Distribution and Anomalies of Coronary Arteries in Tetralogy of Fallot

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SUMMARY The surgical management of tetralogy of Fallot (TF) may be complicated by anomalies in the course and distribution of coronary arteries. Selective coronary angiography was performed in 119 cases of TF in order to prevent injury of aberrant vessels. In 11 patients, anomalies in the origin of coronary branches were revealed (five patients with a single coronary ostium, five with the anterior descending coronary artery arising from the right coronary artery or right sinuses of Valsalva, and one with the circumflex artery arising from the right coronary artery). Other findings include anastomoses between coronary and bronchial arteries, small fistulas between coronary arteries and the pulmonary artery or right atrium, and hypoplasia of the coronary tree.

The onset of left anterior hemiblock after total correction of TF could possibly be explained, in cases with a dominant left pattern, by injury to the coronary system after closure of the interventricular septal defect, rather than by injury to the conduction system.

THE RESULT of the total correction of tetralogy of Fallot (TF) depends not only on the seriousness of the functional changes, but also on the possible presence of certain unfavorable anatomic features.

Anomalies in the course and distribution of the coronary arteries may be present in TF. In a number of patients, the left anterior descending coronary artery (LAD) arises from the right coronary artery (RCA) and crosses over the right ventricular outflow tract, thus complicating the surgical correction. Such an anomalous vessel is not always detectable at the time of ventriculotomy, particularly when it is obscured by the overlying myocardium, by epicardial fat or by pericardial-epicardial adhesions in patients who have previously undergone palliative surgery.

The anomalous origin of the LAD from the RCA is not the only variant found in these patients.

In order to prevent possible consequences that may follow the division of an aberrant vessel, the course and distribution of the coronary arteries must be known before operation. For this reason, we usually carry out selective coronary angiography in patients with TF before corrective surgery. This report describes our observations in 119 patients with TF.

Materials and Methods

Studies were performed on 119 cases with TF, 79 males and 40 females, ages 5 months to 26 years. Cardiac catheterization and selective coronary angiography were carried out. Initially, right ventricular angiocardiography was performed in two projections (sitting-up and hepatoclavicular), with a #6F or 7F NIH catheter introduced into the saphenous or axillary vein. Subsequently, selective coronary angiography was carried out via the right axillary artery, using a Sones 6.5F catheter in patients weighing under 13 kg and a Sones 7.5F catheter in those weighing over 13 kg. For the left coronary artery (LCA), anteroposterior and right anterior oblique projections were always used, and for the RCA, usually the anteroposterior projection was used. In patients in whom an aberrant artery was detected, we also used the left anterior oblique or lateral projections. Injections were carried out by manual pressure,
TABLE 1. Distribution of Coronary Circulation in Tetralogy of Fallot

<table>
<thead>
<tr>
<th></th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dominant right type of coronary distribution</td>
<td>64</td>
<td>53.8</td>
</tr>
<tr>
<td>Dominant left type of coronary distribution</td>
<td>39</td>
<td>32.8</td>
</tr>
<tr>
<td>Balanced coronary distribution</td>
<td>16</td>
<td>13.4</td>
</tr>
<tr>
<td>Total</td>
<td>119</td>
<td></td>
</tr>
</tbody>
</table>

TABLE 2. Coronary Artery Anomalies in Tetralogy of Fallot

<table>
<thead>
<tr>
<th></th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Single coronary ostium</td>
<td>5</td>
<td>4.2</td>
</tr>
<tr>
<td>LAD from RCA or right sinus of Valsalva</td>
<td>5</td>
<td>4.2</td>
</tr>
<tr>
<td>LCX from RCA</td>
<td>1</td>
<td>0.8</td>
</tr>
<tr>
<td>&quot;Large&quot; conus artery</td>
<td>23</td>
<td>19.3</td>
</tr>
<tr>
<td>Fistulas between coronary arteries and PA or right atrium</td>
<td>14</td>
<td>11.7</td>
</tr>
<tr>
<td>Anastomoses between coronary and bronchial arteries</td>
<td>31</td>
<td>26.0</td>
</tr>
<tr>
<td>Hypoplastic coronary arteries</td>
<td>1</td>
<td>0.8</td>
</tr>
</tbody>
</table>

Abbreviations: LAD = left anterior descending artery; RCA = right coronary artery; RSV = right sinus of Valsalva; LCX = left circumflex artery; PA = pulmonary artery.

using 0.25–0.33 ml/kg of contrast material (Urographin 76%).

