tacosis CFT titre remained high. Liver enzymes became normal; WBC remained normal; brain stem evoked potential inter-peak latency I-V decreased slightly to 4.8 ms. The patient was discharged after 25 days in King Faisal Specialist Hospital, able to walk independently but with difficulty.

On follow up the patient gradually returned to normal; mild subjective dysaesthesia in the feet persisted at one year. Psittacosis CFT titres on eight persons at risk in the household were negative. The parrot was sacrificed and disposed of without necropsy.

The criteria for the diagnosis of psittacosis are met fully by the history of exposure to a parrot, onset with pulmonary symptoms, a compatible chest radiograph, normal WBC, high serum CFT titres, and a dramatic response to the appropriate antibiotic after failure of potent but non-specific antibiotics. Unfortunately confirmation by necropsy of the bird was denied.

It is known that psittacosis affects many organs¹ including liver,² heart,³ kidneys,⁴ skeletal muscles⁵ and the CNS.^{6,7} The neurological signs in our patient were those of brain stem involvement, the nature of which remains speculative in the absence of pathological confirmation. One may argue that this is a form of central pontine myelinolysis caused by nutritional derangement or fluctuating electrolytes during the early days of the illness. But the CT scan did not show the unenhancing radiolucency usually seen in this condition;⁸ the near complete recovery argues against pontine myelinolysis which is usually fatal or has significant neurologic sequelae.⁹ The time was too short for a previously healthy patient to have been depleted of thiamine, and no significant hyponatraemia or rapid electrolyte correction had occurred during his illness.

The psittacosis surveillance reported by the United States Center for disease control in the 1970s showed that CNS manifestations occurred in 5–9% of cases.¹⁰ Symptoms are variable. Encephalitis,⁷ polyradiculitis¹¹ and cerebellar symptoms have all been reported in psittacosis, and in other infections such as *Mycoplasma pneumoniae*,^{12,13} legionellosis¹⁴ and mononucleosis.¹⁵ Neuropathologic study of cases of "psittacosis encephalopathy" showed mainly vascular changes including congestion, hyalin thrombi and perivascular monocyctic infiltrate with small foci or necrosis or demyelination.^{6,7} Invasion of the CNS as evidenced by the presence of intracytoplasmic inclusions (LCL bodies) had been found only rarely in cases of meningitis.¹⁶ The term brain stem encephalitis

was first used by Bickerstaffe¹⁷ to describe neurological manifestations predominantly referable to the brain stem and occurring in the course of an acute or subacute infection. The term had been suggested to include a variety of symptoms, such as those of Fisher's syndrome.¹⁸ The non-specificity of the initial infection suggests that the underlying pathophysiology involves the immune system or causes changes in the blood vessels which lead to the symptoms of brain stem involvement. The absence of an immunological event is not inconsistent with the diagnosis as concluded by Waxman *et al*¹⁹ who reported subacute brain stem encephalitis leading to respiratory death without an obvious antecedent infection, immunological event or neoplastic disease. In our case, intense activation of the immune system early in the course of the infection may have led to a focal vasculitis involving the brain stem. Although vascular changes have been reported in psittacosis, the cause of preferential involvement of the stem vasculature requires further study.

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Relapsing Fisher's syndrome

Sir: Fisher's syndrome, characterised by the triad of ophthalmoplegia, areflexia, and ataxia,^{1–2} is considered by many neurologists to be a variant of the Guillain-Barré syndrome. Approximately 3% of patients with Guillain-Barré syndrome will have one or more relapses,³ but the relapse rate for Fisher's syndrome is unknown. We wish to report a patient with a relapsing illness which fits the classical description of Fisher's syndrome.

A 58-year-old woman was first evaluated in November 1977 for the complaints of dizziness and unsteadiness. She had a 'flu-like illness four weeks previously, but was otherwise in good health. Her past history was unremarkable for prior neurological illness. The unsteadiness increased over the ensuing two days, and paraesthesias of the feet and horizontal diplopia developed. General examination, including vital signs, was normal. On neurological examination she was anxious. The visual fields were normal. The pupils were 5–6 mm and reacted minimally to light, although they did not constrict during attempted convergence. Horizontal gaze and upgaze was absent, and on attempted downgaze the eyes would move only a few degrees. There was no

