Recognition of Congenital Heart Disease in the Fifth to Eighth Decades of Life

Diagnostic Criteria and Natural History

By J. M. Fisher, M.D., W. R. Wilson, M.D., and E. O. Theilen, M.D.

CONGENITAL HEART DISEASE used to be a stepchild and a curiosity for a few clinical collectors of the rare. Most clinicians were content to make a diagnosis of "congenital heart disease" and let the matter rest there. A few like Maud Abbott and Helen Taussig correlated and collated their own considerable experience but still there was surprisingly little information on the natural history of congenital heart disease when a new era of surgery suddenly emerged. Since so many people now have partially or sometimes completely corrective procedures done, it will be increasingly difficult to accumulate information on the natural history of various forms of congenital heart disease. For this reason we have examined the incidence of congenital heart lesions in our hospital and have reviewed the clinical course in patients who have survived to middle age and beyond. Congenital heart disease may be overlooked in adults because of the preponderance of acquired heart lesions and the almost unavoidable bias to which this difference in frequency predisposes. Since misdiagnoses are fairly common, the physician must be alert and avoid casual mistakes. Lack of symptoms until middle or later years does not exclude congenital heart disease and an indifferent attitude about precise diagnosis is not warranted because of the possibility of surgical correction with low risk and with reasonable expectation of clinical improvement.

Material and Results

We have reviewed the autopsy records in our hospital from 1950 to 1959. Two hundred and ten cases of congenital heart disease were found in 4,936 examinations. The incidence of various lesions is given in table 1. Atrial septal and ventricular septal defects were the most common congenital heart abnormalities and occurred as isolated lesions or as part of more complex malformations. The majority died before 1 year of age. We found only 17 patients with congenital heart disease between 20 and 40 years of age. The most common lesions were atrial septal defects. There were only eight patients over 40 years of age with congenital lesions, and again atrial septal defects predominated. Children with asymptomatic ventricular septal defects are not uncommon in this hospital, and yet very few adults can be found on our wards or in our clinics with ventricular septal defects. What becomes of these patients in later life? Autopsy data do not tell us, and this is still an unanswered question. Reports of spontaneous closure of ventricular septal defects have appeared in the literature recently.1,2

We have also reviewed 1,400 cardiac catheterizations done in this hospital since 1951 and have found 50 patients over 40 years of age with congenital heart defects. Table 2 shows the incidence of the various congenital heart lesions with age range and sex distribution. The youngest was 40 years old and the oldest 78 years old when first seen in our hospital.

Atrial Septal Defects

Atrial septal defects were the most common, occurring in 25 patients. Three also had partial anomalous venous return. No other associated lesions were found. The difficulties in diagnosis in older people were apparent

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Table 1

**Incidence of Lesions in 210 Patients with Congenital Heart Disease among 4,936 Autopsies (1950-1959)**

<table>
<thead>
<tr>
<th>Lesion</th>
<th>Total number</th>
<th>Age range (years)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Under 1</td>
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<tr>
<td>Ventricular septal defect</td>
<td>69</td>
<td>53</td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>67</td>
<td>42</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>27</td>
<td>5</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>24</td>
<td>22</td>
</tr>
<tr>
<td>Transposition of great vessels</td>
<td>24</td>
<td>21</td>
</tr>
<tr>
<td>Coarctation of aorta</td>
<td>23</td>
<td>19</td>
</tr>
<tr>
<td>Pulmonic stenosis (valvular)</td>
<td>14</td>
<td>5</td>
</tr>
<tr>
<td>Aortic valve anomalies</td>
<td>12</td>
<td>3</td>
</tr>
<tr>
<td>Others*</td>
<td>16</td>
<td>11</td>
</tr>
</tbody>
</table>

*Includes single ventricle, truncus, tricuspid atresia, a-v communis.

Table 2

**Congenital Heart Lesions in 50 Patients Over 40 Years of Age**

<table>
<thead>
<tr>
<th>Lesion</th>
<th>Number of patients</th>
<th>Age range (years)</th>
<th>Sex</th>
<th>Male</th>
<th>Female</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrial septal defect</td>
<td>25</td>
<td>40-78</td>
<td>7</td>
<td>18</td>
<td></td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>12</td>
<td>40-61</td>
<td>3</td>
<td>9</td>
<td></td>
</tr>
<tr>
<td>Pulmonic stenosis</td>
<td>6</td>
<td>40-70</td>
<td>4</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>6</td>
<td>40-57</td>
<td>2</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>1</td>
<td>44</td>
<td></td>
<td></td>
<td>1</td>
</tr>
</tbody>
</table>

in this group. Twelve had been misdiagnosed as rheumatic heart disease before admission to our hospital, two as cor pulmonale and one as arteriosclerotic heart disease. The correct diagnosis had been suspected in only four patients.

