

## Peroxin 6 (P-17): sc-67754

### BACKGROUND

Peroxisomes are single-membrane bound organelles present in virtually all eukaryotic cells. They are involved in numerous catabolic and anabolic pathways, including  $\beta$ -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

- 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.
- Tsakamoto, 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.
- Moser, A.B., et al. 1995. Phenotype of patients with peroxisomal disorders subdivided into sixteen complementation groups. *J. Pediatr.* 127: 13-22.
- 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.
- Distel, B., et al. 1996. A unified nomenclature for peroxisome biogenesis factors. *J. Cell Biol.* 135: 1-3.
- 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.
- 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.
- Krause, C., et al. 2006. Identification of novel mutations in PEX2, PEX6, PEX10, PEX12, and PEX13 in Zellweger spectrum patients. *Hum. Mutat.* 27: 1157.
- Furuki, S., et al. 2006. Mutations in the peroxin Pex26p responsible for peroxisome biogenesis disorders of complementation group 8 impair its stability, peroxisomal localization, and interaction with the Pex1p x Pex6p complex. *J. Biol. Chem.* 281: 1317-1323.

### CHROMOSOMAL LOCATION

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

### SOURCE

Peroxin 6 (P-17) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of Peroxin 6 of human origin.

### PRODUCT

Each vial contains 200  $\mu$ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-67754 P, (100  $\mu$ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

### APPLICATIONS

Peroxin 6 (P-17) is recommended for detection of Peroxin 6 of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for Peroxin 6 siRNA (h): sc-62775, Peroxin 6 shRNA Plasmid (h): sc-62775-SH and Peroxin 6 shRNA (h) Lentiviral Particles: sc-62775-V.

Molecular Weight of Peroxin 6: 116 kDa.

Positive Controls: Jurkat whole cell lysate: sc-2204 or K-562 whole cell lysate: sc-2203.

### RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

### STORAGE

Store at 4° C, **\*\*DO NOT FREEZE\*\***. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

### RESEARCH USE

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

### PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.