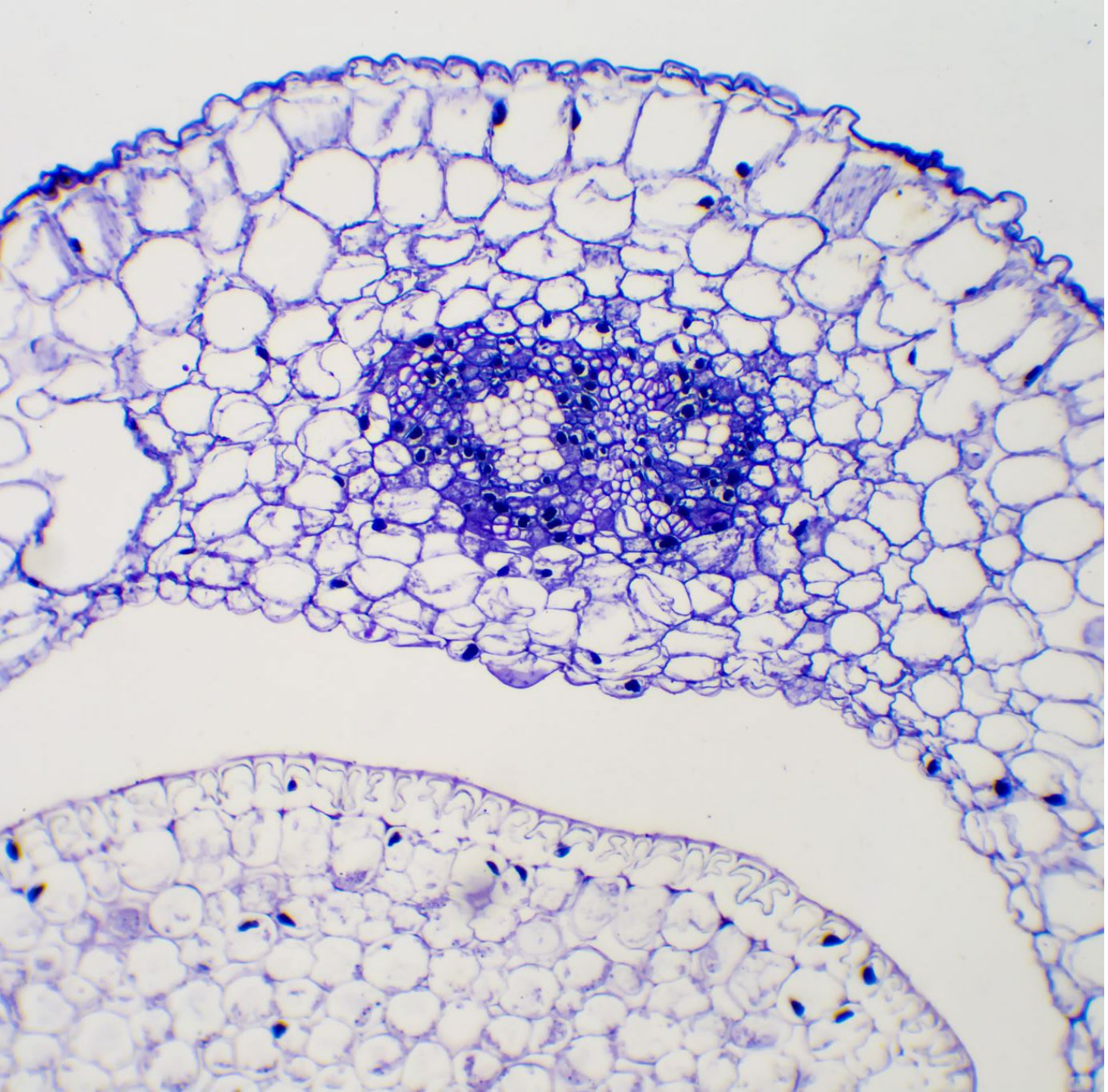


HEMATOPOIETIC part I

FCDS

MARCH 18/2026

BETTY MALANOWSKI,

HEMATOPOLYMPHOID 2026

-Last MAJOR update was in 2014

-Who 5th Edition Hematolymphoid Tumors 5th Edition Volume 11 (2 volumes instead of 1)

Part A

1. General introduction to haematolymphoid tumors
2. Myeloid proliferations and neoplasms
3. Histiocytic/dendritic cell neoplasms

Part B

1. B-cell lymphoid proliferations and lymphomas
2. T-cell and NK-cell lymphoid proliferations and lymphomas
3. Stroma-derived neoplasms of lymphoid tissues
4. Genetic tumour syndromes associated with haematolymphoid tumors

