Congenital H-type Tracheoesophageal Fistula : A Case Report

SOMCHAI WAIKITTIPONG, M.D.*

Abstract

A case of congenital H-type tracheoesophageal fistula was reported. A five-year-old male presented with the symptom of coughing and choking during eating. He had experienced the symptom since the neonatal period. He was frequently admitted with the diagnosis of pneumonia but the definite diagnosis has never been established. The esophagogram revealed communication between cervical esophagus and trachea. The operation was performed by division and suture of the fistula uneventfully. Postoperatively, he could take meals normally. The literature of this condition was also reviewed.

Isolated (H-type) tracheoesophageal fistula is a rare variant of congenital malformation of the esophagus and trachea, of which the most common type is esophageal atresia and distal tracheoesophageal fistula. The incidence is about 4 per cent of esophageal anomalies. Although the surgical repair is simple and the prognosis is good, it may be difficult to make a diagnosis. This report presents a case of this condition and review of the literature.

CASE REPORT

A five-year-old male presented with coughing and choking during eating. He had experienced these symptoms since the neonatal period. At that time he was admitted frequently and given the diagnosis of pneumonia. When he was two months old, an esophagogram was performed but could not demonstrate any abnormality. After that he continued to suffer from the symptoms and was admitted occasionally with developing pneumonia. On physical examination, the child appeared normal with no other abnormal findings. Chest X-ray examination revealed mild peribronchial infiltrations on both sides. Esophagogram was performed and revealed the fistular tract between the trachea and lower cervical esophagus (Fig. 1). The diagnosis of congenital H-type tracheoesophageal fistula was then established. The operation was performed through a left cervical approach. The fistula

* Department of Surgery, Yala Hospital, Yala 95000, Thailand.

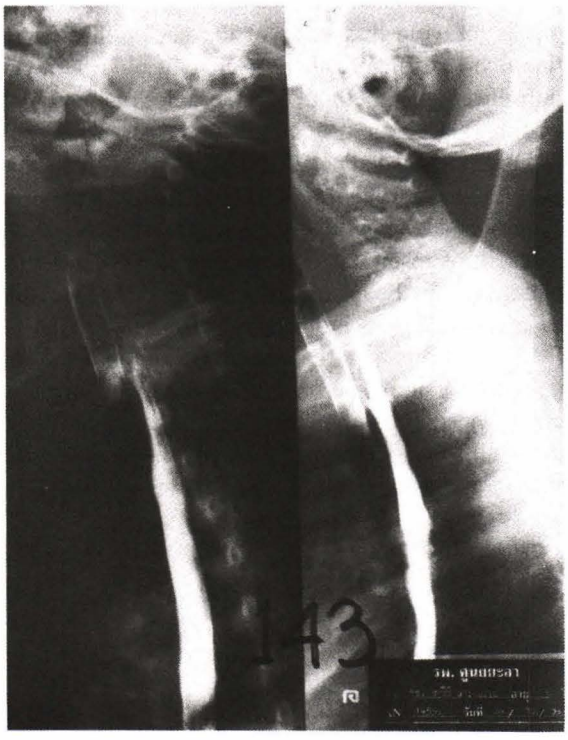

Fig. 1. Esophagogram demonstates fistular tract between trachea and distal part of the cervical esophagus.

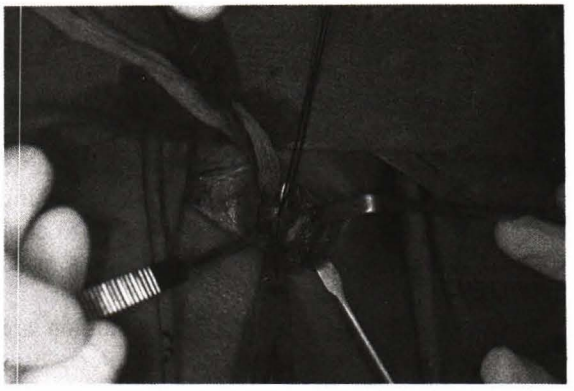

Fig. 2. By right transcervical approach, the fistula was visualized and mobilized.

was visualized, divided, and sutured on both ends (Fig. 2). His postoperative course was uneventful. On follow-up, he could eat very well.

