Dr. Engle: When pulmonic stenosis occurs as an isolated congenital malformation of the heart, it usually is due to fusion of the valve cusps into a dome with a small hole in the center. In Figure 1 the pulmonary artery has been laid open so that one can see the three leaflets of the pulmonary valve are completely fused, and that there is only a small, central, pinpoint opening which permits blood to leave the right ventricle and enter the pulmonary circulation.

Valvular pulmonic stenosis is much more common than subvalvular or infundibular stenosis, where the obstruction to pulmonary blood flow lies within the substance of the right ventricle. There it may be due to a diaphragm of tissue which obstructs the outflow of the right ventricle, or to an elongated narrow tunnel lined with thickened endocardium, or to a ridge of fibrous or muscular tissue just beneath the pulmonary valve.

The changes in the cardiovascular system which result from obstructed pulmonary blood flow are so characteristic that they permit the ready recognition of this condition. Proximal to the constriction, these changes manifest the burden placed on the right ventricle, which enlarges and hypertrophies. On physical examination this is demonstrated by the precordial bulge and tapping impulse just to the left of the sternum, where the rib cage overlies the anterior (right) ventricle. Radiographically, both by fluoroscopy and in roentgenograms in the frontal and both oblique views, right ventricular enlargement is seen. In the electrocardiogram, the precordial leads show a pattern of right ventricular hypertrophy. On cardiac catheterization this burden is evidenced by an elevated pressure within the right ventricle, in contrast to the normal or low pressure in the pulmonary artery beyond the stenosis. In an angiocardiogram, a big right ventricle which is slow in emptying reflects the burden on the heart proximal to the obstruction.

At the level of the obstruction the physical sign is a long, smooth, systolic murmur, which in the presence of valvular pulmonic stenosis tends to be heard high up in the first and second intercostal spaces to the left of the sternum, but with infundibular stenosis is heard more often in the third and fourth intercostal spaces. Also, on auscultation there is a diminished pulmonary second sound, which reflects the low pressure, or low flow, at this level.

Distal to the obstruction, the signs of the slowed or reduced pulmonary flow are chiefly radiographic: excessively clear peripheral lung fields and diminished pulsations of the right and left branches of the pulmonary artery. On cardiac catheterization there is a low pressure in the pulmonary artery, and in angiocardograms the depleted circulation through the periphery of the lung may be evident.

Whatever the site of the stenosis, these changes occur. Additional features help to localize the point of narrowing. This is of more than academic interest, because for one site (valvular stenosis) the correction is relatively simple and quite effective, whereas for stenosis occurring below the valve, surgery is much more difficult.

In the differentiation of valvular from subvalvular stenosis, the location of the murmur is helpful, and one characteristic feature of the roentgenogram is important. When the obstruction occurs at the pulmonary valve, one sees poststenotic dilatation of the main pulmonary artery, as illustrated in Figure 2. In contrast, in infundibular stenosis there is a concave main pulmonary artery segment and a scooped-out appearance of the right ventricular outflow tract, as seen in Figure 3. The right auricular and ventricular enlargement, the small right branch of the pulmonary
artery, and the clear peripheral lung fields are evident in both figures.

By cardiac catheterization, differentiation of valvular from subvalvular stenosis may be possible by passage of a catheter into the pulmonary artery and recording of pressures as the catheter is withdrawn into the ventricle. The area of obstruction is located at the site where the pressure changes from the low pressure beyond the stenosis to a high pressure proximal to the stenosis. And the angiocardiogram may help because it can show a jet of opaque material going through the conical valve, or it may outline the infundibular narrowing. Later, two patients who were recently subjected to surgery and who illustrate some of these features will be presented.

One may remember that not too many years ago the condition of isolated pulmonary stenosis was considered rare, and diagnosis during life was thought to be impossible. Then it became recognized in its more severe form, and finally with the advent and widespread use of cardiac catheterization in people who had cardiac murmurs, many were found to have mild or moderate degrees of pulmonary stenosis. It is now realized that this is quite a common lesion; indeed, it is one of the most common malformations of the heart.

We think of it as a progressive condition, one where the severity of the stenosis increases as the patient becomes older. It is the severity of the stenosis that determines the symptoms and the degree of cardiac enlargement and other abnormal signs which the patient presents.

