

fibrillin-2 (H-10): sc-393968

BACKGROUND

Extracellular glycoproteins fibrillin-1 and -2 are major components of connective tissue microfibrils. Fibrillin-2 containing microfibrils regulate the early process of elastic fiber assembly in tissue. Mutations in the fibrillin-2 gene resulting in impaired assembly of fibrillin-2 may lead to molecular congenital contractural arachnodactyly. Fibrillin-2 constitutes the backbone of microfibrils which insert directly into the lamina densa of basement membranes. Epithelial cells primarily deposit fibrillin into the extracellular matrix in a nonfibrillar form. Mutations in the 8-cysteine motif of fibrillin-2 alters its binding to microfibril-associated glycoprotein-1 (MAGP-1), which may increase the severity of congenital contractural arachnodactyly.

CHROMOSOMAL LOCATION

Genetic locus: FBN2 (human) mapping to 5q23.3; Fbn2 (mouse) mapping to 18 D3.

SOURCE

fibrillin-2 (H-10) is a mouse monoclonal antibody raised against amino acids 397-436 mapping within an internal region of fibrillin-2 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.

fibrillin-2 (H-10) is available conjugated to agarose (sc-393968 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-393968 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-393968 PE), fluorescein (sc-393968 FITC), Alexa Fluor[®] 488 (sc-393968 AF488), Alexa Fluor[®] 546 (sc-393968 AF546), Alexa Fluor[®] 594 (sc-393968 AF594) or Alexa Fluor[®] 647 (sc-393968 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor[®] 680 (sc-393968 AF680) or Alexa Fluor[®] 790 (sc-393968 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

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RESEARCH USE

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

APPLICATIONS

fibrillin-2 (H-10) is recommended for detection of fibrillin-2 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).

Suitable for use as control antibody for fibrillin-2 siRNA (h): sc-45971, fibrillin-2 siRNA (m): sc-45972, fibrillin-2 shRNA Plasmid (h): sc-45971-SH, fibrillin-2 shRNA Plasmid (m): sc-45972-SH, fibrillin-2 shRNA (h) Lentiviral Particles: sc-45971-V and fibrillin-2 shRNA (m) Lentiviral Particles: sc-45972-V.

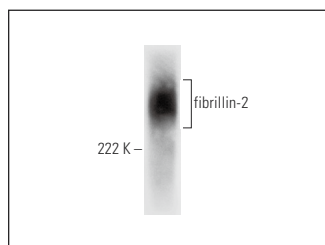
Molecular Weight of fibrillin-2: 350 kDa.

Positive Controls: mouse embryo extract: sc-364239.

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). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz[®] Mounting Medium: sc-24941 or UltraCruz[®] Hard-set Mounting Medium: sc-359850.

DATA



fibrillin-2 (H-10): sc-393968. Western blot analysis of fibrillin-2 expression in mouse embryo tissue extract.

SELECT PRODUCT CITATIONS

1. Mendoza-Topaz, C., et al. 2018. Cells respond to deletion of CAV1 by increasing synthesis of extracellular matrix. PLoS ONE 13: e0205306.
2. Yu, X., et al. 2021. Nanophthalmos-associated MYRF gene mutation causes ciliary zonule defects in mice. Invest. Ophthalmol. Vis. Sci. 62: 1.
3. Zhang, S., et al. 2021. RNA polymerase II is required for spatial chromatin reorganization following exit from mitosis. Sci. Adv. 7: eabg8205.
4. Xu, F., et al. 2022. Association between anti-fibrillin-2 protein induced retinal degeneration via intravitreal delivery and activated TGF-β signalling in mice. Clin. Exp. Pharmacol. Physiol. 49: 586-595.
5. Zhang, R.X., et al. 2023. Intravitreal injection of fibrillin-2 (Fbn2) recombinant protein for therapy of retinopathy in a retina-specific Fbn2 knock-down mouse model. Sci. Rep. 13: 6865.
6. Chrisochoidou, Y., et al. 2023. Crosstalk with lung fibroblasts shapes the growth and therapeutic response of mesothelioma cells. Cell Death Dis. 14: 725.

STORAGE

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

PROTOCOLS

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