CONGENITAL HEART DISEASE

Correlation of Pathologic Anatomy and Angiocardiography

VOLUME II

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DECREASED PULMONARY ARTERIAL VASCULATURE; CYANOSIS PRESENT

DECREASED PULMONARY ARTERIAL VASCULATURE; CYANOSIS PRESENT

The association of diminution in pulmonary arterial vasculature roentgenographically with cyanosis or peripheral arterial desaturation is common. Anatomically, except for pulmonary arteriovenous fistula, each of the entities considered in this section exhibits: (1) an abnormal communication between the lesser and greater circulations, and (2) an obstruction to the flow of blood into the pulmonary circulation. As a result, there is a right-to-left shunt through the abnormal communication.

This combination of anatomic and physiologic features forms the basis for the coexistence of diminution in pulmonary blood flow (decreased pulmonary arterial vasculature) and cyanosis.

Cyanosis associated with increased pulmonary arterial vasculature, on the other hand, results from an admixture of blood from both sides of the heart. These lesions have been covered in Volume I.

Among the conditions to be covered in this section are the tetralogy of Fallot, pulmonary stenosis or atresia with intact ventricular septum and with right-to-left shunt at the atrial level, tricuspid atresia, congenital corrected transposition with pulmonary atresia or stenosis and ventricular septal defect as well as other functionally related anomalies.

Tetralogy of Fallot

General Considerations

The anatomic combination of ventricular septal defect, biventricular origin of the aorta, pulmonary or subpulmonary stenosis and right ventricular hypertrophy is commonly called the tetralogy of Fallot.

Obstruction to pulmonary blood flow (so-called pulmonary stenosis) may result from a narrow state of one or several anatomic structures, including the right ventricular infundibulum, the pulmonary valve and/or the pulmonary trunk. The obstruction may take the form of atresia, in which no channel for the flow of blood is present, or stenosis, in which a narrow opening is present.

In the tetralogy of Fallot it is common that cases with pulmonary atresia and pulmonary stenosis be considered together. Since, however, the potential for surgical therapy is different, we shall present separate coverage for these two types of the condition. Tetralogy of Fallot with pulmonary stenosis will be presented in the immediately following pages under the title of Tetralogy of Fallot.

Tetralogy of Fallot with pulmonary atresia will then follow under the title of Pseudotruncus Arteriosus.

Tetralogy of Fallot

Pathologic Anatomy

In the tetralogy of Fallot the external relationship between the ascending aorta and the pulmonary trunk is nearly normal. Usually, however, there is an obvious disproportion in size between these two vessels, the aorta being wider than normal while the pulmonary trunk is narrow. The heart tends to display a rounded shape as the blunt apex of the heart is formed by the right ventricle.

Within the heart the specific elements of the entity are apparent. The ventricular septal defect lies in a basal position and is of the "large" variety, being sufficiently wide to allow equalization of pressure between the two ventricles. The aorta straddles the ventricular septal defect and arises from both ventricles. The proportion of the aorta which arises from one ventricle and that which arises from the other varies. The feature of the aorta taking some of its origin from the right ventricle has been called "dextroposition of the aorta."

The infundibulum forms a narrow subdivision of the right ventricle and usually contributes to obstruction to pulmonary blood flow. Beyond the narrowed area there may be some dilatation of the infundibulum (the so-called third ventricle). The pulmonary valve is usually bicuspid and may be stenotic. The pulmonary trunk is more narrow than normal, but usually contributes less to pulmonary stenosis than do the changes in the right ventricular infundibulum or the pulmonary valve.

The right ventricular hypertrophy is concentric, the wall of this chamber being of about the same thickness as is the wall of the left ventricle.

Conditions that commonly accompany the tetralogy of Fallot include (1) a patent foramen ovale and (2) a right aortic arch.

Tetralogy of Fallot

Pathologic Anatomy: General Relations

On this page are shown two examples of the tetralogy of Fallot to illustrate typical structural changes of the malformation. In each instance the interior of the right ventricle is exposed and an incision has been carried from this chamber into the ascending aorta. Each shows a narrow infundibular chamber, the ventricular septal defect overhung by the aorta and the right ventricular hypertrophy

