SERIAL CARDIAC CATHETERIZATIONS

but because of the patient’s minimal symptoms and absence of documented pneumonia, it was decided to follow the patient medically.

The patient was placed on a regimen of isoniazid, 300 mg, orally, daily and ethambutol, 1,200 mg, orally, daily; his course was uncomplicated. After four months of initial isolation, he was returned to duty with no physical limitations.

DISCUSSION

The association of pulmonary tuberculosis and bronchopulmonary sequestration is not a recent phenomenon. One theory contends that a sequestration is an acquired disease and that infection early in life initiates the development of this anomaly, which later in its formation captures an aberrant vessel; tuberculosis is postulated as being one such silent etiologic agent. Frequent reference is made to this simultaneous occurrence. However, review of the literature discloses only two patients with the simultaneous occurrence of bronchopulmonary sequestration and tuberculosis.

Smith describes a young woman presenting with a left lower lobe infiltrate and positive sputum for acid-fast bacilli. She responded to antituberculosis chemotherapy. However, a persistent shadow remained in the left lower lobe. One year after the detection of pulmonary tuberculosis, because of an inability to convert her sputum, the patient was subjected to resection of her left lower lobe. Histologic as well as gross examination revealed bronchopulmonary sequestration with a tuberculous lodged within it.

Tosatti and Gravel describe a 23-year-old man who presented with a 17-year history of “bronchitic attacks.” A chest roentgenogram revealed a thin-walled calcified cyst in the right lower lobe. Lobectomy was performed and revealed a bronchopulmonary sequestration. Histologic examination revealed two minute tubercles in the posterior basal segment. Our patient, unlike the young woman in the first case described, did not experience any difficulty in converting her sputum; unlike the man in the second case he did not have a history of intractable pyogenic infections. For these reasons it was felt that surgical intervention could be avoided.

The discovery of pulmonary tuberculosis in conjunction with a bronchopulmonary sequestration has rarely been reported. The benign course which tuberculosis adopts in the face of antituberculosis chemotherapy may mask the necessity for investigating the incomplete resolution of an atypical pulmonary infiltrate. This case underscores the importance of ruling out underlying pulmonary disorders whenever pulmonary tuberculosis appears in atypical sites.

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CHEST, VOL. 62, NO. 3, SEPTEMBER, 1972

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Serial Cardiac Catheterizations in
Infundibular and Pulmonic Stenosis

Lawrence Gould, M.D.; Farsad Unna, M.D.; Mohammad Zohar, M.D.; Robert F. Compere, M.D.; and Robert Frester, M.D.

A 43-year-old man with infundibular and pulmonic stenosis underwent two cardiac catheterizations over a seven-year period. With passage of time a decrease in cardiac index and severity of the infundibular stenosis was observed. Little change was observed in the valvular stenosis gradient. Reduction in obstruction at the infundibular level can probably be ascribed to an increase in the right ventricular volume.

The natural history of isolated pulmonic stenosis is still largely unknown. This is mainly caused by relatively recent clinical awareness of the frequency of this condition as well as the effectiveness of cardiac surgery.

Four groups have performed serial cardiac catheterizations in children, and have concluded that the right ventricular pressure remains essentially unchanged. These studies suggest that the orifice of the stenotic pulmonic valve increases with growth of the patient with mild to moderate pulmonic valvular stenosis. The symptoms that develop in many patients during the second decade of life have therefore been ascribed to the development of right ventricular myocardial fibrosis and infundibular hypertrophy. Indeed, in studies of patients with pulmonic valvular stenosis, the severity of infundibular hypertrophy increases with each decade. This finding suggests that the infundibular stenosis may develop and lead to a secondary and, at times, a major site of ventricular outflow obstruction. This concept, however, has remained conjectural since no serial cardiac catheterization studies have been performed in such a patient. We report the serial cardiac catheterization results in an adult with infundibular and pulmonic stenosis.

CASE REPORT

A 43-year-old man was seen in the Fordham Hospital cardiac clinic because of progressive dyspnea of six months’ duration.

*From the Department of Medicine, Misericordia-Fordham Hospitals, Bronx, New York. Report requests: Dr. Gould, Misericordia Hospital, Bronx, New York 10465.
The patient was informed of a cardiac murmur due to congenital abnormality, at the age of six. At age 18, he was rejected from military service because of the cardiac condition; however, at this time he was completely asymptomatic.

