Pulmonary Stenosis and Ventricular Septal Defect with Arteriovenous Shunts
A Clinical and Hemodynamic Study of Eleven Patients

By John A. Callahan, M.D., Robert O. Brandenburg, M.D. and H. J. C. Swan, M.B., Ph.D., M.R.C.P. (Lond.)

Patients who have pulmonary stenosis and ventricular septal defect usually have a right-to-left intracardiac shunt. This paper presents hemodynamic and clinical data on 11 patients who have pulmonary stenosis, ventricular septal defect and left-to-right shunts. Four of these patients also have defects in the atrial septum, and three have demonstrable bidirectional shunts. The level and direction of shunts were determined by blood oxygen-saturation data and indicator-dilution studies. Choice of the form of treatment to be used in patients such as these should be based largely on hemodynamic considerations. Many resemble patients with tetralogy of Fallot who have had a successful pulmonary valvotomy.

In patients who have stenosis of the pulmonary valve or outflow portion of the right ventricle and a ventricular septal defect, blood is usually shunted through the defect to the left ventricle or the aorta. Recent publications have called attention to a group of patients who have pulmonary stenosis and ventricular septal defect without cyanosis, in whom the blood is shunted from left to right (arteriovenous) through the defect.

It is the purpose of this paper to present hemodynamic and clinical data in seven cases of pulmonary stenosis and ventricular septal defect in which a left-to-right shunt was the major hemodynamic anomaly and in four cases of pulmonary stenosis with both atrial and ventricular septal defects, all of which had left-to-right shunts but three had right-to-left (venoarterial) shunts in addition. Two of the first group of patients had been previously reported by Broadbent and associates.

The patients were studied by cardiac catheterization, employing the method of Courmand and Ranges as modified in apparatus and techniques by Wood and associates. The methods of analysis used have been described in greater detail in a recent paper by the present authors.

Arterial Indicator-Dilution Curves

Because of the value of indicator-dilution curves in the definition of normal and abnormal circulatory states, a statement outlining the principles which are the basis for the use of dilution curves in the study of congenital heart disease with intracardiac and intravascular shunts is appropriate. The changing concentration of an indicator with time, at the sampling site in the arterial system, defines a contour which depends to a major extent on the pathway or pathways taken by the indicator from the injection site to the sampling site. Dilution curves do not define defects as such but indicate the nature of pathways of blood flow, normal or abnormal. Two distinct and fundamental dilution patterns are associated with the presence of a right-to-left and of a left-to-right shunt respectively. When a right-to-left shunt exists, a portion of the injected dye traverses the defect and reaches the systemic arterial sampling site before that portion of the injected dye which passes through the pulmonary circulation. The resultant dilution curve is characterized by a short appearance time and an abnormal initial deflection which is proportional to the amount of dye shunted and hence to the volume of the shunt. When a left-to-right shunt is present, the blood follows a normal pathway through the lungs, but then a part of the blood is
shunted to recirculate through the pulmonary vessels. In this situation the appearance time and the slope of increasing dye concentration of the dilution curve are normal. The magnitude of the peak of dye concentration is reduced in comparison to normal, and the concentration of dye at the sampling site declines slowly because with each recirculation of the indicator remaining in the pulmonary circulation, a constant fraction of the dye passes into the systemic circulation.11 The site of injection of the dye is important in the localization of a shunt. Since the dye follows the pathway taken by the blood, a shunt will be defined only if it occurs at or beyond the site of injection of the dye. Injections of dye into different locations in the heart and great vessels frequently permit the localization of a defect through which a shunt is occurring.12

**CLINICAL DATA**

The clinical data are summarized in table 1. Representative roentgenograms of the chest are reproduced in figure 1.

