Anaesthetic management of a child with Varadi-Papp (orofacial digital syndrome type VI) syndrome

Sir,

Oro-facial digital syndrome type VI (OFDS type VI) (Varadi–Papp syndrome) poses unique perioperative and anaesthesia challenges. It is a rare genetic disorder with autosomal recessive pattern of inheritance presents with facial and oral malformations, central polydactyly, cerebellar abnormality and delayed psychomotor development as its common phenotypic spectrum. A 4-year-old female child with the diagnosis of OFDS type VI was posted for cleft palate repair. In addition to the typical clinical features of OFSD type VI, the child also had macrocephaly with a head circumference of 60 cm and BMI of 30 kg/m^2 on examination. Airway examination revealed grade-II cleft palate, high-arched palate and micrognathia. Preoperative electrocardiography and echocardiography revealed no abnormality. Magnetic resonance imaging of the brain revealed complete corpus callosum agenesis Dandy–Walker variant with hypoplasia of left frontal and parietal lobes. On the day of surgery, the child was premedicated with chlorpheneramine maleate and shifted to operating room. Anaesthesia was induced with 8% sevoflurane oxygen mixture. During induction, there was a brief period of bradycardia, which was treated with injection atropine. Though difficult airway cart was kept ready in view of anticipated difficult airway, the trachea was intubated using direct laryngoscope (Cormack–Lehane grade IIb) with the aid of Frova intubating catheter in a head-elevated position. Intraoperative course was uneventful and the trachea was extubated at the end of the surgery. The same child was posted 6 months later for syndactyly release. The surgery went uneventful though there was a similar bradycardia event at induction, which was recurrent yet responded to injection atropine. Orofacial digital syndrome is caused by a defective OFD gene which can be present either in sex chromosome (X-linked dominant inheritance; OFDS type I) or autosomes (autosomal recessive inheritance; OFDS type VI). It presents with specific patterns of signs and symptoms. Based on its pattern, there are 13 different types of OFDS[1] and they have oral, facial and digital anomalies as a common feature. Varadi–Papp syndrome is characterised by central polydactyly apart from the typical features of OFDS. Head-to-toe examination of the child revealed hypertelorism, micrognathia, crowded teeth, split tongue, cleft palate, depressed nasal bridge, central polydactyly, syndactyly, clubfoot, delayed psychomotor development, brain anomalies such as corpus callosum agenesis and features suggestive of Dandy–Walker malformation[2,3] [Figure 1]. Micrognathia, psychomotor

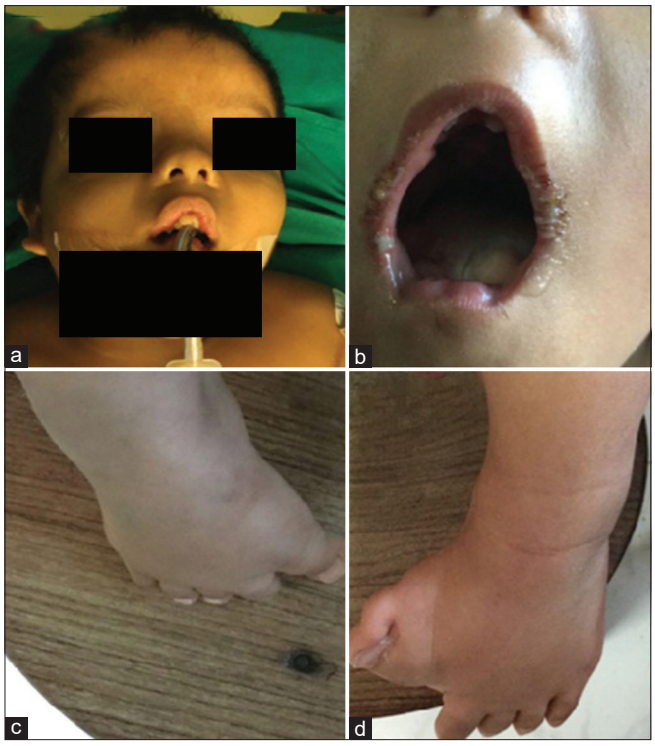

Figure 1: Clinical features of Varadi–Papp syndrome. (a) Depressed nasal bridge, hypertelorisim and crowded teeth; (b) Split tongue, tongue hamartoma and cleft palate; (c) Club foot; (d) Syndactyly

retardation along with macrocephaly and high BMI in this child posed unique anaesthesia challenge at the time of induction and intubation. Though preoperative electrocardiogram and echocardiography revealed no abnormality, OFDS has been associated with various cardiac malformations. The child had bradycardia episodes during induction on two different occasions. This may be attributed directly to Dandy–walker malformation causing raised intracranial pressure during induction rather due to a cardiac cause. There was one case report in a neonate with OFDS presenting with refractory bradycardia.[2] Corpus callosum agenesis has been associated with delayed recovery from anaesthesia.[4] Anticipating this, we used short acting opioids and muscle relaxant doses guided by neuromuscular monitor. Therefore, OFDS type VI needs careful preoperative evaluation to look for the central nervous system, cardiovascular and airway anomalies. Apart from routine perioperative paediatric concerns, particular focus must be on managing induction and intubation due to micrognathia, ensuring adequate anaesthesia, analgesia and neuromuscular recovery. With adequate preoperative evaluation and preparation, safe management of such a rare genetic disease would not be a herculean task.

Declaration of patient consent

The authors certify that they have obtained all appropriate consent forms from the parents of the patient. In the form, the parent/s has/have given consent for their child's images and other clinical information to be reported in the journal. The parent/s understands that her name and initial will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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