

Peroxin 5 (h): 293T Lysate: sc-173622

BACKGROUND

Peroxisomes are single-membrane bound organelles present in virtually all eukaryotic cells. They are involved in numerous catabolic and anabolic pathways, including β -oxidation of very long chain fatty acids, metabolism of hydrogen peroxide, plasmalogen biosynthesis and bile acid synthesis. The Peroxin gene family, which includes more than 20 members, is required for peroxisome biogenesis. Two members of this family, Peroxin 5 (Pex5) and Peroxin 7 (Pex7), are receptors for proteins that contain the peroxisome targeting signal 1 (PTS1) and 2 (PTS2), respectively, and shuttle these proteins from the cytosol to the peroxisome. Peroxin 5, also designated PTS1 receptor, is expressed as two isoforms, Pex5L and Pex5S. Pex5L transports PTS1 and Pex7-PTS2 cargo complexes to the initial Pex5 docking site, Pex14, while Pex5S transports only PTS1 cargoes. Pex5 and Pex7 also require either direct or indirect interaction with Peroxin 13 (Pex13) for proper import into peroxisomes. Mutations in the Peroxin genes result in peroxisome biogenesis disorders (PBDs). Defects in the Pex5 gene are linked to Zellweger syndrome (cerebro-hapato-renal syndrome) of complementation group 2 (CG2), the most severe form of PBDs. Zellweger syndrome is characterized by abnormal neuronal migration in the central nervous system and severe neurologic dysfunction, which leads to early death.

REFERENCES

1. Girzasky, W., et al. 1999. Involvement of Pex13p in Pex14p localization and peroxisomal targeting signal 2-dependent protein import into peroxisomes. *J. Cell Biol.* 144: 1151-1162.
2. Gartner, J. 2000. Organelle disease: peroxisomal disorders. *Eur. J. Pediatr.* 159: S236-S239.
3. Collins, C.S., et al. 2000. The peroxisome biogenesis factors Pex4p, Pex22p, Pex1p and Pex6p act in the terminal steps of peroxisomal matrix protein import. *Mol. Cell. Biol.* 20: 7516-7526.
4. Fujiki, Y. 2000. Peroxisome biogenesis and peroxisome biogenesis disorders. *FEBS Lett.* 476: 42-46.
5. Dodt, G., et al. 2001. Domain mapping of human Pex5 reveals functional and structural similarities to *Saccharomyces cerevisiae* Pex18p and Pex21p. *J. Biol. Chem.* 276: 41769-41781.
6. Baumgart, E., et al. 2001. Mitochondrial alterations caused by defective peroxisomal biogenesis in a mouse model for Zellweger syndrome (Pex5 knockout mouse). *Am. J. Pathol.* 159: 1477-1494.
7. Faust, P.L., et al. 2001. The peroxisome deficient Pex2 Zellweger mouse: pathologic and biochemical correlates of lipid dysfunction. *J. Mol. Neurosci.* 16: 289-297.

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: PEX5 (human) mapping to 12p13.31.

PRODUCT

Peroxin 5 (h): 293T Lysate represents a lysate of human Peroxin 5 transfected 293T cells and is provided as 100 μ g protein in 200 μ l SDS-PAGE buffer.

APPLICATIONS

Peroxin 5 (h): 293T Lysate is suitable as a Western Blotting positive control for human reactive Peroxin 5 antibodies. Recommended use: 10-20 μ l per lane.

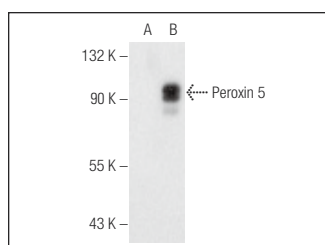
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

Peroxin 5 (F-10): sc-365983 is recommended as a positive control antibody for Western Blot analysis of enhanced human Peroxin 5 expression in Peroxin 5 transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

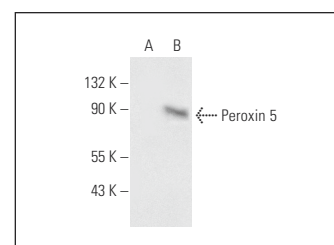
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended:
1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048.

DATA



Peroxin 5 (F-10): sc-365983. Western blot analysis of Peroxin 5 expression in non-transfected: sc-117752 (A) and human Peroxin 5 transfected: sc-173622 (B) 293T whole cell lysates.



Peroxin 5 (B-3): sc-365629. Western blot analysis of Peroxin 5 expression in non-transfected: sc-117752 (A) and human Peroxin 5 transfected: sc-173622 (B) 293T whole cell lysates.

RESEARCH USE

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