Tetralogy of Fallot with Absent Pulmonary Valve
Natural History and Hemodynamic Considerations

By Jeffrey B. Lakier, M.D., Paul Stanger, M.D., Michael A. Heymann, M.D.,
Julien I. E. Hoffman, M.D., and Abraham M. Rudolph, M.D.

SUMMARY
Absent or hypoplastic pulmonary valve is an uncommon condition usually associated with a ventricular septal defect and an obstructive pulmonary valve ring. Secondary abnormalities involving the pulmonary arteries and major bronchi are common, and many result in severe pulmonary complications. Eight patients with absent pulmonary valve, ventricular septal defect and obstructed pulmonary valve ring are herein described. The clinical features, cardiac catheterization, cineangiographic and postmortem findings are presented. Possible fetal hemodynamics and their influences are discussed, as are postnatal hemodynamics and their clinical consequences. Patients with this combination of lesions have been regarded as having a very poor prognosis, and recent publications have advocated early surgical intervention; however, most published series are small, and only 106 cases have been documented. These cases have been reviewed for clinical and anatomic features as well as an overview of prognosis and results of surgical intervention.

Additional Indexing Words:
Ventricular septal defect
Bronchial compression
Pulmonic stenosis
Pulmonary regurgitation
Pulmonary artery aneurysms

A BSENT or hypoplastic pulmonary valve is an uncommon condition usually associated with a ventricular septal defect, an obstructive pulmonary valve ring and massively dilated pulmonary arteries. The latter often result in severe pulmonary problems and are largely responsible for the poor prognosis. Various surgical procedures have been suggested; however, in general, mortality has been high. All reported series are small so that evaluation of treatment has been difficult. In total, only 106 cases have been reported.1-22

The purpose of this communication is threefold: 1) to describe the clinical, hemodynamic, and angiographic findings and natural history of eight cases of absent pulmonary valve, ventricular septal defect and obstructive pulmonary valve ring; 2) to postulate the fetal hemodynamics and their consequences; and 3) to review all reported cases with particular reference to the anatomic details, varied clinical course and results of therapy. Recommendations for management based on our experience, and a review of the literature are included.

Material and Methods
Between June 1966 and June 1973 eight patients with absent pulmonary valve, ventricular septal defect and right ventricular outflow obstruction were diagnosed and their conditions verified at cardiac catheterization. At the time of initial catheterization their ages ranged from 1 day to 1½ years (table 1). Five patients underwent a second catheterization at 2, 20, 29, 30 and 68 months respectively after the initial study. The physical findings, roentgenograms, electrocardiograms, cardiac catheterization data and cineangiograms were analyzed and compared with those of 106 patients reported in the literature. Two of our patients died and were studied at autopsy.

Results
Physical findings were similar in all eight cases and were essentially the same as those reported previously.11, 14, 15, 22, 29 Cyanosis and a cardiac murmur were noted at birth or shortly after birth. Cyanosis became less apparent during the first week of the newborn period in 7 of the 8 patients but persisted throughout a stormy hospital course in the eighth.

A prominent left parasternal impulse suggestive of right ventricular hypertrophy was present in each.
The first heart sound was of normal or slightly increased intensity. There was a grade 3-5/6 harsh, basal systolic ejection murmur which radiated down the left parasternal border. The second heart sound was single, and the audible component was accentuated. A prominent, grade 2-3/4 low frequency decrescendo early diastolic murmur was present in the second-third interspace at the left sternal edge. The combination of a systolic ejection murmur and a rough early diastolic murmur resulted in a characteristic to-and-fro quality. Apical mid-diastolic flow rumbles were absent. Three of the patients had congestive cardiac failure as evidenced by diaphoresis and marked hepatomegaly. Pulmonary complications were present in five of the patients, and in each respiratory infection and/or atelectasis involved predominantly the right middle and right upper lobes. Hyperinflation with air trapping and wheezing was noted in one patient.

The electrocardiographic features were consistent. The mean frontal plane QRS axis was between +80° and +160° and all patients had evidence of isolated right ventricular hypertrophy. In one instance there was evidence suggestive of right atrial enlargement.

