CONGENITAL ATRESIA OF THE ESOPHAGUS WITH TRACHEOESOPHAGEAL FISTULA*

RECONSTRUCTION OF ESOPHAGEAL CONTINUITY BY PRIMARY ANASTOMOSIS

CAMERON HAIGHT, M.D.
ANN ARBOR, MICH.

FROM THE DEPARTMENT OF SURGERY, UNIVERSITY OF MICHIGAN MEDICAL SCHOOL, ANN ARBOR, MICH.

IN THE ANOMALY of congenital atresia of the esophagus, a dysplasia that is more extensive than an atresia is found at a level corresponding to the junction of the upper and middle thirds of the esophagus. The upper portion of the esophagus ends as a blind pouch in almost all instances. The upper end of the lower esophageal segment usually enters the trachea, thus forming a tracheoesophageal fistula; this occurred in 29 of the 32 patients in this series. The condition is incompatible with life, as feedings can not enter the stomach. Furthermore, the feedings and accumulated secretions in the blind upper segment overflow into the trachea, and death soon results from aspiration pneumonia. The surgically ideal plan for the correction of the anomaly is one that accomplishes ligation of the fistula and reestablishment of esophageal continuity by a primary anastomosis of the esophageal segments. This article will be concerned primarily with the anatomic problems and surgical experiences that have been encountered in 24 patients for whom an exploration of the anomaly was undertaken. Reconstruction of the esophagus was accomplished in 16 patients, of whom six are now living from seven months to three years and one month after operation.

THE ANATOMIC PROBLEM

The suspicion of a congenital obstruction of the esophagus is aroused by attacks of choking, dyspnea and cyanosis on attempts to swallow fluids. An actual obstruction of the esophagus can be demonstrated by the inability to pass a catheter into the stomach. To be certain that the obstruction is complete, iodized oil is given by mouth under fluoroscopic observation. A dilated blind upper esophageal segment, terminating usually at the level of the second or third dorsal vertebra, is then evident. In all the 32 cases in this series, the upper segment ended as a blind pouch. In an additional recent case of tracheoesophageal fistula without esophageal artesia, the esophageal lumen was patent and of normal caliber, but a fistulous communication was present between the trachea and the esophagus.

In the presence of a verified complete obstruction of the upper esophagus, the existence of air in the stomach is indicative of a communication between the lower esophageal segment and the trachea or a bronchus. Air was noted

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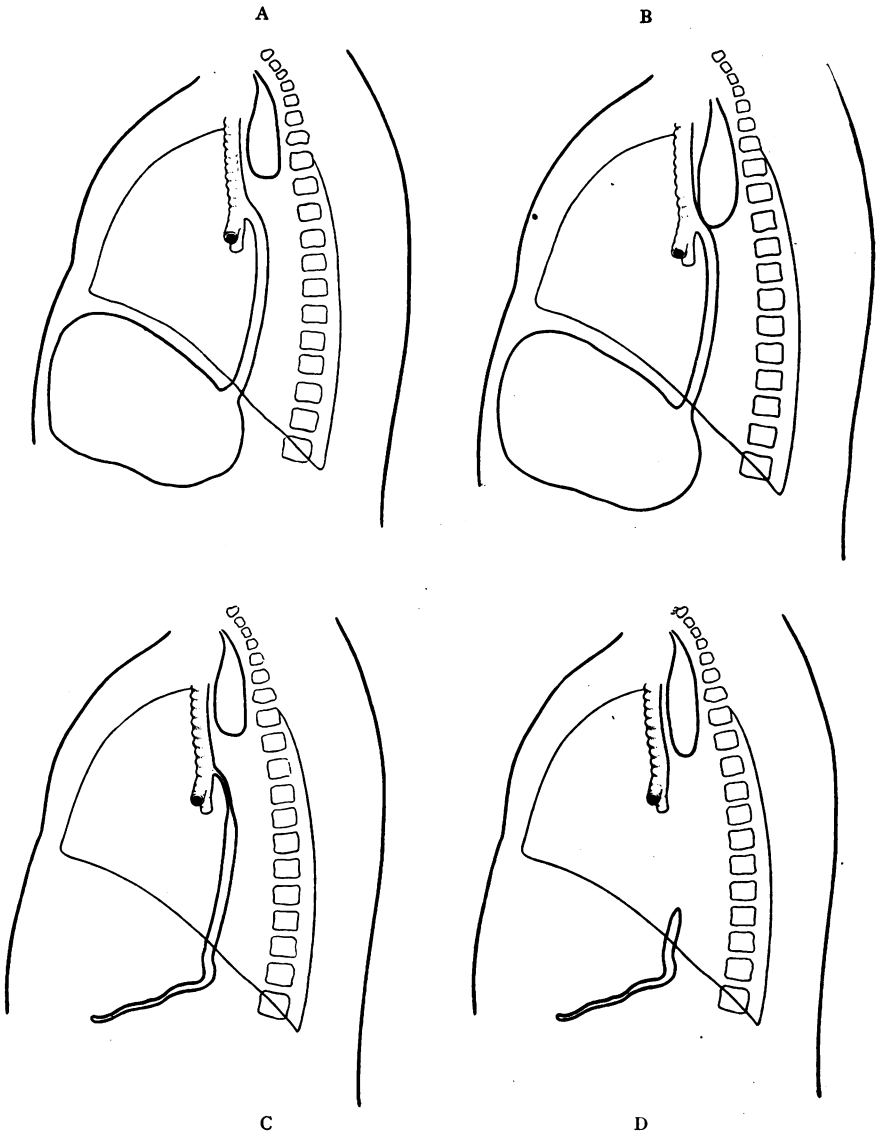
in the stomach or intestines in 26 of the 31 cases studied by roentgenologic examination, and air was present in the stomach before operation in 20 of the 24 patients for whom a thoracic exploration was undertaken. An anastomosis was performed in 15 of the patients with air in the stomach and could have been performed readily in another similar case, thus demonstrating the fact that an anastomosis was possible in 80 per cent of the patients in whom the diagnosis of a tracheoesophageal fistula could be made by the preoperative roentgenologic examination.

The absence of air in the stomach is suggestive, but not necessarily diagnostic, of an absence of a tracheoesophageal fistula. Of the five patients without air in the stomach two patients had a small but patent fistulous communication between the lower esophageal segment and the trachea. In another patient, the exact condition of the lower segment could not be ascertained as surgical exploration was not performed and a postmortem examination was not made. The two remaining patients had an actual agenesis of the esophagus, in that the lower segment of the esophagus extended for only about 1.5 cm. above the level of the diaphragm. Operation was performed in four of the five patients without air in the stomach and an anastomosis was possible in only one case (25 per cent).

The absence of a tracheoesophageal fistula can also be determined by bronchoscopic examination, as advised by Shaw.[22] A preoperative diagnostic bronchoscopic examination has not been undertaken in this clinic as yet, because it has been believed that the final criterion for operability depends upon the result of surgical exploration of the anomaly. It has been definitely established that a fistula was absent in only two of the cases; both were operated upon recently and in both there was agenesis of the lower esophageal segment. In view of this correlation, it would appear that the absence of a fistula which is suspected by the absence of air in the stomach and which could be confirmed by bronchoscopy, is highly suggestive that an anastomosis cannot be done.

The diameter of the upper esophageal segment is usually between 1.2 and 1.5 cm. Although the diameter has been slightly less than 1 cm. in several instances, it has always been of ample size to permit an anastomosis. The length of the upper segment is of greater significance than its diameter, as a short upper segment may interfere considerably with the performance of an anastomosis. If the upper segment is short, and if the lower segment reaches only to the tracheal bifurcation or to a main bronchus, the difficulties are augmented. The gap between the two segments has usually varied between 1.5 and 3 cm. before an attempt has been made to approximate them. Partial muscular continuity of the two segments without any separation between them has been present, however, in five of the 24 patients for whom an exploration was performed. In these cases the muscular attachment was divided and the fistula was ligated at a high level before making the anastomosis.

An unusually narrow diameter of the lower segment, a short length of the



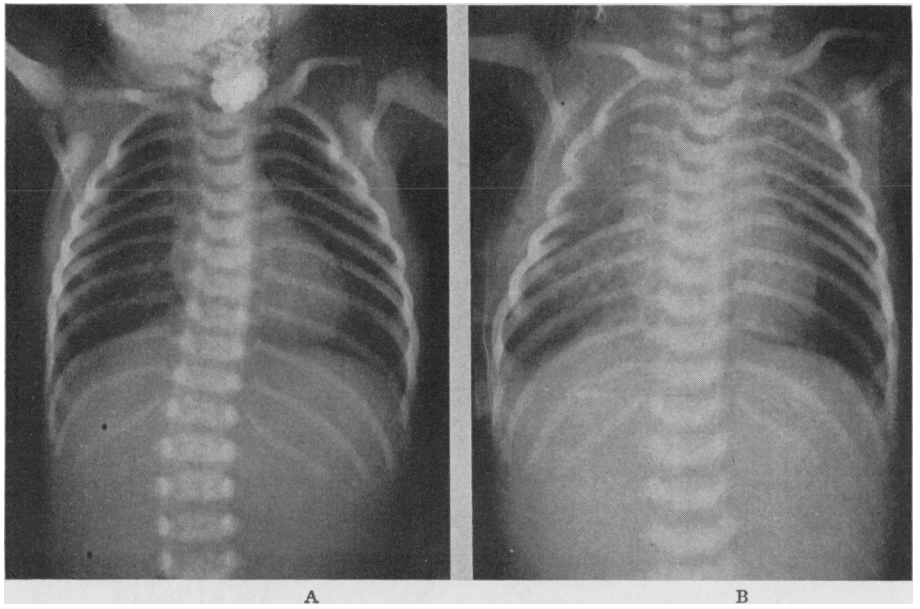
FIG, 1.—The types of the anomaly that have been encountered at operation in 23 of the 24 cases from whom exploratory operation was performed are illustrated. (One patient died during operation before the nature of the lower segment was determined.)

(A) The most frequent type of malformation of the esophagus. The upper esophagus ends as a blind pouch. The lower esophagus arises from the trachea at a level that is usually between 0.5 and 1 cm. above the bifurcation of the trachea. Air is present in the stomach indicating the presence of a tracheoesophageal fistula. This type of anomaly was present in 13 of the 24 patients from whom a primary exploration of the anomaly was performed. Variations in the location of the fistula were found, and in two cases the lower esophagus arose from the right stem bronchus.

(B) Partial muscular continuity of the two segments. The anomaly otherwise is the same as in A (five cases).

(C) The lower segment is greatly contracted for a variable distance below its origin from the trachea and air has not entered the stomach, although a tracheoesophageal fistula was present (two cases). In a third case, the lower segment was contracted, but air was present in the stomach (not illustrated). (Figures 1, A, B, and C, Haight and Towsley,[5] By permission of Surgery, Gynecology and Obstetrics.)

(D) Atresia of the upper esophagus and agenesia of the lower esophagus. The stomach does not contain air (two cases).



A B

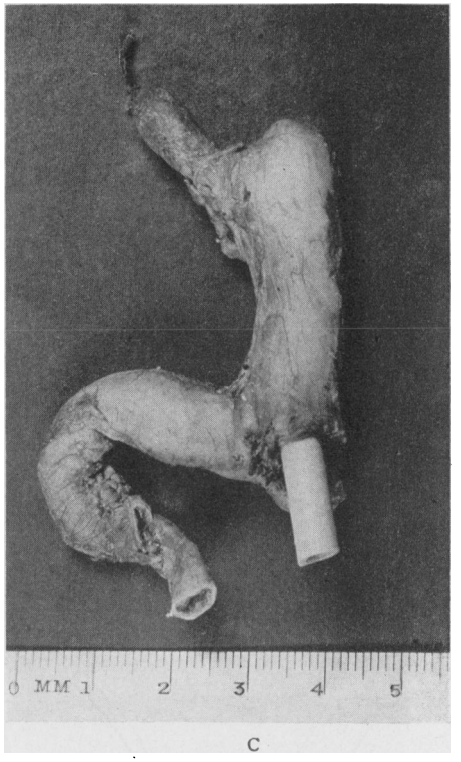


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FIG. 2.—R. R., Case 25: (A) Admission roentgenogram showing absence of air in stomach and intestines. The upper esophageal segment has been visualized by the ingestion of iodized oil. The amount of the oil has been carefully limited, and no oil has entered the trachea or lungs.

