PULMONARY STENOSIS WITH NORMAL AORTIC ROOT*

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Normally systolic pressure is the same in the pulmonary artery and in the right ventricle. If systolic pressure is lower in the pulmonary artery than in the right ventricle, pulmonary stenosis is present. The term "pulmonary stenosis with normal aortic root" is that of Wood(1) and provides an adequate definition of the disease. Pulmonary stenosis may be of variable degree and severity. As a rule the lowest pressure gradient indicative of pulmonary stenosis is considered to be 10 mm. Hg. in the absence of a left to right shunt. A prerequisite for the occurence of a pressure gradient is a narrowing at some point between the cavity of the right ventricle and the pulmonary artery.

MATERIAL AND METHODS

This paper deals with a series of 14 cases of pulmonary stenosis with normal aortic root. The diagnosis was confirmed by cardiac catheterization in all of them. Pulmonary stenosis with a left to right shunt or a right to left shunt and an overriding aorta were excluded from this study. Full clinical examination was carried out in all of them. Particular attention was paid to the following points: the degree of right ventricular hypertrophy as indicated by a left parasternal heave, the presence or absence of an ejection click, the length, duration and intensity of the systolic murmur at the site of its maximum intensity and the degree and width of splitting of the second heart sound in expiration.

Routine electrocardiograms included standard and unipolar limb leads and chest leads V_1 to V_6. The degree of right ventricular hypertrophy was graded from 1 to 4 in severity. Routine radiological examination included a skiagram of chest and fluoroscopy during catheterization. The severity of valvular or infundibular stenosis was assessed by the right ventricular pressure measured during conventional right heart catheterization and the following grading was used: when the systolic pressure was under 60 mm. Hg. stenosis was considered mild; between 60 and 100 mm. Hg. moderate; between 100 and 185 mm. Hg. severe. Cardiac output was calculated by Fick's method. Phonocardiograms were taken in all cases.

RESULTS

Age and sex: The age and sex distribution is shown in Table I. There were 10 males and 4 females. Maximum numbers were between the ages of 11 and 20 years.

Table I.

Pulmonary Stenosis with Normal Aortic root
Age and Sex distribution

Age in years	No. of cases	Male	Female
0 - 5	—		
6 - 10	2	1	1
11 - 15	6	4	2
16 - 20	5	4	1
21 - 25	1	1	
Total	14	10	4

Clinical features: There were no symptoms in 10 patients. Three had effort dyspnoea, of these two had severe pulmonary stenosis. One patient complained of chest pain, palpitation and syncopal attacks. Pulmonary stenosis was very severe in this patient. Peripheral cyanosis of the lips and fingers was seen in one patient, particularly in the cold during cardiac catheterization. Giant 'a' wave was detected in the jugular venous pulse in three patients. The volume of the radial pulse was recorded as small only in four patients.

The cardiac impulse was of normal character in five cases. Marked parasternal heave was recorded in six cases indicating marked right ventricular hypertrophy and in three cases it was considered between mild to moderate.

* Based on the lecture delivered at the Cardiac Society of Singapore on 7 May 1965.

Table 2.

Clinical Features

No Symptoms	10
Dyspnoea on exertion	3
Syncope, Chest pain & Palpitation	1
'a' Wave in Neck	3
R.V. thrust	
No R.V. thrust	5
Slight	1
Moderate	2
Marked	6
Systolic thrill	14
Systolic murmur over PA or left sternal edge (3)	14
Split S_2	
Normal	5
Delayed	6
Single	3
Ejection click	3

Table 3.

