Valvular Pulmonary Stenosis with Intact Ventricular Septum

Clinical and Physiologic Response to Open Valvuloplasty

By S. Gilbert Blount, Jr., M.D., Jack van Elk, M.D., Oscar J. Balchum, M.D., Ph.D., and Henry Swan, M.D.

Clinical and physiologic studies have been carried out in 25 patients following surgical correction of congenital valvular pulmonary stenosis. The patients were operated upon under conditions of hypothermia and circulatory occlusion and the approach to the valve was transarterial, permitting plastic repair of the stenotic pulmonary valve with unimpaired vision and a dry operative field. The systolic pressure gradient between the right ventricle and the pulmonary artery has been completely abolished in 17 of the 25 patients. The results as reported in this series are considered to be superior to those obtained with the blind transventricular approach and the operative mortality certainly compares favorably with the transventricular approach.

Valvular pulmonary stenosis with an intact ventricular septum is a common congenital anomaly. Frequently this defect is not detected early in life as the completely asymptomatic course does not bring the child to the attention of a physician. The increase in periodic physical examinations of school children in recent years has led to the discovery of a greater number of individuals with this type of congenital heart disease. However, the clinical findings are easily misinterpreted, and because of the complete lack of symptoms and the normal heart size the severity of this anomaly has frequently not been appreciated. The true significance of this entity and its potential danger, however, is now more generally realized.

Several forms of corrective surgery for valvular pulmonary stenosis have been developed. The transventricular approach of Brock has been widely employed; however, evaluation of the operative results both at this institution and elsewhere has led to the conclusion that this approach often does not satisfactorily relieve the stenosis.1-5

The application of hypothermia with circulatory occlusion has permitted open heart surgery, whereby the type and the degree of the stenosis can be evaluated and valvuloplasty performed with deliberation. Our experience with this approach in 38 patients has demonstrated that the mortality is no greater than with the blind ventricular approach and that results are superior.

Materials and Methods

A total of 38 patients has been operated on by this technic. Ten patients were cyanotic. The 2 deaths in the series were in this group.

This study concerns itself mainly with the first 25 consecutive patients who have been evaluated before and after surgery from a clinical and physiologic standpoint. The remaining 13 patients have been operated upon too recently for follow-up studies. These 25 patients can be divided into 2 groups, 15 patients with an intact atrial septum and 10 patients with associated functional or anatomic atrial septal defect (tables 1 and 2).

All patients were operated via a transarterial approach under hypothermia and circulatory occlusion.6 The stenotic valve was incised and partially excised in the first 3 patients. In all other patients the stenotic valve was incised along the commissures, without removal of valve tissue. The first 3 patients have now been followed for 3 to 3½ years, while all other patients have been followed for a period of from a few months to 3 years.

All patients except 1 were catheterized prior to operation, and all were re catheterized following surgery. Strain gages and a Hathaway oscillograph were used for the recording of pressures.
Table 1.—Pressures in Fifteen Patients with Valvular Pulmonary Stenosis and Intact Atrial Septum

<table>
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PA = Pressures in mm. Hg in pulmonary artery (PA), right ventricle (RV) and systolic pressure gradient (Grad.) between them.

Sat. = Peripheral arterial blood oxygen saturation in percentages.

* Second reading following exercise.
† Second postoperative catheterization 1 year following surgery.

Table 2.—Pressures in Ten Patients with Valvular Pulmonary Stenosis and Atrial Defect

<table>
<thead>
<tr>
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<td>M</td>
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</table>

PA = Pressures in mm. Hg in pulmonary artery (PA), right ventricle (RV) and systolic pressure gradient (Grad.) between them.

Sat. = Peripheral arterial blood oxygen saturation in percentages.

* Second postoperative catheterization 1 year following surgery.

Results

Clinical Observations

The majority of the patients were young children, only 3 were older than 15 years.

Of the 15 patients with an intact atrial septum (table 1), 7 patients were asymptomatic while the other 8 revealed a slight degree of dyspnea and fatigue, only with sustained exertion. The dyspnea was never severe, and the fatigue was the more prominent complaint,
although this was usually difficult to evaluate, particularly in children. No cyanosis was noted in this group. Following operation all 8 patients reported an improvement in exercise tolerance, and all but 1 were considered to be normal in this respect.

