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Case Reports

Diagnosis and Surgical Treatment of "H-Type" Tracheoesophageal Fistulas

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Four cases of H-type tracheoesophageal fistula are reported. The patients all had chronic aspiration from the esophagus. Although serious symptoms were present in all, definitive diagnoses were not made until the patients had reached 1, 6, 12, and 50 years of age, because ordinary radiologic methods failed to establish the diagnosis. The angulation of the fistula usually prevents contrast medium in the esophagus from entering the trachea, especially with the subject upright. On the other hand, air easily passes from the trachea to the esophagus, eventually producing megaesophagus which may be confused with the picture of achalasia. An ill-advised Heller esophagomyotomy was done on 1 patient. All 4 patients eventually had successful closure of the fistulas. Three operations were by the transthoracic route, and 1 high fistula in an infant was closed through a cervical approach.

In the most common variant of atresia of the esophagus, the origin of the distal segment of the esophagus is from the trachea near its bifurcation (Fig. 1A). This part of the anomaly is so striking that the term "T-E fistula" is frequently used to mean the entire anomaly, which is incompatible with life because no food can reach the stomach. In the variant with no atresia of the esophagus but only a fistula, the condition is compatible with life for long periods, but is associated with chronic aspiration from the esophagus. The fistula usually has appreciable length, and forms the horizontal part of the "H" (Fig. 1B). The "H" is imperfect, since the crossbar is not truly horizontal but is slanted; the opening in the trachea invariably lies above that in the esophagus. This explains the "valve" mechanism that allows material in the esophagus to pass preferentially into the stomach without entering the trachea. On the other hand, air from the trachea can easily get into the esophagus and, hence, into the stomach, thereby producing the radiologic and clinical picture which should arouse suspicion of the existence of the anomaly.

Congenital H-type fistulas are rare, and this rarity helps to explain the fact that many symptomatic patients are followed for years without suspicion of the correct diagnosis by clinicians and radiologists. When Haight [1] reported the successful closure of an isolated fistula, he noted that it was only the second example of such an anomaly in his series of 65 cases of tracheoesophageal fistula, the others having also had atresia. When Haight [2] presented his presidential address before the American Association for Thoracic Surgery in 1957, his total series of esophageal anomalies was 208 cases, of which 7 had the fistula without atresia. His paper contains a photograph of the specimen of the first reported case by Lamb [3] in 1873. In 1962, Waterston [4] of the Hospital for Sick Children of London reported only 3 H-type fistulas in his series of 218 babies. A survey of the experience of members of the surgical section of

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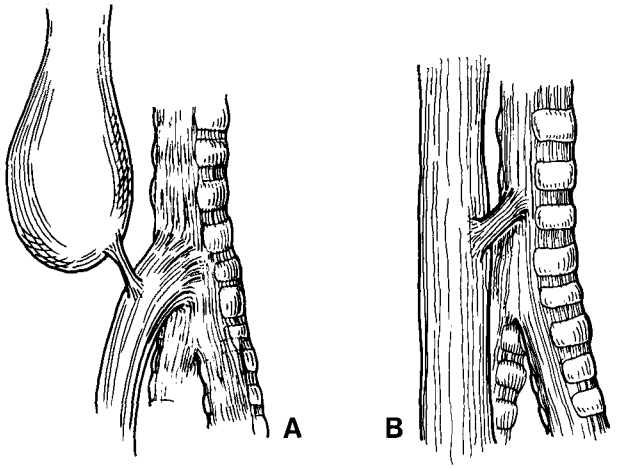


Fig. 1 A. Common type of tracheoesophageal fistula with atresia of the upper segment of the esophagus. **B.** H-type tracheoesophageal fistula. Note the obliquity of the fistula.

the American Academy of Pediatrics in 1964 [5] indicated that of 1,058 patients with tracheoesophageal fistula, only 44, or 4.2%, had the fistula as an isolated anomaly. Other papers on H-type fistulas have been reports of from 1 to 4 cases [6–11].

The anomaly resists diagnosis, as will be seen from the following 4 case reports. In particular, the story of the first case is an almost unbelievable saga of misinterpretation of signs and symptoms, with the result that a child had 4 major operations before the definitive one was done when he was 12 years old! The other 3 patients had surgical correction at the ages of 1 year, 6 years, and 50 years. The symptoms in the last 2 patients were so mild that operation was postponed electively for several months after the diagnosis was established.

