Endocardial Cushion Defect
Preoperative and Postoperative Survey

By Lewis P. Scott, Lt. Comdr. MC USN, Anna J. Hauck, M.D., Alexander S. Nadas, M.D., and Robert E. Gross, M.D.

The repair of atrial septal defects was first undertaken over a decade ago.1-4 Although secundum-type defects could be closed adequately without the use of cardiopulmonary bypass, it soon became apparent that defects of the endocardial cushion type were not amenable to correction by indirect techniques.5 Accordingly, the need arose to differentiate these two lesions; the clinical, electrocardiographic, and hemodynamic characterization of each emerged subsequently.6-10 With the advent of cardiopulmonary bypass the surgeon has been afforded adequate visualization and the necessary time for repair of endocardial cushion defects. Several reports have appeared in the literature evaluating the immediate results of such correction.11-14 However, no appraisal of long-term results of repair of endocardial cushion defects has been published.

The purpose of this presentation is to report the results of surgical correction of endocardial cushion defects at The Children's Hospital Medical Center in 44 patients followed for at least 1 year postoperatively.

Materials and Methods
Forty-four patients with endocardial cushion defect underwent corrective surgery with cardiopulmonary bypass during the period of June 1, 1957, to January 1, 1960. The indication for correction was congestive failure, significant cardiac enlargement, or the presence of symptoms such as exertional dyspnea and exercise intolerance.

Preoperative physical examinations were performed by at least one of the authors. Fluoroscopic evaluations, x-rays, and electrocardiograms were obtained in all patients. Phonocardiograms and vectorcardiograms were taken in several patients. Cardiac catheterization and cineangiography were performed in 43 of the 44 patients.

Cardiopulmonary bypass was carried out according to techniques previously outlined.15 The pump oxygenator consisted of a DeBakey-type pump and a Kay-Cross disk oxygenator. Perfusion was maintained at 2300 to 2500 ml/min./M.2 under normothermic conditions. The perfusion time varied from 39 to 50 minutes. Effective cardioplegia with potassium citrate was used in 17 patients for periods of 10 to 50 minutes, with an average of 37 minutes.

Repair of the defects was carried out through the right atrium in all patients. Clefts in the mitral and tricuspid valves were sutured with silk; care was taken to approximate only the free margins of the valves. A compressed Ivalon patch was sewn into the defective septum with a continuous silk suture, except in the region of the conduction bundle where interrupted sutures were used. All the survivors were followed for periods of 1 to 2½ years after surgery. At the time of follow-up examinations, x-rays, fluoroscopic examinations, and electrocardiograms were obtained in all patients. When indicated, sound tracings were recorded. Cardiac catheterization was repeated in 10 of the 28 survivors.

Nomenclature
Several terms have been used in connection with the lesions under discussion. Wakai and Edwards16 divided the defects into partial, transitional, and complete forms of persistent common atrioventricular canal. However, because all of the defects are derived embryologically from the endocardial cushion, Gross and Watkins5 coined the term endocardial cushion defect. The latter was adopted by Campbell and Missen,9 who objected to the nomenclature of Wakai and Edwards but retained their three subgroups. Paul10 urged that morphologically specific terminology be used. He classified endocardial cushion defects into (1) persistent ostium primum with
cleft mitral valve, (2) persistent ostium primum with cleft mitral and tricuspid valves, and (3) persistent common atrioventricular canal.

We have found that the clinical distinction between the above three morphologic groups is not always clear-cut. Frequently, a cleft in the mitral valve is accompanied by hypoplasia of the septal leaflet of the tricuspid valve without evidence of cleft formation, although functionally, both valves are incompetent. Accordingly, we have classified the patients as having either an incomplete or complete form of endocardial cushion defect. We have defined the incomplete form as an ostium primum defect with a cleft mitral valve and varying degrees of hypoplasia or cleft of the tricuspid valve associated with an intact atrioventricular annulus. By contrast, the complete form is defined as having a common annulus of the mitral and tricuspid valves in addition to a low-lying atrial septal defect and a ventricular septal defect, i.e., it is a persistent common atrioventricular canal.

