

Product datasheet

Recombinant Human ANT XR2/CMG-2 protein
ab180320

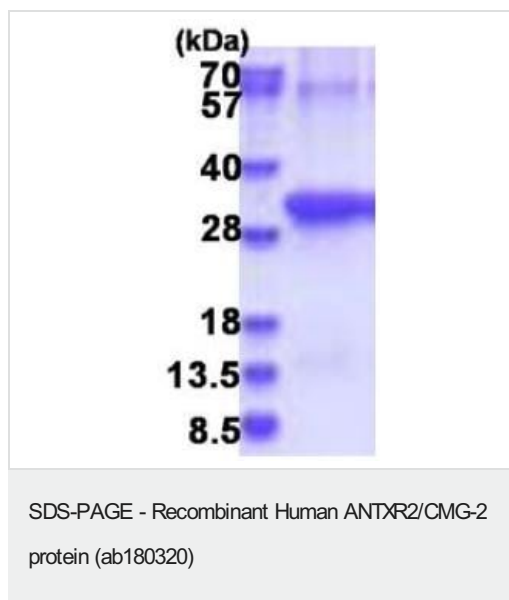
1 Image

Description	
Product name	Recombinant Human ANT XR2/CMG-2 protein
Purity	> 90 % SDS-PAGE. Purified by using conventional chromatography techniques.
Expression system	Escherichia coli
Accession	P5833
Protein length	Protein fragment
Animal free	No
Nature	Recombinant
Species	Human
Sequence	MGSSHHHHHH SSGLVPRGSH MGSQEQPSCR RAFDLYFVLD KSGSVANNWI EYNFVQQLA ERFVSPPEMRL SFMFSSQAT IILPLTGDRG KISKGLEDLK RVSPVGETYI HEGLKLANEQ IQKAGGLKTS SIIIALTDGK LDGLVPSYAE KEAKISRSLG ASVYCVGVLD FEQAQLERIA DSKEQVFPVK GGFQALKGII NSILAQSCTE ILELQPSSVC VGEEFQIVLS GRGFMLGSRN GSVLCITYTN ETYTTSVKPV SVQLNSMLCP APILNKAGET LDVSVSFNGG KSVISGSLIV TATECSN
Predicted molecular weight	33 kDa including tags
Amino acids	34 to 317
Tags	His tag N-Terminus
Additional sequence information	Extracellular domain.

Specifications	
Our Abpromise guarantee covers the use of ab180320 in the following tested applications.	
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.	
Applications	Mass Spectrometry SDS-PAGE
Mass spectrometry	MALDI-TOF

Form	Liquid
Additional notes	Previously labelled as ANT XR2.
Preparation and Storage	
Stability and Storage	<p>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.</p> <p>pH: 8.00</p> <p>Constituents: 10% Glycerol (glycerin, glycerine), 0.58% Sodium chloride, 0.02% DTT, 0.32% Tris HCl</p>
General Info	
Function	Necessary for cellular interactions with laminin and the extracellular matrix.
Tissue specificity	Expressed in prostate, thymus, ovary, testis, pancreas, colon, heart, kidney, lung, liver, peripheral blood leukocytes, placenta, skeletal muscle, small intestine and spleen.
Involvement in disease	<p>Defects in ANT XR2 are the cause of infantile systemic hyalinosis (ISH) [MIM:236490]. This autosomal recessive syndrome is similar to JHF, but has an earlier onset and a more severe course. Symptoms appear at birth or within the first months of life, with painful, swollen joint contractures, osteopenia, osteoporosis and livid red hyperpigmentation over bony prominences. Patients develop multiple subcutaneous skin tumors and gingival hypertrophy. Hyaline deposits in multiple organs, recurrent infections and intractable diarrhea often lead to death within the first 2 years of life. Surviving children may suffer from severely reduced mobility due to joint contractures.</p> <p>Defects in ANT XR2 are the cause of juvenile hyaline fibromatosis (JHF) [MIM:228600]. JHF is an autosomal recessive syndrome that is similar to ISH but takes a milder course. It is characterized by hyaline deposition in the dermis, multiple subcutaneous skin tumors and gingival hypertrophy, followed by progressive joint contractions, osteopenia and osteoporosis that may lead to a severe limitation of mobility.</p>
Sequence similarities	<p>Belongs to the ATR family.</p> <p>Contains 1 VWFA domain.</p>
Domain	Binding to PA seems to be effected through the VWA domain.
Cellular localization	<p>Secreted; Cell membrane. Expressed at the cell surface and Endoplasmic reticulum membrane.</p> <p>Expressed predominantly within the endoplasmic reticulum and not at the plasma membrane.</p>

Images



15% SDS-PAGE analysis of ab180320 (3µg)

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