Dear Editor,

Myotonic dystrophy (MD) is a rare (prevalence rate: 0.003-0.005%) disease inherited genetically as an autosomal dominant pattern. Its onset is most commonly encountered between the second and fourth decades of life. MD affects skeletal, smooth, and cardiac muscles resulting in profound skeletal muscle weakness and degeneration[1]. The disease predisposes patients to cardiac and respiratory problems, malignant hyperthermia (MH) and rhabdomyolysis during anesthesia, making anesthetic management especially important. Cardiopulmonary bypass (CPB) and myocardial protection are complicated by myotonia inducing factors such as hypothermia, medications, potassium, shivering and mechanical or electrical stimulation[2]. We present an anesthetic approach to an MD patient who underwent coronary artery bypass graft operation (CABG).

A 54-year-old male patient with a 6-year history of MD presented with ischemic heart disease and underwent CABG. He was diagnosed with MD-type 1, also known as Steinert disease, after electromyographic studies and muscle biopsy. His myopathy was mild and restricted to minimal weakness of the legs. Preoperatively, his respiratory function tests, serum creatinine kinase (CK), sodium, potassium, urea, and creatinine levels were normal. Dantrolene to be used against MH was made available preoperatively. The anesthetic machine was cleaned with vapor-free fresh gas (10 L/min) for four hours and soda lime was replaced to cleanse anesthetic agents prior to anesthetic induction in order to prevent MH. It was determined that use of propofol and fentanyl citrate together with total intravenous anesthesia and rocuronium bromide-a nondepolarizing muscle relaxant agent-would be a safe method of anesthetic management[3].

The patient was intraoperatively monitored with five-lead EKG, invasive arterial blood pressure, central venous pressure, pulse oximetry, nasopharyngeal temperature, neuromuscular monitoring and transesophageal echocardiography. Regarding neuromuscular monitor, the set point was a TOF ratio G70%. The patient was ventilated with a mixture of oxygen and air (FiO\textsubscript{2} 50%) with a minute volume adequate to maintain a PaCO\textsubscript{2} between 35-40 mmHg. The patient was cooled to 34-36°C on CPB and blood hyperkalemic warm cardioplegia was used for myocardial protection. Hemodynamic parameters were stable throughout the operation. The patient showed no signs of MH such as elevation of the end-tidal CO\textsubscript{2}, cardiac arrhythmia, hyperkalemia, muscle rigidity or metabolic acidosis. The surgical procedure was completed based on routine principles, and the patient was transferred to the postoperative unit afterwards. No cardiac or respiratory complications, muscle contraction,
weakness or shivering were observed during the postoperative period.

There are few reports concerning cardiac operations on patients with MD in the literature \cite{1,2,4}. Sakai et al. reported an atrial septal defect repair in patients with MD with systemic normothermia and off-pump surgery \cite{4}. Gelsominino et al. showed that mild hypothermic (31°C) CPB and hyperkalemic cold cardioplegia could be safely applied in patients with MD requiring mitral valve repair \cite{1}. But the safety of hypothermia and hyperkalemia in MD patients have still not been fully examined and there is a lack of large-scale studies.

In a retrospective study of 219 MD patients who underwent surgery under general anesthesia, Matheiu et al. observed many of known perioperative pulmonary complications \cite{5}.

In summary, an extensive preoperative evaluation, anesthesia maintenance, close monitoring, availability of dantrolene beforehand, and postoperative follow-up of the pulmonary system are of significance in MD patients.

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