Effect of Ventricular Septal Defect and Patent Ductus Arteriosus on Left Ventricular Pressure in Complete Transposition of the Great Vessels


SUMMARY
The relationship between the pressures in the left and right ventricles was examined in 80 infants and children who had complete transposition of the great vessels and in whom the presence of a ventricular septal defect or a patent ductus arteriosus was known. All children with obstruction to left ventricular outflow have been excluded. It was found that 92% (34 of 37) of the children with transposition and a ventricular septal defect had left ventricular pressures greater than 70% of the systemic, while 45% (10 of 22) of those with a patent ductus and only 5% (1 of 21) with a closed ventricular septum and no ductus, had pressures in this range. Seventy-six per cent (16 of 21) of the children in this last group had left ventricular pressures less than 50% of the systemic pressure. It is concluded that infants with transposition of the great vessels and a ventricular septal defect stand in high risk of developing obstructive pulmonary vascular disease and therapy should be guided accordingly.

Additional Indexing Words:
Pulmonary hypertension
Congenital heart disease
Obstructive pulmonary vascular disease

In recent years the outlook for infants born with complete transposition of the great vessels has improved greatly. Whereas formerly 85% of them were dead by the age of 6 months, now the problems of hypoxia and acidosis brought about by the inadequate mixing between the systemic and the pulmonary circulation can be overcome in a proportion of cases by improved palliative surgical techniques, and by the balloon atrial septostomy procedure recently introduced by Rashkind and Miller. Total surgical correction as described by Mustard is now being carried out in many centers and an understanding of the hemodynamics and natural history of children with transposition of the great vessels has become a matter of considerable importance. Ferencz has recently demonstrated the high incidence and early appearance of pulmonary vascular disease in these children. As is the case in uncomplicated ventricular septal defect (VSD) and patent ductus arteriosus (PDA), the severity of pulmonary vascular disease is known to have a profound effect on postoperative morbidity and mortality.

Hemodynamic data on children with transposition of the great vessels are scanty. Since pulmonary hypertension and pulmonary vascular disease are major factors in considering the indications for surgery, as well as prognosis, a study has been made of 80 children with this condition in whom the presence or absence of associated defects, such as ventricular septal defect (VSD), patent ductus arteriosus (PDA), or pulmonary stenosis was known as far as could be determined.
These findings have previously been briefly reported.19

Methods

From the hemodynamic and angiocardiographic data on 173 infants and children studied in the Cardiovascular Laboratories of the Hospital for Sick Children in Toronto, the cases were selected in which both right and left ventricular (RV and LV) pressures had been recorded through the same cardiac catheter in fairly rapid succession and always within 5 minutes. The right and left ventricular angiograms in these cases were studied and the associated defects clearly present were noted, while those cases in which there was some doubt were eliminated. All cases in which there was evidence of obstruction to LV outflow by catheterization, angiocardiography, or postmortem examination, either clear or possible, were also eliminated.

Eighty cases satisfied these criteria; confirmatory evidence was available from postmortem examination of the hearts in 29 (36%) and some information from surgery in 46 (58%). The pulmonary artery had been entered at cardiac catheterization in a small proportion (12 cases). The age of the 80 children who satisfied the criteria ranged from 1 day to 11 years. Eleven children had more than one study and their data are included.

While none of the young infants had any sedation, some of the older infants and children who were not severely cyanosed were sedated with small doses of an ataractic mixture.20 The cardiac catheters were usually no. 5 or no. 6 Lehman catheters (USCI); in some of the small infants a no. 4 plastic catheter (Elecath) was used. The pressures were measured using Statham P23Db transducers and an Electronics for Medicine DR8 recorder.

Results

Of the 80 children with complete transposition of the great vessels included in the study, 37 also had VSD (group I), 22 had an intact ventricular septum and a PDA (group II), and 21 had an intact ventricular septum without PDA (group III). The relationship between right and left ventricular pressures in these three groups is illustrated in figures 1, 2, and 3, and classified in table 1.

