SANTA CRUZ BIOTECHNOLOGY, INC.

Twinkle (H-300): sc-134915

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

Twinkle, also known as PEO1 (progressive external ophthalmoplegia 1 protein), PEOA3, SANDO or TWINL, is a mitochondrial protein that functions as a 5'-3' nucleotide-dependent DNA helicase. Co-localized with mtDNA (mitochondrial DNA) in mitochondrial nucleoids, Twinkle is important in the metabolism and maintenance of mtDNA, playing a crucial role in the regulation of mtDNA copy numbers. Twinkle is expressed at high levels in testis, pancreas and skeletal muscle and exists as three isoforms due to alternative splicing events. Defects in the gene encoding Twinkle are the cause of two conditions: progressive external ophthalmoplegia with mitochondrial DNA deletions autosomal dominant 3 (PEOA3) and sensory ataxic neuropathy dysarthria and ophthalmoparesis (SANDO). PEOA3 is characterized by ptosis and weak muscles, while SANDO is characterized by ophthalmoparesis, dysarthria and sensory ataxic neuropathies.

CHROMOSOMAL LOCATION

Genetic locus: C10orf2 (human) mapping to 10q24.31; Peo1 (mouse) mapping to 19 C3.

SOURCE

Twinkle (H-300) is a rabbit polyclonal antibody raised against amino acids 241-540 mapping within an internal region of Twinkle of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

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.

APPLICATIONS

Twinkle (H-300) is recommended for detection of Twinkle of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Twinkle (H-300) is also recommended for detection of Twinkle in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for Twinkle siRNA (h): sc-63177, Twinkle siRNA (m): sc-63178, Twinkle shRNA Plasmid (h): sc-63177-SH, Twinkle shRNA Plasmid (m): sc-63178-SH, Twinkle shRNA (h) Lentiviral Particles: sc-63177-V and Twinkle shRNA (m) Lentiviral Particles: sc-63178-V.

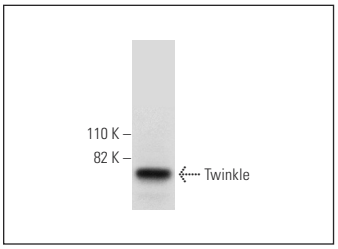
Molecular Weight of Twinkle: 77 kDa.

Positive Controls: human skeletal muscle extract: sc-363776.

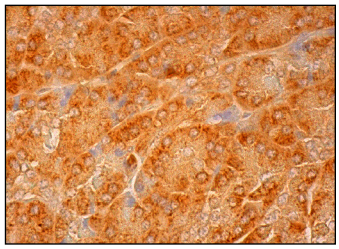
RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941. 4) Immunohistochemistry: use ImmunoCruz™: sc-2051 or ABC: sc-2018 rabbit IgG Staining Systems.

DATA



Twinkle (H-300): sc-134915. Western blot analysis of Twinkle expression in human skeletal muscle tissue extract.



Twinkle (H-300): sc-134915. Immunoperoxidase staining of formalin fixed, paraffin-embedded human pancreas tissue showing cytoplasmic staining of exocrine glandular cells.

PROTOCOLS

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

MONOS Satisfaction Guaranteed

Try **Twinkle (1C5): sc-293368**, our highly recommended monoclonal alternative to Twinkle (H-300).