Dear Editor,

I present total intravenous anesthesia (TIVA) using propofol and remifentanil without neuromuscular blockers (NMBs) was successfully performed in a child with Schwartz Jampel syndrome (SJS).

Bilateral undescended testis surgery was planned for an 18-month-old child weighing 7 kg with SJS. The child had been hospitalized in the intensive care unit due to epilepsy and had been used 200 mg/day carbamazepine. In addition, the child had a seizure again 1 month ago. The child’s Mallampati score was III. There was a possibility of difficult intubation due to abnormal facial defects, the contraction of the jaw muscles, and the limitation of head and neck movements. There was no pathology in the cardiopulmonary system examination and laboratory tests. Due to the risk of developing malignant hyperthermia, necessary preparation (removal of inhalation agents from the anesthesia device, supply of cold serum and drapes, and dantrolene) was provided. Monitoring, including electrocardiogram, pulse oximetry, and body temperature measurement, was applied. The heart rate was 126 beats/min, SpO2 was 99%, and body temperature was 36.8°C. 0.05 mg/kg midazolam and 1 mg/kg ketamine were administered for sedation, and ventilation was provided via the mask. The child was placed in a lateral decubitus position for the caudal block. Following the failed caudal block, TIVA was decided to be applied. Anesthesia was induced with propofol 1 mg/kg and fentanyl 0.5 µg/kg. After then, laryngeal mask airway (LMA) size 1.5 was successfully inserted in the supine position. Maintenance of anesthesia was provided with TIVA using 6–10 mg/kg/h propofol and remifentanil 0.05–0.25 µg/kg/min. During the surgery, the cardiac pulse was 110–140 beats/min, body temperature was 36.6–36.9°C, and EtCO2 level was 32–37 mm Hg. The surgery lasted 115 min, was completed without any problems, and LMA was removed. Paracetamol 10 mg/kg was administered for postoperative analgesia. The child was discharged on the seventh postoperative day.

Schwartz Jampel syndrome is a rare genetic disorder that involves developmental delay, short body structure, phenotypic facial defects, and musculoskeletal system abnormalities.[1] Fatal complications such as malignant hyperthermia, rhabdomyolysis, and hyperkalemic cardiac arrest may be encountered in the perioperative period due to thermoregulatory disorder and the need for mechanical ventilation due to respiratory failure, which is related to existing myotonia and limited chest wall movement, in the postoperative period in children with SJS.[2,3] To avoid the risk of developing these complications, volatile anesthetics and depolarizing NMBs should not be used in general anesthesia.[4,5] There is a possibility of difficult intubation due to abnormal facial defects, including microstomia, micrognathia, and the rigidity of the jaw muscle.[2‑4] The children with SJS may have difficulties due to lordosis, kyphoscoliosis, joint contractures, dysplastic hip, and contracted pelvis while applying regional anesthetic techniques.[1,3] Whether general anesthesia is required, dantrolene should be available in the operating room before the operation, and continuous monitoring, including body temperature and end‑tidal CO2, should be provided during the surgery.[2‑4] In these children, to avoid these dangerous complications, TIVA is recommended,[3,5] and a case of successful and safe administration has been reported.[2]

Volatile anesthetics and depolarizing NMBs were avoided because they have dangerous complications such as malignant hyperthermia, hyperpyrexia, epileptic activity, and hyperkalemia. At firstly, I decided to perform caudal epidural anesthesia. When failed, LMA was inserted, and TIVA was applied using propofol and remifentanil without NMBs. As a result, I suggest that TIVA using propofol and remifentanil can be safely performed in children with SJS.

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Conflicts of interest
There are no conflicts of interest.

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