(B) Roentgenogram two days after operation. The right extrapleural approach was used and the extent of the costal resections (ribs 3, 4 and 5) is seen. The lower esophageal segment was not found, and a gastrostomy was done five days after the extrapleural operation. Death occurred on the fourteenth postoperative day from lobular pneumonia.

(C) Autopsy showed agenesis of the lower esophagus which extended for a distance of only 1.5 cm. above the diaphragm. In the photograph the short length of the lower esophagus and the entire stomach and duodenum are seen. A narrow strand of fibrous tissue extended upward from the lower esophagus for a distance of several centimeters. The stomach is greatly contracted and a Carrell tube has been inserted into the gastrostomy opening to demonstrate its location and the small size of the stomach.

lower segment (agenesis), and a lo wposition of the tracheoesophageal fistula have been the important factors in preventing an anastomosis from being made in six of the seven patients for whom an anastomosis could not be performed. The diameter of the lower segment is usually between 5–7 Mm. In two cases the diameter was so small (3–4 Mm.) that it was technically impossible to construct an anastomosis. In the two cases of actual agenesis of the lower segment, the lower segment extended for such a short distance above the diaphragm that it could not be found within the field of operation (Fig. 2).

The lower margin of the tracheoesophageal fistula is most often situated about 0.5 to 1.0 cm. above the tracheal bifurcation. When the lower segment is short and the tracheoesophageal fistula is present at the level of the tracheal bifurcation, or if the lower segment enters one of the main bronchi, the chance of obtaining a satisfactory anastomosis has been less than when the lower esophagus enters the trachea at a higher level. In two of the six cases for which an anastomosis could not be done, the fistula was located lower than usual. In one of these patients the lower segment extended directly downward from the tracheal bifurcation, and in the other case it arose from the right main bronchus. It was possible, however, to complete an anastomosis in another patient in whom the fistula arose from the right stem bronchus. In view of this case, it may be seen that the preoperative determination of a low position of a fistula, as might be determined by bronchoscopy, does not definitely exclude the possibility of an anastomosis. It is believed, therefore, that an exploratory operation offers the final criterion regarding the possibility of a satisfactory reconstruction of the esophagus in those cases in which the presence of a tracheoesophageal fistula is evident from the fact that air is seen in the stomach. The seventh patient for whom an anastomosis was not done died as a result of injury to the aorta before the lower segment could be identified.

THE SURGICAL PROBLEM

The surgical problem is essentially a threefold one involving the correction of the esophageal obstruction so that aspiration pneumonia will not occur, ligation of the tracheoesophageal fistula so that gastric contents can not enter the trachea, and the provision of some means for feeding the infant. The early attempts to treat this condition surgically have been discussed in a previous communication.[5] Gastrostomy alone failed because of regurgitation of the gastrostomy feedings into the trachea. Various methods have been employed for interruption of the lower esophageal segment in conjunction with, or supplementary to, gastrostomy.[1, 3, 4, 12, 15, 17, 18, 20, 24, 25] These methods have largely been supplanted by two plans that are now fairly well standardized, insofar as surgical principle is concerned.

One plan, the indirect approach, consists of a three-stage procedure, *i.e.*, (1) extrapleural ligation of the tracheoesophageal fistula; (2) exteriorization of the upper esophageal segment; and (3) gastrostomy. This plan, which has

been used successfully by Leven,[12, 13, 14] Ladd,[9] and Humphreys,[7] anticipates the construction of an antethoracic esophagus at a later date so that feedings can eventually be taken by mouth. A modification of this plan has been suggested by Carter[1] and has been employed successfully by Nettrour.[16*] The indirect plan was proposed[12] because it was believed that the operative mortality could be kept low by staging the operations and by avoiding the extensive extrapleural procedure that would be required by a primary reconstruction of the esophagus. Although it is logical to believe that this advantage should hold true, the three patients for whom the indirect method was begun in this clinic, died before the upper segment was exteriorized. The method was used by election in one of the early cases (Case 7) and by necessity in two subsequent cases (Cases 25 and 32). Had the upper segment been exteriorized without delay, the chance of success would probably have been greater. The indirect plan is, however, essential for the maintenance of life if the anatomic conditions are such that a primary reconstruction of the esophagus can not be done.

The other plan, a direct approach to the problem, consists of a reconstruction of esophageal continuity by a single stage extrapleural ligation of the tracheoesophageal fistula and a primary anastomosis of the esophageal segments. The direct plan of primary anastomosis was suggested in principle by Kieth[8] in 1910. The first reported operation of this type was described by Shaw in 1939, [21, 22] who also mentions a case in which the same technic was employed by Samson.[19] Lanman[11] had previously employed this procedure in four cases in 1936 and 1937. The first successful application of this plan was reported by Haight and Towsley[5] in 1943, and the patient is now living three years and one month after operation. The operation of primary anastomosis has subsequently been used successfully in five additional patients in this clinic, and all of the living patients are able to swallow through the reconstructed esophagus. Shaw,[23] Ladd,[9] Humphreys,[7] and Daniel[2] have also mentioned the successful use of this method.

The reconstruction of esophageal continuity by primary anastomosis represents the ideal surgical principle in the correction of the anomaly in that it allows the infant to swallow normally. Feedings by mouth can be started soon after the operation. If leakage of the anastomosis does not result in an external esophageal fistula or a recurrence of a tracheoesophageal fistula, further operations are avoided. The operative procedure is one of considerable magnitude for a new-born infant, but the infants have been able to withstand the operation considerably better than might be expected. The anatomic findings at the time of the operation will not always permit an anastomosis. An anastomosis was found to be possible, however, in 17, or 70.8 per cent, of the 24 patients for whom an exploration was performed

* This patient was living and well at the age of two years and ten months. The antethoracic esophagoplasty has not as yet been performed. (Personal communication from Dr. C. J. Stoecklein, Pittsburgh, Pa., June 11, 1944.)

and it was possible in 16, or 80 per cent, of the 20 patients who had air in the stomach.

The technical objection to the plan of primary anastomosis results from the fact that tension is invariably present at the site of the anastomosis. Tension occurs primarily because of the gap that often has to be overcome when the two segments are approximated. Less important causes of tension are the normal contractility of the longitudinal musculature of the esophagus and the normal mobility of the two esophageal segments. Observations during the preoperative roentgenoscopic examination, as well as during operation in the first several patients when local anesthesia alone was employed, have shown that the upper segment is displaced upward during swallowing, straining or

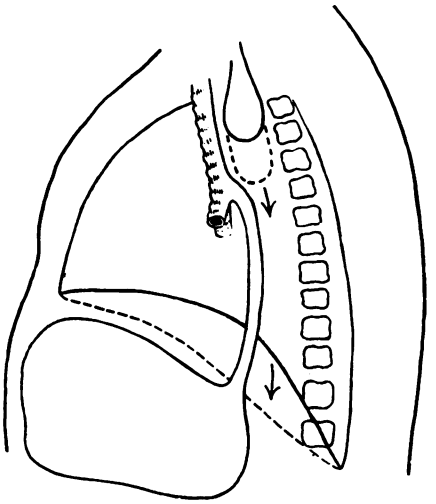

FIG. 3

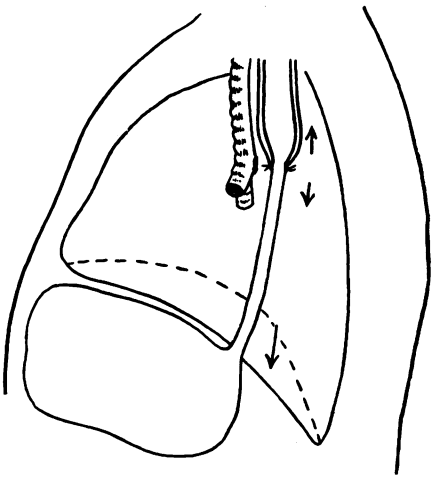

FIG. 4

FIG. 3.—The mobility of the upper esophagus that is seen during roentgenoscopic examination is indicated by the position of the solid and dotted lines. Solid line indicates position of the upper esophagus during the expiratory phase of respiration and during straining and crying with their attending increase in intrathoracic pressure; dotted line indicates position of upper segment during inspiratory phase of respiration. Because of the mobility of the upper esophagus, roentgenoscopic observation allows a more accurate estimation of the position and length of the upper segment than does the roentgenographic examination.

FIG. 4.—Tension unavoidably occurs at the site of the anastomosis because of (1) the gap that usually must be overcome when the two segments are united; (2) the ascent of the upper esophagus during swallowing, crying and the expiratory phase of respiration; (3) the descent of the lower esophagus during the inspiratory phase of respiration; and (4) the normal contractility of the longitudinal musculature of the esophagus.

crying (Fig. 3). The lower segment is normally pulled downward during the inspiratory descent of the diaphragm. Even though tension is not apparent when the patient is under general anesthesia during the operation, it is logical to assume that the mobility of the two segments exerts tension upon the anastomosis during the postoperative period (Fig. 4).

The weak tensile strength of the wall of the underdeveloped lower segment is a further technical disadvantage of the direct plan. The tensile strength of the lower segment is influenced by its diameter and the thickness of its wall. Usually the thickness of the wall varies in proportion to the diameter. In some instances the wall is so extremely thin and delicate that the anastomosis is constructed with considerable difficulty. The weak tensile strength

of the lower segment makes advisable in all cases the complete relaxation that is obtained by general anesthesia, so as to permit the segments to be approximated as readily as possible. Otherwise, there is considerable likelihood that the sutures will pull through the wall of the lower segment during the construction of the anastomosis.

The vascular supply of the upper segment has appeared to be adequate in all cases. The blood supply of the lower segment is less well developed, and there may be a question concerning its adequacy in some cases. For this reason the lower segment is freed from the adjacent tissues only for the minimal distance necessary for its transection. A No. 8-F. catheter, over which the anastomosis is constructed, is removed after completion of the anastomosis, as the catheter may distend the lower segment, if particularly narrow, and so produce ischemic necrosis.

PREOPERATIVE TREATMENT

The extent of the preoperative treatment depends upon the general condition of the patient and the pneumonic complications that may already have occurred. The estimation of the pulmonary status is a matter of immediate concern and should be carried out as soon as the patient is admitted to the hospital. If the infant is cyanotic, mucus should be aspirated from the pharynx and oxygen therapy should be started promptly. During the initial examination, the lung fields will ordinarily show numerous coarse rhonchi resulting from bronchial secretions. The rhonchi will usually disappear after the aspiration of mucus from the pharynx and the hyperventilation of the lungs that is induced by crying or struggling coincident with the aspiration. Aspiration of secretions from the pharynx and upper esophagus should be done at frequent intervals in order to prevent the overflow of secretion into the trachea and to induce hyperventilation so that accumulated secretions in the trachea and bronchi will be effectively expelled.

The pulmonary status is carefully determined by physical and roentgenologic examinations. If a radiopaque solution has not previously been given to establish without doubt the presence of complete esophageal obstruction, it should be used in conjunction with the roentgenologic examination of the chest. The infant is given about 1–2 cc. of iodized oil to swallow. A larger quantity of iodized oil should be avoided, as it is likely to be aspirated into the trachea. The position and length of the upper segment are particularly observed during roentgenoscopic examination. The mobility of the upper segment during respiration is considerable and will usually correspond to the distance occupied by the vertical length of two or three dorsal vertebral bodies. In subsequent film studies, the upper segment may appear to occupy a high position merely because the exposure of the film happens to have been made in the expiratory phase of respiration. An apparently high upper segment, therefore, does not preclude the possibility of a satisfactory anastomosis. At the conclusion of the roentgenologic examination, the iodized oil is aspirated from the upper segment by means of a catheter.

