Bundle of His Recordings in Congenital Complete Heart Block

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SUMMARY
Six children, aged between 3 and 19 years, with congenital complete heart block without associated cardiac disease were studied by His bundle electrography. All were asymptomatic with resting ventricular rates over 40 beats/min, and electrocardiograms that showed a normal axis and QRS complex. His bundle to ventricular activation was within the normal range in all and the block was proximal to the bundle of His. Ventricular and atrial pacing did not result in retrograde or antegrade conduction. All the patients were thought to have a lesion proximal to the site where the bundle of His potential was recorded and, therefore, were unlikely to develop symptoms or require treatment.

Additional Indexing Words:
QRS frontal vector Ventricular conduction Atrial pacing Isoproterenol
Long-term survival

CONGENITAL complete heart block is a rare but important form of atrioventricular (A-V) block. Although most children with it are asymptomatic and the lesion is compatible with longevity, Stokes-Adams attacks and death do occur. A clinical study is in progress to determine the natural history of this entity, but the mortality in infancy and childhood is estimated at not less than 5%. Most reports do not separate heart block with associated cardiac disease from isolated block with a normal heart, and the prognosis of the block itself is thus difficult to determine.

Complete heart block may occur at three major sites: the A-V nodal region, the bundle of His, or the branch bundles. Experimental and histopathologic studies have confirmed lesions at these sites and have shown that the lower the site of block the worse the prognosis. The electrocardiogram in congenital heart block may reflect the long-term prognosis, and patients can be divided into two groups: (1) those with very slow ventricular rate and widened QRS complex, and (2) those who have normal QRS complex and faster ventricular rate, usually over 40 beats/min. The former group is more likely to have Stokes-Adams attacks.

While the electrocardiogram is helpful in assessment, it does not indicate the site of the block in the conduction system. Recently, catheter recordings of specialized conducting fiber potentials for analysis of the A-V conduction system have become possible. Acquired complete heart block has been...
studied with this technic, and the study has confirmed that the lesion associated with a slow rate and a wide QRS complex is usually below the bifurcation of the bundle of His.12

The purpose of this study was to apply this technic to patients with congenital heart block to show more precisely the site of the block.

Methods

Six children with complete heart block and no other clinical cardiac lesion were studied at right heart catheterization. None was thought to have congenital corrected transposition of the great arteries. The block was thought to be congenital as in four it was found at birth or infancy and in the other two before 3 years. No significant illnesses either in the mothers during pregnancy or in the children during infancy were reported, and at investigation no evidence of disease or cardiac anomaly was found. At the time of study their ages ranged from 3.5 to 19 years (mean, 11.2 years); three patients were female and three male. All were asymptomatic; none had previously experienced dizziness or syncope. The basic electrocardiographic pattern was similar in all (fig. 1); viz, a normal axis, a normal QRS frontal vector, normal QRS complex, complete heart block, and a ventricular rate over 40/min. The atrial rate ranged from 65 to 96 at rest (mean, 84) and the ventricular from 42 to 55 (mean, 47). All patients increased their ventricular rate significantly with exercise.

All patients were fasting when studied. Innovar, 0.04 mg/kg was given intramuscularly 1 hour before catheterization. Routine determinations of right heart pressure, indicator-dilution dye curves, and angiography were employed to exclude associated congenital heart disease. His bundle electrograms with a simultaneous conventional ECG lead were recorded on a Honeywell multichannel recorder at 200 mm/sec. Pacing of the right atrium and ventricle from another catheter at rates up to 160 beats/min was recorded as was the rate after intravenous infusion of isoproterenol, 0.7 μg/min for 2 min.

Table 1

**Electrocardiographic Data**

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Age (yr)</th>
<th>Rate</th>
<th>QRS</th>
<th>His bundle electrogram</th>
</tr>
</thead>
<tbody>
<tr>
<td>122 57 27</td>
<td>3</td>
<td>96</td>
<td>0.04</td>
<td>+70</td>
</tr>
<tr>
<td>141 66 74</td>
<td>5</td>
<td>90</td>
<td>0.05</td>
<td>+90</td>
</tr>
<tr>
<td>137 36 73</td>
<td>9</td>
<td>75</td>
<td>0.06</td>
<td>+70</td>
</tr>
<tr>
<td>104 77 66</td>
<td>15 Birth</td>
<td>94</td>
<td>0.07</td>
<td>+60</td>
</tr>
<tr>
<td>90 79 81</td>
<td>17</td>
<td>65</td>
<td>0.06</td>
<td>+30</td>
</tr>
<tr>
<td>76 05 90</td>
<td>19</td>
<td>80</td>
<td>0.06</td>
<td>+75</td>
</tr>
</tbody>
</table>

