Ventricular Septal Defect: A Review

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Henri Roger, in 1879, was the first to accurately describe the clinical manifestations of ventricular septal defect, and the term "maladie de Roger" is often used in connection with these malformations. The lesion and the problems associated with it have attracted a great deal of attention in recent years, particularly with the advent of open heart surgery and a means for affecting a cure.

Embryology of The Ventricular Septum—
The septum develops between the fourth and seventh weeks of gestational life.1 At the end of the fourth week a median partition begins to project inward from the base of the common ventricle. This septum is brought into existence by the enlargement of the future halves of the ventricle on either side of it, and increases in height proportionately as the ventricular sacs become deeper. The anterior part of the septum is derived from a union of the ventral atrioventricular cushion, a local endocardial thickening, with this muscular septum below, and the right and left conus ridges from above. The posterior septum arises from the posterior atrioventricular cushion and the portion of the muscular septum that arises from the apical and dorsal walls of the embryo.2 The central points of fusion of these regions occurs at the pars membranacea. The anlagen of this part of the septum are a matter of dispute.

Classification of Ventricular Septal Defects—
Defects of the septum may involve regions of the septum related to either the outflow or inflow tracts.3 4 The outflow portion of the right ventricle lies between the pulmonary valve above and the nearest part of the tricuspid valve below. Defects in this region are all spoken of as high defects. The inflow tract lies posterior and caudal to the outflow tract.

The outflow tract of the right side of the ventricular septum is divided into superior and inferior regions by the crista supraventricularis. This muscular ridge lies on the floor of the right ventricle between the papillary muscle of the conus below and posteriorly, and the pulmonary valve above and anteriorly. The lesions inferior to the crista represent the most common site for ventricular septal defects, and anatomically lie just below the aortic valve cusps. This is generally spoken of as a membranous ventricular septal defect, but in most cases Becu et al3 found that the defect involved little or none of the membranous septum, but that the size of the defect was mainly related to the muscular part of the septum that was malformed.

The lesions superior to the crista are closely related to the pulmonary valve whose tissue forms their upper border. The membranous septum is generally intact.

Also known as high defects are those lesions in the upper or basal portion of the septum that are in close relation to the mitral and tricuspid valves.

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98
The last group, known as low defects, lie in the muscular septum near the apex and are of the type originally described by Roger. Their chief characteristic is the lack of close relationship to any of the valves.

**Incidence**

Statistics of incidence are difficult to evaluate since an autopsy series implies some degree of selection, and a clinical series leaves some element of doubt in the absence of absolute anatomic proof.

In several large autopsy and clinical series of congenital heart disease, isolated ventricular septal defect represented 5.5 per cent\(^5\) to 21 per cent\(^6\) of cases studied. These series place the defect as either the first or second most common lesion.

This lesion has been found complicating other cardiac anomalies in 22 per cent\(^5\) and 30 per cent\(^6\) of cases in two series.

Males and females appear to be affected equally.\(^3\)\(^,\)\(^7\)

**Diagnosis**

**A. History**

The average patient has one or more symptoms attributable to his condition, fatigue, shortness of breath, and retarded growth and development being the most common.\(^7\) Pneumonia, transient cardiac failure, or transient cyanosis are often found somewhere in the past history, as to a lesser degree are chest pain and chronic cough. Least frequent are hemoptysis, syncope, and paroxysms of rapid heart action.

**B. Physical Examination**

1) Murmur—This of course is the classical sign. In most cases\(^7\) it is systolic alone, heard along the left sternal border, but heard best in the third and fourth interspaces. It radiates widely. The quality of the murmur varies greatly. It may be harsh and low-pitched or blowing and high pitched. The intensity varies from faint to very loud.

A diastolic murmur may also be heard, rarely alone, but most often in combination with the systolic murmur. It may be early or mid-diastolic, the former heard along the left sternal border, and the latter at the apex.

