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CASE REPORT

A case report: Anesthetic management for open-heart surgery in a child with congenital insensitivity to pain with anhidrosis

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Abstract

Congenital insensitivity to pain with anhidrosis (CIPA) is a rare disease also known as hereditary sensory and autonomic neuropathy. CIPA is characterized by a lack of pain sensitivity and impaired development of sweat glands. Surgery is required for patients with self-mutilation and skeletal developmental disorders. Due to the disease's rarity and intricacy, anesthesia poses its challenges. Although there have been a few cases of CIPA patients receiving surgery and anesthesia, the number is very limited. Here, we report a case of a child with CIPA who underwent open-heart surgery and discuss the anesthetic considerations. We conclude that patients with CIPA undergoing open-heart surgery require some opioids, that muscle relaxants and volatile anesthetics should be used with extreme caution, and that airway management and temperature control require special attention.

KEYWORDS
anesthesia, congenital insensitivity to pain with anhidrosis, open-heart surgery

1 | BACKGROUND

Congenital insensitivity to pain with anhidrosis (CIPA) is the most prevalent type of hereditary sensory and autonomic disorder, with an incidence of 1 in 25000.[1,2] CIPA is caused by mutations in the *NTRK1* gene, which result from aberrant development of nociceptive, sensory, and sympathetic neurons.[3,4]

Congenital insensitivity to pain with anhidrosis can be diagnosed via genetic testing during infancy and early childhood. Patients often present pain insensitivity, absent sweating, hyperthermia, mental retardations, abnormal musculoskeletal development, as well as infections and other symptoms. So far, there has been no evidence that patients with CIPA have abnormal airway development. Patients frequently engage in self-mutilation or even bone fractures because of their insensitivity to pain, necessitating one or more surgical procedures. Although there have been some anesthesia case reports of CIPA patients undergoing surgery, due to the limited number of cases, more clinical cases with anesthesia concerns are needed. In this case report, we present a patient with CIPA who underwent atrial septal defect (ASD) repair on the beating heart with cardiopulmonary bypass.

2 | CASE PRESENTATION

A 1-year-old female patient, weighing 10 kg and 80 cm tall, was diagnosed with congenital ASD. The patient fasted for 8 h before the anesthetic procedure. After admission, non-invasive blood pressure monitoring, a 5-lead ECG, and pulse oxygen saturation were conducted on a regular basis. Similar to the bispectral index (BIS), Narcotrend is an EEG monitoring system developed by a research group at Hannover Medical University in Germany to measure the depth of anesthesia. The Narcotrend (MonitorTechnik) was used to measure the depth of anesthesia (post-anesthesia induction). After the induction of anesthesia with sevoflurane with a FiO2 of 80% and placing an intravenous line, the patient was sedated (midazolam

0.5 mg, phencyclidine 0.1 mg, etomidate 6 mg) and paralyzed with cisatracurium 2 mg. Analgesia was provided with sufentanil 5 μg. Endotracheal intubation was performed when anesthetic depth was sufficient. The nasopharyngeal temperature and bladder temperature were monitored. Left radial arterial pressure and internal jugular central venous pressure were monitored via an invasive arterial and central venous catheter. Anesthesia was maintained with propofol (10 mg/Kg/h), sevoflurane (1%–3%), dexmedetomidine (0.4 ug/Kg/h), and cisatracurium (0.5 mg/Kg/h). Dobutamine (2–5 ug/Kg/min) was given after cardiopulmonary bypass, with a perioperative body temperature range of 34.3–36.5°C. Hemodynamics are stable with no obvious bradycardia, tachycardia, severe hypotension, or hypertension. The operation went well, and the patient was transferred to the cardiac surgical ICU. The dobutamine dose was adjusted based on the hemodynamics and terminated 1.5 h after the operation, with hemodynamics remaining stable. The postoperative body temperature ranged from 36 to 37.6°C. The tracheal tube was removed 5 h after the operation, and the patient recovered and was discharged successfully.

3 | DISCUSSION

Congenital insensitivity to pain with anhidrosis (CIPA), also known as hereditary sensory and autonomic neuropathies type IV, is an autosomal disorder with an early onset that is frequently identified at a young age.[5,6] To the best of our knowledge, this is the sole case of CIPA in which the patient underwent intracardiac septal defect repair with cardiopulmonary bypass under direct vision.