Eight of the 10 patients with a defect in the atrial septum (table 2) were cyanotic. Patient no. 24 was acyanotic and on fluoroscopy showed an increased vascularity of the lung fields; at cardiac catheterization a left-to-right shunt was found at the atrial level. Patient no. 25 was not cyanotic; however bidirectional shunting of blood at the atrial level was demonstrated at cardiac catheterization. After surgery 2 patients became acyanotic, the cyanosis decreased significantly in 2, while in the remaining 4 patients no definite change in color was observed. All patients in this group were limited in their exercise tolerance: 6 had slight limitation; 2 patients, (no. 21 and 25) could not walk more than a few blocks and became more deeply cyanotic on exertion; patient no. 20 had spells of severe cyanosis and dyspnea; and patient no. 18, a deeply cyanotic infant, was in congestive failure during the month prior to surgery. Without exception all patients in this group showed marked improvement in their exercise tolerance following surgery.

A palpable thrill was observed in all patients of both groups. Postoperatively the thrill disappeared in all but 3 patients. A harsh, systolic murmur was audible in all patients. It was of grade IV to VI in intensity in 21 and maximum in the first left intercostal space at the sternal border; 4 patients had only a grade II to III murmur. Following operation the murmur decreased in intensity to grade II or III in all but 4 patients, in whom it was grade IV. The murmur never disappeared completely.

A diastolic murmur of pulmonary insufficiency was noted following operation in the 3 patients in whom the pulmonary valve was partially excised and in 8 of the remaining 22 patients.

Electrocardiography

A tall R wave with a ventricular activation time exceeding 0.03 second was present in lead V1R and V7 in all patients in this study. A deep S wave, usually greater than 3 mm. and as great as 14 mm. was present in V5 and ranged from 3 to 12 mm. in V7. A significant decrease in the height of the R wave in leads V3R and V1 and in the depth of the S wave in leads V6 and V7 was observed in all patients followed for a period of more than 6 months after operation (figs. 1 and 2). The late R deflection in lead aVR also diminished. A striking change

![Fig. 1. Electrocardiographic changes in patient J. K. (10 years old) in whom the valve commissures were excised and some valve substance was removed. Note the continuing decrease of the height of the R wave in V3R, V1, and V2, the decrease of the depth of the S wave in V5 and V7, and the decrease in the late R deflection of aVR. (Operation; March 10, 1953.)](http://circ.ahajournals.org/doi/10.1161/01.CIR.22.3.816)
was also noted in 5 patients, with a change from a tall upright deflection in V₁ to an rSR' pattern.

Radiography

Decreased vascularity of the peripheral lung fields was observed prior to surgery in all the cyanotic patients. In 1 acyanotic patient (no. 24), the vascularity was increased, and it was considered normal in the remaining patients. No significant change in the peripheral vascularity has been definitely detected following operation.

The main pulmonary artery was enlarged in all patients and has not changed so far following surgery. The right and left pulmonary arteries were of normal or slightly increased size. The left pulmonary artery was often bigger than the right pulmonary artery.

The main pulmonary artery showed increased activity and pulsations in all patients. This hyperactivity contrasted with the relatively quiet right and left pulmonary arteries. There was an increase in activity of the latter vessels following surgery in 17 patients; in 2 patients the right pulmonary artery did not show pulsations postoperatively and in the remainder it was difficult to estimate whether or not activity of the right and left pulmonary arteries had increased.

All patients demonstrated an enlarged right atrium and a cardiac configuration suggesting right ventricular hypertrophy. This pattern remained unchanged following surgery. A slight increase in over-all heart size was detected following the surgical correction of the pulmonary stenosis in 10 patients. This increase in heart size was slight and occurred immediately following operation; no further enlargement has been observed, and the over-all size of the heart, to date, is considered to be within normal limits.

Pathology

The valvular deformities seem to fall into 2 categories, one perhaps representing a more advanced form of a single developmental process. In type I, more commonly seen in the younger patients and particularly associated with the higher ventricular pressures and larger gradients, the valve is composed of an elongated cone, more or less symmetrical, tapering to a small circular orifice, usually 1 to 3 mm. in diameter. The valve structure is not much thickened or fibrosed. Evenly spaced around the sulcus of the valve ring are 3 tiny elevations of fibrous tissue representing a rudimentary attempt at commissure formation.

