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INTERATRIAL SEPTAL DEFECTS*

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Atrial septal defects occur as the result of abnormal development or incomplete fusion of one or more of the septal constituents. The 3 varieties arise as follows:

1. The septum primum grows down from the dorsocephalic wall of the common atrium to join the upper margin of the atrioventricular cushions which have grown in from the common atrioventricular ring to divide it into tricuspid and mitral valves and themselves to fuse with the interventricular septum. Incomplete development of the septum primum will result in the 'ostium primum' type of interatrial septal defect, while any retardation of the growth of the atrioventricular cushions will produce a partial or complete form of atrioventricularis communis.

2. An ostium secundum develops as a fenestration of the septum primum. This is generally closed by the septum secundum, which grows as a curtain over the ostium. Abnormal development of the septum secundum will result in the ostium secundum remaining patent.

3. The 'sinus venosus' defect is the result of abnormal development of the sinus venosus and lies close to the superior-vena-caval orifice.

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Morbid Anatomy of Atrial Septal Defects

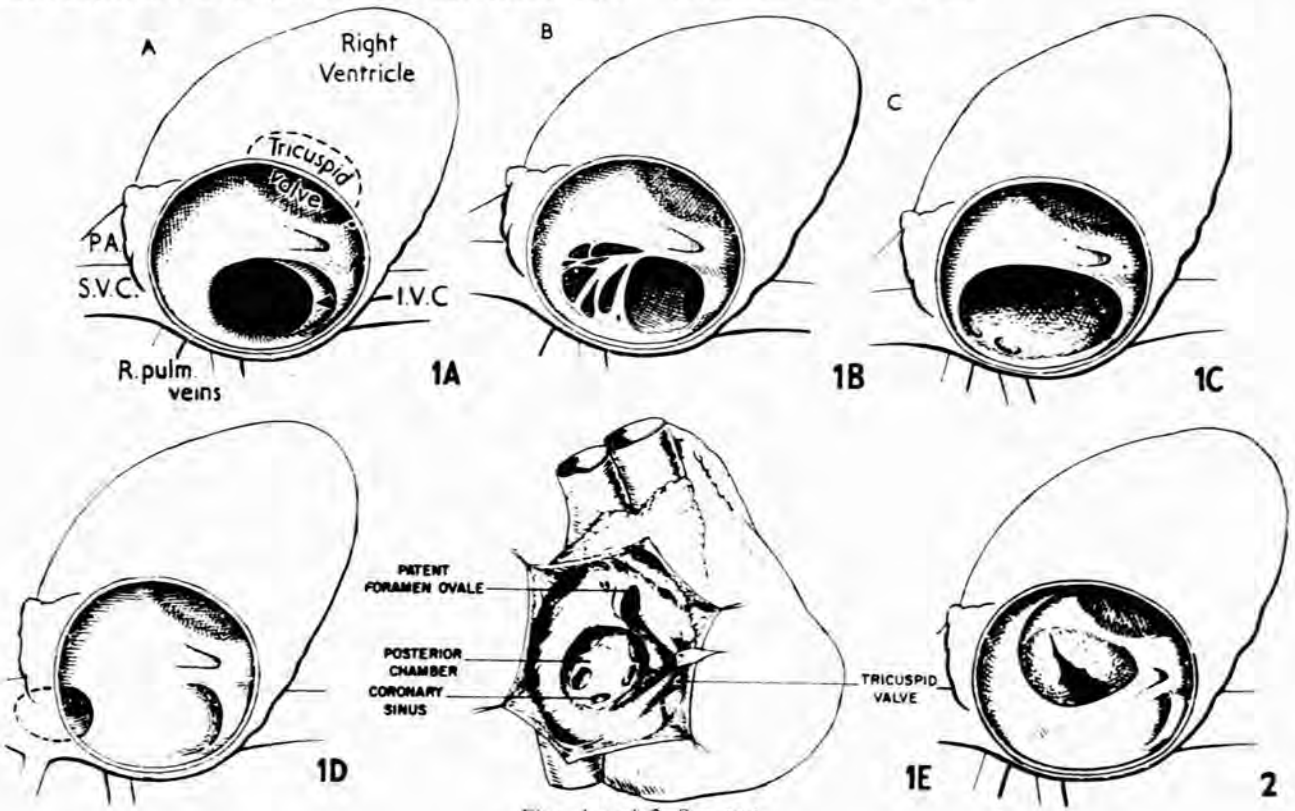
1. *'Septum Secundum' Defects, with which are Included Defects in the Foramen Ovale*

(*a*) Partial defect in the valve of the foramen ovale, which may be represented by a fenestrated membrane (Fig. 1 B).