Results

Preoperative Assessment

Clinical Findings

Of the 44 patients who underwent open-heart surgery, 27 were females and 17 were males. The average age at operation was 10 4/12 years, with a range from 1 10/12 years to 34 years. The incomplete form of endocardial cushion defect was present in 32, the complete form in 12.

Table 1 summarizes the pertinent features of the history and physical examination in the incomplete and complete forms of endocardial cushion defect. These data are presented in tubular form, since the clinical profile of the anomaly has been described amply before.7-10 The table indicates the type of individual undergoing surgery. It may be seen that no absolute differentiation of the complete from the incomplete form is possible on clinical grounds. One may infer, however, that the presence of cyanosis and a narrowly split second sound with an increased pulmonary component raises the question of the complete defect. Only two of the 12 patients with complete atrioventricular canal were asymptomatic at the time of surgery, whereas about half of the individuals with the incomplete deformity were free of symptoms.

Electrocardiograms

It has been stressed repeatedly that the electrocardiogram is the best single tool in the differentiation of the endocardial cushion

| Table 1 |
| Clinical Summary of 44 Patients with Endocardial Cushion Defects |
| History | Incomplete (32) | Complete (12) |
| Congestive heart failure | 9 | 6 |
| Dyspnea on exertion | 14 | 10 |
| Cyanosis | 5 | 8 |
| Physical examination | | |
| Murmur | | |
| Apical systolic | 31 | 9 |
| Diastolic | 29 | 9 |
| S2 split | | |
| Wide, fixed (P2 normal) | 26 | 6 |
| Narrow (P2 increased) | 3 | 5 |
| Congestive heart failure | 8 | 5 |
| Cyanosis | 4 | 7 |

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Figure 2

Distribution of mean electrical axis.

Figure 3

Relation of the mean electrical axis to right ventricular systolic pressure.

defect from secundum-type atrial septal defect. The electrocardiographic features of our patients are summarized in figure 1. The distribution of the mean electrical axis is presented in figure 2. Although there is overlapping of the two groups as demonstrated in these figures the association of right ventricular hypertrophy and a mean electrical axis between –105° and –180° makes the diagnosis of a complete canal likely. These features imply right ventricular hypertension as do the auscultatory findings of a loud pulmonary valve closure and a narrowly split second sound. The correlation between mean electrical axis and measured right ventricular pressure is shown in figure 3.

X-rays

The radiologic features are summarized in figure 4. The high incidence of significant cardiomegaly indicates the type of patient undergoing repair.

Catheterization Findings

The pertinent data at cardiac catheterization are summarized in figure 5; they correspond closely to the profile outlined by other authors. In two patients in the group, right ventricular hypertension was associated with valvular pulmonic stenosis and normal pulmonary arterial pressures. The presence or absence of pulmonary arterial hypertension and peripheral arterial unsaturation was useful in differentiating the complete from the incomplete form of endocardial cushion defect, although considerable overlapping occurred. Mitral and tricuspid regurgitation was evaluated by selective left and right ventricular cineangiograms. This method, although largely qualitative, proved to be more useful than other approaches in our hands.

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EXACT QUANTITATION OF REGURGITATION WAS NOT ATTEMPTED BECAUSE OF THE INHERENT ERRORS IN SUCH METHODS IN THE PRESENCE OF INTRACARDIAC SHUNTS.

OPERATIVE RESULTS

OF THE 32 PATIENTS WITH THE INCOMPLETE FORM OF ENDOCARDIAL CUSHION DEFECT, THREE DIED (9.5 PER CENT) WITHIN THE IMMEDIATE POSTOPERATIVE PERIOD. THREE ADDITIONAL DEATHS (9.5 PER CENT) OCCURRED AT INTERVALS OF 4 TO 14 MONTHS, GIVING AN OVER-ALL MORTALITY RATE OF 19 PER CENT.

IN THE GROUP OF 12 PATIENTS WITH THE COMPLETE FORM OF ENDOCARDIAL CUSHION DEFECT, THERE WERE EIGHT DEATHS IMMEDIATELY AFTER SURGERY AND AN ADDITIONAL DEATH 8 MONTHS LATER WITH AN OVER-ALL MORTALITY RATE OF 75 PER CENT.