The position of the infant is of importance in lessening the possibility of pulmonary complications. From the time that a diagnosis has been made, the patient is placed in the prone position with the foot of the bed elevated. This position favors the drainage of oral secretions to the exterior and lessens the tendency of aspiration of pharyngeal secretions. The position is changed to the alternate lateral positions at frequent intervals to favor drainage from the upper lobes and thereby lessen the tendency for the development of an atelectasis of the upper lobes. If pneumonia or atelectasis has occurred, the use of the alternate lateral positions is continued, but the uninvolved side is kept uppermost for a greater length of time than the uninvolved side. While the involved side is uppermost, the pharynx is aspirated to stimulate coughing and the expulsion of bronchial secretions.

Parenteral fluid consisting of a 3 or 5 per cent solution of dextrose in distilled water is given preferably by the subcutaneous route. The amount of fluid needed to correct dehydration, if present, will necessarily depend upon the extent of the dehydration. The amount needed to maintain the fluid balance should be considerably less than the calculated daily optimal amount for a normal infant of the same weight. Normal saline or Ringer's solution should be avoided because of its sodium chloride content. If pneumonic consolidation has already occurred, sulfonamide therapy should be started by the subcutaneous route. The need for blood or plasma transfusions before operation will depend upon the nutritional level of the infant, as evidenced by the number of days that have elapsed since birth. A blood transfusion is ordinarily not given before operation, and its use is deferred until the time of operation. Ascorbic acid and vitamin K are routinely employed as prcoperative measures.

INDICATIONS FOR OPERATION

A decision in favor of operation should be made as soon as the infant's general condition is satisfactory. If the lung fields are clear, the sooner the operation is undertaken, the less likely is the chance of a pneumonic complication occurring to a degree sufficient to prevent operation. Usually 6 to 12 hours, and ordinarily not more than 24 hours, will be sufficient time to obtain the maximal beneficial response from the preoperative treatment.

The decision regarding the advisability of operation is made on the basis of several criteria: If infants are premature by more than two or three weeks, it is often questionable whether operation should be performed. The size of the infant is another criterion, in that the larger the infant, the better is the chance that he will withstand the operation. Operation should probably not be undertaken in infants whose birth weight is less than four pounds. Those who are weak and listless, and who have a feeble cry or none at all, are more likely to develop pneumonic complications than strong infants with a good cry. The decision regarding operation in weak infants may need to be deferred until the response to general supportive measures is known. Cyanosis which persists after aspiration of mucus from the throat

and after the use of oxygen therapy is a poor prognostic sign and is regarded at present as a positive contraindication to operation. The body temperature is of importance particularly in respect to dehydration or pneumonia. When fever is present, it is preferable to defer operation until the temperature can be reduced to normal, or nearly normal, by treatment of the causative factor.

The presence or absence of pneumonia is one of the most important criteria regarding the advisability of operation, inasmuch as pneumonia is the chief cause of death in these patients. An extensive bilateral pneumonic consolidation is a definite contraindication to immediate operation. A smaller degree of involvement, particularly if limited to one upper lobe, does not necessarily preclude operation, provided that operation is performed on the side of the pneumonic process. If a barium solution has inadvertently been used for roentgenologic diagnosis and if the barium entered the lungs and produced a pneumonic consolidation, the consolidation is likely to persist. An early stage of pulmonary atelectasis usually responds to corrective measures and does not contraindicate operation, particularly if the operation is performed on the side of the atelectasis. The difficulty is often one of determining whether atelectasis or pneumonia is the predominant pulmonary lesion. Although physical and roentgenologic findings are helpful, the differential diagnosis may depend upon the rapidity of clearing of the lesions or the failure of the abnormal signs to clear by the use of posture, carbon dioxide and oxygen inhalations, and enforced crying or coughing. Other congenital anomalies usually do not contraindicate operation, as the anomalies that are most likely to be encountered are ones that are compatible with life.

THE OPERATIVE APPROACH

Ligation of the tracheoesophageal fistula and anastomosis of the esophageal segments can be performed through either a right or left extrapleural approach. In the 24 patients in this series for whom an exploration has been carried out, the left extrapleural approach was used in 12 and the right extrapleural approach in 11. In one patient operated upon early in the series, a right intrapleural approach was used before it was realized that the respiratory embarrassment would be considerably greater with an intrapleural than with an extrapleural approach. In the first cases in this series, the left extrapleural approach was found to be satisfactory but in the more recent cases in this series, the right extrapleural approach has been used in preference to the left, because of the greater ease in obtaining the exposure of the lower esophageal segment.

The upper segment is readily exposed from either the right or left side. The presence of the aorta on the left prevents the exposure of the lower segment until the aorta has been mobilized by ligation and division of the upper two or three intercostal arteries arising from it. Forward, downward and mesial retraction of the aorta is then required to maintain exposure of the lower segment. The pressure upon the aorta during its retraction necessarily reduces its lumen and even though the retraction is intermittent,

it is believed that the retraction contributes to operative shock. With the right extrapleural approach, the azygos vein is divided and thereafter the exposure of the lower segment is not only better than with the left approach, but it is more easily maintained.

As the pulmonary complication plays such an important rôle in the eventual outcome of the case, the side selected for the operation is now chosen according to the preoperative condition of the lungs (Fig. 5). If both lungs are clear, the preferred right approach is elected. If a pneumonic involvement is present

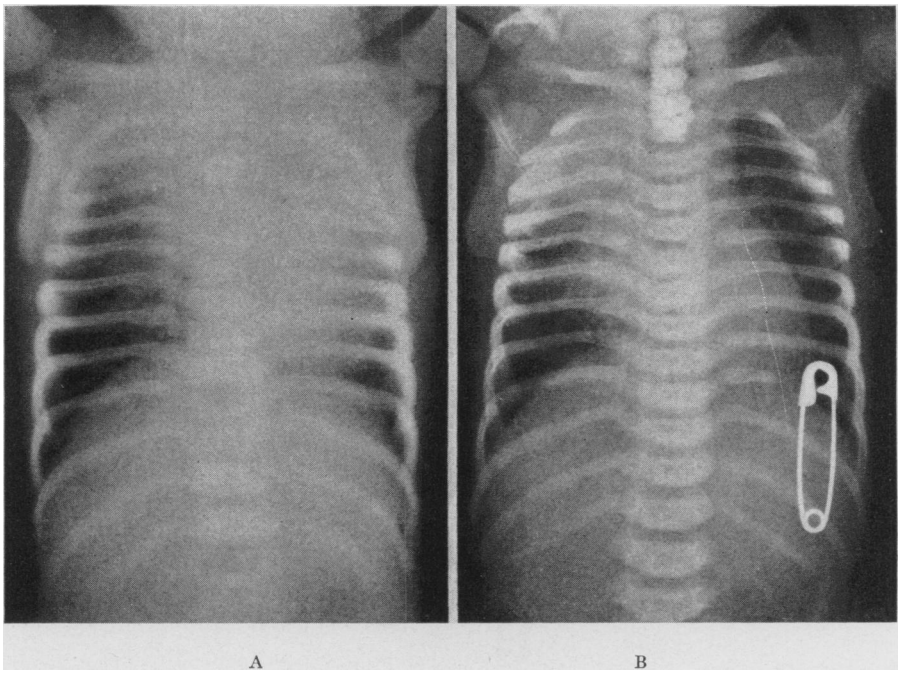

FIG. 5.—V. L., Case 22.—(A) Roentgenogram on admission shows small degree of involvement of right upper lobe and extensive involvement of left upper lobe. When the patient was turned to the right lateral position during the subsequent physical examination, reaeration of the left upper lobe occurred, demonstrating that the involvement of the left upper lobe was on the basis of atelectasis and not a pneumonia. Roentgenogram (B) was made immediately after the physical examination and three hours after roentgenogram (A). The left upper lobe is now clear. The infiltration in the right upper lobe which has persisted in spite of the use of the left lateral position during the physical examination is probably pneumonic in nature. Operation was undertaken one hour after roentgenogram (B). The *right* extrapleural approach was used because of the pulmonary involvement on the right. An anastomosis was completed and the patient is now living 13 months after operation.

in one lung only, the operation is done from the corresponding side. In this way the function of the good contralateral lung will not be impaired by the costal resections and the temporary postoperative extrapleural collapse of the upper portion of the lung. The extrapleural collapse of the upper lobe appears at times to have a beneficial rather than a detrimental effect upon a mild pneumonic process within the collapsed upper lobe. Also, if the pulmonary involvement is due to atelectasis, improvement of the atelectasis may be expected as the result of the uppermost position of the involved lung during the operation. As an upper lobe atelectasis may occur before operation in spite of precautionary measures, the location of the atelectasis can be governed

by the posture of the infant before operation. If during the intervals when the patient is in the lateral position, the left lung is kept uppermost for a longer period than the right lung, the left lung can usually be kept clear. Should an atelectasis develop, it would be likely to occur on the right side and thereby the preferable right-sided approach could be employed.

OPERATIVE TECHNIC
RIGHT EXTRAPLEURAL APPROACH

The neck is shaved posteriorly up to the external occipital protuberance. The preoperative medication of codeine and atropine is adjusted to the

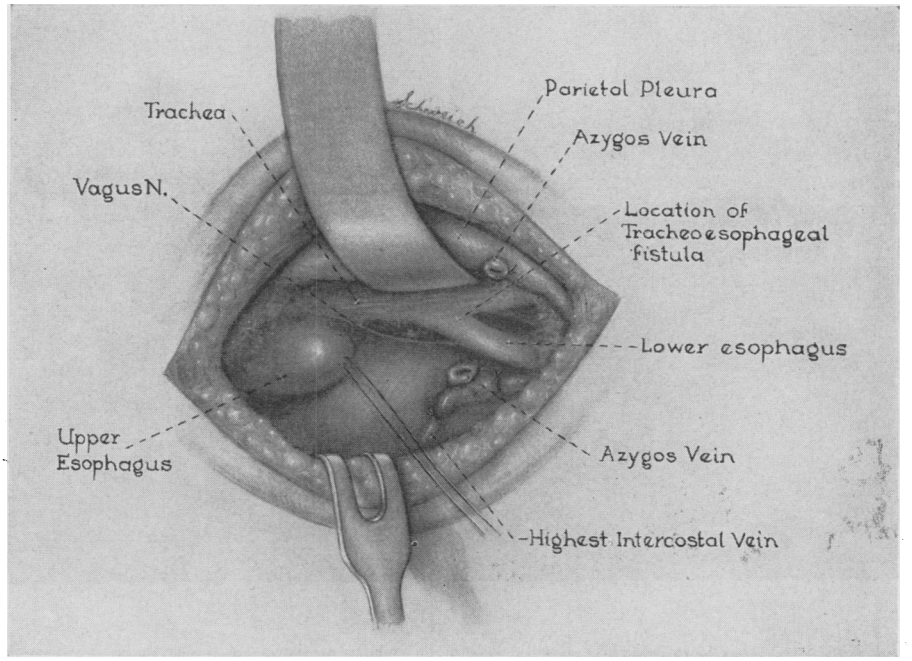

FIG. 6.—The right extrapleural exposure of the anomaly is illustrated. The posterior portions of the third, fourth and fifth ribs have been resected and the parietal pleura has been freed from the thoracic wall. The azygos vein has been divided in order to expose the lower esophagus. The dilated blind upper segment has been partially freed from the posterior wall of the trachea. The relative size of the upper and lower esophagus, and the usual position of the tracheoesophageal fistula are shown. The right vagus nerve serves as a useful guide for locating the lower segment.

weight of the infant. As a satisfactory means for blood replacement is essential, a cannula is inserted in an ankle vein on the side opposite to the side of the operation. The size of the cannula (Nos. 22 F. or 24 F.) depends upon the size of the vein. Between 100 and 150 cc. of whole blood, diluted with an equal amount of physiologic saline or Ringer's solution, is given during the operation and the first several hours after operation. Although the saline or Ringer's solution is undesirable because of its sodium chloride content, a diluent has been employed so that the blood will flow through the small cannula.

