PRESIDENTIAL ADDRESS

THORACIC SURGERY FOR INFANTS

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The Constitution of the Association does not in any way suggest, or limit, the subject matter to be presented by a President at the annual meeting. It has been my choice to dwell upon a topic which long has been of personal interest, and I hope to emphasize that a field—believed by some to be limited—is in reality one of very wide scope and one which should attract the attention of all who would like to extend the borders of thoracic surgery. Within the rather short existence of this Association, measured in only a few decades, chest surgery has made enormous strides in many directions. These are accomplishments of which you as a group can justly be proud because most of the advances have come from men who meet here today. What I want to point out is the need to lower the sites regarding age and size of patients so that your skills can be brought to a large group of little citizens who need your assistance.

Rather than present this subject in a general sort of way, I have chosen to pull out the records and summarize what has happened on our own service. This gives the opportunity to make for myself a score sheet, to see what is good and what needs improvement. It also gives you a descriptive cross-section of the kind of material which is probably coming into community hospitals everywhere. The vast majority of the conditions encountered are emergency problems, and hence the patients are generally not sent far off to some specialized institution. Instead, they usually remain near their homes, and surgical treatment for them must be on or near that base.

In summing up thoracic operations in our hospital for babies under a year of age, I soon found that I had bitten off more than I could chew because the list includes over 1,200 cases. A few of them extend back for as long as I have been in the practice of surgery, but most have appeared in recent years. I am denied the possibility of presenting any part in detail. Time permits little more than giving an idea of the frequency of appearance of the different conditions and making a brief comment or two on each. At once I am conscious of the fact that in most of these divisions there are, in this assembly, men who can show better survival statistics than mine.

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THORACIC TUMORS, CYSTS, AND DUPLICATIONS

Nearly a dozen kinds of lesions can be grouped together under the above heading—none very common—but, taken in their entirety, they form a group of some size and are of considerable interest. Summed up in Table I are the records of 43 infants with primary neoplasms or cystic conditions within the chest. These were lesions of variable size, situated in diverse parts of the thoracic cage, and presented widely different problems in their treatment. To view the entire picture, there was one death from cardiac tumor, two from lung cysts, and two from esophageal duplication. Of 43 babies, 38 have survived and are apparently cured of their abnormality or neoplasm. Some remarks regarding the most important of these are in order.

TABLE I. DISTRIBUTION OF THORACIC OPERATIONS IN INFANTS FOR VARIOUS TUMORS AND CYSTS

2	Hamartoma of lung Hygroma	•
		ฮ
2	Bronchogenic cyst	1
1	Cyst of lung	10
2		10
1	<u></u>	
	2 1 2 1	2 Bronchogenic cyst 1 Cyst of lung 2 Duplication 1 Total-43

Neuroblastoma arising in the thorax is always situated along a paravertebral gutter and arises from some part of the sympathetic chain, bulges outward and forward, impinges on the lung, is broadly attached over bodies of the spine, and infiltrates about the necks of adjacent ribs. No effort should be made to remove all the neoplasm surgically because this implies some damage to vertebral bodies and certainly to the removal of one or two necks of ribs. Such a step can cure tumor but is too apt to produce frightful deformity of spine and thorax during subsequent years of growth. It is much better to cut away that part of tumor which can be comfortably excised and follow this with light x-ray irradiation. Of 8 babies so treated, all have been cured (followed for 7 or more years).

Teratomas of the mediastinum might seem to be forbiddingly large, yet excision is generally not difficult.

Lung cysts can have air trapped within them, build up a considerable pressure therein, and can cause respiratory distress. In some instances a cyst can be removed cleanly; more often a lobe (or even a lung) encasing it must be sacrificed. I have had particular interest in one child from whom the entire left lung was removed 20 years ago; the chest (free from infection or scarring of the thoracic wall) has grown in a symmetrical fashion without scoliosis.

Duplications can occur adjacent to any part of the alimentary tract. We have encountered more than 120 of these, ten of which were in relation to the esophagus (Fig. I). They are cystic structures having smooth muscle coats and some sort of mucosal lining, very often gastric in type. Some communicate with the esophagus, most do not. They should never be drained or marsupialized;

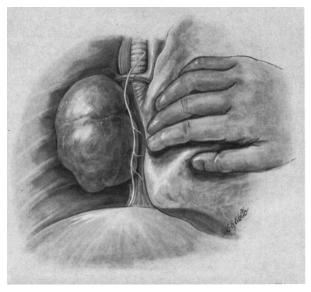


Fig. 1.-Typical duplication of the esophagus in an infant.

it is far better to excise them entirely. They are always attached to the esophageal wall but generally can be cut away without penetrating the esophageal mucosa.

INFANTILE LOBAR EMPHYSEMA

Bronchial obstruction can cause trapping of air in a portion of pulmonary tissue with a build-up in pressure within it so that the mediastinum is pushed to the opposite side, severely compressing all the remaining lung substance (on the same and opposite sides), producing marked respiratory distress, and even sudden death (Fig. 2). Such bronchial obstruction may be from a variety of causes, but most often there is local deficiency of cartilaginous tissue which allows the bronchus to become compressed, angulated, or otherwise blocked. Any lobe can be the seat of this anomaly; the left upper and the right middle lobes are the most frequently involved (Table II). In general, the babies with the more severe forms of the condition are seen early after birth, and almost invariably come to operation within a few weeks thereafter.

