

EBP (T-12): sc-83792

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 Δ^8 -sterols to their corresponding Δ^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

- Hanner, M., Moebius, F.F., Weber, F., Grabner, M., Striessnig, J. and Glossmann, H. 1995. Phenylalkylamine Ca^{2+} antagonist binding protein. Molecular cloning, tissue distribution, and heterologous expression. *J. Biol. Chem.* 270: 7551-7557.
- Derry, J.M., Gormally, E., Means, G.D., Zhao, W., Meindl, A., Kelley, R.I., Boyd, Y. and Herman, G.E. 1999. Mutations in a Δ^8 - Δ^7 sterol isomerase in the tattered mouse and X-linked dominant chondrodysplasia punctata. *Nat. Genet.* 22: 286-290.
- Braverman, N., Lin, P., Moebius, F.F., Obie, C., Moser, A., Glossmann, H., Wilcox, W.R., Rimoin, D.L., Smith, M., Kratz, L., Kelley, R.I. and Valle, D. 1999. Mutations in the gene encoding 3β -hydroxysteroid- Δ^8 , Δ^7 -isomerase cause X-linked dominant Conradi-Hünermann syndrome. *Nat. Genet.* 22: 291-294.
- Has, C., Bruckner-Tuderman, L., Müller, D., Floeth, M., Folkers, E., Donnai, D. and Traupe, H. 2000. The Conradi-Hünermann-Happle syndrome (CDPX2) and emopamil binding protein: novel mutations, and somatic and gonadal mosaicism. *Hum. Mol. Genet.* 9: 1951-1955.
- Becker, K., Csikós, M., Horváth, A. and Kárpáti, S. 2001. Identification of a novel mutation in 3β -hydroxysteroid- Δ^8 - Δ^7 -isomerase in a case of Conradi-Hünermann-Happle syndrome. *Exp. Dermatol.* 10: 286-289.
- Moebius, F.F., Fitzky, B.U., Wietzorrek, G., Haidekker, A., Eder, A. and Glossmann, H. 2003. Cloning of an emopamil-binding protein (EBP)-like protein that lacks sterol Δ^8 - Δ^7 isomerase activity. *Biochem. J.* 374: 229-237.

CHROMOSOMAL LOCATION

Genetic locus: EBP (human) mapping to Xp11.23.

SOURCE

EBP (T-12) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of EBP of human origin.

PRODUCT

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

Blocking peptide available for competition studies, sc-83792 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

EBP (T-12) is recommended for detection of EBP of human and rat 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).

EBP (T-12) is also recommended for detection of EBP in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for EBP siRNA (h): sc-77218, EBP shRNA Plasmid (h): sc-77218-SH and EBP shRNA (h) Lentiviral Particles: sc-77218-V.

Molecular Weight of EBP: 26 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, Jurkat whole cell lysate: sc-2204 or HeLa whole cell lysate: sc-2200.

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.

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.

PROTOCOLS

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



Try **EBP (A-9): sc-374267**, our highly recommended monoclonal alternative to EBP (T-12).