Congenital Aortic Stenosis
Clinical and Hemodynamic Findings, Surgical
Technic, and Results of Operation

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Eugene Braunwald, M.D.

Ten years ago congenital aortic stenosis was generally considered to be uncommon and seldom to carry a grave prognosis. The severity of obstruction of left ventricular outflow could not be directly assessed and suitable surgical procedures for its correction were not available. The changing concepts in respect to this malformation are reflected in the series of 30 patients described. The lesion was found to be relatively common and frequently serious. The severity and site of obstruction could be precisely evaluated and a physiologic basis was provided for the selection of patients for operation providing direct exposure of the aortic valve.

Congenital aortic stenosis has long been considered to be an unusual and relatively benign form of congenital heart disease. However, recent reports1-5 have called attention to the variability of the clinical course in patients presenting the classic physical findings of this lesion. With the development of technics for the surgical correction of aortic stenosis, precise evaluation of the severity of obstruction has become necessary in the proper selection of patients for operation. This has been made possible by the development of safe methods for the measurement of left heart pressure.6,7 The present report summarizes the clinical and hemodynamic findings in 30 patients, in all of whom the diagnosis of aortic stenosis was confirmed by the demonstration of a systolic pressure gradient between the left ventricle and a systemic artery. The technic of operative correction under direct vision and the results of surgical treatment in 18 of these patients are also presented.

Clinical Observations

The pertinent clinical and electrocardiographic findings are summarized in table 1. A history of rheumatic fever was not elicited in any of the 30 patients and none had clinical or hemodynamic evidence of involvement of the mitral valve. A heart murmur had been described in 21 patients before the age of 6 years. Nine patients, though their murmurs had been discovered later, are also included. Two of these 9 patients had other congenital cardiovascular anomalies (J. G. and W. D.). The other 7 patients had never been examined prior to the detection of their murmurs between the ages of 6 and 15 years.

There were 20 males and 10 females; their ages at admission ranged from 4 to 39 years. Three patients had associated patent ductus arteriosus (G. G., J. R., and L. A.), 1 a coarctation (J. G.), and another congenital complete heart block (W. D.). The anatomic site of obstruction was established at the time of operation or preoperative left heart catheterization in 21 patients. Valvular aortic stenosis was found in 13 patients and subvalvular stenosis in 8. In the 9 remaining patients the obstruction has not been localized and the site of stenosis is therefore designated "indeterminate" in the tables.

Of the 13 patients with valvular stenosis, 7 had angina and in 2 of these it was severe; 3 patients had experienced syncopal attacks. Twelve of these 13 patients experienced dyspnea and 4 were severely limited by this symptom. Fatigability was marked and generally associated with dyspnea. Only 1 patient (H. S.) had had right heart failure and 1


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Table 1.—Clinical Features in Congenital Aortic Stenosis

