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Case Report Open Access

Anesthetic Management of Congenital Insensitivity to Pain with Anhidrosis in a Child: A Case Report

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Abstract

Background: Congenital insensitivity to pain with anhidrosis (CIPA) is a rare hereditary sensory autonomic neuropathy. Anesthetic management of those patients is challenging for anesthesiologist because of the potential risks for them including regurgitation and aspiration, sympathetic nervous dysfunction, mandibular osteomyelitis as well as hyperthermic events.

Case report: We present total intravenous anesthesia (TIVA) with spontaneous respiration *via* facemask was applied in a 2 y old child with CIPA who underwent closed reduction and plaster external fixation of a bilateral dislocation of the hip. The patient vital signs remained stable, and did not show any signs of distress by anesthetics during the surgery. And no complication occurred.

Conclusion: TIVA with spontaneous respiration *via* facemask is an effective and safe anesthetic management for patients with CIPA.

Keywords: Case report; Congenital insensitivity to pain with anhidrosis; Anesthetic management; Total intravenous anesthesia with spontaneous respiration

Introduction

Congenital insensitivity to pain with anhidrosis (CIPA, or hereditary sensory and autonomic neuropathy type IV), which was first reported in 1963, is a rare autosomal recessive disease (incidence: 1 in 25,000), related to a mutation in the TrkA gene, characterized by absence of reaction to noxious stimuli, recurrent episodes of unexplained fever, anhidrosis (inability to sweat), mental retardation and self-mutilating behavior [1-4]. Anesthetic management of those patients is challenging for anesthesiologist because they have the potential risks for regurgitation and aspiration, sympathetic nervous dysfunction, mandibular osteomyelitis as well as hyperthermic events [5]. In this case report we present total intravenous anesthesia (TIVA) with spontaneous respiration *via* facemask applied and share the management of anesthetic considerations of CIPA in a child.

Case Report

A 2-year-old Chinese girl with a weight of 11 kg presented for closed reduction and plaster external fixation of a bilateral dislocation of the hip. The young child could not stand up for 20 days before the surgery. She suffered from recurrent episodes of unexplained fever without sweating, and lacked sensation to pain from birth. After establishment of dentition (lower incisors were congenitally missing), she began performing masochistic behaviors of biting her lips and the tips of her fingers. The patient did not have the manifestations of alopecia and visual disturbances. Physical examination didn't display muscle hypotonia and power decline. Basic laboratory testing was normal. This child had no history of surgery or other fractures. Among her known family relationship, there was no genetic disorder in her family members.

During surgery, three-lead ECG with ST-segment analysis, non-invasive blood pressure (NIBP), pulse-oximetry, end-tidal carbondioxide monitoring *via* nasal and temperature were monitored for the child. The axillary and rectal temperature of the patient was monitored in awake and anesthetized state, respectively. After pre-oxygenation, general anesthesia was induced with 2 mg/kg propofol. At the beginning of the operation, 0.1 mg/kg midazolam was intravenously injected. Spontaneous respiration *via* facemask was maintained at a rate of 16-24/min with 2 L/min oxygen. With the monitoring of anesthetic depth using a BIS monitor (The BIS value was kept between 45 and 70), 50 μg/kg/min propofol was continuously administrated thereafter without any analgesic co-administrated. The patient vital signs remained stable, and did not show any signs of distress by anesthetics during the operation. The surgery went smoothly and finished in 45 min. After the surgery, the child was transferred to the post anesthesia care unit without any problem.

Discussion

CIPA is a rare hereditary sensory autonomic neuropathy [1]. Corresponding anesthetic management is therefore different from the

normal. CIPA patients hardly feel pain, but have tactile hyperesthesia. Appropriate anesthesia is necessary for preventing the undue tactile sensation. Generally, patients with CIPA do not require analgesics for the maintenance of anesthesia, and are prone to become hemodynamically unstable with high concentrations of either volatile or intravenous anesthetics. Small doses of sedatives could achieve appropriate anesthetic depth in those patients [6,7]. The anesthetic regimen without opioids could diminish the risk of respiratory depression.

For CIPA patients, the intraoperative basic monitoring (pulse oximetry, capnography, blood pressure, ECG, body temperature) is sufficient for general procedures in children. Temperature should be carefully paid attention to because the thoracic sympathetic nerve which associated with temperature regulation usually present dysfunction in CIPA patients. In our case, the child had the symptoms of recurrent episodes of unexplained fever without sweating. During the surgery, however, her temperature was normal.

The autonomic nervous system abnormalities are common in patients with CIPA, leading to disturbances in gastric myoelectric activity and therefore gastric emptying delay and dyspeptic symptoms, predisposing CIPA patients to an increased risk of regurgitation and aspiration when they are anesthetized [8,9]. In our case the throat reflex was maintained during TIVA with spontaneous respiration *via* facemask, avoiding aspiration if the patient regurgitated. Previous cases report demonstrated that laryngeal mask airway (LMA) is not appropriate for the airway management of CIPA patients, leading regurgitation, aspiration and even hypoxic cardiac arrest [9]. For those patients, surgical procedures can be done with superficial anesthesia but the gag reflex from stimulation of the pharynx with LMA may not be inhibited. Thus laryngeal mask may induce vomiting and reduce the efficiency of vaccum suction in oropharyngeal. That is probably why LMA have high risk of aspiration for CIPA patients. Furthermore, the incidence of mandibular osteomyelitis is very high among patients with CIPA and can result in pathologic fracture [10]. Control airway may cause other oral complications. Compared with tracheal intubation anesthesia or laryngeal mask with mechanical ventilation, TIVA with spontaneous respiration *via* facemask is an effective and safe anesthetic management for patients with CIPA.

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