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Gene Section
Mini Review

PRRX2 (paired related homeobox 2)

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Identity

Hugo: PRRX2
Other names: PRX2; PMX2; MGC19843
Location: 9q34.1

DNA/RNA

Description

57 kb, 4 exons.

Transcription

1327 bp mRNA.

Protein

Note: Paired mesoderm homeobox protein 2, Paired related homeobox protein 2.

PRRX2 protein. HD = Homeodomain.

Description

253 amino acids, 27 kDa, contains an homeobox DNA-binding domain and an OAR domain.

Expression

In embryon, higher levels of transcripts in heart, kidney, lung and skeletal muscle; lower levels in spleen and thymus; barely detecteble levels in brain and liver.
In adult, higher levels in heart, lung, placenta and pancreas; moderate expression in kidney and skeletal muscle.

Localisation

Nuclear.

Function

Fetal skin development, cutaneous regeneration and possible role in cellular proliferation. Transcription factor activity.

Homology

Member of the paired family of homeobox proteins. Murine Prrx2.

Implicated in

t(9;11)(q34;p15)/t-AML ⟶ NUP98-PPRX2

Disease

One case of adult t-AML.

NUP98-PRRX2 fusion cDNA partial sequence.

Structure of the predicted chimeric NUP98-PRRX2 protein. FG = Phe-Gly repeats, GLEBS = RAE1 binding domain, HD = homeodomain.

Cytogenetics

No additional cytogenetic abnormality in this case.

Hybrid/Mutated Gene

5' NUP98 - 3' PRRX2

Abnormal Protein

Fuses the GLFG repeat domains of NUP98 to the homeodomain of PRRX2.

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