The patient became aware of exertional dyspnea at the age of 37 and was hospitalized for evaluation of this symptom. The diagnosis of pulmonic stenosis was made on clinical grounds, and the patient underwent a cardiac catheterization on December 19, 1963 (Table 1). The peak gradient between the right ventricle and pulmonary artery was 102 mm Hg. A systolic gradient of 60 mm Hg was demonstrated between the body of the right ventricle and the right ventriculostenotic chamber. The peak gradient between the infundibular chamber and the pulmonary artery was 36 mm Hg (Fig 1). The cardiac output as determined by the dye dilution method was normal. Dye dilution studies performed by injecting into the left atrium with simultaneous sampling from the pulmonary artery and brachial artery did not reveal a left-to-right shunt. A dye dilution curve recorded with injection into the right atrium and sampling from the brachial artery did not reveal a right-to-left shunt. A right atrial angiogram was compatible with the diagnosis of infundibular and valvular pulmonic stenosis. Surgery was advised but the patient refused.

He remained relatively free of symptoms until the onset of progressive dyspnea brought him to the cardiac clinic in August 1970. Physical examination at that time revealed a blood pressure of 130/70 mm Hg, a respiratory rate of 12 per minute and a pulse rate of 80. The lungs were clear. The point of maximal cardiac impulse was in the sixth intercostal space beyond the midclavicular line. There was a grade IV-VI ejection murmur at the pulmonic area, radiating to the left side of the neck. No gallop was audible. The liver was not enlarged. There was 1+ peripheral edema. Chest roentgenogram revealed cardiomegaly (Fig 2). Electrocardiogram showed right ventricular hypertrophy (Fig 3).

The patient was digitalized and became asymptomatic. A repeat cardiac catheterization was performed on October 8th, 1970 (Table 1). The peak gradient between the right ventricle and pulmonary artery was 40 mm Hg. A systolic gradient of 18 mm Hg was demonstrated between the body of the right ventricle and the right ventriculostenotic chamber. The peak gradient between the infundibular chamber and the pulmonary artery was 23 mm Hg (Fig 4). The cardiac output as determined by the Fick technique was reduced. A left-to-right shunt was not demonstrated through use of a platinum tip catheter in the pulmonary artery and having the patient breathe hydrogen.

**DISCUSSION**

The striking aspect of this case was the patient's progression from an asymptomatic state to congestive heart failure with a concomitant decline in the right ventricular systolic pressure. One would have expected a significant rise in the right ventricular pressure with the passage of time. In general the severity of pulmonic stenosis can be graded according to the signs, symptoms, electrocardiographic and roentgenographic findings. In

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<th>Table 1—Hemodynamic Findings.</th>
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<td>Findings</td>
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<td>Right atrial pressure (mean), mm Hg</td>
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<td>Right ventricular body pressure, mm Hg</td>
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<td>Right ventricular infundibular pressure, mm Hg</td>
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<td>Wedge pressure (mean), mm Hg</td>
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<td>Brachial artery pressure, mm Hg</td>
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**Figure 1.** Pressure recording in the body of the right ventricle of 120/12 mm Hg (upper curve). Note the infundibular pressure of 55/12 (lower left curve) and the pulmonary artery pressure of 23/15 (lower right curve).

