Complete Atrioventricular Block in Patients with Atrioventricular Discordance

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SUMMARY Although patients with atrioventricular (AV) discordance (corrected transposition) have abnormal conduction pathways and may spontaneously develop high-grade AV block, no quantitative assessment of the risk of this happening is available. We reviewed the data on 107 patients with AV discordance ages 2–76 years (mean 22 years) at follow-up. Eighty-two patients (77%) had a ventricular septal defect, 57 (53%) had pulmonary stenosis, 35 (34%) had tricuspid insufficiency and 24 (23%) had dextrocardia.

Twenty-three patients (22%) had complete AV block. This condition was present in four patients at birth and developed in 19 patients at ages 4 months to 53 years (mean 18.1 years). Nine of these patients had permanent pacemaker implantation, four at the onset of complete block and five an average of 11 years later. Nine patients have AV block but no pacemaker. One patient died suddenly.

Detailed data analysis showed that with increasing follow-up the risk of natural onset AV block continued at a rate of approximately 2% per year after diagnosis. The presence of an intact ventricular septum made AV block more likely. We conclude that patients with AV discordance are at risk of developing complete AV block throughout their lives. With increasing age, this risk is approximately constant and is probably not significantly increased by corrective or palliative surgery if acute surgical AV block does not occur. Pacemaker implantation is not necessary in some children with spontaneous AV block.

THE COURSE of the conduction system in congenital heart disease associated with corrected transposition or levotransposition is abnormal.1-3 Impulses arise from the sinus node and pass to the atrioventricular (AV) node, located near the junction of the mitral and pulmonary valves. The infranodal common conduction tissues then pass anterior to the pulmonary valve annulus in the subendocardium and course inferiorly over the anterosuperior aspect of the membranous septum. Several studies have mentioned the increased likelihood of the spontaneous onset of complete AV block with this type of defect, but these have included patients with other lesions, such as univentricular heart.

There is no information available specifically concerned with the expected rate of development of spontaneous complete AV block in patients with AV discordance and two ventricles. The clinical significance of complete AV block in AV discordance is variable and is related to the site of block and the resulting heart rate.4,5 However, it is not clear whether natural-onset (nonsurgical) complete AV block significantly affects survival in these patients and whether associated anatomic lesions increase the risk of this complication. We therefore reviewed the clinical course of 107 patients with this diagnosis. The goal of the study was to provide quantitative information concerning the risk of complete AV block in AV discordance using life-table methods, as well as to search for factors associated with the natural occurrence of this dysrhythmia.

Material and Methods

The study group consisted of 107 patients evaluated at the Mayo Clinic between January 1951 and June 1981. The age at Mayo Clinic diagnosis of AV discordance ranged from birth to 56 years (mean 12.7 years). Sixty-four were males and 43 females. All patients had angiographic or pathologic confirmation of AV discordance with two ventricles. Patients with situs ambiguous were excluded.

Spontaneous complete AV block (third-degree AV block) was defined as the natural onset of persistent complete AV dissociation with the atrial rate faster than the ventricular rate. Other rhythm abnormalities were described using standardized nomenclature.6

Tricuspid (systemic) valve insufficiency was graded angiographically as trivial, mild, moderate or severe and was judged to be present if at least mild (left atrium opacified without filling the pulmonary veins and cleared quickly after the injection).

The data analysis consisted of nonparametric survival estimated by the method of Kaplan and Meier7 from birth and from the time of our diagnosis. Patients were withdrawn if they had spontaneous onset of persistent complete AV block (the event under study) or surgically induced complete AV block, defined as onset within 30 days of surgery, or if they died from any cause.

The date of last follow-up was taken as the date of the ECG documenting that complete AV block was not present, even when further clinical follow-up information may have been available. The association of discrete variables with survival without AV block was tested with the log-rank test.8 The association of continuous variables and combination of continuous and discrete variables was tested using Cox’s life-tables regression model.9

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Associated ECG Abnormalities

Associated ECG abnormalities in AV discordance, other than complete AV block, were present in 48 patients: first-degree block in 23, right or left bundle branch block in nine, atrial fibrillation in six, second-degree block in six, atrial flutter in two and Wolff-Parkinson-White syndrome in two.

