

Rabbit Anti-PAPSS2 antibody

SL9030R

Product Name:	PAPSS2
Chinese Name:	腺苷酸硫酸激酶2抗体
Alias:	ATPSK2; PAPS synthetase 2; PAPSS 2; SK2; Sulfurylase kinase 2; 3'
	phosphoadenosine 5' phosphosulfate synthase 2.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human, Mouse, Rat, Dog, Cow, Horse, Rabbit,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=1:50-
	200 (Paraffin sections need antigen repair)
	not yet tested in other applications.
	optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	68kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	lmg/ml
immunogen:	KLH conjugated synthetic peptide derived from human PAPSS2:351-450/614
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	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.
PubMed:	<u>PubMed</u>
Product Detail:	PAPSS2 is one of the two PAPS synthetases. Three-prime-phosphoadenosine 5-prime-
	phosphosulfate (PAPS) is the sulfate donor cosubstrate for all sulfotransferase (SULT)
	enzymes. SULTs catalyze the sulfate conjugation of many endogenous and exogenous
	compounds, including drugs and other xenobiotics. In humans, PAPS is synthesized
	from adenosine 5-prime triphosphate (ATP) and inorganic sulfate by 2 isoforms,
	PAPSS1 and PAPSS2.

Bifunctional 3'-phosphoadenosine 5'-phosphosulfate synthetases (PAPS synthetase or PAPSS), also designated sulfurylase kinase (SK), are important for sulfate assimilation in the sulfur metabolism pathway. PAPPS proteins are bifunctional enzymes with APS kinase and ATP sulfurylase activity, which mediate two steps in the sulfate activation pathway. The PAPSS proteins belong to the APS kinase family and to the sulfate adenylyltransferase family of proteins. In mammals, PAPSS proteins are the sole source of sulfate. During postnatal growth, PAPSS proteins may play a role in skeletogenesis. Defects in the PAPSS2 gene can cause the Pakistani type of spondyloepimetaphyseal dysplasia (SEMD), an autosomal recessive form of SEMD characterized by short, bowed limbs, enlarged knee joints and mild brachydactyly.

Function:

Bifunctional enzyme with both ATP sulfurylase and APS kinase activity, which mediates two steps in the sulfate activation pathway. The first step is the transfer of a sulfate group to ATP to yield adenosine 5'-phosphosulfate (APS), and the second step is the transfer of a phosphate group from ATP to APS yielding 3'-phosphoadenylylsulfate (PAPS: activated sulfate donor used by sulfotransferase). In mammals, PAPS is the sole source of sulfate; APS appears to be only an intermediate in the sulfate-activation pathway. May have a important role in skeletogenesis during.

Tissue Specificity:

Expressed in cartilage and adrenal gland.

DISEASE:

Defects in PAPSS2 are the cause of spondyloepimetaphyseal dysplasia Pakistani type (SEMD-PA) [MIM:612847]. A bone disease characterized by epiphyseal dysplasia with mild metaphyseal abnormalities. Clinical features include short stature evidenced at birth, short and bowed lower limbs, mild brachydactyly, kyphoscoliosis, abnormal gait, enlarged knee joints. Some patients may manifest premature pubarche and hyperandrogenism associated with skeletal dysplasia and short stature.

Similarity:

In the N-terminal section; belongs to the APS kinase family.

In the C-terminal section; belongs to the sulfate adenylyltransferase family.

SWISS:

O95340

Gene ID:

9060

Database links:

Entrez Gene: 9060 Human

Entrez Gene: 23972 Mouse

Entrez Gene: 294103 Rat

Omim: 603005 Human

SwissProt: O95340 Human

SwissProt: O88428 Mouse

Unigene: 524491 Human

Important Note:

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