



## Case Report

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# General Anesthesia for Dental Treatment in a Patient with Subcortical Band Heterotopia: A Case Report

Nobuhito Kamekura\*, Yukie Nitta, Shigeru Takuma and Toshiaki Fujisawa

Department of Dental Anesthesiology, Faculty of Dental Medicine and Graduate School of Dental Medicine, Hokkaido University, Japan



## Abstract

Subcortical band heterotopia (SBH) or double cortex is a classic malformation associated with deficient neuronal migration. The syndrome is characterized by the presence of symmetrical and bilateral bands of heterotopic grey matter located between the ventricular wall and the cortical mantle, and clearly separated from both. Affected individuals typically present with epilepsy and mental retardation. Dysarthria, hypotonia, pyramidal syndrome, gastroesophageal reflux, aspiration pneumonia, high arched palate and small jaw may be present.

We provided safe anesthesia management to patients with SBH. In such patients, seizure disorders and respiratory complications should be considered. Appropriate evaluation of these associated conditions during the preoperative examination, appropriate intraoperative monitoring, and preparation of countermeasures for epilepsy should allow the safe provision of anesthesia to patients with SBH.

## Keywords

Subcortical band heterotopia, General anesthesia, Intractable epilepsy, Mental retardation

## Introduction

Subcortical band heterotopia (SBH) or double cortex is a classic malformation associated with deficient neuronal migration. The syndrome is characterized by the presence of symmetrical and bilateral bands of heterotopic grey matter located between the ventricular wall and the cortical mantle, and clearly separated from both. It is usually associated with mutations in the double cortin gene [1].

Affected individuals typically present with epilepsy and mental retardation. Dysarthria, hypotonia, pyramidal syndrome, gastroesophageal reflux, aspiration pneumonia, high arched palate and small jaw may be present [2,3].

The prevalence of SBH at birth is estimated to range from 11.7 to 40 per million births [1]. Because of its low incidence, there are few reports on the anesthetic management of patients with SBH [4]. We herein report the conduct of general anesthesia in a patient with SBH.

## Case Report

A 13-year-old girl required general anesthesia for dental treatment due to mental retardation and poorly controlled seizure disorder. Epilepsy and mental retardation were present from early infancy. She was treated with antiepileptic medications at the pediatrics department of a general hospital; however, her epilepsy was intractable. Head MRI

was conducted for the evaluation of epilepsy and SBH was diagnosed based on the observation of a band of heterotopic gray matter under the subcortical white matter.

She was a student at a school for disabled children. She could not express meaningful words and required total assistance for bathing, dressing, urination and defecation. She could not stand or walk without support due to ataxia of her extremities and poor trunk control.

The patient had no swallowing problems or history of aspiration pneumonia. High arched palate and micrognathia were not noted.

She had a tonic seizures and absence seizures. Her tonic seizures with head nodding occurred several times a day. Both arms were raised over her head during seizures. If standing

**\*Corresponding author:** Nobuhito Kamekura, Department of Dental Anesthesiology, Faculty of Dental Medicine and Graduate School of Dental Medicine, Hokkaido University, Nishi 7, Kita 13, Kita-ku, Sapporo, Japan, Tel: +81-11-706-4336

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during an attack, she fell. Absence seizures occurred about ten times a day. Lamotrigine and valproate were prescribed; however, the control of her seizures was insufficient.

Oral hygiene instruction had been continued in the Department of Dentistry for Children and Disabled Persons of Hokkaido University Hospital. However, the treatment of dental caries under general anesthesia was required.

The results of a preoperative examination were as follows: Height, 145 cm; body weight, 28 kg; blood pressure, 107/53 mmHg; heart rate, 106 bpm; SpO<sub>2</sub>, 97%; and body temperature, 36.9 °C. Blood tests, electrocardiography and chest roentgenography revealed no remarkable findings. She did not take any drugs except antiepileptic drugs regularly. It was judged that general anesthesia was possible.

At the beginning of anesthesia, venipuncture was performed in the recovery room, not in the operating room, and 3 mg of midazolam was administered intravenously because her sense of fear was strong. She was sedated and entered the operating room without any expression of fear. In addition to standard electrocardiogram, pulse oximetry, non-invasive blood pressure, and end-tidal CO<sub>2</sub>, bispectral index (BIS) and train of four (TOF) monitoring were performed. General anesthesia was induced with thiamylal (100 mg) and sevoflurane (2.9 MAC) in air and oxygen. After the administration of rocuronium, nasal tracheal intubation was performed with no difficulty. Anesthesia was maintained with sevoflurane (0.9-0.6 MAC) in air and oxygen, and continuous intravenous remifentanyl (0.1 µg/kg/min). A BIS level of approximately 20-40 was maintained during operation. The TOF ratio decreased to zero soon after the administration of rocuronium and it became 75% approximately 30 minutes later. Before extubation, the TOF was confirmed to be 100 and sugammadex was administered.

