SANTA CRUZ BIOTECHNOLOGY, INC.

WBSCR11 (D-19): sc-14711

BACKGROUND

Williams-Beuren syndrome (WBS) is a developmental disorder caused by the hemizygous microdeletion on chromosome 7q11.23. WBS is an autosomal dominant genetic condition that is characterized by physical, cognitive and behavioral traits. The physical traits associated with WBS include facial dysmorphology, vascular stenoses, growth deficiencies, dental anomalies and neurologic and musculoskeletal abnormalities. Mild retardation, a weakness in visual-spatial skills, anxiety and a short attention span are typical cognitive and behavioral traits of WBS patients. The WBSCR11 gene is located within the WBS deletion and may contribute to the developmental symptoms found in WBS because of a loss of the encoded transcription factor. WBSCR11 is also designated GRF2IRD1, GTF3, Cream1, and MusTRD1 in human and BEN in mouse, due to slight differences in gene structure. WBSCR11 is expressed in all adult tissues as several variants and has discrete spatial and temporal expression during embryogenesis. The amino terminus of WBSCR11 interacts with transcriptional machinery proteins, while the carboxy terminus has been shown to bind the retinoblastoma protein to possibly regulate the cell cycle.

REFERENCES

1. Morris, C.A., Demsey, S.A., Leonard, C.O., Dilts, C. and Blackburn, B.L. 1988. Natural history of Williams syndrome: physical characteristics. J. Pediatr. 113: 318-326.
2. Pober, B.R. and Dykens, E.M. 1996. Williams syndrome: An overview of medical, cognitive, and behavioral features. Child Adolesc. Psychiatr. Clin. N. Am. 5: 929-943.
3. Lashkari, A., Smith, A.K. and Graham, J.M. 1999. Williams-Beuren syndrome: an update and review for the primary physician. Clin. Pediatr. (Phila) 38: 189-208.
4. Osborne, L.R., Campbell, T., Daradich, A., Scherer, S.W. and Tsui, L.C. 1999. Identification of a putative transcription factor gene (WBSCR11) that is commonly deleted in Williams-Beuren syndrome. Genomics 57: 279-284.
5. Bellugi, U., Lichtenberger, L., Mills, D., Galaburda, A. and Korenberg, J.R. 1999. Bridging cognition, the brain and molecular genetics: evidence from Williams syndrome. Trends Neurosci. 22: 197-207.
6. Tassabehji, M., Carette, M., Wilmot, C., Donnai, D., Read, A.P. and Metcalfe, K. 1999. A transcription factor involved in skeletal muscle gene expression is deleted in patients with Williams syndrome. Eur. J. Hum. Genet. 7: 737-747.
7. Bayarsaihan, D. and Ruddle, F.H. 2000. Isolation and characterization of BEN, a member of the TFII-I family of DNA-binding proteins containing distinct helix-loop-helix domains. Proc. Natl. Acad. Sci. USA 97: 7342-7347.
8. Yan, X., Zhao, X., Qian, M., Guo, N., Gong, X. and Zhu, X. 2000. Characterization and gene structure of a novel retinoblastoma-protein-associated protein similar to the transcription regulator TFII-I. Biochem. J. 3: 749-757.

CHROMOSOMAL LOCATION

Genetic locus: GTF2IRD1 (human) mapping to 7q11.23; Gtf2ird1 (mouse) mapping to 5 G2.

SOURCE

WBSCR11 (D-19) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the N-terminus of WBSCR11 of mouse origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-14711 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

Available as TransCruz reagent for Gel Supershift and ChIP applications, sc-14711 X, 200 µg/0.1 ml.

APPLICATIONS

WBSCR11 (D-19) is recommended for detection of WBSCR11 of mouse, rat and, to a lesser extent, human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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 WBSCR11 siRNA (m): sc-38622, WBSCR11 siRNA (h): sc-38621, WBSCR11 shRNA Plasmid (m): sc-38622-SH, WBSCR11 shRNA Plasmid (h): sc-38621-SH, WBSCR11 shRNA (m) Lentiviral Particles: sc-38622-V and WBSCR11 shRNA (h) Lentiviral Particles: sc-38621-V.

WBSCR11 (D-19) X TransCruz antibody is recommended for Gel Supershift and ChIP applications.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

SELECT PRODUCT CITATIONS

1. Chimge, N.O., et al. 2008. Identification of the TFII-I family target genes in the vertebrate genome. Proc. Natl. Acad. Sci. USA 105: 9006-9010.

STORAGE

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

RESEARCH USE

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