Chest roentgenograms were very distinctive. The postero-anterior view demonstrated a moderately enlarged heart with massively dilated right and main pulmonary arteries in seven cases and a massively dilated left pulmonary artery in the eighth. The proximal pulmonary arteries were aneurysmal; however, immediately distal to the aneurysmal portion the peripheral vascular markings were either normal or only slightly increased. In one instance (case 2) right-sided vascular markings were distinctly more prominent than the left. All eight cases had a prominent bulge at the upper left cardiac border caused by the dilated right ventricular outflow tract. This was even more apparent in the right oblique view.

The data obtained at cardiac catheterization are listed in table 1. Five of the eight patients underwent cardiac catheterization on more than one occasion. Right ventricular systolic pressure was at systemic levels in all at the initial procedure. The pulmonary artery was entered in six patients, and in these a 46-87 mm Hg systolic pressure drop was recorded at the pulmonary valve level with normal pulmonary arterial pressures in each.

The characteristic pulmonary arterial pressure tracing found in isolated pulmonic regurgitation with intact septum and no stenosis at the level of the pulmonary outflow was not present in any of our cases. The pulmonary arterial diastolic pressure was low but had an arterial configuration and did not approach right ventricular diastolic pressure. Stenosis of the right ventricular outflow tract was presumably severe enough to prevent free reflux during diastole.

### Table 1

<table>
<thead>
<tr>
<th>Case number</th>
<th>Sex</th>
<th>Age at catheterization</th>
<th>LV (mm Hg)</th>
<th>RV (mm Hg)</th>
<th>PA (mm Hg)</th>
<th>RV-PA ∆P</th>
<th>Systemic saturation</th>
<th>Qp/Qs</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>3½ mos</td>
<td>97/55</td>
<td>22/12</td>
<td>70</td>
<td>93</td>
<td>2:1</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>7 yrs*</td>
<td>100/5</td>
<td>24/8</td>
<td>46</td>
<td>93</td>
<td>2:1</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>19 days</td>
<td>75/55</td>
<td>22/14</td>
<td>53</td>
<td>87</td>
<td>1.3:1</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>2 5/12 yrs</td>
<td>100/5</td>
<td>20/10</td>
<td>80</td>
<td>91</td>
<td>1.4:1</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>1½ yrs</td>
<td>110/12</td>
<td>105/8</td>
<td>96</td>
<td></td>
<td>1.7:1</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>3 yrs</td>
<td>105/11</td>
<td>22/12</td>
<td>83</td>
<td>97</td>
<td>1.6:1</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>5 mos</td>
<td>110/15</td>
<td>30/20</td>
<td>80</td>
<td>84</td>
<td>2.1:1</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>7 mos</td>
<td>100/12</td>
<td>40/14</td>
<td>62</td>
<td>93</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>1 day</td>
<td>60/38</td>
<td>60/4</td>
<td></td>
<td>74†</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>2½ yrs</td>
<td>90/10</td>
<td>24/10</td>
<td>66</td>
<td>96</td>
<td>1.3:1</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>6 wks</td>
<td>122/62</td>
<td>35/22</td>
<td>87</td>
<td>93</td>
<td>1.8:1</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>F</td>
<td>4 days</td>
<td>85/8</td>
<td>88/4</td>
<td></td>
<td>89</td>
<td>1:1</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>3 days</td>
<td>65/8</td>
<td>65/8</td>
<td></td>
<td>75</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Abbreviations: LV = left ventricle; RV = right ventricle; PA = pulmonary artery; Qp = pulmonary flow; RV-PA ∆P = systolic pressure difference; Qs = systemic flow.

*Prolapse of the aorta leaflet into ventricular septal defect.
†Ascending aorta.
‡Femoral artery.
¶Left lower pulmonary vein saturation 87.
Systemic arterial desaturation (74%–93%) was present in all seven patients catheterized as infants; however, a dominant left to right ventricular shunt was present in the five of these in whom sufficient data were available for the calculations.

