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Gene Section
Mini Review

RBTN2 (rhombotin-2)

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Identity

Other names: RHOM2; RBTNL1 (rhombotin-like-1); TTG2 (T-cell translocation gene 2); LMO2 (LIM domain only 2)
Location: 11p13
Local order: telomere RBTN1 - NUP98 (11p15) - CD59 - FSHB - RBTN2 - PAX6 - PDX1 - TCL2 centromere.

DNA/RNA

Description

RBTN2 belongs to a multigene family, extremely well conserved during evolution, encoding proteins containing two cystein-rich regions referred to as LIM domains : RTBN1 (11p15), RBTN2 (11p13), RBTN3 (12p); 6 exons.

Transcription

1.7 Kb mRNA (mouse).

Protein

Description

Small cystein rich protein with two tandemly arranged Zinc binding LIM domain motifs: named Lom2; 158 amino acids; 18 kDa; 48% amino-acid identity with RBTN1 protein; Lmo2 contains two transcription activating domains (one in N-term, in a prolin-rich 19 amino acid region, one in C-term) and two LIM domains as transcription repressing domains, selectively inhibiting the N-term activation domain (no effect on the C-term domain).

Expression

Early expressed during development, in all tissues (roughly consistent level in central nervous system, low level in thymus); strongly expressed in the precursors of mixed erythrocyte/macrophage/mast, erythrocyte, megakaryocyte, neutrophil and macrophage colonies, undetectable in the mature progeny; expressed in early B-cells, in leukemias of both the myeloid and lymphoid lineages.

Localisation

Nuclear.

Function

Lmo2 directly interacts with the basic-loop-helix protein Tal1/Scl and the GATA DNA protein Gata-1; Lmo2 has no direct evidence in DNA binding capacity but could act as a bridging molecule bringing together different DNA binding factors (Tal/Scl, Ldb1, E47, Gata-1) in the erythroid complex; this interaction is critical for the regulation of red blood cell development in early stages of hematopoiesis (mouse); because Lmo2 can also bind to GATA-2 protein, a complex Lom2-Gata2 might occur at earlier stages of hematopoiesis when Gata1 is not expressed; Lmo2 has a central role in adult hematopoietic pathway regulation, on bone marrow pluripotential precursor stem cell mainly; Lmo2 also interacts with retinoblastoma-binding protein 2 and elf-2 (ets transcription factor).

Implicated in

t(11;14)(p13;q11)/T-cell leukaemia → RBTN2 - TCRD-A

Disease

Childhood T-cell ALL; found in 5-10% of T-cell ALL.

Cytogenetics

A variant translocation t(7;11)(q35;p13) has been described.

Abnormal Protein

Lmo2 is activated after chromosomal translocation by association either the T-cell receptor a/d (14q11) or b gene (7q35); chromosome breakpoints occur 25 kb upstream RBTN2 gene, in a presumed transcriptional start site, inducing truncation of the promoter/control region and leading to inappropriate Lmo2 level especially in T-cells (abnormal T-cell differentiation).

References

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