


## Product datasheet

# Anti-Collagen VI antibody ab99249

[1 References](#) [1 Image](#)

### Overview

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|                            |  |
|----------------------------|--|
| <b>Product name</b>        | Anti-Collagen VI antibody  |
| <b>Description</b>         | Rabbit polyclonal to Collagen VI   |
| <b>Tested applications</b> | <b>Suitable for:</b> WB  |
| <b>Species reactivity</b>  | <b>Reacts with:</b> Mouse, Human<br><b>Predicted to work with:</b> Rat, Horse, Cow, Dog, Pig, Chimpanzee, Macaque monkey, Gorilla<br> |
| <b>Immunogen</b>           | Synthetic peptide conjugated to KLH derived from within residues 150 - 250 of Human Collagen VI. Read Abcam's proprietary immunogen policy   |
| <b>Positive control</b>    | This antibody gave a positive signal in the following tissue lysates: Human Skeletal Muscle; Human Skin; Human Heart as well as the following whole cell lysates: MEF1; NIH3T3; STO; WI38.                             |

### Properties

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|                             |  |
|-----------------------------|--|
| <b>Form</b>                 | Liquid   |
| <b>Storage instructions</b> | Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle.   |
| <b>Storage buffer</b>       | pH: 7.40<br>Preservative: 0.02% Sodium azide<br>Constituent: PBS<br>Note: Batches of this product that have a concentration < 1mg/ml may have BSA added as a stabilising agent. If you would like information about the formulation of a specific lot, please contact our scientific support team who will be happy to help. |
| <b>Purity</b>               | Immunogen affinity purified  |
| <b>Clonality</b>            | Polyclonal   |
| <b>Isotype</b>              | IgG  |

### Applications

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Our [Abpromise guarantee](#) covers the use of **ab99249** in the following tested applications.

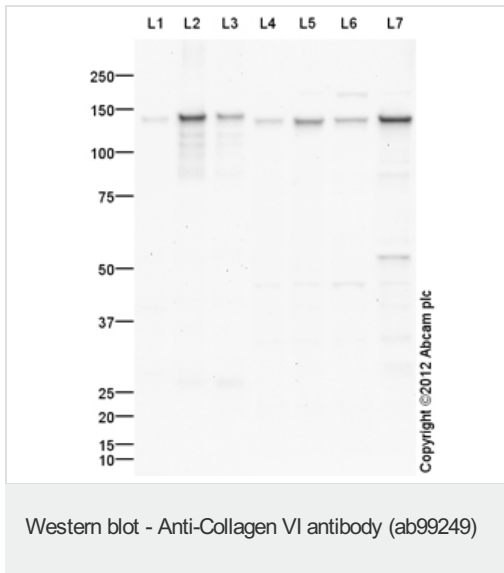
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

| Application | Abreviews | Notes  |
|-------------|-----------|--|
| WB          |           | Use a concentration of 1 µg/ml. Detects a band of approximately 147 kDa (predicted molecular weight: 108 kDa). |

## Target

|   |   |
|---|---|
| <b>Function</b>                         | Collagen VI acts as a cell-binding protein.   |
| <b>Involvement in disease</b>           | <p>Defects in COL6A1 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.</p> <p>Defects in COL6A1 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.</p> |
| <b>Sequence similarities</b>            | <p>Belongs to the type VI collagen family.</p> <p>Contains 3 VWFA domains.</p>  |
| <b>Post-translational modifications</b> | Prolines at the third position of the tripeptide repeating unit (G-X-Y) are hydroxylated in some or all of the chains.  |
| <b>Cellular localization</b>            | Secreted > extracellular space > extracellular matrix.  |

## Images



**All lanes :** Anti-Collagen VI antibody (ab99249) at 1 µg/ml

**Lane 1 :** Human skeletal muscle tissue lysate - total protein (ab29330)

**Lane 2 :** Human thymus tissue lysate - total protein (ab30146)

**Lane 3 :** Human heart tissue lysate - total protein (ab29431)

**Lane 4 :** MEF1 (Mouse embryonic fibroblast cell line) Whole Cell Lysate

**Lane 5 :** NIH 3T3 (Mouse embryonic fibroblast cell line) Whole Cell Lysate

**Lane 6 :** STO (Mouse embryonic fibroblast cell line) Whole Cell Lysate

**Lane 7 :** WI-38 whole cell lysate (ab3960)

Lysates/proteins at 10 µg per lane.

#### Secondary

Goat Anti-Rabbit IgG H&L (HRP) preadsorbed (ab97080) at 1/5000 dilution

Developed using the ECL technique

Performed under reducing conditions.

**Predicted band size :** 108 kDa

**Observed band size :** 147 kDa

**Additional bands at :** 53 kDa. We are unsure as to the identity of these extra bands.

**Exposure time :** 2 minutes

**Please note:** All products are "FOR RESEARCH USE ONLY AND ARE NOT INTENDED FOR DIAGNOSTIC OR THERAPEUTIC USE"

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