(*b*) Complete absence of the valve of the foramen ovale. In this the lower margin may be continuous with the valve of the inferior vena cava and anomalous pulmonary venous drainage is a common association (Figs. 1 A and 1 C).

(*c*) 'Sinus venosus'-type of defects. These are situated high in the septum immediately caudal to the entrance of the superior vena cava and frequently have anomalous veins from the right lung draining into the right atrium or even into the right superior vena cava (Fig. 1 D). The venous drainage from the left lung may return to a persistent left superior vena cava or to the coronary sinus.

(*d*) Combined defects, perhaps with total anomalous venous drainage (Fig. 1 E).



Figs. 1 and 2. See text.

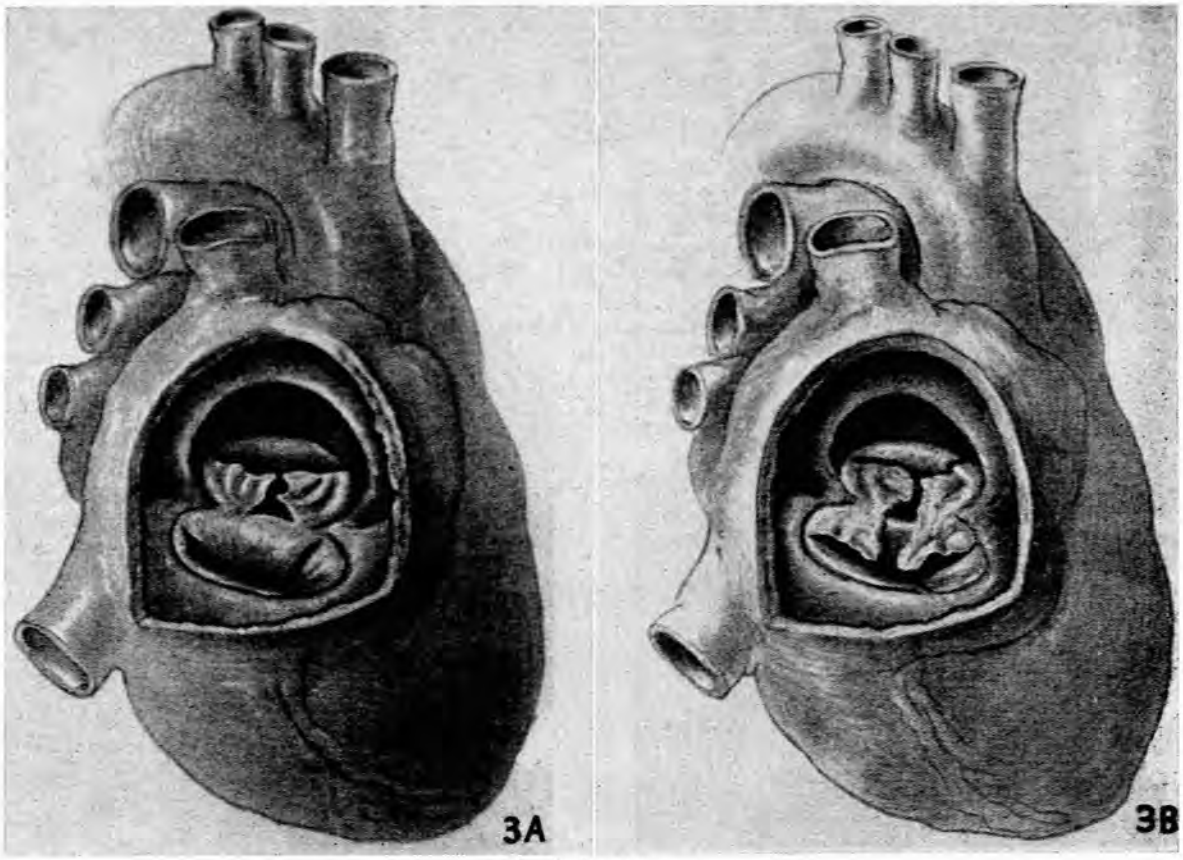


Fig. 3. See text.

