CASE REPORT

General Anesthesia Plus Muscle Relaxant in a Patient with Kugelberg Welander Disease: A Case Report

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Spinal muscular atrophies (SMAs) represent a rare group of inherited disorders that cause progressive degeneration of the anterior horn cells of the spinal cord. The exact cause of the degeneration is unknown. Loss of these cells results in a progressive lower motor neuron disease that has no sensory involvement and that is manifested as hypotonia, weakness, and progressive paralysis. Kugelberg Welander spinal muscular atrophy (also known as Wohlfart-Kugelberg-Welander syndrome or mild SMA) is a milder form of SMA, with symptoms typically presenting after age 18 months. Here in we report a case of anesthetic management of a patient with Kugelberg Welander disease who was refered for squint surgery and also we reviewed some other cases of SMA patients receiving different types of anesthesia.
Keywords: spinal muscular atrophy; squint surgery; muscle relaxant

A 22-year-old man (height 1.73 m; weight 66 kg) with Kugelberg Welander disease was scheduled for squint surgery. The diagnosis of Kugelberg Welander disease was based on neurologist consult. Physical examination revealed that he had proximal muscular atrophy of the limb, muscular strength 3–4 degrees (with 4 degrees in the upper limb and 3 degrees in the lower limb), decreases of tendon reflex in extremities, no pyramidal tract signs, and paraesthesia. Chest X-ray and ECG were normal. The laboratory examinations were normal. On the operating day, no premedication was given to the patient. Electrocardiogram, cuffed blood pressure, SpO2 and EtCO2 were monitored. Anesthesia was induced with remifentanil 0/5mic/kg and propofol 2mg/kg and a size 4 laryngeal mask airway (LMA) was tried, but failed. Then atracurium 30 mg was injected to facilitate the endotracheal intubation. The patient was intubated with an insertion of a size 8 endotracheal tube. Anesthesia was maintained with intravenous remifentanil and propofol. The hemodynamic parameters were stable during surgery. Paracetamole 15mg/kg was given intravenously 30 minutes before the end of operation. The patient woke up quickly after cessation of anesthesia and he could raise his head. Antagonists of muscle relaxant (neostigmine 2.5mg plus atropine 1.25mg) were given. He was extubated and transferred to the intensive care unit. He returned to the general ward on the following day and was discharged from hospital 3 days after operation without complication.

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Discussion

Common characteristic features of SMA disorders are the degeneration of the anterior horn cells in the spinal cord and motor nuclei in the lower brainstem. Depending on the age of onset and clinical manifestations, these diseases can be classified. The incidence of spinal muscular atrophy ranges from 4 to 10 per 100,000 live births, and the carrier frequency of disease-causing SMN1 mutations ranges from 1/90 to 1/50 [1-4].

Classification: SMA type 1 which is known as infantile spinal muscular atrophy or Werdnig-Hoffmann disease, is the most common and severe form of SMA. Its manifestations generally present in the neonatal period. In these neonatal forms, symptoms progress rapidly, and most infants die before one year of age as a result of respiratory failure [5-6].

SMA 2 (intermediate form) and SMA 3 (mild form; Kugelberg-Welander disease) have a later onset and a less severe course [7-8]. SMA 2 presents between 3 and 15 months of age, while presentations of SMA 3, generally presents are muscle weakness at or after one year of age and progression to a chronic course. In a study of children and adolescents with SMA 2 and SMA 3, muscle strength was reduced to a variable extent [9]. The outcome depends primarily on the severity of muscle weakness at presentation rather than the age of onset, but earlier onset tends to correlate with greater weakness [10].

Adult onset of SMA (type 4) usually presents in the second or third decade of life and is otherwise similar to SMA type 3 [11-13].The management of anesthesia in patients with SMA is often challenging due to muscle weakness, anesthesia induced respiratory complications, hypersensitivity to nondepolarizing muscle relaxants, and succinylcholine-induced hyperkalemia. Also neuraxial (epidural or spinal) blocks could worsen the weakness. Very little information is available in the anesthetic textbooks regarding the management of such cases although it was indicated that muscle relaxants, opioids, and thiopental

