



## Rabbit Anti-CLN3 antibody

SL8016R

<b>Product Name:</b>	CLN3
<b>Chinese Name:</b>	神经细胞蜡样质脂褐质沉积病蛋白CLN3抗体
<b>Alias:</b>	Batten disease protein; Battenin; BTS; Ceroid lipofuscinosis neuronal 3; Ceroid lipofuscinosis neuronal 3 juvenile (Batten Spielmeier Vogt disease); Ceroid lipofuscinosis neuronal 3 juvenile; CLN 3; MGC102840; Protein CLN3; CLN3_HUMAN.
<b>Organism Species:</b>	Rabbit
<b>Clonality:</b>	Polyclonal
<b>React Species:</b>	Human,Mouse,Rat,Cow,Horse,Rabbit,Monkey,macaque
<b>Applications:</b>	WB=1:500-2000ELISA=1:500-1000 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
<b>Molecular weight:</b>	48kDa
<b>Cellular localization:</b>	cytoplasmicThe cell membrane
<b>Form:</b>	Lyophilized or Liquid
<b>Concentration:</b>	1mg/ml
<b>immunogen:</b>	KLH conjugated synthetic peptide derived from human CLN3:75-140/438
<b>Lsotype:</b>	IgG
<b>Purification:</b>	affinity purified by Protein A
<b>Storage Buffer:</b>	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
<b>Storage:</b>	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
<b>PubMed:</b>	<a href="#">PubMed</a>
<b>Product Detail:</b>	This gene encodes a protein that is involved in lysosomal function. Mutations in this, as well as other neuronal ceroid-lipofuscinosis (CLN) genes, cause neurodegenerative diseases commonly known as Batten disease or collectively known as neuronal ceroid lipofuscinoses (NCLs). Many alternatively spliced transcript variants have been found for this gene.

**Function:**

Involved in microtubule-dependent, anterograde transport of late endosomes and lysosomes.

**Subunit:**

Interacts with DCTN1 and KIF3A. Interacts with RAB7A and RILP.

**Subcellular Location:**

Lysosome membrane; Multi-pass membrane protein. Late endosome

**Post-translational modifications:**

Highly glycosylated.

Farnesylation is important for trafficking to lysosomes.

**DISEASE:**

Defects in CLN3 are the cause of neuronal ceroid lipofuscinosis type 3 (CLN3) [MIM:204200]; also known as Batten disease. A form of neuronal ceroid lipofuscinosis. Neuronal ceroid lipofuscinoses are progressive neurodegenerative, lysosomal storage diseases characterized by intracellular accumulation of autofluorescent liposomal material, and clinically by seizures, dementia, visual loss, and/or cerebral atrophy. The hallmark of CLN3 is the ultrastructural pattern of lipopigment with a fingerprint profile, which can have 3 different appearances: pure within a lysosomal residual body; in conjunction with curvilinear or rectilinear profiles; and as a small component within large membrane-bound lysosomal vacuoles. The combination of fingerprint profiles within lysosomal vacuoles is a regular feature of blood lymphocytes from patients with CLN3.

**Similarity:**

Belongs to the battenin family.

