Congenital Complete Atrioventricular Block: Problems of Clinical Assessment

By Milton H. Paul, M.D., Abraham M. Rudolph, M.D., and Alexander S. Nadas, M.D.

The diagnosis of congenital complete atrioventricular block usually offers little difficulty. There remains, however, the problem of interpreting certain clinical findings, including systolic and diastolic murmurs and cardiomegaly, which are sometimes falsely suggestive of an associated congenital heart lesion. The clinical, radiologic, and electrocardiographic findings in 27 children with congenital complete heart block have been analyzed in terms of the hemodynamic abnormalities found at cardiac catheterization in 12 of these patients.

The diagnosis of congenital complete atrioventricular block usually offers little difficulty. It requires only electrocardiographic verification of the mechanism of an abnormally slow heart rate that has been recognized in utero or early infancy and has occurred without any known toxic or infectious etiology. There frequently remains, however, the problem of the interpretation of clinical findings that are suggestive of an associated congenital cardiac lesion. Auscultation, in particular, has long been confusing, and it has been a common view that the systolic murmur heard in most patients with congenital heart block is indicative of a ventricular septal defect. Earlier studies also suggested that an underlying structural defect, usually in the ventricular septum, was responsible for the interruption in normal atrioventricular conduction.1,2 More recently, Campbell and Thorne3 and Wood4 have demonstrated that this relationship has undoubtedly been overemphasized.

The purpose of this communication is to present the clinical and hemodynamic findings in a group of children with congenital complete atrioventricular block and to analyze certain perplexing aspects of the clinical profile in relation to the hemodynamic abnormalities.

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Material and Methods

Clinical, electrocardiographic, and radiologic examinations are available in 27 children, observed at the Children's Medical Center during the past 8 years (1949-57). These patients had electrocardiographic evidence of complete heart block without history of associated infection (diphtheria, rheumatic fever, virus) or intoxication (digitalis).

Phonocardiograms with simultaneous electrocardiograms or pulse tracings were obtained on selected patients by means of a dual channel photographic oscillograph. Right heart catheterization according to methods previously described5 was performed in 12 of these patients with special attention directed to the detection of small left-to-right shunts. In 3 patients 2 intracardiac catheters were inserted to provide simultaneous blood sampling from the superior vena cava or right atrium and the main pulmonary artery, and in all patients multiple blood samples were obtained from the superior vena cava, right atrium, right ventricle, and pulmonary artery. In the children on whom ventilatory measurements and oxygen consumption could not be obtained the cardiac outputs were calculated on the basis of an assumed resting oxygen consumption of 180 ml. per minute per M.2

Results

Clinical Profile. The age of these patients (16 male and 11 female) ranged from 5 months to 14 years. In 2 complete atrioventricular block was suspected in utero by the obstetrician. In 12 additional instances the diagnosis was made within the first few months of life and in the remainder, under the age of 4 years.

Two children with definite cyanotic congenital heart disease will not be further discussed,
since in the cyanotic patient with complete congenital heart block the presence of significant congenital heart disease cannot reasonably be doubted. The clinical profile is therefore based on an analysis of the findings in the remaining 25 acyanotic patients, and this group is homogeneous only in terms of the presence of complete atrioventricular block.

Twenty patients had completely normal exercise tolerance and 5 manifested varying degrees of easy fatigability. One of the latter eventually developed congestive heart failure. Respiratory tract infections were reported to be more frequent than usual in 8 patients. It is particularly noteworthy that in this entire group of children, representing approximately 180 patient years of complete heart block, there was no single bona fide episode of an Adams-Stokes attack. Physical development was usually normal although 5 patients were slightly underdeveloped in height or weight.

Jugular venous pulsations showed prominent cannon "a" waves but were otherwise normal. The systolic blood pressure ranged from 85 to 120 mm. Hg (average 110), the diastolic blood pressure from 30 to 70 mm. Hg (average 54), and the pulse pressure from 35 to 100 mm. Hg (average 56).

Auscultation revealed that the first heart sound was frequently variable in intensity from beat to beat; the second heart sound was normally split, and of somewhat increased intensity. A distinct third heart sound was heard at the apex in 12 patients. A fourth heart sound was not heard with certainty but was identifiable in the phonocardiograms (fig. 1).

