Child with Edward's syndrome for radiological procedure: An anesthetic challenge

Sir,

Edward's syndrome is rarely inherited disorder as a result of trisomy of chromosome 18. It usually manifests as serious cardiovascular, renal anomalies and orofacial deformity. We present tailored customized management in a challenging case of Edward's syndrome for magnetic resonance imaging (MRI) under general anesthesia. A 2-year-old male child, weighing 11 kg, presented to hospital with difficulty in walking. He was diagnosed to have Edward's syndrome. Preoperatively, the child was found to be with severe mental retardation and hypotonia. Airway examination could not be assessed other than externally evident facial deformity because of learning deficit of the child. Renal function tests were abnormal (serum creatinine of 1.5 mg/dl). In electrocardiogram (ECG), right bundle branch was evident. Two-dimensional echocardiogram findings were suggestive of moderate atrial septal defect (ASD) of 3.5 mm. MRI of the lungs showed pulmonary hypoplasia of the left lung [Figure 1]. This child was diagnosed to have right-sided kyphotic spine, microstomia, micrognothia, and limb deformities (rocker-bottom foot, fixed flexion deformity, radial hypoplasia, and arachnodactyly) with low-lying tethered cord with syrinx at lumbosacral level in imaging and confirmed to be a case of Edward's syndrome on genetic karyotype analysis. Before surgery, the child needed an MRI examination of the spine. Inside induction room adjacent to MRI suite, the child was induced with sevoflurane 8% and 100% oxygen followed by I-gel™ (size #2) insertion. Maintenance of anesthesia was carried on spontaneous ventilation with sevoflurane (0.4%), O_2+air (50:50 ratio), and intravenous (IV) dexmedetomidine infusion at the rate of 0.2 μg/kg/h. The procedure went uneventfully and lasted 45 min of duration. I-gel™ was removed; sevoflurane and drug infusion terminated at the end of procedure. The child was observed in postanesthesia recovery unit for the next 15 min till full regain of consciousness.

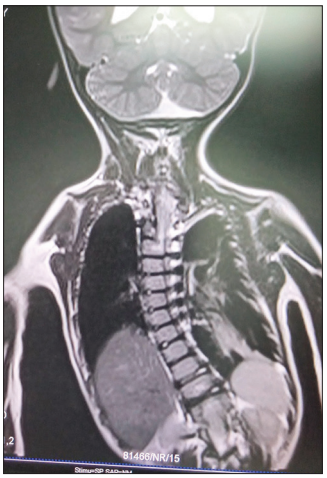

Figure 1: Magnetic resonance image of syndromic child showing dorsolumbar kyphosis of the spine with left pulmonary hypoplasia

Designing or following any anesthetic protocol is impractical in such patients in view of rarity and severity of associated comorbidities. Individualized approach is suggested according to experience and expertise of involved anesthesiologist.[1] ECG artifacts caused by electromagnetic fields of the magnetic resonance system can make detection of any arrhythmia or new onset morphologic changes very difficult. Moreover, I-gel™ is a helpful airway device for MRI in children in routine as well as emergency scenario. We considered the following child as primarily cardiac patient and so it seems prudent to provide anesthetic medications with few cardiac side effects. Hence, sevoflurane was opted which preserves systemic arterial pressure. The use of atracurium as muscle relaxant and sevoflurane among volatile agents would be a safe choice in these patients.[1] In contrast, we maintained anesthesia on spontaneous ventilation with IV sedation (dexmedetomidine) in view of avoiding polypharmacy used perioperatively that may interfere in their excretion with impaired renal functions. In patients with ASD, a potent volatile agent is preferred as the principal anesthetic agent because of increased pulmonary blood flow (PBF). This not just results in faster induction and postoperative extubation than IV agents.[2] We preferred dexmedetomidine infusion in low dose as maintenance drug with gaseous mixture (O_2: Air) because of its central sympatholytic activity which would have helped in condition of congestive heart failure if arisen as untoward event during the procedure unlike any inhalational agent.[3] Moreover, it has shown to significantly attenuate nephropathy (radiocontrast induced) by maintaining renal outer medullary blood flow.[4] However, the hypotonia of the patient allowed us to keep a low concentration/dose of both inhalational and IV agent. Nitrous oxide was avoided in view of further rise in pulmonary vascular resistance. The thought of preserving spontaneous ventilation in this patient was to avoid excessive anesthetic medications in perioperative period.

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Conflicts of interest

There are no conflicts of interest.

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