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Rabbit Anti-NF1 antibody

SL4140R

Product Name:	NF1
Chinese Name:	1型神经纤维瘤抗体
Alias:	Neurofibromin 1; DKFZp686J1293; FLJ21220; Neurofibromatosis Noonan syndrome; Neurofibromatosis related protein NF 1; Neurofibromatosis related protein NF1; neurofibromatosis type I; Neurofibromatosis-related protein NF-1; Neurofibromin 1; Neurofibromin truncated; Neurofibromin1; NF 1; NF; NF1; NF1_HUMAN; NFNS; Type 1 Neurofibromatosis; von Recklinghausen disease neurofibromin; von Recklinghausen disease related protein VRNF; VRNF; WATS; Watson disease related protein WSS; Watson syndrome; WSS.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Horse,Rabbit,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	147/319kDa
Cellular localization:	The nucleus
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Neurofibromin 1:2701-2839/2839
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:	PubMed

Product Detail:	This gene product appears to function as a negative regulator of the ras signal transduction pathway. Mutations in this gene have been linked to neurofibromatosis type 1, juvenile myelomonocytic leukemia and Watson syndrome. The mRNA for this gene is subject to RNA editing (CGA>UGA->Arg1306Term) resulting in premature translation termination. Alternatively spliced transcript variants encoding different isoforms have also been described for this gene. [provided by RefSeq, Jul 2008]. **Function:** Stimulates the GTPase activity of Ras. NF1 shows greater affinity for Ras GAP, but lower specific activity. May be a regulator of Ras activity. **DISEASE:** Neurofibromatosis 1 (NF1) [MIM:162200]: A disease characterized by patches of skin pigmentation (cafe-au-lait spots), Lisch nodules of the iris, tumors in the peripheral nervous system and fibromatous skin tumors. Individuals with the disorder have increased susceptibility to the development of benign and malignant tumors. Note=The disease is caused by mutations affecting the gene represented in this entry. Leukemia, juvenile myelomonocytic (JMML) [MIM:607785]: An aggressive pediatric myelodysplastic syndrome/myeloproliferative disorder characterized by malignant transformation in the hematopoietic stem cell compartment with proliferation of differentiated progeny. Patients have splenomegaly, enlarged lymph nodes, rashes, and hemorrhages. Note=The disease is caused by mutations affecting the gene represented in this entry. Watson syndrome (WS) [MIM:193520]: A syndrome characterized by the presence of pulmonary stenosis, cafe-au-lait spots, and mental retardation. It is considered as an atypical form of neurofibromatosis. Note=The disease is caused by mutations affecting the gene represented in this entry. Familial spinal neurofibromatosis (FSNF) [MIM:162210]: Considered to be an alternative form of neurofibromatosis, showing multiple spinal tumors. Note=The disease is caused by mutations affecting the gene represented in this entry. Neurofibromatosis-Noonan syndrome (NFNS) [MIM:601321]: Characterized by manifestations of both NF1 and Noonan syndrome (NS). NS is a disorder characterized by dysmorphic facial features, short stature, hypertelorism, cardiac anomalies, deafness, motor delay, and a bleeding diathesis. Note=The disease is caused by mutations affecting the gene represented in this entry. Colorectal cancer (CRC) [MIM:114500]: A complex disease characterized by malignant lesions arising from the inner wall of the large intestine (the colon) and the rectum. Genetic alterations are often associated with progression from premalignant lesion (adenoma) to invasive adenocarcinoma. Risk factors for cancer of the colon and rectum include colon polyps, long-standing ulcerative colitis, and genetic family history. Note=The gene represented in this entry may be involved in disease pathogenesis. **Similarity:** Contains 1 CRAL-TRIO domain. Contains 1 Ras-GAP domain.