The patient is secured to a frame and external heat is applied. After preparation of the skin, the location of the incision is indicated by a cutaneous

scratch before the application of the sterile drapes. Without this precaution, the laxity of the skin may be responsible for an incorrect position of the incision. Local anesthesia consisting of one-half of one per cent procaine solution or one-quarter of one per cent metcycaine hydrochloride solution is used until the costal resections are begun. Light ether anesthesia by the drop method is used during the remainder of the operation. An infant-sized face mask for administration of positive pressure oxygen should be available for use in the event that the parietal pleura is inadvertently opened. The incision parallels the lateral border of the sacrospinalis muscle and extends from the second to the sixth ribs, inclusive. Careful hemostasis is essential. The trapezius and rhomboideus major muscles are incised, and the sacrospinalis muscle is retracted mesially. The third, fourth and fifth ribs are resected subperiosteally from the plane of the transverse processes to a point above 1.0–1.5 cm. lateral to the angles of the ribs. The third and fourth intercostal nerves and muscles are divided close to the transverse processes, and the corresponding intercostal vessels are doubly ligated and divided. The periosteum of the resected ribs is incised vertically and the extrapleural separation of the parietal pleura is begun. A small periosteal, or dural, elevator allows the extrapleural separation to be done under direct vision. The fibers of the endothoracic fascia hold the pleura tightly to the intercostal muscles between the heads of the ribs. These fibers must be carefully divided under direct vision to prevent the extremely thin parietal pleura from being torn at this stage of the operation. During the further separation of the parietal pleura from the costovertebral gutter, a small gauze pledget, secured in the jaws of a hemostat, is used as a dissector. A spatula retractor is used to hold the parietal pleura out of the costovertebral gutter and to maintain exposure. The arch of the azygos vein is exposed. The parietal pleura immediately beneath it is especially thin and is in particular danger of being punctured or torn. The azygos vein is ligated centrally with two doubled silk ligatures. The main trunk of the azygos vein below the entrance of the highest intercostal vein is singly ligated, the highest intercostal vein is similarly ligated and the azygos vein is divided central to the entrance of the highest intercostal vein. In the earlier cases in this series, an illuminated retractor was employed, but it is cumbersome in the small operative field. The innominate artery may be exposed at times, but if the operative field is limited to the posterior mediastinum, it is usually not visualized.

The areolar tissue posterior to the trachea is infiltrated with a local anesthetic solution to prevent untoward vagal reflexes and the vagus nerve, if seen, is also blocked. The upper esophageal segment is searched for in the loose areolar tissue posterior to the trachea. If the upper segment has a low position, it is usually found without difficulty. If it is not promptly seen, a catheter is introduced into the mouth by the anesthetist and advanced into the upper segment. Gentle pressure upon the catheter pushes the upper segment downward where the tip of the catheter may be felt within the upper segment. The areolar tissue is then freed from the upper segment and a

tension suture is inserted in the blind end. If the upper segment is unusually short, it may be necessary to resect the posterior portion of the second rib and to divide the second intercostal structures in order to obtain adequate exposure of the upper segment.

The lower esophageal segment is usually found without difficulty when it arises at a point between 1 and 1.5 cm. above the tracheal bifurcation. If the lower segment is not readily identified, the prominent right vagus nerve can often be seen on the right lateral wall of the trachea or in the space posterior to the trachea. The vagus nerve is traced down to the lower segment, which is then readily found unless it should arise in the crotch between the two main bronchi. Occasionally, a strand of fibrous tissue or muscle extends between the two segments and can be traced downward to the lower segment. Considerable difficulty has been experienced in locating the lower segment when it enters the tracheobronchial tree at an extremely low level; that is, when the lower segment passes directly downward from the carina or one of the stem bronchi. In three of the 24 cases, the posterior aspect of both main bronchi was exposed before the lower segment could be definitely lying in the angle between them. In the two cases with agenesis of the lower esophagus, both main bronchi were exposed posteriorly before the absence of a tracheoesophageal fistula could be definitely determined.

A right-angled hemostat is passed around the lower segment just below its entrance into the trachea or bronchus. A narrow tape or thread is then placed around the lower segment to provide tension and to aid in the freeing of the lower segment up to the fistulous communication. After the lower segment has been freed from the trachea, the decision is made regarding the advisability of attempting an anastomosis. The upper segment is pulled downward with the tension suture and the proximity of the two segments is noted. With the upper segment pulled down, a gap of 0.5 to 1 cm. can usually be overcome by the upward mobility of the lower segment after it has been divided. The external diameter of the lower segment should also be estimated. If the lower segment is narrow and its diameter is only three or even four millimeters, the possibility of securing a satisfactory anastomosis is extremely small. If the gap is too great or the lower segment too narrow, the preferable plan, in so far as recovery of the infant is concerned, is to abandon a direct anastomosis and to treat the condition by the indirect method. The lower segment would then be ligated, but not divided so that the factor of possible contamination of the wound would be avoided.

If it seems likely that an anastomosis can be made, the lower segment is ligated with a single or double silk ligature as close as possible to the trachea. A tension suture is placed in the wall of the lower segment distal to the ligature, the lower segment is divided and the mucosa of the two ends is cleansed with a mild antiseptic solution. As a tendency exists for the ligature to roll off the ligated proximal stump, several suture ligatures are used to cover the stump. The lower segment arises from the trachea at a sharp

angle and, as a ligature of the lower segment necessarily lies at a right angle to the direction of the lumen, a several millimeter length of the posterior wall of the lower segment is sacrificed when a ligature is used. If it seems that this additional length posteriorly will be of advantage in the construction of the anastomosis, it can be saved by placing a narrow hemostat across the distal segment almost flush with the trachea and parallel to the direction of the trachea. The lower segment is then divided and the clamp is oversewn with several figure-of-eight plastic silk sutures (Fig. 7). Additional interrupted sutures may be used if needed. This method of dividing the lower segment has been used only in the two most recent cases in this series.

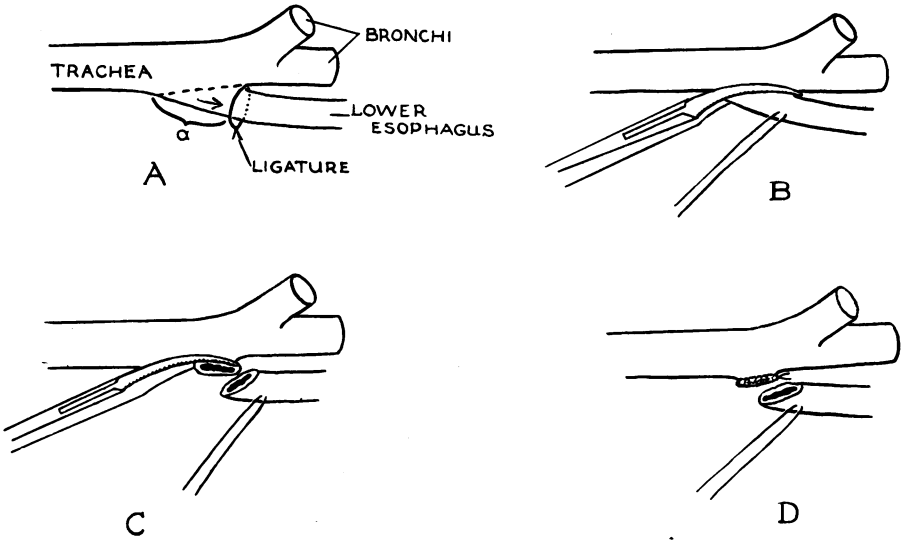

FIG. 7.—(A) The lower esophageal segment arises from the trachea at a sharp angle. A ligature will, of necessity, lie at a right angle to the long axis of the esophagus. Closure of the fistula by a ligature sacrifices the upper portion of the posterior wall indicated by the bracket (a).

B, C and D illustrate the method of closure of the tracheoesophageal fistula that has been used in the two most recent cases.

(B) The upper portion of the posterior wall of the lower segment is preserved by the placement of a mosquito hemostat across the lower segment at its entrance into the trachea.

(C) The lower segment has been divided by an oblique incision. A greater length of the esophageal wall is preserved posteriorly by this method, and a larger circumference is available for the anastomosis.

(D) The hemostat is oversewn with figure-of-eight sutures and is removed. Additional interrupted sutures are used to strengthen the closure. By this method the tracheoesophageal fistula is closed by suture, rather than by ligature. The relative efficacy of the two methods in the prevention of reopening of the fistula has not been established as yet.

The blind end of the upper segment is opened for a distance that corresponds to the diameter of the lower segment. The musculature of the anterior wall of the upper segment is less well developed than it is laterally or posteriorly. As the anterior wall of the upper segment has on occasions split upward during the construction of the anastomosis, the incision in the upper segment carefully avoids the anterior wall. The interior of the upper segment is cleansed with an antiseptic solution. A No. 8 F. catheter with its flared outer end completely removed is introduced into the two segments by way of the wound. One end of the catheter is advanced into the lower segment until it enters the stomach. The other end is advanced through the upper

segment until it enters the pharynx where the anesthetist grasps it with a hemostat and withdraws the end of the catheter from the mouth. The catheter stabilizes the esophagus during the construction of the anastomosis, and the open ends of the two segments are kept from collapsing, thereby allowing the sutures to be placed more readily than when a catheter is not used.

Considerable difficulty is often encountered in the construction of the anastomosis because of the tension that is present and the thinness of the wall of the lower segment. Delicate, atraumatic round-point needles* and the

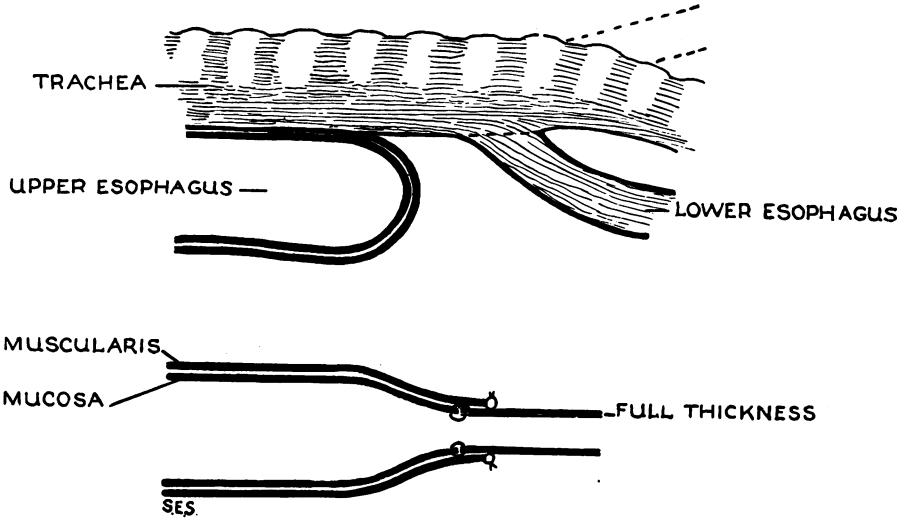

FIG. 8.—The upper drawing shows the blind upper esophageal segment and the lower esophagus arising from the trachea. The relative size of the two segments and the relative thickness of their walls are illustrated.

The lower drawing shows the completed "telescopic" anastomosis. This type of anastomosis is used because of the discrepancy in the size of the two segments and the discrepancy in the thickness of their walls. The relatively thick wall of the dilated upper segment and the extremely thin wall of the smaller lower segment are illustrated. The thickness of the entire wall of the lower segment is usually no greater than the thickness of the mucosa of the upper segment. The inner row of sutures approximates the full thickness of the wall of the lower segment to the mucosa and submucosa of the upper segment. The outer layer of sutures draws the muscularis of the upper segment downward and anchors it to the outer wall of the lower segment at a level several millimeters below the inner layer of sutures. Interrupted sutures of plastic silk are used in the construction of the anastomosis.

smallest available silk sutures (Deknatel A) are employed, so that a minimal amount of trauma is produced. A "telescopic" type of anastomosis (Fig. 8) has been employed for the patients operated upon during the last year and a half. The discrepancy between the thickness of the walls of the two segments readily permits this type of an anastomosis. The thickness of the underdeveloped wall of the lower segment is usually no greater than the thickness of the mucosa of the upper segment. An inner row of about eight interrupted sutures approximates the mucosa and a small amount of submucosa of the upper segment to the full thickness of the wall of the lower segment. As there is considerable mobility of the muscular layer of the upper segment on the submucosa, the muscularis of the upper segment can be pulled downward and sutured to the outer surface of the lower segment, 2 or 3 Mm. below the level of the first row of sutures. This second or outer layer of the anastomosis

* Anchor brand No. 1833–4, ½ circle, taper point.

consists of approximately the same number of interrupted sutures that are used for the inner layer. The catheter is withdrawn after the anastomosis is completed.

