regression line best fitting the points for b versus $\ln(SI)$; where SI is the signal intensity from a region of interest within the images acquired at each b value. Performing this calculation on a pixel by pixel basis created the ADC maps.

Brain MRI performed 3 days after symptom onset showed extensive T2 hyperintensities involving the corona radiata, internal capsule, basal ganglion, thalamus, and midbrain on the right side. Brain diffusion weighted imaging showed slight hyperintensities which were limited to the corona radiata, the medial portion of the basal ganglion, and the thalamus. Four sampled ADCs in the corresponding regions of T2 hyperintensity demonstrated increased diffusion (ranging from 1.17 to $1.26 \times 10^{-5} \text{ cm}^2/\text{s}$), compared with a matching location in the uninvolved contralateral hemisphere (ranging from 0.77 to $0.80 \times 10^{-5} \text{ cm}^2/\text{s}$, figure A and B). Magnetic resonance angiography showed no abnormalities. The patient improved rapidly after treatment with a high dose of corticosteroid. Within 2 weeks all previously noted neurological abnormalities had resolved except for a slight left hemiparesis. An MRI repeated at this time showed a partial decrease in the extent of the T2 hyperintensity. One year later he was readmitted with a slowly progressive bulbar weakness, frontal lobe dysfunction, urinary incontinence, and depressive mood changes. Follow up MRI performed at this time, showed that the previous T2 abnormalities had improved, but the atrophy of the brain stem and basal ganglia became evident with periventricular high signal intensities. Four ADCs sampled in locations corresponding to those of the initially increased ADCs decreased to values which ranged from 0.98 to $1.07 \times 10^{-5} \text{ cm}^2/\text{s}$ (figure C and D).

In our patient the ADC maps and ADC values showed high proton mobility, which suggests vasogenic oedema in acute lesions of neuro-Behçet's disease. Vasogenic oedema develops when the blood-brain barrier is disrupted and is not primarily associated with cellular damage. Discrimination between cytotoxic and vasogenic oedema has important clinical implications because vasogenic oedema can be reversed by proper management.

According to the MRI findings for neuro-Behçet's disease, the most prevalent abnormalities are located in the brain stem or the basal ganglia extending to the diencephalic structures during an acute attack, and brain-stem atrophy in chronic cases.⁴ The reversibility of CT or MRI abnormalities of acute lesions in neuro-Behçet's disease has also been documented and correlated with clinical improvement.^{4,5} The serial MRI findings in our patient were consistent with those described in previous reports. The precise pathomechanism of CNS lesions in Behçet's disease has not been established. Studies of pathology showed that lymphocytic or neurophilic meningoencephalitis with perivascular inflammatory cell cuffing around venules and capillaries were predominant in the brain stem and basal ganglia in neuro-Behçet's disease.¹ However, most studies showed histopathological changes at a chronic stage of the disease and histopathological findings may show various types of lesions according to the age of lesion at the time of examination. A recent pathological report in a fulminant form of neuro-Behçet's disease found no evidence of vasculitis but

an acute destructive inflammatory process.² It has been postulated that at an early stage of the disease, the reversibility of lesions may reflect a reversible breakdown in the blood-brain barrier rather than gliosis or infarction.⁵ The pattern of diffusion changes in the acute lesions in our patient strongly supports the idea that there is increased permeability in the blood-brain barrier as a result of the primary inflammatory process.

We thank Byung Kee Yoo for his assistance with diffusion weighted MR data acquisition.

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Azathioprine and interferon β -1b treatment in relapsing-remitting multiple sclerosis

