Diagnosis of Congenital Tracheoesophageal Fistula in the Adolescent and Adult

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Congenital tracheoesophageal fistula is rarely encountered beyond infancy, but the incidence of this anomaly beyond childhood may be greater than we appreciate. Only the H-type fistula (1.8–4.2 per cent of all tracheoesophageal fistulas)1,2 is compatible with prolonged life. In each of the 13 adult cases reported to date,3 the diagnosis has been made with difficulty. One of the best diagnostic maneuvers is performed daily in the operating theater—endotracheal intubation and positive-pressure ventilation. We recently had an experience in which the anesthesiologist’s observations prompted a more thorough search and the eventual diagnosis of tracheoesophageal fistula in a 15-year-old boy.

**Report of a Case**

The patient had seen numerous physicians for repeated pulmonary infections and frequent coughing and choking spells. In infancy these had been severe enough to precipitate laryngospasm. The diagnosis of tracheoesophageal fistula had been entertained several times since the patient’s birth, but repeated attempts by x-ray (esophagograms) and esophagoscopy to substantiate it had failed. These studies had revealed impaired esophageal motility, delayed gastric emptying, a 2–3-cm hiatal hernia and free esophageal reflux with esophagitis. Also of interest were the numerous congenital anomalies present: omphalocoele, club foot, hypoplasia, horseshoe kidney, a supernumerary rib, and ossous abnormalities of the thoracolumbar spine.

The patient was scheduled for hiatal hernia repair and pyloromyotomy. It was felt that surgical repair of the hiatal hernia and gastric-outlet obstruction would afford relief from apparent reflux and aspiration. The primary anesthetic agents were halothane and nitrous oxide, delivered via endotracheal tube. Ventilation was controlled without the use of muscle relaxants. When the abdomen was opened, gastric dilatation was found despite the presence of a functioning nasogastric tube. Because the distention seemed to increase with each manually assisted ventilation, endotracheal tube placement and cuff patency were rechecked. Advancement of the endotracheal tube to just above the carina abolished the gas leakage. The possible presence of a tracheoesophageal fistula was suggested by the anesthesiologist. After repair of the hernia and lysis of congenital diaphragmatic bands, further attempts were made to demonstrate the suspected fistula by incremenetal withdrawal of the endotracheal tube. However, leakage of air from trachea to esophagus was not again demonstrable despite inflation pressures of 40 to 50 H₂O. We were bewildered!

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**Fig. 1.** The lower illustration shows the oblique pathway of the tracheoesophageal fistula, which resembles an “N” more than an “H”. Redundant tissue at both stomas can have flap-valve action to close the fistula. Insets A and B show that endotracheal tube placement may allow air flow through the fistula (A) or may occlude the fistula.

In the first several postoperative days, pneumonitis of the left lower lobe developed, and the patient later had widespread areas of pneumonitis on the right and the left, suggestive of massive aspiration. Renewed efforts were made to diagnose a tracheoesophageal fistula. Finally, esophagography, coupled with positioning, Valsalva maneuvers and coughing, revealed an H-type fistula beginning in the upper third of the trachea. Bronchoscopy revealed a paralysed stoma on the posterior tracheal wall with a surrounding area of healed-up granulation tissue that was sufficient in size and mobility to occlude the stoma. The esophageal stoma could not be identified by repeat esophagoscopy. Surgical repair of the lesion was accomplished via thoracotomy incision. The patient had a benign postoperative course and has remained free of respiratory tract problems.

**Discussion**

Embryologically, a tracheoesophageal fistula begins in the second month of gestation, as the future larynx
and trachea divide from the foregut. The incidence is about one in four thousand births. Most often this incomplete separation of the respiratory and gastrointestinal tracts is associated with some esophageal atresia. However, in a few people a simple H-type connection is left. Sizes of this structure vary, and it tends to increase in obliquity with age, due to differential growth rates of trachea and esophagus. The final configuration is actually more like an "N" than an "H" (fig. 1). The tracheal stoma of this type of tracheoesophageal fistula is usually quite cephalad, and conceivably could be occluded by the cuff or bypassed during endotracheal intubation. In more than 70 per cent of patients, H-type tracheoesophageal fistulas are found at or above the level of the second thoracic vertebra.

A feature of this anomaly that interferes with diagnostic maneuvers is the flap-valve effect of redundant tissue surrounding both tracheal and esophageal stomas. Periodic closure of the fistula can result, and probably accounted for our inability to demonstrate gastric dilatation after hiatal herniorrhaphy. This flap-valve effect is even more likely at the esophageal stoma due to the collapsible nature of the esophagus, and it may be accentuated by infection, edema, or food particles plugging the fistulous tract.

Gaseous distention of the esophagus and stomach has several implications for diagnosis and symptomatology. Party reported the use of gastric P02 measurements following ventilation with a high concentration of inspired oxygen. Infants with tracheoesophageal fistulas have frothy stools, and older patients have excessive flatus, as a result of this involuntary aerophagia. On plain roentgenograms of the chest pneumoesophagus in the adult, is diagnostic of a respiratory-esophageal communication. An air esophagogram is common for normal infants, however, and may not be helpful in diagnosis in infancy.

Associated congenital anomalies are seen in 27 per cent of patients who have H-type fistulas, as opposed to approximately 50 per cent of those with most other types of tracheoesophageal fistulas. Hence, the other congenital anomalies in the case presented are not surprising, and serve to bear out the adage that the presence of one congenital defect mandates close examination for others. The other diagnostic clue so well demonstrated in this case is the feature of repeated pulmonary infections. Since tracheoesophageal fistula is so hard to demonstrate after infancy by the usual diagnostic methods, it behooves us to entertain the idea in any case of a patient who has chronic pulmonary disease and congenital anomalies. Air flow through the fistula is not a foolproof method of diagnosis, but being alert to this maneuver and the factors affecting it can aid in the anesthetic management of such patients, and may resolve the etiology of chronic pulmonary infection in more patients than we now realize.

References