On examination, we saw a very nervous, profusely perspiring, emotionally labile woman, who demonstrated a retrocollis with stretching of her back and extremities on being spoken to, or when asked to execute simple motor activities. She could not sit up because of the stiffness and when in a supine position, she could be lifted as a board from her bed to the examination table. Because of severe contraction of the sternocleidomastoid muscle, it was difficult to palpate the thyroid gland. Her pulse rate was 120/min. There was a mild tremor of the hands. Motor power was difficult to judge because of generalised hypertonia. Tendon reflexes were brisk, the right plantar response was extensor, the left was equivocal. Laboratory studies revealed hyperthyroidism: T4 167 nmol/l (normal values 60–150 nmol/l); free T4 43 pmol/l (normal 8.0–25.0 pmol/l); free T4 index 184 (normal 59–154). Following administration of iodine containing contrast medium for CT, the hyperthyroidism was aggravated (T4 259 nmol/l, free T4 84 pmol/l). Thyroid stimulating immunoglobulin and antibodies against thyroid mitochondria and parietal cells were present; antibodies against thyroglobulin were absent. Other causes of a pyramidal syndrome were ruled out (normal vitamin B12, skull CT, diameter of cervical vertebral canal and craniocervical junction, Queckenstedt's tests and cerebrospinal fluid), as was myopathy (normal EMG, the paravertebral muscles, however, showed continuous but normal activity, and normal muscle biopsy).

The patient was treated with 3×20 mg thiamazole, 3×40 mg propranolol chloride, and 3×10 mg oxazepam. Five weeks later she was biochemically euthyroid. At that time she was calm, her pulse rate was normal, the hypertonia had decreased and so had the spasms. She could sit and walk again and rise from a chair. It was still difficult for her to rise from a supine position because of some remaining axial spasms. The plantar reflexes had become flexor. After two months, she was given $50 \mu\text{g}$ thyroxine to inhibit the production of TSH. At the same time propranolol and oxazepam were stopped. A few days later she had a relapse of the muscle spasms, the hypertonia and the nervous complaints, but to a milder degree than on admission. The thyroxine was stopped. A week later she had improved to the state prior to the relapse. Two months later the muscle spasms had completely disappeared and no neurological signs were found, except for a mild "clumsy" gait.

The clinical picture shows aspects that are compatible with both dystonia and stiff-man

syndrome. Voluntary movements and emotional stress caused the spasms to occur, which is known in either condition.^{1,2} The absence of spasms during sleep does not differentiate between the two, however; the fact that the spasms were not painful is very uncommon in stiff-man syndrome.² Heavy perspiration and distress can be seen both in stiff-man syndrome² and in a state of hyperthyroidism. Extensor plantar response have been described in both.^{2,3} Choreoathetosis is known to occur in hyperthyroidism.^{4,5} Fahn⁶ mentioned torticollis spastica and Nutt *et al.*⁷ the occurrence of Meige syndrome in hyperthyroidism, which both can be regarded as focal dystonia. Neither muscle spasms nor generalised dystonia or stiff-man syndrome have ever been clearly associated with hyperthyroidism.^{2,8} In our patient the decreasing T4 level was accompanied by disappearance of the muscle spasms and other signs. Administration of a small amount of thyroxine caused them to return. After stopping the thyroxine they again vanished. This suggests a direct or indirect causal relationship between thyroxine (the active T3) and the clinical symptoms.

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Kearns-Sayre syndrome, hypoparathyroidism, and basal ganglia calcification

Sir: Hypoparathyroidism has been rarely described in patients with the Kearns-Sayre syndrome. We describe a young boy who was treated for hypoparathyroidism from the age of 2 years, and had the first features of the Kearns-Sayre syndrome at the age of 6. CT scanning of the brain at the age of 11 years showed no evidence of intracranial calcification which is found in up to 50% of cases of this syndrome¹ and is almost invariably found in idiopathic hypoparathyroidism.²

A West Indian boy presented at the age of 2 years with a 6 day history of diarrhoea and vomiting. He had always been a jittery rather irritable child, tending to shake in all four limbs when handled since birth, and one month prior to presentation had had a febrile fit. Developmental milestones had been normal. He had walked at 10 months and spoke at 11 months. He was found to be hypocalcaemic (1.48 mmol/l), hyperphosphataemic (2.42 mmol/l) and had a low parathormone level (0.18 ng/l). A diagnosis of primary hypoparathyroidism was made and he was commenced on calcium supplements and vitamin D (calciferol 50,000 units daily) with ascorbic acid (500 mg/day).

Over the next 4 years he had a number of upper respiratory tract infections, one episode of oral candidiasis, and one of otitis media. He continued to be irritable, at times bad tempered with occasional short lasting generalised shaking episodes without loss of consciousness. Throughout this period his serum calcium and phosphate levels were normal. An EEG demonstrated an excess of slow forms with episodic slow activity. These episodes were controlled with phenytoin 50 mg twice a day. Testicular descent was not complete until the age of six and he was enuretic until the age of 8 years.