Case Reports

Case 1

R.L. was a 6-week-old baby boy, referred for evaluation of congenital heart disease. The patient had had feeding difficulty for at least 1 week, with a tendency to choke when fed, but he did not regurgitate. Occasionally, these episodes were associated with shortness of breath, but cyanosis was present only when he cried and held his breath. Chest x-ray showed an enlarged heart, and there was a murmur suggestive of patent ductus arteriosus. At cardiac catheterization, this diagnosis was confirmed. In view of the enlarged heart, it was decided to repair the anomaly.

At operation, there was difficulty with the endotracheal tube, and the left lower lobe was found to be atelectatic. Postoperative convalescence was complicated by recurrent aspiration of mucus with atelectasis and pneumonitis. A chest x-ray from that time shows what in retrospect should have been a clue, namely, the stomach was greatly dilated with air (Fig. 2). The patient was discharged from the hospital, but ate poorly, had much mucus, and failed to gain weight. He had coarse rales and gurgling, and was readmitted to the hospital for tube feeding. The tube feedings were tolerated and he was discharged.

Seven weeks later, he was admitted to another hospital with the diagnosis of pneumonitis. After 6 weeks of respiratory distress and recurrent bouts of fever, he was transferred to Henry Ford Hospital. A Lipiodol® swallow showed a posterior indentation of the esophagus. The next day, the baby had his second left thoracotomy, and an aberrant right subclavian artery was divided. At operation, there was difficulty in inflating the left lung. The postoperative course was stormy. Bronchoscopy was required early, and was followed by a tracheostomy to permit repeated aspirations of the trachea. He was discharged in fair condition, but with a residual infiltrate in the right upper lobe. The following year, respiratory symptoms worsened, and he was admitted to the hospital for bronchoscopy and bronchograms. A second tracheostomy was done the day after the studies. The bronchograms showed stenosis of the bronchus to the right upper lobe, and 2 weeks later, there was still retained contrast material in the lobe.

After an uneventful 2 years, he again developed a chronic cough and inability to cough up material. Bronchoscopy confirmed the presence of stenosis of the bronchus to the right upper lobe, and a lobectomy was performed. There was extensive bronchiestasis with retained secretions distal to the stenosis. The subsequent 7 years were characterized by recurrent respiratory infections and repeated hospitalizations. At age 11, he had a widely dilated esophagus suspicious of achalasia. Fluoroscopically, the esophagus was noted to be dilated and showed very little peristaltic activity. There was aspiration of contrast material into the trachea, but no definite evidence of abnormal communication. A Heller cardiomyotomy was done. A postoperative x-ray showed that the dilatation of the esophagus persisted, but contrast passed readily into the stomach (Fig. 2B).

Twelve years after our first contact with him, he still had a troublesome cough. There were rales in both lungs. Drinking water caused fits of coughing. It then became clear that he had a tracheoesophageal fistula, and this was demonstrated rather easily by x-ray. A Levine tube was placed in the esophagus, and contrast medium was injected with the subject in a prone position. A typical H-type

