

## ATRIAL SEPTAL DEFECT—RESULTS OF SURGICAL CORRECTION IN ONE HUNDRED PATIENTS

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The indications for operation in a given condition depend inevitably on the risk of the procedure as weighed against the potential gain from its success. When the procedure is known to be curative and the risk steadily diminishes as experience is gained, a point is eventually reached when the diagnosis of the lesion becomes a firm indication for its correction. That point was arrived at several years ago in the case of the atrial septal defect. We here report the results of operation, based on this precept, in 100 consecutive cases.

The patients ranged in age, at the time of surgery, from 10 months to 45 years. There were 68 females and 32 males in the series. Some were without symptoms, while others were severely disabled. Likewise, the physical and other findings varied from normal to gross derangement.

### Preoperative Evaluation

The assessment of the patient's condition was based on history, physical examination, electrocardiography, fluoroscopy, and, in many cases, cardiac catheterization. It is believed that the diagnosis can be achieved accurately on clinical grounds in over 90% of patients in the age group covered by this report, except, of course, the recognition of anomalous pulmonary venous connections.

It is also considered that the common atrioventricular lesion can be distinguished clinically; none of the patients operated on with the diagnosis of atrial septal defect secundum was found to have the former lesion.<sup>1</sup>

**History.**—The variability of symptoms in atrial septal defect has been frequently emphasized. In this series, incapacity in some patients was severe and in others it was absent. Though there were obvious exceptions, by and large the symptoms appeared in the third decade. Before that they were either absent or slight in most instances. The most prominent complaints were shortness of breath, fatigue, and palpitations ("pounding in the chest"), though there were a number of other symptoms expressive of ill health in general rather than heart disease in particular. Of the 100 patients, 38 were asymptomatic, 29 mildly incapacitated, 23 moderately incapacitated, and 10 severely so. Only eight gave a history indicative of congestive cardiac fail-

*One hundred consecutive patients, ranging in age from 10 months to 45 years, with a diagnosis of atrial septal defect secundum, were operated on. Of the 100 patients, 38 were asymptomatic, 29 mildly incapacitated, 23 moderately incapacitated, and 10 severely so. Only eight gave a history indicative of congestive cardiac failure. The most prominent complaints were shortness of breath, fatigue, and palpitations (pounding in the chest). Operation was performed under conditions of hypothermia and inflow-outflow occlusion. After the second year of life, the earlier the operation is performed the better. Surgery in a relatively asymptomatic child between the ages of 2 and 10 years carries a minimum of risk and in all probability will insure a normal cardiovascular system when maturity is reached. A high pulmonary vascular resistance increases the risk and decreases the gain that can be anticipated from operation.*

ure. We did not find a marked increase in the incidence of respiratory infection in these patients, nor did we note a predilection for rheumatic fever.

**Physical Signs.**—The classic slight build (gracile habitus) was noted in most of the children in this series, and sometimes this was particularly marked. However, it was by no means an invariable finding; a few of the older patients were, in fact, stocky rather than slender.

The normal or small arterial pulse was in most cases in sharp contrast to the marked hyperactivity of the precordium. However, as with so many other findings, this activity tends to be much more pronounced and obvious in the young patient when the chest wall is very thin and less marked in the adult when it is thick. The venous pulse was not considered uniformly remarkable, though when tricuspid insufficiency was present the characteristic systolic wave was seen. A left precordial bulge was present in 40 patients. A systolic thrill was felt in seven, and in most patients the increased activity of the hyperdynamic right ventricle was detectable on palpation to the left of the sternum. Over the pul-

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monary area there was usually a systolic thrust, followed by a shock due to pulmonary valve closure—again, particularly in the younger patients.

A systolic murmur, most prominent in the second or third left intercostal space, was heard in every patient. It was rarely loud or harsh but was rather of the quality of the "flow-murmur." This murmur, resulting from functional stenosis of the pulmonary valve, was easily distinguished from the high-pitched, blowing, pan-systolic murmur of tricuspid insufficiency. The latter was heard in 10 patients, usually at the lower sternal edge. A short, early, low-pitched, diastolic murmur due to increased blood flow through the tricuspid valve was audible in 51 patients, and a high-pitched, blowing, diastolic murmur signifying pulmonary insufficiency was found in 5.

