

# SBDS (D-9): sc-271350

## 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. Majeed, F., et al. 2005. Mutation analysis of SBDS in pediatric acute myeloblastic leukemia. *Pediatr. Blood Cancer* 45: 920-924.
3. 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.
4. Kuijpers, T.W., et al. 2005. Hematologic abnormalities in Shwachman Diamond syndrome: lack of genotype-phenotype relationship. *Blood* 106: 356-361.

## CHROMOSOMAL LOCATION

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

## SOURCE

SBDS (D-9) 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>1</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

SBDS (D-9) is available conjugated to agarose (sc-271350 AC), 500  $\mu$ g/0.25 ml agarose in 1 ml, for IP; to HRP (sc-271350 HRP), 200  $\mu$ g/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-271350 PE), fluorescein (sc-271350 FITC), Alexa Fluor<sup>®</sup> 488 (sc-271350 AF488), Alexa Fluor<sup>®</sup> 546 (sc-271350 AF546), Alexa Fluor<sup>®</sup> 594 (sc-271350 AF594) or Alexa Fluor<sup>®</sup> 647 (sc-271350 AF647), 200  $\mu$ g/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor<sup>®</sup> 680 (sc-271350 AF680) or Alexa Fluor<sup>®</sup> 790 (sc-271350 AF790), 200  $\mu$ g/ml, for Near-Infrared (NIR) WB, IF and FCM.

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## RESEARCH USE

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

## APPLICATIONS

SBDS (D-9) 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).

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.

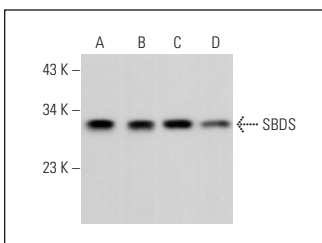
Molecular Weight of SBDS: 31 kDa.

Positive Controls: SK-BR-3 cell lysate: sc-2218, C6 whole cell lysate: sc-364373 or Neuro-2A whole cell lysate: sc-364185.

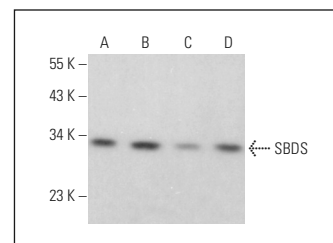
## RECOMMENDED SUPPORT REAGENTS

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<sup>™</sup> Molecular Weight Standards: sc-2035, UltraCruz<sup>®</sup> 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<sup>®</sup> Mounting Medium: sc-24941 or UltraCruz<sup>®</sup> Hard-set Mounting Medium: sc-359850.

## DATA



SBDS (D-9): sc-271350. Western blot analysis of SBDS expression in T-47D (A), SK-BR-3 (B) and C6 (C) whole cell lysates and mouse liver tissue extract (D).



SBDS (D-9): sc-271350. Western blot analysis of SBDS expression in MCF7 (A), Neuro-2A (B), IMR-32 (C) and Caki-1 (D) whole cell lysates.

## SELECT PRODUCT CITATIONS

1. Hao, Q., et al. 2020. Dual regulation of p53 by the ribosome maturation factor SBDS. *Cell Death Dis.* 11: 197.
2. Sera, Y., et al. 2022. SBDS interacts with RNF2 and is degraded through RNF2-dependent ubiquitination. *Biochem. Biophys. Res. Commun.* 598: 119-123.

## STORAGE

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