Esophageal Atresia with Tracheo - Esophageal Fistula

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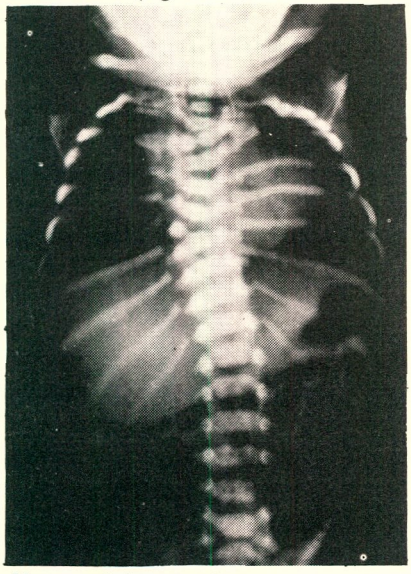
A CASE of esophageal atresia with tracheo-esophageal fistula successfully treated at Salmaniya Medical Center is presented and the management of this condition discussed.

CASE PRESENTATION

H.H.A, a full term male infant, was admitted to Salmaniya Medical Center three hours after birth. A physician, at the Maternity Hospital, noticed excessive salivation at the mouth. An attempt was made to pass a naso-gastric tube but a resistance was encountered 'few inches' from the mouth.

On admission to Salmaniya Medical Center the child weighed 2500 gms and appeared healthy. Apart from a glandular hypospadias no abnormalities could be detected. A film of the chest and abdomen (figure 1) showed the



X-ray of chest and abdomen showing the catheter curled in the upper esophageal pouch. Presence of air in the G.I. tract indicates a distal tracheo-esophageal fistula.

naso-gastric tube curled in the upper esophagus. There was air in the gastro-intestinal tract. The lungs were well expanded and clear. A hemi-vertebra, involving D-4, was noticed. Laboratory determinations were unremarkable. A diagnosis of esophageal atresia with distal tracheo-esophageal fistula was made.

The infant was placed in an incubator. Frequent pharyngeal suction was instituted. No feedings were allowed by mouth. An intravenous line was established and I.V. fluids were started. Intravenous ampicillin was begun.

Twelve hours later the patient was operated and primary correction was done with separation of the tracheo-esophageal fistula and a one layer esophago-esophagostomy. A gastrostomy was established.

The post-operative course was smooth and uneventful. Gastrostomy feedings were started on the sixth post-operative day, the chest tube was removed on the seventh post-operative day. A dionosyl swallow on the ninth post-operative day showed good passage of the dye through the esophagus (Figure 2). There was no leakage and minimal narrowing at the anastomosis was noticed.

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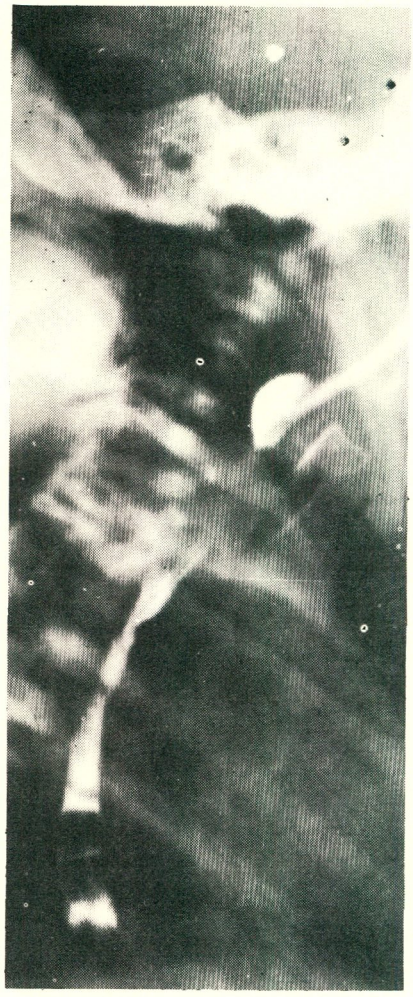


Dionosyl swallow on the 9th post-operative day shows good passage of dye with no leakage and minimal anastomotic narrowing.

Esophageal dilatation to No. 20 French was easily done on the twenty-fifth post-operative day and the gastrostomy tube was removed the same day. The child was discharged on the twenty-ninth post-operative day in very good condition.

