

ABNORMAL RESPONSE TO ANAESTHESIA IN A CASE OF HUNTINGTON'S CHOREA

A Case Report

BY

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SUMMARY

A man, aged 33, suffering from Huntington's chorea, showed an abnormal response to anaesthesia on two different occasions. Prolonged apnoea followed induction. Recovery was slow and complicated by generalized tonic spasm. The abnormal response was probably associated with the neurological disorder. Thiopentone was thought to be mainly responsible for these complications.

Huntington's chorea is a somewhat rare hereditary disease in which there is a slow progressive degeneration of the nerve-cells of the basal ganglia and of the cerebral cortex. It is inherited as a Mendelian dominant and therefore approximately half the family are liable to acquire it. The sexes are equally affected. The onset of the disease is usually relatively late, at about the age of 35 or 40 years.

The first mental symptoms are forgetfulness and restlessness. Physical symptoms, which usually occur at the same time but may occasionally be delayed in their onset, are involuntary spontaneous movements usually of the face, head and hands, ataxia (which may be marked) and slow, slurring articulation. Over the course of years, there is progressive mental failure, often with a suicidal tendency, leading to dementia and death in, perhaps, ten years or more.

There do not appear to have been any reports concerning the response of these patients to general anaesthesia, and the following, the only case of Huntington's chorea anaesthetized by the author, occurred some years ago.

CASE REPORT

The patient, a male aged 33, of average build, gave a history of loss of memory and was seen to have well marked involuntary movements, choreiform in type, and a grossly ataxic gait related to these movements. Family history revealed that his father had developed involuntary movements about the age of 38 and had steadily deteriorated with loss of memory and had died at the age of 48. (It was later elicited that a sister also suffered from the same condition.) The patient

was seen at the Central Middlesex Hospital by Dr. R. J. Porter and Dr. Otho FitzGerald who considered that the patient was, without doubt, suffering from Huntington's chorea with, perhaps, a superadded anxiety state.

Some months later the patient was admitted to the Central Middlesex Hospital following an injury to the hand. He was transferred to the theatre for operation 8 hours after admission. Papaveretum 20 mg and hyoscine 0.45 mg was given 30 minutes before operation. On arrival at the theatre the patient was awake, respiration appeared undepressed and his general condition was quite good. Anaesthesia was induced with thiopentone 600 mg followed by gallamine 60 mg and maintained with nitrous oxide, oxygen and trichloroethylene; he was not intubated. Following induction of anaesthesia the patient did not breathe spontaneously for 60 minutes. Fifty minutes after induction, an intravenous injection of neostigmine 2.5 mg, preceded by atropine 0.6 mg, was given without any effect and then followed by nikethamide 2 ml also without immediate effect. Spontaneous respiration commenced, however, some 2 or 3 minutes after this latter injection. Breathing was at first shallow but was not unduly slow. Over the next 12 minutes ventilation improved considerably and was then considered to be adequate. During this 12-minute period, nalorphine 7 mg was given, but this did not seem to have any very obvious effect. Recovery of consciousness was delayed and during the recovery period the patient suffered from generalized tonic spasms with a tightly clenched jaw producing, each time, deep cyanosis.

Fifteen days later, the patient underwent a further operation on his hand. Premedication was again papaveretum 20 mg and hyoscine 0.45 mg given 30 minutes before operation. Anaesthesia was induced with thiopentone 400 mg and maintained with nitrous oxide, oxygen and trichloroethylene. Muscle relaxants were not given. Following induction of anaesthesia the patient did not breathe spontaneously for 12 minutes. Spontaneous respiration, when it recommenced, was at first shallow but soon improved; breathing was not unduly slow. The recovery period was again prolonged and complicated by occasions of deep cyanosis due to generalized tonic spasm.

The patient underwent a third and final operation for change of dressings 25 days after the date of admission. Premedication was papaveretum 20 mg and hyoscine 0.45 mg and anaesthesia was induced and maintained with nitrous oxide, oxygen and ether alone. The course of the anaesthesia, which was of relatively short duration, was uneventful. The recovery period was also uneventful.

