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SURGICAL TREATMENT OF ATRIAL SEPTAL DEFECTS*

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The treatment of atrial septal defects is essentially surgical. We describe below our experience of the management of a group of 12 cases.

This defect is relatively common. In necropsies on congenital heart cases of all age groups Maude Abbott (1936) found the defect to be the third commonest, and in McGinn and White's series (1936) it was the commonest. Post-mortem examinations in children show a lesser incidence as death often occurs later in life, and Disenhouse *et al.* (1954) found it to be sixth and Keith and Forsyth (1951) thirteenth in frequency. Wood's findings (1950) in 200 patients suffering from congenital heart disease showed atrial septal defects to be the second commonest defect between the ages of 5 and 61 years.

The sex incidence shows a predominance of female patients (Roesler, 1934; Cosby and Griffith, 1949; Bedford *et al.*, 1941).

The morphology of the various types of defect is best understood by a consideration of the embryological development of the septum.

The atrial septum is formed by the fusion and overlapping of two septa which eventually divide the single atrium into two cavities. The first, the septum primum, appears at about the fourth week in the roof of the atrium and grows downwards both ventrally and dorsally in the sagittal plane towards the region of the atrio-ventricular valves and the developing ventricular septum. Growth in its central portion is slower than at the periphery, so that its lower margin becomes crescentic. There is thus left a communication between the two atria which is termed the ostium primum. This foramen then gradually closes, but at the same time the upper part of the septum breaks down to form a second deficiency—the ostium secundum.

To the right of the first a second septum now develops, the septum secundum, which grows downwards to overlap the lower edge of this second foramen. The free edge of the second septum forms the annulus ovalis of the fully developed heart. There remains an oblique cleft bounded above by this free edge and, below and to the left, by the lower edge of the foramen secundum through which blood passes during foetal life from the right atrium to the left. Actual fusion of the two septa occurs before the 12th week of extrauterine life in most cases. Seven types of defect may be described (Fig. 1).

1. A common atrium where there has been no attempt at the formation of any septum or at the most only a small rim of tissue in the sagittal plane.

2. A defect which lies directly above the atrio-ventricular valves and which may vary considerably in size. This represents a persistent ostium primum and arrest of development at this stage.

3. In addition to the persistence of the ostium primum there is a defect in the upper part of the septum which has broken down without prior closure of the ostium primum. The septum secundum has not formed.

4. Multiple small defects are found in the upper part of the septum. This represents the imperfect breakdown of the septum primum in its upper portion after the foramen primum has closed. There is no septum secundum.

5. There is a rounded or oval defect in the upper part of the septum. This is a persistent ostium secundum. There is again either no septum secundum or it is too small to cover the defect.

6. Extensive deficiencies of the septum occur with only strands of tissue crossing it. This may represent either an extreme degree of type 3 or of type 4.

7. There is a small defect beneath the annulus ovalis. This is a persistent foramen ovale.

Pathological Changes

During foetal life blood normally passes from the right atrium to the left. An atrial septal defect therefore does not cause dilatation or hypertrophy of the cardiac chambers *in utero*, and at birth the heart is of normal size and shape. After birth, however, the presence of such a defect results in a flow of blood from the left atrium to the right. This increases the volume passing through the right ventricle, pulmonary artery, and lungs, and the return to the left atrium, which further augments the left-to-right shunt through the defect. The pulmonary arterial flow may be increased up to eight times the systemic flow, which remains normal (Goldberg and Downing, 1955). The pulmonary peripheral resistance is low initially in most patients and the mean pulmonary artery pressure normal or only slightly raised (Wood, 1950). There is apparently no consistent relationship between flow and pressure in the pulmonary arterial system, but a high flow may

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FIG. 1.—Seven types of atrial septal defect—diagrammatic (see text).

ultimately lead to secondary pulmonary hypertension and consequent reduction in flow (Chapman and Frazer, 1953). This abnormal cardio-pulmonary circulation causes enlargement of the right atrium and of the right ventricle; the pulmonary artery becomes large and pulsatile, and dilatation of the vascular system of the lungs occurs. Atheroma of the pulmonary vessels may develop. This sequence of events may be modified by associated congenital cardiac abnormalities.

Clinical Picture

In infancy a large shunt may lead to early cardiac failure and death in the first few months of life, but more commonly failure to gain weight satisfactorily is the only symptom. Children with this lesion may become easily fatigued and suffer from frequent attacks of "congestion of the lungs," but dyspnoea on exertion is not usually severe enough to prevent a reasonably active life, and cyanosis is absent or slight. Many individuals, however, remain symptomless until the third or fourth decade. Once symptoms have become established at any age, congestive cardiac failure is apt to develop, either gradually or suddenly. It may be precipitated by an intercurrent infection and be accompanied by auricular fibrillation.

Examination may show the patient to be thin, with light bones, slender limbs, poor muscular development, and a long narrow chest with a prominent sternum, but some of our patients have been well built and of normal development. Arachnodactyly and a high arched palate are occasional associated abnormalities. The pulse is of small or normal volume; the blood pressure is normal. The apex beat may be displaced outwards and be of the right ventricular tapping type, or diffuse and tumultuous. Heaving of the right ventricular outflow tract is often very marked, and pulsation of the pulmonary artery may be visible in the second left intercostal space, over which a systolic thrill may be palpable. The second sound in the pulmonary area is usually split, and the second pulmonary element may be loud.

