SUMMARY OF NEW CLASSIFICATION NOMENCLATURE OF THE NCLS

Gene	Disease Name	Also Known As	Gene Product (Protein)	Protein Description
CLN1	CLN1 disease, infantile CLN1 disease, late infantile CLN1 disease, juvenile CLN1 disease, adult	Infantile	Palmitoyl protein thioesterase 1, PPT1	soluble lysosomal enzyme deficiency
CLN2	CLN2 disease, late infantile CLN2 disease, juvenile	Late-Infantile	Tripeptidyl peptidase 1, TPP1	soluble lysosomal enzyme deficiency
CLN3	CLN3 disease	Juvenile	CLN3 transmembrane protein	transmembrane protein
CLN4/DNAJC5	CLN4 disease	Adult autosomal dominant Batten Kuf's disease Ceroid Lipofuscinosis Parry type	Cysteine string proteinα	secretory vesicle protein
CLN5	CLN5 disease, late infantile CLN5 disease, juvenile CLN5 disease, adult	Finnish variant late-infantile	Ceroid-lipofuscinosis neuronal protein 5	soluble lysosomal enzyme deficiency
CLN6	CLN6 disease, late infantile CLN6 disease, juvenile CLN6 disease, adult	Early juvenile variant or late-infantile variant	Ceroid-lipofuscinosis neuronal protein 6	transmembrane protein, endoplasmic reticulum
CLN7	CLN7 disease, late infantile	Late-infantile variant	Major facilitator superfamily domain-containing protein 8	transmembrane protein, endolysosomal transporter
CLN8	CLN8 disease, late infantile	Late-infantile variant EPMR (progressive epilepsy with mental retardation)	unknown transmembrane protein, ER, ER-Golgi intermediate complex	transmembrane protein, endoplasmic reticulum , ER-Golgi intermediate complex
CLN10/CTSD	CLN10 disease, congenital CLN10 disease, late infantile CLN10 disease, juvenile CLN10 disease, adult	Congenital classic Late-infantile Adult	Cathepsin D	soluble lysosomal enzyme deficiency
CLN11/GRN	CLN11 disease, adult	Adult (heterozygous mutations cause frontotemporal lobar dementia)	Progranulin	non enzyme; function of protein poorly understood
CLN12/ATP13A2	CLN12 disease, juvenile	CLN12 disease Juvenile (mutations also cause Kufor-Rakeb syndrome)	P-type ATPase	non enzyme; function of protein poorly understood
CLN13	CLN13 disease, adult	Adult Kufs type B	Cathepsin F	soluble lysosomal enzyme deficiency
CLN14/KCTD7	CLN14 disease, infantile	CLN14 disease, infantile	Potassium channel tetramerization domain-containing protein 7	probable transmembrane protein voltage-gated potassium channel complex