DYSTROPHIA MYOTONICA AND ATRACURIUM
A Case Report

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Dystrophia myotonica is the most serious and the most common of the myotonic syndromes, with a prevalence of 5 per 100,000 (Gardner-Medwin, 1980). Inheritance is autosomal dominant. It is characterized by myotonia, especially when cold, and progressive muscular atrophy affecting predominantly the face, neck, pharynx and distal limbs. It is a systemic disease associated with cataracts, frontal balding, gonadal atrophy and cranio-facial abnormalities. Cardiac arrhythmias and cardiomyopathy are common (Cannon, 1962). Pulmonary reserve is often severely diminished and death is usually attributable to cardiorespiratory failure in middle age. Patients are often apathetic, hypersomnolent and of low intelligence. Other abnormalities include altered gut motility—especially difficulty with swallowing (Pierce, Creamer and MacDermot, 1965), various endocrinopathies and hypogammaglobulinaemia as a result of the excess catabolism of IgG (Walton and Gardner-Medwin, 1981).

Many of these abnormalities may be present before the onset of muscle symptoms and diagnosis (de Backer et al., 1976). Patients presenting with systemic manifestations requiring surgery are at particular risk (Kaufman, 1960). Patients with dystrophia myotonica may be sensitive to non-depolarizing neuromuscular blocking agents, and the use of an anticholinesterase drug to antagonize any residual blockade should be avoided (Mitchell, Ali and Savarese, 1978; Buzello, Krieg and Schlickewei, 1982).

SUMMARY

Atracurium was used to produce neuromuscular blockade in a patient with dystrophia myotonica. Operating conditions were excellent and myotonia was not seen. Sensitivity to atracurium was not seen and spontaneous recovery was not prolonged.

Atracurium is a neuromuscular blocking agent with a rapid elimination half-life which may obviate the need for reversal of residual neuromuscular blockade at the end of surgery. It is characterized by cardiovascular stability at normal doses (Payne and Hughes, 1981). These properties suggest that atracurium could be a valuable agent for use in patients with dystrophia myotonica.

There are no reported cases of the use of atracurium in this disease and, hence, we felt that it would be instructive to detail the action of atracurium in such a patient.

CASE REPORT

A 21-year-old girl weighing 55 kg requested sterilization. Dystrophia myotonica had been diagnosed 3 years previously, at which time she was said to have had severe disease for her age. The patient was admitted for assessment 2 days before operation.

At interview she presented a typical picture of dystrophia myotonica with ptosis, facial weakness and slurred speech. She denied all symptoms of cardiorespiratory disease. There was some difficulty with swallowing, but no symptoms of regurgitation.

Past medical history included appendicectomy and a further laparotomy when aged 12 yr. Details
of anaesthetics and postoperative course were not available. Three weeks previously the patient had been admitted with a severe attack of pelvic inflammatory disease requiring bed-rest and antibiotics.

The patient smoked occasionally, drank little alcohol and had no known allergies. Current drug therapy consisted of procainamide 250 mg 6-hourly on occasional days. Her severely affected younger sister had died in the postoperative period following an emergency laparotomy, only 2 weeks before this admission.

Examination revealed the typical features of dystrophia myotonica mentioned earlier. Of note was the weak distal musculature. Arterial pressure was 120/80 mmHg but heart rate was 100 beat min\(^{-1}\) and the pulse of poor volume. The peripheral circulation was poor. Auscultation of the chest showed generalized poor air entry, but no added sounds. The cough effort was noted to be weak.

The following special investigations were normal: full blood count, biochemical profile including liver enzymes, thyroid function tests, glucose tolerance test and blood-gas tensions breathing air. Chest X-ray, ECG, and echocardiogram were within normal limits. Abnormalities were found in serum creatine kinase (159 iu litre\(^{-1}\) (20-130)) and IgG concentrations (7 g litre\(^{-1}\) (8-16)).