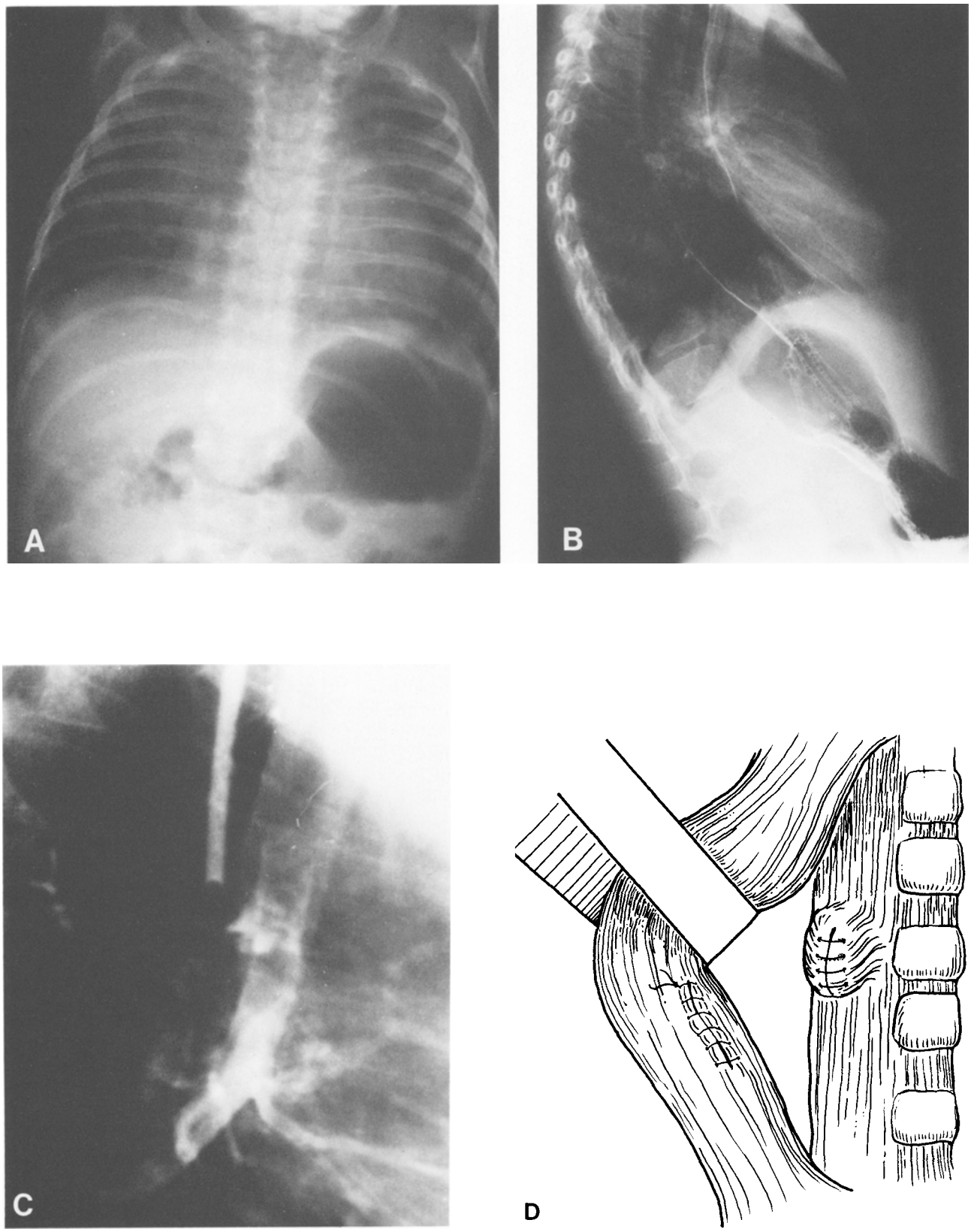


Fig. 2. Case 1. **A.** X-ray of chest at age 2 months. Note the stomach distended with air. **B.** Barium x-ray of esophagus shortly after a Heller operation for "achalasia" was performed at 11 years of age. **C.** X-ray demonstration of H-type tracheoesophageal fistula with contrast medium injected into esophagus at age 12 years. **D.** Division and closure of fistula. The protruding tracheal end will be inverted with another row of sutures.

