

Product datasheet

Anti-PEX14 antibody ab113286

1 Image

Overview

Product name	Anti-PEX14 antibody
Description	Rabbit polyclonal to PEX14
Host species	Rabbit
Tested applications	Suitable for: IP Unsuitable for: WB
Species reactivity	Reacts with: Human Predicted to work with: Chimpanzee, Rhesus monkey, Gorilla, Orangutan
Immunogen	Synthetic peptide corresponding to a region within amino acids 250-300 of Human PEX14 (NP_004556.1).
Positive control	HeLa whole cell lysate

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.
Storage buffer	Preservative: 0.09% Sodium azide Constituent: 99% Tris citrate/phosphate Note: pH range: 7 to 8
Purity	Immunogen affinity purified
Clonality	Polyclonal
Isotype	IgG

Applications

Our [Abpromise guarantee](#) covers the use of **ab113286** 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
IP		Use at 2-5 µg/mg of lysate.

Application notes

Is unsuitable for WB.

Target**Function**

Component of the peroxisomal translocation machinery with PEX13 and PEX17. Interacts with both the PTS1 and PTS2 receptors. Binds directly to PEX17.

Involvement in disease

Defects in PEX14 are the cause of peroxisome biogenesis disorder complementation group K (PBD-CGK) [MIM:601791]. PBD-CGK is a peroxisomal disorder arising from a failure of protein import into the peroxisomal membrane or matrix. The peroxisome biogenesis disorders (PBD group) are genetically heterogeneous with at least 14 distinct genetic groups as concluded from complementation studies. Include disorders are: 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 (PBD-ZSS).

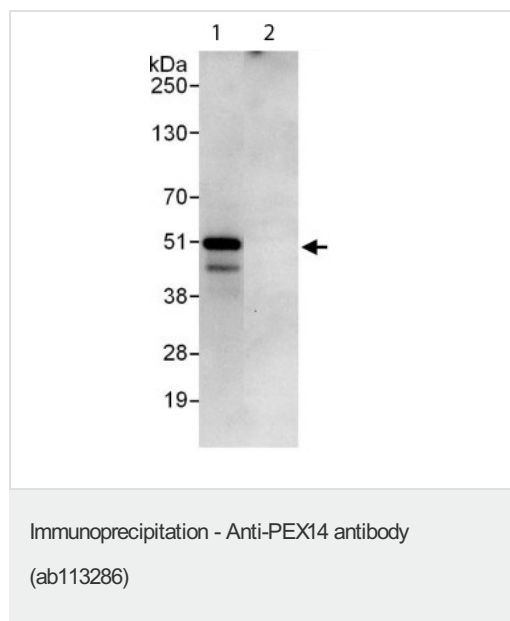
Defects in PEX14 are a cause of Zellweger syndrome (ZWS) [MIM:214100]. ZWS is a fatal peroxisome biogenesis disorder characterized by dysmorphic facial features, hepatomegaly, ocular abnormalities, renal cysts, hearing impairment, profound psychomotor retardation, severe hypotonia and neonatal seizures. Death occurs within the first year of life.

Sequence similarities

Belongs to the peroxin-14 family.

Cellular localization

Peroxisome membrane.

Images

Staining of PEX14 in HeLa whole cell lysate immunoprecipitated using ab113286 at 6 µg/mg lysate (1 mg/IP; 20% of IP loaded/lane). Lane 2: control IgG. Predicted band size : 41 kDa. Exposure time: 10 seconds.

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