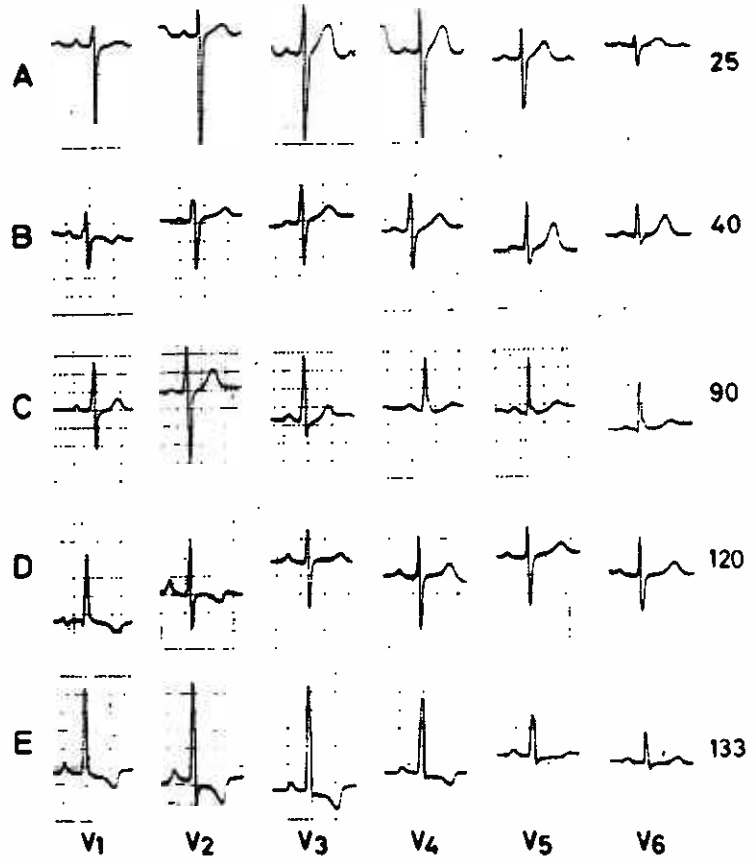
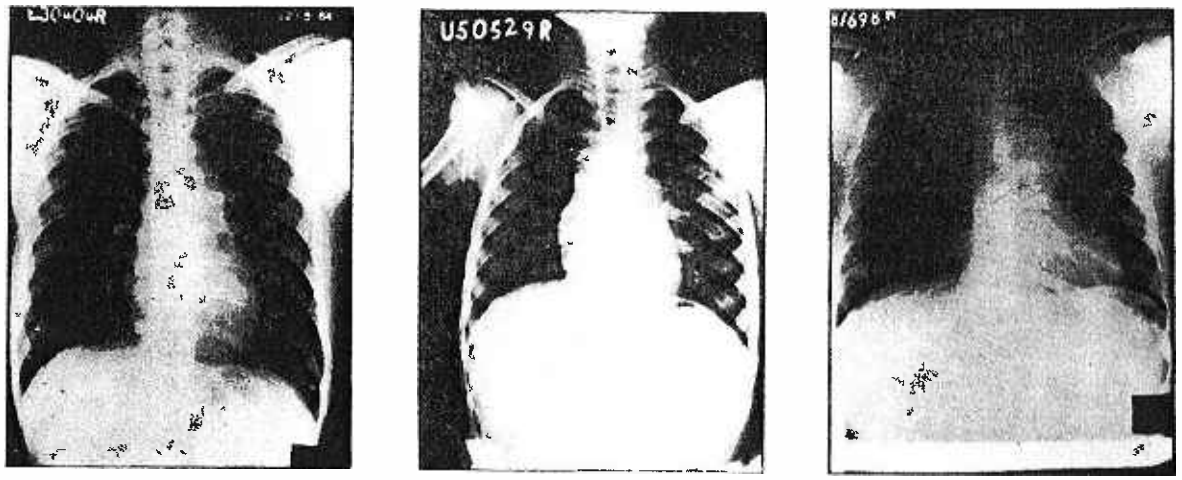


Fig 2.– Chest lead electrocardiograms (V1 to V6) from five cases of simple pulmonary valve stenosis showing increasing grades of right ventricular hypertrophy from none (A) to grade 4 (E).

Fig. 1.



