Multiple Arrhythmias
Detected during Nocturnal Monitoring in Patients with Congenital Complete Heart Block

ARThUR M. LEyY, M.D., A. JOHN CAMM, M.D.,
AND JOHN F. KEANE, M.D.

SUMMARY Twenty patients with congenital complete heart block (CHB) were monitored with ECG tape recordings while awake and asleep. Episodes of marked ventricular slowing during sleep (R-R > 3000 msec., i.e. < 20 beats/min) were noted in 35% (7/20). Most of these sudden R-R prolongations were 2:1 or 3:1 exit block of the junctional focus. Atrial and ventricular rate changes were mostly concordant in 2/3 but the remainder demonstrated fixed ventricular rates while atrial rates varied normally, suggesting a “sick” or “lazy” junctional focus. Other arrhythmias were found in 60% (12/20), including several types of rhythms in three patients. One child with previously unsuspected superimposed multiple arrhythmias later had a syncopal episode despite proven supra-Hisian block.

The multiple patterns of atrial and ventricular rate changes found indicate complex feedback mechanisms, suggesting that congenital CHB is not a single entity. Since a significant number of patients demonstrated one or another of these unexpected and potentially hazardous findings, we recommend that congenital block patients be carefully studied, including Holter monitoring done on a regular basis.

PREVIOUS REPORTS indicate three associated factors can increase the risk of morbidity or mortality in patients with congenital complete heart block: broad QRS complexes, early discovery, and associated cardiac defects. Even in patients with none of these factors, there is a significant incidence of Stokes-Adams attacks and some mortality. In a large International Cooperative Study of congenital complete heart block (599 cases), 418 patients without associated heart disease were mentioned. The study, begun in 1964 and reported in 1972, had already demonstrated an 8% mortality in this subgroup with no other heart disease, with most of the mortality occurring in the first year of life. Close follow-up is recommended for all patients with congenital complete heart block by the authors of this study, but no suggestions are made as to how to follow these children or how to predict those who will develop trouble. Routine examination and ECGs would not detect early warning signs and it is difficult to know how to care for these children.

Electromagnetic tape recordings of cardiac rhythm (Holter Monitoring) over a prolonged period has become generally recognized as the most sensitive method of screening for arrhythmias. Even in the absence of severe prolonged abnormalities with associated syncope, there are often significant clues from abortive arrhythmias to make the test predictive. This test was utilized in 20 patients with congenital complete heart block following the finding in one 4-year-old child with newly discovered CHB of rather marked and unexpected slowing of heart rate at night while undergoing Holter monitoring at home. The results demonstrate a considerable variety and amount of unexpected arrhythmia in this group, as well as more data concerning arrhythmias apparently peculiar to the sleeping hours. Most of the recordings were 12-hour tapes which were turned on at 6 P.M. but toward the end of the study, 24-hour tape recorders were utilized. All 20 patients fulfilled the criteria for congenital heart block as defined in the report of the International Study on congenital heart block, namely that the diagnosis of heart block was established at a relatively early age and there was no history of rheumatic fever or myocarditis. In 17/20 the QRS complex was 0.10 sec or less, in 2/20 it was 0.11 and in 1/20 it was 0.12 (table I). There were four patients with associated cardiac anomalies including two with L-transposition of the great arteries (L-TGA or so-called “corrected” transposition). All of these anomalies were confirmed by angiography. The age of the patients ranged from 3 years to 46 years. Eighteen patients had documented heart block during childhood and the other two stated they had known of slow pulse rates since childhood. Four of the patients did state they had had dizzy spells in the past, but none described syncope prior to the study.

The electrocardiographic tape recordings were analyzed utilizing the scanner module (Avionics) both routinely and then systematically at pre-determined intervals to determine the frequency of associated arrhythmias and circadian rhythms of atrial and ventricular foci.

The ventricular rhythm of one patient was further analyzed by programming a small non-dedicated computer to display sequential R-R intervals as points on the ordinate against lapsed time on the abscissa. This display could be varied from a few minutes to 12 hours. A moving cursor could be utilized to display numerically the exact R-R interval of any two beats or to determine the frequency of a given R-R interval during any given time period.

