

Peroxin 6 (h): 293T Lysate: sc-115993

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. One such member of the Peroxin gene family is Peroxin 6. Of 11 mutations identified in the gene PEX6, most lead to premature termination or large deletions of the Peroxin 6 protein and result in the most severe peroxisome biogenesis disorder phenotype of Zellweger syndrome, a disorder associated with major deformations.

REFERENCES

1. Shimozawa, N., et al. 1993. Standardization of complementation grouping of peroxisome-deficient disorders and the second Zellweger patient with peroxisomal assembly factor-1 (PAF-1) defect. *Am. J. Hum. Genet.* 52: 843-844.
2. Tsukamoto, T., et al. 1995. Peroxisome assembly factor-2, a putative ATPase cloned by functional complementation on a peroxisome-deficient mammalian cell mutant. *Nat. Genet.* 11: 395-401.
3. Moser, A.B., et al. 1995. Phenotype of patients with peroxisomal disorders subdivided into sixteen complementation groups. *J. Pediatr.* 127: 13-22.
4. Fukuda, S., et al. 1996. Human peroxisome assembly factor-2 (PAF-2): a gene responsible for group C peroxisome biogenesis disorder in humans. *Am. J. Hum. Genet.* 59: 1210-1220.
5. Distel, B., et al. 1996. A unified nomenclature for peroxisome biogenesis factors. *J. Cell Biol.* 135: 1-3.
6. Miyata, N., et al. 2005. Shuttling mechanism of peroxisome targeting signal type 1 receptor PEX5: ATP-independent import and ATP-dependent export. *Mol. Cell. Biol.* 25: 10822-10832.
7. Krazy, H., et al. 2006. Identification and characterization of three peroxins —PEX6, PEX10 and PEX12— involved in glycosome biogenesis in *Trypanosoma brucei*. *Biochim. Biophys. Acta* 1763: 6-17.
8. Krause, C., et al. 2006. Identification of novel mutations in PEX2, PEX6, PEX10, PEX12 and PEX13 in Zellweger spectrum patients. *Hum. Mutat.* 27: 1157.

CHROMOSOMAL LOCATION

Genetic locus: PEX6 (human) mapping to 6p21.1.

PRODUCT

Peroxin 6 (h): 293T Lysate represents a lysate of human Peroxin 6 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.

APPLICATIONS

Peroxin 6 (h): 293T Lysate is suitable as a Western Blotting positive control for human reactive Peroxin 6 antibodies. Recommended use: 10-20 μ l per lane.

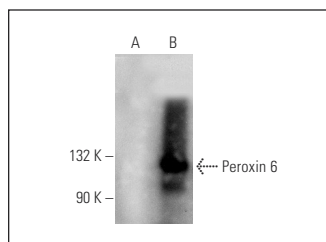
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

Peroxin 6 (F-6): sc-271813 is recommended as a positive control antibody for Western Blot analysis of enhanced human Peroxin 6 expression in Peroxin 6 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 6 (F-6): sc-271813. Western blot analysis of Peroxin 6 expression in non-transfected: sc-117752 (A) and human Peroxin 6 transfected: sc-115993 (B) 293T whole cell lysates.

RESEARCH USE

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

PROTOCOLS

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