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 otothal**mic influence upon head tremor electrophysiological measurements have shown that otothal**mic reflexes may be modulated by both head and body position. Thus there still remains the possibility that a change in head or body position may 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 Polish 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 wheelchair bound.

During the third decade (maybe earlier) dystar**thy 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 syllables, apraxia, aphasia and negativism appeared. She has difficulty swallowing fluids. She often shows spontaneous repeated slowing turning of the head to the right and left and mild tortuous movements of her shoulders. Except for a divergent strabismus there are neither oculo-motor 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 usually brisk and without clonus; ankle jerks were exaggerated. Plantar responses were extensor. Snout reflex and bilateral palmpalant reflexes were present whilst cromandibular reflexes were absent.

The following laboratory investigations showed no significant abnormalities: routine blood examination (except for intermittent elevation of glucose with normal haemoglobin A1C), creatinine, 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 did not show a major 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 men
dranous 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, dystar**thy, mental deterioration and urinary and faecal incontinence; no deficits of sensation or coordination were demonstrated. There was dysp**nea 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 community and McKusick.1-3 There appears to be no similar cases that have been described outside the Amish population.

Ultrason**sitive 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.2 We proposed that hyperthyroidism should be suspected in all Parkinsonian patients when their condition deteriorates.3 Since clinical diagnosis of thy
toxosis is difficult in Parkinsonian patients, they should have a comprehensive thyroid examination and, if there is the slightest suspicion of hyperthyroidism, a hor
monal evaluation of thyroid function (free T₄, ultrason**sitive TSH). Thyroid hormone levels (T₂ and T₃) have been found to be normal in Parkinsonian patients untreated or treated with levodopa.4 However, a decreased response of thyro
tropin (TSH) after stimulation by TRH (thyrotropin releasing hormone) has been reported in Parkinsonian patients treated with levodopa.5 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 ultra
sensitive TSH determination with a mono
clonal antibody assay was available. A low ultrasensitive TSH level has the same sig
nificance as a decreased TSH response to TRH, indicating an increased negative feed-