His bundle electrograms were done on 11 patients in addition to the outpatient ECG tape recordings.

Results All of the patients except the youngest (D.S.) had average daytime ventricular rates between 40 and 55 beats/min, assumed to be junctional by virtue of QRS duration and rate. In eleven this was documented by His studies. However, during the night long R-R intervals (3000-6000 msec, i.e. 10-20 beats/min) were seen in seven (7/20).

From the Cardiology Department, Medical Center Hospital of Vermont, Burlington, Vermont.
Address for reprints: Arthur M. Levy, M.D., Cardiology Department, Medical Center Hospital of Vermont, Mary Fletcher Unit, Burlington, Vermont 05401.
Received June 4, 1976; accepted September 20, 1976.
patients. In six of these, the R-R interval suddenly doubled or tripled. Figure 1 is an example from one of our patients of both a single and a double dropped beat; the prolonged R-R intervals were almost exact multiples of an adjacent R-R interval.

The P-P and R-R intervals derived from the electrocardiogram may be plotted sequentially against lapsed time (fig. 2). The upper plots illustrate single and double dropped beats in two patients. The lower left hand plot illustrates two episodes of single dropped beats in another patient. Except for the loss of ventriculo-phasic sinus arrhythmia, the P-P intervals did not change significantly. Although in four cases dropped beats were rare, in two patients it was a very frequent occurrence. In the six patients in whom the long R-R intervals were double or triple the basic R-R interval, junctional exit block must be the mechanism responsible for this phenomenon.

In the seventh case with long R-R intervals, as illustrated in the lower right hand plot of figure 2 (N.S.), ventricular rates slowed until the R-R interval reached 3000 msec (20 beats/min). This 18-year-old patient was the only one who did not have sudden dropped junctional beats (exit block) as the cause of such a prolonged R-R.

Figure 3 is a dot histogram of the R-R intervals against time as produced by the computer in one of our patients (K.P.) with frequent sudden R-R prolongation. Three distinct populations of R-R intervals are revealed. The bottom cluster of data points represents the basic junctional R-R interval. By means of a moving cursor the average length of the R-R interval in each population was measured. The mid-

