



Rigid Bronchoscopy Through Tracheostome in a Case of Pierre Robin Syndrome – a Case Report

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Abstract: A 3-year-old girl with Pierre Robin Syndrome, having complaints of respiratory distress in supine position and disturbance during sleep due to airway obstruction, was scheduled for adenoidectomy. After ensuring all available precautionary measures along with facilities for emergency tracheostomy, gas induction without muscle relaxation was done in a posture at where her respiratory distress was least. The glottis could not be visualized by direct laryngoscopy in any way. Consecutive three attempts were failed and intubation was successful on fourth attempt with a gum elastic bougie. Then after curarisation, adenoidectomy was performed. Five teeth were avulsed during the instrumentations, 4 out of 5 could be recovered. Fluoroscopic screening located the 5th tooth in right main bronchus. The patient was reversed and kept on spontaneous breathing with ETT in situ. On 1st POD, gas induction was done for removal of the lost tooth. ETT was removed and with muscle relaxation, rigid bronchoscopy was attempted without success. Repeated attempts were made with intermittent ventilation and at one stage bag-mask ventilation became impossible with sharp fall of SpO₂, even down to zero for a while. Emergency tracheostomy was asked for and a needle cricothyroidectomy was done immediately. The patient had a period of anoxia during establishment of safe airway through tracheostome. After ensuring a safe airway through tracheostomy and stabilization of her vital parameters, rigid bronchoscopy was performed through tracheostome and the tooth was removed. Adequate spontaneous ventilation was achieved on the next morning without any neurological deficit. Tracheostome was closed on 8th POD under general anaesthesia and the patient was discharged with excellent cure of her symptoms and kept on regular follow-up in ENT OPD.



Key words: Pierre Robin Syndrome, difficult airway, lost tooth, rigid bronchoscopy, emergency tracheostomy.

Introduction: The Pierre Robin syndrome presents as airway obstruction secondary to micrognathia with retrogenia and glossoptosis (tongue prolapse). An oval or cleft palate is present in 50% of cases. It is usually revealed by acute respiratory distress in the neonate and is a major concern as infants with Pierre Robin syndrome are known to be difficult to intubate. Although many airway management protocols have been described, mostly in case reports and short series, the gold standard for difficult intubation is considered to be fiberoptically assisted intubation¹. With unavailability of a fiberoptic bronchoscope a case of Pierre Robin syndrome was approached to remove an enlarged adenoid which caused difficulties in respiration and severe disturbance during sleep. A paraglossal approach was adopted with a gum elastic-bougie to manage the difficult airway. This clinical report is a narration of the serious consequences those were encountered during the anaesthetic management.

Case Report: A 3-year-old girl, a diagnosed case of Pierre Robin Syndrome was admitted to Combined Military Hospital, Dhaka for adenoidectomy. She had respiratory distress in supine position and had disturbance during sleep due to airway obstruction. On preanaesthetic assessment, she was found to have hypoplastic mandible, large tongue, high arched intact palate, suprasternal & intercostal indrawing with inspiratory stridor.

Relevant investigation reports were within normal limits excepts the X-ray neck lateral view showed mallar bone hypoplasia with hypoplastic mandible, irregular soft tissue shadow occupying oropharynx, narrowing of airway in laryngopharynx. CT scan of brain, face and nasopharynx (non contrast) showed grossly enlarged adenoid with high arched palate.

With full preparations to face difficult intubation, induction was done with preoxygenation in propped up position to minimize her respiratory distress with N₂O+O₂+Halothane. No sedative, opioid or muscle relaxants were given. Direct laryngoscopy revealed an inverse pyramidal shaped oropharynx, there was hardly any space in the oropharynx and glottis could not be visualized because larynx was anteriorly placed. With repeated attempt and with paraglossal approach the child was intubated with the help of a gum elastic bougie by 3.0 mm armoured ETT guided with thread reinforced malleable stylets. After intubation, muscle relaxant and analgesia were given and adenoidectomy was carried out.