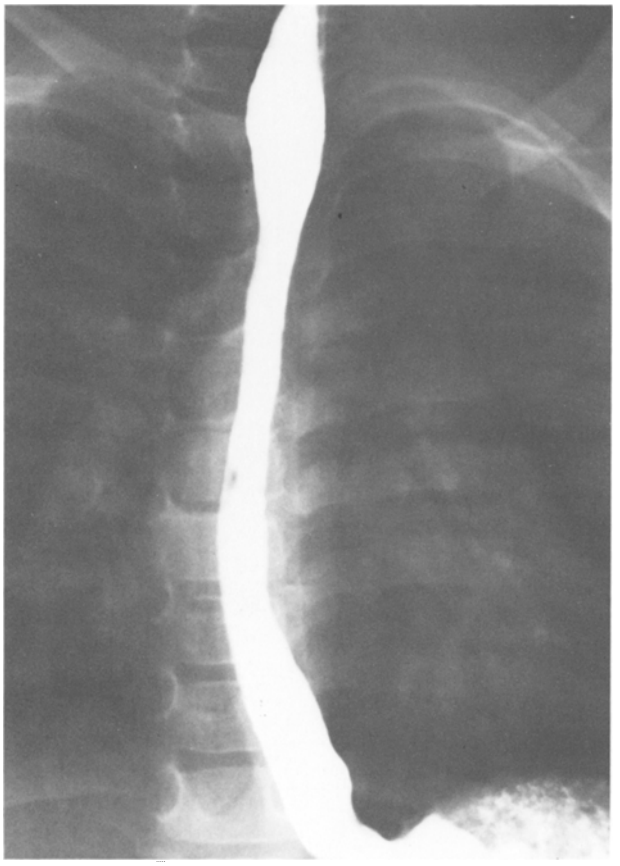


Fig. 3. Case 2. Barium x-ray study of esophagus 5 years after operation shows excellent form and function.

tracheoesophageal fistula was demonstrated going upward toward the trachea (Fig. 2C). At operation, it was noted that the esophagus was inflated by air coming from the endotracheal tube. The fistula was divided, leaving a longer stump on the tracheal side, permitting a double suture line for closure of both ends (Fig. 2D). The postoperative course was uneventful, in contrast to the course after the other operations. The chronic bronchitis gradually resolved, and the patient had no further difficulties.

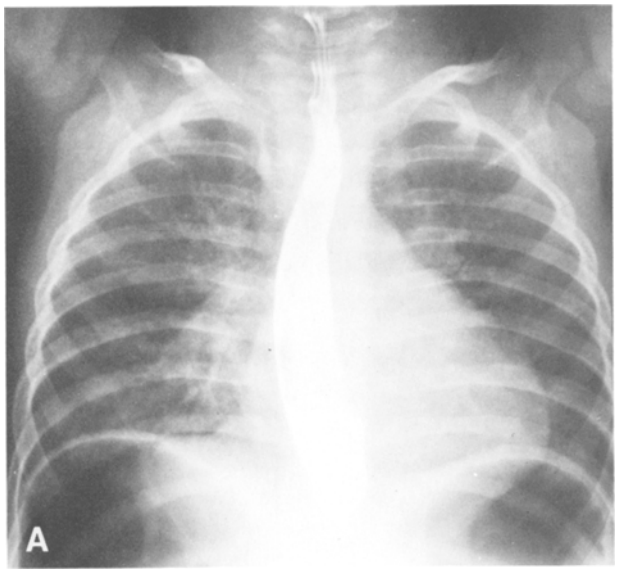
Comments. With knowledge of the correct diagnosis, it is undoubtedly difficult for a reviewer of this 12-year case history to understand why the answer eluded us so long. Excessive amounts of air in the stomach and later a dilated esophagus filled with air should have aroused suspicion. Hints were present repeatedly, but there seemed to be a deeply rooted opposition to accepting the diagnosis of tracheoesophageal fistula. It is evident that ordinary radiologic techniques miss many H-type fistulas. Haight [1] pointed out early that the subject should be prone when the contrast medium is introduced into the esophagus. The author is not the only surgeon to have done a Heller cardiomyotomy in a patient with unrecognized H-type tracheoesophageal fistula. It was also reported by Olivet and Payne [11] from the Mayo Clinic.

