

## Classification of T-Cell disorders

### Identity

**Note** T-cell lymphoid disorders include a variety of disease entities which result from the clonal neoplastic expansion of an uncommitted (thymic) or a committed (post thymic) T-cell. Some of these diseases have distinct cytogenetic/molecular genetic features which allow to better define the various entities and understand their pathogenesis.

### Clinics and Pathology

<b>Disease</b>	<a href="#">T-prolymphocytic leukemia (T-PLL)</a> Variants:small cell and cerebriform cell
Phenotype / cell stem origin	TdT-, CD1a-, CD4+ CD8- CD4 - CD8+ CD4+ CD8+
Clinics	Aggressive course splenomegaly, high WBC with prolymphocytes
Cytogenetics	<a href="#">Inv(14)(q11q32), t(14;14)(q11;q32)</a> Xq28 abnormalities idic(8)(p11), t(8; 8)(p11;q1-2) 11q22-23 abnormalities 12p abnormalities 13q14.3 deletions
Genes	<a href="#">ATM</a> gene (11q22-23) mutated. <a href="#">TCL1</a> (14q32.1) or <a href="#">MTCP1</a> (Xq28) activated
<b>Disease</b>	<a href="#">Large granular lymphocyte leukemia (LGL) - T-cell Type</a>
Phenotype / cell stem origin	TdT-,CD1a CD3+,CD2+,CD8+ CD4 -,CD57+, CD16+/-Cytotoxic or suppressor activity
Clinics	Indolent cytopenias, splenomegaly, lymphocytosis with granular lymphocytes.
Cytogenetics	Clonal abnormalities.in some cases, but no consistent specific abnormalities
Genes	Clonality established by TCR rearrangements
<b>Disease</b>	<a href="#">Large granular lymphocyte leukemia (LGL) - NK type</a>
Phenotype / cell stem origin	TdT-,CD1a CD2+,CD56+, CD16+ ,CD7+/-CD3-, CD5-,TCR-Natural killer Activity.
Clinics	Aggressive or indolent lymphocytosis, splenomegaly, hepatomegaly
Cytogenetics	del(6)(q21-25)
Genes	TCR chain genes in germ line.

**Disease** Sezary syndrome (SS)  
 Phenotype / cell stem origin TdT-, CD1a-, CD3+,CD4+,CD8-, Helper or no functional activity.  
 Clinics Variable clinical course with skin involvement and cells with cerebriform nuclei  
 Cytogenetics Complex, clonal, oligoclonal or nonclonal with variable ploidy  
 Abnormal.2p,  
 Abnormal.6q  
 i(17q),  
 del (13)(q14)  
 Genes [P53](#) gene deletion and protein expression in the absence of gene mutation.  
 Few cases express MDM2

**Disease** Adult T-cell leukemia lymphoma (ATLL)  
 Phenotype / cell stem origin TdT-, CD1a- CD7- CD4+ CD8- CD25+, Suppressor activity  
 Clinics Aggressive , hypercalcaemia, lymphadenopathy, 'flower cells', HTLV-1 Positive.  
 Cytogenetics Complex and often oligoclonal.  
 Numerical abnormalities: 3, 7, X  
 Structural abnormalities: 1q, 3q, 6q, 14q.  
 Genes Oligoclonal/mono clonal integration of HTLV-1 in host DNA  
 Abnormalities of [p53](#), [p16](#) and p15 genes.

**Disease** a/d T-NHL hepatosplenic lymphoma  
 Phenotype / cell stem origin TdT- CD1a- CD3+/- CD56+, CD7+, granzymeA+, TCR g/d+  
 Clinics Aggressive, Hepato splenomegaly  
 Cytogenetics Abnormal.7q, i (7p)  
 Genes TCR genes gamma/delta rearranged but alpha/beta not rearranged

**Disease** Peripheral/post-thymic T cell lymphoma (pleomorphic and immunoblastic subtypes)  
 Phenotype / cell stem origin TdT-, CD1a-, Variable expression of CD4 or CD8  
 Clinics Aggressive; advanced stages.  
 Cytogenetics variable

**Disease** Angio immunoblastic T-cell lymphoma  
 Phenotype / cell stem origin TdT-, CD1a-, CD2+, CD5+, CD3+ CD4+ CD8-  
 Clinics Disproteinemia, lymphadenopathy, immune abnormalities

Cytogenetics      Complex with multiple related or unrelated clones.  
                          [+3](#) or i(3q), +5, del(6q).  
                          Progression from normal karyotype to abnormal clone observed during transition from hyperplasia to neoplasia.

Genes                Integrated EBV sequences present in both B-and T-cells and is unlikely to be the etiological agent.

**Disease**            Angiocentric (nasal) T-cell lymphoma

Phenotype /  
 cell stem            TdT-, CD1a-, T-cell or NK phenotype.  
 origin

Clinics                Prevalent in Asia and south America; extra nodal involvement.

Cytogenetics        i(1q), del(6q), i(6p)

Genes                Majority have no TCR rearrangement; EBV clonally integrated and plays a role in the etiology of the disease

**Disease**            [Anaplastic \(Ki 1+\) large cell lymphoma](#)

Phenotype /  
 cell stem            TdT-, CD1a-, CD3+/- CD30+ (Ki 1+), CD15-, CD25+, HLA-Dr+, CD71+.  
 origin

Clinics                Aggressive with skin nodes and extranodal involvement.