Many young infants with rapidly advancing lobar emphysema present

TABLE II. LOCALIZATION OF INVOLVEMENT OF LOBES BY TENSION EMPHYSEMA (THE LEFT UPPER AND RIGHT MIDDLE LOBES ARE THE MOST COMMON SITES)

RIGHT		LEFT	1
Upper Middle	4	Upper	12
	10		
Lower	0	Lower	1

such a desperate emergency that any delay in operation is dangerous. The mortality of "expectant" treatment is over 60 per cent. Needle aspiration has little to recommend it; indeed, it is apt to be followed by troublesome pneumothorax which complicates the picture. Needle asperation, however, has some place as an emergency measure while the operating room is being readied and, especially, to improve matters temporarily during induction of anesthesia. Immediate lobectomy carries a risk which is virtually zero. When the chest is opened, the ballooned lobe literally pops out of the wound. Lobectomy can be performed quickly and easily because there is rarely infection or thickening around the hilum. Twenty-one babies have been so handled and all have survived.

COMPRESSION OF TRACHEA OR ESOPHAGUS BY AN ANOMALOUS VESSEL OR STRUCTURE IN THE MEDIASTINUM

The fact that 129 babies have come to surgery on our service because of obstruction of the trachea or esophagus (or both) emphasizes that these conditions are by no means rare.

Double Aortic Arch.—The most common form of anomaly to be considered is the double arch, the two limbs of which surround and compress the esophagus and the trachea. Various techniques of roentgenographic investigation, particularly those during which the esophagus and trachea are visualized by opaque materials in their lumina, can identify with a high degree of accuracy the constricting vascular structures. At operation, it is usually the anterior arch which is the smaller of the two and should be divided (Fig. 3); less frequently the posterior limb needs sectioning. It is important to stress that all bands and

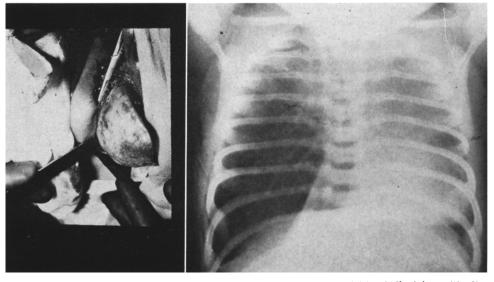


Fig. 2.—Right, Roentgenogram of markedly emphysematous right middle lobe, with displacement of heart to left. Left, Photograph made at operation; the emphysematous lobe protrudes markedly through the operative wound.

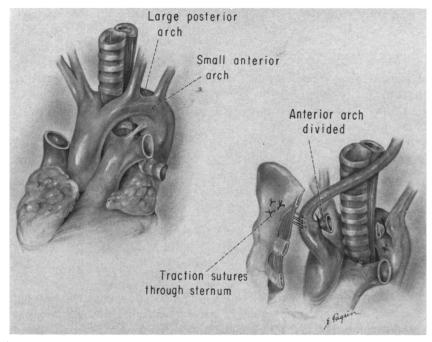


Fig. 3.—Typical double aortic arch, with smaller anterior aortic limb, treated by surgical division of the anterior arch.

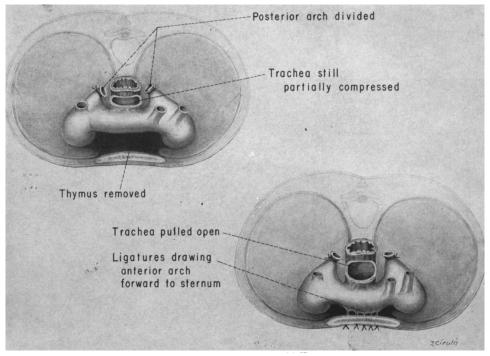


Fig. 4.—Surgical principle of making appropriate opening of the trachea. In this case, the posterior limb of a double aortic arch has been divided. Additional relief of the trachea can be gained by drawing forward the anterior arch.

fasciae which accompany such vessels must likewise be divided to remove all of the constricting mechanism. Further, it is an important additional step (Fig. 4) always to drag forward the left common artery (when an anterior limb has been sectioned) or the anterior aortic limb (when the back one has been cut), and to tack these forward by silk sutures which pierce the sternum. This additional step *pulls* the anterior wall of the trachea forward and thus enlarges its lumen.

Right Arch—Left Ligament.—Another form of constricting mechanism is that in which there is a right aortic arch and a left ligamentum arteriosum (Fig. 5). Division of the ligament (and sometimes the first part of the left subclavian artery) provides sufficient room for the trachea and esophagus.

Anomalous Innominate Artery.—In another small group, we have seen the innominate artery arise from the distal part of the aortic arch or the left common carotid artery arise quite proximally from the aortic arch. In either instance, the ascending vessel lies over and markedly compresses the trachea (there being no interference with the esophagus). It is easy to handle these anomalies by removing the thymus by drawing the offending vessel forward and tacking it to the back of the sternum.

Aberrant Subclavian Artery.—This is very common but generally is symptom free. However, in some cases a large or taut vessel can give dysphagia lusoria. Division of the artery is easily performed.

