

### **Supplemental Figure Legend**

Supplement Figure 1. PAS staining in the cerebral cortex of CD<sup>+/-</sup> (A) and CD<sup>-/-</sup> (B) mice at P23 and PAS staining (C, D) and autofluorescence (E-G) in the cerebral cortex of CB<sup>+/-</sup>-CL<sup>+/-</sup> (C) and CB<sup>-/-</sup>-CL<sup>-/-</sup> (D-G) mice at P13. Positive staining of PAS is intensely detected in both CD<sup>-/-</sup> and CB<sup>-/-</sup>-CL<sup>-/-</sup> mouse cerebral cortical neurons, but not in their littermate control neurons. Autofluorescence is visible in cerebral cortical neurons (arrowheads) and F4/80-positive microglial cells (arrows). Scale bars: 20  $\mu$ m.

Supplement Figure 2. Electron micrographs of accumulated membrane compartments in a dendrite (A) and axon (B) observed in the same cerebellar region of a CB<sup>-/-</sup>-CL<sup>-/-</sup> mouse. The accumulated structures in the dendrite are GROD-like inclusions and AVs, which are seen in the perikarya of Purkinje cells, (A), while those in the axon are largely autophagosomes (AVi) that are encircled by double membrane saccules. Scale bar: 1  $\mu$ m.

Supplement Figure 3. Electron micrographs of neuronal cells from CD<sup>+/-</sup> (A) and CB<sup>+/-</sup>-CL<sup>+/-</sup> (B, C) mouse brains. A, B. Perikaryal regions of cerebellar Purkinje cells obtained from a CD<sup>+/-</sup> mouse at P23 (A) and a CB<sup>+/-</sup>-CL<sup>+/-</sup> mouse at P12 (B). C. Myelinated and unmyelinated axons located in the corpus callosum of the mouse in B. Scale bar: 1  $\mu$ m.





