

Anesthetic management of children with congenital insensitivity to pain with anhidrosis

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INTRODUCTION

Congenital analgesia, also known as hereditary sensory and autonomic neuropathy, is mainly caused by ectodermal dysplasia, which is a rare autosomal recessive hereditary disease.^{1,2} Congenital analgesia is divided into two categories of insensitivity to noxious stimuli and unrecognized noxious stimuli. The former category is consistent with congenital insensitivity to pain with anhidrosis (CIPA), while the latter category has noxious stimulation signals, but they cannot be identified.

CIPA belongs to type IV of hereditary sensory

ABSTRACT

Introduction: Congenital analgesia is a rare autosomal recessive hereditary disease. The primary damage of congenital analgesia is central structure damage of comprehensive pain perception.

Case presentation: A 1-year-old Han Chinese boy was admitted to hospital because of a tongue bite. He had no response to noxious stimulation of the body surface and was diagnosed with congenital analgesia. A small dose of remifentanyl was intravenously injected during anesthetic induction to reduce the stress response caused by endotracheal intubation. A certain depth of anesthesia should be guaranteed during anesthetic induction and surgery to alleviate the stress response induced by endotracheal intubation and the operation.

Conclusion: Opioid analgesics are not required for general anesthesia in patient with congenital insensitivity to pain. With a heat dissipation barrier in patients with congenital insensitivity to pain with anhidrosis, body temperature, end-tidal carbon dioxide and bispectral index should be monitored.

KEYWORDS

Anesthetic, Children, Congenital insensitivity, Pain, Anhidrosis

and autonomic neuropathy (HSAN). The clinical manifestations of CIPA are dull or absent pain, no sweating, high fever, abnormal muscle tone or mental retardation, repeated tongue bites, multiple fractures, infections, and osteomyelitis. Reports on anesthesia in such patients are rare.³ We report here anesthetic management of a child with CIPA.

CASE REPORT

A 1-year-old Han Chinese boy (weight: 10 kg) was admitted to hospital because of a tongue bite. The child had a full-term delivery and normal development. His

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body temperature was usually 38°C to 40°C. Seven months after birth, habitual tongue biting begun to occur without crying, and his tongue had been ulcerated for a long time. The child had no response to noxious stimulation of the body surface and was diagnosed with congenital analgesia. Preoperative diagnosis was confirmed. We planned to perform debridement and suturing of the tongue trauma under general anesthesia for the patient. The preoperative blood routine test, liver and kidney function, electrocardiogram, chest X-ray, and coagulation function were normal. The patient was admitted with intravenous access to the Operating Room. His heart rate was 119 beats/min, noninvasive blood pressure was 92/51 mm Hg, and pulse oxygen saturation (SpO₂) was 98%.

Anesthetic induction

Cisatracurium 1 mg, remifentanyl 10 µg, and propofol 25 mg were intravenously injected. An enhanced 4.0 endotracheal tube was nasally inserted and mechanical ventilation was performed with the pressure control mode. The preset inspiratory peak pressure was 12 cm H₂O, tidal volume was 98 ml, the respiratory rate was 25 times/min, inspiratory-respiratory ratio was 1.0:1.5, end-tidal carbon dioxide (PETCO₂) was between 35–40 and bispectral index (BIS) was between 40–50.

Anesthetic maintenance

The patient inhaled 1.5% to 2.0% sevoflurane with no narcotic analgesic drugs. The duration of the operation was 30 minutes and the vital signs were stable during the operation. The intraoperative temperature was 36.8°C to 37.0°C, the PETCO₂ and BIS were normal. At the end of the operation, the sevoflurane was stopped and the boy was delivered to the postanesthesia care unit. After approximately 10 minutes, the child woke up, with a tidal volume of 100 ml, a respiratory rate of 26 times/min, and SpO₂ of 100%. The tracheal tube was then removed. To prevent the child from biting the tongue again, propofol 10 mg was intravenously administered and the tongue was properly fixed. The patient was then safely returned to the ward without any analgesia. No obvious discomfort, such as pain, occurred by 3 days of follow-up.

