



COL6A3 Polyclonal Antibody

Cat #: ABP53749

Size: 30µl /100µl /200µl

Product Information

	Product Name: COL6A3 Polyclonal Antibody		
	Applications: IHC-P, IF, ELISA		Isotype: Rabbit IgG
	Reactivity: Human, Mouse		
REF	Catalog Number: ABP53749	LOT	Lot Number: Refer to product label
	Formulation: Liquid		Concentration: 1 mg/ml
	Storage: Store at -20°C. Avoid repeated freeze / thaw cycles.		Note: Contain sodium azide.

Background: COL6A3 encodes the alpha-3 chain, one of the three alpha chains of type VI collagen, a beaded filament collagen found in most connective tissues. The alpha-3 chain of type VI collagen is much larger than the alpha-1 and -2 chains. This difference in size is largely due to an increase in the number of subdomains, similar to von Willebrand Factor type A domains, that are found in the amino terminal globular domain of all the alpha chains. These domains have been shown to bind extracellular matrix proteins, an interaction that explains the importance of this collagen in organizing matrix components. Mutations in the type VI collagen genes are associated with Bethlem myopathy, a rare autosomal dominant proximal myopathy with early childhood onset. Mutations in this gene are also a cause of Ullrich congenital muscular dystrophy, also referred to as Ullrich scleroatonic muscular dystrophy, an autosomal recessive congenital myopathy that is more severe than Bethlem myopathy. Multiple transcript variants have been identified, but the full-length nature of only some of these variants has been described.

Application Notes: Optimal working dilutions should be determined experimentally by the investigator. Suggested starting dilutions are as follows: IHC-P (1:100-1:300), IF (1:200-1:1000), ELISA (1:40000). Not yet tested in other applications.

Storage Buffer: PBS containing 50% Glycerol, 0.5% BSA and 0.02% Sodium Azide.

Storage Instructions: Stable for one year at -20°C from date of shipment. For maximum recovery of product, centrifuge the original vial after thawing and prior to removing the cap. Aliquot to avoid repeated freezing and thawing.

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