



## Rabbit Anti-Site 2 protease antibody

SL19356R

<b>Product Name:</b>	Site 2 protease
<b>Chinese Name:</b>	S2P内肽酶抗体
<b>Alias:</b>	BRESEK; Endopeptidase S2P; IFAP; KFSDX; MBTP2_HUMAN; MBTPS2; Membrane bound transcription factor site 2 protease; membrane-bound transcription factor peptidase, site 2; membrane-bound transcription factor protease, site 2; Membrane-bound transcription factor site-2 protease; S2P; S2P endopeptidase; Site 2 protease; SREBPs intramembrane protease; Sterol regulatory element-binding proteins intramembrane protease.
<b>Organism Species:</b>	Rabbit
<b>Clonality:</b>	Polyclonal
<b>React Species:</b>	Human,Mouse,Rat,Cow,Horse,Sheep,
<b>Applications:</b>	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500 (Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
<b>Molecular weight:</b>	57kDa
<b>Cellular localization:</b>	cytoplasmicThe cell membrane
<b>Form:</b>	Lyophilized or Liquid
<b>Concentration:</b>	1mg/ml
<b>immunogen:</b>	KLH conjugated synthetic peptide derived from human Site 2 protease:1-100/519
<b>Lsotype:</b>	IgG
<b>Purification:</b>	affinity purified by Protein A
<b>Storage Buffer:</b>	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
<b>Storage:</b>	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
<b>PubMed:</b>	<a href="#">PubMed</a>
<b>Product Detail:</b>	This gene encodes a intramembrane zinc metalloprotease, which is essential in development. This protease functions in the signal protein activation involved in sterol

control of transcription and the ER stress response. Mutations in this gene have been associated with ichthyosis follicularis with atrichia and photophobia (IFAP syndrome); IFAP syndrome has been quantitatively linked to a reduction in cholesterol homeostasis and ER stress response.[provided by RefSeq, Aug 2009]

**Function:**

Intramembrane proteolysis of sterol-regulatory element-binding proteins (SREBPs) within the first transmembrane segment thereby releasing the N-terminal segment with a portion of the transmembrane segment attached. Site-2 cleavage comes after site-1 cleavage which takes place in the luminal loop.

**Subcellular Location:**

Membrane. Cytoplasm.

**Tissue Specificity:**

Expressed in heart, brain, placenta, lung, liver, muscle, kidney and pancreas.

**DISEASE:**

Defects in MBTPS2 are the cause of ichthyosis follicularis-atrichia-photophobia syndrome (IFAPS) [MIM:308205]. A syndrome characterized by a peculiar triad of follicular ichthyosis, total or subtotal atrichia, and photophobia of varying degree. Histopathologically, the epidermal granular layer is generally well-preserved or thickened at the infundibulum. Hair follicles are poorly developed and tend to be surrounded by an inflammatory infiltrate. A subgroup of patients is described with lamellar rather than follicular ichthyosis. Non-consistent features may include growth and psychomotor retardation, aganglionic megacolon, seizures and nail dystrophy. Defects in MBTPS2 are a cause of keratosis follicularis spinulosa decalvans X-linked (KFSDX) [MIM:308800]. A rare disorder affecting the skin and the eye. Affected men show thickening of the skin of the neck, ears, and extremities, especially the palms and soles, loss of eyebrows, eyelashes and beard, thickening of the eyelids with blepharitis and ectropion, and corneal degeneration.

**Similarity:**

Belongs to the peptidase M50A family.