Both interferon β -1b (IFN β -1b) and azathioprine (AZA) are effective in reducing relapse frequency in relapsing-remitting multiple sclerosis (RRMS).^{1,2} However, no prospective

study has compared the efficacy of the two drugs. To assess their clinical efficacy and impact on the patients' quality of life, we performed a pilot study on a small group of patients with RRMS. Patients with at least two relapses during the previous 2 years and EDSS lower than or equal to 3.5 were offered treatment with IFN β -1b or AZA after information about the efficacy, tolerability, and mode of administration of both drugs, and allocated to one of the two treatments according to the patient's choice. Some patients refused to be treated with either drug, mainly because of the fear of side effects and the negative impact of chronic treatment on their lifestyle; they were followed up according to the same protocol (not treated (NT) group). All patients gave informed consent. Serial neurological evaluations were performed every 3 months for 1 year. At the same times a self administered disease specific questionnaire (MSQOL-54), recently validated in the Italian MS population,³ was filled in and the Hamilton depression rating scale (HD) was administered. Scores for the MSQOL-54 were analysed as previously described³; briefly, the raw scores were linearly transformed into 0-100 scales; the higher the transformed score, the better the patient's quality of life. The two composite scores mental health and physical health were also evaluated. A t test for unpaired samples was used to compare the scores between groups, with adjustment for multiple comparisons. The Kruskal-Wallis test and the two sample Wilcoxon rank sum test were used to compare the change in scores between groups. The main clinical variables were compared using a t test for unpaired and paired data.

Thirty two patients were included in the study (11 IFN β -1b, 10 AZA, 11 NT). The clinical characteristics at entry were similar in the two actively treated groups, whereas in the NT group age was significantly higher than in the AZA group (but not the IFN group) and pretreatment relapse frequency (RF) was lower than in the IFN group (but not the AZA group). After 1 year, RF significantly decreased in both the

Data of patients before and after treatment

	IFN	AZA	NT
No of patients	11	10	11*
Sex (M/F)	3/8	2/8	3/8
Age at entry (y)	33 (6.2)	31.2 (4.9)†	38 (6.3)†
Disease duration (y)	8.3 (5)	6.95 (6.7)	8.4 (6.8*)
EDSS at entry	2.32 (0.9)	2.35 (0.9)	1.83 (1.15)
At 12 months	2.2 (1.05)	2.1 (0.9)	1.9 (1.3)
No of worsened patients at 12 months	1/11	0/10	2/10
RF 2 year pretreatment	2.2 (0.8)†	2 (1)	1.4 (0.3)†
At 12 months	0.8 (0.7)‡	0.9 (0.4)‡	1.0 (0.9)
No of relapse free patients at 12 months	4/11	7/10	4/10
PHC at entry	68.02 (9.1)	61.7 (10.8)	61.7 (11.7)
Change at 12 months	-2.64 (9.26)	+7.9 (9.9)	+3.66 (13.7)
MHC at entry	74.7 (15.7)	61.25 (14.15)	58.7 (16.9)
Change at 12 months	-6.04 (13.9)†	+21.25 (11.9)†	+6.37 (21.8)
RLE at entry	83.34 (32.4)	37.5 (33.05)	55.55 (40.8)
Change at 12 months	-10 (35.3)†	+49.9 (35.6)†	+16.6 (53.4)

IFN=Interferon β -1b treated patients; AZA=azathioprine treated patients; NT=no actively treated patients; RF=relapse frequency (No of relapses/patient/y); worsened=increase of ≥ 1 EDSS point. PHC=physical composite score; MHC=mental composite score; RLE=role limitation for emotional reasons.

*One patient in NT group dropped out at 6 months.

†Significant differences between groups:

Age at entry: NT v AZA $p=0.01$.

RF at entry: NT v IFN $p=0.006$.

MHC change: IFN v AZA $p=0.006$.

RLE change: IFN v AZA $p=0.001$.

‡Significant differences within each group (12 months v entry):

RF: IFN $p<0.001$.

AZA $p=0.005$.

IFN and AZA treated groups without differences between the two treatments, whereas it was unchanged in the NT group. The EDSS remained stable in the three groups (table). Five of 11 patients treated with IFN had flu-like symptoms on one or more occasions, whereas no side effects occurred in the other two groups.