Results

The results are summarized in tables 1 and 2.

The "large" conus artery was considered as an anomaly because of the importance this vessel has when it is greatly enlarged and courses over the right ventricular infundibulum.

Distribution of Coronary Circulation

In 64 patients (53.8%) a dominant right pattern was observed. The RCA gave rise to the artery for the atroventricular (AV) node and to the posterior descending artery (PDA). The left circumflex artery (LCX) was hypoplastic, and in some cases did not pass into the AV groove, but ended on the posterolateral surface of the left ventricle. In one patient, the LCX was absent. This type of dominant right distribution was more frequent in cases of TF with marked hypoplasia of the pulmonary artery trunk.

A dominant left coronary distribution was present in 39 patients (32.8%). A large obtuse marginal branch originated from the LCX, which also gave rise to the PDA, to the branches for the posterior wall of the left ventricle and sometimes to the artery for the AV node. The RCA ended at the crux after giving off a large conus artery or an anterior ventricular branch.

Sixteen patients (13.4%) had a balanced coronary circulation. A small PDA and the artery for the AV node originated from the RCA; the LCX supplied large branches for the posterior wall of the left ventricle and an artery for the obtuse margin of the heart.

Single Coronary Ostium

We considered a single coronary ostium as being right or left depending on its location on the right or left coronary sinus, respectively. A single coronary ostium can be further subdivided, depending on whether the anomalous coronary artery passes in front of or behind the pulmonary artery. We observed five cases of single coronary ostium, two from the left sinus and three from the right.

Single Left Coronary Ostium

In two patients, the LCA had a normal origin in the left sinus of Valsalva. The RCA arose from the proximal portion of the LCA. The RCA passed behind the aorta, reached the AV groove and coursed along it to the crux. The PDA, the two small branches for the posterior wall of the left ventricle and the artery for the AV node originated at the crux. The LAD and the LCX had normal courses. In figures 1A and B, the LAD does not supply the first septal branch and the diagonal branches are poorly developed.

Single Right Coronary Ostium

This anomaly was seen in three patients. The first patient, 2½ years old, had attacks of cyanosis and marked hypoplasia of the pulmonary artery trunk. The second patient, 12 months old, had anoxic spells. We examined the third patient, 3 years old, after a total correction with insertion of a valved conduit had been performed owing to the presence of the LAD coursing over the right ventricular outflow tract. We performed selective coronary angiography to ensure that the anomalous vessel was intact (figs. 2A and B). In these patients, the right sinus of Valsalva, which presented a marked anterior displacement to the left owing to the considerable clockwise rotation of the aorta, gave rise to a common branch that divided into three arteries. One artery followed the normal course of the RCA, while the second one turned to the left and, passing across the infundibulum of the right ventricle, ran into the anterior interventricular groove. The third branch, passing between the aorta and the pulmonary artery, reached the posterior wall of the left ventricle. The lateral projection (fig. 3) shows that the LAD is posteriorly displaced because of the considerable clockwise rotation of the interventricular septum. In two patients, the LAD seemed to have no first septal branch and few diagonal branches.

Left Anterior Descending Coronary Artery Arising from the Right Coronary Artery or the Right Sinus of Valsalva

In accordance with Meng, we subdivided this anomaly into two types, depending on the relationship of the LAD to the base of the pulmonary artery.
Type 1: In three patients, the LAD arose from the proximal portion of the RCA and reached the anterior interventricular groove passing just below the base of the pulmonary artery trunk. In these patients, the LAD did not give off the first septal branch, while the diagonal arteries were very small. One patient had an anomalous origin of the LAD from the right sinus of Valsalva (fig. 4).