---

Table 1. Summary of Patient Data

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age at study</th>
<th>Age at Dx</th>
<th>QRS Duration</th>
<th>Associated Defects</th>
<th>Sleep Bradycardia</th>
<th>Premature Beats and/or Tachycardia</th>
<th>A-V Relationships</th>
</tr>
</thead>
<tbody>
<tr>
<td>L.B.</td>
<td>5½</td>
<td>Birth</td>
<td>0.08</td>
<td>None</td>
<td>—</td>
<td>—</td>
<td>Discordant</td>
</tr>
<tr>
<td>V.C.</td>
<td>10½</td>
<td>Birth</td>
<td>0.08</td>
<td>None</td>
<td>—</td>
<td>—</td>
<td>Discordant</td>
</tr>
<tr>
<td>D.S.</td>
<td>17</td>
<td>Birth</td>
<td>0.08</td>
<td>None</td>
<td>—</td>
<td>JPCs</td>
<td>Intermediate</td>
</tr>
<tr>
<td>K.W.</td>
<td>8</td>
<td>8</td>
<td>0.08</td>
<td>None</td>
<td>—</td>
<td>—</td>
<td>Discordant</td>
</tr>
<tr>
<td>T.L.</td>
<td>5</td>
<td>5</td>
<td>0.08</td>
<td>None</td>
<td>—</td>
<td>—</td>
<td>Discordant</td>
</tr>
<tr>
<td>*K.K.</td>
<td>12½</td>
<td>10</td>
<td>0.08</td>
<td>None</td>
<td>+ VPCs, J. Tach**</td>
<td>Discordant</td>
<td>Discordant</td>
</tr>
<tr>
<td>J.B.</td>
<td>30</td>
<td>Birth</td>
<td>0.08</td>
<td>None</td>
<td>+ VPCs</td>
<td>Concordant</td>
<td>Concordant</td>
</tr>
<tr>
<td>W.F.</td>
<td>18</td>
<td>18</td>
<td>0.06</td>
<td>None</td>
<td>—</td>
<td>VPCs</td>
<td>Concordant</td>
</tr>
<tr>
<td>T.S.</td>
<td>3½</td>
<td>3½</td>
<td>0.12</td>
<td>None</td>
<td>—</td>
<td>—</td>
<td>Discordant</td>
</tr>
<tr>
<td>J.L.</td>
<td>10</td>
<td>10</td>
<td>0.10</td>
<td>None</td>
<td>—</td>
<td>VPCs, JPCs</td>
<td>Intermediate</td>
</tr>
<tr>
<td>K.P.</td>
<td>8</td>
<td>6</td>
<td>0.07</td>
<td>None</td>
<td>+</td>
<td>VPCs</td>
<td>Discordant</td>
</tr>
<tr>
<td>J.D.</td>
<td>3</td>
<td>3</td>
<td>0.08</td>
<td>None</td>
<td>+</td>
<td>VPCs</td>
<td>Discordant</td>
</tr>
<tr>
<td>R.W.</td>
<td>25</td>
<td>10</td>
<td>0.11</td>
<td>None</td>
<td>—</td>
<td>JPCs</td>
<td>Intermediate</td>
</tr>
<tr>
<td>D.M.</td>
<td>3½</td>
<td>Birth</td>
<td>0.08</td>
<td>I-TGA</td>
<td>—</td>
<td>VPCs</td>
<td>Intermediate</td>
</tr>
<tr>
<td>N.S.</td>
<td>18</td>
<td>6</td>
<td>0.08</td>
<td>None</td>
<td>+</td>
<td>VPCs</td>
<td>Concordant</td>
</tr>
<tr>
<td>C.M.</td>
<td>33</td>
<td>14</td>
<td>0.10</td>
<td>None</td>
<td>—</td>
<td>VPCs</td>
<td>Concordant</td>
</tr>
<tr>
<td>D.S.</td>
<td>1½</td>
<td>Birth</td>
<td>0.07</td>
<td>I-TGA</td>
<td>—</td>
<td>—</td>
<td>Concordant</td>
</tr>
<tr>
<td>R.A.</td>
<td>28</td>
<td>Birth</td>
<td>0.07</td>
<td>ASD</td>
<td>—</td>
<td>—</td>
<td>Discordant</td>
</tr>
<tr>
<td>L.B.</td>
<td>46</td>
<td>?</td>
<td>0.11</td>
<td>AS, AR</td>
<td>—</td>
<td>VPCs</td>
<td>Discordant</td>
</tr>
<tr>
<td>S.T.</td>
<td>15</td>
<td>Birth</td>
<td>0.08</td>
<td>None</td>
<td>—</td>
<td>VPCs</td>
<td>Intermediate</td>
</tr>
</tbody>
</table>

Abbreviations: I-TGA = transposition of great arteries; ASD = atrial septal defect; AS = aortic stenosis; AR = aortic regurgitation; J Tach = junctional tachycardia; I-TGA = transposition of the great arteries.

*Later developed syncope and a pacemaker was inserted.

---

Figure 1. ECG from Holter tape recording in one patient showing long pauses and measurements which are compatible with a diagnosis of single and double junctional exit block.
Middle population of R-R intervals measured on average twice and the upper population three times the basic R-R interval. In this same patient, the number of dropped beats was automatically analyzed by computer and the results are displayed as a semilogarithmic plot in figure 4. It is immediately obvious that long R-R intervals occurred only at night. Although there are 1,966 episodes of blocked junctional beats during the night, the effective minute heart rate does not fall below 27 beats/min. The frequency of dropped beats remains high until the patient awakens in the morning. Close scrutiny of the computer display (not illustrated) dramatically demonstrated the sudden and complete cessation of junctional exit block as the patient awakened.