**Figure 2.** Thoracic roentgenogram reveals a greatly enlarged heart.

**Figure 3.** Electrocardiogram showing right ventricular hypertrophy.

**Figure 4.** Right ventriculogram showing the infundibulum in the right ventricle.
children with severe pulmonic stenosis, the mean QRS frontal axis is between +110° and +150°. Lead V1 reveals either an R or a qR pattern and there may be a discordance between the QRS complex and T wave as well as ST segment depression in this lead. In addition, a roentgenogram of the thorax may reveal a greatly enlarged cardiac silhouette in patients with severe pulmonic stenosis. Our patient had a greatly enlarged heart and the electrocardiogram revealed a mean QRS frontal axis of +120°. However, the second cardiac catheterization revealed a peak right ventricular systolic pressure of 60 mm Hg as compared to the peak right ventricular pressure of 125 mm Hg in the first cardiac catheterization. The pressures in the right ventricle and pulmonary artery were obtained on many occasions during the two catheterizations. Catheter entrapment in the right ventricular musculature was therefore eliminated as a possible source of error. Cardiac output was estimated by utilizing dye curves during the first procedure and by utilizing the Fick principle during the second procedure. The values obtained by dye dilution check satisfactorily with those determined by the Fick technique. There was a 23 percent decline in the cardiac index and stroke index in the second catheterization as compared to the first procedure. A 13 percent decrease in the pulmonary artery systolic pressure and a 29 percent decline in the right ventricular systolic pressure was also observed in the second catheterization. However, the decrease in these pressures is in keeping with the fall in the cardiac output.
output. The marked fall of 53 percent in the right ventricular systolic body pressure cannot be explained by a fall in the cardiac index. If one compares the infundibular gradient of 80 mm Hg in the first cardiac catheterization with the gradient of 18 mm Hg in the second procedure then a 67 percent decline in the infundibular stenosis has occurred. It appears inescapable that the severity of the infundibular hypertrophy in this patient did not increase with the passage of time but actually diminished. One can only speculate on the genesis of these findings. However, the clinical deterioration in this patient cannot be explained by the increasing severity of pulmonic stenosis or infundibular stenosis. Perhaps the right ventricular myocardial fibrosis has become so extensive that the muscle in the infundibulum has been extensively replaced with fibrotic material. Unfortunately the patient still refuses to have cardiac surgery performed, and this concept must remain conjectural.

The extensive development of right ventricular myocardial fibrosis has been stressed as an important determinant of the natural history of pulmonic stenosis. Further the right ventricular myocardium of patients with pulmonic stenosis has significantly more fibrosis than the right ventricular myocardium in normal individuals of the same age or in patients with tetralogy of Fallot. However this hypothesis must be rejected in our patient in view of the marked decline in the right ventricular peak pressure which is out of proportion to the decline in the cardiac output. The alternate and more likely explanation is that the right ventricular volume has so increased thereby minimizing obstruction at the infundibular level. Unfortunately right ventricular angiograms were not obtained at both cardiac catheterizations and a comparison of volume of this chamber is therefore not possible.

The first cardiac catheterization in 1963 revealed an elevated right atrial mean pressure and an elevated right ventricular end diastolic pressure. Ayers and Lukas previously observed that this was a frequent hemodynamic finding in mild pulmonic valvular stenosis. Decreased compliance of the right ventricle has been the explanation offered for this finding. The second catheterization in 1970 now revealed a normal right atrial mean pressure, and a normal right ventricular end diastolic pressure. One can only offer a possible explanation for this phenomenon. With the passage of time the volumes in the right atrium and right ventricle may have increased with an increase in compliance. In such a setting, a decrease in the pressure may result.

Obviously one cannot come to definite conclusions about the natural history of pulmonic stenosis in the adult based on one case. However, since this is the first adult with infundibular and pulmonic stenosis who underwent serial cardiac catheterization it does offer additional insight into this disease state.

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Mucopidermoid Tumor of the Trachea*

G. P. Trentini, M.D. and B. Palmieri, M.D.

We describe a rare case of mucopidermoid tumor of the supracricoid region of the trachea in a 25-year-old man, who died three months after resection, following local recurrence of the tumor. On the basis of clinical course and of gross and microscopic examination, the tumor has been classified as a medium-high grade mucopidermoid tumor. Occurrence of this lesion in the trachea has only been reported once, moreover not in pure form, but in combination with squamous cell carcinomas.

Although mucopidermoid tumors as occasional findings are included in bronchial neoplastic pathology, they are not mentioned normally in the classification of tumors of the trachea. Review of the literature on this subject shows only the previous report of Larson, Woolser and Payne.' Their report concerns a 26-year-old woman with a mucopidermoid tumor of the supracricoid region of the trachea, whose symptoms had started three years before, with dyspnea, cough and blood-streaked sputum. The extreme rarity of this pathologic condition prompted us to describe our case of mucopidermoid tumor.

CASE REPORT

History and Clinical Data

The patient was a 25-year-old man, whose past and personal history was not contributory. His illness started four months previously with mild dyspnea. Each episode of dyspnea, cough and blood-streaked sputum. The extreme rarity of this pathologic condition prompted us to describe our case of mucopidermoid tumor.

*Institute of Anatomic and Histologic Pathology, University of Modena, Italy (Chairman: Prof Stigliani). Reprint requests: Prof. Trentini, Via Bertengosa no 4, Modena, Italy 41100

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