In no case was the diagnosis of Lutembacher's syndrome (atrial septal defect with mitral stenosis) seriously considered, nor was a true pulmonary stenosis suspected in any case.

The second sound was widely split and fixed with respect to the respiratory cycle in almost all patients who did not have a raised pulmonary vascular resistance. In the latter event it was louder and the split was narrower; at least this was the impression on auscultation.

*Electrocardiogram.*—The electrocardiogram showed all gradations from normal "crista supraventricularis pattern," through "right ventricular outflow tract hypertrophy," to undoubted "right ventricular hypertrophy."<sup>2</sup> We believe that the rSR' complex so frequently found in patients with atrial septal defect secundum is a manifestation of right ventricular outflow tract hypertrophy or dilatation and that it is not due to a conduction defect. This subject will be dealt with in greater detail in another paper. In only one case was the electrocardiogram normal for age.

*Fluoroscopy.*—Fluoroscopy was, in this series, an indispensable part of the diagnostic study. Together with examination of the x-ray films, it provided valuable information. Several features were found to be particularly helpful. The vascular markings of the lung fields were accentuated and noted well out to the periphery. The heart was enlarged, as were the main, left, and right pulmonary arteries. The increased activity of these structures was also obvious, in contrast to the lack of prominence of the aorta, left atrium, and left ventricle. Taken together, these features make up a pattern which becomes quite characteristic of atrial septal defect; departure from it at once arouses suspicion of either an additional lesion or a different diagnosis.

In this series, the heart was enlarged in 95 cases, sometimes slightly and sometimes massively, though in most cases the enlargement was considerable. In all cases except one, the typical outline of the atrial septal defect was seen.

*Cardiac Catheterization.*—Early in the development of our understanding of the diagnostic pattern, cardiac catheterization was used as an aid to diagnosis and provided a wealth of data with which the postoperative findings could be compared. Ninety-three patients were studied in this fashion before operation, either at this center or at others prior to referral here. At the present time, catheterization is not performed unless there are specific features which require clarification.

### Selection for Operation

On the basis of the considerations outlined briefly above, 100 patients were operated on, each with the diagnosis of atrial septal defect secundum. Two patients were given diagnoses preoperatively of patent ductus arteriosus in addition and eight of anomalous pulmonary venous connections. These lesions were found to be present in each case. Another seven patients were found at operation to have anomalous pulmonary venous connections previously undiagnosed. No cases of the Lutembacher syndrome or of pulmonic stenosis were found.

When the patient is between the ages of 2 and 45 years, the diagnosis of an uncomplicated atrial septal defect is a firm indication for operation. In older patients, other factors tend to weigh more heavily and must be carefully considered before operation is strongly advised. In the younger groups the problem of when not to advise operation is a more difficult one and is centered largely on the level of the pulmonary vascular resistance. The higher this is, and the lower the pulmonary blood flow, the higher is the risk of operation and the less the gain. We believe that the ideal time for operation is when the patient is between the ages of 2 and 10 years, the risk then being minimal and the gain from successful surgery the greatest. With the period of maximum growth still to come, obliteration of the defect gives such patients an excellent chance to have a completely normal circulatory system in adult life.

Operation was performed under conditions of hypothermia and inflow-outflow occlusion, according to techniques previously described.<sup>3</sup> No longer is there any doubt of the superiority of open-heart surgery in the handling of this lesion. It is essential that the exact relationships between the defect and the orifices of the various vessels entering the atriums should be accurately assessed visually. Since the success of the procedure depends on the reestablishment of a completely normal circulation, we are certain that direct vision is vital. Whether this is achieved by the use of hypothermia or the extracorporeal circulation is outside the scope of this article, and opinions will vary from center to center.

### Results

**Operative Mortality.**—Seven patients died as a direct consequence of operation, all in the first 43 cases. There were no operative deaths in the succeeding 57 cases. Three patients died from postoperative thrombosis (one of pulmonary embolism, one of internal carotid artery thrombosis, and one of internal jugular vein thrombosis). One patient died as a result of postoperative bleeding; two patients with a high pulmonary vascular resistance died a few days after operation from congestive cardiac failure. In the remaining case, the cause of death was not clear. The changes in blood-clotting mechanisms during hypothermia and anesthesia are discussed by von Kaulla and Swan.<sup>4</sup> It is felt that cooling to below 30 C (86 F) greatly increases the liability to these complications and also that the early postoperative institution of anticoagulant therapy, as soon as sanguineous drainage from the chest tubes has ceased, minimizes the risk of thrombosis. Of the seven patients who died, four were "good risk" patients and three "poor risk," having a high pulmonary vascular resistance.