More commonly seen in adult patients is what we have termed the type II deformity.
The valve is much shorter, and consists of 3 well-defined cusps, with fused commissures extending the entire distance to the orifice, which is irregular in shape, with diameters of 2 to 6 mm. The defect is asymmetrical so that the jet stream may be ejected obliquely into the enlarged artery beyond. The rim of the defect is irregularly thickened and very firm and rubbery in consistency, offering considerable resistance to the cutting instrument. The commissures are also thickened, and offer the opportunity to the surgeon to create intact semilunar valves by very precise incisions through the center of the commissures.

**Hemodynamics**

The right heart and pulmonary artery pressures are recorded in tables 1 and 2. The postoperative data were obtained within a 3-month period following surgery in all patients. Data obtained at second postoperative catheterization 1 year following surgery in 4 patients are also included.

The preoperative right ventricular systolic pressures were found to range from 100 to 142 mm. Hg in the 3 patients (no. 1, 2, and 16) in whom the valve was partially excised, but became normal in all 3 patients following operation. The systolic pressure gradient between the right ventricle and the pulmonary artery disappeared or was reduced to the minimal value of 14 mm. The pulmonary artery diastolic pressure decreased to lower than normal levels in 1 patient.

Partial excision of valve tissue has been abandoned as unnecessary; therefore our main concern at the present time is with the hemodynamic data obtained before and after operation in the 22 remaining patients.

In them the systolic pressure gradient between the right ventricle and the pulmonary artery, which ranged from 38 to 175 mm. Hg preoperatively, was reduced to less than 20 mm. Hg in 14 patients (fig. 3). Particular attention is to be given to the remaining 8 patients who had a postoperative gradient of over 20 mm. Hg. An additional interesting finding is that the systolic pressure in the pulmonary artery in 6 patients increased to higher than normal levels postoperatively, ranging from 37 to 49 mm. Hg. In a seventh patient (no. 6) the systolic pulmonary artery pressure was normal at rest (27 mm. Hg) but increased during exercise to 56 mm. Hg (table 1).

The right atrial pressure was reduced following surgery (fig. 4) in patients in whom this pressure was high preoperatively, and in whom the pressure record showed a giant a-wave.

In the patients with a defect in the atrial septum, 9 revealed cyanosis or peripheral arterial oxygen unsaturation as evidence of a
right-to-left shunt through the defect. Following operation 2 patients (no. 17 and 21) showed an increase in their peripheral arterial oxygen saturation to normal or nearly normal levels.

No peripheral arterial blood was obtained prior to surgery in patient no. 18, who was severely cyanotic and in congestive failure. Postoperatively the oxygen saturation was 87.7 per cent, probably a very significant increase.

In 2 other patients the oxygen content of the peripheral arterial blood was not determined before and after operation. In the remaining 4 patients the peripheral arterial oxygen saturation has not changed significantly following surgery for periods of a few months to 2 years.

Patient no. 24 demonstrated a decrease in the right ventricular systolic pressure from 70 to 40 mm. Hg postoperatively, but the left-to-right shunt increased. Following closure of the atrial septal defect at a later date the right ventricular pressure was reduced to normal with complete obliteration of the systolic pressure gradient between the right ventricle and pulmonary artery (table 2).

Patient no. 25 also demonstrated an increase in the volume of the left-to-right shunt through the defect in the atrial septum following valvuloplasty.

Discussion

Mortality

The 4 largest groups of patients with valvular pulmonary stenosis and intact ventricular septum that have been operated upon by the transventricular approach are those of Campbell and Brock,\(^2\) Hosier,\(^3\) Lillehei,\(^4\) and Silverman.\(^5\) Campbell and Brock have reported a total of 58 patients with 8 deaths (14 per cent). There was a significant difference in the mortality with the presence or absence of cyanosis. In 33 acyanotic patients with valvular pulmonary stenosis there was but 1 death, while in 25 cyanotic patients there were 7 deaths. Hosier and associates have reported on a series of 86 patients of whom 7 died (8 per cent). Three patients died (14 per cent) of 21 reported by Silverman and associates; these 3 were in congestive failure with marked cardiomegaly. The transventricular approach for the relief of pulmonary valvular stenosis has been performed in only 6 patients in our institution with 1 death. Thus, the over-all mortality from the transventricular approach may be considered to be about 10 per cent.

