OPEN-HEART SURGERY IN ATRIAL SEPTAL DEFECT*

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Atrial septal defects are probably the commonest of the congenital heart lesions, and as the name indicates they consist of some form of abnormal communication between the two atria. The defect may occur as a hole in the interatrial septum itself or, more complexly, as a hole in the lower part of the interatrial septum where it merges into the ventricular septum. This latter may be due to an additional defect of the ventricular septum, and of the leaflets of the tricuspid and mitral valves, which are derived from the endocardial cushions.

An atrial septal defect may therefore be regarded as being interatrial or atrioventricular (an endocardial cushion defect). The different terminology that is applied to these two types, according to embryological descriptions, tends to make the subject unnecessarily difficult to understand.

The defect in the atrial septum may be large or small, single or multiple. Occasionally, it may resemble a cribriform plate instead of a simple hole. If the defect is very large the appearance may be that of a single atrial cavity, the only sign of the septum being a faint rim on the atrial wall.

It is important to remember that an abnormality of the pulmonary veins may coexist with the developmental atrial defect. The drainage of these veins may be anomalous, i.e. they may drain into the right atrium instead of the left; or may appear to be anomalous in the case of a very large atrial septal defect, the impression being that the pulmonary veins are opening into the right atrial cavity. Occasionally the right superior pulmonary vein drains directly into the superior vena cava.

The presence of an atrial septal defect is of no importance during intra-uterine life, because the normal course of all the venous blood then entering the heart is to the left side *via* the fossa ovalis.

The left ventricle shows greater development than the right during the first year of the child's life because of the greater resistance encountered in the peripheral circulation. Consequently, many cases of atrial septal defect remain unrecognized for this length of time. Because of the defect, the blood flows from the left into the right atrium, for there is normally a higher blood pressure in the left chamber. The right atrium, which now has to accommodate all the blood it receives from the two venae cavae, as well as this extra blood from the left atrium, enlarges correspondingly. The right ventricle receives an increased flow from the right atrium, and accordingly its action becomes more forceful. The additional blood volume is pumped into the pulmonary arteries, which enlarge and show pulsation as a result of the extra force transmitted from the right ventricle. This explains the left-to-right shunt with increased strain on the right ventricle. The resultant additional flow into the pulmonary arteries is responsible for the systolic murmur.

The enormously increased strain on the right ventricle is often well tolerated, but eventually failure occurs. This event, which takes 20 - 30 years to develop, is of serious prognostic significance. Of course, if the response to the right-sided over-activity is pulmonary hypertension, reversal of the shunt may take place. A balance may be secured for a time but finally a right-to-left shunt develops with cyanosis, which is an ominous portent.

History of Treatment

Recognition of these cases has become a routine matter within the last 2 decades, and today the diagnosis can be made clinically with great accuracy. Great advances have been made in methods of treatment in the last 10 years. The following résumé of the work that has led to present-day techniques should prove of interest:

In 1947 the closure of an experimental defect by Cohn, by suturing the atrial wall to the margin of the defect, was the forerunner of Murray's successful closure of a defect in a child in 1948. In 1950 Swan attempted to plug the hole by the inversion of the auricular appendage. Bailey devised the doughnut-lumen type of atrioseptopexy in 1952, and a similar method was employed by Hufnagel and Gillespie in 1951, when they used a plastic button to close the opening. Sondergaard described a technique of defect-closure by insertion of an external circumferential suture, and Gross showed that it was possible to work in the left atrium through a rubber well attached to the atrial wall.

Many patients were treated by these methods, but it was obvious that a more accurate means of closure was necessary because of the great variability in size, number and position of these defects. In 1953 Lewis and Taufic were able to visualize and close defects under direct vision, utilizing hypothermia and inflow stasis of blood. This method allowed them about 7 minutes in which to operate, during which time all blood flow to the heart was cut off by ligatures around both venae cavae and a clamp across the aorta and main pulmonary artery. The wall of the right atrium was opened, and the defect was usually accurately and completely repaired. Apart from the time-limit set the surgeon, the disadvantage, of course, was that during this time all cerebral circulation ceased. Simple defects, even if large, could be closed by this procedure with absolute certainty. More complicated cases, however, with venous anomalies or with endocardial cushion defects, could not be treated in the short time at the surgeon's disposal.

Extracorporeal circulation now gives the surgeon ample time to correct all types of defect.

