

The International Consensus Classification of Myeloid and Lymphoid Neoplasms

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Preface

Pathology classifications inexorably must evolve over time as we learn more about specific disease entities. New discoveries provide us information about disorders we thought we knew well but turn out to be more complicated than we had imagined. These discoveries and their influence on disease classification have helped lead to novel therapies and improved outcomes for patients afflicted with hematologic neoplasms. The International Consensus Classification (ICC) presented in this book was developed by over 200 experts in the continued pursuit of defining relevant disease entities. The ICC is the latest phase of a process that started following the publication of the Revised European-American Classification of Lymphoid Neoplasms (REAL classification) created by the International Lymphoma Study Group in 1994. The REAL classification differed from prior lymphoma classifications—which were generally put forth by individuals or small regional groups—and represented a consensus among a large cohort of pathologists from around the world. The classification incorporated all available data and techniques at that time, including morphology, immunophenotype, genetics, and clinical features, to define disease entities. Following the publication of the REAL classification, the World Health Organization (WHO) began planning for a 3rd edition series on *Pathology and Genetics of Tumours*, to be published by its affiliate International Agency for the Research on Cancer (IARC). This series of books ultimately became known as the *WHO Blue Books*. Prior to the 3rd edition, the WHO classification series were not widely used. The first edition contained only a list of disease names and images, while the 2nd edition did not include a book on hematopoietic neoplasms.

Les Sobin, a co-editor with Paul Kleihues of the 3rd edition Blue Book series, approached Elaine Jaffe, a co-author of the REAL Classification, to develop a similar classification for the WHO 3rd edition series. Dr Jaffe at that time was also President of the Society for Hematopathology. Dr Jaffe and the Society for Hematopathology Executive Committee recommended that the 3rd edition WHO effort also include myeloid neoplasms and acute leukemia; be overseen by the two major hematopathology societies, the Society for

Hematopathology (SH) and the European Association for Haematopathology (EA4HP); and incorporate input from a Clinical Advisory Committee (CAC) of leading international pathologists, oncologists, hematologists, and geneticists. Although a significant departure from prior hematopathology classifications, and unique among the 3rd edition series Blue Books, this approach was embraced by the WHO/IARC.

The first CAC meeting was held at Arlie House, Virginia in 1997. The organizers outlined key issues in advance for discussion among pathologists, scientists, and treating physicians to arrive at a broad consensus for diagnostic criteria and definitions of entities. The concept of provisional entities was introduced, in which there was compelling evidence to include in the classification but for which more data were needed before including as a full entity. After the CAC, the lead pathologists were tasked with resolving remaining areas of controversy and finalizing criteria. Once complete, the proceedings of the CAC meetings were published in leading journals, and the results published as the 3rd edition *WHO Classification of Tumors of Hematopoietic and Lymphoid Tissues* in 2001, with 75 contributing authors from across the globe. The 1997 CAC format set the standard for future classifications: similar CACs were held in 2007 and 2014 in Chicago and resulted in WHO Classification publications in 2008 (4th edition) and 2017 (revised 4th edition), respectively. The revised 4th edition, which had more than 200 contributors from 24 countries, was necessary due to delays in finalizing the remaining disease group volumes of the 4th edition Blue Book series, and reflected the significant scientific advances in hematologic neoplasms that had occurred in the interim.

The partnership between the hematopathology societies (including the CAC process that was devised and organized by the societies) and the WHO was an overwhelming success. The 3rd, 4th, and revised 4th editions of *Tumours of Haematopoietic and Lymphoid Tissues* Blue Books were best sellers and moved the field away from the historic classifications to a more widely accepted consensus approach.

As preparations began for the 5th edition WHO Blue Book, it became clear IARC intended to follow

a different process for this series, one that would not involve an initial CAC meeting; would not maintain continuity with the leadership of prior editions; and would not involve a collaboration with the SH and EA4HP. Accordingly, the leaders of the revised 4th edition WHO Blue Book decided to move ahead with a CAC at the University of Chicago in 2021 and to publish the ICC first in the journal *Blood* in 2022 and then in this book as a complete classification.

Thus, development of the ICC classification of hematologic neoplasms followed the same process used for the prior successful REAL classification and 3rd, 4th, and revised 4th edition WHO publications and is intended as an update in continuity with those classifications. The book follows a format similar to the prior Blue Books and is authored by hundreds of experts in the field, most of whom were involved with the revised 4th edition Blue Book. In recognition of their prior work, we thank the editors of those editions, Elias Campo, Nancy Lee Harris, Elaine S. Jaffe, Stefano A. Pileri, Harald Stein, Steven Swerdlow, Jürgen Thiele,

and James W. Vardiman, some of whom participated as co-authors of chapters in this book. The editors hope that this publication will assist pathologists in diagnosing and understanding cases in daily practice, as well as physicians who treat patients with hematologic malignancies. As the readers use the book, they will begin to identify new questions that future classifications will need to address, emphasizing the continued evolution and refinement of the way we categorize myeloid and lymphoid neoplasms.

