Tetralogy of Fallot: An Angiographic–Pathologic Correlative Study

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SUMMARY The anatomic abnormalities observed by cineangiographic axial techniques of 12 patients with tetralogy of Fallot were correlated with anatomic details noted at necropsy. Right ventricular angiograms made in the right anterior oblique view best demonstrated the severity and type of infundibular obstruction and also permitted differentiation of the perimembranous, infundibular muscular and subarterial types of ventricular septal defects. The degree of aortic overriding was best displayed in the long-axis view. Comparison of the intracardiac anatomy of each postmortem specimen with the respective premortem cineangiogram has provided further clarification of the angiographic anatomy displayed by these axial techniques.

VENTRICULAR SEPTAL DEFECT (VSD), dextroposition of the aorta, infundibular stenosis and hypertrophy of the right ventricle were clinically described by Fallot

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in 1888. These cardiac abnormalities form a well-defined entity. Their anatomic characteristics were clarified by Van Rokitansky and further examined by others.2-4 Many angiographic studies of the tetralogy have been published since angiography was developed, and the majority focus on details obtained using standard frontal and lateral projections.5-8 The use of axial projections displays additional anatomic details not seen previously.10

In this report we show the angiographic anatomy of a group of patients with tetralogy of Fallot studied at the University of Alabama Hospitals using axial views and correlate this with the anatomic features seen at necropsy.

Material and Methods

Twelve postmortem specimens of patients with classic tetralogy of Fallot were studied. Each patient had undergone cardiac catheterization and axial angiographic studies a few weeks before death. Each had tetralogy of Fallot with concordant atrioventricular connections, a large VSD in the left ventricular outflow septal area, biventricular origin of the aorta, origin of the pulmonary artery from the right ventricle, infundibular pulmonary stenosis and right ventricular hypertrophy.11

The specimens were selected on the basis of availability of angiographic and anatomic correlation. They had been fixed in formalin and opened in the usual fashion. The right ventricular incision was always extended into the pulmonary artery and the aorta opened from the left ventricle. This allowed clear definition of the VSD and its relation with the arterial and atroventricular valves from either ventricle. The surgically placed patches were removed in all cases. The amount of infundibular and anterior right muscle wall removed by the surgeons varied from case to case according to the need at operation.

The catheterization had been performed under general anesthesia and angiograms made after obtaining hemodynamic and saturation data. Biplane 35-mm cineangiograms were made with injection of Renografin-76 through a large NIH catheter with side holes. Hand injection was used routinely, although power injection was used in larger patients or when smaller catheters were necessary.

Angiograms were made simultaneously in the long-axis and elongated right anterior oblique views after injection into the right ventricle. Left ventricul-
grams were selectively made when further definition of this chamber was desired. If a defect in the posterior ventricular septum was suspected, left ventriculograms in the four-chamber view were also obtained. The long-axis view is defined as a 60° left anterior oblique view combined with 30° craniocaudal angulation. The elongated right oblique view is a 30° right anterior oblique projection with 30° craniocaudal angulation. The four-chamber view is a 45° left anterior oblique projection with 30° craniocaudal angulation. The main pulmonary artery and its bifurcation were defined in each patient using the sitting-up projection with injection of contrast medium into the right ventricular outflow tract or main pulmonary artery. In patients who had undergone previous intrapericardial surgery and in small subjects, coronary artery anatomy was also delineated.

This study was performed by comparing the anatomy observed in the autopsy specimen with that shown by the premortem angiogram. Each component of the malformation was evaluated to correlate the angiographic and pathologic details of the infundibular stenosis, VSD, and the position of the aorta relative to the ventricular septum.

**Results**

**Infundibular Stenosis**

The infundibular septum was abnormally positioned anteriorly and leftward in each case and was the primary cause of the right ventricular outflow tract stenosis. The abnormally positioned infundibular septum was the only cause of stenosis in two cases. In these, the outflow tract was a uniformly narrow channel (fig. 1). Angiographically, this elongated channel, extending from the trabecular portion of the right ventricle to the pulmonary valve, is best seen in the elongated right anterior oblique view (fig. 2).

The right ventricular outflow tract stenosis was accentuated by hypertrophy of the anterior free wall in three specimens. This severe hypertrophy could not be fully appreciated in the anatomic specimens because of previous surgical resection. The angiographic representation of this stenosis appears as a filling defect on the left border of the outflow tract in the elongated right anterior oblique view (fig. 3). Hypertrophy of the trabecula septomarginalis (TSM) contributed significantly to right ventricular outflow tract

**Figure 1.** A right ventricular outflow tract with anterior displacement of the infundibular septum (IS). The outflow tract of the right ventricle has been exposed by incising the anterior wall (W). The posterior aspect of this channel is formed by the infundibular septum, which is related to the ventricular septal defect (VSD) posteriorly and to the trabecular septum (TS) inferiorly. The superior border is formed by the pulmonic valve (PV). The outflow tract has a smooth appearance due to fibrosis of the endocardium. PA = pulmonary artery.

