



CLN2, Ceroid-Lipofuscinoses, Neuronal 2 [domain 1] polyclonal antibody

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Box 1 | Basic Info

Cat. No.	ABP-PAB-11620
Animal ID	RB1115/RB1116
Host	Rabbit
Reactivity	Human, Monkey, Bovine, Dog
Format	Purified
Accession number	NM_000391
Amount	100 µg

Alternative Name(s):

Jansky-Bielschowsky disease protein,
TPP1, LINCL, TPPI, TPP-I

References:

1. Barisic N, Logan P, Pikija S, Skarpa D, Blau N: R208X mutation in CLN2 gene associated with reduced cerebrospinal fluid pterins in a girl with classic late infantile neuronal ceroid lipofuscinosis. *Croat. Med. J.* 44(4): 489-493 (2003).
2. Golabek AA, Kida E, Walus M, Wujek P, Mehta P, Wisniewski KE: Biosynthesis, glycosylation, and enzymatic processing in vivo of human tripeptidyl-peptidase I. *J. Biol. Chem.* 278(9): 7135-7145 (2003).

The neuronal ceroid lipofuscinoses (NCL), also known as Batten disease, are a group of inherited severe neurodegenerative disorders primarily affecting children. They are characterised by the accumulation of autofluorescent storage material in many cells. Six genes have been identified (CLN1, CLN2, CLN3, CLN5, CLN6, CLN8). Children suffer from visual failure, seizures, progressive physical and mental decline and premature death, associated with the loss of cortical neurones. Neuronal ceroid lipofuscinosis (CLN2) is a lysosomal storage disease which results from mutations in the gene that codes for tripeptidyl peptidase-I (TPP-I). CLN2 mutations in humans cause the classic late infantile neuronal ceroid-lipofuscinoses type 2. CLN2 removes tripeptides from the free N termini of small polypeptides and also shows a minor endoprotease activity.

Buffers

Purified rabbit polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein G column and eluted out with both high and low pH buffers and neutralized immediately after elution then followed by dialysis against PBS.

Immunogen

KLH conjugated synthetic peptide comprised of amino acids 19 - 33 [SYSPEPDQRRTLPPG] of the human CLN2 [domain 1] protein.

Application

Tested by peptide-specific ELISA (1:1,000).

Storage

Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C. Avoid repeated freeze-thaw cycles.