ATRIAL SEPTAL DEFECT*

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Atrial septal defect constitutes the commonest of all congenital heart anomalies. In most cases, however, there are associated cardiac anomalies[1] and frequently the defect in the atrial septum is of secondary importance. Isolated atrial septal defect is nevertheless still a common defect and, in our experience in Johannesburg, it occurs as frequently as isolated ventricular septal defect, these two lesions being less common, however, than patent ductus arteriosus.

We present here our experience in 43 patients, 37 with isolated atrial septal defect and 6 with persistent common atrioventricular canal.

Material

The patients were all seen personally by one or more of the authors. The diagnosis was supported by cardiac catheterization in 28 cases. In 17 of these 28 the diagnosis was confirmed at operation for closure of the defect, and in 3 of the 17 necropsy examination added further confirmation. In 7 more cases the diagnosis was confirmed at necropsy. In the remaining 8 cases the diagnosis was made with confidence on clinical grounds and cardiac catheterization was advised.†

Pathological Anatomy

In recent years much confusion has arisen as a result of different anatomical classifications of atrial septal defects. Without entering into any controversy regarding the embryology of the defects, we have attempted to classify our cases into the following 2 groups only. We believe that the mode of surgery should depend on which of the 2 types of defect is present:

1. '*Septum secundum*' *defects*. The defect is usually in the situation of a foramen ovale, but may be higher ('sinus venosus' defect) or lower in the septum. The ventricular septum is intact and the atrioventricular valves have no congenital abnormality. Surgery under hypothermia is the method of choice.

2. '*Septum primum*' *defects*. The defect is always low, and there may be associated congenital anomalies of the mitral valve and/or the tricuspid valve, and possibly a defect of part of the ventricular septum (common atrioventricular canal). Extracorporeal circulation is essential for surgical repair.

While our classification may not satisfy embryologists, we have found it to be satisfactory for clinical application.

In our series, 37 were secundum defects, and 6 (14%) were primum defects.

Lutembacher's syndrome. By definition, this condition is an association of mitral stenosis and a sizable congenital interatrial septal defect. It is now accepted as being rare, and we had no case of it in our series. However, one of our patients had severe rheumatic mitral incompetence with a large atrial septal defect.

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† Two of these 8 patients have been catheterized since this paper was presented and the diagnosis has been confirmed.

Age and Sex Incidence

There were 30 female patients and 13 male (2·3 : 1). The higher incidence in females is well known.

The ages ranged from 3 months to 54 years (Table I). As in most series, the greatest number of cases occurred between 2 and 40 years of age. The fairly high incidence in the 1st year of life is due to the inclusion of 4 patients who were seen for a congenital heart anomaly in infancy, and who all died, the diagnosis being confirmed by necropsy.[2]

TABLE I. AGE INCIDENCE

Age (years)	*Patients*	%
0– 1	4	9·3
1– 2	2	4·6
2–10	12	28·0
10–20	12	28·0
20–30	5	11·6
30–40	6	13·9
40–50	1	2·3
Over 50	1	2·3

Prognosis

It will not be possible to arrive at an accurate assessment of the prognosis in atrial septal defect unless long follow-up studies of large numbers of patients are made. However, 2 facts become evident from analysis of our material: (1) When the patient with an atrial septal defect presents *clinical* evidence of heart disease in early infancy, the prognosis is grave. All 4 of our cases died within the first 2 years of life. We believe, although we have no definite proof, that these cases may be associated with a persistence of 'foetal pulmonary hypertension'. Histological examination of the lung vasculature at necropsy showed thickened media of the smaller arteries in 1 case. In the other 3 the pathologist regarded the vessels as normal. None showed intimal hyperplasia. Our experience would indicate that in those patients with a good prognosis in early life, *clinical* evidence of heart disease is generally not present in infancy. However, in one of our patients, aged 4 years, who was first seen at 6 months of age, although the heart was then regarded clinically as normal, an electrocardiogram showed an rsR′ pattern in VI, and this pattern persisted. (2) Atrial septal defect is rare over the age of 40 years. One of our patients, aged 44, had severe exertional dyspnoea but after corrective surgery her exercise tolerance became normal. Our oldest patient died at 54 of a cerebral thrombosis (? paradoxical embolus) following hysterectomy, and necropsy revealed a small atrial septal defect (0·5 cm. in diameter). Nevertheless there was considerable right ventricular enlargement and the patient had experienced severe dyspnoea.

Of the 6 patients in the 4th decade, 2 were in severe, intractable congestive cardiac failure and died, 1 after corrective surgery.

We consider, therefore, on the available evidence, that atrial septal defect is a serious congenital anomaly with a high mortality in early infancy and early middle age. Campbell *et al.*[3] reported that after infancy has passed the prog-

nosis remains good until the 4th and 5th decades. Similar figures are presented by Bedford *et al.*[4]

The following are factors which appear to affect the prognosis in the present series:

1. *Type of defect.* Of the 11 patients who died, 2 were suffering from primum defects; their ages at death were 2 years and 10 years.*

2. *Pulmonary hypertension.* Of the 9 deaths in patients with secundum defects, 6 probably had well-marked pulmonary hypertension. Of the 6, 4 were under 2 years of age and the other 2 were 22 and 23 years old. The 3 deaths in patients with secundum defects, but no significant pulmonary hypertension, were in patients aged 37, 39 and 54. We believe, therefore, that the prognosis is worse in those with primum defects and in those with significant pulmonary hypertension (Table II).

