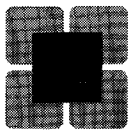

Adults with congenital complete heart block: 25-year follow-up

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■ Complete congenital heart block has been recognized since 1846, yet the prognosis of the disease has not been determined by comprehensive long-term studies. We reviewed 14 cases of complete congenital heart block at The Cleveland Clinic Foundation. The patients were followed for a mean of 25 years (range, 20 to 32 years). Ten patients had structurally normal hearts, and 4 had varying degrees of congenital heart disease. The patients were evaluated with regard to symptoms, and indications for permanent pacemaker implantation were studied. Seven patients received permanent pacemakers. Ages at the time of pacemaker implantation ranged from 15 to 38 years (mean, 25 years). Overall, the patients in our study tended to lead normal productive lives. The main indications for pacemaker implantation were symptoms alone. Guidelines for prophylactic pacemaker implantation have yet to be determined, and the development of these guidelines is difficult due to small numbers of patients at isolated centers. A large international study is needed to set these guidelines.

□ INDEX TERMS: HEART BLOCK; HEART DEFECTS, CONGENITAL; PACEMAKER, ARTIFICIAL; FOLLOW-UP STUDIES
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THE CONGENITAL ETIOLOGY of complete heart block has long been known, and although the criteria for its diagnosis have been well defined, we are unaware of any long-term studies to determine the prognosis of patients with this condition. We reviewed 14 cases of congenital complete heart block (CCHB) at The Cleveland Clinic Foundation in an attempt to determine the long-term consequences of the disease. The mean follow-up was 25 years (range, 20 to 32 years). Patients were also studied to evaluate indications of the need for permanent pacemaker implantation.

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METHODS

To determine long-term outcome, the study included all patients with a diagnosis of CCHB who were followed at the Cleveland Clinic for at least 20 years. The ages of patients at the time of presentation to the Cleveland Clinic ranged from 1 to 23 years (mean, 10.5 years). Eleven of the 14 patients (78%) were referred to the Clinic, having been diagnosed and followed for CCHB at other institutions. Ages at initial diagnosis ranged from 1 to 16 years (mean, 7.5 years). Seven patients were female, and 7 were male.

All patients underwent a complete history and physical examination, including 12-lead electrocardiography and chest roentgenography during the initial evaluation at our institution. The diagnosis of CCHB was initially made on the basis of a slow heart

rate. In all cases, atrioventricular dissociation was confirmed by 12-lead electrocardiography. None of the patients had a history of other arrhythmias, such as atrial fibrillation and atrial flutter, nor were other disease states such as connective tissue disease, myocarditis, or sarcoidosis present. No patients had surgically induced complete heart block. At the time of initial diagnosis, acquired heart block in this age group was not a well-defined condition. We could not rule out all acquired etiologies in this retrospective analysis. There were no known cases of progression to high-degree block.

The criteria used for diagnosis of CCHB were defined by the Association of European Pediatric Cardiologists[1] and state that (1) the atria and ventricles must beat completely and independently of each other; (2) the ventricular rate must be slower than the atrial rate; (3) no other cardiac rhythm can be described; (4) there must be no signs, symptoms, or history of generalized diseases; and (5) the patient must not be over age 20.

Each patient was evaluated with regard to symptoms (syncope, dizziness, shortness of breath, and fatigue) and was assigned a New York Heart Association (NYHA) functional class. Patients with syncope were listed as functional class II. The presence of associated congenital heart defects was noted. Indications for permanent pacemaker implantation were studied in conjunction with age at the time of implantation. After pacemaker implantation, changes in NYHA functional class were evaluated for comparison.

PATIENT DATA

At initial presentation at the Cleveland Clinic, 9 of 14 patients (64%) were NYHA class I, 4 (29%) were class II, and 1 (7%) was class III. No patient presented as class IV.

At initial evaluation, 5 of 14 patients (35.8%) were asymptomatic. Three noted syncope (minimum of two episodes), 3 complained of dyspnea with exertion, 2 had dizziness, and 1 had generalized fatigue as a chief complaint.

All patients underwent cardiac catheterization. Four patients (29%) had associated congenital heart defects: one with a univentricular heart and atrial septal defect, one with corrected transposition of the great arteries, one with atrial septal defect, and one with fusiform dilatation of the aorta. The remaining 10 patients had normal chamber sizes and normal hemodynamic profiles at the time of catheterization. Only the patient with a univentricular heart showed cardiomegaly on the presenting chest radiograph.

Only one patient underwent electrophysiologic testing, which confirmed heart block with a focus below the bundle of His. Echocardiography, Holter monitoring, and graded exercise testing were not performed during the initial evaluation, since these tests were not in routine use at that time.

RESULTS AND FOLLOW-UP

The 14 patients were followed for a mean of 25 years (range, 20 to 32 years). Mean age at follow-up was 38.7 years (range, 20 to 45.3 years). At follow-up, 2 patients (14%) had expired. One was a 22-year-old male with a univentricular heart and atrial septal defect. He had documented ventricular tachycardia degenerating to ventricular fibrillation during hospitalization for congestive heart failure. The second patient was a 43-year-old female who experienced sudden death. She had no associated congenital heart defects. Both of these patients had undergone implantation of a permanent pacemaker. All 12 patients who were alive at follow-up were NYHA class I.