Surgery

Sixty-nine patients had at least one cardiac operation. Complete AV block related to closure of a ventricular septal defect occurred in 12 patients and required early postoperative pacemaker implantation in all 12. Forty-nine patients had an operation with patch closure of a ventricular septal defect. Therefore, the rate of surgical AV block was 24%. The risk is less with current surgical techniques. Six patients developed spontaneous AV block late (more than 30 days) after surgery. The operations included ventricular septal defect closure in two patients, left AV valve replacement in one, systemic-to-pulmonary shunt in one, pulmonary trunk banding in one and exploration in one. Of the 20 survivors of intracardiac surgery alive at follow-up, only one has developed spontaneous complete AV block.

Follow-up and Survival

The median interval for the 62 patients alive at follow-up was 8.7 years. The 10-year survival probability estimated from the date of Mayo Clinic diagnosis was 61%. AV block was not a risk factor for mortality. Of these 62 patients, 74% had been contacted within 1 year of the completion of the study.

Associated Congenital Cardiac Abnormalities

AV discordance was associated with a wide range of other congenital cardiac abnormalities (table 1).

Results

Age Distribution

Because many patients in this study were not followed from birth at our institution, and to allow comparison with previous series, we examined the age distribution at first Mayo Clinic examination for the whole group (median 8.7 years) (fig. 1). The mean age at last follow-up was 23.1 years (range 5–75 years).

Natural-onset Complete AV Block

Natural-onset complete AV block occurred in 23 of the 107 patients and is shown in figure 2, plotted from first Mayo Clinic diagnosis. The risk of complete AV block increased linearly with age at a range of approximately 2% per year. Complete AV block was present at birth in four patients and developed in the other 19 at a mean age of 18.1 years (range 4 months to 53 years). Documented second-degree AV block preceded complete AV block in two patients.

Risk Factors

Tests of the association of risk factors with natural-onset complete AV block were performed for presence or absence of ventricular septal defect, pulmonary stenosis, tricuspid insufficiency, dextrocardia and age at diagnosis. The presence of an intact ventricular septum significantly increased the rate of onset of complete AV block (fig. 3). Twelve of the 25 patients (48%) with intact ventricular septum developed AV block, whereas 11 of the 82 patients (13%) with a ventricular septal defect developed AV block. This finding was also significant when analyzed correcting for age at Mayo diagnosis ($p < 0.03$, beta $-1.28$) and when patients were withdrawn at the time of the first palliative ($p < 0.01$) or corrective ($p < 0.01$) surgery. We could not show that pulmonary stenosis was a significant risk factor when analyzed in combination with other factors. Eleven of 21 patients (52%) with intact septum and no pulmonary stenosis developed complete AV block. There was no detectable effect of dextrocardia, left AV valve insufficiency or age at diagnosis on the risk of complete AV block.

### Table 1. Anatomic Diagnoses Associated with Atrioventricular Discordance in 107 Patients and the Occurrence of Natural-onset Complete Atrioventricular Block

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>No pulmonary stenosis</th>
<th>Pulmonary stenosis</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>VSD</td>
<td>Intact septum</td>
</tr>
<tr>
<td>-----------------------------</td>
<td>-----------------------</td>
<td>--------------------</td>
</tr>
<tr>
<td>Tricuspid insufficiency</td>
<td>11 (2) 12 (6)</td>
<td>11 (2) 1 (0)</td>
</tr>
<tr>
<td>No tricuspid insufficiency</td>
<td>18 (1) 9 (5)</td>
<td>42 (6) 3 (1)</td>
</tr>
<tr>
<td>Levocardia</td>
<td>24 (2) 20 (10)</td>
<td>35 (7) 4 (1)</td>
</tr>
<tr>
<td>Dextrocardia</td>
<td>5 (1) 1 (1)</td>
<td>18 (1) ___________</td>
</tr>
</tbody>
</table>

Numbers in parentheses represents number of patients with natural-onset complete atrioventricular block. Additional abnormalities included atrial septal defect in 11 patients, double-outlet right ventricle in 14, persistent ductus arteriosus in seven, and straddling atrioventricular valve in four. Of the patients with dextrocardia, there were 23 with situs solitus and one with situs inversus.

