General anesthesia for a spinal muscular atrophy type I patient undergoing feeding gastrostomy –A case report–

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Spinal muscular atrophy (SMA) in children leads to progressive muscle weakness, dysphagia, aspiration, and death. The most common and severe form of SMA is designated as type I, also known as Werdnig-Hoffman Disease or Floppy Baby syndrome. We anesthetized an 8 month-old female infant with SMA type I undergoing feeding gastrostomy. We planned to use inhalational anesthesia without muscle relaxants. Anesthesia and surgery were uneventful. We herein report a case of successful peri-operative anesthetic management for SMA type I infant with aspiration pneumonia. (Anesth Pain Med 2010; 5: 329∼332)

Key Words: Gastrostomy, Neuromuscular disease, Spinal muscular atrophy type I.

Spinal muscular atrophy (SMA) with Duchenne muscular dystrophy in children is the most common hereditary neuromuscular disease. The disease occurs when there is loss of myelinated fibers and axonal damage in sensory and mixed nerves in the anterior horn cell. Ninety-eight percent of patients with SMA has genetic deficiency in the 5q13 chromosome [1].

Respiratory complication is the main cause of death in children with spinal muscular atrophy. Hypoventilation, paradoxical movement, decline of coughing ability and atelectasis occur due to the weakening of muscles. As dysphagia and esophageal reflux take places, pneumonia and respiratory failure will occur repeatedly [2]. Specifically, the natural history of SMA type I is such that 80% of children will die within the first year of life. Children affected by SMA type I typically present with the most severe neurological deficits, such as generalized weakness, within the first 3 months of life. Bulbar dysfunction is universal in these patients and typically leads to feeding and swallowing difficulties, which result in aspiration pneumonia and eventual death.

Problems such as muscles weakness, respiratory complications secondary to anesthesia, hypersensitivity towards depolarizing muscle relaxant and hyperkalemia secondary to succinylcholine present challenges in anesthetic management in SMA.

This is the first domestic report on the outcome of general anesthesia without muscle relaxants administered to an 8-month-year old patient who has been diagnosed with SMA type I and required a feeding gastrostomy to manage recurrent aspiration pneumonia and poor oral intake. The case report will be accompanied by a review of the literature.

CASE REPORT

An 8-month-old female patient with a history of recurrent aspiration pneumonia and dysphagia is admitted to our hospital to undergo insertion of feeding gastrostomy.

She was born following cesarean section when her gestational age was 39 weeks and 2 days. Her weight was 2.92 kg at the time, and the Apgar score at 1 minute and 5 minutes were 9 and 10 respectively. At her birth, she was admitted to the neonatal intensive care unit for 5 days due to whole body meconium. When she was about 2 months old, she was suspected of having floppy infant syndrome for which she was investigated with muscle biopsy and electromyogram, which confirmed the diagnosis with SMA type I. She was admitted to this hospital when she was 4 and 5 months old for treatment of recurrent aspiration pneumonia.
At 8-month-old, while she was admitted to our hospital for insertion of feeding gastrostomy to manage recurrent aspiration pneumonia with atelectasis. Her height and weight were 70.7 cm and 7.3 kg respectively, and her blood test and electrocardiogram taken before surgery were normal. There was an impression of pneumonic consolidation in left lower and right lower lobes combined atelectasis in left lung on chest radiograph taken the day before surgery (Fig. 1). There was no movement below the cervical region, muscle power of grade 2 was recorded in the head and neck, but sensory integrity was not measured because the patient did not cooperate. In room air, her SpO2 was 94−96%, her heart rate was 138−145 beats per minute, her respiration rate was 35 beats per minute, and temperature was 37.5°C before surgery. Regular sinus rhythm was recorded by electrocardiogram.

No other medication was administered before anesthesia and after pre-oxygenation was fully performed with 6 L/min of oxygen, induction of anesthesia was attempted with ketamine 10 mg intravenous injection, which failed to induce her unconsciousness. In addition to ketamine 15 mg was injected and after checking unconsciousness of the patient, sevoflurane 2.5%, O2 1.5 L/min and air 1.5 L/min was supplied. After manual bagging for 5 minutes, endotracheal intubation was performed using a 3.0 size reinforced endotracheal tube without administering any muscle relaxant. Analgesic maintenance was achieved solely with sevoflurane 3%, O2 1.5 L/min and air 1.5 L/min. Because spontaneous breathing was weakly preserved in this patient, we elected to perform manual bagging below 20 cmH2O of peak airway pressure while maintaining a respiration rate of 40 times per minute, as it appeared more beneficial than mechanical ventilation.

