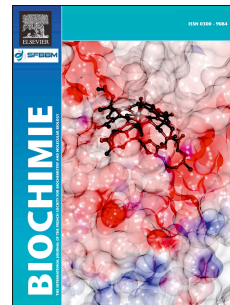


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Membrane lipids and sphingolipid activator proteins regulate lysosomal sphingolipid catabolism

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## Neuronal Sphingolipidoses

### Membrane lipids and sphingolipid activator proteins regulate lysosomal sphingolipid catabolism <sup>1</sup>

#### Abstract

Glycosphingolipids and sphingolipids of cellular plasma membranes (PMs) reach luminal intra-lysosomal vesicles (LVs) for degradation mainly by pathways of endocytosis. After a sorting and maturation process (e. g. degradation of sphingomyelin (SM) and secretion of cholesterol), sphingolipids of the LVs are digested by soluble enzymes with the help of activator (lipid binding and transfer) proteins. Inherited defects of lipid-cleaving enzymes and lipid binding and transfer proteins cause manifold and fatal, often neurodegenerative diseases.

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<sup>1</sup> A short version of the presentation given at the 11th GERLI Lipidomic Meeting, Oct. 25-28, 2015, in Bischoffsheim, France

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