DISCUSSION

The first two anaesthetics, but not the third, were complicated by prolonged apnoea following induction, and by prolonged recovery periods during which generalized tonic spasms occurred. The premedication given to the patient on the three occasions must be regarded as heavy, and the dose of thiopentone administered in the first anaesthetic must be regarded as large. In a man of average build, however, one would not expect any untoward effects from such dosages and it must be assumed that the patient exhibited an abnormal response to one or more anaesthetic drugs possibly directly as a result of the fact that he was, at the time, suffering from Huntington's chorea.

It is possible that thiopentone is mainly implicated as the drug which actually occasioned the episodes of apnoea and the stormy recovery periods. Apnoea followed immediately after its injection in the first two instances; the third anaesthetic, in which thiopentone had not been administered, was not associated with any such complications. The picture is, however, confused by the fact that an opiate had been given before each of the anaesthetics but, although it is probable that the opiate played some contributory part in producing the anaesthetic complications, it is doubtful if it played a significant part in view of the facts that, at the time of induction, the patient did not appear to have been greatly depressed by the opiate, that administration of nalorphine during the time of respiratory inadequacy did not improve the ventilation, and that the third anaesthetic, in which opiate had been administered but thiopentone omitted was, as stated, not complicated by any obvious ventilatory depression, or by difficulty in the recovery period. Gallamine was administered during the first anaesthetic, but probably played only an insignificant role in the causation of apnoea; the dose was relatively small and apnoea was not termin-

ated by the administration of an anticholinesterase.

The incidence of Huntington's chorea throughout the country is not exactly known, but has been investigated throughout Northamptonshire (determined originally by Pleydell (1954, 1955), but amended later by Reid (1960) and has been found to be 7.2 cases per 100,000 population. If the incidence throughout the country is comparable then, as the likelihood of such patients having to undergo surgery is probably the same as that of average individuals in the community, it will mean that, possibly over 100 such patients are anaesthetized in this country every year. Whether many of these respond abnormally to anaesthesia is at present not known, but in anaesthetizing such cases it would seem prudent to administer only a light premedication and to keep the dose of thiopentone to the absolute minimum, or to omit it altogether.

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REPONSE ANORMALE A L'ANESTHESIE DANS UN CAS DE CHOREE DE HUNTINGTON

SOMMAIRE

Un homme de 33 ans souffrant d'une chorée d'Huntington a présenté une réponse anormale à l'anesthésie en deux circonstances différentes. Une apnée prolongée a suivi l'induction. Le réveil a été lent et compliqué par des spasmes toniques généralisés. La réponse anormale était probablement associée aux troubles neurologiques. On pense que la thiopentone est principalement responsable de ces complications.

ABNORME NARKOSEREAKTION BEI EINEM FALL VON HUNTINGTONSCHER CHOREA

ZUSAMMENFASSUNG

Bei zwei verschiedenen Gelegenheiten wurde bei einem 33-jährigen Mann, der an einer Huntingtonschen Chorea leidet, eine abnorme Narkosereaktion beobachtet. Nach der Narkoseeinleitung trat eine verlängerte Apnoe auf. Er erholte sich nur langsam und als Komplikation kam es zu generalisierten tonischen Krämpfen. Die abnorme Reaktion stand möglicherweise mit der neurologischen Erkrankung in Zusammenhang. Als Hauptursache für diese Komplikation wurde das Thiopenton angesehen.

AN UNUSUAL CASE OF LIVE FISH OBSTRUCTING THE AIRWAY

A Case Report

BY

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SUMMARY

A fisherman was brought to hospital with airway obstruction due to a live fish. It proved possible to pass an endotracheal tube, after which the fish was removed, thus saving the life of the patient.

The causes of obstruction of the airway are many and the following is an account of an unusual and interesting case of airway obstruction due to a live fish, which would have caused asphyxiation by its wriggling but for its prompt removal.

CASE REPORT

A male patient, aged 25 years, was brought to the operation theatre on the afternoon of January 15, 1964, choking and fighting for breath.

The history elicited from his friends was that the patient was a fisherman and he was fishing with an angle, which consisted of a line and a hook. After catching one fish he unhooked it from the barb and kept it between his teeth. The fish slipped, wriggled, and made its way into the mouth and oropharynx. Before the patient realized what was happening he found he could not breathe. He was brought to the hospital within a few minutes of the accident. On examination he was cyanosed, anxious, fighting for breath, but co-operative.

