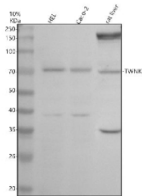


TWNK Antibody / Twinkle (FY12836)

Catalog No.	Formulation	Size
FY12836	Adding 0.2 ml of distilled water will yield a concentration of 500 ug/ml	100 ug

[Bulk quote request](#)

Availability	1-2 days
Species Reactivity	Human, Mouse, Rat
Format	Lyophilized
Host	Rabbit
Clonality	Polyclonal (rabbit origin)
Isotype	Rabbit IgG
Purity	Immunogen affinity purified
Buffer	Each vial contains 4 mg Trehalose, 0.9 mg NaCl, 0.2 mg Na ₂ HPO ₄ .
UniProt	Q96RR1
Applications	Western Blot : 0.25-0.5ug/ml ELISA : 0.1-0.5ug/ml
Limitations	This TWNK antibody is available for research use only.



Western blot analysis of Twinkle/TWNK using anti-TWNK antibody. Electrophoresis was performed on a 10% SDS-PAGE gel at 80V (Stacking gel) / 120V (Resolving gel) for 2 hours. Lane 1: human HEL whole cell lysates, Lane 2: human Caco-2 whole cell lysates, Lane 3: rat liver tissue lysates. After electrophoresis, proteins were transferred to a nitrocellulose membrane at 150 mA for 50-90 minutes. Blocked the membrane with 5% non-fat milk/TBS for 1.5 hour at RT. The membrane was incubated with rabbit anti-TWNK antibody at 0.5 ug/ml overnight at 4°C, then washed with TBS-0.1% Tween 3 times with 5 minutes each and probed with a goat anti-rabbit IgG-HRP secondary antibody at a dilution of 1:5000 for 1.5 hour at RT. The signal was developed using an ECL Plus Western Blotting Substrate. The expected molecular weight of Twinkle/TWNK is at ~77 kDa.

Description

TWNK antibody detects Twinkle, a mitochondrial DNA helicase essential for mtDNA replication and maintenance. Encoded by the TWNK gene on chromosome 10q24.31, this nuclear-encoded protein is imported into mitochondria, where it unwinds double-stranded DNA to facilitate replication of the mitochondrial genome. Twinkle is a member of the

replicative helicase family related to bacteriophage T7 gene 4 helicase-primase, and it plays a vital role in mtDNA stability and copy number control.

Structurally, Twinkle forms a hexameric ring that encircles DNA and uses ATP hydrolysis to unwind duplex strands at replication forks. It cooperates with mitochondrial DNA polymerase gamma (POLG) and mitochondrial single-stranded DNA-binding protein (mtSSB) to sustain replication fidelity and genome integrity. Twinkle localizes to mitochondrial nucleoids, the structures where replication and transcription occur.

The TWNK antibody is widely used in mitochondrial biology, genetics, and metabolic research to study mtDNA replication, maintenance, and disorders of mitochondrial dysfunction. Western blot analysis detects a 77 kilodalton band corresponding to Twinkle, while immunofluorescence reveals mitochondrial localization with a punctate pattern. This antibody supports investigations into mechanisms of mtDNA propagation and repair.

Mutations in TWNK cause autosomal dominant progressive external ophthalmoplegia (PEO) and mitochondrial DNA depletion syndromes, leading to neuromuscular and metabolic disorders. Dysfunctional Twinkle results in replication stalling, deletions, and depletion of mitochondrial DNA. The TWNK antibody provides a reliable tool for studying mitochondrial replication machinery, genome stability, and related diseases. NSJ Bioreagents validates this antibody for western blotting, immunohistochemistry, and immunofluorescence, ensuring accurate detection for mitochondrial research.

Application Notes

Optimal dilution of the TWNK antibody should be determined by the researcher.

Immunogen

E.coli-derived human Twinkle/TWINK recombinant protein (Position: R39-K635) was used as the immunogen for the TWNK antibody.

Storage

After reconstitution, the TWNK antibody can be stored for up to one month at 4°C. For long-term, aliquot and store at -20°C. Avoid repeated freezing and thawing.