CASE REPORT

Anesthesia Management in a Case of Apert Syndrome

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Apert syndrome is a premature fusion of certain skull bones. This early fusion prevents the skull growing normally and affects the shape of the head and face. In addition a varied number of fingers and toes are adhered together. Considering the probability of difficult airway is very important in these patients. Our patient was referred to operating room for syndactyly separation in his lower extremities fingers.
Keywords: apert syndrome; anesthesia

Apert syndrome is a genetic disorder characterized by the premature fusion of certain skull bones. This early fusion prevents the skull growing normally and affects the shape of the head and face. In addition a varied number of fingers and toes are adhered together [1]. Our patient was referred to operating room for syndactyly separation in his lower extremities fingers.

Case Description

A 3 yrs old 12kg child with Apert syndrome was admitted to our hospital for syndactyly separation under general anesthesia. Consent was obtained from his father before surgery. We found many abnormalities such as facial hypoplasia, growth retardation, exophthalmia, multiple syndactilies, hydrocephaly according to brain CT scan. Any other systems anomalies in heart, lung, urogenital system, gastrointestinal system was excluded by medical recording and para clinic evaluations. Standard blood tests were in normal range. Our case was scheduled for elective surgery under general anesthesia. We prescribed for him 0.25 mg/kg midazolam orally 20 minutes before surgery. Our monitoring devices were pulse oximetry, 3 lead electrocardiography, non-invasive blood pressure monitoring, capnography, tympanic membrane temperature monitoring. We started inhalation induction after injection of 1mic/kg fentanyl and 10 cc/kg infusion of lactate ringer solution. We maintained his spontaneous breathing because of probability of difficult airway. We prepared difficult airway equipment such as laryngeal mask airway, different size of laryngoscope blade, tracheostomy set. In a deep level of anesthesia, we intubated patient with 4.5 endotracheal tube without any problem. After intubation we used cisatracurium as a muscle relaxant with a dose of 200mic/kg and maintained anesthesia with isoflurane 1.5 MAC during surgery. Before surgery, we performed caudal block with 12 cc bupivacaine 0.25 % for pain management during and after surgery. We checked heart rate and blood pressure continuously as criteria for analgesia and if we found 15 % increase in these criteria we used fentanyl 1 mic/kg but we didn't use. The surgery lasted about 3 hours uneventful. At the end of surgery we extubated patient without any problems and he was painless. Post operative follow up during next 24 hours did not show the need for opioid and his pain scores according to visual analog scale were 3-4. He was discharged next day without any problems.

Discussion

Patient with Apert syndrome has great challenge to anesthesiologist. The main concern is related to airway management [2]. For this reason, we used inhalation technique with maintaining spontaneous breathing. The possibility of difficult mask ventilation because of facial deformities should be predicted. Another organs involvement should be evaluated [3]. Regional anesthesia if possible provide excellent painless condition for patients and reduce intra and post operative opioid requirement and thus reduce the incidence of opioid side effects.

Conclusion

The main concern for anesthesiologist in Apert syndrome is problem related to difficult airway and appropriate preparation must be thought before induction of anesthesia.

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