Tracheomalacia.—Right or wrong, I add here a group of 15 babies who had considerable respiratory distress and were shown by roentgen examination

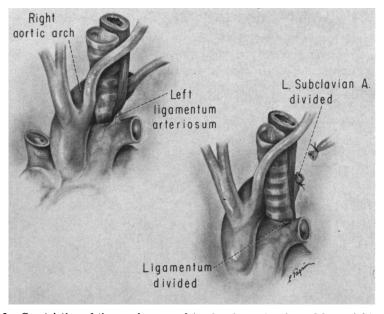


Fig. 5.—Constriction of the esophagus and trachea by a ring formed by a right aortic arch and left ligamentum arteriosum. Surgical management by division of the ligament and first part of the left subclavian artery.

and tracheoscopy to have a soft, collapsing trachea; they had only a narrow transverse slit during some part of the respiratory cycle, particularly in the expiratory phase. There has been no offending external constricting vessel, but we have found at mediastinal exploration what was thought to be a somewhat enlarged thymus and what was certainly a very soft tracheal wall. Considerable relief can be given by removing the thymus and then drawing the aortic arch forward and tacking it to the back of the sternum. The aortic arch (having been left attached to the trachea) will draw the anterior wall of this forward, thus opening up its lumen and giving a better airway.

Table III sums up our surgical experience with 129 babies having the listed kinds of anomalies. There have been six deaths in the double arch group, 1 in the right arch group, and 2 in the last group. In the last 8 years there have been 43 babies operated upon in these various classes, with only one fatality.

TABLE III. PATHOLOGIC CONDITION TREATED IN 129 BABIES OPERATED UPON FOR TRACHEAL OR ESOPHAGEAL COMPRESSION

Double aortic arch	52
Right arch, left ligament	32
Anomalous position of Innominate or left common carotid artery	16
Aberrant subclavian artery	14
Enlarged thymus; tracheomalacia?	15
Total	129

PATENT DUCTUS ARTERIOSUS

A subject so prosaic as patent ductus arteriosus needs little more than passing comment. In general, surgical closure of a ductus is something we all regard with equanimity. In our own series of more than 1,400 operations, patients beyond the first year have carried a risk of only a fraction of one per cent. In contrast, ductal closure in infancy can at times be quite risky because the baby who comes to operation has usually been in failure or on the border of it.

Not infrequently, one is faced with shunts at both ductal and ventricular levels and it might be difficult to decide which is the more disturbing to the baby. The best handling of such a child, who is generally quite ill, is to approach the ductus quickly and close it, and, if there appears to remain a high pulmonary flow from a coexistent ventricular defect, then simultaneously band the pulmonary artery.

From the technical point of view, the thing that has usually bothered me most has been the presence of a very engorged lung which is difficult to get out of the way in order to gain adequate exposure of the mediastinum. This difficulty can be diminished by having the baby turned well up on its side and making the intercostal incision very long and also far laterally. This gives more room to tuck the lung down out of the way.



Fig. 6.—Baby, weighing 4 pounds, 9 ounces, who had been in failure from a large patent ductus arteriosus. Successful treatment by surgical closure of the ductus.

In a series of 119 operations on babies (65 of whom were under 6 months of age), there have been thirteen deaths and 106 survivors. In virtually all of the fatal cases, there were other serious anomalies. The rewards of this operation are great because so many of these youngsters can be instantly and successfully relieved of a severe cardiac overload.

COARCTATION OF THE AORTA

Surgical therapy for coarctation of the aorta in children and adults is widely practiced and is on a sound basis. In our series of nearly 800 operations, the mortality rate for patients over one year of age is between 3 and 4 per cent. Resection of coarctation in babies has been quite a different story.

Whereas most infants do not need surgical correction of their coarctation in the first year of life, there is a definite group who are critically ill and who get little or no relief from medical management. These very often have some complex cardiac or cardiovascular abnormality of a serious form. In the twenty-one babies we have operated upon, four had no other lesion, but ten had a large accompanying ductus, seven a ventricular septal defect, five mitral insufficiency, three aortic stenosis, and one had infundibular stenosis. Such findings explain most of the published surgical fatality rates which range from 30 to 50 per cent. In our own material, 21 babies were operated upon (15 being under 10 weeks of age) with a loss of 40 per cent. Such figures might bring discouragement, but on the brighter side is the fact that probably all the survivors would have died without the surgical intervention.

When operation is required, resection and end-to-end union is the operation of choice (Fig. 7) but, under some circumstances, it is perferable to enlarge the aortic lumen by inlay of a gusset, as employed by Senning, De Bakey, King, and others.

It is well to bear in mind the suggestion of Malm that, after repairing aortic obstruction, any persistent pulmonary hypertension (from a ventricular septal defect) can be improved by simultaneous banding of the pulmonary artery.

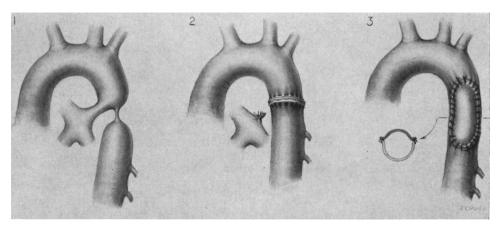


Fig. 7.—Coarctation of the aorta in infancy. 1, This is found very often in combination with a large proximal ductus. 2, Preferred method of treatment by resection and primary anastomosis. 3, Patch repair and enlargement of aortic pathway, preferable under some circumstances.

CONGENITAL DIAPHRAGMATIC HERNIA

Discussion here will be limited to the common, more serious, Bochdalek type of diaphragmatic hernia, and will omit defects at the foramen of Morgagni and those of a hiatal nature.

In 1925, Hedblom reviewed the literature and noted that the general consensus over the United States was that babies with diaphragmatic hernias should not be operated upon. With this "conservative therapy," 75 per cent of patients died before the end of the first month. Shortly before the second World War, the success attending surgical repair of traumatic rupture of the diaphragm in adults influenced surgeons to attempt the correction of the congenital form of diaphragmatic defect. Dr. William Ladd was a leader in this work and, as his assistant, it was a thrilling experience for me to see a small, blue, gasping baby literally pulled from the jaws of death and quickly made well by a dramatic surgical maneuver.