MANUAL changes 2026

2026 Changes

Released November 26, 2025

Hematopoietic and Lymphoid Neoplasm Coding Manual

- No new modules or rules, no modules or rules deleted.
- Overhaul of structure (content not changed).
- New definitive terminology for assigning histology (See Histology Coding Instructions, #3).
- New section introducing Hematopoietic neoplasms (overview of all the groups, lineage tables).
- Overhaul of several sections, including Steps for Using the Heme Manual and DB, Multiple Primaries Calculator, Transformations, Primary site coding instructions (introduction, not the PH rules), and Treatment.
- New section added for Coding Mets at Dx fields for Hematopoietic neoplasms.
- Clarifications added on when and how to code histology based on positive immunophenotyping or genetics.
- Updated lineage tables based on the WHO Blue Book, 5th edition for Hematolymphoid Tumors (2024).
- Primary site assignments for each of the histologies have been removed from the Heme manual. This information is in the Heme DB.
- Grade rules removed. The grade rules are no longer applicable for Heme neoplasms. For cases diagnosed prior to 2018, you can use the Heme DB to help assign grade. Make sure “Help me code for diagnosis year” is 2010-2017.
- New examples added (courtesy of Ask a SEER Registrar questions).
- New Appendix E: List of terms added to the Hematopoietic Database from the WHO 5th edition of Hematolymphoid Neoplasms.

DATABASE Changes 2026

2026 Changes
Released November 26, 2025

Hematopoietic and Lymphoid Neoplasm Database

- New terms from the WHO Blue Book, 5th edition for Hematolymphoid Tumor (2024) added (see Appendix E: Terms added to the Hematopoietic Database from the WHO 5th edition of Hematolymphoid Tumors for a complete list).
- Immunophenotyping and genetics updated based on the WHO Blue Book, 5th edition for Hematolymphoid Tumor. Only the most common immunophenotyping and genetics are included.
- Definitive diagnosis field redone for all histologies (note: field previously included procedures, which were removed).
- Abstractor notes updated based on the WHO Blue Book, 5th edition for Hematolymphoid Tumor (2024).
- Definitions updated based on the WHO Blue Book, 5th edition for Hematolymphoid Tumor (2024).
- Grade field hidden for cases diagnosed 2018+. For cases diagnosed prior to 2018 use "Help me code for diagnosis year" field and change the year to 2017.
- Same primaries changed for Acute Lymphoblastic Leukemias (9811-9819). Any combination of these histologies is the same primary.
- ICD-O-1 and ICD-O-2 fields suppressed for all years.
- ICD-9-CM field hidden for cases diagnosed 2015+. ICD-10-CM codes updated (when applicable).
- Resources updated.

https://seer.cancer.gov/tools/heme/update.html

2025 Changes
Released November 26, 2024

9738/3	Preferred	HHV8-positive diffuse large B-cell lymphoma
9738/3	Synonym	Large B-cell lymphoma arising in HHV8-associated multicentric Castleman disease

- 9738/3 was made obsolete starting 2021 and changed to 9738/1. This was done in error. 9738/3 is still a valid code (2010+), along with 9738/1 (2021+). This is a very rare lymphoma, so registrars will not be required to go back and look for missed cases.
- Post Transplant Lymphoproliferative Disorder (PTLD) was previously reportable as 9971/3 for 2010-2020 when it was the only diagnosis. In 2021, based on the 4th edition of WHO Hematopoietic Blue Book, PTLD became 9971/1, where it was only reportable if it occurred in the brain. Starting in 2025, PTLD as the only diagnosis will become a /3 (malignant) again and will be reportable for all cases.
- A new SSDI has been added to several schemas (Lymphoma, Lymphoma-CLL/SLL, Primary Cutaneous Lymphoma (excluding MF/SS), Plasma Cell Disorders, Plasma Cell Myeloma) for when a PTLD is diagnosed WITH a lymphoma, plasmacytoma, or multiple myeloma. (See the Hematopoietic Manual, Rules M14, PH1). See the SSDI manual for further instructions on coding the new SSDI.

9971/1	Preferred	Post transplant lymphoproliferative disorder, NOS
9971/1	Synonym	PTLD, NOS
9971/1	Related	Polymorphic post transplant lymphoproliferative disorder

9971/3 ✓

9971/ is NOT Reportable prior to 2010
For cases diagnosed for the years 2021-2024 is only reportable for CNS/brain as 9971/1
Starting 1/2/2025 PTLD will be reportable for any site and is coded 9971/3

2026 ICD-O-3.2

New terms/codes for the hemato-lymphoid system are included in the 2026 ICD-O-3.2

ICD-O-3 IMPLEMENTATION GUIDELINES

ICD O 2026 | Previous Guidelines

ICD-O-3.2 Implementation Documents for Implementation in 2026

- 2026 ICD-O-3.2 Coding Guidelines – 4/18/2025
- 2026 ICD-O-3.2 Table 1 Numeric – 4/18/2025
- 2026 ICD-O-3.2 Table 2 Alpha Table – 4/23/2025

WHO IARC ICD-O-3.2

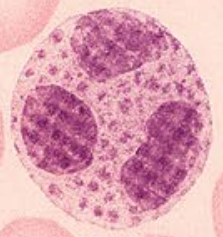
- WHO IARC ICD-O-3.2 Excel Table – 1/1/2021 – *The date North American registries adopted 3.2 for use.*

