

# OSTM1 (E-12): sc-168856

## 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

1. Rajapurohitam, V., et al. 2001. The mouse osteopetrotic grey-lethal mutation induces a defect in osteoclast maturation/function. *Bone* 28: 513-523.
2. Chalhoub, N., et al. 2003. Grey-lethal mutation induces severe malignant autosomal recessive osteopetrosis in mouse and human. *Nat. Med.* 9: 399-406.
3. Ramírez, 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.
6. Lange, P.F., et al. 2006. CLC-7 requires OSTM1 as a  $\beta$ -subunit to support bone resorption and lysosomal function. *Nature* 440: 220-223.

## CHROMOSOMAL LOCATION

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

## SOURCE

OSTM1 (E-12) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an extracellular domain of OSTM1 of human origin.

## PRODUCT

Each vial contains 200  $\mu$ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-168856 P, (100  $\mu$ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## STORAGE

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

## APPLICATIONS

OSTM1 (E-12) 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  $\mu$ g per 100-500  $\mu$ g of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

OSTM1 (E-12) is also recommended for detection of OSTM1 in additional species, including canine and porcine.

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.

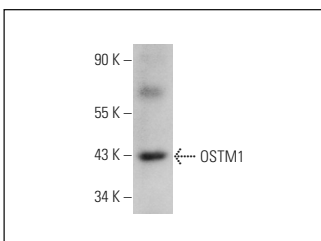
Molecular Weight of OSTM1: 37 kDa.

Positive Controls: mouse heart extract: sc-2254.

## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

## DATA



OSTM1 (E-12): sc-168856. Western blot analysis of OSTM1 expression in mouse heart tissue extract.

## RESEARCH USE

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

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Try **OSTM1 (4H1): sc-293366**, our highly recommended monoclonal alternative to OSTM1 (E-12).