

FLI1 Gene

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Fli-1 proto-oncogene, ETS transcription factor

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1. Normal Function

The *FLI1* gene provides instructions for making the FLI protein, which controls the activity (transcription) of genes. Transcription is the first step in the process of producing proteins. The FLI protein is part of a group of related proteins, called the Ets family of transcription factors, that control transcription. The FLI protein attaches (binds) to certain regions of DNA and turns on (activates) the transcription of nearby genes. The proteins produced from these genes control many important cellular processes, such as cell growth and division (proliferation), maturation (differentiation), and survival. The FLI protein is found primarily in blood cells and is thought to regulate their development.

2. Health Conditions Related to Genetic Changes

2.1 Ewing Sarcoma

Mutations involving the *FLI1* gene cause a type of cancerous tumor known as Ewing sarcoma. These tumors develop in bones or soft tissues such as nerves and cartilage. There are several types of Ewing sarcoma, including Ewing sarcoma of bone, extraosseous Ewing sarcoma, peripheral primitive neuroectodermal tumor, and Askin tumor. The mutations that cause these tumors are acquired during a person's lifetime and are present only in the tumor cells. This type of genetic change, called a somatic mutation, is not inherited. The most common mutation that causes Ewing sarcoma is a rearrangement (translocation) of genetic material between chromosome 11 and chromosome 22. This translocation, written as t(11;22), fuses part of the *FLI1* gene on chromosome 11 with part of another gene called *EWSR1* on chromosome 22, creating an *EWSR1/FLI1* fusion gene.

The protein produced from the *EWSR1/FLI1* fusion gene, called EWS/FLI, has functions of the protein products of both genes. Like the FLI protein, the EWS protein, produced from the *EWSR1* gene, can regulate transcription. The EWS/FLI protein has the DNA-binding function of the FLI protein as well as the transcription regulation function of the EWS protein. It is thought that the EWS/FLI protein turns the transcription of a variety of genes on and off abnormally. This dysregulation of transcription leads to uncontrolled growth and division (proliferation) and abnormal maturation and survival of cells, causing tumor development.

The *EWSR1/FLI1* fusion gene occurs in approximately 85 percent of Ewing sarcomas. The remaining tumors result from translocations that fuse the *EWSR1* gene with other genes.

2.2 Jacobsen Syndrome

3. Other Names for This Gene

- EWSR2
- FLI1_HUMAN
- Friend leukemia integration 1 transcription factor
- Friend leukemia virus integration 1
- proto-oncogene Fli-1

- SIC-1
 - transcription factor ERGB
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References

1. Mao X, Miesfeldt S, Yang H, Leiden JM, Thompson CB. The FLI-1 and chimeric EWS-FLI-1 oncoproteins display similar DNA binding specificities. *J Biol Chem.* 1994 Jul 8;269(27):18216-22.
 2. May WA, Gishizky ML, Lessnick SL, Lunsford LB, Lewis BC, Delattre O, Zucman J, Thomas G, Denny CT. Ewing sarcoma 11;22 translocation produces a chimeric transcription factor that requires the DNA-binding domain encoded by FLI1 for transformation. *Proc Natl Acad Sci U S A.* 1993 Jun 15;90(12):5752-6.
 3. May WA, Lessnick SL, Braun BS, Klemsz M, Lewis BC, Lunsford LB, Hromas R, Denny CT. The Ewing's sarcoma EWS/FLI-1 fusion gene encodes a more potent transcriptional activator and is a more powerful transforming gene than FLI-1. *Mol Cell Biol.* 1993 Dec;13(12):7393-8.
 4. Ohno T, Rao VN, Reddy ES. EWS/Fli-1 chimeric protein is a transcriptional activator. *Cancer Res.* 1993 Dec 15;53(24):5859-63.
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