# Baclofen As a Treatment for Nystagmus

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Three cases of periodic alternating nystagmus treated with baclofen were reported by Halmagyi et al [1]. We have observed the same phenomenon in a 65-year-old man suffering from multiple sclerosis. Since 1948 this patient has experienced five attacks of the disease, which have left him with a bilateral pyramidal syndrome and a moderate static cerebellar syndrome. Periodic alternating nystagmus appeared in 1970. It is regular and changes orientation every 110 seconds.

In October, 1979, a regimen of 20 mg of baclofen per day over 48 hours put an end to the patient's periodic alternating nystagmus. We have been able to observe on several occasions that when treatment was interrupted, the abnormal eye movement reappeared after three days. Under treatment the patient had only a first-degree multidirectional nystagmus. We have tried to replace baclofen with diazepam, 5-hydroxytryptophan, benserazide, thiocolchicoside, valproic acid, or carbamazepine, but none of these agents has had any effect.

Like Halmagyi and his colleagues, we have noticed that baclofen has no effect on congenital nystagmus, whereas 5-hydroxytryptophan has reduced this nystagmus considerably. This reduction appears clearly on oculographic recordings, and the visual acuity of such patients, which is diminished by the abnormal movement, can be improved by baclofen treatment, evolving, for example, from 20/100 to 20/30 [2].

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# Adjunctive Tests for the Mucopolysaccharidoses

Sally Kelly, PhD, MD

The recommendation by Markesbery et al [3] that ultrastructural studies of lysosomes be used as aids to the diagnosis of the mucopolysaccharidoses (MPS) is not well founded, as the features they describe are nonspecific. For example, in both Hunter (MPS II) and Sanfilippo (MPS III) disease they found fingerprint profiles (FP), structures which had been detected earlier in Hurler disease (MPS IH) [5]. Thus, FP were far less distinctive than the metachromatic granules [1], mucopolysaccharide excretion patterns [2], and enzyme defects [4] of the respective diseases. Nor were differences in multiple vacuolation helpful, since the feature appeared not only in MPS VII ($\beta$-glucuronidase deficiency) disease but in other mucopolysaccharidoses as well.

Furthermore, there were no major differences in the frequencies of clear and granular vacuoles in the various mucopolysaccharidoses. The frequency distributions resembled those of a continuous variation between clear and densely granular vacuoles with a peak frequency of faintly granular vacuoles. They suggested equilibria between vacuoles of a heterogeneous, metabolically active population or between vacuoles in a homogeneous, static population exposed to preparative factors that may have caused granulation artifacts.

The resemblance among lysosomes in MPS I, II, and III is not surprising since the three diseases share a metabolic defect, degradation of heparan sulfate. Furthermore, the anticoagulant heparin may have masked differences in these lysosomes, as the systems of heparan sulfate degradation were already compromised.

The ultrastructural differences apparently are subtle and must be searched for systematically. One expects the differences to be few when the biochemical and hematological features are similar, as in Hurler/Scheie and Hunter diseases (as Markesbery et al found), and greater when the features are unique, as in Morquio disease (MPS IV), in which keratan sulfate is excreted, or in Maroteaux-Lamy disease (MPS VI), in which the leukocyte granulation is striking.

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## Reply

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Dr Kelly has made several incorrect assumptions from our report [5]. The purpose of our study was to define the ultrastructural characteristics and frequency of the cytoplas-