

Procedural Sedation for Pediatric Patients with Spinal Muscular Atrophy Undergoing Intrathecal Treatment

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Abstract

Aims: Nusinersen is administered intrathecally for treating spinal muscular atrophy (SMA). Procedural sedation is common with intrathecal treatment in children. This retrospective study presents our experience with procedural sedation during the intrathecal treatment of pediatric patients with SMA I, II, and III. **Methods:** Data were collected from the anesthesia charts and electronic medical records of 14 pediatric patients with SMA types I, II, and III who underwent procedural sedation for repeated intrathecal treatments for SMA. Intravenous induction was performed, and patients were oxygenated with a face mask or nasal cannula while spontaneous breathing continued. **Results:** Fourteen patients were included in the study: one SMA I, eight SMA II, and five SMA III. They underwent 88 intrathecal nusinersen injections totally. In the one SMA I patient, of eight months, the procedure was performed under local anesthesia. In all other patients, the treatments were performed under procedural sedation. Different combinations of midazolam, ketamine, propofol, fentanyl, and remifentanyl were used. The mean doses of the agents used were 0.03 mg/kg, 0.97 mg/kg, 2.71 mg/kg, 0.84 mcg/kg and 0.5 mcg/kg respectively. There were no intraoperative or postoperative complications. **Conclusion:** We found the procedural sedation to be sufficient, effective, and safe in SMA II and III pediatric patients who underwent nusinersen treatment intrathecally, provided anesthetic agents are titrated and administered carefully.

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