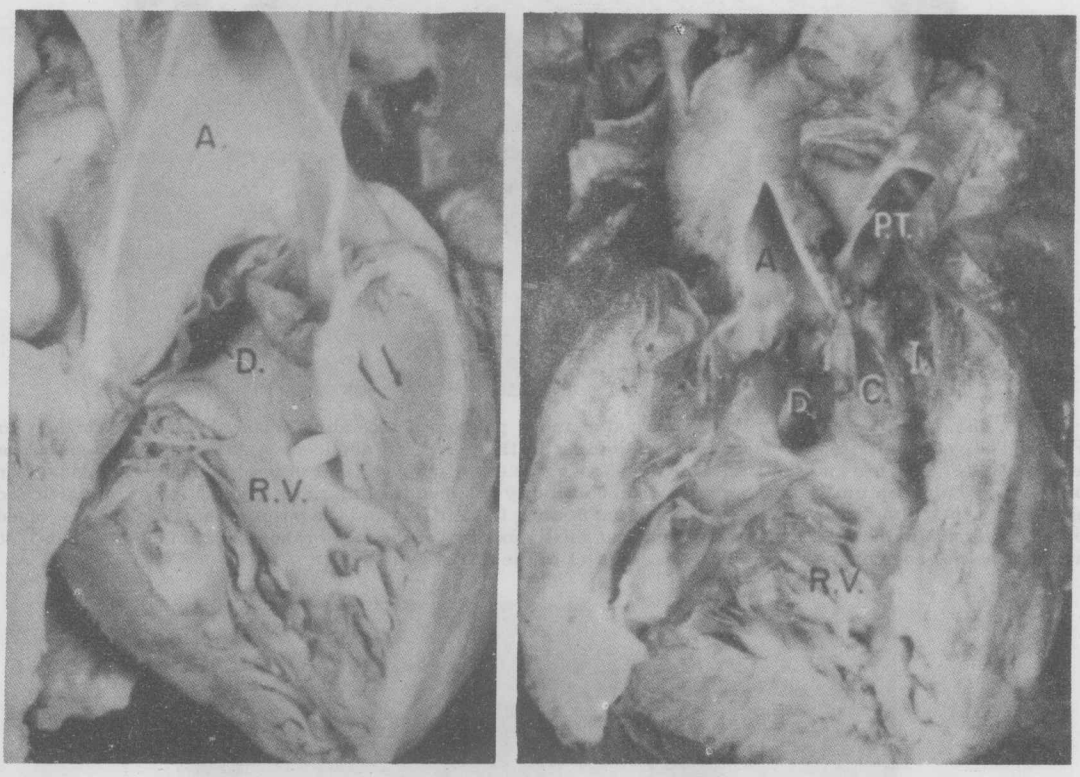

Fig. 484. Interior of right ventricle (R.V.) and ascending aorta (A.) in two cases of the tetralogy of Fallot. *Left.* In this specimen the subpulmonary infundibular channel (probe) has not been opened. The ascending aorta straddles the ventricular septal defect (D.). *Right.* This specimen shows similar relationship between the aorta and the ventricular septal defect. In this instance the infundibular chamber has been opened, as has been the origin of the pulmonary trunk (P.T.). The infundibular chamber (I.) is made narrow by a vertical hypertrophied crista supraventricularis (C.).

Tetralogy of Fallot

Pathologic Anatomy: Relations of Aortic Valve to Ventricular Septum and Mitral Valve

On this and the following page are emphasized the relationships of the aortic valve to the mitral valve and to the ventricular septal defect in typical examples of the tetralogy of Fallot.

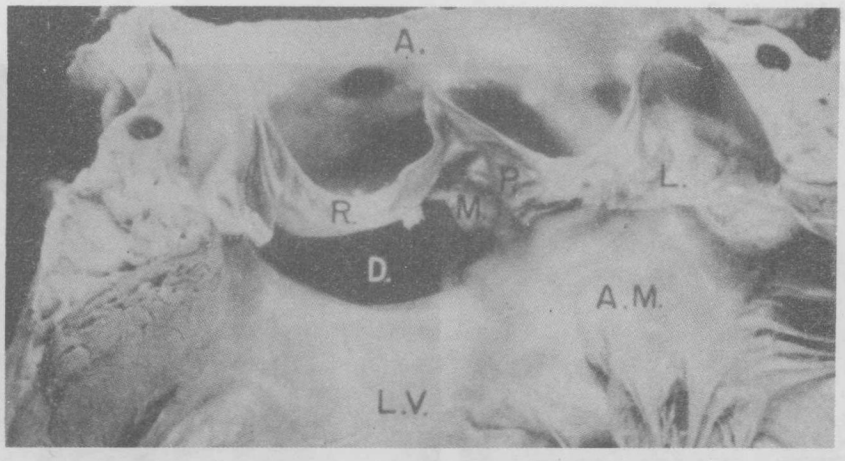

Fig. 485. Left ventricle (L.V.), aortic valve and a portion of ascending aorta (A.) in the tetralogy of Fallot. The ventricular septal defect (D.) lies inferior to the right (R.) aortic cusp. Attached to the posterior aortic cusp (P.) and the anterior leaflet of the mitral valve (A.M.) is a segment of residual membranous portion of the ventricular septum (M.). There is normal continuity between the anterior leaflet of the mitral valve and elements of the pulmonary valve. As an incidental finding, the left (L.) and the posterior aortic cusps are joined by an acquired adhesion. It is not uncommon that small remnants of the membranous septum may be identified at the posterior edge of the ventricular septal defect in the tetralogy of Fallot.