The systolic murmur is probably due to blood flow through the defect. Its character, location, and transmission depend on many factors: a) Size and exact location of the defect, b) Magnitude of flow and systolic pressure gradient between the ventricles, c) The degree of pulmonary resistance, and d) The degree of rotation of the heart secondary to right ventricular hypertrophy.\(^7\)

The early diastolic murmur may represent functional pulmonic insufficiency as a result of dilatation of the pulmonary valve ring secondary to increased pulmonary blood flow. This theory is supported by the fact that most patients with the early diastolic murmur also have significant pulmonary hypertension.\(^7\) Another theory holds that the murmur may result from aortic insufficiency owing to inadequate support of the aortic valve cusps because of the septal defect.\(^14\)

The mid-diastolic murmur at the apex is interpreted as representing functional mitral stenosis as a result of greatly increased blood flow through a normal valve orifice. Dilatation of the left ventricular cavity with rapid filling may also be a factor.\(^14\)
2) Systolic Thrill—There is no diastolic thrill.
3) The second sound at the base to the left of the sternum is often accentuated, and its intensity correlates roughly with the degree of pulmonary hypertension.
4) Thoracic Asymetry—The left anterior chest may be more prominent than the right.
5) Cardiac Enlargement.
6) Visible Precordial Heave.
7) Cyanosis and Clubbing—Occasionally present, but never marked.

C. X-Ray Examination—

The normal fetal circulation involves a shunt, albeit right-to-left, between the pulmonary and systemic circuits through the ductus arteriosus. The addition of another shunt at the ventricular level of the fetal heart will still, as a rule, allow for an adequate fetal circulation. If, because of the defect, the heart grossly fails to sustain the circulation, intra-uterine death supervenes. Thus, Wittenbourg and Neuhauser\(^8\) feel that most infants begin life with a heart of approximately the same external dimensions and that at or shortly after birth, the x-ray will fail to distinguish between a normal heart and the majority of hearts having congenital lesions.

In patients beyond infancy having small defects, with only a small left-to-right shunt, and without pulmonary hypertension, the heart may be normal in size and configuration. With an increase in the size of the lesion and magnitude of the shunt there will be an abnormal load on both ventricles, but x-ray evidence of the lesion is first recognized as right ventricular enlargement. Both ventricles increase in size, but the right ventricle is probably more distensible and enlarges first. The left ventricle enlarges to a considerable degree before there is x-ray evidence of it.\(^8\)

In patients with larger defects the picture may be a changing one which mirrors the progression of the disease. Initially the heart as a whole will be enlarged. In the patient with a large left-to-right shunt this will be predominantly left ventricular, manifested by rounding of the apex and its displacement posteriorly and downward. In the latest stages of the disease, with marked pulmonary hypertension, there will be evidence of enlargement of both ventricles, often with dominance of the right. Normally the right ventricle does not add much to the transverse diameter of the heart, but the hypertrophic ventricle, because of its antero-inferior position, alters the contour of the heart in the postero-anterior film by elevating the left ventricle in a counterclockwise fashion thus elevating the apex, and by increasing the transverse diameter, including the right border, with some increased convexity on this side. As most of the increase in size of the right ventricle is from front to back, both oblique views will show the forward bulge of the anterior contour of the heart encroaching on the retro-sternal space.\(^8\)

In some instances there will be enlargement of the left atrium.\(^14\)

As the disease progresses there will be enlargement of the main pulmonary artery segment and increased vascularity of the peripheral lung fields.\(^14\)
D. Electrocardiogram—

The EKG is of some diagnostic aid, and of significance in establishing the magnitude of the physiologic abnormality, with which it correlates reasonably well.

A normal EKG in a patient with auscultatory evidence of septal defect indicates, if the lesion is truly present, an absence of significant pulmonary hypertension and of significant increase in pulmonary blood flow.