Letters

ptosis; Bell's phenomenon and the oculoccephalic response were absent. The remainder of the cranial nerves were normal. Gait was ataxic, but there was no limb ataxia. There was no weakness, but muscle stretch reflexes were unobtainable. Vibratory and position sense were minimally decreased in the distal lower extremities. The following tests were normal: blood glucose, BUN, serum electrolytes, liver function tests, complete blood count, anti-nuclear factor, rheumatoid factor, monospot, protein electrophoresis, urinalysis, VDRL, vitamin B₁₂ and folate levels. Lumbar puncture revealed CSF under normal pressure with 4 wbc/mm³, a protein of 53 mg/dl, and glucose of 73 mg/dl. Microbiological and cytological examinations were normal. 10 mg of intravenous tensonil did not improve the ocular motility, nor did 100 mg of thiamine. CT with and without contrast was normal. By the second hospital day her eyes were immobile. Over the next week she became less ataxic, and minimal horizontal ocular movements were present. Her ocular motility improved daily, so that over the ensuing week her oculomotor functions returned to normal, first in the horizontal plane, followed by vertical movements. Her ataxia resolved, but she remained areflexic. On ophthalmological examination 8 weeks later extraocular movements were normal, and the pupils were 4 mm and briskly reactive to light. She was evaluated twice a year by her internist who noted no abnormalities on examination; muscle stretch reflexes were present. Repeat examination by the ophthalmologist in 1982 was normal.

In May 1983 the patient developed a viral-like syndrome with upper respiratory tract congestion, nausea, and diarrhoea. Two weeks later she developed gait ataxia and diplopia. She became quite alarmed and sought medical attention stating "it's happening to me again". General medical examination, including vital signs, was normal. Visual fields were intact and optic discs were flat with spontaneous venous pulsations. The pupils were 5 mm and reacted minimally to light; 2 mm ptosis of the left lid was present. The eyes were midline; vertical gaze was absent and the only movements horizontally were abducting nystagmoid jerks in the direction of gaze. Bell's phenomenon and the oculoccephalic reflex were absent. The remainder of the cranial nerves were normal. There was no weakness, abnormal movements, or limb ataxia. Gait was wide-based and ataxic. Muscle stretch reflexes were absent in the lower extremities and hypoaactive in the upper extremities and

there was no Babinski reflex. Proprioception was minimally reduced in both feet. The following laboratory tests were normal: complete blood count, anti-nuclear factor, BUN, VDRL, serum electrolytes, liver function tests, blood glucose, monospot, cold agglutinin titre, vitamin B₁₂, folate, thyroid function tests (T₃, T₄), urinalysis, urinary porphyrin assay, and heavy metal analysis of the urine. CSF was under normal pressure and was acellular with a protein of 55 mg/dl and glucose 89 mg/dl. Cytological and microbiological examinations were normal. CT with and without contrast with brainstem views was normal. A Tensonil test was negative, and intravenous thiamine (100 mg) failed to improve her ocular motility. The ophthalmologist who evaluated her in 1977 confirmed that the ocular findings were the same as during that hospitalisation. Electromyographic (EMG) examination of the upper and lower extremities was normal. Motor nerve conduction velocities were normal as were bilateral ulnar F-wave studies; medium, ulnar, and sural nerve sensory evoked potentials were unobtainable. Two days after admission to the hospital she had no ocular movements and was areflexic. By the 12th hospital day horizontal ocular movements were returning, followed by a gradual return of vertical movements beginning three days later. Pupillary size was still 5 mm, with minimal reaction to light. Pupil testing with 0.06% pilocarpine (on day 15 of the illness) produced prompt constriction bilaterally; no other tests of autonomic function were performed. With physical therapy the patient was able to walk, and ocular motility returned to normal over the ensuing week. She was discharged from the hospital after a total of four weeks of confinement. Eight weeks later ocular examination was normal, the muscle strength reflexes were present. She has remained without neurological symptoms for the past 26 months. Pharmacological testing of the pupils with dilute pilocarpine has not been repeated.

There have been only two prior reports of patients with possible relapsing Fisher's syndrome, but their initial episodes were poorly documented. Elizan *et al*² reported 11 cases of Fisher's syndrome, and they comment that one of their patients (No 11), a 56-year-old woman with limited ophthalmoplegia, ataxia, areflexia, dysphonia, and diffuse sensory abnormalities "29 years previously had an illness of 6 weeks' duration characterised by similar paresthesias, diplopia, and triptopia, and severe trunkal ataxia, which was diagnosed at Bellvue Hospital as polyneuritis". Barontini *et al*⁴ reported a

56-year-old woman with ophthalmoplegia, ataxia and areflexia who had "suffered from similar but less severe symptoms 19 years earlier, without having been admitted to the hospital". Donaghy and Earl⁵ recently reported three patients with minimal oculomotor disturbances who subsequently developed marked weakness and sensory loss (with associated slowing of motor nerve conduction velocities). All three patients suffered relapses of weakness and sensory loss, but not of their oculomotor disturbances. These patients clearly did not have relapsing Fisher's syndrome, but rather chronic relapsing Guillain-Barré syndrome.