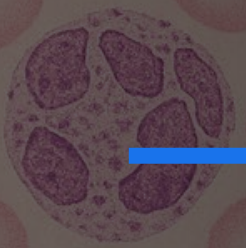
Annotated Histology List

- Annotated Histology List Description and Disclaimer 5/1/2025
- Annotated Histology List – *9/3/2025 (Change from 5/1/2025 posting-8343/2 annotation updated)*

https://www.naaccr.org/icdo3/

Hematolymphoid BASICS





BLOOD AND LYMPHOID TISSUE

Hematopoiesis (Blood formation)

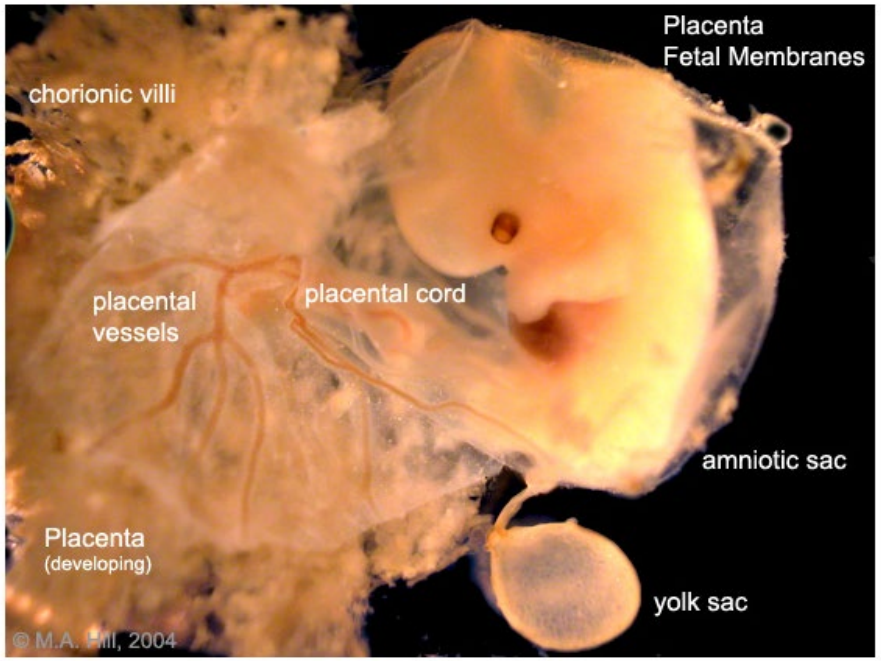
Starts in the YOLK SAC: First blood cells are made in Yolk Sac around 2-3 weeks of gestation
Shifts to the fetal LIVER (6-7 week) and SPLEEN (week 8 to month 7).
Then the bone marrow takes over as the primary site.

Later the **spleen** and **liver** only act as backup (**extramedullary hematopoiesis**) as the bone marrow becomes the primary.

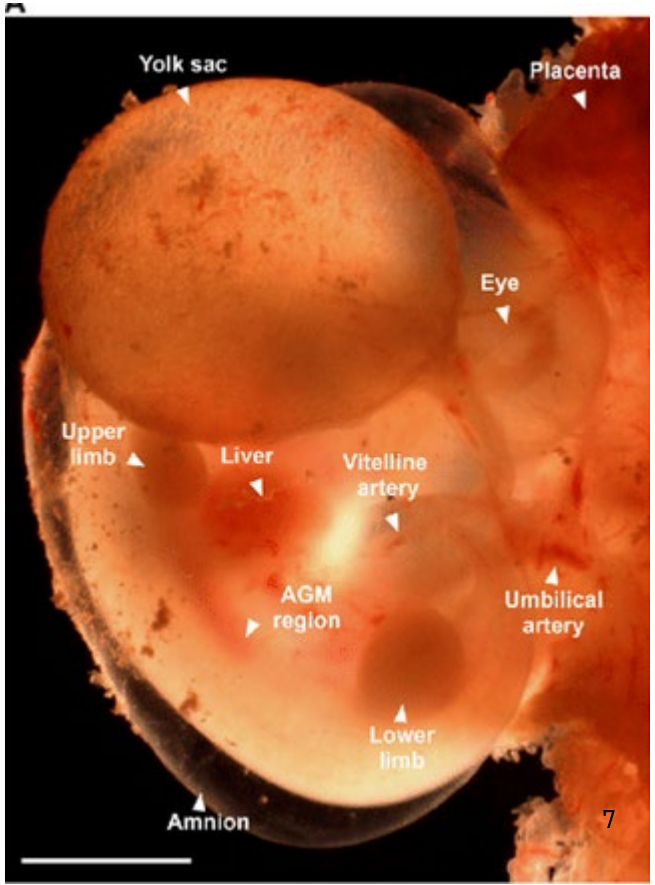
Developing Human Embryo - 4 Weeks from Fertilization

d Swedish photographer Lennart Nilsson

CLO



embryology.med.unsw.edu.au (6 weeks human embryo)

