Clinical Problems Related to Surgical Repair of Intra-cardiac Defects with the Aid of an Extracorporeal Pump-Oxygenator

By Howard B. Burche1l, M.D.

The medical problems in the selection of patients for direct-vision reconstructive operations utilizing the pump-oxygenator of the Gibbon type are discussed. The evaluation of the pulmonary vascular changes is one of the most important medical tasks in many cases. The operative risk for such lesions as ventricular septal defect with high pulmonary flow, tetralogy of Fallot, pulmonary stenosis with intact ventricular septum, and atrial septal defect of the partial atrioventricular canal type is constantly decreasing and is 15 per cent or less at the present time. Heart block and cardiac arrhythmias are still, on occasion, serious postoperative complications. During 1 period of 3 months, subacute bacterial endocarditis (Pseudomonas) occurred in 4 patients but it has not recurred since.

It is an honor to have been asked to give the Henry Jackson Lecture this year. This invitation has afforded me the opportunity to correspond with his son, Henry Jackson, Jr., and hence to learn something of the interests of his father whose 82 years of life spanned the years of the development of the practice and teaching of scientific medicine in America. Henry Jackson came from a famous family of physicians and his first contribution to medical literature was in 1886 as "A Letter from Vienna," indicating one pattern of training of the best physicians at that time. He thereafter returned to Boston where he lived until 1940, dying then just at the dawn of surgical therapy of heart disease. One of his last communications was in 1933, 60 years after graduation, when he contributed to a symposium on the "Care of the Chronic Heart"—speaking on non-valvular heart disease of middle and old age and emphasizing the important principles, particularly the ease and the error of making a poor prognosis in many cases. His son has mentioned that he possesses the original manuscript of this paper, with marginal notes containing the names of contemporary notables who had apparently been under his father's care.

The title of my lecture was suggested by your chairman, Dr. Dexter, and the data I shall present are related to surgical treatment primarily carried out by Dr. J. W. Kirklin but more recently also by Dr. F. H. Ellis, Jr., and the laboratory data have been made freely available to me by Dr. E. H. Wood. I am indebted also to numerous other clinical colleagues for their help with this paper, particularly Dr. J. W. DuShane of the Section of Pediatrics.

Perhaps it now would be a dull story to give you a detailed account of the development of cardiac bypass surgery initiated at our clinic approximately 5 years ago. However, few of us forget that first occasion on March 22, 1955, when the pump-oxygenator was used in the operating room—the interior of the heart of a little girl was exposed, a ventricular septal defect was repaired, and recovery ensued. At the present time, this child's heart is normal on physical examination. The artificial heart-lung apparatus, patterned after, but modified from, that developed for Dr. Gibbon, has continued to serve in a satisfactory way since that time. Since August 1956, the heart has been stopped by potassium during the repair, as originally suggested by Melrose and first done clinically by Effler. It is proper to mention here also the pioneering and continuing stimulating surgical results with direct-vision cardiac surgery of Dr. Lillehei and associates. In the approach to the problems of direct-vision cardiac surgery with the aid of a pump-

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oxygenator, it might be well to review an index of a theoretical monograph, as follows.

1. Introduction—The Need.
2. Technical Problems of the Pump-oxygenator.
3. Technics of Intracardiac Repair.
4. Potassium Asystole.
5. Monitoring of Physiologic Variables during Operation.
7. Pulmonary Hypertension as a Specific Problem.
8. Surgical Mortality.
10. Electrocardiographic Studies.

The specific problems introduced by the "bypass" have been many and varied. Their careful study in animals and in man has provided considerable knowledge of the proper management of whole-body perfusion. The importance of monitoring all the physiologic variables that we could monitor is that it gave us an understanding of basic principles at the time of the experimental work in animals and the early operations in man, and allowed us to discriminate and choose those measurements of promising perpetual value. The contributions of the laboratory of clinical pathology directed by Dr. T. B. Magath, aided by Dr. D. R. Mathieson, in blood grouping, cross matching, and collection of 7 to 10 bottles (500 ml. each) of heparinized blood the morning of operation have been one of the pillars in support of the surgical accomplishments.

To summarize from a clinical viewpoint, the present questions pertinent to bypass intracardiac surgery are as follows:

1. What is the risk of the extracorporeal pump-oxygenator per se?
2. What types of lesion may be cured and what are the specific surgical mortality rates?
3. When is palliative treatment (partial correction of a defect) warranted?
4. What are the evidences of "cure"?
5. What are the economic problems to patient and institution?

Some of these questions will be discussed later.

