

Gene Section

Mini Review

NUP98 (nucleoporin 98 kDa)

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Published in Atlas Database: November 1998

Online updated version : <http://AtlasGeneticsOncology.org/Genes/NUP98.html>
DOI: 10.4267/2042/37478

This article is an update of: Huret JL. NUP98 (nucleoporin 98 kDa). Atlas Genet Cytogenet Oncol Haematol.1998;2(1):7.

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Identity

Location : 11p15

DNA/RNA

Transcription

3.6, 6.5, 7.0 kb mRNA.

Protein

Description

920 amino acids; 97 kDa; contains repeated motifs (GLFG and FG) in N-term and a RNA binding motif in C-term.

Expression

Wide.

Localisation

Nuclear membrane localisation.

Function

Nucleoporin: associated with the nuclear pore complex; role in nucleocytoplasmic transport processes.

Homology

Member of the GLFG nucleoporins.

Implicated in

t(7;11)(p15;p15)/ANLL --> NUP98/HOXA9

Disease

M2-M4 ANLL mostly; occasionally, CML-like cases.

Prognosis

Mean survival: 15 months.

Cytogenetics

Sole anomaly most often.

Hybrid/Mutated gene

5' NUP98 - 3' HOXA9.

Abnormal protein

Fuses the GLFG repeat domains of NUP98 to the HOXA9 homeobox.

inv(11)(p15q22)/MDS or ANLL --> NUP98/DDX10

Disease

Therapy-related MDS and ANLL; de novo ANLL.

Hybrid/Mutated gene

5' NUP98 - 3' DDX10

Abnormal protein

Fuses the GLFG repeat domains of NUP98 to the acidic domain of DDX11.

t(2;11)(q31;p15)/treatment-related leukaemia --> NUP98/HOXD13

Disease

So far, only 1 case of treatment related myelodysplasia evolving towards M6 acute non lymphocytic leukaemia.

Hybrid/Mutated gene

5' NUP98 - 3' HOXD13.

Abnormal protein

Fuses the GLFG repeat domains of NUP98 to the HOXD13 homeodomain.

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This article should be referenced as such:

Huret JL. NUP98 (nucleoporin 98 kDa). *Atlas Genet Cytogenet Oncol Haematol.* 1999; 3(1):15-16.