Although the present series is relatively small, it is the largest one that has been reported in which the direct approach has been applied. Thirty-eight patients have been operated upon with 2 deaths, a mortality of 5 per cent. Both patients who died were cyanotic; there have been no deaths in 30 acyanotic patients.

The first patient who died was a 7-month-old infant with a large heart, marked cyanosis, and congestive failure. The peripheral arterial oxygen saturation was 47 per cent and the pressure in the right ventricle was 108/35 mm. Hg. Operation was considered an emergency measure. A severe valvular pulmonary stenosis was found, the orifice being 1.5 mm. in diameter. Operation was without event, the patient returned to her room in good condition. She died suddenly and unexpectedly 12 hours postoperatively. Unfortunately, a postmortem examination was not permitted. The other patient who died was a 10-year-old boy. He was cyanotic and had a peripheral arterial oxygen saturation of 75 per cent and a hematocrit value of 85 per cent. The right ventricular pressure was 175/9 mm. Hg. Severe valvular pulmonary stenosis was found at operation and was relieved with difficulty. Preoperative cardiac catheterization had revealed a defect in the atrial septum and this finding was confirmed by palpation at operation. After relief of the pulmonary stenosis it was decided to repair the atrial septal defect, but before this was completed the patient developed ventricular fibrillation. The heart beat was restored to a regular rhythm but the patient died 5 hours later of a bleeding diathesis.

Although this series is relatively small, it is considered that the operative risk of 5 per cent with the direct approach under hypothermia is certainly not greater and in fact may be less than that of transventricular approach.
Pulmonary Insufficiency

The creation of insufficiency resulting from the excision of valve substance has been criticized. The 3 patients in whom the valve was partially excised in addition to the incision along the commissures, have now been followed for 3½ years. All have definite auscultatory evidence of pulmonary insufficiency. The degree of pulmonary insufficiency is not considered significant, however, as the diastolic pressure in the pulmonary artery is within normal limits in 2 of these patients and is slightly decreased in the remaining 1. Careful evaluation at this time reveals that all 3 are active, leading entirely normal lives with no symptoms whatsoever. There has been no increase in the size of the heart, which is now within normal limits. The electrocardiogram has revealed regression of the evidence of right ventricular hypertrophy in all 3 patients (fig. 1). These findings suggest that the degree of pulmonary insufficiency in these patients is not of hemodynamic significance.

Experimental excision of pulmonary valve cusps in the dog does not result in any noticeable detrimental effects upon the heart; only if all 3 cusps are completely excised does the right ventricle dilate and the right heart fail. The low diastolic pressure gradient normally existing between the pulmonary artery and the right ventricle, which is due to the low pulmonary vascular resistance, apparently results in an insignificant reflux of blood into the right ventricle, when the insufficiency of the pulmonary valve is mild.

However, it is again emphasized that only a minimal amount of actual valve substance was removed in the first 3 patients, and that no valve tissue has been excised in the last 35 patients. In 8 of the latter patients a murmur of pulmonary insufficiency was also audible. The degree of pulmonary insufficiency in these patients is not considered to be of hemodynamic significance, since the diastolic pressure in the pulmonary artery was over 8 mm. Hg in all. The development of a postoperative pulmonary diastolic murmur has also been reported by authors employing the transventricular technique.

Thus, even when no valve tissue is excised, a slight degree of pulmonary insufficiency may occasionally result in some patients, but even when it occurs, it is not considered to be of hemodynamic nor of clinical significance.

Clinical Observations

Seven patients were asymptomatic prior to surgery. Fourteen patients had symptoms of minimal dyspnea and fatigue on exertion. Two patients (no. 21 and 25) were moderately incapacitated, while the remaining 2 patients (no. 18 and 20) were more severely ill. Patient no. 18 was severely cyanotic and was in congestive failure during the month prior to operation; patient no. 20 had spells of severe dyspnea with increase of the cyanosis.

Postoperatively these latter 2 patients have shown great improvement and are now active and without symptoms. All patients who were symptomatic prior to surgery have reported an increase in their exercise tolerance.