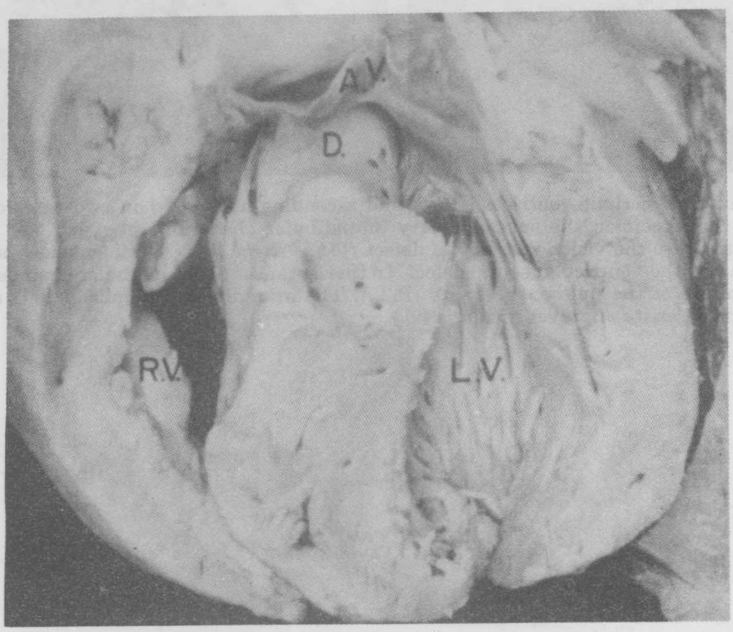

Fig. 486. Sagittal section through the right ventricle (R.V.), ventricular septum and left ventricle (L.V.) in the tetralogy of Fallot. The aortic valve (A.V.) lies above the ventricular septal defect (D.). In this instance it is clear that the anterior leaflet of the mitral valve is continuous with the aortic valve. The aorta arises from both ventricles and straddles the ventricular septal defect, arising about equally from each ventricle.

PATHOLOGIC ANATOMY: RELATIONS OF AORTIC VALVE TO VENTRICULAR SEPTUM AND MITRAL VALVE (*continued*)

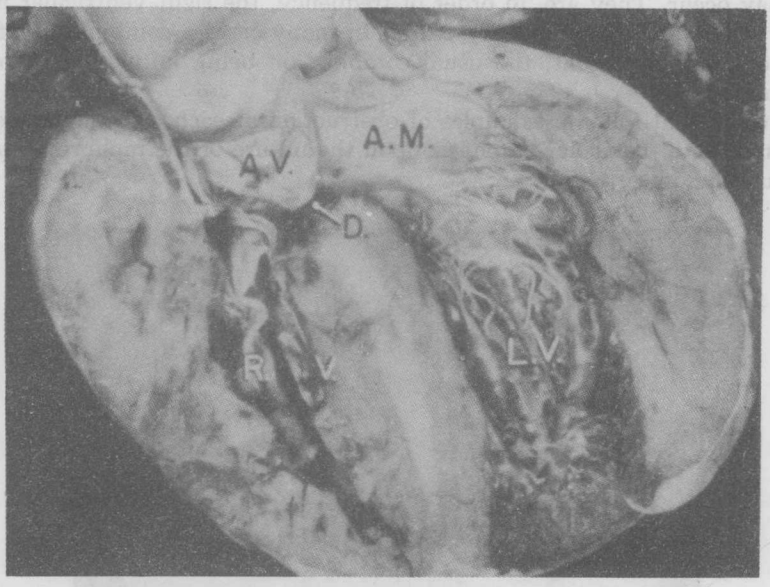

FIG. 487. Heart sectioned as in Figure 486 of the preceding page. In this instance the aortic valve (A.V.) appears to arise more above the left ventricle (L.V.) than above the right (R.V.). D. = Ventricular septal defect. A.M. = Anterior mitral leaflet.

