


# Anti-Rapsyn antibody ab118491

画像数 1

### 製品の概要

製品名	Anti-Rapsyn antibody
製品の詳細	Rabbit polyclonal to Rapsyn
由来種	Rabbit
アプリケーション	<b>適用あり:</b> IHC-P
種交差性	<b>交差種:</b> Human <b>交差が予測される動物種:</b> Mouse, Rat 
免疫原	Synthetic peptide corresponding to Human Rapsyn aa 399-412 (C terminal) conjugated to Keyhole Limpet Haemocyanin (KLH).
ポジティブ・コントロール	Human brain (cortex) tissue.
特記事項	<p>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.</p> <p>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&amp;As</p>

### 製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term.
バッファー	Preservative: 0.01% Sodium azide Constituents: 50% Glycerol, 49% PBS
精製度	Protein A 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ab118491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確認ください。

アプリケーション	Abreviews	特記事項
IHC-P		Use a concentration of 10 - 20 µg/ml. Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.

## ターゲット情報

### 機能

Thought to play some role in anchoring or stabilizing the nicotinic acetylcholine receptor at synaptic sites. It may link the receptor to the underlying postsynaptic cytoskeleton, possibly by direct association with actin or spectrin.

### 関連疾患

Defects in RAPSN are a cause of congenital myasthenic syndrome with acetylcholine receptor deficiency (ACHRDCMS) [MIM:608931]. ACHRDCMS is a post-synaptic congenital myasthenic syndrome. Congenital myasthenic syndromes (CMS) are inherited disorders of neuromuscular transmission that stem from mutations in presynaptic, synaptic, or postsynaptic proteins. Postsynaptic disorders result from mutations in proteins forming the subunits of the muscle acetylcholine receptor (AChR). The kinetic abnormalities of AChR result in either prolonged ion channel activations that underlie 'slow-channel myasthenic syndromes' (SCCMS) or abbreviated channel activations that underlie the abnormally rapid decay of endplate currents in 'fast-channel syndromes' (FCCMS). ACHRDCMS is the third disorder associated with postsynaptic CMS which could result from mutations in the proteins forming the muscle AChR. Mutations underlying AChR deficiency cause a 'loss of function' and show recessive inheritance.

Defects in RAPSN are the cause of fetal akinesia deformation sequence (FADS) [MIM:208150]; also known as Pena-Shokeir syndrome type 1 or fetal akinesia sequence or arthrogryposis multiplex congenita with pulmonary hypoplasia. FADS is a rare condition characterized by decreased intrauterine fetal movement, congenital limb contractures, pulmonary hypoplasia, polyhydramnios and craniofacial abnormalities.

### 配列類似性

Belongs to the RAPsyn family.  
Contains 1 RING-type zinc finger.  
Contains 7 TPR repeats.

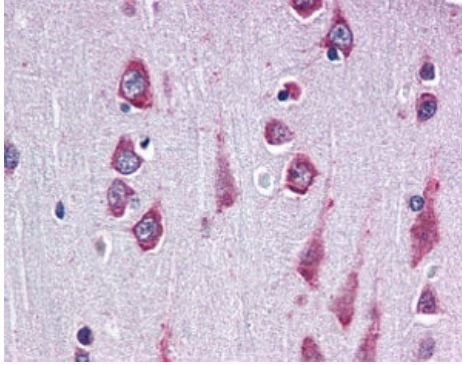
### ドメイン

A cysteine-rich region homologous to part of the regulatory domain of protein kinase C may be important in interactions of this protein with the lipid bilayer.

### 細胞内局在

Cell membrane. Cell junction > synapse > postsynaptic cell membrane. Cytoplasm > cytoskeleton. Cytoplasmic surface of postsynaptic membranes.

## 画像



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Rapsyn antibody (ab118491)

ab118491 at 20µg/ml staining Rapsyn in Formalin-fixed, Paraffin-embedded Human brain (cortex) tissue by Immunohistochemistry, followed by biotinylated secondary antibody, alkaline phosphatase-streptavidin and chromogen.

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

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