Supraglottic airway and caudal epidural for anesthetic management of a child with Larsen syndrome

Sir,

Larsen syndrome (LS) is a rare inherited defect of collagen formation with multiple congenital large joint dislocations associated with characteristic facial, hand and feet anomalies and vertebral malformations resulting in cervical spine instability.[1]

A 5-year-old female, weighing 10 kg, was scheduled for bilateral reduction of congenitally dislocated knees. A diagnosis of LS was confirmed at the genetic clinic of the hospital. She had no history of stridor, breathlessness, syncope or prior hospitalization. The child appeared small for her age. She had a small, flat face with a prominent forehead, wide intercanthal distance, and a Mallampati class II airway. A pectus excavatum was present. The child had generalized hypotonia, but there was no cervical spine instability or any other vertebral anomaly, confirmed radiologically. Clinically, there was no evidence of any cardiac defect and her electrocardiogram (ECG) and two-dimensional Echo was normal.

The child was premedicated with oral midazolam 5 mg 30 min prior to shifting to the operating room, where a difficult airway cart was kept standby. Care of the cervical spine was taken during shifting, and the child was gently positioned on the operating table. Standard noninvasive monitors (automated noninvasive blood pressure, ECG, Pulse Oximeter) were placed. Induction was performed by tidal breathing of sevoflurane 7% with 100% oxygen. Neck extension was avoided during mask ventilation, and no difficulty in ventilation was encountered. An intravenous access was attained during this time. Once adequate depth of anesthesia was achieved, the airway was secured with an LMA Classic (size 2). The inspired concentration of sevoflurane was then decreased to 3% in a mixture of 40% oxygen and air. A caudal block was performed in the left lateral decubitus position with 10 ml of 0.25% bupivacaine with 0.5 mg morphine using a Crawford caudal needle. An assistant ensured that the neck remained in a neutral position when the child was turned in the lateral position, during the procedure and getting back to supine the position. Anesthesia was maintained with sevoflurane (minimum alveolar concentration 1) and a mixture of 40% oxygen and air and the child was allowed to breathe spontaneously to maintain the end tidal CO_2 between 34-38 mmHg. The patient remained comfortable and hemodynamically stable during the surgery that lasted 90 min. Postoperative course remained uneventful. There was no nausea, vomiting, pruritis or urinary retention. Intravenous paracetamol (150 mg) was started 18 h post-operatively when she complained of mild discomfort for the 1st time and repeated at 8 hourly intervals for the next 3 days.

Larsen syndrome is a dysmorphic syndrome that is both genetically and phenotypically variable with both autosomal dominant and recessive patterns. The recessive forms are more severe. It is believed to be a generalized mesenchymal disorder where the formation of mature dense collagen fibers is affected. Typical facies include hypertelorism, frontal bossing, flattened nasal bridge and palate defects. Of prime importance to the anesthesiologist are the associated defects of the airway, including laryngo-tracheomalacia, which may predispose the patient to a difficult airway and perioperative respiratory compromise.[2] These patients should be considered to have an unstable spine unless proved otherwise.[3] Minor trauma leading to quadriplegia and death have been reported.[3] These patients should be handled gently and positioned carefully. The frequent association of this syndrome with cervical spine anomalies mandates an X-ray of the cervical spine with extension and flexion lateral views. Postoperative croup due to the presence of subglottic stenosis is common and has been reported even when a smaller size endotracheal tube (ETT) is used.[3] Whenever possible a supraglottic airway device should be preferred for securing the airway.[4] This not only minimizes manipulation of the neck but also decreases postoperative coughing and the risk of airway collapse on emergence due to airway malacia.[5] The use of muscle relaxants should be avoided till such time the airway is secured. These patients tend to have similar cardiac profile to Marfan's syndrome and therefore a preoperative cardiac evaluation is warranted. Intraoperative cardiac arrest has been reported in a patient with cardiac compromise during spinal surgery.[6] A total intravenous technique with propofol may be a good alternative if general anesthesia cannot be avoided.

Regional analgesia has an important role to play since the surgeries are very painful, and the use of opioids may further contribute to perioperative respiratory problems.[4] In spite of this known advantage, the use of caudal epidurals has not been reported in these children. This is probably because of the high incidence of vertebral instability, presence of spina bifida and also maybe because majority of the cases reported underwent surgery of the spine. Addition of morphine prolonged the analgesic effects considerably. The use of opioids as caudal additives warrant appropriate monitoring in the postoperative period.

LS patients have multisystemic involvement with spinal instability warranting evaluation of vertebral anomalies. Use of supraglottic airways and regional anaesthesia may improve the margin of safety for these patients.

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