The total operating hour was 1 hour and 20 minutes, during which her pulse was maintained at 140−150 beats per minute and her end tidal carbon dioxide tension was maintained at 40 − 45 mmHg. Surgery proceeded without great change in the vital signs. Sevoflurane was reduced to 1.5% before the end of the surgery and was discontinued after the end of surgery, followed by administration of 100% oxygen. When surgery was complete, the patient regained spontaneous breathing (tidal volume 50−60 ml, minute respiratory rate 35 times, end tidal carbon dioxide 40 mmHg) and was subsequently transferred to the recovery room while maintaining the endotracheal tube, as requested by the pediatricians at our hospital.

The patient maintained SpO2 of 100% on 2 L/min of oxygen through a cannula into the endotracheal tube in the recovery room and pulse rate of 180−190 beats per minute. Because there was no change in her vital signs after an observation period of 20 minutes, the patient was transferred to a general ward with oxygen saturation of arterial blood and pulse monitoring. Arterial blood gas analysis of the patient post-surgery was as follows: pH 7.31, PaCO₂ 44 mmHg, PaO₂ 126 mmHg, SpO₂ 99%. The patient maintained SpO₂ of 94−95% with 0.5 L/min of oxygen through a cannula directed into an endotracheal tube in the general ward on the first postoperative day. On the second postoperative day, SpO₂ was maintained as 94−95% with 0.5 L/min of oxygen through a cannula into an endotracheal tube, and subsequently underwent extubation. After the extubation, 1 L/min of oxygen was supplied by a nasal cannula and the SpO₂ was maintained at 96−97%. Arterial blood gas analysis results were as follow: pH 7.44, PaCO₂ 37 mmHg, PaO₂ 73 mmHg, SpO₂ 95%.

Following surgery, the patient was discharged from the hospital on the thirteenth postoperative day with improved aspiration pneumonia and atelectasis and normal vital signs.

**DISCUSSION**

Spinal muscular atrophy is a recessive genetic disorder and occurs when there is the deficit of 5q13 chromosome and atrophic anterior horn cell of the spinal cord [3]. Deficit of a gene occurs in 98% of spinal muscular atrophy patients. The prevalence is approximately 1/50,000 [4]. There are three types based on the age presentation of clinical symptoms, the extent of muscle involvement, age of death, and deficit of genes [5].

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**Fig. 1.** Preoperative chest X-ray shows pneumonic consolidation in the left lung and right lower lung.
The symptom grade is in inverse proportion to the survival of motor neuron in the anterior horn cell of spinal cord [6].

SMA is divided into three types. The type I, Werding-Hoffman disease, is defined when an individual is unable to sit independently [7]. Fifty percent of patients diagnosed with SMA type I die before 7 months after birth, and 80% of patients die before 12 months after birth. Most patients die before the age of two [6]. Upper respiratory infection and dysphagia or aspiration pneumonia due to esophageal reflux or respiratory failure occur frequently [1]. Children with SMA type II develop symptoms of weakness between 6 and 18 months of age, are able to sit independently but not able to stand independently. If no intervention is provided, they may survive into the third decade of life. SMA type II also exists in an intermediate form. Children with SMA type III are symptomatic after 18 months of age, often present with an abnormal gait, and have a normal life expectancy. SMA type III is also known as Kugelberg-Welander disease or juvenile SMA [7].

In this case, a child has a SMA type I and is born with grade 2 cephalic movements. Our patient in particular tired early during feedings and quickly became unable to achieve adequate caloric intake to prevent weight loss, resulting in worsening muscle atrophy. As the disease progressed, bulbar dysfunction, respiratory muscle fatigue, and malnutrition led to recurrent aspiration and respiratory tract infections, each of which could potentially become a life-threatening pneumonia. However, in centers that routinely care for children with complex neuromuscular disorders (NMDs), the life expectancy for children with SMA type I have been extended from less than 1 year to more than 2 years of age. Advances in airway clearance, noninvasive ventilatory support and nutritional supplementation increase the likelihood of improved survival. In addition, systematic integration of pediatric subspecialty management, as recommended by the International SMA Consortium, greatly enhances the care of these patients. Feeding gastrostomy should therefore be performed, because gastrostomy in infants with SMA type I could be performed safely, lead to fewer aspiration events and improved nutritional status.

