both ankles where xanthomas had been removed, with some loss of superficial parts of the Achilles tendons. A xanthoma had recurred at the lower end of the left patellar tendon. There were no xanthomas elsewhere. The lower end of the right radius was thickened. Other physical appearances were normal. Examination of the nervous system showed a tremor of the outstretched left hand and mild inco-ordination of all limbs, more pronounced on the left. All deep reflexes were brisk, with bilateral finger flexion; plantar reflexes were normal. Sensation appeared normal and gait was unsteady. On lumbar puncture C.S.F. was slightly hazy with 60 mg of protein and no cells.

X-ray examination showed a Smith-Petersen nail in the right femoral neck and an old fracture at the lower end of the right radius. All the bones were somewhat porotic. Lung fields were within normal limits. Results of blood counts and standard biochemical tests were normal. Fasting serum cholesterol levels were 295, 288, and 247 mg/100 ml (method of Abell *et al.*, 1952), and triglyceride was 160 mg/100 ml. Paper electrophoresis showed mildly increased betalipoproteins and prebetalipoproteins. Betalipoprotein cholesterol was 202 mg/100 ml. E.C.G. showed slight sag of the ST segment in lateral leads.

Comment

The combination of mental defect, possibly of late onset, with cerebellar ataxia, increased deep reflexes, juvenile cataracts, and massive tendon xanthomas with minimally raised serum cholesterol is characteristic of cerebrotendinous xanthomatosis. This condition was first noted by van Bogaert in 1937 (Phillipart and van Bogaert, 1969). About eight cases have been described since then (Schimstock *et al.*, 1968).

The dementia can present in late childhood or in adult life. Serum cholesterol levels have been normal or transiently

raised. Most of the deaths have been from gradual pseudobulbar paralysis, and one man aged 58 died of myocardial infarction. At necropsy there have been granulomatous lesions and demyelination in the cerebellar white matter lateral to the dentate nuclei, with less severe similar lesions elsewhere in the central nervous system. The disease appears to be inherited as a Mendelian recessive, as in the present case. Accumulation of free and esterified cholestanol—that is, cholesterol without its 5, 6 double bond—is the specific biochemical lesion in the nervous system (Menkes *et al.*, 1968; Phillipart and van Bogaert, 1969), where the saturated sterol makes up a quarter to half of the total sterols. In tendon xanthomas and in serum, however, the percentage of cholestanol is much lower and may (Phillipart and van Bogaert, 1969) or may not (Menkes *et al.*, 1968) be of diagnostic value during life.

We would like to thank Dr. A. B. Daneel, medical superintendent, and Dr. H. E. Bernstein, of the Alexandra Institution, and Dr. Des Shapiro for their help. Dr. E. Lawton performed the ophthalmological procedure, and Dr. F. Ames the neurological examination.

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Prolonged Coma after Tetanus

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British Medical Journal, 1972, **1**, 354-355

Tetanus still carries a considerable morbidity and mortality (Weinberg, 1964), particularly when it is severe and occurs in older subjects (Kerr *et al.*, 1968). We report the case of an elderly woman with severe tetanus successfully treated with pancuronium (a short-acting curare-like drug) and massive doses of diazepam. There were no serious side effects but she remained unconscious for 12 days after treatment was stopped.

Case History

A 72-year-old spinster with no relevant past medical history collapsed in her garden on the evening of 25 October 1970. She was found the next morning somewhat confused, dirty, and with multiple lacerations affecting particularly the knees and the right big toe. She was admitted to the surgical ward of another hospital where the wounds were cleaned and excised and the right big toe was amputated.

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Eight days later she developed trismus. She was given penicillin and tetanus antitoxin before transfer to this hospital. On arrival, about three hours after the onset of trismus, she was also noted to have stiffness of the neck muscles but apart from obesity (weight 79 kg) appeared fit for her age and was mentally fully alert. She was given more antitoxin (100,000 units intravenously) and started on a course of parenteral penicillin. Shortly afterwards she felt nauseated, vomited once, and had a major spasm with respiratory arrest. She was immediately intubated and an emergency tracheostomy was performed. Her wounds were opened and all suspicious tissue was excised. Tetanus bacilli were cultured from debris removed from the right foot.

After tracheostomy she was ventilated artificially, paralyzed with pancuronium, and heavily sedated with diazepam. On the first day she received 220 mg of diazepam intravenously, on the second day 815 mg, and overall a mean dose of 70 mg daily. This dosage did not produce any recognizable side effects and her blood pressure remained stable throughout (110-130/80 mm Hg). During the first two or three days her condition deteriorated and she developed generalized rigidity, with severe spasms as often as every 20 minutes and minor spasms each time she was touched, despite 50-80 mg of pancuronium daily.

Her further course was complicated by bronchospasm, bronchopneumonia, urinary infection, mild renal and cardiac failure, and a deep vein thrombosis. These were treated appropriately.

