Partial Atrioventricular Canal Defect in Adults

JOHN K. HYNES, M.D., ABDUL J. TAJIK, M.D., JAMES B. SEWARD, M.D.,
VALENTIN FUSTER, M.D., DONALD G. RITTER, M.D., ROBERT O. BRANDENBURG, M.D.,
FRANCISCO J. PUGA, M.D., GORDON K. DANIELSON, M.D.,
AND DWIGHT C. McGOON, M.D.

SUMMARY Between March 1955 and March 1981, 52 adult patients (age 20 years or older) with partial atrioventricular canal (PAVC) were examined at the Mayo Clinic. Forty patients were in New York Heart Association (NYHA) functional class I or II. The ECG showed right bundle branch block with left-axis deviation in 48, first-degree atrioventricular block in 33 and atrial fibrillation in nine. Preoperative catheterization was done in 37 patients; the mean pulmonary resistance index was 2.8 U·m², and the mean pulmonary-to-systemic flow ratio was 3.0. Forty-seven patients underwent operation; two required mitral valve replacement. There were three operative deaths (6.4% operative mortality) and two late deaths. All surviving patients were in NYHA class I or II. No patient required subsequent permanent pacemaker implantation. We conclude that repair of PAVC in adults may be performed at low risk and may improve patient longevity and functional status.

PARTIAL atrioventricular canal (PAVC) defect is a form of endocardial cushion defect infrequently encountered in adults; few reported patients have survived into the sixth or seventh decade of life.1-4 Earlier published reports5-9 describing the clinical, electrocardiographic and angiographic features and surgical experience have evaluated this congenital lesion primarily in pediatric patients and have included few adult patients. Therefore we reviewed our experience with 52 adult patients with PAVC. The clinical profile, hemodynamic and angiographic findings, and results of operation are given specific attention.

For this study, PAVC defect was defined as including an ostium primum atrial septal defect but with two separate atrioventricular valve rings, a cleft in the anterior mitral leaflet, and no appreciable interventricular communication.

Patients and Methods

Between March 1955 and March 1981, 52 adult patients with PAVC were examined at the Mayo Clinic. The diagnosis was established by cardiac catheterization (37 patients) or operation (47 patients) or both. The two-dimensional echocardiographic findings were diagnostic in 13 recent patients.

There were 31 women and 21 men. At the initial examination, the patients were 20–75 years old (mean 37 years). Eighteen patients (35%) were age 45 years or older, and six patients (12%) were age 60 years or older.

Clinical Status and Preoperative Evaluation

All patients were categorized on the basis of New York Heart Association (NYHA) functional class at the initial examination. Forty patients (77%) were either asymptomatic or mildly symptomatic, 10 patients (19%) were in NYHA class III, and two patients (4%) were in class IV. All patients age 45 years or older were symptomatic. The most frequently reported symptoms were palpitations, exertional dyspnea and chronic fatigue.

Fifty-one patients (98%) had an abnormal ECG. The most common electrocardiographic abnormality (48 patients [92%]) was right bundle branch block with left-axis deviation associated with a counterclockwise loop in the frontal plane of the vectorcardiogram. Of 42 patients (81%) with normal sinus rhythm, 33 had first-degree atrioventricular block. Nine patients (17%) had chronic atrial fibrillation. One additional patient had intermittent complete atrioventricular block and prior permanent pacemaker implantation. Four patients (8%) had electrocardiographic tracings that were atypical for PAVC and showed (in one patient each) isolated left bundle branch block, right-axis deviation without intraventricular conduction delay, left-axis deviation without intraventricular conduction delay, and completely normal findings. These four patients were ages 20, 76, 36 and 20 years. Three patients underwent cardiac catheterization and had left-to-right shunts of 53%, 54% and 75%, respectively. Two patients had moderate and one had severe mitral regurgitation.

All 52 patients had cardiac enlargement on chest roentgenograms. The cardiac configuration and the size of the main pulmonary arteries varied considerably. In all patients, pulmonary vascular markings were increased, consistent with a left-to-right shunt.

