Atlas of Genetics and Cytogenetics in Oncology and Haematology

OPEN ACCESS JOURNAL AT INIST-CNRS

Gene Section
Mini Review

IGL@ (Immunoglobulin Lambda)

Marie-Paule Lefranc

IMGT, LIGM, IGH, UPR CNRS 1142, 141 rue de la Cardonille, 34396 Montpellier Cedex 5, France (MPL)

Published in Atlas Database: September 2003

Online updated version: http://AtlasGeneticsOncology.org/Genes/IgLID54.html
DOI: 10.4267/2042/38015

This article is an update of :
Lefranc MP. IGL (Immunoglobulin Lambda). Atlas Genet Cytogenet Oncol Haematol 2000;4(3):114-116

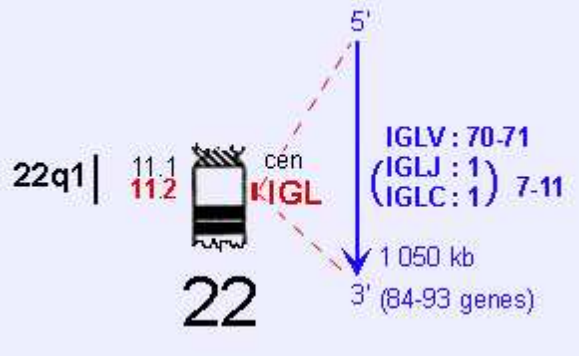
Identity

Other names: IGL (Immunoglobulin Lambda)

HGNC (Hugo): IGL@

Location: 22q11.2

Note: The human IGL locus is located on chromosome 22 on the long arm, at band 22q11.2. The orientation of the locus has been determined by the analysis of translocations, involving the IGL locus, in leukemia and lymphoma. Sequencing of the long arm of chromosome 22 showed that it encompasses about 35 megabases of DNA and that the IGL locus is localized at 6 megabases from the centromere. Although the correlation between DNA sequences and chromosomal bands has not yet been made, the localization of the IGL locus can be refined to 22q11.2.

