LETTER TO EDITOR (VIEWERS CHOICE)

## LARYNGO-TRACHEO-ESOPHAGEAL CLEFT WITH HYPOTHYROIDISM

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A 3 month old male child presented with recurrent episodes of nasal regurgitation, severe choking and respiratory distress that led to repeated admissions in various hospitals. The child was full term, first in birth order and delivered by caesarean section due to failed induction. The neonate needed resuscitation at the time of birth. Due to severe respiratory distress and birth asphyxia, neonate remained in neonatal intensive care unit (NICU) for 7-10 days. Thereafter, the child was having difficulty while taking bottle-feed and invariably developed choking while taking feed. Examination of the child at the time of admission revealed sick, febrile, anemic child with coarse facial features, dry and coarse skin and marked respiratory distress. Child was malnourished (weight = 2.9 kg). Examination of chest revealed marked indrawing of intercostals spaces and retraction of subcostal margins with bilateral extensive coarse crepitations. First heart sound was loud with grade III systolic murmur. Other systems were normal. Laboratory investigations revealed dimorphic anemia and toxic granulation in the peripheral blood film. Blood culture grew staphylococcus aureus. Serum T3 level was 2.0 pg/dl (normal 3.1-8.6 pg/dl and serum T4 was 0.76 g/dl (normal 12-33 g/dl) and TSH level of 18.94 IU/ml (normal level 1.7-9.1 IU/ml) were suggestive of hypothyroidism. X-ray chest showed bilateral patches of infiltration but normal sized heart. MRI brain did not reveal any abnormality. Echocardiography revealed 2mm size ventricular defect. Water soluble non-ionic contrast esophagogram was performed with help of Ryle's tube which was placed at C4-C5 Level. Dye was injected instantaneously with force, which showed normal passage of contrast through esophagus into the stomach. The child did not develop choking or respiration distress. No evidence of tracheo-esophageal fistula was found (Fig 1). After removing Ryle's tube, a few drops of the same contrast when given orally, a communication between hypopharynx (upper part of esophagus) and larynx was visualized. The child developed choking and respiration distress after the procedure (oral intake of dye). These findings suggested the presence of laryngo-tracheo-esophageal cleft (LTEC), a rare malformation (Fig 2). Direct laryngoscopy could not be attempted due to the large tongue. Endoscopy by the pediatric surgeon also confirmed the presence of LTEC.

Laryngotracheoesophageal Cleft is a congenital midline defect of the posterior part of larynx and trachea and the anterior wall of esophagus. It is rare but significant cause of congenital respiratory distress and aspiration (1). The true incidence may be higher, but the approximate incidence of LTEC is one in 10,000 to 20,000 live births and 0.4% in all patients with laryngeal abnormalities (2). Males are affected more commonly than females. There is no consistent pattern of inheritance as most cases are sporadic, however, according to Phelan and Finlay, familial occurrence with autosomal dominant pattern has been reported (3,4).

The trachea and esophagus are foregut derivatives. At 25 days, the laryngotracheal septum develops and begins to fuse in a cephalad direction. It separates the distal trachea from the developing esophagus. The tracheo-esophageal septum completes upto the level of the first ring of trachea by 5th week. LTEC develops as a result of failure of tracheo-esophageal septum development. The phase at interruption determines the length of cleft (5,6). According to Benjamin and Inglis, LTEC is of 4 types (Type I - involving interarytenoid musculature, Type II - involving cricoid only, Type III - involving proximal laryngo-tracheo-esophagus and type IV the whole of thoracic tracheo-esophageal septum (7). As evident from contrast esophagogram and endoscopy, the Type II LTEC was present in the reported case. Associated anomalies like tracheo-esophageal fistula - 20%, gastrointestinal (anal atresia, Meckel's diverticulum, cleft lip and gastroesophageal reflux), gastro-urinary, pulmonary (bronchial and tracheal stenosis) and various types of cardiovascular malformation, may occur with LTEC (8,9). Congenital heart disease, hypoparathyroidism, laryngo-tracheo-esophageal anomalies and autoimmune disorder (late onset autoimmune hypothyroidism) have been described in DiGeorge syndrome. But in the reported case, hypothyroidism along with small sized ventricular septal defect were present though the child did not have other features of DiGeorge syndrome. G-syndrome and Pallister Hall Syndrome, may be associated with LTEC (10).

The infant with LTEC is at risk from repeated aspiration of feed and saliva, recurrent pneumonias and respiratory distress (1,8). Minor LTEC type-I clefts may be asymptomatic. Typically, cleft patients are said to present with classical triad of increased salivation, strider and a low soundless cry.

Management of LTEC includes stabilization of the airway. Healthy patient with LTEC Type-I, only needs intubation and early repair. Tracheostomy is usually required in cleft type II through IV, which allows improved surgical exposure and minimizes post-operative tracheal trauma. Delayed repair results in life threatening pulmonary complications. Various approaches for surgical correction have been adopted and modified to do repair depending upon the associated anomalies (11-13).

Prognosis is always guarded, as review of 85 cases by Roth et al in 1983 revealed overall mortality of 46% (43% type I and II, 42% for type III and 93% for type IV). Of all the patients who died, 67% had other associated serious congenital malformations. Despite secure closure of LTEC, disturbances of swallowing often persist (1,14).

It is therefore recommended the LTEC should be kept in mind in a newborn presenting with: (a) excessive frothiness and drooling of secretions (b) recurrent aspiration and choking during feed (c) repeated attacks of cyanosis and (d) recurrent pneumonias.

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