

Gene Section

Mini Review

ERG (v-ets erythroblastosis virus E26 oncogene like (avian))

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Identity

Hugo: ERG

Location: 21q22.2



Probe(s) - Courtesy Mariano Rocchi.

DNA/RNA

Description

17 exons, 300 kb DNA.

Transcription

There are five isoforms created by alternative splicing and alternative initiation of translation.

Protein

Description

5 different isoforms, 38-55 kDa, all contain the ETS DNA binding domain.

Expression

ERG-3 is expressed in hematopoietic stem cells and in endothelial cells. In the GNF SymAtlas database, the major ERG expression was found to be in CD34+ cells (that include both hematopoietic stem cells and endothelial cells). ERG was also reported to be expressed in during early T and B cell development, and to be down-regulated in later stages of B and T cell differentiation. Furthermore, ERG was found to be expressed in platelets, megakaryoblastic cell lines and in primary megakaryoblastic leukemia (AMKL or M7-AML) from Down syndrome patients.

Localisation

Nuclear

Function

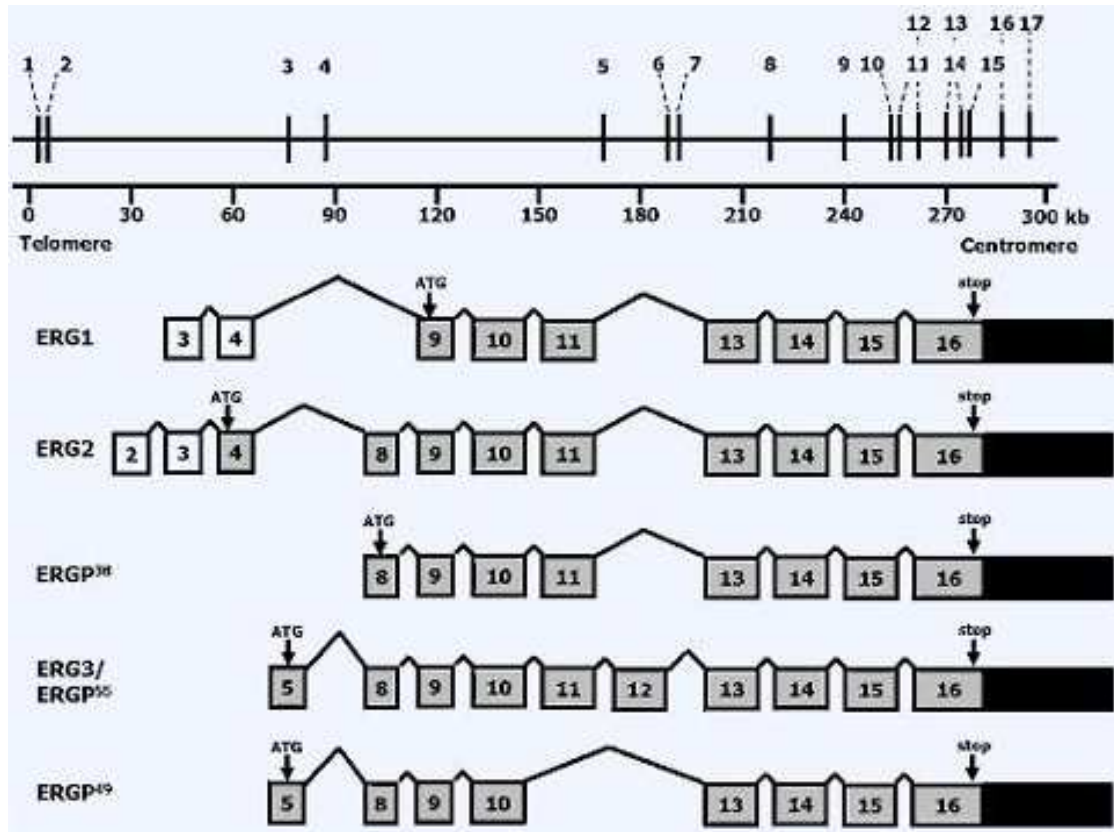
Transcription factor

Homology

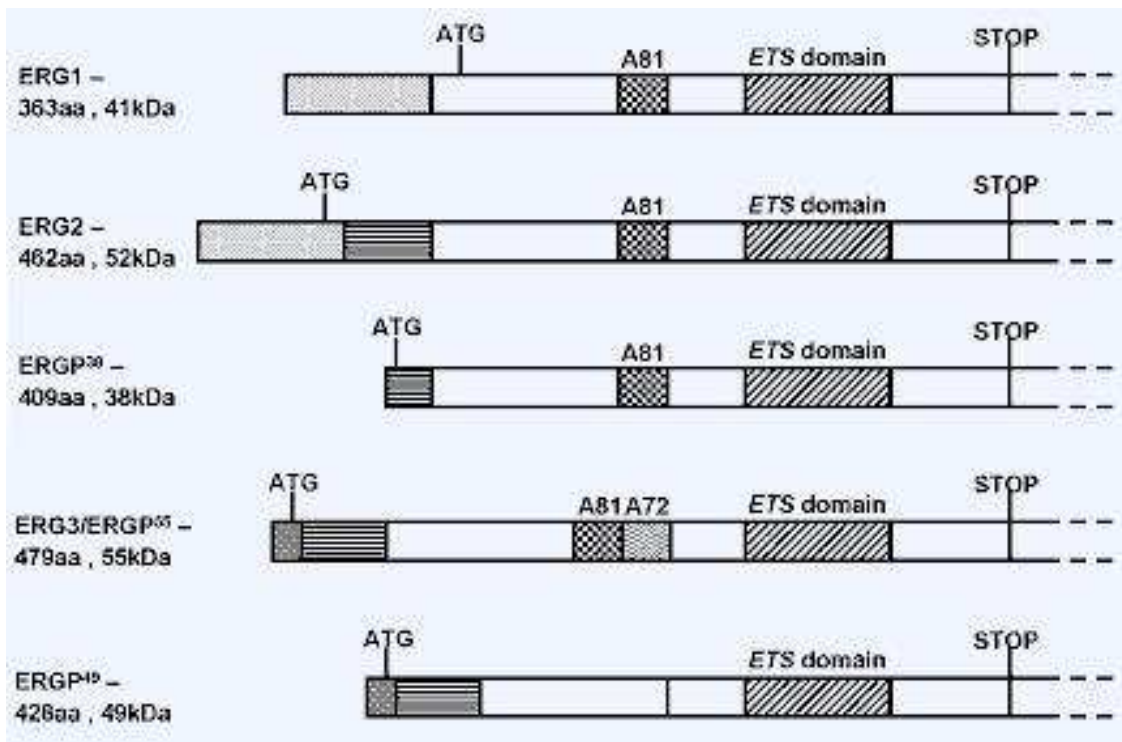
A member of the ETS transcription factors, most homologous to FLI1.

Mutations

Note: No known mutations.



Top : Schematic representation of the ERG gene locus. Exons are depicted as black boxes. Bottom : Structure of alternative transcripts encoded by the ERG gene. Coding exons are shown in grey, 5' UTRs in white and 3' UTRs in black (Adapted from Owczarek et. al., Gene 2004 (324) : 65-77).



The different ERG isoforms (Adapted from Duterque-Coquillaud et al., Oncogene 1993 (8) : 1865-1873).

Implicated in

Ewing sarcoma

Hybrid/Mutated Gene

EWSR1-ERG

Abnormal Protein

The EWS gene fuses with the carboxy terminal of ERG containing the ets DNA binding domain of ERG.

