INFUNDIBULAR PULMONIC STENOSIS WITH INTACT VENTRICULAR SEPTUM*

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Despite an increasing number of reports to the contrary, infundibular pulmonic stenosis with intact ventricular septum is considered a rare congenital anomaly. In 1959, 3 patients with this condition were encountered in a series of 60 patients undergoing open correction of congenital cardiac lesions at the University of Oklahoma Medical Center. A discussion of the findings concerning and, the management of, these patients seems warranted in order to emphasize the special problems encountered in their diagnosis and treatment.

CASE REPORTS

Case 1

D. C. (U.H. No. 181-532), a 12-year-old white boy, was first noted to have a heart murmur in infancy. During early childhood, his development was somewhat slow, and the patient fainted after severe exercise on several occasions. At 8 years of age, cardiac catheterization revealed a right ventricular systolic pressure of 120 mm. Hg and a pulmonary artery systolic pressure of 16 mm. Hg. The transition in the pressure tracing between pulmonary artery and right ventricle was noted to occur rather low and, in retrospect, the pressure tracing suggests infundibular stenosis. Because no definite infundibulum was demonstrated, a diagnosis of valvular pulmonary stenosis was made. There was no evidence of shunting. Transventricular pulmonary valvulotomy was carried out several months later. At the time of operation, no change in the systolic thrill in the pulmonary outflow tract was noted. After the operation, the parents reported some improvement in exercise tolerance; however, the patient continued to have occasional loss of consciousness after severe exercise. Repeat catheterization was performed 6 months before the present admission.

Physical examination. At the time of admission examination revealed an underdeveloped, white boy. His blood pressure was 110/70. There was no cyanosis. There was slight prominence of the left chest with a left parasternal lift. A harsh systolic thrill was palpable along the left sternal border. There was a grade IV harsh systolic murmur which was maximal at the second and third interspaces along the left sternal border. The murmur radiated well to the neck, back and axilla. The second sound was diminished and split.

Laboratory findings. The hemogram and urinalysis were normal. Chest x-ray showed right ventricular enlargement. The pulmonary artery segment was decreased, and the pulmonary vascular shadows were normal. Electrocardiogram revealed right ventricular hypertrophy, and an incomplete right bundle branch block which had not changed after the previous operation. At cardiac catheterization, the right ventricular systolic pressure was 164 mm. Hg. A simultaneously recorded branchial artery pressure was 116/60 mm. Hg. Because of technical difficulties no attempt was made to enter the pulmonary artery. Brachial artery saturation was 96 per cent. Dye curves with right ventricular injection showed no evidence of right to left shunting. A diagnosis of pulmonary stenosis with intact ventricular septum was made.

A second operation was carried out on April 8, 1959, with the use of the midline sternum-splitting incision. The cavae were intubated through the right atrium and an arterial catheter was placed in the right superficial femoral artery. A modified DeWall-Lillehei bubble oxygenator with sigma-motor pump was used, and after establishment of satisfactory bypass, a normal pulmonary valve was exposed through an arteriotomy just above the valve ring. A finger could be passed through the valve and an infundibular stenosis palpated. Right ventriculotomy was carried out, and an infundibular muscular diaphragm excised with scissors. After completion of the cardiac repair and after discontinuing bypass, the right ventricular systolic pressure was 45 mm. Hg, and the brachial artery systolic pressure was 110 mm. Hg (fig. 1).

Postoperative course. The patient's postoperative course was uneventful. He was discharged from the hospital 3 weeks after operation in good

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Fig. 1. Case 1. Upper left, drawing demonstrating site and nature of defect; upper right, operative photograph of right ventrieulectomy. Small opening at center is actual opening in infundibular diaphragm. Lower right shows repair of defect.
Fig. 2. Case 2. Upper left, photograph demonstrating external appearance of infundibular chamber. Drawing at upper right demonstrates site of pathology. Lower right, drawing illustrating placement of outflow patch.
condition. There was diminution of the precordial thrill, but a loud murmur persisted. The right ventricular impulse was diminished. Chest x-ray revealed a normal heart size 3 months postoperatively. Repeat catheterization has not been carried out. Electrocardiogram shows less right ventricular hypertrophy.

Case 2

P. C. (U.H. No. 264-032), an 18-year-old white girl, was first found to have congenital heart disease during a febrile illness at 16 years of age. She gave a history of easy fatigability, but this had not prevented her from leading an active life. Cardiac catheterization was carried out 3 months before admission for surgery.