**Table 1—Clinical Data in 11 Cases of Pulmonary Stenosis With Left-to-Right Intracardiac Shunts**

<table>
<thead>
<tr>
<th></th>
<th>Group I: Pulmonary stenosis and ventricular septal defect*</th>
<th>Group II: Pulmonary stenosis, atrial and ventricular septal defects</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, year</td>
<td>1  2  3  4  5  6  7</td>
<td>1  2  3  4</td>
</tr>
<tr>
<td>Sex</td>
<td>F  M  F  F  M  M</td>
<td>M  F  M  F</td>
</tr>
<tr>
<td>Effort dyspnoea and fatigue</td>
<td>2  1  1  3  3  1?</td>
<td>1  3  3  2</td>
</tr>
<tr>
<td>(grade 0-4)†</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cyanosis (grade 0-4)† since</td>
<td>0  0  0  0  1, 1 yr. 0  0</td>
<td>1, birth 1+, birth† 0  2, 5 yr.†</td>
</tr>
<tr>
<td>Systolic murmur</td>
<td>Gr. 4 with thrill Present with thrill</td>
<td>Gr. 2 with thrill Gr. 3 left 5th space with thrill Gr. 3 with thrill Present with thrill</td>
</tr>
<tr>
<td></td>
<td>(louder right than left)</td>
<td>Gr. 3 with thrill</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other murmur</td>
<td>0 Diastolic lower left sterno. Gr. 1 diastolic pulmonary area Diastolic pulmonary area Diastolic pulmonary area Gr. 2 diastolic 3rd and 4th space</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Indistinct, not split or accent.</td>
<td>Single Accent. Single Accent. Accent. Accent. but single</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Heart enlargement (grade 0-4)†</td>
<td>0  2  2  3  1  2</td>
<td>0  1  3  2</td>
</tr>
<tr>
<td>Pulmonary arterial shadow</td>
<td>2  2  3  2  0 (right-sided aorta) 1  4  0</td>
<td></td>
</tr>
<tr>
<td>(grade 0-4)†</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pulmonary vascular markings</td>
<td>Normal Increased Increased Slight increase Normal Increased Normal Decreased Normal Normal Normal Normal</td>
<td></td>
</tr>
</tbody>
</table>

* Patients 4 and 6 were reported in a previous study.2
† Severity of symptoms and physical signs graded on a 0 (absent) to 4 (severe or marked) basis.
‡ Clubbing present.
Fig. 1. Posteroanterior roentgenograms a and b. Patients 1 and 3, group I. The pulmonary arterial shadow is increased in both patients but particularly in patient 3 (b). The vascular lung markings are markedly increased in patient 3 (b) but are normal in patient 1 (a). c and d. Patients 1 and 4, group II. There is concavity in the region of the pulmonary arterial shadow in both patients and a right-sided aortic arch in patient 1 (c).

The most characteristic physical finding was a loud systolic murmur in the pulmonic area associated with a thrill. In some of the patients, murmurs were noted in other locations. The second heart sound in the second and third left intercostal spaces was absent, single or not accentuated in most of these patients.

The main pulmonary arterial shadow in the postero-anterior roentgenogram of the chest was increased in all patients except patient 7, ni whom it could not be definitely identified. The pulmonary vascular markings were considered to be normal in patients 1, 5 and 7 and increased in patients 2, 3, 4 and 6.

The electrocardiogram in patient 1 disclosed minimal electrocardiographic evidence of delayed activation of the right ventricle (R wave in aV_{n}). In patient 2 there was atrioventricular dissociation with auricular fibrillation. The electrocardiograms in patients 3 and 7 were interpreted to show evidence of biventricular hypertrophy. The electrocardiograms in patients 4 and 6 showed the configuration of right bundle-branch block in lead V_{1} (partial right
bundle-branch block in the latter, with evidence of left ventricular hypertrophy). In patient 5 there was evidence of right ventricular hypertrophy.

The patients in group II were quite similar to those in group I on clinical grounds, except that three of the patients in group II had cyanosis and two had clubbing of the fingers.

The pulmonary arterial shadow was increased in two patients and normal in two. One of the patients in whom the pulmonary arterial shadow was normal was found to have a right-sided aortic arch. The pulmonary vascular markings were normal in three patients and decreased in one.

