骨硬化病相关跨膜蛋白 1 抗体

产品货号： mlR8506

英文名称： OSTM1

中文名称： 骨硬化病相关跨膜蛋白 1 抗体

别　名： GAIP-interacting protein N terminus; GIPN; GL antibody Grey lethal osteopetrosis; HSPC019; OPTB5; Osteopetrosis-associated transmembrane protein 1; Ostm1; OSTM1_HUMAN.

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Pig,

产品应用： WB=1:500-2000 ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 Flow-Cyt=1μg/Test IF=1:50-200 （石蜡切片需做抗原修复）
not yet tested in other applications.
optimal dilutions/concentrations should be determined by the end user.

分 子 量： 33kDa

细胞定位： 细胞膜

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human OSTM1:21-120/334 <Extracellular>

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： 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

产品介绍： OSTM1 (osteopetrosis associated transmembrane protein 1), also known as gl (gray-lethal) or HSPC019, is a 338 amino acid single-pass type I membrane protein that is expressed primarily in osteoclasts and melanocytes as well as brain, kidney and spleen. Bone autosomal recessive osteopetrosis (ARO) is the most severe form of hereditary bone disease whose cellular basis is in the osteoclast and is characterized by abnormally dense bone, due to defective resorption of immature bone. ARO is suggested to be caused by mutations in the OSTM1 gene. The disorder occurs in two forms: a severe autosomal recessive form occurring in utero, infancy, or childhood, and a benign autosomal dominant form occurring in adolescence or adulthood. Defects in the OSTM1 gene are also the cause of the spontaneous gl mutant, which is responsible for a coat color defect in mice.

Function:

Required for osteoclast and melanocyte maturation and function (By similarity).

Subcellular Location:

Membrane; Single-pass type I membrane protein.

DISEASE:

Defects in OSTM1 are the cause of osteopetrosis autosomal recessive type 5 (OPTB5) [MIM:259720]; also called infantile malignant osteopetrosis 3. Osteopetrosis is a rare genetic disease characterized by abnormally dense bone, due to defective resorption of immature bone. The disorder occurs in two forms: a severe autosomal recessive form occurring in utero, infancy, or childhood, and a benign autosomal dominant form occurring in adolescence or adulthood. OPTB5 patients manifest primary central nervous system involvement in addition to

the classical stigmata of severe bone sclerosis, growth failure, anemia, thrombocytopenia and visual impairment with optic atrophy.

SWISS:

Q86WC4

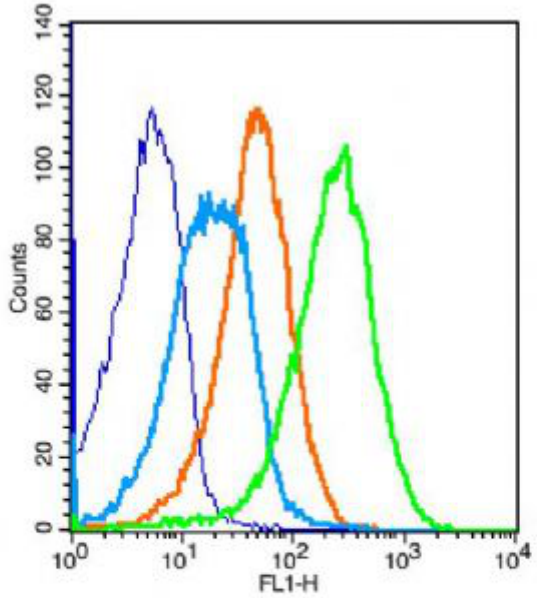
Gene ID:

28962

Important Note:

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

产品图片



Key	Name	Parameter	Gate
	(mo)Splenocyte-blank.049	FL1-H	G1
	bs-0295P(CST)-(FITC)#1E624C.051	FL1-H	G1
	bs-0295G-FITC(CST)-(#1E624A.050	FL1-H	G1
	bs-8506R-(FITC)-(mo)Sple-1.058	FL1-H	G1