Group I—Thirty-seven Cases with VSD

In 27 of the 37 (73%) cases of VSD, left ventricular pressures were greater than 85% of the systemic or the right ventricular pressure. In seven (19%) pressure was between 70% and 85%, and in three (8%) it was less than 50% of the systemic pressure. The children varied in age from 1 day to 8 years; two of those with low LV pressures were less than 1 week old, but others at this age had systemic LV pressures.

Group II—Twenty-two Cases with Intact Ventricular Septum and PDA

In six of these cases (27%) left ventricular pressures were greater than 85% of the systemic pressure, in four (18%) they were between 70% and 85%, in seven (32%) between 50%
The relationship between left and right ventricular pressures in 21 infants and children with complete transposition of the great vessels and intact ventricular septum without patent ductus arteriosus (group III).

and 70%, and in five (23%) less than 50% of the systemic pressure. None of these children was more than 5 months of age.

**Group III—Twenty-one Cases with Intact Septum and No PDA**

Only one patient in this group, 5 weeks of age, had systemic pressures in the left ventricle, four (19%) had LV pressures slightly in excess of 50% of the systemic, and the remaining 16 (76%) had LV pressures between 20% and 40% of the systemic. The ages in this group varied from 1 week to 11 years, the majority being over 4 months of age.

**Serial Studies**

Cardiac catheterization was repeated at intervals which varied from 6 months to 11 years on a total of 11 patients, seven from group I and four from group II.

Figure 4 illustrates the change in the ratio of LV to RV pressure in the seven patients with VSD (group I). Three remained unchanged at systemic levels, two had initial ratios of 80%, and one of these dropped to 45% at 6 months of age, and when the patient died of intercurrent infection at the age of 2 years, the VSD was found to have closed spontaneously. The other, starting at 80%, had reached 100% at 2½ months of age, and at the age of 5½ years the LV pressure exceeded RV pressure with a ratio of 130%. Two patients started at a low level of 44%. One of these at repeat study was found to have the same ratio

<table>
<thead>
<tr>
<th>LV/RV × 100</th>
<th>Group I*</th>
<th>Group II†</th>
<th>Group III‡</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No.</td>
<td>%</td>
<td>No.</td>
</tr>
<tr>
<td>85% or more</td>
<td>27</td>
<td>92</td>
<td>6</td>
</tr>
<tr>
<td>70% to 84%</td>
<td>7</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>50% to 69%</td>
<td>0</td>
<td></td>
<td>7</td>
</tr>
<tr>
<td>Less than 50%</td>
<td>3</td>
<td>8</td>
<td>5</td>
</tr>
<tr>
<td>Total</td>
<td>37</td>
<td>100</td>
<td>22</td>
</tr>
</tbody>
</table>

*Group I: transposition of the great vessels with ventricular septal defect.
†Group II: transposition of the great vessels with closed septum and patent ductus arteriosus.
‡Group III: transposition of the great vessels with closed septum.
but in addition the VSD had closed spontaneously.21 In the other case the ratio increased to 88%.

In figure 5, the serial observations on four patients with a closed ventricular septum and no ductus (group III) are illustrated. All had initial low pressure ratios, between 20% and 45%. Two did not change and one dropped from 40% to 28%. The one child whose ratio had increased from 22% to 65% had a Blalock-Taussig anastomosis between these vessels.

Discussion

In this study the relationship between right and left ventricular pressures in 80 children with complete transposition has been explored and the effects of associated defects determined. It was thought that left ventricular and pulmonary artery systolic pressures were equal since those children who had any obstruction to left ventricular outflow had been eliminated at angiocardiology or postmortem examination.

It is realized that small VSDs and some PDAs are difficult to diagnose in transposition of the great vessels and may be missed; however we were extremely careful with regard to selection, mostly based on both right and left ventricular angiograms as well as by postmortem examination in 36%, and are confidant that the diagnoses are as correct as possible.