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The International Consensus Classification of Myeloid and Lymphoid Neoplasms

Myeloproliferative Neoplasms

Chronic myeloid leukemia
Polycythemia vera
Essential thrombocythemia
Primary myelofibrosis
 Early/prefibrotic primary myelofibrosis
 Overt primary myelofibrosis
Chronic neutrophilic leukemia
Chronic eosinophilic leukemia, not otherwise specified (NOS)
Myeloproliferative neoplasm, unclassifiable

Myeloid/Lymphoid Neoplasms With Eosinophilia and Tyrosine Kinase Gene Fusions

Myeloid/lymphoid neoplasm with *PDGFRA* rearrangement
Myeloid/lymphoid neoplasm with *PDGFRB* rearrangement
Myeloid/lymphoid neoplasm with *FGFR1* rearrangement
Myeloid/lymphoid neoplasm with *JAK2* rearrangement
Myeloid/lymphoid neoplasm with *FLT3* rearrangement
Myeloid/lymphoid neoplasm with *ETV6::ABL1*

Mastocytosis

Myelodysplastic/Myeloproliferative Neoplasms

Chronic myelomonocytic leukemia
 Clonal monocytosis of undetermined significance
Atypical chronic myeloid leukemia
Myelodysplastic/myeloproliferative neoplasm with thrombocytosis and *SF3B1* mutation
Myelodysplastic/myeloproliferative neoplasm with ring sideroblasts and thrombocytosis, NOS
Myelodysplastic/myeloproliferative neoplasm, NOS
 Myelodysplastic/myeloproliferative neoplasm with isolated isochromosome (17q)

Myelodysplastic Syndromes and Premalignant Clonal Cytopenias

Clonal cytopenia of undetermined significance and other clonal cytopenias
Myelodysplastic syndrome with mutated *SF3B1*
Myelodysplastic syndrome with del(5q)
Myelodysplastic syndrome with mutated *TP53*
Myelodysplastic syndrome, NOS (MDS, NOS)
 MDS, NOS without dysplasia
 MDS, NOS with single lineage dysplasia
 MDS, NOS with multilineage dysplasia
Myelodysplastic syndrome with excess blasts
Myelodysplastic syndrome/acute myeloid leukemia (MDS/AML)
 MDS/AML with mutated *TP53*
 MDS/AML with myelodysplasia-related gene mutations
 MDS/AML with myelodysplasia-related cytogenetic abnormalities
 MDS/AML, NOS

Pediatric or Germline Mutation–Associated Disorders

Juvenile myelomonocytic leukemia
Juvenile myelomonocytic leukemia-like neoplasms
Noonan syndrome–associated myeloproliferative disorder
Refractory cytopenia of childhood
Hematologic neoplasms with germline predisposition

Acute Myeloid Leukemias

Acute promyelocytic leukemia (APL) with t(15;17)(q24.1;q21.2)/*PML::RARA*
APL with other *RARA* rearrangements
AML with t(8;21)(q22;q22.1)/*RUNX1::RUNX1T1*
AML with inv(16)(p13.1q22) or t(16;16)(p13.1;q22)/*CBFB::MYH11*
AML with t(9;11)(p21.3;q23.3)/*MLLT3::KMT2A*
AML with other *KMT2A* rearrangements
AML with t(6;9)(p22.3;q34.1)/*DEK::NUP214*
AML with inv(3)(q21.3q26.2) or t(3;3)(q21.3;q26.2)/*GATA2; MECOM(EV1)*
AML with other *MECOM* rearrangements
AML with other rare recurring translocations
AML with t(9;22)(q34.1;q11.2)/*BCR::ABL1*
AML with mutated *NPM1*
AML with in-frame bZIP *CEBPA* mutations
AML and MDS/AML with mutated *TP53*
AML and MDS/AML with myelodysplasia-related gene mutations
AML with myelodysplasia-related cytogenetic abnormalities
AML NOS
Myeloid Sarcoma

Myeloid Proliferations Associated With Down Syndrome

Blastic Plasmacytoid Dendritic Cell Neoplasm

Acute Leukemia of Ambiguous Lineage

Mixed phenotype acute leukemia (MPAL) with t(9;22)(q34.1;q11.2); *BCR::ABL1*
MPAL, with t(v;11q23.3); *KMT2A* rearranged
MPAL with *ZNF384* rearrangement
MPAL with *BCL11B* activation
MPAL with defining immunophenotypic changes
Acute undifferentiated leukemia
Acute leukemia of ambiguous lineage, NOS