It is worth re-emphasizing that symptomatology plays only a minor role in the evaluation of patients for operation, as symptoms occur late in the course of the natural history of this anomaly, and patients with severe stenosis may be relatively asymptomatic.

Electrocardiographic Observations

All patients showed a degree of right ventricular hypertrophy preoperatively. Postoperatively various degrees of regression of the pattern of right ventricular hypertrophy have been observed depending upon the period of observation. All patients who were observed for a period of at least 6 months have revealed a decrease in the height of the R wave in leads V2R, V1, and aV1, and in the depth of the S wave in V6 and V7. In 5 patients the tall upright deflection present in leads V2R and V1 changed to an rSR' pattern. This pattern has been frequently interpreted as indicating incomplete right bundle-branch block. However, a QRS duration of only .08 to .10 second in the presence of an rSR' complex is considered to reflect minimal hypertrophy of the right ventricle mainly in the structures of the outflow tract (figs. 1 and 2). It is expected that
with further regression of the hypertrophy a normal electrocardiographic pattern will develop.

Radiography

There was roentgenologic evidence of significant preoperative cardiac enlargement in only 1 patient in this series (no. 18); this patient was in failure at that time. Although there is enlargement of the right atrium and hypertrophy of the right ventricle, the over-all heart size may be within normal limits despite the presence of severe pulmonary valvular stenosis.

Thus, until dilatation and failure of the heart supervene, the over-all heart size may be within normal limits or only slightly enlarged. There was a reduction in heart size in patient no. 18. There was no significant reduction in heart size in the other patients; to the contrary, a slight increase in over-all heart size following surgery was detected in 10 patients. This increase was minimal and occurred in the immediate postoperative period. The heart size has not increased further in any of these patients, and is considered to be within normal limits at the present time.

Preoperatively the vascularity of the lung fields was considered to be within normal limits in all 15 patients with an intact atrial septum. This observation is in agreement with the hemodynamic observation that the pulmonary blood flow at rest is normal in volume and equal to the peripheral systemic blood flow in these patients despite the stenosis of the pulmonary artery. Postoperatively the vascularity did not change in these patients.

The vascularity of the lung fields appeared decreased in the cyanotic patients; particularly the periphery of the lung fields appeared more translucent than normal. Postoperatively an increase in the pulmonary vascularity was noted in at least 3 of these patients; in the others no definite change has been observed.

Patient no. 24 demonstrated a slightly increased vascularity of the lung fields prior to valvuloplasty. After this operation the vascularity increased considerably; at the present time, however, 20 months following closure of the atrial septal defect, the vascularity of the lung fields is considered to be within normal limits.

Hemodynamic Observations

In 14 patients the preoperative systolic pressure gradient across the pulmonary valve was greater than 100 mm. Hg and in 5 patients it was over 150 mm. Hg. Following surgery the systolic pressure gradient between the right ventricle and pulmonary artery was markedly decreased or completely obliterated in 17 of the 25 patients. We have arbitrarily chosen a gradient of 20 mm. Hg or less as indicative of an ideal result and of the absence of significant residual stenosis. It is with the remaining 8 patients that we are particularly interested. Four of these had residual pressure gradients greater than 30 mm. Hg, the greatest being 59 mm. Hg.

There are several explanations that may be considered in regard to this residual gradient. The first possibility is that the stenosis was inadequately relieved at the time of the surgical procedure. This is considered unlikely for the following reasons. The surgical procedure was performed under direct vision in a bloodless field. The valve was incised from its central aperture down to the annulus, along the lines of the commissures, whenever they were present. When only rudimentary commissures were noted at the base of the valve, the incisions were made from the aperture of the valve to these sites. Following this procedure in all adults and in all children with a pulmonary artery of sufficient size, the surgeon introduced a finger through the valve and palpated the subvalvular area. In patients in whom the size of the pulmonary artery did not allow this maneuver, a surgical clamp was passed through the valve orifice and spread, revealing that the valve had been opened to the limits of the annulus. The subvalvular area was also probed for additional sites of obstruction.

A second consideration is that some valve cusps, being fibroed, were sufficiently rigid to obstruct blood flow although adequate incisions had been made. This possibility exists but is difficult to evaluate.
A third explanation for a residual pressure gradient is the presence of an anatomic infundibular stenosis. At the time of operation in all patients the surgeon explored the subvalvular area with his finger or with an instrument. No evidence of an infundibular stenosis was found; however it remains possible that an infundibular stenosis was present lower in the ventricle.

