The Fate of the Patient with Pulmonic Stenosis

By Mary Allen Engle, M.D., Tomiko Ito, M.D., and Henry P. Goldberg, M.D.

The outlook for the patient with pulmonic stenosis is determined by the severity of the lesion and his age at the time of examination. Regardless of location of the stenosis, whether in the valve as it usually is, or beneath or above the valve as sometimes happens, there is much variation in degree of obstruction. In some, the stenosis is so marked that death in heart failure occurs in the first hours of life. In others the narrowing is less, and the patient may live beyond the average life expectancy.1-3

To judge the effect of age and severity of obstruction, one needs to understand the natural history of the condition. One may attempt to get this information from perusing the literature. A drawback to this approach is that often it is the exceptional case that is reported, or a large series, where individual details must merge into a general picture. Another approach is to examine a current population, noting how many are living at each age period and the condition of each. A third method is to follow individual patients over a long period, noting any changes. These last two technics have been used in the present study in an attempt to add to the knowledge of the fate of the patient with pulmonic stenosis.

The next problem is how to gauge severity of stenosis so that the same criteria can be applied to those who first came under observation in 1940, for example, as those examined today.

Symptoms were noted in all the records. However, even among patients with equally marked obstruction to pulmonary blood flow, there may be differences in expression of cardiac disability. Symptoms are often difficult to evaluate in the patient born with an anomaly who has never known what it is to feel normal. In an individual patient, the severity of stenosis may increase, or it may remain the same while new symptoms appear. Exertional dyspnea and fatigue usually imply that stenosis is severe.

Certain signs, if present, point to marked stenosis, but their absence does not preclude the existence of severe stenosis. These include radiographic evidence of right ventricular enlargement and certain physical findings. The sound of pulmonic valve closure is diminished and delayed as severity of obstruction increases. Cyanosis may appear if the foramen ovale is functionally patent.

The stenosis may best be assessed by measurement of cardiac output and mean systolic pressure gradient at rest and during exercise. While it is not practicable to evaluate the course of each patient over a period of years by means of serial cardiac catheterizations, particularly for those who first came under observation before this test was widely used, another tool that was readily available could be used instead. The correlation of electrocardiographic changes with right ventricular systolic pressures in simple pulmonic stenosis is sufficiently close4,5 that they were used as a guide, in combination with the symptoms and signs mentioned, to estimate the severity of stenosis.

If the patient's electrocardiogram was the same each time as it was at catheterization, his course was drawn as though the level of systolic pressure in the right ventricle remained the same. If, on the other hand, the electrocardiograms showed a changing pattern of right ventricular hypertrophy, the systolic pressure in the right ventricle was estimated for each record. This judgment was

From the Department of Pediatrics, The New York Hospital-Cornell University Medical Center, New York, New York.

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made in accordance with the results of a previous study on the electrocardiographic evaluation of pulmonic stenosis. The study indicated that analysis of multiple features of the electrocardiogram gave a more accurate assessment of severity of stenosis than did any single one. In general, when the systolic pressure in the right ventricle was below 80 mm. Hg, the electrocardiogram was normal or showed a pattern of incomplete right bundle-branch block with an R' of 10 mm. or less. With right ventricular pressure above 80, the R or R’ in V1 exceeded 10 mm. and increased in amplitude with increasing pressure. Usually the normal R and S relationship in precordial leads was reversed when the pressure was greater than 100 mm. Hg. When stenosis was severe, additional abnormalities appeared: depression of the ST segment and inversion of the T waves in right precordial leads, leftward displacement of the transitional zone, and signs of right atrial enlargement in the P wave.

Observations

Included in this study are 100 patients with pulmonic stenosis and intact ventricular septum, selected on the basis that severity of stenosis has been demonstrated by measurement of pressures and that all have been examined within the last 5 years. Most are still in regular attendance at the hospital.

They may be considered representative of present-day patients with pulmonic stenosis under medical supervision, for The New York Hospital is not primarily a heart hospital or children's hospital, but is a 1,000-bed general hospital. The series is weighted to the extent that only those having pressure measurements at cardiac catheterization or surgery have been included. Therefore, some patients (chiefly young children with clinical evidence of only mild pulmonic stenosis) have been omitted. The stenosis was proved at operation to be infundibular in seven. In four others the mild obstruction seems to be infundibular. The remainder have valvular pulmonic stenosis.

Fifty-eight of these patients had serial electrocardiograms and were under observation and without surgical intervention for periods up to 27 years. Eleven were observed for longer than 10 years. These comprise the group for “long-term” analysis.

Cardiac catheterization was performed at varying intervals in the course. Figures 1, 3, and 5 show the age of the patient when pressures were recorded. Dots represent those patients with long-term observations. Figures 2, 4, and 6 depict the course from first recorded electrocardiogram to the most recent one. The age of the patient when each record was obtained is indicated by a vertical mark. Some continued under observation after the catheterization. For those undergoing operation, only the preoperative course is shown. Postoperative changes are discussed later.