Physical examination. The patient was a well developed, slightly obese, white girl who did not appear acutely or chronically ill. There was no evidence of cyanosis. There was a left precardial cardiac impulse with no thoracic deformity. A grade V systolic murmur was audible to the left of the sternal border in the second and third interspaces, and there was a palpable thrill in the same area. The murmur was well transmitted to the neck and back. The second sound was not audible.

Laboratory findings. Hemogram and urinalysis were normal. The chest x-ray revealed no cardiac enlargement; the pulmonary arterial segment was not enlarged, and pulmonary vasculature was normal. The electrocardiogram showed right axis deviation and moderate right ventricular hypertrophy. At cardiac catheterization right ventricular systolic pressure was measured at 127 mm. Hg; pulmonary artery pressure was 15 mm. Hg. There was no evidence of a shunt by oxygen saturation or dye curves. No technically satisfactory pulmonary artery to right ventricle withdrawal tracing was obtained. A diagnosis of pulmonary valvular stenosis with intact ventricular septum was made.

Operation was performed on June 17, 1959. The heart was exposed through a midline sternum-splitting incision and the presence of an infundibular chamber was readily observed upon inspection of the heart. This thin walled chamber was noted to fill and contract slightly after the body of the right ventricle, and there was a constricted area approximately 5 cm. below the pulmonary valve ring. A harsh thrill was present in this portion of the ventricle and did not change in character distal to the pulmonary valve. The cavae were cannulated through the right atrium and arterial cannulation of the right femoral artery was carried out. Connections were completed to the modified DeWall-Lillehei bubble oxygenator with a sigmmamotor pump, and after establishment of satisfactory cardiopulmonary bypass, the infundibular chamber was opened, exposing a thick muscular diaphragm separating the infundibulum from the right ventricle. The opening in this diaphragm was estimated at 8 mm. in diameter. After a large amount of the infundibular muscular ridge was trimmed away with forceps and scissors, there still appeared to be narrowing of the right ventricular outflow tract, and accordingly, a diamond shaped patch of laminated Ivalon and Teflon was sutured into the defect. Aortic cross clamping was carried out for 17 minutes, and after release of the aortic cross clamp ventricular fibrillation occurred. Sinus rhythm was readily restored with electric shock. Good cardiac action was maintained, and after removal of the cannulas, the right ventricular systolic pressure was 25 mm. Hg, and the brachial artery systolic pressure was 100 mm. Hg. No flow measurement was carried out at this time. The bypass was carried out for a total of 27 minutes and perfusion was considered satisfactory (fig. 2).

Postoperative course. The patient’s postoperative course was uneventful. The thrill and murmur were lessened in intensity and there was no change in heart size on x-ray. After discharge from the hospital, the patient continued to improve and was asymptomatic at the time of this report. Postoperative catheterization has not been carried out. The electrocardiogram no longer shows right ventricular hypertrophy and the axis is normal (fig. 3).

Case 3

K. M. (U.H. No. 281-427), a 22-year-old white man, was noted to be slightly cyanotic shortly after birth. Early development was retarded, and the patient had numerous respiratory infections in infancy and childhood. He was first seen at the Medical Center 4 months before the present admission, at which time he was found to have congenital heart disease, mental deficiency, cryptorchidism and nontoxic diffuse goiter. Extensive endocrine work-up revealed no evidence of recognizable endocrine deficiency syndrome, and during this admission, cardiac catheterization was carried out. The patient had never been employed and had led a very sedentary existence.

Physical examination. The patient was a poorly developed, thin white man who appeared younger than 22 years of age. His blood pressure was 130/60. On examination of the head and neck, a high arched palate was noted. There was an increased anteroposterior diameter of the chest with prominence of the left precordium. There was a left parasternal tap. A systolic thrill was
palpable to the left of the sternum in the second and third interspaces, and a grade IV harsh systolic murmur was audible to the left sternum. This murmur was well transmitted to the axilla and neck. The second sound was diminished in the pulmonic area. There was feminine hair distribution with cryptorchidism. Examination of the extremities revealed enlargement and hyperextensibility of both knees and elbows.

Laboratory findings. Routine hemogram and urinalysis gave normal findings. Chest x-ray showed enlargement of the right ventricle. The pulmonary artery was prominent, and the vascular shadows were considered to be within normal limits. Electrocardiogram demonstrated first degree ativoventricular block, marked right axis deviation and marked right ventricular hypertrophy. Catheterization, on April 16, 1959, revealed a right ventricular systolic pressure of 185 mm. Hg, pulmonary artery systolic pressure of 20 mm. Hg, and a brachial artery systolic pressure of 116 mm. Hg. Brachial artery oxygen saturation was 96 per cent, and dye curves revealed a possible small right to left shunt at the atrial level. Withdrawal tracing from the pulmonary artery to the right ventricle was not technically satisfactory. A diagnosis of valvular pulmonic stenosis with intact ventricular septum and small right to left shunt at the atrial level was made.