DISCUSSION

The first case of congenital H-type tracheoesophageal fistula in the literature was reported in 1873 by Lamb[1]. The lesion was diagnosed at autopsy in a 7-week-old infant. The fistula was found one-half inch below the cricoid cartilage. In 1938, Imperatori[2] performed the first successful repair of this condition in a 6-year-old child. The fistula, at the level of the fifth tracheal ring, was diagnosed at bronchoscopy. Closure was performed through a large tracheotomy incision. In 1946, Haight[3], using a thoracic approach, reported the second successful closure of a fistula. From 1946 to 1958, all reported fistulas were repaired through the chest[4]. Miller[5], in 1958, reported the first transcervical approach. The fistula was seen on the esophagogram to be 1.5 cm above the level of the first rib. The fistula was found at the lower pole of the thyroid and successfully repaired.

The location of the fistula in congenital H-type tracheoesophageal fistula varied considerably. In the report of Schneider[4], collecting 32 cases with successful repair from the literature, the fistulas were found to be located at the same levels of the first, second, third, and fourth thoracic vertebras in 9, 11, 10, and 2 cases respectively. Bedard[6] reported 23 cases of congenital H-type tracheoesophageal fistulas, who were seen at the Hospital of Sick Children in Toronto during the period 1939 to 1973. Among the 23 patients, the fistulas were located within 2 cm of the carina in 6 patients, entering the right main stem bronchus in 2 patients, in the cervical esophagus above the suprasternal notch in 9 patients, and at or below the suprasternal notch but well above the carina in 6 patients. Although the fistula may be found at any level, accumulated data emphasize that most of these fistulas occur in the upper portion of the trachea and esophagus. Of the cases in the literature in which the level of the fistula has been carefully determined, at least 70 per cent of the reported H-type fistulas occurred at or above the level of the second thoracic vertebra.

The clinical menifestation of congenital H-type tracheoesophageal fistula consists of the following characteristic symptoms: 1. coughing, choking, and cyanosis with feeding, solids usually

better tolerated than liquids; 2. intermittent abdominal distention, resulting from air forced through the fistula into the stomach during expiration; 3. recurrent pneumonitis, resulting from aspiration of food or gastric juice through the fistula. The important symptom, which usually alerts the physician to suspect this condition, is coughing and choking associated with feedings. The investigation to confirm the diagnosis of this condition has to be made until proved otherwise.

The investigations for this condition are crucial for both the diagnosis and treatment. Esophagogram and bronchoscopy are the two most valuable procedures. In many instances, especially for the small newborns or infants, to make the diagnosis is particulary difficult, when, in spite of a convincing clinical picture, all investigations including an esophagogram and bronchoscopy are negative. One must persist in attempts to confirm the diagnosis. The need for accurate and definite proof of the fistula and its location is obvious if one is to avoid dissection of the neck and mediastinum on suspicion alone. Schneider[6], in 1961, introduced the method of cineradiography which was more accurate than conventional esophagogram to confirm the diagnosis of H-type tracheoesophageal fistula. Bronchoscopy in infants or children should be done with great caution because there is a risk of respiratory obstruction during and after manipulation. Esophagogram alone is not reliable as a method to demonstrate the fistula.

Surgery is the treatment of choice. The operation can be performed through the neck or chest, depending on the location of the fistula. When the fistula is located at or above the level of the second thoracic vertebra, the operation should be