The electrocardiogram in patient 1 showed no definite evidence of ventricular hypertrophy. In patients 2 and 4 there was electrocardiographic evidence of right ventricular hypertrophy. In patient 3 there was the pattern of right bundle-branch block and left ventricular hypertrophy.

**Hemodynamic Data**

**Pulmonary Stenosis and Ventricular Septal Defect** (Table 2). The seven patients with pulmonary stenosis and ventricular septal defects were found to have arterial oxygen saturations of systemic arterial blood within the range of normal, while at rest, breathing air. The average arterial oxygen saturation in these patients, measured by the method of Van Slyke, was 97 per cent (range 95 to 99 per cent). In all but one of these patients, the arterial oxygen saturation was determined while the patient breathed 100 per cent oxygen. The average quantity of oxygen in physical solution (oxygen content minus oxygen capacity) found in arterial blood when 100 per cent oxygen was breathed was 1.6 volumes per 100 cc. of blood (range 1.0 to 2.1 volumes per cent). Under this circumstance, patient 1 was found to have 1.0 volume of oxygen per 100 cc. of blood in physical solution, which is below the range defined by Wood for normal subjects, but the arterial oxygen saturation was within normal limits when this patient breathed air. It is unlikely that there was a significant right-to-left shunt in this patient because of the wide difference between systemic arterial and right ventricular systolic pressures. Also an arterial indicator-dilution curve obtained following injection of T-1824 into the right ventricle while the patient exercised failed to show evidence of a right-to-left shunt.* In patient 5, on the other hand, there was evidence by indicator-dilution curves of a

* Identification of small right-to-left shunts on the basis of minor degrees of desaturation of the systemic arterial blood is limited by the range of variability of arterial saturation encountered in normal subjects of from 94 to 101 per cent by Van Slyke analysis. It is possible for a 15 to 20 per cent right-to-left shunt to exist in the presence of an arterial oxygen saturation within the range of normal. Indicator-dilution curves have proved a more sensitive and a more certain method of diagnosis.  

---

**Table 2.**—Intracardiac Pressures, Arterial Oxygen-Saturation Data, Blood Flow and Shunt Values in Seven Cases of Pulmonary Stenosis and Ventricular Septal Defect With Left-to-Right Shunt

<table>
<thead>
<tr>
<th>Case</th>
<th>Pressure, mm. Hg</th>
<th>% oxygen saturation breathing air</th>
<th>Arterial oxygen* physical solution, vols. per 100 cc.</th>
<th>Blood flow index, L/min/M²</th>
<th>L-R shunt, per cent of pulmonary flow</th>
</tr>
</thead>
<tbody>
<tr>
<td>PT</td>
<td>RV out-flow</td>
<td>Low RV</td>
<td>RA Radial artery Mixed venous blood Pulmonary artery Radial artery</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>18/5</td>
<td>52/4</td>
<td>52/4</td>
<td>8/2</td>
<td>139/71</td>
</tr>
<tr>
<td>2</td>
<td>46/18</td>
<td>48/2</td>
<td>95/2</td>
<td>11/3</td>
<td>150/84</td>
</tr>
<tr>
<td>3†</td>
<td>31/12</td>
<td>63/9</td>
<td>73/3</td>
<td>7/2</td>
<td>120/67</td>
</tr>
<tr>
<td>4</td>
<td>20/6</td>
<td>97/7</td>
<td>104/7</td>
<td>11/7</td>
<td>105/99</td>
</tr>
<tr>
<td>5†</td>
<td>31/12</td>
<td>55/2</td>
<td>90/3</td>
<td>6/3</td>
<td>95/60</td>
</tr>
<tr>
<td>6</td>
<td>48/17</td>
<td>50/3</td>
<td>85/2</td>
<td>11/4</td>
<td>109/60</td>
</tr>
<tr>
<td>7</td>
<td>25/11</td>
<td>26/1</td>
<td>91/1</td>
<td>13/8</td>
<td>198/38</td>
</tr>
</tbody>
</table>