<table>
<thead>
<tr>
<th>Site of stenosis</th>
<th>Patient</th>
<th>Age (yrs.), sex</th>
<th>Age murmur detected, (yrs.)</th>
<th>Fatigability</th>
<th>Dyspnea</th>
<th>Angina</th>
<th>Symptome</th>
<th>Duration symptoms, (yrs.)</th>
<th>Aortic 2nd sound</th>
<th>Diastolic murmur Gr. I-VI</th>
<th>Ventric. hypertrophy</th>
<th>ECG</th>
<th>Operation</th>
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<tr>
<td>V</td>
<td>E. R.</td>
<td>12F</td>
<td>birth</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>D</td>
<td>0</td>
<td>LVH</td>
<td>open, perfusion.</td>
</tr>
<tr>
<td>A</td>
<td>D. S.</td>
<td>13F</td>
<td>birth</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td>0</td>
<td>0.5</td>
<td>D</td>
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<td>LVH</td>
<td>open, perfusion.</td>
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<tr>
<td>L</td>
<td>D. M.</td>
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<td>birth</td>
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<td>+</td>
<td>+</td>
<td>0</td>
<td>1</td>
<td>D</td>
<td>0</td>
<td>LVH</td>
<td>none</td>
<td></td>
</tr>
<tr>
<td>V</td>
<td>C. M.</td>
<td>18M</td>
<td>birth</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td>2</td>
<td>D</td>
<td>0</td>
<td>LVH</td>
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<tr>
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<td>0</td>
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<td>N</td>
<td>2</td>
<td>LVH</td>
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<td></td>
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<tr>
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<td>+</td>
<td>0</td>
<td>0</td>
<td>6</td>
<td>D</td>
<td>0</td>
<td>LVH</td>
<td>closed, transfer.</td>
<td></td>
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<tr>
<td>A</td>
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<td>20F</td>
<td>12</td>
<td>0</td>
<td>+</td>
<td>0</td>
<td>0</td>
<td>14</td>
<td>D</td>
<td>0</td>
<td>LVH</td>
<td>closed, transfer.</td>
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<tr>
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<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>10</td>
<td>D</td>
<td>2</td>
<td>LVH</td>
<td>open, hypotrophy.</td>
<td></td>
</tr>
<tr>
<td>C. W.</td>
<td>32M</td>
<td>8</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>14</td>
<td>N</td>
<td>0</td>
<td>LVH</td>
<td>open, hypotrophy.</td>
<td></td>
</tr>
<tr>
<td>J. T.</td>
<td>3M</td>
<td>5</td>
<td>birth</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>N</td>
<td>0</td>
<td>LVH</td>
<td>open, hypotrophy.</td>
<td></td>
</tr>
<tr>
<td>S. D.</td>
<td>39F</td>
<td>birth</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>3</td>
<td>N</td>
<td>0</td>
<td>LVH</td>
<td>open, perfusion.</td>
<td></td>
</tr>
<tr>
<td>S</td>
<td>C. C.</td>
<td>8F</td>
<td>3</td>
<td>+++</td>
<td>+++</td>
<td>+++</td>
<td>+++</td>
<td>4</td>
<td>D</td>
<td>0</td>
<td>NH</td>
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<tr>
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<td>G. B.</td>
<td>10F</td>
<td>3</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td>+</td>
<td>2</td>
<td>N</td>
<td>1</td>
<td>LVH</td>
<td>operat. elsewhere</td>
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<tr>
<td>B</td>
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<td>0</td>
<td>0</td>
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<td>0</td>
<td>-</td>
<td>D</td>
<td>0</td>
<td>LVH</td>
<td>open, hypotrophy.</td>
</tr>
<tr>
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<td>R. K.</td>
<td>16M</td>
<td>9</td>
<td>+</td>
<td>+</td>
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<td>++</td>
<td>0</td>
<td>9</td>
<td>D</td>
<td>2</td>
<td>LVH</td>
<td>open, hypotrophy.</td>
</tr>
<tr>
<td>A</td>
<td>H. L.</td>
<td>16F</td>
<td>3</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>-</td>
<td>D</td>
<td>3</td>
<td>LVH</td>
<td>open, hypotrophy.</td>
</tr>
<tr>
<td>L</td>
<td>C. G.</td>
<td>20M</td>
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<td>+</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td>4</td>
<td>N</td>
<td>0</td>
<td>LVH</td>
<td>none</td>
<td></td>
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<tr>
<td>V</td>
<td>A. Z.</td>
<td>22F</td>
<td>birth</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>16</td>
<td>N</td>
<td>2</td>
<td>LVH</td>
<td>open, hypotrophy.</td>
<td></td>
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<tr>
<td>U</td>
<td>M. M.</td>
<td>36F</td>
<td>birth</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td>0</td>
<td>4</td>
<td>N</td>
<td>0</td>
<td>LVH</td>
<td>open, hypotrophy.</td>
<td></td>
</tr>
<tr>
<td>I</td>
<td>C. McG.</td>
<td>6F</td>
<td>birth</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>-</td>
<td>N</td>
<td>0</td>
<td>LVH</td>
<td>none</td>
<td></td>
</tr>
<tr>
<td>N</td>
<td>J. R.</td>
<td>6F</td>
<td>birth</td>
<td>+</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>-</td>
<td>N</td>
<td>0</td>
<td>LVH</td>
<td>none</td>
<td></td>
</tr>
<tr>
<td>D</td>
<td>J. D.</td>
<td>8F</td>
<td>½</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>-</td>
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<td>0</td>
<td>LVH</td>
<td>none</td>
<td></td>
</tr>
<tr>
<td>E</td>
<td>W. F.</td>
<td>9M</td>
<td>½</td>
<td>+</td>
<td>0</td>
<td>0</td>
<td>+</td>
<td>4</td>
<td>N</td>
<td>0</td>
<td>LVH</td>
<td>closed, transventr.</td>
<td></td>
</tr>
<tr>
<td>T</td>
<td>D. F.</td>
<td>9M</td>
<td>birth</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>+</td>
<td>-</td>
<td>N</td>
<td>0</td>
<td>NH</td>
<td>none</td>
<td></td>
</tr>
<tr>
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<td>W. D.</td>
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<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>-</td>
<td>-</td>
<td>N</td>
<td>0</td>
<td>CHB</td>
<td>none</td>
<td></td>
</tr>
<tr>
<td>R</td>
<td>M. F.</td>
<td>15F</td>
<td>⅔</td>
<td>+</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>⅔</td>
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<td>0</td>
<td>NH</td>
<td>none</td>
<td></td>
</tr>
<tr>
<td>M</td>
<td>L. A.</td>
<td>15M</td>
<td>birth</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>13</td>
<td>N</td>
<td>Cont.</td>
<td>LVH</td>
<td>none</td>
<td></td>
</tr>
<tr>
<td>I</td>
<td>J. G.</td>
<td>17M</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>-</td>
<td>-</td>
<td>N</td>
<td>0</td>
<td>NH</td>
<td>none</td>
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</table>

0 = Absent, + = mild, ++ = moderate, +++ = severe, D = diminished, N = normal, LVH = left ventricular hypertrophy, NH = no hypertrophy, Cont. = continuous murmur, CHB = complete heart block.