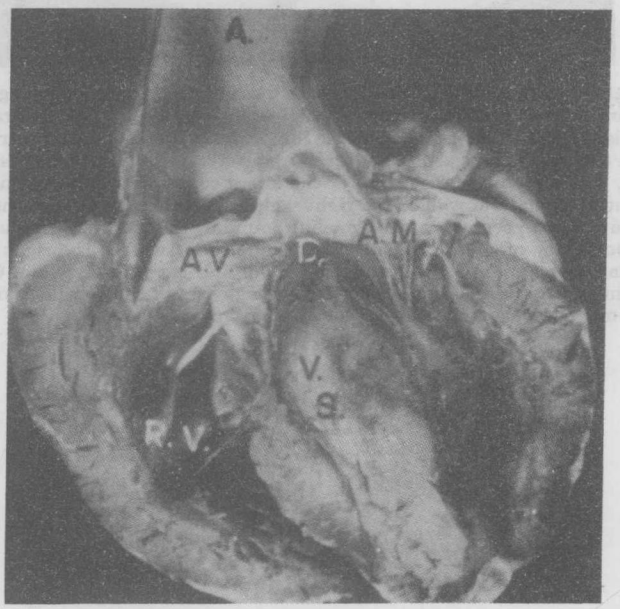

FIG. 488. The sagittal section of the heart in this case of the tetralogy of Fallot shows the aorta (A.) arising over the ventricular septum (V.S.), more from the right ventricle (R.V.) than from the left ventricle. The anatomic relations tend toward those of origin of both great vessels from the right ventricle, but, in the tetralogy of Fallot, with extreme positioning of the aorta over the right ventricle, there is continuity, as in this case, between the anterior leaflet of the mitral valve and the aortic valve. This feature distinguishes the tetralogy of Fallot from origin of both great vessels from the right ventricle.

Tetralogy of Fallot

Pathologic Anatomy: Basis for Pulmonary Stenosis

In the tetralogy of Fallot there are three possible sites at which obstruction to pulmonary arterial flow may occur. They are, in order of frequency, the right ventricular infundibulum, the pulmonary valve and the pulmonary trunk. In many cases a combination of obstructions at more than one site is present, the usual combination being infundibular and pulmonary valvular obstruction.

In the material that follows, examples of obstruction at each of the aforementioned sites will be illustrated, with particular emphasis upon the nature of the malformation of the right ventricular infundibulum.

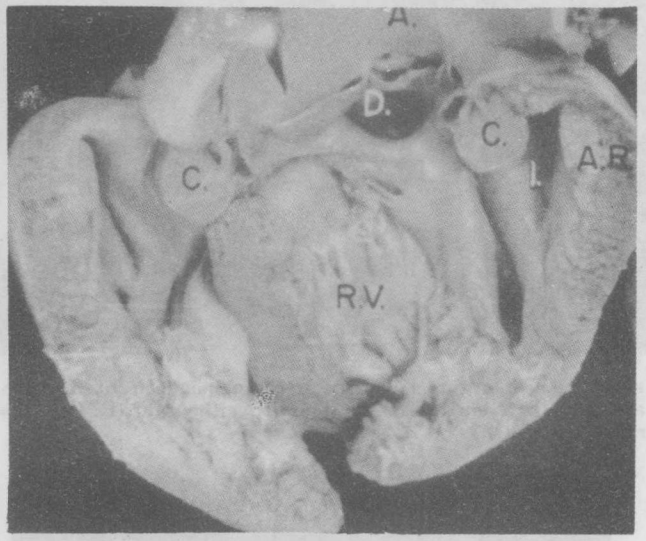

Fig. 489. The right ventricle (R.V.) and ascending aorta (A.) in the tetralogy of Fallot. The fundamental features of the infundibular malformation in the tetralogy of Fallot are illustrated. In the anterior and superior aspect of the right ventricle (right side of the illustration) lies the right ventricular infundibulum (I.). This is separated from the ventricular septal defect (D.) by a vertically oriented parietal limb of the crista supraventricularis (C.). The aforementioned limb of the crista supraventricularis runs from the septal wall of the right ventricle to the anterolateral wall of the right ventricle. In preparation of the specimen the crista has been divided and appears in two segments. The medial boundary of the infundibulum (I.) is the ventricular septum. The anterior boundary of the infundibulum is the anterior wall of the right ventricle (A.R.). In this instance the infundibular chamber is of fairly uniform caliber and is relatively wide. Considerable variation in the contour of this chamber and in its diameter occurs among cases of the tetralogy of Fallot. Other examples of the infundibular chamber will be demonstrated on the next page.

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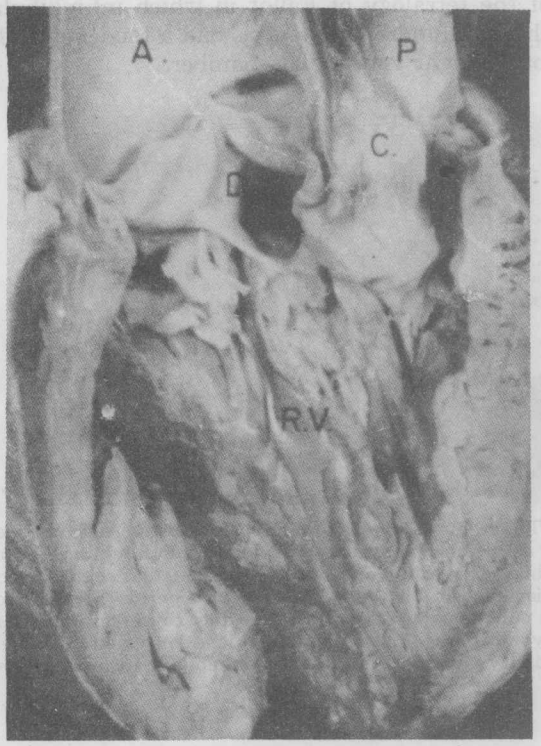
PATHOLOGIC ANATOMY: BASIS FOR PULMONARY STENOSIS (*continued*)



