Table 1 Laboratory and neuropsychological results at presentation (A) and at 12 month follow up (B)

Laboratory (units)	*A*	*B*
Full blood count	Normal	Normal
Erythocyte sedimentation rate	12	6
Urea and electrolytes	Normal	Normal
Liver function tests	Normal	Normal
Antinuclear antibody	Negative	Negative
B12 and folate	Normal	Not tested
VDRL	Negative	Not tested
Thyroid stimulating hormone (mU/l)	58.4	0.87
Free T4 (pmol/l)	7.4	Not tested
Antithyroid microsomal antibody titres	1:25600	1:1600
Psychometric (normal/predicted range):		
Folstein MMSE (>24)	25/30	25
NART IQ	10th percentile	18th percentile
WAIS-R (verbal)	13th percentile	Not tested
WAIS-R (performance)	27th percentile	Not tested
FAS verbal fluency (>30)	25	23
Cognitive estimates test (<6)	10	11
Graded naming test (>15)	10/30	16/30
Digit span forwards (>5)	7	6
Rey-Osterreith complex figure (copy) (36)	25.5	24
Rey-Osterreith complex figure (recall) (30%)	Not tested	75%

became aggressive and threatened them with a saw. The general practitioner was called and suspected either an acute psychosis or a severe depressive illness. Police assistance was requested because of the patient's continuing violent behaviour.

On admission he was unkempt but cooperative and apparently euthymic. He denied depression, but displayed no insight into the irregularity of his behaviour. No psychotic features were seen, although during the admission he consistently rationalised all reported psychotic phenomena. He was aggressive towards staff and made repeated attempts to abscond. General physical examination was unremarkable. Neurological examination was normal except for spoken language, which was fluent and grammatical, but contained word finding pauses, circumlocutions, and occasional semantic errors (for example, "I just want to get my feet back on the table"). Formal neuropsychological testing, and a screen of laboratory tests for reversible causes of encephalopathy, were performed on admission, and results are presented below (column A). Attention is drawn to his mild naming deficit, and poor performance on the Rey figure, which was due to planning rather than visuospatial errors, suggesting a predominantly "dysexecutive" pattern. CT and EEG were both normal, and SPECT disclosed widespread but mild cortical hypoperfusion. Trifluoperazine (2 mg twice daily) was started on admission, and thyroxine (75 µg once daily) added 1 week later. His mental state and behaviour stabilised, leading to discharge after 2 months.

At 6 month follow up the patient had stopped neuroleptic drugs, but continued taking thyroxine. He reported feeling "back to normal", had bought a new house, and was working as a part time shop assistant. He still had subtle word finding difficulties, and was referred to the regional memory clinic for further evaluation, which took place 6 months later. Behavioural assessment showed persisting deficits in delayed recall of verbal material, verbal fluency, and visuospatial function. Formal psychometric testing, blood tests, and SPECT were repeated, 1 year after the original examinations. Laboratory and neuropsychological results are presented in the table. It is of note that, whereas his naming ability had improved, performance on frontal executive tasks remained impaired. The appearance of the follow up SPECT differed minimally, if at all, from the first examination.

In summary, therefore, this patient presented in clear consciousness with a first episode of acute psychosis, and evidence of subtle executive and linguistic neuropsychological disturbance, on the background of gradual behavioural and affective change. He was profoundly hypothyroid due to an autoimmune thyroiditis, but there was no clinical evidence of thyroid failure other than the abnormal mental state. The psychiatric component of his illness recovered fully, and the antithyroid microsomal antibody titre fell markedly after thyroxine replacement, although his mild neuropsychological deficits remained unchanged. Corticosteroids were not used at any stage.

The response to thyroxine does not, in itself, imply that the cerebral illness had an endocrine origin; a recent report described a patient with a subacute encephalopathic illness and compensated hypothyroidism in the presence of increased antimicrosomal antibodies, all of which responded to thyroxine replacement alone.[4] In that case, however, both EEG and SPECT were abnormal, the SPECT showing multiple areas of severely reduced perfusion, which normalised with treatment. By contrast, in the present case the EEG was normal and the SPECT abnormality was marginal and changed little, if at all, with treatment. The evidence for a significant vasculitic component to the illness is, therefore, unconvincing.

