Heart Block after Repair of Ventricular Septal Defect in Children

By Ronald M. Lauer, M.D., Patrick A. Ongley, M.B.Ch.B., James W. DuShane, M.D., and John W. Kirklin, M.D.

The close anatomic relationship of the conduction system of the heart to most ventricular septal defects makes the occurrence of complete heart block a potential risk in the repair of these lesions. In our experience, when complete heart block has developed, it has often proved to be a serious complication. Although reversion to sinus rhythm occurs in the early postoperative period in many patients, others have persistent heart block; some of these die either in the immediate postoperative period or at some time after dismissal from the hospital. Of those who survive with complete heart block, some may be symptom-free but a few are troubled with frequent and potentially lethal Stokes-Adams attacks.

The purpose of this paper is to review an experience with complete heart block following the repair of ventricular septal defect and ventricular septal defect associated with infundibular or valvular pulmonary stenosis or with both in the pediatric age group. The data concerning adult patients are not analyzed as yet. We will mention certain changes in surgical technic which seem to have affected the incidence of heart block and will discuss briefly immediate and long-term therapy.

Material and Methods

From March 1955 to April 1959, 174 patients with ventricular septal defect and 124 patients with ventricular septal defect associated with infundibular or valvular pulmonary stenosis were referred from the Section of Pediatrics for open intracardiac repair. The group of patients designated as "having ventricular septal defect" includes those with isolated ventricular septal defect and those with ventricular septal defect associated with various intracardiac or extracardiac lesions other than pulmonic stenosis. Patients with ventricular septal defect as part of a common atrioventricular canal have been excluded from this study as also have patients with ventricular septal defect associated with corrected transposition of the great vessels. The group of patients described as having ventricular septal defect associated with infundibular or valvular pulmonary stenosis includes those with tetralogy of Fallot as well as those with ventricular septal defect in whom the pulmonic stenosis and the arrangement of the great vessels are not those usually recognized as typical for tetralogy of Fallot.

Included in this study were 94 male and 80 female patients with ventricular septal defect. Their ages ranged from 21/2 months to 15 years. Seventy-nine boys and 45 girls had ventricular septal defects associated with infundibular or valvular pulmonary stenosis or with both. Their ages ranged from 6 months to 15 years.

Three types of surgical technics were employed for the 298 patients included in this study: 1. From March 1955 to July 1956, open intracardiac repair was carried out on the beating heart. 2. From July 1956 to the present, induced asystole has been a standard part of the procedure. During the period from July 1956 to April 1958, ventricular septal defects were closed either by the placement of a prosthetic sponge or by direct suture. 3. In April 1958, the method of repair of the ventricular septal defect was altered by the introduction of a direct suturing technic designed specifically to avoid the conduction system.

The surgical details of perfusion,6,7 the methods of repair of infundibular and valvular pulmonary stenosis,8 and of ventricular septal defect prior to the introduction of the present suturing technic6 have been described previously. The present suturing technic is to be described in a subsequent communication. An artificial pacemaker10 was available to all patients in the series, in early cases with an external electrode and in later cases with an internal electrode.11
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The records of the 298 patients have been reviewed with reference to incidence and to some factors affecting the occurrence of postoperative complete heart block. Data concerning the frequency of reversion to sinus rhythm, and death or survival following the development of complete heart block have been obtained.

Intracardiac pressure was measured either by cardiac catheterization prior to operation or by direct needle puncture at the time of operation.

Remarks concerning symptoms are based on examination while in the hospital or at follow-up visits, and from letters received from the parents or referring physicians since dismissal from our care.

Over-all incidence of complete heart block may not be so important a consideration as the incidence of permanent heart block, that is, heart block present at death or dismissal. Permanent heart block is more likely related to sutures, while temporary heart block may be related to administration of digitalis, sequelae of cardiac anoxia, or to other temporary causes. Therefore, the incidence of block persisting until the time of death or dismissal will be stressed, for temporary heart block is rarely a serious problem when properly treated.