A B C

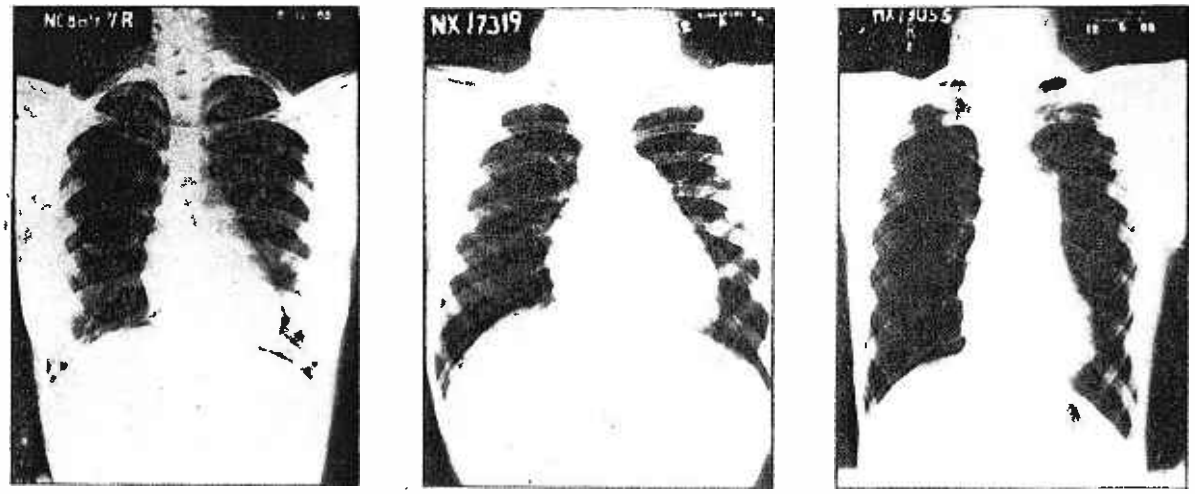
Three Chest X'Rays of Mild Pulmonary Stenosis.

A: R.V. Pressure $\frac{30}{6}$ CTR = 45%.

B: R.V. Pressure $\frac{40}{0}$ CTR = 42%.

C: R.V. Pressure $\frac{50}{2}$ CTR = 56%.

Fig. 1.



A B C

Three Chest X'Rays of Severe Pulmonary Stenosis.

A: R.V. Pressure $\frac{120}{10}$ CTR = 49%.

B: R.V. Pressure $\frac{120}{6}$ CTR = 51%

C: R.V. Pressure $\frac{185}{5}$ CTR = 50%

A pulmonary systolic thrill and murmur were present in the second or second and third left interspace in every case. The second heart sound was normally split in five. P_2 was delayed in six and single in three.

Ejection clicks were heard in three cases, all with mild pulmonary stenosis. Salient clinical features are shown in Table 2.

X-ray appearances: There are three important radiological features of pulmonary stenosis: post-stenotic dilatation of the main pulmonary artery, diminished pulmonary vascular markings in the peripheral lung fields and enlargement of right heart. In this series there was no correlation between the degree of post-stenotic dilatation of the main pulmonary artery and the severity of the pulmonary stenosis. In one of the mild cases there was aneurysmal dilatation of the main pulmonary artery though the right ventricular pressure was only 50 mm. Hg. The degree of pulmonary ischaemia and enlargement of the right heart seemed to correlate more with the severity of the pulmonary stenosis. Pulmonary vascular markings were diminished in eight cases and enlargement of the right heart was seen in seven cases. X-ray appearance of three mild cases and three severe cases of pulmonary stenosis is shown in Fig. 1.

Electrocardiography: The electrocardiographic pattern varied from normal to a very severe degree of right ventricular hypertrophy. In the present analysis electrocardiographic evidence of right ventricular hypertrophy was divided into four grades:

1. Mild: R and S about equal in amplitude in V_1.
2. Moderate: R dominant in lead V_1 but S is still present.
3. Severe: Tall R and no or very small S in V_1. T is usually inverted.
4. Very severe: R tall in V_1 and T inversion extends upto V_4.

This is shown in Table 3.

The electrocardiographic findings are shown in Table 4. Right ventricular hypertrophy was present in 11 cases of which five were severe and three indicated very severe hypertrophy. The most characteristic change is the tall R wave in V_1. The correlation between the pressure in the right ventricle and the amplitude of the R wave in V_1 is shown in Table 5.

Table 4.

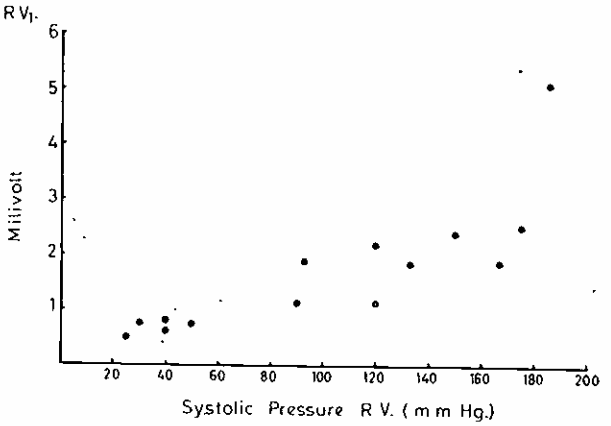
Electrocardiogram.