TABLE II. DEATHS IN RELATION TO AGE, TYPE OF DEFECT AND PULMONARY HYPERTENSION

Age (years)	*Primum defect*	*Secundum defect with pulmonary hypertension*	*Secundum defect without significant pulmonary hypertension*
0–2	1	4	0
2–10	1	0	0
10–30	0	2	0
Over 30	0	0	3
Total deaths ..	2	6	3

CLINICAL PICTURE

The clinical picture of atrial septal defect in infants differs considerably from that in older children and adults; it will not be discussed here because it is the subject of a separate report.[2]

In children and adults, the patients are often of gracile habitus. Arachnodactyly and high arching of the palate are sometimes, though rarely, present; they were noted in only 3 of our cases. Mongolism was not present in any of our cases. Cyanosis and clubbing of the fingers were present in 1 patient, who had severe pulmonary hypertension with reversal of shunt.

The pulse is commonly of small volume. Venous pulsation in the neck is not a noteworthy feature, except in patients with congestive cardiac failure.

In patients with secundum defects there is usually prominent pulsation to the left of the sternum and in the 2nd left intercostal space. A systolic thrill is not often present; it was felt at the pulmonary area in 4 of our patients with secundum defects (10·8%). On auscultation of the heart, the following are classical features in these cases:

1. An ejection systolic murmur, usually of moderate intensity (grade 3 or 4), occasionally soft, and occasionally very loud, is best heard at the pulmonary area and, if loud, may be conducted all over the precordium.

2. The second heart sound is well split (0·03 - 0·06 second, but in 2 patients 0·02 second) and the degree of splitting varies little, if at all, with respiration. The 2nd (pulmonic) component of the 2nd heart sound is usually only slightly louder than the 1st component, but in patients with pulmonary hypertension it is markedly increased in intensity.

3. A systolic ejection click is commonly present; it was found in 23 (62%) of our patients with secundum defects. In some cases the click is difficult to separate clinically from the systolic murmur, but gives the murmur a 'coarseness' of quality. In these cases, the click can be demonstrated with the phonocardiograph, and it often becomes easily audible after surgical closure of the atrial septal defect, when the murmur frequently disappears.

4. A soft, short diastolic murmur, sometimes 'scratchy' in quality and occurring shortly after the 2nd heart sound, is frequently noted at the lower end of the sternum;[6] it was found in 26 (70%) of our cases of secundum defect. This murmur is believed to be due to 'torrential' flow across the tricuspid valve, and is usually best heard during inspiration.

5. A soft blowing early diastolic murmur, due to pulmonic incompetence, may be heard occasionally, especially in patients with pulmonary hypertension.

In patients with primum defects, the above features may all be present. In addition, a localized left ventricular heave may be felt over the cardiac apex. In all our 6 cases, a well-marked pan-systolic murmur could be heard at the apex, and in 4 of them this murmur could be distinguished from the pulmonary ejection murmur. In 1 patient, a loud pan-systolic murmur with associated thrill was best noted along the left sternal edge. In 5 patients, a well-marked mid-diastolic murmur was audible along both edges of the sternum and in 3 of them this murmur was well heard at the cardiac apex.

Symptoms referable to the heart were frequently present, at all ages, but were severer in the older patients and in those with primum defects or pulmonary hypertension. The degree of disability could not be well correlated with the degree of left-to-right shunt. Of the 6 patients with primum defects, 3 were in congestive cardiac failure with its attendant symptoms. Of the remaining 3, 1, aged 3½ years, was asymptomatic and 2, aged 12 and 33, experienced severe exertional dyspnoea and tiredness. Excluding the 4 infants who died, there were 33 patients with secundum defects, of whom 4 were in congestive cardiac failure. Of these 4, 2 were over 30 years of age; 1, aged 11, had associated rheumatic mitral incompetence; and 1, aged 2½, had moderate pulmonary hypertension. Dyspnoea and tiredness on exertion was present in 21 of the remaining 29 patients with secundum

TABLE III. PATIENTS ACCORDING TO SYMPTOMS

Symptom	*Primum defects*	*Secundum defects with pulm. hyperten.*	*Secundum defects without signific. hyperten.*	*Total*
Congestive cardiac failure	3	1	3	7
Dyspnoea and tiredness on exertion..	2	3	18	23
Probable paroxysmal rhythm disturbances	0	2	2	4
Haemoptysis ..	0	1	1	2
Asymptomatic ..	1	0	8	9

* The very poor prognosis in these lesions is commented on by Wakai and Edwards,[5] in whose series of patients with common atrioventricular canal, 21 of 25 died before 6 years of age and only 2 reached adult years.

defects. Of the patients with secundum defects 8 were completely asymptomatic. Haemoptysis was experienced by 2 patients, one with pulmonary hypertension and one in congestive cardiac failure. Symptoms suggestive of attacks of paroxysmal tachycardia were present in 3 patients and 1 patient had several bouts of paroxysmal ventricular tachycardia and died from ventricular fibrillation. Sweating was occasionally noted in the younger patients. The symptoms are tabulated in Table III. The significance of dyspnoea and tiredness is often difficult to assess, because in some cases the patients have become cardiac neurotics.