Results

The data in tables 1 and 2 indicate no difference in the over-all incidence of complete heart block at death or dismissal from the hospital between the patients operated on for ventricular septal defect and those operated on for ventricular septal defect associated with infundibular or pulmonic stenosis, as the incidence was approximately 10 per cent of each group. These tables also relate the incidence of complete heart block at death or dismissal to each of the 3 surgical technics previously mentioned. With the introduction of asystole, this incidence increased in patients with ventricular septal defect (table 1), but more recently with use of the present suture method it has fallen to 5 per cent, nearly as low an incidence as that obtained after operations on the beating heart. Among patients with ventricular septal defect and infundibular or pulmonic stenosis the incidence of complete heart block at death or dismissal changed very little when asystole was introduced, but has diminished with utilization of the present technic (table 2).

No relationship was demonstrated between the incidence of heart block and age. The ages of patients who had complete heart block ranged from 7 months to 11 years. There were 30 boys and 18 girls. In those cases of ventricular septal defect and complete heart block in which postoperative intracardiac pressures were available, right ventricular pressures showed an appreciable fall in most instances (fig. 1).

The relationship between the incidence of postoperative complete heart block and the ratio of preoperative pulmonary artery pressure to systemic systolic pressure is shown in figure 2 for 158 patients with ventricular septal defect for whom data concerning pressures were available. The over-all incidence of postoperative complete heart block was 3 per cent among patients with ratios of less than 0.50, 10 per cent among those with ratios from 0.50 to 0.74, and 30 per cent among those with ratios of 0.75 or more. The incidence of permanent heart block was, of course, lower than this incidence, being for example 21 per cent among the total cases with ratios of 0.75 or more. It can be seen that the over-all incidence of complete heart block was the highest in those patients with severe pulmonary hypertension.12
Table 1

Ventricular Septal Defect

<table>
<thead>
<tr>
<th></th>
<th>Total patients operated on</th>
<th>Complete heart block at some time</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>Total</td>
<td>Reverted to sinus rhythm</td>
</tr>
<tr>
<td></td>
<td>No.  %</td>
<td>No.  %</td>
</tr>
<tr>
<td>Before asystole:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>March 1955—July 1956</td>
<td>38</td>
<td>4 11</td>
</tr>
<tr>
<td>Asystole:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>July 1956—April 1958</td>
<td>81</td>
<td>20 25</td>
</tr>
<tr>
<td>Asystole and new suture technic:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>April 1958—April 1959</td>
<td>55</td>
<td>9 16</td>
</tr>
<tr>
<td>Total</td>
<td>174</td>
<td>33 19</td>
</tr>
</tbody>
</table>

*Patients who were dismissed from the hospital with complete heart block. The patients who died subsequent to dismissal are included in this group.

Table 2

Ventricular Septal Defect Associated with Infundibular or Ventricular Pulmonic Stenosis or Both

<table>
<thead>
<tr>
<th></th>
<th>Total patients operated on</th>
<th>Complete heart block developed</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Total</td>
<td>Reverted to sinus rhythm</td>
</tr>
<tr>
<td></td>
<td>No.  %</td>
<td>No.  %</td>
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<tr>
<td>Before asystole:</td>
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</tr>
<tr>
<td>March 1955—July 1956</td>
<td>20</td>
<td>3 15</td>
</tr>
<tr>
<td>Asystole:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>July 1956—April 1958</td>
<td>50</td>
<td>7 14</td>
</tr>
<tr>
<td>Asystole and new suture technic:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>April 1958—April 1959</td>
<td>54</td>
<td>5 9</td>
</tr>
<tr>
<td>Total</td>
<td>124</td>
<td>15 12</td>
</tr>
</tbody>
</table>

*Patients who were dismissed from the hospital with complete heart block. Patients who died subsequent to dismissal are also included in this group.

