



Anesthesia management for syndromic patients posted for various procedures

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Abstract: Congenital disorders are rare which includes genetic and chromosomal disorders. These disorders present with variety of clinical considerations challenging to anesthesiologist which also includes difficult airway. Essential information can be obtained from preoperative assessment which includes general physical examination, airway examination, laboratory and diagnostic interventions. It is advised to discuss with senior and well experienced anesthesiologist and always have Plan A, B, C before going ahead with challenging cases.

Keywords: Syndrome child, Downs syndrome, Apert syndrome, Prader willi syndrome.

INTRODUCTION

Syndromic child posted for ENT procedures constitutes spectrum of challenges for Anesthesiologists as children usually presents with variety of manifestations which require careful management as children are related to difficult airway and hearing compromise and feeding difficulties. Since last 2 decades survival has been progressively increased due to advances in medical, surgical care. These advances have produced increase incidences of children with congenital malformations. (1)

The approach for evaluation and proper management of syndromic child includes thorough history, airway, neck and head examination and appropriate investigations according to the type of syndrome (2). Management of syndromic child is best achieved by multidisciplinary approach including anesthesiologist, surgeon, rehabilitation team according to the needs of the child.

Case series:

Downs syndrome: A eight years old child weighing 30 kg with Downs syndrome posted for myringotomy and bilateral gromet insertion with history of recurrent ear infections and snoring in night. Patient was evaluated in anesthesia clinic. Patient was anxious, agitated, Not cooperative, difficulty in speech and poor verbal communication. Careful airway examination considering high risk for difficult airway and Bag mask ventilation, revealed macroglossia, microstomia, poor neck mobility. Classified as Mallampati grade IV and typical facial features of Downs syndrome were present. Planned for general anesthesia with supraglottic airway. On the day of surgery Premedication with oral midazolam 0.5 mg/KG was administered 30 minutes before shifting patient to OT. Preoperative patient was little sedated and IV cannula 20 G was secured. At the induction Fentanyl 60 mcg, Propofol 50 mg, was given and I-Gel No 3 was inserted, obtained satisfactory ventilation. Maintenance was achieved with desflurane and mechanical ventilation was set as volume controlled ventilation (VCV) later changed to spontaneous ventilation.

Duration of surgery was 30 minutes, vitals were stable throughout the procedure. Patient was extubated

Pediatric Apert Syndrome (syndromic craniosynostosis): A 16 years old male, 50 KG diagnosed as Apert syndrome since birth was seen in anesthesia clinic with chronic sinusitis posted for Functional endoscopic sinus surgery (FESS). Patient underwent multiple surgeries since childhood which includes craniofacial reconstruction, plastic reconstruction surgery for upper and limbs all under general anesthesia. No mention of difficult airway was mentioned during past surgeries and was intubated with Video laryngoscope. Acrocephaly, brachycephaly, high prominent forehead, Strabismus, proptosis, mandibular prognathism and depressed nasal bridge was noted in Facial examination. Limb defects of upper and lower limbs including fusion of digits of hand and legs. Oral examination revealed hypoplastic maxilla, high arched palate, ectopic tooth eruptions, macrodontia is noted. Premedication with midazolam, Atropine was given according to body weight. As past surgical history was without difficult airway we have induced with IV fentanyl 100 mcg, propofol 100 mg, rocuronium 25 mg was given and was intubated with Glidescope in first attempt. Anesthesia was maintained with 2% sevoflurane and was extubated at end of surgery with no anesthesia complications.

Prader-Willi syndrome: A 4 year old male patient weight 28 kg was scheduled for dental rehabilitation procedure. Preoperative evaluation revealed history of mental retardation, morbid obesity, short neck, sleep apnoea syndrome, type II diabetes mellitus, recurrent episodes of upper respiratory tract infection. Anesthesia was induced with sevoflurane (inhalational) and IV cannula was secured. IV propofol, fentanyl, rocuronium used for intubation. Intubated with 4.0 ETT USING Macintosh blade, Maintained with sevoflurane. At the end of surgery recovery (spontaneous breathing was delayed) and nalbuphine was given to reverse effect of fentanyl to reverse breathing.