Diagnosis

It is not within the province of tonight's discussion to enter deeply into the intricate problems of diagnosis, but rather to summarize some general points that are pertinent to the surgeon's technical plans, to the immediate risks, and to the chances of a good result. In many instances we believe that on the basis of clinical data we can recommend surgical treatment without cardiac catheterization or angiocardiography, or both. If we can be reasonably certain that the patient has 2 ventricles and 2 great vessels, has no gross valvular defect, and has or has not arterial desaturation, dependent upon the presence or absence of pulmonary stenosis, he is potentially a surgical candidate. Even these simple criteria are not iron-clad guides for absolute exclusion of patients from surgical treatment; in a case of a single ventricle Dr. Kirklin has successfully placed an artificial ventricular septum in the heart of 1 patient, whose postoperative life was most satisfactory for 6 months, death then occurring suddenly with effort. Here is an instance in which a Bernheim effect could operate, I suppose, and if it should, it is an exceptional instance in the whole field of cardiologic problems where such an explanation for symptoms could pertain. Dr. Kirklin also was technically able to partition a truncus and do a reconstruction on the single truncus valve, separating it into 2, but the child in this instance survived only 24 hours. Anatomically this reconstruction appeared to have been soundly planned and executed and the ventricular septal defect completely closed.

One may well emphasize that despite our reliance on clinical diagnoses, cardiac catheterization is frequently performed when atypical clinical problems present themselves (and these atypical and complicated cases seem to be more and more frequently encountered). Cardiac catheterization data are valuable in some cases, for instance when special problems of hemodynamics need clarification, particularly when an objective value for pulmonary vascular resistance is needed; or in others, when an anomalous flow pattern needs delineation, as in anomalous pulmonary venous connections. The utilization of dye technics—the
sequential concentration of dye at a sampling site (for example, the radial artery) downstream from the injection site (for example, the pulmonary artery) as developed and applied in our laboratories under the direction of Dr. E. H. Wood—has been of great value in diagnosis.

It probably comes as no surprise to you to learn that the surgeon not uncommonly finds associated defects that were not suspected clinically or even sometimes on cardiac catheterization; an instance of this is that of atrial septal defects in association either with ventricular septal defects or with the tetralogy of Fallot. Also an occasional associated ductus has been encountered in patients in whom operations were scheduled only for repair of the ventricular septal defect. Of hemodynamic interest is the surgeon's observation, in a few cases, of the large size of the ventricular septal defect encountered when the hemodynamic data would have been indicative of a moderate-sized aperture only. With the opened, quiescent heart the surgeon has reported defects approximating the size of the aortic orifice when the moderately increased pressure in the pulmonary artery and the moderate left-to-right shunt logically would have been the basis for prediction of a moderate-sized defect. To me the only available explanation is that the defect must be decreased with systole (and that no significant left-to-right shunt is occurring during diastole). In 1 adult patient, the surgeon estimated the defect ("largest ever seen") as 5 by 2.5 cm., yet the aortic systolic pressure exceeded the right ventricular systolic by 15 mm. Hg. Preoperatively this patient had constant low values for her systemic blood pressure (80 to 85 mm. Hg systolic) and one might conjecture about this having had a benign influence on the pulmonary vasculature. The results of the surgical correction were good, with cardiac catheterization data 5 months later showing normal hemodynamics. The clinical recognition of valvular incompetence in association with septal defects has not always been easy in my experience. The association of aortic insufficiency with a ventricular septal defect represents a well-known syndrome, but fortunately a rare one; at present adequate repair of the prolapsed aortic leaflet would appear technically difficult. If the insufficiency is slight clinically, one does not have the grave concern of sending a child to operation, particularly since the previous potential difficulty of losing the perfusion retrogradely through the aortic orifice has been obviated by the use of potassium asystole and the cross clamping of the ascending aorta.

Of all the hemodynamic problems that engage the attention and the investigative potential of the physician in the field of selection of patients for surgical treatment, that of pulmonary hypertension has the highest priority. The essence of this challenging problem is that there are many patients whose heart can be repaired but whose pulmonary vessels cannot. Colloquially and slightly erroneously in a purely semantic sense, we speak of a lesion as "anatomically operable" but "physiologically inoperable." The inadequately or partly answered questions in this field of pulmonary hypertension in congenital heart disease associated with left-to-right shunts are: 1. How is structure related to pulmonary vascular resistance? 2. Is there any constant recession and secondary accession in the thickness of the vessels in the lungs from birth onward? 3. What is the genesis of the intimal lesions? 4. What retrogression may take place in the varied morphology of the thickened vessels and in the pulmonary hypertension following repair of a defect allowing a left-to-right shunt? 5. Can one identify a factor of dynamic resistance (vasomotion) and differentiate it from the structural resistance? 6. What would be the advantages of pulmonary biopsy in a patient with pulmonary hypertension in regard to the determination of operability and the prediction of postoperative recession in the pulmonary hypertension?