A systolic murmur, which may be pronounced, is usually maximal in the pulmonary area, and less well conducted than the murmurs of pulmonary stenosis, ventricular septal defect, or persistent ductus arteriosus. A short blowing diastolic murmur of pulmonary incompetence may be audible in the pulmonary area. A low-pitched diastolic murmur is also often heard at the apex beat in uncomplicated atrial septal defects.

The electrocardiogram shows the rR' pattern of incomplete right bundle-branch block in the V_1 or V_3R leads so frequently that the absence of this type of record should cast doubt upon the diagnosis. Right axis deviation is usually found.

The radiological appearances are those of right atrial and right ventricular enlargement, with large pulsatile pulmonary arteries, increased peripheral pulmonary vascularity, a well-marked "hilar dance," and a small aorta. In a minority of cases, however, the heart is normal in size and shape.

Cardiac catheterization may suggest the diagnosis by the demonstration of oxygenated blood in the right atrium, but anomalous entry of pulmonary veins into the right atrium, either directly or via the coronary sinus, will give rise to the same finding. The passage of the catheter from the right atrium into the left atrium also suggests the presence of an atrial septal defect but does not prove it, as a catheter will sometimes pass quite freely through a patent foramen ovale. The right ventricular and pulmonary artery pressures are frequently normal but occasionally raised. Under con-

ditions of failure increasing right atrial pressure may at first prevent the left-to-right shunt and later reverse it. That this has occurred may be suggested clinically by the appearance of cyanosis and confirmed by finding a similar oxygen saturation of the blood in the venae cavae and right atrium, and a reduced oxygen saturation in the blood of the femoral artery. It is in this type of case that angiography may be helpful.

Intravenous angiocardiology may reveal a left-to-right shunt by abnormally rapid dilution of the dye in the right atrium. This is best demonstrated in the right posterior oblique position with sufficiently rapid exposures (Lind *et al.*, 1954). In the presence of a reversed shunt or of a very large defect the contrast medium may be seen to pass from the right atrium to the left. The demonstration of a right atrium of normal size should raise doubts on the correctness of the diagnosis.

Prognosis

In 73 cases of atrial septal defect the mean age at death was 28 years, with extremes of 3 months and 70 years (Abbott, 1936). In 62 cases it was 36 years (Roesler, 1934). In 19 cases 49 years (Cosby and Griffith, 1949). In a series of 10 cases the majority of patients died between 30 and 45 years (Bedford *et al.*, 1941). Of 11 children with this defect, however, six died in infancy—two of the older children following operations for surgical correction of the defect, and two others as a result of unassociated conditions (Disenhouse *et al.*, 1954).

The average expectation of life is therefore considerably reduced. Death may occur during the third or fourth decade in patients free of symptoms during childhood, but it may be earlier in children who suffer from recurrent respiratory infections. Once congestive heart failure has developed the patient may survive for only a few months, or, with the assistance of digitalis, perhaps for a few years.

In all cases, therefore, surgical closure of the defect should be considered.

Surgical Treatment

Numerous suggestions have been put forward for accomplishing the closure of an atrial septal defect based on experimental work, and in some cases the method has received clinical trial (Fig. 2).

1. Murray (1948).—Sutures are inserted through the heart in the line from the interatrial septum anteriorly to the interatrial septum posteriorly. There is considerable distortion of the two atria after the sutures are tied, and the veins on the other side may be obstructed. There are difficulties also with the closure of the supraventricular portion of an ostium primum. This method would appear, however, to have some application in the treatment of a common atrium.

2. Cohn (1947).—A portion of the atrial wall is sutured to the margins of the septal defect and is then cut off, the atrial wall being closed by a purse-string suture. Experimental only.

3. Swan *et al.* (1950).—The left and right atrial appendages are plugged through the atrial septal defect and sutured together. Experimental only.

4. Martin and Essex (1948).—A "polythene" sheet is covered with a vein graft and sutured in the septal plane. Experimental only.

5. Dodrill (1949).—A ring clamp is applied over both atria. The right atrial wall is opened over the defect, which is then sutured under direct vision. Suitable only for small defects. Experimental only.

6. Merendino *et al.* (quoted by Bailey *et al.*, 1953).—A plug of fat contained in a bag of pericardium is inserted into the defect. Experimental only.

7. Sndergaard *et al.* (1954).—A groove is dissected between the two atria posteriorly. One suture is anchored to the interatrial septum superiorly close to the aorta and one suture is anchored

to the right ventricle just below the atrio-ventricular groove. The two sutures are tied in the groove over a piece of gelatin foam, and this closes or reduces in size the septal defect. The permanency of this procedure is doubtful as there is only an approximation of the edges of the defect.

8. Bailey *et al.* (1952).—The finger is inserted into the right atrium through the appendage, and after digital exploration the atrial wall is sutured to the margins of the defect around the finger. Exploration of the mitral valve can also be undertaken and the valve split in cases of Lutembacher's syndrome. A large number of patients have now been treated with this operation by Bailey and others.