A pneumothorax resulting from the accidental opening of the pleura during the operation is not well tolerated because of the normally low respiratory reserve of infants. If a pneumothorax should occur, the lung should be promptly reexpanded with positive pressure oxygen administered by a face mask, and the opening through the pleura should be closed if possible. The thin parietal pleura, however, does not lend itself well to suturing, as the puncture wounds caused by the smallest available needle may allow leakage of air or cause a further tear of the pleura. After the parietal pleura has been freed from the thoracic wall, the redundancy of the pleura often allows the torn edges to be grasped and pulled outward. If the tear is not too large, an encircling ligature can be placed beyond the tear, and an air-tight closure is thereby obtained. This maneuver usually can not be employed if the tear is in the region of the arch of the azygos vein. When the tear can not be closed, a small gauze pack is placed against the pleural opening after the lung has been expanded by positive pressure oxygen. Air can be prevented from entering the pleural space by maintaining firm pressure against the pack with the spatula retractor. At the time of closure of the thoracic wall incision, the inner portion of a small gauze drain is placed against the opening in the pleura. Expansion of the lung is maintained by the positive pressure until the wound is closed air-tightly around the drain. The pressure exerted by the gauze keeps the parietal pleura in contact with the lung, and the irritation produced by the gauze causes a rapid adherence of the lung to the parietal pleura at the site of the tear. This method of sealing the opening in the parietal pleura has been successful in the prevention of an empyema, even though the extrapleural wound in some of the cases has become grossly infected as a result of leakage of the anastomosis.

The wound is irrigated with physiologic saline solution before the closure is begun. In the event that the pleura has not been opened, a soft rubber tissue drain is placed near the site of the anastomosis as a safeguard in the event that leakage of the anastomosis should occur. The long free anterior ends of the divided intercostal muscles are sutured to the under surface of the sacrospinalis muscle, and the extracostal wound is closed in layers with interrupted sutures. Fine catgut is used for the buried sutures because of possible suppuration of the wound resulting from leakage of the anastomosis. The skin edges are approximated with interrupted sutures of plastic silk.

The operative technic for the left extrapleural approach[5] varies only in the exposure of the anomaly. After the parietal pleura has been separated from the thoracic wall, the highest intercostal vein is doubly ligated and divided. The left subclavian artery is identified and retracted forward, so as to expose the retrotracheal space. The upper segment of the esophagus is located by the same measures described for the right extrapleural approach. In order to expose the lower esophageal segment, the uppermost two or

three intercostal arteries are doubly ligated and divided close to the aorta. The aorta is then retracted forward, downward and mesially. The left vagus nerve is not seen, but the left recurrent laryngeal nerve may be encountered and injury to it should be avoided.

POSTOPERATIVE MANAGEMENT

If physical or roentgenographic signs of a moderate or considerable pneumothorax are present, the air is aspirated from the pleural space. The infant is placed upon the operated side to reduce the paradoxical motion of the thoracic wall resulting from the costal resections. The foot of the bed is elevated to favor the drainage of secretions from the respiratory tract to the exterior. External heat is applied and oxygen is administered continuously with a face mask while the patient is being returned to the nursery. Prior to the use of oxygen during this interval, intense cyanosis often was present upon the patient's return to the nursery. Oxygen is subsequently administered by means of a tent. The rectal temperature is recorded and, as the temperature is frequently subnormal, external heat is continued. The temperature of the air within the tent is maintained at a considerably higher level than is customary with oxygen tents. If the infant is premature, or if difficulty should be experienced in maintaining the body temperature at a normal level, the patient is placed in an incubator. The intravenous infusion is slowly continued until the required amount of 100 to 150 cc. of blood has been given.

The condition of a few of the infants during the first several hours after operation has been precarious. Sudden attacks of cyanosis with complete cessation of respiration have occurred as a result of obstruction of the airway by secretions. Immediate aspiration of the pharynx is required and, artificial respirations, carbon dioxide and oxygen inhalations, and subcutaneous injection of caffeine sodium benzoate are used to restore respiration. The recovery of the infant from these episodes is almost as sudden and dramatic as is the speed with which they arise. Should the onset of these episodes not be recognized promptly, death may occur suddenly before corrective measures can be instituted. To prevent these attacks insofar as possible, the character of the respirations is carefully determined at frequent intervals, and pharyngeal suction is used at the first appearance of a wet-type of breathing.

Subcutaneous fluid, consisting of a 3 or 5 per cent solution of dextrose in distilled water, is given during the first 12 or 18 hours after operation only if an insufficient amount of fluid has been administered in conjunction with the blood transfusion. The subcutaneous daily amount of subcutaneous fluid is restricted, because of the danger of producing pulmonary edema by the administration of too large a quantity. The amount of fluid ordinarily given is about one and one-half ounces per pound of body weight, instead of the customary two and one-half ounces per pound. The subcutaneous fluids consist of a 3 or 5 per cent solution of dextrose, supplemented with the

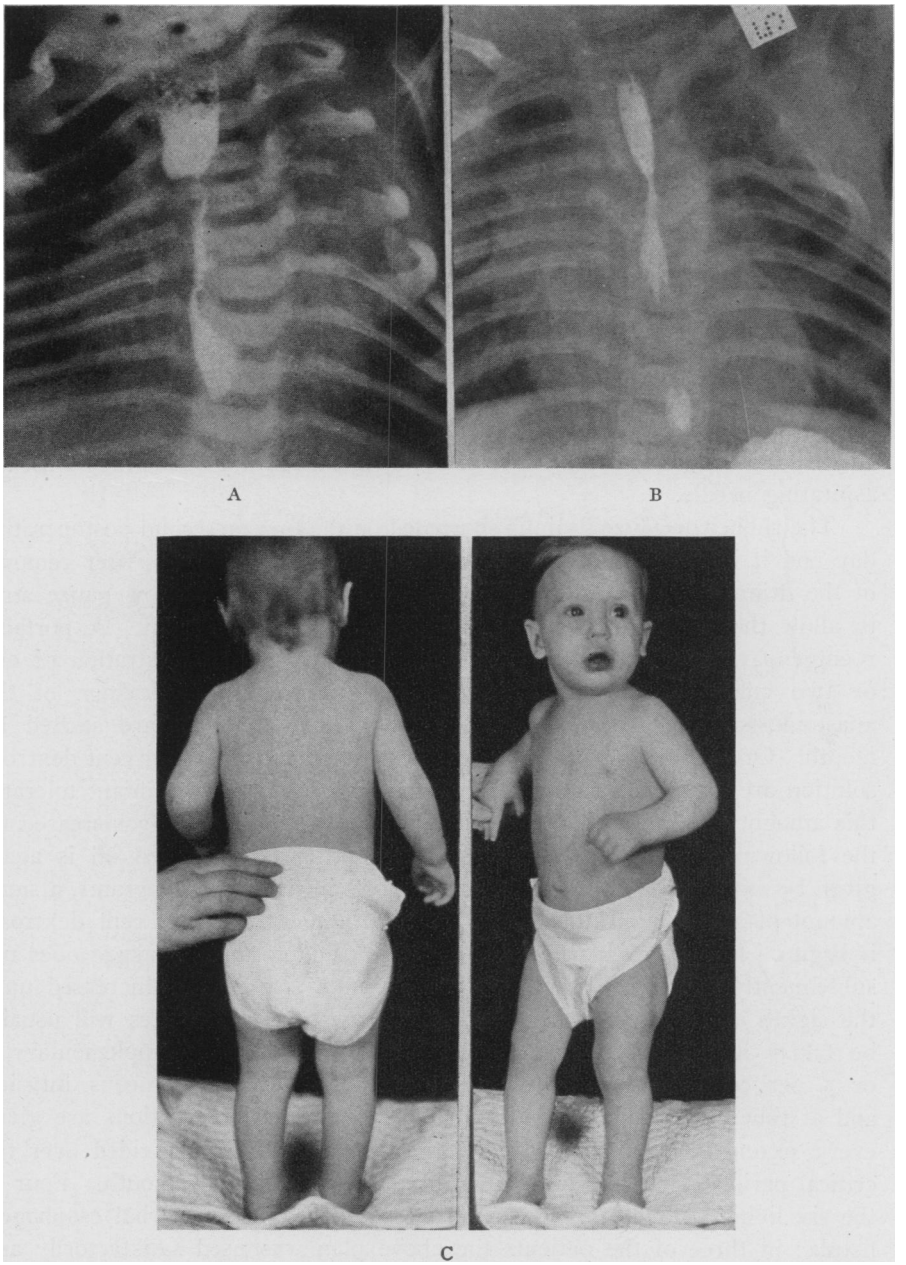

FIG. 9.—S. S., Case 19.—(A) Portable roentgenogram after ingestion of iodized oil five days after left extrapleural ligation of tracheoesophageal fistula and primary anastomosis of esophageal segments. Leakage of the anastomosis did not occur. The upper esophagus is dilated and the lumen at the site of the anastomosis is narrow. All feedings were continued by mouth. Gastrostomy and esophageal dilatation were not necessary.

(B) Roentgenogram 9.5 months after operation. Spontaneous enlargement of the esophageal lumen has occurred at the site of the anastomosis. The upper esophagus is not dilated. In subsequent roentgenologic studies (not illustrated) iodized oil passed through the esophagus so rapidly that satisfactory roentgenograms were not obtained. A slight narrowing was evident at the site of the anastomosis.

(C) Photographs of patient one year after operation show the scar of the healed left extrapleural operation. A gastrostomy has not been necessary.

desired amount of a 0.5 per cent solution of sulfadiazine. Physiologic saline solution is not used, except for the minimal amount required in conjunction with blood transfusions. The onset of edema of the extremities is an indication for a transfusion of whole blood. Blood is used in preference to plasma, as the possibility of producing pulmonary edema appears to be greater when plasma is employed. One of the early cases in this series died from pulmonary edema resulting from the administration of an excessive amount of physiologic saline solution, and a more recent patient died from pulmonary edema following a plasma transfusion.

A portable roentgenogram of the chest is made daily during the first several days after operation. The infant is placed in a sitting position during the exposure of the film. If residual air is present within the pleural space, the indication for withdrawal of the air will depend upon the amount that is present. A moderate amount should be removed; a small amount should probably be allowed to remain because of the danger of injury of the lung with the aspirating needle.

The rubber or gauze drain is shortened on the first or second postoperative day and it is removed completely on about the fourth day. After removal of the drain, the drainage track is packed daily with a narrow gauze strip to allow the wound to heal satisfactorily by second intention. A portable roentgenogram is made on the second day after the administration of one or two cubic centimeters of iodized oil by mouth. If leakage of the anastomosis is not evident, fluids in very small amounts are started by mouth. Ordinarily only one or two cubic centimeters of a 5 per cent dextrose solution are given at hourly or two hourly intervals. If the infant tolerates this amount of fluid without difficulty, the amount is gradually increased on the following day. On the fourth postoperative day, iodized oil is again given by mouth and, if leakage is not apparent in the roentgenograms, a small amount of a formula, diluted with an equal amount of 5 per cent dextrose, is begun. If the formula does not produce choking and if leakage does not subsequently occur, the amount of the formula is gradually increased until the eighth or tenth day when one, or one and one-half ounces will usually be taken at three-hourly intervals. During this period supplementary 3 or 5 per cent dextrose solution is given daily by subcutaneous infusion and at times by rectal instillation. Supportive blood transfusions are given every second or third day. In this way the infants can be tided over the critical period of an insufficient fluid and caloric intake by mouth. Four of the six living patients in this series did not develop an external esophageal fistula; in three of the patients the above plan was used satisfactorily and a gastrostomy was not needed. In the fourth patient a gastrostomy was required because of a recurrence of a tracheoesophageal fistula.

As soon as the infant's condition permits, a roentgenoscopic examination of the reconstructed esophagus is done. Iodized oil is again used and its progress through the esophagus is observed. During the first few weeks after operation, a delay in the passage of oil is present at the site of the

anastomosis, even though a definite stricture may not be present. The narrowness of the lumen at the level of the anastomosis and of the esophagus below the anastomosis apparently accounts for the delay, inasmuch as spontaneous enlargement of the lumen and a more rapid passage of the opaque medium occur during the succeeding weeks (Fig. 9). Later, the passage of the iodized oil has been so rapid in several cases that it has been necessary to use a thick barium paste in order to obtain satisfactory roentgenograms. The roentgenoscopic examination is repeated at intervals, however, in order to ascertain whether a stricture is developing.

