abcam

# Product datasheet

# Recombinant Human TGM1 protein ab159694

1 Image

## Overview

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|---|---|
| **Product name** | Recombinant Human TGM1 protein |
| **Protein length** | Protein fragment |

## Description

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| **Nature** | Recombinant |
| **Source** | Wheat germ |
| **Amino Acid Sequence** | |
| **Species** | Human |
| **Sequence** | PFHMLLLLSRTYESSDRITLELLIGNNPEVGKGTHVIIPV GKGGSGGWKA QVVKASGQNLNLRVHTSPNAIIGKFQFTVRTQSDAGEF QLPFDPRNEIYI |
| **Amino acids** | 146 to 245 |
| **Tags** | GST tag N-Terminus |

## Specifications

Our Abpromise guarantee covers the use of **ab159694** in the following tested applications.

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

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| **Applications** | ELISA<br>Western blot |
| **Form** | Liquid |
| **Additional notes** | Protein concentration is above or equal to 0.05 mg/ml. |

## Preparation and Storage

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| **Stability and Storage** | Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles.<br>pH: 8.00<br>Constituents: 0.31% Glutathione, 0.79% Tris HCl |

## General Info

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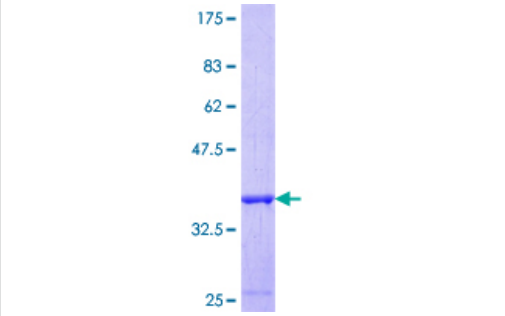
| **Function** | Catalyzes the cross-linking of proteins and the conjugation of polyamines to proteins. Responsible for cross-linking epidermal proteins during formation of the stratum corneum. |
| **Involvement in disease** | Defects in TGM1 are the cause of ichthyosis lamellar type 1 (LI1) [MIM:242300]. LI is a non-bullous ichthyosis, a skin disorder characterized by abnormal cornification of the epidermis. It is one the most severe forms of ichthyoses apparent at birth and persisting throughout life. LI patients are born encased in a tight, shiny, translucent covering called collodion membrane. Over the first weeks of life, the collodion membrane is gradually replaced by generalized large, dark brown, plate-like scales with minimal to no erythroderma. Tautness of facial skin commonly results in ectropion, eclabium and scarring alopecia of the scalp. Common complications are severe heat intolerance and recurrent ear infections.<br>Defects in TGM1 are a cause of non-bullous congenital ichthyosiform erythroderma (NCIE) [MIM:242100]. NCIE is a non-bullous ichthyosis, a skin disorder characterized by abnormal cornification of the epidermis. Most affected individuals are born with a tight, shiny, translucent covering called collodion membrane. The collodion membrane subsequently evolves into generalized scaling and intense redness of the skin. Clinical features are milder than in lamellar ichthyoses and demonstrate a greater variability in the intensity of erythema, size and type of scales. In contrast to lamellar ichthyoses, scales are usually white, fine and powdery, and palms and soles are severely affected. Patients suffer from palmoplantar keratoderma, often with painful fissures, digital contractures, and loss of pulp volume.<br>Defects in TGM1 are the cause of ichthyosis congenital autosomal recessive TGM1-related (ARCI-TGM1) [MIM:242300]. A disorder of keratinization with abnormal differentiation and desquamation of the epidermis resulting in two major clinical entities. Lamellar ichthyosis is a condition often associated with an embedment in a collodion-like membrane at birth; skin scales later develop, covering the entire body surface. Non-bullous congenital ichthyosiform erythroderma characterized by fine whitish scaling on an erythrodermal background; larger brownish scales are present on the buttocks, neck and legs. |
| **Sequence similarities** | Belongs to the transglutaminase superfamily. Transglutaminase family. |
| **Post-translational modifications** | The membrane anchorage region possesses a cluster of five cysteines within which fatty acid(s) may become thioester-linked. It is subject to phorbol ester-stimulated phosphorylation and is hypersensitive to proteolysis, which releases the enzyme in a soluble form. |
| **Cellular localization** | Membrane. |

## Images




SDS-PAGE - Recombinant Human TGM1 protein (ab159694)

ab159694 on a 12.5% SDS-PAGE stained with Coomassie Blue.

**Please note:** All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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