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LONG-TERM RESULTS OF PATIENTS WITH CONGENITAL COMPLETE ATRIOVENTRICULAR BLOCK*

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Congenital complete atrioventricular block is an uncommon disorder with a prognosis which is usually favorable. The disorder is sometimes associated with syncope, sudden death or cardiac pacing. It is difficult to determine the patients at risk of sudden death. In this retrospective study, hospital records of children who had been admitted to the Hacettepe University Children's Hospital between 1970-1990 for evaluation of complete heart block were examined. The study population, consisting of 39 children, 27 males and 12 females, had electrocardiograms consistent with complete heart block. These patients, diagnosed as having congenital complete A-V block, had an otherwise normally structured heart, and were followed up for a period of from one month to 15 years. Age at diagnosis ranged from 27 days to 17 years (mean: 86.9 ± 48 months). Of the 14 patients with symptoms (five with syncope, eight with exercise intolerance and one with presyncope), nine were paced electively and have done well. Clinical and laboratory features of the asymptomatic and symptomatic groups were compared to evaluate potential risk factors. *Key words: congenital complete heart block, isolated risk factors.*

Congenital complete heart block (CCHB) is a rare conduction disturbance. Clinical and laboratory features have been researched in order to determine patients at risk[1-7]. The disorder, originally thought to be benign, is sometimes associated with syncope, sudden death, or implantation of a pacemaker[1,3,8,9].

A persistently low ventricular rate[3,10], a prolonged QT interval[9], and a wide QRS complex have been correlated with an increase in risk among patients with CCHB. Past reviews have often combined subjects with and without structural heart defects, and some have even concentrated on unvariant analysis of the heart rate, without regard to other possible independent risk factors[6].

This study was designed to investigate potentially independent risk factors involved in the development of symptoms related to CCHB, and to report our

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experience in treating a large group of patients in our center afflicted with this disorder. All patients enrolled in this study had a normal cardiac anatomy.

Material and Methods

The hospital records of children who had been admitted to the Hacettepe University Children's Hospital between 1970 and 1990 were examined in retrospect. Patients were enrolled in the study if their electrocardiograms were consistent with complete heart block, and if there was confirmation of a normal cardiac anatomy. All 39 patients met the criteria of CCHB as defined by the International Study on Congenital Heart Block. The children had no previous history of acquired cardiac infection, myopathy or systemic disease.

Cardiac anatomy was evaluated by cardiac catheterization in four patients and by echocardiography in 15 patients. A normal cardiac anatomy was confirmed in the remaining subjects at the initial physical examination, X-ray and electrocardiogram.

Patients were divided into groups according to the presence or absence of one of the following symptoms: (1) sudden cardiac arrest (2) syncope (3) near syncope (4) congestive heart failure and (5) progressive exercise intolerance. Age at diagnosis and age at onset of symptoms were evaluated.

Electrocardiograms were available for 26 of 39 patients (63%). Thirteen patients did not have ECGs and, therefore, were not included in the statistical analysis of certain parameters. Current ECGs were examined for the atrial and ventricular rates, QRS duration and the corrected QT interval ($QT_c = QT / \sqrt{RR}$). A wide QRS was considered ≥ 0.10 sec, and a long $QT_c \geq 0.45$ sec. Heart rate response and exercise-induced ventricular ectopy were determined in some patients during stress tests. Electrophysiologic studies (EPS) were also performed in two subjects.

Risk factors were investigated by comparing the asymptomatic and symptomatic subgroups using the Student's *t* and Mann Whitney *U* tests.

Results

The study population comprised 39 children, 27 boys and 12 girls and the age at diagnosis ranged from 27 days to 17 years of age (mean: 86.54 ± 48.42 months). The follow-up period ranged from one month to 15 years (mean: 40.78 months). Age at last evaluation ranged from one month to 18 years. Nine children were paced. Of the 14 patients with symptoms (five with syncope, eight with progressive exercise intolerance and one with presyncope), one had a ventricular rate of 43 on the ECG and exercise intolerance. Pacing was advised, but her parents refused. The other patients with exercise intolerance had a ventricular

rate of more than 50 beats/min. Of these five patients with syncope, two refused pacer implantation, two were paced, and in one pacing is planned (Fig. 1). Holter monitoring data were available for two subjects, one of whom had a heart rate of less than 50 beats/min and was paced, while the other had a heart rate of more than 50 beats/min.

