carditis in the child of a woman with anti-Ro antibody was less than 3%.

Later studies focused on which antibodies (e.g., SSA/Ro and SSB/La) might be causally related to the development of CCAVB and whether levels of antibodies or degree of symptoms correlated with the development of CCAVB.[10,12] Despite the belief that the maternal antibodies were primarily directed at the fetal conduction system, Taylor[13] and Litsey[14] and coworkers demonstrated that maternal antibodies reacted with all fetal myocardial tissue and were not directed specifically at the conduction system. The diffuse nature of the antibody reaction may help to explain the finding of cardiomyopathy in some patients with isolated CCAVB despite the institution of pacing in the neonatal period.[15] While all agree that the presence of anti-Ro antibody increases the risk of giving birth to a child

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with CCAVB, there is disagreement as to whether the titer of the antibody or the severity of disease in the mother plays a significant role. Many have reported that neither predicts the presence of CCAVB in the children.[11,16] This view has been supported by scattered reports of twin pregnancies in which only 1 twin developed neonatal lupus and CCAVB.[11,16,17] The occurrence of CCAVB in only 1 of HLA (human leukocyte antigen) identical twins in which both the twins and the mother had elevated anti-Ro antibody titers also has been reported.[17] Further confusion as to the interaction between the extent of the disease in the mother, her antibody status, and future development of CCAVB was demonstrated by Waltuck and Buyon.[18] In this series of 52 mothers and their 55 offspring with CCAVB, identification of AV block was made either at birth or in utero. Among this group of mothers, 40% were asymptomatic, 26% had SLE, 19% had an undifferentiated autoimmune syndrome, and 14% had Sjögren's syndrome. Over the ensuing 5 years (mean length of follow-up), 50% of the asymptomatic mothers remained as such and those who became symptomatic did so at a median of 1.5 years following the affected birth. Most of the mothers with SLE at the time of the affected pregnancy did not have progression of their disease. Sera from 100% of the mothers reacted with SSA/Ro and 77% reacted with SSB/La. The antibody profile was similar between asymptomatic mothers who progressed to rheumatic disease and mothers who remained asymptomatic. Children born with CCAVB to these mothers both followed and preceded the birth of an unaffected sibling. Twenty-two of the 52 women (42%) had 25 subsequent pregnancies, of which 4 (16%), each from a different mother, resulted in CCAVB. To date, the exact requirements for development of CCAVB in mothers with anti-Ro or anti-La antibodies remain unknown.

The association between CCAVB and congenital heart disease has been well established. Both Lev[6] and Anderson[19] have reported examples of lack of communication between atrial musculature and the AV node or discontinuity of the AV node-His bundle. Prior to the advent of fetal detection, 30% to 37% of patients with CCAVB were diagnosed with an associated form of congenital heart disease, with L-transposition of the great arteries ("corrected" transposition) being the most frequent anatomic defect.[5,20] The occurrence of CCAVB in patients with L-transposition of the great arteries may be explained by abnormal development of the conduction system. During normal development, the AV node is a posterior structure. In contrast, patients with L-transposition of the great arteries have anterior and posterior AV node structures, but it is the anterior AV node that gives rise to the bundle of His.[6] Whether or not this anterior structure is more unstable or susceptible to injury has not been proven. However, patients with L-transposition of the great arteries have been shown to have a much higher incidence of acquired complete atrioventricular block (CAVB), which can occur at any point during their lives.[21,22] Interestingly, patients with an associated ventricular septal defect have a lower risk of developing CAVB in later life than those with an intact ventricular septum.[23] Presently, with the diagnosis of CCAVB commonly occurring prenatally, associated congenital heart disease is found in up to 53% of fetuses.[24] In this group, the most common heart defect is left atrial isomerism. The majority of patients with left atrial isomerism and CCAVB also have an associated AV septal defect.[25,26] Discordant AV connection was the 2nd most common lesion in patients who were diagnosed in utero.[24]

Natural History

The natural history of patients with CCAVB is largely determined by the presence of congenital heart disease and the time of diagnosis. In patients diagnosed prenatally there is a high rate of termination, fetal hydrops, and fetal and neonatal death. Different series report up to 50% fetal and neonatal death. Mortality was highest in patients with associated structural cardiac defects or fetal hydrops.[24-26] In one series of patients with CCAVB and associated structural cardiac disease, only 14% survived the neonatal period, compared with 85% survival in patients with isolated CCAVB.[24]