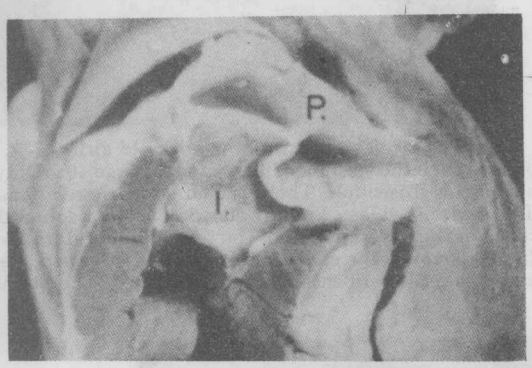


FIG. 490. Infundibular stenosis in the tetralogy of Fallot. *Left.* In this specimen the heart has been dissected as in the illustration on the preceding page and, in addition, the pulmonary valve has been opened to reveal the interior of the pulmonary trunk (P.). In this instance the infundibulum is relatively uniform in caliber, although its narrow state is accentuated somewhat at its inlet by the protrusion of the broad limb of the crista supraventricularis (C.) into the lower aspect of the infundibular chamber. D. = Ventricular septal defect. *Right.* In this instance the infundibular chamber of the right ventricle (I.) has been opened, as have the bicuspid pulmonary valve and the pulmonary trunk (P.). The infundibular chamber is relatively uniform, although, as in the instance in *Left*, the lowermost aspect is somewhat more narrow than is the upper aspect. There is endocardial thickening in the lower half of the infundibular chamber.

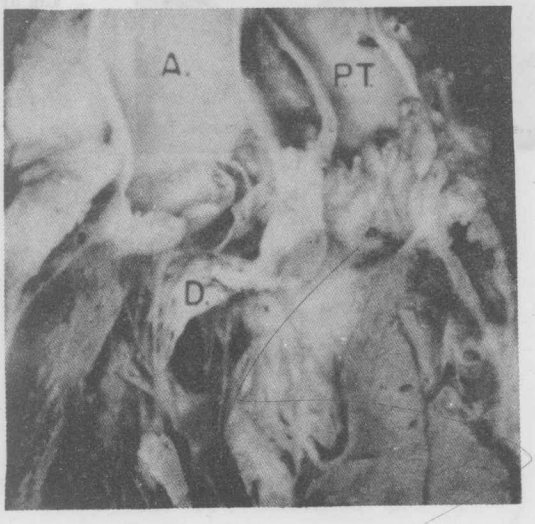


FIG. 491. Sagittal section through the outflow portion of the right ventricle including the infundibulum, the aorta (A.) and the pulmonary trunk (P.T.). The ventricular septal defect (D.) has been closed by the placement of a prosthetic patch. The right ventricular infundibulum is somewhat sigmoid in shape and relatively wide. The pulmonary valve, which was stenotic, had been enlarged surgically.

Tetralogy of Fallot

PATHOLOGIC ANATOMY: BASIS FOR PULMONARY STENOSIS (*continued*)

On this page are emphasized examples of the tetralogy of Fallot in which infundibular stenosis was an important element but in which the infundibular stenosis had a tendency to be localized to the lowermost aspect of this part of the right ventricular chamber.

