

# Recombinant Human COL6A3/Collagen-VI Protein

Catalog Number:PKSH030495

1 Publications



**Note:** Centrifuge before opening to ensure complete recovery of vial contents.

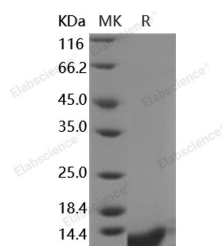
## Description

<b>Synonyms</b>	COL6A3;Collagen-VI
<b>Species</b>	Human
<b>Expression Host</b>	HEK293 Cells
<b>Sequence</b>	Thr3101-Thr3177
<b>Accession</b>	NP_004360.2
<b>Calculated Molecular Weight</b>	8.5 kDa
<b>Tag</b>	None

## Properties

<b>Purity</b>	> 95 % as determined by reducing SDS-PAGE.
<b>Endotoxin</b>	< 1.0 EU per µg of the protein as determined by the LAL method.
<b>Storage</b>	Generally, lyophilized proteins are stable for up to 12 months when stored at -20 to -80°C. Reconstituted protein solution can be stored at 4-8°C for 2-7 days. Aliquots of reconstituted samples are stable at < -20°C for 3 months.
<b>Shipping</b>	This product is provided as lyophilized powder which is shipped with ice packs.
<b>Formulation</b>	Lyophilized from sterile PBS, pH 7.4 Normally 5 % - 8 % trehalose, mannitol and 0.01% Tween80 are added as protectants before lyophilization. Please refer to the specific buffer information in the printed manual.
<b>Reconstitution</b>	Please refer to the printed manual for detailed information.

## Data



> 95 % as determined by reducing SDS-PAGE.

## Background

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.

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