Pierre robin sequence (stickler syndrome): Patient was a 38 day old boy born at 38 weeks of pregnancy with weight of 2.9 kg.

He was diagnosed to have Pierre Robin sequence with symptoms of difficult feeding and respiratory distress. On detailed history revealed History of recovered URTI, features suggestive of reflux and aspiration was noted. On examination micrognathia, Posterior displacement of tongue base(Glossoptosis) and there was difficult visualization of pharyngeal view. Elective tracheostomy was scheduled in view of worsening of airway obstruction. After connecting essential monitors to the patient .Anesthesia was induced with volatile anesthetic agent(sevoflurane) and IV cannula was secured after adequate preoxygenation. Iv fentanyl 6 mcg, Iv propofol 6 mg was administered. It was possible to fit mask for his face and good ventilation was achieved using 2 hand technique and with help of nasal airway .No neuromuscular blocker was administered. Attempted to intubate with 3.0 uncuffed ETT using Glidescope, could be intubated with difficulty using bougie in first attempt. After confirming position of tracheostomy tube ETT was removed, and patient was shifted to NICU for elective ventilation and weaning off was planned later.

Discussion

Anesthesia considerations differ for each syndromic child posted for particular surgery. So preoperative evaluation is key for good perioperative management and outcome. Patient's with downs syndrome have recurrent respiratory tract infections, subglottic stenosis, enlarged tongue, delayed developmental delay, mental retardation, epilepsy, Atlanto axial instability, poor muscle tone, impaired immunity , obesity. Downs syndrome babies are generally gentle and cheerful but have tendency of hyperactivity so it is better to premedicate with anxiolytics before surgery. A detailed cardiovascular evaluation ECG, echocardiography to rule out cardiovascular abnormalities should be advised. Atlantoaxial instability , neck pain, limited neck mobility suggests difficult laryngoscopy and the goal should be kept to keep head and neck in neutral position(3).

Apert syndrome is autosomal dominant inheritance pattern consists of craniofacial synostosis, midline hypoplasia and syndactyly. Craniofacial abnormalities are associated with airway obstruction, and may cause obstructive sleep apnoea.(4) Limited mouth opening, High mallampati score, tracheal stenosis, restricted temporomandibular joint function leads to difficult intubation in children. Mostly patients require induction of anesthesia with inhalational agents without neuromuscular blocking agents to maintain airway patency.

Prader willi syndrome is a genetic disorder characterized by hypotonia, hypopontia, hypogonadism that is associated with obesity and lower life expectancy(5). Airway management may be complicated by poor dentition, micrognathia, palatal abnormalities, limited neck mobility. Anesthesia concerns includes obesity, sleep apnoea, difficult IV access, risk of perioperative respiratory failure, violent behaviour, disturbances in thermos regulation.(6)

Pierre Robin Sequence(PRS): is a triad of micrognathia, posterior inferior displacement of tongue(Glossoptosis) and airway obstruction. PRS can be associated with other syndromes such as stickler syndrome, teacher collin syndrome, velocardiofacial syndrome(7). Patients with PRS have airway obstruction, OSA, chronic hypoxia, increased sensitivity to opioids. Direct laryngoscopy , visualization of vocal cords may be challenging and infants may require awake fiberoptic in some cases. Difficult bag mask ventilation, severe airway obstruction leading to

tracheostomy should be anticipated. If possible consider awake fiberoptic intubation while maintaining spontaneous ventilation.

Conclusion

Syndromic children posted for various surgeries are increasing in all surgical specialities, but children posted for ENT/ Dental procedures are of great importance in view of shared airway, no scope of regional anesthesia, risk of aspiration. Of importance is assessment of mental behavior , assessment of cervical spine, general physical examination, cardiovascular and respiratory system assessment.(8) Perioperative complications like difficult cannulation, difficult airway, bronchospasm, extubation stridor are regular complications to be kept in mind. Post operative management in recovery/pediatric care with good analgesia ensures proper recovery of child(9).

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Nil.

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