**BACKGROUND**

Amyotrophic lateral sclerosis (ALS) is a fatal neurodegenerative disease characterized by progressive limb or bulbar weakness. Mutations in the ALS2 gene result in a number of juvenile recessive motor neuron diseases (MNDs), including juvenile primary lateral sclerosis (JPLS), infantile onset ascending hereditary spastic paraplegia (IAHSP) and a form of complicated hereditary spastic paraplegia (cHSP). The ALS2 gene encodes the Alsin protein, which acts as a guanine nucleotide exchange factor for Rab 5, a modulator of the endocytic pathway. Alsin is a cytosolic protein that is associated with small, punctate membrane structures, suggesting Alsin may mediate membrane transport events, potentially linking endocytic processes and actin cytoskeleton remodeling. ALS2CR4 (amyotrophic lateral sclerosis 2 (juvenile) chromosome region, candidate 4) is a 432 amino acid multi-pass membrane protein highly enriched in retina and localizes to photoreceptor outer segments, ciliary complex and postsynaptic outer plexiform layer. Encoded by a gene that maps to human chromosome 2q33.1, ALS2CR4 exists as two alternatively spliced isoforms.

**REFERENCES**


**CHROMOSOMAL LOCATION**

Genetic locus: TME237 (human) mapping to 2q33.1; Tmem237 (mouse) mapping to 1C1.3.

**SOURCE**

ALS2CR4 (G-8) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 205-226 within an internal region of ALS2CR4 of human origin.

**PRODUCT**

Each vial contains 200 µg IgG κ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

ALS2CR4 (G-8) is available conjugated to agarose (sc-514651 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-514651 HRP), 200 µg/ml, for WB, HRP, and ELISA; to either phycocyanin (sc-514651 PE), fluorescein (sc-514651 FITC), Alexa Fluor® 488 (sc-514651 AF488), Alexa Fluor® 546 (sc-514651 AF546), Alexa Fluor® 594 (sc-514651 AF594) or Alexa Fluor® 647 (sc-514651 AF647), 200 µg/ml, for WB (RGB), IF, IHC and FCM; and to either Alexa Fluor® 680 (sc-514651 AF680) or Alexa Fluor® 790 (sc-514651 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

Blocking peptide available for competition studies, sc-514651 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

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**APPLICATIONS**

ALS2CR4 (G-8) is recommended for detection of ALS2CR4 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).


**RECOMMENDED SUPPORT REAGENTS**

To ensure optimal results, the following support reagents are recommended:


**DATA**

**STORAGE**

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

**RESEARCH USE**

For research use only, not for use in diagnostic procedures.

**PROTOCOLS**

See our web site at www.scbt.com for detailed protocols and support products.