Two-dimensional echocardiography, performed in 13 recent patients, clearly defined the anatomic features of PAVC as described by Hagler et al.10 These include visualization of the primum defect in the inferior or atrial septum and insertion of both atrioventricular valves at the same level onto the crest of the ventricular septum (on the four-chamber view) and identification of the cleft in the anterior mitral leaflet (on short-axis scans).

Hemodynamics and Angiography

Cardiac catheterization was performed in 37 patients (71%). Hemodynamic data were available on 34
patients. The mean pulmonary-to-systemic flow ratio was 3.0 (range 1.6–4.7), the mean left-to-right shunt was 67% (range 43–87%), the mean pulmonary artery pressure was 23 mm Hg (range 11–55 mm Hg), and the mean total pulmonary vascular resistance index (Rp) (mean pulmonary artery pressure/pulmonary index; normal < 4 U·m⁻²) was 2.8 U·m⁻² (range 1.0–9.8 U·m⁻²). Only two patients had an Rp greater than 6 U·m⁻².

Left ventricular angiography was performed in 33 patients. The characteristic “goose-neck” deformity of the left ventricular outflow tract was recognized by the angiographer in 29 patients (88%). Mitral regurgitation was graded as none in six patients (18%), mild in 18 patients (55%), moderate in five patients (15%), and severe in four patients (12%). On the basis of cardiac catheterization and left ventricular angiography, a correct diagnosis of PAVC was made in 31 of 33 patients (94%). The two incorrect diagnoses were secundum atrial septal defect and severe mitral regurgitation.

Surgical Group

Forty-seven of the 52 patients underwent operation. These patients were 20–71 years old (mean 38 years). At operation, 24 patients were age 35 years or older.

Cardiac catheterization was done in 37 of the 47 operative patients. The 10 patients who were referred for surgical repair of PAVC without cardiac catheterization were 23–48 years old (mean 30 years). In nine of these 10 patients, a correct preoperative diagnosis was established on the basis of classic physical findings (hyperdynamic right ventricular impulse, persistently split second heart sound, diastolic tricuspid flow murmur, systolic pulmonary outflow murmur, and mitral insufficiency murmur), electrocardiography (right bundle branch block and left-axis deviation), and chest roentgenography (consistent with increased pulmonary flow). One patient who was referred for operation without cardiac catheterization had an incorrect preoperative diagnosis of severe rheumatic mitral insufficiency. The ECG in this patient showed left-axis deviation without right bundle branch block. The typical features of PAVC were noted at operation. Repair was performed in all 10 patients, and the postoperative course was uneventful.

Surgical Findings, Repair, and Early Course

The ostium primum atrial septal defect was closed with a synthetic patch in 35 patients, with a pericardial patch in nine patients, and by direct suture in three patients. (The defect was more than 2 cm² in all three patients.) The cleft mitral leaflet was repaired by direct suture in 37 patients and with a pericardial patch in one patient. Braunwald-Cutter mitral and aortic prostheses were inserted in one patient who had a double-orifice mitral valve and severe aortic insufficiency. One other patient required mitral valve replacement for severe mitral insufficiency. In six patients, the cleft mitral leaflet was not repaired because the valve was competent on the preoperative ventriculogram and at operation. One patient did not have a cleft mitral leaflet. In addition, two patients had tricuspid annuloplasty and one patient, age 60 years, had one-vessel coronary artery bypass grafting. The complete anatomic findings are shown in table 1.

There were three early deaths (within 30 days after operation), an operative mortality of 6.4%. These three patients had normal pulmonary vascular resistance (Rp less than 3 U·m⁻²) at the time of preoperative catheterization. Two patients (ages 20 and 53 years; NYHA functional class II) died of respiratory insufficiency (acute respiratory distress syndrome) within 72 hours after operation. The third patient (age 20 years; NYHA functional class III) died of refractory ventricular fibrillation 24 hours postoperatively. Since July 1975, 17 patients (36%) have undergone operation without mortality.

Forty-four patients (94%) survived the immediate postoperative period and were dismissed from the hospital a mean of 11 days (range 7–35 days) after operation. In 25 of the 44 patients (57%), the postoperative course was uncomplicated. Postoperative atrial arrhythmia necessitating treatment was noted in 15 patients (34%).

