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LETTER TO THE EDITOR

Bariatric Surgery for a Patient with Kennedy's Disease

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Dear Editor,

Kennedy's disease (KD) is a rare, X-linked recessive inherited, lower motor neuron disease, which was reported firstly by Kennedy in 1968 [1]. As La Spada reported in 1991, KD was associated with an abnormal amplification of the first gene exon trinucleotide repeat sequence in androgen receptor gene [2]. The clinical characteristics of the disease include the slow progression of the weakness, atrophy, and tremor of the medulla and limbs muscles. It possibly accompanies muscle pain spasm, sensation loss, testicular atrophy, and infertility. The prevalence of KD is approximately 2:100,000 males [3].

Patients with morbid obesity who suffer from KD can become much more mobile upon significant weight loss after bariatric surgery. But the risks of bariatric surgery and general anesthesia are much higher due to the abnormal medullary function and muscle weakness. Herein, we report the first case of a patient with KD receiving laparoscopic sleeve gastrectomy under general anesthesia.

The 49-year-old male patient with morbid obesity had a preoperative body mass index (BMI) of 35.5 kg/m^2 and weighed 100.2 kg. He was diagnosed with OSAS, type 2 diabetes mellitus, and hypertension. The patient first observed muscle weakness and fatigue in both the lower limbs 11 years ago. On admission to the hospital, he had completely lost his ability to climb the stairs, but was still able to walk on the flat ground. He also had slurred speech and slight difficulties in swallowing, occasionally accompanied by severe choking.

Physical examination demonstrated waddling gait while walking. He also had dysarthria. The pharyngeal reflex was weakened; fasciculation and atrophy of the tongue muscle were apparently found. The muscle strength of both the lower limbs was grade IV, and tendon reflex was negative. Laboratory tests showed the following values: creatine kinase 2378 U/L, C peptide 5.74 ng/mL, insulin 49.7 uU/mL, fasting glucose 11.94 mmol/L, and glycosylated hemoglobin 6.6%. Blood gas analysis and pulmonary ventilation function were found as normal.

The operation was performed smoothly with the routine procedures. Neuromuscular block was monitored using train-of-four (TOF) stimulation with a muscle relaxation monitor. The TOF value maintained 0 during the operation. After the procedure, neostigmine was given as muscle relaxant antagonist; the TOF value still continued to be unchanged. The patient was transferred into the ICU, and mechanical ventilation was put on. The tracheal catheter was removed 167 min following the anesthesia. The patient's oxygen saturation remained more than 98%. No apparent changes were observed in the muscle strength, respiration, language, and swallowing function following the operation. No postoperative complications were observed as well. The patient was discharged on the fifth day following the operation.

The patient was followed up and percentage of excess weight loss (%EWL) was 57.5% at 6 months postoperatively. His blood pressure and blood glucose were controlled at the normal level without medication. The patient still required CPAP therapy when asleep, but snoring was apparently alleviated. His walking ability was improved but he still had difficulty on stairs.

The genetic test is the gold standard for the diagnosis of KD. The patients' trinucleotide repeats in the first exon of the androgen receptor gene above 35 CAGs, along with the other clinical manifestations and electrophysiological findings, could be diagnosed as KD [4]. KD primarily exerts impact on adult males, and the age of onset typically ranges between 40 and 60 years old [5]. At the beginning of the disease, it is

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usually characterized by the postural tremor and muscle spasm. With the exacerbation of the disease, the patients gradually lose their walking ability and, eventually, become wheelchair-dependent. The symptoms of bulbar dysfunction are also progressively aggravated, which gradually appear in dysphagia and dysarthria. The patients ultimately die of respiratory failure or inhalation pneumonia [5].

The patients have a long course of disease, and the risks of anesthesia and operation are different at various stages. The precise pre-operation assessment of the KD patient constitutes a pivotal section in the determination of the risk of the operation. Pulmonary function test was regarded as a decisive indicator of the surgical risk. The impairment of respiratory muscle function was expected to impact the recovery and maintenance of postoperative spontaneous breathing. In this case, the patient's pulmonary function index was still in the normal range. In consequence, he was able to tolerate the anesthesia and operation well.

In this case, the patient suffered with manifestations of bulbar lower motor neuron injury, including such as tongue atrophy and dysarthria. However, the patient's spontaneous respiration, followed the extubation, was normal. It suggested that the anesthesia and intubation had not given rise to further impairment to the remaining bulbar function. Use of a semireclining position subsequent to the extubation effectively kept the airway unobstructed and made the patient breathe more easily owing to the decline of the diaphragm. The KD patients with the bulbar function damaged are likely to have spontaneous larynx spasms. Due to the malfunction of the bulbar muscle, the pharyngeal reflex is retarded, and the self-cleaning ability of the airway secretions is weakened, which is likely to result in respiratory complications. Late-stage patients usually have pulmonary aspiration. It was reported that a patient with KD developed respiratory distress following general anesthesia [6]. Thus, it is important to pay enough attention to such potential anesthetic risk. The neuromuscular blocker should be cautiously selected as well. The risk of hyperkalemia was significantly increased in the patients with severe lower motor neuron disease when the depolarizing muscle relaxant was utilized [7]. Therefore, we used rocuronium, a non-depolarizing muscle relaxant, to avoid it. Nonetheless, because of the KD patient's low acetylcholine level, the sensitivity of the non-depolarizing muscle relaxant is expected to increase; in other words, even a small dose has the potential to produce an apparent muscle-relaxing effect. As a result, the postoperative residual curarization could lead to complications during the recovery process. As the operation ended, we used neostigmine as antagonist, but the patient still failed to recover from the neuromuscular block. For the patient's safety, the tracheal catheter was not removed immediately.

Bariatric surgery has special implications for the patients with KD. Since the patients have the chance of obtaining significant weight loss following the surgery, they are likely to improve the independent walking ability through the reduction of the lower limb load, delaying the time of wheelchair dependence. Since the symptoms of OSAS are mitigated, the odds of respiratory complications are also reduced.

The incidence of KD is extremely low. It is quite hard to perform the evaluation of the safety of anesthesia and operation with the help of a large sample study. The tolerance of the patient to the anesthesia at various stages of KD is also quite different. That is why evaluation of anesthetic risk should be individualized. If the patient's bulbar and respiratory function have been extensively impaired, it is obvious that the surgery is critical, and we require avoiding the performance of operation if we do not have to. Contrarily, if the patient's bulbar and respiratory function is still good, we consider the surgery comparatively safe.

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Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

Ethical Approval The procedure performed in the study involving human participant was in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed Consent Informed consent was obtained from the individual participant included in the study.

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