The prognosis of patients diagnosed in the neonatal period is significantly better than that of patients diagnosed prenatally. Again, however, the mortality is higher in those with associated congenital heart disease. Before the advent of prenatal diagnosis, 32% to 40% of patients were diagnosed at birth.[5,20,27,28] The mortality of those without associated structural heart disease was 15% but increased to 42% in patients with heart disease. These data preceded the advent of neonatal pacing and therefore may reflect a higher mortality than is currently observed.

The mortality in patients diagnosed outside the neonatal period is significantly lower and ranges from 3% to 18%; again, those with associated structural defects have a higher mortality.[5,20] The overall mortality in patients without structural heart disease has been reported to be 5% to 8% and in those with heart disease 29% to 40%.[5,20] While the mortality of patients both with and without structural heart disease has been affected by the ability to pace patients during infancy and childhood, there has been no study specifically focused on this age group since the advent of early pacing. Additionally, the previously mentioned studies citing higher mortality in those with structural heart disease did not directly

address the severity of that heart disease and its associated mortality alone. For the above reasons, the currently observed mortality may be lower than what is reported.

Many patients, both with and without associated congenital heart disease, have survived into adulthood.[5,20,29-34] Unfortunately, there are few data on adult patients with heart disease, and most of our knowledge is based on small series and case reports.[30,33] There have been 2 large prospective studies looking at the long-term morbidity and mortality of adults with CCAVB without associated congenital heart disease.[32,34] Surprisingly, in some patients, higher degrees of AV block may revert to a lower degree of AV block (i.e., 2nd- or 1st-degree AV block), despite CCAVB that began in the neonatal period and persisted into childhood.[32-34] Patients have reverted to normal sinus rhythm with 1st-degree AV block as late as their 3rd decade of life. Many women with CCAVB have been able to tolerate pregnancy without the benefit of pacemaker therapy; however, some women had their 1st Adams-Stokes attack during pregnancy. Of concern are adult patients with CCAVB and no associated heart disease who were asymptomatic during childhood and adolescence and may have their 1st Adams-Stokes attack in adulthood, which can be fatal. Despite the absence of symptoms in childhood, 50% of patients will develop symptoms in adulthood and 10% will die prematurely.[32,34] A long-term issue in the adult with CCAVB is the development of mitral insufficiency. The etiology of this is unclear; it may be due to the presence of long-standing left ventricular dilatation, or it possibly may be a long-term result of more extensive immunopathologic damage to the fetal heart.[34] Mitral insufficiency can occur in up to 10% of adults with CCAVB, beginning as early as the teenage years or as late as middle adulthood. Resolution of mitral insufficiency after pacemaker implantation has been reported to occur only rarely.[34]

Diagnostic Modalities and Prospective Risk Assessment

No diagnostic modality alone has been shown to predict mortality, and therefore the prospective need for pacing in this population is not well defined. Multiple studies have been performed to elucidate risk factors that can be used to determine who should have a pacemaker implanted.

The surface electrocardiogram has been used to determine resting heart rate, the quality of the escape rhythm, and the QTc (Fig. 1). The mean resting heart rate in patients with CCAVB is reported to be between 40 beats/min and 60 beats/min and slows with increasing age.[5,27,32,34-36] Currently, pacing is recommended for patients with symptomatic bradycardia. Many have tried to use resting heart rate alone to predict syncope or sudden death; Michaëlsson and Engle[5] reported a mortality of 4.3% in 23 infants without heart disease with a ventricular rate greater

