

Problems in the Anesthetic Management of Pierre Robin and Treacher Collin Syndromes

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Babies born with craniofacial disorders can truly challenge the anesthesiologists. Pierre Robin syndrome consists of a recessed and hypoplastic mandible (micrognathia) and glossoptosis(1). The vacuum created in the pharynx exacerbates the obstruction of the airway with each inspiration and sucking(2). This anomaly is mostly associated with incomplete cleft palate (70%), complete cleft palate (10%) and without a cleft palate (20%). There may be associated cardiac and ocular problems.

Babies with Treacher Collin syndrome(4) have hypoplastic maxilla, zygoma and mandible, and narrow nasal, pharyngeal and auditory passages. Deafness is common. External ears are low set and hypoplastic, nose is large and beaklike. Mandibular rami are short with hypoplastic muscles of mastication. Palate is cleft and

high arched. This manuscript outlines the problems in the anesthetic management of Pierre Robin and a Treacher Collin syndrome.

Material and Methods

Three cases of Pierre Robin syndrome and 2 cases of Treacher Collin syndrome of age group from 1 yr 6 mo to 1 yr 8 mo were operated upon for cleft palate. Their average body weight was 8 kg. All were girls. Babies were premedicated with trimeprazine tartarate syrup (3 mg/kg of body weight) given orally 2 hours before operation. Anesthesia was induced with oxygen, nitrous oxide and halothane. Induction was delayed due to increased obstruction by the tongue and soft tissue. So a suture was placed through the tongue and an assistant pulled the tongue forward to help during laryngoscopy and intubation. The intubation in all cases were extremely difficult. Ultimately 3.5-4 mm diameter plain endotracheal tubes were put inside the trachea. Anesthesia was maintained with oxygen, nitrous oxide and halothane through Jackson Rees(5) modification of Ayre's 'T' Piece. Then fresh gas flow was administered (2.5-3 times the minute volume to prevent rebreathing). A small wet throat pack was put around the endotracheal tube to prevent aspiration of blood and mucus into the respiratory tract. The patients were put in Trendelenberg position. 5% dextrose (278 m. mol of glucose/litre) and quarter saline (38 m mol of saline/litre) was given at the rate of 100 ml/kg/day. In 3 cases the blood loss was more than 10% and blood transfusion was given. When the operation was over, pharyngeal suction was done very carefully and the throat pack was removed prior to extubation. After that oxygen was administered. Immediately after extubation the infants were conscious, but when they

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were allowed to breathe, they were cyanotic. Hence, a long silk suture in the tongue was made and administration of oxygen was continued. As soon as the tongue was pulled forward(6) cyanosis improved. In the post-operative period they were kept quiet by small dose of analgesic to prevent bleeding from the operation site. The infants were placed on their side with the head extended and the lower arm placed under the head. Airway obstruction was watched very carefully.

Discussion

Babies born with facio-maxillary anomalies pose a grave danger during induction and maintenance of anesthesia. The airway obstruction is due to hypoplastic mandible and incomplete development of intrinsic muscles of tongue(7). It may give rise to recurrent attacks of respiratory embarrassment in the neonate, persisting for a couple of months and causing permanent cerebral damage from hypoxia. Besides asphyxial mortality, there is still a significant casualty (20%) from the associated cardiovascular anomaly or from aspiration and pneumonia. Prolonged hypoxia and hypercarbia with acidosis can produce pulmonary vasoconstriction and cor pulmonale(3).

In many cases the infant can be nursed through these dangerous weeks by keeping them in face down position in a special frame and a soft rolled towel placed under this shoulder and lower abdomen. But the desirability of relying on this regime depends wholly upon the availability of skilled nurses day and night. One temporary method(8) to relieve airway obstruction is by inserting a catheter through the nose with tip of the catheter placed in the stomach. This procedure prevents the formation of negative pressure in the pharynx and

also prevents suction of the tongue into the pharynx. The other alternatives are to fix the tongue by suturing it to the lip or buccal sulcus, or to perform a tracheostomy. Many suturing operations have been described but none is very satisfactory. Tracheostomy is preferable if surgical intervention is required. Tracheostomy(9) is technically a difficult procedure due to short fat neck and tiny soft trachea.

The cause of airway obstruction in the post-operative period was attributed to the closure of the palatal gap which was present pre-operatively. The reduction of the airway by the operation and the muscular hypotonia following anesthesia and surgery produce conditions in which an emergency can readily set in. After operation, though the infants were fully conscious, the tongue fell back against the soft palate and obstructed the airway causing cyanosis. So, a tongue suture was placed to prevent its falling back.

The frequency and seriousness of post-operative obstruction justifies postponement of closure of the cleft palate in cases of Pierre Robin syndrome for longer than for ordinary cases of cleft palate alone(10).

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