Congenital Aortic Septal Defect with Communication between Aorta and Pulmonary Artery

Case Report and Review of Literature

By Herta Spencer, M.D., and Harvey J. Dworken, M.D.

A case of congenital defect of the aortic septum is reported, together with a summary of thirteen previously described cases. The authors briefly describe their concept of the dynamics of the lesion and suggest a method of possible surgical correction, in instances where the diagnosis is made early.

RECENTLY we have had the opportunity of examining a patient with a congenital defect between the base of the aorta and the pulmonary artery. A careful review of previous reports since the early 1800s revealed only 13 other proved instances of the anomaly, 10 of which were tabulated by Maude Abbott in her extensive monograph on congenital heart disease. As in our patient, the correct anatomic diagnosis was not made in any instance prior to postmortem examination.

CASE REPORT

G. W., an 18 year old white male, was admitted to Mount Sinai Hospital on December 20, 1949 complaining of extreme shortness of breath, orthopnea, and increasing weakness during the previous five weeks. Marked dyspnea first appeared while the patient was walking leisurely, and readily disappeared with rest.

From his father, it was learned that the patient, an only child, was born with "a leakage of the heart" and a hare lip. However, his general development had been normal and, until recently, he had been able to exercise moderately and had never been cyanotic.

There was no history of rheumatic fever except for one episode in 1947, consisting of pain in both legs. This lasted for nine days, but fever or joint swelling did not appear.

The only x-ray film of the chest ever taken was during the Cleveland Tuberculosis Survey in April, 1949 (fig. 1). This showed an elongated chest with increased radiolucency of both lungs, indicating emphysema. The cardiothoracic ratio was 17.8:26.7 cm. The left ventricle was enlarged, extending to the extreme left of the chest, and the right heart border was moderately enlarged. The pulmonary artery and hilar vessels were prominent, and the aorta was slightly dilated.

On examination, the patient appeared undernourished, pale, severely orthopneic and moderately cyanotic. Temperature was 99.2 F., pulse 92 and grossly irregular, respirations 30, blood pressure 130/70. There was a hare lip deformity on the left, contiguous with a complete cleft palate. Neck veins showed marked engorgement. The thorax was increased in anteroposterior diameter, the left chest being more prominent than the right. There were diminished breath sounds and rales at both lung bases.

Precordial activity was marked, and a systolic thrill was felt, most intense in the third left intercostal space adjacent to the sternum. The left cardiac border was in the seventh intercostal space at the posterior axillary line. The rhythm was completely irregular, and a grade IV systolic murmur was present, loudest in the third left intercostal space adjacent to the sternum. The presence of a diastolic murmur in this area was debatable. Peripheral pulses were collapsing.

The abdomen showed a definite fluid wave. The liver edge was felt four fingerbreadths below the costal margin. Marked edema of the scrotum and lower extremities was present. Digital clubbing was absent.

The urine showed a trace of albumin and a few red and white cells in the sediment. Hemoglobin was 15.3 Gm., red blood cell count 5.8 million, white cell count 13,700. The nonprotein nitrogen was 130 mg. per 100 cc, the carbon dioxide combining power 25.8 volumes per cent. Kline test and a blood culture were negative.

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varying from 0.14 to 0.16 second. The pattern was not specific for any type of bundle branch block.

The patient was in extremis and did not respond to diuretic or digitalis therapy. He expired 21 hours after admission. Because of the short hospital course, it was impossible to do complete diagnostic studies. Clinical diagnoses were: severe congestive heart failure, probably due to congenital heart disease, possibly a patent ductus arteriosus; possible chronic rheumatic heart disease; advanced pulmonary emphysema; terminal uremia.

