Anaesthetic Management Of A Child With Arthrogriposis Multiplex Congenita

M Inal, G Kaya, Z Pamukcu

Citation

M Inal, G Kaya, Z Pamukcu. *Anaesthetic Management Of A Child With Arthrogriposis Multiplex Congenita*. The Internet Journal of Anesthesiology. 2006 Volume 14 Number 2.

Abstract

Arthrogriposis multiplex congenita is a rare disease characterized by micrognathia, limited mandibuler opening and multiple joint contractures. Difficult airway due to micrognathia and limited mandibuler opening may complicate anaesthetic management in this syndrome.We describe the anaesthetic management of a child with arthrogriposis multiplex congenita syndrome undergoing repair of club feet.

INTRODUCTION

Arthrogriposis multiplex congenita is an uncommon condition characterized by multiple joint contractures([1]). Although most contractures involve the extremities, patients with arthrogripozis multiplex congenita can have maxillofacial involvement including limited mandibuler opening from temporomandibuler joint involvement and microstomia([2,3]).

Arthrogripozis multiplex congenita also effected other organ systems such as cardiovasculer, respiratory, gastroimtestinal, genitourinary and locomotor system([4]). The incidence of this syndrom is 1 per 3000-10000 live births ([1,2]).

Patients with arthrogripozis multiplex congenita frequently require anesthetic care during surgical procedures to correct the orthopedic deformities or the organ systems asssociate with the disease process. Given the multiple organ system involvement of the arthrogripozis multiplex congenita,the potantiel for dificulties with airway management as well as the implications of the underlying neuromusculer disorder, several perioperative concerns must be addressed in these patients ([4]).

We reviewed our perioperative experience of a child with arthrogripozis multiplex congenita.

CASE REPORT

A 9-month-old 20 kg female infant presented for repair of club feet. The birth history revealed that the infant was born at term. She had multiple contractures so that a diagnosis of artrogripozis multiplex congenita was made when she was 3-months-old.

She underwent multiple previous surgeries such as repair of club feet and release of tendons. Physical examination revealed an awake infant with severe multiple joint contractures involving all limbs and marked micrognathia.

After monitorization of the patient with three way ECG, $Sp0_2$ and NIBP, a 24-gauge scalp peripheral intravenous catheter was secured. After 5 minutes of preoxygenation, anesthesia was induced by propofol (2.5 mg kg^{-1}) and fentanyl (1 µg kg^{-1}). Mask ventilation was attempted without dificulty. After neuromusculer block with mivacurium (0.2 mg kg^{-1}) laryngoscopy was performed. The laryngoscopic grade according to Cormarc Lehane grade was ???; the epiglottis could be visualized but the glottic orifis could not. The first intubation attempt was unsuccessful. Following a further failed intubation attempt, at the third attempt a size 3 uncuffed orotracheal tube inserted into the trachea. Immediately after the insertion, positioning was confirmed by breath sounds. Although intubation had been difficult, the patient did not become hypoxic at any stage. Maintenance of anesthesia consisted of sevoflurane supplemented with fentanyl. At the end of 1.5 hours of anesthesia she awaked and was extubated without complications.

DISCUSSION

This case report describes general anesthesia in a child with arthrogripozis multiplex congenita and highlights a number of potantial anesthetic problems. The primary concern to the anesthesiologist is the potantial for airway involvement thereby making direct laryngoscopy and endotracheal

intubation difficult ([2,3,4,5]).

The most common maxillafacial findings are decreased mandibular opening, micrognathia, a high arced palate, deficient musculature of the orofaringeal complex, limited tongue protusion, cleft palate, Pierre-Robin like sequence, short neck, torticollis and hemangiomas of the upper face ([3,4,5,6]). Steinberg et al. reviewed the findings in 23 patients with artrogripozis multiplex congenita and noted a 22 % incidence of maxillofacial involvement with limited mouth opening in three of the 23 patients ([3]). Martin et al. reviewed 25 % incidence of maxillofacial involvement with limited mouth opening in three of the 12 patients ([6]).

In our case the patient had a limited mouth opening, micrognathia and a short neck. The laryngoscopy was difficult and we intubated the patient in the third attempt.