At operation on July 22, 1959, the heart was exposed through a midline sternum-splitting incision. There were abnormalities of both coronary arteries with aneurysmal dilations in multiple areas. No palpable thrill was palpable over these coronary vessels. The venae cavae were cannulated through the atrium and blood was returned from the modified DeWall-Lillehei bubble oxygenator sigmamotor pump connected to the right femoral artery. Pulmonary arteriotomy revealed a normal pulmonary valve. A finger inserted through the valve revealed diffuse narrowing of the infundibulum. A right infundibulotomy was performed and muscle from the crista removed by sharp scissor dissection. A diamond shaped patch of laminated Ivalon and Teflon was sutured into the ventriculotomy incision in order to enlarge the diffusely narrowed infundibulum. After the cannulas were removed, the pressure in the pulmonary artery rose to 25 mm. Hg, but the right ventricular pressure remained about 100 mm. Hg.

Postoperative course. The patient's postoperative course was complicated by persistent fever for which no cause could be found. He was ultimately discharged from the hospital somewhat improved. The thrill and murmur remained essentially unchanged. After leaving the hospital, the patient felt that his exercise tolerance had increased and his color improved. Although follow-up catheterization has not been carried out,
electrocardiography shows a decrease in right ventricular hypertrophy.

DISCUSSION

The true incidence of infundibular pulmonic stenosis with intact ventricular septum has not been determined. Before 1948, all forms of "pure" pulmonic stenosis were considered very unusual, but since that time, improved clinical and laboratory diagnostic methods have resulted in establishing the diagnosis in an increasing number of patients. It seems likely that pulmonic stenosis with intact ventricular septum comprises about 10 per cent of cases of congenital heart disease, and that of these, approximately 10 per cent are infundibular stenosis. In our small operative series, infundibular stenosis with intact ventricular septum has occurred in 3 of 12 patients with isolated pulmonic stenosis. Swan states that 60 cases of isolated infundibular stenosis have been reported in the literature since 1958.

Keith's explanation that infundibular stenosis occurs because of faulty involution of the bulbus cordis is widely accepted. Acceptance of this thesis does not imply that the pathogenesis is understood. Two pathologic types of infundibular stenosis are described by Lev. In the first type, an fibrotic band is present at the junction of the conus and sinus of the right ventricle which divides the ventricle into two portions. The conus portion is the infundibular chamber. This is the most common variety of infundibular stenosis associated with intact ventricular septum. There may be an associated atrial septal defect or patent foramen ovale. The second type consists of a shrunken conus portion with a long stenotic infundibulum. Johns states that the second type is commonly seen in the transposition complex. In two of our cases, the infundibular stenosis was quite localized with a well formed infundibular chamber. In our third case, a long area of infundibulum was narrowed, corresponding to the second type described by Lev. Kjellberg has discussed the abnormal positions of the muscle bands comprising the crista supraventricularis which may account for some of the anatomical variation in the isolated infundibular stenosis defect.

CLINICAL FEATURES

The history obtained from patients with infundibular pulmonic stenosis is indistinguishable from that in other forms of isolated pulmonic stenosis. Symptomatology ranges from no symptoms in those patients with mild stenosis through mild limitation of activity and progresses to dyspnea, fatigue, syncope and finally, symptoms of right-sided congestive heart failure. Cyanosis may occur in severe cases in patients who have an associated atrial septal defect or in a patent foramen ovale.

On physical examination, there is usually little or no chest deformity. A left parasternal lift or tap is usually present. The heart may be slightly enlarged. The systolic thrill and murmur are usually maximal in the left second and third interspaces, somewhat lower than in valvular pulmonic stenosis. The second sound is almost always well split and of diminished intensity. It is not well transmitted to the right. The long well known fact that the murmur is maximal at a lower interspace in infundibular than in valvular stenosis was obvious only in retrospect in our patients. Certain phonocardiographic characteristics of the murmur are helpful and will be mentioned subsequently.

