Response to vecuronium in a patient with facioscapulohumeral muscular dystrophy

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Increased sensitivity to vecuronium has been noted in patients with Duchenne muscular dystrophy. We report the response to vecuronium in a patient with facioscapulohumeral muscular dystrophy (FSHD), an autosomal dominant disorder with an incidence of 10–20 cases per million. In this patient, sensitivity to an initial dose of vecuronium (0.02 + 0.08 mg kg–1) was normal, but recovery was faster and the effect of incremental doses of vecuronium (0.02 mg kg–1) was less than expected. Onset time and 25% recovery of T1/T0 after the intubating dose of vecuronium were 240 s and 22 min, respectively. Recovery index (spontaneous recovery of T1/T0 from 25% to 75%) was 9 min.

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Facioscapulohumeral muscular dystrophy (FSHD) is an autosomal dominant disorder with an incidence of 10–20 cases per million.1 It is associated with deletions of variable size on chromosome 4q35.2 FSHD is characterized by weakness of the shoulder, pectoral and facial muscles, in particular the orbicularis oculi muscles.3 The pelvic muscles are much less affected than in Duchenne muscular dystrophy. Cardiac muscle is clinically uninvolved,4 although a relatively high susceptibility to atrial fibrillation or flutter together with less frequent conduction abnormalities have been reported during intra-cardiac electrophysiological studies.5 Onset usually occurs in adolescence and progression of the disease is slow. As the life span is reduced only minimally in most patients, FSHD is also known as a benign dystrophy. We report the response to vecuronium at the adductor pollicis muscle in a patient with FSHD.

Case report

A 25-yr-old man (165 cm, 56 kg) with hyperhidrosis was undergoing bilateral thoracoscopic sympathectomy. He presented with a 9-yr history of progressive muscle weakness in the face, shoulders and proximal part of the upper limbs. He had been diagnosed as suffering from FSHD. Physical examination revealed atrophy and weakness of the affected muscles and diminished deep tendon reflexes in the upper extremities. Muscle strength in the forearm and hand muscles was normal (5/5 in the manual muscle test, MMT). Needle electromyogram (EMG) revealed short duration and polyphasic motor units in affected muscles, which is consistent with the diagnosis. Laboratory studies included a creatine phosphokinase concentration of 375 iu litre–1 (normal range 47–195 iu litre –1). Chest x-ray and electrocardiogram were normal. Spirometry revealed a forced vital capacity of 2.35 litre (57% of predicted). He was not receiving any drugs, such as steroids or anticonvulsants.

He was premedicated with atropine 0.5 mg i.m. Before induction of anaesthesia and throughout surgery, neuromuscular transmission was monitored using a Relaxograph (Datex, Helsinki, Finland). The ulnar nerve was stimulated supramaximally (62 mA, train-of-four stimulation) at the wrist every 20 s and the EMG response of the adductor pollicis muscle was monitored. After the calibration period, a stable control EMG response was obtained. The forearm was wrapped in a cotton blanket to minimize cooling.

Anaesthesia was induced with propofol 2 mg kg–1, followed by a constant infusion of 10 mg kg–1 h–1. Vecuronium 0.1 mg kg–1 was then administered in divided doses. First, a small dose of vecuronium 0.02 mg kg–1 was given to test for exquisite sensitivity to vecuronium; no twitch depression was observed. Then vecuronium 0.08 mg kg–1 was given after a 6-min interval and complete neuromuscular block followed (onset time 240 s). The trachea was intubated and anaesthesia was maintained with a continuous infusion of propofol 4–6 mg kg–1 h–1 and fentanyl 50–100 µg h–1. Time from injection of vecuronium 0.1 mg kg–1 to return of T1/T0 to 25% was 22 min. Incremental doses of vecuronium 0.02 mg kg–1 were administered on eight occasions when T1/T0 returned to 25%. However, none of
these doses depressed T1/T0 to less than 10% and four doses did not depress it to less than 25%. Times to 25% recovery of T1/T0 after the four incremental doses which depressed the twitch response below that height were 9.0–14.3 min. Recovery index (spontaneous recovery of T1/T0 from 25% to 75%) was 9 min after the last dose of vecuronium had been given. Residual neuromuscular block was antagonized with neostigmine 2.5 mg and atropine 1.0 mg when T1/T0 and TOF ratio returned to 95% and 50%, respectively. TOF ratio exceeded 95% before the patient awoke, and the trachea was extubated.

Throughout this period, the ECG was normal, \( \text{SpO}_{2} \) was >98%, end-tidal \( \text{PCO}_2 \) was 4.0–4.7 kPa and rectal temperature was 36.7–37.2°C. The postoperative course was uneventful.

**Discussion**

Information on the response to non-depolarizing neuromuscular blocking drugs in patients with muscular dystrophy is mostly limited to those with Duchenne-type dystrophy. Normal sensitivity to gallamine and increased sensitivity to tubocurarine and vecuronium have been reported. We found only one English report on the use of neuromuscular blocking drugs in FSHD where atracurium was used. In this case, the response to atracurium was normal and recovery was rapid.

In our patient, time to 25% recovery after vecuronium 0.1 mg kg\(^{-1}\) was 22 min and recovery index after the last incremental dose of vecuronium was 9 min. These values are considerably shorter than those reported for normal patients under balanced anaesthesia (38.3 (SEM 1.9) min and 15.4 (1.6) min, respectively). Times to 25% recovery after incremental doses of vecuronium 0.02 mg kg\(^{-1}\) in this patient were in the range of 9.0–14.3 min. This is also shorter than the normal range (21.7–29.9 min) using the same dose of vecuronium. Although the intubating dose of vecuronium led to block, incremental doses never depressed T1/T0 to less than 10%. In normal patients, even a smaller dose of vecuronium 0.015 mg kg\(^{-1}\), administered when T1/T0 returned to 25%, has been shown to cause a block exceeding 90% (mean 93.8 (SEM 0.8)%). Thus in the FSHD case report of Dresner and Ali using atracurium, sensitivity to 2×ED95 of vecuronium appeared to be normal but duration of action and effect of incremental vecuronium doses were less than anticipated.

The site of EMG recording may be important in this type of patient. FSHD affects the proximal part of the upper limbs and MMT in our patient revealed normal muscle strength in the forearm and hand muscles (5/5) where we monitored the EMG. Thus the decreased effect of vecuronium was observed in a muscle with normal strength.

The mechanism of the decreased effect of vecuronium is unclear. Pathological changes in muscular dystrophy are believed to occur distal to the neuromuscular junction. However, the co-existence of neurogenic changes has been reported in FSHD patients who showed only myogenic findings on the EMG. Upgrading of acetylcholine receptors as a result of upper or lower motor neurone dysfunction is often regarded as a possible explanation of decreased sensitivity to non-depolarizing neuromuscular blocking drugs. Widespread subclinical neuropathy may have been responsible for the response to vecuronium in our patient.

In many muscle disorders, increased sensitivity to non-depolarizing neuromuscular blocking drugs is common. The short duration of action of vecuronium in this patient with FSHD is noteworthy.

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