

Remifentanil in a patient with Huntington's chorea

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Summary

Relatively few published case reports related to the anaesthetic management of Huntington's chorea (HC) exist. At the time of surgery no publications were found related to remifentanil's use in patients with HC. This case report describes the management of a confirmed HC patient requiring urgent decompression of a spinal tumour associated with quadriparesis. A fibre-optic (FO) assisted naso-tracheal intubation was performed under mild sedation with midazolam and fentanyl. Prior nebulization with lignocaine and further supplementary topical application provided local anaesthesia to the airway. Following intubation the patient was assessed neurologically prior to induction with propofol 100 mg and rocuronium 30 mg. Isoflurane 0,5% to 0,75% and remifentanil, initially infused at the rate of $0,25\mu\text{g kg}^{-1}\text{ min}^{-1}$ rapidly reduced to between $0,04\text{--}0,08\mu\text{g kg}^{-1}\text{ min}^{-1}$, provided intra-operative anaesthesia and analgesia for the three hour procedure. A Bair Hugger warming device was used to guard against the risk of hypothermia and subsequent postoperative shivering known to precipitate life threatening generalized spasms in HC patients. The surgery and anaesthesia had a favourable outcome. Rapid recovery after the termination of the remifentanil and the absence of any untoward intraoperative events related to its use demonstrate that remifentanil may be a valuable adjunct for providing intraoperative analgesia in patients with HC.

Key words: Genetic factors, Anaesthesia, Surgery, Spinal, Remifentanil

Huntington's chorea (HC), a rare hereditary degenerative neurological disorder associated with progressive dementia, ataxia and involuntary choreiform movements, was first described in April 1872 by Dr George Sumner Huntington¹, a year after he graduated from Columbia University Medical School. Published case reports related to the anaesthetic management of HC remain few in number. The disorder usually presents between the ages of 30 and 45 years with a triad of symptoms and signs affecting emotion, cognitive ability and mobility. The reported anaesthetic problems related to HC include increased risk of aspiration², increased sensitivity to a number of anaesthetic drugs including thiopentone³ and suxamethonium⁴ and post operative shivering associated with inhalational agents that may trigger life threatening tonic spasms.⁵

Case report

A 48 year old 62 kg female presented with a three-month history of increasing quadriparesis and bladder retention. Six years previously she had experienced clumsiness as her first

symptom of Huntington's chorea. Over the next four years her symptoms had progressed to include chorea with involuntary oro-facial movements and dysarthria. Her diagnosis of HC was confirmed by DNA analysis. Relevant family history included both her father and one of her brothers dying of complications of Huntington's chorea at the ages of 50 years and 45 years respectively.

The aetiology of her quadriparesis was severe cord compression secondary to an intradural extramedullary lesion unrelated to HC. The lesion was demonstrated by magnetic resonance imaging to extend from the second to the fourth cervical level with evidence of erosion of the lamina of the second and third cervical vertebrae (Figure 1). The patient was presented for excision of the lesion and spinal cord decompression. The anticipated anaesthetic challenges in this instance were problems related to HC along with those related to the severe cord compression and quadriparesis. On examination she was found to have a marked reduction of neck extension in addition to her quadriparesis due to the involvement of her cervical laminae. Her Mallampati score was 2. She had a 30 year history of heavy smoking with clinical and radiological evidence consistent with chronic obstructive pulmonary disease. Her respiratory function was optimized pre-operatively with active chest physiotherapy supplemented with 6-hourly salbutamol and ipratropium bromide nebulization therapy and attention to adequate hydration.

An awake fibre-optic bronchoscope aided intubation was considered as the safest option for securing the airway in the

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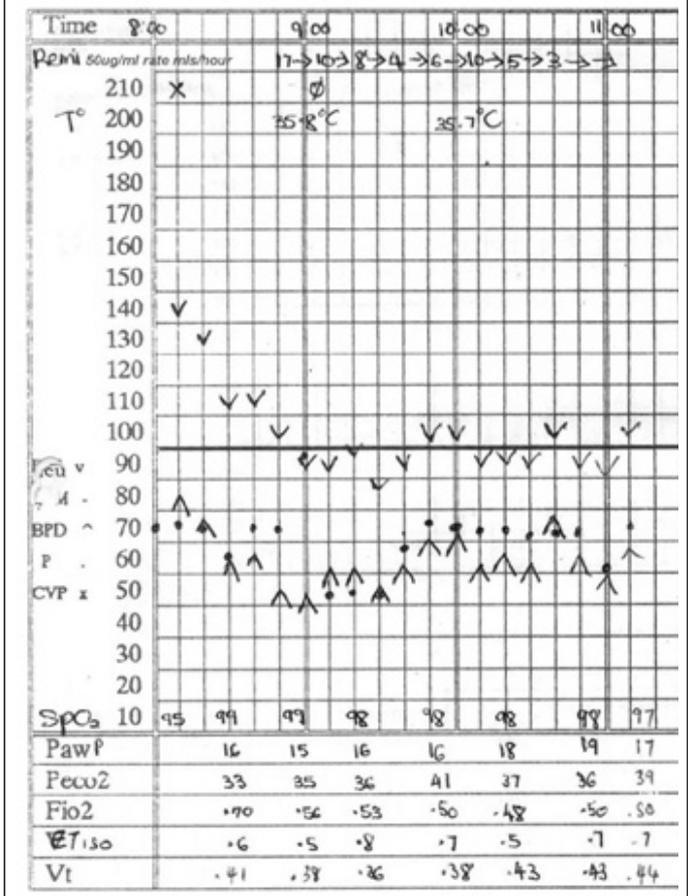
presence of severe cord compression and quadriparesis.⁶⁻⁹ Fortunately dementia, commonly associated with HC, was not present as that may have precluded this option. Topical anaesthesia of the airway was initiated by nebulizing the patient with 4% lignocaine. This is reported to avoid the violent coughing associated with direct instillation of local anaesthetic solutions to the larynx and trachea which in turn may endanger patients with severe cord compression.⁸ Oxymetazol hydrochloride (Drixine[®]) nose drops were instilled into both nostrils prior to the topical administration of lignocaine gel to the nostrils and the nasopharynx. Topical anaesthesia of the larynx and trachea was supplemented by direct administration of 4% lignocaine via the fibre-optic laryngoscope. Intravenous midazolam 2mg and fentanyl administered in 20µg boluses totalling 100µg provided sedation during intubation. A neurological assessment was performed following placement of a 6.5 armoured nasotracheal tube. Following a neurological assessment the patient was induced with propofol and rocuronium. In view of the limited availability of propofol, anaesthesia was maintained with a mean end tidal isoflurane of 0,6%. A remifentanyl infusion, commenced at the rate of 0,25µg kg⁻¹ min⁻¹ and then rapidly reduced to 0,04-0,08µg kg⁻¹ min⁻¹ for the duration of the procedure, provided analgesia. Neuromuscular blockade was maintained by a rocuronium infusion and continuous train-of-four monitoring. An intra-arterial line and an oesophageal stethoscope were placed in addition to standard anaesthetic monitoring. Hydrocortisone was administered as the patient was receiving long-term oral dexamethasone therapy.

Surgical decompression of the spinal cord was achieved by a hemi-laminectomy of the first to the third cervical vertebrae and debulking of the tumour. Histology confirmed the tumour to be a cordoma.

Towards the end of the three-hour procedure the infusion of remifentanyl was reduced and discontinued. Neuromuscular blockade was reversed with neostigmine 2,5 mg and

glycopyrrolate 0,6 mg. After the nasotracheal tube was removed in the recovery room, oxygen administration was continued by facemask. Intravenous morphine was titrated to provide analgesia before transferring the patient to the intensive care unit (ICU) for an uneventful 48-hour stay (Figure 2). A full neurological assessment immediately after the operation demonstrated an increase of power in the upper limbs. After the operation the patient continued to improve and 14 months later was reported to be walking with near normal motor power.