* Oxygen content minus oxygen capacity when breathing 100 per cent oxygen.
† Patient studied under anesthesia.
‡ Radial and right ventricular pressures not measured simultaneously.
§ Small right-to-left shunt found by dye dilution studies.
PULMONARY STENOSIS AND VENTRICULAR SEPTAL DEFECT

Fig. 2. Section of photographic record from the cardiac catheterization of patient 2, group I. In the upper panel the tip of the cardiac catheter was withdrawn successively from the pulmonary artery (1), through a zone of infundibular stenosis (2), into the right ventricle (3) and then advanced through the ventricular septal defect (4) into the left ventricle and into the aorta. This record is indicative of the presence of a normal pulmonary valve and of infundibular pulmonary stenosis. Note the gradient in pressure between the right and left ventricles and the change in the electrocardiogram recorded from the tip of the cardiac catheter as it crosses the ventricular septal defect. In the lower panel, pressures were recorded successively in the right ventricle, left ventricle and aorta and simultaneously in the radial artery. Note that the radial arterial systolic pressure exceeds the simultaneously recorded aortic, systolic pressure.

small right-to-left shunt at ventricular level but 1.9 volumes of oxygen per 100 cc. of blood were present in physical solution when the patient breathed 100 per cent oxygen.

The right ventricular systolic pressure was elevated above normal levels in each patient. The average right ventricular systolic pressure was 88 mm. of mercury (range: 52 to 120 mm.) and the radial arterial systolic pressure exceeded the right ventricular systolic pressure in each case. All but one of the measurements were obtained from simultaneous records of radial arterial and right ventricular pressures (fig. 2). The average difference between systolic pressures in the radial artery and the right ventricle was 41 mm. of mercury.*

The pulmonary arterial pressure was elevated above the normal range in some of the patients in group I. The average pulmonary arterial systolic pressure was 32 mm. of mercury (range: 18 to 48 mm.).

* A comparison of radial arterial and right ventricular systolic pressures does not necessarily reflect the pressure gradient between the right and left ventricles. Kroeker and Wood15 found that in 12 normal male subjects, the peripheral arterial systolic pressure uniformly exceeded the aortic systolic pressure generated by the same heartbeat. The average radial systolic pressure was 112 per cent of aortic systolic pressure in their series.
In each case an attempt was made to define the nature of the pulmonary stenosis by examining the continuous record of the pressure pulses obtained when the tip of the catheter was withdrawn slowly from the pulmonary artery to the right ventricle (fig. 2). Pressure tracings characteristic of valvular, infundibular and combined valvular and infundibular stenosis have been described by Kirklin and associates. One patient (case 1) appeared to have purely valvular stenosis. Three patients (cases 2, 6 and 7) appeared to have only infundibular stenosis. Three patients (cases 3, 4 and 5) appeared to have combined valvular and infundibular stenosis.

Pulmonary blood flow in each case exceeded systemic blood flow and averaged 6.7 liters per minute per square meter of body surface area (range: 3.9 to 11 liters per minute per square meter), while the systemic flow averaged 3.6 liters per minute per square meter (range: 2.2 to 5.5 liters per minute per square meter). There was a large left-to-right shunt in each case, which averaged 43 per cent of pulmonary flow (range: 25 to 60 per cent). Because of the criteria of selection of these patients, no right-to-left shunt large enough to be detected by measurements of blood oxygen saturation could be demonstrated.

Arterial indicator-dilution curves were recorded in each patient by ear oximeter and simultaneously by a cuvette oximeter through which blood from the radial artery was permitted to flow following injection of T-1824 at various sites in the heart and great vessels. The dilution curves recorded in all patients other than patient 5 had normal appearance times and slopes of increasing dye concentration, indicating that no right-to-left shunt existed in these patients. All of the dilution curves had markedly prolonged slopes of declining dye concentration, indicating left-to-right shunts of considerable magnitude. In case 5, an indicator-dilution curve recorded following injection of dye into the right ventricle had a short appearance time and a small abnormal initial deflection, indicating a right-to-left shunt which amounted to approximately 15 per cent of systemic flow. This shunt was not detectable on the basis of oxygen saturation data.