B-Lymphoblastic Leukemias/Lymphomas (B-ALL)

B-ALL with recurrent genetic abnormalities
 B-ALL with t(9;22)(q34.1;q11.2)/*BCR::ABL1*
 with lymphoid only involvement
 with multilineage involvement
 B-ALL with t(v;11q23.3)/*KMT2A* rearranged
 B-ALL with t(12;21)(p13.2;q22.1)/*ETV6::RUNX1*
 B-ALL, hyperdiploid
 B-ALL, low hypodiploid

B-ALL, near haploid
 B-ALL with t(5;14)(q31.1;q32.3)/*IL3::IGH*
 B-ALL with t(1;19)(q23.3;p13.3)/*TCF3::PBX1*
 B-ALL, *BCR::ABL1*-like, ABL-1 class rearranged
 B-ALL, *BCR::ABL1*-like, JAK-STAT activated
 B-ALL, *BCR::ABL1*-like, NOS
 B-ALL with *iAMP21*
 B-ALL with *MYC* rearrangement
 B-ALL with *DUX4* rearrangement
 B-ALL with *MEF2D* rearrangement
 B-ALL with *ZNF384* rearrangement
 B-ALL with *NUTM1* rearrangement
 B-ALL with *HLF* rearrangement
 B-ALL with *UBTF::ATXN7L3/PAN3,CDX2* ("CDX2/UBTF")
 B-ALL with PAX5 P80R
 B-ALL with IKZF1 N159Y
 B-ALL, NOS
 B-ALL, provisional entities*

T-Lymphoblastic Leukemias/Lymphomas

Early T-cell precursor ALL with *BCL11B*-activated
 Early T-cell precursor ALL, NOS
 T-ALL, NOS
 T-ALL, provisional entities*

NK-Lymphoblastic Leukemia/Lymphoma*

Mature B-Cell Neoplasms

Chronic lymphocytic leukemia/small lymphocytic lymphoma
 Monoclonal B-cell lymphocytosis
 CLL type
 Non-CLL type
 B-cell prolymphocytic leukemia
 Splenic marginal zone lymphoma
 Hairy cell leukemia
 Splenic B-cell lymphoma/leukemia, unclassifiable*
 Splenic diffuse red pulp small B-cell lymphoma*
 Hairy cell leukemia-variant*
 Lymphoplasmacytic lymphoma
 Waldenström macroglobulinemia
 IgM monoclonal gammopathy of undetermined significance (MGUS)
 IgM MGUS, plasma cell type
 IgM MGUS, NOS
 Primary cold agglutinin disease
 Heavy chain diseases
 Mu heavy chain disease
 Gamma heavy chain disease
 Alpha heavy chain disease
 Plasma cell neoplasms
 Non-IgM monoclonal gammopathy of undetermined significance
 Multiple myeloma (Plasma cell myeloma)
 Multiple myeloma, NOS
 Multiple myeloma with recurrent genetic abnormalities
 Multiple myeloma with *CCND* family translocation
 Multiple myeloma with *MAF* family translocation
 Multiple myeloma with *NSD2* translocation
 Multiple myeloma with hyperdiploidy
 Solitary plasmacytoma of bone
 Extraosseous plasmacytoma

Monoclonal immunoglobulin deposition diseases
 Immunoglobulin light chain amyloidosis (AL)
 Localized AL amyloidosis
 Light chain and heavy chain deposition disease
 Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma)
 Primary cutaneous marginal zone lymphoproliferative disorder
 Nodal marginal zone lymphoma
 Follicular lymphoma
 In situ follicular neoplasia
 Duodenal-type follicular lymphoma
 BCL2-R-negative, CD23-positive follicle center lymphoma*
 Primary cutaneous follicle center lymphoma
 Pediatric-type follicular lymphoma
 Testicular follicular lymphoma
 Large B-cell lymphoma with *IRF4* rearrangement
 Mantle cell lymphoma
 In situ mantle cell neoplasia
 Leukemic nonnodal mantle cell lymphoma
 Diffuse large B-cell lymphoma (DLBCL), NOS
 Germinal center B-cell subtype
 Activated B-cell subtype
 Large B-cell lymphoma with 11q aberration*
 Nodular lymphocyte predominant B-cell lymphoma
 T cell/histiocyte rich large B-cell lymphoma
 Primary DLBCL of the central nervous system
 Primary DLBCL of the testis
 Primary cutaneous DLBCL, leg type
 Intravascular large B-cell lymphoma
 Human herpesvirus 8 (HHV-8) and Epstein-Barr virus (EBV)-negative primary effusion-based lymphoma*
 EBV-positive mucocutaneous ulcer
 EBV-positive DLBCL, NOS
 DLBCL associated with chronic inflammation
 Fibrin-associated DLBCL
 Lymphomatoid granulomatosis
 EBV-positive polymorphic B-cell lymphoproliferative disorder, NOS
 ALK-positive large B-cell lymphoma
 Plasmablastic lymphoma
 HHV-8-associated lymphoproliferative disorders
 Multicentric Castlemann disease
 HHV-8-positive germinotropic lymphoproliferative disorder
 HHV-8-positive DLBCL, NOS
 Primary effusion lymphoma
 Burkitt lymphoma
 High-grade B-cell lymphoma with *MYC* and *BCL2* rearrangements
 High-grade B-cell lymphoma with *MYC* and *BCL6* rearrangements*
 High-grade B-cell lymphoma, NOS
 Primary mediastinal large B-cell lymphoma
 Mediastinal gray zone lymphoma

