

## SBDS (D-6): sc-271600

### 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

1. Dror, Y., et al. 2005. Shwachman-Diamond syndrome. *Pediatr. Blood Cancer* 45: 892-901.
2. 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.
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.

### CHROMOSOMAL LOCATION

Genetic locus: SBDS (human) mapping to 7q11.21; Sbds (mouse) mapping to 5 G1.3.

### SOURCE

SBDS (D-6) is a mouse monoclonal antibody raised against amino acids 1-250 representing full length SBDS of human origin.

### PRODUCT

Each vial contains 200  $\mu$ g IgG<sub>2b</sub> kappa light chain 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.

### APPLICATIONS

SBDS (D-6) is recommended for detection of SBDS of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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 (D-6) is also recommended for detection of SBDS in additional species, including equine, canine, bovine and porcine.

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.

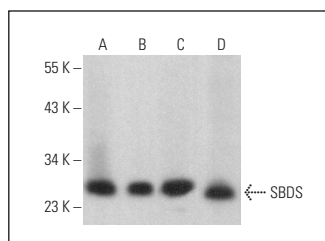
Molecular Weight of SBDS: 31 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, SK-BR-3 cell lysate: sc-2218 or C6 whole cell lysate: sc-364373.

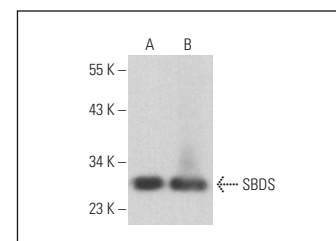
### RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG $\kappa$  BP-HRP: sc-516102 or m-IgG $\kappa$  BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 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 m-IgG $\kappa$  BP-FITC: sc-516140 or m-IgG $\kappa$  BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

### DATA



SBDS (D-6): sc-271600. Western blot analysis of SBDS expression in SK-BR-3 (A), Hep G2 (B) and C6 (C) whole cell lysates and mouse liver tissue extract (D).



SBDS (D-6): sc-271600. Western blot analysis of SBDS expression in NIH/3T3 (A) and T-47D (B) whole cell lysates.

### SELECT PRODUCT CITATIONS

1. Oyarbide, U., et al. 2020. Loss of Sbds in zebrafish leads to neutropenia and pancreas and liver atrophy. *JCI Insight* 5: e134309.

### RESEARCH USE

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