Transposition of the great arteries and narrowing of the aortic arch

Emphasis on right ventricular characteristics

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SUMMARY Of 279 patients with transposition of the great arteries, 14(5%) had narrowing of the aorta, including local coarctation (6), isthmus hypoplasia (6), isthmus atresia (1), and kinking of the aorta (1). There were six deaths in 10 surgically treated patients; in addition four patients died before operation. Two of the four survivors had a subpulmonary malalignment ventricular septal defect with angiocardiographic narrowing of the right ventricular outflow tract; two had evidence of redundant muscle tissue obstructing the right ventricular outflow tract. Necropsy showed the presence of anatomical right ventricular outflow tract or inflow tract obstruction or both in all 10 cases. Outflow tract obstruction was represented by anterior displacement of the infundibular septum and ventriculoinfundibular fold (in hearts with a subpulmonary malalignment ventricular septal defect) or by redundant muscle tissue; inflow tract obstruction was represented by hypoplasia of the tricuspid valve.

It is concluded that the combination of transposition of the great arteries and narrowing of the aorta is always accompanied by right ventricular outflow or inflow tract obstruction or both; the right ventricular abnormalities are probably responsible for the presence of the aortic arch anomalies by reducing aortic flow during morphogenesis. Successful surgical treatment of this complex anomaly is feasible in selected cases.

About one half of all cases of transposition are "simple," the second half being complicated by other anomalies.1 The two commonest associated abnormalities in transposition are ventricular septal defect and left ventricular outflow tract obstruction, occurring separately or together. The incidence of a complicating obstruction of the aortic arch has been estimated to range between 4-6% and 10%.23 More recently, complete transposition of the great arteries with ventricular septal defect and coarctation or atresia of the aortic arch has been described as a separate anatomicoclinical entity which includes inlet or outlet right ventricular obstruction.4

The present study was carried out to define right ventricular characteristics in patients with transposition of the great arteries and obstruction of the aortic arch.

Patients and methods

Two hundred and seventy nine patients with complete transposition of the great arteries were referred to the Leiden University Hospital between 1960 and 1982. Fourteen (5%) of these patients proved to have a complicating aortic arch obstruction. Four patients died before operation and six died after a Senning procedure, a Hanlon operation, banding of the pulmonary artery, coarctation resection, or a combination of these procedures. With one exception, the patients died before 1975; the four survivors were operated on more recently.

The angiograms and heart specimens were studied with special attention to the morphology of the right
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Table 1  Clinical data of patients who underwent successful surgery

<table>
<thead>
<tr>
<th>Case No</th>
<th>Aortic arch anomaly</th>
<th>Right ventricle</th>
<th>Outflow tract</th>
<th>Inflow tract</th>
<th>Ventricular septal defect</th>
<th>Surgical interventions</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>COA</td>
<td>Anterior displacement of IS and prominent VIF</td>
<td>—</td>
<td>Subpulmonary malalignment</td>
<td>Blalock-Hanlon and banding (aged 1 mnth), repair of coarctation (aged 12 yrs), debanding; VSD closed; arterial switch (aged 12½ yrs), closure of residual VSD (aged 12½ yrs)</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>TH</td>
<td>Anterior displacement of IS and prominent VIF</td>
<td>—</td>
<td>Subpulmonary malalignment</td>
<td>VSD closed; arterial switch (aged 3 mnths)</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>TH</td>
<td>Prominent VIF</td>
<td>—</td>
<td>—</td>
<td>Senning operation (aged 6 mnths)</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Kinking</td>
<td>Prominent IS and VIF</td>
<td>—</td>
<td>—</td>
<td>Mustard operation (aged 1 yr)</td>
<td></td>
</tr>
</tbody>
</table>

COA, coarctation of the aorta; IS, infundibular septum; TH, tubular isthmus hypoplasia; VIF, ventriculoinfundibular fold; VSD, ventricular septal defect.

ventricle; angiograms of patients with transposition but without aortic arch anomalies and 15 normal heart specimens from subjects of different ages served as controls. The ventricular septal defects were classified according to Soto et al.; in addition we used the term "subpulmonary malalignment" for defects with rightwards shifting of the infundibular septum and overriding of the pulmonary artery. The right ventricular muscular structures were described according to the terminology proposed by Anderson et al.

Tubular hypoplasia of the aortic isthmus was defined as a combination of abnormally great length and small diameter of that segment according to the criteria given by Moulaert et al.

Using Hegar dilators, the tricuspid valve annulus was measured in relation to the mitral valve annulus. The tricuspid valve was considered to be hypoplastic if its diameter was <0.8 of that of the mitral valve.