When both bradycardias and tachycardias are considered, arrhythmias occurred in 14 of the 20 patients (see table 1). Except for the patient with sinus bradycardia, all the episodes of slowing occurred during sleep and thus no symptoms were noted. Premature beats were found in 12 patients. The premature beats were both ventricular and junctional and were mostly isolated phenomenon, although in three premature ventricular contractions (VPC) were frequent and paired. These rhythm disturbances were present both during sleep and during wakefulness.

In one patient (K.K.) who, incidentally, had no associated defects, a bizarre series of atrial and junctional rhythms were discovered. These occurred throughout the 12-hour recording. Selected strips from her Holter recording (fig. 5) illustrate paroxysmal atrial tachycardia, frequent premature ventricular contractions, and double junctional tachycardia (i.e., two independent junctional foci as noted by different retrograde [inverted P waves] and antegrade [QRS] rates). The basic underlying atrial rhythm was sinus bradycardia with an average rate of 55 beats/min and frequent slowing to 40 beats/min. Thus while the diagnosis from the routine electrocardiogram was uncomplicated congenital complete heart block, the Holter monitor demonstrated a diffuse disorder of sinus node, junctional, and ventricular tissues. Several months following this recording, this 12-year-old patient had a classical Stokes-Adams seizure.

In all 20 patients, atrial and ventricular rates were measured every 15 min throughout the recording. Analysis of these results demonstrated three main patterns: total atrioventricular concordance, partial concordance, and a nonresponsive junctional pacemaker (table 1). In nine patients (9/20), atrial rate changes were matched by proportionate changes of ventricular rates. Patient C.M.'s records are an example of this pattern (fig. 6, top). The graph on the left is a plot of atrial and ventricular rates throughout a 12-hour period. On the right, atrial rates are plotted against corresponding ventricular rates, in this instance demonstrating the high degree of correlation between atrial and ventricular rates. In five patients, the gross diurnal variations of atrial and ventricular response coincided but more rapid and transient fluctuations of atrial rate were not paralleled by a ventricular rate change (fig. 6, middle).

---

Figure 2. R-R and P-P intervals from selected strips of four patients who demonstrated long R-R intervals at night. Patients K.P., K.W. and J.D. show sudden pauses in R-R intervals which are essentially single and double junctional exit block. Ventricular rates range from approximately 38 beats/min to 12 beats/min. Patient N.S. shows progressive slowing of the junctional focus — thus not exit block (ventricular range 60 to 20 beats/min). Note loss of ventriculo-phasis sinus arrhythmia in graphs of P-P intervals during ventricular asystole.
Figure 3. Computerized dot histogram of one hour of R-R intervals from a nocturnal portion of ECG tape recording in patient K.P. (age 6). Numbers along the ordinate indicate R-R intervals in msec. Passage of one hour of time is indicated by a marker on the abscissa. Three populations of R-R intervals are seen. The bottom cluster of data points represents the basic junctional R-R. The middle population of R-R intervals measures twice (single exit block) and the upper population three times (double exit block) the basic R-R interval.

Figure 4. Semilogarithmic bar graph showing frequency of single and double dropped beats at night in same patient as in figure 3. These pauses are totally absent before sleep and end abruptly as patient awakens.
Although in the other six patients, the variation in atrial rate was commensurate with the level of physical activity, the junctional pacemaker did not significantly change its rate of discharge. The plot on the right shows marked atrial rate changes at a fixed ventricular rate (fig. 6, bottom).

**Discussion**

These results reinforce the uneasiness clinicians have traditionally felt over the extent of clinical follow-up needed for young patients with complete heart block. The demonstration of episodes of long R-R intervals in seven of the patients (e.g., fig. 1) was the most striking finding. In six of the seven, the mechanism was exit block of the junctional focus. Measurements of the ECG strips as well as computerized displays (figs. 2 and 3) appear to demonstrate conclusively the mathematic multiples associated with exit block. Thus far, these long pauses do not appear to produce symptoms, even in the two patients with very frequent single and double exit block. However, ventricular asystole has been found only during sleep in all of our patients. There is one case reported in the literature of a patient who had similar sudden pauses of about 4000 msec. This 18-year-old patient complained of blurry vision and occasionally lost consciousness while ECG strips were being recorded although his bradycardia was less severe than in some of our cases. Thus, it appears that while the exit block in our patients is a result of sleep, sleep may well be protective with its associated decreased oxygen demand. Continuous nocturnal EEG monitoring was done on the patient (K.P.) with

