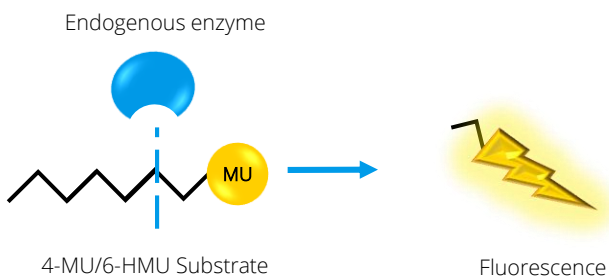


# Fluorogenic Enzyme Substrates

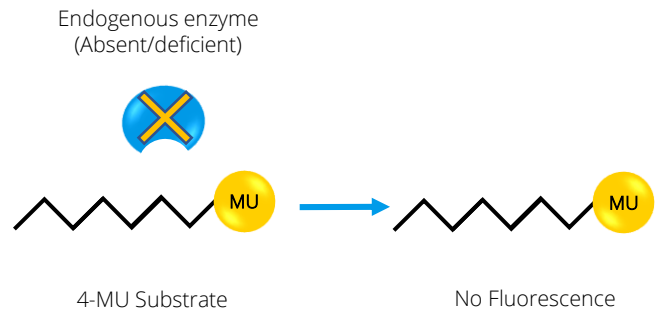
Biosynth Carbosynth offers a range of fluorogenic substrates for lysosomal enzymes that can be employed for diagnosis of lysosomal storage disorders. The products in this range are of high chemical purity, as well as biochemical purity.

Lysosomal storage disorders consists of 50 different genetically inherited metabolic diseases, the majority of which are due to deficiency in lysosomal enzymes. The resulting accumulation of unprocessed metabolites in lysosomes leads to the stimulation of pathogenetic cascades that have implications on multiple systems in the body.

## A. In unaffected individuals



## B. In affected individuals



**Fluorogenic lysosomal enzyme substrates in diagnostic assays.** This scheme displays the mechanism of measuring lysosomal enzyme activity using fluorogenic substrates for diagnosis of lysosomal storage disease. **A.** In unaffected individuals, the 4-MU or 6-HMU labelled enzyme substrate is cleaved by endogenous enzymes. The cleaved fragment elicits a fluorescent signal that can be detected and quantified. **B.** In patients affected by LSD, the endogenous enzyme is deficient and therefore no or reduced fluorescence is detected.

Code	Product	Activity	Quantity	Price (\$)
EM06650	4MU-6-thiopalmityl-β-D-glucoside	Substrate of palmitoyl-protein thioesterase; for diagnosis of INCL	1 mg	60.00
EM03201	4MU-α-L-iduronide-2-sulphate	Substrate of iduronate-2-sulfatase (IDS); for diagnosis of MPS II	0.5 mg	150.00
EM04782	4MU-α-N-acetyl-D-galactosamide	Substrate of N-acetyl-α-D-galactopyranosaminidase; for diagnosis of Schindler disease	5 mg	75.00
EM31027	4MU-α-N-acetyl-D-glucosaminide-6-sulphate	Substrate of N-acetylglucosamine-6-sulphate sulphatase; for diagnosis of MPS II D	1 mg	115.00
EM06602	4MU-α-N-sulpho-D-glucosaminide	Substrate for heparin sulphamidase; for diagnosis of MPS III A	2 mg	60.00
EM05134	4MU-β-D-galactoside-6-sulphate	Substrate of N-acetylgalactosamine-6-sulphatase; for diagnosis of MPS IV A	5 mg	65.00
EM31025	4MU-β-D-glucosaminide	Substrate for heparin sulphamidase; for diagnosis of MPS III C	1 mg	90.00
EH10520	6-Hexadecanoylamino-4-methylumbelliferone	Substrate for galactocerebroside-β-galactosidase; for diagnosis of Krabbe's disease	5 mg	45.00
EH05989	6HMU-β-D-galactoside	Substrate for galactocerebroside; for diagnosis of Krabbe's disease	1 mg	75.00
EH31028	6HMU-phosphorylcholine	Substrate for sphingomyelinase; for diagnosis of Niemann-Pick AVB disease	1 mg	90.00