SWISS:
P21359

Gene ID:
4763

Database links:

Entrez Gene: 4763Human

Entrez Gene: 18015Mouse

Entrez Gene: 24592Rat

Omim: 613113Human

SwissProt: P21359Human

SwissProt: Q04690Mouse

SwissProt: P97526Rat

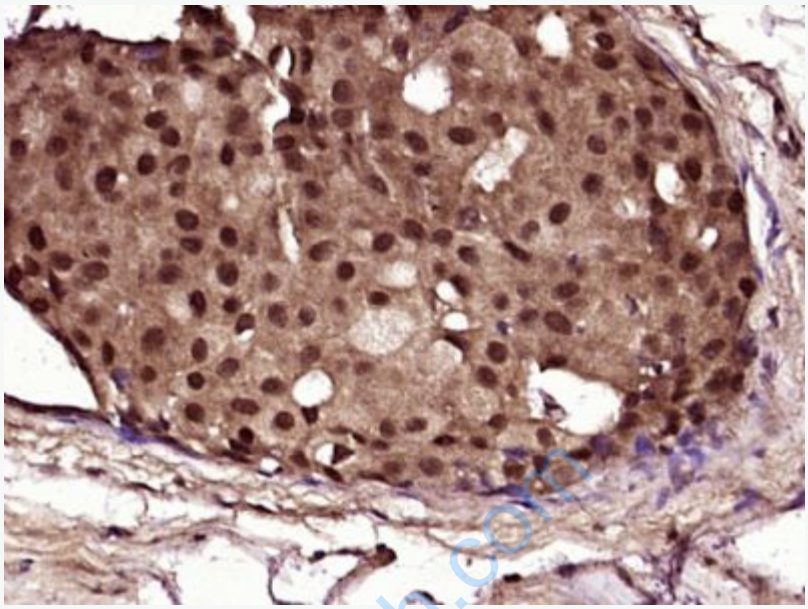
Unigene: 113577Human

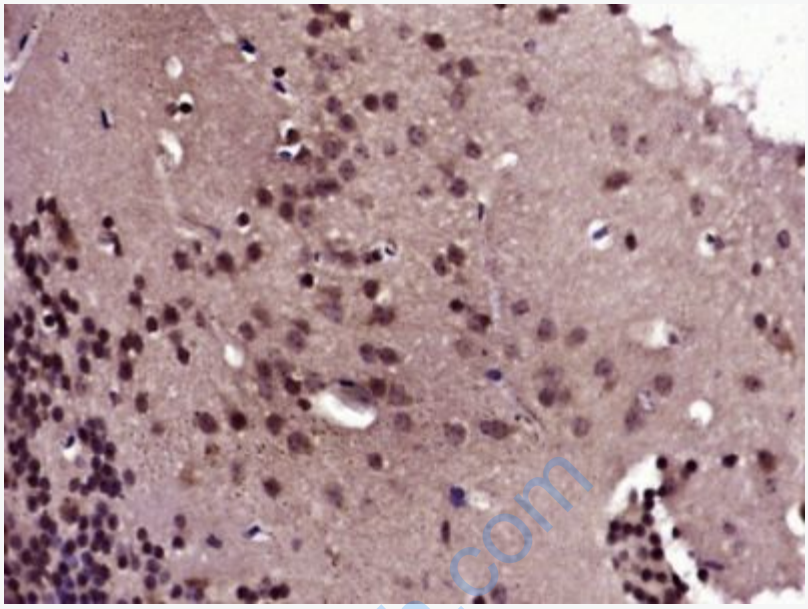
Unigene: 255596Mouse

Unigene: 10686Rat

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

神经纤维素蛋白首先发现于神经细胞，是一种Tumour抑制蛋白，通过调控Ras基因控制异常细胞生长，并且在cAMP信号传导通路中起调节作用.
神经纤维瘤Ⅰ型(neurofibromatosis type 1,NF1)是一种由内分泌紊乱引起的神经纤维瘤,属于常染色体显性遗传病，其发病率为1/3500，主要表现为咖啡斑、神经纤维瘤、Lisch结节(虹膜错构瘤)等。每3，500个新生儿中就有一个是神经纤维细胞瘤I型患者，其临床表现为表皮或皮下多发性神经纤维瘤，良性多于恶性，常沿神经干分布。有时，神经纤维瘤会长大，或者发展到脑和脊髓，大约有一半以上患者智力低下。

Picture:	 Paraformaldehyde-fixed, paraffin embedded (Human breast carcinoma); 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 (NF1) Polyclonal Antibody, Unconjugated (SL4140R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.



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 (NF1) Polyclonal Antibody, Unconjugated (SL4140R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.