(R. S.) had experienced pulmonary edema. Only 1 patient was entirely asymptomatic.

Of the 8 patients with subvalvular stenosis, angina was severe in 3 and mild in another; 2 had histories of syncope. Six of these patients had dyspnea and 2 were severely limited by this symptom. None gave a history of right heart failure or of pulmonary edema. Two patients with subvalvular stenosis were entirely asymptomatic.

It was of interest that the only three physically underdeveloped patients of the entire group all had associated patent ductus arteriosus. Every patient had a harsh systolic mur-
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TABLE 2A.—Left Heart Catheterization Data in Patients with Valvular Congenital Aortic Stenosis

<table>
<thead>
<tr>
<th>Site of Stenosis</th>
<th>Patient</th>
<th>Status</th>
<th>Method</th>
<th>L. V. S/D (mm Hg)</th>
<th>Arterial S/D (mm Hg)</th>
<th>Aortic valve gradient (mm. Hg)</th>
<th>Orifice size (cm.²/BSA)</th>
<th>L. A. Mean (mm. Hg)</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>R. S.</td>
<td>Preop.</td>
<td>P. C.*</td>
<td>270/10</td>
<td>110/61 FA</td>
<td>160</td>
<td>.20</td>
<td>—</td>
</tr>
<tr>
<td></td>
<td>G. G.</td>
<td>Preop.</td>
<td>P. C.*</td>
<td>124/18</td>
<td>101/50 FA</td>
<td>23</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td></td>
<td>E. R.</td>
<td>Preop.</td>
<td>T. B.</td>
<td>221/18</td>
<td>117/68 FA</td>
<td>104</td>
<td>—</td>
<td>13</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Postop.</td>
<td>P. C.*</td>
<td>95/6</td>
<td>90/55 FA</td>
<td>5</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td></td>
<td>D. S.</td>
<td>Preop.</td>
<td>T. B.</td>
<td>200/10</td>
<td>110/65 FA</td>
<td>90</td>
<td>—</td>
<td>10</td>
</tr>
<tr>
<td></td>
<td>V</td>
<td>Preop.</td>
<td>T. B.*</td>
<td>230/12</td>
<td>105/80 AO</td>
<td>125</td>
<td>.26</td>
<td>13</td>
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<tr>
<td></td>
<td>A</td>
<td>Preop.</td>
<td>T. B.</td>
<td>160/8</td>
<td>130/72 BA</td>
<td>30</td>
<td>—</td>
<td>4</td>
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<td>L</td>
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<td>T. B.</td>
<td>184/8</td>
<td>83/54 BA</td>
<td>101</td>
<td>—</td>
<td>12</td>
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<tr>
<td></td>
<td></td>
<td>Postop.</td>
<td>T. B.</td>
<td>145/14</td>
<td>145/64 BA</td>
<td>0</td>
<td>—</td>
<td>—</td>
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<td>V</td>
<td>Preop.</td>
<td>T. B.</td>
<td>227/13</td>
<td>116/76 AO</td>
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<td>—</td>
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<td></td>
<td>U</td>
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<td>T. B.</td>
<td>220/21</td>
<td>134/82 BA</td>
<td>86</td>
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<td>L</td>
<td>Preop.</td>
<td>T. B.</td>
<td>230/17</td>
<td>120/74 BA</td>
<td>110</td>
<td>—</td>
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<tr>
<td></td>
<td></td>
<td>Postop.</td>
<td>T. B.</td>
<td>175/9</td>
<td>125/86 BA</td>
<td>50</td>
<td>—</td>
<td>9</td>
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<tr>
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<td>A</td>
<td>Preop.</td>
<td>T. B.</td>
<td>227/15</td>
<td>83/42 AO</td>
<td>144</td>
<td>—</td>
<td>15</td>
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<td>R</td>
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<td>T. B.</td>
<td>200/12</td>
<td>140/68 BA</td>
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<td>C. W.</td>
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<td>P. C.</td>
<td>146/2</td>
<td>77/60 BA</td>
<td>69</td>
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<td>—</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Postop.</td>
<td>T. B.</td>
<td>176/8</td>
<td>153/74 BA</td>
<td>23</td>
<td>.83</td>
<td>10</td>
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<tr>
<td></td>
<td>J. T.</td>
<td>Preop.</td>
<td>T. B.</td>
<td>210/7</td>
<td>122/72 BA</td>
<td>88</td>
<td>.50</td>
<td>14</td>
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<tr>
<td></td>
<td></td>
<td>Postop.</td>
<td>T. B.</td>
<td>180/18</td>
<td>110/65 AO</td>
<td>70</td>
<td>—</td>
<td>12</td>
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<td>S. D.</td>
<td>Preop.</td>
<td>T. B.</td>
<td>208/10</td>
<td>119/72 BA</td>
<td>89</td>
<td>.57</td>
<td>10</td>
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</tbody>
</table>

T.B. = transbronchial left heart catheterization. P. C. = percutaneous left ventricular puncture. L. V. = left ventricular pressure, FA = femoral arterial pressure. BA = brachial arterial pressure. AO = central aortic pressure.