At autopsy there was evidence of marked congestive failure, with massive peripheral edema and ascites amounting to 2 liters. The heart was markedly enlarged, weighing 960 Gm. The anterior surface was occupied chiefly by the right heart. Both ventricles were hypertrophied and of approximately equal thickness, the left measuring 16 mm., the right 15 mm. maximally. There was a large, round defect in the aorta (fig. 3), 12 mm. above the semilunar cusps. This opening was 20 mm. in diameter,
Fig. 3. Left ventricle, showing aorta with defect 12 mm. above the aortic valve.
(The use of color in this illustration has been made possible by a grant from Wyeth Incorporated to the publication fund of the American Heart Association.)

Fig. 4 (Left). Section of lung showing advanced emphysema. (x85)
Fig. 5 (Right). Marked thickening of the wall of a small pulmonary artery. (x85)
aorta and measured 8 cm. in circumference, the aorta 6.5 cm. at the base. There was no aortic disease, nor were any other cardiac anomalies noted.

The edges of the aortic and pulmonic valves were somewhat thickened and slightly red. The mitral valve leaflets showed rolling and thickening of the edges and there was shortening of the chordae tendineae, suggestive of an old rheumatic process. Neither the ductus arteriosus nor the ligamentum arteriosum could be identified.

The lungs showed abnormal lobulation and were distended and voluminous throughout. The liver weighed 1560 Gm. and showed marked passive congestion. The spleen was enlarged and congested to the extent of the appearance of multiple hemorrhages within the pulp.

Microscopic findings showed hypertrophy of the heart muscle; section through the septal defect showed no evidence of disease. Severe, generalized pulmonary emphysma was present (fig. 4), and the smaller pulmonary arteries and arterioles showed striking sclerotic changes with narrowing of the lumina (fig. 5). In other areas, there was chronic passive congestion evidenced by the presence of heart failure cells and thickened alveolar septa.

Discussion

Thirteen cases of localized congenital defects of the aortic septum have been reported previously to the present. Analysis of table 1 shows the age, sex incidence, size of defect, types of murmur and cause of death in these patients.

The aortopulmonary communication is due to a partial defect in the development of the aortic septum, which under normal conditions completely divides the truncus arteriosus into the two great vessels by the seventh or eighth week of intrauterine life. Usually, it is a round, smooth-edged defect in the anterior wall of the aorta, a short distance above the semilunar valves.

Aortic septal defects are to be differentiated pathologically from a shortened patent ductus arteriosus with approximation of the aorta and pulmonary artery, and the acquired type of communication caused by rupture of an aortic aneurysm into the pulmonary artery. In a shortened ductus, the lesion occurs beyond the origin of the left subclavian artery, while aortic septal defects appear just above the semilunar valves. Both lesions were present in Hektoen's case.

Table 1.—Tabulation of All Cases of Aortic Septal Defect Reported in the Literature, Including the Present Case

<table>
<thead>
<tr>
<th>No.</th>
<th>Author</th>
<th>Age</th>
<th>Sex</th>
<th>Murmurs</th>
<th>Defect</th>
<th>Associated Anomalies</th>
<th>Cause of Death</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Systolic</td>
<td>in mm.</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Diastolic</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>Elliotson</td>
<td>Infant</td>
<td>F</td>
<td>?</td>
<td>10</td>
<td>None</td>
<td>Congestive heart failure.</td>
</tr>
<tr>
<td>4</td>
<td>Gerhardt</td>
<td>5 mos.</td>
<td>F</td>
<td>?</td>
<td>5</td>
<td>None</td>
<td>?</td>
</tr>
<tr>
<td>6</td>
<td>Baginsky</td>
<td>4 yrs.</td>
<td>F</td>
<td>+</td>
<td>10</td>
<td>None</td>
<td>Pertussis, convulsions.</td>
</tr>
<tr>
<td>7</td>
<td>Caesar</td>
<td>9 yrs.</td>
<td>M</td>
<td>+</td>
<td>10</td>
<td>Perforated pulmonary cusps.</td>
<td>TBC-meningitis.</td>
</tr>
<tr>
<td>8</td>
<td>Girard</td>
<td>27 yrs.</td>
<td>M</td>
<td>+</td>
<td>10</td>
<td>None</td>
<td>Brain abscess.</td>
</tr>
<tr>
<td>11</td>
<td>Dadds &amp; Hoyle</td>
<td>14 yrs.</td>
<td>M</td>
<td>+</td>
<td>60</td>
<td>None</td>
<td>Congestive heart failure.</td>
</tr>
<tr>
<td>12</td>
<td>Erf et al.</td>
<td>20 yrs.</td>
<td>M</td>
<td>+</td>
<td>32</td>
<td>Pulmonary Hemangioma.</td>
<td>Ruptd. pulmonary hemangioma.</td>
</tr>
</tbody>
</table>