The limited mouth opening in patients with arthrogripozis multiplex congenita can be severe and may make direct laryngoscopy impossible ([2,3,4,5]). Thomas et al. reported a 3 year old patient with arthrogripozis multiplex congenita who presented for TMJ surgery whose initial maximum interincisal opening was only 9 mm ([5]). In our case the child has a limited mouth opening with an interincisal opening only 10 mm.

The patients with arthrogripozis multiplex congenita may be more susceptible to the respiratory depressant effects of various intravenous and inhalation anaesthetics ([1,2,4]). Respiratory problems in intraoperative period and postoperative period may be related to associated myopathy, pulmonary hypoplasia and spine deformities ([1,4,6]). In our case we noted nothing about postoperative atelectasis or a restrictive respiratory pattern, but we would suggest close monitoring for postoperative respiratory function.

Baires et al. reviewed 396 anaesthetic agents in 67 patients with arthrogripozis multiplex congenita and found no episodes of MH with exposure to known triggering agents ([7]). In our case we are not noted intraoperative hyperthermiae.

The choice of neuromusculer agent is also important in patients with arthrogripozis multiplex congenita. In patients with an identified underlying myopathic disorder,there may be an exaggered hyperkalemic response and there may be a caution to the use of succinylcholine([7]).

As sensitivity to nondepolarizing neuromusculer blocking agents has been reported in patients with underlying neurologic and myopathic conditions, our preference was the use of of a short acting relaxant such as mivacurium ([8,9,10]).

In summary, this report highlights the anaesthetic management of arthrogripozis multiplex congenita, which may be complicated by a difficult airway becouse of micrognathia. Appropriate preoperative evaluation and preparation is required.

CORRESPONDENCE TO

Mehmet Turan Inal Trakya Univercity Faculty of Medicine Department of Anaesthesiology 22030 Edirne, Turkey Tel:+902842357641 Fax:+902842358096 e-mail: mehmetturaninal@yahoo.com

References

1. Thompson GH, Bilenker RM. Compherensive management of artrogripozis multiplex congenita. Clin Orthop 1985;194:6-14
2. Epstein JB, Wittenberg GJ. Maxillofacial manifestations and management of arthrogryposis: literature review and case report. J Oral Maxillofac Surg 1987; 45: 274-279.
3. Steinberg B, Nelson vs, Feinberg SE et al. Incidence of maxillofacial involvement in arthrogryposis multiplex congenita. J Oral Maxillofac Surg 1996; 54: 956-959.
4. Oberoi GS, Kaul HL, Gill IS et al. Anaesthesia in arthrogryposis multiplex congenita: case report. Can J Anaesth 1987; 34: 288-290.
5. Thomas JA, Chiu-Yeh M, Moriconi ES. Maxillofacial implications and surgical treatment of arthrogryposis multiplex congenita. Compend Contin Educ Dent 2001; 22: 588-592.
6. Martin S, Tobias JD. Peroperative care of the child with athrogryposis. Pediatr Anesth 2006 ;16:31-37.
7. Baines DB, Douglas ID, Overton JH. Anaesthesia for patients with arthrogryposis multiplex congenita: what is the risk of malignant hyperthermia? Anaesth Intensive Care 1986; 14: 370-372.
8. Tobias JD, Atwood R. Mivacurium in patients with Duchenne muscular dystrophy. Pediatr Anesth 1994; 4: 57-60.
9. Nguyen NH, Morvant EM, Mayhew JF. Anesthetic management for patients with arthrogryposis multiplex congenita and severe micrognathia: case reports. J Clin Anesth 2000;12:227-30
10. Yingsakmongkol W, Kumar SJ. Scoliosis in arthrogryposis multiplex congenita: results after nonsurgical and surgical treatment. J Pediatr Orthop 2000; 20: 656-661

Author Information

Mehmet Turan Inal, M.D.
Consultant, Department of Anesthesiology and Reanimation, Trakya Univercity Faculty of Medicine

Gaye Kaya, M.D.
Asis. Prof., Department of Anesthesiology and Reanimation, Trakya Univercity Faculty of Medicine

Zafer Pamukcu, M.D.
Professor, Department of Anesthesiology and Reanimation, Trakya Univercity Faculty of Medicine