*Procedure carried out under general anesthesia.
†Calculated effective valve area is in cm.² per M.² of body surface area.

Fluoroscopic and radiographic examinations of the heart revealed some degree of left ventricular enlargement in all but 3 of the 30 patients. Characteristically this was the globular shadow of concentric hypertrophy and was most prominent in the left anterior oblique projection. Poststenotic dilatation of the aorta was a prominent feature in almost every patient. Angiocardiography was carried out in 11 patients and always revealed a thickened left ventricular wall and a small left ventricular cavity. In 1 patient (W.F.)
the left atrium was also enlarged. In all patients without patent ductus, the chambers of the right heart were of normal size. Of the 30 patients, the electrocardiogram revealed left ventricular hypertrophy, according to criteria previously described, in 24. Complete heart block with a normal supraventricular complex was present in 1 patient (W. D.).

**HEMODYNAMIC FINDINGS**

Left heart pressures were measured in all patients and these data are summarized in tables 2A and 2B. Left heart catheterization was performed by the transbronchial route in 19 patients. In the other patients, particularly young children, left ventricular pressures were obtained by means of direct puncture of this chamber through the left anterior
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chest wall, as described by Brock. In these patients, general anesthesia was employed and it was also required in several children catheterized by the transbronchial route. The aortic valve gradients, i.e., the differences between simultaneous peak systolic pressures in the left ventricle and the aorta or a peripheral artery, ranged from 15 to 172 mm. Hg and averaged 82 mm. Hg. Typical left ventricular and arterial pressures, recorded in the course of transbronchial left heart catheterization, are shown in figure 1. Left ventricular end-diastolic pressures ranged from 2 to 28 mm. Hg and averaged 13 mm. Hg. Cardiac outputs were measured by means of the indicator-dilution technic at the time of left heart catheterization, in 10 of the patients in whom there was no evidence of aortic regurgitation. This made possible the application of the Gorlin formula for the estimation of effective orifice size. These areas ranged from 0.20 to 1.12 cm.² per M.² of body surface area (BSA).

In 4 of the patients with subvalvular stenosis the transbronchial catheter was passed from the left ventricle into the aorta; when the catheter was withdrawn, a zone of intermediate pressure was encountered between the aorta and left ventricle that indicated the presence of the subvalvular obstruction (fig. 2). In other patients a single, abrupt pressure change was noted upon withdrawal of the catheter into the left ventricle and the presence of valvular stenosis could thus be established prior to operation.

In the patients without aortic diastolic murmurs, the direct systemic arterial pulse pressures were frequently normal but ranged from 28 to 75 mm. Hg; the average value was 41 mm. Hg. On the other hand, in the 8 patients in whom such a murmur was present, the arterial pulse pressures were often wider and ranged from 40 to 70 mm. Hg; the average value was 55 mm. Hg. In 2 such patients, presenting the clinical findings of associated aortic regurgitation, aortic regurgitant flow was estimated before operation by means of the indicator-dilution method recently described. In H. L., a patient with subvalvular stenosis, the regurgitant flow was 1.4 L./min.; in patient D. S., with valvular stenosis, it was 1.6 L./min.

Right heart catheterizations were carried out in 28 of the 30 patients. These data are summarized in tables 3A and 3B. The mean pulmonary capillary wedge pressure was measured in 25 patients in the resting state was found to be elevated above 12 mm. Hg in only 7 patients without associated patent ductus. Significant pulmonary hypertension was present in only 1 patient without an associated ductus (W. F.). In 6 other patients without patent ductus the mean pulmonary artery pressure was minimally elevated at rest (20 to 24 mm. Hg). One patient, C. G., pre-

Fig. 1 Top. Simultaneous records of central aortic and left ventricular pressure obtained at simultaneous transbronchial left heart and retrograde aortic catheterizations. There is a peak systolic gradient of 120 mm. Hg. (Reproduced by permission from Progress in Cardiovascular Diseases 1: 80, 1958)

Fig. 2 Bottom. Pressure tracings recorded in patient R. R. with subvalvular stenosis. Transbronchial catheter was withdrawn across aortic valve, through subvalvular chamber and then across subvalvular membrane into main left ventricular cavity (L.V.) and left atrium (L.A.). These tracings were made in the course of the same left heart catheterization summarized in table 2B but at a different time. Left ventricular pressure and gradient in table 2B are maximums observed.
sented clear evidence of subvalvular pulmonary stenosis with a systolic pressure gradient of 16 mm. Hg across the right ventricular outflow tract. Several other patients had smaller but significant gradients (tables 3A and 3B). These observations suggest encroachment by the enlarged left ventricle on the interventricular septum and right ventricular outflow tract. In the course of right heart catheterization cardiac outputs were determined by the Fick method. In 10 patients the resting oxygen consumptions ranged from 106 to 156 ml./min./M.\(^2\) BSA; these data were associated with basal respiratory quotients and indicated that the calculated cardiac outputs were representative of the steady basal state. The cardiac indices ranged from 1.50 to 4.61 and averaged 3.28 L./min./M.\(^2\) BSA.