Electrocardiogram

For many years an ECG pattern of so-called incomplete right bundle-branch block has been regarded as usual in atrial septal defects. Cabrera and Monroy[7] referred to this pattern, in which right precordial leads showed an rsR′ or rsR′s′ complex, as diastolic overload of the right ventricle. The failure of this pattern to disappear immediately after successful closure of the atrial septal defect seemed to disprove the 'diastolic overload' concept.

On the basis of the findings in our cases, we believe that this pattern is produced by dilatation of the right ventricular outflow tract. This area of the heart is undoubtedly dilated in cases of atrial septal defect and we believe this dilatation to persist or only slowly to disappear after corrective surgery. If this is so, it is possible that, with the relief of the shunt, the right ventricle may be able to empty before the left ventricle. In 1 patient, aged 44 years, this is suggested by the presence of 'paradoxical splitting' of the 2nd heart sound soon after closure of the defect (Fig. 1).

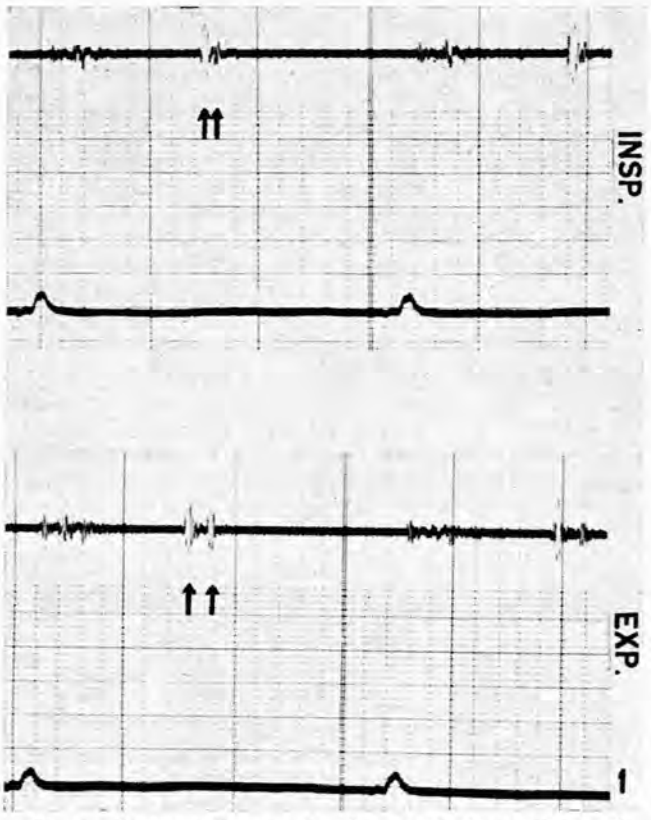

Fig. 1. Phonocardiogram showing paradoxical splitting of the 2nd heart sound after closure of an atrial septal defect.

In patients with pulmonary hypertension, a greater degree of hypertrophy of right ventricular muscle may be present and the ECG features may be somewhat different.

In our series, we recognize the following patterns of right ventricular preponderance in right chest leads (Fig. 2):

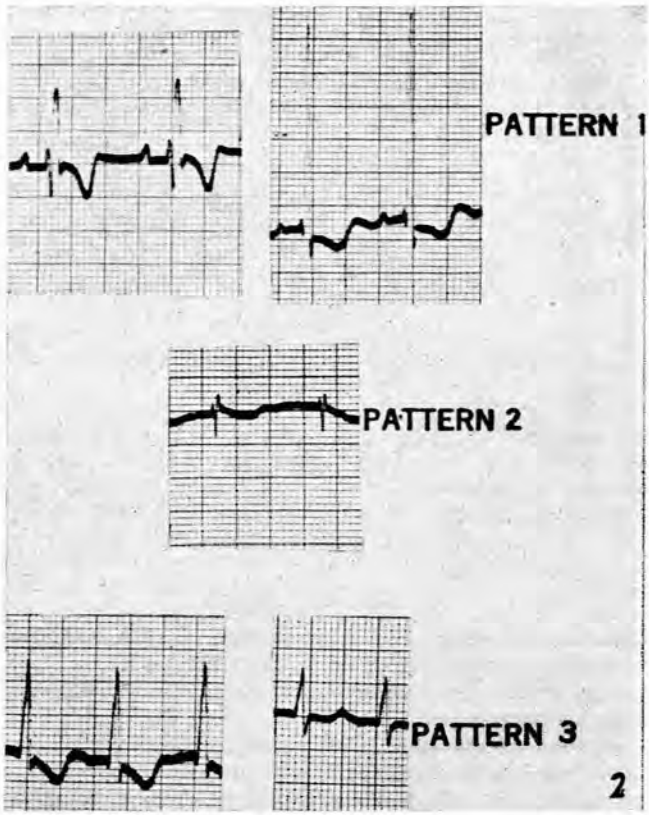

Fig. 2. The 3 basic patterns of the electrocardiogram in atrial septal defect. All tracings are of lead VI. The right-hand tracing in Pattern 1 is from a patient with severe pulmonary hypertension.

1. rsR′ or rsR′s′ where R′ was greater than twice the amplitude of s or s′.
2. rsr′ or rsr′s′ where r′ was less than twice the amplitude of s or s′.
3. qR or Rs or R.

