Gene Section
Mini Review

BCL6 (B-Cell Lymphoma 6)

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Identity

Hugo: BCL6
Other names: LAZ3 (Lymphoma Associated Zinc finger on chromosome 3); ZNF51 (Zinc Finger Protein 51)
Location: 3q27
Local order: gene orientation: telomere - 5' LAZ3 3' - centromere.

DNA/RNA

Description

The gene is encoded by 11 exons that are located on Chromosome 3q27 and is 24.3 kb. The 5' portion encodes for the BTB/POZ domain (broad-complex/tramtrack/bric-a-brac/pox virus/zinc finger), while the 3' end encodes for 6 DNA binding zinc fingers. The first ATG occurs in exon 3.

Transcription

3.8 kb mRNA.

Protein

Description

The protein product is 706 amino acids with an estimated molecular weight of 78.8 kDa.

Expression

Normally expressed in germinal center B and T cells, other lymphoid tissues, in skeletal muscle cells and in keratinocytes.

Localisation

Nuclear paraspeckles/dots.

Function

The protein can bind to sequence specific DNA and repress its transcription in addition to recruiting other protein repressors. The DNA binding is mediated through the consensus sequence: TTCCT(A/C)GAA while the protein-protein interactions are mediated through the BTB/POZ domain and it has been shown to interact with other zinc finger proteins and corepressors (including Histone Deacetylase 1 (HDAC1) and Silencing Mediator of Retinoid and Thyroid Receptor 1 (SMRT1)). The carboxy terminus, on the other hand, is responsible for sequence specific DNA binding through its 6 zinc fingers.

Homology

BTB/POZ - Zinc Finger proteins (PLZF, HIC1, KUP, BAZF, ttk (drosophila), BrC (drosophila)...).
Implicated in

3q27 rearrangements / NHL (non Hodgkin lymphomas)

Disease

B cell non-Hodgkin Lymphoma (B-NHL) carry the greatest number of translocations involving the BCL6 gene locus. Translocations are most commonly detected within 15-40% of Diffuse Large B-Cell Lymphomas (DLBCL), 6-15% of Follicular Lymphomas (FL), and 50% of nodular lymphocyte predominant Hodgkin Lymphomas.

Prognosis

Generally considered to be a better prognosis if there is increased expression of BCL6. The mechanism by which its expression is increased does not seem to matter (ie different translocation partners increasing its expression results in the same prognosis).

Cytogenetics

3q27 rearrangements/aberrations are diverse and include: translocations, micro-deletions, point mutations and hypermutation. Approximately 50% of 3q27 translocations involves Ig genes at 14q32 (IgH), 2p12 (IgK) and 22q12 (IgL) (e.g. t(3;14)(q27;q32)). Less than half (~40%) include a variety of other chromosomal regions (1q21, 2q21, 4p11, 5q31, 6p21, 7p12, 8q24, 9p13, 11q13, 11q23, 12q11, 13q14-21, 14q11, 15q21, 16p11...). In addition, there are frequent bi-allelic alterations (translocation and deletion or mutation on the non-translocated allele).

Hybrid/Mutated Gene

Hybrid gene and transcripts are formed following promoter substitution between BCL6 and its different partners. Chimeric transcripts are generally detected containing the 5’ part of the gene partner fused to the normal BCL6 exon 2 splice acceptor site. In some cases reciprocal chimeric transcripts driven by the 5' regulatory region of BCL6 fused to the partner gene coding region, have been characterised.

t(2;3)(p12;q27) the gene in 2p12 is IGK

t(3;3)(q25;q27) the gene in 3q25 is MBNL1

t(3;3)(q27;q27) the gene in 3q27 is ST6GAL1

t(3;3)(q27;q29) the gene in 3q29 is TFRC

t(3;4)(q27;p13) the gene in 4p13 is RHOH

t(3;6)(q27;p22) the gene in 6p22 is HIST1H4I

t(3;6)(q27;p21) the gene in 6p21 is PIK1

t(3;6)(q27;p21) the gene in 6p21 is SFRS3

t(3;6)(q27;p21) the gene in 6p21 is Histone H4

t(3;6)(q27;p12) the gene in 6p12 is HSP90AB1

t(3;6)(q27;q15) the gene in 6q15 is SNHG5

t(3;7)(q27;p12) the gene in 7p12 is IKZF1

t(3;8)(q27;q24.1) the gene in 8q24.1 is MYC

t(3;9)(q27;p11) the gene in 9p11 is GRHPR

t(3;11)(q27;q23) the gene in 11q23 is POU2AF1

t(3;12)(q27;p13) the gene in 12p13 is GAPDH

t(3;12)(q27;q12) the gene in 12q12 is LRMP

t(3;12)(q27;q23) the gene in 12q23 is NACA

t(3;13)(q27;q14) the gene in 13q14 is LCP1

t(3;14)(q27;q32) the gene in 14q32 is IGH

t(3;14)(q27;q32) the gene in 14q32 is HSP90AA1

t(3;16)(q27;p13) the gene in 16p13 is CIITA

t(3;16)(q27;p11) the gene in 16p11 is IL2R

t(3;19)(q27;q13) the gene in 19q13 is NAPA

t(3;22)(q27;q11) the gene in 22q11 is IGL

Abnormal Protein

No fusion protein.

Breakpoints

Note: Clustered in a 3.3kb EcoRI fragment (MTC) including exon 1A and intron 1.
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References


This article should be referenced as such: