Atlas of Genetics and Cytogenetics in Oncology and Haematology

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Gene Section

Mini Review

TRA@ (T cell Receptor Alpha)

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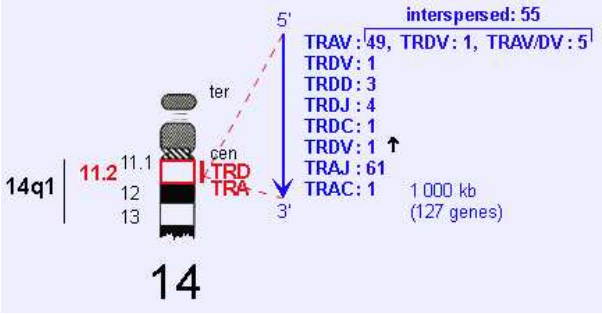
Identity

HGNC (Hugo): TRA@

Location: 14q11.2

Note

The human TRA locus is located on the chromosome 14 on the long arm at band 14q11.2. The orientation of the locus has been determined by the analysis of translocations, involving the TRA and TRD loci, in leukemia and lymphoma.



For complete Figure, see: chromosome 14, IMGT (The International ImMunoGeneTics information system ®) © Copyright 1995-2003 IMGT, IMGT is a CNRS trademark.

Probe(s) - Courtesy Mariano Rocchi, Resources for Molecular Cytogenetics.

DNA/RNA

Description

The human TRA locus at 14q11.2 spans 1000 kilobases (kb). It consists of 54 TRAV genes belonging to 41 subgroups, 61 TRAJ segments localized on 71 kb, and a unique TRAC gene.

The most 5' TRAV genes occupy the most centromeric position, whereas the TRAC genes, 3' of the locus, is the most telomeric gene in the TRA locus.

The organization of the TRAJ segments on a large area is quite unusual and has not been observed in the other immunoglobulin or T cell receptor loci.

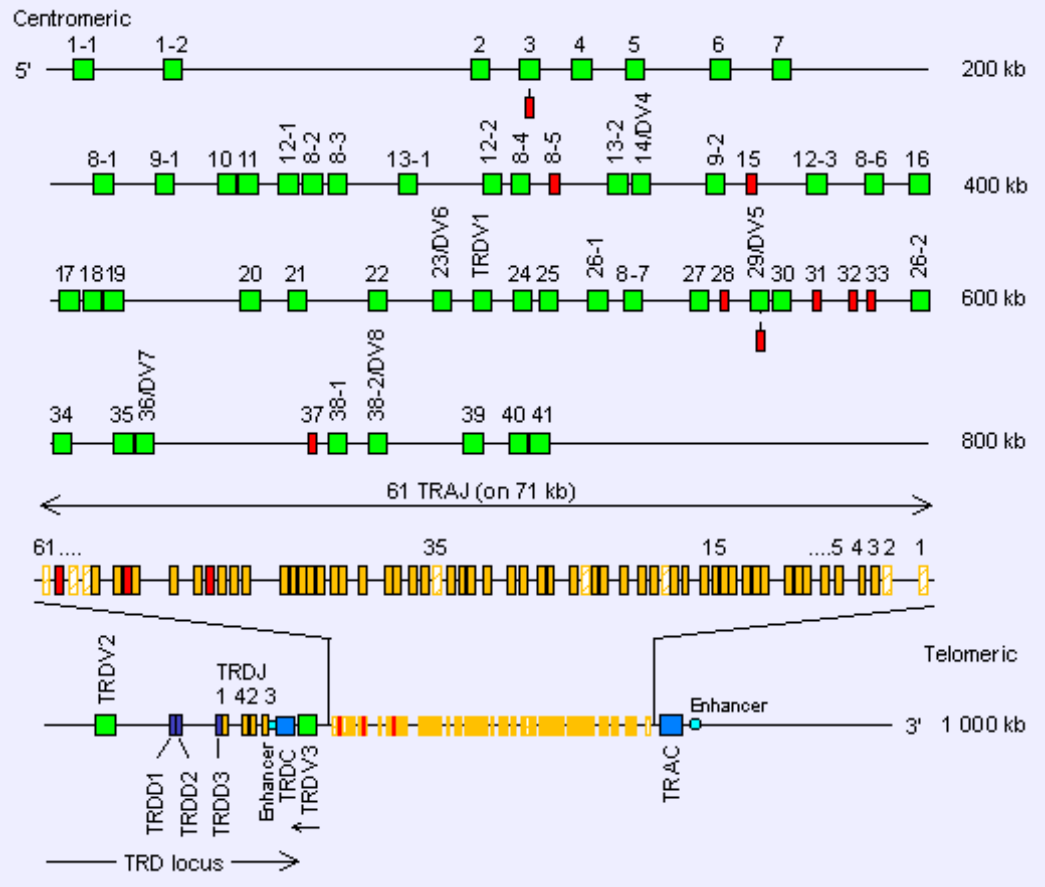
Moreover the TRD locus is nestled in the TRA locus between the TRAV and TRAJ segments. V-J-rearrangements in the TRA locus therefore result in deletion of the TRD genes localized on the same chromosome. That deletion occurs in two steps, that is a deletion of the TRD genes, involving specific sequences located upstream from TRDC (sequence pseudo J alpha) would take place before the TRAV-J rearrangement.