A systolic murmur of at least grade II intensity was heard in all but 1 of the 25 patients, and in 23 it was grade III or louder. This murmur (figs. 1 and 2) was usually of medium or high frequency, described as rough or blowing, generally occurring in the first two thirds of systole, and was heard best at the apex or lower left sternal border. Transmission was usually toward the apex and mid-left sternal border.

A diastolic murmur was noted in 22 patients, and in 19 it was of grade II or III intensity. The diastolic murmur (fig. 3) consisted of medium or low frequency vibrations, sometimes described as rumbling in quality. It was heard shortly after the second heart sound and was maximal at the apex or lower left sternal border. A third heart
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Fig. 3. Phonocardiogram (apex), same patient as figure 1. Diastolic murmur (DM) with medium and low frequency components starting shortly after second sound. Note the markedly increased intensity of the diastolic murmur in the second beat when atrial contraction (P) coincides with the early diastolic murmur.

sound was sometimes the initial vibration of this diastolic murmur, but more often the third sound appeared to be enveloped by the murmur. Notable in some patients was the variable intensity (fig. 3) of the diastolic murmur from beat to beat.

Radiologic examinations, including fluoroscopy, revealed cardiomegaly in 19 of the 25 acyanotic patients. Left ventricular enlargement was present in 16 patients (fig. 4) and in 11 of these some additional right ventricular enlargement was also evident. Left atrial enlargement was present in 10 patients. In only 3 patients was the pulmonary vasculature considered definitely increased, and in each a left-to-right shunt was subsequently proved at catheterization.

The electrocardiograms revealed an average ventricular rate of 54 per minute (range 42 to 85) and an average atrial rate of 112 per minute (range 74 to 180). A normal QRS axis (0° to 90°) was present in 14 patients, right axis deviation in 4 patients, and left axis deviation in 3 patients. Right atrial hypertrophy, as evidenced by p-pulmonale, was present in 8 children. The QRS complexes were of normal duration (0.04 to 0.08 second) in all but 1 patient who had a pattern of complete left bundle-branch block. Characteristic of the group was some tendency to left ventricular preponderance with a rS pattern and deep S in the right precordial lead (V1) and a qRs pattern with tall R in the left precordial leads (V5 and V6). Only 4 patients had an rsR' complex resembling incomplete right bundle-branch block. Cardiac catheterization was performed in 3 of the latter 4 patients, and in each an atrial septal defect was demonstrated.

Catheterization Studies. Twelve of the 25 acyanotic patients were studied by right heart catheterization because the clinical findings (hyperactive apical impulse, cardiac enlargement, systolic and diastolic murmurs) were initially suggestive of an associated left-to-right shunt. In 8 of these patients, however, no intracardiac shunts were demonstrable. Of the remaining 4 patients, 3 had left-to-right shunts at the atrial level, and 1 at the pulmonary artery level.

The pertinent hemodynamic data for the 8 patients without demonstrable intracardiac shunts are presented in table 1. The pressure tracings from this group indicate normal right atrial mean pressures, slightly elevated right ventricular (30 to 65 mm. Hg) but normal end-diastolic (3 to 8 mm. Hg.) pressures, and slightly elevated mean pulmonary artery (12 to 23 mm. Hg) and mean pulmonary 'capillary' pressures (8 to 14 mm. Hg). Giant 'a' waves (cannon waves) were always recorded in right atrial pressure tracings (fig. 5) if atrial systole occurred at a time when the tricuspid valve was closed. A small (15 mm. Hg or less) systolic pressure gradient across the pulmonary valve was demonstrated in 3 patients.

In all but 1 patient (no. 5) the systemic arteriovenous difference and the calculated systemic blood flow were within the normal resting range. The calculated stroke volume, however, was considerably increased (63 to
### TABLE 1. — Cardiac Catheterization Findings in Eight Patients with Congenital Complete Atrioventricular Block and No Demonstrable Left-to-Right Shunt

