Tracheo-Esophageal Fistula and Esophageal Atresia

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Congenital atresia of the esophagus was first described in 1697 by Gibson, who was a grandson of Oliver Cromwell, and physician-general to the army. Neither the clinical history nor the physical findings have changed since his often quoted classical description. It was 245 years later before success was recorded in the management of the condition. Cameron Haight in 1941 first established esophageal continuity successfully in one stage operative procedure, by primary anastomosis of the proximal and distal segments of the esophagus.

The delayed progress in the treatment of the condition was due, in the past, to the misconception that the condition was extremely rare. Guthrie, in searching the records of the Royal Hospital for Sick Children, Glasgow, found 24 proved, and a further six probable, cases in 6,916 necropsies. The late Sir G. Grey-Turner estimated that the malformation occurred about as frequently as hare-lip and cleft palate. At the Postgraduate Medical School of London four cases of atresia were found in 10,543 deliveries (1 in 2,635 deliveries).

It is quite conceivable that the incidence is higher than 1 in 2500 births. Many cases probably are not recognized as such and the deaths are regarded as having been due to atelectasis or pneumonia, without a confirming autopsy.

That the condition can be overlooked is easy to understand, particularly when the infant is not supervised by a pediatrician. The early signs are not always definite or even recognizable. The infant brings up some frothy mucus during the first day or two of life. Attempts at feeding lead to regurgitation, choking and probably cyanosis. Excess of mucus in the naso-pharynx occurs in normal infants, but aspirations soon relieve any cyanosis which may have been present.

Without the diagnosis and treatment of atresia of the esophagus, pulmonary symptoms usually develop by the second day. This may be diagnosed as pneumonia, and further feeding troubles and respiratory distress attributed to the respiratory infection. With the fatal termination in a week or more, even if necropsy is performed, it is quite conceivable that the diagnosis may be overlooked. The burden of early recognition falls largely on the obstetrician, the pediatrician, and the nursing personnel of the new-born nursery. If they are suspicious regarding any child who has excess mucus, episodes of cyanosis, and who does not take fluid properly in the new-born period, more of these cases will be recognized earlier than at present, before a great deal of aspiration has occurred. There is no real substitute for awareness and careful observation.

Although the diagnosis may be made frequently on history alone, physical signs and X-ray film examination are usually necessary to confirm it. The diagnosis is confirmed, when suspected, by the passage of a soft rubber No. 10 or 12 F catheter through the mouth into the pharynx and proximal esophagus. The catheter will be held up 8 to 10 centimeters from the alveolar margin in the usual type of atresia.
To gain positive proof of the anatomic abnormality which is present, a few simple roentgenographic studies are helpful. A straight antero-posterior film of the chest is done to determine the state of the lung fields. Almost all show signs of atelectasis or aspiration pneumonia, characteristically in the right upper lobe. The catheter is again passed into the upper esophagus, and 0.5 to 1.0 cc of iodized oil is injected and its course followed by fluoroscopy. The iodized oil is left in position for a short time to see if any passes through an upper fistula, then is withdrawn. Barium should never be used, as aspiration into the air passages results in a chemical type of pneumonia. The presence of air in the gastrointestinal tract, as shown by x-ray film, is clear-cut evidence of the presence of fistula between the tracheo-bronchial tree and the lower segment of the esophagus. Its absence, however, though suggestive, is not a sure sign of the absence of such a fistula. Fistulae between the upper pouch and the trachea are usually impossible to demonstrate, except by operative dissection.

**Embryology**

To understand the development of congenital tracheo-esophageal fistula and other anomalies of the esophagus it is necessary to review briefly the embryology of the trachea and esophagus. Both these structures develop from part of the foregut. By about the fourth week a definite esophagus has developed as a short tube extending from the pharynx to the stomach. At seven weeks the epithelium thickens by proliferation, then vacuoles appear in it, and in this way the lining becomes irregularly channeled. In the embryos of 3 mm. the laryngotracheal groove appears in the floor of the gut, just caudal to the pharyngeal pouches. This groove deepens and splits off the primordium which is to become pharynx, trachea and lungs. With the close association of the trachea and esophagus in development, it is easy to see how tracheo-esophageal fistula can occur.

**Conditions Which Influence Survival and Prognosis**

**Associated Anomalies:** The possibilities of other anomalies associated with atresia of the esophagus must always be considered. Probably the
majority of these babies have no other important anomalies, but a considerable group have abnormalities which are of an equally serious nature. Congenital heart disease, malformations of the anus and rectum, duodenal obstruction and Meckel's diverticulum are among the commonest associated lesions. Recognition of their existence is imperative to plan a successful surgical attack. Gross found in 233 patients with atresia of the esophagus, 77 had various anomalies, some babies had several. Humphreys et al. in 136 patients found 75 with other congenital malformations, and two-thirds of the infants who died post-operatively were handicapped by other anomalies. Ladd and Swensen (1947) found that of their 82 cases, only 18 had serious additional abnormalities, and these were chiefly in the heart.