When an external esophageal fistula develops as a result of leakage of the anastomosis, a gastrostomy is required for feeding. A gastrostomy, however, subjects the patient to increased risks and it has been an important cause of death in those patients who have required it. One infant died ten hours after the gastrostomy, and no cause of death could be determined other than the fact that the infant had been subjected to an additional operation and had not had a transfusion in conjunction with the gastrostomy. Three infants died of hemorrhage from the region of the gastrostomy. The hemorrhage in one case occurred from ulceration of the umbilical vein and in another instance it resulted from ulceration of the gastric wall in the region of the gastrostomy tube. The third patient died four months after the gastrostomy operation from malnutrition resulting from leakage around the tube and from a terminal hemorrhage from the stomach. Leakage of the oral feedings around the gastrostomy tube and prolapse of the gastric wall adjacent to the gastrostomy are present in one of the living patients. In this case a plastic closure of the gastrostomy will be needed, but this operation has not yet been undertaken because a continuous thread is being kept in place for retrograde dilatation of an esophageal stricture. In the past a Stamm–type of gastrostomy has been used and the catheter has been brought out through the incision. In the last patient in this series, operated upon by Dr. John Alexander, the catheter was brought out through a stab incision. This plan appears preferable because of the thin abdominal wall in infants and the tendency for local disruption of the wound around the catheter when the latter is brought through the main incision. A Witzel-type of gastrostomy is recommended by Ladd and Gross as a satisfactory type of gastrostomy for infants.

Spontaneous healing of an external esophageal fistula occurred in the three patients who survived this complication, but the development of a partial stricture accompanied the process of healing (Cases 10, 22 and 23). In Case 23 a complete stricture occurred after the primary operation, and a partial stricture occurred after correction of the stricture at a secondary operation. As the extent of the stricture can not be determined in advance, the infants who develop an external esophageal fistula should be given a fine silk thread to swallow as soon as the thread can be recovered safely from the stomach by way of the gastrostomy opening. The thread is kept in place for subsequent retrograde dilatations in the event that they are necessary.

A leakage of the anastomosis with a resultant external esophageal fistula is a serious complication with regard to the feeding of the infant. The feedings are given through the gastrostomy tube, but the normal tendency of an infant to regurgitate gastric feedings into the esophagus results in the loss of more or less of the feedings through the external fistula. The gastrostomy feedings will, in such patients, need to be given in smaller amounts and at more frequent intervals than would otherwise be the case, in order to reduce the tendency to regurgitation. In three patients this plan was successful in the prevention of any significant loss of the feedings. In three other patients this plan was not successful and the resulting malnutrition was the chief cause of death. In one of these cases unsuccessful attempts were made to insert a catheter through the track of the gastostomy opening and into the duodenum for the purpose of instituting duodenal feedings. On repeated attempts, the catheter could be guided into the pylorus but it could not be made to enter the duodenum. This maneuver was possible, however, in another patient who developed malnutrition from leakage around the gastrostomy tube, but the patient died before the malnutrition could be corrected. The use of a jejunostomy rather than a gastrostomy has been considered, but has not been employed in the cases in this series. A jejunostomy would not permit retrograde dilatations although it could be used to provide adequate nourishment until the external esophageal fistula had had an opportunity to close.

RESULTS

A reconstruction of esophageal continuity for congenital atresia is an extensive operation for a new-born infant, even when the circumstances pertaining to the operation are entirely favorable. The ability of the infant to withstand an exploratory operation will be influenced largely by its size and general condition on admission. The possibility of restoring esophageal continuity will depend upon the nature of the anatomic findings. As these factors are uncontrollable, they will prevent the operation from being attempted in some cases and in other cases, they will prevent an anastomosis from being completed. In this series of cases, the poor general condition of the patient prevented operation from being undertaken in four, or 14.3 per cent, of the 28 patients who have been seen during the period that the operation of primary anastomosis has been employed. An anastomosis was found to be possible in 17, or 70.8 per cent, of the 24 patients for whom an exploration of the anomaly was performed, and an anastomosis was made in 16, or 66.7 per cent, of the 24 patients.

The details of operative technic and preoperative and postoperative care are factors which are controllable. As an improvement in the management of these cases can be expected, the results in subsequent cases should be superior to those obtained to date during the initial experiences with this operation. In this series of cases, an increase in the recovery rate of the patients for whom an anastomosis was possible occurred with increasing experiences (Table I).

The improved results in the past year have been due to several factors. The use of larger blood transfusions in the replacement of operative blood loss and the use of only the minimal amount of physiologic saline solution that has been necessary in conjunction with the blood transfusions have resulted in a striking decrease in the incidence of postoperative edema, whereas in the earlier cases, edema occurred in all patients who survived the operation for a period of over two days. The difficulty of securing an anastomosis without subsequent leakage has been partially overcome by the

TABLE I

CONGENITAL ATRESIA OF THE ESOPHAGUS

RESULTS IN 28 PATIENTS DURING PERIOD OF USE OF THE OPERATION OF PRIMARY ANASTOMOSIS

	1939	1940	1941	1942	1943	Total
Number of patients	2	2	4	6	14	4
No operation—poor general condition	—	—	1	1	2	8
Anastomosis not possible	—	2*	—	2	4	8*
Anastomosis	2	—	3	3	8	16
Living	—	—	1	1	4	6
Per cent of operative recoveries following anastomosis	0%	0%	33%	33%	50%	37.5%

* One patient (1940) presented ideal anatomic indications for anastomosis, but anastomosis was not attempted.

Four patients admitted 1935-1938 (before use of anastomosis):
Three treated by gastrostomy—Died
One—no operation —Died

use of the telescopic type of anastomosis. The complications arising from pleural perforation that can not be closed have been obviated in the recent cases by the placing of a small gauze pack against the pleural opening during the operation and at the conclusion of the operation.

An analysis of the causes of death shows that pneumonia occurred in most of the fatal cases. Fat stains of the lungs were used in the cases having autopsies and in several of them a lipid pneumonia was found. Atelectasis of the fetal or acquired type was found in five cases in which pneumonia was not present at autopsy examination. Acute pulmonary abscesses were noted in one case. The pulmonary complications have often been accompanied by other complications that have been more important in the cause of death. The other complicating factors have included operative shock and insufficient blood replacement, malnutrition resulting from loss of gastrostomy feedings, sepsis from infection of the operative wound, pulmonary edema from excessive amounts of physiologic saline in one case and from a reaction to a plasma transfusion in another case, hemorrhage from the site of the gastrostomy and hemorrhage from the distal esophagus. Table II summarizes the findings and results in all cases of congenital atresia of the esophagus admitted to the University of Michigan Hospital prior to April 1, 1944.

Six, or 37.5 per cent, of the 16 patients for whom an anastomosis was done are living from seven months to three years and one month after operation. The reconstructed esophagus is patent in all instances. The oldest patient (Case 10) has been reported in detail in a previous communication.[5] This case is the first successful one of reconstruction of the esophagus by

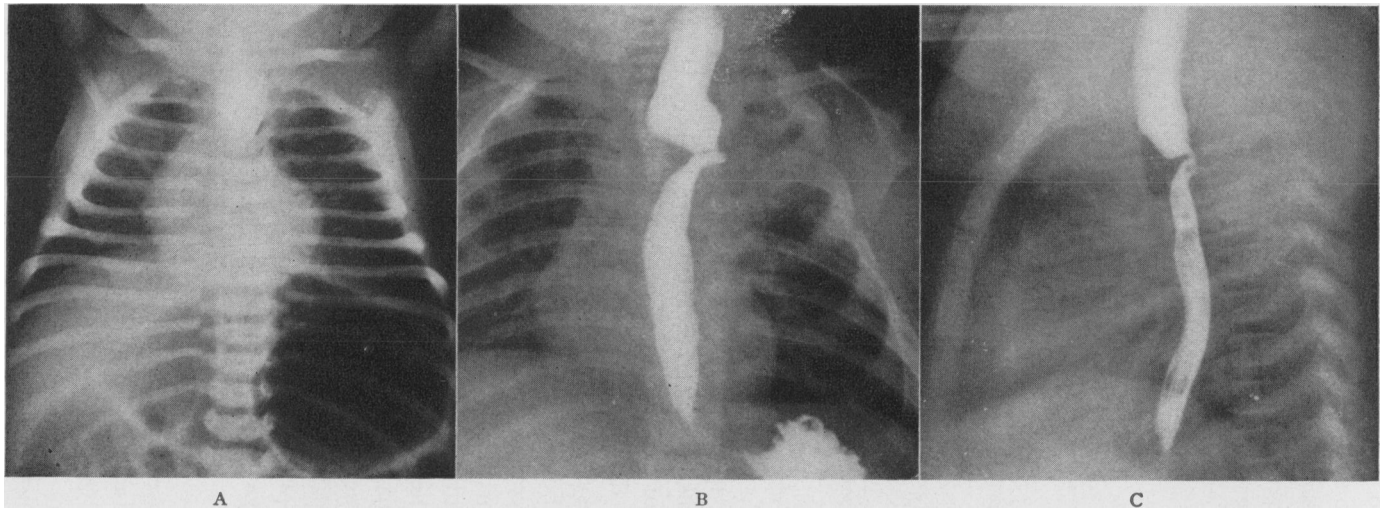

FIG. 10.—J. M., Case 10.—(A) Preoperative roentgenogram showing the dilated blind upper esophagus and the presence of air in the stomach and intestines.

(B) and (C) Roentgenograms following ingestion of sugar four months after operation. The upper portion of the esophagus is moderately dilated, and a short narrow stricture is present at the site of the anastomosis. Leakage of the anastomosis occurred during the second and third weeks after operation, but the external esophageal fistula has healed. (Haight and Towsley,[8] By permission of Surgery, Gynecology and Obstetrics.)

extrapleural ligation of the fistula and primary anastomosis. The patient has developed normally and her weight at the age of three years was 29 pounds. The diet of this patient is not unusual except for the fact that ground meats and sieved vegetables are used instead of coarser foods. In this case a stricture developed subsequent to leakage of the anastomosis, but serial roentgenographic examinations have shown a gradual increase in the size of the lumen at the site of the stricture (Figs. 10 and 11). The stricture was dilated once by Dr. J. H. Maxwell when the patient was 17 months of age. During the first two years after operation, the patient was troubled considerably by accumulations of mucus in her throat but this symptom has greatly lessened during the last year.

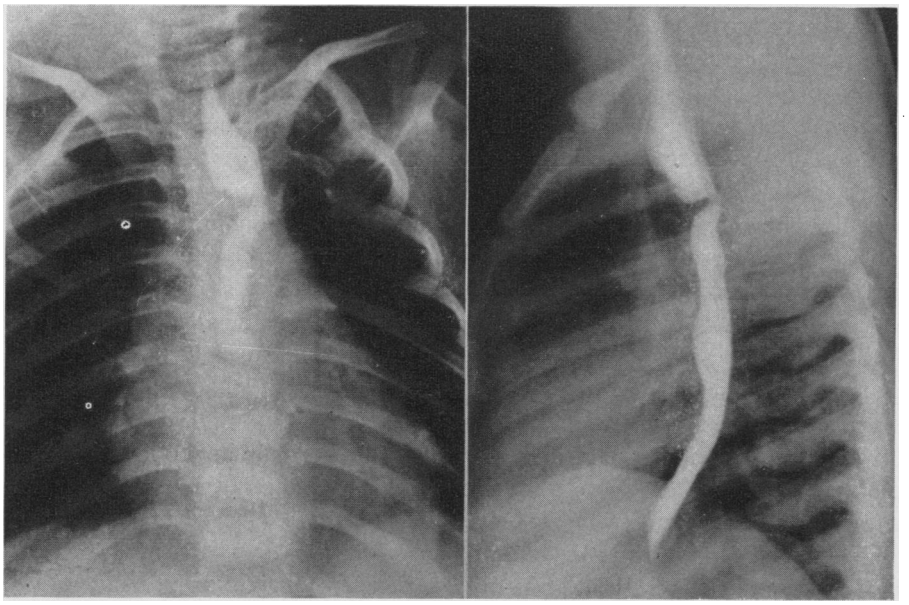

FIG. 11.—J. M., Case 10.—Roentgenograms three years after operation. Progressive enlargement of the lumen of the esophagus has occurred at the site of the anastomosis. (Courtesy of Dr. M. Cooperstock, Northern Michigan Children's Clinic, Marquette, Michigan.)