He was re-admitted one week later with occasional vomiting which, in the hospital, seemed to be associated with coughing. He settled down quickly and was able to retain his feedings. He was kept in the hospital for ten days and discharged tolerating his feedings well. He has stayed well to this date, two months after operation.

DISCUSSION

Esophageal atresia with or without tracheo-esophageal fistula is a rare congenital abnormality, which, in most of its forms, is incompatible with life. The first account of this condition was in 1670 by Durston. Sporadic reports appeared in the literature subsequently but the first exhaustive report appeared in 1940 by Lanman who summarized his experience with thirty-two consecutive failures in the management of this condition. In 1939 Ladd in Boston and Leven in Minneapolis simultaneously reported on the first successes in the surgical repair of the condition. The procedure consisted of gastrostomy, ligation of the tracheo-esophageal fistula and late extra-thoracic esophageal reconstruction. It was Cameron Haight from Ann Arbor, Michigan who in 1941 achieved the first successful primary repair.

The lungs are formed by an outbudding from the foregut at twenty-one to twenty-six days of fetal life. The trachea is separated from the esophagus by a coronal septum which is complete by thirty-two days. Incomplete separation will result in tracheo-esophageal fistula while epitheleal over-growth in the esophagus with incomplete resolution will result in esophageal atresia.

The abnormality presents itself in five anatomic variants which are illustrated in figure 3.

The most common variety is type A, constituting a little over 85% of all the cases. It consists of a blind upper esophageal pouch and a fistula between the distal esophageal segment and the trachea. The next most common type is B constituting about 8% of the cases. In this type there is an atresia with no tracheo-esophageal fistula. Type C, with no atresia but a tracheo-esophageal fistula (also called H type) comes third in frequency with an incidence of about 4%. The remaining two types are very rare, their incidence being less that 1% each.

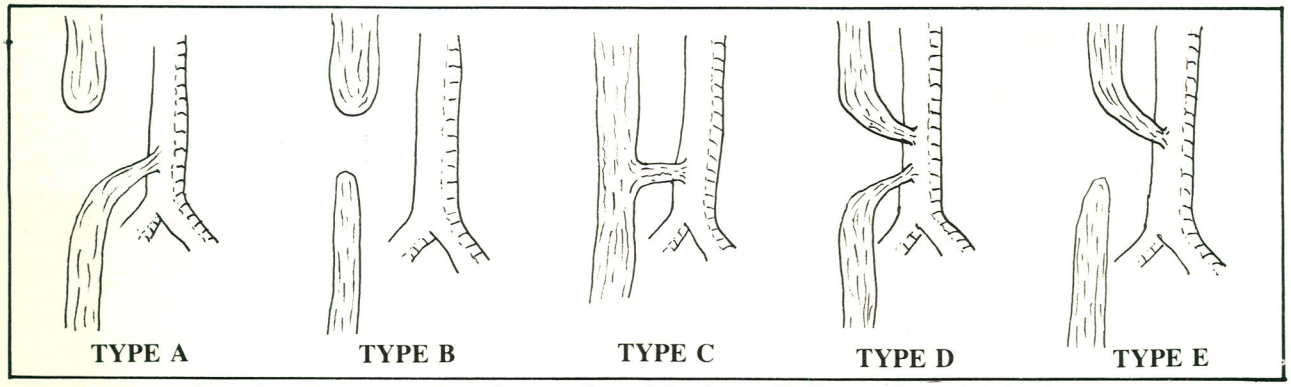
The tracheo-esophageal fistula is commonly with the lower membranous portion of the distal trachea but in rare instances it can occur with other portions of the respiratory tract including the lungs.

The total incidence of these abnormalities is one in three thousand births. The association of other congenital anomalies is fairly high. 20% have some form of congenital heart disease, 10% have imperforate anus, and there are a variety of other, less commonly associated anomalies.

With the exception of type C, all the other types are incompatible with life. Patients with type C and a small fistula may go undetected for years since they can swallow normally. If, however, the fistula is sizeable, chocking and pneumonitis from spillage will set in early.

