Bispectral Index Guided Sedation in Congenital Pain Insensitivity Syndrome

Alper Kılıçaslan, Funda Gök, Eray Yaşar, Ali Başdemirci, Şeref Otelcioğlu

Department of Anaesthesiology and Reanimation, Necmettin Erbakan University Meram Faculty of Medicine, Konya, Turkey

Dear Editor,

Congenital pain insensitivity syndrome (CPIS) is a rare syndrome, characterized by insensitivity to pain, anhidrosis, heat intolerance and different degrees of mental retardation; and due to high morbidity, the cases frequently lose their lives in adolescence (1). Painless fractures, neurogenic arthropathy (Charcot joint) and joint deformities are frequently seen (2). Although these patients are insensitive to pain, general anaesthesia or sedation should be performed during surgical procedures as irritating conditions like tactile hyperalgesia may develop (3). In this paper, we aimed to present the perioperative management of a case with CPIS, who underwent lower extremity surgery under adequate sedation with bispectral index (BIS) monitoring, along with an overview of the relevant literature.

A 10 year old patient weighing 36 kg, who was followed-up by the Department of Paediatrics because of congenital pain insensitivity syndrome, was planned to undergo debridement and abscess drainage by the Department of Orthopaedics due to osteomyelitis of the left knee and septic arthritis. It was learned that the patient was diagnosed with CPIS in his infancy, on noticing that he experienced no pain when he bit his tongue. Preoperative assessment revealed mental retardation, and insensitivity to pain and heat; his sense of touch and other neurological examinations were normal. There were wounds on the hand and fingers of the patient (Figure 1). On airway assessment, Mallampati score (mouth opening) was II, laboratory values and chest X-ray of the patient was normal. In the preoperative period, written informed consent was obtained from the patients' parents after they were informed about the anaesthetic method that would be used. The patient who was taken to the operating room, underwent standard monitoring (pulse oximetry, electrocardiography, non-invasive arterial blood pressure, temperature) and BIS monitoring (BIS VISTA Monitoring System, USA).

As the patient was responsive to tactile stimuli and manipulations, we decided to begin the intervention with sedation and according to the response of the patient, if needed, to switch to general anaesthesia. After administering an IV bolus dose of midazolam (0.05 mg kg^{-1}) and propofol (1 mg kg^{-1}), propofol infusion ($30\text{-}50 \text{ mcg kg}^{-1} \text{ min}^{-1}$) was initiated. While oxygen was delivered by face mask (4 litre min^{-1}), the patient was left on spontaneous breathing and ETCO_2 was monitored with a line connected to the mask. In case of bradypnea, breathing was stimulated by basic airway manoeuvres such as moving the jaw upwards and front or head tilt and ETCO_2 was maintained under 45 mmHg . The temperature of the operating room was maintained at $22^\circ\text{-}24^\circ\text{C}$, and warming blankets were made available in case they were needed. During the surgery, which lasted for approximately 50 minutes, general anaesthesia, positive pressure ventilation or additional airway devices were not required, and the axillary body temperature (-37°C) remained stable. At the end of the surgery, propofol infusion was discontinued and the patient was transferred to the post-anaesthesia care unit. The patient experienced no complications such as desaturation and apnoea during the surgery and in the postoperative period. The haemodynamic parameters and BIS values of the patient, which are within normal ranges, are presented in Figure 2. The patient did not require analgesics in the postoperative period.

The anaesthetist may face some problems in cases with congenital pain insensitivity syndrome, as there is autonomic and nociceptive dysfunction. The importance of anaesthetic approach that should be used in these patients and perioperative temperature control are emphasized in the literature. Although cardiovascular reflexes are preserved, there may be decreases in norepinephrine and epinephrine levels and severe hypotension may develop in the perioperative period (2). As body temper-

Figure 1. The appearance of wounds on the hand and fingers of the patient with congenital pain insensitivity syndrome, made by the patient himself

Figure 2. Perioperative changes in BIS, HR and MAP values
BIS: bispectral index; HR: heart rate; MAP: mean arterial pressure

ature regulation is impaired in patients with anhidrosis, there may be episodes of hyperthermia, and therefore, 20% of the patients die in the first three years of life due to hyperthermia (3). For the control of body temperature, temperature monitoring, maintenance of operating room temperature, cooling or warming blankets may be used.

It should be kept in mind that besides pain insensitivity, these cases may have tactile hyperesthesia. Tomioka et al in their study, in which they reviewed the anaesthetic management of 45 patients with CPIS, reported that they used opioids in 6 patients with tactile hyperesthesia (4). In the literature, while general anaesthesia (3, 4) is preferred in most of the cases undergoing extremity surgery, there are cases that underwent sedation (5). Although general anaesthesia is more comfortable and keeps the patient motionless, it may lead to severe

hypotension. Mental state of the patient, and the localization and size of the surgical site are the other factors used to determine the anaesthetic method.

Bispectral index is an important monitoring tool, used to quantify the sedative and hypnotic effects of anaesthetic drugs. BIS is useful in determining the dose and timing of anaesthetic agents by showing the depth of anaesthesia (3). In this present case, without any requirement for analgesics, depth of anaesthesia was provided with sedative agents under BIS monitoring.

Conclusively, we suggest that, as tactile hyperesthesia may develop in cases with congenital pain insensitivity syndrome, extremity surgery can be performed under adequate sedation without need for general anaesthesia, by objectively monitoring individual awareness levels using BIS monitoring.

Informed Consent: Written informed consent was obtained from patients' parents who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - A.K.; Design - A.K., F.G.; Supervision - Ş.O.; Funding - A.K., E.Y., A.B.; Materials - E.Y., A.B.; Data Collection and/or Processing - A.K., E.Y., A.B.; Analysis and/or Interpretation - A.K., F.G., E.Y., A.B., Ş.O.; Literature Review - A.K., F.G.; Writer - A.K., E.Y.; Critical Review - F.G., Ş.O.; Other - F.G., E.Y., A.B.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

References

1. Rozentsveig V, Katz A, Weksler N. The anaesthetic management of patients with congenital insensitivity to pain with anhidrosis. *Paediatr Anaesth* 2004; 14: 344-8. [\[CrossRef\]](#)
2. Okuda K, Arai T, Miwa T, Hiroki K. Anesthetic management of children with congenital insensitivity to pain with anhidrosis. *Pediatr Anaesth* 2000; 10: 545-8. [\[CrossRef\]](#)
3. Oliveira CR, Paris VC, Pereira RA, Lara FS. Anesthesia in a patient with congenital insensitivity to pain and anhidrosis. *Rev Bras Anesthesiol* 2009; 59: 602-9. [\[CrossRef\]](#)
4. Tomioka T, Awaya Y, Nihei K, Sekiyama H, Sawamura S, Hanaoka K. Anesthesia for patients with congenital insensitivity to pain and anhidrosis: a questionnaire study in Japan. *Anesth Analg* 2002; 94: 271-4. [\[CrossRef\]](#)
5. Layman PR. Anaesthesia for congenital analgesia. A case report. *Anaesthesia* 1986; 41: 395-7. [\[CrossRef\]](#)