# Anaesthetic Management Of A Child With Arthrogyroposis Multiplex Congenita

P Jedge, S Aphale, P Bhosale, S Bapat

**Citation**

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**Abstract**

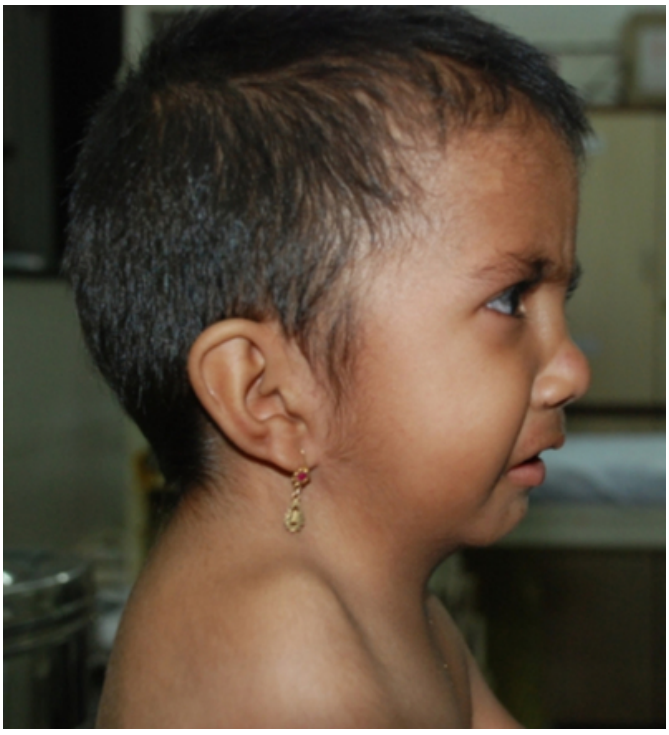
Arthrogryposis, also known as Arthrogryposis Multiplex Congenita, is a rare congenital disorder that causes multiple joint contractures and can be associated with muscle weakness and fibrosis. It is a non-progressive disease. The disease derives its name from Greek; literally meaning 'curved or hooked joints'Anaesthetic issues include difficult intravenous access, difficult airway and association with anterior horn cell disease Although an association with malignant hyperthermia has been suggested, this has not been confirmed. We report the management of a two year-old girl with Arthrogryposis Multiplex Congenita who underwent lower limb surgery with a successful anaesthetic regimen based on an anaesthesia technique with sevoflurane, fentanyl and propofol without neuromuscular blocking agents. The child had an uneventful anaesthetic and postoperative course.

## CASE REPORT

A two year old girl, a known case of Arthrogryposis Multiplex Congenita diagnosed at birth, presented to paediatric department with non healing wound at left lower limb. She was operated for contracture release and debridement for two times under sedation and spinal anaesthesia in other institute. Venous access was achieved via one of the scalp veins. Other intraoperative details of previous surgeries were unavailable. She was posted by pediatric orthopaedic surgeon for debridement and skin grafting.

Thorough general examination revealed multiple contractures almost at every joint. These were present at neck, shoulder, axilla, elbow, wrist and interphalangeal joint in upper extremities while at hip, knee and ankle in lower extremities. Airway examination revealed micrognathia, with receding mandible and protruding upper teeth. Head size was larger with short neck. Neck extension was not possible due to contracture. Mouth opening was adequate and Mallampati classification was class III.

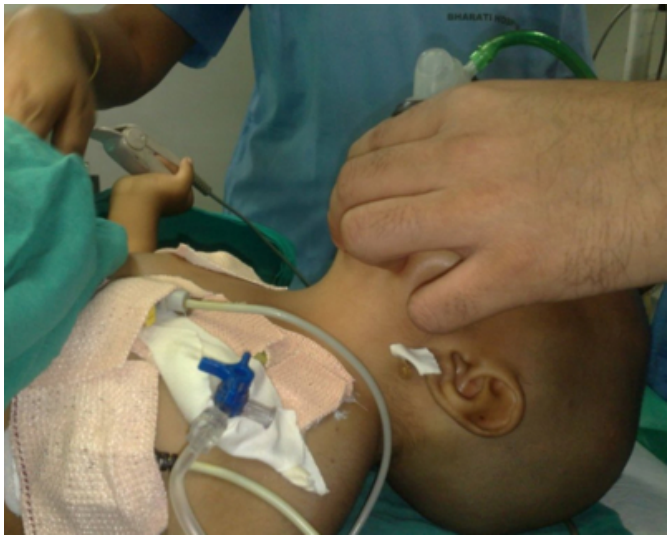
**Figure 1**



She did not have other associated congenital anomalies. Heart sounds were normal with no murmur. Interspinaus spaces were palpable at regular interval with good caudal space. She had no neurological deficit. Her veins were difficult to visualize. External jugular vein had tortuous course due to lateral neck contracture. After necessary

