Case Report

Anesthetic challenges faced in managing multiple surgeries in a child with arthrogryposis multiplex congenita

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Abstract

Background: Arthrogryposis Multiplex Congenita is a syndrome which bring the affected children to the operation room again and again for multiple corrective surgeries. Here we present one such case report of a child who underwent multiple surgeries in our centre and the concomitant anesthetic challenges faced by us.
Key Words: Arthrogryposis Multiplex Congenita, difficult airway, anesthesia, contractures.

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INTRODUCTION

Arthrogryposis is a clinical manifestation of more than 400 different types of disorders, with an incidence of 1:3000 to 1:5100 births, presenting with multiple contractures due to congenital or in utero pathology. The presence of contractures along with associated anomalies like micrognathia, pulmonary hypoplasia, end plate abnormalities, neuromuscular abnormalities and immature gastrointestinal system provide an anesthetic challenge in these patients.

CASE REPORT

A 2 year girl from Oman, known case of Arthrogryposis Multiplex Congenita, was brought to our centre for correction of bilateral hip varus correction and bilateral release of hand skin contracture release. She was born after a lower segment caesarean section due to breech presentation with a birth weight of 2.7 kg. She was noted to have multiple joint deformities. She developed seizures and desaturated two hours after feeds following which she was taken to Intensive Care Unit (ICU), intubated and kept on elective ventilation for 10 days. On further workup, an echocardiogram was done which showed a Ventricular Septal Defect(VSD). She had history of recurrent respiratory tract infections, GERD, sweating on the forehead during feeds, poor oral intake and failure to thrive. Her motor milestones were delayed in view of the multiple large joint deformities, but her speech, hearing and social milestones were not delayed. At 6 months of age, she came for elective bilateral knee contracture release. On Pre anesthetic work up, Echocardiography showed spontaneously closed VSD with normal pulmonary artery pressures and normal systolic and diastolic chamber functions. Chest radiograph showed eventration of diaphragm and further imaging was suggested. For further imaging studies, general anesthesia was required. Previous anesthesia records showed anticipated difficult airway, as the patient had almost nil neck extension, she was shifted to the Operation Room where she was intubated using Glidescope video laryngoscope, and shifted to MRI suite for imaging where anesthesia was maintained with inhalational agent