CASE INVESTIGATION

Clinical Signs

Many cases are known to have had heart murmurs for several years. These may have been discovered in a routine examination, or an examination for some unrelated complaint. Schoolchildren may show some degree of dyspnoea, or complain that they are unable to keep up with the activity of their schoolmates. In many instances, however, the symptoms are minimal. Some children may show minor degrees of physical retardation, while others are normally developed, the heart murmur being the only sign of any

* Paper presented at the 42nd South African Medical Congress (M.A.S.A.), East London, C.P., September-October 1959.

disability. The pulse may be normal, and this is always a striking finding in the presence of cardiac signs. A precordial chest bulge may be present and a systolic thrill may be palpable. A systolic murmur with a widely-split second pulmonary sound is audible over the third left intercostal space.

A diastolic murmur may be heard if pulmonary insufficiency is present. Frequently a diastolic tricuspid murmur is present as a result of the increased flow through the valve. A diastolic mitral murmur suggests an atrioventricular defect (AV type of defect).

Females are affected twice as often as males.

There is usually no history of maternal ill-health during the pregnancy, and the mother may have had normal children before and after the birth of the affected child.

X-ray Screening

This examination is of the utmost value. The heart is enlarged and hyperactive, and the hyperactivity extends to the pulmonary arteries, which may show increased activity into relatively small branches. In contrast, the left atrium, left ventricle and aorta show lack of prominence. This finding has been named 'hilar dance'.

Cardiac Catheterization

Important information is gleaned from this examination, although the diagnosis of an atrial septal defect can usually be made without it. In many cases, however, an AV-canal type of defect, as distinct from the ordinary intra-atrial type, can only be diagnosed after catheterization.

An anomalous venous drainage may be demonstrated by passing the catheter directly from the superior vena cava into the superior pulmonary vein. The position of this catheter will be seen on the straight X-ray film, and the nature of the defect is confirmed by the high oxygen saturation of the blood samples withdrawn through the catheter. A left-sided superior vena cava may be shown on the AP view in an X-ray film. This finding should be confirmed by lateral views, for the catheter may have been passed down the left internal mammary vein giving the appearance of a left superior vena cava.

The passage of the catheter from right to left will confirm the presence of a communication between the two atria. The catheter will pass at a higher level in a case of ordinary intra-atrial defect than in one with an AV type of defect. The passage of the catheter directly from the left atrium into the left ventricle, pressures, position of catheters, and oxygen saturations, all provide confirmatory evidence of the nature and position of the defect.

It is important to remember that though catheterization often demonstrates the anomalous pulmonary veins, the absence of this finding does not preclude their existence.

The surgeon is thus usually provided with an accurate pre-operative picture on which he can base his plan of action.

Electrocardiograph

ECG tracings usually demonstrate partial or complete right bundle-branch block, or right ventricular hypertrophy. Signs of left ventricular hypertrophy or complete heart block are suggestive of an atrioventricular type of defect.

OPERATION FOR ATRIAL SEPTAL DEFECT

The operation is performed under *cardiopulmonary bypass.*

The anaesthesia is a combination of pentothal, relaxant, gas, oxygen, and fluothane, and should be sufficiently light to allow the patient to become wakeful immediately after the operation.

The patient is placed in a supine position with a narrow pad between the shoulders, and a mid-line sternum-splitting incision is made. The sternum is divided by a Gigli wire saw or an electric motor saw, and the pleural cavities are not opened. Alternatively, a right posterolateral incision through the one pleural cavity or a bilateral transthoracic incision through both pleural cavities can be used.

Arterial and venous pressures are monitored through polyethylene tubing in the internal mammary artery and the long saphenous vein. Electrodes in the scalp and on the extremities provide leads to the EEG and ECG machines respectively.

Arterial cannulae of suitable size are prepared, and the pump oxygenator is calibrated according to the patient's weight.

The heart is exposed by a long incision in the pericardium so that good access is obtained to both venae cavae. Appropriate venous catheters are prepared.

Preliminary investigations may have suggested the presence of anomalous veins, but one must remember that the absence of any pre-operative evidence does not exclude their presence.

A persistent left-sided vena cava allows uncontrollable amounts of blood to enter the right atrium during bypass if it is not clamped off. It is thus imperative to examine the left side of the heart as well as the posterior aspect, for if there is such a vein it will enter the coronary sinus.

An aberrant superior pulmonary vein may be seen entering the superior vena cava. Occasionally aberrant pulmonary veins may be seen entering the right atrium, though this abnormality is usually only confirmed when the right atrium has been opened.

