ANAESTHESIA AND HUNTINGTON'S CHOREA
A report of two cases

J. FARINA AND L. A. RAUSCHER

SUMMARY
Two patients with Huntington's chorea were anaesthetized successfully with a technique including thiopentone, nitrous oxide and either a narcotic analgesic or halothane; pancuronium was employed for myoneural blockade.

Huntington's chorea is an hereditary disorder of the nervous system, characterized by progressive dysarthria, ataxia, choreiform movements and dementia. The incidence is approximately 6 : 100 000. The disease is inherited as a single Mendelian dominant autosomal gene. Symptoms appear first between 30 and 45 years of age. There is degeneration of the forebrain and corpus striatum. Histologically, the degeneration is most marked in the putamen, but extensive cellular loss occurs also in the caudate nucleus and cerebral hemispheres. The progress of the disease is variable, but most of the patients die between 10 and 15 years after the onset of symptoms. At present there is no known therapy for reversing the disease or halting its progression. Therapy is directed primarily at decreasing the choreiform movements. The drugs used include haloperidol, diazepam, reserpine, lithium, L-dopa and chlordiapoxide.

A review of the literature revealed only two reports of anaesthesia for patients with Huntington's chorea. The first, based on experience in only one patient, suggested that such patients may be unduly sensitive to barbiturates (Davies, 1966). The second report, also based on experience with only one patient, suggested that there may be abnormal pseudo-cholinesterase activity producing prolonged apnoea after the administration of suxamethonium (Gualandi and Bonfanti, 1968).

The following report describes two different approaches to the anaesthetic management of patients with Huntington's chorea.

CASE REPORTS

Patient 1
A 48-yr-old male was admitted for multiple dental extractions. The patient had suffered progressive ataxia for 5 yr, choreiform movements and mental deterioration, and at the time of admission was completely bedridden, unable to communicate and demented. Treatment included phenobarbitone 30 mg three times a day, chlordiazepoxide 25 mg twice a day and diazepam 10 mg four times a day. The patient's father had died of Huntington's chorea.

Physical examination on admission revealed a disoriented, 48-yr-old male with gross choreiform movements. There were a few coarse râles bilaterally on chest auscultation. There were no abnormalities in the chest x-ray or e.c.g., or in routine haematological and serum biochemical studies.

The patient was prepared for surgery with chest physiotherapy and pulmonary toilet, and on the third day in hospital underwent anaesthesia. Premedication was with atropine 0.5 mg, droperidol 2.5 mg and fentanyl 150 ìg i.m. Anaesthesia was induced with thiopentone 100 mg i.v. followed by pancuronium 4 mg i.v. and tracheal intubation. Anaesthesia was maintained with nitrous oxide 60% and halothane 1% in oxygen. At the end of the procedure, neuro muscular paralysis was antagonized with neostigmine 2.5 mg with atropine 1.2 mg i.v. The duration of anaesthesia was 75 min. Spontaneous respiration returned within 2 min and the tracheal tube was removed without difficulty. The course after operation was uneventful, and the patient was discharged on the 3rd hospital day.

Patient 2
A 53-yr-old female was undergoing exploratory laparotomy because of a pelvic mass. At age 38 yr she first noted abnormalities of speech and gait. The patient had suffered progressive dysarthria, increased choreiform movements and, at admission, was confined mostly to bed with limited periods in a wheelchair. Her mental status was difficult to ascertain because of extreme difficulties of communication. The drug therapy consisted of diazepam 10 mg, orally
thrice daily. The patient’s father had died of Huntington’s chorea.

On admission the patient had gross choreiform movements. Auscultation of the chest revealed bilateral basilar râles and diffuse, coarse rhonchi. The chest x-ray, e.c.g., and routine haematological and serum biochemical measurements were normal.

Chest physiotherapy and vigorous pulmonary toilet were instituted several days before operation. The patient was premedicated with diazepam 10 mg i.m.

