Atlas of Genetics and Cytogenetics in Oncology and Haematology

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Gene Section
Short Communication

FGFR1OP (FGFR1 oncogene partner)

Marie-Josèphe Pébusque

INSERM U119, IFR 57, 27 Blvd Lei Roure, 13009 Marseille, France (MJP)

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Identity

Other names: FOP (Fibroblast Growth Factor Receptor 1 Oncogene Partner**)**

HGNC (Hugo): FGFR1OP

Location: 6q27

FOP (6q27) - Courtesy Mariano Rocchi, Resources for Molecular Cytogenetics.

DNA/RNA

Description

Full length cDNA: 1 627 bp.

Transcription

A single open reading frame of 1 197 bp mRNA; putative ATG: bp 85; stop codon at bp 1 282; alternative splicing: multiple FOP transcript variants resulting from exon 7 or exon 11 splices.

Protein

Description

399 amino acids; predicted molecular mass: 44.3 kDa; Hydrophobic protein containing in its N- and C-termini several regions folding in a-helices with leucine-rich repeats with the consensus sequence L-X2-L-X3-5-L-X3-5-L, in one-third of which the leucine is substituted by either a valine or an isoleucine.

Expression

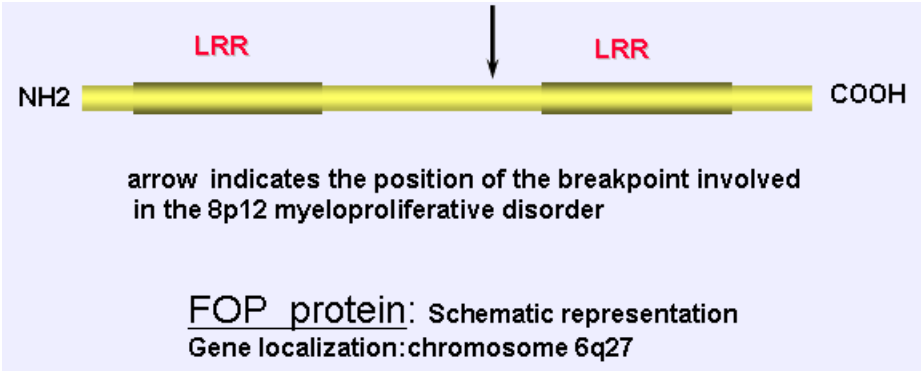
Ubiquitous expression.

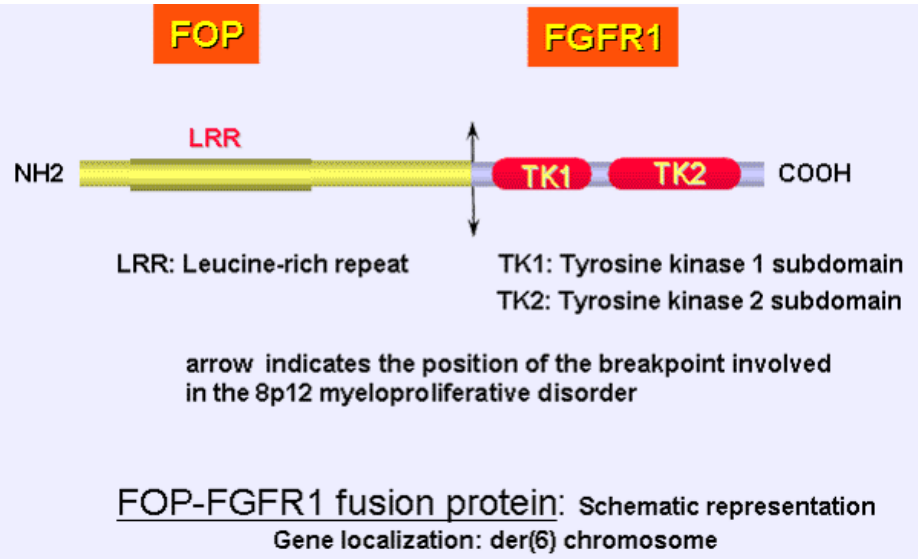
Localisation

Cell cytoplasm.

Function

Unknown.





Implicated in

t(6;8)(q27; p12) myeloproliferative disorder --> FOP - FGFR1; stem-cell myeloproliferative disorder associated with the 8p12 chromosomal translocations with fusions to the catalytic domain of FGFR1.

Disease

Stem-cell myeloproliferative disorder characterized by myeloid hyperplasia, T -cell lymphoblastic leukemia/lymphoma and peripheral blood eosinophilia, and it generally progresses to acute myeloid leukemia; specific to the 8p12 chromosomal region.

Prognosis

Very poor (median survival: 12 mths).

Cytogenetics

Additional abnormalities: 2q+ and +21.

Hybrid/Mutated gene

5' FOP - 3' FGFR1; localisation: der(6).

Abnormal protein

N-term leucine-rich region from FOP fused to the catalytic domain of FGFR1 (FGFR1 intracellular region minus the major part of the juxtamembrane domain).

Oncogenesis

Constitutive kinase activity of FGFR1 through constitutive activation of FGFR1 signal transduction pathways via putative constitutive dimerization capability mediated by the FOP N-term LRR sequences

References

Popovici C, Zhang B, Grégoire MJ, Jonveaux P, Lafage-Pochitaloff M, Birnbaum D, Pébusque MJ. The t(6;8)(q27;p11) translocation in a stem cell myeloproliferative disorder fuses a novel gene, FOP, to fibroblast growth factor receptor 1. Blood. 1999 Feb 15;93(4):1381-9

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