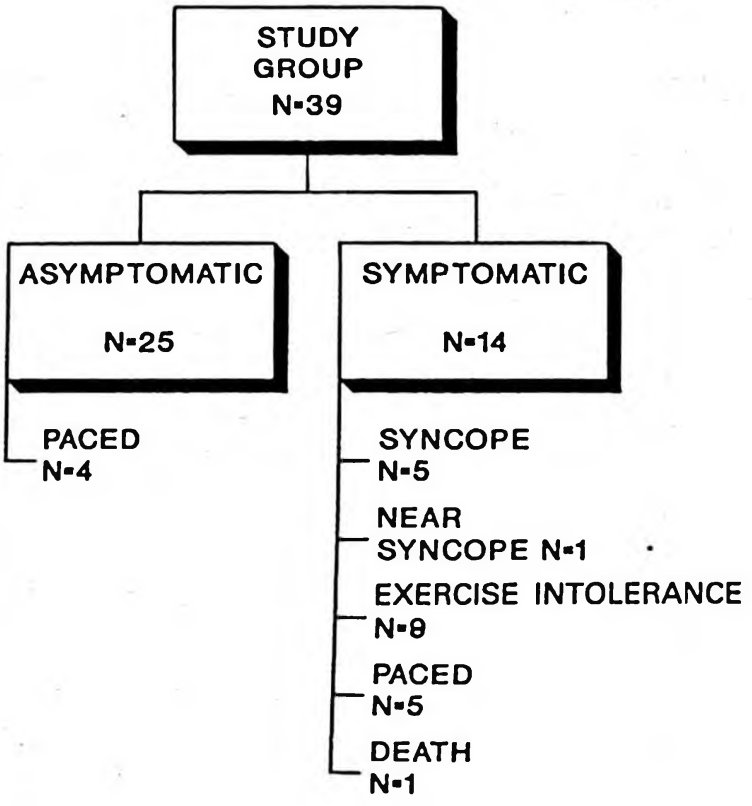

Fig. 1: Clinical outcome of study group.

An EPS was performed in two patients, in one an A-V nodal block was detected and in the other an infrahisian block. Only one patient died, a 27-day-old infant. The cause of death was attributed to noncardiac reasons.

When we compared the clinical features and the laboratory findings of the asymptomatic group with those of the symptomatic group, we found no statistically significant difference between the age at detection of symptoms in these groups (Table I). The development of symptoms did not seem to be related to the age of the patient (mean: 103.10 ± 63 mos.).

When we compared both groups, we were unable to discern any significant difference in respect to heart rate values. Data were also analyzed according to age (five-year) periods. The results remained the same. (Fig. 2).

A wide QRS was detected in two of 16 asymptomatic patients and in three of ten symptomatic patients (30%). A prolonged QT_c was found in five asymptomatic patients (31%) and in seven symptomatic patients (70%).

Table I: Comparison of Risk Factors for Development of Symptoms

	Asymptomatic	Symptomatic	P value
Clinical History	n = 26	n = 14	
• Age at diagnosis (mos)	85.76 ± 42	88.33 ± 55	$p > 0.05$
• Age at 1st symptom (mos)	–	103.10 ± 63	–
Electrocardiogram	n = 16	n = 10	
• Atrial rate (beat/min)	115.87 ± 34	99.20 ± 27	$p > 0.05$
• Ventricular rate (beat/min)	58.43 ± 12	49.40 ± 14	$p > 0.05$
• Wide QRS (≥ 0.10 sec)	11%	30%	–
• Prolonged QT_c (≥ 0.45 sec)	31%	70%	–

