

SUPPLEMENTARY MATERIAL

Unbiased cell-based screening in a neuronal cell model of Batten disease highlights an interaction between Ca²⁺ homeostasis, autophagy, and CLN3 function

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*Running title: *Identification of autophagy modifiers in Batten disease*

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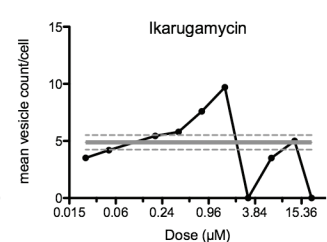
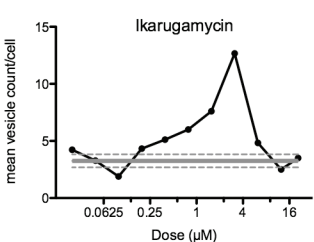
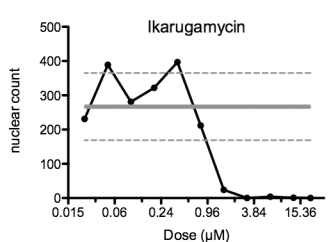
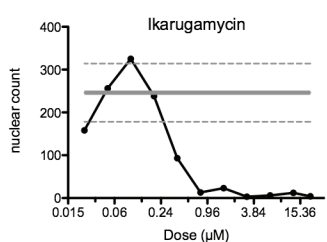
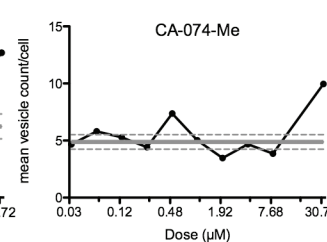
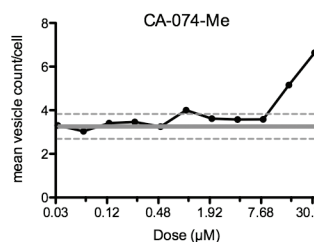
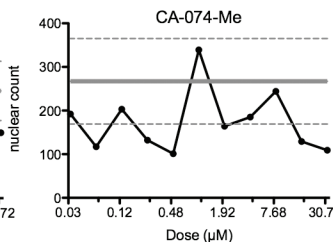
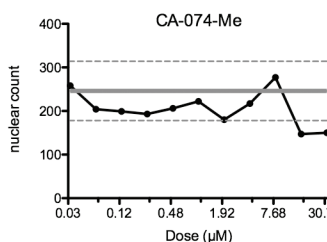
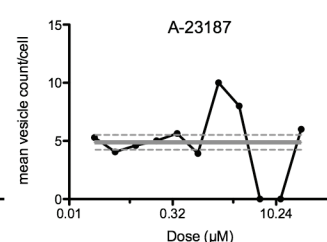
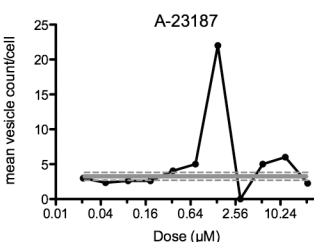
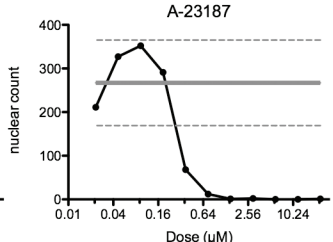
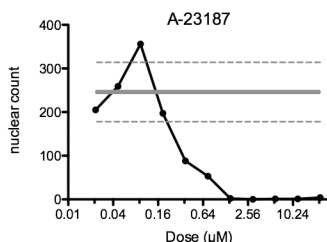
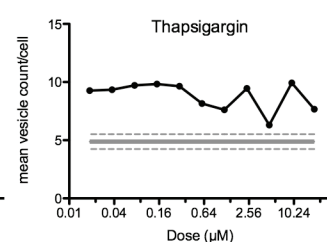
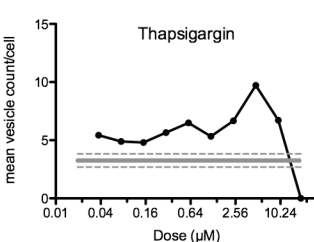
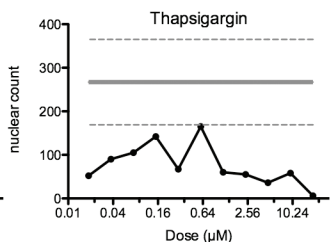
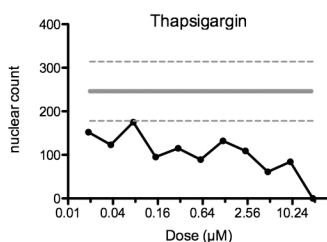
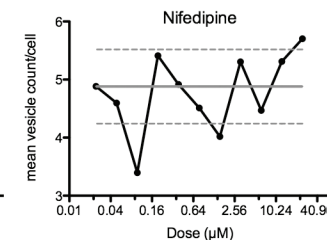
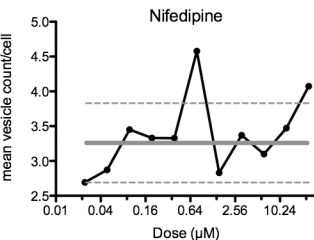
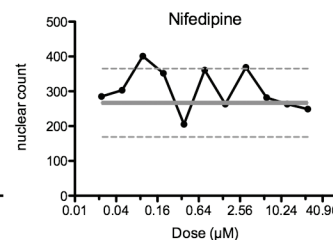
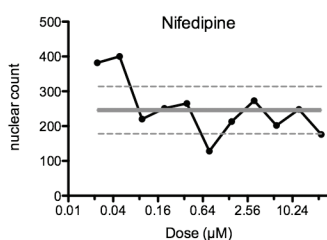
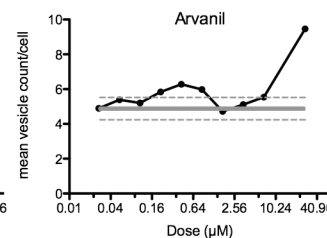
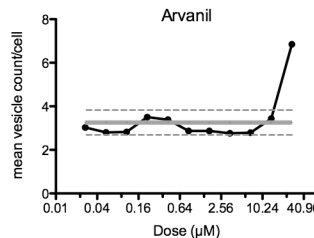
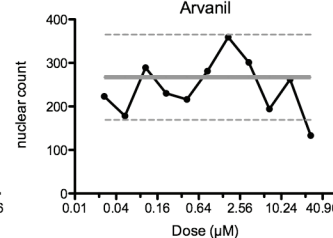
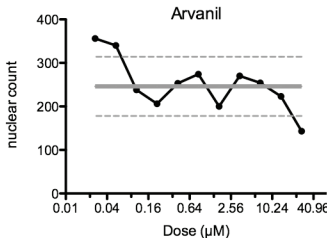
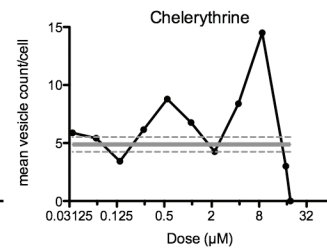
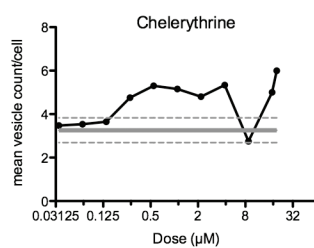
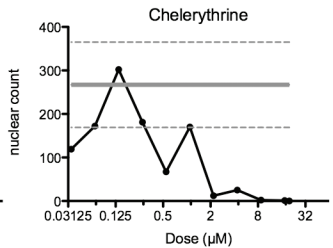
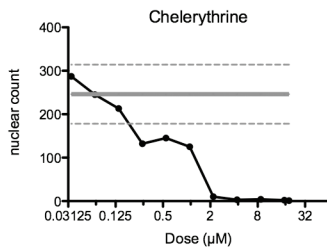
LEGENDS TO SUPPLEMENTARY FIGURES:

Supplementary Figure 1: Follow-up dose response for hit compounds:

Left columns ('Nuclei'): Graphs of nuclear count (y-axis) for each follow-up dose (x-axis, μM) tested are shown in the left set of plots for the indicated compounds, which are provided as an indication of the relative toxicity following the compound dose-response experiment. Right columns ('GFP-LC3 vesicle count per cell'): Graphs of mean vesicle count/cell (y-axis) for each follow-up dose tested (x-axis, μM) are shown in the right set of plots for the indicated compounds. The gray line shown in each graph represents the mean for DMSO-treated wells, and the dashed lines represent \pm two standard deviations from the mean.

Nuclei

GFP-LC3 vesicle count per cell

*CbCln3^{+/+}**CbCln3^{Δex7/8/Δex7/8}**CbCln3^{+/+}**CbCln3^{Δex7/8/Δex7/8}*

SUPPLEMENTARY TABLES and LEGENDS

Supplementary Tables 1 and 2 are uploaded as separate .xls (or .xlsx) files.

File name: Supplementary Table 1.xlsx

Supplementary Table 1. Hit list from primary GFP-LC3 screen run on duplicate plates of wildtype (*CbCln3*^{+/+}) and homozygous mutant (*CbCln3*^{Δex7/8/Δex7/8}) cells.

Predicted mechanism of action information was determined using the PubChem database (pubchem.ncbi.nlm.nih.gov) and/or Enzo Life Sciences (BIOMOL) supplier data (www.enzolifesciences.com).

File name: Supplementary Table 2.xls

Supplementary Table 2. All compounds primary screen data. Per plate z-scores are shown for each of the 320 compounds in the primary screen, for *CbCln3*^{+/+} and for *CbCln3*^{Δex7/8/Δex7/8} cells. The mean z-score for the duplicate plates is also shown for each compound in both the *CbCln3*^{+/+} and for *CbCln3*^{Δex7/8/Δex7/8} cells.