Skull x-ray showed the typical small pituitary fossa and also bilateral subluxation of the temporo-mandibular joints. Diaphragmatic screening revealed inco-ordinate action during quiet breathing. Barium swallow revealed difficulty in initiation of swallowing and poor clearance of barium from the oesophagus, with absence of primary stripping waves. Audio EMG was diagnostic with the typical “dive-bomber” effect. Full scale IQ was 73. Pulmonary function tests are shown in table I. There was a 37% decrease in forced vital capacity (FVC) and a 31% decrease in forced expiratory volume in the first second (FEV\(_1\)). The FEV percentage (FEV\(_1\)/FVC) was 93%. These figures indicate a restrictive ventilatory defect with no evidence of airways obstruction. The patient was unable to perform estimations of residual volume (RV), alveolar volume (VA), specific airway conductance (s.Gaw) and transfer coefficient (Kco).

In view of the probability of a difficult laparoscopy as a result of adhesions and pelvic inflammatory disease the patient’s request for general anaesthesia was accepted.

**Anaesthesia**

No premedication was given. On arrival in theatre the patient was anxious with tachycardia and cool moist palms. A cannula was placed in a vein on the dorsum of the hand. Gel pad electrodes were placed for the continuous monitoring of ECG, frontalis muscle EMG, modified EEG and neuromuscular transmission (NMT). An automatic arterial pressure recorder (Dinamap) was attached to the arm opposite the NMT electrodes.

After baseline measurements of arterial pressure and heart rate had been obtained, anaesthesia was induced with fentanyl 50 ug and thiopentone i.v. All drugs were flushed through with saline. Thiopentone 200 mg was given slowly until apnoea occurred. Ventilation was then controlled using a Magill attachment and face mask. Nitrous oxide 70% in oxygen was given whilst control measurements of NMT were made.

Neuromuscular transmission was monitored continuously using train-of-four stimulation. The ulnar nerve was stimulated at the wrist and the evoked compound muscle action potentials were detected over the adductor pollicis. Four supra-maximal stimuli at a frequency of 2 Hz and pulse duration 0.1 ms were applied every 20 s. The evoked compound muscle action potentials were sensed and analysed automatically (Datex ABM: Anaesthesia and Brain Monitor). The ratios of the first twitch to the control twitch (T1:T0) and of the fourth twitch to the first twitch (T4:T1) were displayed graphically and digitally. A continuous printout was obtained using an Epsom dot matrix printer.

Atracurium 5 mg i.v. was given. After 5 min, pharyngeal secretions were suctioned, but intubation was not possible. This manoeuvre caused an increase in frontalis muscle activity. Enflurane was added to the inspired gases and a further 5 mg of
atrocurium given i.v. Further suctioning was necessary 3 min later and a similar increase in frontalis muscle activity was seen. The inspired concentration of enflurane was increased, fentanyl 50 μg and thiopentone 50 mg were given i.v., and intubation was accomplished with ease.

Pulmonary ventilation using a Brompton Manley ventilator (tidal volume 10 ml kg⁻¹) was adjusted to keep $P_{br}CO_2$ between 4.8 and 5.2 kPa. Oesophageal temperature was maintained between 36.4 and 37°C. At laparoscopy, dense adhesions made visualization of the pelvic organs impossible and laparotomy was necessary. Fentanyl 50 μg and thiopentone 50 mg were given i.v. when skin incision caused an increase in frontalis EMG activity. Further increments of atracurium 2 mg were given i.v. when T1 had recovered to 20% of control. No further increments of atracurium were given after the application of the Filshie clips and spontaneous recovery from neuromuscular blockade was allowed. Enflurane was discontinued when T1 had recovered to 25% of control. When recovery from neuromuscular blockade was deemed adequate, ventilation was discontinued and spontaneous respiration soon ensued. When $P_{br}CO_2$ had stabilized at around 5.3 kPa, nitrous oxide was discontinued. Emergence was rapid, the patient lifting her head and attempting to remove the tracheal tube. Following extubation, the patient was transferred to the recovery room for observation. Shortly after transfer to the recovery room the patient was sitting up and vigorously refusing any oxygen therapy. Observations of arterial pressure, heart rate and respiratory rate remained stable and there was no clinical evidence of a change in muscle tone or power from pre-operative values.