**SWISS:**

Q13286

**Gene ID:**

1201

**Database links:**

[Entrez Gene: 1201](#)Human

[Entrez Gene: 12752](#)Mouse

[Entrez Gene: 293485](#)Rat

[Omim: 607042](#)Human

[SwissProt: Q13286](#)Human

[SwissProt: Q61124](#)Mouse

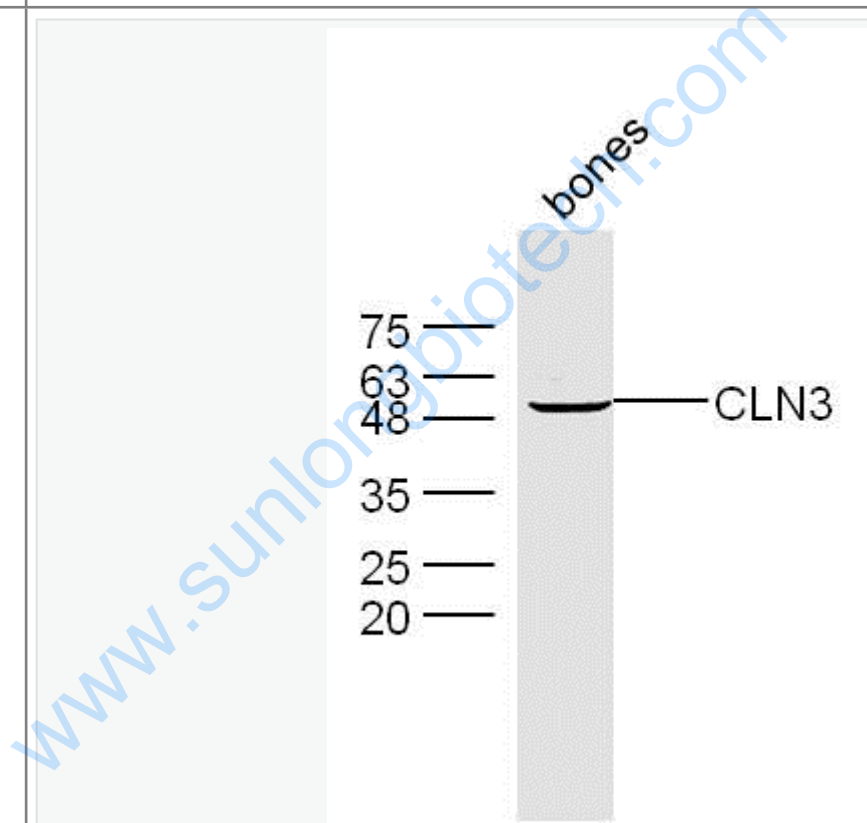
[Unigene: 534667](#)Human

[Unigene: 268930](#)Mouse

**Important Note:**

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

**Picture:**



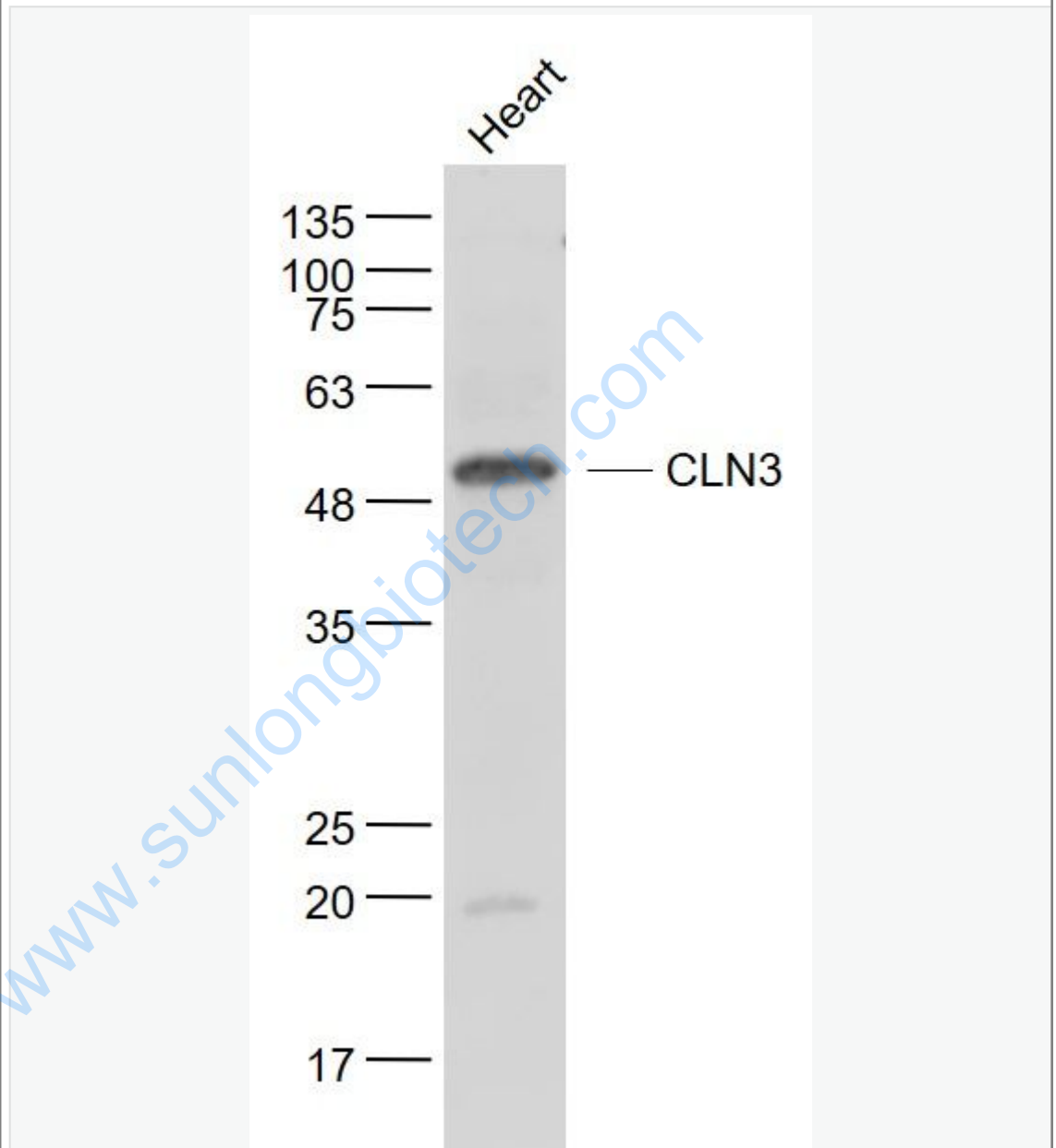
Sample: Bone (Mouse) Lysate at 40 ug

Primary: Anti-CLN3 (SL8016R) at 1/300 dilution

Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

Predicted band size: 48 kD

Observed band size: 50 kD



Sample:

Heart (Mouse) Lysate at 40 ug

Primary: Anti- CLN3 (SL8016R) at 1/1000 dilution

Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

	<p>Predicted band size: 48 kD</p>
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	<p>Observed band size: 50 kD</p>
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