The following principles are worth remembering:

1. Anomalous pulmonary veins must have their blood flow diverted to the left atrium.

2. The blood flow from the venae cavae must be diverted to the right atrium. This applies particularly to the inferior vena cava when there is a low-lying defect and a well-developed eustachian valve, which is sometimes mistaken for part of the rim of the defect. During inflow stasis under hypothermia the valve may be caught up and used to repair the defect, so that inferior-vena-caval blood is diverted into the left atrium. This error, of course, is obviated by the use of the cardiopulmonary bypass.

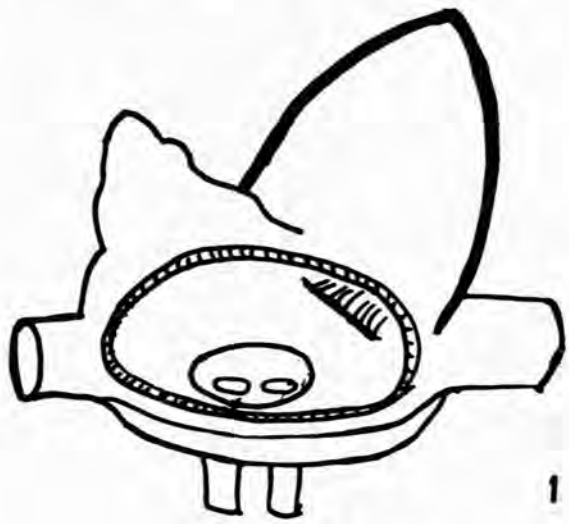
3. A preliminary digital exploration of the defect through the right atrial appendage is always essential. The surgeon may not only establish the presence of mitral stenosis but may also be able simultaneously to perform a digital valvotomy.

4. A piece of prepared ivalon sponge should be ready in case it is required for the closure of a badly situated defect. This applies particularly in the case of an atrioventricular abnormality.

Repair of Interatrial Defects

Interatrial defects can be classified into 3 groups, as follows:

1. *The high or sinus-venosus type* is a defect situated at the lower end of the superior vena cava. This type of defect will require an ivalon patch if the defect extends up into



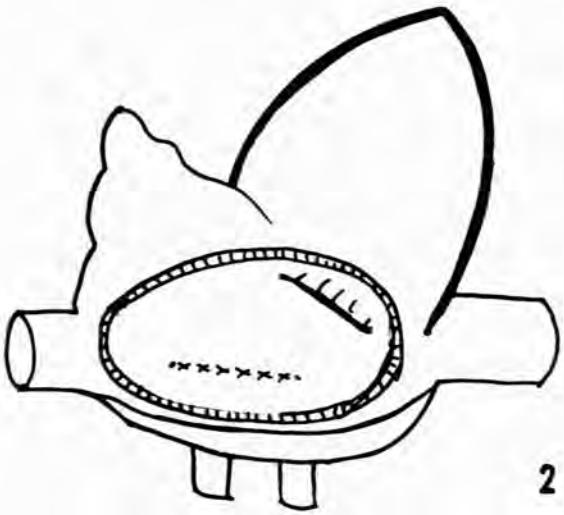


Fig. 1. This illustrates the presence of a septal defect situated in the middle zone of the septum. Through this defect can be seen the opening of the 2 left pulmonary veins, and they open onto the left side of the septum.

Fig. 2. The septal defect has been converted into a horizontal slit by the insertion of a layer of sutures, and when this is over-stitched, the septum is completely closed off.

the superior vena cava. Mattress sutures must be inserted through the wall of the superior vena cava from the outside at the top part of the defect. There is usually a rim of atrial septum in the lower part and the mattress sutures can be inserted from the left atrial side. When the defect has been circumferentially supplied with mattress sutures, the ivalon patch can be slid down into position and fixed. This defect may be associated with an anomalous pulmonary vein, in which case the veins must be diverted before the repair of the defect. Small anomalous veins may be left alone.

2. *The middle type* of defect is probably the commonest variety (Figs. 1 and 2). The defect may be very large, and there is always a rim of atrial septum. This round opening can be converted into a slit-like opening by placing a mattress suture at each end and applying slight traction to each suture. The slit-like opening is then stitched upwards and downwards with a continuous suture, care being taken that a firm hold is obtained with each stitch through the septum. Other openings are treated similarly. An extra opening may be so small that a single mattress suture is sufficient to close it.

