SANTA CRUZ BIOTECHNOLOGY, INC.

KCNQ4 (H-130): sc-50417

SANTA CRUZ BIOTECHNOLOGY
The Power to Question

BACKGROUND

Epilepsy affects about 0.5% of the world's population and has a large genetic component. Epilepsy results from an electrical hyperexcitability in the central nervous system. Potassium channels are important regulators of electrical signaling, determining the firing properties and responsiveness of a variety of neurons. Benign familial neonatal convulsions (BFNC), an autosomal dominant epilepsy of infancy, has been shown to be caused by mutations in the KCNQ2 or the KCNQ3 potassium channel genes. KCNQ2 and KCNQ3 are voltage-gated potassium channel proteins with six putative transmembrane domains. Both proteins display a broad distribution within the brain, with expression patterns that largely overlap. Mutations of KCNQ4 affect the functions of sensory outer hair cells and lead to deafness.

REFERENCES

1. Singh, N.A., et al. 1998. A novel potassium channel gene, KCNQ2, is mutated in an inherited epilepsy of newborns. Nat. Genet. 18: 25-29.
2. Charlier, C., et al. 1998. A pore mutation in a novel KQT-like potassium channel gene in an idiopathic epilepsy family. Nat. Genet. 18: 53-55.
3. Schroeder, B.C., et al. 1998. Moderate loss of function of cyclic-AMP-modulated KCNQ2/KCNQ3 K^+ channels causes epilepsy. Nature 396: 687-690.
4. Biervert, C., et al. 1998. A potassium channel mutation in neonatal human epilepsy. Science 279: 403-406.
5. Wang, H.S., et al. 1998. KCNQ2 and KCNQ3 potassium channel subunits: molecular correlates of the M-channel. Science 282: 1890-1893.
6. Yang, W.P., et al. 1998. Functional expression of two KvLQT1-related potassium channels responsible for an inherited idiopathic epilepsy. J. Biol. Chem. 273: 19419-19423.
7. Tinel, N., et al. 1998. The KCNQ2 potassium channel: splice variants, functional and developmental expression. Brain localization and comparison with KCNQ3. FEBS Lett. 438: 171-176.
8. Kubisch, C., et al. 1999. KCNQ4, a novel potassium channel expressed in sensory outer hair cells, is mutated in dominant deafness. Cell 96: 437-446.

CHROMOSOMAL LOCATION

Genetic locus: KCNQ4 (human) mapping to 1p34.2; Kcnq4 (mouse) mapping to 4 D2.2.

SOURCE

KCNQ4 (H-130) is a rabbit polyclonal antibody raised against amino acids 349-478 mapping within an internal region of KCNQ4 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.

RESEARCH USE

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

APPLICATIONS

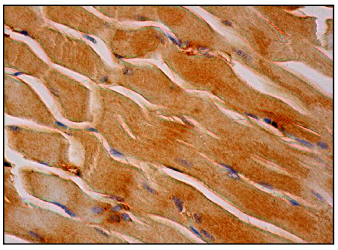
KCNQ4 (H-130) is recommended for detection of KCNQ4 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

KCNQ4 (H-130) is also recommended for detection of KCNQ4 in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for KCNQ4 siRNA (h): sc-42503, KCNQ4 siRNA (m): sc-42504, KCNQ4 shRNA Plasmid (h): sc-42503-SH, KCNQ4 shRNA Plasmid (m): sc-42504-SH, KCNQ4 shRNA (h) Lentiviral Particles: sc-42503-V and KCNQ4 shRNA (m) Lentiviral Particles: sc-42504-V.

Molecular Weight of KCNQ4: 77 kDa.

DATA



KCNQ4 (H-130): sc-50417. Immunoperoxidase staining of formalin fixed, paraffin-embedded human skeletal muscle tissue showing cytoplasmic and membrane staining of myocytes.

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.