

Gene Section

Mini Review

KCNH1 (potassium voltage-gated channel, subfamily H (eag-related), member 1)

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Identity

Hugo: KCNH1

Other names: EAG; EAG1; ether a go-go; h-eag; Kv10.1; MGC142269

Location: 1q32.2

DNA/RNA

Description

450 kb, 11 exons.

Transcription

Main RNA species with 9 kb. An alternatively spliced variant with 81 additional bp.

Protein

Description

Tetramer consisting of subunits with 969 aminoacids each. The protein contains a PAS domain at the N-terminus, "Channel" domain with typical features of K channels (six putatively transmembrane segments and a pore loop between fifth and sixth). Large C-terminal domain with cyclic-nucleotide-binding domain, calmodulin binding site, tetramerizing coiled-coil. The sequence has many potential posttranslational modification sites, but only glycosylation has been reported.

Expression

Abundant in many brain regions, virtually absent from extracranial tissues. Detected in gastric gland chief cells, pancreatic acini, spermatogenic cells, endocervix, secretory endometrium, reactive lymph nodes (germinal centre), mast cells, macrophages, anterior pituitary, adrenal gland.

Localisation

Plasma membrane. Abundant in intracellular pools both in neurons and tumor cells.

Function

Voltage-gated potassium channel.

Homology

Homologous to the "six-transmembrane, one pore" potassium channel superfamily.

Implicated in

Sarcomas (71% positivity).

Disease

Fibrosarcoma, leiomyosarcoma, liposarcoma, malignant fibrous histiocytoma, rhabdomyosarcoma, synovial sarcoma.

Prognosis

Indicative of worse outcome in liposarcoma.

Solid epithelial tumors (70% positivity).

Disease

Esophageal, gastric, colon, hepatocellular, gallbladder, pancreatic, renal cell, transitional, prostate, cervical, endometrial, breast and bronchus carcinoma; cystadenocarcinoma of the ovary, thyroid papillary carcinoma, basalioma, spinalioma, malignant melanoma.

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