In the group of patients in whom data on pressure were available, 60 per cent had ratios of pulmonary artery to systemic systolic pressure of 0.75 or more in the period before asystole, 49 per cent in the period of asystole with the original suture technic, and 56 per cent in the most recent period with the present suture technic. Thus the fall in the incidence of permanent complete heart block with the introduction of the new suture technic cannot be attributed to the fact that more patients selected for operation in the most recent period have had low pressure ratios.

The fate of the 33 patients with ventricular septal defect who had complete heart block at any time is shown in table 1. In 14 of these
(43 per cent) normal sinus rhythm returned; 10 (30 per cent) died with complete heart block, and 9 (27 per cent) were dismissed from the hospital with complete heart block. Complete heart block developed in 15 of the 124 patients operated on for ventricular septal defect associated with pulmonic stenosis (table 2). In 4 of these (27 per cent) normal sinus rhythm was resumed; 8 (53 per cent) died with complete heart block, and 3 (20 per cent) were dismissed from the hospital with complete heart block.

The time of reversion to normal sinus rhythm is shown in figure 3. This occurred on the second to sixteenth postoperative day in 14 patients with ventricular septal defect, and from the second to the twentieth postoperative day in 4 patients with ventricular septal defect and pulmonic stenosis.

The mode of death, which was apparently a direct result of complete heart block, was sudden cardiac asystole refractory to the electric pacemaker. The day of death in those patients with complete heart block who succumbed during the immediate postoperative period is shown in figure 4. Most deaths occurred in the immediate postoperative period. In some patients who died with complete heart block, bradycardia or asystole did not seem to be the sole determining factors resulting in death. Three who died in the immediate postoperative period had respiratory complications and two late deaths occurred, one following a febrile illness, and one following a second operation.

Heart block developed in the operating room in 37 patients, within a few hours of the completion of the operation in 4 and on the first postoperative day in 4. The latest development of heart block was on the eighth postoperative day. Since in the great majority of our patients the heart block developed at the time of operation, we do not have enough cases of delayed block to allow statistical analysis; so far, however, there is no obvious difference in the survival rates or in the percentage of those with reversion to normal sinus rhythm in the two groups.

Course after Dismissal

Twelve patients (cases 1 to 12, fig. 5) in this study were dismissed from the hospital with complete heart block. Three of these died subsequently. The first death was that of a girl (case 12, fig. 5) with ventricular septal defect repaired at the age of 5 years in 1957. Complete heart block developed in the operating room. In the early postoperative period frequent Stokes-Adams attacks occurred. On the twenty-fourth postoperative day sinus rhythm developed but continued to alternate with 2:1 heart block until she left the hospital. The postoperative course was otherwise uneventful, and she was discharged from the hospital without symptoms. On the fifty-fifth postoperative day death occurred suddenly at home.

In the second patient (case 6, fig. 5) a 6-year-old boy with ventricular septal defect and mild infundibular pulmonic stenosis repaired in 1957, complete heart block developed in the operating room. Frequent Stokes-Adams attacks occurred in the hospital after opera-
tion and occasionally after dismissal. About 1½ years after operation a febrile illness developed and death occurred suddenly.

The third patient (case 11, fig. 5) was a girl, 6 years old in early 1958, when a repair of tetralogy of Fallot was made by the use of a prosthesis in the ventricular septal defect and plastic reconstruction of the right ventricular outflow tract and pulmonary artery. Complete heart block occurred in the operating room. However, she had no symptoms of bradycardia, either in the hospital or after dismissal. In the early postoperative period a systolic murmur, which was thought to be the result of an incomplete repair of the ventricular septal defect, was noted at the lower left sternal border. After dismissal the murmur became progressively louder and roentgenograms of the thorax showed progressive dilatation of the pulmonary artery in the region of the prosthesis. Ten months after the first procedure an aneurysm in the region of the reconstructed outflow tract was repaired and the ventricular septal defect was repaired again. The patient died shortly after this operation.

