

# Remifentanyl In Myotonic Dystrophy - Avoiding The Use Of Muscle Relaxants And Long Acting Opioids

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## Abstract

Myotonic dystrophy, a genetic disorder of abnormal muscle function, is associated with many anesthetic hazards. Sensitivity to anesthetic drugs, especially muscle relaxants and opioids, may complicate postoperative care. A period of postoperative ventilation to allow opioids and sedatives to wear off may be required. The use of succinylcholine is contra-indicated because of profound muscle contraction which can make ventilation impossible. Non-depolarising agents can also give unpredictable responses, ranging from decreased sensitivity to exaggerated response. We describe a case where the use of remifentanyl allowed ventilation of a patient with myotonic dystrophy for more than 2 hours and avoided the use of muscle relaxants and longer acting opioids. The short context-sensitive half-time of remifentanyl allowed immediate recovery and safe discharge home on the same day.

## INTRODUCTION

Myotonic dystrophy is a genetic disorder characterised by delayed muscle relaxation following contraction. Anesthetic considerations include undue sensitivity to premedicant drugs, induction agents, opiates and non-depolarizing muscle relaxants.<sup>(1)</sup> Precipitation of myotonia has been reported following administration of depolarizing muscle relaxants and anticholinesterases.<sup>(1,2)</sup> In addition physical factors including cold and shivering can also precipitate an acute attack.

We describe a case where the use of remifentanyl produced optimal surgical conditions while avoiding the use of both muscle relaxants and long acting opioids during general anesthesia for extensive dental surgery.

## CASE REPORT

A 20 year old female, known with myotonic dystrophy required extensive dental work which was to be performed under general anesthesia. Pre-operative assessment revealed clear signs of muscle wasting, ptosis and micrognathia, Mouth opening and neck mobility were adequate. She had no significant cardiorespiratory symptoms and weighed 48 kilograms.

To ensure a safe airway and avoid a period of apnoea, we chose an inhalational induction with oxygen and sevoflurane. Laryngoscopy under deep inhalational

anesthesia revealed a grade 3 laryngoscopy. The trachea was intubated with a size 6.5 endotracheal tube over a gum elastic bougie. A remifentanyl infusion was started at 0.2 mcg.kg-1.min-1 and the patient was ventilated to normocapnia. Oxygen, nitrous oxide and isoflurane were used for maintenance and ketorolac 10mg given for supplementary analgesia. The dentist infiltrated prilocaine 3% with octopressin. Anesthesia was uneventful. At the end of the 150 minute procedure remifentanyl, nitrous oxide and isoflurane were discontinued and spontaneous ventilation returned within 5 minutes. The patient was safely extubated 15 minutes after the end of surgery and was closely observed in the recovery area. Pain was well controlled with paracetamol and ibuprofen and her respiratory pattern was normal. On reassessment 4 hours postoperatively she met the hospital discharge criteria for day surgery patients. In view of patient preference and full recovery she was discharged home later that same day.

## DISCUSSION

Myotonic dystrophy is the most common of the myotonic syndromes with a prevalence of 4:100 000. It exhibits autosomal dominant inheritance. Males and females are equally affected. Myotonia is an intrinsic disorder of muscle due to abnormal closure of sodium and chloride channels.

Characteristic features include difficulty in releasing muscle grip, frontal balding, ptosis, cataracts and "inverted smile".

Apart from generalized muscle wasting, myotonic dystrophy is also a multisystem disorder. Cardiac abnormalities include conduction defects and cardiomyopathy. Ventilatory impairment is multifactorial, partly due to respiratory muscle weakness and partly due to a central component causing hypoventilation and decreased ventilatory response to carbon dioxide.<sup>(1)</sup>

Remifentanil is a selective  $\mu$ -agonist with a rapid onset time and very short duration of action. The drug differs from existing opioids in its rapid metabolism by non-specific red cell and tissue esterases. A further important difference is the consistency of the pharmacokinetics of remifentanil with prolonged infusions. Other opioids accumulate after prolonged infusions, where remifentanil has a context-sensitive half-time of only a few minutes.<sup>(3)</sup> Another benefit is the potent respiratory depressant effect of remifentanil which facilitates ventilation as part of a balanced anesthetic technique, without using muscle relaxants.

A previous report described the use of propofol and alfentanil infusions for both induction and maintenance of anesthesia in a patient with myotonia. <sup>(4)</sup> The procedure lasted 40 minutes and the patient woke up rapidly with no respiratory sequelae. Recent case reports on recurrent respiratory depression after alfentanil infusion <sup>(5)</sup> do cause

concern especially in myotonic patients who are more susceptible to respiratory depression due to opioids. Due to the unique metabolism and rapid offset time of remifentanil it is beneficial in patients with myotonia because delayed opioid induced respiratory depression is avoided.

The use of remifentanil as part of a balanced anesthetic technique ensured an uncomplicated recovery period and safe same day discharge. This case report is, to the best of our knowledge, the first in the English literature, where remifentanil had been used in a patient with myotonia and longer acting opioids and muscle relaxants were both avoided.

### **References**

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