disorder.6 The combination of HLA types in these patients may be a marker for a predisposition to the Guillain-Barré syndrome.

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Adult onset acid maltase deficiency associated with epilepsy and dementia: a case report

Adult onset acid maltase deficiency (AMD) (glycogenosis type IIb) characteristically presents after the age of 20 years with a slowly progressive limb-girdle muscle weakness.1 Marked muscle involvement occurs in a third of cases and may be the presenting feature.2 Although CNS involvement is seen in the more severe infantile form of AMD (Glycogenosis type Ia), it has not been described in adults. We report a case of adult onset AMD associated with epilepsy and dementia.

A 65 year old woman was referred with intractable epilepsy, the onset of which was at 20 years with a generalised tonic-clonic seizure. Seizures had continued without remission despite treatment with pheno- barbitone, phenytoin, and sodium valproate. The family history consisted of a secondarily generalised tonic-clonic seizure twice a month and a complex partial seizure approximately monthly. At the age of 29 years she required psychiatric admission because of attempted suicide and hysterical behaviour. She was thought to have an immature personality and to lack foresight and judgement. From the age of 52 years she had been in residential care, and anti-social behaviour, incontinence of urine and impaired cognition had been noted. From the age of 57 years she developed progressive limb weakness.

She had five siblings. One was reported to have epilepsy but the details were not available. Three of the other siblings had adult onset AMD and their histories have been published previously.7 The parents had no history of any neurological disorder and there was no consanguinity.

On examination cooperation was limited. She was unable to stand. Fundoscopy and eye movements were normal. There was a mild weakness of neck flexion. Wasting of the shoulder girdle muscles and small muscles of the hand was present bilaterally. Wasting was difficult to assess in the legs because of obesity and oedema. There was a grade 4 weakness proximally in the arms, grade 4+ distally, and a grade 2–3 weakness in the legs, more marked proximally. Coordination was normal. Bilateral grasp reflexes were elicited. Upper limb tendon reflexes were normal but knee and ankle jerks were absent bilaterally. Plantar responses were flexor. No sensory deficit was found. The only abnormal examination was unremarkable except for obesity.

The following investigations were normal or negative: full blood count, electrolytes, renal function, liver function, thyroid function, serum electrolytes, autoimmune profile, blood film for acanthocytes, serology for Hepatitis B and C, hepatitis serology, cerebrospinal fluid protein, oligoclonal bands, urinalysis, protein electrophoresis, coagulation screen, hepatitis A, B, and C, Varela J et al. J Neurol Neurosurg Psychiatry 1983;41:7.

A reappraisal of “direction of scratch” test: using somatosensory evoked potentials and vibration perception

We would like to comment on the article by Hankney and Edin8 which proposed “direction of scratch” test for the clinical examination of posterior column function. This method is easy to perform, and the errors in the detection of direction of scratch on the skin were reported to be correlated with impaired position and/or vibration sense tested using a 128 Hz tuning fork. Since somatosensory evoked potential (SEP) and vibration sense was thought to be mediated through the posterior column and lemniscal pathways, reduction of acid maltase deficiency or useof their test together with SEPs and quantitative vibration sense.

Fifty six patients, mean age 43-5, were studied. They consisted of 43 patients with

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