COMPLICATIONS

TABLE 2

<table>
<thead>
<tr>
<th>Complication</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shock</td>
<td>9 (4)*</td>
</tr>
<tr>
<td>Congestive heart failure</td>
<td>15 (8)</td>
</tr>
<tr>
<td>Arrhythmias</td>
<td></td>
</tr>
<tr>
<td>Complete heart block</td>
<td></td>
</tr>
<tr>
<td>Transient</td>
<td></td>
</tr>
<tr>
<td>Persistent</td>
<td>3 (1)</td>
</tr>
<tr>
<td>Ventricular fibrillation</td>
<td>3 (2)</td>
</tr>
<tr>
<td>Supraventricular tachycardia</td>
<td>8 (1)</td>
</tr>
<tr>
<td>Duration of fever</td>
<td></td>
</tr>
<tr>
<td>&gt; 3 days</td>
<td>19 (2)</td>
</tr>
<tr>
<td>&gt; 10 days</td>
<td>3 (0)</td>
</tr>
<tr>
<td>Suspected postpericardiomy syndrome</td>
<td>6 (0)</td>
</tr>
<tr>
<td>Central nervous system damage</td>
<td>3 (2)</td>
</tr>
</tbody>
</table>

*FIGURES IN PARENTHESES ARE THE NUMBER OF PATIENTS WHO DIED.

FOR THIS WAS PROBABLY INADEQUATE CORRECTION OF MITRAL REGURGITATION ASSOCIATED WITH COMPLETE CLOSURE OF THE ATRIAL SEPTAL DEFECT AND AN ABRUPT RISE IN LEFT ATRIAL PRESSURE TO HIGH LEVELS.

THE DEVELOPMENT OF PROFOUND HYPOTENSION, NOT ASSOCIATED WITH CLINICAL FEATURES OF CONGESTIVE FAILURE WAS ENCOUNTERED OCCASIONALLY. ABOUT ONE HALF OF THE PATIENTS WHO DEVELOPED THIS SYNDROME DIED.

THE OCCURRENCE OF ARRHYTHMIA IN THE IMMEDIATE POSTOPERATIVE PERIOD WAS COMMON, BUT IN MOST CASES PROVED TO BE TRANSIENT. PERMANENT COMPLETE HEART BLOCK WAS ENCOUNTERED TWICE; BOTH PATIENTS DIED.

Fever in the first 3 to 4 days following surgery was usually of no consequence and occurred in the majority of patients. The persistence of fever or its delayed onset was seen occasionally and was difficult to evaluate. Of the survivors there were six in whom fever occurred intermittently in periods ranging from 10 days to 6 months. In two of these, fever was associated with pleuritic pain. THESE SIX PATIENTS HAVE BEEN DIAGNOSED TENTATIVELY AS HAVING HAD THE POSTPERICARDIOTOMY SYNDROME.

CENTRAL NERVOUS SYSTEM INJURY WAS APPARENT IN THREE PATIENTS. ONE PATIENT BECAME DECREBRATE FOLLOWING SEVERAL BOUTS OF VENTRICULAR FIBRILLATION AND EVENTUALLY DIED. THE SECOND PATIENT DEVELOPED PERSONALITY CHANGES
that proved to be transient and were thought to be due to air embolization. In the third patient, a subarachnoid hemorrhage was found at autopsy.

Late Results

Of the 29 survivors, 28 have been reevaluated at intervals of 1 to 21/2 years following surgery.* Twenty-five of these had the incomplete and three the complete form of endocardial cushion defect.

Clinical Assessment

At the time of follow-up, all but one of the patients were asymptomatic; this latter patient had mild exercise intolerance and exertional dyspnkea. An appreciable increase in weight occurred in almost half (13/28) and an increase in height in a quarter of the patients. Wide splitting of the second sound persisted in 50 per cent of the patients. This splitting differed from the preoperative state, however, in that respiratory variations of 0.01 to 0.02 second were found phonocardiographically in all. In all but three instances, the wide but variable splitting could be explained by the presence of complete right bundle-branch block. In the remaining 50 per cent the second sound was normally split. On the basis of the above characteristics of

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the second sound, the assumption has been made that no appreciable shunt exists across the atrial septum.18 Nineteen of the 28 patients have persistent regurgitant systolic murmurs, and in all cases these were considered due to residual atrioventricular valve incompetence. The presence of diastolic flow murmurs in 15 patients has been attributed, in the absence of residual shunts, to atrioventricular valve regurgitation or deformity of the valve leaflets.