Operative Technic

Eighteen patients with congenital aortic stenosis have been operated upon at the National Heart Institute. In the first 4 patients
CONGENITAL AORTIC STENOSIS

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Table 3B.—Right Heart Catheterization Data in Patients with Subvalvular Stenosis and with Indeterminate Site of Obstruction*

<table>
<thead>
<tr>
<th>Site of stenosis</th>
<th>Patient</th>
<th>Status</th>
<th>Condition</th>
<th>PC mean (mm. Hg)</th>
<th>PA S/D m (mm. Hg)</th>
<th>RV S/D (mm. Hg)</th>
<th>RA mean (mm. Hg)</th>
<th>Cardiac index (L/min./M.² BSA)</th>
<th>VO₂/M.²</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>C. C.</td>
<td>Preop.</td>
<td>Rest</td>
<td>6</td>
<td>27/10 16</td>
<td>27/0</td>
<td>6</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>M. S.</td>
<td>Preop.</td>
<td>Rest</td>
<td>11</td>
<td>35/7 20</td>
<td>33/5</td>
<td>2</td>
<td>3.60</td>
<td>120</td>
</tr>
<tr>
<td></td>
<td>U</td>
<td>Postop.</td>
<td>Rest</td>
<td>7</td>
<td>28/9 —</td>
<td>35/5</td>
<td>2</td>
<td>2.95</td>
<td>128</td>
</tr>
<tr>
<td></td>
<td>B</td>
<td></td>
<td>Exer.</td>
<td>10</td>
<td>33/14 18</td>
<td>—</td>
<td>—</td>
<td>3.66</td>
<td>360</td>
</tr>
<tr>
<td></td>
<td>A</td>
<td>Preop.</td>
<td>Rest</td>
<td>10</td>
<td>30/11 15</td>
<td>32/3</td>
<td>3</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>L</td>
<td>Postop.</td>
<td>Rest</td>
<td>8</td>
<td>28/9 16</td>
<td>30/2</td>
<td>3</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>V</td>
<td></td>
<td>Exer.</td>
<td>—</td>
<td>33/8 23</td>
<td>—</td>
<td>—</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>L</td>
<td>Preop.</td>
<td>Rest</td>
<td>11</td>
<td>22/9 13</td>
<td>29/4</td>
<td>3</td>
<td>1.50</td>
<td>113</td>
</tr>
<tr>
<td></td>
<td>R</td>
<td>C. G.</td>
<td>Preop.</td>
<td>12</td>
<td>20/12 15</td>
<td>71/4</td>
<td>3</td>
<td>3.00</td>
<td>147</td>
</tr>
<tr>
<td></td>
<td>A. Z.</td>
<td>Preop.</td>
<td>Rest</td>
<td>15</td>
<td>29/12 19</td>
<td>27/5</td>
<td>2</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>M. M.</td>
<td>Preop.</td>
<td>Rest</td>
<td>3</td>
<td>—</td>
<td>35/5</td>
<td>1</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>C. MCG.</td>
<td>Preop.</td>
<td>Rest</td>
<td>8</td>
<td>27/13 15</td>
<td>27/0</td>
<td>2</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>I</td>
<td>J. R.</td>
<td>Preop.</td>
<td>9</td>
<td>45/12 24</td>
<td>65/5</td>
<td>3</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>J. D.</td>
<td>Preop.</td>
<td>7</td>
<td>30/10 16</td>
<td>36/—</td>
<td>—</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>T</td>
<td>W. F.</td>
<td>Preop.</td>
<td>22</td>
<td>58/34 46</td>
<td>58/6</td>
<td>4</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>E</td>
<td>Postop.</td>
<td>Rest</td>
<td>6</td>
<td>22/7 11</td>
<td>22/2</td>
<td>—</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>M</td>
<td>D. F.</td>
<td>Preop.</td>
<td>—</td>
<td>25/10 15</td>
<td>32/4</td>
<td>5</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>I</td>
<td>W. D.</td>
<td>Preop.</td>
<td>—</td>
<td>35/17 26</td>
<td>32/2</td>
<td>—</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>Exer.</td>
<td>—</td>
<td>41/22 31</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>T</td>
<td>M. F.</td>
<td>Preop.</td>
<td>10</td>
<td>23/8 15</td>
<td>25/5</td>
<td>4</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>E</td>
<td>L. A.</td>
<td>Preop.</td>
<td>28</td>
<td>85/55 70</td>
<td>85/6</td>
<td>3</td>
<td>With patent ductus after closure</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Rest</td>
<td>—</td>
<td>7</td>
<td>50/20 28</td>
<td>42/0</td>
<td>2</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

* Symbols and abbreviations as in table 3A.

