Tetralogy of Fallot with Coexisting Type II Aortopulmonary Window
Fred A. Crawford, Jr., M.D., David G. Watson, M.D., and James A. Joransen, M.D.

ABSTRACT An infant had a coexisting tetralogy of Fallot and type II aortopulmonary window between the ascending aorta and the right pulmonary artery in which the communication acted as a palliative systemic-pulmonary shunt. Surgical repair is described, and the appropriate literature is reviewed.

Aortopulmonary windows are unusual cardiac defects, which usually consist of a communication between the ascending aorta and the main pulmonary artery [5, 10, 19]. They have been reported to occasionally coexist with other defects such as ventricular septal defect, patent ductus arteriosus, and more rarely with tetralogy of Fallot [3, 6, 10, 17]. A different type of aortopulmonary window between the ascending aorta and the right pulmonary artery at its origin has recently been described in several patients [3, 14, 19]. This case report describes a patient with tetralogy of Fallot and such an aortopulmonary window which in location and effect was identical to a palliative Waterston-Cooley shunt.

A 3-month-old girl was referred to the University of Mississippi Medical Center in January, 1977, for evaluation of a heart murmur and congestive heart failure. An older brother had recently undergone successful repair of tetralogy of Fallot. Physical examination revealed a small acyanotic infant with a grade 4/6 holosystolic murmur heard along the left sternal border. The liver was 7 cm below the right costal margin. Electrocardiogram demonstrated combined ventricular hypertrophy with right atrial enlargement. Chest roentgenogram showed a moderately enlarged heart with increased pulmonary vascular markings and pulmonary venous congestion. Cardiac catheterization demonstrated a large ventricular septal defect and infundibular pulmonary stenosis, a 3.9:1 left-to-right shunt with most of the step-up in oxygen saturation occurring at the level of the pulmonary artery (Table).

The patient responded well to digitalis and Lasix (furosemide), and was followed in the cardiac clinic without symptoms other than frequent bronchitis. Because of continued cardiomegaly and increased pulmonary flow when she was 2 years old, she underwent repeat cardiac catheterization (see Table). Again there was a gradient between the right ventricle and the main pulmonary artery and between the main pulmonary artery and the right pulmonary artery. A right ventriculogram showed moderately severe infundibular narrowing, and the pulmonary valve annulus was approximately two-thirds the size of the aortic valve annulus. The anteroposterior pulmonary arteriogram revealed a large negative jet entering the pulmonary artery at the junction between the right pulmonary artery and the main pulmonary artery. A diagnosis of tetralogy of Fallot and aortopulmonary window was made, and the child was referred for operation.

At operation, a communication between the posterior ascending aorta and the anterior right pulmonary artery distal to the bifurcation was identified. After isolation of the right pulmonary artery proximal and distal to the communication, cardiopulmonary bypass was begun and the patient cooled to 26°C. The aorta was cross-clamped and a potassium cardioplegic solution infused into the aortic root. A transverse aortotomy was made, and the communication was identified (Figure, A). It was roughly circular in shape, measured 7 mm in diameter, and was well above the coronary ostia and approximately 1 cm distal to the origin of the right pulmonary artery. The defect was easily closed with a Dacron patch without

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Accepted for publication Jan 15, 1980.

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Preoperative Cardiac Catheterization

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<th>Catheter Site</th>
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<th>Saturation (%)</th>
<th>Pressure (mm Hg)</th>
<th>Saturation (%)</th>
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<td>...</td>
<td>63</td>
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</table>

NA = not applicable; QP:QS = pulmonary–systemic perfusion ratio.

(A) Transverse aortotomy exposing the communication between the posterior ascending aorta and the right pulmonary artery (RPA). The tourniquets occlude the RPA proximal and distal to the communication. (B) Communication between the aorta and the RPA has been closed with a Dacron patch.
distortion of the right pulmonary artery (Figure, B). The aortotomy was closed and the tetralogy repaired by closing the ventricular septal defect and patching the right ventricular outflow tract across the pulmonary valve annulus. After cardiopulmonary bypass was discontinued, the ratio of right ventricular pressure to left ventricular pressure was 0.3. Indicator dilution curve showed no residual shunt. The patient had an uneventful hospital course and was discharged on the seventh postoperative day.

Comment

Aortopulmonary windows occur infrequently and represent disorders of development of the truncusconal septum. The anatomy and embryology have been reviewed in detail previously [5, 10, 19]. Typically, the defect consists of an “isolated window-like communication between the adjacent portions of the ascending aorta and the pulmonary arterial trunk” [10]. The defect usually is just above the left coronary artery. In 1975, Todd and co-workers [19] described a single patient in whom the defect was between the ascending aorta and the right pulmonary artery and indicated that no other similar patient had ever been described. Since then, two additional reports have described 3 [14] and 4 [9] patients who demonstrated variants of the typical aortopulmonary window in which the communication was between the aorta and the origin of the right pulmonary artery. Richardson [14], Mori [9], and their associates classified these variants as type II aortopulmonary windows and reviewed the embryology of their formation. None of these patients had associated heart defects.

Associated defects do occur with typical aortopulmonary windows and include patent ductus arteriosus, ventricular septal defect, and coarctation [10, 17]. Cooley and co-workers [6] described a patient with a coexisting typical aortopulmonary window and tetralogy of Fallot in whom both defects were successfully repaired. Tandon and colleagues [17] reported a patient with a typical aortopulmonary window and tetralogy of Fallot who died shortly after cardiac catheterization. Clarke and Richardson [5] reported 2 children with this combination who underwent successful operation, as did Blieden and Moller [1]. In a series of 41 infants undergoing repair of tetralogy of Fallot, Castaneda and associates [2] had 1 12-hour-old infant with a coexisting typical aortopulmonary window and severe congestive heart failure who required operation. This infant and another from the University of Alabama were subsequently reported separately by Castaneda and Kirklin [3], who commented on the rarity of this combination. Such a combination of defects is indeed quite unusual [4, 12, 16].

Our patient is particularly unusual in that the aortopulmonary window was between the ascending aorta and the right pulmonary artery. Such a communication has been described previously in only 8 patients and never, to our knowledge, in a patient with tetralogy of Fallot. The size and location of the communication made it almost exactly analogous to a palliative aortopulmonary shunt of the Waterston-Cooley type [20]. This undoubtedly was the reason for the normal size and development and lack of symptoms manifested by the child. Despite the lack of symptoms, operation was recommended at this time because of the elevated pulmonary artery pressure and the known tendency for such communications—both congenital and man-made—to result in pulmonary vascular changes [10, 11, 13, 18]. Castaneda and Kirklin [3] recommended repair of this combination as soon as the diagnosis is made. Typical aortopulmonary windows are best repaired by a patch through the aorta [5, 7, 21]. While some have advocated complete separation of the aorta and pulmonary artery in repair of tetralogy after aortopulmonary anastomosis, others including ourselves have not [8, 15]. We elected to simply close the communication with a patch from within the aorta because of the large size of the right pulmonary artery and the lack of distortion. We also have preferred to use this technique in uncomplicated aortopulmonary windows. The patient remains well and asymptomatic.

References