## abcam

### Product datasheet

### Anti-PEX19 antibody ab95959

### ★★★☆☆ 1 Abreviews 3 图像

### 概述

产**品名称** Anti-PEX19抗体

描述 兔多克隆抗体to PEX19

**宿主** Rabbit

经测试应用 适用于: WB, IHC-P, ICC/IF

种属反应性 与反应: Human

预测可用于: Mouse, Rat, Cow \_\_\_\_\_

免疫原 Recombinant fragment within Human PEX19 aa 1-269. The exact sequence is proprietary.

Database link: P40855

**阳性**对照 WB: HeLa whole cell lysate (<u>ab29545</u>), MOLT4 whole cell lysate; ICC/IF: A549 cell IHC-P:

SW480 xenograft

常规说明

The Life Science industry has been in the grips of a reproducibility crisis for a number of years.

Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets

your needs before purchasing.

If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be

found below, along with publications, customer reviews and Q&As

性能

形式 Liquid

**存放说明** Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid repeated freeze / thaw cycles.

**存储溶液** pH: 7.00

Preservative: 0.025% Proclin 300

Constituents: 79% PBS, 20% Glycerol (glycerin, glycerine)

纯**度** Immunogen affinity purified

纯**化**说明 Purified by antigen affinity chromatography.

 克隆
 多克隆

 同种型
 lqG

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### The Abpromise guarantee

### Abpromise™承诺保证使用ab95959于以下的经测试应用

"应用说明"部分下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明
WB	<b>★★★</b> ☆☆ <u>(1)</u>	1/500 - 1/3000. Predicted molecular weight: 33 kDa.
IHC-P		1/100 - 1/500.
ICC/IF		1/100 - 1/200.

### 靶标

### 功能

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.

### 组织特异性

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

### 疾病相关

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.

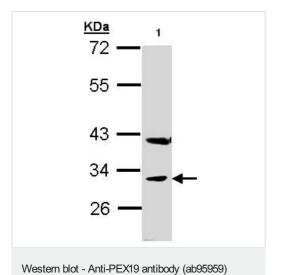
### 序列相似性

### Belongs to the peroxin-19 family.

### 细胞定位

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

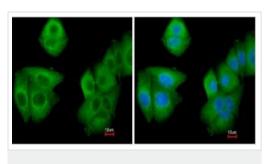
### 图片



Anti-PEX19 antibody (ab95959) at 1/1000 dilution + MOLT4 whole cell lysate at 30  $\mu g$ 

Predicted band size: 33 kDa

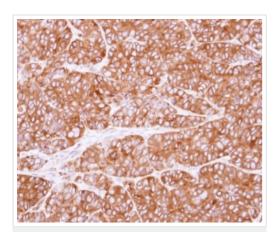
12% SDS PAGE



Immunocytochemistry/ Immunofluorescence - Anti-PEX19 antibody (ab95959)

ab95959, at a 1/200 dilution, staining PEX19 in paraformaldehyde fixed A549 cell by Immunofluorescence analysis.

Image on the right: merged with DNA probe.



Immunohistochemistry (Formalin/PFA-fixed paraffinembedded sections) - Anti-PEX19 antibody (ab95959)

ab95959, at a 1/500 dilution, staining PEX19 in paraffin embedded SW480 xenograft by Immunohistochemistry.

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