9. Swan *et al.* (1950).—Sutures are passed through the atrial defect from the right to the left atrial appendage and a plastic button is drawn into the right atrium over the defect. A similar button is threaded on to the sutures passing through the left atrial appendage and is then sutured down on to the left side of the defect with the wall of the appendage intervening. Experimental only.

10. Gross *et al.* (1952).—A "well" made of rubber is sutured to the side of the atrium, the atrium opened, and the defect sutured under tactile control. A sheet of plastic material can be sutured over the defect if it is large. A number of patients have been successfully treated by this method, but it is technically difficult compared with some others.

11. Husfeldt and Søndergaard (1953).—Søndergaard's method of periseptal sutures is employed with the finger in the atrium to give tactile control.

12. Björk and Crafoord (1953).—A circular suture is passed through the septum around the defect, the needle being directed by a finger in the atrium. Posteriorly the suture passes through a groove as dissected by Søndergaard.

13. Blount *et al.* (1954); Brock and Ross (1955); and others.—With the aid of hypothermia, cardio-pulmonary by-passes, and cross-circulations a number of open operations have been performed in Great Britain and America whereby the atrial septal defect has been sutured under direct vision.

The direct suture of an atrial septal defect with an open heart conforms best to surgical principles, but an open-heart operation is at the moment associated with technical difficulties which make the risks not inconsiderable. The opening of the heart with an interrupted circulation under hypothermia, at any rate in children, has so far been the most successful method, and Blount *et al.* (1954) have had considerable experience of this. The recovery of the heart from trauma at these temperature levels is not as satisfactory as when the operation is performed at normal temperatures.

FIG. 2.—Suggested methods of closure of atrial septal defects (see text).

A tactile non-visual method can be satisfactory, but some preliminary experience is necessary before digital examination can be accurately interpreted. Nevertheless the advantage of the method is that no interruption of the circulation is necessary and no adjuncts are required to the normal form of anaesthesia.

Consideration of the various methods that have been suggested for closing an atrial septal defect led us to the view that the simplest method (for all except the almost completely absent septum) was the external suture suggested by Sndergaard and practised by him and Husfeldt. Some doubt was felt about the permanency of this form of closure, as there is no direct suturing of the defect itself. Furthermore, the possibility of one of the sutures cutting free from its anchorage was envisaged. It seemed that the use of this external suture to approximate the edges of the defect might well simplify the insertion of sutures to close the defect directly.

Technique

The following technique was accordingly worked out and has been applied in 12 cases.

A right thoracotomy is made through the fourth interspace, using a lateral incision extending anteriorly. This incision is long owing to the antero-posterior elongation of the chest, which is so commonly a feature of these patients.

The pericardium is opened throughout its length in front of the phrenic nerve. A Crafoord's clamp is placed on the right atrial appendage and a purse-string suture of linen thread is inserted distal to it. A piece of rubber tubing is slipped over the ends down to the atrial wall. An incision is made in the end of the appendage, and the left forefinger passed through it into the atrium, loosening the clamp at the same time. The purse-string is held taut around the finger and the piece of rubber tubing is clamped around the long ends of thread with a light pair of forceps so that a haemostatic seal is obtained.

The septal defect is examined with the finger, and its size and its relation to the tricuspid and mitral valves and the pulmonary veins are assessed. The anterior edge of the defect is usually thickened and easily felt, but the posterior edge is often little more than a ridge. The size of the defect may appear larger than it actually is owing to a portion of the septum being of diaphanous consistency and practically impalpable. If possible any stenosis of the mitral valve should be estimated and an attempt made to relieve it. Rarely the right pulmonary veins may enter the right atrium, and if this condition is found any closure of the septal defect must be so designed that these veins enter the left atrium.

The finger can now be removed from the atrium and the clamp on the appendage reapplied.

If the defect is large an external Sndergaard suture is applied. Dissection of the groove between the two atria is usually not necessary, as a good temporary approximation of the edges of the defect can be attained without this added procedure. A suture of linen thread is anchored to the right ventricle below the atrio-ventricular groove. Both sutures are then brought together behind the superior and inferior cava respectively. The finger is reinserted into the atrium and the two external sutures are knotted by an assistant, the intra-atrial finger noting the size of the septal defect, and as soon as the walls are in apposition the sutures

are tied. If the opening is very wide it may not be possible to approximate the walls of the defect with the encircling sutures without danger of cutting into the atria, and under these circumstances the sutures are tied as tight as is thought safe. The reduction in size of the defect will be very considerable and the second stage of the operation will be rendered easier. In small defects—that is, 3 cm. or less—the external suture is unnecessary.