Normal	1
R.V. hypertrophy	11
R.B.B. Block	2
Partial	1
Complete	1
P-R interval	Normal in all patients
'P' Pulmonale	3

R. V. hypertrophy

Mild (R = S in V_1)	2
Moderate (R>S V_1)	1
Severe (Dominant R V_1 No or very small s)	5
Very severe (T↓ V_{1-4})	3

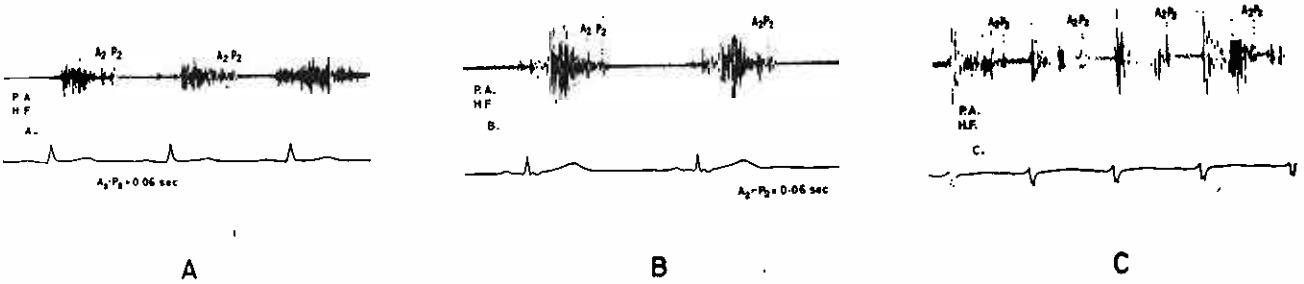
Table 5.



Correlation of the R wave in V_1 with systolic pressure in the right ventricle.

Phonocardiography: The first heart sound was normal in all cases. An ejection click occuring between 0.04 to 0.06 seconds after the first heart sound was detected in three mild cases of pulmonary stenosis. Like Leatham and Vogel Poel (2) I have been unable to detect any ejection click in any of the cases of severe pulmonary stenosis. In severe valvular stenosis the pulmonary component of the second heart sound showed decreased amplitude in all cases. A_2-P_2 distance was increased and recorded between 0.08 seconds to 0.12 seconds. The murmur was

Fig. 2.



A B C

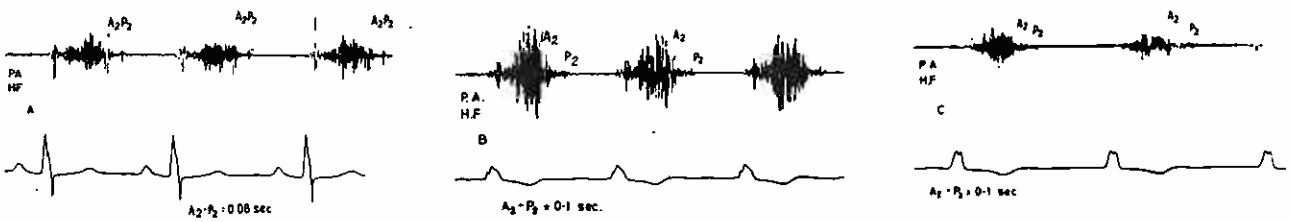
Phonocardiogram in Mild Valvular Pulmonary Stenosis.

A. Boy, aged 18 (N 36797). Right Ventricular pressure 25/4 mm Hg. pulmonary artery pressure 16/10. The murmur is early systolic. $A_2 - P_2 = 0.06$ sec.

B. Boy, aged 14 (N 38736). Right Ventricular pressure 30/6 mm Hg. Pulmonary artery pressure 20/9. The murmur is midsystolic. $A_2 - P_2 = 0.06$ to 0.07 sec.

C. Boy, aged 10 (M 15946). Right Ventricular pressure 40/4 mm Hg. Pulmonary artery pressure 20/14. The murmur is early and midsystolic.

Fig. 2.



A B C

Phonocardiogram in Severe Valvular Pulmonary Stenosis

A. Girl, aged 11 (M 18338). Right Ventricular pressure 120/6 mm Hg. Pulmonary artery pressure 12/7. The murmur is late systolic, diamond shaped and continues with small amplitudes until the pulmonary component of the second sound $A_2 - P_2 = 0.08$ sec.