Electrocardiogram

Figure 6 depicts the electrocardiographic changes in the 28 survivors. A typical tracing before and after operation is shown in figure 7. It is interesting to note the large number of patients in whom complete right bundle-branch block has appeared, despite the fact that no incision was made in the right ventricle. One may speculate that the right branch of the conduction bundle may have been caught up in a suture during the insertion of the Ivalon prosthesis.

X-ray

Figure 8 summarizes the radiographic changes. An example of a typical x-ray before and after surgery is shown in figure 9. A decrease in heart size was most frequently accompanied by a striking diminution in the fullness of the right lower cardiac border, the region of the right atrium.

The disappearance of the pulmonary vascular engorgement on the x-ray, following oblit-
operation of the shunt, occurred in most cases over a period of 6 to 12 months. In less than one fourth of the patients, the pulmonary vasculature was still prominent at the time of their last follow-up. In spite of this, we have assumed, on the basis of the normal heart size and the previously mentioned characteristics of the second sound, that no significant residual shunt was present.

Persistent left atrial enlargement was thought in all cases to be due to varying degrees of mitral regurgitation.

**Cardiac Catheterization**

Of the 28 survivors, 10 have been recatheterized, representing most of the patients in whom a reasonable suspicion existed that there was a residual left-to-right shunt (persistent cardiac enlargement, pulmonary vascular engorgement). In nine of these, no evidence of a residual left-to-right shunt could be demonstrated; in one, a rise in oxygen saturation of 5 to 6 per cent at the atrial level was only questionably significant.

Pulmonary arterial pressures were normal in all but one, a patient with high-grade mitral regurgitation and an elevated pulmonary capillary pressure.

Evidence of mitral regurgitation was present in two patients. Phasic pressure curves from the left atrium or pulmonary capillary position in these patients revealed dominant "V" waves of 17 to 28 mm. with elevated mean pressures. Both patients had clinical,
radiologic, and electrocardiographic evidence of mitral regurgitation.

**Autopsy Findings**

Of the 15 patients who died, postmortem examinations were available in 12. The atrial and ventricular septal defects were found to be closed in all. Both atrioventricular valves were examined and, in all instances, the clefts were found to be adequately sutured. The tricuspid valve was considered competent in all cases. By contrast, the mitral valve was found to be incompetent in nine of the 12, as a result of short chordae attaching the cleft margins to the ventricular septal wall. In one additional patient, inadvertent suture placement resulted in significant mitral stenosis.

Intimal and medial changes in the pulmonary arterioles were graded from 1 to 6 according to the criteria of Edwards et al.19 Generally, the higher preoperative pulmonary arterial mean pressures were associated with the most advanced proliferative arteriolar changes. On the other hand, the patient in whom the highest pulmonary arterial mean pressure was recorded, (a 5½-year-old male with a mean pressure of 60 mm. Hg) had normal-appearing pulmonary arterioles.

Ten of the 12 patients had evidence of varying degrees of acute pulmonary edema and hemorrhage. An additional two patients had microscopic features of chronic passive congestion in the lungs.

Areas of atelectasis, usually confined to the left lung, were found in six patients. Obstruction of the left main or left lower bronchus by a markedly dilated heart frequently contributed to the atelectasis.

**Discussion**

Our immediate results of surgery for the repair of incomplete endocardial cushion defects compare favorably with those reported from other institutions.11-14 Insofar as the reports in the literature deal, for the most part, with the immediate results of surgery only, no comparable over-all mortality statistics are available. One is impressed, however, by the fact that in our series there were as many late as early deaths.

