Laryngotracheoesophageal Cleft; Neonatal Presentation and Diagnostic Challenges

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Abstract
Congenital laryngeal clefts are rare developmental disorders of the upper airway accounting for 0.3% to 0.5% of all congenital anomalies of the larynx. The study is reporting the research team’s first experience with this anomaly at the Armed Force Hospital Southern Region to emphasize the difficulties and the challenges in the diagnosis as well as the treatment of such infants. This is a 32 week preterm female infant with antenatal ultrasound diagnosis of isolated esophageal atresia. After delivery the team faced a great difficulty in intubation and passing nasogastric tube. A large NGT size 12FR passed to the stomach, and contrast study showed abnormal anteriorly located esophagus, which suggested a common esophageotracheal tract. Esophagogram and soft fibro-optic endoscopy was done by ENT consultant which confirmed the presence of long segment laryngo–racheo-esophageal cleft. The infant was transferred to tertiary center where she died from respiratory failure at the age of 6 months.

Keywords: Laryngotracheoesophageal cleft. (LTC), Esophageal Atresia, Intubation, Neonates

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Introduction
Congenital laryngeal clefts are rare developmental disorders of the upper airway accounting for 0.3% to 0.5% of all congenital anomalies of the larynx1. This anomaly results from arrested medial fusion of the lateral aspects of the laryngo-tracheal groove2. The stage of development in which it occurs determines the extent of the cleft. Upper airway clefts range in severity from small soft tissue defects in the interarytenoid region to complete clefts of the larynx, trachea, and esophagus3. Presentations ranged from being virtually asymptomatic throughout life (type I) to being incompatible with life (type IV) 3. Timing of diagnosis is crucial to the successful
treatment of severe clefts. Complete Type IV of laryngotracheoesophageal cleft has mortality rates greater than 90%. Hospital management of these infants has been consistently difficult problem, which need multidisciplinary approach from perinatalogist, neonatologist, pediatric surgeon and pediatric otolaryngologist. This study is reporting its team’s first experience with this anomaly at the Armed Force Hospital southern region in Saudi Arabia to emphasize the difficulties and challenges in diagnosis, as well as the treatment of such infants.

**Case report**

This is a 32 week preterm girl baby born to a 42-year-old G1 P9+1 Saudi mother. She had routine prenatal care, underwent a fetal ultrasonic study at 24 week of gestation, which showed polyhydramnios, a poorly visualized fetal stomach, and a questionable isolated esophageal atresia. She continued her pregnancy with regular antenatal follow up, with no antenatal intervention. The mother was admitted at 32 weeks of gestation because of severe abdominal discomfort secondary to polyhydramnios and received two doses of dexamethasone. Next day, cardiotocography (CTG) revealed severe fetal bradycardia. After emergency caesarean section, a female infant was born with Apgar score 3, 4 and 8 at 1, 5 and 10 minutes respectively. At delivery, she was covered with excessive amniotic fluid, required air way suction and intubation with size 3mm oro-tracheal tube. The endotracheal tube was inserted with great difficulty. On admission to neonatal intensive care unit (NICU), baby maintained normal vital signs with birth weight of 1500gm and other growth parameters on the 10th centile. She had minor distinctive features with hairy fore head, microophthalmia, small cleft soft palate, other systemic examinations were normal. She connected to mechanical ventilation through Oro-tracheal tube size 3mm. To confirm antenatal diagnosis of possible oesophageal atresia, the team faced great difficulties in passing a small nasogastric tube (NGT) size 6-8FR (Figure 1). On one occasion, a large NGT size 10 FR was passed to the right main bronchus (Figure 2). A large NGT size 12FR passed to the stomach, but contrast study showed abnormal anterior located esophagus suggesting a common esophageo-tracheal tract (Figure 3). CT scan chest and upper airway showed difficulties in visualizing the esophagus and normal lower air way. Esophagogram (Figure 4) and soft fibro-optic endoscopy done by ENT consultant confirmed the presence of (type IV) long segment laryngo-tracheoesophageal cleft. Patient was evaluated by the team pediatric cardiologist and genetist. Echo cardiogram showed right sided aortic arch with small non significant patent ductus arteriosus (PDA). Chromosomal study was normal, and the baby did not fit in to any specific syndrome.