The presence of high amplitude equiphasic QRS complexes in the mid-precordial positions is said to be characteristic of, though not specific for, ventricular septal defect, and has been interpreted as left and right ventricular hypertrophy.\(^9,14\) A definite left ventricular hypertrophy pattern has been associated with large left-to-right shunts in the absence of severe pulmonary hypertension. This is in accord with the concept that the left ventricle is subject to a greater work load than the right in the presence of a left-to-right shunt at the ventricular level.\(^14\) As the volume of the shunt becomes small or disappears in the presence of severe pulmonary hypertension, there is less evidence of left sided hypertrophy, and a right ventricular hypertrophy pattern becomes dominant. This finding suggests a late stage in the natural history of the disease.\(^14\)

Evidence of atrial enlargement is not common in the EKG.

Evidence of conduction defects is not too frequent, but certainly does occur, and ranges from right bundle branch block to varying degrees of atrio-ventricular block.\(^7,9,14\) Although no investigation has been made into the course of the conduction system in hearts with ventricular septal defects, Kirklin et al.\(^4\) feel clinically that it is probably the same as in normal hearts. Thus, there may be a close relation of the Bundle of His and its right and left branches to those defects that appear in the outflow tract of the right ventricle inferior to the crista supra-ventricularis. Those defects that lie superior to the crista and immediately inferior to the pulmonic valve, as well as defects located in the apex of the septum, are not closely related to the Bundle of His and its branches.

E. Cardiac Catheterization—

Once again the findings will vary with the stage of the disease process, but qualitatively if not quantitatively, the results will point more specifically towards ventricular septal defect. Occasionally however, it is difficult to distinguish between this lesion and patent ductus arteriosus, persistent ostium atrio-ventricularis communis, persistent truncus arteriosus, and a single ventricle. Diagnostic information comes chiefly from determinations of pressure and oxygen content in the chambers of the right heart and pulmonary artery. Rarely the catheter will pass through the defect and into the left heart or aorta.

A left-to-right shunt at the ventricular level is present if the oxygen content of blood from the right ventricle is one volume per cent or more than that of the right atrium. When a shunt is only left-to-right, the peripheral arterial oxygen saturation should be normal.

In some cases,\(^7\) the left-to-right shunt may appear to be at the atrial level, i.e. there is an increase of more than two volumes per cent from the venae cavae to the atrium without further rise in the ventricle. All the cases in this series were later conclusively shown to have only ventricular septal defect, and most were characterized by having identical pulmonary
and systemic systolic pressures. The probable explanation therefore is that there was tricuspid incompetence, allowing for a shunt from the left ventricle directly into the right atrium. Obviously this finding complicates the differential diagnosis, and the patient may be subjected to surgery for an atrial septal defect.

The most significant pressure finding is an increase in right ventricular systolic pressure. This may be minimal in the small defects, or, in the larger ones with severe pulmonary hypertension it may equal the systemic systolic pressure. The diastolic pressure tends to be normal or only slightly elevated.\textsuperscript{18} In no instance should there be a significant gradient in pressure between the right ventricle and the pulmonary artery, as this would indicate pulmonic stenosis and a probable Tetralogy of Fallot.

Pulmonary artery wedge pressures are generally normal.\textsuperscript{10} Equalization of right and left ventricular pressures, and pulmonic and aortic pressures with a decreased peripheral oxygen saturation, suggests a large ventricular septal defect with a right-to-left shunt.\textsuperscript{11}

F. Contrast Roentgenologic Techniques—

These methods are almost never of value in demonstrating the presence of a ventricular septal defect.

Angio-cardiography is of value only for right-to-left shunts, and therefore is suitable only where there is reversal of flow through the defect.

Retrograde aortography is of value in the differential diagnosis to exclude lesions such as patent ductus arteriosus, and aortic or atrial septal defects.

