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CASE REPORT

# Anesthesia for ambulatory surgery in a child with hyposensitivity to pain

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## Abstract

Congenital hyposensitivity to pain is a condition with predisposition to injury. In these patients, knowledge regarding anesthetic requirements and complications derives from individual case reports, or small case series. Different categories have been described. In patients with hyposensitivity to pain, preventing and treating anxiety as well as insuring immobilization, avoidance of triggering of autonomic reflexes, and sedation are integral aspects for a safe and adequate anesthetic management.



**Key words:** General anesthesia; Child; Pain; Hyposensitivity; Surgery

**Core tip:** Congenital hyposensitivity to pain is a condition with predisposition to injury. In patients with congenital hyposensitivity to pain/Hereditary sensory and autonomic neuropathy (HSAN), knowledge regarding anesthetic requirements and complications derives from individual case reports, or small case series. Different categories of HSAN have been described. Complications in the immediate perioperative period have been described such as mild hypothermia and cardiovascular events, mostly bradycardia and hypotension. The majority of patients with hyposensitivity to pain reported in the literature have received standard anesthesia for surgery. Immobilization, prevention of autonomic reflexes, anxiolysis, and sedation are equally important aspects of the anesthetic management in patients with hyposensitivity to pain.

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## INTRODUCTION

Congenital hyposensitivity to pain is a condition with predisposition to injury, often associated with a delay and difficulty in diagnosis. Anesthesia care of these children may pose a challenge secondary to the rarity of the disease, the presence of unclassified congenital variants of pain hyposensitivity, and the limited information regarding anesthetic management.

## CASE REPORT

The following is a description of a case of a 2-year-old patient, female gender, with mild developmental delay, who presented as a same day case for ear surgery. The duration of the surgery was expected to be 90 min. During the process of the interview, the mother revealed that her daughter is insensitive to pain, no further details were available. Otherwise the review of systems was negative. Premedication with oral midazolam 0.5 mg/kg was administered. Patient received general anesthesia with endotracheal intubation [Sevoflurane mask induction in

$O_2/N_2O$ followed by propofol (3 mg/kg Ⅳ)], and narcotics were titrated to maintain spontaneous ventilation for a total dose of Fentanyl of 1 mcg/kg. No fluctuations in temperature were noticed during the case. Vital signs were stable. There was no delay in emergence from general anesthesia. Recovery room stay was not prolonged and was uneventful. Patient was discharged the same day without further need for narcotics postoperatively.

## DISCUSSION

In patients with congenital hyposensitivity to pain/hereditary sensory and autonomic neuropathy (HSAN), knowledge regarding anesthetic requirements and complications derives from individual case reports, or small case series. Different levels and modalities of autonomic dysfunction and sensory loss have been described[1]. Some patients do have tactile hyperesthesia, or partially preserved nociception with sometimes preserved mechanoreceptor, cooling, and warming sensations. Five types of HSAN have been categorized. HSAN Ⅰ is inherited with autosomal dominance. The age of onset of HSAN Ⅰ is between the 2nd and 4th decade of life while the other types are autosomal recessive with an earlier age of onset, usually at birth in type Ⅲ (familial dysautonomia) or in infancy. HSANs Ⅱ, Ⅳ and Ⅴ usually present with a profound reduction of pain perception, while HSAN Ⅰ has as a milder manifestation. Patients with HSAN Ⅲ's may have intact visceral and peritoneal pain sensation with profound dysautonomia. Thermal perception is severely impaired in all HSAN types. Mild hyperhidrosis is associated in HSAN type Ⅴ, hypohidrosis in type Ⅰ and Ⅱ, while severe anhidrosis with recurrent episodes of severe hyperpyrexia is associated to HSAN type Ⅳ. Type Ⅴ may present with unaffected sensitivity to touch, pressure and vibration. Mutilations may be common in all types of HSAN. The requirements for volatile anesthetics have been described as being within the range of standard population. Intraoperative opioids dosage has been reported to be less than standard, if not negligible. Because some HSAN patients may have anhidrosis, intraoperative hyperthermia cases have been reported[2]. There is no description of malignant hyperthermia in association with the different HSAN types. Patients usually do not require opioids postoperatively. Complications in the perioperative care have been described as cardiovascular, such as bradycardia and hypotension and mild hypothermia. A cardiac arrest following management of a patient with HSAN Ⅳ has been published[3]. Although there is report of an adult patient diagnosed with profound congenital insensitivity to pain who has undergone a major orthopedic surgery without receiving general anesthesia and narcotics; in the literature, patients with hyposensitivity to pain, have been documented to receive standard anesthesia for surgery. In patients with hyposensitivity to pain, preventing and treating anxiety as well as insuring immobilization, avoidance of triggering of autonomic reflexes, and sedation are integral aspects for a safe and adequate anesthetic management.

## COMMENTS

### *Case characteristics*

Pediatric patient with hyposensitivity to pain.

### *Clinical diagnosis*

Anesthesia management of a pediatric patient with hyposensitivity to pain.

### *Differential diagnosis*

Different categories of hereditary sensory and autonomic neuropathy are described.

### *Experiences and lessons*

Immobilization, prevention of autonomic reflexes, anxiolysis, and sedation are equally important aspects of the anesthetic management in patients with hyposensitivity to pain.

### *Peer review*

The author described anesthesia management in a child with congenital hyposensitivity to pain. The diagnosis is based on the history and clinical findings. The paper is good for publication.

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