One other patient, aged 39, died at home eight weeks after operation. The cause of death is said to have been a cerebellar hemorrhage; she was receiving anticoagulant therapy at the time. Of the remaining 92 patients, we have recent information on 91.

**Postoperative Assessment.**—The success of operation is measured by the changes which occur after operation in symptoms, signs, electrocardiogram, radiologic findings, and, in 44 patients, postoperative catheterization. If the defect has been completely closed, the results of evaluation by all these methods show that there has been regression toward normal. If this regression does not occur within a few months, the question must at once be asked: "Why not?"

Eighty-one patients are considered cured by all methods of evaluation. Three others are clinically cured; normal regression of symptoms, signs, electrocardiographic findings, and heart size occurred after operation, but postoperative cardiac catheterization in these three still showed some left-to-right shunt at the atrial level. One of these patients was the last on whom interrupted sutures were used to close the atrial defect, and it is possible that some small gaps remain between sutures. One patient may still have a pulmonary vein draining into the right atrium. The third also had before operation anomalous pulmonary venous connections into the right atrium. In none of these three patients is the shunt of any apparent hemodynamic importance.

Two patients have left-to-right shunts at the atrial level which are of consequence. One is a 12-year-old boy; he was operated on in August,

1954. Postoperative catheterization showed complete obliteration of the shunt, and he has remained without significant symptoms. However, the electrocardiogram failed to reveal any regression toward normal and further catheterization recently has revealed that there is now a shunt. He was also a patient in one of the early cases, on whom interrupted sutures were used. The other patient is a 45-year-old woman with an extremely large heart and tricuspid insufficiency but a low pulmonary vascular resistance. Repeated catheterization seven months after operation showed persistence of the shunt at the atrial level, and she is unimproved.

One patient, now aged 18, did well in all respects for 18 months after operation but since then, over the succeeding two years, has become progressively more disabled by dyspnea and fatigue, accompanied by the development of central cyanosis and finger clubbing. The cause of this has recently been established to be the shunting of inferior vena caval blood directly into the left atrium.

Two patients have been troubled by postoperative arrhythmias. One of these, aged 37, has palpitations and some dyspnea on effort due to atrial flutter. The other, aged 31, had a grossly enlarged heart at operation and had been in cardiac failure. In her case, the arrhythmia has been inimical to her progress. Our latest report, six months after surgery, indicates that she is now improving.

One patient, aged 18, also had a large heart preoperatively and still has some breathlessness on effort one year after operation. However, he has shown improvement. Finally, a housewife 33 years of age, who had severe pulmonary vascular changes before operation, has shown no regression of her pulmonary hypertension in the nine months since operation, despite successful closure of her atrial septal defect. Though she says that she feels better, she is still mildly incapacitated and shows no objective evidence of improvement.

Thus, of the 91 patients of whom we have recent information, only 4 give grounds for serious concern, 2 due to the persistence of a significant left-to-right shunt, one due to the development of central cyanosis, and one due to the presence of apparently irreversible pulmonary vascular disease.

### Summary

At the present time, 125 patients with an atrial septal defect of the secundum type have been operated on at this center under conditions of hypothermia, inflow-outflow occlusion, and direct-vision open-heart surgery. This report briefly analyzes the experience gained in the first 100 consecutive cases.

There were seven deaths due to surgery, all in the first 43 patients. In the last 82 patients there have been no deaths. The indication for operation,

in patients under 45 years of age, is the diagnosis of the uncomplicated anomaly. It is considered that, after the second year of life, the earlier the operation is performed the better. Surgery in a relatively asymptomatic child between the ages of 2 and 10 years carries a minimum of risk (less than 2%) and in all probability will ensure a normal cardiovascular system when maturity is reached.

With use of current methods, closure of the defect is almost assured. Only a small proportion of patients were not improved subjectively or objectively. A high pulmonary vascular resistance increases the risk and decreases the gain that can be anticipated from operation.

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