

Original article

Anesthetic Management for a Patient with Congenital Insensitivity to Pain with Anhidrosis. — Too naked to live alone —

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Summary

We anesthetized a 15-year old boy with congenital insensitivity to pain with anhidrosis (CIPA) for a knee joint surgery. CIPA is a rare disease and is recognized early in infancy, as the patient has high fevers because of the inability to perspire, and is an autosomal-recessive disorder characterized by recurrent episodes of unexplained fever, anhidrosis (absence of sweating) and absence of reaction to noxious stimuli, self-mutilating behavior and mental retardation. The defects of pain and temperature sensations in CIPA are probably due to an almost complete absence of the first-order afferent system that responsible for pain and temperature sensation. The case of CIPA, pathological changes in its homeostatic circumstances should be examined and evaluated preoperatively, required that the temperature of the operating room and the blanket was controlled properly and should be managed with low dose general anesthesia for its low noradrenaline activity combined with rapid induction for his anxiety.

Key words : No-pain, Anhidrosis, General anesthesia

Introduction

Congenital insensitivity to pain with anhidrosis (CIPA) is a rare disease and is recognized early in infancy, as the patient has high fevers because of the inability to perspire, and is an autosomal-recessive disorder characterized by recurrent episodes of unexplained fever, anhidrosis (absence of sweating) and absence of reaction to noxious stimuli, self-mutilating behavior and mental retardation. CIPA is also known as congenital sensory neuropathy with anhidrosis or hereditary sensory and autonomic neuropathy type IV. The defects of pain and temperature sensations in CIPA are probably due to an almost complete absence of the first-order afferent system that responsible for pain and temperature sensation. We scheduled general anesthesia with sevoflurane, nitrous oxide and vecuronium for the patients with CIPA.

Case report

The patient was a 15 years old boy (151cm, 40kg) and was normal delivery and no familial complications. During infancy, the patient had high fevers repeatedly. When he was 5 years old, he sustained right leg fracture and right ankle infectious arthritis, and had casting and conservative therapy. Afterward, he had many painless injuries of extremities. When he was 8 years old, he was pointed out the Charcot's joint of right knee and he had biopsy under general anesthesia. When we saw him the patient could walk only about two feet because of his wounded knee,

he used a wheelchair. Neurological examination showed normal tendon reflexes, while the both Achilles-tendon reflexes were exaggerated. Sensation to pin prick was severely impaired over both the trunk and the extremities. Sensation to touch was intact, except for hypoesthesia in the feet. Sensations of cold, heat, and vibration all were normal. The conduction velocity of sensory and motor neuron was normal. Preoperative laboratory data were within normal range without increasing of CRP for the synovitis. The intelligence quotient was low (60 according to the Suzuki-Binet test). And this time, he was scheduled for open synovectomy and continuous irrigation of left knee joint. Premedication was 0.4mg atropine sulfate and 50mg hydroxyzine hydrochloride intramuscularly injected. We monitored non-invasive blood pressure, Train-of-Four ratio and ECG prior to the induction of anesthesia. General anesthesia was induced with thiopental sodium 200mg, sevoflurane 3.0%, and vecuronium 6mg for tracheal intubation. Anesthesia was maintained with sevoflurane (0.1-0.2%) and 60% nitrous oxide in oxygen supplemented. And the body temperature was monitored continuously and temperature of the operating room was controlled at 24°C, the blanket was controlled at 34°C. There were no hemodynamic changes at intubation and surgical incision in spite of low anesthetic level but there were a hemodynamic change at tourniquet on. He showed decreased response to vecuronium under monitoring of neuromuscular block. Totally we used vecuronium 10mg. And he responded favorably to anticholinesterase. He had little pain and showed no progress in neurological symptoms in the postoperative period. He showed in-

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creased response to sevoflurane under monitoring of non-invasive blood pressure and ECG. During the anesthesia patient's general condition and body temperature was stable and the operation was finished without any episodes. He did not have any complications in the postoperative period.

Discussion

The reported results show that there is a characteristic marked decrease in A-delta myelinated fibers and marked decrease in unmyelinated (C-type) fibers in these patients. These fibers are the afferent fibers for the pathway of sensation of pain. These results can explain the indifference to pain, absence of perspiration, and other clinical signs. The frequent fractures may be attributable either to the patient's inability to prevent trauma or to overuse. Most patients have been mentally retarded, due to a deficit in the development of the nervous system. We should be taken such as the management of the body temperature, concentration of anesthetic agent and dose of muscle relaxants. Totally, general anesthesia maintained with sevoflurane and nitrous oxide should be useful for perioperative management of the patient, and the use of anticholinergic agent was not raise the body temperature for there were no unmyelinated fibers around the sweat glands. The case of CIPA, pathological changes in its homeostatic circumstances should be examined and evaluated preoperatively, required that the temperature of the operating room

and the blanket was controlled properly and should be managed with low dose general anesthesia for its low noradrenaline activity combined with rapid induction for his anxiety.

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原 著

先天性無汗無痛覚症患者の全身麻酔管理

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先天性無汗無痛覚症はまれな感覚性神経疾患であり、精神遅滞、カテコラミン代謝異常を伴い、自損傷、過剰動作、感染等による障害をおこしやすい。今回、われわれは膝関節膜切除、ドレナージを行った15歳少年の全身麻酔管理を経験した。前投薬はアトロピン0.5mg、ヒドロキシジン50mgを筋注し、イソゾール200mg、ベクロニウム6mg、セボフルレン3%で導入、GOSにて維持した。術中は体温の上昇を防ぐため室温、ブランケット温を適宜調節し、血行動態の変化等も低吸入麻酔薬濃度にて最小限におさえることができた。また、アトロピンも体温の上昇をきたさず安全に使用できた。先天性無汗無痛覚症児に対し急速導入、全身麻酔管理は周術期に悪影響を与えることなく安全に行えると思われる。

キーワード：無痛覚、無汗、全身麻酔

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