

<b>Title</b>	Deliberate Awareness Under Anesthesia for Neurosurgery in a Child with Hallervorden-Spatz Disease
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**INTRODUCTION:** Awareness under anesthesia is a complication recognized by anesthesiologists and feared by patients. Patients with recall describe the experience as frightening, and many go on to suffer post-traumatic stress disorder. We describe a case of a child scheduled for a neurosurgical procedure that required muscle relaxation, but could not be performed effectively with administration of amnestic agents. Consequently, the majority of the anesthetic consisted of using only rocuronium and remifentanyl.

**CASE REPORT:** An 11-year-old 16 kg girl with Hallervorden-Spatz disease and severe gastro-esophageal reflux presented for intraneural mapping and left pallidotomy. Rapid sequence induction was performed with 30 mg of propofol and 10 mg of rocuronium, and a 6.0 endotracheal tube was placed. A remifentanyl/ propofol infusion was started at  $100 \text{ mcg} \times \text{kg}^{-1} \times \text{min}^{-1}$  for frame placement and MRI procedure. In the operating room, remifentanyl, nitrous, and rocuronium were used until the start of the mapping. During the five hours of mapping, remifentanyl and rocuronium were continued without nitrous. The patient required gradually increasing doses of remifentanyl during the final hour, from 0.05 to  $0.12 \text{ mcg} \times \text{kg}^{-1} \times \text{min}^{-1}$ . The rocuronium was administered as an infusion at  $7 \text{ mcg} \times \text{kg}^{-1} \times \text{min}^{-1}$  to keep 3 out of 4 twitches on train of four testing. Upon completion of the pallidotomy lesion, a standard general anesthetic was instituted with sevoflurane and nitrous. At the conclusion of the procedure, the patient was awakened and extubated in the operating room.

**DISCUSSION:** Several hundred adult patients have been treated at our institution with either deep brain stimulation or pallidotomy procedures to decrease extraneous movements. For these procedures, local anesthetic and minimal sedation is used for frame placement and MRI. Minimal narcotics are given during the neurosurgical mapping procedure; generally, only remifentanyl is used as needed between electrode passes to alleviate any discomfort. Reassurance from the anesthesiologist aids in alleviating anxiety during the procedure. This anesthetic plan works well for aware adults being treated for Parkinson's disease, essential tremor, or other movement disorders. However, the patient in our case has Hallervorden-Spatz disease, a rare autosomal recessive disorder involving abnormal iron deposition in the globus pallidus, resulting in spasticity, dystonia and severe developmental delay. In this instance, the procedure aims to decrease spasticity in hopes of allowing the child to interact with her environment as well as be cared for at home, rather than require institutionalized care. For the anesthesiologist, there are seemingly conflicting anesthetic goals in this case. Since the child has severe reflux and a history of frequent aspirations, endotracheal intubation is indicated for airway protection. Because of the stereotactic frame, spastic and dystonic movements must be prevented with either deep anesthesia or muscle weakness in order to prevent cervical spine injury. However, benzodiazepines, nitrous oxide, volatile agents, and intravenous induction agents all preclude the ability to perform accurate biometrical mapping. Unfortunately, since the child is non-verbal and uncommunicative, verbal reassurances are unlikely to be effective in her case. Consequently, a balance between patient safety and achieving surgical goals must be sought. Communication with the surgeon as well as the child's family about the anesthetic plan and its implication is essential. The surgeon must be able to map as effectively and accurately as possible, and the family must agree that in providing optimal surgical condition, there is a high risk for intra-operative awareness. Through communication and careful planning, the appropriate balance can be attained, allowing success even in such a challenging situation.