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PAPSS1 Rabbit pAb

Catalog # AP97988

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	O95340
Other Accession	O95340
Reactivity	Mouse
Predicted	Rat, Human, Rabbit, Dog, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	69501
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human PAPSS1
Epitope Specificity	155-260/624
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	In the N-terminal section; belongs to the APS kinase family.In the C-terminal section; belongs to the sulfate adenylyltransferase family.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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.

Additional Information

Gene ID	9060
Other Names	ATPSK2; BCYM4; SK2; 1810018P12Rik; AtpsU2; bm; PAPS2_HUMAN; PAPSS2; PAPS synthase 2; PAPSS 2; Sulfurylase kinase 2 (SK 2 | SK2); PAPS2_MOUSE; 3'-phosphoadenosine 5'-phosphosulfate synthase 1; sulfate adenylyltransferase; adenylyl-sulfate kinase
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name	PAPSS2
Synonyms	ATPSK2
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, the activated sulfate donor used by sulfotransferases (PubMed:11773860, PubMed:19474428, PubMed:23824674, PubMed:25594860). In mammals, PAPS is the sole source of sulfate while APS appears to only be an intermediate in the sulfate-activation pathway (PubMed:11773860, PubMed:19474428, PubMed:23824674, PubMed:25594860). Plays indirectly an important role in skeletogenesis during postnatal growth (PubMed:9771708).
Tissue Location	Expressed in cartilage and adrenal gland.

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