3. *The lower type* of defect extends down to the region of the eustachian valve of the inferior vena cava, and there may be no visible rim of atrial wall. An artificial rim can be made by picking up the atrial wall in sutures, which will then help to convert the rounded defect into a slit-like opening. The presence of the caval catheter will be of great help, for it will ensure that the flow from the inferior vena cava is not diverted into the wrong atrium. This is a point in favour of the use of the cardiopulmonary bypass as opposed to hypothermia.

The septal defect in any one of these 3 types may be so large that it is impossible to suture the margins together without causing great tension on the stitches. The stitches are thus likely to tear out and lead to a recurrence of the defect and it is therefore deemed wiser to stitch an ivalon patch over the defect. A piece of previously prepared ivalon is cut into the correct shape and attached to the margins of the defect by means of mattress sutures.

Care must be taken to allow all air to escape from the left atrium before the final stitch is drawn taut; this applies to direct suture as well as to a patch. During the repair operation the suction apparatus must be used to ensure visibility of the site. Sometimes there is quite a large blood flow into the left atrium, either from bronchial arteries which return their blood to the pulmonary veins, or most commonly from the coronary sinus *via* the right atrium. When the blood is removed from the operation site the left atrium may be emptied, and any air which enters may

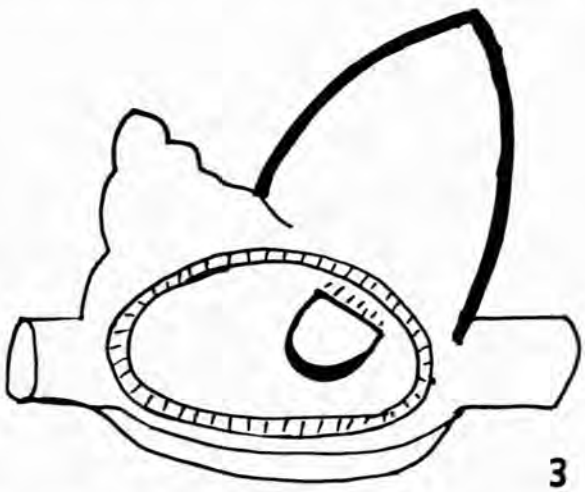


Fig. 3. This illustrates an ostium primum septal defect situated immediately over the tricuspid valve. The tricuspid and mitral valves may be split, in which case the defect would be of atrioventricular-canal type.

pass into the left ventricle. This is extremely dangerous, for this air may be pumped by the ventricle into the aorta, from where it may filter into a coronary artery, or into the cerebral or systemic circulation.

The Atrioventricular Type of Defect

A low defect, with part of its inferior margin formed by the septal leaflets of the mitral and tricuspid valves, is called the atrioventricular type (Fig. 3). These two valves are rarely intact, and their repair must be performed before the closure of the atrioventricular defect. It is usually possible to repair the clefts by simple suturing, but at times it may be necessary to use a narrow strip of ivalon, which can be sewn to both sides of the cleft. It is important to mobilize the two cleft leaflets very thoroughly before repairing them. After repair of the valves the defect in the atrial wall can then be closed by means of an ivalon patch.

Frequently not only are the two valves divided, but there is a failure of development of the upper part of the interventricular septum. This means that there is an interventricular communication as well as an interatrial communication. The valves developed from the cushions of the septum primum are also divided, so that there is an additional communication between the left ventricle and the right atrium. This abnormality can be clearly shown on catheterization, when the catheter can be seen passing from the right atrium directly into the left ventricle through a low-sited defect.

It is important to ascertain that there is no air trapped in the left ventricle when the mitral valve has been repaired. An incompetent mitral valve cannot force the entrapped air in the ventricle into the aorta, but a competent mitral valve after repair will offer such resistance to the entrapped air that it may be forced up into the aorta. This complication is readily recognized on the ECG tracings. The arterial blood enters the patient from the pump oxygenator at the right femoral artery, and the stream of blood is therefore against the normal direction of blood in the aorta. Consider what happens when blood and some air is ejected from the left ventricle through the aortic valves and into the aorta, which is receiving blood in an opposite direction. The mixture will tend to go down the coronaries, and the air in these vessels will show an immediate pattern of damage on the ECG. This accident can be treated by keeping the patient on the pump so that eventually all the air will be driven out of the coronaries, with the consequent restoration of a normal ECG pattern. It is far more serious if air gains entrance to the cerebral vessels. The mitral valve should therefore be rendered incompetent temporarily until the surgeon is reasonably sure that all air has left the ventricle.

