Anesthetic challenges and successful management of a child with Pelizaeus–Merzbacher disease using general and caudal anesthesia

Madam,

We recently took care of a patient with Pelizaeus–Merzbacher disease (PMD) and wanted to highlight anesthetic implications and appropriate management related to this rare condition. Manifestations can include microcephaly, tracheomalacia, nystagmus, hypotonia, spasticity, gastroesophageal reflux, dysphagia, epileptic activity, and cognitive impairment. There is no definite curative treatment, and death usually occurs at an early age.[1] A difficult airway, risk for aspiration, respiratory depression, perioperative seizures, hypothermia, and positional injuries are all possible with PMD. The use of regional anesthesia in particular has been encouraged to avoid respiratory depression,[2] but never reported in literature.

We anesthetized a 30-month-old male baby requiring port-a-cath removal and circumcision using general and caudal anesthesia. His symptoms included choreoathetoid movements, nystagmus, and cognitive/motor dysfunction. Brain magnetic resonance imaging showed diffuse hypomyelination. He was in the 6th percentile for weight and 10th percentile for height. After obtaining a careful history from his mother, we mask-induced with sevoflurane and N_2O, after which a peripheral intravenous (i.v.) line was placed. To deepen the anesthetic and avoid neuromuscular blockade for intubation, a minimal bolus dose of propofol and fentanyl was administered. Glycopyrrolate was given to act as an antisialagogue. Intubation was successfully performed and a single-shot caudal block was performed as part of a multimodal analgesic approach that also included i.v. acetaminophen, dexmedetomidine, and local anesthetic infiltration at the port-a-cath site by the surgeon. We padded bony prominences and an underbody forced-air warmer was used to maintain intraoperative normothermia. After an uneventful procedure, the child was extubated awake, and then discharged home from recovery the same day without needing additional analgesics.

Significant clinical heterogeneity exists with PMD. It comprises a spectrum of three different types: classical, connatal, and intermediate transitional.[3] Our patient exhibited the classical form. After confirming lack of breathing problems with sleeping, nocturnal positional preference, issues with managing secretions, and symptoms of reflux, we believed a mask induction could be safely performed. To deal with potential decreases in pharyngeal tone after induction, appropriately sized oral/nasal airways were readily available. Anticholinergic effects helped mitigate secretion formation while under general anesthesia. Conversely, rapid sequence intubation should be considered if increased secretions and/or uncontrolled reflux are a concern, but as with upper motor neuron diseases and conditions with atrophic musculature, avoidance of succinylcholine due to the theoretical risk of hyperkalemia is advised.[1] Poor nutritional status and growth necessitate extra caution with positioning to prevent nerve injuries. A history of seizures treated with anticonvulsants could lead to altered hepatic drug metabolism, hematologic dysfunction, and hepatotoxicity. However, anticonvulsants should be continued on the day of surgery.[4] Dopaminergic antagonists

could lead to exacerbation of movement disorders and should be avoided. Ondansetron is the antiemetic of choice.

As the first reported use of regional anesthesia in a patient with PMD, our caudal block allowed minimization of anesthetic/analgesic-related respiratory depression. We encourage a thorough preoperative evaluation and perioperative planning as highlighted above, along with the use of regional anesthesia, for any patient with this leukodystrophy.

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Conflicts of interest
There are no conflicts of interest.

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