There is one additional explanation that might account for this residual gradient: marked hypertrophy in the area of the crista supraventricularis and other structures forming the right ventricular outflow tract\footnote{11, 15} may act as an obstruction to the flow of blood into the pulmonary artery. If the residual gradient is chiefly the result of the obstruction offered by the hypertrophied outflow tract, then it is anticipated that this gradient will decrease with regression of the hypertrophy. A definite answer will not be forthcoming until more patients are restudied at longer intervals following surgery. However, in one patient (no. 21) who was catheterized on 2 occasions following surgery, the first time in the immediate postoperative period and the second time a year following surgery, the findings may be interpreted as supporting the hypothesis that the hypertrophied outflow tract of the right ventricle may be the cause of the presence of a residual gradient in some patients (table 2).

This patient (no. 21), was a 13-year-old cyanotic girl in whom a Blalock-Taussig procedure had been performed at the age of 5. This anastomosis thromosed shortly after the operation, and little change was noted in the patient’s clinical condition. At age 8 a transventricular valvulotomy by the Brock technic was performed. Prior to the latter operation the right ventricular pressure was 190/8 mm. Hg, the pressure in the pulmonary artery was 18/10 mm. Hg, and the femoral arterial blood oxygen saturation was 78 per cent. Recatheterization in February 1955 revealed the pressure in the right ventricle to be 120/5 mm. Hg; the femoral arterial blood oxygen saturation was 80 per cent. At the third operation in Denver in June 1955, a long, conical valve with an orifice of 4 mm. in diameter was observed. Three incisions were made down to the valve ring along the lines of the commissures and the surgeon inserted his finger through the valve into the outflow tract of the right ventricle. The outflow tract was narrowed presumably as a result of the severe hypertrophy of the ventricle. No definite sites of infundibular stenosis were considered to be present. The valve leaflets were pliable.

Two weeks later the right ventricular pressure was 60/3 mm. Hg and the pulmonary artery pressure was 28/16 mm. Hg. On withdrawal of the catheter the pressure tracing showed an intermediate pressure zone between the pulmonary artery and the right ventricle (fig. 5). Thus, on withdrawing into the subvalvular area, the systolic pressure remained at the same level as in the pulmonary artery while the diastolic pressure fell to a level of 5 mm. Hg. On continued withdrawal into the lower right ventricle the systolic pressure suddenly rose to 60 mm. Hg. Prior to surgery there was no evidence in the withdrawal pressure tracing of an infundibular chamber. The immediate postoperative electrocardiograms were not significantly different from the preoperative tracings, continuing to reveal marked

\begin{figure}[h]
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\includegraphics[width=0.5\textwidth]{fig5.png}
\caption{Pressure tracing obtained on withdrawing cardiac catheter shortly after the third operation in patient 21, showing an intermediate pressure zone between the pulmonary artery and right ventricle.}
\end{figure}
right ventricular hypertrophy (fig. 2). The peripheral arterial blood oxygen saturation was 73.6 per cent.

Recatheterization in May 1956, 11 months following the last operation, demonstrated a pulmonary artery pressure of 23/13 mm. Hg, a right ventricular pressure of 45/8 mm. Hg, and the continued evidence in the withdrawal pressure tracing of an infundibular chamber with a pressure of 30/8 mm. Hg.* The peripheral arterial oxygen saturation was 91 per cent, which represents a significant increase over the previous values. At this time the electrocardiogram showed a striking change, with evidence of marked regression of the right ventricular hypertrophy (fig. 2).

A second patient (no. 10) also revealed changes in the postoperative withdrawal pressure tracing that suggested the presence of an intermediate pressure zone (fig. 6). Preoperatively the pulmonary artery pressure was 25/13 mm. Hg, with an abrupt change to a right ventricular systolic pressure of 190 mm. Hg. This change occurred high at the level of the pulmonary valve. The peripheral arterial saturation was 93.2 per cent. At operation the valve appeared conical with an orifice 4 to 5 mm. in diameter. The lines of the fused commissures were visible and the valve was incised along these 3 lines to the annulus. Recatheterization 10 days postoperatively revealed a right ventricular pressure of 85/10 mm. Hg and a pulmonary arterial pressure of 28/10 mm. Hg. The withdrawal pressure tracing at this time revealed a pressure tracing suggestive of an intermediate chamber (fig. 6); in the immediate subvalvular area there was a slight rise in the systolic pressure and a fall of the diastolic pressure to zero; on further withdrawal there was an additional abrupt rise in the systolic pressure level to 85 to 90 mm. Hg.