B. Boy, aged 13 (M 20257). Right Ventricular pressure 168/4 mm Hg. Pulmonary artery pressure 16/8. The murmur is late systolic, diamond shaped and continues with small amplitudes until the pulmonary component of the second sound $A_2 - P_2 = 0.1$ sec.

C. Boy, aged 16 (N 16069). Right Ventricular pressure 185/0 mm Hg. Pulmonary artery pressure 12/5. The murmur is late systolic, diamond shaped and continues with small amplitudes until the pulmonary component of the second sound. $A_2 - P_2 = 0.1$ sec.

Table 6.

SUMMARY OF FINDINGS

	Simple Pulmonary Stenosis	
	Mild: R.V. <60	Severe: R V >100.
R. V. Thrust	Nil or Slight	Marked.
Pulmonary ejection click	Common	Uncommon
Second sound – Aortic Component	Normal	Normal but may be masked by murmur at pulmonary area.
Pulmonary Component	Normal	Present but of reduced intensity.
Degree of Splitting	Increased (upto 0·06 sec) in expiration.	Greatly increased (0·06-0·12 sec in expiration)
Pulmonary Systolic murmur	Soft or loud with crescendo at mid systole and ended before A_2.	Loud with great delay in crescendo and duration, murmur extended well beyond A_2, partially or completely obscuring it.
Electrocardiogram		
Right Ventricular hypertrophy	Nil or Slight	Marked. $RV_1 > 1{\cdot}5$ mv. $T \downarrow V_{1-4}$.
Skiagram of Chest		
Pulmonary Ischaemia	Absent	Present
Enlargement of right heart	Absent	Present.

Table 7.

Pulmonary Stenosis

Name	Sex	Age	Race	Unit No.	Pressure (m.m. Hg) P.A.	R.V.	Site of Stenosis	Cardiac Index (L/m/m²)
L.B.K.	M	16	C	N 32605	22/11	150/12	Valvular	—
K.K.C	M	14	C	N 7318	21/11	133/26	Valvular	—
P.A.S.	M	19	I	N 30655	28/7	50/2	Valvular	3·4
M.M.	F	17	I	N 4563	20/8	120/10	? Infundibular	3·8
D.B.S.	F	10	M	M 18338	12/7	120/6	Valvular	4·0
T.Y.K.	F	14	C	M 18820	23/13	90/0	Valvular	2·7
R.B.A.	M	18	M	N 36797	15/10	25/4	Valvular	4·9
L.S.W.	M	14	C	N 38736	20/9	30/6	Valvular	4·2
T.E.C.	M	25	C	M 19489	20/8	40/5	Valvular	3·8
C.S.C.	M	10	C	M 15946	20/14	40/0	Valvular	4·1
L.K.S	M	13	C	M 20257	16/8	168/4	Valvular	3·3
L.K.T.	F	12	C	N 30318	29/18	93/0	Valvular	4·2
O.E.K.	M	12	C	N 2207	8/5	175/5	Valvular	3·2
T.C.H.	M	16	C	N 16069	12/5	185/0	Valvular	2·4

usually late systolic, diamond shaped or kite shaped and continued with small amplitude until the pulmonary component of the second sound. Phonocardiographic recordings in three cases of mild pulmonary valvular stenosis and three cases of severe valvular stenosis are shown in Fig. 2.

The important clinical features, phonocardiographic, electrocardiographic and radiological findings to assess and distinguish mild from severe cases of pulmonary stenosis are summarized in Table 6.

Cardiac catheterization: Our knowledge of the characteristic clinical picture of pulmonary stenosis has been corroborated by this means, so that it is now possible not only to diagnose the condition without cardiac catheterization but to assess the severity of the stenosis also. It is however necessary to resort to cardiac catheterization for an exact determination of the degree and site of stenosis. This was carried out in all the 14 patients. The findings may be summarised as follows:

1. The systolic pressure in the right ventricle, which provides a measure of the degree of stenosis was always higher than in the pulmonary artery.
2. There were five mild cases in which the systolic pressure in the right ventricle was between 25 to 59 mm. Hg.
3. In two moderate cases the right ventricular systolic pressure was between 60 to 99 mm. Hg.
4. In seven severe cases the right ventricular systolic pressure ranged from 100 to 185 mm. Hg.

