EXAGGERATED PHYSIOLOGICAL RESPONSES TO PROPOFOL IN MYOTONIC DYSTROPHY

Sir,—Dr Speedy's case report [1] is at variance with our experiences of propofol in myotonic dystrophy [2]. We used continuous infusions of propofol and atracurium during a 4-h oral surgical procedure; total doses were 2488 mg and 75 mg, respectively. There were no intraoperative problems and operating conditions were excellent. Emergence from anaesthesia was rapid (10 min until eye-opening) and there were no postoperative problems. We have since anaesthetized the same patient for an abdominal hysterectomy and another patient for cataract extraction, using the same technique, without perioperative problems.

Use of a neuromuscular blocking drug may have prevented coughing and ocular myotonia, necessitating the use of a volatile agent with potent vasodilator properties. The hypotensive episode noted by Dr Speedy appears to be related to the introduction of 1% isoflurane, and resolved on reducing the inspired concentration. We have noted that propofol and isoflurane have an additive effect on arterial pressure in normal patients.

Dr Speedy correctly states that hypothermia and recovery from volatile anaesthesia may induce shivering, a potent cause of myotonia and the reason why we avoided volatile agents. Total i.v. anaesthesia using etomidate (although no longer recommended for infusions) has been reported previously in patients with myotonic dystrophy [3].

D. A. WHITE
D. G. SMYTH
Ely

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SIR,—The differences in response to propofol between the patient described by Drs White and Smyth [1] and my patient [2] are interesting, but not surprising. Patients may present with a wide variation in severity of disease from asymptomatic to severely disabled. It is apparent that, while some patients respond more or less normally to a variety of drugs, others show a very abnormal response to the same agents.

The patient described by Drs White and Smyth could walk 100 m and, unless exceptionally tall, had a normal weight and spirometry values. They make no comment on their patient's cerebral status, so presumably it was unremarkable. In contrast, my patient had severe limitation of mobility, marked loss of muscle mass and a forced vital capacity less than 50% of predicted. Furthermore, he suffered from severe lethargy and daytime somnolence and his family had noticed a marked deterioration in mental alertness over the preceding year. Although I did not formally assess psychometric state, the patient had marked impairment of cerebral function, presumably because of degeneration. The differences between our patients in physical and mental status would appear to be marked, and may explain the longer time required for my patient to recover normal ventilatory function and consciousness.

As our patients were different, it is perhaps not surprising that we chose different techniques. I chose to use isoflurane as a means of reducing muscle tone and ensuring immobility without recourse to neuromuscular blockers [3]. Isoflurane has the added advantage of being eliminated more rapidly than i.v. anaesthetic agents and non-depolarizing neuromuscular blockers. I accepted the risk of postoperative shivering which is less with isoflurane than with other volatile agents [4]. In the event, neither shivering nor myotonia occurred after operation. I would agree with Drs White and Smyth that hypotension was an additive effect with propofol; 1% isoflurane was clearly more than was required in this patient after initial loading, and the problem resolved when the inspired concentration was reduced to 0.5%.

If this patient presented again for similar surgery, I would modify my technique in the light of the experience already gained: avoid all i.v. anaesthetic agents and use an inhalation induction with rapidly eliminated gases or vapours; use atracurium, titrated carefully to effect to facilitate tracheal intubation and ensure intraoperative immobility (I have been impressed by other authors' reports of using this drug successfully in myotonic subjects without the need to reverse paralysis with neostigmine [5]); use nitrous oxide administered by controlled ventilation for anaesthetic maintenance, and the minimum amount of isoflurane to ensure adequate depth of anaesthesia and satisfactory operating conditions.

I would make no recommendations on the anaesthetic technique for any other patient with this disease, as the severity of its manifestations is so variable.

H. M. SPEEDY
Leeds

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