In two of the patients (cases 2 and 5) the cardiac catheter passed from the right ventricle into the aorta.

Patient 7 differed from the other patients in certain features. There appeared to be a zone of infundibular stenosis which was defined when the cardiac catheter was withdrawn slowly from the pulmonary artery to the right ventricle. The oxygen saturation of the blood in the infundibular area was higher than that in the outflow portion of the right ventricle. For example, the oxygen saturations of intracardiac blood samples, measured in quick succession by the cuvette oximeter on one of several occasions, were: pulmonary artery 85 per cent, infundibular zone 84 per cent, low right ventricle 73 per cent, right atrium 70 per cent. The radial arterial and aortic pressures were 200/45 and 148/57 respectively, when recorded simultaneously. In this case the final diagnosis rested between pulmonary stenosis, ventricular septal defect and aortic insufficiency on the one hand, and rupture of an aneurysm of a sinus of Valsalva with communication between the aorta and the outflow tract of the right ventricle on the other.

Pulmonary Stenosis, Ventricular Septal Defect and Atrial Septal Defect. In addition to pulmonary stenosis and ventricular septal defect, the four patients in group II have an associated defect in the atrial septum (table 3). In each of these four patients there is a left-to-right shunt, but in three of them there is a right-to-left shunt in addition. The findings for each patient are considered below and shown diagrammatically in figures 3 and 4.

Case 1. The catheter crossed an atrial septal defect and passed into the left atrium and left ventricle. Intracardiac pressures of 9/2 and 79/6 were recorded in these positions. Immediately following this the catheter was withdrawn to the right atrium and advanced into the right ventricle, and pressures of 9/4 and 65/5 respectively were obtained at these sites. The patient's hemodynamic status did not appear to have changed between observations. The cardiac catheter did not enter the pulmonary artery, and the diagnosis of pulmonary stenosis depended on the presence of a coarse systolic murmur and thrill at the pulmonary area and x-ray findings of a slight decrease in pulmonary vascular markings.
associated with right ventricular hypertension. The second sound in the pulmonary area, though accentuated, was single.

The oxygen saturation of the blood in the right ventricle was higher than that in the right atrium, indicating the presence of an arteriovenous shunt into the right ventricle. While the patient breathed air, the oxygen saturation of the right atrial blood was not higher than that of venae cavae blood; but when the patient breathed 100 per cent oxygen, the oxygen saturation of right atrial blood exceeded that of the caval blood by 5 per cent, giving evidence of a left-to-right shunt at atrial level under this circumstance.

An arterial indicator-dilution curve recorded following injection of T-1824 into the right ventricle had a normal appearance time and slope of increasing dye concentration, showing that there was no right-to-left shunt at ventricular level. Dilution curves recorded following injection of dye into the superior and inferior venae cavae had short appearance times and abnormal initial deflections, indicating a right-to-left shunt at atrial level. The initial deflection in the curve recorded following the superior venous caval injection was larger than that recorded following the superior caval injection. It was estimated that 25 per cent of inferior caval blood and 20 per cent of superior caval blood traversed the defect. The difference in the relative magnitudes of the shunts from each cava, as demonstrated by Swan and co-workers, strongly favors an atrial septal defect as the site of the right-to-left shunt in this instance. These dilution curves are also consistent with a left-to-right shunt of considerable magnitude. A dye-dilution curve recorded following injection of dye into the left ventricle had a normal appearance time and slope of increasing dye concentration for this site of injection. The slope of declining dye concentration was less steep than would be expected from the initial portion of the curve, indicating a left-to-right shunt at or distal to the left ventricle. This dilution curve is compatible with the finding of a higher oxygen saturation in the right ventricle than in the right atrium. The curve recorded following injection into the left atrium is characterized by a more abnormal slope of declining dye concentration and suggests the presence of a left-to-right shunt at atrial level.