This study demonstrates that when a ventricular septal defect was present, 96% of the patients had left ventricular pressures greater than 70% of the systemic. Only three of 37 patients had left ventricular pressures less than 50% of the pressure in the right ventricle. In contradistinction, of 21 with intact ventricular septum and no ductus, only one had a pressure in the pulmonary circulation at systemic level. Seventy-six per cent of this group had left ventricular pressures between 20% and 40% of the right ventricular pressure. In the intermediate group (group II), 22 patients with an intact ventricular septum but in whom there was a patent ductus arteriosus, there were intermediate levels of pressure in the pulmonary circulation.

Thus the presence of a ventricular septal defect or patent ductus arteriosus very largely determines the presence of pulmonary hypertension in transposition of the great vessels. However it is likely in an unseleted series of 37 cases of isolated ventricular septal defect that the defect would vary in size from small to large and the level of pulmonary artery pressure would vary also.22 The finding of a high incidence of pulmonary hypertension in those transposition cases with a ventricular septal defect must either mean that the VSDs are nearly all large or that an additional mechanism is operating.

The serial studies are too few to support any firm conclusions, but they appear to suggest that there tends to be little change in the pulmonary artery pressure with age, although in those with high pressure, changes in pulmonary vascular resistance are likely.

The findings are similar to those reported in the literature. The largest series of 50 patients was reported by Noonan and associates.14 Of their 23 patients with VSD in whom both right and left ventricular pressures were recorded, 19 (83%) had systemic pressure in the left ventricle. However as has recently been pointed out by Rahimtoola and associates,18 the diagnosis in some of these cases may be in doubt. Of Noonan and as-

Figure 5

The relationship between left and right ventricular pressures in four children with complete transposition of the great vessels and intact ventricular septum who had serial cardiac catheterizations.
sociates' 12 patients with closed ventricular septum, five (42%) had near systemic pressures in the left ventricle. The condition of the ductus is not defined however, and our data suggest that it is likely that the ductus arteriosus was patent or that there was some obstruction to the outflow tract of the left ventricle in these children.

In the present series we have not attempted to calculate the pulmonary blood flow because firstly, the pulmonary artery was not entered in a large enough proportion of cases and secondly, as has recently been emphasized by Burchell, the pulmonary blood flow is difficult to estimate in transposition of the great vessels when the left ventricular and pulmonary artery saturations are extremely high and there is an unknown and almost indeterminable amount of admixture from the desaturated systemic circulation via the bronchial arteries. Rahimtoola and associates and Carr and Wells have recently demonstrated methods of securing pulmonary artery pressures and saturations. However, the saturations here again cannot take into account the admixture from the bronchial circulation and, therefore, determinations of pulmonary vascular resistance are likely to be inaccurate. We have taken the degree of pulmonary hypertension rather than an estimate of high pulmonary vascular resistance as a pointer to pulmonary vascular disease.

With the introduction of the atrial septal balloonning technique by Rashkind and Miller, a possible line of management of the severely hypoxic newborn infant with transposition of the great vessels presents itself. At diagnostic cardiac catheterization, the presence of a ventricular septal defect and a patent ductus arteriosus can be ascertained. A balloon septostomy should be carried out if there is a pressure gradient between the left and right atria or if there is inadequate communication between the two circulations producing hypoxemia and acidosis. This will produce a major improvement in a vast majority of cases. Those patients who have a ventricular septal defect—40% of our total series of 240 children—should be followed closely with a view to closing this defect as early as possible and possibly improving the size of the atrial opening if this is not adequate. In this way the physiology in all these cases would be converted to resemble that of patients who have a closed ventricular septum to start with and in whom the risk of developing progressive pulmonary vascular disease appears minimal.

Acknowledgment

We wish to acknowledge with gratitude the encouragement and advice of Drs. John D. Keith and William T. Mustard and the secretarial assistance of Miss Elaine Smith.

References


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doi: 10.1161/01.CIR.37.2.232

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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