A closed transaortic or transventricular operation was employed. Operation under direct vision by means of general hypothermia was carried out in the next 11, and the last 3 operations were performed with the aid of extracorporeal circulation and elective cardiac arrest with the patients at normal temperature. The open operation with hypothermia was employed after its safety and simplicity had been demonstrated in this laboratory by Kaiser, Gaertner, and Kay. The clinical application of a similar technic has been reported by Swan and Lewis.

The management of patients undergoing general hypothermia in this clinic has been described. Because of the experimental demonstration of myocardial failure following hypothermia and inflow occlusion, and its prevention by digitalis glycosides, all patients were digitalized preoperatively. Patients were cooled by immersion to an ultimate esophageal temperature of 30 to 32 C. When
the operation under direct vision is carried out at normal temperature and with the aid of extracorporeal circulation, a Kay-Cross oxygenator with occlusive roller pumps is employed.

When either hypothermia or perfusion is used, a complete median sternotomy is made and the right pleural space and pericardium are opened wide (fig. 3). Tapes are passed about the superior and inferior venae cavae within the pericardium and the entire ascending aorta is freed of pericardium and adventitia and separated from the pulmonary artery. This dissection is carried as close as possible to the aortic annulus and is facilitated by retraction of the right atrial appendage and the fat pad in the atrioventricular groove. Care must be taken to avoid injury to the right coronary artery.

A partially occluding clamp is applied to the anterolateral wall of the aorta excluding a segment 2 to 3 cm in length (fig. 4). An incision is made in the excluded aorta and stay sutures are placed. The venae cavae are occluded (hypothermia) or cannulated and occluded (extracorporeal circulation) and the aorta is cross clamped just below the innominate artery. With perfusion the heart is stopped by the injection of 2.5 per cent potassium citrate in blood into the occluded aorta. The aortic incision is opened, its margins are carefully retracted, and the residual blood is aspirated from the aorta and the left ventricle. The valve is easily visualized (fig. 5). If valvular stenosis is present, the commissures are carefully identified and divided in turn with scissors (fig. 6B). The commissures are cut to within a few millimeters of the...
aortic wall and their division is then completed with a Brock or Bailey dilator. If the valve is found not to be stenotic, the leaflets are grasped and retracted by an assistant. The subvalvular obstruction may then be cut, dilated, or resected with a Brock infundibular punch (fig. 6A).

After the obstruction has been relieved, residual air is flushed from the left ventricle and aorta, the partially occluding clamp is reapplied, the cross clamp removed, and the aortic incision closed with a continuous suture. The periods of inflow occlusion have varied from 2 to 5 minutes with hypothermia. Measurements of the valve gradient and cardiac output have ordinarily been made before and after valvulotomy. The details of these technics have been described elsewhere.16

Results of Operation

Of the 18 patients operated upon, 15 are alive and free of cardiovascular symptoms. One patient (D. S.) died of irreversible ventricular fibrillation following the period of inflow occlusion. Another (H. L.) developed Pseudomonas septicemia and died 5 weeks postoperatively; at autopsy infected granulation tissue was present in the aortic suture line. The third patient (S. D.) died 10 days postoperatively from massive intrathoracic hemorrhage following the administration of heparin and Dicumarol given in the treatment of femoral thrombophlebitis. No bleeding point could be identified at autopsy; the aortic incision was well healed. One patient (G. G.) had recurrent right pleural effusions for several months and another, (J. B.) operated upon by the closed transaortic route, developed epileptiform seizures 6 months postoperatively. The relationship between the onset of these seizures and the operation is obscure. One patient (R. R.) exhibits the electrocardiographic configuration of left bundle-branch block in postoperative tracings.

Postoperatively the systolic murmur was usually decreased in intensity but was never abolished and was sometimes unchanged. In the relatively short follow-up period significant reductions in heart size have not consistently been observed. In general, postoperative electrocardiograms have shown a tendency toward normal in the amplitude of RV6 and SV1 (fig. 7).

To date, postoperative measurements of left heart pressures have been carried out in 13 of the patients operated upon. The aortic valve gradient was found to be decreased in every instance (fig. 8), but was abolished in only 3 patients. The aortic valve gradients fell from 18 to 172 mm. Hg and the average decrease was 72 mm. Hg. In the 1 patient

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**Fig. 4.** Aortotomy is made within a partially occluding clamp and its edges are retracted. When extracorporeal circulation is employed, venae cavae and femoral artery are cannulated at this time.
Fig. 5 Top. Subvalvular (A) and valvular (B) congenital aortic stenosis as seen in the course of operation under direct vision. (Reproduced by permission from Progress in Cardiovascular Diseases 1: 80, 1958)

Fig. 6 Bottom. Operative management of subvalvular (A) and valvular (B) obstructions under direct vision. Subvalvular membrane is exposed by retraction of leaflets, and is cut and dilated.