the most slowing and at no time was there evidence of disorders during the ventricular slowing. Nonetheless we remain disturbed by these long pauses and with the possibility that a perpetuation of this exit block with no escape mechanism could result in sudden death during sleep. As for the possibility of a potential shift from nocturnal to more ominous daytime slowing, it would be hoped that intermittent monitoring would detect such a change. This is a partial basis for our recommendation to include monitoring in any program of follow-up.

The finding of tachyarrhythmias as well as bradyarrhythmias superimposed on the baseline heart block is not really surprising. Other studies have shown ventricular premature contractions in this group of patients — and special note is made of VPCs occurring during exercise. In this study, the progress of patient K.K. with multiple arrhythmias (fig. 5) brings out some important implications for these patients. This patient was the only one to develop syncpe after the study was done. She had a true documented syncopal episode at home one day while working with her mother. The mechanism was not documented. In view of her congenital heart block, a pacemaker was implanted after a His bundle electrophysiological study which showed supra-Hisian block. The variety and number of arrhythmias, previously undetected by routine screening techniques, was very striking on her Holter tape. Since she was the only patient with multiple arrhythmias and since she is thus far the only patient who developed syncpe, her future course is uncertain, given that her syncpe could well have been due to tachycardia rather than bradycardia. The
fact that in her case the complete heart block is just one of multiple arrhythmias suggests a very diffuse disorder of the sinoatrial node, junction, and ventricular tissues, as is seen with other tachy-brady syndromes.

The relationships of atrial and ventricular rates in congenital heart block are not well understood. Most of the work has been done on a single patient and in a laborious fashion by taking intermittent ECG rhythm strips for a prolonged period of study. The patient showed concordant atrial and ventricular rate changes. There are many possible feedback mechanisms which could be responsible for the concordance. However, which ones are actually brought into play and which chamber is the primary driving chamber remain unknown.

This pattern of concordant atrial and ventricular rate changes is the most common pattern in our study but is by no means the only pattern (fig. 6). The group who at times maintain atrio-ventricular concordance and at other times show transient fluctuations of atrial rate which are not paralleled by a ventricular rate change may have a difference in innervation of sinus and junctional pacemakers. The group (6/20) with a fixed ventricular rate but marked atrial rate changes is even more complex and may imply inadequate pacemaker function analogous to the "lazy" sinus node function in the so-called sick sinus syndrome. Studies of junctional pacemaker function in these children are being undertaken. Thus far, these have shown a wide variability in junctional recovery times. A normal range for junctional recovery times has not been previously established but there are some patients in the group whose recovery following rapid ventricular pacing is slow enough to cause transient symptoms while others exhibit almost immediate recovery.

These findings all suggest that congenital complete heart block is not a simple single entity. For some time, of course, it has been recognized that some congenital heart block patients have fascicular disease with infra-Hisian block and wide QRS intervals (i.e., idioventricular rhythm). In 1962 this group was separated out as being at increased risk.1 It is interesting that in reviewing this highly-quoted article, one of the three children with fatal Adams-Stokes attacks had narrow QRS (0.09 sec). More recently, a patient with congenital block and narrow QRS has been found to have intra-Hisian block8 and these patients are again felt to be at increased risk, although there are no series of such patients. The pathalogy of this combined group of supra-Hisian and intra-Hisian block with narrow QRS is very variable — and not that of a simple disconnection between atria and ventricles.10 There may be diminished or even absent connection between atria and A-V node due to deficient musculature of the peripheral proportion of the atrial septum. The A-V node itself may be normally developed or may be deficient, absent, or abnormal in shape. The His bundle may be absent, diseased, or cut off from the A-V node. This variability, from absence of structures to deficiencies of structures, accounts for the occasional A-V conduction noted in these patients when carefully looked for. One of our patients, for instance, showed previously unsuspected ability to conduct on the Holter during vigorous exercise, suggesting the presence of a tenuous anatomical connection.

