Complete Transposition of the Great Vessels: Importance of Septal Defects and Patent Ductus Arteriosus

Analysis of 132 Patients Dying Before Age 4

By Ib Boesen, M.D.

Complete transposition of the great vessels combined with other cardiac malformations, principally septal defects, is one of the most frequent congenital cardiac abnormalities. The prognosis is extremely poor, most patients die before the age of 6 months, and only a few survive their first year.

The purpose of this paper is to analyze the relationship between the time of death and the presence of different veno-arterial communications (i.e., atrial septal defect, ventricular septal defect, or patent ductus arteriosus) in patients with complete transposition of the great vessels. Included are only those cases in which the aorta passes in front and to the right of the pulmonary artery (a rotation of 180° of the great vessels) and in which both caval veins enter the right atrium normally.

Case Material

The study is based on a series of 1,145 children with congenital heart disease, who all died before the age of 4. In all cases autopsy was performed. Among these children, 180 were found to have complete transposition of the great vessels. From this total, 35 children were eliminated because, in addition to total transposition, they exhibited other malformations that opposed the purposes of this study. Among those excluded were six children in whom complete transposition was combined with ventricular septal defect with pulmonary stenosis and pulmonary atresia. There remained a group of 145 children, who besides complete transposition exhibited septal defects or persistent ductus, or both, or in whom right-to-left communications could not be demonstrated. In 11 of these children, the cause of death was noncardiac. Since the purpose of the present study is to analyze when the cardiac disorder leads to death, these 11 children and two mongoloid idiots as well were also excluded from the analysis. Remaining were 132 patients with complete transposition whose deaths were caused by heart failure (table 1).

Discussion

The incidence of complete transposition varies greatly in published studies, depending on the circumstances under which the material was collected. Owing to the poor prognosis, the disorder rarely occurs in older children and adults, whereas it is a salient feature of series composed of infants and young children. In the present autopsy series comprising 1,145 patients with congenital heart disease, all dead before reaching the age of 4, a total of 180 were found to have complete transposition, i.e., 16 per cent of the total series.

From a therapeutic and prognostic viewpoint it is, however, erroneous to deal collectively with the group relating solely to the transposition of the great vessels, since the nature of the attending anomalies influences the prognosis and the possibilities of surgical therapy. As already mentioned, these factors necessitated the exclusion of 35 children from our analysis, as well as 11 more, in whom the cause of death was noncardiac, and two mongoloid idiots. In this connection it should be emphasized that, since the analysis comprises only autopsy material, the table is

From The Queen Louise's Childrens Hospital, Copenhagen, Denmark.
no guide to the absolute prognosis of the disorder, inasmuch as all the children were dead before the age of 4. Further, the material was collected from 50 different children's hospitals in Scandinavia (mainly covering the period from 1945 to 1953). The collection of the cases from many hospitals may throw doubt on its value with respect to the detailed findings, since the material relates to autopsies, performed by many pathologists of whom only the minority have been especially skilled in this particular field. Consequently, a number of details possibly were not revealed which the expert would have noted. This fact, at least in part, may be the reason that in 15 of 180 children no veno-arterial communications whatsoever were demonstrated.

Since it may be of surgical interest to examine what combinations of atrial septal defect, ventricular septal defect, and persistent ductus arteriosus lead to the best prognosis, the time of death in relation to these combinations has been given in table 1, where patients with patent foramen ovale and atrial septal defect have been dealt with collectively. Although the figures in the individual spaces are not representative, this table is published in the hope that others will carry out similar analyses so as finally to collect data sufficiently comprehensive to form the basis of valid deductions.

Finally, it should be mentioned that the ratio in the present series of boys to girls of 1.4 differs from that usually published. Only in the group of 13 patients, who were excluded from the analysis because death was unrelated to the heart disorder, was a greater ratio found (i.e. 2.3). No explanation of this variation can be given.

