

Gene Section

Short Communication

RNF213 (ALK lymphoma oligomerization partner on chromosome 17)

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Identity

Other names: ALO17 (ALK lymphoma oligomerization partner on chromosome 17); KIAA1618

HGNC (Hugo): RNF213

Location: 17q25

DNA/RNA

Transcription

Alternate spicing; 5185 and 5332 bp cDNA.

Protein

Description

1550 and 1599 amino acids if the sequence is complete; putative zinc finger in the N term, and AraC motif in the C term.

Implicated in

Anaplasic large cell lymphoma (ALCL) with t(2;17)(p23;q25) --> ALK- ALO17

Disease

ALCL are high grade non Hodgkin lymphomas; ALK+ ALCL are ALCL where ALK is involved in a fusion gene; ALK+ ALCL represent 50 to 60 % of ALCL cases (they are CD30+, ALK+); belong to the "cytoplasmic ALK+" subset.

Prognosis

Althouth presenting as a high grade tumour, a 80% five yr survival is associated with this anomaly.

Hybrid/Mutated gene

5' ALO17 - 3' ALK

Abnormal protein

NH2 ALO17 - COOH ALK.

References

Cools J, Wlodarska I, Somers R, Mentens N, Pedeutour F, Maes B, De Wolf-Peeters C, Pauwels P, Hagemeijer A, Marynen P. Identification of novel fusion partners of ALK, the anaplastic lymphoma kinase, in anaplastic large-cell lymphoma and inflammatory myofibroblastic tumor. Genes Chromosomes Cancer. 2002 Aug;34(4):354-62

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