The next oldest patient (Case 18) is 19.5 months of age. In this case a soft rubber drain, that was introduced into the extrapleural space at the conclusion of the operation, was unintentionally removed on the first postoperative day. The external wound healed *per primam*, but a fistula developed between the esophagus and the trachea as a result of a small leak at the anastomosis and a reopening of the original tracheoesophageal fistula. The lumen of the esophagus remained patent. Gastrostomy feedings were employed for a period of 16 months. The fistula gradually became smaller and it closed spontaneously 14 months after operation. All feedings have been given by mouth for the past two months and the gastrostomy wound is being allowed to close. A bilateral pneumonitis that is present apparently began as a result of secretions within the esophagus having gained entrance into the trachea when

the recurrent tracheoesophageal fistula was still open. The patient's weight is now 16.5 pounds.*

The third oldest patient (Case 19) is now living 14.5 months after operation. In this case the two esophageal segments were smaller than usual, and the lower segment entered the trachea immediately above the bifurcation. This patient's postoperative convalescence was exceptionally satisfactory. Leakage of the anastomosis did not occur, and a gastrostomy was not needed. The patient is able to swallow all foods customarily given to a child of this age, and the present weight is 19 pounds.

In the fourth living patient (Case 22) the anastomosis was less satisfactory than usual, as the anterior wall of the upper segment split in an upward direction during the construction of the anastomosis. A second row of sutures could be used around only half of the circumference of the anastomosis. A large external esophageal fistula developed three days after operation and a gastrostomy was done three days later. The external fistula closed spontaneously six weeks after operation. A very tight stricture developed and feedings by gastrostomy were used for nine months. A thread was subsequently swallowed and is now being used for retrograde dilatations. The patient is now 13 months of age. For the past three months, the feedings consisting of cereals and liquids have been taken entirely by mouth. The gastrostomy opening is unsatisfactory because of its large size and a prolapse of gastric mucosa. The gastrostomy is slowly becoming smaller with the use of a constant pressure dressing over the opening. The weight has increased four pounds during the past month and at present the patient weighs 14 pounds.

The fifth patient (Case 26) (Figs. 12, 13 and 14) is living 10 months after operation. A gap of three centimeters existed between the two segments before the anastomosis was made. Although there was roentgenographic evidence of a small leak at the site of the anastomosis from the fifth to tenth day, an external esophageal fistula did not develop. Feedings by mouth were continued during this interval and a gastrostomy was not necessary. The extrapleural wound was completely healed 10 days after operation, and the patient was discharged from the hospital 35 days after operation. The infant has developed normally and was in excellent condition when last seen at the age of six months.*

The sixth, and most recent living case (Case 30), is now seven months

*Condition on August 10, 1944, of:

1. Case 18. A more recent roentgenologic examination of the esophagus with iodized oil reveals a persistence of the tracheoesophageal fistula. Gastrostomy feedings have been resumed and a plastic closure of the fistula will be required.

2. Case 26. This patient was examined at the age of one year. His general condition continued to be excellent and he experienced no difficulty in swallowing.

3. Case 30. Feedings have been given orally for the past 1.5 months. The wheezing during feedings has largely subsided. The attacks of apnea and respiratory arrest have almost disappeared, only one attack having occurred during the past two months. Esophagoscopic examination shows no evident abnormality of the reconstructed esophagus.

TABLE II

CONGENITAL ATRESIA OF THE ESOPHAGUS
ANALYSIS OF 32 CASES

Case and Year	Operation—Days after Birth	Died—after Operation	Operation	Air in Stomach before Operation	Pneumothorax during Operation	Anomaly Favorable for Anastomosis	Remarks
Case 1 B. G. F. 1935	3 days	22 hrs.	Gastrostomy.	Yes			Premature (8 mos.). Aspiration pneumonia.
Case 2 R. B. 1936	4 days	18 hrs.	Gastrostomy and ligation of cardiac end of esophagus.	Yes			Aspiration pneumonia. Atelectasis.
Case 3 B. B. C. 1937	4 days	5.5 days	Gastrostomy.	Yes			Premature (8-8.5 mos.). Aspiration pneumonia.
Case 4 B. B. S. 7937	No operation	27 hrs. after birth	None.	(No x-ray)			Premature (7.5 mos.). Atresia of esophagus not diagnosed until autopsy.
Case 5 B. B. O. 1939	3 days	17 hrs.	Left extrapleural suture of fistula. Anastomosis of esophageal segments over T-tube.	Yes	Yes	Yes	Upper esophageal segment adherent to and overlapped lower segment. Death due to atelectasis.
Case 6 C. A. B. 1939	7 days	Died during operation	Right intrapleural ligation of fistula. Anastomosis of esophageal segments over T-tube.	Yes	Yes (intentional)	Yes	Respirations became progressively more difficult and shallow after pleural cavity was opened.
Case 7 G. P. 1940	11 days	3 days	Left extrapleural ligation of fistula. Anastomosis not attempted. Gastrostomy.	Yes	No	Yes	Upper esophageal segment adherent to and overlapped lower segment. Death due to overhydration.
Case 8 M. W. 1940	3 days	Died during operation	Right extrapleural exploration. Distal segment not found.	Yes	Yes	No Autopsy	Operation complicated by bilateral pneumothorax and injury to aorta.
Case 9 P. C. 1941	2.5 days	3 days	Left extrapleural ligation of fistula. Anastomosis over catheter. Parietal pleura torn. Air-tight drainage of pleural cavity.	Yes	Yes	Yes	Overlap of esophageal segments present at operation. Death due to sepsis.
Case 10 J. M. 1941	13 days	Living 3 yrs. & 1 mo. after operation	Left extrapleural ligation of fistula. Anastomosis of esophageal segments. Gastrostomy 10 days after operation.	Yes	No	Yes	Leakage of anastomosis. Gastrostomy. Spontaneous healing of external fistula 21 days after operation. Gastrostomy wound healed 1.5 mos. after operation. Progressive enlargement of esophageal stricture.
Case 11 G. W. 1941	No operation	Died 5 days after birth—1 day after admission	None.	Yes	No operation	No Autopsy	Atelectasis present on admission. Condition too poor to permit operation. Atresia of esophagus and air in stomach. Premature.
Case 12 R. W. 1941	6 days	12 hrs.	Left extrapleural ligation of fistula. Anastomosis of esophageal segments.	No	No	No	No air in stomach. Abnormally small distal segment arising from right main bronchus, enlarging to normal size 2.5 cm. below bifurcation of trachea. Open tracheoesophageal fistula found at operation. Anastomosis intact at autopsy. Lobular pneumonia and atelectasis.
Case 13 S. M. 1942	7 days	6 days	Left extrapleural exploration Anastomosis not possible because of contracted lower segment.	Yes	Yes (bilateral)	No	Abnormally small distal segment arising from trachea. Operation terminated without ligation of distal segment because of poor condition. Death due to hemorrhage from acute ulceration of lower end of esophagus.
Case 14 J. T. 1942	No operation	Died 6 days after birth—1 day after admission	None.	No	No operation	No Autopsy	Poor condition of patient prevented operation. Premature.
Case 15 P. G. 1942	Gastrostomy 6 days after birth. Ligation of fistula 15 days after birth	Died 15 days after ligation of fistula	Gastrostomy because of absence of air in stomach. Subsequent left extrapleural ligation of fistula because of eructation of gastric feedings.	No	No	No	Ligation of fistula deferred until 15 days after birth because of atelectasis following gastrostomy. Anastomosis not attempted because of abnormally small distal segment. Death due to hemorrhage from umbilical vein in ligamentum teres at site of gastrostomy.
Case 16 L. B. 1942	11 days	6.5 days	Left extrapleural suture of fistula. Telescopic anastomosis (3 layers).	Yes	Yes	Yes	Lipiodol retained in lungs. Operation delayed 7 days because of bilateral pneumonia and atelectasis, predominated on right. Right approach would have been preferable. Leakage of anastomosis. Death due to pneumonia.
Case 17 A. T. 1942	7 days	2 days	Left extrapleural ligation of fistula. Telescopic anastomosis.	Yes	Yes	Yes	Upper segment adherent to and overlapped lower segment. Death due to pulmonary edema 4 hrs. after plasma transfusion.
Case 18 R. A. 1942	3 days	Living 19.5 mos. after operation	Left extrapleural ligation of fistula. Telescopic anastomosis. Gastrostomy because of recurrent tracheoesophageal fistula.	Yes	No	Yes	Premature (2 weeks). Birth weight, 4 lbs. 15 oz. Primary healing of wound. Recurrent tracheoesophageal fistula closed spontaneously 13 mos. after operation.
Case 19 S. S. 1943	5 days	Living 14.5 mos. after operation	Left extrapleural ligation of fistula. Telescopic anastomosis.	Yes	Yes	Yes	Small upper segment. Low lower segment. No leakage of anastomosis. No gastrostomy.
Case 20 K. M. 1943	4 days	14 days	Left extrapleural ligation of fistula. Telescopic anastomosis. Gastrostomy 13 days after operation.	Yes	Yes	Yes	Leakage of anastomosis. Moderate intra-abdominal hemorrhage and operative shock following gastrostomy. Bilateral atelectasis. Died 10 hrs. after gastrostomy.
Case 21 R. R. 1943	No operation	Died 3 days after birth—1 day after admission	None.	Yes	No operation	No Autopsy	Poor condition prevented operation. Died of progressive pneumonia. Premature; weight, 3 lbs. 9 oz.
Case 22 V. L. 1943	3 days	Living 13 mos. after operation	Right extrapleural ligation of fistula. Partial telescopic anastomosis. Gastrostomy because of leakage of anastomosis.	Yes	Yes	Yes	External esophageal fistula developed 3 days after operation. Retrograde dilatation for tight esophageal stricture. All feedings now taken by mouth.
Case 23 A. S. 1943	2 days	4 mos. after primary operation	Right extrapleural ligation of fistula. Partial telescopic anastomosis. Gastrostomy 8 days after operation.	Yes	Yes	Yes	Leakage of anastomosis and recurrent fistula between trachea and proximal esophagus. External esophageal fistula closed spontaneously. Complete stricture of esophagus.
	3 mos.	1 mo. after reoperation	Reoperation. Ligation of recurrent fistula. One layer anastomosis of esophageal segments.	Yes	Yes (bilateral)	Yes	Leakage of anastomosis. External fistula closed and esophageal lumen patent at death. Died of malnutrition from loss of feedings around gastrostomy tube, hemorrhage from gastrostomy wound and terminal pneumonia.
Case 24 D. M. 1943	3 days	10 hours	Right extrapleural ligation of fistula. Telescopic anastomosis.	Yes	Yes	Yes	Pneumonia on admission. Operation delayed 24 hrs. but pneumonia increased before operation. Died of increasing pneumonia.
Case 25 R. R. 1943	5 days	14 days	Right extrapleural exploration. Lower segment not present. Gastrostomy 5 days after operation.	No	No	No	Agenesis of lower segment. Lobular pneumonia, lipid pneumonia and atelectasis at autopsy. Atresia of gallbladder.
Case 26 F. B. 1943	8 days	Living 10 mos. after operation	Right extrapleural ligation and suture of fistula. Telescopic anastomosis.	Yes	Yes (bilateral)	Yes	Intravenous cannula became obstructed before operation. Unsuccessful attempt to give blood transfusion into azygos vein during operation. No gastrostomy.
Case 27 L. D. 1943	12 days	6 days	Right extrapleural ligation of fistula. Anastomosis not possible. Esophageal catheter left in place after operation.	Yes	Yes	No	Gastrostomy 6 days before admission. Lower segment arose from right main bronchus. Esophageal segments too far apart to permit anastomosis. Lobular pneumonia, horseshoe kidney and single ureter.
Case 28 J. S. 1943	Admitted 3 days after birth	Died 6 hrs. after admission	None.	Yes	No operation	Yes	Bilateral pneumonia. Horseshoe kidney. Partial aortic stenosis and patent interventricular septum. Low position of tracheoesophageal fistula.
Case 29 G. S. 1943	6 days	36 days	Right extrapleural ligation of fistula. Satisfactory anastomosis not possible. Esophageal catheter left in place. Gastrostomy 9 days after operation.	Yes	No	No	Low position of tracheoesophageal fistula. Died of malnutrition from regurgitation of gastrostomy feedings into extrapleural wound.
Case 30 J. P. 1943	4 days	Living 7 mos. after operation	Right extrapleural suture of fistula. Telescopic anastomosis.	Yes	Yes	Yes	Upper segment adherent to and overlapped lower segment. Gavage feedings because of attacks of respiratory obstruction after oral feedings. Esophagus patent. No gastrostomy.
Case 31 R. L. 1943	6 days	13 days	Right extrapleural suture of fistula. Telescopic anastomosis. Gastrostomy because of leakage of anastomosis.	Yes	Yes	Yes	Leakage of anastomosis. Malnutrition from regurgitation of gastrostomy feedings into extrapleural wound. Fetal atelectasis, pulmonary abscesses, gangrenous appendicitis and hemorrhage into intestinal wall.
Case 32 J. S. 1943	5 days	1 day	Right extrapleural exploration. Lower segment not present.	No	Yes	No	Agenesis of lower segment. Pulmonary infiltration before operation. Bilateral atelectasis. Full term twin. Weight, 4 lbs. 6 oz.