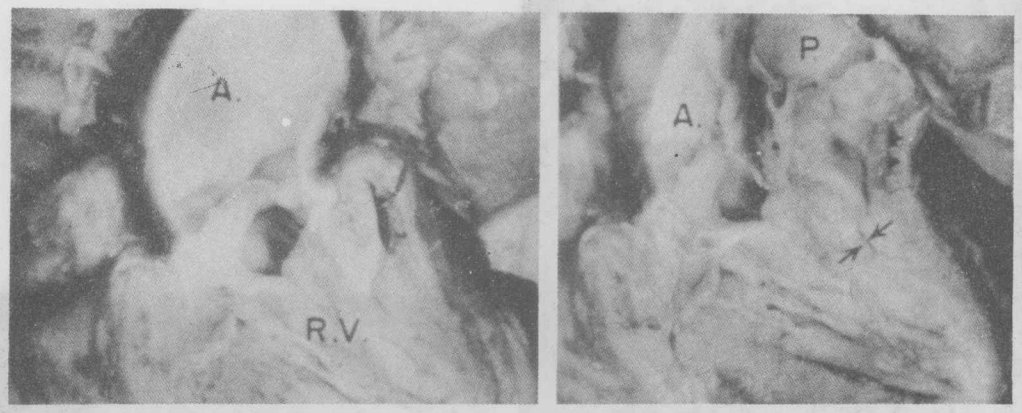


FIG. 492. The heart of a 6-week-old girl. *Left.* The right ventricle (R.V.) and the aorta (A.) have been opened as in the specimen illustrated on page 404. Only a portion of the right ventricular infundibulum is apparent in this dissection. There is characteristic relationship between the aorta and the ventricular septal defect. The aorta is wide. *Right.* Following the dissection displayed in *Left* a longitudinal incision has been carried through the anterior wall of the infundibulum into the pulmonary trunk (P.). At the inlet to the infundibulum (facing arrows) the channel is markedly narrowed. Beyond this the infundibulum widens somewhat, although even here, the caliber is relatively confined. The pulmonary valve is bicuspid. The pulmonary trunk is narrow

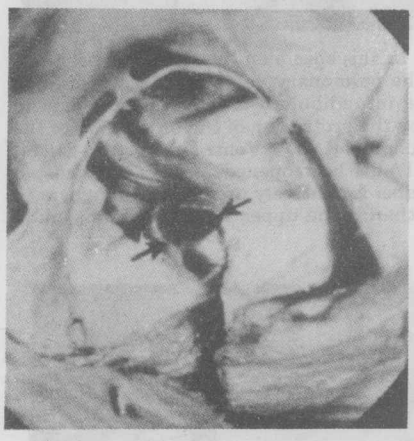


FIG. 493. Localized infundibular stenosis in the tetralogy of Fallot. A 30-year-old man. The right ventricular infundibulum is viewed through the pulmonary valve from above. The valve has three leaflets and the infundibulum immediately beneath the valve is relatively wide. At the lowermost aspect of the infundibulum the caliber of the channel is narrow (facing arrows).

Tetralogy of Fallot

PATHOLOGIC ANATOMY: BASIS FOR PULMONARY STENOSIS (*continued*)

In most examples of the tetralogy of Fallot the pulmonary valve is bicuspid. In many of these the valve contributes an element of pulmonary stenosis. In some instances this contribution is derived from the fact that the leaflets are short and adherent at the commissures, thus creating a more narrow orifice than would be the case with normal leaflets. Another feature related to the bicuspid pulmonary valve is that the orifice of the pulmonary trunk is usually intrinsically narrow and may inherently contribute to the pulmonary stenosis.

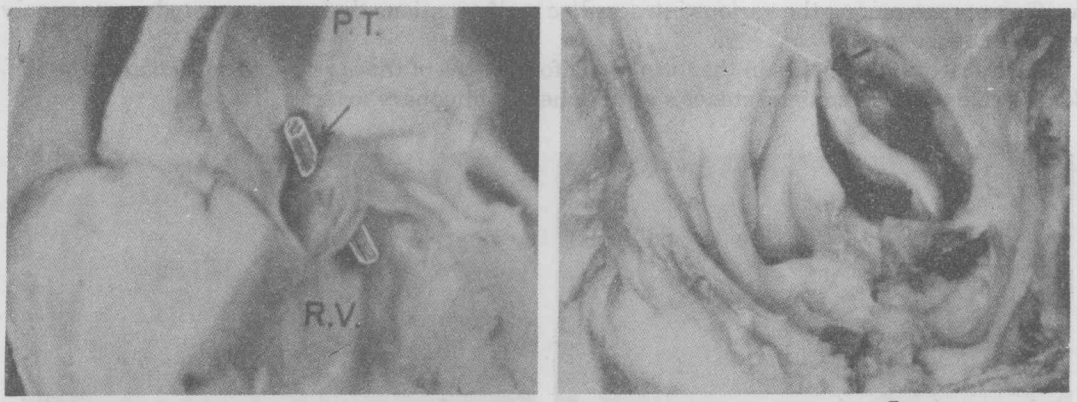


FIG. 494. *Left.* Right ventricular infundibulum (R.V.) and pulmonary trunk (P.T.) in the tetralogy of Fallot. The pulmonary valve is deformed in a dome-shaped manner, yielding a narrow orifice (containing probe). *Right.* The pulmonary valve from above in the tetralogy of Fallot. Superficial resemblance to the bicuspid valve, although there is but one commissure (point of arrow). This is a unicommissural type of pulmonary valve comparable to the deformity seen in some instances of congenital aortic stenosis. The valve is essentially a modified dome-shaped structure.

