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ULTRASOUND GUIDED ILIOINGUINAL AND ILIOHYPOGASTRIC NERVE BLOCK FOR INGUINAL HERNIA REPAIR IN ARTHROGRYPOSIS MULTIPLEX CONGENITA

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ABSTRACT: Arthrogyrosis multiplex congenita (AMC) refers to a syndrome of unknown etiology with multiple congenital contractures in one or more joints with a concomitant inability of passive extension and flexion. The overall prevalence of arthrogyrosis is one in 3000 live births. The extensive contractures, tense skin, minimal muscle mass and subcutaneous tissue pose challenges in anaesthetic management. We report a seven year old boy (15 kg), known case of AMC with congenital talipes equino varus (CTEV) and bilateral hip dislocation posted for right sided herniotomy and orchidopexy. We planned to combine general anaesthesia without muscle relaxants and regional nerve block. The child was induced with propofol and Classic LMA Size 2 was inserted. An ilioinguinal and iliohypogastric nerve block was given under ultrasound guidance using 0.2% ropivacaine. Patient remained hemodynamically stable during surgery with minimal anaesthetic requirement and no analgesics. Analgesia lasted for 8 hours postoperatively. Combining narcosis with regional anaesthesia leads to a reduced demand for anaesthetics, stable circulatory conditions, maintenance of spontaneous breathing, prevention of stress and sufficient postoperative analgesia.

KEYWORDS: Arthrogyrosis Multiplex Congenita, Ultrasound guided nerve block, ilioinguinal and iliohypogastric nerve block.

INTRODUCTION: Arthrogyrosis Multiplex Congenita (AMC) is a congenital syndrome characterized by contracture of multiple joints of the body due to fibrosis of the affected muscles with an intact sensory and normal intellect.⁽¹⁾ Fetal akinesia appears to be the cause of congenital joint contractures.⁽²⁾ Incidence is 1 in 3000 live births.⁽³⁾ The extensive contractures, tense skin, minimal muscle mass and subcutaneous tissue pose challenges in anaesthesia management.

We report a case where ultrasound aided ilioinguinal and iliohypogastric [ILIH] nerve block proved beneficial in AMC patient.

A seven year old boy (height 100 cm weight 15kg) was scheduled for herniotomy and orchidopexy. He was a known case of AMC with multiple contractures. On examination he had a small mouth, malampatti class 3 airway, pectum excavatum, kyphoscoliosis and bowing of upper and lower limbs. After premedication with Midazolam 1mg, child was induced with Propofol 70mg; Classic LMA size 2 was inserted and placed in position. Sevoflurane 2% along with O₂ – N₂O mixture was given as maintenance.

After skin and transducer preparation a linear hockey stick probe (Venue 40 GE) was placed obliquely along a line joining the anterior superior iliac spine and the umbilicus, immediately superior and medial to the ASIS. Three muscular layers of the abdominal wall [the

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external oblique, the internal oblique and transverse abdominalis muscles] were identified. The Ilioinguinal and Iliohypogastric nerves could be seen within the fascial plane between the transverse abdominalis and internal oblique muscles above the ASIS. Colour doppler was performed to check vascular identity at this neurovascular plane.

A 23 G 50mm stimuplex needle (B Braun) was advanced in in plane approach under direct ultrasonographic vision and positioned in the proximity of the nerves. 3ml of 0.5% Ropivacaine was injected after aspiration and fluid expansion in the sheath of the internal oblique and transverse abdominalis muscle layers noted. A skin wheel was raised over the surgical incision site with 2ml of 0.2% ropivacaine.

Incision was put by the surgeon after 20 minutes of instituting the block and the surgery lasted for 45 minutes without any additional analgesics requirement.

In the post-operative period the sensory block lasted for 8hours as per the VAS scores. Tramadol 30 mg and Paracetamol suppository 250mg was given once the child complained of pain. [VAS score >4]. The child was discharged after 24 hours.

DISCUSSION: Arthrogryposis multiplex congenita is a rare congenital syndrome characterized by multiple joint contractures found throughout the body and often accompanied by associated anomalies.

Anaesthetic management may be complicated by difficult intravenous access, difficult positioning, difficult regional blockade and difficult airway.

Primary concern to the anesthesiologist is the potential for airway involvement thereby making direct laryngoscopy and endotracheal intubation difficult.⁽⁴⁾ In most cases of difficult intubation, good conditions for ventilation through a laryngeal mask airway have been reported.

