

COL6A3 (A-5): sc-515335

BACKGROUND

Collagens (COLs) are fibrous, extracellular matrix proteins with high tensile strength that function as the major components of connective tissue, such as tendons and cartilage. All COL proteins contain a triple helix domain and frequently show lateral self-association in order to form complex connective tissues. There are several types of COL proteins, including fibril-forming interstitial COLs (types I, II, III and V), basement membrane COLs (type IV) and beaded filament COLs (type VI). COL6A3 (collagen α -3(VI) chain), is a 3,176 amino acid secreted protein that contains one fibronectin type-III domain, one MPTI inhibitor domain and 12 VWFA domains and functions as the third (and largest) of 3 α chains of the type VI COL protein complex. Existing as a trimer with two other type VI α proteins, COL6A3 acts as a cell-binding protein that plays an important role in the organization of matrix components. Defects in the gene encoding COL6A3 are the cause of Bethlem myopathy (BM), a rare autosomal proximal myopathy, and Ullrich congenital muscular dystrophy (UCMD), an autosomal recessive congenital myopathy. Multiple isoforms of COL6A3 exist due to alternative splicing events.

REFERENCES

- Weil, D., et al. 1988. Cloning and chromosomal localization of human genes encoding the three chains of type VI collagen. *Am. J. Hum. Genet.* 42: 435-445.
- Demir, E., et al. 2002. Mutations in COL6A3 cause severe and mild phenotypes of Ullrich congenital muscular dystrophy. *Am. J. Hum. Genet.* 70: 1446-1458.

CHROMOSOMAL LOCATION

Genetic locus: COL6A3 (human) mapping to 2q37.3.

SOURCE

COL6A3 (A-5) is a mouse monoclonal antibody raised against amino acids 301-600 mapping within an internal region of Collagen α 3 Type VI of human origin.

PRODUCT

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

COL6A3 (A-5) is available conjugated to agarose (sc-515335 AC), 500 μ g/0.25 ml agarose in 1 ml, for IP; to HRP (sc-515335 HRP), 200 μ g/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-515335 PE), fluorescein (sc-515335 FITC), Alexa Fluor[®] 488 (sc-515335 AF488), Alexa Fluor[®] 546 (sc-515335 AF546), Alexa Fluor[®] 594 (sc-515335 AF594) or Alexa Fluor[®] 647 (sc-515335 AF647), 200 μ g/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor[®] 680 (sc-515335 AF680) or Alexa Fluor[®] 790 (sc-515335 AF790), 200 μ g/ml, for Near-Infrared (NIR) WB, IF and FCM.

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STORAGE

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

APPLICATIONS

COL6A3 (A-5) is recommended for detection of Collagen α 3 Type VI isoforms 1 and 2 of human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g 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 COL6A3 siRNA (h): sc-94560, COL6A3 shRNA Plasmid (h): sc-94560-SH and COL6A3 shRNA (h) Lentiviral Particles: sc-94560-V.

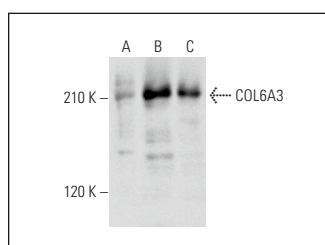
Molecular Weight of COL6A3 isoforms: 260-300 kDa.

Positive Controls: human heart extract: sc-363763, human skeletal muscle extract: sc-363776 or human thymus extract: sc-516711.

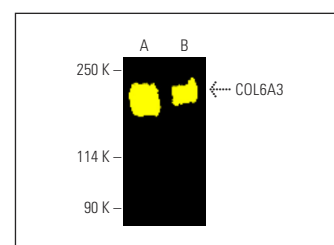
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ 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-IgG κ BP-FITC: sc-516140 or m-IgG κ 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



COL6A3 (A-5): sc-515335. Western blot analysis of COL6A3 expression in human skeletal muscle (A), human thymus (B) and human heart (C) tissue extracts.



COL6A3 (A-5): sc-515335. Fluorescent western blot analysis of COL6A3 expression in human thymus tissue extract (A) and Hs67 whole cell lysate (B). Blocked with UltraCruz[®] Blocking Reagent: sc-516214. Detection reagent used: m-IgG₁ BP-CFL 488: sc-533661.

SELECT PRODUCT CITATIONS

- Marakhonov, A.V., et al. 2018. Two novel COL6A3 mutations disrupt extracellular matrix formation and lead to myopathy from Ullrich congenital muscular dystrophy and Bethlem myopathy spectrum. *Gene* 672: 165-171.
- Kashpur, O., et al. 2019. Differentiation of diabetic foot ulcer-derived induced pluripotent stem cells reveals distinct cellular and tissue phenotypes. *FASEB J.* 33: 1262-1277.

RESEARCH USE

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