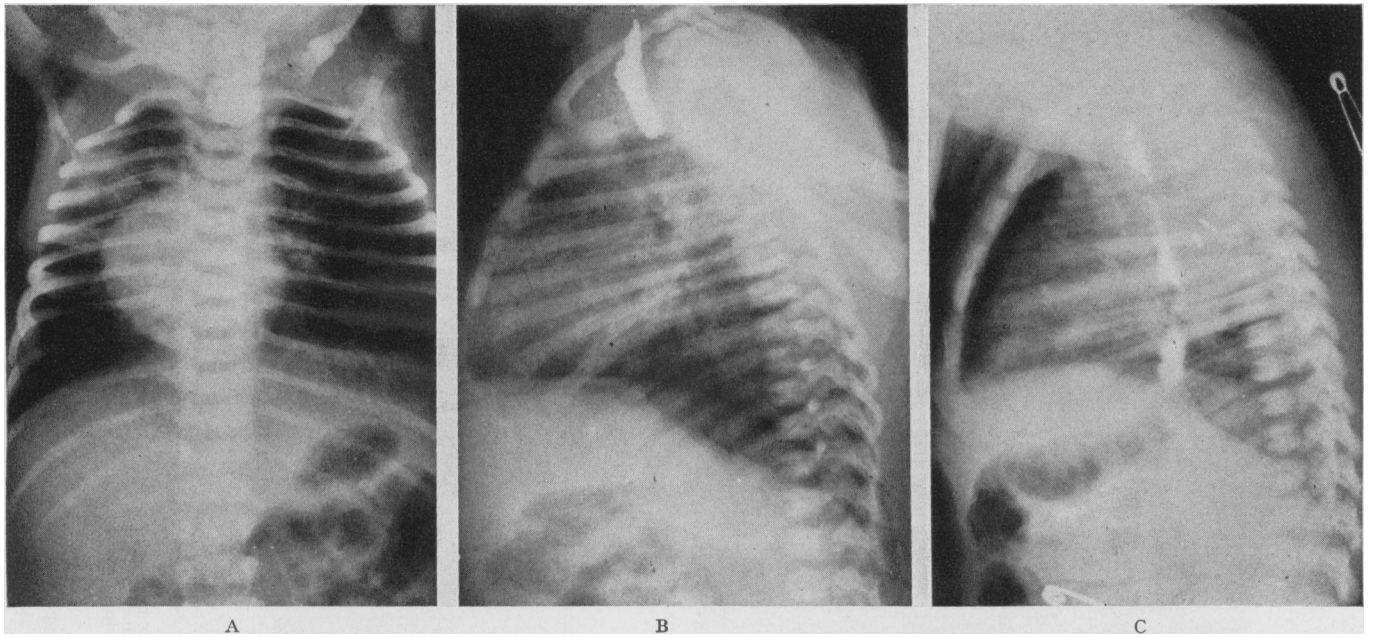

FIG. 12.—F. B., Case 26.—(A) and (B) Roentgenograms after ingestion of iodized oil show the upper esophageal segment to be smaller than usual. Air is present in the stomach, demonstrating the presence of a tracheoesophageal fistula. Congenital dextrocardia.

(C) Portable lateral roentgenogram two days following right extrapleural ligation of fistula and primary anastomosis. The iodized oil demonstrates the esophagus to be patent, although the lumen is narrow at the site of the anastomosis. A small, temporary extravasation of oil was noted on roentgenograms (not illustrated) on the fifth and tenth postoperative days, but an external fistula did not occur. Feedings by mouth were continued and a gastrostomy was not done.

old. The patient has not been discharged from the hospital because respiratory difficulties occur with oral and gavage feedings. In this case the anastomosis healed without the development of an external fistula, although a very small temporary leakage of the anastomosis was seen in the roentgeno-

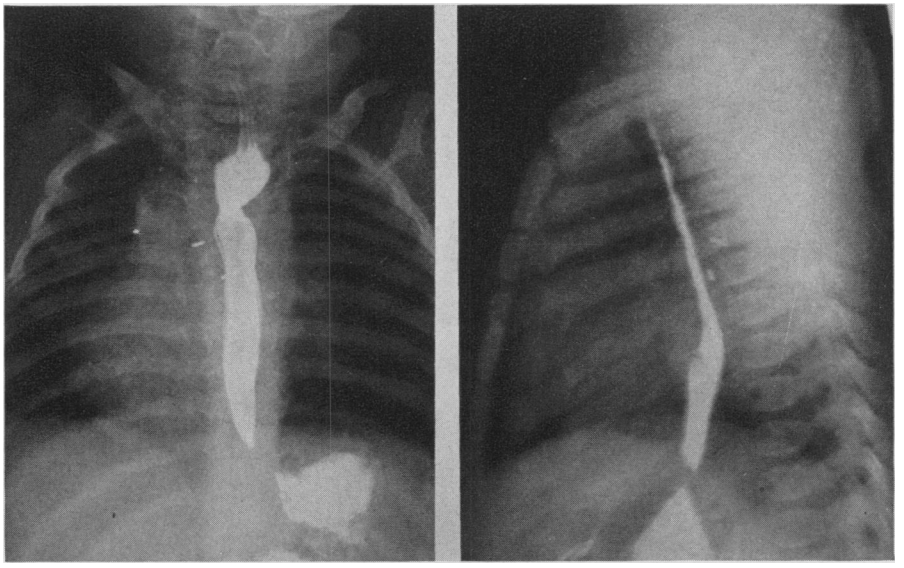

FIG. 13.—F. B., Case 26.—(A and B) Roentgenograms, six months after operation show the lumen of the esophagus to have enlarged. There was no delay in the passage of rugar through the esophagus.

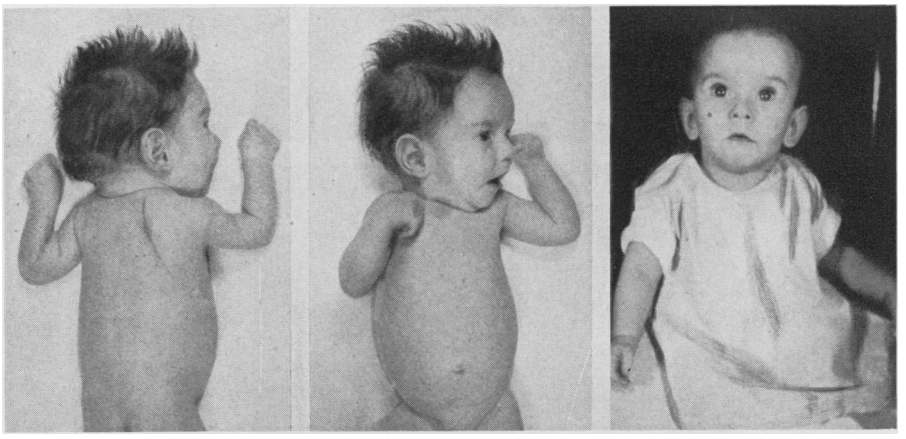

FIG. 14.—F. B., Case 26.—(A and B) Photographs of patient one month after operation. The healed right parascapular incision is seen. A gastrostomy has not been needed.
(C) Photograph of patient six months after operation.

grams. Roentgenoscopic examination shows only a slight delay in the passage of lipiodol through the region of the anastomosis, and a No. 18 F. catheter can be easily introduced into the stomach. Wheezing occurs during all feedings, but it is more severe during oral feedings. The respiratory

difficulty is characterized by apnea, cyanosis and respiratory arrest which occasionally occur shortly after the conclusion of a feeding, whether it be by the oral route or by gavage. Immediate resuscitative measures are needed to restore respirations. As the gavage feedings provoke the symptoms less often than do the oral feedings, the gavage feedings have been used almost exclusively and are being continued. The explanation for the respiratory attacks is not clear. A recurrence of the tracheoesophageal fistula has been considered as a possible cause for the symptoms, but the roentgen examinations reveal no evidence of a fistula. A faulty mechanism of deglutition has also been considered as a possible cause, although the epiglottis appears to be normal on laryngoscopic examination.*

SUMMARY

The anatomic and surgical problems concerned with the correction of congenital atresia of the esophagus with tracheoesophageal fistula are presented. Reconstruction of esophageal continuity by a single-stage operation consisting of an extrapleural closure of the tracheoesophageal fistula and an anastomosis of the esophageal segments offers the most satisfactory approach to the correction of the anomaly. A primary anastomosis of the upper and lower esophageal segments was done in 16 of the 24 patients for whom an exploration of the anomaly was undertaken. Six, or 37.5 per cent, of the 16 patients for whom an anastomosis was performed are living from seven months to three years and one month after operation, and the reconstructed esophagus is patent in all instances.

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DISCUSSION.—DR. WILLIAM E. LADD, Boston: Doctor Haight justly remarks that congenital atresia of the esophagus has proven amenable to surgical correction only in very recent years. As far as I know, Doctor Leven, of St. Paul, has the oldest living patient, and my oldest living patient is 24 hours younger than his. They are both now over four and one-half years old.

Doctor Haight is certainly to be congratulated on his presentation of the subject and on his splendid results. He has made a real contribution to this baffling problem. I am in complete sympathy with his idea that extrapleural ligation of the fistula with primary anastomosis is the operation of choice when feasible. The feasibility of the plan depends on the findings in the individual case, and on the surgeon's ability. The high percentage of cases that Doctor Haight considers feasible for primary anastomosis makes it unnecessary to comment on his ability.

At The Children's Hospital we have had the comparatively large series of 72 cases. Up until 1939 the experience was valuable largely for furnishing pathologic material, and for teaching us what not to do. Since that time, however, there has been a rift in the clouds, and we now have 11 living patients. During the past year we have operated upon 13 patients, of whom eight are living. In five of these patients a primary anastomosis was attempted, and two are living. In the other eight patients the fistula was tied off, an esophagostomy and gastrostomy performed, and six of these eight patients are living. Two of this latter group were a little premature, weighing 4 pounds 6 ounces and 4 pounds 8 ounces, respectively. In our oldest patient, now 4.5 years old, the anterior esophagus is continuing to function satisfactorily.

I believe there always will be some patients in whom a primary anastomosis will not be feasible. We have had at least two patients in whom the lower segment of the esophagus extended not over a half inch above the diaphragm and the upper segment was not below the second or third dorsal vertebra. One of these patients is among our group of living cases.

From data at hand to date, it would seem that the multiple-stage operation is a considerably safer procedure, though in some ways less desirable. The advantages of primary anastomosis make this operation preferable in patients in whom the two segments are close together.