In the early work of Dr. J. E. Edwards and myself it was considered that the correlation between pulmonary pressure (and pulmonary vascular resistance) and pulmonary vascular morphology was a crude one. The extremes of pressure and vascular change could be seen to be related, but it was difficult or impossible to predict the extent of a left-to-right shunt, or to predict the weighting of a bidirectional shunt, in the presence of pulmonary blood pressure
equivalent or nearly equivalent to systemic blood pressure. Dr. Edwards' has proceeded to refine the categories of pulmonary vascular changes and has introduced the useful concept of the high-resistance, high-reserve, and the high-resistance, low-reserve lung, the term "reserve" being related to the capacity of the vessels of the lung to carry the flow of blood. A lung with low reserve is one in which there are advanced obstructive changes of the intima. Despite these advances, it is our belief that a study of vascular changes, as might be done by pulmonary biopsy, would not furnish a sound basis for determining operability, if this were already in grave doubt prior to this examination. As a special case in point, a child had a good result from closure of multiple ventricular septal defects, but died 4 months later from hemorrhage of the subclavian artery. If a biopsy had been done, the vascular-bed vessels would have been assessed as severely obstructed, and with such a criterion for selection for operation it would have excluded the child from the successful surgical correction of the intracardiac defect. It is of note (and perhaps a little disappointing) that the pulmonary vascularity in this case showed no recognizable regression although pressure in the pulmonary artery was normal at the end of the repair.

Occasionally pulmonary biopsy will be of value. In one instance of a cyanotic young man, the pulmonary artery had not been entered on cardiac catheterization and although there was no characteristic accentuation of the second sound in the pulmonary area, the lung fields were of about normal vascularity and one was at a loss to know with certainty whether the patient had, with the demonstrated right-to-left shunt through a ventricular septal defect, a pulmonary stenosis or a severe pulmonary hypertension; in the former instance, operable; in the latter, nonoperable. Pulmonary biopsy was urged by Dr. D. C. McGoon, of the thoracic surgical team; it revealed thickening of the vessels that could only reflect severe pulmonary hypertension.

In patients with ventricular septal defects the main criterion for surgical treatment is the presence of a left-to-right shunt. The surgeon can repair this leak; surgical treatment is not mandatory if the leak is small due to a small orifice, but if the leak is small due to right ventricular hypertension and pulmonary vascular changes, surgical treatment is fraught with danger and the late results are not predictable at this time.

**Electrocardiograms**

Electrocardiographic studies are of inestimable help both in the differential diagnosis of intracardiac defects and in the estimation of the past and present hemodynamic states of patients having a ventricular septal defect. When the pulmonary fields are plethoric, patients who have a single ventricle or tricuspid atresia may simulate the patient with a simple ventricular septal defect, but the electrocardiogram may suggest the correct conditions. Patients with the atrioventricularis commune defect (common atrioventricular canal or so-called ostium primum defect) often have a characteristic electrocardiogram and vectorcardiogram. The left axis deviation in the standard leads and an apparent partial right bundle-branch block in the precordial leads are characteristic and are only very occasionally seen in a patient with an atrial or ventricular septal defect. Gross mitral incompetence is not necessary in the genesis of the nearly diagnostic tracing. In patients with ventricular septal defect one would like to demand evidence of left ventricular preponderance in the electrocardiogram if operation is to be recommended, but one cannot use the electrocardiogram as an infallible guide to operability, in my opinion. In this regard, while I am enthusiastic concerning the electrocardiogram as an index of operability for patients with ventricular septal defect, it cannot stand alone as an interdiction of surgical treatment if it should show predominant right ventricular hypertrophy.

Following surgical repair of a ventricular septal defect, either as an isolated lesion or as a component of the tetralogy of Fallot, it is expected that a right bundle-branch block will probably have developed, and that this is of classic conformation (fig. 1). In patients with defects of the ventricular septum in the muscular portion, one would not expect postoperative bundle-branch block and indeed this has hap-
Surgical Repair of Intracardiac Defects

Fig. 1. Electrocardiograms from a young man before (left) and after (right) successful operation for pulmonary stenosis and a ventricular septal defect. The preoperative right ventricular systolic pressure averaged 90 mm. Hg, and a left-to-right shunt was present.

Fig. 2. Electrocardiograms taken on a girl, 19 years of age, 2 weeks after successful repair of tetralogy of Fallot. Complete heart block occurred postoperatively. The upper tracing shows interference dissociation, while the lower one shows an interference beat followed by 2:1 heart block. Arrows denote interference beats.