No significant differences in the HD scores and quality of life profile were found between the three groups at entry. At 6 (data not shown) and 12 months the mental health composite score significantly increased in patients treated with AZA compared with the patients treated with IFN, mainly due to the increase in role limitation for emotional reasons item; no significant differences between the NT group and actively treated groups were seen. No significant changes in HD scores in the three groups were found at 12 months. These results suggest that both AZA and IFN β -1b are effective in reducing relapse frequency in patients with RRMS. The treatment effect on quality of life has been rarely investigated, with conflicting results: no significant change after 1 year of IFN β -1b treatment was found by Schwartz *et al.*,⁴ whereas an improvement on physical items after 5 years was reported by Rice *et al.*⁵ In our study, the impact on quality of life was better in patients treated with AZA than in those treated with IFN, mainly due to the improvement in mental score. A direct effect of the drugs on the CNS seems unlikely: no symptoms of neurotoxicity were found in either treatment group and no patients developed depression according to the HD scale. Most likely the improvement of quality of life in patients treated with AZA might be related to different tolerability or to differences in treatment schedules, resulting in a more pronounced and persistent perception of the disease in patients treated with IFN. Due to the few patients, the results of this study need to be verified by a larger randomised comparative trial.

We are indebted to Dr Alessandra Solari, Laboratory of Epidemiology, C Besta National Neurological Institute, Milan, Italy, for performing the statistical analysis of the data.

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Unilateral caudate head lesion simulating brain tumour in X-linked adult onset adrenoleukodystrophy

The appearance of X-linked adrenomyeloneuropathy (AMN)/adrenoleukodystrophy (ALD) on MRI is usually specific, with bilateral symmetric areas of white matter abnormality surrounding the posterior horns of the lateral ventricles with various degrees of atrophy of the spinal cord.¹ Our patient with AMN, however, showed a lesion in the right caudate head simulating a brain tumour, which has not been a feature in this disease.

At the age of 25 the patient started to have progressive spastic paraparesis and mild ataxia with genitourinary dysfunction (urge urinary incontinence and erectile dysfunction).² On admission to our hospital at the age of 34, T2 weighted MR images showed small lesions in the bilateral internal capsule although no abnormality was seen in the spinal cord. Nerve conduction studies and the sural nerve biopsy showed evidence of

Figure 1 Brain MRI of the patient at the age of 37. T2 weighted MR images showed a high signal mass lesion in the right caudate head and the ipsilateral anterior internal capsule which simulated an intracranial tumour, without marked demyelination in the surrounding deep white matter.

Figure 2 Microscopic section of the right caudate head (haematoxylin-eosin staining, originally $\times 50$). This shows neuronal loss and tissue rarefaction with fibrillary gliosis, presenting as spongy with little inflammation.

peripheral nerve involvement. A low serum cortisol response to intravenous adrenocorticotrophic hormone and increased concentration of plasma very long chain fatty acids were consistent with a diagnosis of AMN. Three years later he showed marked emotional lability. T2 weighted MRI showed a high signal mass lesion in the right caudate head and the ipsilateral anterior internal capsule which simulated an intracranial tumour, without marked demyelination in the surrounding deep white matter (fig 1). A year later he became wheelchair bound, apathetic, and demented. Brain MRI showed right sided dominant white matter abnormalities and atrophy of the spinal cord. Three years later he died of respiratory infection and necropsy was performed. Pathological examination showed frontotemporal cortical atrophy with diffuse white matter demyelination including bilateral internal capsules, where astrocytes proliferated and lipid laden macrophages infiltrated around the small vessels. Neurons were moderately shrunken and the neuropil showed tissue rarefaction. Demyelination was also seen in the cerebellar white matter. The caudate head showed bilateral but right side dominant atrophy, where neuronal loss and tissue rarefaction with fibrillary gliosis (spongy state) were seen (fig 2).

Previous reports of X-ALD/AMN showed occasional unilateral basal ganglia involvement. Afifi *et al.* reported on a 4.8 year old boy whose MRI showed a right anterior white matter lesion extending into the ipsilateral putamen and the thalamus.³ Close *et al.* described an 8 year old boy who had a left occipitotemporal white matter lesion extending into the ipsilateral thalamus on MRI.⁴ However, the imaging pattern in our patient is unique because of the high signal mass lesion in the right caudate head and the ipsilateral anterior internal capsule without marked demyelination in the surrounding white matter, falsely suggestive of a brain tumour. There are also other demyelinating disorders simulating brain tumour which include multiple sclerosis.⁵ The findings indicate that plasma very long chain fatty acid