



CASE REPORT

Medicine Science 2016;5(Supp):116-8

Airway management in an osteogenesis imperfecta case in the lateral decubitus position

Muharrem Ucar, Ahmet Selim Ozkan, Duygu Demiroz, Mehmet Ali Erdogan, Ulku Ozgul, Mahmut Durmus

Inonu University, School of Medicine, Department of Anesthesiology and Reanimation, Malatya, Turkey

Received 15 February 2016; Accepted 16 March 2016

Available online 22 March 2016 with doi: 10.5455/medscience.2016.05.8439

Abstract

Osteogenesis imperfecta that is a connective tissue disorder is a rare autosomal disease. Osteogenesis imperfecta in the presence spinal deformity, respiratory disorders, cardiac anomalies and bone deformities may create several problems in the management of anesthesia. The risks of teeth, mandible and cervical trauma and difficulty making positions due to bone deformity challenge difficult airway management. We aim to provide airway management with ProSeal Laryngeal Mask Airway in left lateral decubitus position in a child patient who has severe thoracic spine deformity and can not be given supine position.

Keywords: Osteogenesis imperfecta, PLMA, airway management

Introduction

Osteogenesis imperfecta (OI), which is a connective tissue disease, is a rarely seen autosomal disease. It is known that its frequency is from 1: 5000 to 1: 20.000 [1]. OI is a syndrome involving many skeletal infiltrations such as skeletal involvement, loose joints, thinning of the skin, congenital heart disease, valvular heart disease, core pulmonale, malignant or non-malignant hyperthermia, hyperhidrosis, blue sclera, otosclerosis-related hearing loss, broken tooth structure and platelet dysfunction syndrome [2].

The presence of spinal deformities, respiratory disorders, cardiac abnormalities, bone deformities in OI patients can create a variety of problems in the management of anesthesia. Management of anesthesia carries the challenges of difficult ventilation, difficult intubation, dental and mandibular fractures, the risk of cervical trauma, difficulty in making positions. OI requires a careful management of anesthesia due to the fact of both its being a rare disease and its having features such as difficult airway in terms of anesthetic and malignant hyperthermia risk [3].

The very fragile bone structures in those patients make position-making process and particularly intubation attempts more difficult. Due to short neck, limited neck motion, chest deformity, fragile tissues, abnormal tooth

structure and inability to provide appropriate position for airway control, airway management has an importance [4,5].

We aim to present airway control on the left side by ProSeal Laryngeal Mask (PLMA) for children for whom supine position cannot be given and having OI with severe thoracic vertebrae deformities.

Case Reports

An operation was planned for a thirteen-year-old boy having 35 kg weight due to broken right femur. On physical examination, growth retardation, pectus carinatum, scoliosis and major deformities in lower extremities were available (Figure 1). His head and neck movements were restricted. His mallampati scoring was evaluated as II-III. His preoperative complete blood count, coagulation profile, biochemical values and chest X-ray were normal. His Echocardiography (ECHO) did not reveal any pathology. The environment of his tibial and femoral knee joint was markedly expansive and his joint integrity had deteriorated. For the possibility of development of malignant hyperthermia during the preparation for anesthesia; dantrolene sodium, sodium bicarbonate and cold intravenous serum were prepared. We have avoided volatile agents such as halothane and enflurane, and succinylcholine not to trigger malignant hyperthermia. He was taken to the operating room before the premedication of the case. Electrocardiogram, pulse oximetry, noninvasive blood pressure, temperature monitoring procedures has been carried out. It has been found that the patient's initial heart rate is 120 beats / min,

***Corresponding Author:** Muharrem Ucar, Inonu University, School of Medicine, Department of Anesthesiology and Reanimation, Malatya, Turkey
E-mail: umuharrem@hotmail.com

noninvasive blood pressure is 100/70 mmHg and oxygen saturation is (SpO₂) 93%, body temperature is 36.5 °C. After five minutes preoxygenation, anesthesia induction with 2.5 mg / kg propofol and 1 µg/kg remifentanyl. Carefully keeping the head on the left side position, without harming the lower jaw and teeth, PLMA-3 was placed on the first try. Anesthesia was maintained with total intravenous anesthesia (TIVA) using 4-10 mg / kg / h propofol and 0.25-0.5 mg / kg / h remifentanyl infusion. 50% O₂ and 50% air mixture was ventilated with mechanical ventilator. The operation lasted about 90 minutes. Intraoperative significant hemodynamic changes were not observed. Any increase in body temperature was not observed. At the end of the operation, the PLMA of the patient, whose spontaneous breathing and protective airway reflexes have already formed were, was taken out by lowering its cuff. The patient was taken to the recovery room and was returned to service without problems at his follow-up process.



