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CLINICAL REPORT

Anesthetic management of a child with Aicardi syndrome undergoing laparoscopic Nissen's fundoplication: a case report

Yui Terakawa · Takaaki Miwa · Yoshiko Mizuno · Tatsuya Ichinohe · Yuzuru Kaneko · Koui Ka

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Abstract Aicardi syndrome (AS) is a rare congenital syndrome and is characterized by the triad of infantile spasm, agenesis of the corpus callosum, and anomaly of chorioretinal lacunae. We here report a case of a patient with AS under general anesthesia. Although there is no report in which muscle relaxants were used in AS patients, vecuronium bromide was used for artificial pneumoperitoneum in this case. Careful management is important for AS patients during an operation that significantly affects respiratory function. In addition, it is possible that muscle relaxants be administered safely in AS patients. Careful monitoring such as epileptiform electroencephalogram and bispectral index monitors may be needed for the early detection of epileptic activities.

Keywords Aicardi syndrome · Pediatric · Anesthetic management · Muscle relaxant

Introduction

Aicardi syndrome (AS) is a rare congenital syndrome and is characterized by the triad of infantile spasm, agenesis of the corpus callosum, and anomaly of chorioretinal lacunae [1]. Severe mental retardation and multiple anomalies accompany AS [1]. AS is not familial, and most AS patients are female [1, 2]. The incidence of AS is reported to be 1 in 10,000 to 1 in 1,000,000 [3]. Attention concerning respiratory management, epileptic seizure, and multiple anomalies should be paid to AS patients undergoing general anesthesia [3–5]. However, appropriate anesthetic management for AS patients remains unclear, and there is no report in which muscle relaxants were used for AS patients.

We here report a case of laparoscopic Nissen fundoplication for a patient with AS under general anesthesia. In this case, vecuronium bromide was used for artificial pneumoperitoneum (AP).

Y. Terakawa (✉) · T. Ichinohe · Y. Kaneko
Department of Dental Anesthesiology, Tokyo Dental College, 1-2-2 Masago, Mihama-ku, Chiba 261-8502, Japan
e-mail: terakawayui@tdc.ac.jp

T. Miwa · Y. Mizuno · K. Ka
Department of Anesthesia, Kanagawa Children's Medical Center, Yokohama, Japan

The case

The patient was a girl aged 6 years and 3 months, 11.5 kg in body weight, and 98 cm in height. She was delivered normally at 42 gestational weeks, and her weight at birth was 3,600 g. She was diagnosed as AS on the basis of symptoms including agenesis of the corpus callosum, left anophthalmia, right nanophthalmia, and infantile spasm.

Phenytoin, clonazepam, and baclofen were administered to control epileptic seizure; petit and grand mal seizures occurred 3–4 times a day and 5–6 times a month, respectively. Additional rectal diazepam was administered when the patient had a grand mal seizure. The patient had recurrent respiratory tract infection and was diagnosed as gastroesophageal reflux (GER) based on an esophageal pH index of 4.3%. Therefore, laparoscopic Nissen fundoplication was planned. Sodium cromoglicate inhalant liquid, salbutamol sulfate inhalant liquid, ambroxol hydrochloride, procaterol hydrochloride hydrate, L-carbocisteine, and tulobuterol tape were used in the usual fashion. Laboratory findings in preoperative assessment were within normal limits, except for rib cage deformity seen on chest X- ray.