(W. F.) who had significant pulmonary hypertension preoperatively, a right heart catheterization 9 months after operation revealed that all right heart pressures were normal.

Peripheral arterial pulse pressures were generally increased postoperatively. Three patients have abnormally low diastolic arterial pressures and the auscultatory findings of aortic regurgitation. Two of these patients (N. S. and E. R.) had aortic diastolic murmurs before operation but it is believed that the commissurotomy increased the pre-existing
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Fig. 7 Top. Electrocardiograms made before operation (upper) and 1 year after operation (lower) in patient A. Z. with subvalvular obstruction.

Fig. 8 Bottom. Aortic valve gradients before and after operation in the 13 patients in whom both measurements were made.

Aortic regurgitation. The aortic regurgitant flows in these 2 patients were found postoperatively to be 4.7 and 1.6 L./min. respectively. In the other patient (G. G.) an aortic diastolic murmur was heard for the first time after operation. Patient R. R. had a grade II diastolic murmur, which was unchanged after operation. His postoperative regurgitant flow was estimated at 0.6 L./min. These patients are all asymptomatic although 1 (E. R.) has shown a slight increase in heart size roentgenographically.

DISCUSSION

There is now abundant evidence that congenital stenosis is a lesion that may pose a threat to life. In the present series, 2 patients (D. M. and J. R.) died suddenly on the ward while awaiting operation. Some observers have had similar experiences.1-3 Hemodynamic evidences of severe outflow obstruction and elevated left ventricular filling pressure were frequent in the patients studied. Many patients in this and other series were severely incapacitated by angina, syncope, and dyspnea, and the incidence of sudden death has been estimated at nearly 10 per cent.3 Subvalvular stenosis has, in particular, been regarded as a benign lesion, offering little obstruction to left ventricular outflow in the vast majority of cases.17 Three of the largest aortic gradients in the present series, however, were recorded in patients with subvalvular obstruction and 2 of them were severely limited by angina and dyspnea.

The diagnosis of aortic stenosis is ordinarily apparent from the characteristic harsh systolic murmur and thrill at the base of the heart that are transmitted to the carotid arteries. The lesion may be considered congenital if the murmur is detected in infancy or in early childhood and if there is no evidence of other valve involvement or a history of rheumatic fever. Congenital aortic stenosis may be masked by the presence of associated anomalies, the most common of which are patent ductus arteriosus18 and coarctation of the aorta. The chief value of right heart catheterization is in the detection of patent ductus and in the differentiation of aortic stenosis from ventricular septal defect or pulmonic stenosis. This technic is of little value in estimating the severity of the aortic lesion itself. With 1 exception the resting cardiac indices measured were within normal limits. It was of interest that no correlation existed between the aortic valve gradient and the resting cardiac output.

Subaortic stenosis has been considered to be a very rare congenital anomaly of the heart; a total of 45 cases had appeared in the medical literature before 1949.17 It was
therefore of considerable interest that the obstruction was subvalvular in 8 of the 21 patients in whom the diagnosis was proved (at operation in 17 and by left heart catheterization in 4). The valvular and subvalvular form of congenital aortic stenosis, in the present series, could not be distinguished on the basis of clinical findings, electrocardiography, or standard roentgenographic techniques. Poststenotic dilatation of the aorta was present in both groups. Intravenous or selective angiocardiograms with pulmonary artery injection were carried out in 11 of the patients in this series. In no instance could the site of obstruction be precisely located. Direct injections of contrast substances into the left heart have not been employed in patients with aortic stenosis in this clinic. The only method by which the site of obstruction could be localized before operation was transbronchial left heart catheterization. The presence of subvalvular stenosis was established in 4 of the 8 patients with this lesion when the catheter tip was withdrawn from the aorta through an area where the systolic pressure was equal to that in the aorta and the diastolic pressure was that of the main left ventricular chamber. In the other 3 patients with subvalvular stenosis in whom the transbronchial route was employed the catheter could not be manipulated into the aorta.

The incidence of aortic regurgitation among the patients with subvalvular obstruction is high. Aortic diastolic murmurs were present preoperatively in only 4 of the 13 patients with valvular stenosis, while 4 of the 8 patients with proved subvalvular obstruction had the murmur of aortic regurgitation. In 1 of the latter (H. L.) the presence of significant aortic reflux was proved preoperatively by an indicator-dilution technic. When aortic valvular incompetence is encountered in association with a subvalvular membrane, its etiology must remain a matter for speculation. At operation the valve leaflets have frequently been noted to be thickened and fibrotic. This observation was further confirmed by the autopsy findings in patient H. L.; the leaflets were considerably thickened, their edges rolled, and they did not approximate centrally. Similar pathologic observations have been made by Burchell and Edwards, who attributed the valvular damage to the impact of the jet of blood from the subaortic orifice. Deformity of the valve may also result from a previous, unrecognized and healed bacterial endocarditis. Edwards believes that this complication may also be related to the trauma of the jet impact on the valve.

