

# Laryngotracheo-oesophageal Cleft

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## Introduction

Laryngotracheo-oesophageal cleft should be considered in the differential diagnosis of any newborn presenting with respiratory distress and choking with feeding. The history and presenting symptoms may be identical with oesophageal atresia with or without tracheo-oesophageal fistula. Laryngotracheo-oesophageal cleft is a condition that varies greatly in anatomical extent and clinical severity. In the mildest form the only abnormality is the absence of the interarytenoid muscle and the patient may survive into adult life, with the history of hoarseness and recurrent respiratory infections. The more severe forms in which the cricoid and some or all of the tracheal cartilaginous rings are incomplete posteriorly are fatal unless corrected surgically.

LTOC is a rare anomaly, thus seldom suspected. This delays the diagnosis and cause high mortality. This paper includes a detailed case report of one patient, illustrating an important manoeuvre at endoscopy, which helped in early diagnosis and subsequent surgical management of this child.

## Case Report

Three month old male infant, came to us with complains of choking, dusky facial discolouration on feeds with respiratory distress and recurrent episodes of respiratory infections.

On examination, general condition was fair with respiratory rate of 40/minute, with audible crepts on right side of lung.

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Haemogram was within normal range, with X-ray showing right side pneumonitis. Cine-oesophagogram, using barium with patient in prone position and gradually withdrawing the end hole catheter, revealed spillage of dye into the trachea (Fig. 1).

Bronchoscopy was done, using cystoscope with camera which showed a common channel extending beyond the cricoid lamina to the cervical trachea i.e. type 2 variety of tracheo oesophageal cleft.

Right side cervical incision at the level of cricoid cartilage was taken. Laryngotracheo-oesophageal cleft was approached through lateral pharyngotomy. Normal oesophagus and trachea were identified and separation of the two tubes were completed by incising the cleft in such a manner that more tissue was left towards the tracheal side to create neo membranous portion of the trachea and neo oesophagus (Fig. 2).

While dissecting, vagus and the recurrent laryngeal nerves were taken care of. Closure of Oesophagus and trachea was done from caudal to cranial direction using absorbable vicryl sutures. Flap of sternocleidomastoid was kept in between trachea and oesophagus. Drain was left in situ.

Post operatively patient had leak. Gastrostomy was

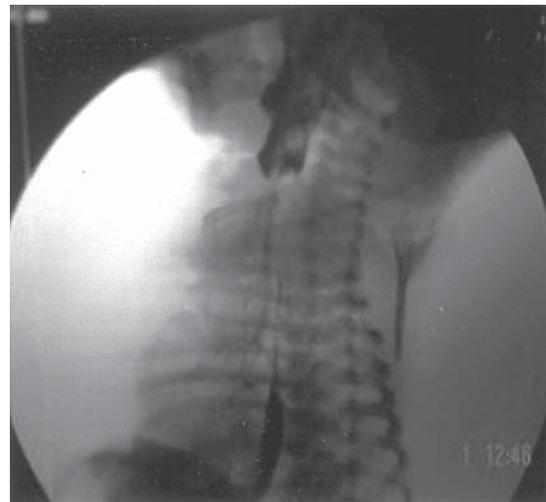


Fig. 1 : Showing cine oesophagogram of our patient

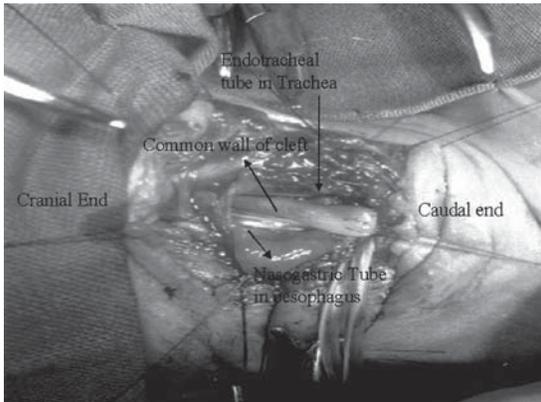


Fig. 2 : Peroperative photograph showing type 2 cleft

done and later patient was discharged.

### Discussion

The incidence of congenital anomalies of the larynx is approximately 1 in 2000 live births.<sup>1</sup> Laryngotracheo-oesophageal clefts, comprise only 0.3% of these anomalies.<sup>2</sup>

As with the embryogenesis of LTOC, firstly there is arrest of the cranial advancement of the tracheoesophageal septum, which subsequently, prevents, the dorsal fusion of the creoid cartilage, which develops from the fifth or sixth branchial arch. Points against this accepted notion are, LTOC's would have been commoner as compared to TEF, but review of literature reveals vice versa. Secondly clefts, do not have tissue deficiency, rather the edges herniate into the larynx or trachea as a soft tissue masses.<sup>3</sup>

Petterson<sup>4</sup> recognized three types of clefts as shown in Figure 3.