Right ventricular cineangiograms demonstrated several interesting features concerning the level of stenosis, orientation of the infundibulum, and the dilatation of the pulmonary arteries. The outflow tract stenosis was at the level of the pulmonary valve annulus where a discrete ridge of tissue was present. Valve leaflets could not be identified. There was no stenosis in the infundibulum which, in fact, was dilated. The infundibulum was abnormally oriented in six of eight patients. Normally the infundibulum is fairly short and vertical in the newborn (fig. 1A) whereas in these six infants the infundibulum was long and horizontal. The entire infundibulum was directed from left to right with the distal portion also being directed posteriorly (fig. 1B). In the seventh patient the orientation and length of the infundibulum was normal, while in the remaining patient the infundibulum was oriented slightly toward the left (fig. 1C). In six patients the right pulmonary artery was aneurysmal, and in one of these the left pulmonary artery was also interrupted. In one patient both right and left pulmonary arteries were massively dilated while in another the left pulmonary artery was aneurysmal. A striking association was found between the lateral orientation of the infundibulum and the pulmonary artery which was most dilated. Those patients with displacement of the right ventricle towards the left and a long infundibulum that was oriented from left to right had an aneurysmal right pulmonary artery. Of the two patients with vertical and leftward orientation, one had dilatation of both right and left pulmonary arteries and the other of the left pulmonary artery.

Pulmonary arteriograms were performed in the six patients in whom the pulmonary artery was entered. They all demonstrated dilatation of the proximal portion of the pulmonary arteries and moderate pulmonary insufficiency which are the hallmarks of this condition. The distal pulmonary arteries were normal.

The massively dilated right pulmonary artery caused depression of the roof of the left atrium in two cases and displaced the pulmonary veins anteriorly in a third. In the two patients in whom the pulmonary artery could not be entered, slow clearing of contrast material from the right ventricle suggested pulmonary insufficiency.

The aortic arch was normally situated in five patients and was right sided in three. In all the ascending aorta was dilated and the aortic isthmus was of normal diameter. In the patient with interruption of the left pulmonary artery a small left-to-right ductal shunt was present at the initial study; however, there was no evidence of a patent ductus arteriosus when restudied at age 2½ years, and the left lung was supplied entirely by well developed bronchial collateral vessels.

Follow-up information is available on seven of the eight patients. Two have been subjected to operation because of repeated pulmonary infections and right sided heart failure. Case 5 underwent a right middle lobectomy because of marked emphysematous changes in that lobe, the result of compression of the right main bronchus by the dilated right main pulmonary artery. He has progressed well since then, is no longer in cardiac failure nor prone to recurrent pulmonary infections. Case 4 underwent surgical closure of the ventricular septal defect and a small atrial septal defect with an excellent symptomatic result. The oldest patient (case 1), now aged 7 years, has progressed well and is acyanotic at rest. Pulmonary infections have become less of a problem with increasing age and at present are confined to an occasional episode of bronchitis. When restudied a 20 mm Hg systolic pressure difference was noted across

**Figure 1**

Right ventricular cineangiograms demonstrating the association of infundibular orientation and pulmonary arterial dilatation in patients with tetralogy and absent pulmonary valve. A. Normal. B. The infundibulum is oriented toward the right, and there is aneurysmal dilatation of the right pulmonary artery. C. The infundibulum is oriented slightly toward the left, and there is preferential dilatation of the left pulmonary artery. RPA = right pulmonary artery; LPA = left pulmonary artery; Inf = infundibulum; RV = right ventricle.

*Circulation, Volume 50, July 1974*
the ventricular septal defect. Cineangiography demonstrated partial prolapse of the right cusp of the aortic valve resulting in partial closure of the ventricular septal defect and mild aortic regurgitation. The other three patients on whom information is available have followed a similar course, pulmonary complications decreasing with increasing age, particularly after the first year.

