Taussig-Bing Malformation, Coarctation of the Aorta, and Reversed Patent Ductus Arteriosus

Operative Correction in an Infant

By ANNE LITTLE WEDEMeyer, M.D., RUSSELL V. LUCAS, JR., M.D., and ALDO R. CASTANEDA, M.D.

SUMMARY

Operative correction of a Taussig-Bing malformation was successfully accomplished in a 23-mo-old boy, weighing 6.8 kg. Associated coarctation of the aorta and reversed patent ductus arteriosus were corrected in a preliminary operation. The patient's management was complicated by severe preoperative thrombocytopenia which responded to multiple phlebotomies. Surgical repair of the Taussig-Bing malformation consisted in patching the ventricular septal defect to create physiologically complete transposition of great vessels. Then a Mustard procedure was done.

Additional Indexing Words:
Phlebotomy  Thrombocytopenia  Platelet counts  Coagulation data

THE Taussig-Bing malformation, one of several forms of origin of both great vessels from the right ventricle, has eluded successful operative correction until recently.

The subject of this report is a 6.8-kg infant with the Taussig-Bing malformation who has survived successful surgical correction. The coexistence in this infant of coarctation of the aorta and reversed patent ductus arteriosus added to the diagnostic and surgical problems. Significant hematologic abnormalities further complicated his care.

The purpose of this communication is to define the anatomic features of his defect, to relate these to the other varieties of origin of both great vessels from the right ventricle, and to discuss methods of operative correction.

Report of Case

This male infant was the product of a full-term pregnancy, uncomplicated by infections or drugs. He weighed 3.4 kg at birth. Cyanosis occurred shortly after birth, and a systolic heart murmur was heard at 2 weeks of age. At age 4 mo congestive heart failure was recognized, and he responded to digitalis.

At age 13 mo (January 1968), when he was referred to the University of Minnesota Medical Center, the mother reported increasing cyanosis, mild tachypnea, and delayed growth and development.

His weight of 6.4 kg and length of 68 cm were less than the third percentile for his age. Flush blood pressures were 90 mm Hg in the right arm and 80 mm Hg in the left arm and leg; all peripheral pulses were strong. Reversed differential cyanosis was noted, with moderate cyanosis of his lips and upper extremities and mild cyanosis of the lower extremities. All fingers and toes were clubbed. A precordial bulge approached a carinate deformity. S1 was loud, and the second sound was very loud and thought to be single. A systolic ejection click and a grade III/VI harsh holosystolic murmur were heard maximally at the left midsternal border. The liver was palpable 3 cm below the right costal margin.
Preoperative electrocardiogram. The QRS axis is 110°, an unusual finding in origin of both great vessels in the right ventricle. Note the evidence of biventricular hypertrophy.

The hemoglobin value was 14.1 g/100 ml, and the hematocrit was 51%. The electrocardiogram revealed a mean QRS frontal plane axis of +110°, mild right atrial enlargement, and biventricular hypertrophy (fig. 1). Thoracic roentgenograms revealed cardiomegaly, massively increased pulmonary vascular markings, a prominent left atrium, and an enlarged pulmonary artery segment (fig. 2).

The clinical impression was either a variant of transposition of the great vessels and reversed patent ductus arteriosus or truncus arteriosus.

Angiocardiography and cardiac catheterization were performed. The right ventriculogram (fig. 3) demonstrated a hypertrophied right ventricle. The dilated pulmonary artery arose from the right ventricular infundibulum in its usual location. The aorta opacified more densely than the pulmonary artery and arose via a subaortic conus from the right ventricle immediately to the right of the pulmonary artery. The semilunar valves were at the same plane and the great vessels were side by side. There was no continuity between semimlar and ativoventricular valves. In addition, mild tubular hypoplasia of the distal aortic arch and isolated severe coarctation of the aorta distal to the left subclavian artery were present. Blood flow through a large patent ductus was predominantly right to left. A pulmonary arteriogram revealed regurgitation of contrast material into both ventricles. The descending aorta opacified via the reversed patent ductus arteriosus.

The hemodynamic features (table 1) further defined the defect. The high (82%) O2 saturation in the main pulmonary artery and low (47%) O2 saturation in the right brachial artery (obtained via cannulation though not simultaneously) were compatible with origin of both great vessels from the right ventricle (type IIA Taussig-Bing malformation).

The equal pressures proximal and distal to the coarctation and the equal (and high) O2 saturation in main pulmonary artery and descend-
Figure 2

Preoperative thoracic roentgenograms. (A) Posteroanterior view. Pulmonary vascular markings are increased. There is generalized cardiomegaly and prominence of pulmonary artery. The left atrial appendage is apparent at the left cardiac border. (B) Left lateral view. The posterior deviation of the esophagus confirms the presence of left atrial enlargement.