With the forefinger of the left hand in the atrium the portion of the right atrium lying between the superior and the inferior vena cava can be approximated to the septal defect with the left thumb. The position in which to insert the sutures which will close and fix the outer wall of the atrium over the defect and at the same time firmly approximate the edges will then become obvious. No. 30 linen thread mounted upon half-circle Mayo needles is used double, and inserted first through the outer wall of the atrium and then the lower part of the septum. The point of the needle is now in the left atrium and is turned so that it picks up a good piece of the upper part of the septum and comes out again through the outer wall of the right atrium close to the original point of entry (Fig. 3). It is picked up and the thread drawn through and tied by the assistant, the surgeon controlling the tension with the intra-atrial finger. Further sutures are inserted at intervals of 5 mm. in the same position and gradually the defect is closed. The lower sutures will be medial to the inferior vena cava. The portion of right atrial space lost is quite small, but it is important to see that the upper and lowermost sutures do not encroach upon the venae cavae, and this point can be checked by inserting the intra-atrial finger into their orifices or noting any increased tension in palpating the vessel. It is important, when suturing the lowermost portion of the defect, that the electrocardiographic record is studied continuously for evidence of any abnormality of heart action due to the malplacement of a suture affecting the bundle of His or the coronary sinus. No suture is tied tightly, as firm approximation of the structures is adequate and there is less likelihood of subsequent cutting out of the stitches.

With a persistent ostium primum it may not be possible to close completely the last few millimetres above the mitral and tricuspid valves, but the size of the foramen left is so small that it is unlikely to have any significant effect upon subsequent cardiac function.

The amount of blood lost in the suturing procedure is surprisingly small and the heart tolerates the finger in the atrium for at least one hour quite comfortably provided the myocardium is reasonably healthy. In hearts that have been in failure or otherwise demonstrated signs of severe strain, however, the toleration of intracardiac manipulation is reduced and there is a greater tendency to extrasystoles.

Finally, the finger is removed from the right atrium, the Crafoord clamp reapplied, and the appendage sutured. The external suture is left in place, for it is believed that it may relieve tension on the suture line in the septum during the post-operative phase and lead to sounder healing.

The beneficial effect of closure of the atrial septal defect is immediately obvious in the less vigorous action of the heart and the reduced distension of the right atrium.

The pericardium is loosely sutured and the chest closed with drainage.

FIG. 3.—Method of closure of atrial septal defect employed in the present series.

Anaesthesia

Anaesthesia for the operation of closure of an atrial septal defect has presented no special difficulties. The technique has been based on an induction of anaesthesia with thiopentone, D-tubocurarine chloride, and pethidine, and maintenance with nitrous oxide.

As in all cases exhibiting ventricular strain, the desiderata are the early establishment of an infallible means of pulmonary ventilation with the use of minimal quantities of drugs that produce either a lowering of peripheral resistance or a reduction in the capacity of the myocardium to do work. Despite the effect of thiopentone on the myocardium we have considered that the rapid and smooth induction of anaesthesia by this drug justifies its use in children undergoing this operation. It is also considered imperative that when intermittent positive-pressure breathing is established the positive pressure used should be minimal. There is a high pulmonary blood flow, and in some cases a high pulmonary vascular resistance, which will be further increased if high inflation pressures are used. In order that adequate ventilation may be obtained with the use of low intermittent positive pressure it is essential that good muscular relaxation be obtained.

The patients are premedicated with morphine, 1/40 gr. per stone (0.25 mg. per kg.) of body weight, and with atropine.

An intravenous infusion is set up pre-operatively, and on arrival in the theatre narcosis is induced with thiopentone. The dose of thiopentone is minimal and even in the younger children does not exceed 35 mg. per stone (5.5 mg. per kg.). This is followed by D-tubocurarine, 3 mg. per stone (0.47 mg. per kg.), and pethidine, 2 mg. per stone (0.31 mg. per kg.). Prior to intubation the patients were ventilated with a 75% and 25% mixture of nitrous oxide and oxygen for five minutes to ensure full saturation with nitrous oxide before intubation. Thereafter anaesthesia is maintained by intermittent positive-pressure breathing with 50% nitrous oxide and oxygen, and supplementary doses of D-tubocurarine as indicated by increased resistance to inflation or a return of spontaneous respiration.

In no case has procaine or procainamide been used, as it is considered that both these drugs are contraindicated in the presence of right ventricular strain. Despite this, the only cardiac arrhythmias which have been observed, other than those associated with the insertion of sutures into the region of the conducting bundle, have been occasional extrasystoles and, in several cases, nodal rhythm, which was associated with pericardial traction and which reverted to normal after the injection of intravenous atropine.

At the end of the operation spontaneous respiration is permitted to return, and the patient given intravenous atropine, 1/500 gr. per stone (0.02 mg. per kg.), followed by neostigmine, 0.5 mg. per stone (0.075 mg. per kg.). All recovered consciousness in the operating theatre.

Occasionally, when a finger was in the right atrium an increase in venous pressure was indicated by a slowing of the intravenous infusion, a fall in blood pressure, and reduction in the voltage of the E.C.G. These effects are presumed to result from the reduced cardiac output due to deficient filling of the atrium whilst the finger is *in situ*.

With the exception of Cases 2 and 5 there were no major post-operative pulmonary complications.