**Case 2.** The left side of the heart was not catheterized in this patient. The blood oxygen saturation in the right atrium was found to be higher than that in the venae cavae and the saturation in the right ventricle was slightly higher than that in the right atrium. This was evidence for the existence of left-to-right shunts at both atrial and ventricular levels.

Spontaneous fluctuations in oxygen saturation were present in this patient and affected the recording of dilution curves of T-1824. When dye was injected into the right pulmonary artery the resultant curve showed a normal appearance time and slowed build-up and disappearance phases. When dye was injected into the venae cavae and right ventricle the resultant curves had short appearance times and abnormal initial deflections, demonstrating the existence of a right-to-left shunt. There was no substantial difference in the initial deflections in the dilution curves recorded following the caval injections, as had been noted in case 1, and these deflections were not larger than the initial deflection in the dilution curve recorded following the right ventricular injection. This demonstrated that a large right-to-left shunt existed at ventricular level and did not support but did not completely exclude the presence of a right-to-left shunt at atrial level.

**Case 3.** Pressures were measured in all four chambers of the heart. As in case 1, the cardiac catheter passed through an atrial septal defect. The pressures obtained were as follows: right atrium 5/3, left atrium 9/4, right ventricle 66/2 and left ventricle 83/3 mm. of mercury. The blood oxygen saturation was found to be higher in the right atrium than in the venae cavae and higher in the

### Table 3.—Intracardiac Pressures, Blood Oxygen-Saturation Data, Blood Flow and Shunt Values in Four Cases of Pulmonary Stenosis With Atrial and Ventricular Septal Defects

<table>
<thead>
<tr>
<th>Case</th>
<th>Pressure, mm. Hg</th>
<th>% oxygen saturation breathing air</th>
<th>Arterial oxygen,* physical solution, vols. per 100 cc.</th>
<th>Blood flow index L./min./M²</th>
<th>Shunt</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Pulmonary artery</td>
<td>RV outflow</td>
<td>Low RV</td>
<td>LV</td>
<td>RA</td>
</tr>
<tr>
<td>1†</td>
<td>—</td>
<td>63/6 65/579/6</td>
<td>9/4 9/2100/71</td>
<td>68</td>
<td>74</td>
</tr>
<tr>
<td>2</td>
<td>12/8</td>
<td>75/6 106/5</td>
<td>7/3 115/67</td>
<td>70</td>
<td>75</td>
</tr>
<tr>
<td>3‡</td>
<td>22/13</td>
<td>50/3 66/2 84/3</td>
<td>4/3 11/4 97/62</td>
<td>73</td>
<td>86</td>
</tr>
<tr>
<td>4‡‡</td>
<td>20/12</td>
<td>106/10</td>
<td>93/8 96/6 10/11/7 103/67</td>
<td>72</td>
<td>80</td>
</tr>
</tbody>
</table>

* Oxygen content minus oxygen capacity when breathing 99.6 per cent oxygen.
† Patient studied under anesthesia.
‡ Oxygen uptake estimated from tables of normal values.¹⁷
right ventricle than in the right atrium. This was evidence for left-to-right shunts at both levels.

Indicator-dilution curves recorded following injection of dye into the venae cavae and the right ventricle had normal appearance times and normal slopes of increasing dye concentration, showing that there was no right-to-left shunt in this patient. The curves all had prolonged and flattened slopes of declining dye concentration, showing that a large left-to-right shunt existed. A dye-dilution curve was also recorded following injection into the left ventricle. The appearance time and slope of increasing dye concentration were normal, but there was marked prolongation of the slope of decreasing dye concentration, compatible with a large left-to-right shunt from or distal to the left ventricle.
TABLE 4.—Comparison of Intravascular Systolic Pressures in 26 Normal Subjects,19 11 Patients With Tetralogy of Fallot18 and Seven Patients With Pulmonary Stenosis, Ventricular Septal Defect and Arteriovenous Shunts

