or large, are all autofluorescent, stain with Sudan stains, resist lipid solvents, and have characteristic ultrastructure. Cases of GM₁ gangliosidosis also have extraneuronal lesions (notably in liver, kidney,⁴ fibroblasts, and endothelium⁵); nonautofluorescent; easily dissolved out, therefore hard to stain except on frozen sections; seen by electronmicroscopy as clear vacuoles; and sometimes containing tubular profiles. In the light of these facts, there is no reason why anyone should have to speculate that the patient of Lowden et al may have had two diseases.

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Reply from the Authors: Unfortunately, Dr. Carpenter has misinterpreted what we are saying. Our case report is not that of Batten disease but type 2 GM₁ gangliosidosis with the unusual features of long survival and accumulation of ceroid lipofuscin.^{1,2}

The recognized criteria^{3,4} for morphologic demonstration of ceroid lipofuscin are: (1) autofluorescence, (2) insolubility in lipid solvents (acetone, chloroform: methanol, absolute methanol), (3) positivity on frozen and paraffin sections with Sudan black B, Ziehl-Neelsen, oil red O, and PAS stains, and (4) variable ultrastructural patterns (fingerprint profiles and osmiophilic amorphous and granular deposits).

In the 17-year-old girl with GM₁ gangliosidosis, material had accumulated within neurons that met each of these four criteria. Therefore, there was no doubt that there was accumulation of ceroid lipofuscin. We did not state that the fingerprint profiles were identical to those of Batten disease, and we were careful to say "fingerprint-like" profiles. Parallel single lamellae were illustrated although, in areas, parallel paired lines could also be demonstrated along with osmiophilic granular deposits. The additional presence of autofluorescent material in lymph nodes and spleen could result in confusion, not necessarily with Batten disease but with other poorly delineated neurologic disorders⁵ in which

there is an unexplained accumulation of ceroid lipofuscin.

We maintain that diagnostic problems of variable degree can occur in such a case. This can be significantly reduced by widespread tissue sampling for histochemistry, electronmicroscopy, and biochemical analysis. We believe it is worthwhile to freeze portions of the brain in cases of severe cerebral atrophy to rule out an enzymatic defect.

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Correction

O.H. PERURENA should be added as the first author for "Insulin Receptors (IR) and Insulin Sensitivity (IS) in Amyotrophic Lateral Sclerosis (ALS)," April (2) 1982, p. A105.

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Correction

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