Anesthesia in a child with arthrogryposis multiplex congenita – how to proceed: A case report

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ABSTRACT

Arthrogryposis multiplex congenita (AMC) consists of a heterogeneous group of disorders characterized by non-progressive congenital joint contractures. They have tense skin, minimal subcutaneous tissue, and muscle mass. The anesthetic management of these children is complicated by associated congenital abnormalities, airway anomalies, congenital heart disease, pulmonary hypoplasia, and vertebral anomalies. We managed a case of AMC with bilateral contractures of both upper and lower limb and neck who presented for correction of bilateral equino varus.

Keywords: Congenital joint contractures, Pediatric anesthesia, Caudal epidural analgesia, Difficult airway

A rthrogryposis multiplex congenita (AMC), a rare congenital disease, is characterized by multiple joint contractures throughout the body, often requiring multiple corrective surgeries [1]. The incidence is 1 in 3000 live births [2]. Although primarily a disease of the musculoskeletal system, it may be associated with neurological or other anomalies such as congenital heart disease, gastrochisis, cleft palate, and genitourinary disease [3-5]. The affected children are born with polyarticular contractures, especially spine and distal extremities, leading to abnormal bone growth, and restriction of mobility [6]. Children with AMC require multiple corrective surgeries to improve the mobility of joints and, thus, their lifestyle.

We present one such case of a child with AMC with bilateral contractures of both upper and lower limbs and neck, which were managed successfully in our hospital. Written informed consent was taken from parents for the publication of the manuscript. This manuscript adheres to the applicable EQUATOR guidelines.

CASE REPORT

A 6-month-old male child weighing 5 kg, a known case of AMC presented to our hospital for bilateral Congenital Talipes Equino Varus correction. The child was born at term by normal vaginal delivery, cried immediately after birth with an appearance, pulse, grimace, activity, and respiration score of 9 out of 10, and a birth weight of 2300 g.

On examination, the patient had multiple contractures all over the body, including bilateral elbow, axillae, hip, and knee, so much so that the hands and legs could not be extended at these joints. The right-sided chest had some bony protrusion of ribs. The neck contracture limited the extension of the neck (Fig. 1).

All investigations, including 12 lead electrocardiograph (ECG) and chest X-ray, were normal. The patient was taken with due risk explained and consent and assent were taken from the parents for the surgery.

In the operation theater, all monitors are attached, including ECG, pulse oximetry (SpO₂), non-invasive blood pressure, end-tidal CO₂, and temperature. The patient was induced with sevoflurane 8% and 100% O₂. A laryngeal mask airway (LMA) was inserted to buy time for securing intravenous access due to anticipated difficult cannulation due to multiple contractures. Only a scalp vein could be secured with great difficulty after 1 h. Considering the anticipated difficult airway, the difficult airway cart was kept ready aside. LMA was removed and a check laryngoscopy was done, Cormack and Lehane 2b was found. Injection of thiopentone 20 mg and fentanyl 15 mcg was given intravenously. The trachea was intubated with uncuffed endotracheal tube size 4.5 with slight external manipulation of the neck. The neck contracture limited the extension of the neck.

Intravenous access was given 5 ml of 0.25% bupivacaine. Anesthesia was maintained with O₂ 50% and N₂O 50% with 2%
At the end of the surgery, once the spontaneous respiratory efforts were present, neuromuscular blockade was reversed with neostigmine and glycopyrrolate and the trachea was extubated. The total surgical duration was 2½ h and the intraoperative period was uneventful. In the post-operative room, after about 1 h, the patient started desaturating at room air but was maintaining saturation $\text{SpO}_2$ 97–98% with a normal face mask at 4 L/min. On examination, the patient was awake, moving all four limbs with no signs of respiratory distress except a respiratory rate of 38/min. On auscultation, bilateral air entry was clear. The patient was, therefore, shifted to the pediatric intensive care unit (ICU) for further care. On day two in ICU, the patient remained awake, maintaining heart rate and $\text{SpO}_2$ on room air. The patient was shifted to the ward and was subsequently discharged on day 4 post-surgery.

DISCUSSION

AMC is a rare heterogeneous group of disorders characterized by congenital, non-progressive, multiple, and persistent joint contractures with generalized muscle wasting. Apart from extensive contractures, tense skin, and minimal muscle mass and subcutaneous tissue, the anesthetic management of children with AMC is complicated by associated congenital anomalies including congenital heart disease, pulmonary hypertension, cor pulmonale, upper airway abnormalities and positioning difficulty, deformities of skeletal, and genitourinary systems [7,8]. Pulmonary hypoplasia and scoliotic restrictive lung changes may contribute to hypoxemia risk and may necessitate post-operative ventilator support [7,9].

The anesthesiologist should be ready for the possibility of a difficult airway. Contractures at multiple sites, including the neck, markedly limit neck mobility during airway maintenance, decreased mandibular opening, micrognathia, a high arched palate, and deficient musculature of the orofacial complex. For these reasons, there may be difficulty in mask ventilation, placing a supraglottic device, laryngoscopy and intubation, and difficulty in surgical access to the airway. Direct laryngoscopy or tracheal intubation may be difficult or impossible [10]. Patients with AMC are also vulnerable to perioperative aspiration due to loss of normal muscle movement and gastroesophageal reflux.

Malignant hyperthermia (MH) is yet another concern in the intraoperative and perioperative period, though the association of AMC with MH is controversial [11]. We used inhalational induction in our case as securing an intravenous line was anticipated to be difficult. However, several myopathies themselves have strong relations with MH. About 8% of the etiology of AMC is related to neuromuscular disorders, myopathic processes such as congenital muscular dystrophies, mitochondrial disturbances, or abnormalities of the biochemistry of the muscles [12]. Therefore, anesthesiologists should know the risk of MH in patients with AMC and should closely monitor these patients.

In addition, multiple contractures of the extremities make it difficult to maintain intraoperative positioning. Careful attention is required and appropriate measures, such as padding for reducing compression pressure. Moreover, peripheral intravenous access may be difficult in these patients due to tense, glossy skin, and scanty subcutaneous tissues.

In our patient, we anticipated difficulty in intravenous cannulation, airway management, and positioning. The child was taken for the surgery with due care for these factors. Furthermore, the patient was continuously monitored for hypermetabolic syndrome throughout the surgery with $\text{EtCO}_2$ and temperature monitoring.

CONCLUSION

To summarize, perioperative care in patients with AMC may be challenging for anesthesiologists. The most important concerns are the possibilities of difficult airway management and securing intravenous access, difficult positioning, and difficult regional blockade. Pre-operative evaluation to rule out congenital anomalies, anticipating difficulties, and preparation for emergencies such as perioperative monitoring of body temperature, tachycardia, and hypercarbia are needed to avoid critical events.

REFERENCES


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