Figure 1.

Discussion

The patients diagnosed with osteogenesis imperfecta need anesthesia for surgery due to various reasons including most frequently multiple bone fractures. The management of anesthesia to the patients with OI has the challenges such as difficult ventilation, difficult intubation, dental and mandibular fractures, the risk of cervical trauma, difficulty in making positions. An operation was scheduled for right femur fracture in our case. The management of airway had a major importance due to limited head and neck movements, pectus carinatum, scoliosis and the presence

of major deformities at the lower extremities and having difficulty in taking the supine position.

There are a number of studies showing the application of LMA in the control of airways with success for the patients with OI depending on the duration and type of operation. LME application is less invasive than endotracheal intubation, moreover, since it prevents the bone and soft tissue trauma that may occur during intubation for patients with OI, LMA application may be preferred [4-6].

In some studies, it has been reported that the endotracheal intubation of the patients in the lateral decubitus position can be done with the help of light light guided intubation. However, it is recommended that this method be carried out by experienced anesthesiologists. The general anesthesia was administered with success together with the LMA for those patients having difficult airways and in the lateral decubitus position [7].

Considering that the operation duration is not going to be more than 2 hours in our case, in order to reduce possible complications during intubation and extubation, we have performed PLMA in the lateral decubitus position. By using the PLMA, we have avoided complications that can occur with endotracheal intubation and provided a safe airway.

Many publications suggest the application of TIVA in order to prevent intraoperative temperature increases that may occur by different mechanisms including malignant hyperthermia [8]. We also have stayed away from the inhalational agents during the induction and maintenance of our case, and we have implemented the method TIVA with remifentanyl and propofol.

Kill C *et al.* [9] have reported that propofol infusion syndrome is seen in a child having OI after propofol anesthetic (TIVA) lasting about 150 min. In our case, we did not have any problem due to the propofol infusion.

As a result; we have successfully management general anesthesia with PLMA in the lateral decubitus position for those OI patients who cannot be brought to the supine position and have difficult intubation, cervical trauma, dental and mandibular fracture risk, the limited head and neck motion, pectus carinatum, scoliosis, major deformities at the lower extremities.

References

1. Huber MA, Antino S. Osteogenesis imperfecta. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2007;103(3):314-20.
2. Chiarello E, Donati D, Tedesco G, Cevolani L, Frisoni T, Cadossi M, Hoque M, Spazzoli B, Giannini S. Conservative versus surgical treatment of osteogenesis imperfecta: a retrospective analysis of 29 patients. Clin Cases Miner Bone Metab. 2012;9(3):191-4.

3. Ogawa S, Okutani R, Suehiro K. Anesthetic management using total intravenous anesthesia with remifentanyl in a child with osteogenesis imperfecta. *J Anesth*. 2009;23(1):123-5.
4. Karabiyik L, Parpucu M, Kurtipek O. Total intravenous anaesthesia and the use of an intubating laryngeal mask in a patient with osteogenesis imperfecta. *Acta Anaesthesiol Scand*. 2002;46(5):618-9.
5. Kostopanagiotou G, Coussi T, Tsaroucha N, Voros D. Anaesthesia using a laryngeal mask airway in a patient with osteogenesis imperfecta. *Anaesthesia*. 2000;55(5):506.
6. Erdoğan MA, Sanli M, Ersoy MO. Anesthesia management in a child with osteogenesis imperfecta and epidural hemorrhage. *Braz J Anesthesiol*. 2013;63(4):366-8.
7. Komatsu R, Nagata O, Sessler DI, Ozaki M. The intubating laryngeal mask airway facilitates tracheal intubation in the lateral position. *Anesth Analg*. 2004;98(3):858-61.
8. Szmuk P, Ezri T, Soroker D. Total intravenous anaesthesia for patients with osteogenesis imperfecta. *Paediatr Anaesth*. 1994;4:344.
9. Kill C, Leonhardt A, Wulf H. Lactic acidosis after shortterm infusion of propofol for anaesthesia in a child with osteogenesis imperfecta. *Paediatr Anaesth*. 2003;13(9):823-6.