Abbreviation: VSD = ventricular septal defect.

### Figure 1. Age distribution of patients with atrioventricular discordance and two ventricles (corrected transposition) at the time of Mayo Clinic diagnosis.
Survivorship free of heartblock

Figure 2. Overall survivorship free of atrioventricular block analyzed from diagnosis at the Mayo Clinic.

Treatment

A permanent pacemaker was implanted in nine of the 23 patients with natural onset of complete AV block (39%). The implantation was performed at the onset of complete AV block in four patients and at an average of 11.4 years later in the others (1, 6, 12, 17 and 21 years later). In all but two of these patients, the pacemaker insertion was not associated with any other cardiac surgery. Five of the nine patients had a history of syncope. Six patients have subsequently required at least one pacemaker replacement.

None of the 14 patients who did not have permanent pacemaker implantation had syncope in the follow-up period. One died suddenly at age 28 years while playing tennis, 4 years after the onset of spontaneous complete AV block, with a resting heart rate of 40 beats/min.

Discussion

The spontaneous onset of complete AV block has long been recognized to be a complication of AV discordance. Walmsley reported a case of a 30-year-old man who died with complete AV block and congestive heart failure in 1931 and noted the long course of the AV bundle and fibrosis in the bundle in his patient. The progression from partial to complete AV block in some patients was suggested by Schiebler et al. and was documented in two patients in this study. Several recent reports of late-onset spontaneous complete AV block with syncope in middle age have appeared in the last decade and raised the likelihood that this complication is one which is acquired. Anderson et al. suggested that the fibrosis in the region where the bundle makes contact with the AV node may be due to the excursions of that region of mitral pulmonary continuity.

Our data demonstrated that the spontaneous onset of complete AV block may occur in utero, during infancy, childhood, adolescence or at older ages and that the probability of this complication increased nearly linearly with age. Although previous reports were weighted toward younger patients, the important result from this study is that spontaneous AV block can occur at any time during the natural or unnatural history of AV discordance.

The risk of natural-onset complete AV block with AV discordance was increased if the ventricular septum was intact. We are not aware of anatomic information in AV discordance with intact septum that may help to explain this finding. It cannot be explained by longer follow-up of the patients with intact septum, for this observation was significant when analyzed from birth or Mayo Clinic diagnosis and was not affected by age at diagnosis. If stress or tension related to the AV node is a factor in the development of fibrosis, perhaps these forces are more intense when the ventricular septum is intact than when a ventricular septal defect is present.

Only six of 23 patients with spontaneous complete AV block developed it later after an operation. Late-onset complete AV block as a sequela of cardiac surgery has been reported in other types of congenital heart disease. Based on our limited long-term follow-up, ventricular septal defect closure without surgical AV block did not confer risk of late-onset complete AV block in excess of that expected without surgery. The risk of complete AV block occurring naturally even after an excellent result of definitive surgical repair of associated anatomic abnormalities will require vigilance on the part of physicians caring for these patients, but should rarely impede or delay otherwise indicated surgery.

The management of patients with AV discordance and narrow-complex complete AV block and an adequate heart rate remains controversial. Based on our experience, we favor avoiding pacemaker implantation during childhood if there are no symptoms and the heart rate is over 50 beats/min, as suggested also by Karpawich et al. Sudden death related to complete AV block occurred in one adult patient in this series. This death emphasizes the potential risk of treating complete AV block expectantly despite an adequate heart rate.

We conclude that patients with AV discordance are at increased risk of developing complete AV block...
throughout their lives. Increasingly more sophisticated pacing systems and insertion techniques should allow prompt and safe management of AV block in these patients.

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References
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