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

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

Disease
ALCL are high grade non Hodkgin 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


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