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (years)</th>
<th>Ventricular rate (per min.)</th>
<th>Systemic arterial (mm.Hg)</th>
<th>Right ventricle (mm.Hg)</th>
<th>Pulmonary artery (mm.Hg)</th>
<th>Mean Pulmonary &quot;capillary&quot; (mm.Hg)</th>
<th>Arteriovenous Oxygen difference (ml/O2/100 ml)</th>
<th>Oxygen consumption (ml/min./M.2)</th>
<th>Cardiac Index (L./min./M.2)</th>
<th>Stroke Index (ml/M2)</th>
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<tr>
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<td>8/12</td>
<td>45</td>
<td>96/39</td>
<td>36/6</td>
<td>33/11</td>
<td>12</td>
<td>3.7</td>
<td>*</td>
<td>4.9</td>
<td>109</td>
</tr>
<tr>
<td>2</td>
<td>3</td>
<td>42</td>
<td>128/65</td>
<td>65/12</td>
<td>40/19</td>
<td>14</td>
<td>4.3</td>
<td>*</td>
<td>4.2</td>
<td>100</td>
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<tr>
<td>3</td>
<td>5</td>
<td>59</td>
<td>140/55</td>
<td>26/6</td>
<td>23/9</td>
<td>10</td>
<td>3.3</td>
<td>156</td>
<td>4.7</td>
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<td>6</td>
<td>85</td>
<td>122/60</td>
<td>50/5</td>
<td>40/12</td>
<td>10</td>
<td>4.9</td>
<td>182</td>
<td>3.7</td>
<td>44</td>
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<tr>
<td>5</td>
<td>7</td>
<td>62</td>
<td>100/45</td>
<td>40/2</td>
<td>28/5</td>
<td>8</td>
<td>2.9</td>
<td>196</td>
<td>6.8</td>
<td>109</td>
</tr>
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<td>7</td>
<td>67</td>
<td>150/75</td>
<td>33/3</td>
<td>33/4</td>
<td>8</td>
<td>4.3</td>
<td>180</td>
<td>4.2</td>
<td>63</td>
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<tr>
<td>7</td>
<td>7</td>
<td>60</td>
<td>125/55</td>
<td>35/7</td>
<td>35/13</td>
<td>10</td>
<td>3.7</td>
<td>192</td>
<td>5.2</td>
<td>87</td>
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<tr>
<td>8</td>
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<td>28/9</td>
<td>10</td>
<td>4.5</td>
<td>122†</td>
<td>2.7</td>
<td>68</td>
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</tbody>
</table>

* Assumed at 180 ml. O2/min./M.2
† Patient anesthetized.

**Fig. 4.** Representative x-rays of 3 patients with congenital atrioventricular block and no evidence of associated atrial or ventricular septal lesions by cardiac catheterization. Note cardiomegaly and left ventricular dominance. **Top, left.** Patient E.G., 8 months. **Top, right.** Patient S.C., 3 years. **Bottom, left and right.** Left anterior oblique and anteroposterior x-rays of patient A.W., 5 years.
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119 ml. per stroke per M.²), except in 1 patient (no. 4) who had an unusually rapid ventricular rate (85 per minute) during the catheterization procedure. This increase in calculated stroke volume per square meter of body surface area (SV/M.²) was the most consistent hemodynamic abnormality found at right heart catheterization in the patients without demonstrable intracardiac shunts.

DISCUSSION

Children with congenital complete atrioventricular block can present perplexing diagnostic and prognostic problems. In a few individuals an obvious cardiac defect such as cyanotic congenital heart disease or patent ductus arteriosus can be diagnosed readily by clinical methods. In others the findings are so minimal as to indicate at once the absence of any significant associated congenital heart disease. In a considerable number of patients, however, the clinical findings are highly suggestive of an associated intracardiac left-to-right shunt, specifically a ventricular or atrial septal defect, without being completely diagnostic of such. Right heart catheterization and long-term follow-up studies have failed to substantiate the clinical diagnosis of a septal defect in a considerable number of this latter group.

The present study suggests that certain clinical findings in complete heart block might be related directly to the altered hemodynamics associated with the slow ventricular rate. It would appear that the most significant physiologic consequences of the slow cardiac rate are the increased stroke volume, and the increased end-diastolic heart volume. The former may be responsible for some of the auscultatory findings and the latter for the frequently observed cardiomegaly.

A systolic murmur is almost always present in congenital complete heart block and its presence suggests the diagnosis of a ventricular or atrial septal defect in children. This murmur may equally well be explained by the large stroke volume and the associated high ejection velocity of the blood flow in complete heart block.⁶ ⁷

Phonocardiograms in our patients without demonstrable left-to-right shunts indicate that the systolic murmur is often restricted to the first half of systole and is high in frequency. These characteristics are suggestively similar to a form of "innocent systolic murmur" described by Wells⁸ and can be related to increased turbulence in the blood stream at the time of rapid increase in the rate of blood flow. This murmur can be classified with the systolic murmurs heard in high cardiac output states, i.e., severe anemia, thyrotoxicosis, pregnancy, and the basal systolic murmur in atrial septal defects ("relative pulmonary stenosis").