The potentiel genomic TRA repertoire comprises 45-47 functional TRAV genes belonging to 33-35 subgroups, 50 functional TRAJ segments, and the unique TRAC gene.

Among the variable genes are included five genes designated as TRAV/DV which belong to five different subgroups and which have been found rearranged either to TRAJ or to TRDD segments and can therefore be used in the synthesis of alpha or delta chains.

The total number of human TRA genes per haploid genome is 116 of which 96 to 98 genes are functional. Enhancer sequences have been characterized 4.5kb 3' from TRAC.

List of the human TRA genes.



TRA/TRD
V-GENE: Green box: Functional; Red: Pseudogene.
D-GENE: Blue: Functional.
J-GENE: Yellow: Functional; Pale yellow: Open reading frame; Red: pseudogene.
C-GENE: Blue: Functional.
For complete Figure, see: locus TRA, IMGT (The International ImMunoGeneTics information system ®) © Copyright 1995-2003 IMGT, IMGT is a CNRS trademark

Protein

Description

Proteins encoded by the TRA locus are the T cell receptor alpha chains. They result from the recombination (or rearrangement), at the DNA level, of two genes: TRAV and TRAJ, with deletion of the intermediary DNA to create a rearranged TRAV-J gene. The rearranged TRAV-J gene is transcribed with the TRAC gene and translated into an T cell receptor alpha chain.

Translation of the variable germline genes involved in the TRAV-J rearrangements are available at IMGT Repertoire Protein displays. TRA V-J rearrangements can be analysed using the IMGT/V-QUEST tool.

Mutations

Note

Mutations which correspond to allelic poly-morphisms of the functional germline TRAV, TRAJ and TRAC genes are described in the IMGT database: (IMGT Repertoire>Alignments of alleles).

Implicated in

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

t(1;14)(p32;q11); involve TAL1 in 1p32

Prognosis

Median survival > 5 yrs in children.

t(8;14)(q24;q11); involve MYC in 8q24

Disease

T-cell acute lymphocytic leukemia (ALL); rare.

t(10;14)(q24;q11); involve HOX11 in 10q24

Disease
T-cell acute lymphoblastic leukemia (ALL) and non-Hodgkin lymphoma (NHL).

Prognosis
Not unfavourable.

t(11;14)(p13;q11); involve RBTN2 in 11p13

Disease
T-cell acute lymphocytic leukemia (ALL).

t(14;14)(q11;q32), inv(14)(q11q32); involve TCL1 in 14q32

Disease
T-cell prolymphocytic leukemia (T-PLL) and adult T cell leukemia/lymphoma.

Prognosis
Poor.

t(14;21)(q11;q22); involve OLIG2 in 21q22

Disease
T-cell acute lymphoblastic leukemia (ALL)

Prognosis
Unknown.

References

Hoh F, Yang YS, Guignard L, Padilla A, Stern MH, Lhoste JM, van Tilbeurgh H. Crystal structure of p14TCL1, an oncogene product involved in T-cell prolymphocytic leukemia, reveals a novel beta-barrel topology. Structure. 1998 Feb 15;6(2):147-55

Lefranc MP. Nomenclature of the human T cell Receptor genes (Review) Current Protocols in Immunology. 2000, Wiley, J. and Sons, New York, Supplement 40

Lefranc M-P.. Locus Map and Genomic repertoire of the Human Immunoglobulin Genes (Review) The immunologist. 2000; 8: 80-8.

Lefranc MP and Lefranc G. The T cell Receptor FactsBook (Review) Academic Press, London, UK (2001) ISBN:0124413528.

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Lefranc MP. TRA@ (T cell Receptor Alpha). Atlas Genet Cytogenet Oncol Haematol. 2003; 7(4):244-246.