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Continuous propofol anaesthesia for patients with myotonic dystrophy

M. Bennun[1], B. Goldstein[1], Y. Finkelstein[2] and R. Jedeikin[1]*

[1]Department of Anaesthesiology and Critical Care Medicine, Meir Hospital, 44281, Kfar Saba, Sackler School of Medicine, Tel-Aviv University, Tel-Aviv, Israel, and [2]Department of Otolaryngology, Head and Neck Surgery, Meir Hospital, 44281, Kfar Saba, Sackler School of Medicine, Tel-Aviv University, Tel-Aviv, Israel

**Corresponding author*

Myotonic dystrophy, a rare genetic disorder, may pose a serious problem to the anaesthesiologist due to muscular and extramuscular involvement. Thirteen patients, median age 21 yr were anaesthetized by continuous propofol infusion, fentanyl, atracurium and N_2O to evaluate this combination in myotonic dystrophy. Intraoperatively, neither exaggerated reactions nor haemodynamic instability was observed. Recovery was smooth and quick. Although there was a significant decrease in mean postoperative vital capacity (965 (349) ml) from the preoperative value (1664 (566) ml, P=0.0028), there was no change in mean postoperative Sp_{O_2} and there were no perioperative respiratory complications. Only two patients complained of nausea and vomiting. Similarly, muscular hypertonia and shivering were not observed. We conclude that the combination of continuous propofol infusion and fentanyl was a successful anaesthetic technique in these young myotonic dystrophy patients undergoing peripheral surgery.

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Myotonic dystrophy, an autosomal dominant disorder, first described by Steinert in 1909, is the most common of the myotonic syndromes with a prevalence of three to five per 100 000.[1 2] Although a rare congenital form of the disease exists, in most patients the onset is between the second and fourth decades of life. Myotonic dystrophy is characterized by myotonia (incomplete muscle relaxation), marked wasting of the muscles of mastication, neck, pharynx and distal limbs, ptosis, frontal baldness, low intelligence and multisystem involvement. Extramuscular features include cataracts, cardiomyopathy, conduction abnormalities, restrictive lung disease, central and obstructive sleep apnoea syndrome, dysphagia, delayed gastric emptying and endocrine abnormalities such as hypothyroidism, primary hypogonadism, infertility and diabetes mellitus.[2]

Anaesthetic management of these patients is challenging and may pose a serious problem to the anaesthesiologist. Myotonia may be precipitated by hypothermia, shivering, and mechanical or electrical stimulation. Furthermore, sensitivity to sedative, anaesthetic and neuromuscular blocking agents may result in intraoperative and early postoperative cardiovascular and respiratory complications, as well as prolonged recovery from anaesthesia. We therefore performed a study to assess the adequacy of an anaesthetic regimen based on i.v. propofol in myotonic dystrophy patients undergoing oropharyngeal and neck surgery.

Patients and methods

Between 1993 and 1997, 13 patients with myotonic dystrophy scheduled for oropharyngeal and neck surgery were enrolled in an open prospective study. All patients complained of peripheral muscle weakness, which started at a median age of 6 yr (range 3–35) but were able to function adequately, with good physical mobility and without handicap. There was no history of systemic disease and none was taking medication. Physical examination showed wasting of thenar and masseteric muscles. Preoperative haematological and biochemical investigations, ECG, echocardiograpy and lung function tests were normal.