**Figure 2.** Right ventriculogram in the elongated right anterior oblique view showing the type of outflow tract stenosis seen in figure 1. The right ventricular outflow tract is formed by a uniform channel formed by the anterior wall of the right ventricle (AW) and the infundibular septum (IS). The upper limit of this channel is the pulmonary valve (arrowhead). RV = right ventricle; PA = pulmonary artery; AO = aorta; TV = tricuspid valve.
stenosis in three cases. The body of the TSM was the most prominent cause of the inferior component of the infundibular stenosis (fig. 4). This is angiographically demonstrated by the elongated right anterior oblique view of the right ventriculogram as a prominent inferior filling defect (fig. 5). In three additional cases, a lesser degree of hypertrophy of the TSM was accompanied by hypertrophy of the infundibular septum and/or anterior wall.

The infundibular septum was very small in one case in which the main cause of pulmonary stenosis was a small pulmonary valve annulus and hypertrophy of the TSM (fig. 6). A diagram of each type of infundibular pulmonic stenosis found in this group of cases is shown in figure 7 and the causative factors are listed in table 1.

### Ventricular Septal Defect

Three types of VSDs were identified, using the classifications listed by Soto et al. This classification system is very simple and easily applied anatomically because it is based on the relations of the defect to the

<table>
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<th>TABLE 1. Right Ventricular Outflow Tract Stenosis</th>
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<td>Cause</td>
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<td>Anteriorly displaced infundibular septum</td>
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<td>Additional factors:</td>
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<td>Hypertrophy</td>
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<td>Anterior infundibular wall</td>
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<td>Trabecula septomarginalis</td>
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classic structures of the ventricular septum and is also easily defined by angiographic studies.

The three types are:

**Perimembranous with extension into the infundibulum.** This is the classic and most common location of the VSD in tetralogy of Fallot and was present in 10 of these 12 hearts. Viewed from the right ventricle, the defect lies anterior and superior to the tricuspid valve. The posterosuperior border of the defect is formed by the fibrous continuity between the aortic and tricuspid valves (fig. 8A). The TSM buttressed posteriorly and leftward by the trabecular septum, the infundibular septum and the ventriculo-infundibular fold form the other boundaries. Viewed from the left ventricle, the VSD was located beneath the right and noncoronary aortic valve cusps. The defect was contiguous with the anterior mitral valve leaflet (fig. 8B). Angiographically, this type of VSD is best demonstrated in the elongated right anterior oblique view of the right ventriculogram. The defect itself appears as a channel that connects the right ventricle and the aorta during systole (fig. 9A). In diastole, the aortic valve (right and noncoronary cusps) forms the ceiling of the defect. The negative shadow beneath the aortic valve is produced by non-opacified blood from the left ventricle passing into the right ventricular chamber and indicates the size of the defect (fig. 9B). Continuity between the VSD and the tricuspid annulus can be demonstrated during both systole and diastole.

**Infundibular muscular defect.** Classically, this is called an “intracristal" VSD and is much less common than the perimembranous defect. This was present in only one of our 12 cases. The defect is located in

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**Figure 6.** Right ventricular angiogram in the elongated right anterior oblique view. The stenosis is mainly produced by the hypertrophied trabecula septomarginalis (TSM) and by hypertrophy of the anterior infundibular wall (W). The infundibular septum (IS) contributes to the stenosis. AO = aorta; PA = pulmonary artery; RV = right ventricle.

**Figure 7.** Types of right ventricular outflow tract stenosis observed in the elongated right anterior oblique view of the right ventricular angiogram. (A) Stenosis predominantly due to an anteriorly displaced infundibular septum. (B) Hypertrophy of the trabecula septomarginalis. (C) Hypertrophy of the anterior wall of the infundibulum. (D and E) Mixed type of stenosis produced by hypertrophy of the anterior wall, hypertrophy of trabecula septomarginalis and hypertrophy of infundibular septum. (F) Stenosis produced by a small pulmonary annulus.
Figure 8. Anatomy of the perimembranous ventricular septal defect (VSD). (A) Right ventricular view. A portion (arrowheads) of the tricuspid annulus is in continuity with the defect. The borders of the defect are the trabecula septomarginalis (TSM) anteriorly, infundibular septum (IS) superiorly and ventriculoinfundibular fold (VIF) posteriorly. The entire noncoronary cusp of the aortic valve (NCC) and part of the right coronary cusp are directly related to the defect. PV = pulmonary valve; ATL = anterior tricuspid leaflet, STL = septal tricuspid leaflet; TS = trabecular septum. (B) Left ventricular view. The ventricular septal defect is beneath the noncoronary aortic cusp (NCC), extending anteriorly beneath the right coronary aortic cusp (RCC). The inferior contour is in continuity between fibrous elements of mitral, aortic and tricuspid valves (arrowheads).

