

# Anesthetic management of a neonate with arthrogryposis multiplex congenita for emergency laparotomy

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## Abstract

Arthrogryposis multiplex congenita is a rare disease, characterized by non-progressive, multiple joint contractures since birth. Anesthetic issues include difficult intravenous access, difficult airway management and regional anesthesia. We report the anesthetic management of a six-day-old neonate presenting to the emergency with features of intestinal obstruction, who was detected for the first time to have arthrogryposis multiplex congenita. General anesthesia along with caudal analgesia for preoperative and postoperative pain relief was used. There was an episode of intraoperative hyperthermia, which was tackled successfully. The child had an uneventful post-anesthesia recovery.

**Key words:** Arthrogryposis multiplex congenita, caudal analgesia, perioperative hyperthermia

## Introduction

Arthrogryposis multiplex congenita (AMC) is a syndrome characterized by multiple joint contractures and can include muscle weakness and fibrosis. Incidence of arthrogryposis is one in 3000 live births.<sup>[1]</sup> Anomalies with diverse etiologies are associated which include congenital heart defects, cleft palate, gastroschisis, genitourinary defects, spinal deformities and concomitant restrictive pulmonary disease.<sup>[1-2]</sup>

Patients of AMC require anesthesia mostly to correct orthopedic deformities and for muscle biopsy for disease confirmation, but rarely present for exploratory laparotomy at the neonatal age. AMC is associated with difficult intravenous access, difficult airway, difficult positioning and a variable response to different anesthetic agents.

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## Case Report

A six-day-old, 2.6-kg full-term male neonate with features of intestinal obstruction was posted for emergency exploratory laparotomy. The neonate was in a hyper-extended posture with multiple contractures [Figure 1] involving different joints and bilateral clubfoot [Figure 2]. He had normal cry and reflexes, micrognathia and high-arched palate, but no vertebral anomalies were detected. His veins were difficult to visualize. He was diagnosed as a case of AMC [Figures 1 and 2].

Hemogram, liver function, ion status were normal but mild metabolic alkalosis with mild hypoxemia was noted on arterial blood gas. Intravenous access was secured in a scalp vein with 24G intravenous cannula with difficulty. 1/5 Normal saline in 10% Dextrose with 1 ml KCl/100 ml was started as intravenous fluid. A 5-Fr Foley's catheter was inserted and the neonate was kept warm with the help of a warmer. The child was premedicated with 50 µg atropine and fentanyl 2.5 µg IV. After preoxygenation with 100% oxygen, anesthesia was induced with 8% sevoflurane in oxygen. Tracheal intubation was conducted after neuromuscular blockade with 0.3 mg/kg atracurium intravenous. Laryngoscopy was slightly difficult (Cormack-Lehane Grade II) but endotracheal tube was inserted in one attempt. Anesthesia was maintained with atracurium and sevoflurane in oxygen. Gentle ventilation using small tidal volume and minimum pressure along with permissive hypercapnia was used. Caudal epidural catheter was placed, with the baby held in left lateral position, by insertion of 18G intravenous cannula at sacral hiatus through which a 20G

Portex epidural catheter was threaded; 2 ml of 0.125% isobaric bupivacaine was injected and analgesia was maintained with 0.125% bupivacaine @ 1-1.5 ml/h., EtCO<sub>2</sub>, SpO<sub>2</sub>, urine output, neuromuscular monitor, non-invasive blood pressure, and nasopharyngeal/axillary temperature were monitored. Glucose levels were estimated every 30 min and were found normal.

Thirty minutes after induction, as the core temperature was 38.5°C and peripheral 38°C, active external cooling was started. Hyperthermia was not associated with rigidity and temperature returned to normal by the time of completion of the Santulli's operation, which took one hour. Jejunio-ileal atresia was detected and proximal ileostomy was done. Neuromuscular blockade was reversed with 0.13 mg neostigmine and 0.01 mg glycopyrrolate intravenous. As the baby had normal protective reflexes with active pelvic movements after reversal and was calm and quiet, trachea was extubated and the baby was shifted to the neonatal intensive care unit (NICU). Postoperatively 50 ml whole blood was administered; caudal analgesia was maintained with 0.125% bupivacaine @ 1ml/h; and paracetamol 40 mg suppositories were administered thrice daily.

