SIMPLE PULMONARY STENOSIS

BY

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Simple pulmonary stenosis is relatively common. Wood (1950) considers it to account for 12 per cent of cases of congenital heart disease and Campbell (1954) gives the figure of 10 per cent. The two largest series of cases to be reported are those of Abrahams and Wood (1951) 69 cases, and Campbell (1954) 100 cases.

This paper deals with a consecutive series of 33 cases. The clinical features are briefly reviewed and evidence is sought to define more clearly the natural history of the disease.

The age and sex distribution of the 33 cases are shown in Table I. All but one of them were originally referred to hospital as a result of a routine medical examination at school or at ante-natal

TABLE I
SIMPLE PULMONARY STENOSIS
AGES AT THE TIME OF REVIEW, AND SEX DISTRIBUTION

Age in years	No. of cases	Male	Female
0-10	9	3	6
11-20	15	12	3
21-37	9	1	8
Total	33	16	17

clinics. Fifteen have been followed for more than 5 years and a further eleven for from 10 to 21 years. The children were seen at six-monthly or yearly intervals until they left school and thereafter less frequently. Three were referred back to the clinic during pregnancy and others returned specifically for the present review.

SYMPTOMS

Twenty-three patients had no symptoms. They led unrestricted lives and took part in every activity, including competitive sport, without handicap. Of the remaining ten patients nine complained of breathlessness. As in other types of heart disease the assessment of breathlessness was sometimes difficult; in every case heart disease had been diagnosed early, and because of this, some of the children had been restricted at school and at home, and in two patients other features of cardiac neurosis were present. It seemed clear, however, that the breathlessness was genuine in at least six, and three were considerably handicapped. In one of these three, physical capacity was limited, not only by breathlessness, but also by central chest pain, which was constantly related to more severe exertion. A second also complained of fainting attacks after effort.

Additional features were failure to gain weight (1 case) and mental defectiveness (2 cases). Pulmonary stenosis was not considered to be the cause of these two conditions.

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SIGNS

The external appearance was usually normal. Peripheral cyanosis of the lips and fingers was seen in a few cases, particularly in the cold. In the warm a rather bright pinkness of the cheeks was sometimes noted. Central cyanosis was observed in 4 cases. In three, slight finger clubbing was present, but one of the three had subacute bacterial endocarditis and the absence of clubbing had been noted at her previous attendance. All four with central cyanosis had polycythæmia with hæmoglobin levels of over 115 per cent.

Conspicuous exaggeration of the venous "a" wave was observed in 7 cases. This sign was seen to disappear in at least one case with rest in bed.

The cardiac impulse was of normal character in 22 cases. In the remaining 11 a right ventricular lift or heave was felt in the third left interspace midway between the apex beat and the centre of the sternum. There was as a rule no pulsation in the second left interspace, nor was pulmonary valve closure ever felt.

A pulmonary systolic thrill and murmur were present in the second or second and third left interspace in every case, and were usually coarse. The second heart sound was split in 11, and single in 22 cases.

Radiological Appearances. These are described by Abrahams and Wood (1951). Post-stenotic dilatation of the main pulmonary artery segment was well marked in 19 of this series, less well marked in 4, and absent in 10 cases (Fig. 1-2). Diminution of the pulmonary vascular markings was observed in three and considered probable in two others. Considerable increase in the transverse diameter of the heart was seen only once.

The Electrocardiogram. The electrocardiographic pattern varied from normal to the most severe degree of right ventricular dominance (Fig. 4). The tall sharp P waves of right auricular hypertrophy were seen in seven instances. For convenience the electrocardiograms were divided into four grades as follows.

- Grade 1. Normal.
- Grade 2. Incomplete right bundle branch block.
- Grade 3. Right ventricular dominance without widespread inversion of the T waves.
- Grade 4. Extreme right ventricular dominance with widespread inversion of the T waves in the præcordial leads, usually accompanied by T wave inversion in leads II, III, and VF.

Cardiac Catheterization. This was carried out in 8 cases and the pulmonary artery was reached in seven. Systolic pressures in the right ventricle lay between 40 and 65 mm. in four cases and between 130 and 180 mm. Hg in four cases.

CLINICAL COURSE

There has been no symptomatic deterioration in most of these patients in the first three decades of life. A clear history of progressive dyspnœa was obtained in only 4 of the 10 with symptoms. Three of these were women of 24, 26, and 37 years: two of them had borne full-term babies without symptoms but subsequently became disabled by breathlessness, which was accompanied in one by anal pain, and both are becoming increasingly cyanosed. The fourth patient, a man of 19, noticed increasing dyspnœa and cyanosis on exertion and twice fainted after cycling uphill.

In no case has there been radiological evidence of increasing heart size. In those where right ventricular enlargement was evident it was present in the earliest films and did not increase. In the only patient with gross cardiac enlargement, a girl of 23 years, the first film 5 years before showed the same degree of enlargement.