At the age of 6 years it was first noticed he had drooping of the eyelids. Over the next two years he developed difficulty with walking due to unsteadiness and had frequent falls. From the age of 8 he developed a progressive proximal muscle weakness. At the age of 11 bilateral fascia lata slings were inserted to correct the bilateral ptosis.

On examination at the age of 11 he had atypical bilateral diffuse pigmentary changes in both fundi with normal choroid and retinal vessels. There was marked limitation of abduction of both eyes, bilateral ptosis and facial weakness, and a myopathic facies. He had a proximal muscle weakness with normal upper but brisk lower limb reflexes and

bilateral flexor plantar responses. There was a mild finger/nose, heel/shin and truncal ataxia. There were no sensory abnormalities and general examination was normal but his height was between the 5th–10th percentile for his age.

Routine investigations at the age of 11 were normal and included a full blood count, ESR, urea and electrolytes, calcium, phosphate, alkaline phosphatase, liver function tests, blood sugar, glucose tolerance test, serum lipids, CK, vitamin B12, folate, thyroid function, prolactin, cortisol, growth hormone, urinary protein and respiratory function tests. The serum magnesium was low at 0.5 mmol/l (normal 0.7–0.95) with a 24 hour urinary magnesium excretion of 1.46 mmol/l (normal 2.1–6.2). The cerebrospinal fluid protein was 1.13 g/l. An ECG revealed right bundle branch block with left axis deviation. An EEG showed a slight excess of slow forms for his age but no evidence of epilepsy. Visual evoked responses were normal. Electromyography (EMG) showed normal motor and sensory conduction velocities. Concentric needle electromyography showed short duration, polyphasic motor unit action potentials. On maximal volition there was a full interference pattern of small amplitude with maximum amplitude of 750 μ V. A right quadriceps muscle biopsy demonstrated a large number of “ragged red fibres” affecting only type I fibres. A CT scan was normal with no evidence of intracranial calcification.

There have been few previous reports of the association of the Kearns-Sayre syndrome with hypoparathyroidism.^{1–5} The hypoparathyroidism may predate or post-date the onset of symptoms and signs typical of the Kearns-Sayre syndrome and may or may not be associated with seizures (table). In the three previous cases reports in which CT scans were performed intracranial calcification was found, localised to the basal ganglia. Seigal *et al*¹ found intracranial calcification in four of eight patients

with the Kearns-Sayre syndrome, one had hypoparathyroidism, one pseudo-hypoparathyroidism, and the remaining two had normal calcium, phosphate and parathormone levels as did all the patients without intracranial calcification.

The absence of basal ganglia calcification in our patient with both hypoparathyroidism and the Kearns-Sayre syndrome may be due to the early treatment with calcium and vitamin D and the return of the serum calcium and phosphate levels to normal. This suggestion would be supported by the observation that intracranial calcification does not occur in secondary hypoparathyroidism due to previous thyroid surgery where treatment is started early with the onset of tetany.³ The predilection of the basal ganglia for calcification in the recognised disorders associated with intracranial calcification may be due to disorders of calcium metabolism, increased vascular permeability, the preferential perfusion of grey matter and the high rate of blood flow to the basal ganglia. Furthermore alkaline phosphatase activity may be regionally elevated in the basal ganglia in patients developing intracranial calcification.⁶

The early treatment of hypoparathyroidism in cases of Kearns-Sayre syndrome is required to control the symptoms of hypocalcaemia and may prevent basal ganglia calcification, as in our case. Whether the calcification in the basal ganglia has a clinical consequence in this syndrome is uncertain and where it is found a careful search for hypoparathyroidism should be made.

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Table

Author	Age of presentation of hypoparathyroidism	Age of presentation of Kearns-Sayre syndrome	Epilepsy	CT scan calcification
Seigal ¹	?	1	Absent	Present
Toppet <i>et al</i> ²	7	9	Absent	—
Sachs ³	?	?	—	Present
Horwitz ⁴	13	11	Absent	—
Pellock ⁵	9	7	Present	Present
Present case	2	6	Present	Absent

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Meige's syndrome and palatal myoclonus associated with brain stem stroke. A common mechanism?

Sir: Meige's syndrome (blepharospasm and oro-facial dystonia), and palatal myoclonus are two uncommon movement disorders whose pathophysiology is poorly understood. Isolated reports have associated each with focal brain stem lesions and have suggested denervation hypersensitivity of brain stem nuclei as the underlying mechanism.^{1–3} We report a patient in whom both Meige's syndrome and palatal myoclonus developed following upper brain stem strokes. A related origin for these movement disorders is postulated.

A 78-year-old right-handed woman, who was recovering from a stroke was first seen because of involuntary facial movements. Five years previously she had experienced the sudden onset of rotational vertigo, vomiting and loss of balance, resulting in falling. A coarse action and intention tremor of the right upper limb appeared after this episode, causing difficulty with writing and bringing food to the mouth. The tremor apparently remained unchanged over the next five years but was totally abolished following the recent stroke. Involuntary facial grimacing and intermittent forced eye closure had also been present for five years, being aggravated by concentration or anxiety. Six weeks previously she developed sudden loss of speech (anarthria) with right hemiplegia. There was