Oncogenesis

In a transgenic mouse model expression of the EWS-ERG in lymphoid progenitors induced T-cell leukemia.

Prostate cancer

Hybrid/Mutated Gene

Fusion of the the 5' untranslated region of an androgen regulated gene TMPRSS2 and all the coding region of the ERG protein.

Abnormal Protein

This translocation results in androgen dependent overexpression of ERG.

Acute myeloid leukemia (AML) and Ewing sarcoma

Prognosis

Poorer prognosis.

Hybrid/Mutated Gene

The FUS gene fuses with the carboxy terminal of ERG containing the ets DNA binding domain of ERG.

Childhood ALL with either TEL-AML1 (ETV6-RUNX1) translocation or extra copies of chromosome 21 (where ERG is located); AML with complex karyotype and AML with t(X;21)(q25-26;q22) translocation.

Note: Overexpression of ERG.

Prognosis

Overexpression of ERG was reported to predict a worse outcome in acute myeloid leukemia with normal karyotype.

Oncogenesis

Overexpression of ERG in the different leukemias and in prostate cancer suggests that overexpression of the full-length ERG may be oncogenic.

AMKL: Association of ERG expression with normal and malignant megakaryocytic differentiation

Oncogenesis

ERG was found to be expressed megakaryoblastic leukemic cell lines and in primary leukemic cells from DS patients.

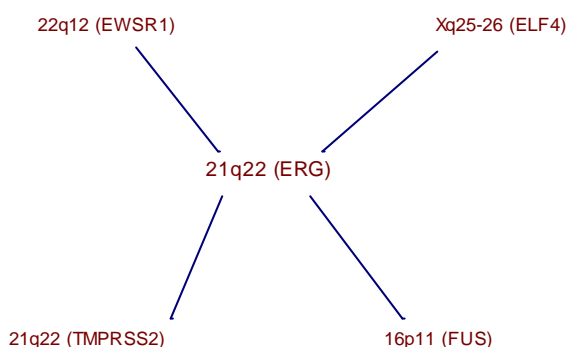
ERG involvement in endothelial development.

Note: ERG has been reported to regulate genes involved in chondrogenesis and angiogenesis and functions as a modulator of endothelial cell differentiation.

ERG involvement in lymphoid development.

Note: ERG was reported to be expressed in during early T and B cell development, and to be down-regulated in later stages of B and T cell differentiation.

Breakpoints



ERG and partners. Editor 08/2005.

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