abcam

Product datasheet

Anti-FKRP antibody ab65243

画像数 1

製品の概要

製品名	Anti-FKRP antibody
製品の詳細	Rabbit polyclonal to FKRP
由来種	Rabbit
アプリケーション	**適用あり:** WB
種交差性	**交差種:** Human **交差が予測される動物種:** Mouse, Rat
免疫原	Synthetic peptide derived from N terminal of human FKRP.
ポジティブ・コントロール	Jurkat and COLO205 cell extracts.
特記事項	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

製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Store at -20°C. Stable for 12 months at -20°C.
バッファー	pH: 7.40 Preservative: 0.02% Sodium azide Constituents: PBS, 50% Glycerol (glycerin, glycerine), 0.87% Sodium chloride Without Mg2+ and Ca2+
精製度	Immunogen affinity purified
ポリ/モノ	ポリクローナル
アイソタイプ	IgG

アプリケーション

The Abpromise guarantee **Abpromise保証は、** 次のテスト済みアプリケーションにおけるab65243の使用に適用されます

アプリケーションノートには、推奨の開始希釈率がありますが、適切な希釈率につきましてはご検討ください。

アプリケーション	Abreviews	特記事項
WB		1/500 - 1/1000. Detects a band of approximately 55 kDa (predicted molecular weight: 55 kDa).

ターゲット情報

機能

Could be a transferase involved in the modification of glycan moieties of alpha-dystroglycan (DAG1).

組織特異性

Expressed predominantly in skeletal muscle, placenta, and heart and relatively weakly in brain, lung, liver kidney and pancreas.

関連疾患

Defects in FKRP are the cause of muscular dystrophy-dystroglycanopathy congenital with brain and eye anomalies type A5 (MDDGA5) [MIM:613153]. MDDGA5 is an autosomal recessive disorder characterized by congenital muscular dystrophy associated with cobblestone lissencephaly and other brain anomalies, eye malformations, profound mental retardation, and death usually in the first years of life. Included diseases are the more severe Walker-Warburg syndrome and the slightly less severe muscle-eye-brain disease.
Defects in FKRP are the cause of muscular dystrophy-dystroglycanopathy congenital with or without mental retardation type B5 (MDDGB5) [MIM:606612]. MDDGB5 is a congenital muscular dystrophy characterized by a severe phenotype with inability to walk, muscle hypertrophy, marked elevation of serum creatine kinase, a secondary deficiency of laminin alpha2, and a marked reduction in alpha-dystroglycan expression. Only a subset of MDDGB5 patients have brain involvements.
Defects in FKRP are the cause of muscular dystrophy-dystroglycanopathy limb-girdle type C5 (MDDGC5) [MIM:607155]; also known as limb-girdle muscular dystrophy type 2I. MDDGC5 is an autosomal recessive disorder with age of onset ranging from childhood to adult life, and variable severity. Clinical features include proximal muscle weakness, waddling gait, calf hypertrophy, cardiomyopathy and respiratory insufficiency. A reduction of alpha-dystroglycan and laminin alpha-2 expression can be observed on skeletal muscle biopsy from MDDGC5 patients.

配列類似性

Belongs to the licD transferase family.

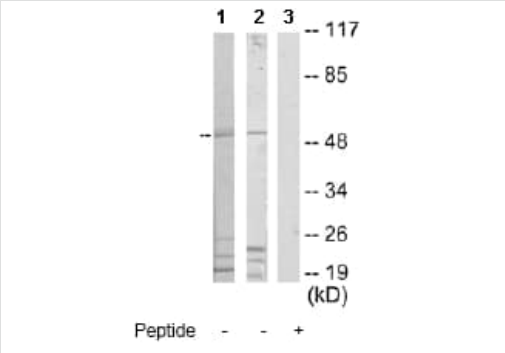
翻訳後修飾

N-glycosylated.

細胞内局在

Golgi apparatus. Secreted. Cell membrane > sarcolemma. Rough endoplasmic reticulum. According to some studies the N-terminal hydrophobic domain is cleaved after translocation to the Golgi apparatus and the protein is secreted. According to others the N-terminal hydrophobic domain is a transmembrane domain and the protein is a type II transmembrane type targeted to the Golgi apparatus by a non-cleavable signal anchor sequence. Localization at the cell membrane may require the presence of dystroglycan. At the Golgi apparatus localizes most likely at the cis-compartment. Detected in rough endoplasmic reticulum in myocytes. In general, mutants associated with severe clinical phenotypes are retained within the endoplasmic reticulum.

画像



Western blot - Anti-FKRP antibody (ab65243)

All lanes : Anti-FKRP antibody (ab65243) at 1/500 dilution

Lane 1 : Jurkat cell extract
Lane 2 : COLO205 cell extract
Lane 3 : COLO205 cell extract with immunising peptide

Predicted band size: 55 kDa
Observed band size: 55 kDa
Additional bands at: 22 kDa, 24 kDa. We are unsure as to the identity of these extra bands.

The amount of positive control loading for the WB is 5-30 ug of total protein. The amount of the peptide for the WB is 5-10 ug.

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