J Anesth (2005) 19:309–310
DOI 10.1007/s00540-005-0329-x

Journal of
Anesthesia

Clinical reports

Anesthetic and airway management of general anesthesia in a patient with Meckel-Gruber syndrome

Mitsunori Miyazu, Kazuya Sobue, Hiroaki Ito, Takafumi Azami, Shoji Ito, Akinori Takeuchi, Hiroshi Sasano, Takako Tsuda, and Hirotada Katsuya

Department of Anesthesiology and Medical Crisis Management, Nagoya City University Graduate School of Medical Sciences, 1 Kawasumi, Mizuho-cho, Mizuho-ku, Nagoya 467-8602, Japan

Abstract
Meckel-Gruber syndrome, characterized by occipital encephalocele, microcephaly, polydactyly, cleft lip or palate, mandibular micrognathism, and anatomical abnormality of the larynx and tongue, along with other associated malformations, is in the list of diseases associated with difficult airway. However, there has been no report on the management of general anesthesia and airway management for such patients. A 2-year-old girl with Meckel-Gruber syndrome was scheduled for cardioplasty and gastrostomy for gastroesophageal reflux under general anesthesia. Preoperative examination revealed obesity, microgenia, dysspondylism, proteinuria, hypoplastic kidneys, and stenosis of the anal canal. Although we anticipated some difficulty with the intubation and prepared several alternative methods for intubation, such as a bronchofiberscope and a laryngeal mask airway, tracheal intubation was completed without difficulty using conventional laryngoscopy after inhalational induction with sevoflurane. Because most patients with this syndrome die before and shortly after delivery, those who survive to some age might have less severe deformities.

Key words Meckel-Gruber syndrome · Sevoflurane · Airway

Introduction

Meckel-Gruber syndrome is a rare autosomal recessive disease. Most babies with it are either medically aborted or stillborn, and the rest die shortly after birth. This syndrome is characterized by occipital encephalocele, microcephaly, polydactyly, cleft lip or palate, mandibular micrognathism, anatomical abnormality of the larynx and tongue, abnormal genitalia, and polycystic kidneys along with other associated malformations [1].

Address correspondence to: K. Sobue
Received: August 9, 2004 / Accepted: April 30, 2005

Although this syndrome is thought to be associated with difficult airway [2], there has been no report on the management of general anesthesia for such a patient. We describe the successful anesthetic management of cardioplasty for a child with Meckel-Gruber syndrome.

Case report

A 2-year-old girl (height 53 cm, weight 4.2 kg) was scheduled for cardioplasty and gastrostomy for repair of gastroesophageal reflux under general anesthesia. She had been diagnosed with Meckel-Gruber syndrome at 34 weeks after birth. Preoperative examination revealed obesity, microgenia, dysspondylism, proteinuria, hypoplastic kidneys, and stenosis of the anal canal. She was premedicated with chloral hydrate (50 mg/kg), which made her sleepy and moderately sedated. Because we anticipated some difficulties with tracheal intubation, a bronchofiberscope and laryngeal masks (sizes 1.0 and 1.5) were made available for intubation. General anesthesia was induced slowly with sevoflurane, nitrous oxide, and oxygen; and a tracheal tube (4.0 mm diameter; Portex) was inserted orotracheally with no difficulty using direct laryngoscopy under adequate muscle relaxation with 1 mg of vecuronium. At the end of 4.5 h of anesthesia, neuromuscular blockade was antagonized with neostigmine, and she awakened and was extubated without complications. The subsequent hospital course and recovery were uneventful. She was discharged on postoperative day 42.

Discussion

In 1822, Meckel first described a rare syndrome consisting of occipital encephalocele, polydactyly, and polycystic kidneys. In 1934, Gruber reported a similar case of

dysencephalic splanchocystosis as Gruber syndrome. In 1969, Opitz and Howe showed that Gruber syndrome was identical to Meckel syndrome, and the syndrome has since been termed Meckel-Gruber syndrome [1].

As far as we know, there are only two previous reports concerning surgical procedures in a patient with Meckel-Gruber syndrome: one for plastic surgery for polydactyly [3] and the other a neurosurgical report on a newborn with occipital encephalocele [4]. Unfortunately, neither described the anesthetic management used. Intubation for a patient with microgenia is generally considered difficult. It is noted that Meckel-Gruber syndrome and Larsen syndrome are occasionally associated with upper airway deformity [2]. Fortunately, we had performed anesthetic management of a patient with Larsen syndrome including airway management, and we reported that sevoflurane is useful for inhalational induction and maintenance of general anesthesia for a patient with the syndrome [5]. Because Larsen syndrome and Meckel-Gruber syndrome are closely related, we selected sevoflurane for induction and maintenance of general anesthesia in this patient, with satisfactory results and without encountering any difficult airway problems. Sevoflurane is a preferred anesthetic agent for pediatric anesthesia because of its rapid induction and emergence (which enable precise control over the depth of anesthesia) and its lack of pungency and agreeable smell (which permit easy administration without discomfort or airway irritation) [6].

We predicted difficulty with orotracheal intubation because of the mandibular micrognathism and anatomical abnormalities of the larynx, tongue, and cervical vertebrae. A bronchofiberscope and laryngeal mask airways were made available before induction of anesthesia for difficult airway problem according to the practice guidelines for management of the difficult airway established by the American Society of Anesthesiologists (ASA) [7–9]. Finally, we could intubate her trachea using conventional direct laryngoscopy without difficulty. This is probably because patients with this syndrome usually die before or shortly after birth, and those who survive to some age may have less severe deformities. However, a safe and successful method of anesthetic management for Meckel-Gruber syndrome has yet to be established, and further case reports are needed.

References

1. McMillan J, Deangelis C, Feigin D, Warshaw J (1999) The nervous system; congenital anomalies of the central nervous system. In: Oski's pediatrics. 3rd edn. Principles and practice. Lippincott Williams & Wilkins, Philadelphia, p1806
2. Motoyama E, Davis P (1990) Clinical management of special surgical problems; anesthesia for general, urologic, and plastic surgery. In: Smith's anesthesia for infants and children. 5th edn. Mosby, St. Louis, p607
3. De Chalain T, Hudgins R, Burstein F, Armfield K (1994) Combination of unilateral polydactyly, syndactyly, and clinodactyly with occipitocervical encephalocele and vertebral fusion. Ann Plast Surg 33:86–89
4. Gazioglu N, Vural M, Seckin MS, Tuysuz B, Akpir E, Kuday C, Ilikkan B, Erginel A, Cenani A (1998) Meckel-Gruber syndrome. Childs Nerv Syst 14:142–145
5. Morishima T, Sobue K, Tanaka S, So M, Arima H, Ando H, Katsuya H (2004) Sevoflurane for general anesthetic management in a patient with Larsen syndrome. Paediatr Anaesth 14:194–195
6. Goa K, Noble S, Spencer C (1999) Sevoflurane in paediatric anaesthesia: a review. Paediatr Drugs 1:127–153
7. Infosino A (2002) Pediatric upper airway and congenital anomalies. Anesthesiol Clin North Am 20:747–766
8. Selim M, Mowafi H, Al-Ghamdi A, Adu-Gyamfi Y (1999) Intubation via LMA in pediatric patients with difficult airways. Can J Anesth 46:891–893
9. American Society of Anesthesiologists Task Force on Management of the Difficult Airway (2003) Practice guidelines for management of the difficult airway: an updated report by the American Society of Anesthesiologists Task Force on Management of the Difficult Airway. Anesthesiology 98:1269–1277