

Schiffmiller M, Pandya Shah S

Rutgers University - New Jersey Medical School , Newark , NJ, United states

Introduction:

Adams-Oliver Syndrome (AOS) is a rare, congenital syndrome with variable expression characterized by aplasia cutis congenita and transverse defects of the limbs. Sometimes it is associated with cardiovascular malformations and cleft lip/palate. The deformities in this syndrome are possibly a result of vascular disruption. Despite the numerous descriptions of this syndrome in the literature, little is mentioned regarding its anesthetic management and airway concerns. The following case illustrates the anesthetic management of a child with AOS with a perceived, potentially-challenging airway.

Case:

A five year-old girl with AOS presented for repeat surgical release of left clubfoot deformity and right tenotomy and osteotomy for calcaneovalgus foot deformity. Her history included full-term birth, amniotic band syndrome in all four extremities, syndactyly, global developmental delay, craniosynostosis, encephalocele, hydrocephalus, tethered spinal cord, and uncontrolled seizures with prior status epilepticus. Her surgical history included multiple cranial surgeries, VP shunt and revisions, and several foot surgeries. Her only medications were oxcarbazepine and polyethylene glycol. She weighed 25kg.

On the day of surgery, the patient was noted to have self-destructive behavior and a retrognathic jaw. Further airway exam was unobtainable. Given her behavior and seizure history, premedication with 10mg oral midazolam was given. Despite no history of difficult intubations in the past, the decision was made to use a video laryngoscope for intubation given the child's cranial deformities, retrognathic jaw, and obvious facial dysmorphism. Inhalation induction was performed using 8% sevoflurane and 70% nitrous oxide. Mask ventilation was easy with the use of a 7cm oral airway. A 22g intravenous (IV) catheter was placed peripherally, and 100mg propofol was given to facilitate tracheal intubation with 30 mcg fentanyl to blunt the sympathetic response to laryngoscopy. No neuromuscular blocking agent was used until the airway was secured. Intubation with a 5.0 cuffed endotracheal tube using the #2 video laryngoscope blade provided a Cormack-Lehane grade II view of the vocal cords. Maintenance of anesthesia was performed with sevoflurane and intermittent 10 mcg boluses of IV fentanyl. Rocuronium 10 mg was used for surgical muscle relaxation. Maintenance and emergence were uneventful, and the patient was successfully extubated at the end of the surgery in the operating room.

Discussion:

Aside from surgical correction of cranial or cardiovascular defects, which inherently affect anesthetic management due to physical, physiological, or hemodynamic concerns, little is published regarding the anesthetic management of children with AOS who may present for other surgeries. Specifically, those with associated jaw or airway deformities may potentially pose a challenge for intubation. We recommend a careful review of the patient's anesthetic history, a comprehensive airway evaluation, and having appropriate rescue equipment available to secure the airway.

Reference:

Baum VC, O'Flaherty JE. (2007) Anesthesia for Genetic, Metabolic, and Dysmorphic Syndromes of Childhood 2nd edition. Philadelphia, PA. Lippincott Williams & Wilkins. p. 15.
