
LETTER TO EDITOR**Perioperative anaesthesia management in a parturient with arthrogryposis multiplex congenital posted for caesarean delivery***Sangharsh S¹, Sushama T¹, Sanyogita N¹**¹Department of Anaesthesia, B. J. Govt. Medical College and Sassoon General Hospitals, Pune-411030 (Maharashtra) India*

Abstract

Arthrogryposis multiplex congenita is usually associated with multiple congenital abnormalities, including skeletal deformities, congenital heart disease and sometimes alterations of the respiratory and genitourinary systems. Challenges faced by the anesthesiologist when dealing with patients with this syndrome include potential difficult airway, risk of malignant hyperthermia associated with myopathy, cardiorespiratory problems, difficult regional blockade, difficult IV access, and difficult positioning. Hence these patients need meticulous care perioperatively. Though regional anaesthesia is technically challenging in them, it has been used successfully in them.

Keywords: Arthrogryposis Multiplex Congenita, Caesarean delivery, Regional anaesthesia

Introduction

Arthrogryposis Multiplex Congenita (AMC) is a term used to describe a group of musculoskeletal disorders characterized by congenital, non-progressive, and symmetric joint contractures involving both upper and lower limbs [1]. The etiology of AMC is unclear and may include genetic and environmental factors, affecting 1:3000 births [2]. Patients with AMC have higher incidence of associated neuromuscular, cardiac, respiratory, genitourinary diseases and myopathy [3]. They have pelvic deformities hence caesarean delivery is the preferred mode of delivery for them [4].

Case Report

A 31 year old full term primigravida female presented in labour and posted for caesarean delivery in view of cephalopelvic disproportion. Patient was a known case of AMC since birth with multiple joint contracture, thoracic kyphoscoliosis and short

stature having height 130 cm. Patient's obstetric history was uneventful. She denied history of any medical comorbidity or surgical intervention done in the past. Airway examination revealed short neck with MPC-I, jaw-slux +1, with limited neck extension. Vital parameters and system examination were unremarkable. Breath holding time was 22 seconds. Hematological and biochemical investigations were within normal limits. Proposed plan of anaesthesia was spinal anaesthesia in view of reduced neck movement and potential for hyperthermia and post-operative pulmonary complications following general anaesthesia. Technically central neuraxial blocks are also challenging in view of abnormal curvature of spine and abnormal bony landmark of spine. Also the height of block and duration of anaesthesia is unpredictable with spinal anaesthesia. We decided to go ahead with spinal anaesthesia and in case of its failure general

anaesthesia was planned with adequate preparation for difficult airway. Standard ASA monitors were attached. All vital parameters were normal. Local infiltration with 2cc 2% lignocaine was given with hypodermic needle at L3- L4 space. Spinal anaesthesia was given with 23G spinal needle via median approach and 1.4 cc of 0.5% heavy bupivacaine was injected in intrathecal space with great difficulty. Adequate anaesthesia level at T6 was achieved. Sedation was not given to the patient as she was comfortable throughout the procedure. Procedure went uneventful with 500ml blood loss. Weaning of spinal anaesthesia was noted at 45 minutes though procedure was completed before it. Postoperatively analgesia continued with intravenous paracetamol 1 gm 6 hourly and diclofenac suppository 100 mg 12 hourly. Patient had uneventful recovery thereafter.

Discussion

General anaesthesia is definitely a concern in patients with AMC as many of these patients have facial abnormalities such as microcephaly, mandibular hypoplasia, craniosynostosis, and cleft palate including tracheal or laryngeal stenosis. Abnormal range of cervical spine movement makes their neck manipulation difficult during laryngoscopy [3]. Forward subluxation of cervical spine can happen with forcible movement leading to quadriplegia [5]. Hence they are potential candidate for difficult airway during general anaesthesia. Thoracic kyphoscoliosis may cause loss of lung volume and restrictive lung disease which may result in post-operative pulmonary dysfunction following general anaesthesia [3].

These patients also demonstrate an increased sensitivity to neuromuscular blockers and opioids resulting in delayed recovery from general anaesthesia. Though association of AMC with malignant hyperthermia is not supported in the literature but many of them exhibit the hypermetabolic response with hyperthermia even without exposure to triggering agents which can be treated with active cooling only [6]. Hence temperature monitoring is essential in order to detect early changes and correct accordingly. Abnormal spine curvature, short stature, difficult positioning due to contracture may result in abnormal CSF dynamic with unpredictable action of local anaesthetics following regional anaesthesia [7]. In spite of the known structural abnormalities of the spine, the spinal anaesthesia was chosen due to risks associated with general anaesthesia [6]. Epidural anaesthesia was not opted in our patient as she did not give consent for same. Regional anaesthesia has been successfully used in these patients [7] with better success rate and accuracy, if done under ultrasound guidance. As ours was elective surgery, we had additional time for performing spinal block with additional preparation of difficult airway. Unpredictable block height and variable duration of block was kept in mind. Surgeon was requested to keep the procedure time minimal. We also had difficulty in positioning of patient on table in supine position lots of padding and pillows were required to make her lie down comfortably. There was a difficulty in securing intravenous access due to shiny skin and swollen extremity.



Figure 1: Both extremity contracture
Figure 2: Difficult positioning with short neck
Conclusion

Conclusion

These patients should be assessed early by the anaesthesiologists to identify the relevant comorbidities for better perioperative care. Regional anesthesia is a safe option in parturient patient having AMC posted for caesarean delivery though it can be technically challenging.

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