

CEP290 (B-7): sc-390462

BACKGROUND

Centrosomes are the major microtubule-organizing centers of mammalian cells. They are composed of a centriole pair and surrounding microtubule-nucleating material, termed pericentriolar material (PCM). Bipolar mitotic spindle assembly relies on two intertwined processes: centriole duplication and centrosome maturation. Failure to properly orchestrate centrosome duplication and maturation is subsequently linked to spindle defects, which can result in aneuploidy and promote cancer progression. The CEP290 (centrosomal protein of 290 kDa) gene encodes a protein of 2,479 amino acids that activates CREB-2-mediated transcription. Specifically, CEP290 ensures the correct localization of ciliary and phototransduction proteins in retinal photoreceptor cells. Mutations in the CEP290 gene have been identified in several diseases, including Joubert syndrome type 5 (JBTS5), Senior-Løken syndrome type 6 (SLSN6), Leber congenital amaurosis type 10 (LCA10) and Meckel syndrome type 4 (MKS4).

CHROMOSOMAL LOCATION

Genetic locus: CEP290 (human) mapping to 12q21.32.

SOURCE

CEP290 (B-7) is a mouse monoclonal antibody raised against amino acids 1441-1740 mapping within an internal region of CEP290 of human origin.

PRODUCT

Each vial contains 200 µg IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

CEP290 (B-7) is available conjugated to agarose (sc-390462 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-390462 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-390462 PE), fluorescein (sc-390462 FITC), Alexa Fluor® 488 (sc-390462 AF488), Alexa Fluor® 546 (sc-390462 AF546), Alexa Fluor® 594 (sc-390462 AF594) or Alexa Fluor® 647 (sc-390462 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-390462 AF680) or Alexa Fluor® 790 (sc-390462 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

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APPLICATIONS

CEP290 (B-7) is recommended for detection of CEP290 of 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).

Suitable for use as control antibody for CEP290 siRNA (h): sc-72865, CEP290 shRNA Plasmid (h): sc-72865-SH and CEP290 shRNA (h) Lentiviral Particles: sc-72865-V.

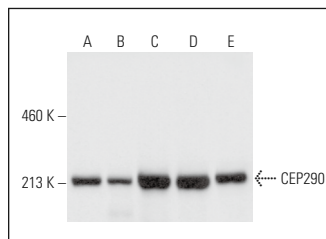
Molecular Weight of CEP290: 290 kDa.

Positive Controls: CCRF-CEM cell lysate: sc-2225, HeLa whole cell lysate: sc-2200 or HEK293 whole cell lysate: sc-45136.

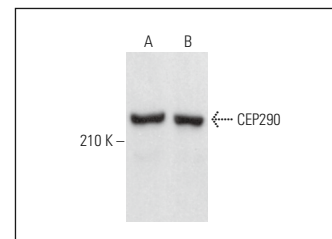
STORAGE

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

DATA



CEP290 (B-7): sc-390462. Western blot analysis of CEP290 expression in HeLa (A), HEK293 (B), K-562 (C), MCF7 (D) and NTERA-2 cl.D1 (E) whole cell lysates.



CEP290 (B-7): sc-390462. Western blot analysis of CEP290 expression in CCRF-CEM (A) and HEL 92.1.7 (B) whole cell lysates.

SELECT PRODUCT CITATIONS

- Barbelanne, M., et al. 2015. Nephrocystin proteins NPHP5 and CEP290 regulate BBSome integrity, ciliary trafficking and cargo delivery. *Hum. Mol. Genet.* 24: 2185-2200.
- Han, K.J., et al. 2019. Deubiquitylase USP9X maintains centriolar satellite integrity by stabilizing pericentriolar material 1 protein. *J. Cell Sci.* 132: jcs221663.
- Zhang, R., et al. 2020. Whole exome sequencing identified a homozygous novel variant in CEP290 gene causes Meckel syndrome. *J. Cell. Mol. Med.* 24: 1906-1916.
- Chahine Karam, F., et al. 2022. Human iPSC-derived retinal organoids and retinal pigment epithelium for novel intronic RPGR variant assessment for therapy suitability. *J. Pers. Med.* 12: 502.
- Gaudin, N., et al. 2022. Evolutionary conservation of centriole rotational asymmetry in the human centrosome. *Elife* 11: e72382.
- Chahine Karam, F., et al. 2022. Human iPSC-derived retinal organoids and retinal pigment epithelium for novel intronic RPGR variant assessment for therapy suitability. *J. Pers. Med.* 12: 502.
- Renaud, C.C.N., et al. 2023. The centrosomal protein 131 participates in the regulation of mitochondrial apoptosis. *Commun. Biol.* 6: 1271.

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.