The lowest pressure recorded was 25 mm. Hg. and the highest 185 mm. Hg. The smallest pressure gradient between the pulmonary artery and the right ventricle was 10 mm. Hg. The cardiac output was normal in all but two cases. In two cases cardiac output was not calculated. The site of stenosis was valvular in all cases except in one in which it was doubtful. The details of the cardiac catheterization findings are shown in Table 7. The pressure tracings from the pulmonary artery and right ventricle in a case of severe pulmonary stenosis are shown in Fig. 3.

Angiocardiography: Angiocardiography is an important and useful method of investigation in pulmonary stenosis. It is especially important

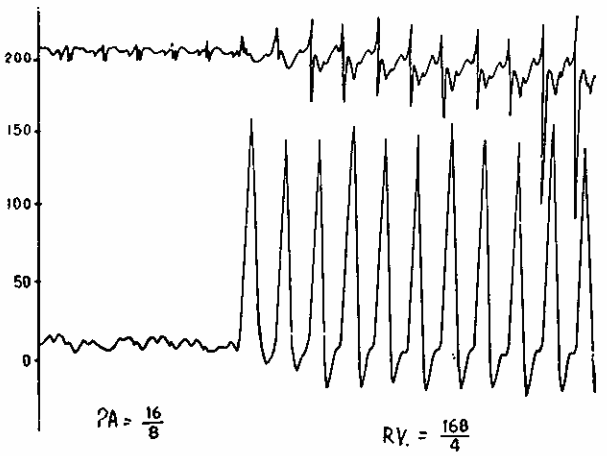


Fig. 3. Withdrawal Curve in Pulmonary Stenosis. Boy, aged 13. Valvular stenosis with sudden change from low pulmonary pressure (16/8 mm Hg) to high ventricular pressure (168/4 mm Hg) P.A., Pulmonary artery; R.V., Right ventricle.

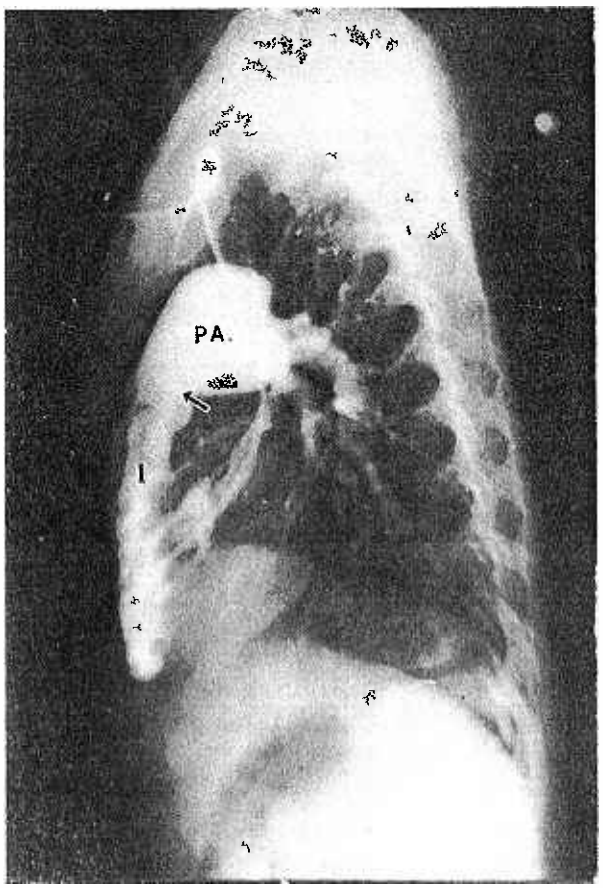


Fig. 4. R. V. angiocardiogram showing valvular pulmonary stenosis. Boy, aged 14. Arrow points to valvular plane. I, infundibulum; P.A. pulmonary artery.

from the surgical point of view aimed mainly at elucidating the anatomy of the outflow tract of the right ventricle and in the pulmonary orifice. Visualisation is facilitated by injecting the contrast medium selectively into the right

ventricle. This was carried out in five patients. Right ventricular angiocardiographic findings in a case with a right ventricular systolic pressure of 133 mm. Hg. are shown in Fig. 4.

