



Rabbit Anti Human COL6A3 monoclonal Antibody, Clone 4 (CABT-L0599YC)

This product is for research use only and is not intended for diagnostic use.

PRODUCT INFORMATION

Product Overview	Recombinant Rabbit anti Human COL6A3-Specific Monoclonal Antibody
Specificity	Human COL6A3
Immunogen	Recombinant protein
Isotype	IgG
Source/Host	Rabbit
Species Reactivity	Human
Clone	4
Purification	Affinity Purification
Conjugate	Unconjugated
Applications	Sandwich ELISA, ELISA(Cap), Cytometric bead array We recommend the following for sandwich ELISA (Capture - Detection): CABT-L0599YC - CABT-L0599YD Each laboratory should determine an optimum working titer for use in its particular application. Other applications have not been tested but use in such assays should not necessarily be excluded.
Format	Purified, Liquid
Concentration	Lot specific
Size	100 μg

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Buffer	PBS
Storage	For short-term storage, store at 2-8°C. For long-term storage, aliquot and store at -20°C. Avoid multiple freeze-thaw cycles.
Ship	Wet ice

BACKGROUND

Introduction

This gene 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.

Keywords

COL6A3, Collagen Type VI, Collagen alpha-3(VI) chain, Collagen alpha 3(VI) chain, 230540E3

GENE INFORMATION

Entrez Gene ID	<u>1293</u>
UniProt ID	<u>P12111</u>