

WBSCR18 (P-16): sc-165882

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 WBSCR18 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. WBSCR18 (Williams-Beuren syndrome chromosomal region 18 protein) is a 226 amino acid protein containing one J domain. WBSCR18 is expressed in brain, heart, kidney, liver, lung, spleen, stomach and testis.

REFERENCES

- Morris, C.A., et al. 1988. Natural history of Williams syndrome: physical characteristics. *J. Pediatr.* 113: 318-326.
- Lashkari, A., et al. 1999. Williams-Beuren syndrome: an update and review for the primary physician. *Clin. Pediatr. (Phila)* 38: 189-208.
- Bellugi, U., et al. 1999. Bridging cognition, the brain and molecular genetics: evidence from Williams syndrome. *Trends Neurosci.* 22: 197-207.
- Pober, B.R., et al. 1996. Child Adolesc. Psychiatr. Clin. North Am. 5: 929-943.
- Merla, G., et al. 2002. Identification of additional transcripts in the Williams-Beuren syndrome critical region. *Hum. Genet.* 110: 429-438.
- Ota, T., et al. 2004. Complete sequencing and characterization of 21,243 full-length human cDNAs. *Nat. Genet.* 36: 40-45.
- Hillier, L.W., et al. 2003. The DNA sequence of human chromosome 7. *Nature* 424: 157-164.
- Gerhard, D.S., et al. 2004. The status, quality, and expansion of the NIH full-length cDNA project: the Mammalian Gene Collection (MGC). *Genome Res.* 14: 2121-2127.

CHROMOSOMAL LOCATION

Genetic locus: DNAJC30 (human) mapping to 7q11.23; Dnajc30 (mouse) mapping to 5 G2.

SOURCE

WBSCR18 (P-16) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of WBSCR18 of human origin.

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 or our catalog for detailed protocols and support products.

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-165882 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

WBSCR18 (P-16) is recommended for detection of WBSCR18 of mouse, rat and 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); non cross-reactive with other WBSCR family members.

WBSCR18 (P-16) is also recommended for detection of WBSCR18 in additional species, including canine, bovine and porcine.

Suitable for use as control antibody for WBSCR18 siRNA (h): sc-89463, WBSCR18 siRNA (m): sc-155247, WBSCR18 shRNA Plasmid (h): sc-89463-SH, WBSCR18 shRNA Plasmid (m): sc-155247-SH, WBSCR18 shRNA (h) Lentiviral Particles: sc-89463-V and WBSCR18 shRNA (m) Lentiviral Particles: sc-155247-V.

Molecular Weight of WBSCR18: 26 kDa.

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.

RESEARCH USE

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