Two patients died, one (case 7) at the age of five months after chronic and recurrent pulmonary problems, and the other (case 8) at age one month after prolonged and difficult hospitalization after birth. The latter patient was treated with digitalis and diuretics and was maintained in supplemental oxygen with little improvement in clinical status. Marked tachypnea, cyanosis and tachycardia during feedings necessitated a gastrostomy. Persistent atelectasis, thought to be the result of bronchial compression by the aneurysmal right pulmonary artery prompted plication of the right pulmonary artery with an estimated narrowing of that artery to 60–70% of its original diameter. Postoperatively the patient required endotracheal intubation and positive pressure respiration to maintain adequate blood gases. On the tenth postoperative day a chest roentgenogram demonstrated atelectasis of the entire left lung with herniation of the right lung across the midline. Plication of the dilated left main pulmonary artery produced little significant improvement. She was never able to tolerate extubation, deteriorated gradually and died on the sixteenth postoperative day.

Both patients were studied at necropsy. In case 8 both right atrial enlargement and marked right ventricular hypertrophy were found. There was no infundibular obstruction, but a moderate constriction at the pulmonary valve level was evident. The pulmonary valve leaflets were markedly hypoplastic and consisted only of tiny sessile nubbins of tissue. Postvalvar pulmonary artery dilatation was present with an aneurysmally dilated right pulmonary artery and slightly small left pulmonary artery. Evidence of plication of both right and left pulmonary arteries was present. The dilated pulmonary arteries compressed the lower trachea and the right and left mainstem bronchi in an antero-posterior direction (fig. 2). There was a 0.6 cm diameter membranous ventricular septal defect. The aortic valve was noted to override both ventricles and all branches of the coronary arteries originated from a single ostium. The right lower lobe had a pinkish-yellow color and appeared to be the only portion of the lung which was adequately aerated.

In the other patient (case 7) the findings were identical except for minimal infundibular obstruction and pulmonary arteries in their natural position. The lower trachea and right mainstem bronchus were flattened.

Discussion

The earliest report of the association of absent pulmonary valve and ventricular septal defect was by Royer and Wilson in 1908. Their case was studied at autopsy and was unique in that the patient also had corrected transposition of the great arteries in situs inversus (d-transposition in situs inversus). In 1927 Kurtz, Sprague and White described the physical findings and clinical course of a patient who at necropsy was found to have a ventricular septal defect, stenosis at the pulmonary valve ring and an absent pulmonary valve. Since then 104 cases of hypoplastic pulmonary valve with a ventricular septal defect have been documented. All but three of these had pulmonic stenosis. The clinical features have been described in 80 of these cases, whereas the remaining 38 were included in surgical or autopsy series and detailed clinical findings were lacking.

Eight cases of absent pulmonary valve with intact

Figure 2

Diagrammatic portrayal of anatomic causes of bronchial compression in congenital heart disease. A. Normal interrelationships of tracheobronchial tree and great arteries. B. Relationships in tetralogy with absent pulmonary valve. Note compression of lower trachea and proximal bronchi. Compression is in an antero-posterior direction. C. Sites of compression in patients with hypertensive, distended pulmonary arteries are indicated by arrows. The left pulmonary artery compresses the left main bronchus as it courses superiorly across it, and the left upper lobe bronchus as it passes posteriorly to it. The right pulmonary artery compresses the origin of the right middle bronchus. D. Bronchial compression by an enlarged left atrium. This may be isolated or complicate other forms of bronchial compression. Tr. = trachea; Ao. = aorta; L.U.Br. = left upper lobe bronchus; I.Br. = intermediate bronchus; R.M.Br. = right middle lobe bronchus; R.P.A. = right pulmonary artery; Inf. = infundibulum; L.A. = left atrium.
ventricular septum and no pulmonary stenosis have been reported. It is unlikely that a ventricular septal defect which had been present in infancy closed spontaneously, since the combination of a large ventricular septal defect with free pulmonary regurgitation would impose a particularly severe hemodynamic burden postnatally. Furthermore, one of these patients died in the neonatal period. It would therefore appear that this represents a distinct group.