<table>
<thead>
<tr>
<th></th>
<th>Pressure, mm. Hg</th>
<th>Right ventricle minus pulmonary artery</th>
<th>Radial artery minus right ventricle</th>
</tr>
</thead>
<tbody>
<tr>
<td>Present series...</td>
<td>89 31 131</td>
<td>58</td>
<td>42</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>104 17 118</td>
<td>87</td>
<td>14</td>
</tr>
<tr>
<td>Normal subjects*</td>
<td>27 22 135</td>
<td>5</td>
<td>Not relevant</td>
</tr>
</tbody>
</table>

* Pressures are averaged from series and do not necessarily indicate the exact magnitudes of the gradient of pressure across the pulmonary valve in the normal subjects.

Case 4. Pressures were measured in all four chambers of the heart as in cases 1 and 3. In contrast to the other patients, the hemodynamic status in this patient underwent alteration during the course of study, owing to the occurrence of episodes of nodal rhythm. The pressures in the atria were found to be virtually identical when recorded in quick succession, averaging 10/7 and 13/6 when the rhythm was normal and nodal respectively. The ventricular pressures were also considered to be nearly identical. The oxygen saturation of the blood in the right atrium was slightly higher than that in the venae cavae, and a further increase in oxygen saturation was found in the right ventricle, demonstrating left-to-right shunts at both levels.

Dye-dilution curves recorded following injection of T-1824 into the venae cavae and right ventricle had short appearance times and abnormal initial deflections. The dilution curve recorded following injection of dye into the right ventricle indicated that approximately 20 per cent of the systemic blood was shunted from the right ventricle. The caval curves indicated that approximately 35 per cent of systemic blood was shunted from the venae cavae or sites distal thereto. As in case 1, the initial deflection recorded following the inferior caval injection was larger than that recorded following the superior caval injection. A dilution curve recorded following injection of dye into the left ventricle was similar in contour to that obtained from this injection site in case 1, supporting the evidence for a left-to-right shunt from the left ventricle.

**Comment**

The important determinants of the direction of flow across a ventricular septal defect are the pressure differential between the ventricles and the size of the defect. A major factor controlling right ventricular pressure is the resistance to pulmonary flow. In patients with pulmonary stenosis the main resistance to pulmonary flow is proximal to the lungs, at a stenotic pulmonary valve or in the outflow tract of the right ventricle. Variation in the relative magnitudes of the resistances to pulmonary and systemic blood flow in these patients allows most of the intermediate hemodynamic situations to exist ranging from pure left-to-right shunting, seen in the cases of this report, to predominant right-to-left shunting, as seen in the more classic cases of tetralogy of Fallot.

The basic cardiac malformations in the patients in group I have a certain resemblance to the hearts of patients with tetralogy of Fallot as noted by Moffitt and associates in a group of similar patients. In spite of the anatomic similarity of the congenital anomalies, increased pulmonary blood flow is a feature of the hemodynamic state in these patients, and they may be properly designated as examples of acyanotic congenital heart disease. Comparisons were made between the present group of patients, a series of patients with tetralogy of Fallot and a series of normal subjects studied by cardiac catheterization in this laboratory (table 4). Several interesting features are apparent. In the patients presented in this paper the average pulmonary arterial systolic pressure exceeded that pressure in the patients with tetralogy of Fallot and also in the normal subjects. This suggests that the pulmonary arterial systolic pressure is a function of the pulmonary blood flow. However, comparing the present series of patients with the cases of tetralogy of Fallot, there was a smaller pressure gradient across the pulmonary valve (58 versus 87 mm. Hg) in the presence of a greater pulmonary blood flow (6.7 versus 2.3 L. per minute per square meter). The degree of pulmonary stenosis in the present series of patients must be relatively mild. Barratt-Boyes and Wood have demonstrated a small systolic pressure gradient between the pulmonary artery and the right ventricle in normal subjects.
A considerable pressure gradient existed (average 42 mm. Hg) between the right ventricular systolic pressure and the radial arterial systolic pressure in the present series, whereas there was a much smaller difference between these pressures in the patients with tetralogy of Fallot (average 14 mm. Hg). The radial arterial systolic pressures in the patients with tetralogy of Fallot averaged 114 per cent of the right ventricular systolic pressures. It is interesting to note that Kroeker and Wood found radial arterial systolic pressures to be 112 per cent of aortic systolic pressures in a series of normal subjects.

