Laryngo-tracheo-oesophageal cleft

A cleft larynx is a rare but significant cause of congenital respiratory distress and aspiration. Management of this defect is complex and recent advances in the overall mortality remains unacceptable high. Although over 179 cases have been reported to date there continues to be significant delay in diagnosis, perhaps due to a reluctance to perform neonatal bronchoscopy, and appropriate intervention which contributes to increased morbidity and mortality. However these patients often have other significant abnormalities which may independently affect their outcome. Of these gastro-oesophageal reflux, microgastria, ventricular septal defect, bronchial and tracheal stenosis are among the most clinically significant.

The true incidence is one in 10,000 to 20,000 live births, although the true incidence may be higher. There is no consistent pattern of inheritance but males are affected more commonly than females and sporadic familial occurrences have been described. Laryngeal clefts have also been reported with the G syndrome (X linked or autosomal dominant) and in the Pallister-Hall syndrome. Clefts are classified into four types based on the length of involvement of the trachea. The more extensive the defect the greater the potential for fatal pulmonary complications and poor outcome. The defect extends to the cricoid in type 1, involves the cricoid and/or proximal trachea in type 2, extends to the carina in type 3, and into one or both main stem bronchi in type 4. Fortunately types 1 and 2 clefts constitute the most common abnormality seen.

A laryngeal cleft arises from abnormal separation of the larynx-trachea and the oesophagus. At 25 days the laryngo-tracheal septum develops and begins to fuse in a cephalad direction and so separate the distal trachea from the developing oesophagus. At the same time the cricoid cartilage develops as two lateral centres of condensation from the sixth branchial arch. Dorsal fusion of the cricoid plate is complete by day 50-54 and laryngeal muscular development then ensues. The range of abnormalities seen results from defects in cricoid condensation or fusion (types 1 and 2) and or failure of fusion of the laryngotracheal septum (types 3 and 4). This embryological difference is highlighted by the occurrence of isolated tracheo-oesophageal fistula that occurs in the absence of any laryngeal abnormality. The exact cause of the disorder is unknown but substance abuse in the first trimester and neural crest abnormalities have been cited as potential factors.

The result is an abnormal communication between the larynx, trachea, and oesophagus. The infant is thus at risk from repeated aspiration of food and saliva, recurrent pneumonia and respiratory distress, although minor type 1 clefts may be asymptomatic. A laryngeal cleft should be considered in any infant who develops aspiration and cyanosis after feeding and respiratory distress, although other causes such as choanal atresia, tracheo-oesophageal fistula, tracheomalacia, and laryngopharyngeal dysmotility should also be considered. Typically, cleft patients are said to present with a classical triad of increased salivation, stridor, and a low soundless cry. However this triad is rare and most infants develop aspiration and cyanosis after feeding and this combined with postpartum respiratory distress should stimulate urgent bronchoscopy. The cry is usually harsh and not silent and stridor is uncommon. Secretions are increased and there is significant difficulty with feeding. Contrast radiography may show the abnormal communication but this is often difficult to distinguish from simple spil over. A lateral neck x ray film may show an anteriorly displaced nasogastric tube or a posteriorly displaced endotracheal tube. Successful management of these infants requires a low threshold for bronchoscopy and thus early diagnosis, a secure and stable airway, and provision of adequate nutrition. Most require a tracheostomy and gastrostomy at the outset. A fundoplication may be required if there is troublesome gastro-oesophageal reflux. Other procedures such as gastric transsection are not
routinely required since the availability of total parenteral nutrition. Alternatively a feeding jejunostomy or transpyloric jejunal tube may be used. There is considerable controversy over the timing of surgical intervention. Should these patients undergo early single staged correction or is it appropriate to plan a staged procedure with an initial tracheostomy, gastrostomy, and fundoplication followed by delayed repair? If the diagnosis can be made shortly after birth and the patient is stable then early repair should be considered; however many infants experience a delay in diagnosis and require considerable respiratory and nutritional treatment before correction. Early repair reduces the severity of associated pulmonary dysfunction and facilitates postoperative weaning from the ventilator. Conversely delayed repair facilitates weight gain and may make surgical correction easier. Whatever the timing of surgery it is essential that it be performed in a centre with paediatric surgical expertise.

Type I defects may be corrected endoscopically if not responding to standard antireflux treatment. There is no consensus as to the appropriate surgical approach for the remaining type 2, 3, and 4 clefts. An anterior approach may exacerbate coexistent laryngeal weakness and may be difficult in the presence of a tracheostomy, whereas a lateral approach may potentially injure the recurrent laryngeal nerve. Correction of type 4 defects is more difficult and requires a cervical-thoracic approach in addition to innovative anaesthetic techniques to maintain and secure the airway during and after surgery. Further surgery is often required to correct gastro-oesophageal reflux, tracheobronchomalacia, and microgastria. Recurrent tracheo-oesophageal fistula remains a problem and wound breakdown may occur in as many as 50% of patients. Interposition of a vascularised muscle flap may help to prevent breakdown.

Overall survival with aggressive treatment is 70% but is significantly less for those patients with extensive type IV defects and severe associated anomalies. While noteworthy advances have been made in surgical and anaesthetic care there remains an unwillingness to refer neonates for diagnostic endoscopy. A lower threshold for intervention coupled with further improvements in surgical technique especially in the treatment of microgastria and tracheobronchial stenosis may further improve survival.

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