A gastrostomy tube was inserted after Nissen fundoplication and she received full enteral feeding with infant formula. She was referred to higher tertiary center to be operated after she achieved good size. Unfortunately the infant died at 6 month of age from respiratory failure secondary to aspiration pneumonia.

**Discussion**

Laryngotracheoesophageal clefts are rare developmental anomalies of the upper aero digestive tract. Richter was the first to describe this disorder in 1792 based on his examination of a newborn with feeding difficulties. It was not until 150 years later that additional cases were reported, including the first successful repair of a laryngeal cleft by Pettersson in 1955. This case is the first case reported in Saudi Arabia. There are no pathognomonic prenatal findings associated with laryngeal clefts and/or laryngotracheoesophageal clefts. The nonspecific finding of polyhydramnios observed in this case had been reported in many other reported cases with airway clefts. The baby in this case born prematurely at 32 weeks of gestation: Moungthong and Holinger found an association between laryngeal clefts and prematurity in six of eight cases.
A high index of suspicion is required for early diagnosis to reduce morbidity and mortality.

Post natal diagnosis is often difficult to establish. Successful passage of nasogastric tube to the stomach will rule out esophageal atresia and associated tracheoesophageal fistula which is present in 20-37% of cases\textsuperscript{10}. Passing of the tube to the right or left main bronchus or anterior position of the nasogastric tube on X-ray examination may suggest the defect\textsuperscript{10}. Barium (Gastrografin) esophagogram may also show aspiration through posterior laryngeal spillage or a common proximal laryngotracheoesophageal lumen\textsuperscript{11}. The research team confirmed the diagnosis after seeing the barium spillage in to the airway. Rigid Endoscopy, contrast radiographic studies and fibroptic Endoscopic evaluation of swallowing (FEES) are the golden standard for diagnosis. The use of rigid endoscopy for confirming the diagnosis, and evaluation of minor defects had been reported by Hilinger LD in 1985\textsuperscript{12}. It will show the height of the posterior cricoids lamina and laryngo-oesophageal common cavity. Palpation of the inter-arytenoid area With forceps is needed in case of occult or
type 1 cleft. Association with Cardiac and genetic disorders have been associated with this anomaly in 68%-78%, which include digestive 16 -67%, genitourinary 14 -44%, cardiac 16-33% and pulmonary in 2 -9%. Gastrostomy was performed after Nissen fundoplication to prevent the complication of Gastric reflux, and to control any air leak during positive pressure ventilation. In addition to surgical management of the reflux, these patients often require aggressive anti-reflux protocols at the time of diagnosis including medical management of proton pump inhibitors, pro-kinetics, and histamine type 2 antagonists, reverse Trendelenburg positioning during aggressive feeding.

Nutritional status is another significant aspect to the preoperative and postoperative care of these patients with complete airway clefts. They must have sufficient caloric and protein intake to meet their higher metabolic demands associated with wound healing and neonatal growth. We achieved that through gastrostomy feeding, but some cases may require feeding through jejunostomies.

The time elapsed from diagnosis to reconstruction should be as short as possible to decrease the degree of reflux-induced esophageal tracheitis and pulmonary aspiration. During this time, it is imperative to incorporate multiple specialties in the management of the patient. The multidisciplinary team should be composed of an otolaryngologist, pediatric surgeon, anesthesiologist, and neonatal intensive care specialist. A cardiologist should be implored to assess any cardiac malformations. A geneticist may be called on to evaluate any syndromic or genetic disorders. This team of physicians can then formulate the best approach for the surgical correction of the cleft based on the specific anatomical defect, other congenital defects, and the general status of the patient. Despite early detection and diagnosis the prognosis remains unfavorable with a mortality of 46% secondary to recurrent pulmonary aspiration.

**Conclusion**

Laryngeal clefts and laryngotracheoesophageal clefts are rare developmental defects of the upper aerodigestive tract. Critical factors in the successful management of these patients include a multi-disciplinary team-oriented approach with experience neonatologist, safe management of the airway, early and aggressive management of gastric reflux, nutritional sustenance, and early surgical intervention. Through increased awareness of clinical presentation and diagnostic techniques of laryngeal and laryngotracheoesophageal clefts, otolaryngologists can make earlier diagnoses of laryngeal clefts and/or laryngotracheoesophageal clefts and can implement prompt and comprehensive treatment.

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**References**