**SWISS:**

O43462

**Gene ID:**

51360

**Database links:**

[Entrez Gene: 51360](#) Human

[Entrez Gene: 270669](#) Mouse

[Entrez Gene: 302705](#) Rat

[Omim: 300294](#) Human

[SwissProt: O43462](#) Human

[SwissProt: Q8CHX6](#) Mouse

[Unigene: 443490](#) Human

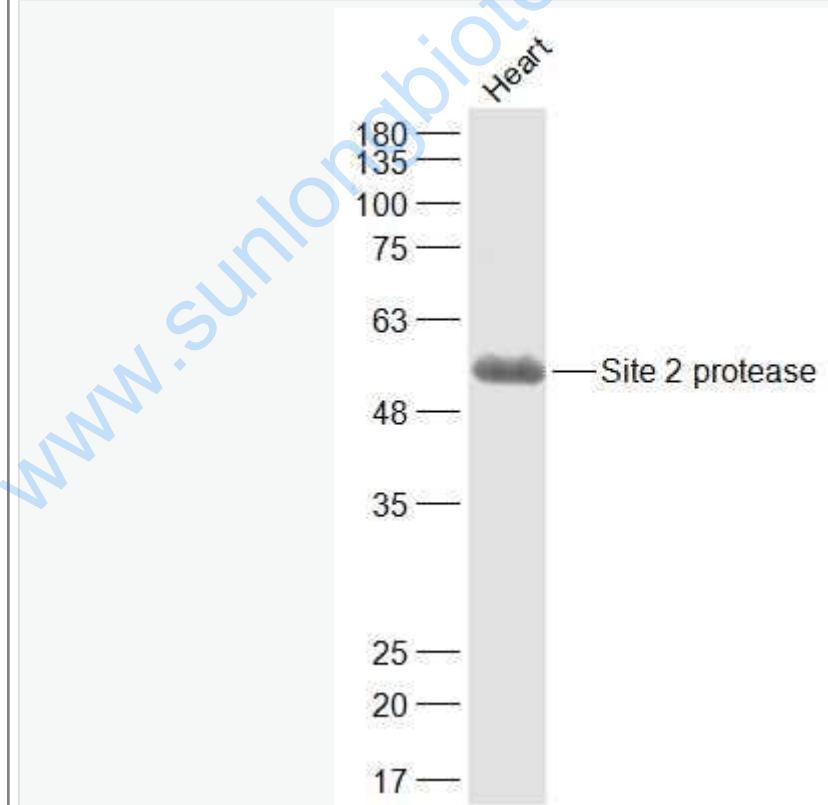
[Unigene: 37577](#) Mouse

[Unigene: 212224](#) Rat

**Important Note:**

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

**Picture:**



Sample:

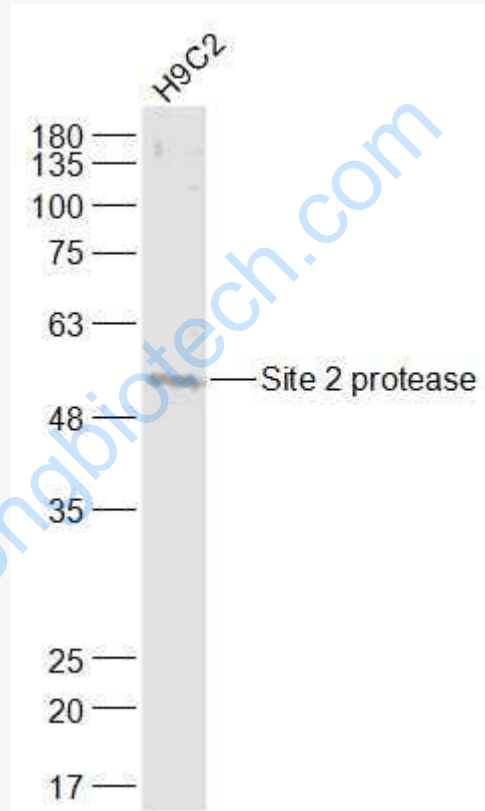
Heart (Mouse) Lysate at 40 ug

Primary: Anti-Site 2 protease? (SL19356R) at 1/1000 dilution

Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

Predicted band size: 57 kD

Observed band size: 57 kD



Sample:

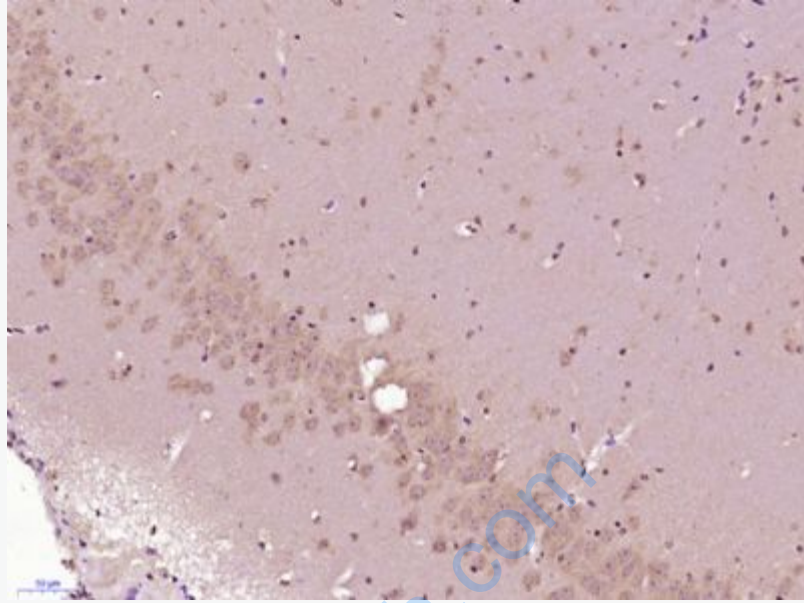
H9C2(Rat) Cell Lysate at 30 ug

Primary: Anti-Site 2 protease? (SL19356R) at 1/1000 dilution

Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

Predicted band size: 57 kD

Observed band size: 57 kD



Paraformaldehyde-fixed, paraffin embedded (Mouse brain); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Site 2 protease) Polyclonal Antibody, Unconjugated (SL19356R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructions and DAB staining.