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Physiologically, the vestibular pathways make contact with the ocular motor system, the spinal cord, and the vestibular cortex, contributing to the stabilization of posture and perception of verticality and self motion. The tonic bilateral vestibular input builds up the actual central vestibular tone in the three major planes: horizontal or "yaw", sagittal or "pitch", and frontal or "roll". It seems that central pathways that mediate vestibular function in either of the three planes travel independently of each other, so that a specific lesion could cause a disorder restricted to one of them. The vestibular tone in the frontal or "roll" plane allows a correct perceptual, ocular, and postural alignment to the "gravitational vertical," an imbalance in this tone causes a tilt that is proportional in degree to the hypothetical lesion. Therefore it is not likely that an associated medullary ischaemia could cause the tilt effects in our patient. A mesencephalic injury could cause this clinical picture but there were no other upper brainstem symptoms and MRI was normal at this level. A supratentorial disorder is unlikely because there were no MRI alterations and there were associated postural tilt effects. In this patient, we think that cerebellar dysfunction could be responsible for the tilt effects.

The present report confirms a previously hypothesised role for the cerebellar structures in the control of perception of verticality, and may contribute to a better knowledge of the pathophysiology and the topographic diagnosis of the central vestibular syndromes.

Low striatal D2 receptor binding as assessed by [3H]IBZM SPECT in patients with writer's cramp

Writer's cramp is a form of idiopathic focal task specific dystonia. In accord with other studies on idiopathic and symptomatic dystonia, Tempel and Perlmutter suggested the presence of an abnormal striatothalamocortical drive in writer's cramp. 1 In view of the

Axial T2 weighted (SE 2000/80) MRI shows an increased signal intensity in the right hemispheric cerebellar region without mass effect, corresponding to an ischaemic infarction in the territory of the posteroinferior cerebellar artery.
possible involvement of dopaminergic striatal receptors in dystonia, we measured the availability of striatal D2 receptors in patients with writer’s cramp using [123I]IBZM SPECT.

Ten consecutive right handed patients (eight male and two female) were classified into four groups: (a) six with dystonic writer’s cramp, (b) two with dystonic writer’s cramp depending on whether or not the symptoms appeared only during writing. None of the patients had been treated with neuroleptic, dopaminergic, or anticholinergic drugs or botulinum toxin. Hypokinesia, rigidity, tremor, and bradykinesia were present in all patients. Bradykinesia of the hands was assessed with a pegboard test, measuring the time (s) required to invert eight pegs. Pegboard performance of patients was compared with that of 46 age matched controls. Results from [123I]IBZM SPECT were compared with 12 other age matched controls from an earlier study.1 A brain dedicated SPECT system, the Strichman Medical Equipment 810X, was used. Two hours after intravenous injection of approximately 185 MBq [123I]IBZM (Cygne BV, Technical University, Eindhoven), tomographic SPECT studies were performed. A maximum of 12 slices was made, starting at the orbitomeatal line and proceeding parallel to it (300 s/lice; inter­slice distance 5 mm). For analysis of specific striatal [123I]IBZM binding, two slices with the highest striatal activity were sum­mated and a template with fixed regions of interest for the striatum and occipital cortex was placed bilaterally on the summated image.2 The ratio of the striatal binding to occipital binding quantities specific binding.

The mean ages (table) did not differ among the three groups (t tests). Left and right [123I]IBZM striatal : occipital ratios were significantly lower in patients than in controls (t tests). There was no asymmetry between [123I]IBZM ratios for the hemispheres in patients or controls (repeated measures multivariate analysis of variance (MANOVA) tests involving side, group, and group by side: P > 0.05). The pegboard test was performed with writer’s cramp and controls in either hand (t tests), showing that the patients with writer’s cramp did not have bradykinesia. There was no correlation between age or duration of disease and [123I]IBZM ratios (Pearson’s correlation coefficients). None of the variables differed between patients with simple and dystonic writer’s cramp.