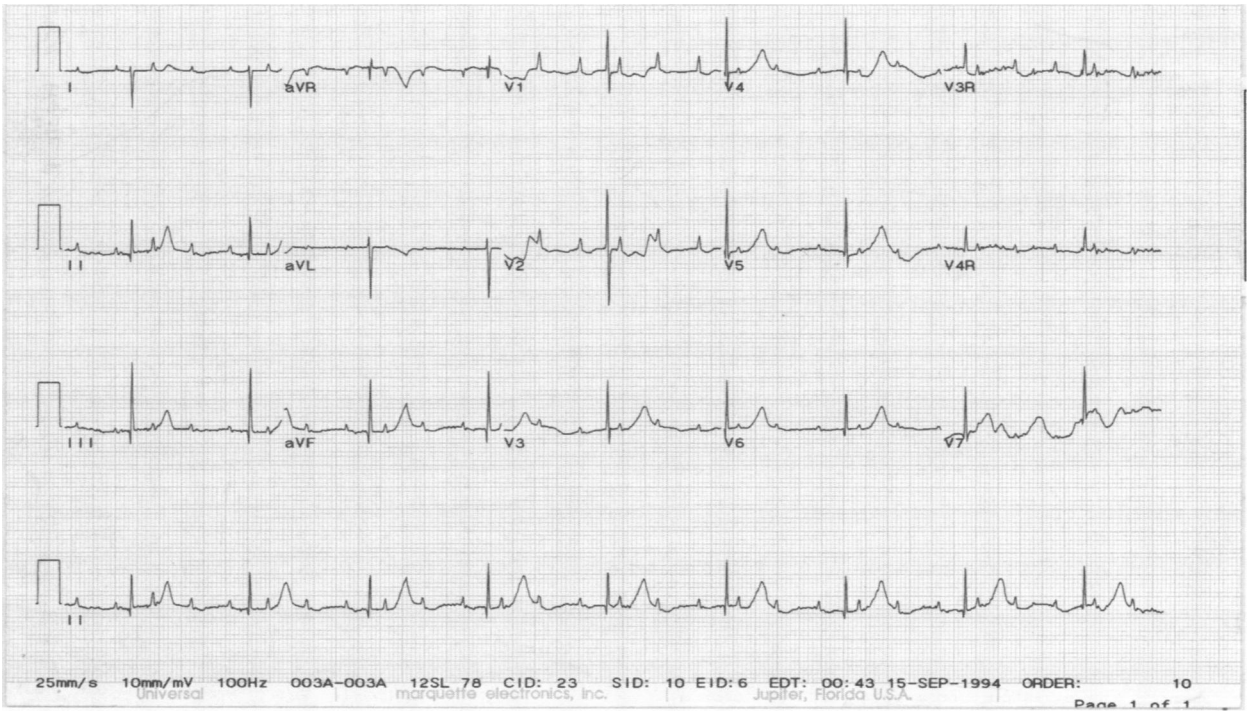

Fig. 1 *A 1-month-old infant with congenital complete atrioventricular block.*

than 55 beats/min and an atrial rate less than 140 beats/min, as opposed to a mortality of 29% in 17 infants with slower ventricular rates and faster atrial rates. Further studies did not find a difference in mean heart rates in patients who subsequently developed syncope or died.[27,34] Other studies, however, have found a statistical difference in mean resting heart rates,[32,35] although the actual differences in heart rates are quite small. In one study, the heart rate of patients with symptoms was 42 beats/min versus 46 beats/min in those without symptoms, and in another study it was 44 beats/min versus 55 beats/min. It is important to mention that these risk assessments have been based on resting heart rates and not heart rates during sleep or the lowest heart rate recorded during Holter monitoring. Interestingly, a slowing of patients' heart rates not only occurs throughout childhood but also has been documented in adults with CCAVB.[34] The width of the QRS has been used to infer the stability of the underlying escape rhythm, with the presence of a wide QRS escape rhythm interpreted as evidence of an unstable pacemaker (Fig. 2). Despite this assumption, studies have failed to document an increased risk of syncope or mortality in patients with a wide QRS escape rhythm in the absence of heart disease.[27,32,34] These data must be evaluated with the knowledge of the small number of patients without heart disease and a wide QRS escape rhythm. Prolonged QTc has been reported to occur in 7% to 22% of patients with CCAVB and is an unfavorable prognostic sign (Fig. 3).[34,37] It can 1st appear in adulthood in patients who have previous documentation of a normal QTc.[32,34]