Twenty-one days after her admission the major spasms had stopped, though she remained rigid and had spasms if touched. The pancuronium was stopped and she was maintained on diazepam. She was weaned off the ventilator gradually until she was breathing completely unaided nine days later. Diazepam had been stopped 28 days after the onset of trismus. She remained rigid and unconscious for a further 12 days though capable of spontaneous respiration through the tracheostomy, and only then made some response to commands such as "open your mouth" or "move your fingers." The muscular stiffness persisted and passive movements of the limbs were made difficult by pronounced cog-wheel rigidity. There was little evidence of any further improvement in mental function

until a week later, when on provocation she muttered two expletives. Thereafter she was mobilized and made satisfactory progress. The tracheostomy was closed and it subsequently healed. Four weeks after stopping the diazepam it was clear that her mental faculties were unimpaired and that she retained a remarkably good sense of humour.

She was discharged home three months after her admission walking well, thinking clearly, and with her original wounds almost healed. When seen as an outpatient two months after discharge she was completely cured and had no appreciable physical, haematological, or biochemical defect. She was living alone and managing her home without difficulty.

Comment

Tetanus is a serious disease, the mortality of which was as high as 70% before Lassen *et al.* (1954) described the use of total paralysis and intermittent positive-pressure ventilation.

This case is of particular interest because of the huge doses of diazepam used and the prolonged recovery time. Even when a patient is satisfactorily paralyzed with curare-like drugs direct stimulation of a nerve at operation will provoke a muscular contraction. Diazepam is a centrally-acting muscle relaxant and seems a reasonable drug to use to combat the effects of tetanus. Despite doses of pancuronium which allowed us to ventilate the patient artificially and controlled most of her spasms, she continued to have intermittent serious spasms. Faced with the choice of increasing either the pancuronium or the diazepam we chose the latter. Our reasons

were that pancuronium may have cardiovascular side effects (Kelman and Kennedy, 1971), while diazepam is largely free from side effects in high doses. At the time we did not anticipate any long-term effects of diazepam.

We suggest that the prolonged delay in regaining consciousness may be accounted for by the cumulative body stores in this obese patient. Fortunately there were no mental or neurological sequelae to this severe attack of tetanus and the 40 days of therapeutic "unconsciousness," despite her age and the possibility of cerebrovascular disease.

In further cases of tetanus treated with large doses of diazepam the possibility of a similar prolonged recovery phase of cerebral activity should be borne in mind and meanwhile other supportive treatment continued.

We thank Professor Sir W. Melville Arnott for permission to report this case, Dr. C. G. Burgess, of Roche products, for his help and co-operation, and members of the department of anaesthesia and the intensive care unit for their help with the patient.

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Pulmonary Embolectomy by Flying Squad

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British Medical Journal, 1972, **1**, 355-356

Massive pulmonary embolism is a grave surgical emergency. In most peripheral hospitals, however, facilities for pulmonary embolectomy with cardiopulmonary bypass are not available. Reported below are two cases treated successfully by a flying squad which brought both the surgeon and the bypass apparatus to the patient.

Case 1

A 34-year-old housewife taking a contraceptive pill with a high oestrogen content had been confined to bed for several weeks with a respiratory tract infection. During that time her left calf became swollen and tender and one week before admission she had a sudden onset of severe dyspnoea, substernal pain, and palpitations lasting 20 minutes. Seven days later she had woken up severely dyspnoeic and was admitted to hospital.

On examination she was found to be anxious, pale, and sweaty and had severe dyspnoea and peripheral cyanosis. The blood pressure was not recordable by auscultation but by palpation the systolic pressure was 70 mm Hg. The pulse rate was 150/min and regular. The jugular venous pressure was not raised but there was a pronounced parasternal heave and a right ventricular triple rhythm. Heparin and warfarin were immediately given but five hours after admission her condition remained critical. The

systolic blood pressure was 100 mm Hg by palpation, the jugular venous pressure was raised to the angle of the jaw, and a loud friction rub was heard over a large area of the precordium.

Eight hours after admission pulmonary embolectomy was carried out under cardiopulmonary bypass by a surgeon who had come with the bypass apparatus from the nearest thoracic surgical centre 40 miles (64 km) away. At operation both the main pulmonary trunk and the right ventricle were tense and scarcely pulsating, and masses of clot were removed from both main pulmonary arteries and their branches. There was no evidence of femoral vein thrombosis on either side on exploration. The patient recovered well and remained in good health. There were no residual symptoms or signs referable to her chest or cardiovascular system.

Case 2

A 61-year-old woman had a vagotomy and gastroenterostomy for a duodenal ulcer. Seven days later she was transferred from a nursing home to hospital because of the onset of severe breathlessness. On admission she was dyspnoeic, sweaty, and pale. The blood pressure was 100/60 mm Hg and the pulse regular at 110/min. The jugular venous pressure was raised to the ear lobes and there was a pronounced parasternal heave and a right ventricular triple rhythm. An electrocardiogram showed complete right bundle-branch block. X-ray examination of the chest showed no definite abnormality. Four hours later pulmonary embolectomy was carried out under cardiopulmonary bypass, the surgeon and the apparatus again coming from the regional thoracic surgical centre. A large, pencil-like clot 9 to 12 in (22.5 to 30 cm) long was removed from the main pulmonary artery and small pieces of clot were removed from each main branch. Neither femoral vein seemed to contain clot. She recovered well and remained in good health.

Comment

Reviewing 271 cases of death from massive pulmonary embolism, Donaldson *et al.* (1963) concluded that about one in five patients suffering from massive fatal embolism may

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