

Product datasheet

Recombinant Human BOLA3 protein (His tag) ab201887

[1 Image](#)

Description

Product name	Recombinant Human BOLA3 protein (His tag)
Purity	> 95 % SDS-PAGE. ab201887 was purified using conventional chromatography.
Expression system	Escherichia coli
Accession	<u>Q53S33</u>
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Sequence	MGSSHHHHHSSGLVPRGSHMGSMAAWSPAAAAPLLR GIRGLPLHHRMFA TQTEGELRVTLKKEKFPRTAIKVTDISGGCGAMYKIES EEFKEKRTVQQHQMVNQALKEEIKEMHGLRIFTSVPKR
Predicted molecular weight	15 kDa including tags
Amino acids	1 to 107
Tags	His tag N-Terminus
Additional sequence information	NCBI Accession No.: NP_997717

Specifications

Our **Abpromise guarantee** covers the use of **ab201887** in the following tested applications.

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

Applications	SDS-PAGE Mass Spectrometry
Mass spectrometry	MALDI-TOF
Form	Liquid

Preparation and Storage

Stability and Storage

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.

pH: 7.40

Constituents: 89% PBS, 10% Glycerol (glycerin, glycerine)

General Info

Tissue specificity

Widely expressed.

Involvement in disease

Defects in BOLA3 are the cause of multiple mitochondrial dysfunctions syndrome type 2 (MMDS2) [MIM:614299]. A severe disorder of systemic energy metabolism, resulting in weakness, respiratory failure, lack of neurologic development, lactic acidosis, hyperglycemia and early death. Some patients show failure to thrive, pulmonary hypertension, hypotonia and irritability. Biochemical features include severe combined deficiency of the 2-oxoacid dehydrogenases, defective lipoic acid synthesis and reduction in activity of mitochondrial respiratory chain complexes.

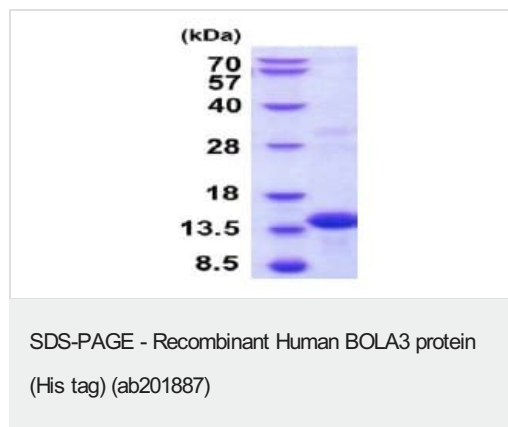
Sequence similarities

Belongs to the bolA/yrbA family.

Cellular localization

Secreted. Could be secreted via a non-classical export pathway.

Images



15% SDS-PAGE analysis of ab201887 (3 µg)

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

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