Children with arthrogryposis may have altered responses to neuromuscular relaxants.⁽⁵⁾ Response to nondepolarizing relaxants has been reported to be extremely variable, careful monitoring of neuromuscular function has been advocated in these patients. In patients with underlying myopathic disorders, there may be hyperkalemic response with the use of succinylcholine.⁽⁶⁾ We avoided administration of succinylcholine despite a predicted difficult airway. Succinylcholine can also trigger malignant hyperthermia (MH). As a part of standard monitoring, the surveillance of temperature and CO₂ is of particular importance, even in case of small interventions. The reason is that AMC is frequently accompanied by an unspecific intraoperative hypermetabolic reaction (hyperthermia). 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.⁽⁷⁾ Under these circumstances, the patient possibly develops acidosis and hyperkalemia. In patients with AMC, a slightly increased responsiveness towards respiratory depressant drugs is assumed. We avoided opioids in this case as it can also cause hypermetabolic response and can induce rise in temperature preoperatively.⁽⁸⁾

In AMC due to existing spinal abnormalities, regional anaesthesia methods are partly described to be impossible. Severe deformities of the spinal column can increase the difficulty in placing a spinal anesthetic.

Whenever possible, it is very reasonable to combine narcosis with regional anaesthesia for AMC patients. This leads to a reduced demand for anaesthetics, stable circulatory conditions,

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maintenance of spontaneous breathing, prevention of stress and sufficient perioperative analgesia.

Variable neural anatomy and proximity of vascular structures in children make landmark based and nerve stimulated approach for peripheral blocks more challenging. Adding to the difficulty in children is the concern of local anaesthetic toxicity secondary to a narrow therapeutic window.

In AMC due to contractures, reduced muscle mass makes these procedures practically impossible. Moreover classical motor response to nerve stimulation also cannot be obtained.

Of the peripheral nerve blocks, ilioinguinal and iliohypogastric (ILIH) nerve blockade remains one of the most commonly placed blocks in infants and children. ILIH nerve blockade is used commonly to provide perioperative pain relief for children undergoing inguinal procedures. ILIH nerve blockade is frequently applicable even in patients in whom neuraxial analgesia including caudal block may be contraindicated. It has historically proven to be safe and effective with landmark-based administration in the pediatric population.⁽⁹⁾ Although placement of the ILIH nerve block was initially accomplished using anatomical surface landmarks, the introduction of ultrasonography has been shown to improve the accuracy and consistency of the ILIH nerve block and local anaesthetic placement.⁽¹⁰⁾

The risk of intraneural or intravascular injection is reduced. Lower doses of a local anaesthetic can be used as the quantity required is less. We planned for ultrasound guided block as conventional block was difficult in this patient because of muscle wasting.

CONCLUSION: We conclude that combining narcosis with regional anaesthesia [ILIH nerve block] in AMC patients leads to a reduced demand for anaesthetics, stable circulatory conditions, maintenance of spontaneous breathing, prevention of stress and sufficient postoperative analgesia.



Fig. 1: Arthrogryposis Multiplex Congenita showing Pectus excavatum



Fig. 2: Leg deformity in Arthrogryposis Multiplex Congenita

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Fig. 3: Scoliosis in Arthrogyrosis Multiplex Congenita

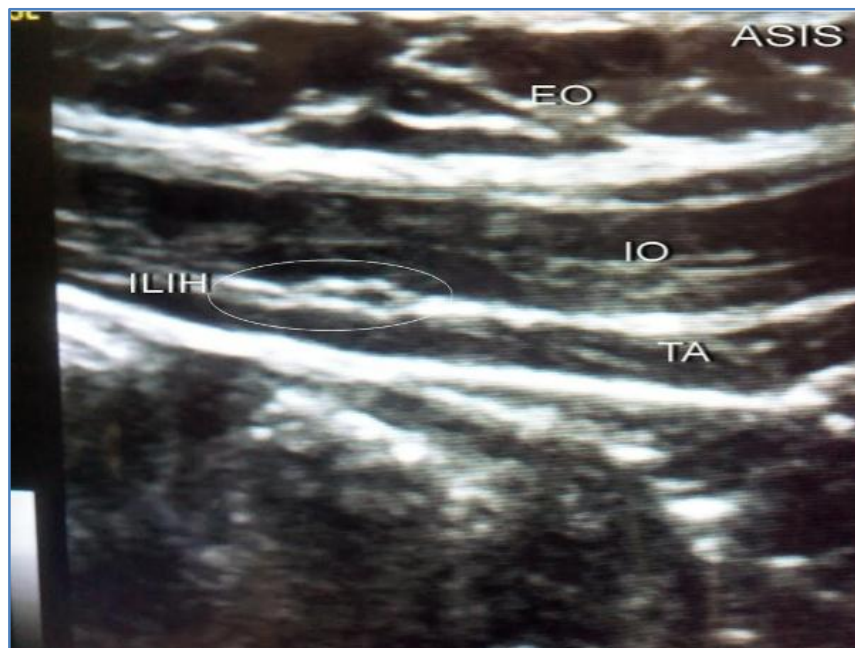


Fig. 4: Ultrasound Image of Ilioinguinal and Iliohypogastric Nerves

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