Three patients among the 9 surviving with complete heart block have had symptoms of heart block since they left the hospital. One (case 4, fig. 5) was a boy aged 2 years in 1957 when he was operated on for ventricular septal defect. Heart block developed in the operating room. Both in the early postoperative period and after discharge frequent Stokes-Adams attacks occurred. Because of the frequency of these spells it was necessary to readmit him for observation and adjustment of therapy. For the last 14 months he has had no Stokes-Adams attacks, but still complains of headache or chest pain when his pulse rate becomes slower than 30 per minute. He is given isoproterenol (Isuprel) hydrochloride\textsuperscript{12} daily.

In the second patient, a girl (case 5, fig. 5), whose ventricular septal defect was repaired at the age of 2 years in 1957, complete heart block also occurred in the operating room. In the postoperative period the rhythm varied from normal sinus to complete heart block, often changing abruptly from the one to the other. When the rhythm changed from normal sinus to heart block, Stokes-Adams attacks would occur, requiring the use of the electric pacemaker. The child was readmitted to the hospital 2 months after dismissal because of frequent spells. She was digitalized and has since remained in complete heart block. Following her second discharge, Stokes-Adams attacks continued for about 2 weeks and then ceased. For the past 15 months, her pulse rate has been around 50 per minute and she appears to be in good health.

The ventricular septal defect of the third surviving patient (case 1, fig. 5) who has had symptoms after dismissal was repaired at the age of 3 years in 1957. Two Stokes-Adams attacks occurred during postoperative hospitalization, and another during the first month after dismissal. For the first 6 months following operation congestive heart failure persisted. Since then he has been without symptoms, fully active, with a pulse rate of about 40 beats per minute.

The remaining 6 patients (cases 2, 3, 7 to 10, fig. 5) who survive with complete heart block have no symptoms from bradycardia and no periods of asystole.

**Surgical Technics and Discussion**

During the period when ventricular septal defects were repaired while the heart was
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beating, the incidence of complete heart block present at death or dismissal was low. This is attributed to the fact that the beating heart allowed observation of the effect of each suture on ventricular rhythm. If block occurred, the offending suture was removed and replaced so that conduction would not be permanently affected. In some instances, however, the complete heart block persisted many days even when the stitch had been immediately removed. Although the introduction of induced asystole allowed a higher incidence of accurate closure of the defects, the incidence of permanent heart block increased from 3 to 19 per cent among patients operated on for ventricular septal defect. Others\textsuperscript{14} have had similar experiences. Although not all would agree, it is our belief that this increase was due to interference with the conduction bundle which passed unnoticed with the heart at rest. Paradoxically, the increase in incidence of heart block on changing from the beating heart technic to asystole was not seen in the patients operated on for ventricular septal defect associated with pulmonic stenosis.

In order to minimize the occurrence of postoperative permanent heart block and to continue using asystole because of the more accurate repair obtainable, the present suture technic was evolved specifically to avoid the conduction system. Since adoption of this technic, the incidence of permanent heart block at death or dismissal has fallen from 19 to 5 per cent in patients with ventricular septal defect; and from 12 to 6 per cent in patients with ventricular septal defect and infundibular or pulmonary valvular stenosis. It is apparent that none of the technics utilized has eliminated the occurrence of permanent heart block. This is not surprising, in view of the probable anatomic variations in distribution of the conduction system and human frailties in application of surgical technics. Also unforeseen complications, such as spread of hemorrhage from a suture site into conduction tissue,\textsuperscript{15} are always possible. Yet a low incidence (5 to 6 per cent) of permanent heart block, present at death or dismissal, has been attained.

\begin{center}
\textbf{Figure 4}
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Postoperative day of death of 18 patients with complete heart block.