Cytogenetics        [t\(2;5\)\(p23;q35\)](#)

Genes                Fusion gene NPM-ALK; 2p23 -Nucleolar phosphoprotein- [NPM](#); 5q35 - Anaplastic lymphoma kinase- [ALK](#)

**Disease**            Intestinal T-cell lymphoma

Phenotype /  
 cell stem            TdT CD1a -, CD3+, CD8+, CD103+, CD4-, CD8-  
 origin

Clinics                Bone pain, coeliac disease, mesenteric nodes.

Genes                EBV genome present in mexican population but not in the europeans.

**Disease**            T-lymphoblastic Lymphoma/leukaemia (T-Lbly/T-ALL)

Phenotype /  
 cell stem            TDT+, CD1a+, CD7+, cytCD3+ or +/-, other T-cell antigens.Thymic  
 origin                uncommitted T-cell.

Clinics                Aggressive; course similar to ALL  
                          mediastinal mass, high WBC

Cytogenetics        del(6)(q21-q22)  
                          [t\(11;14\)\(p13;q11\)](#)  
                          [t\(1;14\)\(p34;q11\)](#); 1p34: [tal-1gene](#); 14q11: [TCR alpha](#)

Genes                TCR chain genes rearranged.

## **Bibliography**

**CD30-positive large cell lymphomas (Ki-1 lymphoma) are associated with chromosomal translocation involving 5q35.**

Mason DY, Bastard C, Rimokh R, Dastugue N, Huret JL, Kristoffersson U, Magaud J-P et al.

Br J Haematol 1990; 74: 161-168.

Medline [2156548](#)

**Chromosome abnormalities in adult T-cell Leukemia/Lymphoma : A karyotype review committee report.**

Kamada N, Sakurai M, Miyamoto K, Sanada I, Sadamori N, Fukuhara S, Abe S, Shiraishi Y, and Shimoyama M.  
Cancer Res 1992; 52:1481-1493.  
Medline [1540956](#)

**Clonal diseases of large granular lymphocytes.**

Loughran TP.  
Blood 1993; 82:1-14.  
Medline [8324214](#)

**Cytogenetic findings in peripheral T-cell lymphomas as a basis for distinguishing low- grade and high-grade lymphomas.**

Schlegelberger B, Himmler A, Godde E, Grote W, Feller AC, and Lennert K.  
Blood 1994; 83: 505-511.  
Medline [8286748](#)

**Detection of aberrant clones in nearly all cases of angioimmunoblastic lymphadenopathy with dysproteinemia type T-cell lymphoma by combined interphase and metaphase cytogenetics.**

Schlegelberger B, Zhang Y, Weber-Natthiesen K, Grote W.  
Blood 1994; 84:2640-2648.  
Medline [7919378](#)

**Consistent presence of isochromosome 7p in Hepatosplenic T-gamma/delta lymphoma: a new cytogenetic-clinicopathologic entity.**

Wang CC, Tien HF, Lin MT, Su IJ, Wang CH, Chuang SM, Shen MC, Liu CH.  
Genes Chromosom Cancer 1995;.12:161-164.  
Medline [7536454](#)

**Response to : "presence of t(2;5) in primary CD30+ cutaneous lymphoproliferative disorders".**

DeCocteau JF, Lowsky R, Kinney MC, Kadin ME..  
Blood 1996; 88:3251

**Classification of natural killer(NK) cell and NK- like T-cell malignancies.**

Jaffe E. .  
Blood 1996; 87: 1207-1210.  
Medline [8608206](#)

**Relationship of T- cell leukaemias with cerebriform nuclei to T-Prolymphocytic leukaemia: A cytogenetic analysis with in situ hybridisation.**

Brito-Babapulle V, Maljaie SH, Matutes E, Hedges M, Yuille M, Catovsky D.  
Br J Haematol 1997; 96 : 724-732.  
Medline [9074412](#)

**Identification of del(6)(q21q25) as a recurring chromosomal abnormality in putative NK cell lymphoma /leukaemia.**

Wong KF, Chan JKC, Kwong YL..

Br J Haematol 1997; 98:922-926.  
Medline [9326190](#)

**p53 allele deletion and protein expression occurs in the absence of p53 gene mutation in T-Prolymphocytic leukaemia and Sezary syndrome.**

Brito-Babapulle V, Hamoudi R, Matutes E, Watson S, Kaczmarek P, H.Maljaie, and Catovsky.

Br J Haematol 2000; 110:180-187.

Medline [10930996](#)

**The D13S25 locus mapping to 13q14.3 locus is deleted in T-Prolymphocytic leukemia.**

Brito-Babapulle V, Baou M, Atkinson S, Catovsky D.

Int.Natl.J Hematol.2000; Suppl1. 72:167 (abstract 15780).

**T-Cell Lymphoproliferative Disorders. Classification, Clinical and Laboratory Aspects..**

Matutes E.

Advances in Blood Disorders 2000. Ed.:A.Polliak. Harwood Academic Publishers

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