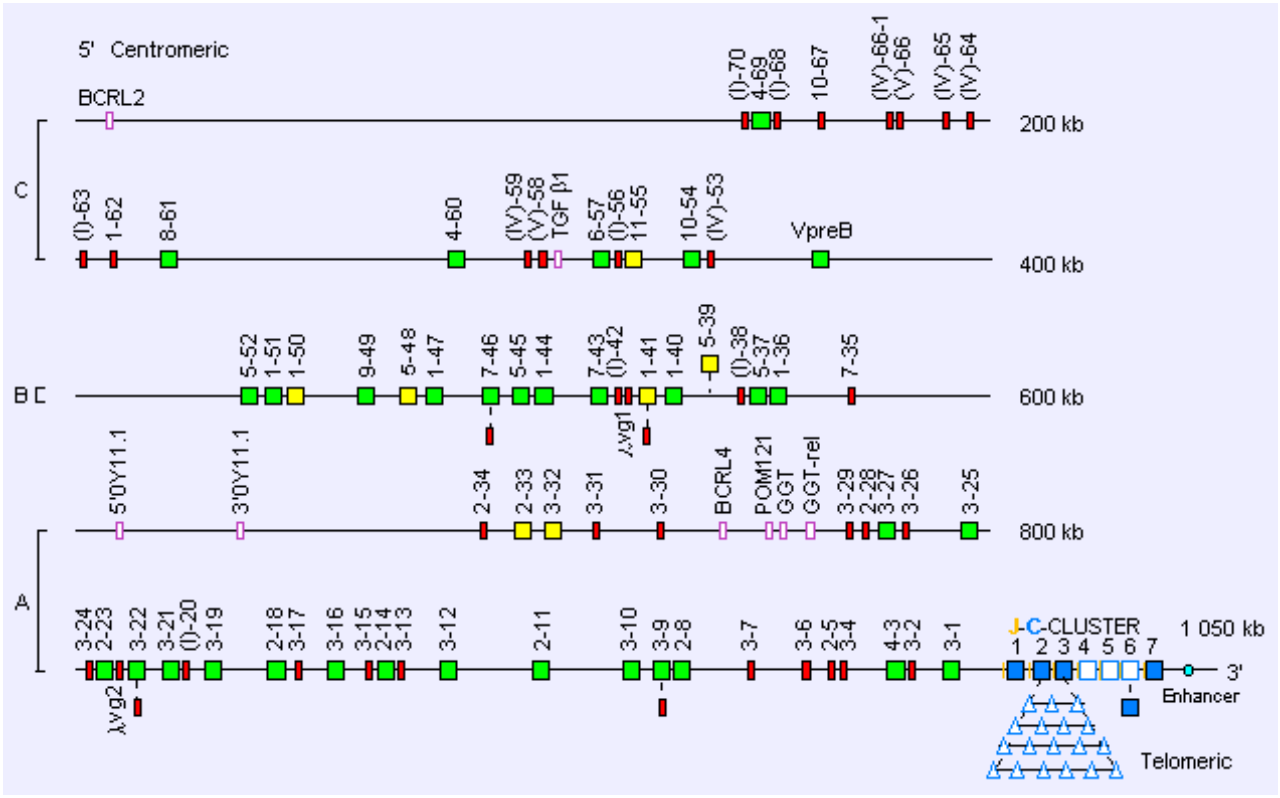


For complete Figure, see: chromosome 22, IMGT (The International ImMunoGeneTics information system ®) © Copyright 1995-2003 IMGT, IMGT is a CNRS trademark.

DNA/RNA

Description

The human IGL locus at 22q11.2 spans 1050 kb. It consists of 70 to 71 IGLV genes, localized on 900 kb, 7 to 11 IGLJ and 7 to 11 IGLC genes depending on the haplotypes, each IGLC gene being preceded by one IGLJ segment.
Fifty-six to 57 genes belong to 11 subgroups, whereas 14 pseudogenes which are too divergent to be assigned to subgroups have been assigned to 3 clans.
The most 5' IGLV genes occupy the more centromeric position, whereas the IGLC genes, in 3' of the locus, are the most telomeric genes in the IGL locus.
The potential genomic IGL repertoire comprises 29 to 32 functional IGLV genes belonging to 10 subgroups, 4 to 5 IGLJ, and 4 to 5 IGLC functional genes in the 7-IGLC gene haplotype. One, 2, 3 or 4 additional IGLC genes, each one probably preceded by one IGLJ, have been shown to characterize IGLC haplotypes with 8, 9, 10 or 11 genes, but these genes have not yet been sequenced.
Two IGLV orphons have been identified on chromosome 8 at 8q11.2 and one of them belonging to subgroup 8 has been sequenced.
The recent sequencing of the chromosome 22q showed that the IGL locus is localized at 6 megabases from the centromere. Two IGLC orphons and two IGLV orphons have also been characterized on 22q outside of the major IGL locus (See also IMGT Repertoire).
The total number of human IGL genes per haploid genome is 84-93 (90-99 genes, if the orphons are included) of which 37-42 genes are functional
List of the human IGL genes.



IGL
V-GENE: Green box: Functional; Yellow box: Open reading frame; Red: Pseudogene.
J-GENE: Grey: Functional .
C-GENE: Blue: Functional; Blue open box: Pseudogene; Blue triangle: Not sequenced.
GENES NOT RELATED: Purple open box: Pseudogene
For complete Figure, see: locus IGL, IMGT (The International ImMunoGeneTics information system ®) © Copyright 1995-2003 IMGT, IMGT is a CNRS trademark.

Protein

Description

Proteins encoded by the IGL locus are the immunoglobulin lambda chains. They result from the recombination (or rearrangement), at the DNA level, of two genes: IGLV and IGLJ, with deletion of the intermediary DNA to create a rearranged IGLV-J gene. The rearranged IGLV-J gene is trans-cribed with one of the IGLC genes and translated into an immunoglobulin lambda chain.

Translation of the variable germline genes involved in the IGLV-J rearrangements are available at IMGT Repertoire Protein displays.

Compared to the germline genes, the rearranged variable genes will acquire somatic mutations during the B cell differentiation in the lymph nodes, which will considerably increase their diversity. These somatic mutations can be analysed using the IMGT/V-QUEST tool available at http://imgt.cines.fr