G. Indicator-Dilution Curves—

This technique is based on the theory that the dilution of an indicator substance by the blood stream during its initial passage through the central circulation proceeds in a manner determined by several hemodynamic variables. These include, among other things, the velocity, volume, and pathway of the circulating blood.\textsuperscript{13} The actual technique involves intra-venous injection of the dye, and then recording from a peripheral artery the varying concentrations of the dye in the arterial blood.

When of sufficient magnitude, a centrally located left-to-right shunt produces an alteration in the dye-dilution pattern which is characterized by:

1. An appearance time and buildup time of normal duration.
2. An initial peak of concentration of dye that is lower than would be anticipated in a normal person for the amount of dye injected.
3. A prolongation of the disappearance slope of the curve.
4. The absence of the normal secondary peak of dye concentration due to systemic recirculation of dyed blood.

Presumably these alterations are due to central recirculation and to arterio-venous admixture.\textsuperscript{13}

Braunwald et al.\textsuperscript{12} suggest a similar technique, but inject the dye through a catheter directly into the left ventricle, left atrium, and aorta, and by determining the chamber in which the dye pattern becomes abnormal, they can localize the shunt.

The dye-dilution methods do not seem to be in any way superior to catheterization studies, and in several respects are less reliable. The
limitations include: a) Wide variability in contour of the curve in normal subjects; b) Left-to-right shunts of small magnitude cause only minor, and perhaps unrecognizable abnormalities; c) Without introducing the dye by catheter it is not possible to predict the anatomic location of the defect.  

H. Nitrous Oxide Test—

Grant et al. have devised this test in lieu of certain shortcomings of standard catheterization. They found that large differences in oxygen content of blood from the chambers of the right heart may be normal, and even multiple samples from a single chamber may vary widely. Owing to this, the diagnosis of a shunt into the right ventricle or pulmonary artery requires an increase in oxygen content of at least one volume per cent, and therefore small shunts may be missed.

The nitrous oxide test is performed during right heart catheterization. After a sample of blood has been drawn for nitrogen blank determination, the patient breaths a mixture of 15 per cent nitrous oxide and air for one minute. During this time blood samples are drawn simultaneously from the right heart and femoral artery, and analyzed for the gas. Nitrous oxide is a soluble inert gas, and is used for the test because of the wide arterio-venous difference that exists during the first minute of its administration. The arterial level rises quickly while the venous level lags because of absorption by the tissues.

In the presence of a left-to-right shunt, left heart blood rich in nitrous oxide increases the nitrous oxide content of right heart blood. Because the arterial level is so variable, the nitrous oxide content of the right heart is expressed as a percentage of the arterial content—the nitrous oxide ratio. A left-to-right shunt is present if this ratio in the right ventricle or pulmonary artery exceeds 20 per cent, or if it exceeds 30 per cent in the right atrium.

Hemodynamics—

A. Flow Through the Defect—

Flow through a defect, both in magnitude and direction, depends on its size and on resistance to the movement of the blood. Normally, pulmonary resistance is low, being only 15-20 per cent that of the systemic circuit. Since, for an adequate circulation, left and right sided output must be equal, it follows that the force of contraction of the left ventricle must be greater than that of the right. Therefore, a communication between the two ventricles will favor a left-to-right shunt. If the defect is small, relatively little blood may be shunted because the remaining septum offers greater resistance to flow than does the peripheral arterial circulation. A larger defect on the other hand means less septal resistance and more blood shunted.

Downing and Goldberg point out that with increasing pulmonary resistance, pressure in the right ventricle gradually increases, and as right heart pressure increases, the magnitude of the left-to-right shunt must decrease. The resistance becomes progressively greater, and eventually a state of balance is reached in which, resistance in both circuits being equal under basal conditions, there is no appreciable flow through the
defect. With activity and increased demand there may be a slight shunt in either direction. Even at degrees of resistance less than this a right-to-left shunt may occur under stress, accounting for the transient cyanosis seen in patients with a wide difference between pulmonary and systemic pressures. Ultimately, however, resistance to pulmonary flow becomes greater than resistance to systemic flow and a persistent right-to-left shunt develops, with the appearance of cyanosis. Pulmonary artery pressure does not then mirror pulmonary resistance because the septal defect now acts as an escape vent. Thus, there may develop the clinical features of an Eisenmenger Complex.

