gramme. This is in keeping with the tremor of other body parts, for example the wing-beating arm tremor of Wilson's disease and task-specific tremors such as writing and other occupational tremors.[3]

We have examined clinically, three other patients with clear dystonic head tremor in whom head tremor was altered by the position of the head in the anterior-posterior plane. In none of the others could we demonstrate a vestibular (otolith) mechanism. Despite these negative observations for a clear otolith influence upon head tremor electrophysiological measurements have shown that otolith spinal reflexes may be modulated by both head and body position.[6] Thus there still remains the possibility that a change in head or body position may in itself modify vestibular influences upon body movement.

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Probable cases of Mast syndrome in a non-Amish family

Complicated forms of hereditary spastic paraplegia are rare. A 58 year old Flemish woman was admitted with a clinical picture of slowly progressive spastic paraplegia, dysarthria, presenile dementia and mild athetosis.

At the age of 16 her gait became shuffling. Before the age of 30 she used a walking stick, since the age of 35 years she has needed a walking frame and from the age of 40 she has become more wheelchair bound. During the third decade (maybe earlier) dysarthria, apathy and negativism appeared. Towards her 45th year, urinary incontinence began. On admission, aged 48, significant bradyphrenia and comprehension difficulties were noted and during the following years she presented a further mental deterioration. She is now bedridden and her speech restricted to rare, usually inappropriate, single syllable answers, which are sometimes repeated. She has difficulty swallowing fluids. She often shows spontaneous repeated slow turning of the head to the right and left and mild tortuous movements of the shoulders. Except for a divergent strabismus there are neither oculomotor abnormalities, nor fundoscopic anomalies. There is a slight bilateral facial weakness. The fine motor hand skills are lost without clear paresis of the upper limbs. The strength of leg muscles measures about 1 to 2/5. There were neither sensory deficits nor cerebellar signs. Deep tendon reflexes in the upper limbs were increased slightly. Knee jerks were unusually brisk and without clonus; ankle jerks were decreased. Plantar responses were extensor. Snout reflex and bilateral palmomental reflexes were present whilst corneomandibular reflexes were absent.

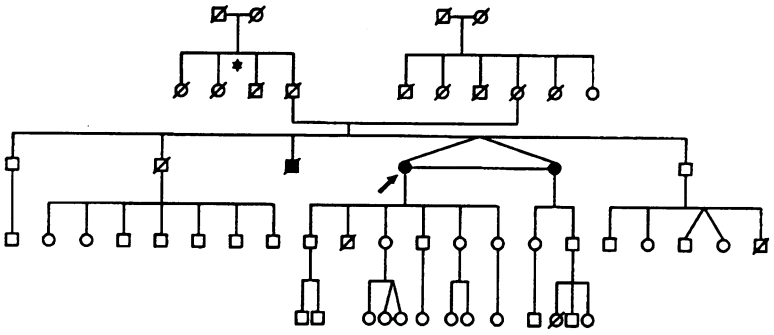

Figure Pedigree

The following laboratory investigations showed no significant abnormalities: routine blood examination (except for intermittent elevation of glucose with normal haemoglobin AlC), creatine kinase, copper, lipids, very long chain fatty acids ratios C24/C22 and C26/C22, vitamin E, vitamin B12, folate, cortisol, adrenocorticotropic hormone, thyroid hormones, arylsulfatase A and hexosaminidase A + B; the CSF protein was 590 mg/l with normal electrophoretic pattern.

The EEG showed mild general slowing. Nerve conduction studies and needle electromyography showed a mild axonal polyneuropathy. The somato-sensory evoked potentials demonstrated a slightly prolonged central conduction time. An electrocardiogram was normal. MRI of the brain showed diffuse cortico-subcortical atrophy, periventricular hyperintensities, thin corpus callosum and less marked atrophy of the brainstem and cerebellum. Light microscopic and electron microscopic examination of conjunctiva and skin showed some membranous cytoplasmic body-like inclusions (Professor J J Martin, Dr C Ceuterick-de Groote, University Hospital Antwerp).

The family history revealed two similar cases (figure). There was no known consanguinity.

The monozygotic twin has an almost identical medical history and clinical picture. She is able to stammer a few simple words. Her answers are sometimes slightly more appropriate, particularly for old memories. Her knee jerks and ankle jerks are both unusually brisk, and the plantar responses are extensor. One brother died at the age of 53. The medical records and relatives described difficulties with walking from the age of 20 (maybe earlier). During the following decades he presented a progressive spastic paraparesis, dysarthria, mental deterioration and urinary and faecal incontinence; no deficits of sensation or coordination were demonstrated. There was dysphagia in his last years. Death was due to pneumonia.

The pedigree suggests an autosomal recessive inheritance. The neurodegenerative syndrome in this family seems fully comparable to the Mast syndrome, described in 1967 in an Ohio Amish isolate by Cross and McKusick.[1–3] There appears to be no similar cases that have been described outside the Amish population.

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Ultrasensitive TSH assay and anti-Parkinsonian treatment with levodopa

We have recently reported the association of Parkinson's disease and hyperthyroidism in a group of 10 patients.[1] In that report, symptoms of Parkinson's disease were always significantly exacerbated by the development of hyperthyroidism and improved by its successful treatment.[1] We proposed that hyperthyroidism should be suspected in all Parkinsonian patients when their condition deteriorates.[1] Since clinical diagnosis of thyrotoxicosis is difficult in Parkinsonian patients, they should have a comprehensive thyroid examination and, if there is the slightest suspicion of hyperthyroidism, a hormonal evaluation of thyroid function (free T_4, ultrasensitive TSH).

Thyroid hormone levels (T_3 and T_4) have been found to be normal in Parkinsonian patients untreated or treated with levodopa.[2] However, a decreased response of thyrotropin (TSH) after stimulation by TRH (thyrotropin releasing hormone) has been reported in Parkinsonian patients treated with levadopa.[3] Such a decreased TSH response after TRH stimulation is observed during hyperthyroidism, and is sometimes the only hormonal abnormality, especially in elderly patients with autonomous thyroid nodules. Thus the decreased TSH response after TRH-stimulation in patients treated with levodopa could be responsible for a false diagnosis of hyperthyroidism. These results, however, were obtained before the ultrasensitive TSH determination with a monoclonal antibody assay was available. A low ultrasensitive TSH level has the same significance as a decreased TSH response to TRH, indicating an increased negative feed-