Our results suggest that the striatal dopaminergic system is involved in writer’s cramp given that patients with writer’s cramp had a significantly lower [123I]IBZM binding than controls. Unfortunately, lack of an accurate measure of the severity of writer’s cramp itself prevented us from studying the relation between severity of dystonia and [123I]IBZM. We did not find a correlation between [123I]IBZM ratios and duration of disease or age. This probably means that the decline in striatal D2 receptors is not linearly progressive but remains stable over many years, which accords with our clinical impression. However, because the preclinical [123I]IBZM ratios of the individual patients were not known, it is hazardous to assess rates of decline in a small cross sectional sample. The results raise some questions. Firstly, there was bilateral reduction of available striatal D2 receptors, whereas the symptoms were unilateral and there was no asymmetry between the hemispheres. Bilateral abnormality of D2 receptor activity has also been found by others.3 This bilaterality probably only means that the abnormalities found are related to particular motor dys­functions which pass undetected if not properly challenged, as shown by the fact that many patients develop writer’s cramp on the left side, if they change to writing with the left hand. Accordingly, it is also not uncommon to find involvement of the left, or fingering hand, in musicians playing keyboards, guitars, or other stringed instruments.4

A second question is why the reduced availability of [123I]IBZM binding on striatal D2 receptors is not accompanied by parkinsonism in our patients. According to well known models of basal ganglia function, overactivity of the indirect striatopallidal pathway is usually associated with parkinsonism. Because D2 receptor stimulation inhibits the indirect pathway, by contrast with the D1 receptor driven direct pathway, this decreased striatal D2 receptor binding in writer’s cramp indicates disinhibi­tion of the indirect pathway which might be expected to be accompanied by parkinsonism. In this view, we found a mean [123I]IBZM ratio to be 1.43 (SD 0.16) in patients with definite hypokinetic-rigid symptoms due to multiple system atrophy or progressive supranuclear palsy,5 which is in the same range as the values obtained in most of the present patients with writer’s cramp (1.27-1.59).

Therefore, our finding is probably better explained by loss of D2 receptors on cholinergic striatal interneurons rather than D2 receptors on striatal output neurons. This is consistent with recent evidence suggesting cholinergic interneurons is sufficient to account for the decreased density of D2 receptors in our patients.6 Furthermore, striatal cholinergic interneurons are highly represented in the sensorimotor part of the striatum and are predominantly innervated by fibres from the thalamus suggesting a feed forward modulation from thalamus to striatum.7 A dysfunction of such thalamo-striatal sensorimotor function—caused by increased activity of striatal cholinergic interneurons—might be due to D2 receptor loss—fits the suggestion that central sensorial processing in dystonia is impaired.8 Our hypothesis that writer’s cramp dystonia could be related to an increased activity of striatal cholinergic interneurons is also consistent with the increased density of striatal cholinergic interneurons in dystonia after peripheral anticholinergic injury and with the well known efficacy of anticholinergic therapy in dystonia.

CA HOFSTINK
Department of Neurology
Utrecht University Hospital Nijmegen, The Netherlands

HJC BERGER
Academic Medical Centre, Amsterdam, The Netherlands, Department of Nuclear Medicine, University Hospital Nijmegen, The Netherlands

Correspondence to: Dr. CA Hofstink, Department of Neurology, University Hospital Nijmegen, PO Box 9010, 6500 HB Nijmegen, The Netherlands.


Chronical sensory ataxic neuropathy and ophthalmoplegia with oculomotor nerve hypertrophy associated with IgM antibodies against gangliosides containing disialosyl groups

Recent studies have shown that serum anti-ganglioside antibodies may be involved in various immune mediated peripheral neuropathies. We report an investigation of anti-ganglioside antibodies in a patient with chronic sensory neuropathy and ophthalmoplegia associated with oculomotor nerve hypertrophy.

A 40 year old man had been in good health when he developed subacute diplopia in 1983. At the age of 45, he developed a moderately unsteady gait after an infection of the upper respiratory tract. The symptoms gradually worsened and he was unable to run at the age of 46. At the age of 48, he was admitted to hospital because of progressive numbness in all limbs and difficulty in performing fine motor movements. Treatment...