DISCUSSION

Pain, the temperature perception test, and the iodine starch method for sweating are used for diagnosing CIPA. The current view is that CIPA is caused by mutations in the tyrosine receptor kinase 1 (*NTRK1*) gene. The normal human *NTRK1* gene is located on chromosome 1 (1q21–q22), and its encoded tyrosine receptor is required for nerve growth factor. Because of mutation of the *NTRK1* gene, the encoded protein is abnormal, which in turn produces a series of clinical symptoms.

In our case, tracheal intubation and general anesthesia were required to ensure the safety of the child. The primary damage of congenital analgesia is central structure damage of comprehensive pain perception. Although such children have no pain perception, they still have a stress response to other stimuli, such as endotracheal

intubation. Therefore, a certain depth of anesthesia should be guaranteed during anesthetic induction and surgery to alleviate the stress response induced by endotracheal intubation and the operation. In our case, a small dose of remifentanyl was provided intravenously during anesthetic induction to reduce the stress response caused by endotracheal intubation. The child did not require analgesia and there was no continuous intravenous infusion during the operation. Because our patient did not require analgesia, a small dose was favorable for early extubation. Tongue bites may cause oral hemorrhage. Therefore, rapid induction was adopted in our patient and a tracheal catheter was inserted as soon as possible to avoid regurgitation and aspiration.

Congenital analgesia is a rare autosomal recessive hereditary disease that is characterized by the disappearance or dullness of pain, which can be combined with anhidrosis. Our case showed this combination with anhidrosis, and his peripheral nervous and central nervous systems had normal sensory signal transduction pathways, such as sweating, tearing, the corneal reflex, and the axonal reflex. In this situation, these systems can sense and react to noxious stimuli correctly, but cannot identify the nature of a noxious stimulus, and thus cannot make a defensive response or evasive behavior.

Because of mutation of the *NTRK1* gene, CIPA patient cannot encode neurotrophic tyrosine kinase receptors. This affects formation of autonomic and sensory neurons in the dorsal root ganglion, and eventually leads to loss of sensory nerve fibers in the skin and peripheral nerve fibers in the sweat glands. Recent studies have shown that patients with CIPA have low plasma norepinephrine levels, which is characterized by loss of postganglionic sympathetic neurons.⁴ Because of the limited number of cases, there have been few reports on anesthetic management of such patients. With regard to use of intraoperative analgesics, Okuda et al⁵ considered that patients who was HSAN Type IV may have unpleasant feelings during the operation because of tactile allergies. Therefore, these patients should be provided standard doses of inhalation anesthetics, but not opioids. Oliveira et al⁶ also believed that patients with CIPA do not require opioids because of loss of peripheral sensory nerve fibers. In our case, we used small doses of opioids conservatively because various operational stimuli may induce tactile hypersensitivity and discomfort. However, in view of stability of monitoring indicators during the operation and the absence of pain and other discomfort after the operation, we believe that opioid analgesics are not required in general anesthesia. Because of a heat-dissipating disorder in patients with CIPA, we monitored body temperature, PETCO₂ and BIS at the same time. Under the premise of sufficient sedation, no significant fluctuations in body temperature were observed. Because of a lack of patients with CIPA, especially surgical cases, more research and discussion on how to provide reasonable intraoperative anesthetic management for such patients are required.

In conclusion, opioid analgesics are not required in congenital insensitivity to pain with general anesthesia. A certain depth of anesthesia should be achieved during anesthetic induction and surgery to alleviate the stress response induced by endotracheal intubation and the operation. When there is a heat-dissipating disorder in patients with CIPA, body temperature, PETCO₂, and BIS should be monitored.

CONSENT FOR PUBLICATION

Parental consent obtained. [Correction added on 17 March 2020, after first online publication: The Consent for Publication section was added.]

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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