2. *'Septum Primum' Defects*

These are less common. They are situated in the lower portion of the septum in front of the orifice of the coronary sinus and their inferior rim is the upper edge of the ventricular septum (Fig. 2). In some classifications these are included as a sub-group of the following category.

Of surgical significance in this group is the proximity of the AV conducting bundle to the posterior free edge of the defect.

3. *Atrioventricularis Communis*

(*a*) Partial. The interventricular septum is intact but, associated with the 'ostium primum' defect, there is a cleft or defect in the septal cusp of either or both mitral and tricuspid valves (Figs. 3 A and 3 B).

(*b*) Complete. There is a defect of the upper portion of the ventricular septum as well.

Associated Lesions

Atrial septal defects of the 'secundum' type often occur as isolated phenomena. The association of atrial septal defect with the tall asthenic build, high arched palate, and arachnodactyly, is too well known to elaborate further. A patent foramen ovale may be of no haemodynamic significance unless an accompanying obstruction to the pulmonary outflow causes raised right atrial pressure and the shunting of desaturated blood to the left atrium. As the fifth component of the pentalogy of Fallot it has no great significance symptomatically but naturally adds to the complexity of the repair. From case reports in the literature the association with mitral stenosis (Lutembacher's syndrome) would appear to be uncommon. Simultaneous separate defects in atrial and ventricular septa occur frequently; so do associated anomalous pulmonary veins. Indeed any congenital cardiovascular abnormality may have an associated interatrial defect.

The 'primum' type of defect, by its very nature, is often associated with abnormalities in the development of both mitral and tricuspid valves as well as of the ventricular septum.

Symptomatology and Diagnosis

It is not the purpose of this communication to discuss either symptomatology or diagnosis.* Suffice it to say that in the neonatal period atrial septal defect occurring alone may produce severe cardiac symptoms and lead to early death, but it is more usual for the secundum defect to remain undetected for a number of years, although there may be radiological evidence of considerable cardiomegaly. In spite of being asymptomatic it is noticeable how many of these patients have had no inclination to participate in strenuous activities. Progressive symptoms of disability generally commence in the third decade and continue for several years, until a severe degree of pulmonary hypertension destroys any chance of effective

* See article by Drs. Zion, Bradlow and Braudo at page 810 of this issue.

TABLE I. RESULTS OF OPERATIVE TREATMENT IN THE FIRST 23 CASES

Case	Age	Sex	Date	Method	Defect	Pressures RA	Pressures RV	Pressures PA	Result	Comment
1.	9	M	3.2.58	Hypothermia	Secundum + anom. R. pulm. veins	5	54/4	44/12	Cured	See Table II
2.	14	F	4.6.58	Hypothermia	Secundum	—	28/7	—	Cured	
3.	27	F	22.8.58	Pump	Secundum (*a*)	—	—	—	Good	Portion of defect not closed
4.	39	F	9.9.58	Pump	Secundum (*b*)	—	40/0	45/20	Died	Postop. haemorrhage (*e*)
5.	9	F	18.9.58	Pump	Secundum + anom. veins (*c*)	8/2	55/0	55/22	Persistent hypertension (*f*)	
6.	11	F	29.10.58	Pump	Secundum + MI	8(*d*)	56/20	56/42	Cured	
7.	19	F	4.11.58	Hypothermia	Secundum	6/0	34/5	28/5	Cured	
8.	2	M	12.11.58	Pump	Total anom. pulm. veins	8/2	54/0	45/11	Died	Postop. haemorrhage (surg.)
9.	2	M	31.12.58	Hypothermia	Secundum + VSD	11/4	60/25	65/35	Improved	VSD not closed
10.	9	F	7.1.59	Pump	Trilogy	11	185/15	15/8	Cured	
11.	31	F	17.1.59	Hypothermia	Secundum	8/3	29/6	25/9	Cured	
12.	5	F	13.2.59	Hypothermia	Secundum	5/0	25/1	23/8	Cured	
13.	45	F	20.3.59	Hypothermia	Secundum	8/-	40/2	28/12	Cured	
14.	22	F	13.3.59	Hypothermia	Secundum	15/4	100/10	102/50	Died (*g*)	(*h*)
15.	10	M	16.4.59	Pump	AV communis	7/3	35/1	35/15	Died	Haemolysis (*i*)
16.	16	M	28.4.59	Hypothermia	Secundum	—	—	35/15	Cured	
17.	19	F	6.5.59	Hypothermia	Secundum	9/3	50/0	40/15	Cured	
18.	4	F	12.6.59	Hypothermia	Secundum +	6	40/2	45/22	Cured	
19.	9	M	6.8.59	Pump	Trilogy	2	100/12	44/27	Cured	
20.	4	F	20.8.59	Pump	AV communis	10/5	—	50/25	Cured	
21.	16	F	25.8.59	Hypothermia	Secundum	6	30/5	26/12	Cured	
22.	16	F	8.9.59	Hypothermia	Secundum	5/3	42/0	42/10	Cured	
23.	21	F	10.9.59	Pump	Trilogy	10/8	125/4	18/11	Cured	