DISCUSSION

Fourteen cases of pulmonary stenosis with normal aortic root amongst 280 cases examined by cardiac catheterization showed an incidence of 5%. There was a preponderance of males over females. In Gwee's (3) small series of eight cases there were three males and five females. The exact incidence however is difficult to determine, as this is a very selective group and may not represent the true incidence of the disease.

There were no symptoms of any kind among 10 out of 14 patients. Physically all the patients were of normal build. On the whole most of these patients had very little incapacity. The general appearance of the patients did not help to suspect the nature of the heart disease. Only one case showed mild peripheral cyanosis. Right ventricular hypertrophy indicated by a left parasternal heave was the most important sign on palpation. This was present in all except the mild cases and helped to determine the severity of the pulmonary stenosis. Like Leatham and Vogel Poel(2) I have been unable to detect an ejection click in any of the cases with severe pulmonary stenosis. The pulmonary component of the second heart sound exhibited a decreased amplitude in severe cases but this seemed difficult to be sure from auscultation because the aortic component is masked by the long systolic murmur and the pulmonary component was not always easy to recognise.

In this series there was no correlation between the degree of dilatation of the main pulmonary artery or its right or left branch and the severity of the stenosis. Pulmonary ischaemia and enlargement of the right heart seemed to correlate better with the degree of stenosis. Pulmonary vascular markings were diminished in all cases of severe stenosis. As Kjellberg (4) has pointed out the lack of objective criteria for evaluating the vascular markings and their normally wide range of variation, makes it difficult to make a definite statement in mild cases. The right heart was enlarged in six out of seven cases of severe stenosis.

The analysis of the electrocardiograms showed that normal curves occured only in mild cases and grade 4 right ventricular hypertrophy occured only in severe cases. The most characteristic change was the tall R wave in V_1. There was a definite correlation between the height of the R wave in V_1 and the pressure in the right ventricle. Whenever the amplitude of the R wave in V_1 was higher than 1.5 millivolt the right ventricular pressure was always above 90 mm. Hg.

Phonocardiographic recordings showed the following characteristics. In mild pulmonary stenosis the systolic murmur is characteristically early or mid systolic. With severe stenosis the murmur is diamond or kite shaped and late systolic. This observation tallies with those of Vogel Poel(5) and Kjellberg(4). The length of the murmur seemed to bear an important relation to the severity of stenosis. The width of splitting of the second sound at the base also was important in assessing the severity of stenosis. In severe stenosis A_2-P_2 interval varied between 0.07 seconds and 0.12 seconds in this series. Ejection click was absent in all cases of severe pulmonary stenosis. The amplitude of the pulmonary second sound was also very small in all cases of severe stenosis.

Cardiac catheterization was carried out in all the cases. Of these 14 cases five were mild valvular stenosis where the right ventricular systolic pressure varied between 25 to 59 mm. Hg., in two moderate cases it ranged between 60 and 99 and in seven severe cases the pressure ranged between 100 and 185 mm. Hg. All of them were valvular stenosis except one where the site of stenosis remained doubtful. The site of stenosis was confirmed in all cases by direct fluoroscopic visualization of the withdrawing catheter, in four cases by intracardiac electrography in addition to fluoroscopic visualization. Angiocardiographic visualization was obtained in five cases. It seemed from the analysis of this small series that intracardiac electrography helped to determine the site of stenosis more accurately than fluoroscopic visualization. Cardiac output was normal in all cases except two and in two cases the cardiac output was not calculated.

SUMMARY

Fourteen cases of pulmonary stenosis with normal aortic root are presented. The incidence of the anomaly was 5% amongst 280 cases of cardiac catheterization. Their clinical features

are described and severe cases are distinguished from mild pulmonary stenosis by the presence of a marked parasternal heave, the absence of an ejection click, a loud and long systolic murmur over the pulmonary area and the characteristic reduced intensity and wide splitting of the second heart sound. Skiagram of the chest showed post-stenotic dilatation of the main pulmonary artery, pulmonary ischaemia and enlargement of rightventricle and atrium. The electrocardiogram varied from normal to marked right ventricular hypertrophy. Phonocardiogram confirmed the auscultatory findings and revealed the shape of the systolic murmur, the wide splitting of the second sound and the reduced intensity of P_2. Cardiac catheterization revealed right ventricular pressures, the site of stenosis and the cardiac output. Selective angiocardiography elucidated the anatomic condition in the outflow tract of the right ventricle and pulmonary orifice and is of vital importance from the surgical point of view.

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