Pattern 1 was present in 28 patients, of whom 19 were cases of secundum defect without pulmonary hypertension, 5 of secundum defect with pulmonary systolic pressures above 40 mm. Hg, and 4 of primum defect. In 18 of the 19 secundum defects without pulmonary hypertension the duration of the initial r was greater than 0·025 seconds while in 4 of the 5 with pulmonary hypertension the duration was less than 0·025 seconds.

Pattern 2 was present in 6 patients, 5 being cases of secundum defect and 1 of primum defect. In 2 of the 5 with secundum defects, the left-to-right shunt was large, while in 3 it was small as measured by cardiac catheterization

or presumed (in 1) by virtue of the small size of the defect found at necropsy.

Pattern 3 was present in 8 patients, all cases of secundum defect, 7 of whom were believed or known to have pulmonary hypertension.

In 1 patient no right ventricular 'preponderance' was found and it is believed that a 'balanced ventricular pattern' was produced by combined ventricular enlargement. This patient had an unusual form of 'septum primum' defect and will be reported separately.

Thus, while we have been unable to correlate a typical ECG pattern with pulmonary artery pressure, pulmonary flow, or pulmonary vascular resistance, we believe that careful appraisal of the ECG enables the type of case to be fairly accurately classified.

ECG evidence of right atrial enlargement was a common feature, being present in 38 (88%) of our cases. Left atrial enlargement was present in 3 cases, 1 of a primum defect, 1 associated with systemic hypertension, and 1 in severe congestive cardiac failure.

Evidence of left ventricular enlargement and the presence of left axis deviation constituted the most important single evidence of the defect being of the primum type. Thus, of 6 cases of primum defect, 5 showed left ventricular enlargement and the remaining 1, already referred to, was believed to have a 'balanced electrocardiogram'. One patient with a secundum defect showed left ventricular enlargement, attributable to systemic hypertension.

Radiological Features

In past years the detection of a so-called 'hilar dance' on fluoroscopy was regarded as an important feature of atrial septal defect. In our series, while pulsation in the main pulmonary arteries was often vigorous, a true 'hilar dance' was seldom present, and we pay little attention to this finding.

Roentgen films were taken in the customary views, namely postero-anterior, left and right anterior oblique, and lateral. With few exceptions, we have obtained all the information we require from routine PA films alone. However, we believe that none of these views are quite satisfactory for determination of right atrial enlargement. In view of the lateral situation of the right atrium and its tendency to enlarge in an anterior direction, we feel that examination in a slightly left anterior oblique position (about 10° rotation) will be advantageous; we are at present studying the value of this view. The following radiological features were analysed:

1. Size of the heart, as assessed by the cardiothoracic ratio.

2. Right atrial enlargement, assessed in the PA view and divided into 4 grades, viz. grade 1 (slight enlargement), grade 2 (moderate enlargement), grade 3 (marked enlargement), and grade 4 (aneurysmal enlargement).

3. Prominence of the right pulmonary artery on the one hand and main and left pulmonary arteries on the other. This is graded as grade 1, 2 or 3 for slight, moderate or marked prominence.

4. Pulmonary plethora, and particularly the visibility of large 'end-on' vessels well out in the lung fields. It is graded as grade 1, 2 or 3 for slight, moderate or marked plethora.

5. The so-called 'pulmonary hypertension picture', with sudden attentuation of pulmonary arteries towards the lung periphery.

6. Right and left ventricular enlargement.

Detailed Analysis

1. *Size of heart.* The cardiothoracic ratio was 53% or more in 25 cases. In general, those with primum defects had larger hearts (CTR over 60%), although the two largest hearts were in patients with secundum defects, one being in intractable heart failure, and the other having marked pulmonary hypertension. A normal-sized heart may be present even with large left-to-right shunts, and in 6 patients the CTR was under 50%, although the pulmonary flow was twice or more the systemic flow. The average CTR was 55%.

2. *Right atrial enlargement* of greater or lesser degree was always present (Fig. 3) and, in one patient in intractable heart failure, the right atrium was aneurysmal. The average grade of right atrial enlargement was 2·4, with no difference between primum and secundum defects.

3. *Prominence of pulmonary artery.* Because of 'overlapping' of the main and left pulmonary arterial shadows in the PA projection, these were analysed together; they were prominent in every case. The average grade of prominence was 2·5, with no difference between primum and secundum defects. Prominence of the right pulmonary artery was a feature, and is felt to be more notable than in other types of congenital heart disease with left-to-right shunt (Fig. 3). The average grade of prominence was 2·3, with no difference between primum and secundum defects.