FIG. 1.—Examples of the radiological picture. (A) Normal. (B) Typical post-stenotic dilatation of the main pulmonary artery segment.

FIG. 2.—Example of the radiological picture. Post-stenotic dilatation, small aorta, prominence of the right atrium and diminished pulmonary vascular markings.

FIG. 3.—Calcification of the pulmonary valve. By courtesy of the Cardiac Department, St. Thomas' Hospital.

FIG. 4.—The four grades from normal to right ventricular strain in the electrocardiogram.

Limb lead electrocardiograms were available for study from the date of first attendance and præcordial leads more recently. In 28 cases no changes were found in the cardiograms over the years. This included all 17 cases with grade 1 and grade 2, and 10 of the 11 with grade 3 tracings. In 4 of the 5 cases with grade 4 tracings, increasing degrees of right ventricular dominance could be seen. A characteristic example is shown in Fig. 5 where T wave inversion is seen to develop successively in leads III, V2 to V4, and VF.

Subacute bacterial endocarditis has occurred three times in two patients. The first, a girl of 13 had an attack five years ago. The second, a woman of 37, also had an attack five years ago and is now being treated for bacterial endocarditis again. In both these patients there is evidence of deterioration, taking the form of progressive dyspnoea and polycythæmia in one, and increasing cardiographic abnormality in the other.

No case of pulmonary tuberculosis has come to light.

DISCUSSION

No attempt has been made in this series to distinguish between valvular and infundibular stenosis. In every case but one the thrill was high, being felt in the second or second and third left interspaces. In six of the seven where pressure tracings were obtained there was a single, abrupt rise of pressure at or near valve level when the catheter was withdrawn from the pulmonary artery to the right ventricle: in the seventh there was clear evidence of valvular and high infundibular

FIG. 5.—Increasing right ventricular dominance in the electrocardiogram.

stenosis and this was confirmed at operation. It is probable, therefore, that this series does not include a case of low infundibular stenosis. Brock and Campbell (1950), Dow *et al.* (1950), and Kirklin *et al.* (1953) have all emphasized the difficulty or impossibility of distinguishing between valvular and high infundibular stenosis by pressure tracings.

The clinical course of most of these patients has been uneventful. Usually there was no symptomatic, radiological, or cardiographic evidence of deterioration. The relative youth of the series as a whole is largely explained by the fact that nearly all came under observation while at school, and that in the course of years it becomes increasingly difficult to keep in touch with symptomless patients. It seems likely, however, that many of them will reach the later decades relatively symptomless.

Such a case was recently brought to my notice by Dr. Michael Mathews of St. Thomas' Hospital. This was a man of 65 years who was known to have an abnormal heart when he was discharged from the services in the first world war. In youth he played football regularly and he remained free of symptoms until the last year or two when he began to suffer from bronchitis and became breathless on hurrying. The cardiogram showed right bundle branch block and on fluoroscopy the pulmonary valve was seen to be heavily calcified (Fig. 3).

Symptomatic deterioration has occurred in four patients, in each of whom increasing dyspnoea was paramount. Two of the four complained, in addition, one of fainting on exertion, and one of anginal pain. Progressive electrocardiographic changes have occurred in two patients who remain symptomless.

The decision when to operate in this relatively symptomless condition has recently been discussed by Campbell (1954). We have taken as evidence of a severe stenosis, breathlessness, central cyanosis, an accentuated "a" wave in the neck, a heaving right ventricle, diminished pulmonary vascular markings, and extreme right ventricular dominance in the cardiogram. It is assumed that central cyanosis can only occur in those patients with a communication between the atria.

In the presence of increasing symptoms there is usually no difficulty in deciding to submit the patient to the risk of cardiomy. Further observation may well show that better results are obtained by performing valvotomy when there are signs of increasing right heart hypertrophy rather than by waiting until breathlessness becomes evident, age advances, and right heart hypertrophy becomes extreme.

There seems at present little indication for operation in those patients who show no real evidence of right heart stress.

SUMMARY

Thirty-three patients with simple pulmonary stenosis are presented, the majority having been under observation for from 5 to 21 years. Two-thirds of them were symptomless. Six were breathless on exertion, four being markedly handicapped.

The physical signs, radiological appearances, and electrocardiographic features are briefly reviewed.

The clinical course has been studied. There was no evidence of deterioration in the majority. The question of operative intervention is briefly discussed.

I am grateful to Professor C. Bruce Perry for permission to study these cases and to him and to Dr. D. H. Davies for their advice and criticism. Professor Neale and Dr. Apley have also allowed me to quote their cases. Dr. Michael Mathews and Dr. Ernest Jellinek of the Cardiac Department of St. Thomas' Hospital kindly arranged for me to see one of their patients and provided Fig. 3.

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