Ideal management of the patient with congenital aortic stenosis requires an accurate estimate of the severity of the obstruction. In the present series prominent symptoms were always associated with severe hemodynamic alterations. On the other hand, both hemodynamic and anatomic evidence of severe obstruction was present in several patients who had not yet developed symptoms. In general, electrocardiographic evidence of left ventricular hypertrophy was accompanied by a large pressure gradient but exceptions to this were occasionally noted. Precise evaluation of the degree of obstruction to left ventricular outflow depends not only upon the measurement of the pressure gradient but also upon the simultaneous determination of blood flow across the aortic valve. The recent combination of the technics of left heart catheterization and measurement of cardiac output by the indicator-dilution method has made possible the preoperative estimation of orifice size. Such measurements materially aid in the selection of patients for operation. It is currently the policy at the National Heart Institute to recommend operation to patients with congenital aortic stenosis who, regardless of symptoms, have aortic valve gradients in excess of 50 mm. Hg or calculated orifice areas less than 0.5 cm.²/M.² BSA. Further experience may indicate changes in these arbitrary standards.

Congenital valvular aortic stenosis is, in general, a more favorable anatomic lesion than acquired stenosis. Calcification may occur in the congenitally narrowed valve, and was present in the 5 oldest patients in this series. The transventricular and the closed transaortic technics for aortic valvulotomy seem particularly unsuitable for the treatment of
CONGENITAL AORTIC STENOSIS

Congenital stenosis, since accurate division of congenitally fused commissures would appear to be difficult without direct vision. This conclusion is borne out by the experiences of Marquis and Logan,22 who reported severe aortic insufficiency in 4 of 6 patients operated upon through the left ventricle. Relief of subvalvular obstruction through an aortic pouch has not been reported and would seem to be difficult or impossible.

The open operation described provides excellent exposure of the valve and permits precise control of the commissurotomy. Complete anatomic correction with separation of the commissures to the annulus was at first attempted and was probably responsible for the development or aggravation of aortic insufficiency in some patients. At present the commissures are divided only partially and the valve is opened further by dilatation. The valve in 1 patient (G. G.) appeared to be bicuspid, and the possible occurrence of this variation must be borne in mind lest a leaflet be damaged in an attempt to open a nonexistent commissure. Among the 7 patients with subvalvular stenosis, the obstruction was due to a fibrous ring in 6; it was easily cut and dilated. In the other patient (A. Z.) a thick musculofibrous ridge occluded the outflow tract, and division and dilatation were carried out. In this instance, however, resection of the obstructing tissue might have been preferable.

Although most patients surviving operation have had gratifying clinical results, postoperative hemodynamic studies in all but 5 indicate that some degree of residual stenosis exists. The necessarily brief period allowed for visualization of the valve is the chief disadvantage of the open operation with hypothermia. A logical extension of the method has been the application of a similar technic during elective cardiac arrest or retroperfusion of the coronary arteries with circulatory support by means of extracorporeal circulation.23, 24

Summary

The clinical and hemodynamic findings in 30 patients with congenital aortic stenosis are summarized, and the potentially serious nature of this defect is emphasized. The use of left heart catheterization in the precise assessment of the severity of the obstruction and in the detection of subvalvular stenosis is described. Eighteen patients were operated upon, 14 with direct exposure of the aortic valve. The technic, advantages, and limitations of the open operation are discussed, and the clinical and hemodynamic findings in patients studied after operation are presented.

Summario in Interlingua

Es summarisate le constataiones clinic e hemodynamic in 30 patientes con congenite stenosis aortice. Es sublineate le potentialmente grave character de iste defecto. Es des Ribarite le uso de catheterismo sinistro-cardiaco in le evaluatation precise del severitate del obstructione in le detection del presenta de stenosis subvalvular. Dec-octo patientes esseva operate, 14 con exposition directe del valvula aortice. Le technica, le avantages, e le limitationes del operation aperte es discutite, e le constataiones clinic e hemodynamic in patientes studiate post le operation es presentate.

REFERENCES


TRUTH AND LIBERTY

JOHN MILTON

English Puritan poet, 1608-1674

Where there is much desire to learn, there of necessity will be much arguing, much writing, many opinions; for opinion in good men is but knowledge in the making.— Areopagitica. From Great Companions. Readings on the Meaning and Conduct of Life from Ancient and Modern Sources. Vol. I, Boston, The Beacon Press, 1952.
Congenital Aortic Stenosis: Clinical and Hemodynamic Findings, Surgical Technic, and Results of Operation
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