Figure 3

Selective right ventriculograms. (A) The right ventricle is dilated and heavily trabeculated. The aorta and pulmonary artery fill simultaneously. The aorta is more densely opacified than the pulmonary artery. The aortic and pulmonary valve planes are at the same level and the aorta does not take its usual leftward swing in this view.

(B) Lateral view at the same time as A. The aorta and pulmonary artery lie in the same plane and the valves are at the same level. The aorta clearly rises anterior to the ventricular septum. In both views the descending aorta is poorly opacified. Coarctation of the aorta and reversing ductus arteriosus were demonstrated by aortography.
Table 1

<table>
<thead>
<tr>
<th>Site</th>
<th>O₂ saturation (%)</th>
<th>Pressure (mm Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>54</td>
<td>—</td>
</tr>
<tr>
<td>Right atrium</td>
<td>55 a = 6 v = 5 m = 5.5</td>
<td>—</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>59 94/0, m = 12</td>
<td></td>
</tr>
<tr>
<td>Main pulmonary artery</td>
<td>82 100/43, m = 68</td>
<td></td>
</tr>
<tr>
<td>Descending aorta</td>
<td>80 95/40, m = 73</td>
<td></td>
</tr>
<tr>
<td>Right brachial artery</td>
<td>47</td>
<td>—</td>
</tr>
</tbody>
</table>

ing aorta defined the reversed patent ductus arteriosus.

As a prelude to corrective intracardiac surgery, the child underwent division and suture of the patent ductus arteriosus and repair of the coarctation of the aorta in February 1968. Following the procedure, cyanosis appeared to be equal in his head and upper and lower extremities. He was discharged but was to continue on digoxin 8 days after surgery.

In May 1968, a relative anemia was treated with iron given orally. A month later hemoglobin was 18.9 g/100 ml, hematocrit was 75%, and platelet count, 6,000/cu mm. Hematologic findings (table 2) were not consistent with typical intravascular coagulation.

Heparin therapy failed to correct the thrombocytopenia. Peripheral platelet destruction was ruled out by normal platelet survival after platelet transfusion.

Continued iron therapy over the next 2 weeks resulted in an increase in hematocrit (81%) and further decrease in platelets (4,000/cu mm).

Table 2

<table>
<thead>
<tr>
<th>Test</th>
<th>Patient's values</th>
<th>Normal values</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prothrombin time (sec)</td>
<td>10.3</td>
<td>11-13</td>
</tr>
<tr>
<td>Kaolin partial thromboplastin time (sec)</td>
<td>56.4</td>
<td>35-45</td>
</tr>
<tr>
<td>Thrombin time (sec)</td>
<td>18.0</td>
<td>14-16</td>
</tr>
<tr>
<td>Venous platelet count</td>
<td>17,000</td>
<td>200,000-400,000</td>
</tr>
<tr>
<td>Fibrinogen (mg/100 ml)</td>
<td>210</td>
<td>200-400</td>
</tr>
<tr>
<td>Bleeding time (min)</td>
<td>5</td>
<td>3-6</td>
</tr>
<tr>
<td>Coagulation factors</td>
<td></td>
<td></td>
</tr>
<tr>
<td>V (%)</td>
<td>100</td>
<td>70-130</td>
</tr>
<tr>
<td>VIII (%)</td>
<td>115</td>
<td>70-130</td>
</tr>
<tr>
<td>IX (%)</td>
<td>50</td>
<td>70-130</td>
</tr>
<tr>
<td>XI (%)</td>
<td>29</td>
<td>70-130</td>
</tr>
</tbody>
</table>

Relationship of platelet count (solid dots) to hematocrit (open dots).

Because of the reciprocal relationship between hematocrit and platelets, the infant was subjected to multiple small phlebotomies. During a 3-week period, approximately half his total blood volume was withdrawn without replacement. He tolerated these procedures well. The platelet count (80,000 on the day of definitive surgery) had increased after phlebotomy as the hematocrit fell (fig. 4).

On October 10, 1968, he underwent total physiologic correction of his cardiac defect with the aid of a pump oxygenator.* The heart was exposed via a median sternotomy. On total cardiopulmonary bypass a transverse right ventriculotomy was performed. A large ventricular septal defect was found above the crista supraventricularis, its upper border being in continuity with pulmonary valvar tissue. The aortic valve was to the right of this defect separated from it by a prominent muscle mass. The left lateral margin of the ventricular septal defect was formed by the parietal band of the crista and the inferior margin by the muscular septum. Interrupted 4-0 Tefdack stitches were placed along the free margin of the ventricular septum, the parietal band, and the right lateral margin. These stitches were then passed through a 1/16-inch thick Teflon felt patch and tied. The remaining free edge of the prosthetic patch was sutured to the upper lip of the transverse ventriculotomy incision. Left ventricular-pulmonary artery continuity or a functional transposition of the great vessels was thereby created (fig. 5). The final physiologic correction of this defect was accomplished by the Mustard transatrial

*Olson Medical Products, Inc., Ashland, Massachusetts, and Bentley Laboratories, Santa Ana, California.