Only a few studies have focused on the use of Holter monitoring to provide a broader overview of heart rate profiles and, therefore, some indication of the risk of sudden death or syncope in this population. One study[27] reported no difference in minimum heart rate, maximum heart rate, incidence of pauses greater than 3 seconds, or amount of ventricular ectopy between symptomatic and asymptomatic patients. Levy and coworkers[38] found that some patients had concordant atrial and ventricular rate changes, though there were others who had fixed ventricular rates, the implication of which was unclear. Dewey and coauthors[39] prospectively followed patients with CCAVB who had Holter monitoring every 6 months for a mean of 8 years. Patients were divided into 2 groups on the basis of their mean daytime heart rates. All patients who developed either syncope or sudden death had a mean daytime heart rate of less than 50 beats/min. In addition, the majority who developed adverse outcomes had 1 of the following: frequent episodes of junctional exit block, a flat junctional response, or associated tachyarrhythmias. The authors felt that the association of a slow junctional rate with junctional exit block or a flat junctional response may be a manifestation of junctional instability.

Treadmill exercise testing has provided some interesting yet conflicting insights into CCAVB. A sur-

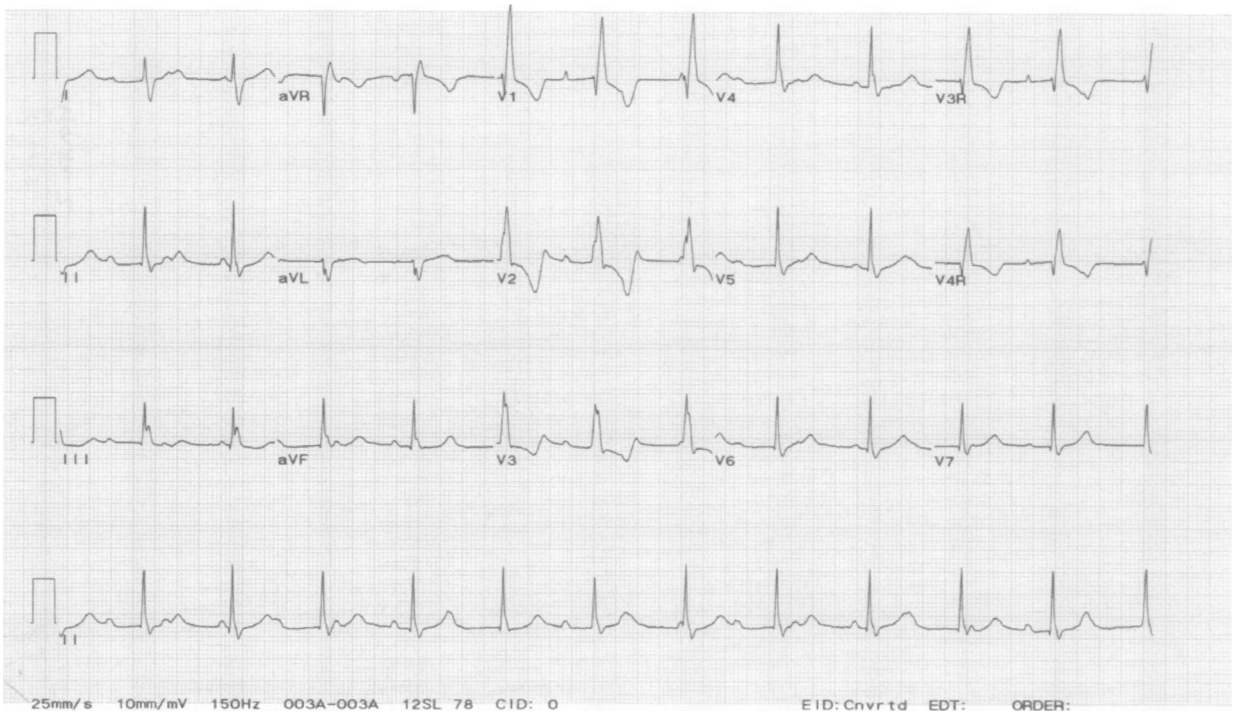

Fig. 2 *A 12-year-old child with congenital complete atrioventricular block and a wide QRS escape rhythm.*

