# SANTA CRUZ BIOTECHNOLOGY, INC.

# Peroxin 14 (1G12): sc-293383



# BACKGROUND

Peroxisomes are single-membrane bounds 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

- Girzalsky, 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.
- Gartner, J. 2000. Organelle disease: peroxisomal disorders. Eur. J. Pediatr. 159: S236-S239.
- 3. Fujiki, Y. 2000. Peroxisome biogenesis and peroxisome biogenesis disorders. FEBS Lett. 476: 42-46.
- 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.
- 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.
- 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.
- 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.
- Brosius, U. and Gartner, J. 2002. Cellular and molecular aspects of Zellweger syndrome and other peroxisome biogenesis disorders. Cell. Mol. Life Sci. 59: 1058-1069.
- Otera, H., et al. 2002. Peroxisomal targeting signal receptor Pex5p interacts with cargoes and import machinery components in a spatiotemporally differentiated manner: conserved Pex5p WXXXF/Y motifs are critical for matrix protein import. Mol. Cell. Biol. 22: 1639-1655.

## CHROMOSOMAL LOCATION

Genetic locus: PEX14 (human) mapping to 1p36.22.

# SOURCE

Peroxin 14 (1G12) is a mouse monoclonal antibody raised against amino acids 293-375 of Peroxin 14 of human origin.

#### PRODUCT

Each vial contains 100  $\mu$ g IgG<sub>2a</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

## **APPLICATIONS**

Peroxin 14 (1G12) is recommended for detection of Peroxin 14 of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2  $\mu$ g per 100-500  $\mu$ g of total protein (1 ml of cell lysate)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for Peroxin 14 siRNA (h): sc-40827, Peroxin 14 shRNA Plasmid (h): sc-40827-SH and Peroxin 14 shRNA (h) Lentiviral Particles: sc-40827-V.

Molecular Weight (predicted) of Peroxin 14: 34 kDa.

Molecular Weight (observed) of Peroxin 14: 34-43 kDa.

#### **RECOMMENDED SUPPORT REAGENTS**

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz<sup>®</sup> Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml).

#### DATA



of human recombinant Peroxin 14 fusion protein.

# **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.