The mild and relatively circumscribed neuropsychological deficits coupled with florid psychotic phenomena, also contrast with the profound global disturbance of cognition usually associated with Hashimoto's encephalopathy.[3] This distinction suggests that microvascular disruption and thyroid hormone depletion may emphasise different aspects of the clinical range in Hashimoto's encephalopathy. Although the present case would support Asher's conclusion that the psychiatric features of Hashimoto's encephalitis typically respond to thyroid replacement, it additionally suggests that subtle neuropsychological deficits may be apparent even in the absence of obvious cerebral perfusion deficits, and that these may not be fully reversible.

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Alien hand sign in Creutzfeldt-Jakob disease

The clinical picture of Creutzfeldt-Jakob disease (CJD) includes various movement disorders such as myoclonus, parkinsonism, hemiballism, and dystonia. We report on a patient with CJD who manifested the alien hand sign. We suggest that CJD should be included in the differential diagnosis of diseases which present with an alien hand.

Creutztfeldt-Jakob disease, one of the human prion diseases, is characterised by rapidly progressive mental and motor deterioration.[1] Involuntary movements occur in above 90% of the patients in the course of the disease, the most common being myoclonus.[1] Other movement disorders range from tremor to chorea, athetosis, dystonia, and hemiballism.[1] We report on a patient with CJD who presented with an alien hand.

Alien hand is a rare and striking phenomenon defined as "a patient's failure to recognise the action of one of his hands as his own".[2] One of the patient's hands acts as a stranger to the body and is uncooperative. Thus, there is loss of feeling of ownership but not loss of sensation in the affected hand. Originally described in callosal tumours,[3] the aetiology of alien hand also includes surgical callosotomy,[4] infarction of the medial frontal cortex, occipitotemporal lobe, and thalamus,[1 5] infection,[6] and corticobasal degeneration.[5 7]

A 70 year old, right handed Jewish man born in Argentina, living in Israel for the past 20 years, was admitted to the Neurology Department. Until a month before his admission, he was apparently healthy and helped in the accounting office of the village where he lived. His neurological illness had presented insidiously during the past month with unsteadiness of gait and frequent falls. He also manifested behavioural changes, became aggressive, and had visual hallucinations, perceiving insects and mice moving through his visual field. Often, he expressed his fear from seeing that the "ceiling was

falling over him". His wife mentioned bizarre, useless movements of his left hand which were present from the beginning of the disease.

On admission, he was awake, bradyphrenic, and partially collaborative. His conversation was often disrupted by hallucinations. The affect was sad and he had partial insight for his mental dysfunction. He was disoriented for time, place, and situation. He could understand speech and was able to follow oral instructions involving two consecutive components. Naming was preserved. Prominent dysgraphia and dyscalculia were noticed. Immediate recall and short term memory were severely disturbed, whereas long term memory, especially for personal life events, was relatively spared. Abstract thinking was severely affected. Bimanual movements, such as clapping, were extremely difficult.

The cranial nerves were normal as were ocular fundi. The motor examination showed normal force. Deep reflexes were symmetric and plantar responses were flexor. The right arm had a dystonic posture. His gait was ataxic on a wide base.

At times, the left arm would spontaneously rise in front of the patient during speaking or while using his right hand. He was unaware of these movements until they were brought to his attention. When questioned about their purpose, the patient denied that they were voluntary. No grasping of either hand or foot was found. The patient had no cortical sensory loss.

The laboratory data including blood chemistry, haematology, and sedimentation rate were normal, as were folic acid, vitamin B_{12} concentrations, and thyroid function.Venereal disease research laboratory and HIV tests were negative. The cerebrospinal fluid had normal content. Brain CT showed mild cerebral atrophy. An EEG showed severe diffuse slowing at admission. Within a week, repeated EEGs showed triphasic waves with a periodic pattern of 1- 1.5 Hz.

During the next 2 weeks, the patient developed myoclonic jerks. Severe dysphasia and cognitive decline were accompanied by confusion and aggression. He became grossly ataxic, and unable to walk and perform any of his daily activities even with help. Transferred to a chronic care hospital, he died few weeks later. Postmortem examination was not allowed.

This short fatal neurological disease manifested by fulminant dementia, myoclonic jerks, and extrapyramidal and cerebellar dysfunction was strongly suggestive of CJD. The periodic EEG pattern reinforced this diagnosis. Our patient's alien hand was part of the otherwise characteristic clinical picture of CJD, but it occurred early in the disease course when no myoclonic jerks were present. We are aware of only one report of alien hand in CJD. MacGowan *et al*[8] described two patients with CJD with a myoclonic alien hand syndrome. In one patient the left arm "was noted to have spontaneous movements which appeared purposeful…wandered out of her view". In the second, the alien limb performed complex actions such as unbuttoning her blouse and removing a hair pin. Although our patient had no myoclonus or pyramidal signs when the alien hand appeared, in their patients it was associated with spontaneous or stimulus sensitive myoclonus, spastic hemiparesis, and cortical sensory loss.