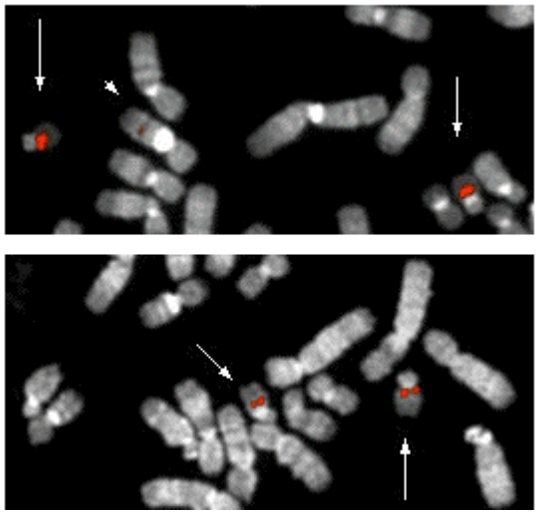
Mutations

Note

Mutations which correspond to allelic polymo-rphisms of the functional germline IGLV, IGLJ and IGLC genes are described in the IMGT database: (IMGT Repertoire>Alignments of alleles).

Implicated in

Translocations which frequently result from errors of the recombinase enzyme complexe (RAG1, RAG2, etc.), responsable of the Immunoglobulin and T cell receptor V-J and V-D-J rearrangements. IGLV or IGLJ recombination signals or isolated heptamer are observed at the breakpoints.



c-Immunoglobulin gene IgL at 22q11, in normal cells: PAC 1019H10 and PAC 869I1 - Courtesy Mariano Rocchi, Resources for Molecular Cytogenetics.

t(3;22)(q27;q11); involve BCL6 in 3q27

Disease

B-cell non-Hodgkin lymphomas (NHL), mainly diffuse large cell lymphoma; adult aggressive lymphoma.

Prognosis

Controversial.

t(8;22)(q24;q11); involve C-MYC in 8q24

Disease

B-cell acute lymphoblastic leukemia (ALL3) and non-Hodgkin lymphomas (NHL), especially in the Burkitt lymphoma.

Prognosis

The prognosis has evolved with new treatments.

References

Emanuel BS, Cannizzaro LA, Magrath I, Tsujimoto Y, Nowell PC, Croce CM. Chromosomal orientation of the lambda light chain locus: V lambda is proximal to C lambda in 22q11. Nucleic Acids Res. 1985 Jan 25;13(2):381-7

Dariavach P, Lefranc G, Lefranc MP. Human immunoglobulin C lambda 6 gene encodes the Kern+Oz-lambda chain and C lambda 4 and C lambda 5 are pseudogenes. Proc Natl Acad Sci U S A. 1987 Dec;84(24):9074-8

Ghanem N, Dariavach P, Bensmana M, Chibani J, Lefranc G, Lefranc MP. Polymorphism of immunoglobulin lambda constant region genes in populations from France, Lebanon and Tunisia. Exp Clin Immunogenet. 1988;5(4):186-95

Vasicek TJ, Leder P. Structure and expression of the human immunoglobulin lambda genes. J Exp Med. 1990 Aug 1;172(2):609-20

Frippiat JP, Williams SC, Tomlinson IM, Cook GP, Cherif D, Le Paslier D, Collins JE, Dunham I, Winter G, Lefranc MP. Organization of the human immunoglobulin lambda light-chain locus on chromosome 22q11.2. Hum Mol Genet. 1995 Jun;4(6):983-91

Kawasaki K, Minoshima S, Schooler K, Kudoh J, Asakawa S, de Jong PJ, Shimizu N. The organization of the human immunoglobulin lambda gene locus. Genome Res. 1995 Sep;5(2):125-35

Kawasaki K, Minoshima S, Nakato E, Shibuya K, Shintani A, Schmeits JL, Wang J, Shimizu N. One-megabase sequence analysis of the human immunoglobulin lambda gene locus. Genome Res. 1997 Mar;7(3):250-61

Lefranc MP, Pallarès N, Frippiat JP. Allelic polymorphisms and RFLP in the human immunoglobulin lambda light chain locus. Hum Genet. 1999 May;104(5):361-9

Lefranc MP, Pallarès N, Frippiat JP. Allelic polymorphisms and RFLP in the human immunoglobulin lambda light chain locus. Hum Genet. 1999 May;104(5):361-9

Scaviner D, Barbié V, Ruiz M, Lefranc MP. Protein displays of the human immunoglobulin heavy, kappa and lambda variable and joining regions. Exp Clin Immunogenet. 1999;16(4):234-40

Lefranc MP. Nomenclature of the human immunoglobulin lambda (IGL) genes. Exp Clin Immunogenet. 2001;18(4):242-54

This article should be referenced as such:

Lefranc MP. IGL@ (Immunoglobulin Lambda). Atlas Genet Cytogenet Oncol Haematol. 2003; 7(4):241-243.