One aspect not yet mentioned is the factor determining appearance of cyanosis in pulmonic stenosis. When the cardiac septa are intact, there is no true oxygen unsaturation of the arterial blood. But if there is a patent foramen ovale, this opening may be stretched so that some blood may pass from the right auricle (which now has higher than normal pressure) over into the left auricle. This permits venous blood to enter the systemic circu-
lation and causes the cyanosis some of these patients present. Less often a true defect of the auricular septum may function in the same way in the presence of severe stenosis.

The patients to be presented were operated on about 9 months ago by a new technique in surgery—pulmonary valvulotomy under direct vision with the aid of hypothermia.

The first approach to surgery of this condition was in 1948. Two British surgeons inde-
pendently devised the technique of opening the right ventricle of the heart and introducing a cutting instrument up through the valve into the pulmonary artery and withdrawing it along the same route. Then a dilator could be pushed through, to be sure that the opening was adequate and the stenosis relieved.

This technique was quite widely adopted. Many patients were subjected to this procedure, and many were benefited. However, as data on the postoperative course has accumulated, it has become clear that what this procedure did was to convert a severe degree of stenosis (which produced symptoms) into a milder degree of stenosis (with few or no symptoms). The right ventricular pressure has usually been reduced by half as a result of this operation, and although a reduction from 200 to 100 mm of mercury, for example, is quite an achievement, the lower figure is still four or five times the normal. So it became desirable to find a better procedure—one that would avoid the right ventricle and the blind approach to the stenosis, if possible.

The anterior location of the pulmonary artery and, in the case of valvular stenosis, its dilatation, make possible a different access to the pulmonary valve. As can be seen in Figure 1, if the surgeon could open the pulmonary artery and under direct vision could see the fused cusps and cut them as did the pathologist in the specimen, as well as along two other lines to make three leaflets as in the normal tricuspid pulmonary valve, then there would be a much greater chance of restoring function of the valve and the heart to normal.

With the advent of hypothermia this became possible. As the temperature of the body is reduced, the circulation can be interrupted for brief periods of time, sufficient to allow visualization of the pulmonary valve and cutting of the fused cusps. This technique was pioneered by Dr. Henry Swan. The results from the combined procedure of moderate hypothermia and direct-vision correction of the stenosis of the pulmonary valve have, in our experience, been quite gratifying.

The first patient to be presented was seen at the age of 12 months with congestive heart failure. A cardiac murmur was heard at birth. She had been cyanotic from birth, with episodes of increased cyanosis whenever she cried, and a great deal of breathlessness on minor exertion.

The findings were characteristic of severe stenosis of the pulmonary valve with a patent foramen ovale. The electrocardiogram showed marked right ventricular hypertrophy and “strain.” Roentgenograms of the chest (Fig. 4) revealed increased size of the heart, affecting the right auricle and right ventricle, and excessively clear lung fields. The enlargement of the heart prevents one from seeing the left and right pulmonary arteries in the frontal view.

This baby was treated in the usual manner for heart failure, and 1 month later, in January, 1956, was operated upon by Dr. Frank Glenn and Dr. George Holswade. The body
The temperature was reduced to 30°C and during two 3-minute periods of occlusion of the circulation, a cone-shaped valve with a tiny central opening was grasped and cuts were made which extended all the way to the valve ring.

Pressure determinations in the operating room showed a decrease in right ventricular pressure from over 200 mm of mercury before the valvulotomy, to 50 mm after the valve was opened, and a rise in the pulmonary artery pressure from 15 to about 30 mm of mercury.

This patient could not walk prior to surgery. She was just able to sit up, and even that made her breathless, yet within 2 weeks after the operation she was not only sitting and standing, but walking. Her color became perfectly pink. The clubbing that was evident in the fingers and toes disappeared. The mother stated that her general activity had become normal and the cyanosis had disappeared.

There were two other evidences of improvement. One is a recent roentgenogram of the chest which Dr. Baker will show (Fig. 5).

Dr. Baker: It can be seen quite clearly, compared with Figure 4, that the lung fields are no longer overly clear. The pulmonary radicals are somewhat less conspicuous than usual, but much more within normal limits. A shadow is seen in the left superior mediastinum with a sharp lip on it, and this represents the thymus gland; the actual cardiac silhouette emerges beneath the thymic shadow and the prominent bulge of the main pulmonary artery has now disappeared.

Dr. Engle: I don't believe Dr. Baker mentioned the cardiothoracic ratio is normal now; preoperatively it was 60% and now is 45%.