the body of the septum and separates the right and left ventricular outflow tracts and is surrounded entirely by muscle. It is anterior and to the left of the membranous septum. Viewed from the right ventricle, its superior rim is formed by the abnormally positioned infundibular septum and its right border by the ventriculoinfundibular fold. Inferiorly and anteriorly it is separated from the tricuspid annulus by a muscle bar.
that is the posterior arm of the TSM (fig. 10A). Viewed from the left ventricle, the defect is located immediately beneath the noncoronary cusp and may extend anteriorly to the commissure between the noncoronary and right coronary cusps. It is separated from the mitral valve by a segment of muscle (fig. 10B). The angiographic representation of this kind of defect is similar to that of the perimembranous type except that it is separated from the tricuspid valve by a filling defect caused by the posterior arm of the TSM (figs. 11A and B).

**Subarterial ventricular septal defect.** This type of defect was present in one specimen. The TSM anteriorly and inferiorly and the tricuspidinfundibular fold posteriorly form the borders of the VSD as seen through the right ventricle. The semilunar valves form the ceiling of the defect. The infundibular septum in this heart was very small and was mostly replaced by a bridge of fibrous tissue that separated the defect from the actual pulmonary valve annulus (fig. 12A). The pulmonary valve ring and main pulmonary artery also were hypoplastic. Viewed from the left ventricle, the defect is beneath the right coronary cusp extending to its leftward commissure. The hypertrophied TSM formed the posterior border and separated the defect from the mitral valve (fig. 12B). The elongated right anterior oblique view of the right ventriculogram also provides the best demonstration of this type of VSD. The negative shadow produced by the infundibular septum is almost absent (fig. 13).

Each type of VSD is shown diagrammatically in figure 14.

**Overriding of the Aorta**

The degree of aortic dextroposition was assessed by considering the amount of aortic valve tissue present within the right ventricle. Severe overriding was present when the right and noncoronary cusps formed the

Figure 9. Right ventriculogram in elongated right anterior oblique view showing the perimembranous type of ventricular septal defect. (A) Systolic frame. The ventricular septal defect (VSD) is behind the infundibular septum (IS). The posterior border of the VSD is the junction of the tricuspid and aortic annuli (large arrow head). The noncoronary cusp (NCC) is directly above this junction. The contrast medium passes from the right ventricle (RV) into the aorta (AO). (B) Diastolic frame. The VSD appears as a negative shadow located beneath the aortic valve (arrow heads) and posterior to the infundibular septum (IS). Notice the continuity of the defect and the tricuspid valve annulus (TV). RV = right ventricle, RA = right atrium, PA = pulmonary artery.

Figure 10. Anatomy of the infundibular muscular ventricular septal defect (VSD). (A) Right ventricular view. The defect is surrounded entirely by muscle. The inferior and anterior borders are formed by the posterior and superior arms (PA and SA) of the trabecula septomarginalis (TSM). The posterior arm is interposed between VSD and tricuspid valve annulus (TV). The left border is formed by the tricusuloinfundibular fold (VIF), the superior border by the anteriorly placed and partially resected infundibular septum (IS). The white arrow shows the RV outflow tract. ATL = anterior tricuspid leaflet; STL = septal tricuspid leaflet. (B) Left ventricular view. The VSD is located beneath the noncoronary aortic cusp (NCC). The inferior and anterior borders are formed by the trabecular septum (TS). The mitral valve (MV) is separated from the VSD by the posterior arm of the trabecula septomarginalis (arrow head). RCC = right coronary cusp. LCC = left coronary cusp.
FIGURE 11. Ventricular septal defect (VSD) of infundibular muscular type. Right ventricular angiogram in the elongated right anterior oblique view. (A) Diastolic frame. The right ventricle (RV) is in continuity with the aorta (AO) through the VSD. Notice the discontinuity between the defect and the tricuspid valve (TV) by a filling defect (arrow), which represents the posterior arm of the trabecula septomarginalis seen anatomically in figure 10A. The infundibular septum (IS) is partially obscured by the left ventricular outflow tract. (B) Systolic frame. The VSD is much better visualized and lies behind the infundibular septum. Again, the discontinuity (arrow) between the defect and the tricuspid valve annulus appears. RV = right ventricle; AO = aorta; PA = pulmonary artery.