Type 2: In one patient, the LAD arose from the proximal portion of the hypoplastic RCA and entered the anterior interventricular groove passing across the outflow tract of the right ventricle, about 2-3 cm below the base of the pulmonary artery trunk.

In all five cases, the LCA originated normally from the left sinus of Valsalva, giving off only the LCX, which ran along the AV groove. In two patients, a small, short accessory LAD arose from the LCA and ended before it could enter the anterior interventricular groove.

Left Circumflex Artery from Right Coronary Artery

In one patient, 4 years of age, a common right coronary ostium gave rise both to the RCA, with a normal course along the AV groove, and to a large LCX, which bent to the right and passed behind the aorta and the pulmonary artery, ending in a long branch for the posterolateral wall of the left ventricle. The LAD originated normally from the left sinus of Valsalva and ran along the anterior interventricular groove.
supplying septal and diagonal branches (figs. 5A and B). In accordance with other authors, we have observed this anomaly even in normal subjects.

Large Conus Artery

In 23 patients, this vessel was remarkably developed, but its size was not related to the type of coronary distribution. A large conus artery was, in fact, present in both right and left dominant patterns. It usually originated from the proximal portion of the RCA, but in two cases, it originated from a separate ostium in the right sinus of Valsalva (fig. 6). In 10 patients, this vessel had a diameter equal to or greater than that of the RCA and, crossing over the outflow tract of the right ventricle, reached the heart apex, so that in anteroposterior projections it could be mistaken for the LAD (figs. 7A and B).

Fistulas from Coronary Arteries to the Pulmonary Artery or Right Atrium

Small fistulas between the coronary arteries and the pulmonary artery were seen in 13 patients. In eight patients, the fistulas originated from the left main coronary artery or the initial portion of the LCA or LAD. In three cases they arose from the RCA and in two from both the RCA and LCA. Figure 8 shows a case of TF with a left dominant pattern whose left main coronary artery gives rise to a branch that communicates with the pulmonary artery trunk. A communication between the coronary arterial bed (anterior branch arising from the hypoplastic RCA) and the right atrium was present in one patient (fig. 9). In all cases the fistulas were small.

Anastomoses Between the Bronchial and Coronary Arteries

We observed anastomotic channels between the RCA or LCA and the bronchial arteries in 31 cases of severe TF with hypoplasia of the pulmonary artery branches. Figure 10 shows a case of TF in which the LCA gives rise to branches running superiorly and posteriorly to anastomose with the bronchial arteries. This patient had agenesis of the left branch of the pulmonary artery. Figure 11 shows the RCA in the emptying phase. Anastomoses between the RCA and the bronchial arteries are also evident.

Hypoplasia of the Coronary Artery Tree

One patient had hypoplasia of all the coronary branches (fig. 12), and died of a low cardiac output syndrome during surgical correction.

Discussion

The low statistical incidence of coronary anomalies in TF reported by surgical teams can be explained by the fact that many of these anomalies are difficult to detect intraoperatively. Postmortem examinations indicate a higher incidence, probably due to a greater possibility of carefully examining the coronary arteries.

Angiographic studies of the coronary arteries in TF, apart from one of the two cases reported by White, have been performed by aortography only. Fellows, in his study of 84 patients with TF, visualized the coronary arteries by injecting contrast material into the aorta (79 cases) or into the right ventricle (five cases). He found a 5% incidence of anomalies in coronary distribution (4% LAD from the RCA, 1% single coronary).
Figure 4. Cineangiograms of anomalous origin of the left anterior descending artery (LAD) from the right sinus of Valsalva (RSV). A and B) Left selective coronary angiography in the anteroposterior and lateral projections. The left circumflex artery runs posteriorly in the lateral projection. C and D) Right selective coronary angiography. The right coronary artery, conus artery and anomalous LAD from the RSV are evident in anteroposterior (C) and lateral (D) projections.