FIG. 495. The right ventricular infundibulum, pulmonary trunk (P.) and left (L.) and right (R.) pulmonary arteries. The pulmonary valvular orifice is narrow, as are the pulmonary trunk and its major branches. The latter two features contribute to pulmonary stenosis in this instance. A. = Exterior of ascending aorta.

Tetralogy of Fallot

Pathologic Anatomy: Associated Anomalies

In the patient with the tetralogy of Fallot anomalies or variants other than those forming part of the complex may be encountered. The most common is a patent foramen ovale of the valvular competent variety, a condition that is not truly a malformation. True atrial septal defect, however, may exist in association with the tetralogy of Fallot. Rarely is there a muscular ventricular septal defect in addition to the usual ventricular septal defect of the tetralogy. Partial anomalous venous connection, hypoplasia or absence of the pulmonary valve, persistent common atrioventricular canal, partial anomalous pulmonary venous connection and tricuspid insufficiency are among the various intracardiac malformations that are associated uncommonly with the tetralogy.

Extracardiac anomalies in the form of malformations of the aortic arch system are common. Considerably less common is stenosis of peripheral pulmonary arteries.

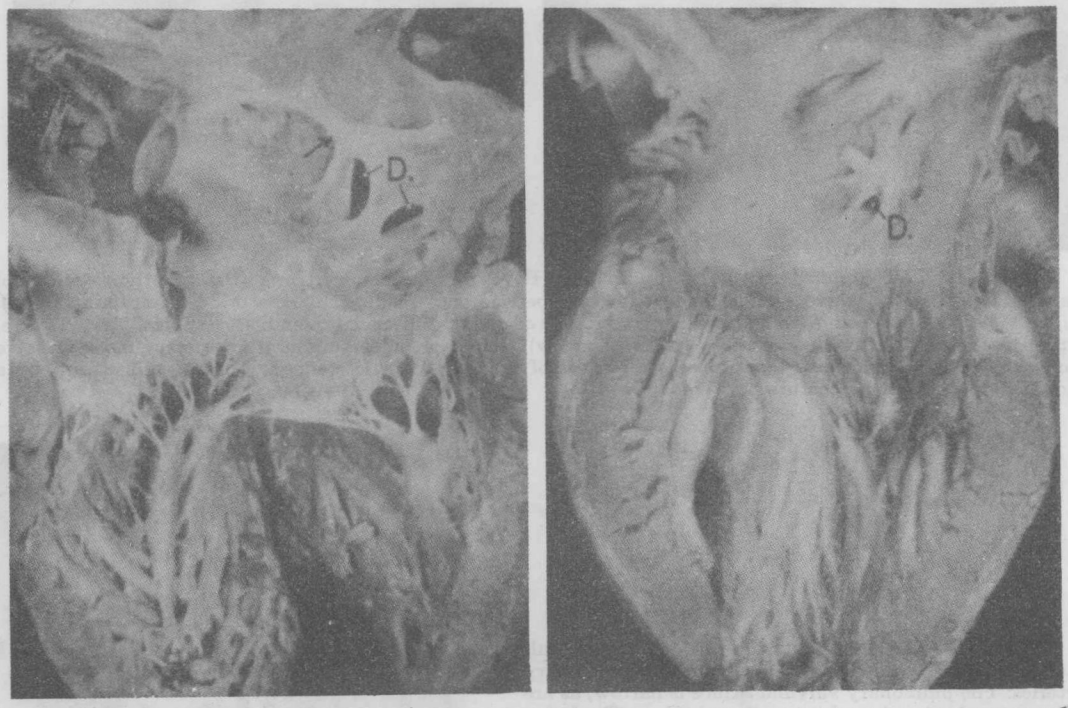


Fig. 496. Two examples of valvular competent patent foramen ovale and atrial septal defect in the tetralogy of Fallot. *Left.* From a 13-year-old boy. Left atrium and left ventricle. In addition to a valvular competent patent foramen ovale (arrow) there are two defects (D.) in the floor of the fossa ovalis. *Right.* Left atrium and left ventricle in a male infant 8 months of age. The probe lies in a valvular competent patent foramen ovale. Below this is a small true atrial septal defect (D.) at the fossa ovalis. In the tetralogy of Fallot with an interatrial communication some of the right-to-left shunt characteristic of this condition may occur at the atrial level.