Although both patients (no. 10 and 21) were not considered to have an anatomic infundibular stenosis preoperatively and showed an abrupt transition in their withdrawal tracings without evidence of an intermediate pressure

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* We are greatly indebted to Professor P. Soulié for his kindness in sending to us the results of his studies on this patient and for his permission to include these data in this paper.
zone, their postoperative pressure records have revealed a wave form commonly considered suggestive of infundibular stenosis. The explanation is offered that preoperatively these patients had 2 sites of obstruction in series. The distal one, being the more severe, obscured the presence of the less significant proximal obstruction. Following removal of the distal obstruction of the valvular stenosis, however, the proximal obstruction due to the hypertrophied outflow tract became manifest. The pressure tracing noted in these 2 patients postoperatively is considered to reflect obstruction to blood flow by the structures of the hypertrophied outflow tract of the right ventricle, which developed as a result of the valvular stenosis.

A true anatomic infundibular stenosis must of course be considered in some cases. However this does not appear to be the case in patient no. 21 in view of the continued fall in the right ventricle systolic pressure level attending the regression of the right ventricular hypertrophy.

Postoperative Pulmonary Hypertension

Slight to moderate pulmonary hypertension was encountered postoperatively in 6 patients (no. 8, 11, 12, in table 1; and 19, 23, and 25, in table 2), although the pulmonary artery pressures were normal preoperatively. Exercise resulted in an additional increase in the pulmonary artery pressure in patient no. 8 (to 64/29 mm. Hg) and in patient no. 12 (to 59/43 mm. Hg). The pulmonary artery pressure during exercise was not recorded postoperatively in the remaining 4 patients. In one additional patient pulmonary hypertension was manifest only during exercise.

The explanation for the development of the postoperative hypertension in these patients is not evident. It might be considered to reflect an increase in the pulmonary artery blood flow in the face of minimal structural changes in the pulmonary vessels. However, there is no definite evidence to support this supposition. The pulmonary artery blood flow at rest has not increased significantly in any of these patients, and, as to the possibility of structural changes, no pulmonary biopsy specimens were obtained.

Multiple thromboses with recanalization have been observed by Rich in the pulmonary vessels of patients with a tetralogy of Fallot. Recently this subject has been reviewed by Dammann and Ferencz and it was found that the small pulmonary arteries in patients with pulmonary stenosis were thin walled with wide lumens and could not be distinguished from those found in normal lungs. Lung sections obtained from patients of an older age group, however, frequently revealed pulmonary arteries containing thrombi similar to those described by Rich. These changes were thought to be the results of polycythemia, hypoxia, and inadequate pulmonary blood flow due to the pulmonary stenosis. However none of the patients with postoperative pulmonary hypertension was polycythemic; slight peripheral arterial oxygen unsaturation was present in only 3 (patients 19, 23, and 25, table 2), while the pulmonary arterial oxygen saturation varied between 52 and 72 per cent. Only the 1 patient (no. 6) with a normal resting pulmonary artery pressure and a rise above normal during exercise demonstrated a very low (38 per cent) oxygen saturation of the pulmonary artery blood. Therefore it seems unlikely that the postoperative hypertension was the result of hypoxia.

A definite explanation for this postoperative phenomenon must therefore await further investigation.

Evaluation of Surgical Therapy

Patients with valvular pulmonary stenosis have been divided into 2 groups on the basis of the presence or absence of an associated defect in the atrial septum.

We are satisfied at the present time that the procedure of choice in patients with an intact atrial septum is the direct approach to the stenotic valve through an incision in the pulmonary artery. However, in the patients in whom there is an associated defect in the atrial septum, the surgical approach is not so evident. In the patients with a left-to-right shunt through the defect, it is considered that the atrial septal defect should be closed first, and the pulmonary stenosis should be relieved at the same operation if the condition of the pa-
tient permits. Should the condition of the patient not warrant it, then pulmonary valvuloplasty can be performed at a later date.

