Anaesthesia for Charcot-Marie-Tooth disease: a review of 86 cases

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Operative charts were reviewed in 86 patients with Charcot-Marie-Tooth disease, a condition characterized by chronic muscular denervation. A total of 161 surgical procedures was performed. Major complications were few, and only one operative death occurred, unrelated to anaesthesia. Succinylcholine and malignant hyperthermia triggering agents were used in 41 (48%) and 77 (90%) patients, respectively, without untoward effects. Contrary to previous reports, this survey supports the safe use of succinylcholine and MH triggering agents in this disease.

La maladie de Charcot-Marie-Tooth se caractérise par une dénervation musculaire chronique. Les dossiers anesthésiques de 86 patients atteints de cette maladie ont été revus. Les patients avaient subi un total de 161 interventions chirurgicales. Il y à eu peu de complications majeures, et un seul décès périopératoire sans relation avec l'anesthésie. La succinylcholine a été utilisée chez 41 (48%) patients et d'autres agents susceptibles d'induire une hyperthermie maligne ont servi pour anesthésier 77 (90%) patients. Aucun effet indésirable dû à ces médicaments n'a été observé. Contrairement à d'autres observations cliniques, cette étude rétrospective suggère que la succinylcholine et les agents inducteurs d'hyperthermie maligne peuvent être utilisés sans augmenter le risque de complication chez les patients atteints de cette maladie.

Key words
ANAESTHESIA:
MUSCLE: denervation;
SYNDROMES: Charcot-Marie-Tooth disease.

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Charcot-Marie-Tooth disease (CMTD), also known as hereditary motor and sensory neuropathy, is a rare neurological condition that affects peripheral nerves, primarily of the distal musculature.[1,2] Denervation with subsequent muscular atrophy is the hallmark of this disease. There are only five reports of anaesthesia in patients with CMTD,[3–7] with some raising the possibility of succinylcholine-(SCH) induced hyperkalaemia, and one[4] suggesting that these patients may be susceptible to malignant hyperthermia (MH). This report summarizes the anaesthetic management of 86 patients with CMTD, many of whom received SCH and MH triggering agents safely.

Methods

Questionnaires and medical release forms were sent to approximately 1,000 members of Charcot-Marie-Tooth International, an organization dedicated to helping patients with CMTD. Questions included age, anaesthetic/surgical history, dates of diagnosis and onset of symptoms, severity of disease and history of familial problems with anaesthesia and surgery. Most patients could not remember the disease severity adequately at the time of each procedure, so current severity is reported.

Medical records (anaesthetic record, discharge summary, history and physical, recovery room record and preoperative anaesthetic evaluation) were obtained and information regarding surgery, type of anaesthesia and complications was extracted. Complete records could not be obtained on all of the respondents; however, no patient was included if an anaesthetic record was unavailable or if the surgical procedure preceded the onset of symptoms.

Results

One hundred ninety-three questionnaires were returned. For several reasons (foreign hospital, surgery in remote past, incomplete records, etc.), medical records were obtained on only 86 of these patients, with 161 surgical procedures performed. For each procedure the age was 41 ± 17 yr (mean ± SD), range 2–75 yr; duration of symptoms was 23 ± 14 yr, range 0–59 yr. Sixty-nine of 86 patients (80%) had symptoms in all extremities.

The majority (53%) of the procedures were orthopaedic.

Other surgery included peripheral (17%), intra-abdominal (7%), obstetrical/gynaecological (7%), and miscellaneous (17%). General anaesthesia was used for 139 procedures in 78 patients, while regional and local anaesthesia were used for 22 procedures in 18 patients. Malignant hyperthermia triggering agents (succinylcholine and/or potent inhalational anaesthetics) were given to 77 (90%) patients for 130 procedures.

Succinylcholine was used during 56 operations in 41 (48%) patients. Pre-treatment with a "defasciculating" muscle relaxant was used in 32 of these 56 exposures. A paralyzing dose of a nondepolarizing agent was used during 50 episodes in 39 (45%) patients: of these, 26 (30%) had pharmacological reversal.