In the past the hemodynamic features of absent pulmonary valve with intact ventricular septum have not been separated from those of absent pulmonary valve with obstructive pulmonary valve ring. As both the fetal and postnatal hemodynamics of these two conditions probably differ, separate discussion is warranted.

It has been postulated that because of conditions present in the fetus, massive pulmonary regurgitation could produce prenatal congestive cardiac failure in those with absent pulmonary valve and intact ventricular septum. In the fetus, pulmonary arterial pressure is at systemic levels, and there is free communication with the systemic arterial circuit through the patent ductus arteriosus. Under these circumstances absence of the pulmonary valve could lead to an aortic runoff through the patent ductus arteriosus and pulmonary trunk into the low pressure right ventricle during diastole. The amount of aortic runoff would depend on the resistance offered by the patent ductus and the compliance of the right ventricle. It is likely that the low resistance placental circulation in the fetus would tend to favor forward flow. This may in part explain the rarity of intrauterine congestive failure in this condition. Should tricuspid regurgitation supervene as a result of the right ventricular diastolic volume overload, it is conceivable that retrograde ductal flow would be substantially increased with ensuing intrauterine congestive cardiac failure. The latter has been reported in only one of eight cases of absent pulmonary valve with intact septum.

The hemodynamic considerations are quite different in fetuses with absent pulmonary valve, ventricular septal defect and obstructed pulmonary valve ring. As the ventricular septal defect is invariably large, the pressures in the right and left ventricles should be equal. Obstruction to pulmonary outflow and the relatively low resistance placental vascular bed favors systolic right to left shunting at the ventricular level and possibly increased right to left shunting at the foramen level. The latter may be augmented by impaired diastolic filling of the right ventricle secondary to pulmonary insufficiency, right ventricular dilatation and decreased right ventricular compliance.

The ductus arteriosus is of interest in this condition. In the presence of severe pulmonary outflow obstruction ductal flow in utero would tend to be reversed, i.e., from the aorta to pulmonary artery. Depending on the severity of the stenosis, flow through the ductus would be equal to or less than flow through the pulmonary circulation.

In patients with severe pulmonary obstruction the ductus is narrow and connects with the descending aorta to make an inferior acute angle in contrast to the inferior obtuse angle found in normals. The association of a small patent ductus arteriosus and isolated absent pulmonary valve has been reported in three cases, but has not been documented in those cases with obstructed pulmonary valve ring and a ventricular defect unless there is also interruption of the left pulmonary artery. It would thus seem that retrograde ductal flow does not contribute significantly to postnatal hemodynamics except in those cases with interruption of the left pulmonary artery where it may be the main supply of blood to the left lung.

The predilection for interruption of the pulmonary artery in this combination of lesions suggests that altered fetal flow patterns may be the cause of the atretic portion, i.e., direction of flow from the main pulmonary artery toward the right may predispose to the development of an atretic portion at the origin of the left pulmonary artery. In these patients the left pulmonary artery is supplied by a small patent ductus, and in them it is not possible for aorta to pulmonary artery flow to enhance pulmonary insufficiency or to produce right pulmonary artery dilatation. The finding of dilatation of the right pulmonary artery and not of the left suggests that the dilated pulmonary arteries are related to the pulmonary stenosis and/or insufficiency and not to ductal shunting.

Owing to the relative rarity of absent pulmonary valve and ventricular septal defect most reports have been of isolated cases or small groups of cases. Our series of eight patients has been studied with particular emphasis directed towards assessing the natural history of the disease. We have combined our experience with that reported in the literature and have been able to draw certain conclusions.

Although a male preponderance has been commented on, this is not confirmed by the reported cases. Of the 116 reported cases, the sex is indicated in 77, and in those the male:female ratio is 1:1 (37 male:40 female).

Little attention has been directed to the course of cyanosis in these patients. It is of considerable interest that although cyanosis was usually present in the neonatal period, cardiac catheterization after four weeks almost invariably revealed arterial oxygen...
saturation above 89% except in those with severe continuing pulmonary disease. All our patients were observed to have moderate cyanosis in the newborn period, but this was not apparent after one week of age except in the one patient with pulmonary problems who had progressive cyanosis.