The symptoms were nonspecific in general, but the rapid progression after onset of symptoms in some patients should be emphasized. Shortness of breath on exertion, increased fatigability, and palpitation were common complaints. These symptoms had been present for only a short period in many patients although a heart murmur had been heard in some during childhood. No history of frequent respiratory infections or slow growth in childhood could be obtained from any of these patients with atrial septal defects. Cardiac findings were quite typical and included right ventricular over-accessibility, a low-pitched systolic murmur in the second left intercostal space along the sternal border, and splitting of the second sound in the pulmonic area, with accentuation of the second component. A low-pitched diastolic flow rumble was heard at times in the third and fourth left intercostal spaces, and occasionally at the apex. It is possible that the diastolic flow rumble may have been misinterpreted as the murmur of mitral stenosis by some, as this was the most common admission diagnosis. Mitral insufficiency was another common misdiagnosis before admission to our hospital. This should not be a common error, since the systolic murmur in patients with atrial septal defects is usually heard best in the pulmonic area. Left ventricular enlargement is usually not present as it is in mitral insufficiency unless the defect is of the ostium primum type, or unless the patient has superimposed heart disease.

Patients with atrial septal defects also present a fairly typical picture radiographically. The x-ray changes in a patient with an atrial septal defect of the secundum type are shown in figure 1. They consist of increased pulmonary vasculature, a prominent pulmonary artery segment, and cardiac enlargement due primarily to right ventricular hypotrophy and some right atrial enlargement. Left atrial enlargement is not seen often. At cardiac fluoroscopy a hilar dance is seen unless pulmonary hypertension is severe. Posteroanterior chest x-rays of patients with severe mitral stenosis may resemble superficially those of patients with atrial septal defects, but
failure to find signs of a left-to-right shunt, and enlargement of the left atrium and B-
lines of Kerly should aid in differentiating these two lesions. Figure 2 shows a chest
x-ray of our oldest patient who is now nearly 80 years of age. She had no cardiac symp-
toms and the atrial septal defect was found at the time of admission for gallbladder dis-
eease. The electrocardiogram showed an in-
complete right bundle-branch-block pattern seen in the majority of patients with atrial septal defects. Cardiac catheterization re-
vealed a left-to-right shunt of 47 per cent and essen-
tially normal pulmonary arterial pres-
sures. A moderate-sized secundum defect is certainly compatible sometimes with long life
without any incapacity.

Table 3 shows hemodynamic data on our patients with atrial septal defects. The left-
to-right shunts were significant in all cases and very large in some. Pulmonary artery
pressures on the other hand were normal or
only moderately increased in the majority.
These findings suggest that even large shunts into low pressure areas need not produce se-
vere or irreversible pulmonary hypertension.
Two of our patients with moderate pulmonary
artery hypertension (70 mm. Hg systolic) had
normal pulmonary artery pressures on recath-
ereterization after closure of the defects. A
total of 10 patients over age 40 have had sur-
gical closure of their atrial septal defects. All
have shown excellent clinical improvement.
This is in accord with the reports of others.3-5

Patent Ductus Arteriosus

Patent ductus arteriosus was the second
most common congenital heart lesion, occur-
ing 12 times in this series. The age range
was from 40 to 61 years. Fewer diagnostic
difficulties were encountered in this group,
since all the patients had typical machinery-
like murmurs in the pulmonic area. Two
patients were thought to have aortic insuffi-
ciency and aortic stenosis. Aneurysm of the
aorta, possible pulmonary A-V fistula, and
mitral valve disease were erroneous admission
diagnoses in three instances. The heart mur-
mur had been heard in childhood in the others
and congenital heart disease was suspected.
Symptoms included shortness of breath on
exertion, palpitation, and fatigability; these
had been present for 3 to 50 years.
Enlargement of the left ventricle and left atrium, a prominent pulmonary artery segment, and evidence of a left-to-right shunt are the usual radiographic signs of this lesion. The right ventricle may be enlarged also if there is pulmonary hypertension. The 45-year-old patient whose film is shown in figure 3 was asymptomatic despite cardiac enlargement and moderate pulmonary hypertension. Preoperative cardiac catheterization revealed a pulmonary artery pressure of 60/35 mm Hg; this decreased to 39/11 mm Hg after operation. The electrocardiogram in such patients is not specific but usually shows left ventricular hypertrophy or combined ventricular hypertrophy, if pulmonary hypertension is severe. Table 3 shows the hemodynamic data on the 12 patients with patent ductus arteriosus. A significant left-to-right shunt was present in all patients, and in general these patients showed a greater increase in pulmonary artery pressure than the patients with atrial septal defects with the same degree of shunt. Four of these patients have had cardiac surgery. Three had definite clinical improvement, despite significant pulmonary hypertension before operation. The fourth patient was explored but the ductus arteriosus was not closed because of severe calcification.

