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CASE REPORT

Neuromuscular blockade in dystrophia myotonica with atracurium besylate

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Summary

The anaesthetic management of a young woman with dystrophia myotonica is described. The use of atracurium and monitoring of neuromuscular block allowed suxamethonium and neostigmine to be avoided, and thereby to reduce the risk of myotonic contractions. No adverse sequelae were observed.

Key words

Muscle relaxants; atracurium besylate.
Complications; Dystrophia myotonica.

Dystrophia myotonica is an inherited, autosomal dominant, disorder of muscle characterised by weakness, sustained contractures and wasting. Abnormalities in smooth and cardiac muscle and disorders of neurological and endocrine function are also reported.[1] It can present problems perioperatively for the anaesthetist due to sustained contractions which make intubation, ventilation and surgery difficult, or postoperatively when weakness and respiratory inadequacy may persist.[2]

We describe the anaesthetic management of a woman who had a history of a complicated postoperative recovery after previous anaesthesia, before she had been diagnosed as having dystrophia myotonica. She presented for laparoscopic sterilisation and was managed with atracurium and monitoring of neuromuscular function.

Case history

A 33-year-old woman (weighing 47 kg) with dystrophia myotonica, presented for laparoscopic sterilisation. In 1981, following treatment of infertility with clomiphene, she became pregnant and was admitted to another hospital suffering from polyhydramnios and hypertension. She was delivered of a floppy infant by Caesarean section. The general anaesthetic sequence was thiopentone, suxamethonium, tubocurarine and neostigmine with atropine. One hour postoperatively the nursing staff were concerned because she was noted to have 'muscular twitching' and ventilatory insufficiency with sputum retention. The infant was transferred to King's College Hospital but died 10 days later. Postmortem examination diagnosed infantile dystrophia myotonica. The mother was investigated and dys-

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trophia myotonica was diagnosed. She was warned about the hazards of future anaesthetics before leaving hospital.

On admission, she was found to have myopathic facies, weakness of the neck muscles and demonstrated the classical sustained grip upon handshake. Clinical examination revealed no gross cardiovascular or respiratory pathology, although an electrocardiogram (ECG) demonstrated right bundle branch block. She was, naturally, quite apprehensive, but premedication was avoided.

In the anaesthetic room continuous ECG, indirect blood pressure (by Dinamap, Critikon Corp.) recording was begun. Neuromuscular function was assessed by recording the combined electromyograph (EMG) in the hypothenar muscles (Relaxograph, Datex, Vickers Medical, Hampshire) (Fig. 1), after applying supramaximal stimuli to the ulnar nerve by surface electrodes. A train of four, 2 Hz stimuli, was given every 15 seconds. The control height (T0) was recorded while the patient was awake and the subsequent values (T1 and T4) based upon T0 being 100%.

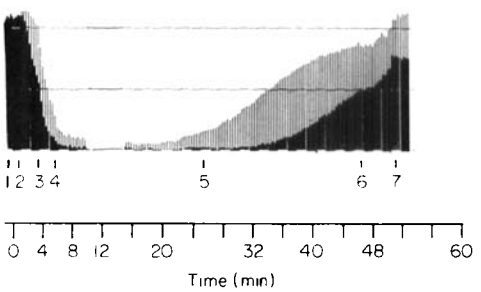

Fig. 1. Response to atracurium in a patient with dystrophia myotonica. Using the Datex 'Relaxograph', a train of four stimuli was applied every 15 seconds to the ulnar nerve and the effect recorded as the first and fourth EMG response in the hypothenar muscles. 1 = induction with thiopentone (3.2 mg/kg). 2, 3, 4 = atracurium 0.32 mg/kg as three equal incremental doses. 5 = termination of surgery, halothane discontinued. 6 = spontaneous ventilation. 7 = extubated, moving arm.

The patient was kept warm and an intravenous infusion of Ringer's lactate (at 37°C) was established. She was pre-oxygenated for 5 minutes and anaesthesia was induced by a small dose of thiopentone (3.2 mg/kg) and maintained with 70% nitrous oxide, oxygen and 0.5% halothane. Intubation and surgical relaxation were provided by atracurium besylate which was injected in 5 mg bolus doses (0.11 mg/kg) until T1 had decreased to 5%. She received a total dose of 15 mg (0.32 mg/kg) and intubation was completed without difficulty. Ventilation was readily controlled and maintained within the physiological range with a capnograph used to measure end tidal CO_2 concentrations.

At the completion of the uneventful surgery, which lasted 20 minutes (5 in Fig. 1), the halothane vaporizer was turned off and artificial ventilation of her lungs was continued until recovery of neuromuscular function was observed (6 in Fig. 1), and the nitrous oxide was then discontinued. After 46 minutes, 80% of T1 was noted and the patient breathed adequately. Neostigmine was therefore unnecessary. Five minutes later (7 in Fig. 1), she was fully awake, able to sustain a straight leg raise to 80° and was extubated. The cough reflex was good and brisk. At no time did we observe sustained contractions in the hypothenar or adductor pollicis muscles.

Her postoperative course was uncomplicated and she left hospital the following day.

Discussion

The anaesthetic management of patients with dystrophia myotonica requires special consideration of neuromuscular, respiratory and cardiac function before, during and after surgery. A characteristic of the disease is the development of sustained contractions in skeletal muscle which can be severe and generalised. These not only make surgery difficult but tracheal intubation and artificial ventilation become impossible. The contractions may be elicited by stimuli which can be mechanical (manipulation and voluntary contractions), electrical (nerve stimulation), chemical (hyperkalaemia) or hypothermic (shivering).[3]

The use of neuromuscular blocking agents in patients with neuromuscular disease has recently been reviewed:[2] suxamethonium or anticholinesterase agents may produce sustained contractures[4–6] and suxamethonium may also cause a transient hyperkalaemia. The non-depolarising relaxants appear to act normally, but even minimal residual weakness (insignificant in normal health) may contribute to postoperative respiratory inadequacy.

The myotonic patient tends to develop apnoea in response to respiratory depressants, especially opiates and benzodiazepines, and barbiturates,

including thiopentone;[7] volatile agents have also been implicated. Undue myocardial depression may result from incautious use of volatile agents.

In this case, known respiratory depressants (opiates and benzodiazepines) and the known 'trigger' agents (suxamethonium) were avoided. A small dose of thiopentone (150 mg) induced anaesthesia without apnoea. Halothane (0.5%) was preferred to the use of an opiate; this may be effective in decreasing the possibility of mechanically induced contractions. Dantrolene sodium, which acts distal to the motor end plate within the muscle fibres, directly produces muscular relaxation, but has provided inadequate relaxation for surgery in dystrophia myotonia when used alone.[8] In this case intubation was easily achieved after atracurium, obviating the need to use suxamethonium. The rapid spontaneous recovery ensured that neostigmine could be avoided. The recovery time (Fig. 1), T1 25% to T1 75% was approximately 12 minutes, which was within the normal range (unpublished data). It was interesting that contractures were not observed clinically or by EMG in response to repetitive nerve stimulation.

In conclusion, we consider that a neuromuscular blocking agent of intermediate duration is the logical choice for relaxation in an adult patient with dystrophia myotonica. In babies, however, local or regional anaesthesia may be preferable.[9]

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