Certainly everyone now agrees that infants with diaphragmatic hernias should be operated upon, and this should be done promptly. While some still advocate a thoracic exposure, I have always believed that an abdominal approach is generally far better because there are often anomalies of intestinal malrota-

tion which have to be currently treated. Also, in some babies the viscera cannot be crammed back into the small abdominal cavity and it may be necessary to employ temporary relief for some days by merely letting intestines extrude through an abdominal decompression, covered only by skin flaps.

It is essential at operation to keep the anesthesiologist from attempting inflation of a collapsed lung by positive pressure because of the danger of producing blebs on the lung surface which can rupture at this time or within the subsequent day or two. We have had several tragic losses of this sort. At no time should the intrapulmonary pressure be raised above 15 cm. of water. It is far safer to gently and slowly unfold and expand the lung by providing a negative pressure in the pleural space. We have come to feel that a small tube should always be left in the pleural cavity postoperatively and that very gentle constant suction (10 to 15 cm. of water vacuum) should be applied to it. Expansion of the lung can in many instances be accomplished in a few hours, but in others it might require several days.

As we look back over our material, it is disturbing to find that surgical cure rates are falling off. As shown in Table IV, the recovery rate in 65 cases (1940 through 1950) was 87 per cent whereas in 71 cases from 1951 through 1963 the rate was only 68 per cent. Restudy of records (Table V) now shows important differences in the two groups with reference to the ages of the babies. In the material of the forties, 91 per cent were over 24 hours old whereas 9 per cent were under 24 hours of age. In marked contrast, from 1951 through 1963, babies under 24 hours of age had risen to 52 per cent of the series. Apparently, in former times, the most seriously ill infants were dying on the first day of life and were not being transferred to a surgical service. In contrast, the present alertness of physicians now brings to us a much higher proportion of subjects on the first day after delivery. Obviously, any improvement in this

Table IV. Congenital Diaphragmatic Hernia (Comparison of Surgical Recovery Rates of 1940-1950 With Those of 1951 Through 1963)

1940 through 1950	
Cases	65
Alive	55
$\operatorname{Recovery}$	87%
1951 through 1963	
Cases	71
Alive	48
Recovery	68%

TABLE V. STATISTICS—DIAPHRAGMATIC HERNIA*

	OVER 24 HR. OLD		LESS THAN 24	4 HR. OLD
	% of series	ALIVE	% of series	ALIVE
1941 through 1950	91	91%	9	50%
1951 through 1963	48	100%	52	47%

^{*}Diaphragmatic hernia. Comparison of material of 1940-1950 with that of 1951-1963. In the more recent series there is a very much higher proportion of babies under 24 hours of age, which apparently accounts for the higher surgical death rates in the recent series, as shown in Table IV.

branch of surgery must come from closer attention to the desperately ill infants who are less than 24 hours old and who are now coming to us in greater numbers.

It is obvious that some of these patients, only a few hours old, are going to die because of other coexisting anomalies or because of severe anoxia and irreparable brain damage. However, some can probably be salvaged. I would like to give an example of what can be accomplished (Fig. 8). A very ill child was referred at 12 hours of age literally gasping for breath because of a large left diaphragmatic hernia. He was immediately turned with the left side down, a nasogastric tube for deflation of the stomach was inserted, and the baby was placed in a closed box with 95 per cent oxygen while the operating room was being set up. Through an abdominal approach, the displaced viscera were withdrawn from the chest and pulled out through the abdominal wound. The baby's condition promptly improved and the diaphragm was repaired. It was now impossible to get all the intestines back into the abdomen; therefore only skin was drawn over the bulging intestinal mass. Five days later, the cutaneous wound was reopened and the abdominal cavity now was found to be stretched enough to receive all its viscera. Complete repair of the peritoneum, abdominal wall muscles, and fasciae was now possible. I am sure that, with a little ingenuity and careful attention, many of the seriously ill babies who have been dying on the first day of life can be saved.

HIATUS HERNIA

About one sixth of all congenital diaphragmatic defects are of the esophageal hiatus hernia type. Of these, only 3 or 4 per cent are of the paraesophageal form; the remainder are of the sliding variety. Most babies with hiatal weakening will vomit, which generally begins in the early weeks of life. About one half of them have hematemesis, and a fourth of them have some degree of anemia. Malnutrition is common. Aspiration pneumonia occurs in some. All patients have gastroesophageal reflux and a few have varying degrees of esophagitis; actual stricture of the esophagus is seen only rarely. Poor gastric empyting is common. About two thirds of the patients we have had were below the third percentile in weight.

In most of these babies, the hernia allows 2 to 4 cm. of stomach to ride up above the level of the diaphragm but we have seen as much as 90 per cent of the stomach in the thorax. Oddly enough, the *size* of the hernia has no definite relationship to the *severity* of symptoms; indeed, some of our youngsters with the most marked symptoms have had hernias which extended only 1 cm. or so above the diaphragm.

