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LETTER TO THE EDITOR

Dexmedetomidine for a patient with Hallervorden–Spatz syndrome during magnetic resonance imaging: a case report

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To the Editor:

A 17-year-old boy of 160 cm height and 30 kg weight, diagnosed with Hallervorden–Spatz syndrome (HSS), also known as pantothenate kinase-associated neurodegeneration (PKAN), presented as severe opisthotonus, retrocollis, deteriorating intelligence level, thoracolumbar spine scoliosis, and tongue dystonia and was scheduled for brain magnetic resonance imaging (MRI) under general anesthesia. On preanesthesia examination, his heart rate was 127 bpm, respiratory rate 16/min, blood pressure 80/45 mm Hg, and oxygen saturation 95 % on room air. A loading dose of 1 μg/kg dexmedetomidine (Dexem; Themis Medicals, India) was infused over 15 min. With this, posturing and rigidity disappeared and the Ramsay sedation score (RSS) reached 5. Dexmedetomidine infusion was continued at a rate of 0.5 μg/kg/h using an MRI-compatible infusion pump (Perfusor; Space B/Braun, Germany) during MR imaging that lasted 1 h. Oxygen was administered through a face mask. He was hemodynamically stable throughout the procedure. An RSS of 2 was reached and opisthotonus posturing and rigidity reappeared 2 h after stopping the infusion.

This case report highlights successful use of dexmedetomidine as a sole sedative agent for MRI in a patient with HSS. HSS is a rare autosomal recessive disorder caused by pantothenate kinase-associated neurodegeneration 2 (PKAN2) gene mutations resulting in accumulation of iron in the brain. The disease is also called neurodegeneration with brain iron accumulation (NBIA) [1]. Presenting features of HSS include progressive generalized dystonia involving the oromandibular muscles, rigidity, and choreoathetoid movement [1]. Airway management is critical during anesthesia in these patients. Oromandibular dystonia results in impaired swallowing, malnourishment, and high risk for pulmonary aspiration. Involvement of the pharyngeal muscles results in breathing difficulty and dynamic airway obstruction. These patients develop contractures and joint stiffness, including the jaw and cervical spine, leading to difficult airway. Associated mental retardation renders preoperative airway assessment difficult. Airway problems necessitate careful postoperative monitoring. MRI studies demand a quiet patient for better image quality. Severe dystonia necessitated sedation/general anesthesia in our patient. In a previous case report, volatile anesthetic was used to sedate a child with HSS, but postoperatively the patient developed airway obstruction requiring emergency intubation [2]. In our patient, we chose dexmedetomidine to provide sedation without respiratory depression [3]. As dystonia completely disappeared and an RSS of 5 was achieved after a bolus infusion of dexmedetomidine, we continued dexmedetomidine infusion during the MR imaging. Dexmedetomidine, by alpha-2 adrenergic modulation, induces prolonged non-REM sleep. As muscle atonia has been described during non-REM sleep, patients receiving dexmedetomidine infusion tend to be in a relaxed state with reduced muscle tone [4]. This mechanism could have suppressed the dystonia in our patient. Although propofol has been used in such patients, we avoided propofol as it causes dystonic reactions and opisthotonus [5]. Dystonia suppression by

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dexmedetomidine, although described in animal studies, has not been described in humans. Upon awakening, the patient's dystonia reappeared, which again confirms reversible suppression of dystonia by dexmedetomidine. In conclusion, dexmedetomidine can be a viable option to sedate dystonic children for MRI studies, without comprising airway and hemodynamics.

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