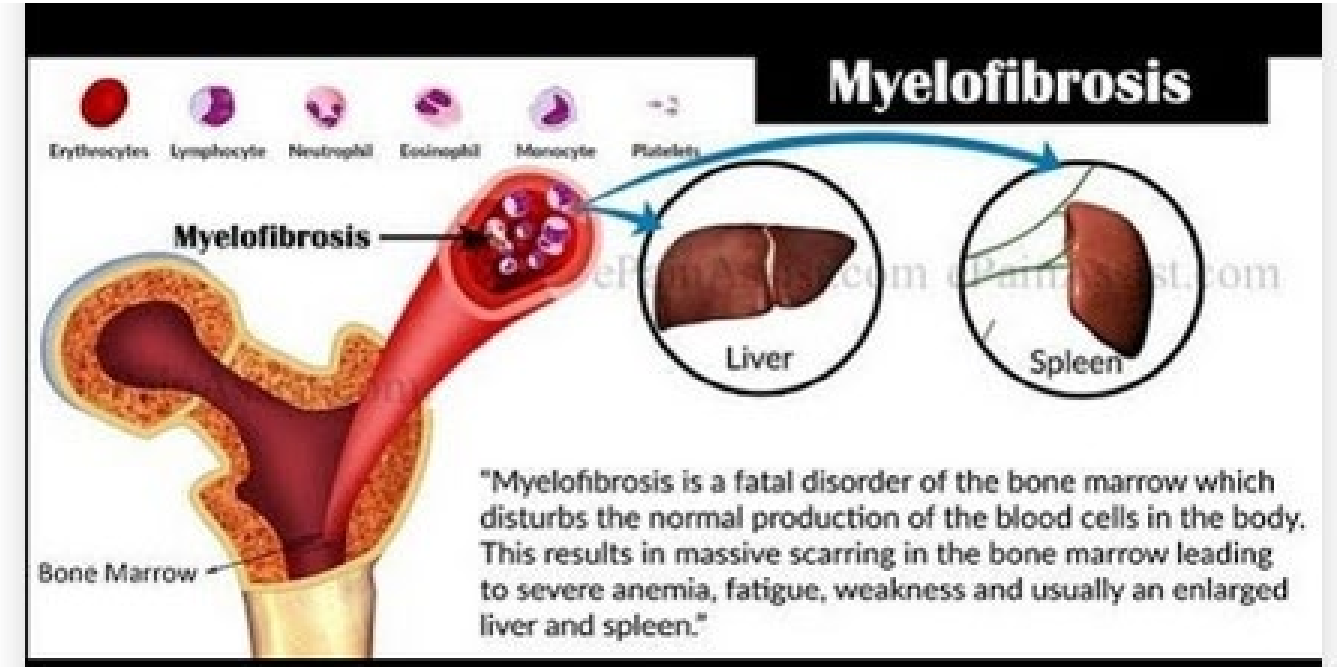


What is Myelofibrosis?

Myelofibrosis occurs when scar tissue (**fibrosis**) replaces bone marrow.
It can be:

- **Primary** (Primary Myelofibrosis)
- **Secondary**

  Secondary to conditions as MM, Leukemia, Lymphoma, Post Polycythemia Vera Myelofibrosis, Post Essential Thrombocythemia Myelofibrosis, or other diseases no related to cancer.



https://www.youtube.com/watch?v=AxbdizRHaCg

EXTRAMEDULLARY Hematopoiesis: Is the production of blood cells outside the bone marrow, primarily occurring in the **spleen** and **liver** during fetal development and under pathological conditions in adults.