4. *Pulmonary plethora.* Contrary to popular opinion,

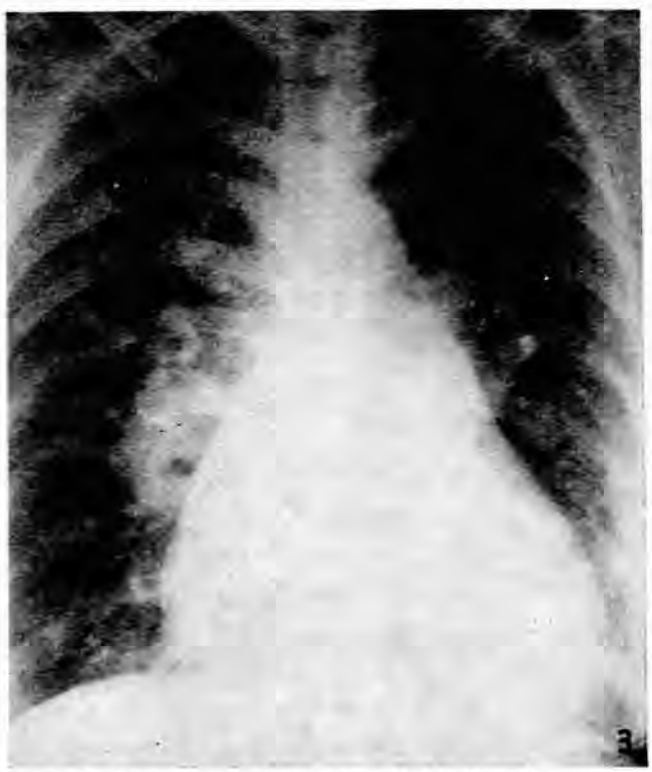

Fig. 3. Posterior-anterior radiograph showing enlargement of right atrium, both main pulmonary arteries, and right ventricle.

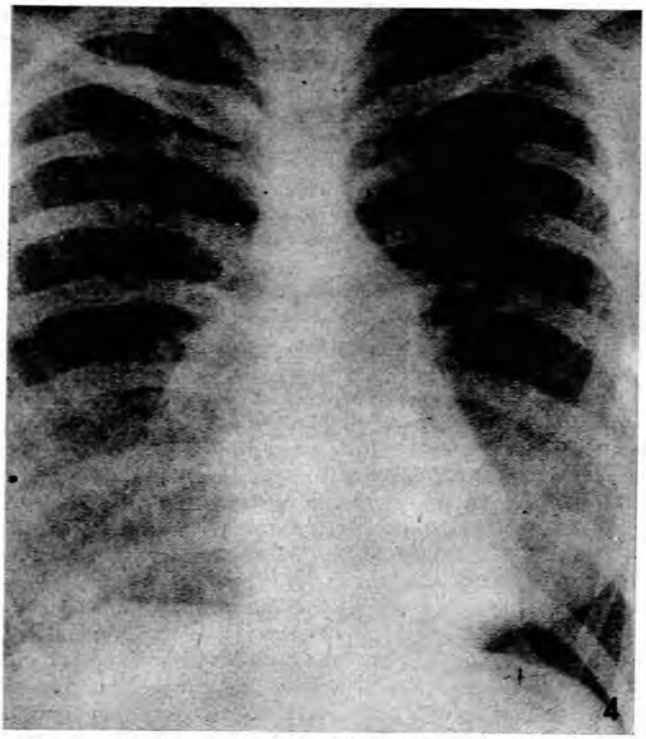

Fig. 4. Radiograph showing minimal 'plethora' of the lungs.

this was not a notable feature in the majority of our cases, the average grade being only 1·3 (Fig. 4). Since in most cases of atrial septal defect the pulmonary-artery pressure is normal or only very slightly elevated, the pulmonary vascular resistance must be diminished. This will allow a greater rate of flow through the lungs, and pulmonary blood volume may therefore not be increased proportionately to the increased flow. Much of the increased volume, too, will be taken up in the arteriolar and capillary bed. Hence, marked plethora would not occur. However, were the pulmonary-artery pressure to be moderately elevated, these factors would not apply, and pulmonary plethora would be more marked. Moreover, at low pressures, as exist in the pulmonary circuit, arterial distensibility increases strikingly with increased pressure (Fig. 5). Thus, in those patients with pulmonary-artery systolic pressure above 40 mm. Hg, the average grade of plethora was 2·1 (Fig. 6).

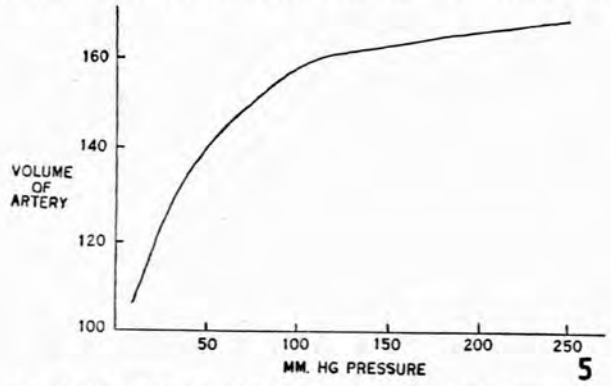

Fig. 5. Relationship of arterial volume to blood pressure.

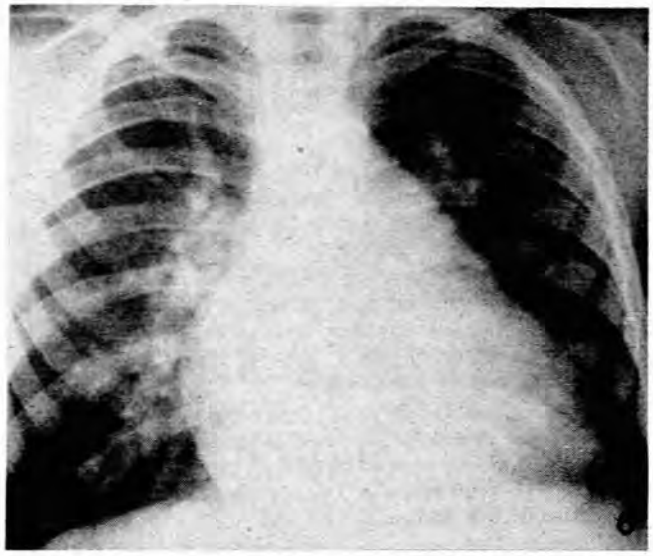

Fig. 6. Radiograph showing well-marked pulmonary plethora in a patient with moderate pulmonary hypertension.