As for precautions during general anesthesia, it was difficult to decide on the recovery of muscular strength, because there is no motor function beneath the cephalic area, and there was worsen of aspiration pneumonia due to increased risks of airway reflux based on dysphagia. There was also the possibility of admission to intensive care unit and mechanical ventilation due to weakening of respiratory muscles after the operation.

Paying attention to use of muscle relaxants is important not only in spinal muscular atrophy but also in other neuromuscular disease, because extension of muscle relaxation and delayed recovery can occur. Extension of muscle relaxation secondary to reduction in the density of acetylcholine at neuromuscular junction endplate, which causes reduction in the secretion of cholineacetyltransferase and acetylcholinesterase and delay recovery when depolarizing muscle relaxant, such as succinylcholine, is used. As such, it is essential to pay extra-attention to respiratory muscle recovery after the operation [8,9].

In this case, we anticipated prolonged recovery of spontaneous breathing if muscle relaxant was to be used to reduce the possibility of worsening of atelectasis, aspiration pneumonia and the necessity of mechanical ventilation. Accordingly, we did not use any non-depolarizing muscle relaxants and also did not use succinylcholine, because it has been reported to cause rhabdomyolysis and hyperkalemia from denervated muscles. Therefore, that is contraindicated in SMA [10]. This event occurs secondary to proliferation of extra-junctional receptors in these patients with lower motor neuropathy. Our patient had mild but secured spontaneous breathing but also pneumatic consolidation with atelectasis and excessive secretion in the oral cavity. As such, endotracheal intubation was performed to reduce the risks of pulmonary aspiration. Laryngeal mask can be used during general anesthesia if the SMA patients without bulbar involvement. However, in patients with paralysis secondary to bulbar involvement, like our patient with SMA type I, use of laryngeal mask should be considered carefully. However, we thought tracheal intubation with manual bagging would provide secure and controlled ventilation, and would be tolerated with deep anesthesia without muscle relaxants.

If SMA patients develop respiratory failure during the operation, the degree of respiratory failure needs to be assessed by arterial blood gas analysis. It is certainly necessary to use an instrument to detect the level of muscle relaxation, such as the train-of-four stimulation (TOF) measuring instrument, while muscle relaxant is being used. However, we believed that ‘classical’ TOF monitoring of neuromuscular block with a nerve stimulator is unreliable in patients with lower motor neuron disease, because SMA is usually more prominent in the proximal muscles of the limbs [11]. Furthermore, differential involvement of single muscles by SMA made TOF monitoring equivocal during recovery of all muscles from the muscle.
relaxant [12].

Very little information is available in the anesthetic literature regarding the management of patients with SMA, except to confirm that muscle relaxants, opioids and thiopental could all have a prolonged duration of action [13]. Watts JC said that total intravenous anesthesia using short acting drugs may provide an ideal way of avoiding longer acting medication, the action of which can be unpredictably prolonged by the underlying condition [14]. Also, perioperative care can be provided for children with SMA safely and effectively with inhaled anesthetics (e.g. sevoflurane, isoﬂurane) to improve patient comfort without increased morbidity [15].

SMA type I results in diffuse respiratory muscle weakness with greater weakness developing in the intercostal muscles, compared to the diaphragm. Respiratory muscle weakness results in increased difficulty in clearing lower respiratory secretions and hypoventilation during sleep, especially during viral respiratory infections. Interventions, including airway-secretion mobilization and clearance techniques, and respiratory support with noninvasive ventilation (NIV) with bilevel positive airway pressure may be used. During acute illnesses, airway-clearance techniques and NIV are increased. In addition, nutrition and hydration should be maintained, and there is a low threshold for antibiotic administration. Important to the success of home management is education of families to provide respiratory care to their children on a long-term basis punctuated with acute illnesses.

There were reports on several other neuromuscular diseases, but there is no domestic report on the anesthetic care in patients with SMA type I. This is the first report on the insertion of feeding gastrostomy implementation on the patient who has SMA type I. The patient safely underwent general anesthesia with sevoflurane. Extubation should be performed thorough observation of vital signs during the operation, checking the definite recovery of spontaneous breathing after the operation and evaluation of ventilation ability. However, the anesthetist should be vigilant, against the background of bulbar dysfunction and the possibility of aspiration. Although no complications from the use of short acting non-depolarizing neuromuscular blockade were reported, SMA patients may still have an increased sensitivity to muscle relaxants. These agents should be used judiciously and with close monitoring of clinical effects.

REFERENCES