



CLN5 (h5): 293T Lysate: sc-372537

BACKGROUND

Neuronal ceroid-lipofuscinosis (NCL), also designated Batten disease, comprises a group of recessively inherited, progressive neurodegenerative diseases found in children. NCL is characterized by atrophy of the brain and an accumulation of lysosome derived fluorescent bodies found in many cells, especially neurons. Symptoms of NCL include a failure of psychomotor development, seizures, impaired vision and premature death. The eight genes/proteins associated with NCL are designated CLN1-CLN8. Mutations in six of these genes results in a distinct type of NCL-disease; the six genes/proteins are CLN1 (encoding PPT1, a protein thioesterase), CLN2 (encoding the serine protease TPP1), CLN3, CLN5, CLN6 and CLN8. A single base duplication mutation in canine and bovine CLN5 has been shown to cause NCL.

REFERENCES

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STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.

CHROMOSOMAL LOCATION

Genetic locus: CLN5 (human) mapping to 13q22.3.

PRODUCT

CLN5 (h5): 293T Lysate represents a lysate of human CLN5 transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

CLN5 (h5): 293T Lysate is suitable as a Western Blotting positive control for human reactive CLN5 antibodies. Recommended use: 10-20 µl per lane.

Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

RESEARCH USE

For research use only, not for use in diagnostic procedures.