[p. 980]

When a patient has atrioventricular heart block, it is to be noted that only rarely, and then in the late hospitalization period, is there an increase in the P-R interval—atrioventricular conduction being in a normal range or no conduction occurring at all. In accordance with this conduction characteristic, one may often see sequences of 2:1 block followed by 1:1 conduction without difference in the P-R interval. Frequently, as a premonitory sign of resumption of normal sinus rhythm in a patient with heart block, one may see interference dissociation; that is, there may be a conducted beat if the P wave falls in a time-specific zone at the end of the T wave or the beginning of the U wave (fig. 2). This phenomenon is in accord with basic physiologic observations on the increased (or supranormal) conductivity during the late recovery period of injured excitable tissues; and in this instance of surgical injury to the junctional tissue of the heart, its manifestation is a welcome herald of the return of normal sinus mechanism. In patients with atrioventricular dissociation, the QRS configuration is usually that of left bundle-branch block, indicating that the most active tissue from the point of view of automaticity is in the right bundle area. As it is
expected that permanent right bundle-branch block is present, the possibility exists that complete heart block, when it occurs, is due to bilateral bundle-branch block and not necessarily to a lesion of the main bundle. In 1 patient, for a period just before resumption of normal atrioventricular conduction, there was retrograde (R-P period) conduction with the atria following the ventricular pacemaker and the QRS being of the left bundle-branch type. Following the repair of atrial septal defects, but not characteristically after other reconstructive intracardiac operations, nodal rhythm has been relatively common, usually at a moderately slow rate, and this has been of benign nature, perhaps even of salutary influence because of the control of the heart rate.

As yet, the full involution of the electrocardiogram to normal following surgical procedures has not been observed in patients, disregarding those having surgically induced right bundle-branch block, but one would expect a complete transition to normal to take some years if it is to occur at all. The electrocardiographic changes due to pericarditis following operation are unexpectedly minimal in most cases, in both the early and the late postoperative periods, and no residual electrocardiographic changes attributable to pericarditis have been seen.

One may mention that with the opened heart, there was the tempting opportunity to explore the pattern of excitation of the endocardium but such studies were deferred because of the possibility of increasing risk, and now with the almost universal utilization of potassium-created asystole, this electrocardiographic opportunity has vanished.

**Postoperative Management**

Before and after the surgical procedure, infants and children are weighed, and postoperatively they are brought to a recovery ward where they are under constant surveillance by experienced nurses. Special attention is paid to the drainage from either thoracic cavity, urinary flow, and any other loss of fluid, and the fluids are replaced as seems necessary on the basis of an accumulative chart with entries every 15 minutes for the first hour or so, then less frequently. The requirements for water (considered as 750 ml. per M.² of body surface per 24 hours) are met, but no electrolytes are given for the first 2 days or more.

Dr. G. S. Sturtz, with Drs. Kirklin, E. C. Burke, and M. H. Power,¹ has studied various aspects of water metabolism in the first 3 days of the postoperative period in 21 patients who have undergone intracardiac operations with the aid of the extracorporeal pump-oxygenator. The weight of the child appeared to be the simplest guide to the requirements for water, but the studies on obligatory water losses were most informative in helping to arrive at a figure for probable water needs. The daily obligatory losses of water were considered as the obligatory urinary water (1 ml. of urine per mol of solute), insensible water loss, and the abnormal losses. The insensible water loss was assumed to be 90 per cent of insensible weight loss, according to the method of Newburgh. The daily water needs were considered as the obligatory water loss minus the metabolic water, assumed to be 270 ml. per M.² of body surface per day. These calculations gave the water requirement as 500 ml. per square meter for the first day and 750 ml. per square meter for the second and third days. Of some interest is the finding that the serum concentration of water was low immediately after perfusion but returned to normal in 7 to 10 hours, and that it did not always conform to values expected from the water balance data. The data on the urine revealed that most patients in the first 36 hours excreted predominantly obligatory urinary water, which is believed strong evidence of an existing state of antidiuresis during this period.

Excessive bleeding has rarely been a problem and in only 2 patients did it seem that it was a serious hazard to recovery. The patients sometimes are in need of care because of severe hypotension in the early postoperative period, but this is uncommon in recent months. In such instances norepinephrine (Levophed) is still the drug that is favored and undoubtedly has been lifesaving in some instances.