The literature seems to describe distinct forms of alien hand. All share the occurrence of involuntary movements contrary to the patient's stated intent, but the types of movement differ. In the callosal form, there are purposeful movements of the non-dominant hand.[9] In the frontal form, there is grasping and utilisation behaviour of the dominant hand.[9] In the corticobasal degeneration, there are aimless movements of either hand.[5 7] When a consequence of tumorous or vascular pathology,[9] alien hands can perform complex acts such as trying to tear clothes or undoing buttons. The description by MacGowan *et al*[8] has characteristics of the callosal form (especially in patient 2). However, our case suggests that the alien hand sign in CJD may appear in a different type, performing less complex movements which resemble those reported by Riley *et al* in corticobasal degeneration.[7] These authors described the alien limb as " involuntarily rising and touching the mouth and eyes" (patient 1). The patient thought that she "was powerless to stop this movement" and when directed to stop responded that "she can't". Another patient's left arm was at times "elevated in front of him", while he was "unaware of this situation until his attention was called to it" (patient 10).

Another related phenomenon coined as "arm levitation" was reported in progressive supranuclear palsy. In these patients the arm involuntarily raised and performed semi-purposeful movements.[10]

One common denominator between CJD, corticobasal degeneration, and progressive multifocal leukoencephalopathy,[6] in which an alien hand sign has also been described, is multifocality. In corticobasal degeneration, it was proposed that more than one site is affected or that a "release" phenomenon occurs accounting for the aetiology of alien hand.[7] In CJD, bilateral cortical damage to motor areas might be the origin of their subsequent isolation and disconnection.

We suggest that CJD should be added to the differential diagnosis of diseases presenting with an alien hand with or without myoclonus.

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Recurrent peripheral neuropathy in a girl with celiac disease

The involvement of the peripheral nervous system (PNS) in children with celiac disease is particularly rare. Furthermore, in both children and adults with celiac disease, neurological complications are chronic and progressive.[1]

We report on a 12 year old girl affected by celiac disease, who on two separate occasions presented with an acute peripheral neurological syndrome after accidental reintroduction of gluten in her diet.

This patient was born uneventfully to healthy non-consanguineous parents with no family history of neurological or metabolic diseases. At the age of 6 months she was diagnosed as having celiac disease according to the European Society of Paediatric Gastroenterology and Nutrition (ESPGAN) criteria. Since then she was on a strict gluten free diet and was asymptomatic until the age of 10 years when severe diarrhoea, vomiting, and abdominal pain manifested 6 days after the intake of corn flakes erroneously thought to be gluten free. No previous infections had been noticed. One week after the onset of these symptoms she experienced acute weakness and pins and needles sensation confined to her legs. At that time her parents stopped her intake of corn flakes on the suspicion that these were responsible for the symptoms. Despite this, symptoms worsened during the next 2 days, confining her to bed.

At hospital admission, she was alert and mentally stable. Results of general physical examination were unremarkable. Neurological examination disclosed symmetric, predominantly distal, weakness of the legs; the knee jerks and ankle reflexes were depressed; plantar reflexes were flexor. Distal stocking glove decreased in pin prick and temperature with sparing of propioception and light touch. Coordination tests were normal.

Laboratory investigations showed a white cell count of 9300/mm^3. The results of the following investigations were within the normal limits: haemogram, erythrocyte sedimentation rate, serum urea, nitrogen, electrolytes, creatinine, glucose, transaminase, bilirubin, immunoglobulins (Igs), lead, iron, copper, urinalysis, urinary porphyrin, folic acid, and vitamins A, B_1, B_6, B_{12}, and E. Antibodies to *Campylobacter jejuni*, neurotropic antivirus antibodies, specific and non-specific organ autoantibodies, IgA and IgG antigliadin antibodies (AGAs), IgA antiendomesium antibodies (EMAs), and IgA antireticulum antibodies (ARA), assayed by enzyme linked immunoadsorbent assay (ELISA) and immunofluorescence (IF) were also negative. Lumbar puncture was not performed. Antibodies against gangliosides GM1 and GQ1b, myelin associated glycoprotein and myelin