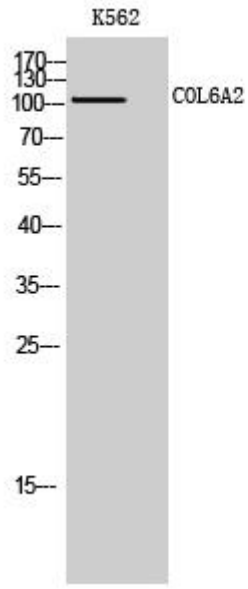
COL6A2 Monoclonal Antibody

Catalog No	BYmab-16994
Isotype	IgG
Reactivity	Human;Mouse;Monkey
Applications	WB
Gene Name	COL6A2
Protein Name	Collagen alpha-2(VI) chain
Immunogen	The antiserum was produced against synthesized peptide derived from human Collagen VI alpha2. AA range:691-740
Specificity	COL6A2 Monoclonal Antibody detects endogenous levels of COL6A2 protein.
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	COL6A2; Collagen alpha-2(VI) chain
Observed Band	109kD
Cell Pathway	Secreted, extracellular space, extracellular matrix . Membrane ; Peripheral membrane protein . Recruited on membranes by CSPG4.
Tissue Specificity	Fibroblast,Kidney,Liver,Ovary,Placenta,Uterus,
Function	disease:Defects in COL6A2 are a cause of Bethlem myopathy (BM) [MIM:158810]. 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.,disease:Defects in COL6A2 are a cause of Ullrich congenital muscular dystrophy (UCMD) [MIM:254090]; also known as Ullrich scleroatonic muscular dystrophy. 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.,function:Collagen VI acts as a cell-binding protein.,PTM:Prolines at the third position of the tripeptide repeating unit (G-X-Y) are hydroxylated in some or

	all of the chains.,similarity:Belongs to the type VI collagen family.,similarity:Contains 3
Background	This gene encodes one of the three alpha chains of type VI collagen, a beaded filament collagen found in most connective tissues. The product of this gene contains several domains similar to von Willebrand Factor type A domains. 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 this gene are associated with Bethlem myopathy and Ullrich scleroatonic muscular dystrophy. Three transcript variants have been identified for this gene. [provided by RefSeq, Jul 2008],
matters needing attention	Avoid repeated freezing and thawing!
Usage suggestions	This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



Western Blot analysis of various cells using COL6A2 Monoclonal Antibody