The finding of an esophageal hiatus hernia in an infant does not necessarily imply a need for operation. Conservative measures, largely concerned with maintaining a propped-up position for some months, will often give good relief of symptoms and sometimes disappearance of smaller hernias. But when there is a large hernia, chronic bleeding, or failure of medical management, operation has much to offer. We prefer to make the reconstruction transabdominally. The repair is essentially that described by Allison. After incision of the hernial sac



Fig. 8.—Two-stage repair of diaphragmatic hernia in a 7-pound baby. Transabdominal correction of diaphragm; since the abdomen was not large enough to receive all the viscera, loops of intestine were allowed to protrude through the abdominal wound and were covered only by skin. Five days later, the wound was reopened, the abdominal cavity now was found to be large enough to receive all the intestines which permitted complete closure of all layers of the abdominal wall.

and cutting of the phrenoesophageal ligaments, the stomach and esophagus are freed from the surrounding mediastinal bed and drawn downward, after which the widened hiatus is narrowed appropriately, posterior to the esophagus. It is well to sew the lower part of the esophagus up against the gastric fundus to give an acute angle and create a better pinchcock mechanism at the cardia. Furthermore, the fundus should be anchored to the undersurface of the diaphragm. We believe it best in all cases to perform a pyloroplasty to insure good gastric emptying.

In our series of 30 patients, 20 were treated by medical means, 13 cases were regarded as successes and 6 as failures (one death). Surgical repair was carried out in 15 (4 of whom had esophageal stricture which required concomitant therapy). All had successful corrections. Severe decline in nutritional status before operation is generally followed by a marked improvement in weight following surgical therapy.

ANOMALOUS LEFT CORONARY ARTERY ARISING FROM THE PULMONARY ARTERY

Origin of the left coronary artery from the pulmonary artery carries a high fatality rate in babies. As suggested by Brooks, and emphasized by Edwards, this vessel might not receive blood from the pulmonary system but instead can act as a drain-off from the myocardium into the pulmonary artery. Such pathologic physiology is well illustrated by sketches in Sabiston's article of 1960. The concept gave logical basis for surgical ligation of an anomalous coronary, first successfully performed by Sabiston, which stops the flow from the myocardium back into the pulmonary artery and thus gives better perfusion of the heart muscle from the other coronary artery.

This therapeutic advance has led many to believe that a coronary arising from the pulmonary artery always has a reversal of flow in it and always should be tied. It is now evident that ligature should not universally be used; indeed in some cases it should be definitely avoided. There are really two forms of abnormal coronary blood flow (Fig. 9), sometimes identifiable before operation but certainly always discernible at the operating table. In one, the right and left coronary systems have many intercommunications within the musculature of the heart; here ligation of the anomalous vessel prevents blood from seeping back into the pulmonary system and thus improves the myocardial supply. In the second, there is little or no intercommunication between the two coronary systems; visible, extensive myocardial infarction and a bluishappearing first part of the left coronary artery suggest that the flow is coming from the pulmonary artery. This can be rapidly confirmed by taking samples from the coronary and the pulmonary arteries, which shows that they have the same dark color and low oxygen content. With these findings, ligation of the anomalous coronary is a fatal step because it cuts off the only vascular supply to the regional heart muscle. Here it is far better to band and constrict the distal part of the pulmonary artery, thus forcing more blood into the right coronary, even though this carries blood of low oxygen content to the myocardium—a situation we know gives reasonable myocardial nourishment in cyanotic states such as the tetralogy of Fallot. If the child survives this initial step, possibly the anomalous coronary artery could subsequently be transferred to the aorta by some procedure such as that suggested by Shumacker.

VENTRICULAR SEPTAL DEFECTS

For children beyond a few years of age, there is universal agreement regarding the desirability and efficacy of open repair of ventricular septal defects, first brilliantly performed by Lillehei in 1955. For babies, employment of cardio-pulmonary bypass to close ventricular septal defects has been successfully performed by Kirklin, Cooley, Sloan, Sauvage, and a few others, but the mortality rate is still considerable. For the infant who demands some immediate surgical relief, there is widespread adoption of the principle, initiated by Muller and Dammann, of protecting the lungs by constricting the pulmonary artery. This operation is indicated in a child who is not thriving properly in the

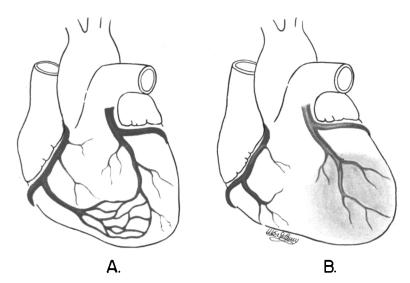


Fig. 9.—Forms of coronary circulation, in the presence of a left coronary artery arising from the pulmonary artery. A, Intercommunications between the two coronary systems which permit arterialized blood to flow back into the pulmonary artery. B, No intercommunications between the two coronary systems; the left ventricle receives bluish blood from the pulmonary artery.

artery. With the anatomical arrangement in A, ligation of the left coronary artery at the pulmonary artery provides improved perfusion of the myocardium. With the arrangement in B, ligation of the left coronary artery should not be done.

presence of a ventricular septal defect and in whom cardiac failure is manifest by pulmonary congestion. There should be included also those babies who have cardiac enlargement which is producing at electasis of the left lung.

It is of considerable importance always to have catheterization studies before banding a pulmonary artery: first, to make sure that there is an interventricular communication, and, second, to determine accurately the degree of pulmonary vascular resistance. We have had a fatal outcome in two babies in whom banding was done and insufficient attention had been paid to the presence of a high pulmonary vascular resistance. The operative step diminished the thrust into the pulmonary bed and an insufficient pulmonary flow led to rapid death. The finding of a high vascular resistance contraindicates the use of this operation.