PATHOLOGIC ANATOMY: ASSOCIATED ANOMALIES (*continued*)

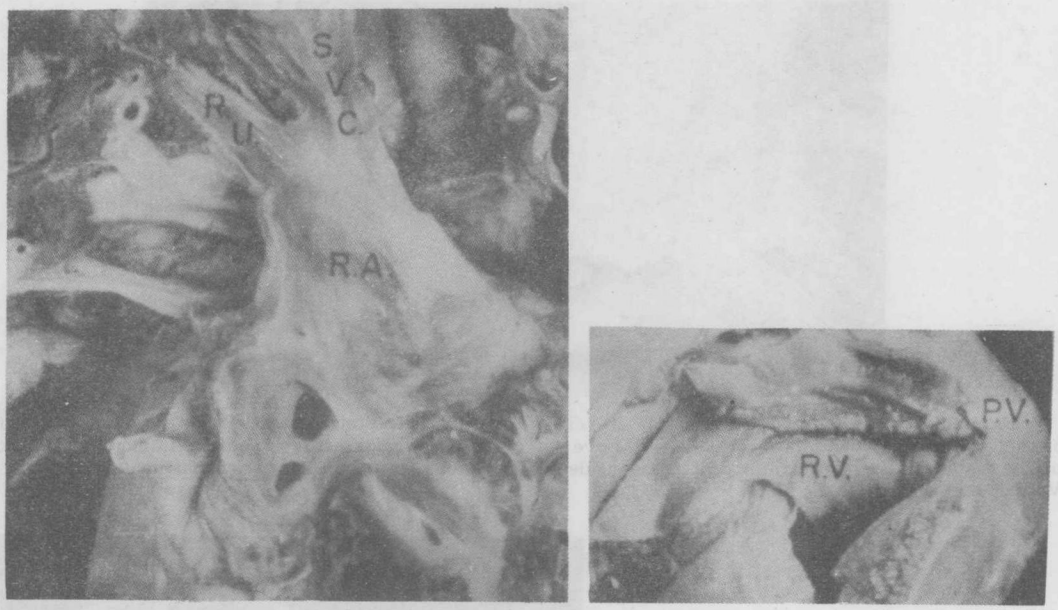


FIG. 497. *Left.* Right atrium in the 13-year-old boy from whom the left side of the heart is illustrated in Figure 496, *Left*, page 408. In addition to the two defects at the fossa ovalis, there is partial anomalous pulmonary venous connection in that the right upper pulmonary vein (R.U.) communicates with the right atrium (R.A.) at the level of the entrance of the superior vena cava (S.V.C.). *Right.* Tetralogy of Fallot with hypoplasia of pulmonary valve in an 8-week-old girl. Illustrated are the upper portion of the right ventricular infundibulum (R.V.), the pulmonary valve (P.V.) and the beginning of the pulmonary trunk. The pulmonary valve is represented by a rudimentary rim of fibrous tissue. The subject of congenital pulmonary insufficiency in association with the tetralogy of Fallot was presented in Volume I, pages 145 to 148.

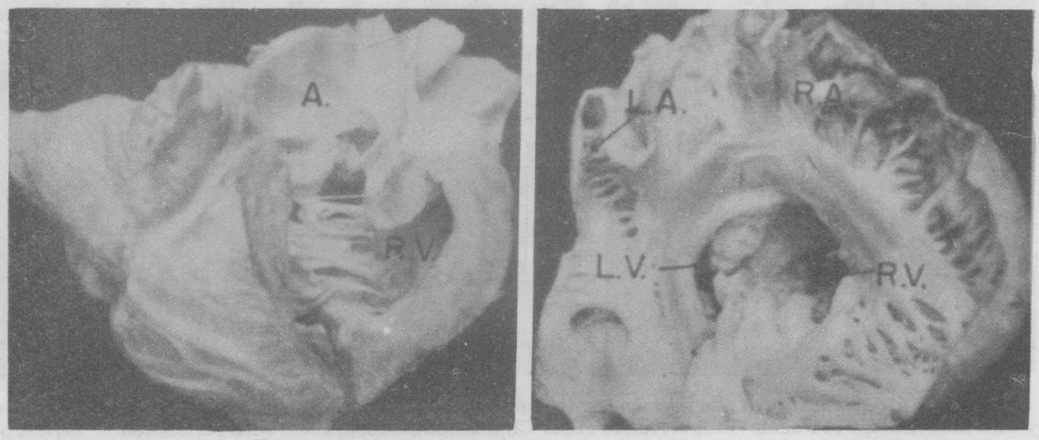


FIG. 498. Tetralogy of Fallot and persistent common atrioventricular canal in an 8-month-old male infant. *Left.* Right ventricle (R.V.) and ascending aorta (A.). Features of the tetralogy of Fallot, the biventricular origin of the aorta and a stenotic infundibular chamber, are displayed. *Right.* The atrial septum has been divided and retracted to reveal the common atrioventricular valve from above. To the right, the valve leads into the right ventricle (R.V.), and to the left, into the left ventricle (L.V.). R.A. = Right atrial appendage. L.A. = Left atrial appendage.