

## EBP (V-14): sc-83793

### BACKGROUND

EBP (emopamil-binding protein), also known as CPX, CHO2, CPXD or CDPX2, is a 230 amino acid multi-pass membrane protein that localizes to the endoplasmic reticulum (ER) and is involved in steroid biosynthesis. Functioning to catalyze the conversion of  $\Delta^8$ -sterols to their corresponding  $\Delta^7$ -isomers, EBP plays an important role in drug transport and cholesterol metabolism within the ER. Defects in the gene encoding EBP are the cause of chondrodysplasia punctata X-linked dominant type 2 (CDPX2), a heterogeneous disorder that is caused by defective cholesterol biosynthesis. CDPX2 is characterized by punctiform calcification of the bones leading to linear ichthyosis, cataracts and short stature.

### REFERENCES

1. Hanner, M., et al. 1995. Phenylalkylamine  $\text{Ca}^{2+}$  antagonist binding protein. Molecular cloning, tissue distribution, and heterologous expression. *J. Biol. Chem.* 270: 7551-7557.
2. Derry, J.M., et al. 1999. Mutations in a  $\Delta^8$ - $\Delta^7$  sterol isomerase in the tattered mouse and X-linked dominant chondrodysplasia punctata. *Nat. Genet.* 22: 286-290.
3. Braverman, N., et al. 1999. Mutations in the gene encoding  $3\beta$ -hydroxysteroid- $\Delta^8$ ,  $\Delta^7$ -isomerase cause X-linked dominant Conradi-Hünermann syndrome. *Nat. Genet.* 22: 291-294.
4. Has, C., et al. 2000. The Conradi-Hünermann-Happle syndrome (CDPX2) and emopamil binding protein: novel mutations, and somatic and gonadal mosaicism. *Hum. Mol. Genet.* 9: 1951-1955.
5. Becker, K., et al. 2001. Identification of a novel mutation in  $3\beta$ -hydroxysteroid- $\Delta^8$ - $\Delta^7$ -isomerase in a case of Conradi-Hünermann-Happle syndrome. *Exp. Dermatol.* 10: 286-289.
6. Moebius, F.F., et al. 2003. Cloning of an emopamil-binding protein (EBP)-like protein that lacks sterol  $\Delta^8$ - $\Delta^7$  isomerase activity. *Biochem. J.* 374: 229-237.

### CHROMOSOMAL LOCATION

Genetic locus: Ebp (mouse) mapping to X A1.1.

### SOURCE

EBP (V-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the N-terminus of EBP of human origin.

### PRODUCT

Each vial contains 200  $\mu\text{g}$  IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-83793 P, (100  $\mu\text{g}$  peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

### STORAGE

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

### APPLICATIONS

EBP (V-14) is recommended for detection of EBP of mouse and rat origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2  $\mu\text{g}$  per 100-500  $\mu\text{g}$  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).

EBP (V-14) is also recommended for detection of EBP in additional species, including canine and bovine.

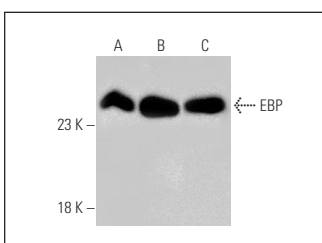
Suitable for use as control antibody for EBP siRNA (m): sc-77219, EBP shRNA Plasmid (m): sc-77219-SH and EBP shRNA (m) Lentiviral Particles: sc-77219-V.

Molecular Weight of EBP: 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) 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



EBP (V-14): sc-83793. Western blot analysis of EBP expression in Jurkat (A), HeLa (B) and Hep G2 (C) whole cell lysates.

### RESEARCH USE

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

### PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.