Surgical Management in Tetralogy of Fallot and Vascular Ring

Inderjit S. Virdi, Barry R. Keeton, Darryl F. Shore, and James L. Monro

Wessex Cardiothoracic Centre, Southampton General Hospital, Southampton, England

SUMMARY. The management of three infants born with a combination of tetralogy of Fallot and a vascular ring causing tracheoesophageal compression is described. There was a double aortic arch in two patients and an aberrant left subclavian artery with left ligamentum arteriosum and right aortic arch in one. Single-stage corrective surgery of both lesions during infancy, performed under profound hypothermia and circulatory arrest, was successful. In our opinion, this is the treatment of choice, when the anatomy of the tetralogy is favorable for primary correction.

KEY WORDS: Tetralogy of Fallot — Vascular ring — Surgery — Infants

The term vascular ring has been used to describe a variety of vascular anomalies which result from an abnormal development of the aortic arch complex [14] and produce tracheoesophageal compression. These are relatively uncommon anomalies comprising less than 1% of operable congenital cardiac malformations [8]. In association with other serious congenital cardiac lesions, such as tetralogy of Fallot, they are even rarer and have only rarely been reported [2, 8, 13, 14]. Successful single-stage repair of the two lesions during infancy has not, to our best knowledge, been reported, and surgery in other reports has involved division of the vascular ring with a palliative systemic-pulmonary shunt performed through a left thoracotomy [2, 13, 14].

Surgical Technique

The three infants were operated upon using standard anesthetic technique and surface cooling to 25°C. Using a midline thoracotomy, the innominate vein and the aortic arch and its branches were mobilized, enabling division of the appropriate vessel to release compression from the vascular ring. Cardiopulmonary bypass was then established and the infant cooled further on bypass to 18°C. At this temperature, the circulation was arrested and the intracardiac repair carried out during a single period of circulatory arrest lasting from 37 to 65 min (Table 1).

Case Reports

Case 1

A baby girl was referred to another hospital for a cardiological opinion on the second day of life when a murmur was heard, associated with cyanosis and laryngeal stridor. She initially did fairly well, but with worsening cyanotic spells was referred to us at 9 months of age. Angiography confirmed the clinical diagnosis of tetralogy of Fallot and also showed the abnormal arrangement of the great arteries (Fig. 1). There was a dominant right aortic arch with the left common carotid, right common carotid, and right subclavian arteries, coming off this arch separately. The nondominant left arch arose from the aorta close to the origin of the left carotid artery, passed to the left, anterior to the trachea, and gave rise to the left subclavian artery. It joined the descending thoracic aorta posterior to the oesophagus, thus forming a double aortic arch. No duct was seen (Fig. 2).

Surgery was performed at 9 months of age. After dissecting out the great arteries and clamping the anterior left arch close to its origin from the ascending aorta, a good pulse could still be felt in the left arm. Because of this, the left arch was divided proximally, thus relieving compression of the ring. The tetralogy of
A 6-week-old boy was referred to us with severe cyanosis and failure to thrive. He had been noted at birth to have a murmur and a rather unusual high-pitched cry. Angiography confirmed the clinical diagnosis of tetralogy of Fallot and demonstrated the abnormal arrangement of the great arteries (Figs. 3 and 4). There was a right-sided aortic arch; the common carotid arteries originated close together from the arch followed by the right subclavian artery. The nondominant left arch came off to the left of these vessels and ran across the trachea anteriorly. It gave rise to the left subclavian artery and ductus arteriosus before continuing to join the descending aorta, thus completing the ring and causing tracheoesophageal compression (Fig. 4).

Fig. 1. Aortogram in case 1 in left anterior oblique projection showing the double aortic arch: DRAA, dominant right aortic arch; LAR, left arch remnant; LCC, left common carotid artery; LS, left subclavian artery; RCC, right common carotid artery; and RS, right subclavian artery.

Fig. 2. Diagram of anatomy in case 1: abbreviations as for Fig. 1.

Fig. 3. Aortogram in case 2 in frontal view showing the double aortic arch: PDA, persistent ductus arteriosus; other abbreviations as for Fig. 1.

stridor was much better and had disappeared when she was reviewed after 2 months.

Case 2

Fallot was then corrected, the ventricular septal defect being closed through the right atrium, with resection of obstructing muscle in the right ventricular outflow. A transangular patch was not necessary and postoperatively she made a good recovery, being extubated 6 h later. Particular care was needed to keep her trachea free of secretions, but on discharge, 13 days later, the

Table 1. Operative details

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Sex</th>
<th>Weight (kg)</th>
<th>Date of operation</th>
<th>Age at operation</th>
<th>Duration of circulatory arrest (min)</th>
<th>Outflow patch</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Female</td>
<td>6.3</td>
<td>16.07.80</td>
<td>9 months</td>
<td>48</td>
<td>None</td>
</tr>
<tr>
<td>2</td>
<td>Male</td>
<td>2.9</td>
<td>15.09.80</td>
<td>6 weeks</td>
<td>65</td>
<td>Pericardium</td>
</tr>
<tr>
<td>3</td>
<td>Male</td>
<td>5.5</td>
<td>03.08.85</td>
<td>4 months</td>
<td>37</td>
<td>Durameter</td>
</tr>
</tbody>
</table>

Angiography showed the double aortic arch with the common carotid arteries originating close together from the arch followed by the right subclavian artery. The nondominant left arch came off to the left of these vessels and ran across the trachea anteriorly. It gave rise to the left subclavian artery and ductus arteriosus before continuing to join the descending aorta, thus completing the ring and causing tracheoesophageal compression (Fig. 4).