

# WBSCR11 (G-16): sc-14710

## 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 dysmorphism, 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.* 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.

## CHROMOSOMAL LOCATION

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

## SOURCE

WBSCR11 (G-16) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of WBSCR11 of human 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-14714 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

WBSCR11 (G-16) is recommended for detection of WBSCR11 of human and, to a lesser extent, mouse and rat origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg 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).

WBSCR11 (G-16) is also recommended for detection of WBSCR11 in additional species, including equine.

Suitable for use as control antibody for WBSCR11 siRNA (h): sc-38621, WBSCR11 siRNA (m): sc-38622, WBSCR11 shRNA Plasmid (h): sc-38621-SH, WBSCR11 shRNA Plasmid (m): sc-38622-SH, WBSCR11 shRNA (h) Lentiviral Particles: sc-38621-V and WBSCR11 shRNA (m) Lentiviral Particles: sc-38622-V.

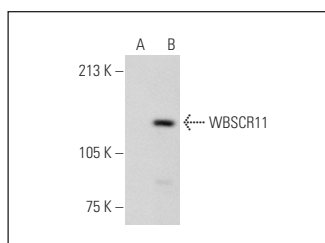
WBSCR11 (G-16) X TransCruz antibody is recommended for Gel Supershift and ChIP applications.

Positive Controls: HeLa whole cell lysate: sc-2200 or WBSCR11 (h): 293 Lysate: sc-113214.

## 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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) 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.

## DATA



WBSCR11 (G-16): sc-14710. Western blot analysis of WBSCR11 expression in non-transfected: sc-117750 (A) and human WBSCR11 transfected: sc-113214 (B) whole cell lysates.

## 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.