

DAX-1 (3G8): sc-293452

BACKGROUND

Adrenal hypoplasia congenita (AHC) is an X-linked disorder characterized by primary adrenal insufficiency. The disorder, which is lethal if untreated, results in adrenal insufficiency early in infancy and is characterized by low serum concentration of glucocorticoids, mineralocorticoids and androgens and failure to respond to ACTH. AHC has been mapped to chromosome Xp21.2 at the same or close to an X-linked locus involved in sex determination, DSS (for dosage-sensitive sex reversal). The gene corresponding to DSS and AHC (designated DAX-1 for DSS-AHC critical region on the X chromosome, gene 1) has been cloned and shown to be deleted in AHC deletion patients and mutated in AHC non-deletion patients. The carboxy terminal 250 amino acids of the DAX-1-encoded protein, DAX-1, exhibits approximately 50% continuous similarity to the ligand-binding domain of the members of the nuclear hormone receptor superfamily while the amino terminal domain contains a putative DNA-binding motif. DAX-1 binds to retinoic acid responsive elements and down regulates retinoic acid receptor-mediated transcriptional activation.

REFERENCES

1. Walker, A.P., et al. 1993. Isolation of the human Xp21 glycerol kinase gene by positional cloning. *Hum. Mol. Genet.* 2: 107-114.
2. Worley, K.C., et al. 1993. Yeast artificial chromosome cloning in the glycerol kinase and adrenal hypoplasia congenita region of Xp21. *Genomics* 16: 407-416.
3. Bardoni, B., et al. 1994. A dosage sensitive locus at chromosome Xp21 is involved in male to female sex reversal. *Nat. Genet.* 7: 497-501.
4. Zanaria, E., et al. 1994. An unusual member of the nuclear hormone receptor superfamily responsible for X-linked adrenal hypoplasia congenita. *Nature* 372: 635-641.
5. Muscatelli, F., et al. 1994. Mutations in the DAX-1 gene give rise to both X-linked adrenal hypoplasia congenita and hypogonadotropic hypogonadism. *Nature* 372: 672-676.

CHROMOSOMAL LOCATION

Genetic locus: NR0B1 (human) mapping to Xp21.2.

SOURCE

DAX-1 (3G8) is a mouse monoclonal antibody raised against amino acids 361-470 representing partial length DAX-1 of human origin.

PRODUCT

Each vial contains 100 µg IgG_{2a} 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.

RESEARCH USE

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

APPLICATIONS

DAX-1 (3G8) is recommended for detection of DAX-1 of human 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)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for DAX-1 siRNA (h): sc-35175, DAX-1 shRNA Plasmid (h): sc-35175-SH and DAX-1 shRNA (h) Lentiviral Particles: sc-35175-V.

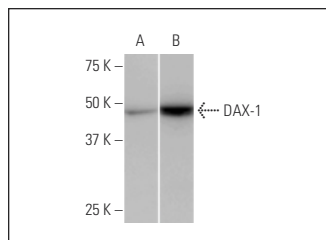
Molecular Weight of DAX-1: 60 kDa.

Positive Controls: DAX-1 transfected 293T whole cell lysates.

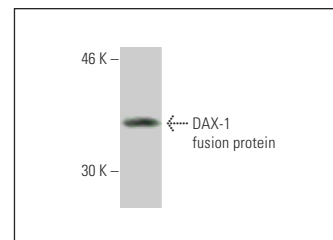
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ 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).

DATA



DAX-1 (3G8): sc-293452. Western blot analysis of DAX-1 expression in non-transfected (A) and DAX-1 transfected (B) 293T whole cell lysates.



DAX-1 (3G8): sc-293452. Western blot analysis of human recombinant DAX-1 fusion protein.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.