The inordinately high mortality rates (60 to 75 per cent) in most reported series11,12 as well as in our group of complete atrioventricular canals are quite unacceptable. Among the various factors contributing to these figures are the presence of pre-existing congestive heart failure, marked pulmonary hypertension, large defects in the septa with markedly deformed and cleft valves, long perfusion time, and, finally, the danger of heart block. The outlook was particularly
Electrocardiograms, preoperative (September 30, 1957) and postoperative (March 4, 1959) of patient J.M.

Figure 10A

Electrocardiograms, preoperative (September 30, 1957) and postoperative (March 4, 1959) of patient J.M.

forbidding when repairs were undertaken in children less than 35 pounds in weight. Newer technics, especially with respect to better relief of mitral regurgitation, will improve operative results.

Among the 28 survivors in our series, effective obliteration of intracardiac shunts has been proved at catheterization in nine instances. In one recatheterized patient, a questionable shunt exists. In the remaining 18, clinical data indicate that no appreciable shunt persists postoperatively.

On the other hand, residual mitral incompetency was not infrequently encountered. Figure 10A and B are the electrocardiograms and x-rays before and after surgery on a patient in whom moderately severe mitral regurgitation persists. Figure 11 summarizes the clinical, radiographic, and electrocardiographic features used to assess the severity of mitral regurgitation. There were 18 patients in whom a significant apical systolic murmur indicated the presence of mitral regurgitation. Of these, 12 had enlargement of the left atrium by x-ray. Among the latter groups, seven had evidence of left atrial hypertrophy by electrocardiogram. Thus, we may summarize by saying that of the total number of patients with mitral regurgitation, there were seven who have significant disease.

The etiology of residual mitral regurgitation in patients with endocardial cushion defects in whom the mitral cleft has been successfully repaired, was demonstrated by Edwards.20 In reviewing autopsy material, he found short chordae attaching the closed cleft margin of the mitral valve to the ventricular septum, and preventing adequate coaptation of the valve during systole. In nine of 12 cases examined post mortem, we
were able to demonstrate such chordae. Presently, surgical exploration of the under surface of the cleft margin for chordae with attachments to the ventricular septum is carried out. If they are found, connections to the cleft margin are severed. It is our belief that this maneuver may reduce the incidence and severity of mitral regurgitation considerably.

Clinical evidence of tricuspid insufficiency was not seen postoperatively. The 10 recatheterized patients had no evidence of tricuspid regurgitation by pressure-curve analysis. Angiographic evaluation of tricuspid regurgitation with injection of dye into the right ventricle was not conclusive.

**Summary**

Preoperative and postoperative data on 44 patients with endocardial defect, 32 with the incomplete and 12 with the complete form are presented. Nineteen per cent of the incomplete form died with 9.5 per cent of the deaths occurring early and 9.5 per cent late. The mortality rate for the complete form was 75 per cent. *

Late follow-up results were evaluated in 28 patients 1 to 2½ years following surgery.

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*See Addendum 2.
Catheterization data in nine of 10 and clinical observations in 18 indicated no residual shunt. One patient showed questionable evidence of a left-to-right shunt by cardiac catheterization. Evidence of mitral regurgitation was present in seven (25 per cent) of the survivors. Short chordae attaching the closed cleft margin of the mitral leaflet to the ventricular septum rendered the valve incompetent in nine of 12 autopsied specimens. Surgical exploration of the underside of the cleft margins for such chordae is recommended. No evidence of residual tricuspid regurgitation was found.

Addendum

1. Since these findings were compiled, a follow-up of the twenty-eighth patient, a boy with incomplete canal, has been obtained. He was reevaluated elsewhere 1 year postoperatively. At that time he still showed evidence of mitral regurgitation, but his heart size was appreciably smaller than it was preoperatively. (The cardiothoracic ratio dropped from 0.72 to 0.55.) He showed symptomatic improvement.

2. Since the close of this series, January 1, 1960, an additional 11 patients with endocardial cushion defect have been treated surgically. Six of the nine patients with the incomplete form and both patients with the complete form of endocardial cushion defect survived. The addition of these patients to our original series changes the mortality figures of the total group to 22 per cent in respect to incomplete canal and 64 per cent in respect to complete canal.

References

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