

Mouse Anti-Collagen VI [3C4]: MC0579, MC0579RTU7

Intended Use: For Research Use Only

Description: Collagen VI is a major structural component of microfibrils. Collagen VI was found to be present throughout the connective tissue and in the extracellular matrix of cultured fibroblasts. It is a heterotrimer composed of three different chains: alpha-1, alpha-2, and alpha-3 or alpha-5 or alpha-6. Defects in Collagen VI are a cause of Bethlem myopathy (BM). BM is a rare autosomal dominant proximal myopathy characterized by early childhood onset (complete penetrance by the age of 5) and joint contractures most frequently affecting the elbows and ankles. Defects in Collagen VI are a cause of Ullrich congenital muscular dystrophy (UCMD). UCMD is an autosomal recessive congenital myopathy characterized by muscle weakness and multiple joint contractures, generally noted at birth or early infancy. The clinical course is more severe than in Bethlem myopathy. Mutations in this gene result in Bethlem myopathy and Ullrich congenital muscular dystrophy.

Specifications:

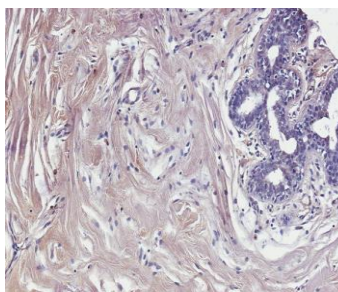
Clone: 3C4
 Source: Mouse
 Isotype: IgG1k
 Reactivity: Human
 Immunogen: Purified human Collagen VI
 Localization: Secreted, extracellular matrix
 Formulation: Antibody in PBS pH7.4, containing BSA and $\leq 0.09\%$ sodium azide (NaN₃)
 Storage: Store at 2°- 8°C
 Applications: IHC, IF, WB
 Package:

Description	Catalog No.	Size
Collagen VI Concentrated	MC0579	1 ml
Collagen VI Prediluted	MC0579RTU7	7 ml

IHC Procedure*:

Positive Control Tissue: Liver, cardiac muscle, kidney, skin, stomach, placenta
 Concentrated Dilution: 10-100
 Pretreatment: Tris EDTA pH9.0, 15 minutes Pressure Cooker or 30-60 minutes water bath at 95°-99°C
 Incubation Time and Temp: Overnight @ 4°C
 Detection: Refer to the detection system manual

* Result should be confirmed by an established diagnostic procedure.



FFPE human breast stained with anti-Collagen VI using DAB

References:

1. Changes in stiffness and biochemical composition of the pericellular matrix as a function of spatial chondrocyte organisation in osteoarthritic cartilage. Danalache M, et al. Osteoarthritis Cartilage N/A:N/A, 2019.
2. Perfect chronic skeletal muscle regeneration in adult spiny mice, *Acomys cahirinus*. Maden M. Sci Rep 8:8920, 2018.
3. Authentication of collagen VI antibodies. Endicott J, et al. BMC Res Notes 10:358, 2017.
4. De novo mutations of TUBA3D are associated with keratoconus. Hao XD, et al. Sci Rep 7:13570, 2017.