Cleft Larynx
Laryngotracheoesophageal Cleft
Persistent Esophagotrachea

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Richter is credited with the first diagnosis of cleft larynx in 1792 when he inserted his finger in the throat of an infant and found the gullet and larynx to be a common cavity. Since then, reports of about 40 proven cases of laryngotracheoesophageal cleft have been published, the majority dating from the last years. The apparent rarity of this anomaly may be due in part to a general unawareness of its existence and to the difficulties in making the correct diagnosis. Haight and Williams (1962) found the lesion in four patients out of approximately 2000 autopsies. Because of the small number of reported cases it is virtually impossible to establish positive conclusions with respect to incidence, clinical pattern or associated anomalies. As with other congenital anomalies of the esophagus, there seems to be a relation with both immaturity and hydramnios (Zachary and Emery, 1961; Haight and Williams, 1962; Blumberg et al., 1965; Frates, 1967; Fisher, 1969). The sex distribution is approximately equal and a familial tendency has been described (Crooks, 1954; Zachary and Emery, 1961).

The associated anomalies include esophageal atresia with fistula (Welch and Husain, 1958; Haight and Williams, 1962; Daum et al., 1965; Delahunt and Cherry, 1969), Meckel’s diverticulum (Blumberg et al., 1965; Fisher, 1969; Delahunt and Cherry, 1969), absence of a kidney (Welch and Husain, 1958; Delahunt and Cherry, 1969), extrophy of the bladder (Griscom, 1966), hiatus hernia (Jahrsoerfer et al., 1967), severe cleft palate and persistence of a left vena cava (Haight and Williams, 1962), incomplete iris (Fisher, 1969), ventricle septal defect (Daum et al., 1965), bifid spleen, imperforate anus, annular pancreas, patent ductus arteriosus, coarctation of the aorta, urethrectal fistula (Delahunt and Cherry, 1969).

The respiratory tract is first recognizable as a pouch arising from the anterior aspect of the primitive foregut. As the gut elongates the pouch also progresses caudad but it becomes separated from the developing esophagus by the union, from below upward, of infoldings of mesodermal origin from each side of the common tube; this separation of esophagus and trachea up to the level of the larynx is completed in the embryo of 9 to 10 mm (± 33 days). The cricoid cartilage, coming from the fifth or sixth branchial arch, develops as two lateral centers of cartilage that fuse first ventrally and then dorsally at about the forty-fifth gestational day. The development and functioning of the vocal cords and the ability of the epiglottis to cover the glottis depend on the dorsal fusion of the cricoid cartilages. The pathogenesis of a laryngotracheoesophageal cleft seems to be due to an arrest in the rostral development of the tracheoesophageal septum which in turn prevents the dorsal fusion of the cricoid cartilages. The defect apparently begins to appear around the thirty-fifth gestational day (this is
approximately 10 days before the normal fusion of the cricoid cartilage). The severity of the cleft will vary with the stage of development reached by the embryo at the time the growth of the tracheoesophageal septum was arrested. It may involve only the cricoarytenoid area (Finlay, 1949; Crooks, 1954; Daum et al., 1965; Harrison et al., 1965; Shapiro et al., 1966; Jahrsdoerfer et al., 1967; Imbrie and Doyle, 1969; Delahunty and Cherry, 1969; Fisher, 1969); it may extend downward beyond the cricoid cartilage into the trachea (Petersen, 1955; Zachary and Emery, 1961; Blumberg et al., 1965; Geiger et al., 1970); or it may be so extensive that the septum is completely absent, resulting in a persistent esophagotrachea (Welch and Husain, 1958; Tribolletti, 1958; Zachary and Emery, 1961; Griscom, 1966; Fisher, 1969).

The clinical picture chiefly includes respiratory distress, sometimes with pronounced mucous secretions and marked episodes of cyanosis, and feeding difficulties (with incoordination of swallowing and aspiration of food). There is also a high incidence of laryngeal stridor and vocal changes in the form of a weakness of voice or frank aphonia, due to an air leak through the cleft and the inability to approximate the vocal cords. The symptoms develop immediately after birth and, if no stridor occurs, may be indistinguishable from esophageal atresia with tracheoesophageal fistula. Untreated the infants will almost always succumb to aspiration pneumonia or sudden respiratory death. The more extensive the communication between airway and gullet, the freer the aspiration and the sooner the death of the infant may be expected. Except for the case described by Jahrsdoerfer (1967), which is exceptional in that the infant lived 46 months in the hospital before a proper diagnosis was made and surgery undertaken, all other cases without successful surgical correction have been fatal in early infancy.

Diagnosis is extremely difficult. This can be attributed in some cases to intermittent opening and closure of the posterior margins of the larynx and trachea during respiration and in other cases to a lack of knowledge of the condition. Routine chest films show only the secondary changes of aspiration pneumonia, although the shadows of the air contained in trachea and esophagus may arouse suspicion. Esophagograms show spillage of contrast material into the trachea but by themselves do not allow a correct diagnosis. In cases of persistent esophagotrachea cineradiography during swallowing can lead to a correct preoperative diagnosis if a persistent fusion of the tracheal and esophageal lumen is found (Frates, 1967). Endoscopic procedures (laryngoscopy, tracheoscopy, esophagoscopy) might be expected to reveal a greater percentage of cases but frequently the larynx will appear surprisingly normal. Neither roentgenographic evaluation, nor endoscopy, nor surgical exploration have been uniformly successful in reaching a correct diagnosis. All patients in whom the diagnosis was made preoperatively had undergone intensive investigation, including X-ray examinations, endoscopic explorations and even surgical procedures. Often a right diagnosis could only be made at the time of surgical exploration (Blumberg et al., 1965; Griscom, 1966; Delahunty and Cherry, 1969; Fisher, 1969; Geiger et al., 1970) or at post-mortem examination (Welch and Husain, 1958; Tribolletti, 1958; Haight and Williams, 1962; Daum et al., 1965; Harrison et al., 1965; Fisher, 1969).

The surgical approach depends on the size of the cleft. If it extends downward for a few tracheal rings only, repair of the defect can be attempted by an anterior (Jahrsdoerfer et al., 1967; Delahunty and Cherry, 1969) or a lateral pharyngeal (Shapiro et al., 1966) approach. Large defects extending down to the carina require a combined thoracic-cervical approach. Although a prolonged delay in total correction of the lesion does not seem advisable, a short waiting period to clear an eventual pneumonic process and to improve the nutritional state will