**Pulmonic Stenosis**

Six patients had isolated pulmonic stenosis. Symptoms referable to heart disease were present in only two of the six, and both of these had congestive failure. One was 70 years old and was thought to have coronary artery disease in addition to his congenital lesion; the other had severe infundibular stenosis and had only mild heart failure. Figure 4 shows an angiocardiogram of an asymptomatic 46-year-old man who had been sent to the hospital with a diagnosis of a bronchogenic cyst. The angiogram revealed aneurysmal dilatation of the left pulmonary artery secondary to the pulmonic stenosis. The pulmonary artery segment was prominent in all our patients, and four had obvious right ventricular enlargement. Three had some radiographic evidence of left ventricular enlargement, which was probably associated with degenerative heart disease and heart failure. The electrocardiogram did not always exhibit the typical right ventricular hypertrophy seen commonly in younger patients with moderate hypertension.

### Table 3

<table>
<thead>
<tr>
<th>Lesion</th>
<th>Number of patients</th>
<th>Systolic Range</th>
<th>Mean Range</th>
<th>Left-to-right shunt Range</th>
<th>Average</th>
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<tr>
<td>Atrial septal defect</td>
<td>25</td>
<td>20-104</td>
<td>51</td>
<td>16-70</td>
<td>51</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
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<td>17-116</td>
<td>56</td>
<td>12-96</td>
<td>42</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>5</td>
<td>18-37</td>
<td>26</td>
<td>11-23</td>
<td>16</td>
</tr>
</tbody>
</table>

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to severe pulmonic stenosis. Some may have been modified by associated left ventricular hypertrophy.

Table 4 shows the ages and systolic gradients across the pulmonic valve of our six patients. The age range was 46 to 70 years. The patient with infundibular stenosis who was in mild congestive heart failure had a gradient of 85 mm. Hg between the pulmonary artery and right ventricle. Clinical signs of mild heart failure were present. The right ventricular pressure decreased to normal after surgery and there was no systolic gradient. Clinical improvement was dramatic. The second patient had an initial gradient of 92 mm. Hg even though he was asymptomatic and had a normal electrocardiogram. Following valvulotomy his gradient fell to 21 mm. Hg. The gradients were slight in the next three patients, but the last patient had moderately severe pulmonic stenosis with symptoms at the age of 61 years.

Ventricular Septal Defects

Only six patients with ventricular septal defects were found past the age of 40 years. Four of these had no symptoms. Heart murmurs were heard in early childhood in five, and they were thought to have congenital heart disease. The murmur was discovered on routine physical examination for herniorrhaphy in one patient at age 57. Cardiac catheterization in five patients revealed normal pulmonary artery pressures and small left-to-right shunts. The sixth patient was in congestive failure with intermittent cyanosis and severe pulmonary artery hypertension.

Electrocardiograms and chest x-rays were normal in the five patients with small shunts and normal pulmonary arterial pressures. The patient with severe pulmonary hypertension had right ventricular hypertrophy on electrocardiogram and chest x-rays.

Tetralogy of Fallot

One patient was seen recently with a tetralogy of Fallot. He had a Blalock operation 12 years before and did quite well clinically until 2 years before his examination here. No record of any other patients with cyanotic congenital heart disease over the age of 40.

Table 4

<table>
<thead>
<tr>
<th>Patients</th>
<th>Age (years)</th>
<th>Functional class</th>
<th>Right ventricular systolic pressure (mm. Hg)</th>
<th>Gradient (mm. Hg)</th>
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<tr>
<td>1</td>
<td>50</td>
<td>III</td>
<td>100</td>
<td>85</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>Post-op. 28</td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>110</td>
<td>92</td>
</tr>
<tr>
<td></td>
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<td></td>
<td>Post-op. 45</td>
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<td>46</td>
<td>I</td>
<td>67</td>
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<td>57</td>
<td>I</td>
<td></td>
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<tr>
<td>6</td>
<td>61</td>
<td>I</td>
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</tbody>
</table>

*Figure 4*

Angiogram showing aneurysmal dilatation of pulmonary artery.
Summary and Conclusions

Fifty patients with congenital heart disease in the fifth to eighth decades of life have been reviewed. Atrial septal defects were the most common lesions and were compatible with long life. Symptoms may occur late but are often rapidly progressive. Despite large left-to-right shunts, pulmonary artery pressures are often normal or only moderately increased. Misdiagnoses are common. Atrial septal defects are confused most frequently with rheumatic heart disease. A diagnosis of mitral stenosis or mitral insufficiency was made in over 50 per cent of this group before admission to our hospital. Some patients with a patent ductus arteriosus may survive to old age. The pulmonary artery pressure tends to be higher in this group than in those with atrial defects with a similar magnitude of left-to-right shunt. Patients with moderately severe pulmonic stenosis may be asymptomatic. Patients with ventricular septal defects show a high attrition rate before 40 years of age, and those surviving past 40 seem to have small and dynamically insignificant lesions. Cyanotic congenital heart disease is very rare after 40 years of age.

Age alone should not be a deterrent to surgical correction of congenital heart defects.

Moderate pulmonary artery hypertension is not a contraindication either, and was reversible in a number of our patients. In view of these findings, it would seem important to consider congenital heart disease in the differential diagnosis of all patients with suspected cardiac disease, even in individuals of middle age and beyond. An accurate diagnosis should be sought, since surgical correction may be possible with considerable benefit to the patient.

References


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