Postoperative discomfort was treated with dihydrocodeine 25 mg i.m. Deep breathing and coughing were encouraged by nursing staff and physiotherapists. There were no postoperative respiratory problems and the patient was discharged on the 4th day after operation.

Figure 1 illustrates the course of events.
Neuromuscular transmission

Following each dose of atracurium, the "fade" pattern produced by non-depolarizing agents was seen. Five minutes following atracurium 5 mg, T1 was 50% and T4:T1 was 48%. Four minutes after a further 5 mg, T1 was 14%, T4:T1 was 0 and intubation was possible. Time to complete blockade of T1 was 10 min from the initial 5-mg dose. Two further increments of atracurium were necessary. The first was given 25 min after the second 5-mg dose. The second increment was given after a further 11 min. The time for T4:T1 to reach 75% of control after the last increment was 32 min. Spontaneous breathing returned when T4:T1 was 70% and extubation of the trachea was performed when T4:T1 was 78%.

Cardiovascular variables

The ECG showed sinus rhythm throughout. The preoperative tachycardia, presumably caused by marked anxiety, decreased rapidly following the induction of anaesthesia. There were increases in heart rate associated with intubation, skin incision and emergence. Arterial pressure was stable during surgery. An increase in arterial pressure was seen following induction and again at extubation, but not during intubation. The sharp decrease in arterial pressure shortly following skin incision was presumably the result of the rapid injection of fentanyl and thiopentone plus an increase in the inspired enflurane concentration.

DISCUSSION

Patients with dystrophia myotonica may be expected to be sensitive to non-depolarizing neuromuscular blocking drugs. Many reports, however, indicate a normal response (Dalai et al., 1972; Mitchell, Ali and Savarese, 1978). Certainly, these patients are sensitive to many of the drugs used in anaesthesia.

The rapid breakdown of atracurium by "Hofmann" elimination leads to a predictable recovery (Payne and Hughes, 1981), and was chosen with the hope of avoiding the use of neostigmine which may precipitate myotonia (Buzello, Krieg and Shlickewei, 1982).

Continuous neuromuscular monitoring enabled an accurate assessment of the degree of blockade and allowed observation of the rate of recovery. The type of monitoring enabled us to estimate that almost total recovery would have taken place by the end of surgery and we feel it is essential in the proper management of all cases of muscle dysfunction receiving neuromuscular blocking drugs. The ED₉₅ for atracurium is approximately 0.2 mg kg⁻¹, depending upon the technique used (Basta et al., 1982; Goudsouzian et al., 1983; Black et al., 1985). The patient described here developed total block for 15 min following the initial doses of atracurium, which amounted to 0.18 mg kg⁻¹.

Enflurane is known to potentiate the action of neuromuscular blocking agents (Fogdall and Miller, 1975) and was indicated as an inhalation adjunct, so that smaller doses of a neuromuscular blocker could be used. Despite the use of enflurane, there was no obvious increased sensitivity to atracurium.

It was not possible to utilize T1 to monitor recovery, as a result of changes in the baseline—a problem well recognized with EMG recording. However, it is known that T1 is a poor indicator of recovery from non-depolarizing blockade and that recovery of T4:T1 is a more sensitive index. Recovery of T4:T1 to 75% reliably indicates recovery of T1 to control values (Ali et al., 1981; Calvey et al., 1983). The time for recovery of T4:T1 to 75% following the last increment of atracurium was 32 min, which is a normal rate of recovery.

It is our clinical impression that the incidence of postoperative shivering is low with enflurane. Shivering, especially when associated with a decrease in temperature, is known to induce myotonia (Ravin, Newmark and Saviello, 1975). Additionally, the use of an inhalation agent enables the dose of any narcotic to be kept to a minimum, thus decreasing the possibility of postoperative respiratory depression.

The case reported suggests that patients with dystrophia myotonica retain a normal sensitivity to atracurium. In view of its rapid elimination, obviating the need for an anticholinesterase drug, it is felt that atracurium should be considered when neuromuscular blockade is required for such patients. In addition, we believe that NMT monitoring is an integral part of the anaesthetic management of these patients, and full supportive facilities must be available.

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REFERENCES