ONE OF THE GREAT advances in the treatment of congenital heart disease is the surgical correction of tetralogy of Fallot.1-3 Tetralogy implies: (1) right ventricular outflow tract (pulmonary infundibulum) stenosis or atresia; (2) ventricular septal defect; (3) a dextroposed aorta which overrides the ventricular septum; and (4) right ventricular hypertrophy.

Diagnostic angiocardiography combined with cardiac catheterization is essential prior to surgical correction of tetralogy of Fallot and is highly desirable prior to any palliative surgical procedure used for the treatment of this condition.4 The main objectives of angiocardiography are to: (1) demonstrate the right ventricular outflow tract and

Figure 1

Diagram of the right ventricular outflow tract in tetralogy of Fallot. (A) Frontal projection. (B) Lateral projection. INF = infundibular stenosis; A = annulus of pulmonary valve; V = site of stenosis in domed pulmonary valve; VSD = ventricular septal defect; RV = right ventricle; LV = left ventricle; CS = crista supraventricularis; MV = mitral valve orifice (linear structure adjacent to MV is anterical mitral (leaflet); MPA = main pulmonary artery; RPA = right pulmonary artery; LPA = left pulmonary artery; LA = site of ligamentum arteriosus at apex of ductus stump which arises from proximal left pulmonary artery.
pulmonary arteries including sites of obstruction and other anatomic abnormalities; (2) demonstrate the presence of a ventricular septal defect—most crucial and difficult is the documentation, if possible, that two ventricles (not a single ventricle) and two atrioventricular valves are present; (3) rule out other anomalies such as transposition or congenitally corrected transposition of the great vessels. Ebstein's anomaly of the tricuspid valve, anomalous pulmonary venous connection, patent ductus arteriosus, and aortic obstruction; (4) demonstrate the status of any previous palliative operations including secondary effects on the pulmonary vasculature; and (5) show any major anomalies in the coronary arteries. In severe tetralogy of Fallot the right coronary artery often supplies large conus and ventricular branches or rarely the major anterior descending coronary artery, vessels which may be injured at ventriculotomy or prevent adequate surgical exposure.

Selective right ventricular angiography, particularly with biplane serial filming, is optimal for the preoperative evaluation of the right ventricular outflow tract.

Figure 2
Pulmonary valve and infundibular stenosis in tetralogy. Early systole showing domed valve (V), annulus (A), and long, moderately narrow infundibulum (INF).
**Right Ventricular Infundibulum**

Atresia or hypoplasia of the conus of the right ventricle is the basic developmental defect in tetralogy of Fallot. Not only does this basic defect narrow the right ventricular outflow tract, but it also results in a ventricular septal defect as well as abnormal anterior and leftward positioning of the conus septum, the crista supraventricularis (fig. 1). Secondary muscle hypertrophy further narrows the infundibulum (figs. 2 and 3).

Between the narrowed mouth of the infundibulum and the pulmonary valve is a variably sized infundibular chamber, the “third ventricle.” The part of this chamber with muscular walls can be identified on the angiocardiogram as it constricts during ventricular systole. Often, the largest part of this apparent chamber is a region enclosed by the stenotic pulmonary valve as it balloons upward during systole (fig. 2).

Numerous variations in pathologic anatomy are found in patients carrying the clinical diagnosis of tetralogy of Fallot, about which the surgeon must be forewarned. For example narrowing of the right ventricular infundibulum can also be produced by a

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**Figure 3**

Pulmonary valve and infundibular stenosis in tetralogy. (Same case as fig. 2.) Later in systole the infundibulum has markedly narrowed. CS = crista supraventricularis; LV = left ventricle.
bulging aneurysm of an aortic sinus of Valsalva or an aortic valve leaflet prolapsing into a supracristal ventricular septal defect. Hypertrophied aberrant muscle trabeculae sometimes obstruct the right ventricle proximal to the os infundibulum. Anomalous coronary arteries should be particularly looked for in the vicinity of the right ventricular outflow tract, the customary site for ventriculotomy. Accidental interruption of one of these vessels can cause an operative mortality. Even if recognized such anomalous vessels may prevent optimal resection of infundibular muscle.