**Summary**

In 1,145 autopsies performed on children with congenital heart disease who died before the age of 4, 180 were found to have a completely transposed aorta and pulmonary artery. Of these, 35 were excluded from the analysis because of other severe congenital cardiac malformations (among which were six with pulmonary stenosis); 11 who died from noncardiac disorders and two mongoloid

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**Table 1**

One Hundred and Thirty-two Cases of Complete Transposition of the Great Vessels with Various Combinations of Septal Defects and Patent Ductus Arteriosus. Excluded are 35 Cases Combined with Other Severe Malformations of the Heart and Great Vessels and 13 Cases Who Died from Noncardiac Disease

<table>
<thead>
<tr>
<th>Additional anomalies</th>
<th>Males</th>
<th>Females</th>
<th>Totals</th>
<th>Survival to age (days)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>8</td>
<td>7</td>
<td>15</td>
<td>10</td>
</tr>
<tr>
<td>ASD</td>
<td>10</td>
<td>3</td>
<td>13</td>
<td>10</td>
</tr>
<tr>
<td>PDA</td>
<td>16</td>
<td>6</td>
<td>22</td>
<td>10</td>
</tr>
<tr>
<td>ASD + VSD</td>
<td>2</td>
<td>4</td>
<td>6</td>
<td>6</td>
</tr>
<tr>
<td>ASD + PDA</td>
<td>16</td>
<td>21</td>
<td>37</td>
<td>20</td>
</tr>
<tr>
<td>VSD + PDA</td>
<td>7</td>
<td>4</td>
<td>11</td>
<td>5</td>
</tr>
<tr>
<td>ASD + VSD + PDA</td>
<td>3</td>
<td>4</td>
<td>7</td>
<td>3</td>
</tr>
<tr>
<td>AVC</td>
<td>2</td>
<td></td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>AVC + PDA</td>
<td>1</td>
<td>1</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>SV</td>
<td>1</td>
<td></td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>SV + PDA</td>
<td>1</td>
<td>1</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>No shunts demonstrated</td>
<td>8</td>
<td>7</td>
<td>15</td>
<td>8</td>
</tr>
</tbody>
</table>

**Totals** | 74 | 58 | 132 | 77 | 36 | 21 | 11 | 3

**Abbreviations:** ASD, atrial septal defect, included patent foramen ovale (small or large); VSD, ventricular septal defect, excluded single ventricle; PDA, patent ductus arteriosus; AVC, atroioventricularis communis; SV, single ventricle.
idiots were also excluded. The remaining 132 were analyzed with reference to the relationship between the time of death and the presence of veno-arterial communications. Forty-two per cent of the children died within 1 month and 73 per cent within the first 3 months of life. Therefore, diagnostic and therapeutic problems chiefly arise during the first months of life in patients with complete transposition of the great vessels.

References

Aneurysm

Among the diseases of the cardiovascular system aneurysm stands out as having by its obvious features forced due recognition long before the circulation was discovered, thus contrasting with cardiac disease which up to the eighteenth century was thought not to occur. Antyllos (A.D. 55-118) operated upon it, and Galen (A.D. 130-200) described two forms, the traumatic and spontaneous. The traumatic and superficial aneurysm was that mainly seen until syphilis became widespread after the return in 1493 of Columbus from the New World and the occupation of Naples by the army of Charles VIII in 1495; there was then what has been called an unprecedented occurrence of aortic aneurysm. The association with syphilis did not escape Fernelius who in 1542 first recognized internal aneurysms, Vesalius who about 1557 diagnosed both thoracic and abdominal aneurysms, and Ambroise Paré (1582) who incriminated mercurial treatment as a cause of aneurysm.

Early in this century the Wassermann reaction (1906) facilitated the detection of clinically latent syphilis, and early changes in and around the vasa vasorum of theorta (mesaortitis, Heller) and the presence there of the Spiroæma pallidum (Schaudinn and Hoffmann, 1905) fully established the paramount importance of syphilis in the etiological relationship of aneurysm.

In aneurysm, particularly intrathoracic, though syphilis is in the vast number of instances the important underlying factor, the influence of strain as an immediate exciting cause must not be overlooked; further, in some instances, luetic infection is not concerned: in old people dissecting aneurysms and abdominal aneurysms may be due to ordinary arteriosclerosis.—Sir Humphrey Davy Rolleston. The Harveian Oration. Great Britain, Cambridge University Press, 1928, p. 56.
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