Follow-up

Follow-up data were obtained in 42 of the 44 patients (95%) who survived surgical repair. The mean follow-up period was 83 months (range 1–269 months). There have been two late deaths. One patient, age 32 years, died with recurrent left ventricular failure in 1972 at 31 months after operation. Preoperatively, she had severe mitral insufficiency and was in NYHA class IV congestive heart failure. Autopsy confirmed that the repair was intact, with continuity of the anterior mitral leaflet and an intact atrial septal patch. The second late death occurred in 1972, 3 months postoperatively, in a 52-year-old man with severe hemolysis and intravascular coagulopathy, presumably related to his mechanical (Braunwald-Cutter) mitral and aortic valve prostheses.

Table 1. Anatomic Findings in 47 Surgical Patients with Partial Atrioventricular Canal Defect

<table>
<thead>
<tr>
<th>Defect</th>
<th>No. of pts</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ostium primum defect</td>
<td>47</td>
</tr>
<tr>
<td>Cleft anterior mitral leaflet</td>
<td>46</td>
</tr>
<tr>
<td>Complete cleft</td>
<td>37</td>
</tr>
<tr>
<td>Incomplete cleft</td>
<td>9</td>
</tr>
<tr>
<td>Tricuspid leaflet abnormality</td>
<td>20</td>
</tr>
<tr>
<td>&quot;Cleft&quot; leaflet</td>
<td>3</td>
</tr>
<tr>
<td>Diminutive portion of septal leaflet</td>
<td>17</td>
</tr>
<tr>
<td>Secundum atrial septal defect</td>
<td>8</td>
</tr>
<tr>
<td>Persistent left superior vena cava</td>
<td>2</td>
</tr>
<tr>
<td>Left atrial orifice to coronary sinus</td>
<td>2</td>
</tr>
<tr>
<td>Double-orifice mitral valve</td>
<td>1</td>
</tr>
<tr>
<td>Prolapsed aortic cusp with severe</td>
<td>1</td>
</tr>
<tr>
<td>aortic insufficiency</td>
<td></td>
</tr>
</tbody>
</table>
Thirty-three of 40 postoperative long-term survivors are entirely asymptomatic. Improvement in NYHA classification is shown in figure 1. Seven patients are in NYHA class II and have mild fatigue and exertional dyspnea. Ten patients are receiving antiarrhythmic therapy for paroxysmal atrial arrhythmia. No patient has required permanent pacemaker implantation or reoperation for repair of recurrent atrial septal defect or mitral valve replacement. Repeat catheterization was performed in only two patients, and there was no evidence of residual shunt or appreciable mitral insufficiency.

Discussion

Atrial septal defect is the third most common form of congenital heart disease in adult patients, after mitral valve prolapse and bicuspid aortic valve in prevalence. The incidence of atrial septal defect in adults has been estimated by Seldon et al. to be 1 in 5700 persons. On review of all patients who had repair of atrial septal defect at our institution between 1970 and 1980, ostium primum defect accounted for 33% of patients age 20 years or younger and 9.5% of patients older than age 20 years.

We report the first large series of adult patients with PAVC defect. Martin et al. reported a surgical series of seven adult patients with PAVC (mean age 41 years, range 34–48 years), and Goodman et al. reported a surgical series of 12 adult patients with PAVC. Larger series of PAVC have been reported but have included relatively few adult patients. The 101 patients in the operative series of McMullan et al. included only 15 patients age 20 years or older. Those 15 patients are also included in the present report.

The typical electrocardiographic features of PAVC are the association of right bundle branch block, left-axis deviation to more than –30 degrees, and counterclockwise rotation of the vector loop in the frontal plane. In our adult series, 48 of 52 patients (92%) had these typical electrocardiographic findings. The high prevalence of first-degree atrioventricular block (79% of patients with normal sinus rhythm) and atrial fibrillation (17%) in our adult series differs from that reported in earlier studies. In four of six patients with atrial fibrillation who were studied angiographically, moderate or severe mitral regurgitation was present. As noted by Somerville, established cardiac arrhythmia was associated with considerable clinical deterioration; 50% of our patients with arrhythmia were in NYHA functional class III or IV. Severe pulmonary hypertension was extremely rare in our patients, an observation also consistent with Somerville’s experience.

