General Anesthesia for a Child with Congenital Insensitivity to Pain with Anhidrosis

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Introduction

- Congenital insensitivity to pain with anhidrosis (CIPA), also known as hereditary sensory and autonomic neuropathy (HSAN) type IV with anhidrosis, is a rare autosomal-recessive disorder featuring the inability to feel pain and temperature, and decreased or absent sweating (anhidrosis).

![Fig 1: Five different clinical entities have been described under hereditary sensory and autonomic neuropathies – all characterized by progressive loss of function that predominantly affects the peripheral sensory nerves. Their incidence has been estimated to be about 1 in 25,000.]

- Less than 50 cases have been reported.
- CIPA results from a defective development of the neural crest, with loss of the nociceptive neurons in the dorsal root ganglia, and in the sympathetic ganglia. Nerve growth factor (NGF) guarantees survival of these cells, through the neuronal TRKA receptor. Mutations in the TRKA gene, found in HSAN patients, correlate well with the defective development of the nociceptive neurons.

![Case description

- A 3-year-old female with CIPA, global developmental delay, ADHD, failure to thrive requiring gastric tube feeding, right thumb amputation due to self-mutilation, scarlet fever, and recurrent otitis is scheduled for incision and drainage (I&D) of the left elbow due to osteomyelitis.
- After a 2-month antibiotic course for methicillin-resistant Staph aureus and Group A Streptococcus infection of her right thumb and left elbow, she is brought to the pre-op clinic wearing a helmet to protect from self-injuries.
- Because of her anxiety to face masks that developed from her previous anesthesias, mask induction was avoided.
- In the pre-op area, she patiently watched an anesthesiology team member place her IV and tolerated the procedure without any signs of pain.
- In the OR, anesthesia was induced with 2 mg/kg intravenous propofol and was maintained with sevoflurane via laryngeal mask airway.
- A rectal temperature probe was placed, in addition to standard monitors. She remained normothermic throughout her perioperative course.
- Apart from sevoflurane, she did not require additional analgesia, and she did well in the recovery room, with no opiate requirement.
- The child underwent the same procedure twice, and no opiates were administered in either case, although she received rectal acetaminophen prior to incision.

Discussion

- Common features of CIPA are the lack of pain sensation, painless injuries of the extremities from self-mutilation, which often lead to osteomyelitis that requires repeat procedures, corneal abrasions and injuries of oral structures, mental retardation, and fever due to anhidrosis. As the child grows older, bone fractures, joint deformities, and limb amputation are common too.
- CIPA differs from more common familial dysautonomias by complete insensitivity to superficial and deep painful stimuli and normal lacrimation, much milder autonomic dysfunction, with absent postural hypotension or dysphagia.
- Most children with CIPA present for an I&D, but usually not until the infection is much worse or they have developed a systemic fever.

- Decreased central and peripheral norepinephrine in addition to anhidrosis may cause hyperpyrexia in perioperative children. Death from hyperpyrexia occurs within the first 3 years of life in almost 20% of these patients.

References