

Produktinformation



Forschungsprodukte & Biochemikalien



Zellkultur & Verbrauchsmaterial



Diagnostik & molekulare Diagnostik



Laborgeräte & Service

Weitere Information auf den folgenden Seiten! See the following pages for more information!



Lieferung & Zahlungsart

siehe unsere Liefer- und Versandbedingungen

Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

SZABO-SCANDIC HandelsgmbH

Quellenstraße 110, A-1100 Wien

T. +43(0)1 489 3961-0

F. +43(0)1 489 3961-7

mail@szabo-scandic.com

www.szabo-scandic.com

linkedin.com/company/szaboscandic in



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



The Power to Question

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

- Dror, Y., et al. 2005. Shwachman-Diamond syndrome. Pediatr. Blood Cancer 45: 892-901.
- Kawakami, T., et al. 2005. Genetic analysis of Shwachman-Diamond syndrome: phenotypic heterogeneity in patients carrying identical SBDS mutations. Tohoku J. Exp. Med. 206: 253-259.
- 3. Kuijpers, T.W., et al. 2005. Hematologic abnormalities in Shwachman Diamond syndrome: lack of genotype-phenotype relationship. Blood 106: 356-361.
- 4. Majeed, F., et al. 2005. Mutation analysis of SBDS in pediatric acute myeloblastic leukemia. Pediatr. Blood Cancer 45: 920-924.
- 5. Nicolis, E., et al. 2005. Identification of novel mutations in patients with Shwachman-Diamond syndrome. Hum. Mutat. 25: 410.
- Savchenko, A., et al. 2005. The Shwachman-Bodian-Diamond syndrome protein family is involved in RNA metabolism. J. Biol. Chem. 280: 19213-19220.
- 7. Shammas, C., et al. 2005. Structural and mutational analysis of the SBDS protein family. Insight into the leukemia-associated Shwachman-Diamond Syndrome. J. Biol. Chem. 280: 19221-19229.
- 8. Austin, K.M., et al. 2005. The Shwachman-Diamond SBDS protein localizes to the nucleolus. Blood 106: 1253-1258.
- Costa, E., et al. 2007. Identification of a novel AluSx-mediated deletion of exon 3 in the SBDS gene in a patient with Shwachman-Diamond syndrome. Blood Cells Mol. Dis. 39: 96-101.

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 µg protein in 200 µl SDS-PAGE buffer.

STORAGE

Store at -20 $^{\circ}$ 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 µl 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.

Santa Cruz Biotechnology, Inc. 1.800.457.3801 831.457.3801 fax 831.457.3801 Europe +00800 4573 8000 49 6221 4503 0 www.scbt.com