Obviously, secondary operations in these precarious infants raises the mortality rate, but from a perusal of the literature, a considerable number of infants have survived the correction of the additional deformities.

Prematurity: There seems to be a higher incidence of prematurity with this type of anomaly than in other malformations of the body. Approximately 25 per cent of these infants are less than five pounds in weight. Prematurity added to pulmonary complications greatly increases the risk of surgery to these vulnerable infants. Humphreys et al. attempted in their 136 cases to relate mortality following surgery at birth weight. They found that prematurity, as judged by weight, though a determining factor at the lower extremes of size, did not preclude survival.

Delay in Diagnosis: This is probably the largest single factor in contributing to the mortality rate of the condition. An infant who comes to major surgery exhausted, and with an overwhelming pulmonary infection, is obviously a bad risk, in spite of the best pre-operative preparation. Probably most infants where the diagnosis has been delayed for four days or more, will not survive surgery, in spite of all measures. Cases that are diagnosed in the first few hours or the first day, should be brought to operation immediately. After the first day, much can be gained by an appropriate period of preparation, to get the infant into better condition. Adequate time should be expended to correct ketosis, dehydration and pulmonary infection. This period may extend up to 12 hours. Awareness of the condition, early diagnosis, and prompt instigation of pre-operative

FIGURE 2: The common type of esophageal malformation. There is esophageal atresia. The upper pouch ends blindly. The lower pouch communicates with the back of the trachea, just at or slightly above its bifurcation. (Courtesy from Gross: Surgery of Infancy and Childhood, W. B. Saunders Company, Philadelphia, 1953.)
measures will greatly enhance the infant’s chance of successful surgery. The mortality will be less if the operation is done as soon as possible after birth.

Pre-operative Care: This is most important, and a pediatrician should be in attendance. When the condition is suspected, after passage of a soft rubber catheter into the proximal esophagus, it is our opinion, from experience with over-zealous radiologists, that opaque media should not be used to confirm the diagnosis. Iodized oil that overflows or is introduced into the tracheo-bronchial tree is hazardous to an already embarrassed system. One can obtain a satisfactory contrast x-ray film by simply distending the upper pouch with air and avoiding soilage of the lungs.

The infant is placed in an incubator or isolette and moist oxygen administered with controlled temperature. The pharynx is suctioned with a soft rubber catheter every 15 to 30 minutes. The potential of the bacterial content in the upper stump of the esophagus should be emphasized. It often has a high content of anaerobic streptococci. Penicillin and streptomycin plus a wide spectrum antibiotic should be administered. Of recent years we have been making more frequent use of tracheal suction in order to prepare these cases for surgery and carry them past the crucial phase in the post-operative period. Respiratory complications are among the main lethal factors: and in their debilitated condition they are unable to satisfactorily clear their tracheo-bronchial tree. By means of a laryngoscope and catheter suction, introduced through the vocal cords, a simple and effective means is available to protect and drain the respiratory system and reduce mortality.

There is a good argument for nursing these patients in the Fowler’s rather than the Trendelenburg position. The gastric secretion in the first few hours of life is highly acid, and the chemical pneumonia it produces on gaining entrance to the tracheo-bronchial tree through the distal fistula is added to that pneumonia already present from aspirated saliva and other liquids.

Adequate hydration should be maintained by intravenous fluids, which also correct ketosis. Great care should be taken to avoid overhydration. Infants that are jaundiced should receive vitamin K pre-operatively. Blood is obtained to the amount of 15 to 20 cc per pound, in preparation for operative procedure.

Operative Procedure

Provided a skilled anesthetist is available, intra-tracheal anesthesia, controlled respiration and a right trans-pleural approach, offers certain advantages over other methods of attack. Certainly this approach offers better visualization of the segments and an easier, time-saving anastomosis. It gives an excellent view from the top to the bottom of the chest, and it allows the widest possible dissection of the esophagus. It also permits opening of the diaphragm and mobilization of the stomach when this is required.

A standard postero-lateral incision is made, entering the pleural cavity through the right fourth interspace. The mediastinal pleura is entered by dividing the azygos vein, the bifurcation of the trachea is easily identified, and the fistulous communication with the distal esophagus dissected free. The distal esophagus may be mobilized to the hiatus of the diaphragm if
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Post-operatively, A a frequent cause of pulmonary edema and fatality in these cases. The gastrostomy tube is left open for 24 to 48 hours, to keep the stomach deflated. On the second or third day feedings can be cautiously started through the gastrostomy tube, starting with 10 per cent glucose and water in small quantities. Oral feedings are begun on the sixth to the eighth day, after satisfactory conditions are demonstrated by a iodized oil swallow.
The gastrostomy tube should be removed only when oral feedings have been well taken for at least one week and radiologically there is an adequate esophageal lumen.