*Figure 1*

The electrocardiogram (patient 122 57 77) shows complete A-V dissociation. The atrial rate is 80 and the ventricular 48. The QRS complex is normal.
Results

In all six patients a bundle of His electrogram was recorded. Atrial depolarization (A) was not followed by a His bundle deflection (H) in any instance. The time from the bundle of His deflection to the beginning of ventricular depolarization (H-Q time) was constant in each case and within the normal range (table 1). The H to Q times ranged from 28 to 42 msec (mean, 35.1 msec) (normal, 35 to 45 msec). Examples are shown in figure 2. Atrial activity was not related to ventricular and thus the lesion in all cases was above or in the upper part of the bundle of His.

Right ventricular pacing did not cause retrograde conduction to the atrium in any of the patients. Atrial pacing did not alter ventricular rate and in no instance was any A-V conduction observed. Isoproterenol given intravenously increased the ventricular and atrial rates, but complete heart block persisted.

Discussion

The diagnosis of congenital complete heart block is usually accepted when complete block is demonstrated in fetal life or in infancy. James has shown that considerable change occurs histologically in the junctional region in the early postnatal period and thus occasionally complete block may not be established till then. Although acquired heart block is found most in elderly people, it may rarely occur in children and, if no previous history is available, may be termed "congenital." However, if serial electrocardiograms beyond infancy show progressive disease of the conduction system such as appearance of branch-bundle block, the etiology is probably not congenital as pathologic examination shows the lesion in congenital block is around the A-V node with no degeneration in the branch bundles. In rare instances complete heart block and Stokes-Adams attacks in a young person may, therefore, not be congenital.

Our patients are a small but homogenous group that is representative of the majority of children seen with congenital complete heart block. The major features are an absence of symptoms, a ventricular rate of usually over 40/min that increases significantly with exercise, and a normal QRS complex in the electrocardiogram. A normal QRS complex does not always localize the block above the bundle of His. However, we have shown in all patients that the conduction pathway was interrupted proximal to the site where the His bundle potential was recorded. Similar findings have been obtained by others. The pacemaker thus appears to be junctional rather than ventricular. Interventions such as atrial pacing to rates of 160/min and isoproterenol infusion did not alter the basic conduction defect. The H-Q time in each case was constant, and all were within the normal range. With the technic we are using in this study, it is not possible to determine whether the H potential is from the upper or lower part of the His bundle, and thus pathologically the lesion could be in the superior portion.

Figure 2

His bundle electrogram (HBE 1 and 2) with lead 2 of the standard electrocardiogram (ECG) in same patient as in figure 1. The atrial potentials are independent of H and Q, and H-Q time is 35 msec.

Circulation, Volume XLV, February 1972
of the bundle of His. These electrophysiologic findings agree with the site found histologically in the few patients who have come to autopsy.\textsuperscript{3, 15, 16}

A slow resting atrial rate has been mentioned as a possible factor in prognosis.\textsuperscript{8} However, as conduction is blocked between the A-V node and bundle of His, the inherent rate of the focus that causes ventricular depolarization plus its ability to increase with exercise seems to be more important.

Isoproterenol increased the ventricular rate significantly in all patients studied. To us this is not a useful index in prognosis as the ventricular rate also will increase when the pacemaker is ventricular in origin.

A further type of congenital heart block with a left axis, prolonged QRS, and a very slow rate of less than 40/min has been described.\textsuperscript{5, 14} Death occurs usually in infancy or shortly afterward. No His bundle recordings have been made of this rare form of heart block, but a lesion below the bundle of His in the bundle branches would be expected. Ventricular pacing for such patients would seem to be indicated.

Although the collection of more data, both electrophysiologic and histologic, is required, we tentatively suggest that children who have congenital heart block with no other cardiac lesion, a resting ventricular rate over 40/min which can be augmented by exercise, a normal QRS complex and frontal axis in the electrocardiogram, and a normal His bundle-to-ventricular activation time measured as determined by electrography are less likely to develop Stokes-Adams attacks or other symptoms referable to their block than children with other findings associated with heart block. Infants and children with no associated cardiac disease who died as a result of the heart block all show widened QRS on the electrocardiogram and a slow rate.\textsuperscript{5}

If children with congenital heart block show a conduction defect on the electrocardiogram and have a rate of under 40 beats/min which does not respond to exercise, His bundle recordings should be done. If the lesion is below the bundle of His, ventricular pacing may be advisable to prevent Stokes-Adams attacks.

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Circulation. 1972;45:277-281
doi: 10.1161/01.CIR.45.2.277
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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