Other important information supplied by the selective right ventricular angiograms include demonstration of the ventricular septal defect, especially in lateral view (fig. 4, left), as well as the left ventricular outflow tract and the characteristically large ascending aorta. In the lateral projection the relationship of the aortic root to the ventricular septum (i.e. the degree of aortic overriding) can be evaluated. Also in this view continuity between the aortic root and the anterior mitral leaflet may be demonstrated (fig. 4, right), a relationship of critical importance in distinguishing the dextropositioned aorta of tetralogy from kindred forms of double-outlet right ventricle and transposition of the great vessels. A small cavity of the right ventricle suggests a small stroke output and that right-to-left shunting may be occurring proximal to the right ventricle, for example, by way of an atrial septal defect or rarely by an anomalous connection of systemic veins to the left atrium.

**Pulmonary Valve**

The pulmonary valve may or may not be stenotic in tetralogy of Fallot. When stenotic (about 60% of the cases) the thickened, fused valve cusps balloon in systole and collapse in diastole. Occasionally, the valve is so deformed or dysplastic that little motion is possible. During ventricular systole, a jet of contrast material may be seen spurting through the stenotic valve. In this phase, the abnormal, domed cusps are readily shown in the lateral view since not only is the tissue thick but the X-ray beam is tangential to some aspect of the domed valve (figs. 2–8). In contrast, normal pulmonary valve cusps usually are not seen during systole.

In about 15% of clinical cases, the pulmonary valve stenosis is the only site of outflow tract

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**Figure 4**

Infundibular and valvular stenosis in tetralogy. (Left) Lateral view in diastole showing severely stenotic infundibulum (INF) and domed valve (V) with large right-to-left shunt via ventricular septal defect (VSD) opacifying undersurface of aortic valve leaflets. (Right) Succeeding diastole shows aortic opacification obscuring much of right ventricular outflow tract. The straight margin of the open anterior mitral valve leaflet (MV) is in continuity with aortic valve (AV), ruling out double-outlet right ventricle and transposition of the great vessels.
obstruction, although considerable infundibular narrowing may occur in late systole (fig. 8). In many of the other cases in which the valve is not stenotic, it is bicuspid. This abnormality can also

Figure 5
Infundibular and valvular stenosis in severe tetralogy. Early demonstration of severe infundibular stenosis (INF) and jet (arrow) of contrast material emerging from stenotic pulmonary valve (V). Contrast material also opacifies undersurface of aortic valve (AV), because of large ventricular defect and right to left shunt.
produce a domed configuration on the angiogram, during systole. Bicuspid valves are frequently associated with a small annulus and pulmonary artery.

Figure 6
Infundibular and valvular stenosis in severe tetrology. (Same case as fig. 5) Diastole after aortic opacification by large right-to-left shunt at ventricular level reveals persistent infundibular stenosis (INF) and collapsed pulmonary valve at level of small pulmonary annulus (A). A Blalock anastomosis (B) connects left subclavian artery to left pulmonary artery.
**Pulmonary Valve Annulus**

The site of basal attachment of the pulmonary valve cusps is the valve ring or annulus. Observation of the valve cusps in both closed and open positions is helpful in identifying the annulus and in judging its size (fig. 7). The diameters of the annulus and the adjacent pulmonary artery are very important. If the two are of adequate size it is not necessary to widen the region by grafting and thus gross pulmonary valve regurgitation will not be produced by the surgical correction.

**Main Pulmonary Artery**

The main pulmonary artery is often sharply narrowed (fig. 7) just beyond the level of the ballooned pulmonary valve. This narrowing may be the consequence of low pressures lateral to the jet of blood emerging from the stenotic valve. Some narrowing or hypoplasia is also commonly present more distally at the bifurcation of the main pulmonary artery, especially at the origin of the right branch—though these regions are usually distensible enough so that enlargement by surgery is not required.

