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Short Communication

Anesthetic Care in Special Clinical Situations: Pierre Robin Sequence, Mitochondrial Disease and Mucopolysaccharidoses

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There are clinical situations in which the anesthetic care are difficult and need to pay more attention because of risk of serious complications. These diseases are Pierre Robin sequence, mucopolysaccharidoses and mitochondrial disorder.

What are the best way to reduce or avoid this risk?

The clinical triad of *Pierre Robin sequence* (PRS) is micrognathia (small mandible), glossoptosis (backward, downward displacement of the base of the tongue) and airway obstruction . Some authors have described as micrognathia, glossoptosis, and cleft palate. The incidence is 1:10000 births. When we saw patients with PRS, we need to exclude some syndromes (Table 1). How we manage these children with PRS in anesthetic procedures?

Syndrome	Associated anomalies	Anesthetic concerns
Fetal alcohol syndrome	Microcephaly, maxillary hypoplasia, micrognathia, short neck, ventricular septal defect, cognitive developmental delay, hyperactivity	May be difficult to ventilate and intubate. Preoperative echo may be indicated. Consider subacute bacterial endocarditis prophylaxis. May be uncooperative
Stickler	Marfanoid appearance, airway obstruction, micrognathia, joint laxity, mitral valve prolapse	May be difficult to ventilate and intubate. Care with positioning
Treacher Collins	Craniofacial clefting, mandibular hypoplasia. May have obstructive sleep apnea. May have congenital heart disease	May be difficult or impossible to intubate. Preoperative echo may be indicated. Consider subacute bacterial endocarditis prophylaxis
Velocardiofacial	Microdeletion of chromosome 22, microcephaly, micrognathia, congenital cardiac disease. May have developmental delay, neonatal hypocalcemia, T-cell immune deficiency	May be difficult to intubate. Preoperative echo may be indicated. Consider subacute bacterial endocarditis prophylaxis. Blood products need to be irradiated

Table 1: *Syndromes associated with Pierre Robin sequence.*

PRS can present significant challenges for the anesthesia provider including airway obstruction and difficult intubation. This may result in intraoperative and postoperative respiratory complications. Patients who cannot tolerate supine positioning will be more difficult to mask ventilate and may require airway adjuncts such as oral pharyngeal airways, NPA (nasopharyngeal airways) and laryngeal mask

airways (LMAs) (Figure 1). The primary concern in the postoperative management of the patient with PRS is airway obstruction that may result in hypoxia, negative pressure pulmonary edema, and death. There are several cases reports of significant tongue edema after cleft palate surgery resulting in postoperative airway obstruction. The mouth retractor should be released every 1 to 2 hours to reduce the risk of tongue edema.

