

Dpl (m): 293T Lysate: sc-126739

BACKGROUND

Prion diseases, or transmissible spongiform encephalopathies (TSEs), are manifested as genetic, infectious or sporadic, lethal neurodegenerative disorders involving alterations of the prion protein (PrP). Infectious PrP^{Sc} is highly expressed in the brain of animals affected by TSEs, including scrapie in sheep, BSE in cattle and Cruetzfeldt-Jacob disease in humans. The PRND gene locus, located on human chromosome 20p, encodes for the doppel protein (Dpl), which exhibits approximately 25% sequence homology with PrP. Dpl is characterized by an α -helical conformation, intramolecular disulfide bonds and two N-linked oligosaccharides, and it is presented on the cell surface by a glycosylphosphatidylinositol anchor. Dpl is highly expressed in adult testis and heart and is detectable in the brain of neonatal mice. Dpl does not appear to contribute to prion disease progression, but ectopic expression of Dpl is implicated in neuronal degeneration of ataxic PrP-deficient mice. Dpl is also thought to play a role in angiogenesis, specifically maturation of the blood-brain barrier.

REFERENCES

1. Prusiner, S.B. 1998. Prions. *Proc. Natl. Acad. Sci. USA* 95: 13363-13383.
2. Lee, I.Y., et al. 1998. Complete genomic sequence and analysis of the prion protein gene region from three mammalian species. *Genome. Res.* 8: 1022-1037.
3. Mead, S., et al. 2000. Examination of the human prion protein-like gene doppel for genetic susceptibility to sporadic and variant Creutzfeldt-Jakob disease. *Neurosci. Lett.* 290: 117-120.
4. Silverman, G.L., et al. 2000. Doppel is an N-glycosylated, glycosylphosphatidylinositol-anchored protein. Expression in testis and ectopic production in the brains of Prnp(0/0) mice predisposed to Purkinje cell loss. *J. Biol. Chem.* 275: 26834-26841.
5. Li, A., et al. 2000. Physiological expression of the gene for PrP-like protein, PrPLP/Dpl, by brain endothelial cells and its ectopic expression in neurons of PrP-deficient mice ataxic due to Purkinje cell degeneration. *Am. J. Pathol.* 157: 1447-1452.
6. Mo, H., et al. 2001. Two different neurodegenerative diseases caused by proteins with similar structures. *Proc. Natl. Acad. Sci. USA* 98: 2352-2357.

CHROMOSOMAL LOCATION

Genetic locus: Prnd (mouse) mapping to 2 F2.

PRODUCT

Dpl (m): 293T Lysate represents a lysate of mouse Dpl transfected 293T cells and is provided as 100 μ g protein in 200 μ l SDS-PAGE buffer.

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.

RESEARCH USE

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

APPLICATIONS

Dpl (m): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive Dpl antibodies. Recommended use: 10-20 μ l per lane.

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

PROTOCOLS

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