

Leukaemia Section

Short Communication

t(3;12)(q27;p12) LRMP/BCL6

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Published in Atlas Database: December 2012

Online updated version : <http://AtlasGeneticsOncology.org/Anomalies/t0312q27p12ID2129.html>
DOI: 10.4267/2042/48871

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Clinics and pathology

Disease

Non Hodgkin lymphoma

Clinics

The t(3;12)(q27;p12) was found in a case of follicular lymphoma transformed to diffuse aggressive lymphoma, from a study with no individual data (Akasaka et al., 2003).

Genes involved and proteins

BCL6

Location

3q27.3

Protein

706 amino acids; composed of a NH₂-term BTB/POZ domain (amino acids 1-130 (32-99 according to Swiss-Prot) which mediates homodimerization and protein-protein interactions with other corepressors (including HDAC1 and NCOR2/SMRT to constitute a large repressing complex, another transcription repression domain (191-386), PEST sequences (300-417) with a KKYK motif (375-379), and six zinc finger at the C-term (518-541, 546-568, 574-596, 602-624, 630-652, 658-681), responsible for sequence specific DNA binding. Transcription repressor; recognizes the consensus sequence: TTCCT(A/C)GAA (Albagli-Curiel, 2003). Role in germinal centers of lymphoid follicles. BCL6 prevents ATM and TP53 to induce apoptosis in response to DNA rearrangements such as somatic hypermutation and class switch recombination. Therefore essential for normal B cell development.

LRMP

Location

12p12.1

Protein

LRMP is an integral membrane protein of the endoplasmic reticulum (ER), oriented on the ER membrane facing the cytosol.

It is a lymphoid-restricted gene that is expressed in a developmentally regulated fashion (Behrens et al., 1994).

It may be involved in diabetes susceptibility (Grimm et al., 2003). A V141L polymorphism of LRMP has been found associated with a higher mortality in patients with lung cancer (Manenti et al., 2006).

LRMP has recently been found involved in a translocation t(8;12)(q24;p12) with a LRMP/MYC fusion gene in non Hodgkin lymphoma (Singh et al., 2012).

Result of the chromosomal anomaly

Hybrid gene

Description

Breakpoint in BCL6 first intron.

References

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This article should be referenced as such:

Huret JL. t(3;12)(q27;p12) LRMP/BCL6. *Atlas Genet Cytogenet Oncol Haematol*. 2013; 17(4):275-276.