The existence of this type of defect gives further support to the theory that heart valves develop from the endocardial cushions. These two cushions develop ventrally, and dorsally, and they then fuse. If there is failure to fuse, then either the left-side valve, i.e. the aortic leaflet of the mitral valve, or the right-side valve, i.e. the septal leaflet of the tricuspid, will remain cleft. In many instances, both remain cleft.

The septum primum grows down to join with these fused cushions, dividing the space into the two atria. Directly beneath the two atrioventricular cushions is the muscular part of the ventricular septum. The interventricular opening is obliterated when these cushions fuse to form part of the membranous septum between the two ventricles.

The malformation may be complete, that is, it may involve both mitral and tricuspid valves, or partial, involving only the mitral valve. The complete form shows interventricular communication as well as insufficiency of both mitral and tricuspid valves. The partial form may show insufficiency of the mitral valve.

Modern treatment of these defects of atrioventricular type is carried out under direct vision and can be performed in two ways, viz. (1) under hypothermia with inflow stasis or (2) under cardiopulmonary bypass.

Hypothermia allows the surgeon a limited time in which to do the repair. The atrium is opened and the defect is visualized and then repaired. Occasionally more time is required, and it may be necessary to have two periods of inflow stasis. Obviously this method can only be employed for the simpler types of interatrial defects. Repair of the more complicated types, particularly the AV-canal type of defect, requires considerably more time, and *cardiopulmonary bypass* becomes essential.

Anomalous Venous Drainage

Anomalous pulmonary veins drain either into the superior vena cava or into the right atrium. The presence of these anomalous veins with an interatrial septal defect increases the difficulty of repair. All pulmonary venous drainage should pass into the left atrium, and closure of an interatrial defect without correction of the pulmonary venous drainage would result in a continuation of the left-to-right shunt; that is to say, the original state would still be present, though to a lesser degree.

The operation for the correction of the anomalous venous drainage must be so devised that the flow from the anomalous veins is directed into the correct atrium, i.e. the left atrium. In the case where the superior pulmonary vein drains into the superior vena cava, the aim of operation is to separate the oxygenated blood in the pulmonary vein from the de-oxygenated blood in the superior vena cava. A row of mattress sutures can be inserted into the superior vena cava, dividing it into two streams. The stream from the pulmonary vein can then be diverted into the left atrium by bringing a flap of atrial wall over the newly-made venous orifice. This flap is formed by making a curved incision in the atrial septum at the new opening of the pulmonary vein, and stitching this flap to the atrial wall. The atrial septal defect is closed in the usual way.

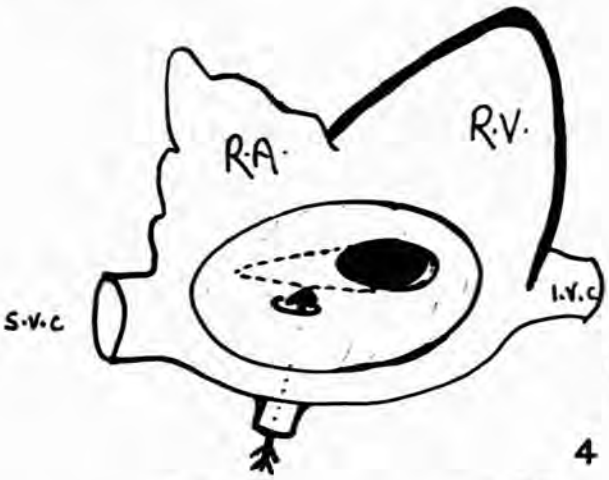
In the case where a pulmonary vein drains into the right atrium, a flap is fashioned from the atrial wall adjacent to the septal defect, and this flap is then stitched to the atrial wall (Figs. 4, 5 and 6).

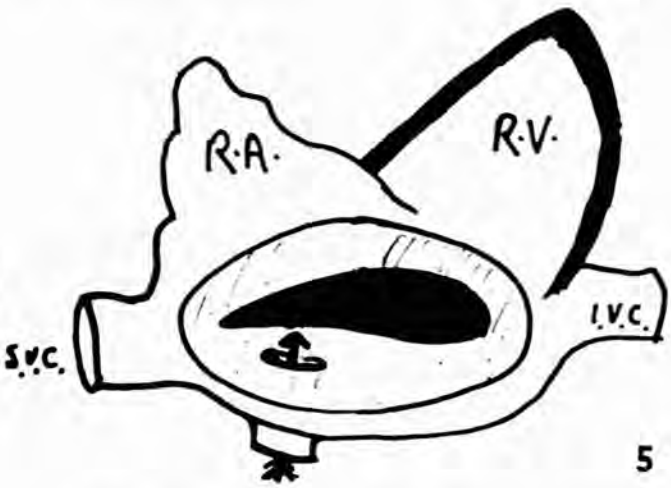
In both cases, if the newly fashioned septal flap is not large enough to be attached to the atrial wall, a section of compressed ivalon can be used as a patch.

