[NM-248] General anesthesia for a child with congenital insensitivity to pain with anhidrosis

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GENERAL ANESTHESIA FOR CHILD WITH CONGENITAL INSENSITIVITY TO PAIN WITH ANHIDROSIS
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Introduction: Hereditary sensory and autonomic neuropathy type IV with anhidrosis, is a rare autosomal-recessive disorder featured by the inability to feel pain and temperature, and decreased or absent sweating (anhidrosis). Also known as congenital insensitivity to pain with anhidrosis (CIPA), less than fifty cases have been reported. This disorder results from a defective development of the neural crest, with loss of the nociceptive neurons in the dorsal root ganglia, and loss of cells of the sympathetic ganglia. Survival of these cells is stimulated by the nerve growth factor (NGF) 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: 3 year old female with CIPA, global developmental delay, ADHD, failure to thrive requiring feeds via gastric tube, right thumb amputation secondary to self-mutilation scheduled for an incision and drainage of the left elbow complicated by osteomyelitis. She was brought to the pre-op clinic wearing a helmet to protect her from self-injuries, after a 2-month antibiotic course for methicillin resistant staph aureus and Group A Streptococcus infection of her right thumb and left elbow. She had a history of anxiety to face masks although she was happy to have her helmet removed. In the pre-op area, she was patiently watched an anesthesiology team member place her IV. In the OR, anesthesia was induced with 2 mg/kg intravenous propofol, and a laryngeal mask airway was placed. A rectal temperature probe was placed in addition to standard monitors. Apart from sevoflurane, she did not require additional analgesia, and she did well in recovery with no opiate requirement. The child underwent the same procedure twice, 7 and 14 days after. Both times no opiates were administered, although she received acetaminophen prior to incision.
Discussion: Because of the patient’s history of anxiety with mask induction, this was avoided, which is unique in a three-year old. Common features of CIPA are corneal abrasions and painless injuries of the extremities from self-mutilation, often leading to osteomyelitis requiring repeat procedures. 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 the patients. CIPA differs from FD by complete insensitivity to superficial and deep painful stimuli and normal lacrimation, much milder autonomic dysfunction, with absent postural hypotension or dysphagia.

References:1)Congenital insensitivity to pain with anhidrosis. Sztriha L. Pediatr Neurol 2001;25:63-66. 2)Disease mechanisms in hereditary sensory and autonomic neuropathies. Nathalie Verpoortena. Neurobiology of Disease. Volume 21, Issue 2, February 2006, Pages 247–255. 3)Congenital insensitivity to pain with anhidrosis (hereditary sensory and autonomic neuropathy type IV). Sérgio Rosemberg. Pediatric Neurology. Volume 11, Issue 1, July 1994, Pages 50–56