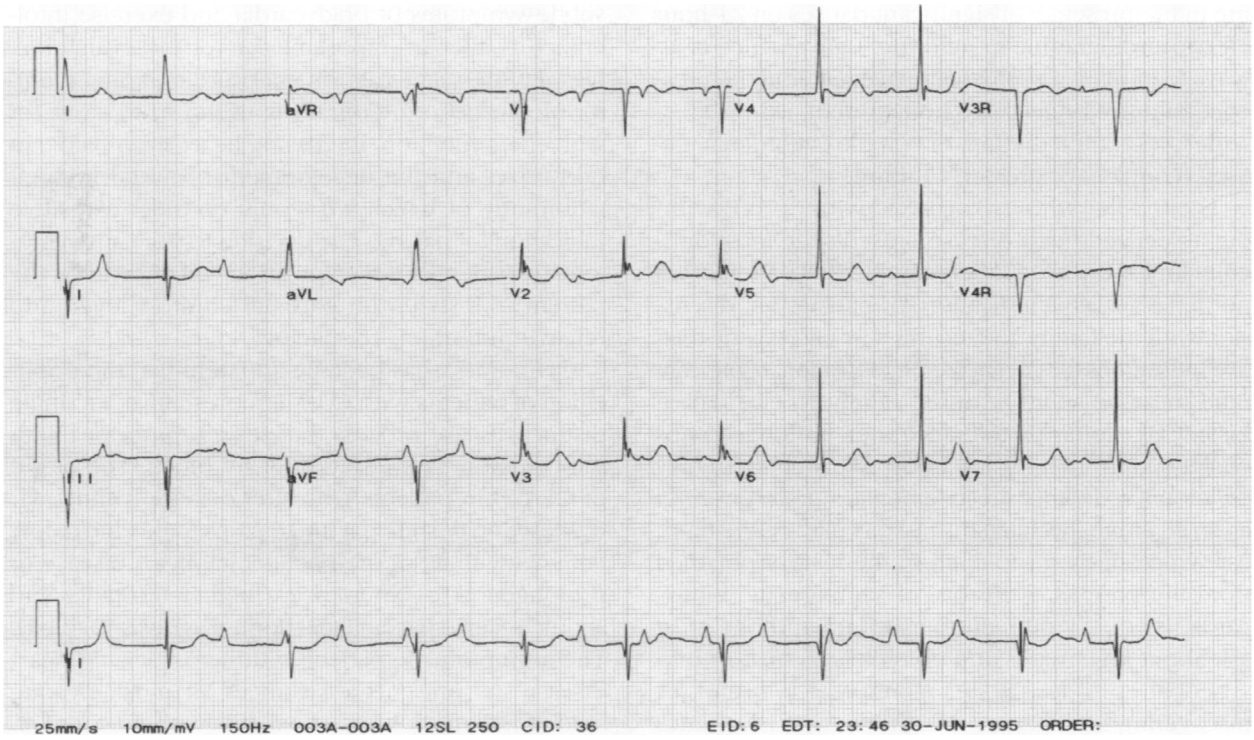

Fig. 3 *An 11-year-old child with congenital complete atrioventricular block and prolonged QTc (620 msec).*

prisingly high number of patients, as high as 90% in some reports, may have a normal exercise treadmill evaluation, especially those without associated heart disease.[32,40,41] Neither exercise tolerance nor the maximum heart rate achieved with exercise can be predicted from the resting heart rate.[40-42] A radionuclide study[43] in a small group of patients with CCAVB demonstrated that the mechanism of cardiac output augmentation with exercise is an increase in heart rate alone and not changes in left ventricular ejection fraction. Ectopy with exercise occurs in 50% to 70% of patients but its prognostic implication is unclear.[32,34,35,40-42] Exercise-induced ectopy has been correlated with increasing age but not with resting heart rate, exercise tolerance, syncope, or sudden death.[34,35,40]

Electrophysiologic studies have been performed on a minimal number of patients with CCAVB. Supra-, infra-, and intrahisian block have been demonstrated, with suprahisian block being most common.[20,35] The location of block has not predicted syncope or sudden death. Electrophysiologic testing has shown that patients may have prolonged junctional recovery times, which could become manifest if patients develop tachycardia and overdrive suppress their intrinsic rhythm.[44] However, this finding has not been correlated with sudden death or syncope.