Table 1 illustrates the distribution of patients according to severity of stenosis at the time pressures were measured. Mild pulmonic stenosis was defined as a pressure gradient between right ventricle and pulmonary artery of 10 mm. Hg or more in patients with a right ventricular systolic pressure at rest of less than 70 mm. Hg. The stenosis was considered moderate if the systolic pressure in the right ventricle was between 70 and 130 mm. Hg, and severe if higher than 130 mm.

The 39 patients with mild stenosis ranged

<table>
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<th>Right ventricular systolic pressure (mm. Hg)</th>
<th>No. of patients</th>
<th>Long-term follow-up</th>
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<tr>
<td>Mild</td>
<td>-70</td>
<td>39</td>
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<tr>
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<td>131-200+</td>
<td>36</td>
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<td>Total</td>
<td>100</td>
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MILD PULMONIC STENOSIS

Age range of patients with mild stenosis at time of measurement of right ventricular systolic pressures. In this figure, and in figures 3 and 5, open circles indicate those with long-term follow-up; closed circles show those with no previous data or those whose course after catheterization was altered by surgery.

Figure 2

Long-term observations on 21 patients with mild stenosis. The pressure scale, 0 to 100 mm. Hg is expanded for this group, in comparison to figures 4 and 6, where the scale is 0 to 200 mm. Hg. In this figure, as in figures 4 and 6, the ages at which electrocardiograms were obtained before and after catheterization are indicated by vertical marks.

in age up to 54 years (fig. 1). They were free from symptoms on unrestricted activity. They had no cardiac enlargement although the main pulmonary artery was often strikingly prominent, so much so that the oldest patient was referred because of the question of a mediastinal tumor. Observations on 21 patients (fig. 2) indicate that the stenosis in them remained mild. However, none of these has as yet reached adult life. For the eight adults who had mild stenosis, it may be presumed that the obstruction had been insignificant throughout their earlier years. For the group who have been followed only through childhood or early adolescence, it may be hoped that the stenosis will continue to be mild, but they will need a longer period of observation to determine this point. Review of the course of those with more significant obstruction at the time of cardiac catheterization indicates that two of them had only mild stenosis in early childhood.

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The severity of obstruction apparently did increase as they grew, for the electrocardiographic expression of the right ventricular burden showed progression of hypertrophy.

While cardiac disability and congestive heart failure posed no threat to this group of patients with mild stenosis, subacute bacterial endocarditis did occur in one child. Remarkably, her obstruction diminished after the endocarditis. Coincident with the infection at the age of 10 years, a murmur of pulmonic insufficiency developed and has persisted. Over the months following recovery, her electrocardiogram showed the same pattern of regression of hypertrophy that is observed following successful surgery,6 and two years later cardiac catheterization demonstrated only slightly elevated right ventricular pressure.

Moderate stenosis was defined as a right ventricular systolic pressure between 70 and 130 mm. Hg. These 35 patients ranged in age up to 49 years (fig. 3). Age distribution was similar to that with mild obstruction (fig. 1). Though unusual exertion was often curtailed by parents or physicians, the patients experienced no symptoms on ordinary activity. One adult was a truck driver who lifted heavy loads, and one woman golfer took part in tournaments until age 47, when she began to tire easily. Three women went through normal pregnancies without compli-
SEVERE PULMONIC STENOSIS

Figure 5
Pressure and age at study of patients with severe and extreme stenosis. Age distribution is comparable to patients with less severe stenosis.

Figure 6
Long-term observations of 21 patients with moderate and extreme stenosis. Note increase in severity, especially during childhood years, in several.

cations; two in their early twenties, and the third in her mid-thirties. None showed signs of cardiac failure, and none were cyanotic. One young adult was treated successfully for subacute bacterial endocarditis. The heart size was average to slightly increased; marked enlargement was not seen.

Examination of serial electrocardiograms of 16 patients with moderate stenosis (fig. 4) indicated no change in 10 and a pattern of increasing severity in six children.

Thirty-six patients had severe stenosis with a right ventricular systolic pressure over 130 mm. Hg. The six patients with pressure in excess of 200 mm. Hg were considered to have exceptionally marked stenosis (fig. 5). The age distribution was the same as that for patients with less marked obstruction (figs. 1 and 3). The oldest patient was 51 years old, and two others were in their forties. One of these had a right ventricular pressure of 197/9 mm. Hg when first seen at age 45. She had undergone two uneventful pregnancies when she was 18 and 20 years old. A recent increase in fatigue on exertion brought her to the hospital.