The change in the electrocardiogram is shown in Figure 6, and is just as dramatic. Noteworthy are the initial marked right axis deviation, the high peaked P waves which reflect the right auricular enlargement and, in the precordial leads, the signs of right ventricular hypertrophy. The V-leads, 2-6, had to be taken at half-standardization because the amplitude of the deflections was so great. One can see the markedly tall R wave over the right ventricle and, in contrast, over the left ventricle where one would expect a tall R wave and a small S wave, just the reverse.

Beyond this evidence of right ventricular hypertrophy, there is evidence of "strain" in the T waves and the ST segments over the right ventricle. This implies a very high pressure in the right ventricle. However, in the electrocardiogram taken 6 months postoperatively, there no longer is right axis deviation. The P waves are normal in height. The T waves and the ST segments are normal throughout. The tall R wave in V-1 is now gone. Instead, there is only a slight conduction disturbance over the right ventricle, which may be found in normal children. The deep S wave has disappeared in V-5 and V-6, and there is now normal dominance of the left ventricle.

The next patient demonstrates some of the problems in differentiating valvular from sub-valvular stenosis, but also emphasizes one form of obstruction to the outflow-tract which can be helped by this type of surgery. The little girl was born in this hospital and was cyanotic for the first 3 months of life. There was a loud systolic cardiac murmur and enlargement involving the right side of the heart.

![Figure 6](http://pediatrics.aappublications.org/)
After the age of 3 months, cyanosis disappeared, cardiac enlargement remained stationary, and for the first 4 years she did rather well in the limited sphere of activities which is normal for that age.

However, once she became older it was evident that she could not keep up with other children. She could not run or skip. She could not climb a flight of stairs. As we followed her we noted progression of cardiac enlargement and the development of an electrocardiographic pattern of right ventricular "strain" in addition to that of right ventricular hypertrophy.

In a roentgenogram of the chest (Fig. 7), one can see the degree of cardiac enlargement, the remarkably clear peripheral lung fields, and the right auricular and right ventricular enlargement. Beneath the aorta one notes a smaller pulmonary artery than in the previous patient.

Furthermore, the murmur in this patient was not localized high up in the pulmonary area, but extended farther down the left sternal border. So we were worried about the possibility of infundibular obstruction in addition to stenosis of the pulmonary valve.

At cardiac catheterization the pressure in the pulmonary artery was low (16/7 mm) and the right ventricular pressure elevated (178/7 mmHg). The change in pressure was interpreted to occur at the pulmonary valve. No evidence of infundibular involvement was detected. Arterial oxygen saturation was normal and there was no shunt.

In January, 1956, surgery was performed at 5½ years of age under hypothermia. A cone-shaped, stenotic valve was seen and incisions were made to the valve ring under direct vision. When the pressure was measured in the operating room after the valvuloplasty, it was found to our great dismay that the right
ventricular pressure was reduced only from 170 to 130 mm of Hg. This meant that there was still obstruction to the flow of blood. Pressures were recorded at various levels in the outflow-tract of the right ventricle, and an area was found just proximal to the valve where the systolic pressure was 40 mm of Hg. Thus, there was infundibular stenosis as well.

With hypothermia it is unwise to open the right ventricle to resect any obstruction there, because of the danger of ventricular fibrillation, which may be uncontrollable. So it was decided not to proceed further, and to see what benefits there would be from relief of the valvular stenosis. If another operation were needed to remove the remaining obstruction, it would be done with the aid of a heart-lung machine.

A very gratifying development occurred—the apparent resolution of the infundibular stenosis over a 6-month period. Before operation she was quite a limited little girl. Since then she has been to camp during the summer, and she goes to school now, where she plays as actively as her friends. The cardiac murmur, as in the other child, has become subdued, and she too had an excellent result from the surgery.

What has happened to this patient postoperatively has surprised and pleased us all, and I think it can best be demonstrated by tracings from serial electrocardiograms (Fig. 8). The higher the R wave over the right ventricle, the higher the right ventricular pressure. These tracings show the progressive decrease in the size and duration of this wave. The unipolar precordial leads were taken 1 week preoperatively and in the postoperative period at 2 weeks, 6 weeks, 3 months, and 6 months. In the preoperative tracing can be seen the very tall R wave in V-1 of about 39 mm, and the evidence of right ventricular “strain” in the right precordial leads. V-2 to V-6 were taken at half-standardization, because the deflections were so large.