Complications included 19 (22%) patients who complained of "weakness" postoperatively, one unexpected admission to the intensive care unit secondary to postoperative hypotension, and two patients who developed pneumonia, one of whom died. There were no other deaths reported by family members. No complications occurred as a result of muscle relaxants, i.e., objective weakness, prolonged intubation or reintubation.

Discussion

Anaesthesia in these CMTD patients appeared to be tolerated well with few complications. The only perioperative death was in a young girl who had severe restrictive lung disease and developed pneumonia subsequent to spinal fusion. Other complications were relatively minor. However, this survey is necessarily biased because information was obtained from patients who were able to respond, or from their parents. Since CMTD tends to be familial, this bias was minimized by requesting information regarding any family members who had had problems with surgery and anaesthesia. Nonetheless, complications and deaths may have been under-reported. Also, this survey was retrospective, with sometimes incomplete data obtained from differing institutions. Such a data source could have obscured important trends in perioperative morbidity and mortality in CMTD.

Patients with CMTD have chronic denervation, often of all extremities. Since denervation is one of the most potent predisposing factors for release of potassium after exposure to SCH,[8,9] previous reports have cautioned against its use in CMTD. This survey indicates that SCH is well tolerated. While a small "defasciculating" dose of a nondepolarizing muscle relaxant may lessen the potassium release from diseased muscle,[8] patients who had SCH alone had no apparent problems.

The massive release of potassium resulting from SCH is not an all-or-none phenomenon. A group of patients susceptible to this complication develop varying degrees of hyperkalaemia;[10] some may manifest subtle ECG changes, some may develop peaked T waves, while others can have malignant arrhythmias and cardiovascular collapse. If CMTD patients were sensitive to the hyperkalaemic effect of SCH, some ECG and haemodynamic changes should have been found. Their absence indicates that SCH is probably safe in CMTD. However, because the plasma potassium concentration was not measured in these patients, the true risk is unknown. In addition, any acute exacerbation in CMTD may alter the amount of involved muscle and therefore change the sensitivity to SCH.

The range of age and symptom duration was wide, indicating that the degree of chronicity was not an important influence on outcome. Also, symptom severity was not a factor. Nearly 80% of all patients had involvement of all extremities. Thus, regardless of whether a patient has had recent onset of symptoms or has had long-standing disease, SCH is well tolerated. Presumably, the process of denervation is much slower than the atrophic process, such that the amount of muscle which can release potassium is relatively small.

Very few patients developed pulmonary complications, i.e., pneumonia. In the past, pulmonary involvement in CMTD was thought to be uncommon. However, more recent data suggest that respiratory muscles may be affected, with a restrictive lung pattern predominating.[11,12] The patient of the report by Brian *et al.*[3] was ventilator-dependent for approximately one month following a Caesarean section. Involvement of the phrenic nerve[13] and the nerves subserving expiratory muscles[14] may have been responsible. Patients may have few or no symptoms despite considerable abnormalities in pulmonary function. The presence of proximal muscle weakness of the arms may be a predictor for respiratory muscle weakness.[11]

Theoretically, muscle weakness related to loss of motor units might sensitize a patient to nondepolarizing muscle relaxants. In this survey, however, no patient appeared to have any complications vis-a-vis muscle relaxants, i.e., prolonged block. This possible complication was probably adequately evaluated, as the nerves which are used clinically to monitor neuromuscular function may be affected by CMTD, including the posterior tibial, ulnar and facial nerves.

One report has raised the issue of MH.[4] While several neuromuscular diseases are associated with MH, based on our understanding of the pathophysiology of CMTD and MH, there is no reason to suspect that a connection between the two exists. Most patients received MH triggering agents without untoward effects. However, the relatively small sample size of this survey does not exclude a potential link.

The CMTD patients evaluated in this survey appeared to tolerate anaesthesia well. Although SCH has been

considered to be contraindicated in this disease, no complications occurred from its use in these patients who had chronic symptoms. An acute exacerbation, however, might render SCH use inadvisable. Other risks, including sensitivity to neuromuscular blocking agents and MH, are probably minimal. The possibility of occult pulmonary dysfunction should be considered, but, in general, the anaesthetic management in CMTD can be adjusted to the needs of the individual patient.

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