5. '*Pulmonary hypertension picture*'. The striking attenuation of peripheral pulmonary arteries characteristic of pulmonary hypertension was noted in 8 patients (Fig. 7).

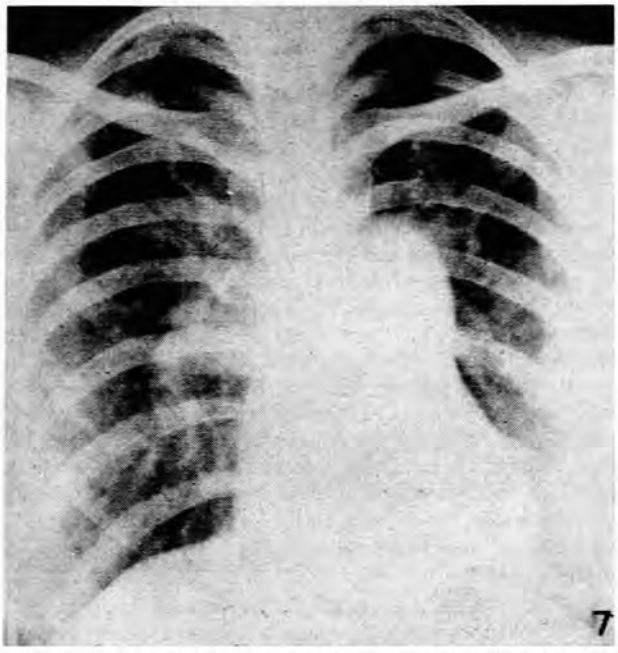

Fig. 7. Radiograph showing striking attenuation of peripheral pulmonary arteries in a patient with severe pulmonary hypertension.

In 3 of them pulmonary systolic pressure was over 65 mm. Hg, in 1 it was 45, and in the other 4 (all infants) cardiac catheterization was not performed. Pulmonary 'wedge angiography' showed a typical 'leafless tree' appearance in these cases (Fig. 8).

6. *Ventricular enlargement* was noted on the right side in most cases, usually of moderate degree, but marked in

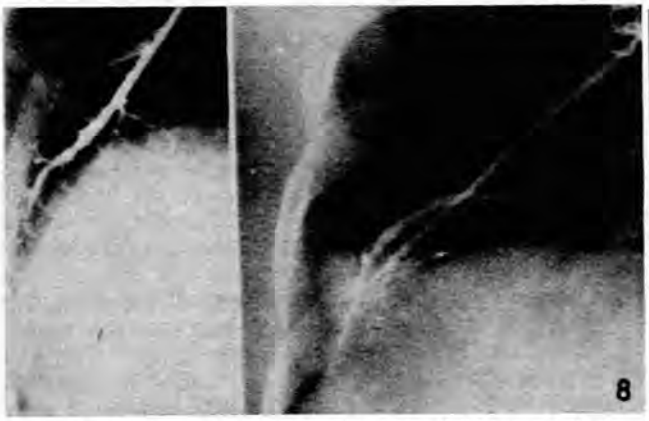

Fig. 8. Pulmonary 'wedge' angiographs (left) in a patient with severe pulmonary hypertension and (right) in a patient with normal pulmonary arterial pressure.

those in congestive heart failure. Left ventricular enlargement was difficult to assess, but was felt to be definite in 4 patients, all with primum defects.

Phonocardiography

This procedure was a valuable auxiliary to auscultation, particularly for determining the degree of splitting of the 2nd heart sound and the presence or absence of an ejection click (Fig. 9).

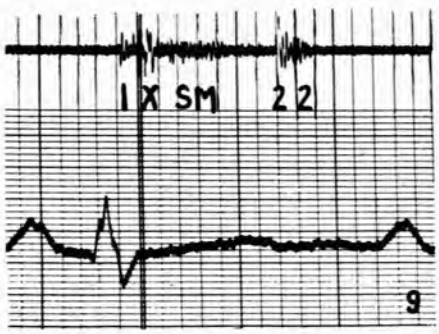

Fig. 9. Phonocardiogram demonstrating systolic ejection click.

Cardiac Catheterization

Cardiac catheterization was performed in 28 cases, by the usual techniques, the catheter being introduced *via* a vein in an arm, usually the left arm. In 16 cases the catheterization was personally performed by the authors, and in 13 of these the catheter could be passed *via* the septal defect into the left atrium (Fig. 10). Of the 3 cases where we did not enter the left atrium, 2 were *not* thought clinically to be atrial septal defects and exploration was not thorough, while in the 3rd a paroxysm of atrial tachycardia precluded exploration of the septum.*

The following features were analysed from data obtained at cardiac catheterization:

1. Pulmonary-artery pressure (mm. Hg).
2. Pulmonary arterial resistance; arbitrary units, viz.

* Since this paper was presented we have catheterized 12 more patients *via* the left arm and in each case we have been able to introduce the catheter into the left atrium and frequently into the left ventricle.

