

Appendix

Table A1: MALDI-TOF MS and LC-MS/MS analyses of lysosome-enriched fraction

Coomassie Blue-stained Spots (#) ^a		Man6P eluate ^b	Non bound ^c
Known lysosomal proteins			
Legumain	(#59→68)	Legumain	Legumain
Dipeptidyl peptidase 1	(#56→58)	Dipeptidyl peptidase 1	Dipeptidyl peptidase 1
Protein CREG1	(#46→51)	Protein CREG1	Protein CREG1
Cathepsin D	(#26→38)	Cathepsin D	Cathepsin D
Cathepsin Z	(#38→44)	Cathepsin Z	
α -N-acetylgalactosaminidase	(#16→25)	α -N-acetylgalactosaminidase	α -N-acetylgalactosaminidase
Cathepsin A	(#45)	β -hexosaminidase subunit α	β -hexosaminidase subunit α
Dipeptidyl peptidase 2	(#9→15)	Dipeptidyl peptidase 2	Dipeptidyl peptidase 2
Epididymal secretory protein E1 (Npc2)	(#51→54)	Lysosomal α -glucosidase	Lysosomal α -glucosidase
Sphingomyelin phosphodiesterase, acid like 3a	(#1→3)	Lysosomal α -mannosidase	Lysosomal α -mannosidase
Palmitoyl-protein Thioesterase 1	(#69→71)	Palmitoyl-protein Thioesterase 1	Palmitoyl-protein Thioesterase 1
		Group XV phospholipase A ₂	Group XV phospholipase A ₂
		Epididymis-specific α -mannosidase	
		Phospholipase D4	Phospholipase D4
Non known lysosomal proteins		β -glucuronidase	
Serpin1AC	(#72)	Retinoid-inducible carboxypeptidase	serine
Fibrinogen γ	(#4→8)	Putative phospholipase B-like 2	
		β -galactosidase	
		Pro-cathepsin H	
		Sphingomyelin phosphodiesterase	
		Plasma glutamate carboxypeptidase	
		β -hexosaminidase subunit β	
		Lysosomal acid lipase/cholesteryl ester hydrolase	

		Cathepsin B
Mammalian	ependymin-related	
protein 1		
N(4)-(β-N-acetylglucosaminy1)-L-asparaginase		N(4)-(β-N-acetylglucosaminy1)-L-asparaginase
Phospholipase B-like 1		
Serine protease inhibitor A3C		
Ferritin light chain 1		
		Apolipoprotein E
Hemoglobin subunit epsilon-Y2		Hemoglobin subunit epsilon-Y2
Ribonuclease UK114		
Ribonuclease T2		
Prefoldin subunit 1		
Elongation factor 2		
AT-rich	interactive	domain-
containing protein 1A		
		ATP synthase subunit □, mitochondrial
		ATP synthase subunit □, mitochondrial
		Copine-3
		Peroxisomal bifunctional enzyme
		Lysosomal protein NCU-G1
		Lysosome-associated membrane glycoprotein 2
		Probable lysosomal cobalamin transporter
		Prolow-density lipoprotein receptor-related protein
		Long-chain-fatty-acid-CoA ligase 1
		Ig γ-2B chain C r
		V-type proton ATPase subunit E

Transmembrane protein 106

α -soluble NSF attachment protein

Hydroxyacyl-coenzyme A dehydrogenase, mitochondrial

V-type proton ATPase subunit β , brain isoform

β -actin-like protein 2

Succinate dehydrogenase [ubiquinone] flavoprotein subunit, mitochondrial

Receptor-type tyrosine-protein phosphatase C

Protein disulfide-isomerase A3

ATP-binding cassette sub-family B member 6, mitochondrial

Phospholipid transfer protein

Amine oxidase [flavin-containing] B

Serum albumin

Fibronectin

Serotransferrin

^a2D separation of 200 μ g of lysosome-enriched fraction prepared from *Acp2/Acp5* ^{-/-} mice and stained by Coomassie Blue (n=3). The numbering (#) corresponds to identified spots encircled in Supplementary Figure 2

^bMan6P eluate fraction after Man6P-affinity chromatography of lysosome-enriched fraction (150 μ g) prepared from *Acp2/Acp5* ^{-/-} mice

^cNon bound fraction after Man6P-affinity chromatography of lysosome-enriched fraction prepared from *Acp2/Acp5* ^{-/-} mice