The patient was induced with general anesthesia after fasting. Although the patient had fasted for a sufficient period, we still had to consider the patient in a full stomach state, which is worth noting. Airway management has been reported in three cases of CIPA patients. All three patients showed regurgitation or aspiration and even asphyxia-induced cardiac arrest.[7] In other case reports, the incidence of regurgitation or aspiration was also high.[8] This may be related to impaired gastric emptying as a result of autonomic disorders. Therefore, for CIPA patients undergoing general anesthesia, the duration of fasting should be appropriately prolonged, and all patients should be considered in a full stomach state. Rapid induction followed by tracheal intubation is preferred. Ketamine inhibits serotonin uptake at synaptic terminals[9] and increases the risk of nausea and vomiting, which may lead to regurgitation and aspiration.[10,11] Therefore, it should be avoided for anesthesia induction. The patient should be fasted for 6 h according to age. However, we prolonged the fasting duration to 8 h to avoid aspiration or regurgitation due to the disease's peculiarity. Establishing peripheral venous access with intranasal dexmedetomidine followed by induction of intravenous anesthesia, as well as assessing gastric contents with the ultrasonic method before anesthesia should be recommended in the future for these patients.

The need for perioperative opioid administration in CIPA patients is still controversial. According to current literature, some patients can be operated on while sedated[12]; however, it is believed that while these patients are insensitive to pain, paradoxically, they may have hyperalgesia. Some CIPA cases have reported perioperative somatotropic reactions caused by increasing surgical stimulation and postoperative pain complaints.[13,14] Therefore, it has been proposed that opioid analgesics should be used in surgical treatment for this patient population. In addition, it has been found that heart rate, blood pressure, and body temperature all rise significantly when patients are intubated without opioid analgesics.[15] Although epinephrine, norepinephrine, and cortisol levels were not significantly elevated throughout the procedure,[16] it was not ruled out that the patient's stress levels were raised. The patient's stress level may rise due to inadequate perioperative depth of anesthesia, increasing the risk of high body temperature and even cardiac arrest. In this case report, we administered a dose of the opioid sufentanil due to the consideration of high surgical stimulation, hyperalgesia, and endotracheal intubation. Currently, remifentanil[17] and fentanyl[18] analgesics have been used in CIPA patients without causing serious side effects.

Patients with CIPA are unable to sweat due to abnormal sweat gland development, compromising their thermoregulation. Hyperthermia is the primary cause of death in this group of patients.[19] Therefore, better perioperative body temperature management is necessary. Some studies found that the Bispectral index (BIS) and Narcotrend are useful tools for monitoring the depth of anesthesia in such patients.[15,20] Despite the necessity to warm our patient to prevent hypothermia, the temperature must be closely monitored to detect and manage hyperthermia. There is no evidence of a correlation between CIPA patients and malignant hyperthermia. When hyperthermia occurs, it is critical to distinguish between malignant hyperthermia and a temperature imbalance caused by the disease. Malignant hyperthermia is linked to depolarizing muscle relaxants and potent volatile anesthetic agents. To date, there has been no indication that non-depolarizing neuromuscular blocking agents are associated with malignant hyperthermia.

The patient was given sevoflurane inhalation for anesthesia induction and cisatracurium neuromuscular blockade during induction and maintenance. It is noteworthy that patients who use inhaled anesthetics may develop malignant hyperthermia. Current muscle relaxants such as succinylcholine, vecuronium bromide,[14] pancuronium bromide,[21] atracurium,[22] cisatracurium,[15] and mivacurium chloride[8] have been reported to be utilized in CIPA patients without side effects. However, succinylcholine has been closely related to malignant hyperthermia and should be used with caution. Cholinergic antagonists are often used in children as a preoperative medication to minimize perioperative secretions. The safety of atropine administration should be considered in patients with abnormal sweat gland development. According to the present research, atropine has not resulted in serious side effects, but the drug should still be used with caution. There is a report that scopolamine may cause tachycardia and hypertension.[23]

CIPA patients have low concentrations of epinephrine and norepinephrine and the impaired metabolism of catecholamines. Bradycardia, tachycardia, hypotension, hypertension, and even cardiac arrest could occur after general anesthesia. If there is trouble

with cardiac resumption, severe hypotension, or bradycardia, an appropriate increase in vasoactive medications needs to be considered, especially in cardiopulmonary bypass surgery. Some case reports have found a strong correlation between post-anesthesia bradycardia and propofol use.[8] With a standard dose of dobutamine (5 ug/Kg/min), the patient did not develop severe hypotension or intractable hypotension, heralding that dobutamine medicines may be employed as an effective vasoactive agent to maintain hemodynamics.

In this patient population, discomfort or anxiety is mainly assessed in the ICU by monitoring the patient's blood pressure and heart rate. If the patient is in severe pain or if the patient's heart rate and blood pressure change due to anxiety, propofol, and sufentanil may be administered for sedation and analgesia.

In conclusion, some opioids are required for open-heart surgery in patients with congenital insensitivity to pain with anhidrosis. Regurgitation and inadvertent aspiration should be avoided during anesthetic induction, and perioperative temperature management should be strengthened. Hyperthermia should be avoided when using anesthetic medications, and cholinergic antagonists should be used with caution.

DATA AVAILABILITY STATEMENT

There is no data statement.

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