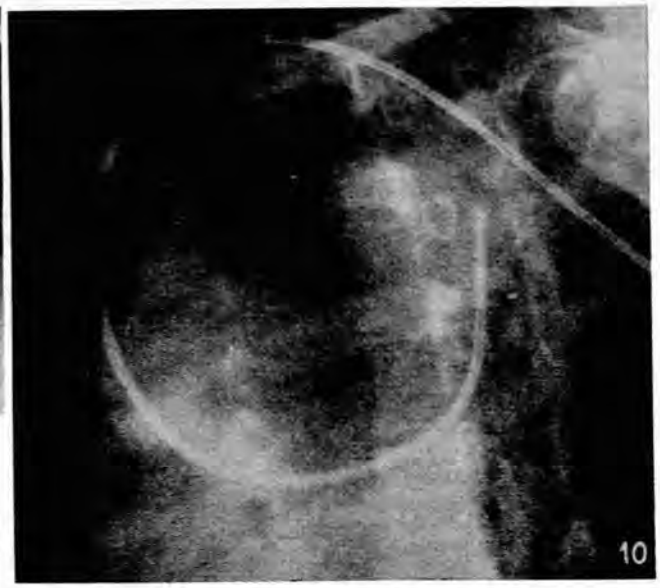

Fig. 10. Left anterior-oblique radiograph showing cardiac catheter in a pulmonary vein, *via* the atrial septal defect and left atrium.

$\frac{MPA - MPCP,}{CO}$ where MPA = mean pulmonary arterial pressure, MPCP = mean pulmonary 'capillary' pressure or 'wedge' pressure, and CO = pulmonary blood flow in litres per minute.

3. Degree of left-to-right shunt expressed as the ratio shunt flow: pulmonary flow.
4. A pressure gradient across the pulmonic valve.
5. Right atrial mean pressure (mm. Hg).

The haemodynamic data are summarized in Table IV.

TABLE IV. HAEMODYNAMIC DATA

Type of defect	*Pulm. art. press. (mm. Hg)**			*R. atrial press. (mean) mm. Hg**	*Shunt flow/ pulm. flow*	*Pulm. art. resist-ance (units)*	*Pressure gradient at pulm. valve mm. Hg*
	Sys.	*Dias.*	*Mean*				
S	38	22	30	8	0·5	4·0	0
S†	56	40	48	8	0·81	2·5	0
S	35	15	21	4	0·5	2·0	0
P	50	25	38	7	0·5	10·0	0
S	35	5	20	2	0·8	1·4	3
S	102	50	66	7	0·44	29·0	0
S	25	3	10	0	0·30	2·0	0
S	30	12	19	4	0·8	0·4	12
S	30	12	20	2	0·8	1·0	0
P	52	35	40	4	0·7	4·0	0
P	25	8	14	5	0·65	1·5	20
S	25	8	15	5	0·64	0·7	0
P	35	15	25	5	0·88	1·5	0
S	25	10	14	7	0·7	1·0	0
S	38	13	25	5	0·73	5·0	0
S	60	‡	‡	4	‡	‡	‡
P	70	20	42	0	0·7	16·0	0
S	30	10	18	1	0·8	1·1	0
S	23	8	15	2	0·61	2·0	0
S	28	10	17	0	0·7	1·6	0
S	50	22	35	5	0·7	7·0	5
S	45	20	30	12	0·74	1·8	0
S	38	10	20	4	0·9	0·4	0
S	38	16	24	4	0·57	2·0	0
P	30	14	20	3	0·8	1·6	0
S	28	7	17	5	0·65	1·4	0
S	65	35	48	4	0·7	8·0	0
S	28	5	15	3	0·6	3·0	8

S — Secundum. P — Primum.

* Reference point for all pressures is mid-axillary line.

† Plus mitral incompetence.

‡ Pulmonary artery not entered.

1. *Pulmonary-artery pressure.* Pulmonary-artery systolic pressure above 40 mm. Hg was found in 6 of 22 secundum defects (27%) and in 3 of 6 primum defects (50%). In 2 patients the pulmonary-artery systolic pressure was equal to systemic systolic pressure, measured simultaneously, and both patients showed well-marked right-to-left shunt at atrial level. The ages of the patients with pulmonary hypertension ranged from 2½ years to 39 years. Repeat catheterization was performed in 2 of the patients with pulmonary hypertension. In 1, a second study 1 year after the first showed a rise of systolic pressure from 65 to 73 mm. Hg. In the other, corrective surgery was performed 3 years after catheterization had shown a systolic pressure of 50 mm. Hg. As she failed to improve after complete closure of the defect, repeat catheterization was performed, which showed a systolic pressure of 75 mm. Hg. There is thus evidence that pulmonary hypertension in patients with atrial septal defect may be a rapidly progressive phenomenon. This evidence is further substantiated by clinical evidence in a 3rd patient, with the highest pressure in our series (systolic 102 mm. Hg); 6 months before catheterization, the pulmonic element of the 2nd heart sound was only moderately accentuated, but at the time of catheterization it was extremely loud.

