CASE REPORT

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

Jialong Jiang | Xuefeng Wang | Jicheng Hu | Sheng Wang

The First Affiliated Hospital of USTC, Division of Life Sciences and Medicine, University of Science and Technology of China, Hefei, Anhui, 230036, China

Correspondence
Sheng Wang, The First Affiliated Hospital of USTC, Division of Life Sciences and Medicine, University of Science and Technology of China, Hefei, Anhui, 230036, China
Email: iamsheng2006@163.com

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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 cardio-pulmonary 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 μg/
Kg/h), and cisatracurium (0.5 mg/Kg/h). Dobutamine (2–5 μg/Kg/
min) was given after cardiopulmonary bypass, with a periopera-
discharge successfully.

removed 5 h after the operation, and the patient recovered and was
body temperature ranged from 36 to 37.6°C. The tracheal tube was
transferred to the cardiac surgical ICU. The dobutamine dose was
sion, or hypertension. The operation went well, and the patient was
stable with no obvious bradycardia, tachycardia, severe hypoten-
sion, 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 au-
tosomal 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 pa-

tients. 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 im-
paired gastric emptying as a result of autonomic disorders. Therefore,
for CIPA patients undergoing general anesthesia, the duration of fast-
ing should be appropriately prolonged, and all patients should be con-
sidered in a full stomach state. Rapid induction followed by tracheal
intubation is preferred. Ketamine inhibits serotonin uptake at synap-
tic terminals9 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 dexmedetomi-
dine followed by induction of intravenous anesthesia, as well as as-
sessing 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 sedated12; however, it is believed that while
these patients are insensitive to pain, paradoxically, they may have
hyperalgesia. Some CIPA cases have reported perioperative somato-
trropic reactions caused by increasing surgical stimulation and post-
operative 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 pres-
sure, and body temperature all rise significantly when patients are
intubated without opioid analgesics.15 Although epinephrine, norepi-
nephrine, and cortisol levels were not significantly elevated through-
out 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 inade-
quate perioperative depth of anesthesia, increasing the risk of high
body temperature and even cardiac arrest. In this case report, we ad-
ministered a dose of the opioid sufentanil due to the consideration of
high surgical stimulation, hyperalgesia, and endotracheal intubation.
Currently, remifentanil17 and fentanyl18 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 pa-
tients.19 Therefore, better perioperative body temperature man-
agement 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 hyperthermia, 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 ma-
lignant 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 in-
duction 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,14 atracurium,12 cisatracurium,15 and mivaca-
rium chloride8 have been reported to be utilized in CIPA patients
without side effects. However, succinylcholine has been closely re-
lated 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 atro-
pine administration should be considered in patients with abnormal
sweat gland development. According to the present research, atro-
pine 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 car-
diac 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. With a standard dose of dobutamine (5µg/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.

**ORCID**

Sheng Wang https://orcid.org/0000-0002-1187-3240

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