Five of seven females (71.4%) underwent a total of 10 pregnancies. Nine of these pregnancies were well tolerated. One pregnancy was complicated by bradycardia and hypotension occurring in the third trimester. This was the fourth pregnancy for a patient with corrected transposition of the great arteries. No patients required permanent pacemaker implantation before or during their pregnancies. All but one underwent permanent pacemaker implantation at a later date. No deaths occurred during pregnancy.

Seven patients underwent permanent pacemaker implantation. Ages at the time of pacemaker implantation ranged from 15 to 38 years (mean, 25 years). Indications for pacemaker implantation included syncope (minimum of one episode) in five patients (71%) and dizziness in two (29%). The five patients who had syncope included the three patients who originally presented with syncope. Five of the seven patients reported subjective improvement following permanent pacemaker implantation. Two continued to have fatigue that did not appear to be related to pacemaker function. Six patients received VVI pacemakers, four via a transvenous approach and two with implantation of epicardial leads. Only one patient received a DDD pacemaker at the initial implantation.

Patients in the pacemaker and no-pacemaker groups were followed for an equal length of time. The ages of

the pacemaker group ranged from 23 to 45 years (mean, 37.8 years) at follow-up: the ages of the no-pacemaker group ranged from 25 to 42 years (mean, 34.9 years).

DISCUSSION

The congenital etiology of complete heart block was recognized in 1846,[2] and the first case was documented electrocardiographically by White and Eustis in 1921.[3] Complete heart block is either congenital or acquired. This 25-year retrospective analysis could not entirely rule out the diagnosis of acquired nonsurgical complete heart block in this patient population. Many of the diagnostic modalities in routine use today (eg, Holter monitoring, two-dimensional echocardiography, graded exercise testing) were not available when our patients were initially evaluated 25 years ago.

Patients with CCHB fall into two categories: those with congenitally malformed hearts, and those with otherwise anatomically normal hearts.[4] The approach to treatment in patients with normal hearts, eg, permanent pacemaker implantation, has not been well defined. Initially, clinicians were reluctant to implant pacemakers in pediatric patients. This was most likely due to the tendency of these patients to remain symptom-free at an early age, combined with the increased morbidity associated with the procedure of inserting pacing systems 10 to 20 years ago. The generators were bulky and required frequent replacement for battery depletion, and the lead systems did not allow for the rapid somatic growth of pediatric patients. Children received pacemakers primarily for syncope.[5] More recently, the trend has been towards earlier use of pacemakers, both to improve physical working capacity as these patients age, and in response to several reports of sudden death as the initial symptom.[6,7] Currently, our indications for pacemaker use include symptoms, frequent or multiform ectopy, impaired left ventricular function, and prolonged corrected QT interval.[7]

No clear-cut guidelines exist to identify all high-risk pediatric or adult patients.[1] Ventricular rates less than 50 to 55 bpm for neonates[8] and less than 40 bpm for children[8,9] have been proposed as predictive of the development of symptoms. Using this criterion, three patients in the pacemaker group would have been excluded, as their initial heart rates were greater than 50 bpm. Further evaluation by Holter monitoring may have uncovered lower resting-awake ventricular rates in these three patients. We found no significant correlation between ventricular rates and symptoms.

Graded exercise testing has been used to evaluate the response of the ventricular rate to strenuous exercise[7] and, thus, may be a useful guide in selecting patients for pacemaker implantation. Bradycardia was originally believed to be the cause of symptoms, and even death, in this patient population. More recently, severe ectopy leading to ventricular tachycardia and fibrillation has been thought to account for many of these episodes. Ventricular ectopy is more likely to occur and is more severe among older patients, independent of other variables (eg, QRS duration, QT intervals). This may indicate a progression of the disease. The more severe arrhythmias occur during the recovery phase of exercise testing, as opposed to exercise, supporting the concept that ectopy may be a response to stress relative to the individual, rather than a predictive norm.[7] Our sample size prohibited a firm conclusion with respect to the variable of exercise test response.

An international cooperative study[8] sought to determine the natural history of patients with CCHB. Unlike the present study, this study did not follow patients into adulthood: the median age at follow-up was 13 years. Permanent pacemaker implantation was recommended for Stokes-Adams attacks only. After following these patients for a longer time, we have demonstrated the need for more widespread use of pacemaker implantation once patients reach adulthood (mean age of 25 years).

The patients in our series tolerated pregnancy well. There were no deaths or syncopal attacks during pregnancy. Other studies[5] have documented Stokes-Adams attacks during pregnancy. No patient in our study was noted to change to other rhythms. Previous reports indicate rare cases with conversion to normal sinus rhythm.[5]

CONCLUSION

Overall, the patients in our study tended to lead normal, productive lives, and the main indication for pacemaker implantation was symptoms alone. We feel that guidelines for prophylactic pacemaker implantation have yet to be established. Because of the relatively small number of patients in our study, a large cooperative study is needed to make this determination.

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