\textbf{Treatment of Complete Heart Block}

\textbf{Early Postoperative Period}

Unlike congenital heart block,\textsuperscript{16–19} which is frequently asymptomatic, heart block resulting suddenly from the surgical repair of a ventricular septal defect may cause serious symptoms. These vary in severity from a feeling of discomfort and restlessness to Stokes-Adams attacks characterized by unconsciousness, seizures, and occasionally sudden death. Likewise it appears likely that in the postoperative patient, cardiac output and coronary blood flow may be adversely affected by a very slow heart rate, possibly with fatal sequelae.

During the 4-year period covered by this study, several items have been altered in the immediate postoperative care of patients. Thus it is not possible to establish what is the optimal treatment from a review of the methods used. Certain points that seemed important have been incorporated into a routine of treatment that appears rational and is usually effective.

When complete heart block persists for more than a few minutes after the heart is restarted in the operating room, a wire electrode is imbedded into the right ventricle over the outflow tract and an external electrode is inserted into the skin of the chest wall.\textsuperscript{11} The electrode wires are connected to an appropriate electric pacemaker. The units
supplied by the Electrodyne Co., Norwood, Massachusetts, and by Medtronic, Inc., Minneapolis, Minnesota, have been used with satisfaction. The heart rate is adjusted to a level approximating that seen postoperatively in patients of the same age as the child being treated. The amplitude of the stimulus is adjusted until an adequate response occurs with each electric impulse.

When complete heart block first occurs in the postoperative period, external electrodes applied to the chest wall are used, or an internal myocardial wire is placed percutaneously and used as just described.

A patient whose heart is being driven by a pacemaker or has been driven by a pacemaker recently is watched continuously for a number of days by special nurses who also watch an oscillographic electrocardiographic monitor. If the heart is not responding to the pacemaker, the amplitude of the stimulus is increased until an adequate cardiac response occurs. A patient whose heart is no longer under pacemaker control is put back immediately on it by the nurse if an indication arises.

The heart is driven by the electric pacemaker continuously for 72 hours after operation unless the electrocardiograms show that the heart is in sinus rhythm. When reversion to sinus rhythm is suspected, the rate of the pacemaker is cautiously diminished to observe the rhythm of the patient's heart. If there is sinus rhythm, use of the electric pacemaker is discontinued, but the electrodes are left in place so that the pacemaker can be made to function immediately if heart block again develops. Isoproterenol is not administered while the patient's heart is being driven by the pacemaker.

When the heart has been driven by the electric pacemaker for 3 to 4 days, and the block is still present, the pacemaker rate is cautiously reduced to see whether the heart itself can maintain an adequate idioventricular rate. If the rate is adequate, the electrodes are left in place and the patient is observed closely for signs of bradycardia, systemic hypotension, and inadequate cardiac output. It is important that the patient never be left alone during this period.

If the heart rate is inadequate to maintain the patient when the electric pacemaker is stopped, it is turned on immediately and used for another 24 hours. On the following day the patient is given isoproterenol by rectum about a half an hour before the rate of the pacemaker is decreased again. This procedure is repeated daily until the heart rate stabilizes at an adequate rate.

For the patients who continue in complete heart block after the pacemaker is stopped, isoproterenol is administered by rectum every 2 to 6 hours if necessary to maintain a stable and adequate ventricular rate. In some children 40 to 50 beats per minute is an adequate rate, while others are uncomfortable unless the rate is somewhat faster. A slow heart rate is better tolerated 10 to 14 days after operation than it is 4 to 5 days after it.

Isoproterenol hydrochloride is supplied in 10 and 15 mg. tablets for sublingual use. We have given these tablets by rectum to children in the following dosage: 2.5 mg. for infants and small children, 5 to 15 mg. for larger children. Prior to insertion into the rectum the nurse checks the time of the last bowel

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Figure 5
Course to May 1959 of 12 patients who had complete heart block at time of dismissal from the hospital.
movement. If there has been no recent evacuation, the child is given a glycerine suppository to cause evacuation of the rectum before the medication is administered.