RA = right atrium. RV = right ventricle. PA = pulmonary artery. MI = mitral incompetence. VSD = ventricular septal defect. (*a*) 4 litres L - R shunt. (*b*) Catheter 1954. (*c*) Catheter 1955 (see Table III). (*d*) Clean pressure. (*e*) Christmas factor. (*f*) See Table III. (*g*) Died suddenly 12 hrs. postop. (*h*) 6 months pregnant. Pulm. artery pressure greater than systemic. See text. (*i*) Too high line-pressure during bypass.

relief. Associated cardiac abnormalities naturally make the symptoms and prognosis much worse.

Natural History of Atrial Septal Defect

During foetal life the normal blood flow passes from right to left through the foramen ovale. After birth the increased pressure in the left atrium closes the valve of the foramen ovale and prevents left-to-right flow. However, if a large defect is present the two ventricles fill from a common chamber and the quantity of blood entering each ventricle depends upon the resistance to filling which each offers. At birth the two ventricles are of equal muscular development and the pressures in the pulmonary and systemic circuits are about the same. With the physiological fall in pulmonary vascular resistance and rise in the systemic resistance more blood passes through the right side than the left. At first the right ventricle dilates and the pulmonary vascular bed increases, but as time passes secondary changes occur in the pulmonary vasculature. This takes time and generally in early life few symptoms develop. As the degree of left-to-right shunt increases, enormous amounts of blood may pass through the pulmonary circuit and perhaps only one-tenth as much passes through the systemic. With the development of pulmonary arteriolar changes the amount of this shunt diminishes and ultimately, in the extreme case with severe pulmonary hypertension, the pressure in the pulmonary artery may actually exceed that in the aorta and blood may shunt in the reverse direction through the defect (see case 14, Table I). The ages at which these changes occur cannot be predicted. In some infants in whom the normal foetal pulmonary vascular resistance persists, death may occur at an early age from congestive cardiac failure. More commonly symptoms develop slowly and may not be severe until the 2nd or 3rd decade. There are some instances of patients living out a normal life span with relative ease, while there are many instances of severe secondary pulmonary hypertension being encountered under the age of 10 years. Swan[2] reports, 'We have seen patients pass from the first group to the second in a period as short as a year or two' (Table III). The significance of this lies in the graver prognosis in the second group and in the possibility that permanent obliterative vascular changes will militate against the beneficial effect that surgery may otherwise have offered. The mean life expectancy is not much over 37 or 38 years.