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sevoflurane. Temperature monitoring was not done. MRI Brain study was normal, Neurosonogram also showed normal study, MRI Spine was done which showed tethered spinal cord, CT Thorax showed eventration of diaphragm. She was advised immediate corrective surgery for the eventration of diaphragm and was hence not extubated and sent back to ICU for elective ventilation, the following day she underwent thoracoscopic repair of eventration of diaphragm and kept in Pediatric Intensive Care Unit for 5 days following which she was extubated and sent to the ward. A muscle biopsy was also taken and the results were normal. In the present admission, she was active, moving all four limbs, able to walk and weighed 10 kilograms. Heart rate of about 110 beats per minute, respiratory rate of 20/minute at rest, afebrile , blood pressure of 95/56 mm hg in the left upper limb and. room air saturation of 98%. . Her airway examination showed Mallampati grade 3 with restricted neck movements Grade II (22-34 degree). There was no micrognathia, microglossia or high arched palate. She had normal intelligence for her age. On investigating, Hemoglobin was 12.4 gm%, chest radiograph showed right eventration of diaphragm. MRI of both hands was planned under general anesthesia. In view of history of previous difficult airway, the airway of the child was secured in the operating room premises. After connecting standard monitors, baseline vitals were noted. Due to unavailability of MRI compatible temperature probe, temperature monitoring was not done. Inhalational induction was done with 8% sevoflurane in 100% oxygen using bag and . After attaining adequate depth of anesthesia, intravenous line was secured in the lower limb and 10 mcgs of Fentanyl and 20 milligram Propofol given, mask ventilation checked, then atracurium 5 mg given. Macintosh blade #2 was used and the Cormack Lehane view was IIB. Trachea was intubated with a PVC ETT ID 4.5 uncuffed under vision and fixed at 13cm. She was then shifted to the MRI suite with ventilator backup where anesthesia was maintained with air-oxygen(FiO2 between 0.3-0.4) and sevoflurane between 1-2%. Pre and post procedure temperatures were increased (37.8 degrees celcius). She was moved back to OT complex, residual neuromuscular blockade reversed at the end of the procedure and trachea extubated. 8 days later, she underwent Right hip varus osteotomy and the following week left hip varus osteotomy was done under general anesthesia. Similar induction technique was followed as for the previous procedure. Trachea was intubated with #4.0 PVC ETT under direct vision after obtaining a laryngoscopic view of CL 2B using Macintosh #2 blade. Intraoperative hyperthermia (38 degrees celsius) was observed for one hour after induction which resolved spontaneously in both the procedures. The Procedure was uneventful. Neuromuscular blockade was reversed at the end of the procedure and the patient's trachea extubated once she was awake. Post operatively she was given syrup ibugesic for analgesia. The subsequent week she underwent Right hand contracture release with split skin grafting. Anesthesia was induced as in the previous two surgeries, fentanyl was given as the analgesic at 2 mcg/kg and maintained with sevoflurane in air and oxygen mixture. Several attempts had to be made to secure intravenous access. Trachea was intubated using Macintosh size 2 blade with a laryngoscopic view of CL 2b , #4.5 PVC ETT was inserted. Due to the presence of hip spica, slight difficulty in positioning was faced. Intraoperative temperature monitoring showed a drop in temperature from 36.4 to 35.1 which was brought back to 36.2 at the end of procedure with use of forced air warmer. Procedure and post operative recovery periods were otherwise uneventful. Another week later, she underwent surgery for left hand contracture release during which hyperthermia occurred (36.5 to 37.9) and tepid sponging was done intraoperatively to achieve normothermia.

DISCUSSION

The varying clinical presentation of arthrogryposis provides a challenge for perioperative anesthetic management. The clinical manifestation depends on the causative factor which can be neurological or non neurological. The presence of tethered cord in this patient could suggest a neurological cause for the contractures. But the involvement of upper limb and neck muscles with normal cognition suggest a non neurological cause. Muscle biopsy rules out a muscular cause for the contractures. Airway difficulties in these patients include, decreased mouth opening (Freeman Sheldon Syndrome), mandibular hypoplasia, cervical rigidity, micrognathia, high arched palate, limited tongue protrusion, Pierre-Robin-Sequence like, short neck, torticollis and hemangiomas of the neck.[1] Multiple procedures in this child helped in the understanding of the pathology. With the growth of the child there was a mild improvement in the neck movement. The correction of eventration in a growing lung improved her pulmonary reserve. In cases where central nervous system cause can be ruled out and a normal sleep history is obtained, premedication can be safely given. Preoperative identification of venous access, use of topical anesthesia can be useful in these patients. The presence of tethered cord precluded any neuraxial anesthesia in this patient. Neuromuscular monitoring could not be performed due to the presence of contractures, which also cause difficulty in positioning.[2,3] Intraoperative hyperthermia has been observed in these patients and in some cases have been treated with dantrolene. Intraoperative hyperthermia was observed in three out of four procedures and was not observed during hand contracture release though similar anesthetic management was done. Hypermetabolism has

been suggested as a cause for hyperthermia. The understanding of mechanism of hyperthermia will help in the intraoperative management.[4] In contrast to previous reports, no increase in sensitivity to anesthetic agents or opioids were observed. Peripheral nerve blocks have been used in these patients for both muscle biopsy and postoperative analgesia.[5] Several orthopedic surgeries are usually required for the correction of multiple contractures and deformities in these children.[6]

CONCLUSION

AMC is a condition all anesthesiologists should be aware of as they come with a myriad of problems including difficult airway, hypermetabolism, hyperthermia, positioning and contracture difficulties, myopathies etc. Different types of anesthesia can be given keeping this in mind and patient can be managed successfully.

PERMISSION: The parents consented to us sharing the above information, but did not permit us to share the photographs or radiological investigations.

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