**Long-Term Treatment**

Patients who still have complete heart block at dismissal but do not have symptoms or unfavorable signs related to it are not maintained on special treatment. For those patients with symptoms treatment has been a matter of trial of various drugs that have the properties of increasing the rate and maintaining the stability of idioventricular rhythm, namely isoproterenol and ephedrine. We have also tried atropine by parenteral administration in several patients with disappointing results. The use of digitalis is apparently not contraindicated under these circumstances and has seemed to be beneficial at times.

For patients in whom the bradycardia and asystoles recur in a pattern, isoproterenol is administered about half an hour prior to the time of expected trouble. Oral administration of the drug is effective for some children, although for others rectal administration is better.

It is important to stress the value of careful, patient management of these children. Although the threat of sudden death may always be present, some children with serious symptoms in the early months after operation eventually stabilize and become well adjusted to a slow ventricular rate.

**Summary**

From March 1955 to April 1959, 174 patients with ventricular septal defect and 124 patients with ventricular septal defect associated with pulmonic stenosis were referred from the Section of Pediatrics of the Mayo Clinic for open intracardiac repair.

Forty-eight of these 298 patients had complete heart block at some time following operation. Eighteen of the patients with heart block died in the immediate postoperative period, 18 had reversion to normal sinus rhythm, and 12 were discharged from the hospital with complete heart block. Three of these 12 patients died subsequently. Two died presumably as a direct result of complete heart block and a third died following a second open cardiac operation. Of the 9 surviving patients, 3 have had serious bradycardia or periods of asystole and 6 have no symptoms from heart block.

One of 3 surgical technics was employed for all the patients in this study with variation in incidence of block with the use of each technic. Other factors that might relate to the incidence of heart block are discussed.

The management of the patient with complete heart block both in the immediate postoperative period and after dismissal from the hospital is described.

**Addendum**

From April 1959 to April 1960, 90 children with ventricular septal defect and 75 with tetralogy of Fallot were operated on. The suture technic was the same as that used from April 1958 to April 1959.

Of the 90 children with ventricular septal defect, 6 (7 per cent) had complete heart block at some time during the postoperative period, and normal sinus rhythm returned to 5 of these while they were still in the hospital, leaving only 1 patient (1 per cent) with complete block at death or dismissal. Of the 75 patients with ventricular septal defects and pulmonic stenosis, 10 (13 per cent) had heart block at some time during the postoperative period and 6 (8 per cent) were still in block at death or dismissal.

In addition to the 3 patients who died in block following dismissal, reported in the main paper, one other died at Christmastime, 1959, while sitting at the dinner table. This means that 4 of the 12 patients originally discharged in block have now died.

**Summario in Interlingua**

Inter martio 1955 e April 1959, 174 patientes con defectos ventriculo-septal e 124 patientes con defectos ventriculo-septal in association con stenosis pulmonic esseva destinate per le Section de Pediatria del Clinica Mayo al reparo per un aperte operation intracardiac.

Quaranta-octo de iste 298 patientes experienciava un complete bloco cardíac a un tempore o un altere post le operation. Deec-octo del patientes con bloco cardíac moriva immediatemente post le operation; 18 reverteva a normal rhythmos sinusal; e 12 esseva dimittite ab le hospital con complete bloco cardíac. Tres de iste 12 moriva subsecuentemente. Duo moriva presumitamente como resultato directe del complete bloco cardíac, e un tertie moriva post un secunde aperte operation cardíac. Inter le 9 patientes vivente, 3 ha manifestate serie formas de bradycardia o periods de asystole, e 6 ha monstrate nulle symp-tomas atribuibile al bloco cardíac.
Un de 3 differente technicas chirurgic esseva em
pleate pro omne le patientes in iste studio. Le
incidentia de bloco cardiac differeva secundo le tech
nica usate. Altere factores que es possibilemente relationate al incidentia de bloco cardiac es discutite.
Es descritte le programma de tractamento pro
patientes con complete bloco cardine, tanto durante le periodo immediatemente post le operation como etiam al tempore subsequente al dimission ab le
hospital.

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