A loud apical diastolic murmur was heard in 20 of the 25 children studied including each of the 8 patients who had no shunts demonstrated on right heart catheterization. The murmur (fig. 6) occurs early in diastole and is synchronous with the period of rapid ventricular filling. When the atria contract during this period, a summation effect can occur with a resultant increase in the intensity of the murmur (fig. 3).

The mechanism of this diastolic murmur in complete heart block may again be related to a slow cardiac rate and the resultant increased stroke volume. Although the total diastolic period is prolonged in bradycardia, the duration of the rapid ventricular filling
Fig. 6. Phonocardiogram and apexcardiogram from a patient with no demonstrable left-to-right shunt illustrating a low-frequency diastolic murmur beginning almost immediately after the second sound and enveloping a third heart sound during the period of rapid ventricular filling.

The period is relatively constant over a wide range of heart rates. In 5 patients with congenital complete heart block (mean ventricular rate 42 per minute) the duration of the rapid ventricular filling period as estimated from the phonocardiogram (S₂ to S₃ interval) was 0.12 to 0.14 second. In 4 patients with tachycardia (mean ventricular rate 130 per minute) this period ranged from 0.11 to 0.13 second.

In complete heart block the abnormally large stroke volume that traverses the atrioventricular valves during early diastole may result in a high blood flow velocity, increased turbulence, and a diastolic murmur. A similar hypothesis of increased blood flow traversing a normal valve orifice has been advanced as the mechanism of the apical diastolic rumble in large left-to-right shunts ("relative atrioventricular valve stenosis").

Rytand\(^9\) has described a diastolic apical murmur related to atrial activity in elderly patients with heart block. In the adult patients, as in the children reported here, the murmur was loudest early in diastole whenever atrial systole more or less coincided with the rapid diastolic filling period. It was postulated that after the atrioventricular valves have been floated nearly together by early diastolic ventricular filling they constitute a relatively narrowed orifice for the continuing and accelerated (atrial contraction) transvalvular blood flow. The murmur disappears or is replaced by short atrial contraction sounds in late diastole (fig. 3) because the transvalvular blood flow has markedly decreased.

It should be clearly appreciated that other possible origins of these murmurs have not been definitely excluded even in those patients with essentially normal findings on right heart catheterization. Organic mitral valve lesions, possibly in association with endocardial fibroelastosis, are not completely excluded even though the pulmonary "capillary" pressures were only minimally elevated. Furthermore, a small septal defect could possibly not be detected because of the limitation of oxygen saturation methods in measuring small left-to-right shunts.

The radiologic and electrocardiographic findings in children with congenital complete heart block must similarly be interpreted with caution in regard to the diagnosis of an associated intracardiac lesion. In our study slight or moderate cardiomegaly was a frequent finding, occurring in 19 of the 25 acyanotic patients. It is general clinical experience that cardiac enlargement is common with bradycardia in adults,\(^10\) and thus cardiomegaly might be expected in complete heart block even in the absence of any associated congenital heart lesion. It has also been demonstrated that experimental induction of complete heart block in the dog\(^11\) frequently results in generalized cardiac enlargement and myocardial hypertrophy. Fortunately, the status of the pulmonary vasculature provides a relatively reliable finding in assessing this group for associated congenital heart lesions. The pulmonary vasculature was described as normal by the roentgenologist in each of the 8 patients without demonstrable left-to-right shunts on catheterization, whereas 3 of the 4 patients with left-to-right shunts had radiologic evidence of pulmonary vascular engorgement. The electrocardiogram is invaluable in establishing the diagnosis of congenital complete heart block. In addition considerable information can probably also be derived from the QRS complex, despite the fact that the conduction pathway is not intact. In contrast to acquired complete heart block in adults where over half of the
patients have idioventricular QRS complexes,12 the electrocardiogram in congenital complete heart block almost always has a QRS complex that is supraventricular in form.3 In 24 of our 25 acyanotic patients the QRS complexes were of normal duration (0.04 to 0.08 second) and supraventricular in form. This finding would indicate that the dominant impulse center is located above the bifurcation of the bundle of His, and that the excitatory process propagates through the ventricular myocardium in a relatively normal sequence. Under these circumstances the usual criteria for ventricular hypertrophy should be applicable. Right ventricular hypertrophy was present in only 1 of the 25 acyanotic patients. Left ventricular hypertrophy was noted in 8 patients; cardiac catheterization in 3 of these did not disclose any significant associated congenital heart lesion. Incomplete right bundle-branch block was diagnosed in 4 patients; catheterization in 3 of this group demonstrated an atrial septal defect. These electrocardiographic data, although limited, suggest that left ventricular hypertrophy patterns are not uncommon in congenital complete heart block and that incomplete right bundle-branch block might possibly indicate the presence of an associated septal defect.

