

Recombinant human ABCD1/ALD protein (Active)
ab235011

2 Images

| Description | |
|---------------------|--|
| Product name | Recombinant human ABCD1/ALD protein (Active) |
| Biological activity | Measured by its binding ability in a functional ELISA. Immobilized ab235011 at 5 µg/ml can bind human PEX19, the EC ₅₀ of human PEX19 protein is 22.96-33.00 µg/ml. |
| Purity | > 90 % SDS-PAGE. |
| Expression system | Escherichia coli |
| Accession | <u>P33897</u> |
| Protein length | Full length protein |
| Animal free | No |
| Nature | Recombinant |
| Species | Human |
| Sequence | MPVLSRPRPWGNTLKRTAVLLALAAYGAHKVYPLVRQC LAPARGLQAPA GEPTQEASGVAAAKAGMNRVFLQRLLWLLRLLFPRVLCR ETGLLALHSAA LVSRTFLSVYVARLDGRLARCIVRKDPRAFGWQLLQWLLI ALPATFVNSA IRYLEGQLALSFRSRLVAHAYRLYFSQQTYYRVSNMDGRLR NPDQSLTED VVAFAASVAHLYSNLTKPLLDVAVTSYLLRAARSRGAGT AWPSAIGLV VFLTANVLRAFSPKFGELVAEEARRKGELRYMHSRVVAN SEEIAFYGGHE VELALLQRSYQDLASQINLILLERLWYVMLEQFLMKYVWSA SGLLMVAVP IITATGYSESDAEAVKKAALKKEEELVSE RTEAFTIARNLL TAAADAIE RIMSSYKEVTELAGYTARVHEMFQVFEDVQRCHFKRPRE LEDAQAGSGTI GRSGVRVEGPLKIRGQVVDVEQGIICENIPVTPSGEVVVA SLNIRVEEG MHLLITGPNGCGKSSLFRILGGLWPTYGGVLYKPPPQRMF |

YIPQRPYMSV
 GSLRDQVIYPDSVEDMQRKGYSEQDLEALDVVHLHHILQ
 REGGWEAMCD
 WKDVLSGGEKQRIGMARMFYHRPKYALLDECTSAVSIDV
 EGKIFQAAKDA
 GIALLSITHRPSLWKYHTHLLQFDGEGGWKFEKLDSAARL
 SLTEEKQRLE
 QQLAGIPKMQRRLQELCQILGEAVAPAHVPAPSPQGP
 LQGAST

| | |
|--|--------------------------|
| Predicted molecular weight | 85 kDa including tags |
| Amino acids | 1 to 745 |
| Tags | His tag C-Terminus |
| Additional sequence information | C-terminal 10xHis-tagged |

Specifications

Our **Abpromise guarantee** covers the use of **ab235011** in the following tested applications.

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

| | |
|-------------------------|--|
| Applications | Functional Studies SDS-PAGE |
| Form | Liquid |
| Additional notes | Produced in an in vitro E.coli expression system. Previously labelled as ABCD1. |

Preparation and Storage

| | |
|------------------------------|---|
| Stability and Storage | Shipped at 4°C. Store at -20°C or -80°C. Avoid freeze / thaw cycle. pH: 7.2 Constituents: Tris buffer, 50% Glycerol (glycerin, glycerine) This product is an active protein and may elicit a biological response in vivo, handle with caution. |
|------------------------------|---|

General Info

| | |
|-------------------------------|---|
| Function | Probable transporter. The nucleotide-binding fold acts as an ATP-binding subunit with ATPase activity. |
| Involvement in disease | Defects in ABCD1 are the cause of adrenoleukodystrophy X-linked (X-ALD) [MIM:300100]. X-ALD is a peroxisomal metabolic disorder characterized by progressive multifocal demyelination of the central nervous system and by peripheral adrenal insufficiency (Addison disease). It results in mental deterioration, corticospinal tract dysfunction, and cortical blindness. Different clinical manifestations exist like: cerebral childhood ALD (CALD), adult cerebral ALD (ACALD), adrenomyeloneuropathy (AMN) and 'Addison disease only' (ADO) phenotype. Note=The promoter region of ABCD1 is deleted in the chromosome Xq28 deletion syndrome which involves ABCD1 and the neighboring gene BCAP31. |
| Sequence similarities | Belongs to the ABC transporter superfamily. ABCD family. Peroxisomal fatty acyl CoA transporter (TC 3.A.1.203) subfamily. |

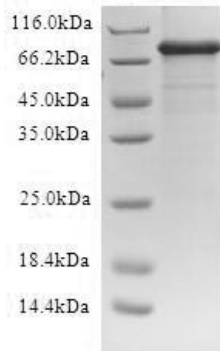
Contains 1 ABC transmembrane type-1 domain.

Contains 1 ABC transporter domain.

Cellular localization

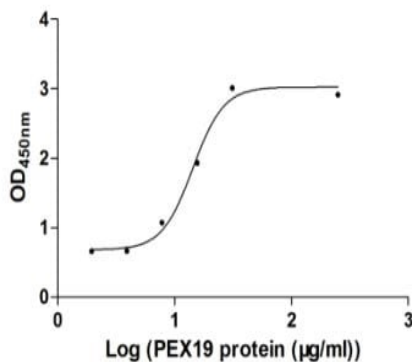
Peroxisome membrane.

Images



SDS-PAGE - Recombinant human ABCD1/ALD protein (Active) (ab235011)

ab235011 analyzed by (Tris-Glycine gel) discontinuous SDS-PAGE (reduced) with 5% enrichment gel and 15% separation gel.



Functional Studies - Recombinant human ABCD1/ALD protein (Active) (ab235011)

Immobilized ab235011 at 5 µg/ml can bind human PEX19, the EC₅₀ of human PEX19 is 22.96-33.00 µg/ml.

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

Our Abpromise to you: Quality guaranteed and expert technical support

- Replacement or refund for products not performing as stated on the datasheet
- Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours
- We provide support in Chinese, English, French, German, Japanese and Spanish

- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit <https://www.abcam.com/abpromise> or contact our technical team.

Terms and conditions

- Guarantee only valid for products bought direct from Abcam or one of our authorized distributors