DIAGNOSTIC PROCEDURES

Chest x-ray usually shows no increase in transverse diameter of the heart. The right ventricular enlargement is best seen in a direct lateral projection and is frequently marked, although this may be less prominent in infundibular than in valvular stenosis. Of more importance is the fact that the pulmonary artery, which is almost always enlarged because of poststenotic dilation in valvular pulmonic stenosis, is usually of normal size in infundibular pulmonic stenosis. The decrease in size of peripheral pulmonary arteries has been reported to be less marked in the infundibular form of the disease. Absence of an enlarged main pulmonary artery in an x-ray of a patient with the clinical findings of pulmonic stenosis should arouse serious suspicion that the stenotic area is infundibular. Electrocardiography in both valvular and infundibular stenosis shows right ventricular hypertrophy. The degree of right ventricular hypertrophy has been shown to correlate with the height of right ventricular systolic pressure in patients with valvular pulmonic stenosis, and in our experience, this is also true of infundibular pulmonic stenosis. The phonocardiogram may be of help in differentiating isolated valvular...
from infundibular pulmonic stenosis. The murmur of infundibular pulmonic stenosis has been reported to start immediately after the first sound and to extend only to the aortic component of the second sound, in contrast to the murmur of valvular pulmonic stenosis which starts later and extends through the sound of aortic closure. Pre-and postoperative phonocardiographic studies in our patients indicate that the difference in characteristics of the murmur depends more on the severity of the stenosis than on the site.

At cardiac catheterization, withdrawal of the cardiac catheter from pulmonary artery to infundibular chamber to sinus portion of right ventricle should result in a pressure tracing diagnostic of infundibular pulmonic stenosis. Typically this tracing reveals no change in systolic pressure on withdrawal from the pulmonary artery into the infundibular chamber, but rather, a sudden drop in diastolic pressure. When the catheter is further withdrawn into the body of the right ventricle, the systolic pressure should increase with relatively little change in diastolic pressure. It was not possible to make the diagnosis of infundibular stenosis in our patients by this technique because technically satisfactory withdrawal tracing could not be obtained. Kirklin has pointed out that even when careful withdrawal tracings are recorded, the diagnosis of infundibular stenosis may be missed. The possibility that catheterization data alone may not prove or disprove a diagnosis of infundibular stenosis is of considerable significance to the surgeon.

The injection of contrast media into the right ventricle with clear visualization of the outflow path is often successful in differentiating infundibular from valvular stenosis. The importance of differentiating infundibular hypertrophy from true infundibular stenosis has been emphasized by Kjellberg. This procedure was not carried out with any of the patients reported herein, but has been of considerable value in subsequent studies on other patients.

SURGICAL TREATMENT

Although attempts have been made to resect the tissue responsible for infundibular stenosis by closed method, there is little question that the direct visual approach allows more accurate and thorough correction. Swan has recommended that direct vision surgery be carried out with the use of inflow occlusion and hypothermia, and has emphasized the minimal risk of ventricular fibrillation with careful hypothermic technique. Resection of the infundibulum has been accomplished in 4 to 5 minutes in Swan's series, and the results have been quite satisfactory. Other surgeons have preferred the use of cardiopulmonary bypass to allow more time for infundibular resection and to prevent the danger of ventricular fibrillation, however slight. In our 3 patients, the heart was bypassed for 19, 25 and 63 minutes, and in two instances, revision of the outflow tract with a plastic patch was considered advisable. The risk of cardiopulmonary bypass has steadily diminished until at the present time there is little hesitation in employing this method if there appears to be a definite advantage gained by its use. The fact that bypass techniques safely allow the intracardiac operating time necessary for extensive reconstruction of the ventricular outflow tract or for repair of unsuspected associated defects has made the approach the procedure of choice at our institution for operations on all cases of pulmonic stenosis except for patients operated on in infancy.

SUMMARY AND CONCLUSIONS

1. Infundibular pulmonic stenosis in the presence of intact ventricular septum is not a rare congenital cardiac anomaly, and probably comprises about 10 per cent of all cases of pulmonic stenosis with intact ventricular septum.

2. In 1969, 3 of 12 patients operated on for isolated pulmonic stenosis at the University of Oklahoma Medical Center have had infundibular stenosis. In none of these patients was the diagnosis of infundibular stenosis definitely established before operation.

3. Important points in preoperative differentiation of this form of pulmonic stenosis from the usual valvular stenosis are as follows: (a) Physical examination reveals a thrill and murmur in the second and third interspaces to the left of the sternum rather than the first and second. (b) Chest x-ray fails to show poststenotic dilation of the pulmonary artery. (c) Cardiac catheterization reveals an area of low systolic pressure proximal to the pulmonic valve. (d) Selective right ventricular angiogram is of definite value in visualizing the site of stenosis.

4. Treatment of this condition by infundibular resection can be accomplished with good clinical
results and a low mortality rate. Because of the

time required to carefully repair the right ven-

tricular outflow tract, cardiopulmonary bypass

is the method of choice at this center.

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