B. Pulmonary Hypertension—

It has been found by various authors\(^5\),\(^14\),\(^16\) that patients with ventricular septal defect do not generally die in the pediatric age range, i.e. 1 to 15 years. These authors point out that most children with this defect seen at autopsy are less than six months of age. They generally die of left and right sided congestive heart failure with varying degrees of pulmonary congestion and edema, and hepatic enlargement.\(^3\) Lillehei\(^1\) estimates that perhaps as many as 50 per cent die as a result of their defect in the first twelve months of life. The remainder go on to adult life to present with symptoms at a later date. Many of the early deaths, and most of the later symptoms are due, directly or indirectly, to the one progressive element in the disease process—pulmonary hypertension.

In summing up the present state of affairs, Lillehei has aptly pointed out that “patients with ventricular septal defects have, in reality, two diseases. The cardiac lesion is being well handled, but the problem is in the management of the secondary disease—pulmonary hypertension and arteriosclerosis.”

Swan et al.\(^10\) feel there are three factors which may control pulmonary artery pressure:

1. Increased resistance to pulmonary flow beyond the pulmonary capillaries, as in mitral stenosis or congestive heart failure. In such an event the pulmonary artery wedge pressure would be increased, but in ventricular septal defect this is characteristically normal, so this mechanism may be excluded.

2. Increased pulmonary flow in the presence of unchanged resistance. This may well be a factor since it has been found that in patients with patent ductus arteriosis and high pulmonary pressures that manual closure of the ductus results in a decrease in pulmonary artery pressure. Swan has found increased pulmonary flow in the 20 patients he studied, and the average proportion of pulmonary flow representing the shunted blood was 50-60 per cent, i.e. pulmonary flow was about double the systemic.

3. Increased pulmonary resistance. Calculations of resistance are based on measurements of pressure and flow (Poiseuille's Law), and the finding of increased pressure means that the observed increase in pulmonary artery pressure must be disproportionately greater than the increase in pulmonary blood flow. These relative alterations in flow and pressure can best be quantitated in terms of resistance, and as indicated earlier, this must be proximal to the pulmonary capillaries. This is undoubtedly the most important factor in the pathogenesis of pulmonary hypertension.
The most widely accepted views as to the cause of this increased resistance are those of Civin and Edwards. In the fetus the distribution of blood from the right ventricle to the lungs on the one hand, and through the ductus arteriosus to the systemic circuit on the other, is largely dependent on the state of the smaller pulmonary vessels, i.e. at the level of these vessels the resistance to flow is similar to that in the systemic circulation. Were resistance to flow in the pulmonary arterial vessels lower than that in the systemic vessels, as is the normal adult condition, blood from the right ventricle would be almost completely distributed to the lungs. Little or no blood would flow through the ductus into the aorta. In a later article they gave substance to this theory by tracing the development of the pulmonary arteries and arterioles in a series of sections from different age levels. They found that in the muscular fetal arteries the lumen of the vessel is less than the transverse diameter of the vessel wall. Most of this thickening was due to hypertrophy of the medial musculature. By the second half of the first month of life this thickening begins to diminish, and by the time the infant reaches six months of age the small arteries have become dilated and resistance to flow is low. They then propose that the pulmonary hypertension seen at birth represents a continuation of the fetal vascular state, and indeed they have demonstrated muscular arteries of the fetal type in the lungs of patients dying from Eisenmenger's complex. This they consider to be a compensatory mechanism which permits the patient to remain alive. However, the patient pays a price for this, since the mechanism has pulmonary hypertension as a component part.