HEMATOPOIETIC System and LYMPHOID system

Lymphocytes B and Lymphocytes T

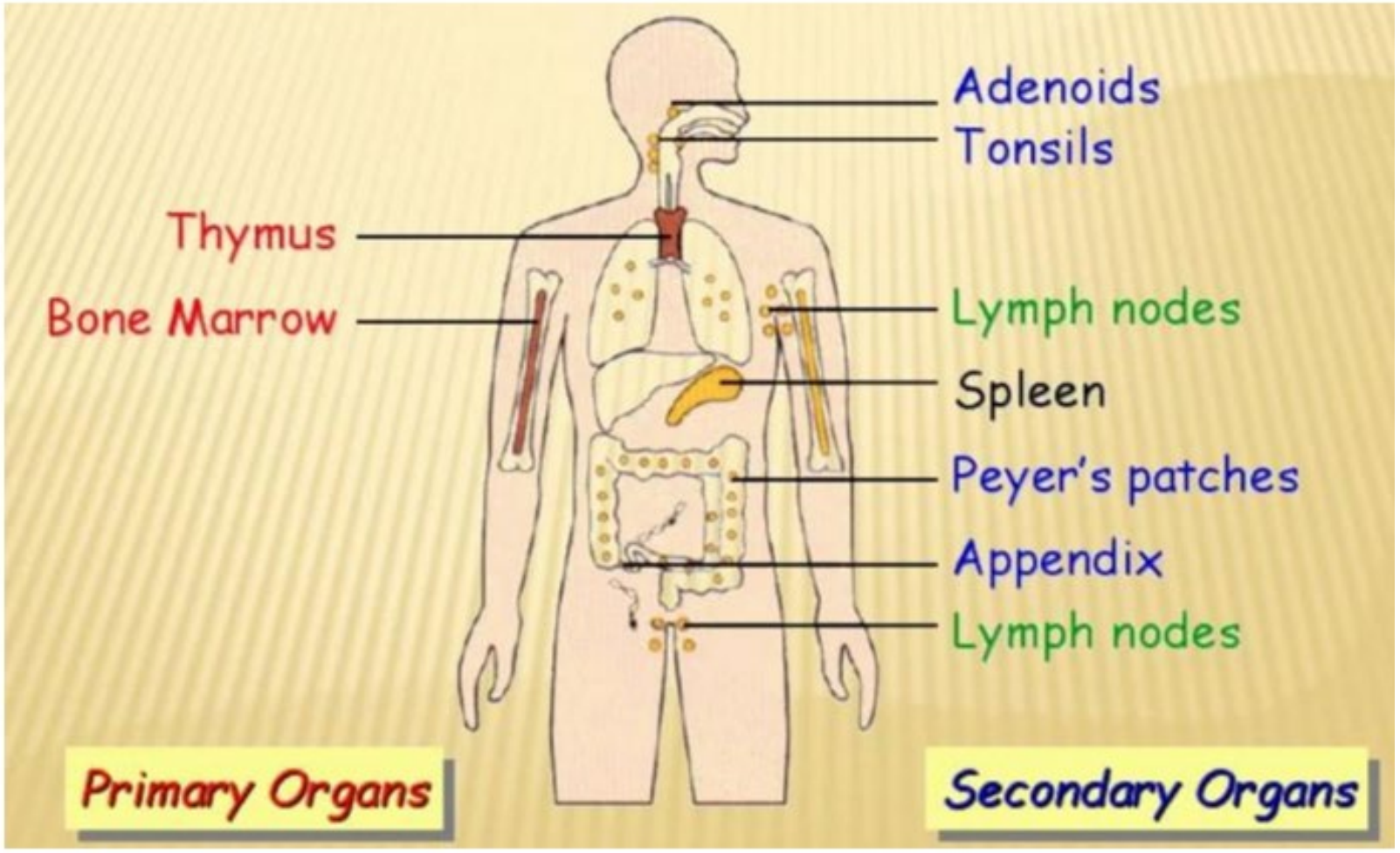
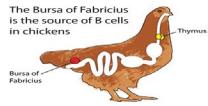


Diagram of the Immune System; the primary and secondary lymphoid organs (from SlideShare).

PRIMARY lymphoid organs (where lymphocytes B and T develop and mature)
-Thymus
-Bone Marrow

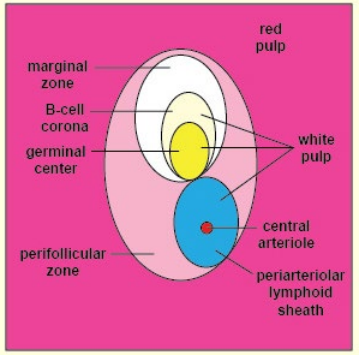
-Bone marrow (Lymphocytes B are **named after** the Bursa of Fabricius, a bird organ where they were first discovered and found to mature there. But in humans, cells mature in the BM).



-Thymus (school where lymphocytes T learn to distinguish b/w an invader/foreign and between our own cells).

SECONDARY lymphoid organs including **MALT**

(where mature lymphocytes encounter pathogens and mount immune responses).



SPLEEN

RED PULP: Filters blood, removing old or damaged cells, Also tries to filter out cancer cells. Due to the increased demand for blood cell filtration and extramedullary hematopoiesis primary myeloid.

Splenomegaly

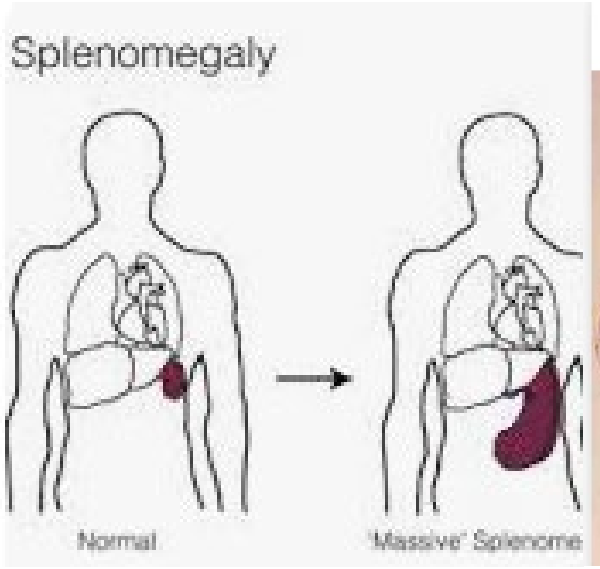
Due to the increased demand for blood cell filtration and extramedullary hematopoiesis when trying to compensate for a failing bone marrow. Splenic **infiltration** does not mean it is the primary site!