The use of digitalis in our surgical cardiac cases has led to many discussions within our group, and I cannot claim exact scientific data to support our present practice utilizing this
valuable medication. Any patient who has, or has had, heart failure or who has atrial fibrillation receives full doses of digitalis preoperatively and maintenance doses following operation. Those infants and children who have had no digitalis preoperatively may receive it postoperatively on suspicion of heart failure. This practice is based primarily on the fact that some infants and children have developed heart failure postoperatively. In addition, there is suggestive evidence for early use of digitalis from observations on the ease with which nonsurgical infants have developed heart failure with paroxysmal tachycardia. Adults who have not had digitalis prior to operation receive it postoperatively only as the need presents itself. A longer period of observation before its administration is warranted in adults as compared to children. In a few instances in which rapid arrhythmias have appeared postoperatively it has been most difficult to know whether digitalis had already been given in excess or the patient needed more. In one instance it was felt reasonably certain that the atrial tachycardia with varying heart block was related to digitalis. No response to the administration of potassium occurred, but there was alleviation by procaine amide, and recovery ensued without more digitalis being administered. In another instance late in the postoperative period a sinus tachycardia was associated with a 3:2 block with the Wenckebach phenomenon following the administration of only 2.5 mg. of digoxin in a period of 36 hours.

Complete heart block has continued to be an occasional, troublesome occurrence in the postoperative period in respect to its contribution to both morbidity and mortality. Transient atrioventricular dissociation as the patient’s own heart takes over the circulation in the operating room is fairly frequent and not particularly worrisome, but if from the time of the operation heart block persists with an idioventricular rate less than 50 per minute, it is of ill omen as to survival. I had believed that if the ventricular rate was more than 60, with heart block, there would be no circulatory embarrassment; while this appears to be true, the prognosis must be guarded. To illustrate, in 1 child who had a satisfactory postoperative course with heart block with an idioventricular rate between 60 and 90, episodes of tachycardia developed on the ninth postoperative day with resumption of normal conduction, and the outlook was considered excellent for a normal sinus mechanism at the time of the approaching dismissal. The child died suddenly and unexpectedly, presumably of an arrhythmia. This patient was 1 of 2 dying in the late hospital period after episodes of arrhythmia; the second, however, did not have heart block. Heart block has appeared in 1 patient some hours after the surgical procedure and in this case it would be reasonable to attribute the block to edema or hemorrhage in the ventricular septum.

In complete heart block the utilization of the artificial pacemaker* as developed by Dr. Paul Zoll has been a boon to adequate management, and I believe lifesaving in a dramatic way in 2 patients: in 1, an adequate heart beat was maintained continuously for 4 days, and in the other for 8 days (fig. 3). The successful application of this technic lies not only in the excellence of the equipment but in the disciplined devotion to duty of the nurses and the resident surgical staff. The combination of the monitor and the pacemaker is such a valuable apparatus that I do not believe we could do without it. It may be mentioned that the automatic transfer to pacemaker by the monitor is not relied upon completely, as the attending physician is in a better position to utilize the pacemaker if he sees the rhythm mechanism and he may set the amplitude of stimulation and heart rate. If a rapid arrhythmia occurs during heart block, it is thought that the pacemaker’s action would be voided or ineffective, and this is the explanation of the sudden death of an individual having episodes of tachycardia despite being connected to monitor and pacemaker.

Isoproterenol (Isuprel) has been a most valuable drug when the ventricular pacemaker begins to fail; it can be used rectally and sublingually as well as intravenously. In 1 child one quarter of an Isuprel linguet (2.5 mg.) every hour maintained an adequate ventricular

lactate have also been given on a number of occasions, but in the cases in which it was administered no change in the block was seen. Multiple observations on the effect of molar sodium lactate have been impossible because of the belief that an excessive sodium load would be detrimental to the general welfare of these patients shortly after the surgical procedure.

Of incidental interest is the fact that with one probable exception we have not observed any characteristic late postoperative syndromes such as associated pleurisy, pericarditis, and fever, which compose the typical postcommis-suromony syndrome.

**Nature of Postoperative Deaths**

When one of our patients has died after an operation in which extracorporeal circulation has been used, the postmortem examination—at least the discussion concerning claims and conjectures as to cause—persists long after the actual anatomic examination of the body. One must admit that the true mechanism of death is rarely established despite the close co-operative work of our section of pathology, particularly the efforts of Dr. Edwards, and that the valiant efforts of the clinical staff have resulted in no dearth of theories. A few conclusions concerning deaths are in order. In the first place, assuming the role of the sharpest critic, I have been unable to see definite evidence of a death being directly, only, and immediately attributable to perfusion by the artificial pump-oxygenator. In the early cases, deaths were not easily categorized etiologically, but with increasing experience the deaths early in the series can be reviewed and valuable lessons learned. 1. **Pulmonary complications** were distressingly common in the first 30 cases, but have now become relatively uncommon. This has resulted from careful attention to the ventilation of the lungs at operation, avoidance of hypervolemia, and precise surgical procedures. 2. **Apparently “sudden” deaths**, although seemingly mysterious, have been eliminated through the gradual perfection and refinement of technics, the use of excellent perfusion flow, and thus the avoidance of tissue anoxia and metabolic acidosis. 3. **Incomplete repairs**, especially in cases of ventricular septal defect and tetralogy, unquestionably