Case Reports

Twelve patients, ten children and two adults, have been submitted to this surgical procedure. In only nine of these (Cases 1, 2, 4, 6, 7, 8, 9, 10, and 12) was an atrial septal defect diagnosed on clinical grounds, including radiological and electrocardiographic findings, alone. In the other three the provisional diagnoses were a septal defect, with pulmonary stenosis (Case 5); Eisenmenger's complex (Case 3); and pulmonary hypertension of doubtful origin (Case 11). The presence of an atrial septal defect, however, was confirmed before operation in eleven of the cases by cardiac catheterization, and in the twelfth by angiocardiology. Catheterization also revealed pulmonary hypertension with systolic pressure over 40 mm. Hg in Cases 5, 9, and 11, and suggested it in a fourth in which the pulmonary artery was not catheterized but the right ventricular systolic pressure was 53 mm. Hg. In Cases 2, 3, 6, 8, and 12 the systolic pressure in the pulmonary artery was less than 25 mm. Hg, in Case 1 the pressure measurements were inaccurate, and in Case 10 a sudden drop in systolic pressure at the pulmonary valve of 29 mm. Hg suggested pulmonary stenosis.

Case 1.—Boy aged 12. Frequent bronchitis since infancy. Easily fatigued from the age of 5. Dyspnoea on exertion from the age of 8. X-ray: Cardiac enlargement and pulmonary plethora marked (Fig. 4 (A)). E.C.G.: Lead V₁, rsR', V.A.T. 0.05 sec., QRS 0.08 sec. P₂ 4 mm., peaked (Fig. 5 (A)). Provisional diagnosis: Atrial septal defect and ventricular septal defect. Cardiac catheterization: Pulmonary artery pressure not accurately recorded; superior vena cava—R.A. O₂ increase 35%. Operation: Ostium primum, 6 by 2 cm. (type 2 defect), almost completely closed with five sutures. Result: Systolic thrill ceased—immediate. Loud systolic and apical diastolic murmurs became soft—immediate. X-ray: R.A. contour less prominent—first day. General cardiac enlargement less and pulmonary plethora diminished—tenth week (Fig. 4 (B)). E.C.G.: P₂ smaller

FIG. 4.—Radiographs, Case 1. A, Pre-operative. B, 10 weeks after operation, showing decrease in size of heart and in pulmonary vascularity.

FIG. 5.—Case 1. Electrocardiograms showing smaller bifid P waves in Lead II and changes in T waves following operation. A, Before operation. B, After operation.

and bifid, R.V. dominance less—tenth week (Fig. 5 (B)). T₂ inverted for 14 weeks, upright at eight months. Dyspnoea on exertion less—tenth week. Dyspnoea on exertion still diminishing—eighth month. Had first winter since birth without bronchitis. Follow-up eight months.

Case 2.—Girl aged 16. Mentally retarded. Dyspnoea probable from age of 2 years. Could run 25 yds. at 15. Lymphangiectasia of left leg. X-ray: Cardiac enlargement moderate; pulmonary plethora moderate. E.C.G.: Lead V₁R, rsR', V.A.T. 0.04 sec., QRS 0.09 sec. P₂ 3 mm., peaked. Provisional diagnosis: Atrial septal defect. Cardiac catheterization: Pulmonary artery pressure 18/11 mm. Hg; superior vena cava—R.A. O₂ increase 17%. Operation: Ostium primum, 2 by 1.2 cm.; ostium secundum, 1.5 by 1 cm. (type 3 defect). Ostium secundum completely closed and ostium primum partly closed with two sutures. Result: Moderately loud systolic murmur became very soft on first day (Fig. 6). Right chylothorax from seventh day, persisting at eighth month. Chylothorax associated with mediastinal lymphangiectasia. Follow-up, eight months.

FIG. 6.—Case 2. Phonocardiograms showing cessation of systolic murmur (SM) in pulmonary area (PA) following operation. A, Before operation. B, After operation.

Case 3.—Girl aged 10½. No symptoms. X-ray: Cardiac enlargement moderate; pulmonary plethora moderate. E.C.G.: Lead V₁R, rsR'S', V.A.T. 0.05 sec., QRS 0.07 sec. P₂ 3 mm., peaked. Provisional diagnosis: Eisenmenger's complex. Cardiac catheterization: Pulmonary artery pressure 15/10 mm. Hg; superior vena cava—R.A. O₂ increase 19%. Operation: Atrial septum very thin; ostium secundum, 2 by 1.5 cm. (type 5 defect), completely closed with two sutures. Result: Loud systolic murmur became softer—tenth day. X-ray: Pulmonary arc and pulmonary plethora slightly less—sixth week. Exercise tolerance unchanged. Follow-up, 15 weeks.

Case 4.—Girl aged 13. Dyspnoea and slight cyanosis of lips and extremities on extreme exertion, and easily tired, from 10 years of age. X-ray: Cardiac enlargement moderate; pulmonary plethora moderate. E.C.G.: Lead V₁, rsR', V.A.T. 0.07 sec., QRS 0.08 sec. P₂ 3 mm., peaked. Provisional diagnosis: Atrial septal defect. Cardiac catheterization: Pulmonary artery not catheterized; R.V. pressure 55/0 mm. Hg; superior vena cava—R.A. O₂ increase 15%. Operation: Ostium secundum, 4 by 3 cm. (type 5 defect), almost completely closed with three sutures. Lowest suture caused transient a-v dissociation. Result: Convalescence uneventful for six days. Loud systolic murmur and thrill less marked—sixth day. Hepatic enlargement—sixth day. Collapsing pulse. B.P. 170/50—sixteenth day. Death—seventh week. Necropsy: Aortico-atrial fistula caused by suture.