Intimal fibrous changes may be present in the pulmonary vessels of older patients and produce further narrowing. These changes seem to arise after the medial changes are evident, and are considered to be a reaction to the traumatic influence of existing pulmonary hypertension. These changes are thought to be progressive and to increase pulmonary resistance even further to a point where it equals or exceeds systemic resistance, producing reversal of shunt and cyanosis.

Clinically, the increased pulmonary artery pressure seen at birth may be minimal or severe. After birth, the pressure may remain stable, increase slowly, or rise rapidly. Where pressure is high initially or rises rapidly an early death is likely.

C. Relation of the Defect to Pulmonary Hypertension—

The size of the defect is obviously of prime importance in the development of pulmonary hypertension. It has been pointed out that in a small lesion the factor limiting pulmonary blood flow is the size of the defect. The resistance regulating the major portion of the pulmonary blood flow is at the ventricular level in contrast to large defects where the limiting factor is the capacity of the pulmonary vascular bed. In accord with this the former group is found to have little pulmonary hypertension.

Warden et al. feel that a factor at least as important in this regard is the precise anatomic relation of the defect to the orifice of the pulmonary artery. They believe that even relatively small defects, wherever they are located, situated so that the shunt of blood from the left ventricle projects directly into the pulmonary artery are hemodynamically more deleterious than considerably larger defects situated so that the left
ventricular jet strikes the opposite right ventricular wall in such a way as to create turbulence in the outflow tract of this ventricle. The consequent increased resistance due to this turbulence, creates, in effect, a functional pulmonic stenosis tending to dampen the physiological consequences of the defect. Most authors\textsuperscript{8,14} believe that this is correct in theory but is not borne out in practice, and that the size of the defect is the sole factor governing the clinical picture.

Relation To Other Complexes—

A. The Eisenmenger Complex—

This is a cyanotic type of congenital heart disease comprising a high ventricular septal defect, overriding of the defect by the aorta, pulmonary hypertension, and right ventricular hypertrophy. The only difference between this and isolated septal defect is the overriding aorta and consequent cyanosis. The mal-position of the aorta may range from mere overriding to actual and complete dextraposition. In the former type, Bailey\textsuperscript{11} feels that the complex is a misnomer. He states that a simple defect located high in the septum will in any heart give the impression of overriding. Others\textsuperscript{14} take a less demanding point of view and feel that when pulmonary hypertension is severe in an isolated defect and reversal of flow through the defect occurs, that there is created a physiologic, albeit not anatomic, overriding of the defect. Thus, this group prefers to think of the Eisenmenger Complex not as an anatomic entity, but rather as a stage in the natural history of an isolated septal defect. It must be pointed out however, that the existence of an anatomic Eisenmenger Complex, i.e. when there is actual dextraposition of the aorta, cannot be denied.

B. The Tetralogy of Fallot—

Gasul et al.\textsuperscript{16} in studying a group of 15 infants with ventricular septal defects over a course of several years, have in four cases observed the development of pulmonic stenosis. This change was clinically manifested in all by a diminished second heart sound at the pulmonic area, and in two by the development of the clinical findings of Tetralogy of Fallot.

These authors suggest that the child with ventricular septal defect may protect himself in at least two ways—either through the development of pulmonary hypertension, or in some cases through the development of pulmonic stenosis. They make no estimate as to how many cases go this way, but do feel that if this change is to occur it does so at an early age or not at all.

This is the only report of such a phenomenon this author has seen in the recent literature.

Prognosis Without Surgery—

Most of the work in this paper has been directed toward the commoner class of ventricular septal defects, i.e. those variously situated high in the septum, but mention should also be made of the less common but more benign small defect located low in the septum and unrelated to any valve orifice. This is the lesion referred to as maladie de Roger, and it carries a rather good prognosis. The patients develop almost no pulmonary hypertension, and except for the risk of sub-acute bacterial endocarditis they may expect to live a normal life.
As for the larger, higher defects, Lillehei believes that probably 50 per cent die in the first year of life because of the lesion. Most of these die in left ventricular failure, some probably in conjunction with pulmonary infection. As the remainder of the patients reach adult age and develop progressive pulmonary hypertension, right ventricular failure contributes a rapidly increasing element to this picture. Just how long these patients may live is difficult to say, but this author has rarely read of a patient surviving beyond 55 to 60 years of age.

