

**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.

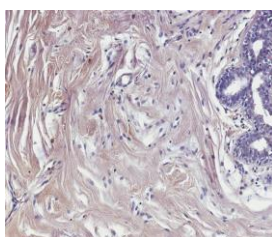
**Specifications:**

Clone: 3C4  
 Source: Mouse  
 Isotype: IgG1k  
 Reactivity: Human  
 Localization: Secreted. extracellular matrix  
 Formulation: Antibody in PBS pH7.4, containing BSA and ≤ 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: Skin, placenta, kidney  
 Concentrated Dilution: 25-100  
 Pretreatment: Tris EDTA pH9.0, 15 minutes using Pressure Cooker, or 30-60 minutes using water bath at 95°-99°C  
 Incubation Time and Temp: 30-60 minutes @ RT  
 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, et al. 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.