Laryngo-tracheo-oesophageal cleft

Clinical features, diagnosis and therapy

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Abstract. The laryngo-tracheo-oesophageal cleft is marked by a missing anatomical separation of the oesophagus and the larynx. The cleft can be restricted to the dorsal part of the larynx (type I), extend to the upper area of the trachea (type II) or involve the whole of the trachea (type III). In reviewing our three cases and 82 cases in the literature, clinical features, diagnosis, and therapy of this rare condition are presented. The condition leads almost always to life-threatening disturbances in the form of asphyxia and aspiration pneumonia. The diagnosis is made through direct laryngoscopy. Radiological demonstration of swallowing and tracheo-oesophagoscopy are necessary additional investigations. In many cases further severe malformations of organs are found, i.e., oesophageal atresia and tracheo-oesophageal fistulae. Despite early detection the prognosis remains unfavourable with a mortality of 46%. With few exceptions, an early operative correction of the defect is required. Access to the dorsal laryngeal area is achieved usually by a lateral pharyngotomy. Difficulties arise in securing adequate closure of the dorsal larynx, because of lack of sufficient tissue material. Despite secure closure of the laryngo-tracheo-oesophageal cleft, disturbances of swallowing often persist. Development of speech is not impaired.

Key words: Larynx malformation – Oesophageal malformation – Congenital stridor – Asphyxia – Swallowing dysfunction

Introduction

Congenital laryngo-tracheo-oesophageal cleft is characterised by deficient anatomical separation of the oesophagus and respiratory system at the level of the larynx and trachea. Depending on the extent of the cleft in the larynx and trachea, life-threatening disturbances of respiration can occur immediately after birth. Petterson [46] distinguishes three types (Fig. 1): In type I named cleft larynx, a cleft is found in the Lamina cricoidea and the inter-arytenoidal musculature system of the larynx is missing. In types II and III there is also malformation of the trachea, which can be limited to the six proximal tracheal rings (type II), or extend to the carina (type III). In many cases other complex organ malformations are found.

Probably the first description was given by Richter in 1772 [52], a second by Mackenzie in 1880 [38]. More recently cases were reported by Bowman and Jackson [8] and by Finlay [22].

Case reports

1—Patient K.-B.S., male: The mother was a 31-year-old para-three of Indian origin. A 3-year-old brother and a 5-year-old sister are in good health. After uneventful pregnancy a Caesarian section was performed in the 38th week, because of foetal distress. The birth weight was 2890 g, length 52 cm, and the amount of amniotic fluid normal. Apgar rate was 1 after 1 min. Immediate nasotracheal intubation and ventilation were given. Under direct laryngoscopy a wide inter-arytenoidal cleft was seen which extended to below the glottis level. Radiologically both lungs showed signs of aspiration. Extubation was performed on day 7. During cine-radiography of swallowing, contrast medium was seen to enter the upper trachea (Fig. 2). At tracheo-oesophagoscopy the suspicion of a cleft larynx was confirmed. Widely separated arytenoids with partial cleft of the cricoïd lamina were found. Crying was faint and the flow of saliva excessive. At first nutrition was maintained via duodenal tube only. Oral nutrition was started in the fourth month. The subsequent course was uneventful. Surgical correction of the larynx defect was not necessary.
Except for a ventricular septal defect, no other deformities were found.

2—Patient C.B., male: The mother was a 25-year-old para-one, who had taken thiamazole during the first 6 weeks of pregnancy and confinement was uneventful. The apgar score was 9-9-9 and the amount of amniotic fluid normal. Birth weight 3200 g, length 51 cm, no visible external malformations. Cyanosis and shock developed after the first hour of life. The husky cry was striking. Naso-tracheal intubation relieved the respiratory insufficiency immediately. Extubation, after transfer to a Children's Hospital, was followed by renewed tachyypnoe, stridor on inspiration and expiration and cyanosis. Under therapy with 35% oxygen and antibiotics because of suspected septicaemia, the patient's condition stabilised. On day 4, after feeding, sudden worsening of the condition and cyanosis necessitated further intubation. No anomaly of the larynx was seen at any of the intubations under direct laryngoscopy. On day 15, after feeding by duodenal tube, milk was aspirated beside the endotracheal tube. Direct laryngoscopy with careful inspection of the larynx demonstrated a cleft larynx. Through this defect the endotracheal tube appeared to be shifted dorsally towards the oesophagus. A cine-radiograph of swallowing showed the passage of contrast medium from the larynx to the trachea (Fig. 3). Tracheo-oesophagoscopy under general anaesthetic showed a cleft reaching the 6th tracheal segment. On day 20 after tracheostomy surgical closure was performed through a right-sided lateral pharyngotomy. The cleft extended in the posterior mid-line from above the larynx to the 6th tracheal segment. After widespread incision of the oesophagus and tracheal mucous membrane of the defective area, and after preparation of a middle layer of muscular tissue of constrictor pharyngis and oesophagus, a three-layered closure was made with Dexon Single Strand sutures 6 × 0 using fibrin glue. The healing process was free from complication. The post-operative course was complicated by marked gastro-oesophageal reflux, which resulted in repeated food aspiration. This was overcome by an open gastric tube and gradually increasing feeds through a duodenal tube. When aged 10 months the child was discharged. Decannulation was not yet possible, but the tracheostomy was helpful for bronchial toilet. The child's development is progressing normally.

3—Patient P.S., male: was the child of a 32-year-old mother. He had one 6-year-old healthy sister. A Caesarian section was performed because of maternal indication. Amniotic fluid was normal. The apgar index was 6-3-9. Because of repeated attacks of asphyxia, intubation was undertaken. After transfer from the maternity department the following findings were reported: at 2 h old, a mature newborn infant of 3220 g; diffuse cyanosis; inspiratory stridor and epigastric indrawings; hypertelorism; marked cleft lip and palate; crepes over both lungs; hypoplasias over the full length of the penis. The abdomen was unremarkable and both testicles had descended. Because of increasing respiratory insufficiency the patient was reintubated and placed on assisted ventilation. The first feed through a gastric tube was immediately followed by symptoms of aspiration. At an X-ray investigation with instillation of a water soluble contrast medium, a transit of medium into the trachea was shown. On day 33 the child was transferred to the Kinder-chirurgische Klinik des Kinderkrankenhauses der Stadt Köln, FRG (Prof. Dr. D. Helbig), and a surgical closure of the tracheo-oesophageal cleft was performed from the middle of the trachea to the 3rd tracheal segment using a left-sided cervical approach. Simultaneously a gastrostomy was performed and nutrition was started through a duodenal tube. The child was transferred to us for closure of the remaining laryngo-tracheo-oesophageal cleft. On the 58th day the defect of the dorsal larynx and upper tracheal area was closed through a right-sided, lateral pharyngotomy and a tracheostomy was formed. The cleft was closed after incision and preparation of the tracheal and oesophageal mucous membrane and after insertion of a muscle layer of constrictor and oesophageal muscle tissue between the mucous membranes. After that a three layered closure was made with atraumatic Dexon Single Strand sutures 6 × 0 and two instillations of 0.5 ml fibrin glue.