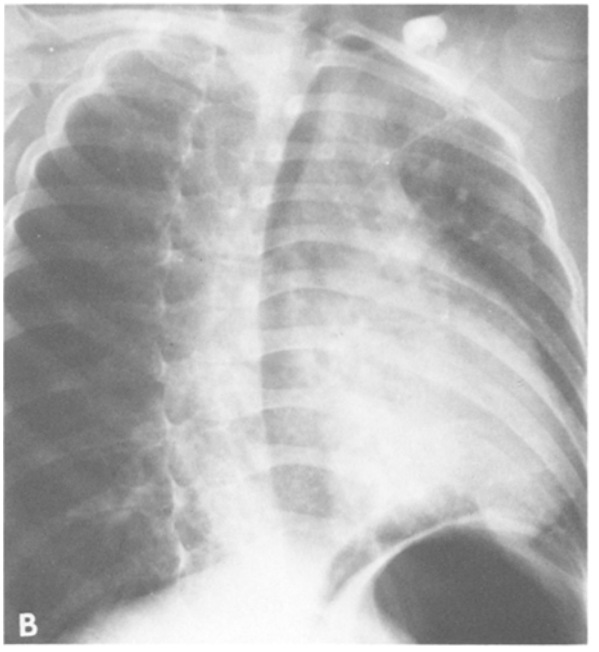
Case 2

G.D. was a 1-year-old boy, with a history of pulmonary problems since birth. There had always been a cough with choking when he was fed. He tolerated solids better than liquids. A tracheoesophageal fistula had been suspected, but radiologic studies at the age of 5 months failed to demonstrate one. He had recently been admitted to another hospital with areas of consolidation in the left upper and right lower lobes of the lung. A barium swallow x-ray did not show a fistula. At admission to Henry Ford Hospital, there was clearing of the pneumonitis, and abundant air in the gastrointestinal tract. Cine radiography revealed a small tracheoesophageal fistula from the cervical esophagus. The fistula was closed through a cervical incision. The patient was discharged in good condition 3 weeks after the operation. Five years later, a barium swallow revealed a normal esophagus (Fig. 3).

Case 3

K.E. was 1 year old when she was admitted for cardiac catheterization to evaluate a heart murmur. She had had rattling sounds in her chest since birth and pneumonia in early infancy. On admission, she had noisy breathing with much mucus. A barium swallow x-ray showed moderate cardiomegaly and a large amount of air in the gastrointestinal tract (Fig. 4A). Cardiac catheterization confirmed the presence of a ventricular septal defect. It was suggested that closure of the cardiac defect might be done when she was 5 years old. In the subsequent 4 years, she had recurrent respiratory infections and chronic cough. These symptoms progressed and the patient had the most difficulty with cough when lying down. X-rays of the chest showed the esophagus dilated and filled with air (Fig. 4B). Review of the x-rays taken at age 1 year showed some barium in the tracheoesophageal tree in the lateral projection, without definite evidence of fistula (Fig. 4C). A cine esophagogram with a tube placed in the esophagus demonstrated the fistula high in the thoracic region. It could not be seen in any position when the x-ray was taken with the patient swallowing barium. Operation was done through the right chest. The esophagus was distended with air, and there was marked expansion every time the anesthetist compressed the bag. The fistula was located high in





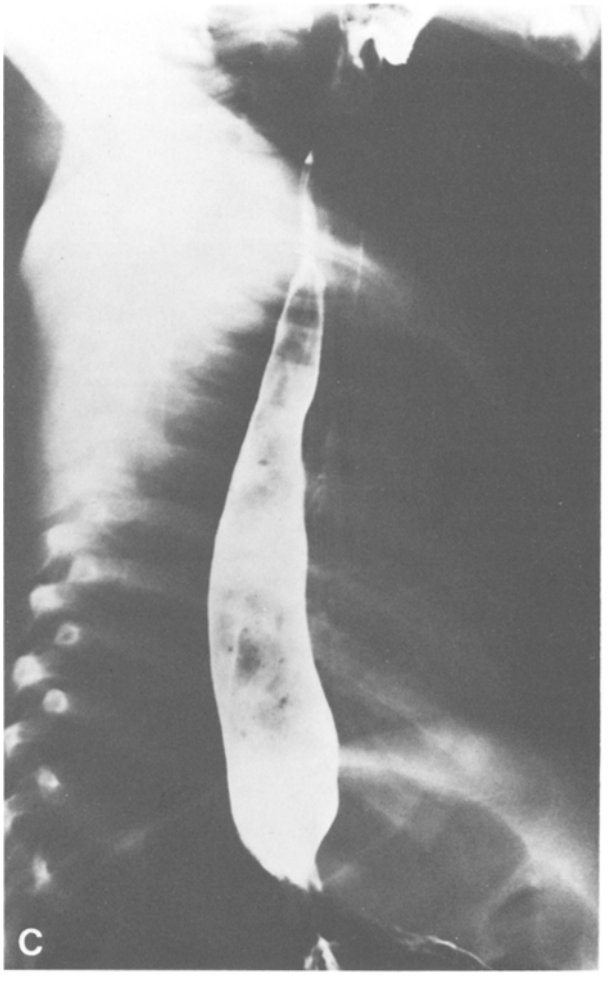


Fig. 4. Case 3. **A.** X-ray at 1 year of age shows the esophagus is well filled with barium and no opacification of the trachea. Note the large amount of air in the gastrointestinal tract. **B.** Chest x-ray at 5 years of age showing dilated, air-filled esophagus and stomach. **C.** Barium x-ray study of esophagus at age 1 year, which, on review, showed barium in the trachea. It was assumed at the time the x-ray was performed that the opacification of the trachea was from aspiration from the pharynx!

the chest; after it was divided, the trachea and esophagus were closed with a double row of sutures. The patient had no further pulmonary difficulties thereafter.