Post-Operative Stricture

It is difficult to predict which subjects are going to develop stricture at the anastomotic site after discharge with a satisfactory radiological picture. That it can be a serious late complication is illustrated by fatalities in two of our cases after an excellent early surgical result. The later development of stricture may be due to prior leakage and mediastinitis. When the anastomosis has been done under great tension or the union has been poor, presumably stricture formation is likely to develop. It has generally been found that narrowing at the point of anastomosis has been most troublesome within the first year of life, requiring often, many dilatations. After this period the stricture seems to grow adequately with the esophagus, to necessitate only infrequent dilatations.

Early strictures that are visualized before the gastrostomy tube is removed are best treated by retrograde dilatation with small soft Tucker's dilators. Later strictures should be inspected under general anesthesia to determine the character of the tissue at the stricture site and subsequently dilated with a gum webbing type of dilator, passed after an attached filiform leader.

Most strictures require multiple instrumentation, though some are well treated by one dilatation. Occasionally a more radical attack is necessary,
and excellent results have been obtained from a secondary operation, where removal of scar tissue and a re-anastomosis were performed, usually between one and two years of age.

Results

In 1956 we treated surgically 10 cases of tracheo-esophageal fistula. There were six long-term survivors of this group, although one of these subsequently died of acute staphylococcal pneumonia at the age of three months. Two of the deaths were due to stenosis at the site of anastomosis, occurring at six weeks and two months respectively. In one of these there was a sudden onset of dysphagia in an infant who had previously been swallowing quite well. The iodized oil swallow in this baby showed an almost complete obstruction. Death occurred from pneumonia after dilatation under general anesthesia. The second one had intermittent difficulty in feeding, with aspiration of feedings. A recurrence of fistula was suspected, but lipiodol swallow showed what appeared to be a normal esophagus. Death was due to pneumonia, and at autopsy a partial obstruction was found by a baffle of fibrous tissue at the site of anastomosis. This baby had developed a post-operative disruption of the suture line of the esophagus, with tension pneumothorax, which had been controlled by the insertion of an intercostal catheter. The other patients not surviving the immediate post-operative period died of pneumonia. One of these was operated on two days after birth, and the other eight hours after birth. Both were premature infants, and at autopsy both were found to have multiple abnormalities, including duodenal atresia in one case, and a large atrial septal defect in the other.

It is probably reasonable to conclude that there were in this series, 60 per cent survivors of the operative procedure for correction of congenital tracheo-esophageal fistula.

FIGURE 4: Pre-operative radiographic picture of Case 2. Note the unusual length of the upper pouch.
Of the surviving group there were two unusual cases, which are to be reported in detail. One of these is a type which, to our knowledge, has not been previously described.

Case Reports

We were called in consultation on May 17th, following the iodized oil swallow which resulted in the radiological picture (Fig. 3). The infant’s history had been briefly as follows: The second day after birth the attending nurse noticed the infant had stertorous breathing, and seemed to have excessive mucus. Examination at this time revealed moist rales in both lung fields, which were thought to be due to aspiration or atelectasis. Feedings resulted in moderate cyanosis, therefore the infant was fed by gavage of breast milk and supplemented with interstitials of glucose and saline. Terramycin was started on the third day of life.

This regime improved the infant’s condition to some degree. However, on May 11, cyanosis became troublesome at gavage, and the infant was placed in a croupette. Abnormal amounts of mucus secretion and respiratory difficulty were prominent features until operative repair.

The x-ray film on May 17, easily established the diagnosis of tracheoesophageal fistula, without disruption of the continuity of the esophagus. Due to bilateral aspiration pneumonia and poor general condition of the infant, it was necessary to spend a further three days in trying to improve its desperate condition.

Surgery was performed on May 20, as rapidly as possible. As the fistula was high, opposite the first and second thoracic vertebrae, the upper four ribs were exposed through a right postero-lateral incision. The second rib was resected subperiosteally in the main, and the first and third ribs were shingled posteriorly. By gentle dissection, the parietal pleura was dissected off the chest wall and the apex of the lung depressed. The esophagus and trachea were cleaned and the fistula found with some difficulty. It ran tangentially downward from the esophagus to the trachea, opposite

FIGURE 5: Schematic representation of the findings at operation in Case 2. It was realized after operative exposure that a right sided approach would have been easier. The aortic arch interfered with easy division of the fistula and closure of the same. However, from the pre-operative radiologic picture there was a suspicion of an atretic area of some length of the lower esophagus.
the first thoracic vertebra. It was divided and the ends closed with fine interrupted silk sutures. Following closure of the chest wall the post-operative condition was better than we expected. Intravenous fluids were continued until the fifth day and on the fourth day small amounts of sterile glucose feedings were taken by mouth. Subsequently, to our amazement, the child progressed rapidly toward normality. Although there had been a delay of fifteen days before the operative procedure, respiratory complications post-operatively were not of serious concern. A gastrostomy was not indicated in this case, we felt, because of the normal continuity of the esophagus.