Reports of this operation give testimony to its effectiveness as a palliative and lifesaving measure—a step to use with the thought in mind that intracardiac repair can be undertaken some years later. In 1961, Morrow and Brunwald reported the cases of 11 infants under a year of age who had had the pulmonary artery banded; there were 10 survivors. In a series of 38, reported by Craig and Sirak, there were 10 patients under a year of age at the time of operation with but one death. In our own material there have been 25 babies with a loss of 6 at or immediately following operation. Dammann and Muller in 1961 reviewed 63 patients of their series and presented late studies to show that pulmonary vascular disease can be reversible in some cases.

AORTIC STENOSIS

Marked aortic stenosis, severe enough to give devastating symptoms in the early months of life, is a lesion which is almost always fatal if untreated. Attempts at surgical relief of this advanced form of obstruction have been very dismal in outlook for two reasons: First, the small size of the thickened and deformed valve gives little hope of making a competent structure, even though the surgeon is lucky enough to open it up somewhat. Second, these babies are in a desperate state and will stand little in the way of operative manipulation. Most of them do not survive the operative procedure in which the surgeon opens the ascending aorta, looks down upon and works on a constricted valve in a very limited time.

Our attempts in 12 young patients led to only two recoveries. In these there was mild hypothermia, inflow occlusion, aortotomy, and some poor stabs at opening up the valve under direct vision. Deaths came on the table or within a few hours thereafter.

Our greatest hope comes from a case, data of which are shown in Fig. 10. The baby was 7 days of age and in severe left-sided failure. He was placed in a compression chamber under 3 atmospheres pressure, which raised the arterial PO_2 from 50 to 1,550 in a few minutes; this allowed a suitably long period of inflow occlusion so that there was a more reasonable length of time for the aortotomy and work on the valve. This child recovered and did amazingly well. Where facilities of a compression chamber are available, there is little doubt that this technique offers the best chance of recovery.

PULMONARY STENOSIS

While pulmonary stenosis in older subjects can be handled very well by open approach to the valve, babies with high degrees of obstruction can be so critically ill that they will not tolerate such extensive procedures. Indeed, many will quickly develop cardiac dilatation and arrest as soon as the limiting cardiac envelope is opened. Hence it is generally necessary to employ rapidly some relief of a Brock type.

For the valvulotomy, we have had the best luck with a No. 13 blunt needle or trocar, in which is carried a small iridectomy knife. When introduced into the right ventricle, the blunt needle (carrying within it the knife) can be positioned up against the dome-shaped obstructed valve, and then the knife is pushed upward and through the valve. After the heart action has improved for a few minutes, larger instruments, such as a Pott's knife or dilator, can be inserted and used for more adequate opening of the valve structure.

Of 19 babies in whom we have made this attempt, 13 have survived. Others have had much better figures—there comes to mind the series of Mustard who had only 3 deaths in 17 cases.

PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM

Complete block at the pulmonary valve, with an intact ventricular septum, is rare. The right ventricle is apt to be rather small, in contrast to the enlarged

size seen with pulmonary stenosis. These babies are living because of an open ductus, and, as soon as it closes, death comes rapidly. Keith found that one quarter of such babies were dead by the end of the first week of life and about three quarters by 6 months.

Prior to 1962 we attempted operation in 10 of these infants; most were less than 2 weeks of age. The efforts were to pierce and attempt opening of the valve; only 2 infants survived. We are now convinced that procedures in this direction should be abandoned. Glenn operations do not promise much because of the small size of the subjects.

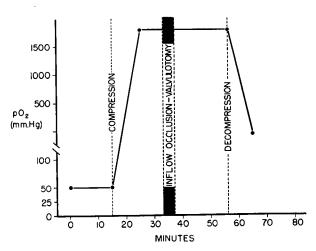


Fig. 10.—Chart of arterial blood oxygen tension of a baby with aortic stenosis who was subjected to hyperbaric conditions in a compression chamber for surgical aortic valvulotomy. Under conditions of compression to 3 atmospheres, the enormous increase in arterial PO_2 allows a very long safe period of inflow occlusion and circulatory arrest.

In the last year, our policy in all instances has been: (1) performance of operation in a compression chamber to improve the condition of the baby temporarily by markedly stepping up the oxygenation of tissues, (2) complete avoidance of any attempt to penetrate the valve, and (3) establishment of a right Blalock shunt (generally performed in an end-to-end fashion). In five such attempts by Dr. William Bernhard, all survived. Four of these patients are shown in Fig. 11.

TRICUSPID ATRESIA OR STENOSIS

In this malformation the primary lesion is obliteration (or marked hypoplasia) of the tricuspid valve. The right ventricle is always diminutive and often some degree of pulmonary stenosis or atresia is present. Of necessity, there is an interatrial septal defect, usually (but not always) of adequate size to empty the right atrium into the left side of the heart. In occasional cases, there may be an interventricular septal defect which allows some blood to enter the right ventricle and flow up into the pulmonary circuit. More and more of these babies



Fig. 11.—Four babies with pulmonary atresia and intact ventricular septum. All were successfully operated upon under hyperbaric conditions in a compression chamber for establishment of aorticopulmonary shunts.

are coming to the attention of surgeons in the early weeks or months of life, when there is sudden emergency because a ductus is closing off. Riker has described well attempts to operate on these small, critically ill babies when he states "that an anastomosis is made amid the asphyxial gasps of the patient, the dire warnings of the anesthesiologist, and the ominous slowing of the heart." These can be life-and-death struggles on the table.