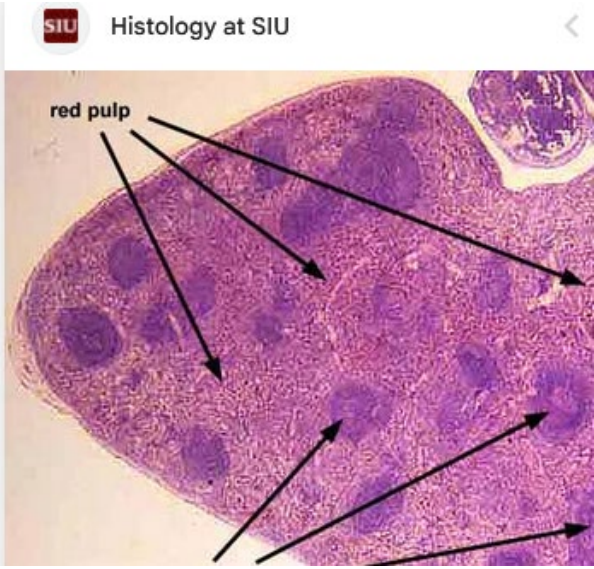
WHITE PULP: Immune system support, acts as a defense center producing Lymphocytes (WBCs) and antibodies.

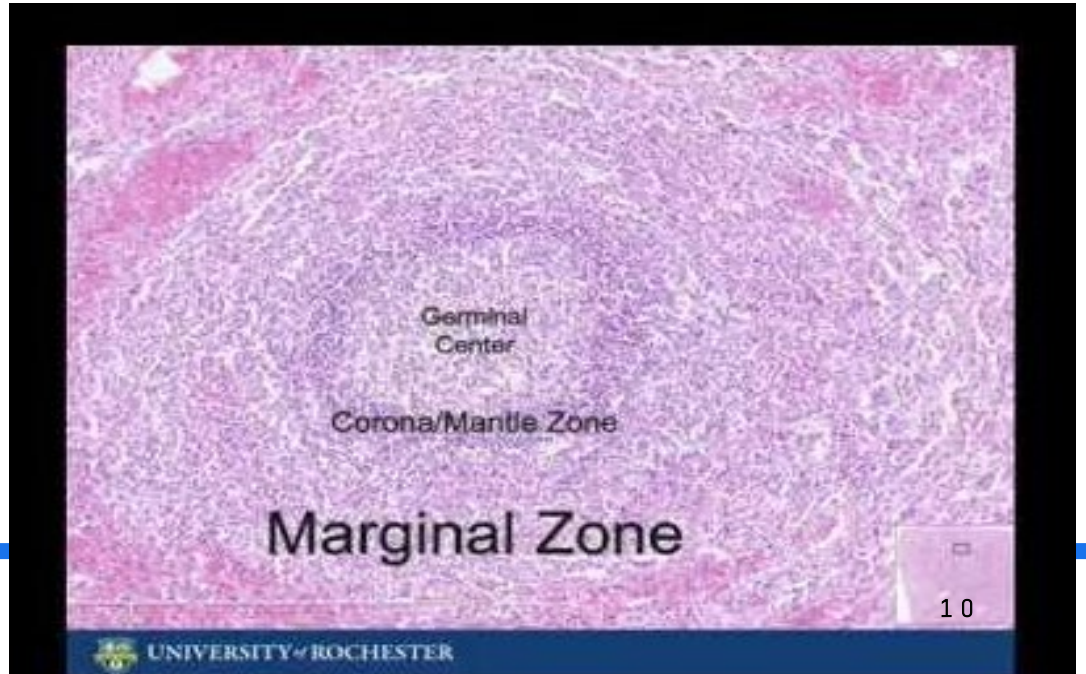
Beware: Primary site is not usually the Spleen unless it is SPLENIC LYMPHOMA, Splenic MARGINAL zone lymphoma,

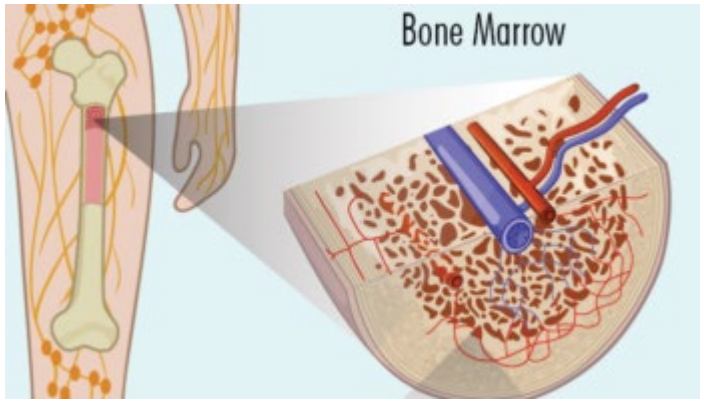
Hepato**Splenic** lymphoma …

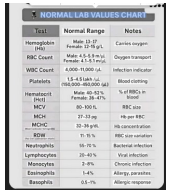
In cancer the liver and spleen may grow so much to the point to give abdominal discomfort.