There also remains the possibility that a significant number of so-called "congenital" complete heart block actually appears sometime after birth. There is pathologic evidence that A-V block could result from an overgrowth of collagen directly through the His bundle.11 This type of overgrowth would only occur in infancy and not in utero. And, of course, there is always the possibility of acquired interruption due to some infectious or inflammatory process. So many of the cases of congenital heart block are picked up in early childhood or young adulthood — rather than at birth — that the definition of congenital heart block of the large Natural History Study Group includes "heart block established by graphic records in relatively young individuals."12 The point we want to make is that since the

**Figure 6.** Examples of three types of atrial and ventricular rate changes. Upper graphs (C.M.) show almost perfect concordance between atrial and ventricular rates (9/20 patients). Middle graph (D.S.) shows general concordance but definite periods of discordance (5/20) and lower graph (T.S.) shows flat ventricular response despite marked atrial changes (6/20).
pathologic anatomy is seemingly so varied, one would expect the physiology to be somewhat varied and that prognosis might be related to a number of factors hitherto unrecognized in these patients.

It is our feeling that a number of potential risk factors have been recognized in this study — all requiring long-term follow-up to see which, if any, do indeed increase risk. The abnormalities identified in this study, most of which were totally unsuspected from routine ECG studies and which are suggested as possible risk factors include: 1) nocturnal bradycardia mostly due to junctional exit block, noted in 7/20; 2) tachyarrhythmias (12/20) and especially major tachyarrhythmias (3/20); and 3) unresponsive or "lazy" junctional pacemakers which did not vary discharge rate. Several patients had more than one of these potential risk factors but the only patient with all three suffered a serious syncopal attack. In order to determine whether or not these really are risk factors, we would recommend that follow-up of all patients with congenital heart block should include, in addition to a His study, electromagnetic ECG tape recordings at fairly frequent intervals. The meaning of these described abnormalities is still not sufficiently understood to change recommendations for insertion of permanent pacemakers. These indications, at present, are symptomatic block or evidence of distal block.

Atrial Tachycardia without P Waves
Masquerading as an A-V Junctional Tachycardia

DOUGLAS P. ZIPE, M.D., WINSTON E. GAUM, M.D., BASIL C. GENETOS, M.D., ROBERT D. GLASSMAN, M.D., R. JOE NOBLE, M.D., AND CHARLES FISCH, M.D.

SUMMARY Two patients who presented by scalar ECG with an A-V junctional tachycardia were demonstrated during an electrophysiologic evaluation to have an atrial tachycardia without P waves in the surface ECG. Case 1 had an atrial tachycardia that conducted through the A-V node with a Wenckebach block. Atrial activity was recorded only from the proximal portion of the coronary sinus and from right atrial areas near the tricuspid valve. Case 2 had an atrial tachycardia that abruptly began and terminated following carotid sinus massage. Atrial activity was recorded only in the coronary sinus os, and pacing at that site resulted in atrial capture, with Wenckebach conduction to the ventricles. These observations demonstrate that an atrial tachycardia without P waves can simulate A-V junctional tachycardia with or without Wenckebach block. Such findings may have a bearing on some important electrophysiologic concepts such as the origin of A-V junctional rhythms and the need for atrial participation in A-V nodal re-entry.

RHYTHMS CHARACTERIZED in the scalar electrocardiogram (ECG) by a normal QRS complex and the absence of P waves are thought to originate in or near the bundle of His. Similarly, when a regular ventricular rhythm with normal QRS complexes occurs during atrial fibril-
Multiple arrhythmias detected during nocturnal monitoring in patients with congenital complete heart block.
A M Levy, A J Camm and J F Keane

doi: 10.1161/01.CIR.55.2.247

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1977 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/55/2/247

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/