



SBDS (h): 293T Lysate: sc-117277

BACKGROUND

The 249 amino acid Shwachman-Bodian-Diamond syndrome (SBDS) protein belongs to the UPF0023 family. SBDS is widely expressed and may be involved in RNA metabolism. SBDS contains a C-terminal domain, a central domain and an N-terminal domain. The C-terminal domain has a ferredoxin-like fold and is structurally homologous with known RNA-binding domains. The central domain contains a three-helical bundle. The N-terminal domain consists of a three-dimensional α/β fold and is the most frequent target of disease-linked mutations. Mutations in the SBDS gene cause Shwachman-Diamond syndrome (SDS), an autosomal recessive marrow failure disorder marked by hematologic dysfunction, skeletal abnormalities and pancreatic exocrine insufficiency. SDS is also characterized by an increased risk of leukemia and myelodysplasia in as many as one third of affected individuals.

REFERENCES

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CHROMOSOMAL LOCATION

Genetic locus: SBDS (human) mapping to 7q11.21.

PRODUCT

SBDS (h): 293T Lysate represents a lysate of human SBDS transfected 293T cells and is provided as 100 μ g protein in 200 μ l SDS-PAGE buffer.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

APPLICATIONS

SBDS (h): 293T Lysate is suitable as a Western Blotting positive control for human reactive SBDS antibodies. Recommended use: 10-20 μ l per lane.

Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

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.