Anesthesia lasted 1 hour 34 minutes and was completed uneventfully. There were no significant changes in hemodynamics and the patient did not experience delayed emergence from anesthesia.

Because of postoperative nausea, she was not able to take antiepileptic drugs orally. Diazepam was administered rectally, and there was no change in her seizure attacks. On the following day, she became able to eat orally and left the hospital.

## Discussion

The first symptoms of patients with SBH are neurologic deficits consisting of poor feeding, mild hypotonia, and abnormal arching behavior or opisthotonus in infancy. Thereafter, delayed motor milestones are recognized and seizures occur in most cases. In all affected children, the major medical problems encountered are feeding problems and gastroesophageal reflux, epilepsy of many different types, and recurrent aspiration and pneumonia due to feeding problems and epilepsy [5]. Cognitive performance ranges from normal to learning disability and/or severe intellectual disability [6]. High arched, small jaw and protruding tongue may be present [2].

Our patient had lower extremity-predominant movement disorder in addition to epilepsy and mental retardation. However, we considered that the likelihood of respiratory complications was low because she did not have symptoms such as gastroesophageal reflux, aspiration pneumonia, high arched palate, or small jaw.

In this case, consideration of epilepsy was most important when administering general anesthesia. It is necessary to avoid factors causing epilepsy.

Perioperative discontinuation of oral antiepileptic drugs may be a factor that predisposes a patient to seizures. Perioperative starvation periods should be kept to a minimum and routine medications should be given preoperatively and postoperatively in order to minimize missed doses of oral antiepileptic drugs. If oral intake is impossible, the intravenous or rectal administration of the drugs should be considered [7]. The patient took the routine antiepileptic drugs in the morning before surgery. However, she was not able to take the drugs at night after surgery because of postoperative nausea and vomiting, and the intrarectal administration of benzodiazepine was performed. During hospitalization, there was no change in her seizure patterns.

General anesthetics have both proconvulsant and anticonvulsant properties [8]. Attention to the occurrence of seizures is necessary for any general anesthetic agent. In this case, we administered midazolam, thiamylal and sevoflurane. Among these drugs, particular care is necessary during the use of sevoflurane, especially when used in high concentrations, due to reports that deep anesthesia with sevoflurane is a risk factor for excitation of the central nervous system. Furthermore, hypocapnia is associated with electroencephalogram (EEG) changes [9,10]. When administering sevoflurane, it may be necessary to pay attention to hyperventilation and high concentrations.

Drug interactions between oral antiepileptic drugs and anesthetic agents should be considered. Her regular drugs were lamotrigine and valproate. Lamotrigine does not induce hepatic enzymes and has fewer drug interactions [7]. Valproate is mainly metabolized by CYP2B6 and CYP2C9. These enzymes contribute to the metabolism of propofol. If total intravenous anesthesia is performed, the required dose of propofol might be lower [11].

In addition to standard electrocardiography, pulse oximetry, non-invasive blood pressure, and end-tidal CO<sub>2</sub>, TOF monitoring and BIS monitoring were performed in this case. Her reaction to rocuronium, a non-depolarizing muscle relaxant, was normal. As patients with SBH may have neurological symptoms, such as dysarthria and hypotonia, the TOF monitoring is desirable [12].

The patients BIS values ranged from approximately 20 to 40 during the operation in this case. Taking the inspired concentration of sevoflurane and the remifentanyl infusion rate into consideration, this range of BIS values might be lower than expected. Many clinical conditions influence BIS values [13]; da Costa, et al. reported that the chronic use of anticonvulsants associated with oral midazolam as pre-

anesthetic medication could lead to a decrease in BIS values [14].

In this case, the habitual use of anticonvulsants and preoperative administration of midazolam could affect BIS values. In addition, the underlying cerebral pathology could also affect BIS values. Valkenburg, et al. reported that the BIS values were low in patients with lissencephaly and Miller Dieker syndrome, which are classified as the same migration disorder as SBH [4]. They also reported that BIS values were low in intellectually disabled patients [15]. In general anesthesia of a patient with neuronal migration disorder, BIS values should be interpreted carefully. It is conceivable that BIS values should not be used as an absolute value, and rather as a reference of the relative change in the depth of anesthesia.

## Conclusion

We provided safe anesthesia management to a patient with SBH. In such patients, seizure disorders and respiratory complications should be considered. The appropriate evaluation of these associated conditions during the preoperative examination, appropriate intraoperative monitoring, and the preparation of countermeasures for epilepsy should allow the safe provision of anesthesia to patients with SBH.

## Conflicts of Interest

The authors declare no conflicts of interest in association with the present study.

## Declarations

1. All authors of this paper have contributed to the study.
2. All authors have read and approved the version submitted to the Journal.
3. The content of the manuscript is original and have not been published earlier.

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