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**Fig. 3 a.** Electrocardiograms in a 5-year-old girl 8 days after operation for ventricular septal defect and after use of an artificial pacemaker (Zoll) for 7 days. They show dependence of the ventricular beat upon the artificial pacemaker, but increase in rate of the ventricular pacemaker with administration of isoproterenol hydrochloride and the establishment of 3:1 heart block. Note change in QRS configuration from left bundle-branch form to right bundle-branch form when the idioventricular rhythm is replaced by conducted beats. **b.** Diagram illustrating the duration of time patient was maintained on the external pacemaker. At the time the graph ends the patient was maintaining a 2:1 heart block.

rate, that is, 50 to 80 per minute instead of less than 30 during many periods of several hours each.

When a potential catastrophe related to complete heart block is feared, I must admit that one does not wait for the dictates of scientific medicine to be formulated but may change, perhaps illogically, to a “do everything” philosophy, and may give the adrenal corticosteroids. I cannot report any unequivocal value despite the fact that the 2 patients who survived long periods of heart block were given these steroids. Molar solutions of sodium
accounted for some deaths. 4. Hypervolemia was probably a factor in some cases. 5. Heart block has been an infrequent cause of death, but probably representing 8 per cent of deaths in recent months.

In deaths of patients with tetralogy of Fallot, the early convenient explanation for the death and the pulmonary edema often found was inadequate size of the left ventricle, but in the light of our total experience and the smooth postoperative course of some patients with this condition, such an easy explanation is hardly tenable.

The late postoperative deaths have been few but trying. Two patients with tetralogy of Fallot who underwent intracardiac reconstruction died of congestive heart failure after a few months. While postmortem examinations were not performed, it is accepted as most probable that these 2 patients, without cyanosis postoperatively, died because of an inadequately relieved pulmonary stenosis in the face of complete repair of the ventricular septal defect. No cases of this type have occurred recently, which is compatible with improvements in technics of relieving pulmonary stenosis to the extent of using a doubly pointed ovoid expansion patch on the outflow tract of the right ventricle and root of the pulmonary artery in some cases. Pulmonary valvular incompetence, if present after operation, was not expected to be a problem and so far, practically speaking, it has not proved to be. A few patients have shown mild transient signs of heart failure after repair of a simple ventricular septal defect. One case in which such failure was most perceptible was that of a child who had intermittent heart block following operation and from whom digitalis had been withheld. One adult patient with severe pulmonary stenosis and heart failure had recurrent thromboembolic complications and died 3½ months after operation from these complications.

In the period from last November to January we witnessed the distressing occurrence of bacterial endocarditis in 4 patients, with Pseudomonas as the infecting bacterium, which was resistant to the usual antibiotics. Polymyxin was not of definite value. Two patients died and a third has probably died by this time. Because of these, the sterilizing technics for the pump-oxygenator were altered to what would seem to be the most thorough methods possible, and it is hoped that this problem has been eliminated.

Mortality

In a field of endeavor changing as rapidly as that of direct-vision cardiac surgery and in view of the numerous problems presented by the variations in the defects attacked, mortality rates for the heterogeneous total group mean little. At present, the mortality rate from utilization of the pump-oxygenator to supply the body with a normal systemic flow up to an hour under conditions of bilateral thoracotomy can be assumed to be less than 5 per cent, I believe.

The most homogeneous group of cases are patients with a ventricular septal defect and a large pulmonary flow, and in this group, excluding infants, the mortality rate has been approximately 15 per cent. In the infants operated upon for ventricular septal defect, the mortality rate has been much higher in the past than at present. Although these were all very sick youngsters, the reason for advising operation at this age, the mortality rate was great enough in the fall of 1956 to cause us to revert to the “banding” operation on the pulmonary artery in these very small patients. Refinements in surgical and perfusion technics have resulted in the resumption of reparative operations in even these small infants, with a decline in mortality rate that is gratifying. To this date nearly 100 patients with ventricular septal defect have been operated upon.