Figure 2



Discussion

Wherever possible anaesthetic agents reported to have been used safely in patients with HC were utilized. Several authors reported the safe use of propofol for the induction and maintenance of anaesthesia in patients with HC.⁵⁻⁶ Similarly midazolam was reported as safe¹⁰⁻¹¹ although the possibility of increased sensitivity to the drug may exist.¹⁰ A wide variety of non-depolarizing neuromuscular blocking drugs have been used successfully in HC including atracurium⁶, pancuronium¹², mivacurium² and rocuronium.¹³ Although the use of remifentanyl in patients suffering from HC had not been reported at the time of this procedure, we had not expected any untoward reaction to the drug as virtually all the opiates including morphine¹⁴, pethidine^{2,6}, fentanyl^{10,11,13,15} and alfentanil¹⁶ have been used safely in HC. When selecting remifentanyl for analgesia, we considered its ultra short action and rapid recovery to have potential advantage in patients with HC where delayed recovery from anaesthesia had previously been reported.^{3,4} The rapid recovery after termination of the anaesthetic allowed for

the performance of the full neurological examination immediately after the operation.

Cholinesterase deficiencies including a higher incidence of the rare E₁^f gene associated with abnormal plasma cholinesterase¹⁵ have been reported with HC. This has been suggested as a possible cause of reports of prolonged duration of suxamethonium in HC patients. Although remifentanyl is broken down by cholinesterases, deficiencies per se do not appear to effect the duration of its effect¹⁵ and therefore the suspected higher incidence of plasma cholinesterase abnormalities associated with HC bears little relevance to its use.⁴

The whole concept of an abnormal sensitivity to certain anaesthetic drugs is being questioned in recent publications.^{2,17} There is a view is that the lowered plasma cholinesterase activity referred to above may in fact merely reflect the poor state of nutrition of patients with advanced HC as a result of feeding difficulties and difficulty in swallowing.¹⁷ In our patient there was no evidence of malnutrition and plasma albumen and total serum protein levels were normal.

Although the incidence of postoperative shivering that could give rise to dangerous clonic contractions in HC is reportedly lower with isoflurane than with halothane², great care was taken to ensure that the patient remained warm. A forced air warming device was used and no post operative shivering was encountered. The use of isoflurane as opposed to propofol is justifiable as no significant differences in postoperative shivering were found when using low concentrations of isoflurane, 0,6% end tidal, in comparison to using propofol, for maintenance of anaesthesia in combination with remifentanyl.¹⁸ Studies comparing combinations of remifentanyl with either propofol, as part of a total intravenous anaesthetic (TIVA) technique, or isoflurane, found them to be clinically equivalent in terms of emergence and recovery¹⁸ with no differences noted in the times to extubation and postanaesthesia recovery room discharge.¹⁹ Remifentanyl's effect on reducing the MAC of isoflurane has been demonstrated to be similar to that of other opiates.²⁰ The dosage requirements of remifentanyl in this case were comparatively low.

Finally, a comprehensive review of the anaesthetic management of patients with HC providing clear guidelines is well overdue. It would appear that many of the untoward reactions ascribed to HC in earlier publications may be due to non-specific secondary effects associated with the disease process. We therefore support the views expressed by Leng and Gupta¹⁵ in calling for an end to the myths related to anaesthetic drugs and HC.

Conclusion

Remifentanyl in this case used in combination with low concentrations of isoflurane provided a complication free and rapidly reversible alternative to TIVA techniques recommended for use in patients with Huntington's chorea.^{4,9,21} Precautions need to be taken to prevent intraoperative patient cooling if the serious complications associated with postoperative shivering in patients with Huntington's chorea are to be avoided. The possibility of an increased sensitivity to remifentanyl requiring reduced dosages should also be considered.

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Additional extracts

Early symptoms of Huntington's disease consist of the triad affecting emotion, cognitive ability and mobility. Symptoms and Signs include: depression, mood swings, forgetfulness, clumsiness, lack of coordination and involuntary twitching.

Huntington's Chorea is characterised by Hyperkinesia - abnormal, purposeless, involuntary motor repetitive or non-repetitive movements.

Patients suffer from motor dysfunction and progressive mental deterioration.

22. The optimum mode of intubation of a patient with an unstable spine is an awake, fibre-optic bronchoscope aided intubation, following topical anaesthesia and sedation, when indicated." Crosby HT, Lui ? *CASJ* 1990;37:1:77-93.