SURGERY

Assessment for Surgery

The first requirement is adequate diagnosis. In Johannesburg we are fortunate in having the excellent diagnostic facilities and clinical acumen of a first-class privately-run Cardiac and Catheter Unit, as well as the Cardiac Clinic of the Johannesburg General Hospital. These groups of clinicians are able to present the surgeon with a thoroughly prepared assessment of the patient. This includes clinical history, physical signs, ECG, phonocardiogram, and radiology. Final confirmation is obtained from cardiac catheterization when the right heart chambers have been thoroughly explored with the catheter, where anomalous pulmonary venous drainage can be detected

TABLE II. CATHETER DATA IN CASE 1 (J.R., AGED 9, OPERATION 3.2.58)

	Pre-operative		*Postoperative*	
Site	*mm.Hg*	*Oxymetry*	*mm.Hg*	*Oxymetry*
Main PA	44/12	90.5%	28/10	71%
RV	54/4	90.2%	28/2	70%
Mid RA	5	92.5%	4	71%
Low RA		68.0%		65%
Peripheral flow—1·45 litres per minute			No evidence of left-to-right shunt	
Pulmonary flow—7·8 litres per minute				

TABLE III. CATHETER DATA IN CASE 5 (V.S., AGED 9, OPERATION 18.9.58)

	Pre-operative (8.7.55)		*Postoperative (2.7.59)*	
Site	*mm.Hg*	*Oxymetry*	*mm.Hg*	*Oxymetry*
Main PA	50/22	76%	70/50	56%
RV	55/0	78%	70/5	56%
Mid RA	8/2		8	56%
Low RA		80%		58%
Brachial artery			100/70	

with a high degree of certainty and where shunts can be picked up and pressures recorded. Other associated anomalies are confirmed or excluded. Should circumstances require it, selective angiocardiography can be performed. The end-result of these investigations is the estimation of the flow across the defect, the pulmonary vascular resistance and the degree of pulmonary hypertension, if any.

The amount of flow and degree of pulmonary hypertension are of prime importance. A large left-to-right shunt is an encouraging finding and is an indication of increased flow through the lungs, while a raised pulmonary vascular resistance diminishes this flow and is a pointer to probable structural pulmonary vascular changes. So important are these changes that, when they become severe, corrective surgery is unlikely to succeed and, indeed, may be dangerous. Dammann and Ferencz,[3] in a study of hypertensive vascular disease complicating congenital cardiac anomalies, state: 'We do not know when, in the course of the disease, pulmonary vascular changes become so severe that operation is no longer indicated'.

It is because we do not know when pulmonary vascular changes may develop that surgery is recommended in an early phase; it should not be postponed until symptoms progress, as is so often done. When the vascular changes have passed from the phase of intimal fibrosis associated with medial hypertrophy to one of dilatation, as is found in grade-4 and grade-5 pulmonary hypertension, then closure of the defect is unlikely to result in lowering of the pulmonary blood pressure. In the Mayo Clinic series of cases with hypertension, the ratio of pulmonary systolic pressure to systemic pressure rose in all of the grade-5 cases after closure of the defect.[4] It is also stated from the Mayo Clinic: 'Analysis of accumulated experience with the repair of atrial septal defect in 119 adults has revealed several factors determinable pre-operatively that appear to have a strong influence on the operative risk. These factors are related to the presence of pulmonary vascular disease or heart failure and contribute importantly to the selection of patients for operation'.[5] They also found that if congestive heart failure had occurred the mortality was 39%, whereas in its absence the rate was 7%. Patients whose right atrial pressure reached a peak of 15 mm.Hg or more had a 50% hospital mortality; so did patients with a right-to-left shunt of 10% or more. They list 5 factors in determining the influence of abnormal values on the outcome of an operation, viz. (*a*) congestive cardiac failure, (*b*) markedly elevated right atrial pressure, (*c*) a large right-to-left shunt, (*d*) severely elevated pressure in the pulmonary artery, and (*e*) markedly increased pulmonary vascular resistance.[5] If none of these unfavourable factors, or only one of them, exists, the risk of operation is well acceptable. The risk is high if 2 of them are present, and extreme if 3 or more coexist.

It is generally accepted that a diagnosis of an uncomplicated lesion in patients under 40 years of age is sufficient to be a firm indication for surgery irrespective of whether symptoms are present or not.

In Swan's series[2] of over 100 cases only 1 death occurred in a patient under the age of 18 years, while Kirklin[6] reported no death under the age of 20. These figures are comparable with those of Holmes Sellors.[7] In view of these low mortality figures the decision is not *whether* surgery should be advised, but *when*. In the absence of clear contra-indications surgery should be advised in all cases. Swan[2] has stated that when pulmonary blood flow is no more than 1 litre per minute higher than the systemic, serious doubts about the advisability of surgery should be entertained. Unfortunately, when such a problem arises, knowing that no other treatment will help the patient, the surgeon is often persuaded to operate in the hope of affording some measure of relief; and more often than not this does not materialize (see case 14, Table I).

