

Bispectral index monitoring in a patient with combination of congenital insensitivity to pain with anhidrosis (CIPA) and Shwachman–Diamond syndrome

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Received: 29 May 2013 / Accepted: 20 June 2013 / Published online: 5 July 2013
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Keywords Congenital insensitivity to pain with anhidrosis · Shwachman–Diamond syndrome · Pediatric anesthesia · Total intravenous anesthesia · Bispectral index monitoring

To the Editor:

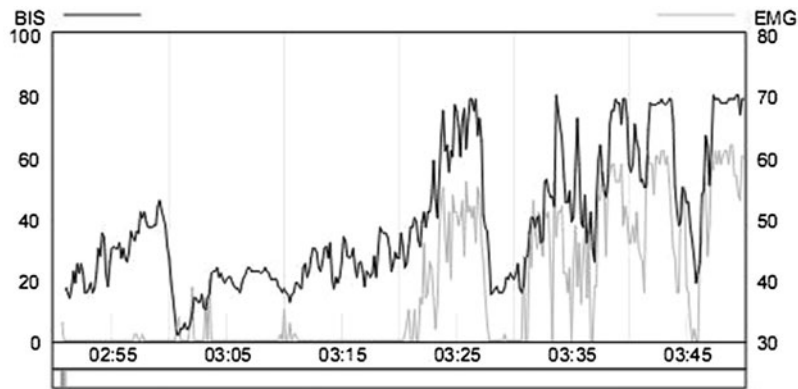
Congenital insensitivity to pain with anhidrosis (CIPA) is an autosomal recessive disorder that is characterized by lack of pain sensation, mental retardation, and changes in temperature control [1]. Shwachman–Diamond syndrome (SDS) is a rare autosomal recessive disorder. It is characterized by exocrine pancreatic insufficiency, bone marrow dysfunction, leukemia predisposition, and skeletal abnormalities [2]. The association of these two syndromes is very rare. We present a total intravenous anesthesia (TIVA) practice guided by bispectral index monitoring (BIS) of a patient with the combination of CIPA and SDS.

A 5-year-old male patient was admitted to hospital following fracture of his tibial bone. He was diagnosed with CIPA when he was 7 months old after a scratched cornea occurred because of corneal hypoesthesia and insensitivity. He had also a variety of injuries, such as produced by beating his tongue, and a sweating disorder. SDS was diagnosed after detection of an exocrine pancreatic insufficiency. After a 5-h preoperative fast, the patient was taken to the operating room without any sedation agent. Because of thermoregulatory dysfunctions in patients with CIPA, the temperature of the operating

room was adjusted to 22 °C. To monitor electrocardiography, noninvasive blood pressure, oxygen saturation, capnography, temperature monitoring, and BIS were applied. The anesthesia induction drugs were propofol 2–3 mg/kg and vecuronium 0.1 mg/kg. After tracheal intubation with a 5-mm endotracheal tube, the ventilator was adjusted to tidal volume 6–8 ml/kg, respiratory rate 20/min, and fresh gas flow 6 l/min. TIVA with propofol 50–150 µg/kg/min was applied to keep the value of BIS between 40 and 60 without any analgesic agent (Fig. 1). Tracheal extubation was performed with neostigmine 50 µg/kg and atropine 0.01 mg/kg. During the operation the patient did not have any sensitivity to pain, although he was reacting to temperature changes. While he was in contact with a warm sponge, BIS increased to 60 and higher. However, no reaction was detected to cold contact. Therefore, any hot contact was avoided. After extubation the patient was followed up for about 30 min at the postanesthesia care unit without any problems.

So far, reported cases about anesthesia management in patients with CIPA give a perspective about inhaled anesthesia [1]. In one of these, BIS was successfully used during an inhalation anesthesia for a CIPA patient [3]. BIS monitoring is an effective way for providing general anesthesia to the patients with combination of CIPA and SDS. In this patient we have successfully used BIS during TIVA with propofol. Coadministration of these agents provides many advantages such as prevention of excessive drug implementation and prevention of recovery delay. Another issue with the combination of CIPA and SDS is the increased aspiration risk during general anesthesia practices [2, 4]. Therefore, it is necessary to secure the airway with an endotracheal tube from the beginning of anesthesia induction. Consequently, to provide a balanced TIVA, BIS can assist with other monitoring techniques,

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Time	BIS	SQI	EMG	Time	BIS	SQI	EMG
02:51	16	59	27	03:21	37	79	29
02:52	22	85	29	03:22	42	96	46
02:53	19	80	30	03:23	46	95	32
02:54	35	90	29	03:24	64	97	43
02:55	31	90	28	03:25	70	97	49
02:56	27	87	29	03:26	79	92	52
02:57	36	95	31	03:27	65	92	46
02:58	37	100	29	03:28	16	80	28
02:59	46	95	29	03:29	17	67	31
03:00	19	95	24	03:30	22	69	28
03:01	4	83	25	03:31	28	74	20
03:02	14	79	25	03:32	41	75	51
03:03	10	90	37	03:33	47	87	42
03:04	24	86	26	03:34	66	94	52
03:05	21	100	25	03:35	40	95	43
03:06	16	100	27	03:36	48	84	49
03:07	23	100	27	03:37	46	97	39
03:08	23	100	27	03:38	52	95	57
03:09	20	100	26	03:39	78	100	56
03:10	16	94	30	03:40	55	97	46
03:11	19	67	28	03:41	53	100	43
03:12	23	80	27	03:42	77	74	54
03:13	23	71	29	03:43	77	90	59
03:14	20	88	28	03:44	43	95	38
03:15	34	81	28	03:45	45	79	39
03:16	24	92	29	03:46	22	85	29
03:17	23	100	28	03:47	51	86	49
03:18	29	100	29	03:48	78	95	59
03:19	30	95	28	03:49	79	95	62
03:20	35	100	29				

Fig. 1 Trend graph of bispectral index (BIS) shows changes in the depth of anesthesia under total intravenous anesthesia (TIVA) with propofol

especially in pediatric patients with a combination of CIPA and SDS.

References

1. Varshney M, Girdhar KK, Taneja S, Agarwal M, Varshney PG. Anesthetic management of a pediatric patient with congenital insensitivity to pain. *Paediatr Anaesth*. 2009;19:552–3.
2. Tamhane P, Newton NI, White S. Anaesthetic management of quinsy in a patient with Shwachman–Diamond syndrome. *Anaesthesia*. 2003;58:821.
3. Brandes IF, Stuth EA. Use of BIS monitor in a child with congenital insensitivity to pain with anhidrosis. *Paediatr Anaesth*. 2006;16:466–70.
4. Zlotnik A, Gruenbaum SE, Rozet I, Zhumadilov A, Shapira Y. Risk of aspiration during anesthesia in patients with congenital insensitivity to pain with anhidrosis: case reports and review of the literature. *J Anesth*. 2010;24:778–82.