While Glenn shunts are excellent for tricuspid blockages in older children, we have had very poor luck with them in subjects under 15 to 18 pounds in weight. With pulmonary arteries so small, the lungs can be perfused better by turning into them a flow at higher pressure in either a Blalock or Potts fashion. The best series I know of is that reported by Potts in which 48 babies (under a year of age) were treated by Potts anastomoses; 38 survived. We believe we have our greatest probability of success if this is done in a compression chamber, for this greatly increases the availability of oxygen to the body tissues and greatly improves temporarily the status of the baby. Thus the surgeon is given a much better opportunity to perform the anastomosis successfully.

TETRALOGY OF FALLOT

For a small baby with a severe form of tetralogy of Fallot, who has syncopal attacks and is urgently in need of surgical help, the technical problem of relief can be considerable. Because of obvious practical limitations, total repair is rarely possible. A Brock approach to open up the infundibulum or pulmonary valve (by Fell and others) has some theoretical advantage, but has had only

limited adoption. The dramatic palliative effects of a surgically established shunt are still as evident today as they were 20 years ago, when that magnificent step was made by Blalock and Taussig.

To subject a small, extremely cyanotic baby to operation for establishment of a shunt is all too frequently followed by a deterioration during operation or immediately thereafter. Under hurried and harrassing conditions it can be quite a task to get a good vessel union, which is so important in keeping the shunt open postoperatively. The various difficulties are reflected in a high mortality rate (24 deaths in our 71 cases) when compared to a risk of only a few per cent for the same operation in older children.

To improve results, we have now found it tremendously helpful to operate on these babies in a compression chamber (this work is being done by Dr. William Bernhard of our staff) for this usually dramatically improves the condition of the baby temporarily and thus allows the surgeon to be more deliberate and more careful in establishing an excellent anastomosis (which gives the best assurance that it will stay open). The temporary benefits of compression can be regarded as a short but golden hour, during which operation can be carried out in a less hurried and in a more precise way. Anastomoses of a Blalock type are preferred, but a Potts union is sometimes still employed. On a few occasions we have brought the distal end of the right pulmonary artery to the side of the ascending aorta. In both of the latter kinds of union there is a real risk in making a communication too large, thereby flooding the lungs and leading to pulmonary edema which is rapidly fatal. Our percentage of survivors has risen noticeably with the performance of operation under hyperbaric conditions; in 1963, 15 patients were treated thus, with 13 survivors.

TRANSPOSITION OF THE GREAT VESSELS

Here, the fundamental defect is that the right ventricle pumps bluish blood into the aorta and the left ventricle delivers well-oxygenated blood into the pulmonary arterial tree. This is incompatible with life unless there is some cross-communication between the two systems through an interventricular septal defect, an interatrial orifice, or a patent ductus. Concurrently, there might be blockage at some valve, such as the tricuspid or, particularly, the pulmonic. While a few of these patients can live to older childhood, the vast majority die in the first year. Keith finds that 52 per cent are dead at 1 month and 86 per cent at 6 months. Since transposition is seen in 30 or 40 per cent of all babies born with cyanotic congenital heart disease, the magnitude of the challenge to surgeons is evident.

For the occasional older child of larger size, therapy can range through a variety of approaches, such as a Senning revision or modification thereof, the Baffes procedure, the Glenn anastomosis, or even the shifting of the great arteries. During the better part of a decade we have had discouraging losses with these various attempts. All such maneuvers are too extensive for handling the average sort of case which confronts us, which is that of a baby only a few

weeks or months of age, intensely cyanotic, desperately needing surgical treatment but who is not going to stand much of it.

Our success has been in two directions. First, there are a few babies who have a large intracardiac shunt and whose lungs are being flooded; banding of the pulmonary artery can be very helpful. Second, the most common situation we face is that wherein an insufficient communication exists between the two circulations; this is best handled by making a larger interatrial opening. For this, there has been widespread use of the Blalock-Hanlon procedure, but recently we have preferred to handle these by placing the child in a compression chamber, temporarily improving the status of the child, using inflow occlusion (Fig. 12), opening the right auriele and, under direct vision, excising a piece of septum. This can be done with only 2 or $2\frac{1}{2}$ minutes of suspension of the circulation, which is well within the limits of safety. During the last year and a half, this approach (by Dr. Bernhard) has given an 80 per cent survival rate. This has been a very worth-while step to give important palliation; hopefully, more extensive reconstruction can be offered at an older age.

ESOPHAGEAL ATRESIA

As a final chapter, a few remarks about esophageal atresia. My fascination with this anomaly stems from the fact that I have lived through the entire period of development of surgery for it. Through the thirties, on our service and under Dr. Ladd's direction, there were more than 50 surgical attempts, all ending fatally. His first success came in 1939. In the early forties, Ladd brought 12 children through multiple-stage antethoracic repairs, now only of historical interest, yet they formed an opening wedge in surgical attack on the problem. The first successful anastomosis of the esophagus for atresia was accomplished by Cameron Haight in 1941, and this quickly focused attention on repair within the mediastinum as the preferred method of treatment, an approach which is now widely used whenever possible. Today, throughout the land, 70 to 80 per cent of these babies can have a reconstruction of the esophagus and live. Our survival rates for 1961 and 1962 were 80 and 81 per cent.

Of all the techniques of union which can be employed, I am firmly convinced that by far the best is Haight's two-layered repair, with the muscularis of the upper pouch pulled down in a sleeve-like manner to cover over the inner line of sutures.