Type 1 in which the cleft involves the interarytenoid muscles and the cricoid lamina; Type 2 - in which the cleft extends caudal to include some of the tracheal rings; and Type 3 - in which the cleft extends to the carina and involves all the tracheal rings. Ryan *et al*<sup>9</sup> suggested type 4 cleft which extends beyond the carina to involve one or

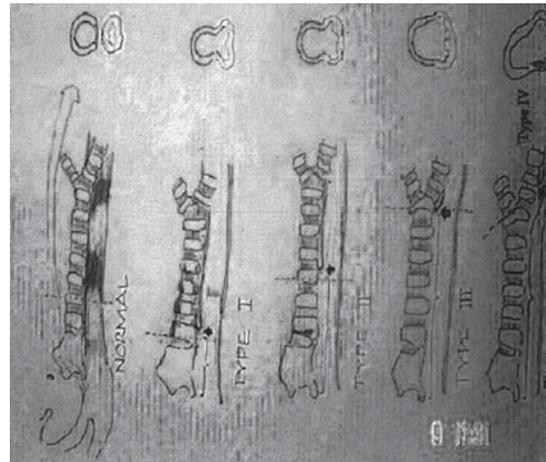


Fig. 3 : Types of Cleft

both main stem bronchi.

Laryngotracheo-oesophageal clefts should be suspected in an infant with a hoarse cry, stridor, and aspiration pneumonia, this condition is sometimes familial as reported by Cameron and Williams.<sup>5</sup> They also report that it is associated with harelip, cleft palate, and oesophageal atresia with tracheo-oesophageal fistula. LTOC symptoms closely resemble those of H type fistula as stated by Armitage.<sup>6</sup>

Barium oesophagogram and bronchoscopy are the important investigations for diagnosis of cleft. Barium oesophagogram under image intensifier screening, will show, contrast in oesophagus and trachea, however, it is difficult to differentiate whether there is spillage of dye due to cleft or spillover, at the level of larynx. For this purpose, it is better to do, oesophagogram in prone position, with end hole catheter, which was slowly withdrawn, hence we will see the caudal most part of the cleft. Bronchoscopy will, delineate anatomy of cleft clearly. Under general anaesthesia during intubation preferential slippage of endotracheal tube into oesophagus raises suspicion of cleft. Endoscopic visualization under general anaesthesia will

confirm the cleft. In the presence of cleft, it is easy to negotiate, bronchoscope into airway, in the presence of endotracheal tube. For endoscopy in our patients we have used, cystoscope with camera. Under general anaesthesia with endotracheal intubation, it is easy to negotiate scope, in the subglottic region. This manoeuvre, would open the edges of the cleft. Temporary tracheostomy and decompression gastrostomy offers stabilization of child. Definite treatment depends on the type of the cleft. Endoscopic management is successful in type 1 clefts as advocated by Dubois.<sup>7</sup>

There are two approaches for the repair of clefts anterior cervical and lateral cervical approach. Anterior cervical approach is the approach of choice for type 2 as advocated by Lipshutz *et al.*<sup>8</sup> Depending on type of cleft, approach can be either cervical, or combined cervicothoracic.

Anterior cervical approach for repair of clefts has advantage of, direct visibility due to good exposure, minimum dissection in the neck and reduced risk of damage to the recurrent laryngeal nerve. Disadvantage of this is overlapping suture lines and need for tracheostomy. The lateral approach poses a risk for damage to recurrent laryngeal nerve but it is easy way of access to the cleft and avoids contiguous suture lines and subsequent refistulization. Sternocleidomastoid interposition is required to prevent refistulization. Technical aspect of surgery involves going to normal oesophagus first, and then dissect cranially, divide the cleft in a manner so that more tissue is left, with trachea, this ensures better

closure of trachea and prevents stenosis.

Type 3 and 4 requires combined thoracic and cervical approach reported by Ryan *et al*<sup>9</sup> and Simpson *et al.*<sup>10</sup> Leaks accounts for 50% of repair. In long term, tracheal stenosis, pharyngo-oesophageal dysfunction and GOR are to be monitored.

The prognosis depends upon the extent of cleft and associated malformations.

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