Case 5.—Girl aged 7½. Dyspnoea on exertion from 2½ years. At 7 years could run 40 yds. Recurrent bronchitis with pneumonia once a year since infancy. Bronchiectasis of left lower lobe. X-ray: Cardiac enlargement marked; pulmonary plethora moderate. E.C.G.: Lead V₁, rR's, V.A.T. 0.05 sec., QRS 0.09 sec. P₂ 3 mm., peaked. Cardiac catheterization: Pulmonary artery pressure 42/14 mm. Hg; superior vena cava—R.A. O₂ increase 19%. Provisional diagnosis: Septal defect and pulmonary stenosis. Operation: Ostium secundum, 4 by 2 cm. (type 5 defect); right pulmonary veins entering right atrium; defect almost completely closed and pulmonary veins incorporated into left atrium by three sutures between anterior margin of defect and atrial wall anterior to pulmonary veins. Result: Transient pneumonitis. E.C.G.: P₂ smaller—fourth day. X-ray: R.A. contour less prominent and pulmonary plethora less—fourth week. Dyspnoea on exertion less; less easily tired—fourteenth week. Follow-up, 21 weeks.

Case 6.—Boy aged 7½. Dyspnoea on exertion from 6½ years. Congestive cardiac failure at 7 years which responded to digitalization. X-ray: Cardiac enlargement marked; pulmonary plethora moderate. E.C.G.: Lead V₁, rR's, V.A.T. 0.07 sec., QRS 0.10 sec. P₂ 3.5 mm., peaked. Provisional diagnosis: Atrial septal defect. Cardiac catheterization: Pulmonary artery pressure 22/12 mm. Hg; superior vena cava—R.A. O₂ increase 15%. Operation: Ostium primum, 4 by 2 cm. (type 2 defect), almost completely closed with four sutures. Result: Loud systolic murmur became very soft—immediate. Apex beat became less forceful during first three weeks. X-ray: R.A. contour less prominent—immediate. E.C.G.: P₂ smaller—sixth day. s-t V₁ depression marked—sixth day. s-t V₅ depression almost normal—fourteenth week. Digoxin stopped on sixteenth day. Dyspnoea on exertion much less—twelfth week; normal at five months, though still easily tired. Follow-up, five months.

Case 7.—Girl aged 16½. Dyspnoea on exertion since age of 13. Congestive cardiac failure relieved by digoxin at 13. Digitalization maintained until 16½ years, when unable to run or walk uphill without extreme dyspnoea. X-ray: Cardiac enlargement marked; pulmonary plethora moderate. E.C.G.: Lead V₁, qR, R 22 mm., QRS 0.08 sec. P₂ 3.5 mm., peaked. Provisional diagnosis: Lutembacher's syndrome. Angiocardiography: right-to-left shunt through atrial septal defect. Operation: Defect 8 by 5 cm., replacing almost whole septum (type 1 defect). Closure impossible. Mitral regurgitation but no stenosis. Result: Condition unchanged at sixth week.

Case 8.—Woman aged 22. Exercise tolerance normal. X-ray: Cardiac enlargement slight. Hilar vessels very prominent. Peripheral pulmonary plethora moderate. E.C.G.: Lead III, RR', V.A.T. 0.05 sec., QRS 0.10 sec. P₂ 2.5 mm. Provisional diagnosis: Atrial septal defect. Cardiac catheterization: Pulmonary artery pressure, mean 10 mm. Hg; R.A. high—R.A. mid O₂ increase 14%. Operation: Ostium primum, 5 by 3 cm. (type 2 defect), almost completely closed with four sutures. Result: Loud systolic murmur became soft—immediate. X-ray: R.A. contour less prominent. Pulmonary plethora slightly diminished—sixth week. Pulmonary plethora further diminished—fifteenth week. E.C.G.: P₂ smaller (1.5 mm.)—fourteenth day. Exercise tolerance unchanged. Follow-up, 15 weeks.

Case 9.—Woman aged 35. Increasing dyspnoea on exertion, effort angina, occasional haemoptysis, and oedema of feet from age 31. X-ray: Cardiac enlargement and pulmonary plethora marked. E.C.G.: Lead III, rR'S', V.A.T. 0.05 sec., QRS 0.12 sec. P₂ 3.5 mm., peaked. Provisional diagnosis: Atrial septal defect. Cardiac catheterization: Pulmonary artery pressure 63/30 mm. Hg; superior vena cava—R.A. O₂ increase 14%. Operation: Ostium secundum, 6 by 4 cm. (type 5 defect), partly closed with one suture. Result: Condition satisfactory until sudden death 18 hours after operation. Necropsy: Ostium secundum incompletely closed by suture; marked sclerosis of pulmonary vessels; thrombosis of right middle cerebral artery; pulmonary oedema and collapse.