Case 4

A 40-year-old female presented with hemoptysis present for a few days and severe coughing spells lasting for 3–6 hours. She had had colds all of the previous winter. Examination revealed coarse rales at the right base which cleared on coughing. A chest x-ray was reported as negative. Eight years later, she appeared in the Emergency Room complaining of chest pain. The lateral chest x-ray showed the esophagus to be enlarged and filled with air (Fig. 5). A cine radiographic study of the esophagus demonstrated a small high communication between the trachea and esophagus. Repair was done through a right thoracotomy and was difficult because of the high location of the fistula. Postoperatively, the patient had no more coughing spells or abdominal distention when coughing or straining.

Comment. Although this fistula was successfully repaired with a thoracic approach, a cervical approach might have been preferable. The group from the

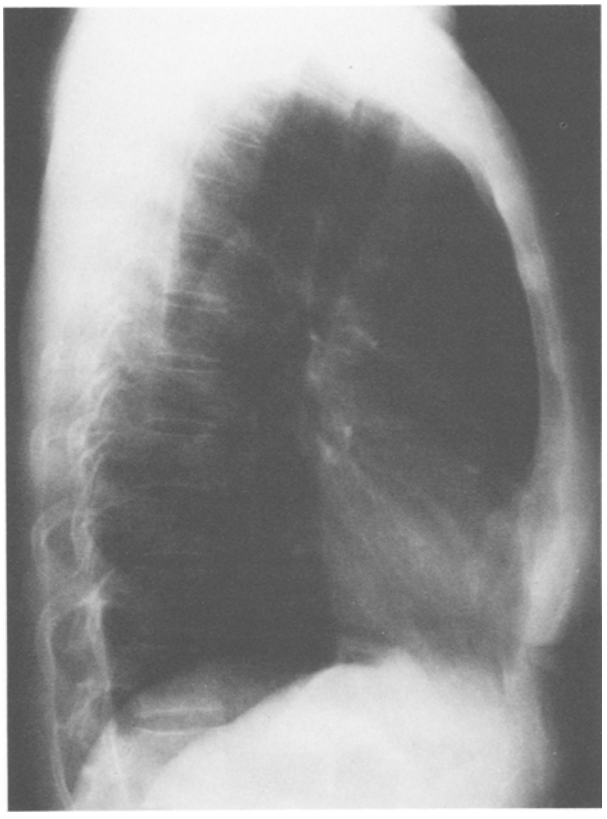


Fig. 5. Case 4. Lateral chest x-ray at age 49, showing a moderately dilated esophagus filled with air.

Children's Hospital of Los Angeles [12] reported a remarkable series of 15 cases, which included 11 neonates. There was no mortality in the 8 most recent cases, all of which had been repaired by the cervical approach, irrespective of the level of the fistula.

Résumé

Présentation de 4 cas de fistule trachéo-oesophagienne en H. Les 4 malades présentaient des symptomes d'aspiration du contenu oesophagien vers l'arbre respiratoire. Ces symptomes étaient graves dans tous les cas et pourtant le diagnostic n'a été posé qu'aux âges de 1, 6, 12 et 50 ans, parce que les examens radiologiques standard n'avaient pu mettre la lésion en évidence. L'angulation de la fistule empêche, en général, le produit opaque de passer d l'oesophage vers la trachée, surtout en position debout. Par contre, l'air passe aisément de la trachée vers l'oesophage, pouvant donner un aspect de mégaoesophage qui peut être pris pour une achalasie. Chez 1 malade, une oesophagomyotomie de Heller fut réalisée par erreur. Chez les 4 patients, la fistule a pu être fermée avec succès. L'opération a été faite par voie thoracique dans 3 cas; chez 1 enfant, la fistule haut située a été fermée par voie cervicale.

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