# abcam

## Product datasheet

# Anti-CHRND antibody [1H1F9] ab233758

### 4 Images

#### Overview

Product name Anti-CHRND antibody [1H1F9]

**Description** Mouse monoclonal [1H1F9] to CHRND

Host species Mouse

Tested applications
Suitable for: WB, Flow Cyt
Species reactivity
Reacts with: Rat, Human

Immunogen Recombinant fragment corresponding to Human CHRND aa 1-250. Expressed in E.coli.

Database link: Q07001

Run BLAST with
Run BLAST with

Positive control WB: Recombinant human CHRND protein (AA: extra 22-245); C6 and CHRND (AA: extra 22-

245)-hlgGFc transfected HEK-293 whole cell lysate. Flow: SK-N-SH cells.

General notes

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

#### **Properties**

Form Liquid

Storage instructions Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long

term. Avoid freeze / thaw cycle.

**Storage buffer** Preservative: 0.05% Sodium azide

Constituent: PBS

**Purity** Protein G purified

**Clonality** Monoclonal

Clone number 1H1F9

**Isotype** IgG1

#### **Applications**

The Abpromise guarantee

Our **Abpromise guarantee** covers the use of ab233758 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
WB		1/500 - 1/2000.
Flow Cyt		1/200 - 1/400.

-	_		
П	<b>a</b>	ra	Δt

**Function** 

After binding acetylcholine, the AChR responds by an extensive change in conformation that affects all subunits and leads to opening of an ion-conducting channel across the plasma membrane.

Involvement in disease

Defects in CHRND are a cause of multiple pterygium syndrome lethal type (MUPSL) [MIM:253290]. Multiple pterygia are found infrequently in children with arthrogryposis and in fetuses with fetal akinesia syndrome. In lethal multiple pterygium syndrome there is intrauterine growth retardation, multiple pterygia, and flexion contractures causing severe arthrogryposis and fetal akinesia. Subcutaneous edema can be severe, causing fetal hydrops with cystic hygroma and lung hypoplasia. Oligohydramnios and facial anomalies are frequent.

Defects in CHRND are a cause of congenital myasthenic syndrome slow-channel type (SCCMS) [MIM:601462]. SCCMS is the most common congenital myasthenic syndrome. Congenital myasthenic syndromes are characterized by muscle weakness affecting the axial and limb muscles (with hypotonia in early-onset forms), the ocular muscles (leading to ptosis and ophthalmoplegia), and the facial and bulbar musculature (affecting sucking and swallowing, and leading to dysphonia). The symptoms fluctuate and worsen with physical effort. SCCMS is caused by kinetic abnormalities of the AChR, resulting in prolonged endplate currents and prolonged AChR channel opening episodes.

Defects in CHRND are a cause of congenital myasthenic syndrome fast-channel type (FCCMS) [MIM:608930]. FCCMS is a congenital myasthenic syndrome characterized by kinetic abnormalities of the AChR. In most cases, FCCMS is due to mutations that decrease activity of the AChR by slowing the rate of opening of the receptor channel, speeding the rate of closure of the channel, or decreasing the number of openings of the channel during ACh occupancy. The result is failure to achieve threshold depolarization of the endplate and consequent failure to fire an action potential.

Sequence similarities

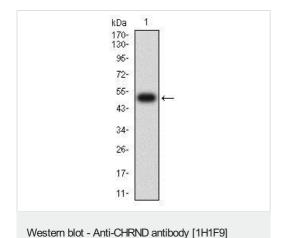
Belongs to the ligand-gated ion channel (TC 1.A.9) family. Acetylcholine receptor (TC 1.A.9.1)

subfamily. Delta/CHRND sub-subfamily.

**Cellular localization** 

Cell junction > synapse > postsynaptic cell membrane. Cell membrane.

#### **Images**

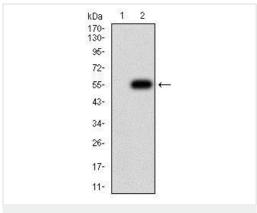


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Anti-CHRND antibody [1H1F9] (ab233758) at 1/500 dilution + Recombinant human CHRND (AA: extra 22-245)

Developed using the ECL technique.



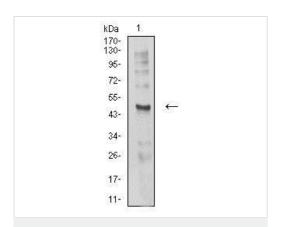
Western blot - Anti-CHRND antibody [1H1F9]

**All lanes :** Anti-CHRND antibody [1H1F9] (ab233758) at 1/500 dilution

**Lane 1 :** HEK-293 (human epithelial cell line from embryonic kidney) whole cell lysate

**Lane 2 :** CHRND (AA: extra 22-245)-hlgGFc transfected HEK-293 whole cell lysate

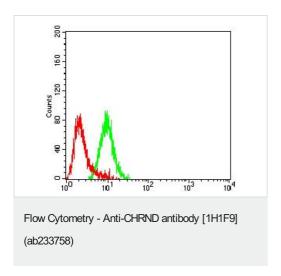
Developed using the ECL technique.



Western blot - Anti-CHRND antibody [1H1F9] (ab233758)

Anti-CHRND antibody [1H1F9] (ab233758) at 1/500 dilution + C6 (rat glial tumor cell line) whole cell lysate

Developed using the ECL technique.



Flow cytometric analysis of SK-N-SH (human neuroblastoma cell line) cell line labeling CHRND with ab233758 at 1/200 dilution (green) compared with a negative control (green).

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

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