Since type A is most commonly encountered we shall limit the discussion to this variant. Diagnosis is easily established early in life. When the first feeding is attempted the child will recurgitate the milk and will cough and choke. There will be excessive salivation at the mouth. These should immediately alert doctors and nurses to the possibility of esophageal atresia and a fine catheter should be passed orally into the esophagus. It is preferable to have a radio-opaque catheter so that it can be visualized by X-ray. Failure of the catheter to pass indicates a blind proximal esophageal pouch. The diagnosis therefore will immediately be narrowed to types A and B. It is usually not necessary to instill radio-opaque dye through the catheter to demonstrate the blind proximal pouch but this study can be done if desired for purposes of documentation. It may give useful information about the length of the proximal pouch. The next step consists of a plain X-ray including the chest and abdomen. The presence of gas in the G.I. Tract will clinch the diagnosis of type A since no gas can possibly enter the G.I. tract in type B. The film will also show the catheter curled in the proximal blind pouch as was the case in our patient.

Within the first few hours of life abdominal distention sets in due to



The five types of esophageal atresia and tracheo-esophageal fistula are illustrated. Type A is by far the most common.

continued passage of air through the tracheo-esophageal fistula into the G.I. tract. This may be marked enough to interfere with ventilation. Reflux of gastric fluid into the tracheo-bronchial tree will result in chemical pneumonitis which, in untreated cases, will be followed rapidly by secondary bacterial pneumonia. Dehydration and inanition are unavoidable with eventual toxemia due to overwhelming pulmonary infection. Untreated infants are not expected to survive more than six or seven days at most.

As soon as the diagnosis is made these infants should be transferred to a surgical unit where such cases can be handled. The immediate measures that should be instituted are the following.

* The patient should be placed in a warm incubator and given moist oxygen.
* The bed should be inclined to 30% head up position. This will minimize gastric reflux through the fistula.
* No attempt should be made at further oral feeding. An intravenous line should be established with adequate fluid administered.
* Continuous pharyngeal suction should be instituted to minimize aspiration of saliva.
* Antibiotics should be started routinely, even in the absence of demonstrable pneumonia, since the lung has already suffered a chemical insult. Bacterial super-infection can thereby be prevented.

The timing and type of surgical procedure instituted will depend on a number of factors including prematurity, the babys general condition, and the presence or absence of associated anomalies and their severity. In full term babies where the diagnosis has been made early and the lungs are clear, and who have no associated anomalies, primary total correction is the procedure of choice.

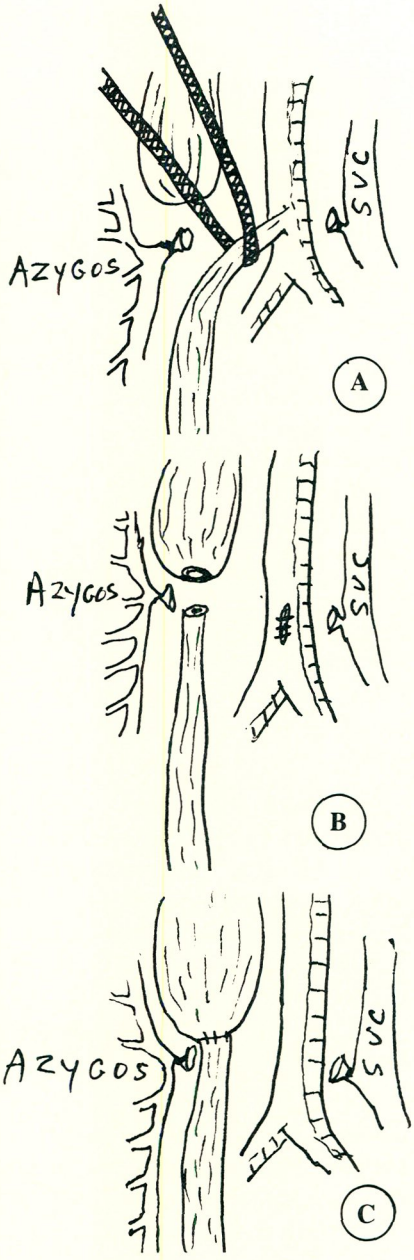
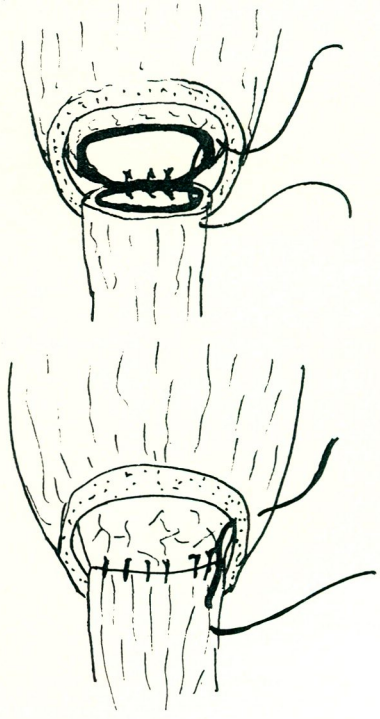