When the subject of pulmonary stenosis and ventricular septal defect is considered, the relation of the origin of the aorta to the right ventricle is of interest. The aorta was catheterized in two of the seven patients in group I of the present series and in 10 of the 21 cases of tetralogy of Fallot. Chapman and associates considered overriding of the aorta a diagnosis to be made only at necropsy. Gordon and associates believed that overriding of the aorta could be diagnosed if a cardiac catheter passed into the aorta. This conclusion is not necessarily correct. As was shown by Eisenmenger in dog hearts and by Edwards and associates the human heart, if an artificial defect is made in the membranous portion of the ventricular septum of a normal heart, the aorta communicates directly with both ventricles and a probe or cardiac catheter can be passed from the right ventricle into the aorta.

The four patients in group II were included in this report because they satisfied the basic criteria for selection in that they had pulmonary stenosis, ventricular septal defect and left-to-right shunt. It is felt that anatomically the four patients resembled each other but their circulatory dynamics were quite dissimilar. The nature of the disorder in these four patients is complex and a description of the hemodynamic as well as the anatomic situation is necessary for an adequate assessment of the condition. Any decision about the form of treatment to be used in patients such as these should be based largely on hemodynamic considerations.

Summary

Clinical and hemodynamic data have been presented in 11 cases of pulmonary stenosis and ventricular septal defect with left-to-right shunt. Four of these patients also had interatrial communications and three of these had bidirectional shunts.

The clinical diagnosis depends primarily on the diagnostician's being aware of the possible existence of a left-to-right intracardiac shunt with increased pulmonary blood flow in the presence of signs of pulmonary stenosis. All except one of the first seven patients had a harsh systolic murmur and thrill in the pulmonary area. The pulmonary arterial shadow was increased in all except one of these patients. Other physical signs, the pulmonary vascular markings and the electrocardiograms were more varied.

The technic of selective injections of T-1824, used in conjunction with measurements of blood oxygen saturation in the chambers of the heart and great vessels by cuvette oximeter, was of great value in the definition of the level, direction and magnitude of intracardiac shunts.

Acknowledgment

The authors gratefully acknowledge the critical interest of Drs. H. B. Burchell, J. E. Edwards and E. H. Wood in the preparation of this paper.

Summario in Interlingua

Patientes con stenose pulmonar e defeoto ventriculo-septal ha usalmente un dextero-sinistre derivation intracardiac. Le presente reporto offere datos hemodynamic e clinic in re 11 patientes qui ha stenose pulmonar, defeo ventriculo-septal, e sinistro-dextere derivationes. Quatro de iste patientes ha etiam defeectos in le septo atrial, e tres ha demonstrabile derivationes ambidirectional. Le nivello e le direction del derivationes esseva determinate super le base de datos de saturation oxygenic del sanguine e de studios de dilution de injections indicatori. Le selection del forma de tractamento pro patientes del typo hic discutete debe esser basate in grande mesura super considerationes hemodynamic. Multes resimila patientes con tetralogia de Fallot post succedite valvulotomia pulmonar.
REFERENCES


Pulmonary Stenosis and Ventricular Septal Defect with Arteriovenous Shunts: A Clinical and Hemodynamic Study of Eleven Patients
JOHN A. CALLAHAN, ROBERT O. BRANDENBURG and H. J. C. SWAN

Circulation. 1955;12:994-1004
doi: 10.1161/01.CIR.12.6.994
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1955 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/12/6/994

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/