[NM-250] Use of dexmedetomidine for prevention of post-operative agitation in a 14 year-old male with Angelman's Syndrome

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Introduction: Angelman's Syndrome is a genetic disease caused by a chromosomal deletion between 15q11 and 15q13. Historically, children with Angelman's were termed "happy puppets" due to their frequent smiling and jerky gait. Main clinical features are severe developmental delay, speech impairment, ataxia, and epilepsy. Patients with Angelman's generally have favorable airways due to their wide mouth, protruding tongue, and prognathia. Severe intellectual impairment and delayed speech present challenges in the perioperative period. Use of antiepileptic medications may alter metabolism of anesthetic drugs(1).

Case Report: 14 yo (54kg) M with Angelman's Syndrome who had heel cord contractures presented for bilateral tendon lengthening. Patient was very playful and smiling, displaying typical characteristics of Angelman's. Previous anesthetics were complicated by significant agitation and combativeness on induction and emergence. Twenty mg of PO midazolam was given preoperatively. Still encountered difficulty restraining patient during mask induction, but thereafter underwent general anesthesia with LMA uneventfully. The patient was given dexmedetomidine as a slow bolus (0.2 mcg/kg) over 10 minutes prior to the end of the case. Patient emerged from anesthesia comfortable and required no additional sedation, analgesics, or restraint.

Discussion: Children with Angelman's pose certain anesthetic challenges. While their rambunctious nature may be charming in any other setting, it may lead to difficulty in the perioperative period. Dexmedetomidine, an alpha-2 agonist, has been widely used for pediatric procedural sedation and for sedation in ICU settings. In addition, several clinical trials have shown that dexmedetomidine can aid in preventing emergence agitation in pediatric patients(2,3). Dexmedetomidine was administered in doses ranging from 0.15 to 1 mcg/kg as a bolus dose or an infusion of 0.2 mcg/kg/hr in those studies. One of the possible side effects is bradycardia, which is undesirable in children. However, literature suggests bradycardia is infrequently encountered at the aforementioned doses and hemodynamically insignificant if it is encountered. Dexmedetomidine has also been shown to decrease opioid and anesthetic requirements and not have significant respiratory depression(2).

Conclusion: A low dose intraoperative dexmedetomidine bolus can help provide a smooth emergence from general anesthesia in pediatric patients. Dexmedetomidine can be used not only in healthy children but in those who by nature are boisterous such as patients with Angelman's, and in older children in whom restraint may be difficult.

References:
1. Baum et al. Anesthesia for Genetic, Metabolic, and Dysmorphic Syndrome of Childhood, Second Edition. Philadelphia: Lippincott Williams & Wilkins, 2007.
2. Patel et al. Dexmedetomidine Infusion for Analgesia and Prevention of Emergence Agitation in Children with Obstructive Sleep Apnea Syndrome Undergoing Tonsillectomy and Adenoidectomy. Anesth Analg. 2010 Oct;111(4):1004-10.
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