For the 15 years up to 1957, primary anastomosis was the invariable routine in our hospital. Successes were intermingled with complications and also some deaths. Yet there was the pleasing aspect that 140 babies were alive and doing well after a one-step operation. It was thrilling to find that a tiny infant, even as small as 2 pounds, 14 ounces, could be brought through with a single procedure. But the occasional brilliant result in premature infants tended to obscure the fact that many small fragile infants died after this rather extensive surgery. A review of our records from 1953 through 1957 showed 62 babies handled by a one-stage anastomosis. For those over 7 pounds, there was 100 per cent survival; for those between 6 and 7 pounds it was 63 per cent, for those 5 to 6 pounds

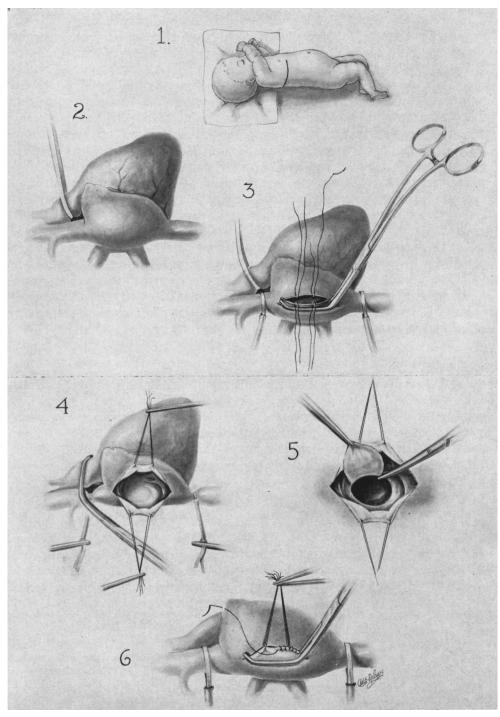


Fig. 12.—Treatment of transposition of the great vessels when there is inadequate mixing between the pulmonary and peripheral circulations. Operation is performed under hyperbaric conditions in a compression chamber, an interatrial communication is enlarged by inflow occlusion, and septal wall is removed under direct vision.

it fell to 53 per cent, and for those between 2 and 5 pounds it was only 28 per cent. Therefore in 1958 we adopted a policy of using staged procedures for infants under 5 pounds. On hospital entry, constant nasoesophageal suction is instituted for evacuation of saliva from the upper esophageal pouch, a gastrostomy is performed for feeding, and in the next day or two the tracheoesophageal fistula is divided. After the child reaches 6 or 7 pounds in weight, an anastomosis of the esophageal ends is made in the posterior mediastinum. Such a delayed, or staged, attack for premature babies was presented before this Association 2 years ago by Drs. Holder, McDonald, and Woolley. The effectiveness of this policy can now be judged from the fact that prior to 1958 (when primary anastomosis was performed on all babies regardless of size) there was recovery of only 28 per cent of premature infants under 5 pounds, whereas in the 5 years since then, the staged procedures have raised this to 62 per cent.

At times, when depressed about some aspect of medicine or surgery, I get a great lift by pulling out pictures of children who have survived repairs for esophageal atresia. These healthy growing youngsters are a delight to see; here are a few. I will not take time to show photographs of *all* who have recovered, because we now have 220 of these happy faces. Certainly, esophageal surgery for babies has been one of the great advances of our generation.

MISCELLANEOUS

And then there must be the inevitable group called "miscellaneous." It is sizable because it includes a wide variety of conditions; sternal anomalies, esophageal stenosis, tracheoesophageal fistula without atresia, empyema, spontaneous pneumothorax, chylothorax, cor triatriatum, ventricular diverticulum, sequestered lobe, foreign bodies, remnants of the neuroenteric canal, etc. Some of these are fairly common and merit interesting discussion but we will have to pass them by.

SUMMARY AND CONCLUSIONS

Table VI sums up our material from 1,212 infants who have undergone some thoracic operation. Almost half have been of a cardiovascular nature.

We have made no counting of babies who died in our institution who were potentially salvageable by surgery which for one reason or another was not at-

TABLE VI. SUMMARY OF 1,212 THORACIC OPERATIONS FOR INFANTS IN THE FIRST YEAR OF LIFE

Tumors and cysts	43	Pulmonary stenosis	19
Lobar emphysema	21	Pulmonary atresia	16
Diaphragmatic hernia	136	Aortic stenosis	13
Hiatus hernia	19	Ventricular septal defect	25
Esophageal atresia	402	Tetralogy of Fallot	71
Vascular ring	129	Patent ductus arteriosus	119
Transposition of great vessels	53	Coarctation	21
Tricuspid atresia	18	Miscellaneous	107

tempted, but in looking backward there come to mind literally scores of instances when autopsy disclosed a pathologic condition which technically could have been corrected. We are left with the feeling that a more aggressive surgical approach could have saved many more lives. Immediate action is often necessary, for a considerable proportion of these babies present emergency situations which require prompt attention if the moment of opportunity is not to be lost and if success is to be courted.

Much could be said regarding diagnostic techniques, technical aspects of surgery on tiny subjects, points in anesthetic management, supportive care in the postoperative period, etc., all of which are highly significant in the successful practice of this branch of surgery. Rather than deal here with these details, important as they all are, it is my purpose to paint a broad picture and emphasize that many babies, often in the newborn period, have urgent need for the skills of thoracic surgeons. What I have been able to see in one small hospital makes it completely evident that there must be extensive need for this kind of work across our entire continent. Many of you have engaged in this already and have contributed greatly to it. There is little doubt that virtually all of you will be called upon in the future for this activity and there is much that you can accomplish. The challenge is great; the rewards are enormous. For me there have been few things in life which have been more satisfying than to face a small child, struggling for his very existence, to perform some corrective surgical maneuver, and later to see the youngster, thriving and healthy, starting out in life, sound in body and mind.

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