

Gene Section

Mini Review

STK11 (serine/threonine kinase 11)

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Identity

Other names: LKB1; PJS (Peutz-Jeghers syndrome)

HGNC (Hugo): STK11

Location: 19p13.3

DNA/RNA

Description

Spans 23 kb; 10 exons, transcribed in telomere to centromere direction.

Transcription

Transcripts of 3.0 and 3.3 kb mainly.

Protein

Description

436 amino acids, 48 kDa; N-term with a nuclear localization domain and a putative cytoplasmic retention signal, a kinase domain, and a C- term domain that is phosphorylated on a Ser by the cAMP-dependant protein kinase.

Expression

Wide, in particular during embryonic development.

Localisation

Found in both the nucleus and the cytoplasm, but predominantly nuclear.

Function

Serine/threonine protein kinase of unknown function; expression of LKB1 results in an inhibition of cell growth by inducing G1 arrest.

Mutations

Germinal

Most mutations are null alleles; they are dispersed through the entire gene.

Somatic

Many of the polyps that develop in Peutz-Jeghers syndrome (see below) show loss of heterozygosity; somatic mutations have been tested and rarely found in the following sporadic cancers: pancreas, colon, stomach, breast, uterine cervix, ovary, testis, melanomas. The inactivation of the LKB1 can also occur through promoter hypermethylation.

Implicated in

Peutz-Jeghers syndrome (PJS)

Disease

Syndrome associating mucocutaneous melanotic pigmentation, intestinal polyposis, and an increased risk of cancers (small intestine, stomach, pancreas, colon, esophagus, ovary, uterus, breast, and lung).

Hybrid/Mutated gene

A majority of Peutz-Jeghers patients show mutation in STK11; there is however genetic heterogeneity in this disease.

Oncogenesis

STK11 is affected by biallelic inactivation in tumors of Peutz-Jeghers syndrome patients.

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