In the group of patients with tetralogy of Fallot there is even greater lack of homogeneity in the clinical and laboratory picture, and the initial experience with surgical treatment was discouraging, but the mortality rate for a group of 38 patients operated on in 1956 and to May 1, 1957, has been 26 per cent. Of particular interest is 1 adult patient with pulmonary stenosis and intact septum who came with class IV heart failure, apparently in his last days of life, phenomenally having no murmur at times, with a cardiac index of 1.6 L. and a pulmonary valvular maximal gradient of 200 mm. Hg. He responded to medical therapy sufficiently to allow surgical treatment. With the use of the
pump-oxygenator to bypass the heart, the pulmonary valve was opened and the right ventricular pressure reduced to 40 mm. Hg. Remarkable improvement occurred.

**Criteria for Cure**

The majority of the patients who have survived operation are believed to be cured of their primary defects, and confidence in this regard is greater in the later patients than in the first dozen or so that were treated. We have not considered it justifiable to recatheterize routinely the hearts of many children during and after their convalescence from operation, but eventually over the years the plans are to collect the data such a procedure provides.

Evidence for the cure of the defect may be submitted as follows:

**Direct.** 1. The surgeon's critical appraisal of his repair following its conclusion. This has had more value since asystole has been used. 2. The pathologist's appraisal of the repair in the patients who have not survived. As nearly all the deaths occur early, it is difficult to assess what might have developed in the area of the healed defect, but this consideration aside, the results are anatomically good. In the early experience, 1 or 2 patients had multiple defects of the ventricular septum, and the surgeon had apparently overlooked 1 of them. Dr. Kirklin believes it fair to say that in the "beating heart era" about 15 to 20 per cent of the patients had repairs that were not anatomically perfect; with asystole a complete repair is nearly always possible in the hands of an experienced surgeon.

3. Normal dye curves and decreases in pulmonary artery pressure obtained in the operating room.

**Indirect (Clinical).** 1. Improvement in the child's strength, weight, and tolerance to exercise. 2. Absence of cyanosis or hypoxemia under conditions of exercise. 3. Decrease in heart size (and, in septal defects, the pulmonary vascular shadows).

Some of the children have a persistent systolic murmur after convalescence from repair of a ventricular septal defect, and many patients who have had surgical treatment for pulmonary stenosis have a systolic murmur and occasionally a soft diastolic one. It is a dramatic auscultatory effect when murmurs are absent after operation, but the persistence of a murmur of mild intensity is not considered as definite evidence of a persistent septal defect. In many of the patients who had severe pulmonary hypertension the second sound in the pulmonary area remains accentuated following repair of their ventricular septal defects, but this is hard or impossible to evaluate quantitatively as a sign of persistent pulmonary hypertension.

In an infant, previously thin and disabled, ability to exercise normally, gain in weight, and replacement of tumultuous heart action and murmur by a quiet heart action are evidence of clinical cure. In a few it has been proved by catheterization that the hemodynamics are also normal (table 1). However, it will take some years to establish the degree of regression of the pulmonary vascular resistance in those patients in whom it was markedly elevated pre-operatively and to establish the preoperative clinical and hemodynamic profiles of those patients who do, and those who do not, have a "complete cure" after closure of the intracardiac defect.

The cardiac defects that have been cured by direct-vision intracardiac operations utilizing an extracorporeal pump-oxygenator include (1) ventricular septal defect, (2) atrial septal defect, (3) common atroventricular canal (when mitral valve is adequate), (4) anomalous pulmonary venous connection with atypical atrial septal defect (superior vena cava syndrome), (5) complete anomalous pulmonary venous connection, (6) pulmonary stenosis—infundibular or valvular with intact ventricular septum or both, (7) tetralogy of Fallot, cyanotic and noncyanotic, (8) aortic sinus aneu-

### Table 1.—Repair of Ventricular Septal Defect on August 26, 1955, in a Patient, Age 6 Years

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<td><strong>Pulmonary pressure, mm. Hg.</strong></td>
<td>109/74</td>
<td>33/14</td>
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<td><strong>Flow, L./min.</strong></td>
<td>5.4</td>
<td>2.3</td>
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<tr>
<td><strong>Systemic pressure, mm. Hg.</strong></td>
<td>122/67</td>
<td>113/87</td>
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<tr>
<td><strong>Flow, L./min.</strong></td>
<td>—</td>
<td>2.3</td>
</tr>
<tr>
<td><strong>O₂ saturation, %</strong></td>
<td>92</td>
<td>97</td>
</tr>
<tr>
<td><strong>Shunts, left-to-right</strong></td>
<td>Large</td>
<td>0</td>
</tr>
<tr>
<td><strong>Right-to-left</strong></td>
<td>Small</td>
<td>0</td>
</tr>
</tbody>
</table>
rysm with rupture, with and without ventricular septal defect, (9) congenital aortic stenosis, and (10) combined subaortic and subpulmonary valvular stenosis.