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Schattenberg and Harris\(^\text{37}\) state that three per cent of all ruptured syphilitic aortic aneurysms open into the pulmonary artery. They differ from congenital defects in that they have irregular edges and are situated higher in the aorta.\(^\text{17-19}\) Aneurysms of the sinus of Valsalva almost invariably rupture into the right heart, and are easily differentiated from aortic septal defects.\(^\text{16}\)

The cardiac history dating back to infancy, the associated malformations elsewhere in the body, and the fact that the aorta and pulmonary artery could not be dissected apart, all favor a congenital origin of the defect in the present case. The smooth edges on the opening and the absence of disease in the aorta indicate that it had not formed on an inflammatory basis.

The communication in aortic septal defects leads to circulatory changes similar to those seen in patent ductus arteriosus.\(^\text{20}\) Because of the high pressure differential, oxygenated blood flows from the aorta into the pulmonary artery. This additional flow, estimated by Eppingher, Burwell and Gross\(^\text{21}\) to be almost 50 per cent of the total output of the left ventricle, causes pulmonary hypertension, dilatation of the pulmonary artery, and eventually leads to left ventricular hypertrophy.

The right ventricular hypertrophy is a direct result of the prolonged pulmonary hypertension, which in turn was probably caused by a combination of factors: the arteriovenous shunt, the severe pulmonary emphysema and associated vascular sclerosis, and the marked congestive heart failure terminally.\(^\text{21-23}\)

The terminal cyanosis in the present case was probably due to a reversal of the arteriovenous shunt associated with congestive failure, and the advanced emphysema. The abnormal pulmonary lobulations found suggest that the emphysema had formed on a congenital basis. The marked arteriosclerosis in the lungs may have been due to pulmonary hypertension, since Parker\(^\text{24}\) found that 80 per cent of his patients with emphysema had this and ascribed it to the high pulmonary pressure in that disease.

Auricular fibrillation is rare in congenital heart diseases other than auricular septal defects or types associated with valvular disease.\(^\text{25}\) Changes of the auricular wall, anoxia or other metabolic factors may have accounted for the fibrillation in our case. Cardiac arrhythmia has been reported in only one other case of aortic septal defect.\(^\text{7}\)

Since angiocardiography has proved its value as an aid in the specific diagnosis of certain types of congenital heart disease, it may also be useful in confirming the presence of an aortic septal defect. Burford\(^\text{26}\) has introduced the retrograde arterial technic which enables one to demonstrate a patent ductus together with the aorta and pulmonary artery, and this method could conceivably outline other aortic anomalies as well. According to Cournand,\(^\text{27}\) such a method is essential in distinguishing a patent ductus from an aortic septal defect, since this cannot be done by cardiac catheterization alone.

In view of the present success of cardiac surgery, it is possible that aortic septal defects may be relieved by operation. Due to the fusion of the aorta and pulmonary artery, the communication could not be ligated in the manner of a patent ductus. A technically difficult procedure involving removal of the lesion and separately suturing the wall of the aorta and the pulmonary artery would be necessary.

**Summary**

The physical and pathologic findings in a case of an aortic septal defect are described. A careful review of the literature revealed that only 13 similar cases have been reported previously. An attempt is made to outline clinical and laboratory methods which may prove of value in the differential diagnosis of this condition. Suggestions are made for accomplishing possible surgical correction of the defect.

**Acknowledgment**

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**References**

9 Girard, E.: Quoted by Hektoen. See reference 11.
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