Tetralogy of Fallot Associated with a Double Aortic Arch
Palliation with a Prosthetic Tube Graft

Tetralogy of Fallot associated with a double aortic arch is an exceptionally rare congenital anomaly. Among the 785 patients who underwent definitive repair of tetralogy of Fallot between 1969 and 1985 at our center, only 1—a 19-month-old girl—had a double aortic arch. This report describes her case history, which included successful palliative repair with a polytetrafluoroethylene (PTFE) tube graft. (Texas Heart Institute Journal 1988;15:131-133)

Tetralogy of Fallot associated with a double aortic arch is an exceptionally uncommon congenital disorder. To our knowledge, only 12 cases of this anomaly have been reported in the literature: 8 of these have involved a right-sided descending aorta.1-6

This report describes an additional case, in which palliative repair was achieved by using a polytetrafluoroethylene (PTFE) tube graft to create a shunt from the aortic arch to the pulmonary artery.

Case Report

A 19-month-old girl was referred to our institution because of increasing cyanosis and numerous episodes of stridor. Physical examination upon admission showed a well-nourished, pink-complexioned child with mild-to-moderate cyanosis during meals and during crying spells. A grade 3 6 pansystolic murmur could be heard over the centrum cordis, and a grade 3 6 ejection systolic murmur was audible over the pulmonary artery. Breath sounds were normal, and there was no evidence of cardiac failure; the liver and spleen were not enlarged. Electrocardiography revealed sinus rhythm with right-axis deviation, and right ventricular hypertrophy. Chest radiography disclosed a boot-shaped heart and normal lung fields. These findings were consistent with a diagnosis of tetralogy of Fallot.

Cardiac catheterization confirmed the suspected diagnosis. Except for severe stenosis at the infundibular level, the pulmonary anulus appeared normal in size; the pulmonary trunk was mildly hypoplastic. Further stenosis was present at the level of the bifurcation. The pulmonary artery branches were normal. Right ventriculography (Fig. 1) revealed the presence of a double aortic arch: the left carotid and subclavian arteries arose from the smaller, anterior arch, whereas the right carotid and subclavian arteries arose from the larger, posterior arch (Fig. 2). Posteriorly, the two arches rejoined each other on the left side of the spine. A patent ductus arteriosus was seen to arise from the right arch. Because we suspected that this case involved an anomalous origin of the left anterior descending coronary artery from the right coronary artery, we decided to postpone definitive repair in favor of palliation.

Palliative Surgery
At age 23 months, the patient underwent palliative surgery. We performed a left posterolateral thoracotomy through the fourth intercostal space. The
suspected extracardiac anatomy was confirmed, and the vascular ring was seen to compress the esophagus tightly. We first divided the long, narrow, patent ductus arteriosus. Next we divided the anterior aortic arch at its junction with the descending aorta, and freed its adhesions to the esophagus. The proximal end of the arch was anastomosed end-to-end to a 6-mm PTFE tube graft, whose distal end was then anastomosed end-to-side to the left pulmonary artery, creating an unusual (perhaps unique) aorto-pulmonary shunt (Fig. 3).

The patient's postoperative course was uneventful. A continuous murmur could be heard parasternally over the second intercostal space, and chest radiography showed increased vascular markings bilaterally; these were evidence of a well-functioning shunt.

**Definitive Repair**

During the 4 years after palliation, the patient's growth rate was satisfactory. At 6 years of age, she was readmitted for repeat cardiac catheterization, which confirmed the previous diagnosis, as well as the presence of a well-functioning aorto-
pulmonary shunt (Fig. 4). Selective coronary angiography revealed a normal coronary artery pattern. The patient then underwent definitive repair, during which the PTFE graft was carefully isolated, and closed with two metallic clips. After resection of many of the parietal bands and closure of the ventricular septal defect, it was necessary to perform a pulmonary commissurotomy and to place a pericardial patch over the pulmonary artery to the point of bifurcation.

The patient recovered uneventfully. At her most recent follow-up examination, 14 months after definitive repair, she was asymptomatic and in good general condition. Electrocardiography revealed sinus rhythm, with a right bundle-branch block. A grade 1-2/6 systolic ejection murmur could be heard over the pulmonary artery.

**Discussion**

Use of the smaller arch for creating an aorto-pulmonary shunt in cases of double aortic arch was first described by Blalock and Bahnson;6 Binet and associates7 subsequently proposed a method by which the proximal aortic stump could be anastomosed directly to the left pulmonary artery, after adequate length had been obtained by dividing the left subclavian artery.

The use of a PTFE graft to elongate the proximal stump of the divided arch, as in our case, offers the unique advantage of preserving the subclavian artery.8 Although rare, arm ischemia has been reported after the Blalock procedure.9 Moreover, the division of one subclavian artery can result in unequal growth of the arms.10 Because of these considerations, we chose to spare the subclavian artery of our young patient by using a prosthetic tube graft, and to perform subsequent total correction. Choosing the correct conduit size proved difficult: in the Blalock procedure, the diameter of the subclavian artery usually imposes a natural limitation upon shunt flow;11 but in this instance the aortic diameter might have permitted excessive pulmonary flow. Close follow-up, including radiography of the lung fields, has since confirmed the adequacy of flow—an observation further confirmed by recatheterization, which indicated a mean pulmonary arterial pressure of 14 mmHg, with no evidence of increased vascular resistance. At the time of definitive repair, no relevant complications were encountered.

On the basis of these results, we consider the use of a PTFE graft to be a safe, effective method for palliation of this rare congenital malformation.

**References**