[NM-131] Anesthetic Management of Children with Arthrogryposis Syndromes

Gleich S, Weingarten T, Flick R, Nemergut M
Mayo Clinic , Rochester , MN, USA

Introduction: Arthrogryposis syndromes are characterized by multiple joint contractures secondary to extrinsic factors or intrinsic neuromuscular disease.[1] Currently, there is a paucity of studies examining anesthetic management in these children. Previous reports noted difficulties with intravenous catheter (IV) placement, airway management, and intraoperative hyperthermia in children with arthrogryposis syndromes.[2] The purpose of this study was to review our institution's anesthetic experience in children with arthrogryposis syndromes.

Methods: We performed a retrospective review of all patients 0-20 years of age with arthrogryposis syndromes who underwent anesthesia at Mayo Clinic in Rochester, Minnesota from 2008-2013. The medical records were examined for arthrogryposis diagnosis, procedure type, anesthetic type (general vs. sedation), anesthetic agents (volatile, succinylcholine, or nondepolarizing muscle relaxant administration), airway management, difficult IV placement, intraoperative hyperthermia (temperature >37.0°C), and evidence of malignant hyperthermia.

Results: During the study period, 18 patients underwent 48 anesthetics (81% general anesthesia, 19% sedation). There were 12 males (67%), with a median age of 4.1 [1.6, 10.7] years. Fifteen patients (83%) were diagnosed with Arthrogryposis Multiplex Congenita, with the remainder (17%) diagnosed with distal arthrogryposis syndromes. The most common type of surgery was orthopedic procedures (46%), followed by diagnostic studies (29%), otolaryngological and oral procedures (13%), and general surgical procedures (10%). The majority of patients were classified as ASA Physical Status 2 and 3 (58%, 31%). There were 5 instances of difficult endotracheal intubation. Four patients were successfully intubated with multiple direct laryngoscopies. There was 1 failed intubation, requiring LMA placement. Two patients had difficult IV placement, with 1 requiring ultrasound-guided peripheral IV placement, and another with placement of a scalp peripheral IV following failed attempts on the extremities. Due to concerns of possible malignant hyperthermia, 3 patients were treated with a non-triggering technique. Interestingly, 14 patients (78%) developed intraoperative temperatures >37°C and 5 patients (28%) developed intraoperative temperature >38°C. No patients developed malignant hyperthermia.

Conclusions: Children with arthrogryposis syndromes pose unique challenges to the pediatric anesthesiologist, including difficult airway management and IV placement. Although there was no evidence of malignant hyperthermia, intraoperative hyperthermia was common in our cohort. Further studies, with larger numbers of patients are needed to elucidate risk factors and potential complications of hyperthermia.

References:
1. Baum VC, O'Flaherty JE. Anesthesia for Genetic, Metabolic, and Dysmorphic Syndromes of Childhood. Second ed: Lippincott Williams & Wilkins; 2007.
2. Martin S, Tobias JD. Perioperative care of the child with arthrogryposis. Paediatric anaesthesia. Jan 2006;16(1):31-37.
