

ARL13B (6F11): sc-293467

BACKGROUND

ADP-ribosylation factors (ARFs) are highly conserved guanine nucleotide binding proteins that enhance the ADP-ribosyltransferase activity of Cholera Toxin. ARFs are important in eukaryotic vesicular trafficking pathways and they play an essential role in the activation of phospholipase D (PC-PLD). ARL13B (ADP-ribosylation factor-like 13B), also known as ARL2L1 or JBTS8, is a 428 amino acid protein that belongs to the ARL subfamily of ARF-like GTPases and is thought to be involved in cilia formation. Defects in the gene encoding ARL13B are associated with Joubert syndrome (JS), a rare genetic disorder of the brain that is characterized by an underdeveloped cerebellum and brain stem and often leads to ataxia, abnormal breathing and seizures.

REFERENCES

1. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 608922. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
2. Fan, Y., et al. 2004. Mutations in a member of the Ras superfamily of small GTP-binding proteins causes Bardet-Biedl syndrome. *Nat. Genet.* 36: 989-993.
3. Caspary, T., et al. 2007. The graded response to Sonic hedgehog depends on cilia architecture. *Dev. Cell* 12: 767-778.
4. Ray, J., et al. 2007. Joubert syndrome: a major brain malformation. *J. Indian Med. Assoc.* 105: 392-394.
5. Cantagrel, V., et al. 2008. Mutations in the cilia gene ARL13B lead to the classical form of Joubert syndrome. *Am. J. Hum. Genet.* 83: 170-179.
6. Hori, Y., et al. 2008. Domain architecture of the atypical Arf-family GTPase Arl13b involved in cilia formation. *Biochem. Biophys. Res. Commun.* 373: 119-124.
7. Millen, K.J., et al. 2008. Cerebellar development and disease. *Curr. Opin. Neurobiol.* 18: 12-19.
8. Sampathkumar, K., et al. 2008. Joubert syndrome. *Kidney Int.* 74: 1222.

CHROMOSOMAL LOCATION

Genetic locus: ARL13B (human) mapping to 3q11.1.

SOURCE

ARL13B (6F11) is a mouse monoclonal antibody raised against amino acids 329-428 representing partial length ARL13B of human origin.

PRODUCT

Each vial contains 100 µg IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

STORAGE

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

APPLICATIONS

ARL13B (6F11) is recommended for detection of ARL13B of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

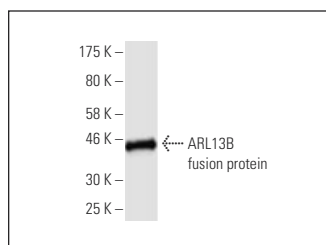
Suitable for use as control antibody for ARL13B siRNA (h): sc-78165, ARL13B shRNA Plasmid (h): sc-78165-SH and ARL13B shRNA (h) Lentiviral Particles: sc-78165-V.

Molecular Weight of ARL13B: 49 kDa.

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml).

DATA



ARL13B (6F11): sc-293467. Western blot analysis of human recombinant ARL13B fusion protein.

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.