Most of those with severe obstruction were tired and dyspneic after walking fast or climbing stairs, and they preferred quiet or sedentary activities. Nevertheless, even
among those with high-grade obstruction, some did not recognize or acknowledge symptoms. The parents of a 5-year-old boy with a right ventricular pressure of 220/13 and pulmonary artery pressure of 15/9 mm. Hg could not understand why we recommended an operation for their son, who seemed healthy to them. Eight patients were cyanotic. Congestive heart failure occurred in seven patients at the ages of 3 and 11 months, and 3, 4, 14, 21, and 43 years. Although more patients with severe stenosis showed greater cardiac enlargement than those with lesser obstruction, even so, the heart size by roentgenogram was in normal limits in several, including a 27-year-old woman with a pressure of 197/9 mm. Hg.

A girl with extreme infundibular pulmonic stenosis recovered from subacute bacterial endocarditis at age 10. Interestingly, though her obstruction was subvalvular, a murmur of pulmonic regurgitation developed temporarily during the illness. Four other patients with severe valvular pulmonic stenosis were treated successfully for subacute bacterial endocarditis at the ages of 5, 8, 18, and 26 years.

Of the 21 patients in the group with severe stenosis and several examinations (fig. 6), the electrocardiograms of 10 changed over the years. In three young children with moderate obstruction and in one with only mild stenosis, judged by the first tracings, electrocardiographic evidence of increasing right ventricular hypertrophy developed over a period of a few months or years. In six with findings of severe obstruction when first seen, the electrocardiograms showed an even more marked hypertrophy pattern during childhood in four and in adolescent or adult life in two others.

Effect of Operation

Open-heart surgery has dramatically changed the fate of the patient with severe and with moderate stenosis. It was performed on all those with severe stenosis and on 24

### Table 2

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* Infundibular pulmonic stenosis

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patients with moderate stenosis. Table 2 indicates the improvement achieved by open valvotomy in the first six patients who were recatheterized after they were operated upon under hypothermia. Table 3 shows similar results in the first eight recatheterized patients undergoing open-heart surgery with use of the pump oxygenator. Although the risk of operation was greater in those with unusually severe stenosis, the surgical relief of obstruction was just as complete as in those with more moderate stenosis. Three deaths occurred among the six patients with a right ventricular systolic pressure over 200 mm Hg and three among the other 30 with severe stenosis, but none in the 24 patients with moderate stenosis. There were no deaths from other causes in these 100 patients. Although the longest postoperative follow-up in those undergoing open-heart surgery is only 8 years, all patients have maintained improvement commensurate with the hemodynamic changes delineated by recatheterization 1 or 2 years after the operation. There has been no instance of re-stenosis. Bacterial endocarditis has not occurred in these postoperative patients, but this threat has not been eliminated by surgery. We believe that they should continue to be protected with antibiotics at times of known risk, just as for the unoperated patient.

A common sequel to complete relief of valvular pulmonic stenosis is the development of a murmur of pulmonic insufficiency (fig. 7). This murmur was usually present immediately after operation, but in two patients operated upon in infancy, the murmur developed as they grew older (about 2 years after operation in one, and 4 years after surgery in the other). Although the natural history of patients with such pulmonic insufficiency is not known, it would appear to be benign. Factors that increase the physiologically deleterious effects of pulmonic regurgitation, such as complete ablation or congenital absence of the valve, coexistence of a left-to-right shunt, and pulmonary arterial hypertension, are not present in these patients.

Conclusions and Recommendations

From the observations on these 100 present-day patients, it appears that pulmonary stenosis may become progressively more severe with growth of the young child, but it is impossible to predict which patient will...
PULMONIC STENOSIS

follow that course. Frequent observations at regular intervals are indicated.

If the obstruction has remained mild through adolescence and into adult life, the chief risk to such a patient is not cardiac dysfunction but bacterial endocarditis. Like patients with more severe stenosis and those who are postoperative, he should be protected against this infection at times of predictable risk. Curtailment of physical activities is not necessary for patients with mild stenosis, and surgery is not indicated.

While some adults with moderate obstruction continue to lead a near-normal existence, others in mid-adult life may begin to experience easy fatigue and shortness of breath. Operation in such an adult is indicated. However, the older the asymptomatic adult with moderate stenosis, the less is the operation indicated. In the child, operation is elective but advisable. In the infant with moderate stenosis, surgery can usually be deferred until childhood.

The patient with severe stenosis may lead a quiet life without discomfort but carries a risk of premature heart failure and death. The results of open-heart surgery have shown that the patient with severe stenosis can be relieved of his cardiac burden. Operation is indicated, regardless of his age. Especially if the stenosis is severe enough to have caused heart failure, operation should be undertaken without delay as soon as medical measures have controlled the failure.

Summary

The prognosis for the patient with pulmonic stenosis was analyzed from a study of 100 current patients, with long-term observations on 58 of them. Congestive heart failure occurred in seven, all of whom had severe stenosis. Subacute bacterial endocarditis developed in another seven, even when the stenosis was less marked. With appropriate medical supervision of those with mild stenosis who have no cardiac disability, and open-heart surgery for those with more severe obstruction, the outlook for the patient with pulmonary stenosis should be a full and active life.

Acknowledgment

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References


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