There is a paucity of echocardiographic data in patients with CCAVB. Cecconi[45] reported normal shortening fractions in patients with CCAVB. We performed a longitudinal study of patients with CCAVB in collaboration with The Children's Hospital of Boston in which we examined ventricular geometry and function and performed wall stress analysis. We found that the left ventricle in patients with CCAVB is moderately dilated, with normal geometry and enhanced systolic function. The degree of left ventricular dilatation and enhanced function did not significantly change with the age of the patient.[46]

To Pace or Not To Pace

The remaining controversy is over whom to pace and when to pace. There is no argument that any symptomatic patient (i.e., a patient with syncopal episodes or congestive heart failure) requires pacing. However, this does not represent the majority of the CCAVB population. The consensus among electrophysiologists is that the presence of significant congenital heart disease is an indication for pacing even though it currently is not listed as a Class I indication in the American Heart Association/American College of Cardiology guidelines.[47] Current debate centers on what to do with asymptomatic patients who present as neonates, children, and young adults. In the newborn patient, most electrophysiologists use the current heart rate guidelines for pacing an asymptomatic neonate (i.e., any infant with an awake resting heart rate less than 55 beats/min). Strong consideration would be given to pac-

ing in the presence of significant pauses on 24-hour electrocardiographic monitoring. Our practice is always to implant a permanent pacemaker in the presence of a prolonged QTc (longer than 460 msec), a wide QRS escape rhythm, or complex ventricular ectopy (couplets or greater) (Table I).

The toddler who is diagnosed with CCAVB without associated heart disease presents the most difficult clinical decision. Many times these patients will not have a Class I indication for pacing, but discussion with the family and testing may elicit many relative indications for pacemaker implantation. Significant time must be spent with the family to elucidate a history of exercise intolerance (i.e., prolonged naps or sleep, or irritability at school). Many children may develop a sedentary lifestyle not as a preference but as an adaptive mechanism, and this may be the hardest symptom by far to elicit in the history obtained from the parents. A history of symptomatic bradycardia is equally difficult to ascertain and one must ask about nightmares and possible syncopal or presyncopal episodes. Holter monitoring is useful for discovering long pauses and monitoring heart rates during sleep. Many times the decision to pace will depend upon clinical signs and symptoms as well as parental comfort and access to health care. The patient who does not receive a pacemaker should be reevaluated yearly.

We recommend that adolescents who have been asymptomatic and have no evidence of cardiac dysfunction be evaluated yearly with an electrocardiogram, Holter monitoring, and a treadmill exercise test. A careful history in which the clinician looks for subtle symptoms of bradycardia and exercise intolerance is also an important part of the evaluation. Despite the scant data on echocardiographic monitoring, an echocardiogram is useful in looking for worsening function, increasing heart size relative to body size, and the development of mitral regurgitation, any of which we would consider a relative indication for pacing. The decision to pace an asymptomatic patient in this population will depend on the patient's lifestyle, the parental comfort level, and relative indications for pacing.

Over time it has become apparent that the population of patients with CCAVB represents not a single distinct disease process, but several processes with the common manifestation of AV block. The origin of CCAVB in patients who later develop ventricular dysfunction despite pacing remains to be elucidated. Patients with associated congenital heart disease or prolonged QTc experience a higher mortality and should receive pacemakers upon diagnosis (Table I). The asymptomatic patient with no associated heart disease still presents a treatment dilemma. Despite studies in large cohorts of patients without congenital heart disease, reliable indicators of syncope or sudden death have yet to be determined.

TABLE I. Indications for Pacing in Patients with Congenital Complete Atrioventricular Block at Texas Children's Hospital

Class	Indications
I	Heart rate <55 beats/min in a neonate or <40 beats/min in a child or adolescent
	Associated congenital heart disease
	Symptomatic bradycardia
	Long naps
	Nightmares
	Exercise intolerance
	Pauses >3 seconds while awake or >5 seconds while sleeping
	Wide QRS escape rhythm
	Prolonged QTc
	Complex ventricular ectopy (couplets or greater) regardless of escape rhythm
II	Ventricular ectopy with exercise
	Prolonged subsidiary pacemaker recovery time

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