The purpose is to ensure that all oxygenated blood is diverted to the left atrium, and that the septal defect is closed.

Completion of Bypass Operation

The use of cardiopulmonary bypass allows the surgeon ample time to examine the defect thoroughly and to close it either by direct suture or by means of ivalon patches. Adequate perfusion and blood oxygenation can be carried out





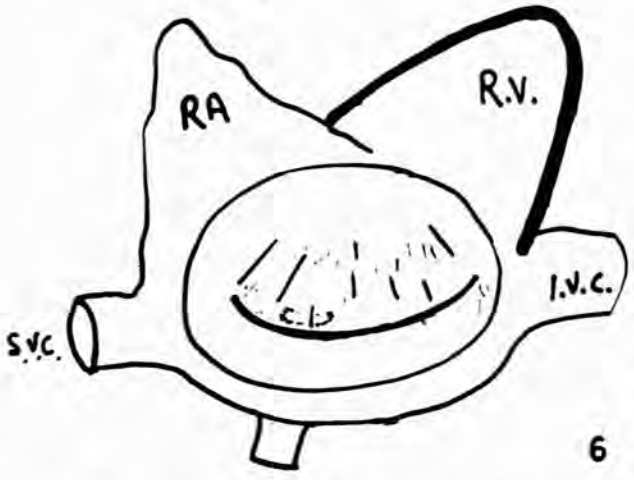


Fig. 4. This shows an oval-shaped defect with an anomalous pulmonary vein opening onto the right side of the septum. The correction of this defect is started by enlarging it along the dotted line.

Fig. 5. The defect has now been enlarged considerably and the upper margin has to be brought over the opening of the pulmonary vein so as to divert its flow into the left atrium.

Fig. 6. This shows the completed repair with the anomalous vein now draining into the correct atrium.

with any of the different types of pump oxygenator. The Cape Town unit at Groote Schuur Hospital uses the bubble oxygenator.

After closure of the defect, the right atrium is allowed to fill with blood while the atriotomy incision is repaired. Haemostasis is all-important, and a careful closure is essential.

When preparations are made to go off bypass, the superior-vena-caval snare is released and then the inferior. The effect on the heart beat is noted, and particularly the appearance of the ECG pattern. If all is well, the machine is stopped and the arterial and venous lines are clamped. The superior-vena-caval catheter is removed and the previously applied purse-string suture is drawn taut and then tied off. The inferior-vena-caval catheter is similarly removed and an extra purse-string suture is usually inserted because this catheter is placed in the wall of the atrium. The superior-vena-caval catheter is placed through the appendage of the atrium.

When all the incisions have been closed off and inspected, the internal-mammary cannula is removed and the vessel is tied off. The chest is then ready for the placing of drainage tubes. The anterior mediastinum should be drained by 2 catheters and, if the pleural cavity has been opened, a drainage tube should be inserted into the cavity.

Dressings are applied, the patient is weighed to check blood loss or gain, and an X-ray film is taken of the chest to confirm that the lungs are fully expanded and the drainage tubes correctly placed.

Postoperative care is not to be discussed in this paper, but it should be stated that these patients remain under constant supervision by senior medical and nursing staff for the first 48 hours of the postoperative period.

ANALYSIS OF CASES

The technique of cardiopulmonary bypass has been used at Cape Town for 52 cases, 20 of which were suffering from atrial septal defects. Of the 20 cases, 16 were in the group of interatrial septal defects, i.e. the 'ostium secundum' group, and 4 were in the 'ostium primum' or endocardial-cushion group. There was 1 death, which occurred in a case of the 'ostium primum' type. All cases catheterized since operation, so far, show complete closure of the defects.

CONCLUSIONS AND SUMMARY

This report deals with the surgical treatment of 20 cases of atrial septal defect. A panorama of treatment is given. The closed methods should not be used today, although the open methods under hypothermia with a limited operating time are still used by some surgeons. Here I have discussed the precise surgical technique which is possible with cardiopulmonary bypass.

The technique which we prefer and is in use at present, viz. cardiopulmonary bypass, allows us to repair completely all types of defect with minimum risk, and without the strain on the surgeon of an imposed time limit.