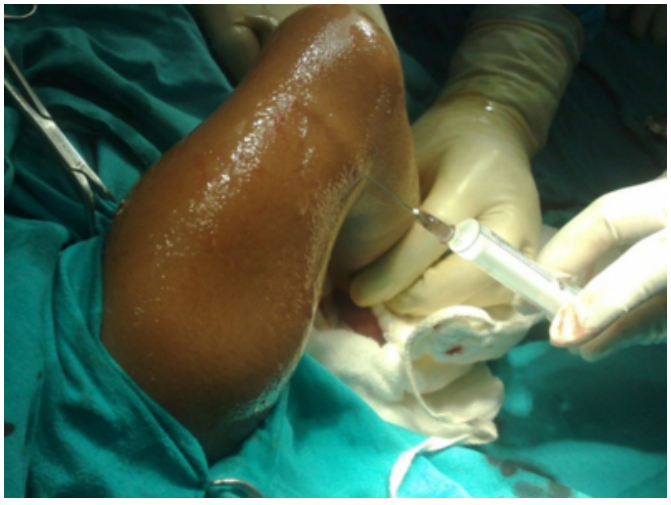
investigation she was posted for debridement and skin grafting as ASA Gr. II with difficult airway consent. Premedication was given with inj. Glycopyrrolate 0.04 mg/kg intra muscular. Venous access was secured with difficulty over anterior chest wall. Inj fentanyl 1.5gm/kg was given. The patient was induced with sevoflurane and inj. Propofol 1mg/kg iv and maintained on spontaneous respiration with titrated doses of inj. Propofol and sevoflurane.

**Figure 2**



Field block was performed with inj. Lignocaine 4mg/kg in 0.5 % concentration at the site of skin graft.

**Figure 3**



Surgery underwent uneventfully without any complications. Postoperative course was uneventful. Postoperative pain was managed with NSAIDS. The patient was discharged on 7th post operative day with good wound healing.

## DISCUSSION

Arthrogryposis Multiplex Congenita (AMC) is a non-progressive congenital syndrome complex characterized by contracture of several joints in different parts of the body due to varying degrees of fibrosis of the affected muscles, thickening and shortening of periarticular capsular and ligamentous tissue with an intact sensory system and normal intellect ([1]).

Etiology of AMC is multi-factorial. It has been accepted that within the context of heterogeneity, there are two main types: the neuropathic and the myopathic. Studies by Banker,([2]) prove that the neuropathic form is seen in almost 90% of affected children compared to the myopathic type (< 10%).

The most common orthopaedic deformities include talipes equinovarus, dislocated hip, dislocated patella, and scoliosis. Surgical management is directed at correcting all lower extremity deformities that delay ambulation. These are ideally performed before the patient is 2 years old.

The anaesthetic management is complicated by associated congenital abnormalities and an abnormal upper airway. Arthrogryposis is associated with congenital heart disease, pulmonary hypertension-cor pulmonale, and urogenital anomalies ([5]).

Patients with arthrogryposis have micrognathia, a high arched palate, and a short and rigid neck making tracheal intubation difficult and at times impossible ([3]). The primary concern to the anesthesiologist is the potential for airway involvement thereby making direct laryngoscopy and endotracheal intubation difficult ([5]).

Direct laryngoscopy and intubation become more difficult as the patient ages because craniofacial involvement often progresses with growth. Alternatives to direct laryngoscopy and tracheal intubation, such as the use of the laryngeal mask airway, with or without the use of a tube exchanger, or fiberoptics, have been used successfully in this disorder. In such difficult airway scenario we decided to avoid intubation and maintained her on spontaneous ventilation. We kept difficult intubation cart ready in case required to secure airway in emergency.

The extensive contractures, tense skin, and minimal muscle mass and subcutaneous tissue pose challenges for intraoperative positioning and intravenous access. Children with arthrogryposis may have altered responses to

neuromuscular relaxants and are akin to other patients with anterior horn cell diseases(3).

Response to nondepolarizing relaxants has been reported to be extremely variable, the use of short-acting nondepolarizing agents in association with careful monitoring of neuromuscular function has been advocated in these patients.

Baines et al. reviewed 396 anaesthetic agents in 67 patients with arthrogryposis multiplex congenita and found no episodes of MH with exposure to known triggering agents (4). In our patient we avoided the triggering anesthetic agents as Halothane, Succinyl choline.

In the past, self-limited anterior horn cell disease was proposed as one of the etiology of AMC (3).

Spinal muscular atrophy (SMA) remains the most common cause of neurogenic arthrogryposis. Congenital Hypomyelinating Neuropathy is a rare potentially reversible cause of infantile neuromuscular weakness and its occurrence in association with AMC has been reported making it an important differential diagnosis (6).

Neurogenic arthrogryposis is used to denote the association of arthrogryposis with several conditions including infantile SMA and primary degeneration of anterior horn cells, anterior roots or peripheral nerve(7).

Therefore the regional anaesthesia in the form of central neuroaxial blockade was not tried.

In summary, this report highlights the anaesthetic management of arthrogryposis multiplex congenita, which may be complicated by a difficult airway. Appropriate preoperative evaluation to rule out congenital abnormalities, anticipating difficulties and keeping alternate arrangements and preparation is required.

## References

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