Sub-acute bacterial endocarditis is a risk that all patients must carry, no matter how large or small the defect. The organisms may implant either on the defect margins, or on the free wall of the right ventricle opposite the defect. Selzer estimates that 25 per cent of patients with this defect develop bacterial endocarditis. Others believe the figure is lower, possibly below 5 per cent.

**Selection of Patients for Corrective Surgery**

Lillehei has outlined some criteria for surgery:

1. Infants who fail to gain weight.
2. Infants with recurrent or persistent cardiac decompensation that is resistant to medical therapy.
3. Infants who suffer repeated respiratory infections.

On the other hand, in infants who are gaining weight normally or nearly normally and who appear to be responding well to medical therapy, he prefers to delay corrective surgery till at least 18 months when the child is perhaps better able to withstand major surgery. He feels that waiting longer than this increases the risk, since in some patients the pulmonary artery pressure can double in one year. On the other hand there are some patients whose pulmonary pressures may remain stable for years, however, there is at present no way of telling into which group any given patient will fall.

From a technical aspect early operation is also desirable. Lillehei claims that the defects are nearly always small and easily closed by stitches only. To delay surgery would be technically undesirable since these defects grow in size as the septum grows.

In the adult the selection of surgical patients becomes a great deal more difficult. The chief problem is the existence and degree of pulmonary hypertension. It has been shown that in the presence of severe pulmonary hypertension the operative mortality is markedly increased, and the immediate and prolonged prognosis after surgery are not good. Presumably the pulmonary vascular changes have progressed to a point where they are no longer reversible. The problem may eventually resolve itself into the selection of patients on the basis of lung biopsy. Apparently the state of the pulmonary vessels seen in these sections correlates rather well with operative risk and prognosis.

**Surgical Correction**

The most immediately apparent method of therapy for a patient with this lesion is closure of the defect. For many years this was mere phantasy, but in the 1930's Claude Beck opened a new era with the demonstration that the heart could be approached surgically. The barrier had been crossed, and progress toward repair on intra-cardiac lesions was not far off. The technique of closed cardiac surgery however, still was
not satisfactory for repair of ventricular septal defects. The most frank approach, i.e. suturing the edges of the defect, was rarely successful. Blind implantation of sutures was both dangerous and difficult, especially in those defects whose margins lay in close proximity to the aortic valve ring. Alternatives to direct suture were suggested. Bailey\textsuperscript{11} favored plugging the defect by pulling a tapered, pedicled, viable pericardial tube through it from the right ventricular side and suturing it to the free wall of the left ventricle, hoping for fibrous union between the tube and the defect margins. However, this and other techniques suggested still fell short of the results anticipated with the ideal "direct" approach. This approach was finally made possible with the advent of open heart surgery.

Surgical correction of the lesion by open cardiectomy is mainly dependent upon the use of either hypothermia or extra-corporeal circulation, the most popular variant of which is the mechanical pump oxygenator. Both techniques are successful, although the pump oxygenator is somewhat more in favor since it allows for longer operations than does hypothermia. There are certain problems inherent in each method, but these are beyond the scope of this paper.

If the heart is beating actively during the cardiectomy air may be aspirated, pass to the left side of the heart and be ejected into the aorta at the next contraction. To avoid this, drugs such as acetyl choline, prostigmine, or potassium citrate are administered via coronary artery perfusion to produce a flaccid heart.

