CLN3 Research Tools and Resources

Original References

Amoeba
1. Dictyostelium discoideum
   - Huber RJ, Myre MA, Cotman SL. Loss of CLN3 function in the social amoeba Dictyostelium discoideum causes pleiotropic effects that are rescued by human CLN3.

Yeast Models
1. Schizosaccharomyces cerevisiae
2. Schizosaccharomyces pombe

Roundworm
1. Caenorhabditis elegans

Fruitfly
1. Drosophila melanogaster - Genetic gain-of-function system

Zebrafish
1. Danio rerio - Morphant

Mouse Models
1. Mus musculus - Deletion of exons 2-6 and most of exon 1 on 129/Sv inbred and 129Sv3 Black Swiss outbred lines
2. *Mus musculus* - Cln3Δex7/8 knock-in mice bearing the common ~1 kb deletion on an out-bred 129Sv-CD1 background

3. *Mus musculus* - Cln3Δex7/8 knock-out Exons 7-8 replacement insertion on C57BL/6J-129Sv and C57BL/6J congenic

4. *Mus musculus* - Targeted replacement of most of exon1 and all of exons 2-8 of the CLN3 gene with β-galactosidase on SV129 and onto C57BL/6

5. *Mus musculus* – CLN3Δex7/8 mice carrying the leucine variant of RPE65 on a C57BL/6J background
   - Dannhausen, K., Moble, C., Langmann T. Immunomodulation with minocycline rescues retinal degeneration in juvenile neuronal ceroid lipofuscinosis mice highly susceptible to light damage. Dis ModelMech. 2018. Sep 5;11(9).

6. *Mus musculus* – TgCRND8/Cln3Δex7/8 mice homozygous for Cln3Δex7/8 with the hAPP transgene

Porcine Model
1. CLN3Δex7-8/Δex7-8 porcine model of CLN3-Batten

Animal Cell lines
1. Hela CLN3 KO line

2. *Mus musculus* - Homozygous ChCln3Deltaex7/8 precursor cells

3. *Mus musculus* - Mouse Brain Endothelial Cells
   - Tecedor L, Stein CS, Schultz MS, et al. CLN3 Loss Disturbs Membrane Microdomain Properties and Protein Transport in Brain Endothelial Cells

Human
1. CLN3 deficient
   - Homo sapien - Human CLN3 Knockout cell line 19 bp deletion
   - Homo sapien - Human CLN3 Knockout cell line 8 bp deletion
   - Panoply™ Human CLN3 Knockdown Stable Cell Line (CSC-DC003300)
• Homo sapien - Induced pluripotent stem cells

2. CLN3 overexpression
• Homo sapien - Cancer cell lines
    • Glioblastoma (U-373G, T98g)
    • Neuroblastoma (IMR-32, SH-SY5Y, SK-N-MC)
    • Prostate (Du145, PC-3, LNCaP)
    • Ovarian (SK-OV-3, SW626, PA-1)
    • Breast (BT-20, BT-549, BT-474)
    • Colon (SW1116, SW480, HCT 116)
• Panoply™ Human CLN3 Knockdown Stable Cell Line(CSC-DC003300)

NCL Mutation and Patient Database http://www.ucl.ac.uk/ncl/mutation.shtml
“"This database contains published mutations and sequence variations in genes that cause NCL together with unpublished data included with permission. It follows the mutation nomenclature recommendations of the Human Genome Variation Society. From mid-2012, there are now two sets of tables for each human NCL disease gene - the new Patient Database listing all published or reported patients and families, and the Mutation Database listing all published or reported mutations and many sequence variants as before, and now cross-referenced to the patient table. These are available to view via this web site and also to download as excel files for off-site use to aid local needs or interests (e.g. sorting according to occurrence in particular countries).”"