

Product datasheet

Anti-PEX19 antibody ab123315

1 Image

Overview

Product name	Anti-PEX19 antibody
Description	Rabbit polyclonal to PEX19
Host species	Rabbit
Tested applications	Suitable for: WB
Species reactivity	Reacts with: Human
Immunogen	Synthetic peptide conjugated to KLH, corresponding to a region within N terminal amino acids 32-61 of Human PEX19 (NP_002848.1, NP_001180573.1).
Positive control	HeLa cell lysate.

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Store at 4°C (up to 6 months). Store at -20°C long term.
Storage buffer	Preservative: 0.09% Sodium azide Constituent: 99% PBS
Purity	Immunogen affinity purified
Purification notes	ab123315 is purified through a protein A column, followed by peptide affinity purification.
Clonality	Polyclonal
Isotype	IgG

Applications

Our [Abpromise guarantee](#) covers the use of **ab123315** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
WB		1/100 - 1/500. Predicted molecular weight: 33 kDa.

Target

Function

Necessary for early peroxisomal biogenesis. Acts both as a cytosolic chaperone and as an import receptor for peroxisomal membrane proteins (PMPs). Binds and stabilizes newly synthesized PMPs in the cytoplasm by interacting with their hydrophobic membrane-spanning domains, and targets them to the peroxisome membrane by binding to the integral membrane protein PEX3. Excludes CDKN2A from the nucleus and prevents its interaction with MDM2, which results in active degradation of TP53.

Tissue specificity

Ubiquitously expressed. Isoform 1 is strongly predominant in all tissues except in utero where isoform 2 is the main form.

Involvement in disease

Defects in PEX19 are the cause of peroxisome biogenesis disorder complementation group 14 (PBD-CG14) [MIM:600279]; also known as PBD-CGJ. PBD refers to a group of peroxisomal disorders arising from a failure of protein import into the peroxisomal membrane or matrix. The PBD group is comprised of four disorders: Zellweger syndrome (ZWS), neonatal adrenoleukodystrophy (NALD), infantile Refsum disease (IRD), and classical rhizomelic chondrodysplasia punctata (RCDP). ZWS, NALD and IRD are distinct from RCDP and constitute a clinical continuum of overlapping phenotypes known as the Zellweger spectrum. The PBD group is genetically heterogeneous with at least 14 distinct genetic groups as concluded from complementation studies.

Defects in PEX19 are a cause of Zellweger syndrome (ZWS) [MIM:214100]. ZWS is a fatal peroxisome biogenesis disorder characterized by dysmorphic facial features, hepatomegaly, ocular abnormalities, renal cysts, hearing impairment, profound psychomotor retardation, severe hypotonia and neonatal seizures. Death occurs within the first year of life.

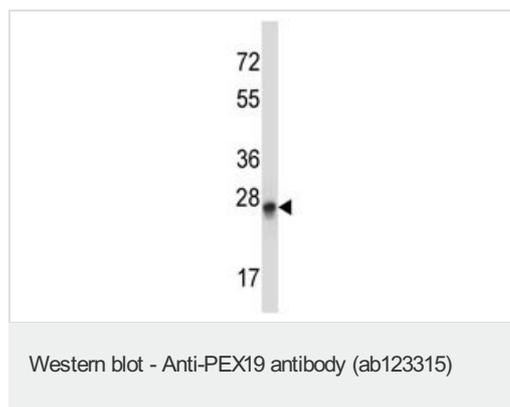
Sequence similarities

Belongs to the peroxin-19 family.

Cellular localization

Cytoplasm. Peroxisome membrane. Mainly cytoplasmic. Some fraction membrane-associated to the outer surface of peroxisomes.

Images



Anti-PEX19 antibody (ab123315) at 1/100 dilution + HeLa cell lysate at 35 µg

Developed using the ECL technique.

Predicted band size: 33 kDa

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