Figure 5. Cineangiograms of the left circumflex artery (LCX) from the right coronary artery. A) Left selective coronary angiography in the right anterior oblique projection. The LCX is absent. B) Right selective coronary angiography in the lateral projection. The LCX is evident and runs behind the aorta.
FIGURE 6. Cineangiogram of a "large" conus artery arising from the anteriorly displaced right sinus of Valsalva.

The use of selective coronary angiography allowed us to visualize anomalies that cannot be shown by surgical examination and aortography, such as coronary-bronchial anastomoses or small fistulas between the coronary arteries and the pulmonary artery.

In TF, for the reason mentioned previously, the anomalies in distribution and origin of the coronary arteries involving the anterior wall of the right ventricle become very significant. These anomalies, if not detected by angiography, may lead to errors at operation.

Thus, an aberrant vessel that courses over the outflow tract of the right ventricle is important.

We observed a large conus artery in 23 patients (19.3%), in 10 of whom the vessel was as large as or larger than the RCA. This anomaly should therefore be sought for by the surgeon.

The three patients with a single right coronary ostium and the five with the LAD arising from the RCA confirmed that the LAD runs over the outflow tract of the right ventricle in 6.7% of our cases.

Eleven patients (9.2%) had anomalies in the origin of the coronary arteries. Of these, five had a single coronary ostium, five had the LAD originating from the RCA or the right sinus of Valsalva and one patient had an anomalous origin of the LCX from the RCA.

The absence of the first septal branch and the scarcity of diagonal arteries in almost all the patients with the LAD arising from the RCA or with a single coronary ostium are quite interesting. In all the other patients, we found that the first septal branch was more or less long. A clear visualization of the other septal branches was not always possible, but the blood supply of the interventricular septum was similar to that of normal subjects.

As already described by James,17 the interventricular septum and the conduction system can be divided into two portions. The distal tract of the AV node, the bundle of His, and the proximal portion of the two branches of the bundle of His lie in the superior and posterior portion of the interventricular septum. These structures are supplied by the artery of the AV node, by the septal branches arising from the PDA and by the first septal branches of the LAD. The lower and apical portion of the interventricular sep-
tum includes the two branches of the bundle of His and the most distal portion of the bundle branches. These structures are mostly supplied by septal branches of the LAD and, to a lesser extent, by septal branches of the PDA.

In patients with dominant left pattern, the LCA supplies the whole interventricular septum. In the presence of a subcristal interventricular septal defect (as often occurs in TF), the septal branches arising from the LAD can be displaced on the upper and anterior margin of the defect, while those arising from the PDA and coursing in a posteroanterior direction can be displaced on the lower and posterior margin of the defect.

Considering this type of blood supply of the interventricular septum, an injury to the arterial system supplying the septum could probably explain some of the left anterior hemiblocks that occur after total correction of TF, without necessarily implying an injury to the conduction pathways.

Finally, the incidence of the dominant left pattern (32.8%) is higher than in normal subjects (10–12%).

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**Figure 8.** Left selective coronary angiography in the anteroposterior projection, showing a fistula between the coronary and pulmonary arteries.

**Figure 9.** A fistula between an anterior ventricular branch, arising from a hypoplastic right coronary artery, and the right atrium.

**Figure 10.** Left selective coronary angiography in the anteroposterior projection, showing coronary-bronchial artery anastomoses.

**Figure 11.** Right selective coronary angiography in the anteroposterior projection during the emptying phase, showing anastomoses between the right coronary artery and the bronchial arteries.
cases, when an expected coronary branch is not visualized by selective coronary angiography, aortography may be advisable. There were no complications from selective coronary angiography during our investigation. Selective coronary angiography, moreover, requires injecting only small amounts of contrast material.

References

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