

# 25 Years Ago in the Canadian Journal of Neurological Sciences

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## HL-A FREQUENCIES IN PATIENTS WITH MULTIPLE SCLEROSIS

D. W. Paty, H. Mervart, B. Campling, C.G. Rand, C. R. Stiller

**SUMMARY:** The histocompatibility antigens (HL-A) have been determined in 100 multiple sclerosis (M.S.) patients and 143 randomly selected controls. In the M.S. group there was a statistically significant increase in the frequency of HL-A 7 and W 18 with an insignificant increase in HL-A 3. The variance from normal HL-A patterns in the M.S. population may play some role in establishing the substrate for this disease. Studies in experimental animals have shown that susceptibility to autoimmune disease and to virus infection is linked to the major histocompatibility locus. This has interesting implications for both the "slow virus" and the "autoimmune" theories of the etiology of multiple sclerosis.

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## SUPPRESSIVE EFFECTS OF VARIOUS AMINO ACIDS AGAINST OUABAIN-INDUCED SEIZURES IN RATS

Y. Tsukada, N. Inque, J. Donaldson A. Barbeau

**SUMMARY:** The suppressive effect of various amino acids against ouabain-induced seizures was investigated in young female rats. The amino acids were injected into the left lateral ventricle 10 minutes prior to the intraventricular administration of 5 $\mu$ g. of ouabain. Animals receiving  $1.9 \times 10^{-1}$  M solutions of hypotaurine and of B-alanine were almost completely protected from the ouabain seizures. Administration of L-alanine and of glycine was also effective, although running and leaping seizures still occurred to some extent. Betaine reduced only clonic-tonic and whole body flexion and extension seizures. In contrast, L-proline exclusively suppressed clonic-tonic and focal clonic seizures. Rats injected with isethionic acid showed increases in incidence of running and leaping seizures while L-arginine in high concentrations caused aggravation in clonic-tonic seizures. L-cysteine, even in low concentrations, also brought about an increase in the occurrence and incidence of clonic-tonic seizures. The ED<sub>50</sub> of hypotaurine was  $10.11 \times 10^{-2}$  M for running seizures and  $4.63 \times 10^{-2}$  M for clonic-tonic seizures; that of B-alanine was  $14.01 \times 10^{-2}$  M for running seizures and  $5.5 \times 10^{-2}$  M for clonic-tonic seizures. However, hypotaurine and B-alanine, the most effective compounds tested in the present studies, provided less protection than taurine previously examined by us under similar conditions (Izumi et al., 1973)

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## ELECTRICAL STIMULATION OF THE HUMAN VISUAL CORTEX Preliminary Report

Andrew Talalla, Leo Bullara, Robert Pudenz

**SUMMARY:** A feasibility study for the development of a human visual prosthesis has led several workers to observe the effects of electrical stimulation of the human visual cortex. Experience with such stimulations of three normal-sighted patients is reported. The results confirm some of the findings of other workers, but do not show that multiple phosphenes were experienced by our patients, using strictly limited parameters of stimulation.

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## SEX-LINKED HEREDITARY ATAXIC DIPLEGIA, THE BORDERLAND BETWEEN CEREBRAL PALSY AND PELIZSAEUS-MERZBACHER DISEASE

H. G. Dunn, Margaret W. Thompson, Elizabeth Bandler, L. G. Andrews

**SUMMARY:** After a review of the literature concerning hereditary cases of cerebral palsy, a family is reported in which ataxic diplegia appears to be inherited as a sex-linked and probably recessive condition occurring in 3 males in successive generations. This ataxic diplegia, occurring after an unremarkable perinatal course, is associated with mild to moderate mental retardation, congenital nystagmus and significantly small stature and prevents the acquisition of free walking. Associated extrapyramidal features may gradually become more marked, while the nystagmus may subside. The condition is similar to that described in three previous reports in the literature. No evidence of linkage with other sex-linked disorders has been found. Xga typing showed that recombination between the Xg locus and the locus for hereditary ataxic diplegia has occurred once out of three possible opportunities. In the absence of neuropathological findings or specific biochemical tests, the differential diagnosis from Pelizaeus-Merzbacher disease cannot be made with certainty. The differentiation from other progressive sex-linked neurological disorders is discussed.

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## SPINAL MYOCLONUS IN ASSOCIATION WITH HERPES ZOSTER INFECTION: Two Case Reports

G. S. Dhaliwal, D. A. McGreal

**SUMMARY:** Two cases of segmental spinal myoclonus, attributed to herpes zoster infection, are presented. The findings support the suggestion made by Campbell and Garland (1956) that "subacute myoclonic spinal neuronitis" is of viral origin. Both patients were receiving immuno-suppressive treatment when the myoclonus developed. The value of carbamazepine in therapy is mentioned.

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## LARGE ELECTROENCEPHALOGRAPHIC RESPONSES AND THEIR RELATIONSHIP TO CLEIDO-CRANIAL DYSPLASIA

Adrian Upton, Sarah Bunday, Susan Sanders

**SUMMARY:** We have reported six individuals (five certain heterozygotes for cleido-cranial-dysostosis and one possible heterozygote) who have unusual EEG findings, consisting of very large responses to photic flash stimulation at very low stimulus rates.

Such visual responses are extremely rare and have not been seen before in the experience of an EEG department over 12 years and they were not seen in 98 control subjects. It is likely that these responses are an irregular manifestation of the gene for cleido-cranial-dysplasia, and that the responses are independent of skull deformity. One importance of these responses is their demonstration in neurologically normal individuals for previously such large responses have only been reported in association with neurolipidosis. They may have neurophysiological significance in that they may reflect an unusual balance between inhibitory and excitatory mechanisms in the nervous system.

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## POLYARTERITIS NODOSA COMPLICATED BY A MULTIPLE SCLEROSIS LIKE SYNDROME

H. Waisburg, K. L. Meloff, R. Buncic

**SUMMARY:** A case is presented of a 16-year-old boy with angiographically proven polyarteritis nodosa who developed a multiple sclerosis like syndrome affecting the brain stem and cerebrum. His serum demyelinated nerve in tissue culture. The case is reviewed in detail and the mechanism of myelotoxicity is discussed.

Can. J. Neurol. Sci. 1974; 4:250