FIGURE 12. Subarterial ventricular septal defect (VSD) anatomy. (A) Right ventricular view. The defect is located immediately beneath the arterial valves. The boundaries of the defect are the posterior and superior arms (PA and SA) of the trabecula septomarginalis (TSM), the ventriculo-infundibular fold (VIF) and the ceiling is formed by the pulmonic valve (PV). The aortic valve is not shown. The abnormally located infundibular septum (IS) is very short and it contributes little to the stenosis. The defect is separated from the tricuspid valve annulus (TV) by the posterior arm of the trabecula septomarginalis. (B) Left ventricular view. The VSD is bound by the trabecular septum (TS) inferiorly, the posterior arm of the trabecula septomarginalis posteriorly, and the right coronary aortic cusp (RCC) above. The ceiling of the defect is formed by arterial valves (aorta on left and pulmonary on right). The mitral valve (MV) is separated from the defect by the posterior arm of the trabecula septomarginalis. NCC = noncoronary cusp; LCC = left coronary cusp (arrow heads).
ceiling of the VSD as seen from the right ventricular chamber. The degree of overriding was related to the magnitude of malalignment of the plane of the infundibular septum with respect to the plane of the trabecular septum (fig. 15) and also to the degree of elongation of the left ventricular outflow tract. In these 12 specimens, one or two aortic valve cusps were over the right ventricle. The angiographic demonstration of aortic overriding is best obtained from the long-axis view of the right ventriculogram (fig. 16). In this projection, the anterior muscular interventricular septum is seen in profile and the aortic valve is best identified in diastole. The plane of the ventricular septum may be extrapolated superiority to determine the degree of overriding.

Discussion

Angiograms obtained using axial projections in tetralogy of Fallot provide greater anatomic definition than those obtained from standard views. The morphology of the right ventricular outflow tract obstruction, the type of VSD and the degree of aortic dextroposition are well demonstrated. The pulmonary
ing the right and left ventricular outflow tracts, and is completely surrounded by muscle. Angiographically, its separation from the tricuspid valve annulus by the posterior arm of the TSM can usually be demonstrated. Certainly, if the muscular separation were very small it would not be angiographically detected and the defect would be inaccurately described as perimembranous. The subarterial defect is in direct continuity with the annulus of the aortic and pulmonary valves and is away from the tricuspid valve annulus. It can usually be distinguished from the infundibular muscular defect because muscular tissue separates the latter from the semilunar valves.

The degree of aortic dextroposition is best shown using the long-axis view. The plane of the ventricular septum can be extrapolated superiorly to provide an estimation of the magnitude of aortic override.

In conclusion, we believe the long-axis and elongated right anterior oblique views of the right ventriculogram, compared with standard projections, significantly improve the angiographic definition of the intracardiac anatomy of patients with tetralogy of Fallot. This study has provided anatomic correlation of the details observed angiographically using these axial views.

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Figure 16. Right ventricular angiogram in long-axis view showing the degree of aortic overriding. The degree of aortic valve dextroposition is estimated by extrapolation of the septal axis. All of the right coronary cusp (RC) lies to the right. The ventricular septal defect is indicated by the arrowheads. RV = right ventricle; LV =left ventricle; AO = aorta; LC = left coronary cusp.

valve annulus and the main pulmonary artery and its bifurcation and branch vessels are also accurately defined, as previously reported. In this study of 12 patients, right ventricular infundibular stenosis was always produced by anterior displacement of the infundibular septum. The stenosis was further magnified by hypertrophy of the anterior infundibular wall, hypertrophy of the TSM or combinations of each. The anatomical observations were similar to those described by others. Angiographic demonstration of the infundibular portion of the right ventricle is best obtained using the elongated right anterior oblique view of the right ventriculogram. This projection frees the outflow tract from the superimposition of the opacified aorta noted on the posteroanterior projection and from the shoulder of the right ventricle found on the lateral projection. The three types of VSD previously described as perimembranous, infundibular muscular and subarterial were observed. They were best demonstrated using the elongated right anterior oblique view of the right ventriculogram. The perimembranous defect is more easily defined angiographically because of its relation to the tricuspid valve annulus, which is in direct continuity with the aortic valve. The infundibular muscular defect resides in the septum, separat-
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