BABY cells

Bone marrow

10 µm

Multipotential hematopoietic stem cell (Hemocytoblast)

Common myeloid progenitor

Common lymphoid progenitor

Megakaryoblast

Promegakaryocyte

Megakaryocyte

Proerythroblast (Pronormoblast)

Basophilic erythroblast

Polychromatic erythroblast

Orthochromatic erythroblast (Normoblast)

Polychromatic erythtocyte [1] (Reticulocyte)

Myeloblast

B. promyelocyte

B. myelocyte

B. metamyelocyte

B. band

N. promyelocyte

N. myelocyte

N. metamyelocyte

N. band

E. promyelocyte

E. myelocyte

E. metamyelocyte

E. band

Monoblast

Promonocyte

Lymphoblast

Prolymphocyte

Stem cells

Committed progenitor cells

Mature cells

Blood

Tissue

Thrombocytes (Platelets)

Thrombopoiesis

Erythrocyte [2] (Red blood cell)

Erythropoiesis

Basophil

Neutrophil

Eosinophil

Granulopoiesis

Monocyte

Monocytopoiesis

Small lymphocyte [4]

Natural killer cell (Large granular lymphocyte)

B lymphocyte

T lymphocyte

Lymphopoiesis

Plasma cell

Lymphoid dendritic cell [3]

Mast cell

Macrophage

Myeloid dendritic

Conclusion: The Critical Role of CBC in Leukemia Detection

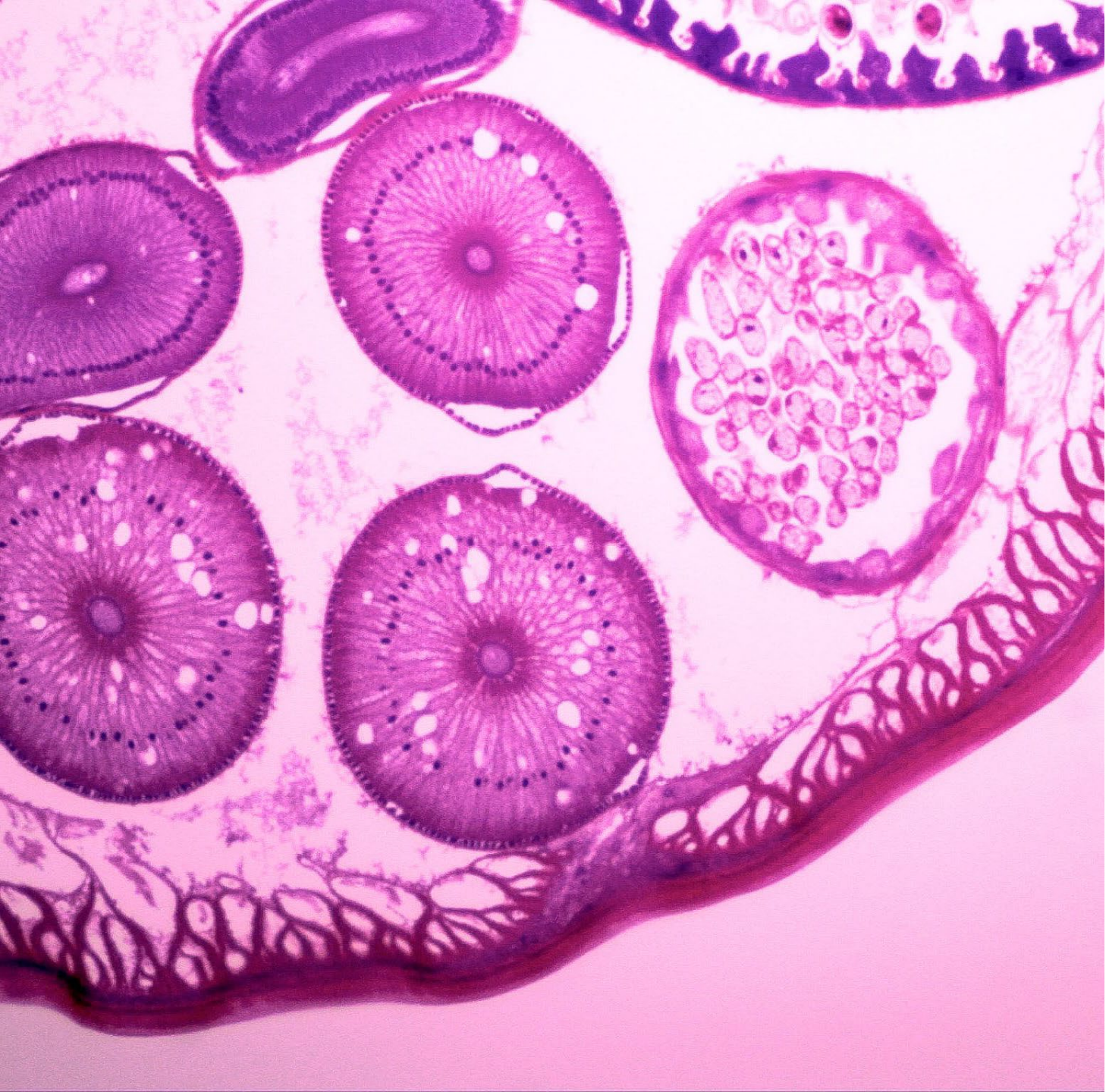
A Complete Blood Count (CBC) test is key in finding leukemia. It gives clues that might suggest leukemia. These clues come from blood cell counts and how the cells look.

