tract signs (11 patients) or areflexia (6 patients) to suggest a spinal or peripheral nerve component to their cerebellar deficit. Their ages at the onset of symptoms ranged from 16 to 65 years. Five of them had first degree relatives suffering from similar disabilities, and none had any evidence of fat malabsorption or liver disease.

Vitamin E concentrations were estimated in non-fasting samples of serum using a fluorometric method.⁴ The mean result was 27.2 $\mu\text{mol/l}$, range 17.4–56.7 $\mu\text{mol/l}$. The reference range for our laboratory based on 300 apparently healthy individuals was 11.5–45.6 $\mu\text{mol/l}$. None of the patients therefore had a below-normal vitamin E concentration, but one had an above-normal level.

Our failure to demonstrate a deficiency of vitamin E in a group of 14 patients suggests that such deficiency is uncommon among adult-onset cases of spinocerebellar degeneration, but nevertheless worth seeking as the condition is otherwise untreatable.

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- malabsorptive disorders such as biliary atresia, cystic fibrosis and after extensive intestinal resection,² but patients with an identical neurological syndrome and an apparently genetically determined selective deficiency of vitamin E have also been reported.^{3,4} Although most of the cases of selective vitamin E deficiency have developed symptoms before the age of 20 years and inheritance is assumed to be autosomal recessive, a family containing patients with onset in the sixth decade or later and possibly exhibiting X-linked inheritance has been described.⁵
- Serum vitamin E concentrations are normal in Friedreich's ataxia, a disorder resembling the early onset neurological syndrome associated with isolated vitamin E deficiency.⁶ We have now measured serum vitamin E concentrations, using high performance liquid chromatography and fluorimetry⁷ (normal range 11.5–35.0 $\mu\text{mol/l}$), in 50 patients with a variety of degenerative ataxic disorders.⁸ None of them were known to be taking vitamin E supplements or had any clinical evidence of malabsorption.
- There were 12 with early onset cerebellar ataxia with retained reflexes (median serum vitamin E concentration 22.4, range 11.0–35.0 $\mu\text{mol/l}$), 11 with other early onset ataxias (median 21.7, range 15.8–48.4 $\mu\text{mol/l}$), 4 with autosomal dominant cerebellar ataxia (median 29.6, range 13.1–46.6 $\mu\text{mol/l}$), and 23 with idiopathic late onset cerebellar ataxia (median 24.4, range 12.6–37.7 $\mu\text{mol/l}$). Seven of the cases of idiopathic late onset cerebellar ataxia, one with autosomal dominant cerebellar ataxia, and 10 of those with other early onset ataxias had depressed or absent tendon reflexes, usually combined with loss of vibration and joint position sense. Not all of these patients had electrophysiological evidence of a peripheral sensory neuropathy. Over the same period of time we have identified two patients with apparently isolated deficiency of vitamin E, both of whom had previously been diagnosed as having Friedreich's ataxia, with ataxia developing before the age of 20 years, areflexia, marked loss of joint position sense, and undetectable serum vitamin E concentrations.³

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Serum Vitamin E concentrations in degenerative ataxias

Sir: Prolonged and severe vitamin E deficiency is known to cause a progressive neurological disorder comprising limb and gait ataxia, distal loss of proprioception and vibration sense, and areflexia.¹ This usually occurs in abetalipoproteinaemia or other fat

malabsorptive disorders such as biliary atresia, cystic fibrosis and after extensive intestinal resection,² but patients with an identical neurological syndrome and an apparently genetically determined selective deficiency of vitamin E have also been reported.^{3,4} Although most of the cases of selective vitamin E deficiency have developed symptoms before the age of 20 years and inheritance is assumed to be autosomal recessive, a family containing patients with onset in the sixth decade or later and possibly exhibiting X-linked inheritance has been described.⁵

Serum vitamin E concentrations are normal in Friedreich's ataxia, a disorder resembling the early onset neurological syndrome associated with isolated vitamin E deficiency.⁶ We have now measured serum vitamin E concentrations, using high performance liquid chromatography and fluorimetry⁷ (normal range 11.5–35.0 $\mu\text{mol/l}$), in 50 patients with a variety of degenerative ataxic disorders.⁸ None of them were known to be taking vitamin E supplements or had any clinical evidence of malabsorption.

There were 12 with early onset cerebellar ataxia with retained reflexes (median serum vitamin E concentration 22.4, range 11.0–35.0 $\mu\text{mol/l}$), 11 with other early onset ataxias (median 21.7, range 15.8–48.4 $\mu\text{mol/l}$), 4 with autosomal dominant cerebellar ataxia (median 29.6, range 13.1–46.6 $\mu\text{mol/l}$), and 23 with idiopathic late onset cerebellar ataxia (median 24.4, range 12.6–37.7 $\mu\text{mol/l}$). Seven of the cases of idiopathic late onset cerebellar ataxia, one with autosomal dominant cerebellar ataxia, and 10 of those with other early onset ataxias had depressed or absent tendon reflexes, usually combined with loss of vibration and joint position sense. Not all of these patients had electrophysiological evidence of a peripheral sensory neuropathy. Over the same period of time we have identified two patients with apparently isolated deficiency of vitamin E, both of whom had previously been diagnosed as having Friedreich's ataxia, with ataxia developing before the age of 20 years, areflexia, marked loss of joint position sense, and undetectable serum vitamin E concentrations.³

Neurologists need to know which clinical circumstances dictate that serum vitamin E concentrations should be measured in patients with progressive ataxic syndromes. It is clearly important not to overlook treatable causes of these disorders such as vitamin E deficiency, even if they can be identified in only a very small proportion of patients. The neurological consequences of severe vitamin E deficiency appear to be remarkably consis-

tent, reflecting the pathological findings of a central-peripheral distal sensory axonopathy preferentially affecting centrally directed axons.¹ On the basis of this and our data, we suggest that the measurement of serum vitamin E concentrations should be confined to degenerative ataxias associated with depressed or absent tendon reflexes and abnormal proprioception, regardless of whether or not peripheral sensory nerve action potentials are abnormal.³ This applies to patients presenting at any age, in view of the late age of onset in some deficient cases.

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