2. *Pulmonary arterial resistance* was above 5 units in 5 patients, all with well-marked pulmonary hypertension, and in one of them* it was extremely high (29 units). This patient had severe pulmonary hypertension, with well-marked bi-directional shunting of blood, clinical cyanosis, and clubbing of the fingers. There was no direct correlation between pulmonary resistance and either pulmonary flow or pulmonary arterial pressure. However, all 5 patients with pulmonary arterial resistance above 5 units had mean pulmonary arterial pressures of more than 30 mm. Hg, whereas only 2 of 22 patients with pulmonary arterial resistance below 5 units had pressures above 30 mm. Hg (Fig. 11). There is, therefore, evidence that pulmonary

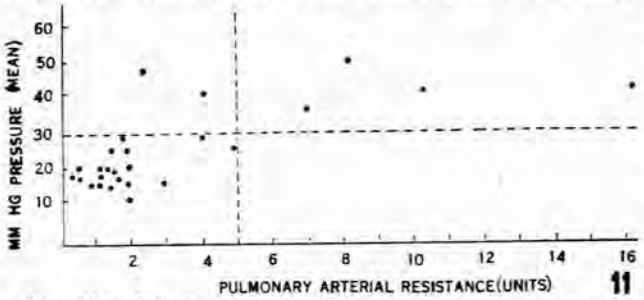

Fig. 11. Relationship of pulmonary arterial pressure to resistance.

hypertension is due to increased pulmonary arteriolar resistance. Paucity of material for histological examination does not permit us to say whether the increased pulmonary arteriolar resistance is due to organic vascular disease or simply increased tone.

3. *Degree of left-to-right shunt.* In all but 2 of the patients catheterized, shunt flow constituted 50% or more of pulmonary flow. In one of these 2 patients the defect was regarded as being small, while in the other, in whom operation confirmed a large defect, the small left-to-right shunt was attributable to severe pulmonary hypertension.

* Not shown in Fig. 11.

4. *A pressure gradient across the pulmonary valve* was found in 5 patients, ranging from 3 to 20 mm. Hg. The degree of gradient was not proportional to the amount of shunt or total pulmonary-artery flow.

5. *Right atrial mean pressure* higher than 5 mm. Hg was found in 6 patients, 5 of whom showed clinical evidence of congestive cardiac failure. In those patients the average pulmonary-artery systolic pressure was 53 mm. Hg, while in those whose mean atrial pressure was less than 5 mm. Hg the average pulmonary artery systolic pressure was 37 mm. Hg. However, 2 of the 6 with elevated right atrial pressure had normal pulmonary-artery pressures, while 5 of 22 with normal right atrial pressures had pulmonary hypertension. In those cases where the left atrium was entered, the left atrial mean pressure was found to be equal to the right atrial pressure, or up to 3 mm. higher. Anomalous entry of a pulmonary vein into the right atrium was shown to be present in only 1 patient in our series; it was not diagnosed pre-operatively.

TREATMENT

Seventeen patients in the present series were subjected to surgical treatment for closure of the septal defect, 1 by a 'closed' technique, and 16 by open-heart surgery with either hypothermia or extracorporeal circulation.*

Four of the 17 patients died. They comprised one patient with a primum defect who was subjected to surgery against our advice and who died after closure by a closed technique; one patient with a complete common atrioventricular canal who died immediately after repair under extracorporeal circulation; one patient in gross congestive heart failure who died from haemorrhage due to a bleeding diathesis several hours after closure under extracorporeal circulation; and one patient with equalization of pulmonary and systemic blood pressures and well-marked bi-directional shunt, who died several hours after closure of the defect under hypothermia.

Of the 13 survivors, in all but one the result has been good, with a follow-up of 3 - 16 months. The one patient who failed to improve was found at re-catheterization to have persistent pulmonary hypertension but no residual shunt.

DISCUSSION

Atrial septal defect is a common congenital cardiac anomaly, the diagnosis of which is usually possible on clinical grounds and can be confirmed by cardiac catheterization. In early infancy the clinical picture is atypical, and the anomaly should be considered in all infants with evidence of heart disease.

The mortality rate in infancy is high. In the patients who survive infancy, the presence of primum defect indicates a persistently poor prognosis, and so does the advent of pulmonary hypertension in those with secundum defects. Even in those with secundum defects and normal pulmonary-artery pressures, the life span is limited and cardiac failure may develop in the 4th and 5th decades of life. The surgical repair of secundum defect in patients without pulmonary hypertension or cardiac failure has an extremely low mortality,[4] but once these two complications develop the risks of surgery increase alarmingly.[5] For these reasons, operation should be advised *on diagnosis* in uncomplicated secundum

* See article by Mr. D. Fuller, at page 797 of this issue, which deals with the 16 cases operated on by 'open' technique.

defects. There is no justification for awaiting the development of symptoms. As surgery produces an anatomical cure, there is every reason to predict a normal life span in these cases.

The risks of surgery are significantly greater in primum defects, but the natural history of the disease is such as to warrant facing this higher risk.

We believe, in conclusion, that atrial septal defect should be likened to patent ductus arteriosus and that early diagnosis and early treatment are strongly indicated.

SUMMARY

The clinical, electrocardiographic, phonocardiographic, radiological and haemodynamic features in 43 cases of atrial septal defect are described. The prognosis of the disease is discussed and the safety of surgery in uncomplicated cases is stressed. A plea is made for early diagnosis and treatment of this defect.

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