The reason for this sequence has been demonstrated by the course of patient no. 24, in whom removal of the pulmonary stenosis was followed by increased left-to-right shunt and congestive failure. This patient slowly responded to medical therapy and closure of the atrial septal defect was accomplished 6 months later. At the present time her heart size has decreased to normal and she is asymptomatic.

The problem of the surgical approach is different in patients with a right-to-left shunt through the defect in the atrial septum. If the defect is small and due to a foramen ovale, closure may occur following surgery. The relief of the pulmonary stenosis and subsequent regression of the hypertrophy of the right ventricle will result in a decrease in the right atrial pressure. The re-establishment of the normal relationship between the right and left atrial pressures then might result in closure of the foramen ovale. This apparently has occurred in 2 patients, (no. 17 and 21) in whom there has been a marked regression of the hypertrophy of the right ventricle, as reflected by the electrocardiogram and the complete or nearly complete obliteration of the right-to-left shunt. When the flow of blood is from right to left through the defect in the atrial septum, then it is considered that the stenosis of the pulmonary valve should be relieved first in contrast to the order of repair in patients with a left-to-right shunt. The indications for closing the defect in the atrial septum at the time of valvuloplasty are not clearly established. At the present time it is our plan to open the right atrium following relief of the pulmonary stenosis, and if the defect in the atrial septum appears to be of considerable size and if the patient’s condition permits, the defect will be closed. However, should the defect feel small and if there is considered to be a velum, such as to suggest dilatation of a foramen ovale, then further surgery will be postponed.

**Summary**

Clinical and physiologic studies have been carried out in 25 patients following surgical correction of congenital valvular pulmonary stenosis. The patients were operated upon under conditions of hypothermia and circulatory occlusion, permitting plastic repair of the stenotic pulmonary valve with unimpaired vision and a dry operative field. The results are considered to be superior to those obtained with the blind transventricular approach. The operative mortality certainly compares favorably with the transventricular approach. Auscultatory evidence of insufficiency of the pulmonary valve is occasionally noted; however, it is emphasized that no valvular substance has been excised in the last 35 patients. The first 3 patients, who have been previously reported, have now been followed for a period of 3½ years and there has been no clinical nor physiologic evidence that this minimal degree of pulmonary insufficiency is significant.

The systolic pressure gradient between the right ventricle and the pulmonary artery has been completely abolished in 17 of the 25 patients. A residual pressure gradient of more than 20 mm. Hg was present in 8 patients. Possible explanations for the residual pressure gradient are presented.

Finally, the recording of pulmonary hypertension has been reported following relief of the pulmonary stenosis and explanations for this are considered.

**Summario in Interlingua**

Studios clinic e physiologic ha essite executate in 25 patientes post correction chirurgic de congenite stenosis del valvula pulmonar. Le operationes essave effectuate sub conditiones de hypothermia e occlusion circulatori de manera que le reparo del stenotic valvula pulmonar poteva facer se con visualisation complete in un non-inundate campo chirurgic. Le resultatos obtenite es considerate como superior a illos possibile per le occulte methodo transventricular. Isto es clarmente evidente ab un comparation del procentages de mortalitate. Indicios auscultatori de insufficientia del valvula pulmonar es a vices a notar, sed le autores sublinea le facto que nulle excision de substantia valvular essave effectuate in le ultime 35 patientes de lor serie. Le prime 3 pa-
tientes, reportate in un previe publication, ha nunc esiste sub observation post-operatori durante un periodo de 3½ annos, e nulle indication clinic o physiologic existe a suggerer que iste grado minimal de insufficientia pulmonar es significative.

Le gradiente de pression systolic inter le ventriculo dextere e le arteria pulmonar esseva completamente abolite in 17 del 35 patientes. Un residuo de gradient de plus que 20 mm Hg persisteva in 8 casos. Es presentate explicationes possibile del residue gradient de pression.

Es reportate, finalmente, le registration de hypertension pulmonar post alleviamento del stenosis pulmonar. Explicationes es considerate.

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Science increases our power in proportion as it lowers our pride.—Claude Bernard, 1813–1878.
Valvular Pulmonary Stenosis with Intact Ventricular Septum: Clinical and Physiologic Response to Open Valvuloplasty
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