Operative Techniques

As with most surgical procedures, the techniques have passed through varied evolutionary phases. Older and cruder methods have given way to the present direct approach. It will serve no purpose to enumerate the many earlier types of operation. At present it is almost universally accepted that direct visualization of the defect with occlusion of the venous inflow is the most desirable method. This can only suffice under conditions of hypothermia or with the use of the pump-oxygenator. The first closure of an atrial septal defect by direct suture in a bloodless field was performed by Lewis, using hypothermia, in 1953. Since that time many thousands of such defects have been closed successfully, with a mortality in the good-risk patient of about 1%.

Hypothermia has the advantage of simplicity and does not require the elaborate set-up that is used with a pump oxygenator. It has, however, the great disadvantage that the surgeon is gravely limited in time. At the usual tempe-

ratures employed, 6 - 8 minutes of circulatory occlusion is all the time available. This may not allow of the more complicated defects, or associated defects, being effectively cured. Nevertheless in the usual 'secundum' type of interatrial defect, which comprise about 90% of such defects, the allotted time is quite adequate for complete closure. If need be, after waiting a sufficient time to re-establish the circulation, the surgeon may make a second entry.

Pump oxygenation should be employed in all cases where the diagnosis may be in doubt, in all cases of 'ostium primum' or 'atrioventricularis communis' defect, and where other abnormalities coexist. It cannot be denied that this method may be the ultimate ideal in all cases of interatrial defect. However, for two reasons we have not been using it, thus: Firstly, where the diagnosis is not in doubt, even though one has to work against time, direct closure under hypothermia can be effected completely and with small risk.[2,7] Secondly, with our present available opportunities, where we have a long waiting list of more complicated conditions, precedence with the pump must be given to these. Possibly if we are able to eliminate our present backlog of cases (not a very likely prospect) we may change. We have indeed operated upon 3 pure secundum defects using the pump, but this was early on when we chose to operate upon uncomplicated abnormalities. Two further cases in whom the diagnosis was in doubt have been operated on using the pump.

Results of Operative Treatment

The first case of atrial septal defect was operated upon under hypothermia on 3 February 1958. Since then 27 more cases have been operated upon under hypothermia and 12 with pump oxygenation.

Two cases have been catheterized after operation and their pre- and postoperative findings are presented (Tables II and III). These two cases illustrate dramatically the complete restoration to normality in J.R. and the persistence of pulmonary hypertension in V.S., whose condition was allowed to persist too long before surgical correction.

SUMMARY

Twenty-eight cases of secundum defect have been operated upon under hypothermia, 1 of whom has subsequently been found to have an associated ventricular septal defect. In 13 cases the defect has been closed with the aid of the pump oxygenator — 5 of these having uncomplicated secundum defect; 1, secundum defect associated with rheumatic mitral incompetence, which was corrected at the same time; 3, secundum defects associated with pulmonary valvular stenosis; 3, atrioventricularis communis; and 1, total anomalous pulmonary venous return.

Among the pure secundum defects 2 patients have died, one under hypothermia, the other with the pump. The first (case 4, Table I), who died a haemorrhagic death[8] after being adequately repaired on the pump, was a 39-year-old woman who had pulmonary hypertension and severe disability, could climb only 8 steps, and had been having haemoptysis for 6 months. The other was a 22-year-old woman, 7 months pregnant, weighing 78 lb, who had an extreme degree of pulmonary hypertension, the pulmonary-artery pressure being 10 mm.Hg higher than the systemic, with a left-to-right flow of half a litre per minute; in the light of present knowledge she should not have been operated upon.

Of the 3 cases of atrioventricularis communis, 1 died of a technical fault during the bypass phase of the pump. The child with total anomalous venous drainage died from a technical fault connected with the surgery. These 2 deaths should have been avoided. However, in cases of pulmonary hypertension high mortality must be expected if the operation is performed.

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