Anaesthesia was induced with the i.v. administration of 400 μg fentanyl in four equally divided doses. At this point choreiform movements stopped, and breathing was regular at a rate of 12 b.p.m. Diazepam 15 mg i.v. was followed by pancuronium 4 mg i.v. and an endotracheal tube was inserted. Anaesthesia was maintained with 67% nitrous oxide and 33% oxygen. The duration of anaesthesia was 2 h. The total doses were fentanyl 600 μg, diazepam 20 mg and pancuronium 6 mg. Neuromuscular paralysis was antagonized with the i.v. administration of neostigmine 2.5 mg with atropine 1.2 mg i.v. Spontaneous breathing returned normally and the endotracheal tube was removed without incident.

The course after operation was complicated by a low grade fever (maximum rectal temperature 39.2 °C), which was thought to be secondary to pneumonia or bronchitis. This was treated successfully with ampicillin. Auscultatory findings did not change from those found before operation and the chest x-ray remained normal.

DISCUSSION

Davies (1966) suggested that patients with Huntington’s chorea might be abnormally sensitive to barbiturates. He described one patient who was anaesthetized on three occasions within 25 days. The first anaesthetic consisted of premedication with papaveretum and hyoscine, induction with thiopentone and maintenance with nitrous oxide and trichloroethylene with gallamine for myoneural blockade. The return of spontaneous respiration was greatly retarded and the period immediately after operation was complicated by episodes of generalized tonic spasms and deep cyanosis. The second anaesthetic was similar to the first but gallamine was not given; similar postoperative respiratory depression, tonic spasms and episodes of cyanosis were encountered. The third anaesthetic was very brief and consisted of premedication with papaveretum and hyoscine, induction and maintenance with nitrous oxide and diethyl ether. Recovery from anaesthesia was normal on this occasion.

Gualandi and Bonfanti (1968) suggested that patients with Huntington’s chorea might be unduly sensitive to depolarizing muscle relaxants. They reported one patient who was anaesthetized twice in one week. On the first occasion, the anaesthetic consisted of premedication with pethidine, atropine and haloperidol, induction with droperidol and maintenance with fentanyl and nitrous oxide. Recovery after operation was normal. On the second occasion, the anaesthetic management was similar to the first except for the addition of suxamethonium for muscle relaxation. Following this anaesthetic, the patient was apnoeic for 2 h but subsequently recovered normally. No mention was made in this report of any familial incidence of abnormal pseudocholinesterase in this patient’s relatives.

Our two case reports describe the successful management of myoneural block with pancuronium and of anaesthesia with nitrous oxide–narcotic and nitrous oxide–halothane techniques. These drugs should be able to provide all the anaesthetic requirements for patients with Huntington’s chorea who require surgery. We present this report in the hope of stimulating further reports to enlarge the known experience of anaesthesia in these patients.

REFERENCES


ANESTHESIE ET CHOREE D'HUNTINGTON

Rapport sur deux cas

RESUME

On a anesthesié avec succès deux malades atteintes de choree d'Huntington (danse de Saint-Guy) grâce à une technique comprenant du thiopentone, du protoxyde d'azote et soit un narcotique analgésique, soit de l'halothane; le pancuronium a été utilisé pour le blocage myoneural.

ANÄSTHESIE UND HUNTINGTON'S CHOREA

Zwei Krankengeschichten

ZUSAMMENFASSUNG

Zwei Patienten mit Huntington's Chorea wurden erfolgreich narkotisiert, unter Verwendung einer Methode mit Thiopent, Stickoxyd und entweder einem narkotischen Analgeticum oder Halothan. Für myoneurale Blockierung wurde Pancuronium verwendet.

ANESTESIA Y COREA DE HUNTINGTON

Un informe de dos casos

SUMARIO

Se anestesizaron con éxito dos pacientes que sufrían de corea de Huntington mediante una técnica que incluyó tiopentona, óxido nitroso, y ya fuera un analgésico narcótico o halotano; se empleó pancuronio para el bloqueo mio neural.
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References
Tinker, J.H., and Milchenfelder, J.D., Anesthesiology, 1978, 45, No. 3, 340

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