

Anesthetic approach to patient with Down syndrome and Hallervorden-Spatz disease who underwent tooth extraction

Sir,

Hallervorden-Spatz disease (HSD) is a rare neurological condition, caused by iron accumulation, which causes degeneration of the central nervous system. Blood and cerebrospinal fluid iron levels are normal. Accompanying Down syndrome (DS) is very rare.^[1,2] We present our anesthetic approach with midazolam, metoclopramide, and ranitidine combination for tooth extractions of a mentally retarded, DS patient with HSD. We did not come across the use of this combination for HSD patients during the literature search.

A 16-year-old male with DS, dystonia, dysarthria, gait disturbances, and sudden onset of psychotic symptoms with the diagnosis of HSD referred to Ankara University, Faculty of Dentistry, Oral and Maxillofacial Surgery Department (Turkey, 2015), for tooth extraction. General anesthesia was considered because of cooperation difficulty, agitation, and motion disorders. At preoperative examination, thyromental distance = 5 cm, sternomental distance = 10 cm, mandibulothyroid distance = 3 cm, Mallampati class was IV, atlanto-occipital joint extension was limited, and he had macroglossia. The patient was considered as a difficult intubation case. Oxygen saturation, heart beat rate, and noninvasive blood pressure were tracked. The patient with University of Michigan Sedation Scale (UMSS)-1 was oxygenized and was administered 2 mg of intravenous midazolam and metoclopramide intravenous 10 mg and ranitidine intravenous 50 mg to increase lower esophagus sphincter pressure and neutralize stomach acid. Anesthesia was maintained with 1 mg/kg of intravenous ketamine. Convenient mouth opening was achieved at UMSS 3. Extractions were carried out with no additional anesthetic agents or methods. After anesthesia termination, UMSS 1 and sufficient respiration was achieved in 4 min, general condition was good, and the patient was sent to service.

In conclusion, treatment plans for HSD patients require general anesthesia when difficult intubation criteria and cooperation difficulty are present.^[1,2] Sedoanalgesia under monitored anesthesia care, with titration of short-term anesthetics, and achieving UMSS 3 provides sufficient mouth opening for minor dental surgery procedures. Probable complications are prevented; mortality is decreased, fast recovery and early discharge is provided.

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Conflicts of interest

There are no conflicts of interest.

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