During instrumentation, 5 deciduous teeth from upper jaw were avulsed, 4 out of 5 teeth could be taken out but 5th could not be located in spite of thorough search. Post-operative fluoroscopic screening showed the tooth was in and around the right principal bronchus (Fig. 1). She was kept intubated in ICU under sedation and analgesia. Intravenous steroids were given 6 hourly.

On next morning she was scheduled for rigid bronchoscopy for removal of accidental impacted tooth in right main bronchus. Repeated attempts of rigid bronchoscopy through glottis were made without success. At one stage bag mask ventilation became impossible, arterial O₂ saturation came down to zero, the child developed bradycardia, there was no effective circulation

and cardiac arrest was inevitable. To maintain oxygenation & ventilation cricothyroid puncture was done by two wide bore cannula and emergency tracheostomy was done as a life saving procedure. Thiopentone sodium 50 mg/kg was given iv after the hypoxic episode for cerebral protection.

After ensuring a safe airway through tracheostomy and stabilization of her vital parameters, rigid bronchoscopy was performed through tracheostomy stoma and the tooth was taken out. She was then nursed in post-operative ward on controlled mechanical ventilation for 24 hours with thiopentone sodium 10 µg/kg/min, fentanyl 2.5 µg iv 2 hourly, dexamethasone 2.5 mg iv 8 hourly. On the following day, the baby was fully recovered found no neurological deficit and she was haemodynamically stable. Then she was weaned off the mechanical ventilator (Fig. 2). Tracheostomy wound was closed on the following day under GA with spontaneous ventilation without any complications. She was nursed in ICU for next 24 hours. Her HR, BP, RR and SPO₂ were monitored closely, the period was uneventful. She was then discharged from hospital (Fig. 3). Now she is on regular follow-up in ENT OPD, her difficulty in breathing and snoring has reduced and she can sleep undisturbed.



Fig. 1 Fluoroscopic view of chest showing a tooth in the right bronchus. This is one of the five teeth those were avulsed accidentally during instrumentation while managing the difficult airway.



Fig. 2 The patient in the post-operative ward after emergency tracheostomy.



Fig. 3 The photograph of the patient after closure of the tracheostome and before discharge.

Discussion: Congenital micrognathia with glossoptosis is a relatively rare condition that occurs in 50,000 births and is referred to as Pierre Robin syndrome. Patients should be screened for Stickler syndrome, which includes myopia in infancy, retinal detachment and preventable blindness. Associated coanal atresia should be excluded by passing a catheter through each nostril².

Depending on the degree of microretrognathia, severe respiratory difficulties may be present soon after birth. The tongue is displaced posteriorly and may be hyperplastic. In the supine position, this causes obstruction of the oral and nasal pharynx thus preventing normal breathing³. Increasing inspiratory efforts create negative pressure in the pharynx, which further enhances already severe obstruction. Unrelieved upper airway obstruction may result in the development of cor pulmonale and congestive heart failure⁴. These respiratory problems are frequently compounded by aspiration when the child attempts to feed. Placing the child in prone position allows the tongue to fall forward and partially relieves the airway obstruction. Swallowing is impaired and aspiration pneumonia is frequent, weight gain usually poor, and "glossoptotic cachectia" may develop⁵.

Pierre Robin, in his original report, considered this lesion incompatible with survival after the second year of life. As recently as 20 years ago, mortality in infants with this syndrome was as high as 50% despite treatment. With intensive nursing care in a constantly maintained prone position and gavage feeding, the majority of these infants can be managed conservatively^{6,7}. Surgical intervention becomes necessary when retrognathia is more than 1 cm in relation to the maxilla and/or when recurrent obstruction or aspiration episodes occur despite proper positioning. Glossopexy by creation of a lip-tongue adhesion has become the preferred method of surgical treatment. Tracheostomy, with its inherent problems in the neonate, is rarely performed. Because most micrognathic mandibles have normal growth potential, to a large extent the deformity tends to correct itself within the first few months of life if nutrition can be maintained and if hypoxic episodes can be avoided⁸.