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**Method of surgical correction.** The patent ductus arteriosus and coarctation of the aorta were repaired at a previous operation. (Left) This illustrates the preoperative anatomy and hemodynamic features. (Right) Operative management. First, a Teflon patch (lower patch) was positioned in such a way as to create a tunnel from the ventricular septal defect to the base of the pulmonary artery. This maneuver resulted in the hemodynamic pattern of complete transposition of the great vessels and intact ventricular septum. Second, the atrial septum was excised and a pericardial baffle positioned within the atria in the fashion described by Mustard. As a consequence systemic venous blood passed through the pericardial tunnel into the anatomic left ventricle, then through the ventricular septal defect and Teflon tunnel into the pulmonary artery. Pulmonary venous blood was channeled into the anatomic right ventricle and thence out through the aorta.

**Discussion**

Origin of both great vessels from the right ventricle is a relatively uncommon cardiac malformation. Neufeld and associates emphasized that the location of the ventricular septal defect in this anomaly is largely responsible
for the physiologic features. In type I, the ventricular septal defect is located subjacent to the crista supraventricularis; this location allows blood from the left ventricle to flow preferentially into the aorta. In type IIA Taussig-Bing malformation, on the other hand, the ventricular septal defect lies above the crista and is closely related to the pulmonary valve. Left ventricular blood thus preferentially enters the pulmonary artery and systemic venous blood is directed into the aorta.

There are, however, a number of anatomic variations among patients with origin of both great vessels from the right ventricle. The ventricular septal defect may be above the crista supraventricularis and confluent with the pulmonary valve (our case, Neufeld and associates\(^3\)). The defect may be supracristal but separated from the pulmonary valve by the subpulmonary conal free wall (the original Taussig-Bing heart,\(^6\) the cases of Hightower and associates\(^4\)). The defect may be supracristal, underlying the pulmonary valve, and extending under the aortic valve as well (type IIB of Neufeld and associates\(^3\)). Finally the defect may be subcristal and immediately opposite the subaortic conus.\(^3\)

The location of the ventricular septal defect in origin of both great vessels from the right ventricle is of critical importance in the operative correction. When the defect is located subjacent to the crista supraventricularis, a patch can be sewn to establish continuity between left ventricle and the subaortic conus of the right ventricle, thereby completely correcting the defects.\(^7\)

When the ventricular septal defect is supracristal, this simple internal rerouting is not possible. In our patient the high subpulmonary position of the ventricular septal defect and the large muscle mass separating the defect from the aortic valve prevented the internal routing of left ventricular blood into the aorta. On the other hand, it was a simple matter to establish continuity between the left ventricle and the pulmonary artery with a patch without compromising the outflow to either pulmonary artery or aorta. This physio-

logic pattern of complete transposition of the great vessels with intact ventricular septum was then amenable to correction utilizing the technic pioneered by Mustard.

This same technic was employed by Kirklin's group.\(^4\) Their patients differed from our patient in that the aorta was anterior, and the ventricular septal defect was located lower, and was separated from the pulmonary valve by infundibular muscle. They suggested that a tunnel from ventricular septal defect to aorta could have been made and an aortic homograft utilized to connect the right ventricle to the pulmonary artery. This procedure could not have been employed in our patient.

In those patients in whom the supracristal defect underlies both pulmonary artery and aorta, a review of the anatomy suggests that the subaortic portion of the ventricular septal defect could be enlarged and then an intraventricular tunnel constructed without obstructing aortic pulmonary outflow.

Thus the surgeon, preparing to correct origin of both great vessels from the right ventricle with a supracristal defect, must have an accurate preoperative anatomic and physiologic assessment and be prepared at operation to utilize a variety of operative technics, depending on the precise anatomic features he encounters.

The association of coarctation of the aorta and patent ductus arteriosus is common in the Taussig-Bing malformation.\(^5\) When these are present, technical considerations of cardiopulmonary bypass necessitate preliminary surgical elimination of the coarctation of the aorta and patent ductus arteriosus.

Utilization of multiple phlebotomies to treat thrombocytopenia was based on observations reported by Hartmann.\(^8\) The dramatic response of the hematologic abnormalities in our patient confirms the usefulness of multiple phlebotomies as preoperative preparation in this clinical setting.

The majority of patients with the Taussig-Bing malformation die in infancy. Further, the presence of high pressure and high flow within the pulmonary circuit may result in
early pulmonary vascular disease. Thus, corrective operation in infancy would be highly desirable. Our patient weighed only 6.8 kg. Since we are accumulating encouraging experience with open heart surgery in infants weighing 3 to 4 kg, we propose that the infant with the Taussig-Bing malformation have corrective operation performed as soon as the diagnosis can be established.

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