The study was approved by the Institutional Ethics Committee and consent was obtained from all patients or their parents. No pre-anaesthetic medication was adminis-

tered. Prior to induction of anaesthesia, with the patient in the supine position, vital capacity and tidal volume were measured using an Ohmeda Modulus II anaesthetic machine pneumotachograph with a tight-fitting face mask. Intraoperative monitoring included electrocardiogram, non-invasive arterial pressure, pulse oximeter, capnograph, nerve stimulator and rectal temperature. The highest and lowest intraoperative arterial pressure and heart rate were documented. Anaesthesia was induced with fentanyl 0.05 mg, propofol 2.5 mg kg^{-1} and atracurium 0.5 mg kg^{-1}. After intubation of the trachea, the patients' lungs were ventilated with 70% N_2O in oxygen and anaesthesia was maintained with a continuous infusion of propofol 6 mg kg^{-1} h^{-1}, bolus fentanyl 2 μg kg^{-1} and incremental doses of atracurium 0.2 mg kg^{-1}. After patients had regained consciousness and four equal 'train of four' twitches were observed, the trachea was extubated. A 20% or greater decrease in systolic blood pressure was treated with i.v. ephedrine (5 mg).

Duration of anaesthesia (start of anaesthesia–extubation), recovery time (end of surgery–extubation), post-extubation tidal volume and vital capacity were recorded. Patients were admitted for 2–3 h to the post-anaesthetic care unit (PACU) prior to being discharged to the ward. On admission to PACU, ECG, NIBP, Sp_{O_2} as well as the occurrence of postoperative nausea and vomiting were recorded. Postoperative respiratory and cardiac complications and length of hospital admission were noted.

The mean highest and lowest intraoperative heart rate, systolic blood pressure, post-extubation tidal volume and vital capacity, and lowest postoperative Sp_{O_2}were compared to baseline values and analysed using the Mann–Whitney U-test. A P-value of <0.05 was considered significant.

Results

The following surgical procedures were performed: pharyngeal flap for correction of velopharyngeal incompetence (eight patients), pharyngeal flap and tonsillectomy (two patients), endoscopic surgery of the sinuses (FESS) for pansinusitis (one patient), cervical lymph node biopsy (one patient) and total thyroidectomy for papillary carcinoma of thyroid (one patient). The median age was 21 yr (range: 11–42 yr). The mean (SD) duration of anaesthesia was 104 (44) min, and the mean (SD) recovery time was 12 (11.5) min. The intraoperative highest and lowest systolic blood pressures were significantly different from the baseline (135 (14.5), 100 (18.7) versus 122 (8)) (P=0.01, P<0.001). On the other hand, only the lowest intraoperative heart rate differed significantly from the baseline (64.7 (14.2) versus 78.2 (13)) (P=0.01). Ephedrine was not required in any case. The mean postoperative Sp_{O_2} (97 (2.9)) did not differ from the preoperative (98 (1)) values. The median hospital admission was 2 days (range: 1–4 days).

There was no difference between mean preoperative and postoperative tidal volumes (334 (119) ml versus 330 (122) ml) (P=0.97). However, there was a significant decrease in mean postoperative vital capacity (965 (349) ml) from the preoperative value (1664 (566) ml) (P=0.0028). There were no perioperative respiratory or cardiac complications and bronchial secretions were not a problem. Only two patients complained of nausea and vomiting, but no treatment was required. Similarly, muscular hypertonia and shivering were not observed.

Discussion

Because of the marked sensitivity of myotonic dystrophy patients to sedative and anaesthetic agents and the increased risk of precipitating generalized myotonia by cold, shivering and drugs, the safest anaesthetic technique has still to be established. Dundee and thereafter Bourke initially used thiopental as an induction agent for myotonic dystrophy patients.[3 4] Both reported thiopental to induce prolonged apnoea and respiratory depression. However, later studies failed to confirm these reports.[6 7] In a recent retrospective study, 152 out of 219 myotonic dystrophy patients received thiopental without adverse effects.[5] These conflicting reports suggest that the prolonged apnoea attributed to thiopental may be due to an idiosyncratic peripheral effect and central nervous system depression commonly associated with the development of the disease.