Anaesthetic management presents several challenges. Foremost is the maintenance of an adequate airway prior to endotracheal intubation since these patients usually become obstructed immediately when they are turned supine. In addition, all the problems of anaesthesia in the neonate have to be considered, including transitional circulation, sensitivity to inhalation agents and muscle relaxants, the danger of hyperoxia, and thermal instability. Sedation should not be administered preoperatively because relaxation during sleep tends to aggravate the existing airway



obstruction. Atropine 0.01 to 0.02 mg/kg or a minimum of 0.1 mg should be administered intramuscularly 30 minutes prior to induction to block parasympathetic reflexes and reduce oral secretions, which are frequently copious⁹.

The infant should be brought to the operating room in the prone position. Infants with micrognathia often present a formidable obstacle to endotracheal intubation. Prior to any attempt at laryngoscopy, a surgeon capable of performing tracheostomy in an infant must be present in the operating room with appropriate sets. A Robertshaw or Wis-hipple blade, slightly larger than predicted by age, could be useful. Introduction of the laryngoscope blade on the extreme right side (retromolar) with concurrent adjustment of the larynx by cricoid pressure often results in improved visualization. In small infants an awake intubation should be performed. The infant is preoxygenated in the prone position and then turned supine and restrained with the head extended by an assistant. Complete airway obstruction frequently occurs the moment the infant is turned supine. Insertion of an oropharyngeal airway or introduction of the laryngoscope blade displaces the tongue anteriorly and allows the infant to breathe. Oxygen can be insufflated via a catheter held next to the laryngoscope blade or with an Oxy blade. The larynx is often at a lower position than anticipated, and with gentle pressure on the cricoid by an assistant the arytenoids and the posterior aspect of the cords may become visible. Because of the anterior position of the larynx, the endotracheal tube may have to be curved slightly, with the help of a stylet, in order to place it through the cords. If the larynx cannot be visualized the endotracheal tube can be placed blindly by following breath sounds. Special care is needed during blind intubation to avoid trauma to the laryngeal structures. With experience, an ultra thin fiber -optic bronchoscope can be used after topical anaesthesia of the nose and pharynx to perform an awake nasal intubation with surprising ease. As soon as proper endotracheal tube position has been ascertained by auscultation of the chest, anaesthesia is induced with halothane and nitrous oxide via a non-rebreathing system. The Jackson Rees modification of the Ayres T-piece with a heated in-line cascade humidifier is preferred. The infant then is allowed to breathe spontaneously and the endotracheal tube securely taped^{10, 11}.

In older infants, inhalation induction with halothane and oxygen should be carried out with an oropharyngeal or nasopharyngeal airway in place. Adequate time for the laryngoscopy can be achieved by deepening the level of anaesthesia. Muscle relaxants should not be used since airway obstruction is more likely to occur when soft tissues are relaxed. After intubation nitrous oxide can be added to the anaesthetic mixture. In the neonate, halothane may cause severe hypotension, particularly in the presence of dehydration. This can occur despite an inadequate depth of anaesthesia. Nitrous oxide and oxygen and ketamine, 0.5 to 1.0 mg/kg intravenously, with controlled ventilation but without the use of relaxants provide adequate anaesthesia. The children usually extubated in the operating room and observed for airway obstruction. Only after patency of the airway has been ascertained, the patient is transferred to the recovery room and placed in a high-humidity hood for several hours. During this interval the child is cared for in either a prone or lateral position. From the recovery room, the patient should be transferred to an intensive care unit. The nasopharyngeal airways are left in place for 3 to 4 days until all oedema has resolved. Oral feeding is withheld and the infant fed by gavages. These children are kept in the hospital 2-4 weeks until they



feed well orally, show satisfactory weight gain and have no current episodes of respiratory obstruction^{12, 13}.

Conclusion: Nothing can be more embarrassing for an anaesthesiologist than being stuck up into a failure to intubate an already induced patient. Developments of respiratory obstruction, ventilatory failure, hypoxia, cardiac arrest or losing the patient on the operation table are the worst possible experiences for an anaesthesiologist. A high degree of suspicion, careful individual assessment and to ensure the most appropriate technique remains the key to successful management of patients with anticipated difficult airway.

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