The child was discharged from hospital on June 6, in good condition and has remained so since, gaining and developing normally.

Baby L.: This infant was a full term, six pound girl. Delivery was essentially normal, but there was some difficulty in making the child start breathing, and she was taken to the observation nursery. Here it was noted that there was an excessive amount of mucus, necessitating frequent aspiration of the pharynx. A tracheoesophageal fistula was suspected, and a catheter was passed into the esophagus. An obstruction was encountered somewhat lower than in the usual type of tracheoesophageal fistula. A small amount of lipiodol was injected through the catheter, and x-ray films taken. As is seen in Figure 4, the esophageal "pouch" extends into the lower one-third of the thorax. Because of this, it was decided to do a left, rather than a right, thoracotomy. At operation, eight hours after birth, it was found that there was a partial reduplication of the esophagus, with the lower segment communicating with the trachea, as is shown in the diagram. It was a simple matter to establish continuity of the esophagus, and obliterate the remaining fistulous tract into the trachea, with interrupted sutures. The post-operative course was quite uneventful. Oral feedings were commenced with glucose and water on the fifth post-operative day, and formula feedings begun the following day. Follow-up at six months of age showed the child to be normally developed for its age, and having no difficulty in swallowing.

SUMMARY

1. Since the outstanding contribution by Cameron Haight in 1941 in the surgical management of tracheoesophageal fistulae and esophageal atresia, the picture has changed from one of almost invariably fatal outcome, to that of a high survival rate in the surgically corrected infants. Prematurity and associated anomalies are uncontrollable factors in this condition, but do not necessarily, in themselves, preclude a successful outcome. It is the awareness of the condition by doctors and the nursing profession, and the establishment of an early diagnosis that is stressed. If, following early diagnosis, proper pre- and post-operative care (including late post-operative supervision) is maintained, a recovery rate in the neighborhood of 70-80 per cent can be expected, where prematurity and concomitant anomalies are not a factor.

2. Reported here are two rare types of this anomaly, from our series of ten cases in 1966, with a 60 per cent survival rate.

RESUMEN

1. Desde que Cameron Haight en 1941 hizo su notable contribución al tratamiento quirúrgico de la fistula tráqueo-esofágica y en la atresia del esófago el cuadro ha cambiado desde aquella época en que casi había resultado fatal invariablemente hasta ahora que hay un sobrevida en proporción elevada en los niños tratados quirúrgicamente. La prematuridad y las anomalías que le acompañan, son factores no dominables de esta anomalía, pero no por eso impide un resultado satisfactorio. Se recalca la importancia de que los médicos y las enfermeras estén alertas ante esta malformación.

Si después de hacerse el diagnóstico se hace un cuidado pre y postoperatorio bien mantenidos (incluyendo el postoperatorio tardio), es de esperarse una recuperación de 70-80 por ciento cuando no se trata de prematuros y de otras anomalías coexistentes.

2. Se relatan dos casos típicos de esta anomalía de nuestra serie de diez en 1956 con 60 por ciento de sobrevivida.

RESUME

1. Depuis l'éminente contribution de Cameron Haight, en 1941, au traitement chirurgical de la fistule trachéoesophagienne et de l'atrésie oesophagienne, le tableau a changé. Alors qu'autrefois l'issue était presque invariablement fatale, on peut parler maintenant d'un taux élevé de survie chez les enfants traités chirurgicalement. La prematurité et les anomalies congénitales et associées sont des facteurs contre lesquels on ne peut rien dans cet état, mais ne sont pas en elles-mêmes un obstacle à une issue satisfaisante. C'est la nécessité pour les médecins et les personnes qui s'occupent des enfants, de reconnaître cet état et d'établir un diagnostic précoce que souligne l'auteur.

Si, après ce diagnostic précoce, une surveillance convenable, pré- et post-opératoire (comprenant une observation post-opératoire éloignée) est maintenue, on peut espérer un taux de guérison de 70 à 80% environ, à condition que les anomalies congénitales associées ne soient pas essentielles.

2. L'auteur rapporte ici deux types habituels de cette anomalie, d'après une série de 10 cas suivis en 1966, avec un taux de survie de 60%.
TRACHEO-ESOPHAGEAL FISTULA

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REFERENCES