In those cases, of course, where the main pulmonary artery or the pulmonary valve annulus is very hypoplastic or even atretic more radical methods for surgical correction are necessary. In such situations demonstration of the existing pulmonary arteries and their sources of blood supply is critical to the planning of surgical correction.

**Peripheral Pulmonary Arteries**

Significant peripheral stenoses of the right or left pulmonary arteries, especially as these vessels divide in the hilar regions, are occasionally encountered in tetralogy of Fallot. In severe tetralogy episodes of thrombosis may occur in the pulmonary arteries with or without secondary infarcts or residual pulmonary arterial stenosis. The significance of such lesions must be judged by pressure measurements and observations concerning relative blood flow. An artery carrying blood at low pressure usually appears smaller than when distended with blood at higher pressure. In each

*Figure 7*

*Pulmonary valve stenosis in tetralogy. (Left) Lateral view in late systole showing domed stenotic valve (V) and narrowed infundibulum (INF). Mild narrowing of the main pulmonary artery is seen above valve. (Right) In diastole the valve is closed and both the annulus (A) and the infundibulum (INF) are wide.*

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case, the pulmonary vascularity in the various regions of the lung, as shown in plain chest roentgenograms, should be correlated with the findings at cardiac catheterization and angiocardiography in the search for abnormal regions of lung perfusion. Congenital absence of the proximal left pulmonary artery, though characteristic of tetralogy of Fallot, is only occasionally encountered (fig. 8). The peripheral left pulmonary artery in these cases is usually supplied by a patent ductus, which, in the case of a right aortic arch, typically arises from the left subclavian artery. Exceptionally, one of the pulmonary arteries arises directly from the ascending aorta.

Collateral arterial blood supply to the lung in tetralogy of Fallot is usually conspicuous. Previous palliative operations tend to increase the collateral arterial circulation. As mentioned, large arteries arising directly from the aorta or bronchiocephalic vessels may supply portions of the lung in an anomalous fashion. Catastrophe can be averted if such variations are appreciated preoperatively.

Effects of Previous Palliative Surgery

The effects of previous palliative surgical procedures upon the pulmonary arteries must be known prior to corrective surgery. In the neonatal period palliative augmentation of pulmonary blood flow is usually achieved by side-to-side anastomoses between the dorsal aspect of the ascending aorta and the right pulmonary artery (Waterston). The Potts anastomosis (between the left pulmonary artery and the proximal descending aorta) can also give excellent palliation but is notoriously difficult to

Figure 8
Absent proximal left pulmonary artery in tetralogy. Early opacification of right ventricular outflow tract shows infundibular (INF) stenosis and domed bicuspid pulmonary valve (V). The proximal left pulmonary artery is absent. (The peripheral left pulmonary artery is of good size and is supplied by a patent ductus arteriosus.)

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close and thus is not used in patients who might be expected to have subsequent corrective open-heart surgery. When technically feasible in older infants and children the Blalock-Taussig operation (end-to-side subclavian artery-to-pulmonary artery) is the preferred palliative operation (Fig. 6). Blood flow by way of any of these shunts may be sufficiently large that interruption of the connection is required as a part of the corrective surgery. Frequently the pulmonary artery used in a palliative anastomosis is significantly obstructed just proximal or distal to the anastomosis site. If these arterial obstructions are clearly demonstrated preoperatively, successful repair at operation may be possible with important preservation of lung function.

A successful palliative systemic-to-pulmonary shunt occasionally is complicated by a gradual obliteration of the lumen of the right ventricular infundibulum by hypertrophied muscle so that pulmonary atresia is acquired. Fortunately, surgical correction of this condition by extensive infundibular muscle excision is often possible.

References


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