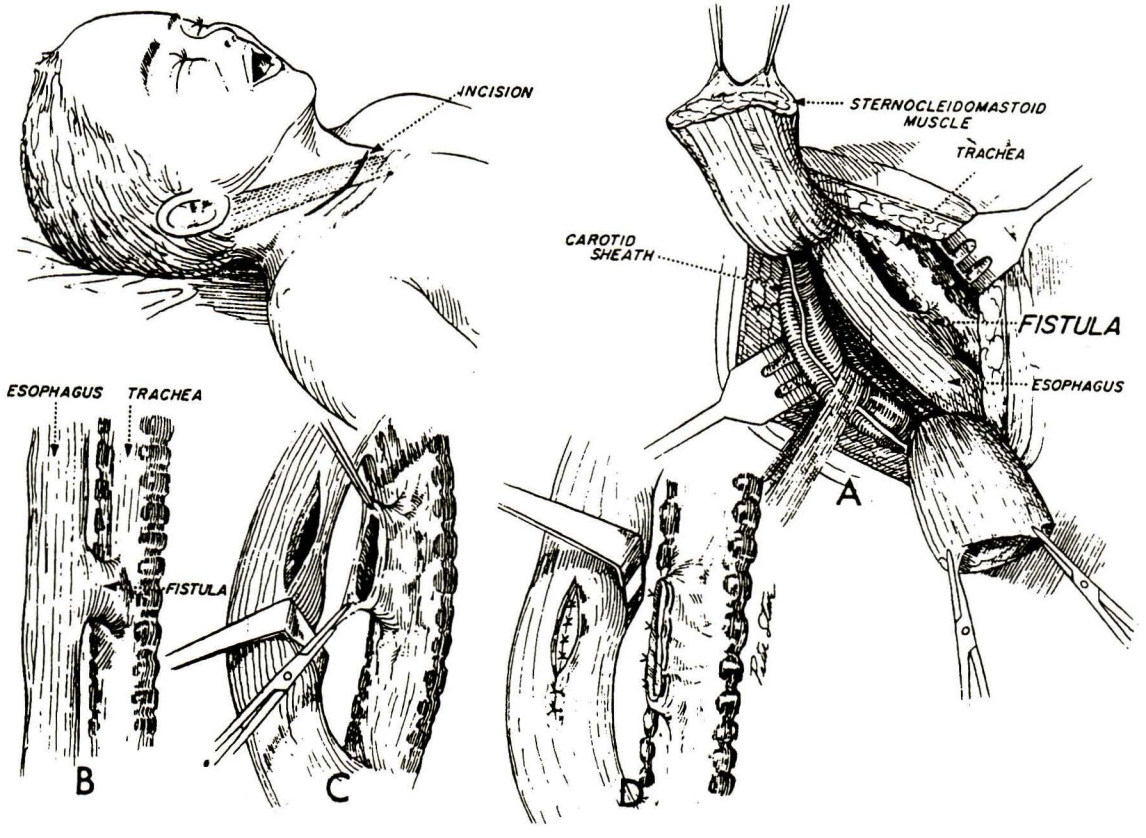

Fig. 3. Diagram demonstates the transcervical approach for congenital H-type tracheoesophageal fistula.

performed through the cervical approach. In spite of the fact that the esophagus lies slightly to the left of the midline at the thoracic inlet, the position of the fistula warrants a right-sided approach. A curving incision in the skin line low in the neck is made from the midline and extended laterally beyond the sternocleidomastoid muscle (Fig. 3). Division and suturing of the fistula is best accomplished by severing the fistula nearly flush on the esophageal side. The additional length achieved by leaving some of the fistula wall on the tracheal side assures a secure closure without compromise of the tracheal lumen. When the fistula is located below the second thoracic vertebra, right thoracotomy approach should be performed. In some instances, it may be difficult to find the fistula and thus extensive dissection is performed, which can lead to further morbidity. Stephen[7] developed a technique which combinded diagnostic and surgical management for this condition. He used the bronchoscopy with rod lens technique to identify the fistula through the trachea, then a small Fogarty catheter is threaded through the opening in the trachea under direct vision and passed through the fistula into the esophagus. The balloon is inflated and bronchoscopy removed, leaving the catheter in place. A proper incision is then made in the neck or chest, depending on the location of the fistula, and the fistula tract is quickly located by palpation of the balloon and catheter. Then with minimal and accurate dissection, correction is carried out.

SUMMARY

A case of congenital H-type tracheoesophageal fistula was presented. This condition is a rare variant of congenital malformation of the esophagus and trachea. In spite of its convincing symptoms, it may be difficult to make a diagnosis. One must persist in attempts to confirm the diagnosis. Repeated investigations, both esophagogram and bronchoscopy, should be done. Result of surgical correction is excellent.

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ความพิการแต่กำเนิดของหลอดอาหารชนิดที่มีทางติดต่อกับหลอดลมโดยที่ไม่มีการตีบตันของหลอดอาหาร : รายงานผู้ป่วย

*สมชาย ไวกิตติพงษ์, พ.บ.**

รายงานผู้ป่วยเด็กชายอายุ 5 ปี มีอาการไอและสำลักเวลากลืนอาหารมาตั้งแต่แรกเกิด มีประวัติต้องเข้ารับการรักษาในโรงพยาบาลหลายครั้งโดยได้รับการวินิจฉัยโรคเป็นปอดอักเสบ ได้ส่งตรวจภาพถ่ายรังสีของหลอดอาหาร พบเป็น congenital H-type tracheoesophageal fistula ผู้ป่วยได้รับการผ่าตัด หลังผ่าตัดผู้ป่วยรับประทานอาหารได้เป็นปกติ เนื่องจากความพิการแต่กำเนิดของหลอดอาหารชนิดนี้พบได้น้อยจึงได้ทบทวนความรู้เกี่ยวกับการวินิจฉัยและการรักษาของภาวะนี้

* กลุ่มงานศัลยกรรม, โรงพยาบาลยะลา, จ.ยะลา 95000