SUMMARY

The general problems of direct-vision intracardiac surgery utilizing the extracorporeal pump-oxygenator from the viewpoint of the cardiologist are those of proper selection of patients and the care of the patient in the postoperative state. The surgeons have advanced their techniques until there are few remaining lesions that may not be regarded as eventually operable. The techniques of extracorporeal circulation may be regarded as established and no longer on trial. The apparatus patterned after the pump-oxygenator designed for Dr. Gibbon has established its worth, adaptability, and reliability in our clinic, and there is no reason to change to any other type at this time. Potassium asystole has worked out exceptionally well.

The mortality rate for surgical procedures in young infants has been distressingly high, but the general condition of these babies has been critical even before operation. It may be emphasized that there is reason to believe that rapid progress is being made both in regard to reduction in mortality and in regard to the certainty with which the defects are being adequately repaired. Complete heart block occasionally occurs, and medically we may not have adequate means to maintain an adequate ventricular rate. The artificial pacemaker of Zoll has been of great assistance in maintaining an adequate ventricular rate and life in some patients.

The problem of pulmonary vascular disease remains one of the most important, and many aspects of it are puzzling. The degree of regression of the obstructive changes in the pulmonary vascular system in surgically treated patients with pulmonary hypertension awaits some years of study, but there are promising signs of involution in some cases.

SUMMARIO IN INTERLINGUA

Ab le puncto de vista del cardiologo, le problemas general del chirurgia intracardiac a vision directe con le utilisation de un pumpa-oxygenator extra le corpore concern le correcte selection del patientes e lor curatura durante le periodo postoperatori. Le chirurgos ha perfectionate lor technicas usque a un puncto ubi pauc lesiones remane que non pote esser considerate como potentialmente operabile. Le technicas del circulation extracorporee pote esser reguardate como establite e ultra lor stadio experimental. In nostre clinica, un apparatura construite secundo le modello disveloppate sub le direction de Dr. Gibbon ha demonstrate su valor, adaptabilitate, e regularitate de servizio, e il existe a iste tempore nulle ration pro transiri a un altere typo. Le asystole a kalium ha functionate exceptionally ben.

Le mortalitate in interventiones chirurgic in juvene infantes ha essite discoragiantemente alte, sed le condition general de iste babys esseva critic mesmo ante le operation. Il es a signalar que nos ha rationes a creder que rapidi avantiamentos es in progresso si ben con respecto al reduction del mortalitate como etiam con respecto al grado de certitude que on possede de haber reparate le defectos adequatamente. Il occurre casos de complete bloco cardiae, e il es probablemente ver que como medicos nos non ha medios adequate pro mantenir un satisfacente rapiditate ventricular. Le pacemaker artificial de Zoll ha essite de grande valor in mantenir un adequate rapiditate ventricular e assi le vita de cete patientes.

Le problema del morbo pulmono-vascular remane un del plus importante, e multes de su aspectos es enigmatic. Le grado de regression del alterationes obstructive in le systema pulmono-vascular in chirurgicament tractate patientes con hypertension pulmonar attende annos de studio, sed in cete cases il ha signos promittente de involution.

REFERENCES

**Medical Eponyms**

*By Robert W. Buck, M.D.*


“I have observed that it is possible to see the movements of the diaphragm...on the thorax in all healthy men. The phenomenon takes the form of a wave motion which, beginning on both sides at the height of the sixth intercostal space, travels downward with maximum inspiration in the form of a straight line or shallow furrow (which makes an acute angle with the ribs) over several intercostal spaces, at times as far as the costal margin. With expiration it rises again over the same area.”

A more detailed account of the phenomenon is given in the paper “The Diaphragm Phenomenon and Its Significance from the Physiological and Clinical Standpoint” (Das “Zwerchfellphänomen” und seine Bedeutung von physiologischen und klinischen Standpunkte aus) which appears in the Verhandlungen des Congresses fur Innere Medicin 13: 309–319, Wiesbaden, 1895.

“The patient to be studied is placed in a horizontal position...with his feet toward the window, while the examiner, standing three or four steps away with his back toward the window, observes him from an angle of about forty-five degrees...We are convinced that in a dark room the phenomenon is only visible if a source of light is so placed that the beams strike the thorax from the direction of the feet at an acute angle with the thorax.”
Clinical Problems Related to Surgical Repair of Intracardiac Defects with the Aid of an Extracorporeal Pump-Oxygenator

HOWARD B. BURCHELL

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