Clear fluid and usual doses of the anticonvulsants were administered through the feeding tube 3 and 2 h, respectively, before induction of anesthesia without premedication. ECG, pulse oximetry, and invasive blood pressure were monitored. Before induction of anesthesia, heart rate, blood pressure, and SpO_2 were 73 bpm, 65/35 mmHg, and 98%, respectively. Epileptic seizure was not observed. Anesthesia was induced with 1.5 monitored anesthesia care (MAC) of sevoflurane, 4 l/min nitrous oxide, and 2 l/min oxygen. Atropine sulfate, fentanyl citrate and 1 mg vecuronium bromide were administrated after a 22-gauge intravenous line was secured. Thereafter, the trachea was intubated with a 5.0-mm uncuffed tracheal tube with its tip 3 cm below the vocal cord. The lung was mechanically ventilated with air, oxygen, and 1–1.5 MAC sevoflurane. The size of the tracheal tube was determined based on a chest X-ray shadow. It was confirmed that the tip of the tracheal tube was not located in the main bronchus by chest auscultation. The vocal cord was clearly visible with a laryngoscope (Cormack grade II), and the trachea was intubated without difficulty. After tracheal intubation, inhalational salbutamol sulfate was administered because wheezing was confirmed throughout inspiration and expiration. However, no significant improvement was observed. Caudal anesthesia was performed using 0.375% ropivacaine. Vecuronium bromide at 0.5 mg was added at a train-of-four (TOF) ratio of approximately 20%; the total vecuronium bromide dosage was 2.5 mg.

After 2 h of AP, SpO_2 was decreased to 94–96%. Manual inflation, tracheal suctioning, and salbutamol sulfate spraying were administered to minimize alveolar collapses. SpO_2 was increased to 98%, and the lung was mechanically ventilated again. These treatments were repeated when SpO_2 tended to decrease. FIO_2 was controlled between 0.4 and 1.0. After AP, SpO_2 was maintained between 97% and 99% under $FIO_2 = 0.4$. At the end of anesthesia, the trachea was suctioned and the patient was extubated under adequate spontaneous breathing. SpO_2 was maintained between 97% and 99% under O_2 inhalation of 4 l/min, and chest X-ray examination was within normal limits. The postoperative lung rale showed no change in comparison with the preoperative state. Epileptic seizure was not observed. Duration of AP, and operation and anesthesia time, were 3 h and 2 min, and 4 h and 50 min, respectively.

Postoperative management was intensively performed by suctioning the trachea and spraying salbutamol sulfate and epinephrine. Petit mal seizure occurred 3–4 times a day, whereas grand mal seizure was not observed. The patient was discharged 7 days after surgery.

Discussion

AS is an X-linked dominant disorder [1], but the etiology remains unknown. Brain, face, rib, and vertebral anomalies occur frequently in addition to the triad of anomalies [1]. Infantile spasm is often drug resistant and intractable [2]. Swallowing function is undeveloped, based on agenesis of the corpus callosum [2]. Therefore, an eating disorder and recurrent aspiration pneumonia are highly possible. Prognosis for AS is poor, and it is said that the most common cause of death is respiratory disease such as aspiration pneumonia. The important anesthetic implications for AS patient are reported to be respiratory management, control of epileptic seizure, and difficult airway as a consequence of facial anomaly [3–5].

In this case, an increase in airway resistance, and decreases in PaO_2 and tidal volume, were anticipated because of recurrent respiratory infection and application of AP. If SpO_2 decreased during anesthesia, we had planned application of tracheal suctioning and alveolar recruitment maneuver to prevent alveolar collapse. In fact, respiratory management was frequently required during the entire hospital stay because of respiratory secretion. Although the need of respiratory management was indicated in a previous report [3], it is suggested that more careful management is important during an operation, which significantly affects respiratory function such as AP, lateral position, and one-lung ventilation.

The main effects of AP on the respiratory system are increases in airway pressure and $PaCO_2$ and a decrease in PaO_2 [6]. In contrast, there are no effects on lung compliance and airway resistance [7]. Control of tidal volume, respiratory rate, and positive end-expiratory pressure (PEEP) are usually necessary to maintain appropriate ventilation during AP. In this case, tidal volume and respiratory rate were controlled at approximately 10 ml/kg and 15 bpm, respectively. Although there may be no specific anesthetic management during AP for the AS patient, tracheal suctioning and alveolar recruitment maneuver might be needed to a greater extent in comparison with an uncomplicated patient.