Etomidate was used successfully in a myotonic dystrophy patient undergoing maxillo-facial surgery.[8] However, because etomidate has been shown to cause adrenal suppression, its use by continuous infusion is now questioned.[9]

The use of propofol as an induction and maintenance agent in myotonic dystrophy is controversial. Milligan was the first to report the uneventful use of propofol as an induction agent in myotonic dystrophy,[10] while White and Smyth administered a continuous infusion of propofol in a case undergoing prolonged oral surgery without mishap.[11] Speedy found that less than 1 mg kg^{-1} of propofol was sufficient to induce hypnosis and permit tracheal intubation.[12] However, when isoflurane was added to maintain anaesthesia, the patient developed severe intraoperative hypotension and prolonged postoperative unconsciousness and apnoea. Similarly, in a patient in whom a target-controlled propofol infusion system was used, recovery was prolonged for 1 h after the completion of surgery.[13] A generalized myotonic state was also reported after the i.v. administration of propofol.[14] As with thiopental, the cause of these variable exaggerated physiological responses to propofol can be associated with the severity of the disease.

Inhalation agents may be deleterious in patients suffering from myotonic dystrophy and should be used with caution. Deep inhalation anaesthesia may further risk the already compromised cardiovascular and respiratory systems of these patients, while postoperative shivering may precipitate myotonia. However, large concentrations of volatile

anaesthetics have been shown to resolve myotonic contractions albeit at the expense of cardiorespiratory depression.[2]

Neuromuscular blocking drugs pose a particular problem in myotonic dystrophy. These patients are particularly sensitive to succinylcholine. Succinylcholine has an initial stimulating action on muscle.[15 16] This may result in the development of myotonia leading to difficulty in intubation and ventilation. Therefore, the use of succinylcholine is not recommended in myotonic dystrophy patients.[2 7 17]

In contrast, the non-depolarizing neuromuscular blocking drugs usually evoke a normal response. However, if muscle wasting exists, a prolonged response may occur necessitating the reversal of the muscle relaxant. The anticholinesterase drugs used for this purpose may also precipitate myotonia, presumably a result of increased sensitivity to the stimulatory effect of acetylcholine.[18 19] Although neostigmine has been found to be safe in some instances,[16 20] it would be prudent to rather use short-acting non-depolarizing muscle relaxants which do not require antagonism. In this context, atracurium, an intermediate-acting muscle relaxant, which is metabolized by Hoffman degradation may be the most suitable. Spontaneous recovery from atracurium has been shown to occur in under an hour.[17 21–23]

We found the combination of a continuous propofol infusion, fentanyl, atracurium and nitrous oxide in oxygen to be successful for patients undergoing oropharyngeal and neck surgery. Exaggerated reactions to drugs and myotonia were not observed. The recovery period was smooth and relatively short (12 min), and only two patients had nausea and vomiting. There were no serious postoperative respiratory or cardiac complications.

Anaesthetic and surgical complication rates range between 8.2 and 42.9%.[5–7] The stable intraoperative period, uneventful recovery and lack of postoperative complications in our study may be explained by two factors. Firstly, our patients were young (median age 21 yr) and only had minor symptoms of the disease. Myotonic dystrophy is a slowly progressing multi-system disease with death occurring usually in the sixth decade of life.[17] Secondly, the surgical procedures in this study were peripheral. The prevalence of postoperative pulmonary complications has been reported to be as high as 38.1% after upper abdominal surgery in patients with myotonic dystrophy.[5] However, in none of these cases was propofol used as the main anaesthetic. In contrast, White and Smyth reported an uneventful perioperative period in a patient anaesthetized using a continuous infusion of propofol for abdominal hysterectomy.[11] Therefore, the effect of propofol-based anaesthesia on the surgical outcome in myotonic dystrophy patients undergoing major surgery needs further clarification.

In conclusion, the combination of propofol infusion, fentanyl, and atracurium with nitrous oxide in oxygen appears to be a suitable anaesthetic technique for young myotonic dystrophy patients undergoing peripheral surgery. Further studies are required to evaluate this anaesthetic technique in elderly patients suffering from myotonic dystrophy as well as those undergoing major surgery.

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