Summary

The clinical, radiologic, and electrocardiographic features of congenital complete atrioventricular block have been reviewed in 27 infants and children, and cardiac catheterization was performed in 12 of them.

In this group certain clinical findings including a hyperactive apical impulse, prominent systolic and diastolic murmurs, and cardiomegaly are frequently suggestive of an associated congenital cardiac lesion with a left-to-right shunting of blood. Eight of the 12 patients studied by cardiac catheterization had no demonstrable congenital cardiac lesion except complete heart block, despite the presence of the above findings. In the remaining 4 patients with left-to-right shunts demonstrated by cardiac catheterization the only reliable sign of the associated congenital cardiac lesion was the radiologic demonstration of an engorged pulmonary vasculature.

It is suggested that the difficulty in interpreting certain auscultatory findings in patients with complete congenital heart block is related to the large stroke volume associated with the slow heart rate. This results in a high velocity of blood flow through the semilunar and atrioventricular valves and can produce murmurs suggesting functional stenosis. The large stroke volume is also associated with a large end-diastolic heart volume, probably leading to cardiomegaly.

Since long-term studies indicate that the prognosis in complete congenital heart block is dependent primarily on the nature of the associated congenital cardiac lesion that may be present, it is important to establish a correct anatomic diagnosis of any septal defect with a view to surgical correction when indicated.

Summario in Interlingua

Le aspectos clinic, radiologic, e electrocardiographic de congenite bloco atrioventricular complete esseva revidite in 27 infantes e juveniles. Catheterisation cardiac esseva interprendite in 12 de illes.

In iste grupo certe constitutiones clinic—incluse un hyperactive impulso apical, prominente murmures systolico e diastolic, e cardiomegalia—es frequentemente indicios de un associate lesion cardiac congenite con derivation sinistro-dextere de sanguine. Octo del 12 patientes studiate per catheterisation cardiac habeva nulle congenite lesion cardiac excepte le complete bloco cardiac, ben que le supra-listate indicios esseva presente. In le altere 4 patientes con derivationes sinistro-dextere demonstrate per catheterisation cardiac, le sol convineente signo de un associate lesion cardiac congenite esseva le demonstration radiologic de un turgide vasculatura pulmonar.

Es exprimite le opinion que le difficultate del interpretation de certe constatationes auscultatori in patientes con congenite bloco cardiac complete es relationate al grande volumine per pulso que es associate con le basse frequentia cardiac. Isto resulta in un alte
velocitate del fluxo de sanguine a transverso le valvulas semilunar e atrioventricular e pote producir murmures que pare indicar stenosis functional. Le grande volumine per pulso es etiam associate con un grande volumine cardiac termino-diastolic, e isto resulta probablemente in cardiomegalia.

Proque studios a longe vista indica que le prognose in congenite bloco cardiac complete depende primarimente del natura del possiblemente associate lesion cardiac congenite, il es importante establir un correcte diagnose anatomic con respecto al presentia de un defecto septal e corriger lo chirurgicamente si un tal manovra es indicate.

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J. ARTHUR THOMSON
British: professor of natural history, editor and author; 1861-1933

In any scientific inquiry the first step is to get at the facts, and this requires precision, patience, impartiality, watchfulness against the illusions of the senses and the mind, and carefulness to keep inferences from mingling with observations. The second step is accurate registration of the data. A third step is arranging the data in workable form. The fourth step is when a whole series of occurrences is seen to have a uniformity, which is called their law.—The Outline of Science, Vol. 4, p. 1165. From Great Companions. Readings on the Meaning and Conduct of Life from Ancient and Modern Sources. Vol. I, Boston, The Beacon Press, 1952.
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