GER is frequently accompanied with aspiration pneumonia, and intensive attention should be paid to the risk of aspiration during anesthesia induction and intubation [8]. In addition, cricoid pressure should be necessary for tracheal intubation for the severe GER patient [8].

In this case, gastric content was sufficiently suctioned before anesthesia induction because a gastric tube was inserted before surgery. However, venous cannulation was predicted to be difficult, and epileptic seizure might be induced by repetitive venous punctures. Furthermore, reflux

of gastric content might be provoked by increased muscle tension. Therefore, in this case, we considered that slow induction was appropriate for the prevention of epileptic seizure and reflux of gastric contents. In this case, anesthesia was maintained with sevoflurane, because it is a potent bronchodilator [9] with no effects on GER [10].

Vecuronium bromide was used because of AP. There are no reports on metabolic abnormalities in AS patients. In this case, additional vecuronium bromide was administered at a TOF ratio of approximately 20%, and the administration interval was approximately every 60 min. Adequate spontaneous breathing was restored at the time of extubation, and no antagonist of muscular relaxant was used. The effect of a muscle relaxant is dose-dependently enhanced by inhalation anesthetics [11], although the duration of action of a muscular relaxant is shortened by anticonvulsants [12]. The duration of action of vecuronium bromide is approximately 40 min [13] to 60 min [14]. Therefore, it is suggested that vecuronium bromide may be safely used in AS patients as well as in patients without complications, although the metabolism of muscular relaxant is not completely understood.

Epileptic seizure was not observed during general anesthesia in previous cases [3–5] or in the present case. General anesthesia for epileptic seizure patients is desirable when epileptic seizure is well controlled and blood concentration of anticonvulsant is appropriately maintained [15]. Administration of the anticonvulsants should be continued until the morning of surgery and resumed immediately postoperatively. In addition, minimally invasive surgery such as laparoscopic surgery is indicated to prevent epileptic seizure.

In this case, anesthesia was maintained with 1–1.5 MAC of sevoflurane. Sevoflurane may induce an epileptiform electroencephalogram (EEG) change [16]. In a previous report, minimal seizure-like activity was observed in 80% of pediatric patients during slow induction under 8% sevoflurane in 50% O_2 and 50% nitrous oxide [17]. In contrast, in another report, no epileptiform EEG was observed during similar conditions [18]. Although the effect of sevoflurane on epileptiform EEG change remains to be clarified, it has been said that a high concentration of sevoflurane is one of the risk factors of epileptiform EEG change [16]. For this reason, concentration of sevoflurane is recommended to be maintained at about 1.5 MAC or less using benzodiazepine premedication, nitrous oxide, and opioids during maintenance anesthesia [16]. In this case, sevoflurane was maintained at 1–1.5 MAC using nitrous oxide, fentanyl, and caudal anesthesia. Epileptic seizure was not observed throughout anesthesia. However, muscle relaxants could cancel body movement including epileptic seizures because EEG and bispectral index (BIS) monitoring might be appropriate to detect epileptic activities.

In the present case, intubation was easy, although craniofacial dysplasia was observed. In a previous report, although nasal intubation was planned, fiberoptic oral intubation was performed because of craniofacial dysplasia [3]. Therefore, enough attention should be paid to craniofacial dysplasia and a possible difficult airway in AS patients.

Anesthetic management appropriate for AS patients has not been established. Previously, nitrous oxide [4], sevoflurane [5], and remifentani [4] have been successfully used for AS patients. It is suggested that the anesthetic management of systemic complications should be important for several types of anesthesia for AS patients.

In conclusion, careful management is important for AS patients during an operation that significantly affects respiratory function. In addition, muscle relaxants may be administered safely in AS patients. Careful monitoring such as EEG and BIS monitors may be needed for the early detection of epileptic activities.

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