Case 10.—Boy aged 15. Cyanosis on exertion and easily tired since birth. Dyspnoea on exertion since 18 months. At 15 years could run 50 yds. and walk any distance. X-ray: Cardiac enlargement slight; pulmonary plethora moderate. E.C.G.: Lead V₁R, rsR', V.A.T. 0.06 sec., QRS 0.10 sec. P₂ 2.5 mm. Provisional diagnosis: Atrial septal defect and pulmonary stenosis. Cardiac catheterization: Pulmonary artery pressure 11/8 mm. Hg; R.V. pressure 44/0 mm. Hg; superior vena cava—R.A. O₂ increase 15%. Operation: Ostium secundum, 5 by 2 cm. (type 5 defect). Right superior pulmonary vein entering superior vena cava. Thrill in pulmonary artery suggesting organic pulmonary stenosis, but no post-stenotic dilatation of artery. Defect almost completely closed with four sutures. Result: Systolic thrill and loud murmur diminished—second day. Dyspnoea less. Runs 150 yds.—fourth week. X-ray: Cardiac enlargement slightly increased, pulmonary plethora unchanged—fifth and ninth weeks. Follow-up, nine weeks.

Case 11.—Boy aged 8½. Frequent bronchitis and easily tired since whooping-cough at 3 years of age. Dyspnoea on exertion from 5 years. At 8½ could run 30 yds. or walk slowly for half a mile. X-ray: Cardiac enlargement slight; pulmonary plethora moderate. E.C.G.: Lead V₁R, rsR', R' 22 mm., V.A.T. 0.05 sec., QRS 0.08 sec. P₂ 2 mm. Provisional diagnosis: Pulmonary hypertension. Cardiac catheterization: Pulmonary artery pressure 80/40 mm. Hg; superior vena cava—R.A. O₂ increase 9%. Operation: Ostium secundum, 2.5 by 2 cm. (type 5 defect), completely closed with two sutures. Results: Moderately loud systolic murmur became very soft—first day. X-ray: R.A. contour less prominent—first day. Pulmonary plethora slightly diminished—thirteenth day. Cardiac enlargement slightly increased—seventh week. E.C.G.: T_v inversion increased with

s-t depression increased. Tv, formerly upright, inverted—seventh week. Exercise tolerance unchanged. Palpitation increased. Follow-up, seven weeks.

Case 12.—Boy aged 13 years 11 months. Easily tired and slight occasional peripheral cyanosis since pneumonia at 2 years of age. At 13 years and 11 months, dyspnoea on prolonged exertion only. Frequent epistaxis since birth. X-ray: Cardiac enlargement slight; pulmonary plethora moderate. E.C.G.: Lead V₁, rsr'r³, V.A.T. 0.07 sec., QRS 0.08 sec. P₂ 1.5 mm. Cardiac catheterization: Pulmonary artery pressure 23/7 mm. Hg (recording doubtful); superior vena cava—R.A. O₂ increase 18%. Operation: Ostium secundum, 5 by 2.5 cm. (type 5 defect), closed with four sutures. Result: Moderately loud systolic murmur became very soft—third day. Apical mid-diastolic murmur ceased—seventeenth day. X-ray: Pulmonary vascularity diminished—sixteenth day. Dyspnoea and tiredness less—fourth week. Follow-up, six weeks.

Discussion

Twelve patients have been operated upon. In one case, with a common atrium (Case 7), no closure was attempted. In the remaining 11 the procedure described above was undertaken. Surgical closure of the defect as judged by the exploring finger at the end of the operation was thought to be complete or almost complete in 10 of these. Complete closure of those defects which approach the atrio-ventricular valves was limited by interference with the conduction mechanism as indicated by the electrocardiographic control. Two of the 11 died in the post-operative period.

Of the two fatal cases, one (Case 9) was that of a woman aged 35 who had had recurrent attacks of failure over a period of four years and had developed cyanosis, suggesting some reversal of the shunt. The right atrium was very large, the defect measured 6 by 4 cm., and there was marked sclerosis of the pulmonary vessels. The suturing appeared only partially effective and recovery was satisfactory for 18 hours, when sudden death occurred. At necropsy, occlusion of the right middle cerebral artery was found, but the cause of death was thought to be of cardiac origin. This was an unfavourable case for operation, and supports the view that prolonged procedures are not well tolerated on hearts that have been in failure. Two of the other cases in this series had been in failure, but for shorter periods, and their comparative youth probably enabled them to tolerate the operation.

The second fatality was due to a technical error during the operation. The defect was a large one, the anterior portion of the septum was narrow, and one of the stitches was inserted through an aortic sinus. Six days later the patient's liver enlarged and shortly after that her pulse became collapsing. A superficial to-and-fro murmur became audible over the base of the heart, and, although the possibility of an aortico-atrial fistula was considered, it was thought improbable, as the murmur was atypical. Death took place five weeks later following an exploratory operation, and necropsy revealed the presence of such a fistula and that the greatly raised atrial pressure had torn out the remaining sutures.

One patient developed a chylothorax which has persisted for nine months after operation. She had a congenital lymphangiectasia of the left leg, and it is thought that there may have been abnormal lymphatic channels in the mediastinum, some of which may have been damaged at the time of the operation. No exposure or operative procedure was undertaken anywhere near the normal position of the thoracic ducts.

