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GNAS Rabbit pAb

Catalog # AP98514

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	P63092
Other Accession	P63092
Reactivity	Rat, Human
Predicted	Pig, Mouse, Rabbit, Chicken, Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	45665
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human GNAS
Epitope Specificity	901-1037/1037
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the G-alpha family. G(s) subfamily. membrane protein.
SUBUNIT	G proteins are composed of 3 units; alpha, beta and gamma. The alpha chain contains the guanine nucleotide binding site. Interacts through its N-terminal region with ALEX which is produced from the same locus in a different open reading frame. This interaction may inhibit its adenylyl cyclase-stimulating activity (By similarity).
DISEASE	Defects in GNAS are the cause of GNAS hyperfunction (GNASHYP) [MIM:139320]. This condition is characterized by increased trauma-related bleeding tendency, prolonged bleeding time, brachydactyly and mental retardation. Both the XLas isoforms and the ALEX protein are mutated which strongly reduces the interaction between them and this may allow unimpeded activation of the XLas isoforms. Defects in GNAS are a cause of ACTH-independent macronodular adrenal hyperplasia (AIMAH) [MIM:219080]; also known as adrenal Cushing syndrome due to AIMAH. A rare adrenal defect characterized by multiple, bilateral, non-pigmented, benign, adrenocortical nodules. It results in excessive production of cortisol leading to ACTH-independent Cushing syndrome. Clinical manifestations of Cushing syndrome include facial and trunkal obesity, abdominal striae, muscular weakness, osteoporosis, arterial hypertension, diabetes. Genetic variations in GNAS are the cause of pseudohypoparathyroidism type 1B (PHP1B) [MIM:603233]. PHP1B is characterized by parathyroid hormone (PTH)-resistant hypocalcemia and hyperphosphatemia. Patients affected with PHP1B have normal activity of the product of GNAS, lack developmental defects characteristic of AHO, and typically show no other endocrine abnormalities besides resistance to PTH. Note=Most affected individuals have defects in methylation of the gene. In some cases microdeletions involving the STX16 appear to cause loss of methylation at exon A/B of GNAS, resulting in PHP1B. Paternal uniparental isodisomy have also been observed. Defects in GNAS are the cause of pseudohypoparathyroidism type 1C (PHP1C) [MIM:612462]. It is a disorder characterized by end-organ resistance to parathyroid hormone, hypocalcemia and hyperphosphatemia. It is commonly

	associated with Albright hereditary osteodystrophy whose features are short stature, obesity, round facies, short metacarpals and ectopic calcification.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Guanine nucleotide-binding proteins (G proteins) are involved as modulators or transducers in various transmembrane signaling systems. The Gs protein is involved in hormonal regulation of adenylate cyclase: it activates the cyclase in response to beta-adrenergic stimuli.

Additional Information

Gene ID	2778
Other Names	GNAS1; NESP55; SCG6; AHO; AIMAH1; C20orf45; GPSA; GSA; GSP; NESP; PITA3; POH; SgVI; 5530400H20Rik; A930027G11Rik; C130027O20Rik; Galphas; Gnasxl; Gs-alpha; Nespl; Oed-Sml; Oedsml; P1; P2; P3; PHP1A; PHP1B; PORGSA1; ALEX; G-alpha-8; Gnpas; GNAS2_BOVIN; GNAS; Adenylate cyclase-stimulating G alpha protein; 3.6.5.-; GNAS_CANLF; GNAS2_HUMAN; ALEX_HUMAN; Alternative gene product encoded by XL-exon; GNAS1_HUMAN; Extra large alphas protein (XLalphas); GNAS2_MOUSE; GNAS1_MOUSE; GNAS_PIG; GNAS2_RAT; GNAS1_RAT; GNAS complex locus; guanine nucleotide binding protein (G protein), alpha stimulating activity polypeptide 1; secretogranin VI; G protein subunit alpha S
Dilution	WB=1:500-2000,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	GNAS
Synonyms	GNAS1, GSP
Function	Guanine nucleotide-binding proteins (G proteins) function as transducers in numerous signaling pathways controlled by G protein- coupled receptors (GPCRs) (PubMed:12391161, PubMed:17110384, PubMed:21488135, PubMed:26206488, PubMed:8702665, PubMed:10200251). The alpha chain contains the guanine nucleotide binding site and alternates between an active, GTP-bound state and an inactive, GDP-bound state (PubMed:12391161, PubMed:17110384, PubMed:10200251). Signaling by an activated GPCR promotes GDP release and GTP binding (PubMed:12391161, PubMed:17110384, PubMed:10200251). The alpha subunit has a low GTPase activity that converts bound GTP to GDP, thereby terminating the signal (PubMed:12391161, PubMed:17110384, PubMed:10200251). Both GDP release and GTP hydrolysis are modulated by numerous regulatory proteins (PubMed:12391161, PubMed:17110384, PubMed:10200251). Signaling involves the activation of adenylyl cyclases, resulting in increased levels of the signaling molecule cAMP (PubMed:17110384, PubMed:26206488, PubMed:26206488, PubMed:8702665). Functions downstream of beta-adrenergic receptors (PubMed:21488135). Stimulates the Ras signaling pathway via RAPGEF2 (PubMed:12391161).
Cellular Location	Cell membrane; Peripheral membrane protein; Cytoplasmic side.

Note=Associated to the plasma membrane via lipid-anchor.

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