Technical Data Sheet

Purified Mouse Anti-PEX1

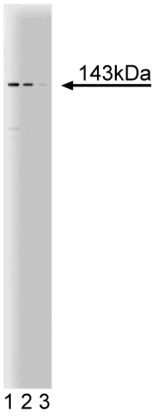
Product Information

Material Number:	**611719**
Size:	150 µg
Concentration:	250 µg/ml
Clone:	1/PEX
Immunogen:	Human PEX1 aa. 1049-1256
Isotype:	Mouse IgG1
Reactivity:	QC Testing: Human Tested in Development: Mouse, Rat, Chicken, Dog
Target MW:	143 kDa
Storage Buffer:	Aqueous buffered solution containing BSA, glycerol, and ≤0.09% sodium azide.

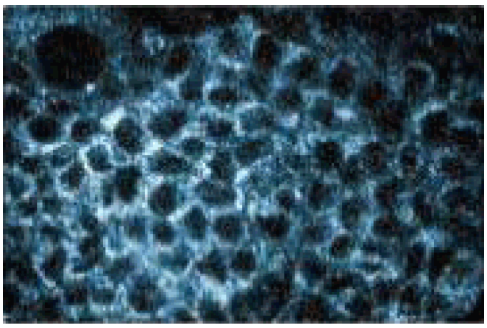
Description

Peroxisomes, ubiquitous organelles of eukaryotic cells, are involved in a number of metabolic processes. Their formation involves membrane generation, targeting and insertion of peroxisomal membrane proteins (PMPs) into the membrane, and transport of matrix proteins across the newly formed membrane. Import of PMPs and synthesis of peroxisomal membranes may involve as many as 17 different PEX proteins. Mutation in any of 12 different Pex genes causes Zellweger syndrome (ZS), a disease characterized by loss of peroxisome biogenesis leading to severe neurologic, hepatic, and renal abnormalities. Mutations in two peroxisomal AAA ATPases, PEX1 and PEX6, are commonly associated with this and other neurological disorders. These ATPases form a complex in vitro and are required for normal import of proteins targeted to the peroxisome, as well as for maintaining the stability of PEX5, a peroxisomal receptor required for protein import. Substitution of aspartate for glycine at position 843 in PEX1 is the most common cause of peroxisome biogenesis disorders. Thus, PEX1 has an essential role in peroxisome biogenesis and mutation leads to Zwellweger syndrome-type diseases.

This antibody is routinely tested by western blot analysis. Other applications were tested at BD Biosciences Pharmingen during antibody development only or reported in the literature.



Western blot analysis of PEX1 on a HCT-8 cell lysate (Human colorectal adenocarcinoma; ATCC CCL-244). *Lane 1: 1:250, lane 2: 1:500, lane 3: 1:1000 dilution of the mouse anti-PEX1 antibody.*



Immunofluorescence staining of A431 cells (Human epithelial carcinoma; ATCC CRL-1555).

BD Biosciences

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Preparation and Storage

The monoclonal antibody was purified from tissue culture supernatant or ascites by affinity chromatography.
Store undiluted at -20° C.

Application Notes

Application

Western blot	Routinely Tested
Immunofluorescence	Tested During Development

Recommended Assay Procedure:

Western blot: Please refer to http://www.bdbiosciences.com/pharmingen/protocols/Western_Blotting.shtml

Suggested Companion Products

Catalog Number	Name	Size	Clone
611474	HCT-8 Cell Lysate	500 µg	(none)
554002	HRP Goat Anti-Mouse Ig	1.0 ml	(none)
554001	FITC Goat Anti-Mouse Ig	0.5 mg	Polyclonal

Product Notices

1. Since applications vary, each investigator should titrate the reagent to obtain optimal results.
2. Please refer to www.bdbiosciences.com/pharmingen/protocols for technical protocols.
3. Caution: Sodium azide yields highly toxic hydrazoic acid under acidic conditions. Dilute azide compounds in running water before discarding to avoid accumulation of potentially explosive deposits in plumbing.
4. Source of all serum proteins is from USDA inspected abattoirs located in the United States.

References

Collins CS, Gould SJ. Identification of a common PEX1 mutation in Zellweger syndrome. *Hum Mutat.* 1999; 14(1):45-53.(Biology)
Geisbrecht BV, Collins CS, Reuber BE, Gould SJ. Disruption of a PEX1-PEX6 interaction is the most common cause of the neurologic disorders Zellweger syndrome, neonatal adrenoleukodystrophy, and infantile Refsum disease. *Proc Natl Acad Sci U S A.* 1998; 95(15):8630-8635.(Biology)
Reuber BE, Germain-Lee E, Collins CS, et al. Mutations in PEX1 are the most common cause of peroxisome biogenesis disorders. *Nat Genet.* 1997; 17(4):445-448.(Biology)