Fig. 4

Steps in the total surgical correction of the common type of anomaly. A: The azygos vein has been divided and the distal esophageal segment has been isolated with a tape. B: The tracheo-esophageal fistula has been divided close to the trachea and the opening in the latter has been closed with interrupted sutures. A small opening has been made dependently in the upper esophageal pouch. C: A one layer esophago-esophagostomy has been completed with interrupted sutures.

The method of correction pioneered by Haight is still the standard procedure (Figure 4)

A high fourth inter-space postero-lateral right thoracotomy will provide excellent exposure of the upper mediastinum. An extra-pleural approach is desirable though not mandatory. Its main advantage is that in case postoperative leak occurs from the esophageal anastomosis, the infection will be limited to the extra-pleural space. It is necessary to divide the azygos vein. The fistula is divided close to the trachea and the opening in the latter closed with fine interrupted non absorable sutures. The proximal pouch is then dissected and a small opening made in it dependently approximating in size the diameter of the distal esophagus. Haight originally advocated a double layer fish-mouth type of anastomosis (Figure 5).



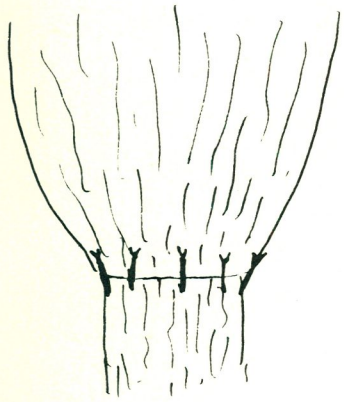


Fig. 5

The two layer fish-mouth type of anastomosis with an initial layer between the mucosa of the upper pouch and the full thickness of the distal esophagus. The second suture layer includes the muscularis of the proximal pouch and the outer muscle layer of distal esophagus thereby overlapping the first suture line.

But most surgeons today prefer a single layer anastomosis with very fine, non-absorbable, atraumatic, interrupted sutures. It is imperative to handle the anastomotic edges with utmost gentleness. The factors that contribute to anastomotic leak are rough tissue handling, tension on the suture line and excessive mobilization of both esophageal ends resulting in interference with blood supply. We prefer to establish a pre-operative or post-operative gastrostomy to decompress the stomach and feed the patient post-operatively.

The chest drainage tube can usually be removed on the 5th or 6th post-operative day. If the patient has been doing well contrast material can be instilled at this time into the upper esophagus to check for leaks. If none are present oral feedings can be started by the 8th to 10th day. The gastrostomy provides a convenient alternate route of feeding until oral feeding becomes fessible. It should not be removed until esophageal patency has been documented and the child can take oral feedings adequately. If esophageal leak does develop the gastrostomy will of-course have to be kept. Most children will develop slight narrowing at the anastomotic site which may require periodic dilatation every three to four weeks for up to two years in some cases. Eventually all of them will stay open.

In sick children with pneumonia, or in severe prematures, staged procedures may be necessary consisting of gastrostomy and division of the fistula followed by delayed repair of the atresia. In patients with short esophagus or in those where disruption of the anastomosis has taken place a cervical esophagostomy is also necessary followed, later in life, by colon interposition . The mortality rate in this group of patients is very high. Prematurity alone (a patient weighing less that 1500 grams) carries a very high operative mortality even when the diagnosis has been made early.

One of the greatest factors in success, assuming an ideal patient, is experience gained from a large volume of cases. However, a competent surgeon, aware of tissue handling techniques and using fine suture materials should have a high rate of success, approaching 90%, in otherwise healthy infants.

SUMMARY

Esophageal atresia with tracheo-esophageal fistula is a condition that is expected to be encountered, though, rarely in any general acute hospital. Based on the number of live births in the Maternity Hospital alone, we would expect to see atleast two cases a year in our health delivery system. Since a sizable portion of these lesions are surgically correctible, a favourable outcome would depend to a great extent on early diagnosis. The case discussed in this presentation was picked up by an alert physician in the Maternity Hospital. Awareness of the presenting symptoms should therefore be an important attribute of any physician or nurse involved in the care of new borns.