In the second case, the fourfold increase in sulphatides of the white matter was accompanied by an increase of hexosamine in the white matter and a slight loss of total phospholipid content (less significant in cortex than in white matter) and of sphingomyelin and cholesterol. Neuraminic acid was slightly increased in the cortex.

The biochemical studies suggest that both cases fall into the group of metachromatic leukodystrophy and that the derangement is one in which there is some abnormality in one of the myelin components. This abnormality would appear less likely to be one resulting from an unusual demyelination than a consequence of a faulty formation of myelin in that a great amount of a lipid material normally present is found in the cerebral white matter.

These cases are considered as showing the histopathological and histochemical features of both metachromatic leukodystrophy and amaurotic idiocy. We would suppose that the inborn enzymatic disorder underlying the two conditions may produce changes in the whole group of sphingolipids, leading to disease states in which the biochemical and histochemical features varied according to the position of the defect in the enzymatic system. It might, for example, result in alteration mainly of cellular lipids as in amaurotic idiocy or of myelin lipids as in metachromatic leukodystrophy. Such a hypothesis suggests the possible existence of intermediate forms of the disease showing simultaneously alteration in both cellular and myelin lipids. We consider our cases exemplify just such a possibility.