The remaining nine have been observed for periods varying from six weeks to eleven months. Exercise tolerance has improved in four of the seven patients in whom it was previously reduced; in two of these (Cases 11 and 12) the period of observation is too short for this factor to be assessed. The systolic murmur in the pulmonary area has ceased or become softer in eight of the nine cases, suggesting a reduced blood flow through the pulmonary artery. Cardiac enlargement on x-ray examination has become less in four cases, but has increased slightly in the case in which

there was associated pulmonary stenosis; the pulmonary arc has become smaller in three cases and pulmonary vascularity has diminished in seven. The P wave in lead II of the electrocardiogram has decreased in size in four cases.

In these nine patients, therefore, the procedure appears to have achieved immediate satisfactory closure of the defect. Whether such closure is permanent will of course be learned only after a longer period of observation and by repeating cardiac catheterization after a sufficient interval.

If it is accepted that a cardiovascular abnormality is likely to shorten life and a reasonably safe operation will correct the defect, then that operation should be undertaken as early as possible even in the asymptomatic stage. This principle is now generally accepted in the treatment of patent ductus arteriosus, coarctation of the aorta, and pulmonary stenosis. We believe it should apply with even greater force to cases of atrial septal defect where, in addition to increasing strain on the cardiac musculature, the progressive enlargement of the right atrium appears to result in an increase in the size of the defect and the difficulties of closure.

Summary

The embryology and clinical features of atrial septal defects are reviewed.

Twelve cases have been explored and in 11 closure was attempted. Two patients died. In nine cases the defect was sutured.

Closure was undertaken by a direct suture of the defect, including the portion of the atrial wall between the two venae cavae. Tactile control of the suturing is maintained by a finger in the right atrium.

Results suggest that in an ostium secundum complete closure may be expected. Closure by suture of an ostium primum is apt to produce damage to the conduction bundle, but a partial closure can bring great clinical improvement.

It is advisable to undertake surgical treatment before severe symptoms and failure have supervened.

Standard forms of anaesthesia only are required.

A description is given of the cases so far treated, with follow-up.

ADDENDUM.—Since this article was written four further cases have been operated upon by this method, with success. One patient had a coincidental open pulmonary valvotomy for pulmonary stenosis.

REFERENCES

- Abbott, M. E. (1936). *Atlas of Congenital Heart Disease*. New York.
Bailey, C. P., Bolton, H. E., Jamison, W. L., and Neptune, W. B. (1953). *J. thorac. Surg.*, **26**, 184.
— et al. (1952). *Ann. intern. Med.*, **37**, 888.
Bedford, D. E., Papp, C., and Parkinson, J. (1941). *Brit. Heart J.*, **3**, 37.
Björk, V. O., and Crafoord, C. (1953). *J. thorac. Surg.*, **26**, 300.
Blount, S. G., Swan, H., Gensini, G. I., and McCord, M. C. (1954). *Circulation*, **9**, 801.
Brock, Sir Russell, and Ross, D. N. (1955). *Guy's Hosp. Rep.*, **104**, 99.
Chapman, C. B., and Frazer, R. (1953). *Amer. Heart J.*, **46**, 352.
Cohn, R. (1947). *Ibid.*, **33**, 453.
Cosby, R. S., and Griffith, G. C. (1949). *Ibid.*, **38**, 80.
Disenhouse, R. B., Anderson, R. C., Adams, P., Novick, R., Jorgens, J., and Lavin, B. (1954). *J. Pediatr.*, **44**, 269.
Dodrill, F. D. (1949). *J. thorac. Surg.*, **18**, 652.
Goldberg, H., and Downing, D. F. (1955). *Amer. Heart J.*, **49**, 862.
Gross, R. E., Pomeranz, A. A., Watkins, E., and Goldsmith, E. I. (1952). *New Engl. J. Med.*, **247**, 455.
Husfeldt, E., and Søndergaard, T. (1953). Personal communication.
Keith, J. D., and Forsyth, C. G. (1951). *J. Pediatr.*, **38**, 172.
Keriluk, L. B., Hoag, E. W., and Merendino, K. A. (Quoted by Bailey et al., 1953.)
Lind, J., Spencer, R., and Wegelius, C. (1954). *Brit. Heart J.*, **16**, 407.
McGinn, S., and White, P. D. (1933). *Amer. Heart J.*, **9**, 1.
— (1936). *New Engl. J. Med.*, **214**, 763.
Martin, W. B., and Essex, H. E. (1948). *Amer. J. Physiol.*, **155**, 453.
Murray, G. (1948). *Ann. Surg.*, **128**, 843.
Roessler, H. (1934). *Arch. intern. Med.*, **54**, 339.
Søndergaard, T., Sørensen, H. R., Poulsen, T., and Andersen, L. (1954). *Acta chir. scand.*, **107**, 485.
Swan, H., Maresh, G., Johnson, M. E., and Warner, G. (1950). *J. thorac. Surg.*, **20**, 542.
Wood, P. (1950). *British Medical Journal*, **2**, 693.