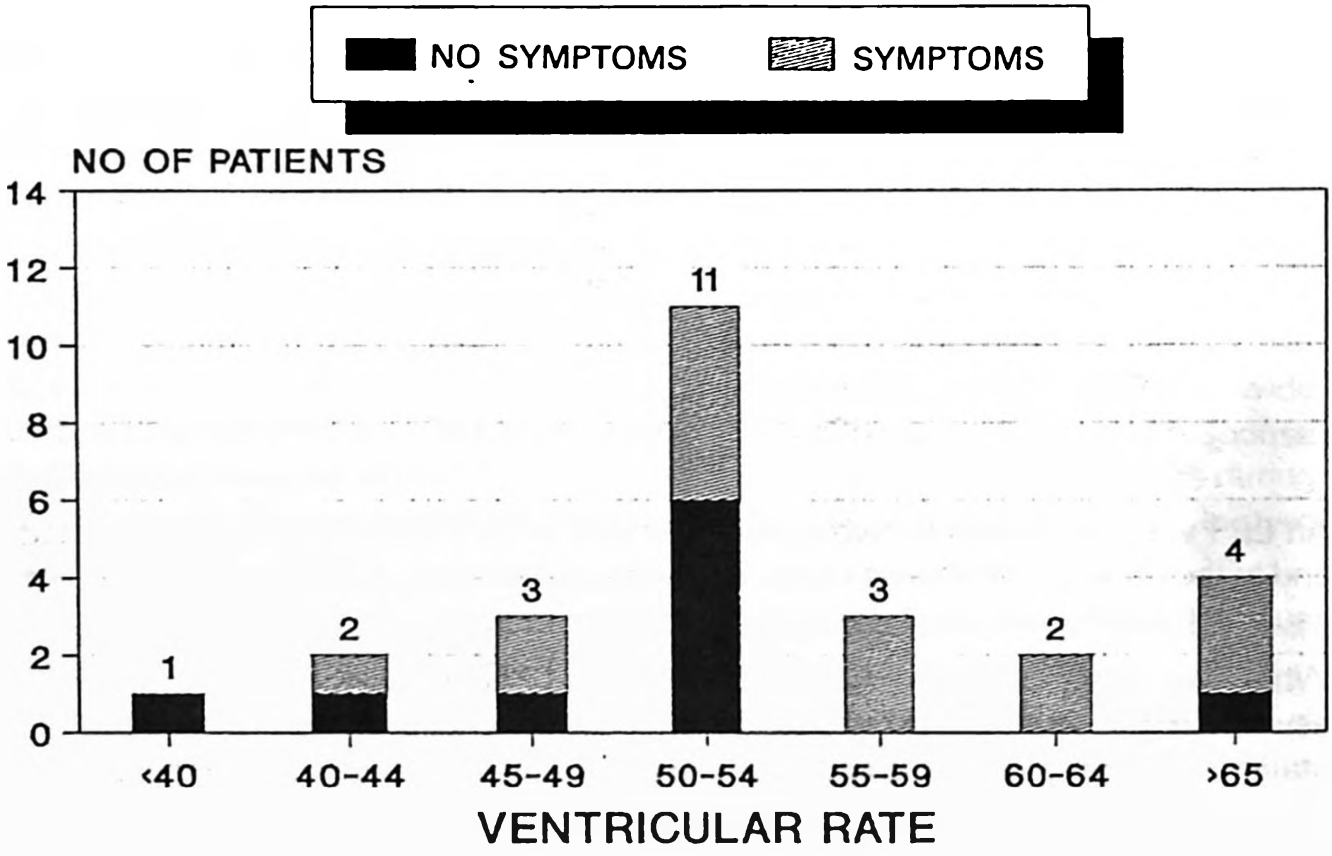

Fig. 2: Relationship between ventricular rate on ECG and clinical symptoms.

Discussion

Congenital complete A-V block is an uncommon disorder and the prognosis is usually regarded as favorable[11]. However, it is sometimes associated with such complications as deterioration, bradycardia, syncope or sudden death[3,8].

Complete heart block was well tolerated in most of the patients in this study. Our results are similar to those reported by Michaëlsson and Engle[12] of large multicenter series where approximately 86 percent of the subjects were alive at the time of the study.

A certain ventricular rate has been most frequently advocated as an indication for pacing in congenital heart block. A rate less than 55 beats/min for neonates and a rate between 40 and 50 beats/min for older individuals are correlated with disabling symptoms or death[13,1,6].

In the present study, the ventricular rate was less than 50 in only six of the 14 patients in the symptomatic group. Of the five patients with syncope, only three had a ventricular rate of 50 or less. On the other hand, the mean ventricular rate was 49.40 ± 14.2 in the symptomatic group and 58.43 ± 12.3 in the asymptomatic group. Thus there was no heart rate variable which distinguished the symptomatic patients. The atrial and ventricular rates were very similar in both groups. These results were obtained from surface ECGs, but if we had used Holter monitoring in all subjects, we could have gotten contrast results. In 1987 Dewey et al[6] reported a Holter monitoring study in CCHB patients with follow-ups for a mean of 8 ± 3 years which revealed that eight of the 13 patients with a mean heart rate of less than 50 beats/min developed cardiac complications of sudden death, syncope, presyncope or excessive fatique while awake. Based on this data, serious consideration should be given to recommending pacemaker implantation in asymptomatic patients with CCHB who have a mean awake heart rate of less than 50 beats/min.

In 1989 Sholler et al[14] reported a retrospective study of 43 patients and were unable to discern any significant difference in heart rate variation between the symptomatic and asymptomatic groups. They explained this discrepancy by the exclusion of patients with anatomic cardiac defects. However, these and our findings raise some doubt as to whether the ventricular rate is the single best index of a patient's compensatory response to CCHB.

With regard to the prevalence of a long QT interval, the first such study was conducted six years ago by Esscher and Michaëlsson[5] of Sweden. They found that 50 of 273 CCHB patients (22%) had a long QT_c interval. In the study of Sholler et al[14] done in 1989, prolonged QT_c was a rare finding, but all patients were symptomatic. In our present study group, 70 percent of symptomatic and 31 percent of asymptomatic patients had a long corrected QT interval. This result indicates a poor prognostic sign of the long corrected QT interval in patients with congenital complete atrioventricular block.

Some reports have focused attention on QRS duration as an alternate risk factor[9]. In 1982 Pinsky et al[1] reported a study conducted in a series of 65 patients with

CCHB. They found a QRS duration of 100 msec or greater for patients with block in or below the bundle of His, and noted that there was a trend toward frequent severe ectopy in children who had a prolonged QRS duration at rest. In this study we found a wide QRS duration in only one patient in the asymptomatic group and in three of the 16 symptomatic patients.

In conclusion, the consideration of a long QT_c as an alternate risk factor is supported by this study. This retrospective study does not invalidate the use of the heart rate as a criteria for prophylactic pacing in cases of isolated CCHB but suggests that other factors are strong independent predictors of symptoms.

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