

Anesthetic management of a patient with congenital insensitivity to pain and anhidrosis using remifentanil

Takeuchi Y.¹, Fujita Y.¹, Shimomura T.¹, Kurokawa S.², Noguchi H.², Fujiwara Y.¹

1) Department of Anesthesiology, Aichi Medical University School of Medicine

2) Department of Anesthesiology, JA Aichikoseiren Konan Kosei Hospital

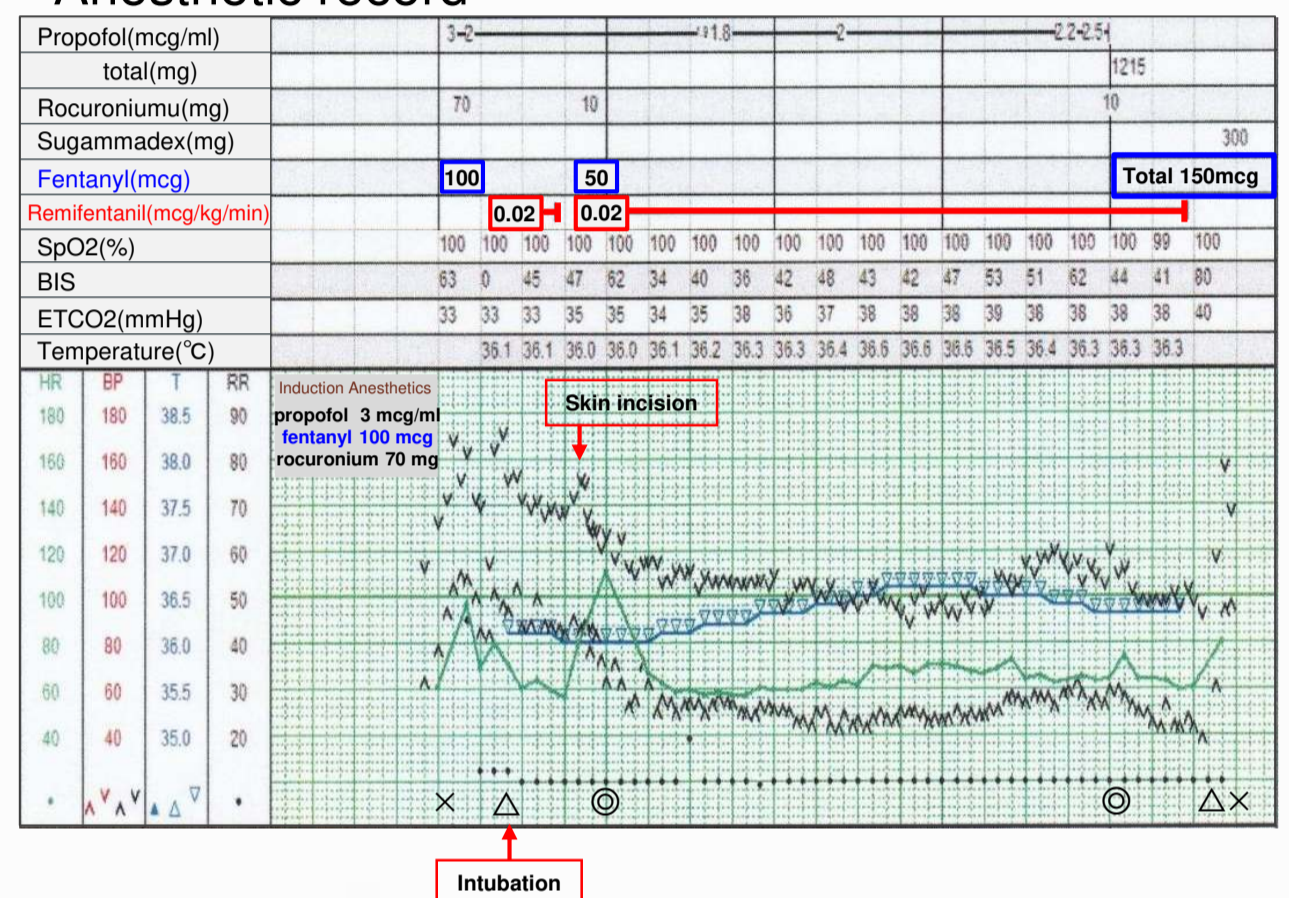
Summary:

We administered general anesthesia for a 37-year-old male with congenital insensitivity to pain and anhidrosis (CIPA). Despite the lack of pain sensation, the blood pressure and heart rate increased at the time of intubation and skin incision. This case suggests that we are able to maintain anesthesia of CIPA safely with a low dose of remifentanil without hyperalgesia or shivering.

Background:

- Congenital insensitivity to pain and anhidrosis (CIPA) is a rare autosomal disease characterized by loss of pain and thermal sensation and mental distress.
- A few reports of the anesthetic management of patients with CIPA have been published from Japan and Israel.¹
- The number of CIPA patients in Japan is reported from 130 to 210 people.¹
- Insensitivity to pain leads to bone fractures, burns and self-mutilation such as tongue, lips or finger biting, with some patients requiring surgical procedures.

Anesthetic record



Case Report:

- A 37-year-old male (153 cm, 69 kg) with CIPA underwent posterior spinal fusion for thoracic spondylotic myelopathy.
- Anesthetic induction involved target controlled infusion (TCI) of propofol (site-effect 3.0 mcg/ml), fentanyl (100 mcg), and rocuronium (70 mg), and anesthetic maintenance was achieved by propofol at a bispectral index (BIS) of 40 to 60.
- After intubation and at the time of skin incision, the patient's blood pressure and heart rate increased. We administered 50 mcg of fentanyl and maintained anesthesia with propofol (2.0–2.5 mcg/ml) and remifentanil (0.02 mcg/kg/min). The blood pressure and heart rate then remained stable during surgery.
- We also continuously monitored the rectal temperature and maintained it at 36° C to 37° C using an air conditioner and warming device.
- After extubation, this patient felt discomfort of the throat but did not have any pain without any postoperative analgesic drugs.

Discussion:

- The most important points of management of patients with CIPA are selection of the appropriate analgesic drug and maintenance of body temperature.
- We used fentanyl and remifentanil as the analgesic drugs in the present case. To our knowledge, no reports have described the anesthetic management of patients with CIPA using remifentanil. Because of their insensitivity to pain, anesthesia can be maintained without analgesia in patients with CIPA.
- However, despite the lack of pain sensation, perceptions of touch, pressure and vibration are more sensitive. So some require fentanyl for airway manipulation.² In our patient, the blood pressure and heart rate increased after intubation and at the time of skin incision, fentanyl and remifentanil were therefore administered.
- We were otherwise able to maintain anesthesia with a low dose of remifentanil to prevent reaction to surgical stimuli without hyperalgesia³ or shivering.
- CIPA is an infrequent disease and it is difficult to estimate the sensory responses. Therefore, further study is required on the safety of anesthesia on patients with CIPA.

References:

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3. Zlotnik A et.al. Anesthetic management of patients with congenital insensitivity to pain with anhidrosis: a retrospective analysis of 358 procedures performed under general anesthesia. *Anesth Analg.* 2015; 121: 1316-20