Classic Hodgkin Lymphoma

Nodular sclerosis classic Hodgkin lymphoma
 Lymphocyte-rich classic Hodgkin lymphoma
 Mixed cellularity classic Hodgkin lymphoma
 Lymphocyte-depleted classic Hodgkin lymphoma

Mature T-Cell and NK-Cell Neoplasms

T-cell prolymphocytic leukemia
 T-cell large granular lymphocytic leukemia
 Chronic lymphoproliferative disorder of NK cells*
 Adult T-cell leukemia/lymphoma
 EBV-positive T/NK lymphoproliferative disorders of childhood
 Hydroa vacciniforme LPD
 Classic
 Systemic
 Severe mosquito bite allergy
 Chronic active EBV disease (T-cell and NK-cell phenotype)
 Systemic EBV-positive T-cell lymphoma of childhood
 Extranodal NK/T-cell lymphoma, nasal type
 Aggressive NK-cell leukemia
 Primary nodal EBV-positive T/NK-cell lymphoma*
 Enteropathy-associated T-cell lymphoma
 Type II refractory celiac disease
 Monomorphic epitheliotropic intestinal T-cell lymphoma
 Intestinal T-cell lymphoma, NOS
 Indolent clonal T-cell lymphoproliferative disorder of the gastrointestinal tract
 Indolent NK-cell lymphoproliferative disorder of the gastrointestinal tract
 Hepatosplenic T-cell lymphoma
 Mycosis fungoides
 Sézary syndrome
 Primary cutaneous CD30-positive T-cell lymphoproliferative disorders
 Lymphomatoid papulosis
 Primary cutaneous anaplastic large cell lymphoma
 Primary cutaneous CD4-positive small/medium T-cell lymphoproliferative disorder
 Subcutaneous panniculitis-like T-cell lymphoma
 Primary cutaneous gamma-delta T-cell lymphoma
 Primary cutaneous acral CD8-positive T-cell lymphoproliferative disorder
 Primary cutaneous CD8-positive aggressive epidermotropic cytotoxic T-cell lymphoma
 Peripheral T-cell lymphoma, NOS

* Provisional entity

Follicular helper T-cell lymphoma
 Follicular helper T-cell lymphoma, angioimmunoblastic type
 (Angioimmunoblastic T-cell lymphoma)
 Follicular helper T-cell lymphoma, follicular type
 Follicular helper T-cell lymphoma, NOS
 Anaplastic large cell lymphoma, ALK positive
 Anaplastic large cell lymphoma, ALK negative
 Breast implant-associated anaplastic large cell lymphoma

Immunodeficiency-Associated Lymphoproliferative Disorders

Posttransplant lymphoproliferative disorders (PTLD)
 Nondestructive PTLD
 Plasmacytic hyperplasia PTLD
 Infectious mononucleosis PTLD
 Florid follicular hyperplasia PTLD
 Polymorphic PTLD
 Monomorphic PTLD (B-cell and T/NK-cell types)
 Classic Hodgkin lymphoma PTLD
 Other iatrogenic immunodeficiency-associated lymphoproliferative disorders

Histiocytic and Dendritic Cell Neoplasms

Histiocytic sarcoma
 Langerhans cell histiocytosis
 Langerhans cell sarcoma
 Indeterminate dendritic cell histiocytosis
 Interdigitating dendritic cell sarcoma
 ALK-positive histiocytosis
 Disseminated juvenile xanthogranuloma
 Erdheim-Chester disease
 Rosai-Dorfman-Destombes disease
 Follicular dendritic cell sarcoma
 Fibroblastic reticular cell sarcoma
 EBV-positive inflammatory follicular dendritic cell/fibroblastic reticular cell tumor

