Abstract
We described the anesthetic management of a 17-year-old male patient with Fukuyama congenital muscular dystrophy (FCMD) who underwent surgical repair for scoliosis under total intravenous anesthesia. The patient had severe constructive lung disease (%VC 18.6%). Left ventricular wall motion was reduced (left ventricular ejection fraction 40%). Propofol and remifentanil were continuously infused to maintain anesthesia, but we did not use any muscle relaxant throughout the course. We used arterial pressure-based cardiac output and stroke volume variation as a guide for circulatory management. We could not find any congestion on chest X-ray after the surgery. The emergence and recovery from the anesthesia was rapid and muscle strength was enough, and we could extubate the patient just after the end of the surgery. No respiratory and cardiac complications occurred during the postoperative period. Even though he was in the young age in FCMD, respiratory and cardiac complications were severely impaired. For successful anesthetic management in FCMD patient, we should take care of rapid emergence from anesthesia and also we should not impair muscle strength for good postoperative respiratory function. Appropriate hemodynamic monitoring to avoid postoperative cardiac congestion is also required.

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