could all have a prolonged duration of action [14]. Several cases were reported about anesthetic management of patients with SMA without using muscle relaxants. Watts reported a 25-year-old female with SMA type III presented for urgent corneal grafting due to keratoglobus under general anesthesia. Anesthesia was induced using alfentanil and propofol. After insertion of laryngeal mask airway (LMA), anesthesia was maintained with propofol and remifentanil infusions; controlled ventilation was maintained throughout the operation without muscle relaxants. When the infusions were stopped, spontaneous respiration returned almost immediately and LMA was removed within 5min [15]. The author suggested that total intravenous anesthesia (TIVA) may provide an ideal anesthetic regimen for such patients. Habib et al. reported a 23-year-old parturient of SMA type II that underwent elective cesarean section (CS) under general anesthesia. After induction of anesthesia with alfentanil and propofol, cricoid pressure was applied, and endotracheal tube was easily inserted without muscle relaxants. Anesthesia was maintained using 50% nitrous oxide in oxygen and 0.5–1% isoflurane; for intraoperative analgesia morphine (6mg/IV) was given after delivery. The patient was extubated at the end of the procedure, and was transferred to the intensive care unit (ICU). Following an overnight stay in the ICU and being moved to general labor ward, she was discharged 5 days later [16]. The authors suggested that although succinylcholine was classically used for rapid-sequence induction of anesthesia in the obstetric patients, its use was contraindicated in patients with SMA because of the high risk of life-threatening hyperkalemia and rhabdomyolysis, and the use of nondepolarizing muscle relaxants should also be avoided. Propofol-remifentanil technique was alternatively used instead of rapid sequence induction without muscle relaxants. Kitson et al. reported a 38-year-old type III SMA parturient receiving CS who had a history of tracheostomy during previous pregnancy. Because of the history of failed intubation she received awake fibreoptic intubation (FOI). Anesthesia was induced with alfentanil and propofol and maintained with isoflurane, nitrous oxide, and oxygen without muscle relaxants. She was extubated soon after the operation. She had a smooth uncomplicated recovery and was discharged home 2 days later [17]. Additionally, several case reports exist about using muscle relaxants during general anesthesia. A 24-year-old female with type III SMA at 38 weeks' gestation was admitted for CS. For premedication oral ranitidine 300 mg plus metoclopramide 10 mg were administered. Anesthesia was induced with thiopental and rocuronium; laryngoscopy and tracheal intubation proceeded uneventfully. Anesthesia was maintained by intermittent positive pressure ventilation with 50% nitrous oxide in oxygen with isoflurane 1%.With the exception of fentanyl 100 µg administered after the delivery of the baby, no opiates were given intraoperatively. Reversal of neuromuscular blockage was not attempted until at least 40min after the administration of rocuronium. After reversal of neuromuscular blockage, spontaneous respiration quickly returned and anesthesia was discontinued. After regaining consciousness and despite adequate spontaneous respiration without dyspnea, there was significant residual weakness of the muscles of the upper limbs, head, and neck. The patient was transferred to SICU where she regained her preanesthetic pattern of muscle weakness after 8 h. She was transferred to the postnatal ward 24 h later and was discharged home after one week [18]. Stucke and Stuth reported the use of a nondepolarizing neuromuscular blocking agent in an 18-month-old child with SMA [19]. For anesthetic induction thiopental and alfentanil were used; a premature attempt at direct laryngoscopy provoked laryngospasm and inability to ventilate, so rapacuronium was administered. Within 60 s, mask ventilation was restored and the child was intubated without difficulties. Within 15min, the authors observed some diaphragmatic recovery, and after emergence from anesthesia, the child showed adequate respiratory effects but the strength of the upper extremity muscles was diminised. Small dose of midazolam was given to reduce the anxiety, and the patient was extubated within 5 h without any complications. They suggested that the significant difference in recovery times of the diaphragm and the upper limb muscles in the patient were most likely due to the different involvement of these muscle groups in the primary disease. In other cases, spinal, epidural, or combined spinal and epidural (CSE) anesthesia was selected for SMA patients during labor (either for CS or for labor analgesia). It was believed that parturient with SMA presented several problems for anesthesiologists. General anesthesia was complicated by underlying restrictive lung disease, sensitivity to nondepolarizing muscle relaxants, potential for hyperkalemia with succinylcholine and likelihood of difficult intubation. Regional anesthesia can be technically difficult. Epidural anesthesia may fail due to inadequate spread of local anesthetics, particularly if there had been corrective back surgery. Dose requirements for spinal anesthesia were difficult to predict, increasing the risk of either a failed or high block. CSE or continuous spinal technique may allow the block height to be titrated more carefully and should be considered ideal [20-24]. In our case, nondepolarizing muscle relaxant was used, and the time of onset and recovery of atracurium remained in the normal range. The reason may be that these patients had different types of SMA, in which our patient had type 3 SMA, with later onset and a milder impairment of motor function. In conclusion, nondepolarizing muscle relaxants could be used safely in SMA type 3 patients, and the combined infusion of remifentanil and propofol (TIVA), providing an ideal condition for anesthetic induction and maintenance. Regarding recovery of muscle strength after anesthesia, it is advisable that train of four (TOF) monitoring should be carried out to secure the patients with SMA.

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