On the second postoperative day an ultrasonography (USG)-guided subclavian venous central line was placed for total parenteral nutrition. Muscle biopsy report from gastrocnemius muscle revealed small-for-age muscle fibers, with fibrous and fatty degeneration of the muscle. Oral liquid feeds were started almost 14 days later and the child was discharged home with an advice to report for ileostomy revision a month later.

## Discussion

AMC is a non-progressive congenital syndrome characterized by contracture of several joints in different parts of the body due to fibrosis of the affected muscles, thickening and shortening of periarticular capsular tissue with an intact sensory and normal intellect.<sup>[3]</sup> The etiology of AMC is multifactorial: neurogenic, myogenic, structural central nervous system and genetic disorders.<sup>[4]</sup> Electromyogram (EMG) and muscle biopsy help distinguish these forms.

The anesthetic management is complicated by associated congenital abnormalities of the upper airway, congenital heart defects, pulmonary hypertension, cor-pulmonale and urogenital anomalies.<sup>[2]</sup> Primary concerns for airway management are related to mandibular hypoplasia, cleft palate, temporomandibular and cervical rigidity,<sup>[5]</sup> decreased mouth opening, micrognathia, high arched palate, deficient musculature of the oropharyngeal complex, limited tongue



**Figure 1:** Picture of neonate showing multiple joint contractures



**Figure 2:** Image showing bilateral clubfoot

protrusion, Pierre-Robin-like sequence, short neck, torticollis and hemangiomas of the neck,<sup>[2,3,6,7]</sup> which make direct laryngoscopy and endotracheal intubation difficult.<sup>[1,2,6]</sup> The extensive contractures, tense skin, minimal muscle mass and subcutaneous tissue may make intraoperative positioning and intravenous access difficult. Children with AMC have altered responses to neuromuscular relaxants and are more susceptible to the depressant effects of various intravenous and inhalation anesthetics.<sup>[1,2]</sup> As sensitivity to nondepolarising neuromuscular blocking agents has been reported due to underlying neurologic and myopathic conditions, use of relaxants such as mivacurium<sup>[8]</sup> is preferred but it was not available in our centre. Atracurium was administered in a reduced dose (0.3 mg/kg) with neuromuscular function monitoring. A reduced train of four and marked fade during prolonged titanic stimulation was noted. Close monitoring of respiratory function is advisable due to congenital myohypoplasia and spine deformities and associated restrictive respiratory pattern.<sup>[3,7]</sup> Regional techniques may be technically difficult due to scoliosis

and associated spine anomalies.<sup>[2,7]</sup> We used a multimodal analgesic approach, including caudal analgesia, to use smaller opioid doses pre and postoperatively.

In patients with underlying myopathic disorders, there may be hyperkalemic response with the use of succinylcholine.<sup>[7]</sup> We avoided administration of succinylcholine despite a predicted difficult airway. Halothane and succinylcholine can also trigger malignant hyperthermia (MH). Since the risk of MH is unproven, it is suggested that the use of volatile agents in these patients is justified; particularly if alternative agents may place the child at an even greater risk.<sup>[7]</sup> We used sevoflurane and there was per-operative hyperthermia, which responded to conservative measures. The probable causes of per-operative rise in temperature may be infection, MH, excessive warming etc. In this case, hypermetabolic response to fentanyl<sup>[9]</sup> was probably the offending cause as other causes were ruled out. During general anesthesia, the patient should be observed and specifically monitored for signs of hypermetabolic response. Should this occur, active cooling should be started immediately.<sup>[10]</sup>

We plan to conduct the ileostomy revision surgery under balanced anesthesia using sevoflurane, atracurium supplemented with caudal analgesia.

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