Direct suture may be used in small defects, but in the larger ones it is often followed by partial disruption along the suture line. It is believed that the living ventricular septum allows tight stitches to cut through rather readily. Thus, in the larger lesions, Kirklin\textsuperscript{4} recommends the use of a securely anchored polyvinyl-formal (Ivalon) sponge cut to conform to the shape of the defect. Mattress sutures are placed for repair so that they grasp one side of the defect, pass through the sponge, and are then anchored to the other side of the defect. Thus, the sponge helps to fill the tissue defect, and allows a snug repair without the stitches having to be drawn too tightly.

\textit{Complications of Surgery—}

Some degree of post-operative conduction defect is common,\textsuperscript{22} but generally well tolerated. The most serious conduction defect, and the one that is a threat to life is complete heart block. Lillehei\textsuperscript{4} estimates the incidence of complete heart block following surgery to be about 12 per cent, probably because in the most frequent location of defects there is a close relation of the conduction system to the area for placement of sutures. He also believes that in patients with severe pulmonary hypertension and diminution of cardiac reserve there is a predisposition to block. This element is difficult to control, but interruption of the conduction pathway by a suture can be guarded against if certain precautions are taken. If, with the placement of a single stitch, complete heart block occurs, Lillehei removes the offending stitch and places it in a slightly different place. If this maneuver is not of value he continues regardless since the primary objective, if the patient is to live, is closure of the defect. He then attempts to correct the condition, if possible, by post-
operative medical therapy with Isuprel, or with the combined use of myocardial electrodes and the artificial pacemaker.\textsuperscript{23}

In all age groups, but particularly in children under two years of age, one of the chief factors involved in post-operative morbidity and mortality is the development of respiratory complications.\textsuperscript{17} Even normal infants have limited respiratory reserve, but in those infants with large defects and severe pulmonary hypertension the reserve is even further diminished. Thus the management of the secretions that are inevitably retained in their bronchial tree, and the consequent frequent development of pneumonia, becomes a major problem, and one that is as yet unsolved.

Another factor complicating surgery in patients with advanced pulmonary pathology results from the altered hemodynamics following closure of the defect. Some patients will die due to circulatory failure resulting from the high resistance in the pulmonary circuit.\textsuperscript{23}

\textbf{Mortality Rates—}

In the largest single series reported on at this time, Lillehei et al.\textsuperscript{24} have operated on 154 patients for closure of ventricular septal defects. Their overall mortality rate was 29.3 per cent. The figures for specific groups are significant, and are therefore presented:

\begin{center}
\begin{tabular}{|c|c|c|}
\hline
 & Moderate* & Severe** \\
 & Per Cent & Per Cent \\
Under 2 Years of Age & 29 & 48 \\
Over 2 Years of Age & 6.8 & 37 \\
Total & 14 & 40 \\
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*Pulmonary artery pressure less than 70 per cent of aortic pressure.
**Pulmonary artery pressure greater than 70 per cent of aortic pressure.

From these data it is evident that the "ideal" group from the standpoint of surgical risk are patients over two years of age, and with only moderate or minimal pulmonary hypertension.

It is also of interest to note that this group found that the type of cardio-pulmonary by-pass used does not seem to have an effect on mortality.

\textbf{Therapeutic Results—}

In catheterization studies done on patients surviving surgery,\textsuperscript{24} most patients appear to have a significant decrease in pulmonary artery pressure and blood flow. Some degree of residual shunt is, however, not uncommon.

The various murmurs may or may not disappear. In all likelihood this will depend on the degree of pre-operative cardio-pulmonary aberrations in all parameters, as well as the completeness of closure of the defect.

Adams et al.\textsuperscript{22} found that pre-operative growth failure which is so frequently observed, is remedied and nearly all their patients returned to average growth levels post-operatively when the shunt had been closed and pulmonary hypertension was not a prominent feature.

They also found that increased pulmonary vascularity, as estimated by x-ray, soon returns to normal, while the cardiomegaly disappears less strikingly.

In this same series all patients were found post-operatively to have complete or incomplete right bundle branch block.
REFERENCES