At our institution, two-dimensional echocardiography is the preferred diagnostic procedure. Cardiac catheterization or angiography may not be necessary when the clinical findings do not suggest significant mitral incompetence or pulmonary hypertension.

The natural history of PAVC defect is poorly defined, and the prognosis has been extrapolated from adults with secundum atrial septal defect or children with PAVC. Several factors may account for the very low incidence of PAVC in adults. Obviously, this congenital lesion is rare. In addition, most of these patients have had heart murmurs detected as children; because of additional electrocardiographic abnormalities, specialized consultation is often obtained, leading to the correct diagnosis and early operation. Finally, the patient attrition rate is unknown. Clinically, ostium primum atrial septal defect with associated mitral insufficiency and direct shunting of blood from the left ventricle into the right atrium would be expected to impose a more severe hemodynamic burden on the right and left ventricles and the pulmonary circuit than would uncomplicated secundum atrial septal defect.

Campbell reviewed the natural history of atrial septal defect and calculated mortality rates for each patient decade from a series of 289 patients with secundum atrial septal defect. Table 2 shows the predicted yearly mortality rate (per decade), the total number of patient-years of follow-up in our series, and the expected

<table>
<thead>
<tr>
<th>NYHA Class</th>
<th>Preoperatively</th>
<th>Postoperatively</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>22</td>
<td>21</td>
</tr>
<tr>
<td>II</td>
<td>10</td>
<td>9</td>
</tr>
<tr>
<td>III</td>
<td>7</td>
<td>4</td>
</tr>
<tr>
<td>IV</td>
<td>1</td>
<td>1</td>
</tr>
</tbody>
</table>

**Figure 1. Preoperative and postoperative New York Heart Association (NYHA) functional class in surviving operative patients. (Excludes three early and two late deaths; two additional patients were lost to follow-up.)**

<table>
<thead>
<tr>
<th>Age (years)</th>
<th>Predicted yearly mortality (%)</th>
<th>Patient-years observed</th>
<th>Predicted no. of deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>20–29</td>
<td>2.7</td>
<td>67.7</td>
<td>1.8</td>
</tr>
<tr>
<td>30–39</td>
<td>4.5</td>
<td>99.2</td>
<td>4.5</td>
</tr>
<tr>
<td>40–49</td>
<td>5.4</td>
<td>61.8</td>
<td>3.3</td>
</tr>
<tr>
<td>50–59</td>
<td>7.5</td>
<td>46.8</td>
<td>3.5</td>
</tr>
<tr>
<td>60–69</td>
<td>7.5</td>
<td>29.0</td>
<td>2.2</td>
</tr>
<tr>
<td>70–79</td>
<td>7.5</td>
<td>10.9</td>
<td>0.8</td>
</tr>
<tr>
<td>Overall</td>
<td>315.4</td>
<td>16.1</td>
<td></td>
</tr>
</tbody>
</table>

**Table 2. Mortality Rates for Atrial Septal Defect in Patients with Partial Atrioventricular Canal Defect**

Expected deaths 16
Observed deaths 5
Operative 3
Postoperative 2

*Data from Campbell.
ed total mortality (predicted yearly mortality rate × patient-years observed). If one assumes that the natural history of primum atrial septal defect is at least similar to (if not worse than) the natural history of secundum atrial septal defect, we would have predicted at least 16 deaths in our population of 40 postoperative patients who were followed up for a mean of 83 months. Because five deaths (three early and two late deaths) occurred in our surgically treated patients (p < 0.01), we believe that operation substantially improved patient longevity. Our data support early operation for asymptomatic adult patients with PAVC.

Acknowledgment

The authors thank Peter C. O’Brien, Ph.D., for assistance with the statistical analysis.

References

Partial atrioventricular canal defect in adults.


_Circulation_. 1982;66:284-287
doi: 10.1161/01.CIR.66.2.284

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1982 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/66/2/284

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in _Circulation_ can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to _Circulation_ is online at:
http://circ.ahajournals.org/subscriptions/