The loss of cyanosis and the occurrence of predominant left to right shunts is probably the result of the decrease in pulmonary vascular resistance which normally occurs in the neonatal period.\textsuperscript{39-41} Two mechanisms are probably operative, one related to systolic afterload, the other to diastolic overload. The high pulmonary vascular resistance immediately after birth together with pulmonic stenosis probably imposes a total pulmonary resistance sufficient to cause right to left ventricular systolic shunting. Diastolic right to left ventricular shunting would be the result of increased diastolic pressure associated with the diastolic overload imposed by the pulmonary insufficiency. A reduction in pulmonary vascular resistance would be associated with a reduction in total right ventricular outflow resistance to a degree which permits left to right ventricular systolic shunting. Pulmonary regurgitation would also diminish with a resultant decrease in right ventricular diastolic overload and less right to left diastolic shunting. A small amount of diastolic right to left shunting may persist even in the presence of a moderate left to right ventricular systolic shunt.

Most of our patients, as well as the vast majority of those reported, had predominant left to right shunts; however, mild systemic arterial desaturation was common, particularly in infants with pulmonary problems. In the absence of pulmonary venous samples one cannot state whether the arterial desaturation was the result of right to left intracardiac shunting and/or the result of pulmonary disease.

Pulmonary complications were the direct result of tracheobronchial compression by dilated pulmonary arteries. The compression involved the anterior aspects of the lower trachea and major bronchi, particularly the right (fig. 2B). This pattern of bronchial compression differs markedly from that seen in patients with large left to right shunts and pulmonary hypertension (fig. 2C) or in those patients with enlarged left atrium (fig. 2D).\textsuperscript{42} Depending on the severity of bronchial obstruction, emphysema and/or atelectasis were commonly seen.

Bronchial compression by distended pulmonary arteries is a particular problem in early infancy and may improve spontaneously towards the end of the first year. Several factors account for this improvement, and these are related primarily to maturational changes in the tracheobronchial tree and the lungs.\textsuperscript{42, 43} In the infant the cartilagenous, muscular and elastic support of the airways is weak. Normally there is partial collapse of the lower airways during expiration, and this collapse may be exaggerated with increased expiratory excursion. These weak-walled structures are particularly susceptible to compression by distended pulmonary arteries. With growth the bronchi become firmer and more resistant to deformation. In addition the bronchial lumina increase in caliber, consequently the same degree of compression would produce less airway obstruction than in the smaller bronchi of young infants. Furthermore, the pores of Kohn allow collateral ventilation in the event of bronchial obstruction; however, these structures are poorly developed in infants. Altered hemodynamics may also play a role in decreasing pulmonary problems with increasing age. An increase in the severity of outflow obstruction with a resulting lessening of pulmonary arterial pressure and/or insufficiency would tend to decrease the distension of the proximal pulmonary arteries.

The pulmonary arterial tree shows several interesting features in these patients. The main and proximal branch pulmonary arteries are usually dilated with frequent aneurysmal dilatation of one branch of the pulmonary artery, more commonly the right. In contrast, the peripheral pulmonary vasculature is either normal or slightly increased. Ventricular left to right shunts are usually present in these patients. Since the stenosis at the pulmonary valve ring is only of moderate degree, right to left shunts are unusual beyond infancy. The combination of a left to right shunt and pulmonary insufficiency results in a large flow across the stenotic pulmonary valve ring and would account for the marked systolic pressure drop resulting in severe post-stenotic turbulence. It is probable that this turbulence is the cause of the dilated pulmonary arteries in most instances, although it has been suggested that the proximal pulmonary arteries are structurally abnormal,\textsuperscript{7} and cystic medial degeneration of the pulmonary arteries has been documented in one patient with absent pulmonary valve and Marfan's syndrome.\textsuperscript{18}

There was a striking association between pulmonary artery dilatation and the orientation of the infundibulum. When the infundibulum was oriented toward the right, the right pulmonary artery was aneurysmal. Similarly, vertical and leftward orientations resulted in bilateral pulmonary artery dilatation and left pulmonary artery dilatation, respectively. This suggests that the orientation of the post-stenotic jet determined which pulmonary arteries were dilated.