Results

SURVIVORS
Four patients were alive and well six months to five years after operation (Table 1). In two of these patients (cases 1 and 2) a subpulmonary malalignment ventricular septum defect was closed, and the other two had an intact ventricular septum. The subpulmonary malalignment defects were angiographically characterised by overriding of the pulmonary artery; in addition, right ventriculography showed muscular subaortic stenosis caused by an anteriorly displaced infundibular septum and prominence of the ventriculoinfundibular fold (Figs. 1 and 2).

The angiocardiogram of the patient with kinking of the aorta and an intact ventricular septum (case 4)
showed prominence of the infundibular septum in the lateral view (Fig. 3a) and of the ventriculo-infundibular fold in the left oblique view (Fig. 3b). Another example of prominence of the ventriculo-infundibular fold is shown in Fig. 4. Regarding the aortic arch of the survivors, one local coarctation has been resected, one hypoplastic isthmus has grown to normal dimensions, and two of the patients await an eventual operation.

**NON-SURVIVORS**

Ten patients died before or after operation (Table 2). Six of the necropsy specimens showed a subpulmonary ventricular septal defect (cases 5–10). This type of defect is, as has been described before, characterised by anterior displacement of the infundibular septum and prominence of the ventriculo-infundibular fold; this results in a reduction in distance between these structures and the anterior right ventricular wall and thus in narrowing of the outflow tract (Fig. 5). This close relation between (type of) ventricular septal defect and outflow tract narrowing was absent in the other four specimens, of which one showed a perimembranous trabecular defect, one a very small muscular inlet defect, and two an intact septum. In these four cases either redundant muscle tissue obstructed the outflow tract (three hearts) or the inflow tract was obstructed by a hypoplastic tricuspid valve or both. Two cases (cases 13 and 10) of hypoplastic tricuspid valves and redundant outflow tract muscle tissue (a prominent ventriculo-infundibular fold and a trabecula in front of the aortic ostium) are shown in Figs. 6 and 7.

Valid catheterisation data were available for eight patients. In only one of these patients a pressure gradient (of 30 mmHg) was present across the right ventricular outflow tract.

**Discussion**

The findings in this study show that in all cases of transposition of the great arteries and narrowing of the aorta, anatomical right ventricular outflow or inflow tract obstruction or both are present. Milanesi et al came to the same conclusion, and we agree with these authors that unequal partitioning of the blood flow as a consequence of right ventricular obstruction may be the fetal pathogenetic mechanism for the
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underdevelopment of the aortic arch, as postulated for left ventricular outflow tract obstruction in normally related great arteries. Eight out of 14 patients proved to have a subpulmonary malalignment ventricular septal defect with anterior and rightward displacement of the infundibular septum. A rightward shift of the infundibular septum is also an essential feature of double outlet right ventricle with subpulmonary ventricular septal defect; the difference between transposition and double outlet right ventricle is evident by the degree of rightward shifting of the infundibular septum and thus by the degree of pulmonary artery overriding. Since the frequent concurrence of double outlet right ventricle and aortic arch anomalies has been established before (Kurosawa H, van Mierop LHS. World Congress of Paediatric Cardiology, London, 1980, abstract No 339), we selected for the present study cases of transposition according to the 50% rule—that is, in our cases the pulmonary artery was mainly committed to the left ventricle.

Table 2  Anatomical features of cases examined at necropsy

<table>
<thead>
<tr>
<th>Case No</th>
<th>Age</th>
<th>Aortic arch anomaly</th>
<th>Right ventricle</th>
<th>VSD</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>Outflow tract</td>
<td>Inflow tract</td>
</tr>
<tr>
<td>5</td>
<td>5 mth</td>
<td>TH</td>
<td>Anterior displacement of IS and VIF</td>
<td>—</td>
</tr>
<tr>
<td>6</td>
<td>4 mth</td>
<td>TH</td>
<td>Anterior displacement of IS and VIF</td>
<td>—</td>
</tr>
<tr>
<td>7</td>
<td>2 wk</td>
<td>TH</td>
<td>Anterior displacement of IS and VIF</td>
<td>—</td>
</tr>
<tr>
<td>8</td>
<td>1½ yr</td>
<td>COA</td>
<td>Anterior displacement of IS and VIF</td>
<td>—</td>
</tr>
<tr>
<td>9</td>
<td>1 mth</td>
<td>COA</td>
<td>Anterior displacement of IS and VIF</td>
<td>—</td>
</tr>
<tr>
<td>10</td>
<td>1 mth</td>
<td>COA</td>
<td>Prominent VIF</td>
<td>Hypoplastic TV Subpulmonary malalignment</td>
</tr>
<tr>
<td>11</td>
<td>1 mth</td>
<td>COA</td>
<td>Prominent VIF</td>
<td>Hypoplastic TV Subpulmonary malalignment</td>
</tr>
<tr>
<td>12</td>
<td>8 mth</td>
<td>COA</td>
<td>Prominent VIF</td>
<td>Hypoplastic TV Subpulmonary malalignment</td>
</tr>
<tr>
<td>13</td>
<td>2 mth</td>
<td>TH</td>
<td>Trabecula ventral to aortic ostium</td>
<td>—</td>
</tr>
<tr>
<td>14</td>
<td>3 mth</td>
<td>Isthmus atresia</td>
<td>Prominent IS and VIF</td>
<td>Hypoplastic TV Perimembranous trabecula</td>
</tr>
</tbody>
</table>