Without delay, and within approximately 10 minutes of the accident, laryngoscopy was attempted. The cephalic end of the fish was found in the hypopharynx with its tail fin just visible over the dorsum of the tongue. Its size prevented it from entering the oesophagus or trachea fully. In this position the body of the fish was found to be pressing upon the larynx (fig. 1), obstructing the air entry into it. Attempts to remove the fish, by catching the tail with a Kocher's forceps was futile as its side fins were by now embedded in the pharyngeal wall. By depressing the body of the fish backwards, however, the larynx could be visualized. The patient got immediate relief by this procedure. A plain endotracheal tube (Magill No. 6) was introduced into the trachea and the lungs filled with 100 per cent oxygen. After disappearance of the cyanosis and to facilitate removal of the fish, a sleep dose of thiopentone was given, and anaesthesia was maintained by nitrous oxide and oxygen. The general condition of the patient was good. With considerable difficulty the whole fish, which measured 9 cm, was removed piecemeal. A prophylactic

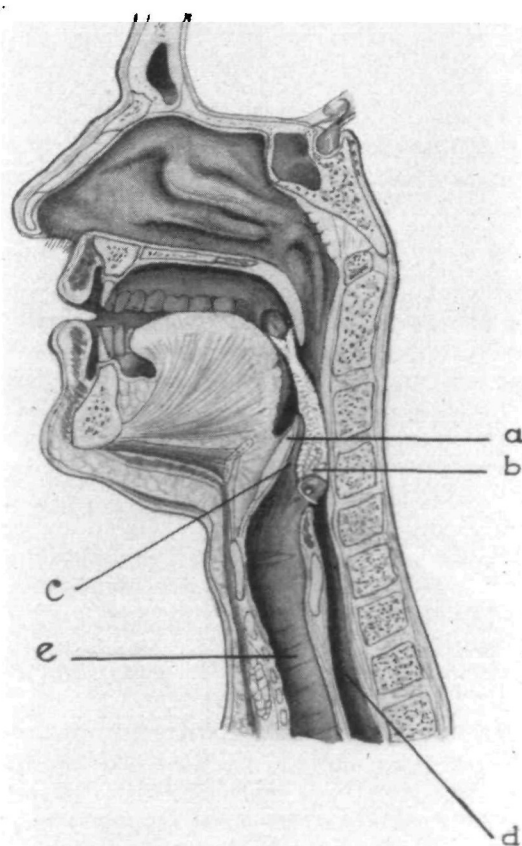


FIG. 1

Compression of the airway by the fish.

(a) Epiglottis. (c) Aryepiglottic fold.
(b) Fish. (d) Oesophagus.

(e) Trachea.

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tracheostomy was done in case cellulitis of the pharynx developed as a result of laceration of the pharyngeal mucosa by the spine-like lateral fins of the fish, and it was retained for three days.

He was given a course of antibiotics and was discharged after an uneventful postoperative period.

DISCUSSION

This peculiar habit of keeping fish between the teeth is quite common amongst the fishermen on our coastal areas. To our knowledge there has been no previous report of the successful management of such an accident. Although there have been few cases with a similar history, autopsy has revealed small fish in the trachea or main bronchus.

Under similar circumstances life can be saved if the other fishermen realize what is happening and have the presence of mind to transport him immediately to the nearest hospital. It will then depend upon the speed with which a patent airway is established.

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UN CAS EXTRAORDINAIRE DE POISSON VIVANT OBSTRUANT LES VOIES RESPIRATOIRES

SOMMAIRE

Un pêcheur a été amené à l'hôpital pour une obstruction des voies respiratoires due à un poisson vivant. Il a été possible de passer une sonde endotrachéale après quoi le poisson a été enlevé.

EIN UNGEWÖHNLICHER FALL VON ATEMWEGSVERLEGUNG DURCH EINEN LEBENDEN FISCH

ZUSAMMENFASSUNG

Ein Fischer wurde mit einer Verlegung der Atemwege durch einen lebenden Fisch in das Krankenhaus gebracht. Die Einführung eines Endotrachealtubus erwies sich als möglich, worauf der Fisch entfernt werden konnte.