

## SBDS (FL-250): sc-134735

### 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  $\alpha/\beta$  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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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.
6. 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.
9. 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; Sbds (mouse) mapping to 5 G1.3.

### SOURCE

SBDS (FL-250) is a rabbit polyclonal antibody raised against amino acids 1-250 representing full length SBDS of human origin.

### RESEARCH USE

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

### PRODUCT

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

### APPLICATIONS

SBDS (FL-250) is recommended for detection of SBDS 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).

SBDS (FL-250) is also recommended for detection of SBDS in additional species, including equine, canine, bovine, porcine and avian.

Suitable for use as control antibody for SBDS siRNA (h): sc-61493, SBDS siRNA (m): sc-61494, SBDS shRNA Plasmid (h): sc-61493-SH, SBDS shRNA Plasmid (m): sc-61494-SH, SBDS shRNA (h) Lentiviral Particles: sc-61493-V and SBDS shRNA (m) Lentiviral Particles: sc-61494-V.

Molecular Weight of SBDS: 31 kDa.

Positive Controls: NIH/3T3 whole cell lysate: sc-2210 or HeLa whole cell lysate: sc-2200.

### 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<sup>™</sup> compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker<sup>™</sup> 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<sup>™</sup> Mounting Medium: sc-24941.

### STORAGE

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

### PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.



Try **SBDS (D-9): sc-271350** or **SBDS (D-6): sc-271600**, our highly recommended monoclonal alternatives to SBDS (FL-250).