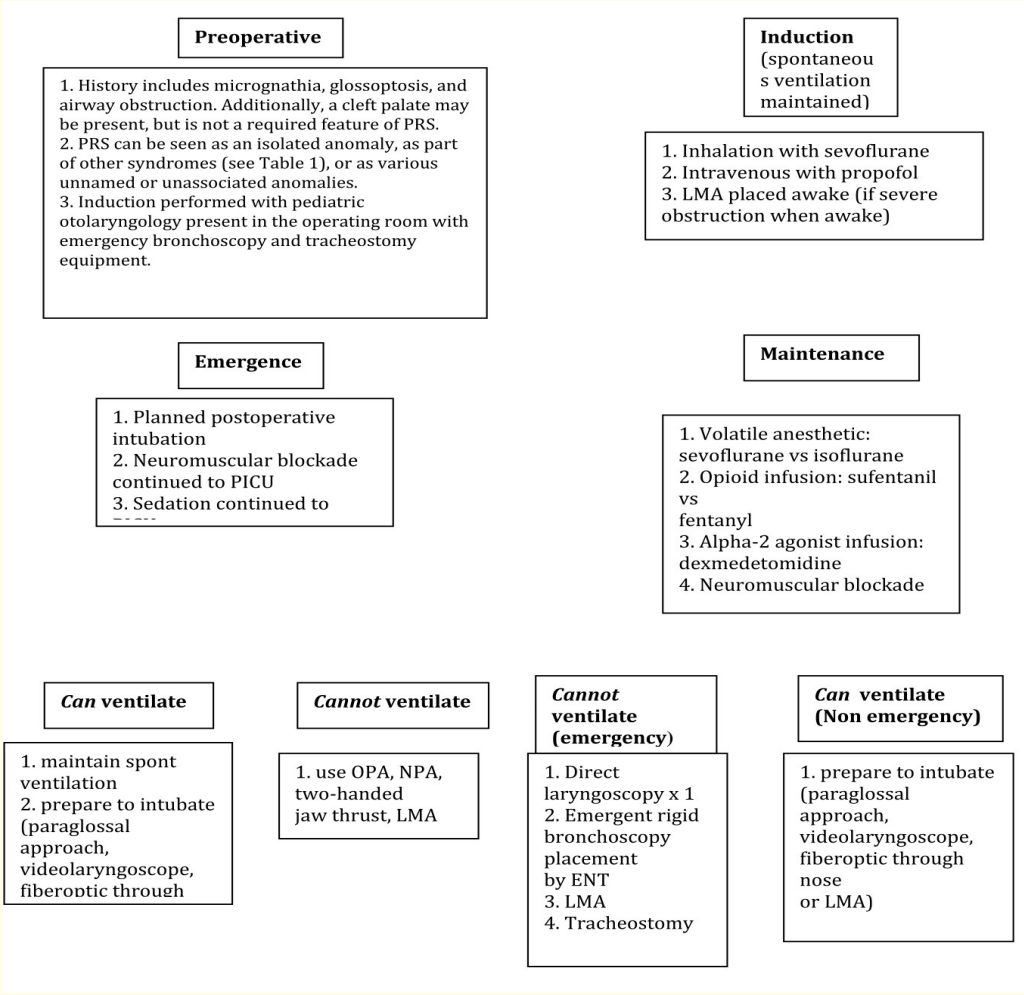

Figure 1: *Guidelines for Anesthesia Management of Neonates with Pierre Robin Sequence (PRS) or Severe Micrognathia for TLA (tongue lip adhesion), MDO (mandibular distraction osteogenesis) or Tracheostomy.*

Mucopolysaccharidoses (MPS) is caused by a deficiency of glycosaminoglycan (GAG) metabolism enzymes. Involvements of heart and lungs are the causes of mortality. The incidence is 1:25000 newborns. Despite breakthroughs in the anesthetic and surgical approach, perioperative mortality is still high among patients. Preoperative evaluation includes a thorough clinical history and note of prior surgical procedures or anesthesias. Is important to do imaging to identify odontoid dysplasia or atlantoaxial subluxation. The risk of airway obstruction mostly is because of hypertrophy of the tonsils and adenoids, macroglossia, anterior larynx, decreased tracheal gauge, stiff-

ness of the temporomandibular joint, short neck and cervical instability. If we can choose locoregional techniques will be better but if not possible, orotracheal intubation, either by laryngoscopy or fiberoptic techniques, laryngeal mask or tracheostomy are the main choice. As MPS progresses in spite of enzyme replacement therapy, techniques that previously had been successful to manage the airway may not be successful for the current procedure. Following major surgery recovery should be performed in the intensive care unit with extubation in a controlled environment. In relation to preanesthetic medication, the sedatives drugs should be avoided because of the risk of respiratory depression. For decreased secretions, anticholinergics may be administered. There is a risk of upper airway obstruction and there have been reports of post extubation pulmonary edema presumably due to forced expiration against a narrowed and thickened glottis. The degree of postoperative monitoring is dependent on the surgical procedure and preoperative condition of the patient. Intensive care is not mandatory, but intensive care facilities should be on site. If sleep apnea is present use regional local anaesthetic blocks and avoid excessive intraoperative opiates. Continuous oximetry monitoring to detect airway obstruction episodes and desaturation. Consider applying CPAP (Continuous Positive Airway Pressure) or BiPAP (Bilevel positive airway pressure). Postoperative chest physiotherapy has a role in reducing respiratory complications.

Mitochondrial disorder (MD), is now recognized as an important cause of a wide range of neurological, cardiac, muscle and endocrine disorders. The incidence is about 1 per 4 - 5,000 live births. Muscle biopsy or other tests needing anesthesia are required for diagnosis. All general anesthetic depressed mitochondrial function. Among them, are the volatile anesthetics and propofol. This last one cause propofol infusion syndrome, characterized by a severe lactic acidosis, rhabdomyolysis and lipidemia, which can lead to death (Table 2). Parenteral sedatives like, etomidate, ketamine and barbiturates are also complex I inhibitors. Fortunately, there are many reports of patients with mitochondrial disease that can tolerate volatile anesthetics, propofol and local anesthetics. These patients needs tight control during and after general anesthesia.

Young age
Dosages of > 5 mg/kg per hour, duration of therapy exceeding 48 hours
Underlying illness (respiratory or neurologic)
Concomitant catecholamine or steroid in the setting of acute neurologic or inflammatory diseases

Table 2: *Risk Factors for Development of Propofol Infusion Syndrome.*

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