Letter to the editor
The use of bispectral index monitoring in the anesthetic management of a patient with Rett syndrome undergoing scoliosis surgery

To the Editor:

Rett syndrome is a genetically inherited disorder that almost exclusively affects females. It is characterized by severe mental retardation after a period of apparent normal development for the first 6 to 18 months of life [1]. Other characteristics of the disorder are autistic behavior, myoclonic movements, seizures, apneic spells, aerophagia, tachybradyarrhythmias, serotonin imbalance, scoliosis, bruxism (teeth grinding), and autonomic nervous system dysfunction [2].

Prevalence studies show that it is not a rare phenomenon among the population of severely retarded females, and that the prevalence may be as high as 25% [3]. Prevalence rates range from 1:10 000 in Sweden to 1:25 000 in Japan, rendering Rett syndrome as the leading cause of profound cognitive impairment in females [2]. In general, survival of these patients is quite prolonged; 75% of patients survive to age 35 years compared with 27% for other causes of profound mental retardation. In young patients, deaths occur primarily during sleep, are unrelated to seizures, and largely remain unexplained [4]. Cardiac conduction abnormalities are a possible factor in these deaths; longer corrected QT intervals and T-wave abnormalities [5], as well as severe sinus bradycardia [6], have been described.

With increased life expectancy of patients with Rett syndrome, it has become more common to encounter these patients in the operating room (OR). These patients have been reported to be unduly sensitive to sedative drugs, and they exhibit slow recovery from anesthesia [7,8]. We describe and discuss the anesthetic management of a 12-year-old girl with Rett syndrome, undergoing spinal fusion, who could not be awakened after surgery despite strict titration of anesthetic agents to the range of bispectral index (BIS) numbers that were previously reported to lead to successful awakening after anesthesia.

1. Case report

A 12-year-old, 43-kg girl with confirmed Rett syndrome was scheduled for a T3-L4 posterior spinal fusion. Her history included scoliosis, poorly controlled seizure disorder, episodes of irregular breathing, and reflux. She had a history of slow awakening after anesthesia for dental work performed three years earlier. Her medications included topiramate, levetiracetam, lamotrigine, and buspirone. On physical examination, the patient was noted to be cognitively impaired, with purposeless movement of the upper extremities and atrophic lower extremities. Preoperative workup was significant for prolonged QT intervals on electrocardiography.

One hour before surgery, the patient’s father reported that she had a seizure. No premedication was administered. In the OR, routine monitors and BIS monitors were placed. Anesthesia was induced via the inhalational route, with sevoflurane. Once the BIS reading reached 40, an 18-gauge intravenous catheter was placed and the sevoflurane was discontinued. For somatosensory evoked potential monitoring, the patient was then given 50 mg of propofol, 100 μg of fentanyl, and 60 mg of rocuronium. Her trachea was then intubated. Because the father reported preoperative seizures, the surgeon requested that 750 mg of phenytoin and 65 mg of phenobarbital be administered during the first three hours of the procedure. Anesthesia was maintained with propofol between 100 and 125 μg/kg per minute and remifentanil between 0.1 and 0.15 μg/kg per minute. Both drugs were titrated to maintain BIS values between 30 and 50. After 6.5 hours of anesthesia, the case ended and the patient was transported to the pediatric intensive care unit registering a BIS of 60. The anesthetic agents and the BIS monitoring were discontinued in the OR before transport. Despite being hemodynamically stable, normothermic, normoglycemic, and normocapnic, the patient was slow to awaken. Four hours after arrival at the intensive care unit, the patient’s trachea was successfully extubated. No electroencephalograms or further workup was done to evaluate the delayed awakening.

2. Discussion

This case report illustrates the difficulty of proper anesthetic management of a patient with Rett syndrome who underwent surgery to correct scoliosis. Although these patients are known to be possibly excessively sensitive to both sedative drugs and volatile anesthetics [7,9], one recent
case report described normal awakening after premedication and anesthesia [10]. Our patient had a history of slow awakening when chloral hydrate was given for her prior dental procedure, and we thus decided to abstain from giving her any preoperative medication. The patient was anesthetized with relatively low maintenance doses of propofol and remifentanil. However, although BIS readings were maintained similar to those noted in a previous case report [9], our patient did not awaken in timely fashion at the end of the surgical procedure. Our patient received phenobarbital and phenytoin intraoperatively, her surgery lasted 6 hours, and her father reported seizure activity before the procedure. Furthermore, her postoperative stay was characterized by multiple seizurelike episodes. Future studies are required to determine the range at which BIS should be maintained for appropriate hypnotic depth in patients with Rett syndrome.

Although BIS monitoring was used in this case, the BIS values tended to remain at the lower end of the recommended range (30-45). The BIS response to total intravenous anesthesia with propofol can often exhibit a plateau of about 40 and may not go significantly lower until a burst-suppression pattern appears on the electroencephalogram. This situation can allow a significant excess to saturate tissues of the body, rendering rapid emergence more difficult. The BIS should be allowed to increase slowly toward a value of 50 to prevent excess tissue accumulation during a long case.

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References