Myotonic dystrophy and inhalational anesthesia: Is it time to shun the fear?

Dear Editor,

Myotonic dystrophy type 1 (DM1, Steinert’s disease) is an autosomal dominant disorder of the neuromuscular system.

A 10-year old, 25-kg male child with DM1 was posted for mastoid exploration under general anesthesia. The child had decreased tone in jaw muscles and upper limbs, elongated face, micrognathia, maloccluded teeth, prominent incisors, and a high arched palate [Figure 1]. He had muscular impairment rating scale (MIRS) grade 2 and no neurodevelopmental delay or cardiac abnormalities. A written informed consent was taken by the child’s parents and the possibility of postoperative ventilation was explained.

As the child was uncooperative for IV cannula insertion, inhalational induction was done with sevoflurane and air-oxygen mixture (50:50). SpO2, electrocardiogram, and NIBP monitors were attached. Intravenous line was secured with a 22G cannula after achieving adequate anesthetic depth. Bispectral index (BIS), train of four (TOF) neuromuscular monitor, and an esophageal temperature probe were placed thereafter. Anesthesia was induced with intravenous fentanyl 50 μg and propofol 20 mg. Airway was secured after giving atracurium 12.5 mg IV with 5.5 mm cuffed endotracheal tube using C-MAC™ video laryngoscope blade #2 at TOF count zero (Cormack-Lehane grade 1).
Anesthesia was maintained with desflurane, oxygen-air 50:50 mixture (titrated to BIS of 40–60) with pressure control ventilation. Neuromuscular blockade (NMB) duration was 70 min with the first atracurium bolus dose. Hence, subsequently reduced (1 mg) doses were given at TOF count > 1. The surgery lasted for 4 h. NMB was reversed with neostigmine (1.25 mg IV) and glycopyrrolate (0.2 mg IV) when TOF count was ≥ 3. After ensuring the adequate return of motor power and airway reflexes, trachea was extubated in a fully awake state. Patient was monitored for 24 h postoperatively for muscle weakness or respiratory distress. Hourly oral temperature charting was done to monitor for hypothermia or hyperthermia.

Preferential use of intravenous anesthesia (IVA) without muscle relaxants is the general paradigm for anesthetic management in Steinert’s disease, but IVA use is also not devoid of side effects. There is an increased sensitivity to anesthetic agents and every drug should be titrated to effect. Use of inhalational anesthesia has been feared due to the presumed risk of malignant hyperthermia (MH). However, the current literature projects the risk of developing MH equivalent to the general population. Saitoh et al. used sevoflurane for induction and maintenance in a 14-year-old DM1 patient for orthopedic surgery without any adverse reactions. Gandhi et al. used desflurane anesthesia in a 33-year-old DM1 patient for laparoscopic cholecystectomy and did not report any significant adverse events. Hence, we used sevoflurane for induction and desflurane for maintenance of anesthesia under BIS guidance, especially because our patient did not have significant muscle weakness.

To conclude, inhalational agents may be used along with monitoring of the depth of anesthesia, neuromuscular function, and temperature in DM1 patients with lower MIRS grades.

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Conflicts of interest
There are no conflicts of interest.

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