COA, coarctation of the aorta; TH, tabular isthmus hypoplasia; IS, infundibular septum; VIF, ventriculoinfundibular fold; VSD, ventricular septal defect; TV, tricuspid valve.

Fig. 4  Right ventriculogram in 8 month old child (case 12) with transposition of the great arteries and local coarctation, which is hidden behind the ascending aorta in this frontal view. Note the subaortic narrowing by undue prominence of the ventriculoinfundibular fold (arrow).

Fig. 5  Morphological appearance of the right ventricle in a patient (case 5) with subpulmonary ventricular septal defect (arrow). Anterior displacement of the infundibular septum (IS) and prominence of the ventriculoinfundibular fold (VIF) narrow the outflow tract (curved arrow). AW, anterior wall of the right ventricle.
In a previous anatomical study we found aortic arch anomalies in 35% of hearts with transposition of the great arteries and right ventricular outflow tract obstruction. It should be noted that the diagnosis of right ventricular outflow tract obstruction is an estimation, and in that as well as in the present study it means an obstruction relative to normal. In this respect it is important to realise that in only a few patients with an obvious, angiographically demonstrated, narrow subaortic area may a pressure gradient be recorded across it. Nevertheless, a poorly expanded or relatively narrow subaortic conus should be viewed as potentially obstructive according to Freedom et al. In our view more studies of the relation between anatomical right ventricular obstructions and catheterisation data are needed to establish the functional implications and natural history of these anomalies.

Milanesi et al suggest that the combination of transposition of the great arteries and coarctation of the aorta is not so rare and that the clinical manifestations and the pathological findings of this association are sufficiently distinctive as to be considered a separate nosographic entity among transposition of the great arteries. According to these authors, the entity should include the following morphological features: (a) a ventricular septal defect, mostly subpulmonary; (b) underdevelopment of the right ventricular trabecular component; and (c) right ventricular outflow or inflow obstruction.

Our study confirms the presence of angiographic or anatomical outflow or inflow obstruction or both in all cases. Nevertheless, underdevelopment of the right ventricle was diagnosed only once as a separate feature; in all other cases the trabecular part was not hypoplastic, although in some cases outflow tract obstructing muscle bundles protruded into the trabecular part as well, thus reducing the trabecular lumen.

Another difference between our findings and those of Milanesi et al applies to the obligatory presence of a ventricular septal defect in their cases; in our patients four (28%) proved to have an intact ventricular sep-
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Since, in addition, the type of aortic arch anomaly may differ (from atresia via tubular hypoplasia and local coarctation to kinking of the aorta) we conclude that the variety of these findings does not justify the definition of a nosographic entity.

This conclusion implies that a general prognosis cannot be given for the group as a whole and that the surgical results depend on the presence or absence of an intact ventricular septum and the type and degree of aortic arch anomaly. In this respect it is important to realise that, although most cases had distinct narrowing of the aorta, in other cases the narrowing may be mild; this was the case in some patients with tubular hypoplasia of the isthmus, in one of which the isthmus grew spontaneously to normal dimensions.

The hospital mortality has been high in the earlier years of our experience; recent results support the feasibility of successful surgical treatment. In this context, however, the maintained systemic right ventricular blood pressure may trigger hypertrophy, eventually resulting in progression of the outflow obstruction; as we indicated before this progression might be prevented by performing an arterial switch procedure with resulting normalisation of right ventricular pressure.

Indications for surgical treatment of right ventricular outflow and inflow tract obstruction should be based on the results of functional and natural history studies, which have not been performed as yet.

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References

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