

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	Q53S33	
Protein length	Full length protein	
Animal free	No	
Nature	Recombinant	
Species	Human	
Sequence	MGSSHHHHHHSSGLVPRGSHMGSMMAAWSPAAAAPL LRGIRGLPLHHRMFA TQTEGELRVTQLKEKFPRTAIKVTDISGGCGAMYEIKI ESEEFKEKRT VQQHQMVNQALKEEIKEMHGLRIFTSVPKR	
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

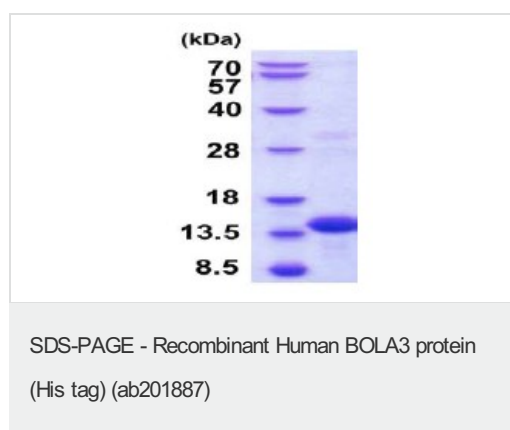
Preparation and Storage

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: 7.4 Constituents: 89% PBS, 10% Glycerol
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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, hyperglycinemia 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)

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