

OSTM1 (4H1): sc-293366



The Power to Question

BACKGROUND

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.

REFERENCES

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3. Ramirez, A., et al. 2004. Identification of a novel mutation in the coding region of the grey-lethal gene OSTM1 in human malignant infantile osteopetrosis. *Hum. Mutat.* 23: 471-476.
4. Blin-Wakkach, C., et al. 2004. Osteopetrosis, from mouse to man. *Med. Sci.* 20: 61-67.
5. Pangrazio, A., et al. 2006. Mutations in OSTM1 (grey lethal) define a particularly severe form of autosomal recessive osteopetrosis with neural involvement. *J. Bone Miner. Res.* 21: 1098-1105.
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8. Pata, M., et al. 2008. OSTM1 bone defect reveals an intercellular hematopoietic crosstalk. *J. Biol. Chem.* 283: 30522-30530.
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CHROMOSOMAL LOCATION

Genetic locus: OSTM1 (human) mapping to 6q21; Ostm1 (mouse) mapping to 10 B2.

SOURCE

OSTM1 (4H1) is a mouse monoclonal antibody raised against amino acids 183-282 of OSTM1 of human origin.

PRODUCT

Each vial contains 100 μ g IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

OSTM1 (4H1) is recommended for detection of OSTM1 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for OSTM1 siRNA (h): sc-95126, OSTM1 siRNA (m): sc-151337, OSTM1 shRNA Plasmid (h): sc-95126-SH, OSTM1 shRNA Plasmid (m): sc-151337-SH, OSTM1 shRNA (h) Lentiviral Particles: sc-95126-V and OSTM1 shRNA (m) Lentiviral Particles: sc-151337-V.

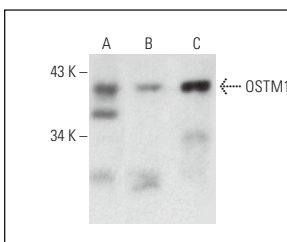
Molecular Weight of OSTM1: 37 kDa.

Positive Controls: NRK whole cell lysate: sc-364197, LADMAC whole cell lysate: sc-364189 or A-375 cell lysate: sc-3811.

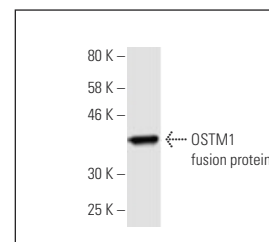
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml).

DATA



OSTM1 (4H1): sc-293366. Western blot analysis of OSTM1 expression in A-375 (A), LADMAC (B) and NRK (C) whole cell lysates.



OSTM1 (4H1): sc-293366. Western blot analysis of human recombinant OSTM1 fusion protein.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.