The abnormal orientation of the infundibulum may make catheterization of the pulmonary artery difficult, particularly in early infancy. A conventional semi-
rigid catheter may not make the sharp turn from the right ventricle into the long, horizontal infundibulum and from there through the posteriorly positioned pulmonary valve ring (fig. 3). Because the pulmonary valve ring is quite posterior, the catheter may appear to be in the pulmonary artery when it is, in fact, still within the infundibulum. Pulmonary insufficiency may prevent passage of a balloon catheter into the pulmonary artery.

In attempting to assess the relative merits of medical or surgical management, it becomes apparent that one is dealing with two separate patient populations: those with and those without severe pulmonary complications. The recorded cases would indicate that infants with severe pulmonary complications have an extremely high mortality if treated only medically. Our estimated mortality of 76% represents a minimum since follow-up information is not available in many of the reported cases. Those infants with pulmonary problems who underwent surgical intervention also had a high mortality (41%) but somewhat less than with medical management.

A variety of palliative procedures have been performed in an attempt to relieve the pulmonary problems in infants and include plication of aneurysmal pulmonary arteries, a sling around the dilated pulmonary artery attached to the retrosternal fascia, and resection of the involved pulmonary artery with graft replacement. A total of 18 palliative procedures have been performed; consequently, the numbers for each procedure are small. The mortality for aneurysmorrhaphy and the various miscellaneous palliative procedures is shown in figure 4. Patients with mild or no pulmonary problems usually survived infancy. Most deaths in this group (10) were operative. Thirty-two patients have had corrective procedures (fig. 4) with a mortality of 31%.

Twenty-eight patients have had the ventricular septal defect repaired with resection of the hypertrophied infundibulum. Ten of these have died. In four patients, including one in our series, only the ventricular septal defect was repaired, with no mortality in this small group. It is debatable whether infundibular resection is necessary. It would appear both from our own small series, as well as from a review of the pathological data in the literature, that infundibular stenosis is rare, and that stenosis at the valve ring is usual in this condition.

It would seem reasonable to recommend closure of the ventricular septal defect as the primary procedure, where corrective surgery is indicated, unless definite evidence of infundibular stenosis is present at cineangiography.

Our own experience indicates that those patients without serious pulmonary problems do quite well on medical management. Both patients in our series who died had severe pulmonary complications. Of the remaining six, four are doing well with no therapy, one had the ventricular septal defect closed, and one is scheduled for surgery because of a prolapsed aortic valve cusp.

The prognosis of patients with absent pulmonary valve and ventricular septal defect is variable. We believe that the prognosis for those patients without severe pulmonary problems is not as dismal as has been previously reported for the entire group. With adequate medical management these patients appear...
to run a course similar to that seen in patients with mild Fallot's tetralogy. As in the latter condition, surgery can be performed at a time when the patient is adequately compensated and has grown to a reasonable size. Those patients with pulmonary complications continue to be therapeutic problems, and the best approach to management has not been determined.

Acknowledgment

The authors are grateful to Drs. S. J. Robinson, M. Auerbach and S. Sevy for permission to include their patients in this report, and for their helpful suggestions.

References


Circulation, Volume 50, July 1974
40. HEYMANN MA, RUDOLPH AM: Effects of congenital heart disease on fetal and neonatal circulations. Prog Cardiovasc Dis 15: 115, 1972
Tetralogy of Fallot with Absent Pulmonary Valve: Natural History and Hemodynamic Considerations

JEFFREY B. LAKIER, PAUL STANGER, MICHAEL A. HEYMANN, JULIEN I. E. HOFFMAN and ABRAHAM M. RUDOLPH

Circulation. 1974;50:167-175
doi: 10.1161/01.CIR.50.1.167

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1974 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/50/1/167

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/