Postoperatively there was an immediate reduction of the R wave in V-1, but the T wave abnormalities persisted. At 6 weeks postoperatively there was a more marked decrease in the amplitude of the R wave, to about half of the preoperative value, and perhaps less negativity of the T waves as compared to the preoperative record. At 3 months postoperatively the right ventricular “strain” pattern was gone. T waves were upright as they should be, but the R wave stayed the same. In the period of time between 3 months and 6 months postoperatively, the pattern of right ventricular hypertrophy disappeared. There is now normal dominance of the left ventricle, with no evidence of right ventricular “strain” or hypertrophy. She too has a slight conduction disturbance over the right ventricle.

Our interpretation of the improvement in this child is that the great thickness of the walls of the hypertrophied right ventricle caused some obstruction during systole to the free flow of blood through the slit valve into the pulmonary artery. With relief of the tight obstruction at the valve and the reduction of the work-load of the right ventricle, the hypertrophy lessened. As it did, the opening at the most distal part of the outflow-tract of the right ventricle became progressively wider during ventricular systole. With continued decrease in thickness of the ventricular wall there followed a melting away of the subvalvular obstruction.

Cardiac catheterization is planned for both of these children at 1 year postoperative, but with the data already available and particularly with respect to the return to normal limits of the electrocardiogram, one may confidently expect that the right ventricular pressure will be at normal or near normal level and the gradient across the valve will be minimal or abolished.

It is our present belief that when patients present with signs of stenosis of the pulmonary valve and have cyanosis, or a pattern of right ventricular “strain” in the electrocardiogram, or progressive cardiac enlargement or increasing symptoms, that they should be operated upon by this technique. The right ventricular pressure at this time will usually exceed 100 mm of mercury. If one can demonstrate that the stenosis is purely subvalvular, then I think operation should be deferred until it can be done as an open procedure with the aid of a heart-lung machine. There are some patients where this differentiation is difficult, particularly when the infundibular stenosis occurs just beneath the valvular stenosis, as in the second girl. If one encounters this situation in the operating room it may be of some comfort to know that, in some instances at least, the infundibular narrowing may be due to the thickness of the right ventricular mus-
culature, and that it can disappear after the relief of the valvular stenosis, when the two coexist.

Dr. Levine: Thank you, Dr. Engle. Are there any questions?

Question: Do these patients develop valvular insufficiency?

Dr. Engle: A murmur of pulmonary insufficiency has been heard in a small number of patients following blind valvulotomy and also after the open technique. If part of the pulmonary valve is resected at the time that the stenosis is relieved, then insufficiency is likely to develop. However, it seems to us unreasonable to remove a section of pulmonary valve. So far, in our patients, there has been no evidence of pulmonary insufficiency. If it should occur, the one consoling factor is that this is a low pressure system on the right side of the heart, and probably would not produce symptoms, as would an aortic insufficiency on the left side of the heart.

Question: Is there an optimum age for this procedure?
DR. ENGLE: Hypothermia is tolerated better in the pediatric age period than it is by the adult. I think there is no optimal time for the valvulotomy itself, based on present knowledge. Instead, the procedure is done on indication. Probably as its safety is established and the beneficial results are shown to be lasting, these patients will be subjected to surgery before signs and symptoms become as marked as in patients seen at present.

QUESTION: Is there any danger of untoward effect as a result of this length of the period of hypothermia?

DR. ENGLE: Only a moderate degree of hypothermia is needed in this particular operation because only a short period of interruption of blood flow, for about 3 or 4 minutes, is needed. With hypothermia of 30°C a period of occlusion up to 8 or 10 minutes is within safe limits, so that there is a relatively wide margin of safety.

As soon as the patient’s temperature has reached 30°C—and the cooling is planned so that the surgeon is ready to occlude the circulation at the time this occurs—rewarming is begun and within 30 minutes he is getting warm again. This depth or duration of hypothermia has not produced any untoward effects, in our experience.

QUESTION: How do you manage the patients with infundibular stenosis?

DR. ENGLE: Fortunately, infundibular stenosis is so much rarer than valvular stenosis that the problem does not happen too often. If the patient has the indications for surgery that were just mentioned, such as cyanosis or right ventricular “strain,” what we would do at the present time is refer him to a surgical unit that has available a heart-lung machine that would permit opening the right ventricle to relieve the obstruction. Blind approaches have been unsatisfactory, but the more widespread availability of heart-lung machines should make it possible within a few years for these patients who are worries to us now to be helped.
CLINICAL CONFERENCE: Pulmonic Stenosis
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