Oculomotor abnormalities occur in 5-24% of patients with Guillain-Barré syndrome.^{2,6,7} Twelve of the 50 patients with Guillain-Barré syndrome reported by Haymaker and Kernohan⁶ had varying degrees of extraocular defects, but only one had total ophthalmoplegia. A necropsy study of 19 patients with Guillain-Barré syndrome reported by Asbury *et al*⁷ included three patients with total ophthalmoplegia, and three others with pupillary paresis or restrictions of gaze. Neuropathological findings in true Fisher's syndrome have only recently been described. This patient, reported by Phillips *et al*⁸ was a 67-year-old woman with total bilateral external ophthalmoplegia, partial ptosis, moderate facial weakness, marked ataxia and generalised areflexia. She died suddenly 27 days after admission (from bronchopneumonia). Microscopic examination of the spinal nerve roots and peripheral nerve roots revealed patchy, but extensive, recent segmental demyelination. The changes appeared to be approximately the same age in all the nerves. Examination of the cranial nerves revealed the same patchy demyelination and mild cellular infiltration involving the 7th, 10th, and 11th cranial nerves. The distal portion of the 3rd, 4th, and 6th cranial nerves were not available for examination, but no histological abnormalities of their proximal portions were found. The brainstem and cerebellum were histologically normal. These changes are all characteristics of a demyelinating peripheral neuropathy of the Guillain-Barré type and tend to support the inclusion of Fisher's syndrome within the spectrum of the Guillain-Barré syndrome.

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Pseudotumour cerebri with amiodarone

Sir: Amiodarone is a relatively safe anti-arrhythmic agent, but is also has some extracardiac side effects, which may involve the cornea, the skin, the thyroid, the lungs, the gut and the nervous system. The neurological side effects include tremor, sleep disturbances, headaches¹ and, less commonly, peripheral neuropathy,² proximal weakness and cerebellar dysfunction.³ We report a case of pseudotumour cerebri (PTC) as another possible side effect of amiodarone.

A 58-year-old man had been treated for 6 months with amiodarone (400 mg/day) for supraventricular arrhythmias on exercise. He had also received pindolol (5 mg/day), allopurinol (100 mg/day) and clofibrate (500 mg/day) for several months prior to

amiodarone. He was referred to us after he developed acute blurring of vision in the right eye. The general examination was normal except for a moderate obesity (81 kg, height 162 cm). The blood pressure was 180/90 mm Hg and the ECG was normal. The neurological examination showed bilateral papilloedema, which predominated in the right eye, with normal visual acuity (1.0 in both eyes). A partial inferior field defect was present in the right eye. The CSF pressure was 300 mm H₂O with normal protein (305 mg/l), leucocytes (2.8.10⁶/l) and glucose (5.0 mmol/l). A brain CT scan, including a survey of the pituitary and orbital regions, was entirely normal. Standard blood and urine tests were normal. A diagnosis of pseudotumour cerebri was made. Acetazolamide (3 × 250 mg/day) was administered for 5 months and prednisone (60 mg/day) for 1 month, but both were discontinued in the absence of improvement. The CSF pressure also remained elevated (range 270 to 300 mm H₂O) on follow up 2, 5 and 12 months later. At 12 months, neurological examination still showed bilateral papilloedema with developing disc atrophy. Visual acuity was 1.25 (OD) and 0.8 (OS). Corneal deposits typical of amiodarone were present. Serum levels of amiodarone were determined by HPLC and were within the optimal range,⁴ that is 1.5 mg/l for the parent drug (n = 1.93 ± 0.80) and 0.9 mg/l for its major metabolite, desethyl-amiodarone. Because of the possible side effects, amiodarone as well as any other medication were withdrawn. Sequential spinal taps after 4 and 7 days showed a decrease in CSF pressure (180 and 80 mm H₂O). During the following months the visual acuity improved (1.5 (OD) and 1.0 (OS)) with disappearance of the papilloedema. Some degree of optic disc atrophy persisted bilaterally. A partial field defect also remained in the inferior nasal quadrant of the right eye.

This observation suggests that pseudotumour cerebri was induced by amiodarone because it developed shortly after amiodarone was administered and resolved after the drug was discontinued. The other drugs had been administered previously for a much longer time. The role of amiodarone in the pathogenesis of pseudotumour cerebri is also suggested by the fact that the toxicity of this drug is similar to that of perhexiline, which indeed may produce pseudotumour cerebri.^{5,6} The same types of keratopathy^{1,5} and peripheral neuropathy with lysosomal inclusions^{2,7} have also been reported as side effects of both drugs. This is possibly due to the fact that these drugs are amphiphilic.⁸ A

rise in venous pressure or an impairment in CSF outflow have been proposed to explain the elevation of intracranial pressure in pseudotumour cerebri.⁹ In the cases of pseudotumour cerebri associated with perhexiline and in our patient the exact mechanism remains unsettled.

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Adult onset spinal muscular atrophy with atrophic testes: report of two cases

Sir: Adult onset progressive spinal muscular atrophy is often considered to represent a variant of amyotrophic lateral sclerosis¹ or, less commonly, a hereditary entity.²⁻⁴ Here we report two sporadic cases of severe adult onset progressive spinal muscular atrophy associated with testicular atrophy and normal hormone levels.

Patient 1, a 31-year-old male native of the Ivory Coast, presented with a 2 year history