The CBC helps spot odd white blood cell counts and blast cells. It also finds other issues with blood cell making. These signs can lead to more tests like bone marrow biopsies and molecular tests to confirm leukemia.

The CBC is a vital first step in finding leukemia. It helps doctors spot leukemia early. This way, they can start treatment right away.

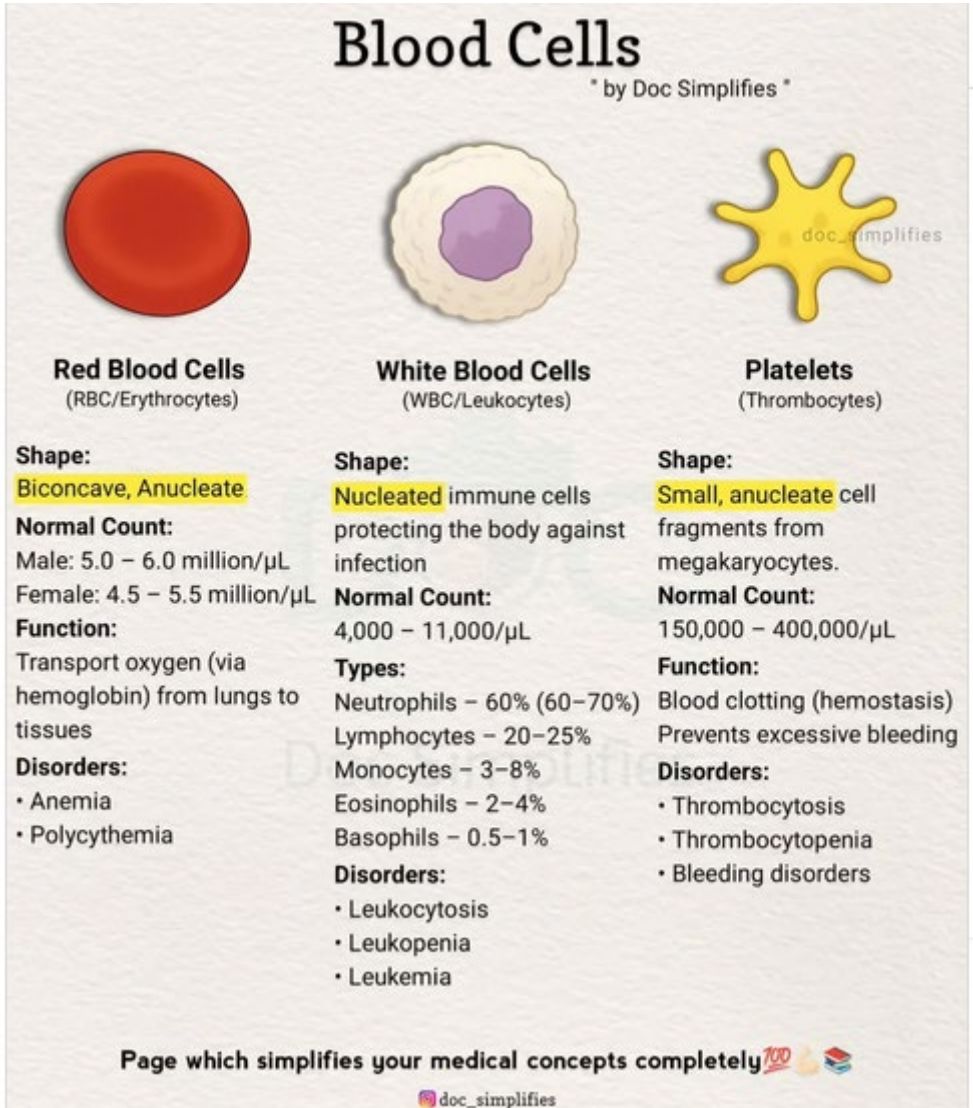
https://int.livhospital.com

Mast cell leukemia	9742/3



MDS, MPN, LEUKEMIAS, LYMPHOMAS…

Myelodysplastic syndromes formerly known as "preleukemia" or "lazy bone marrow" and can progress to Acute Myeloid Leukemia

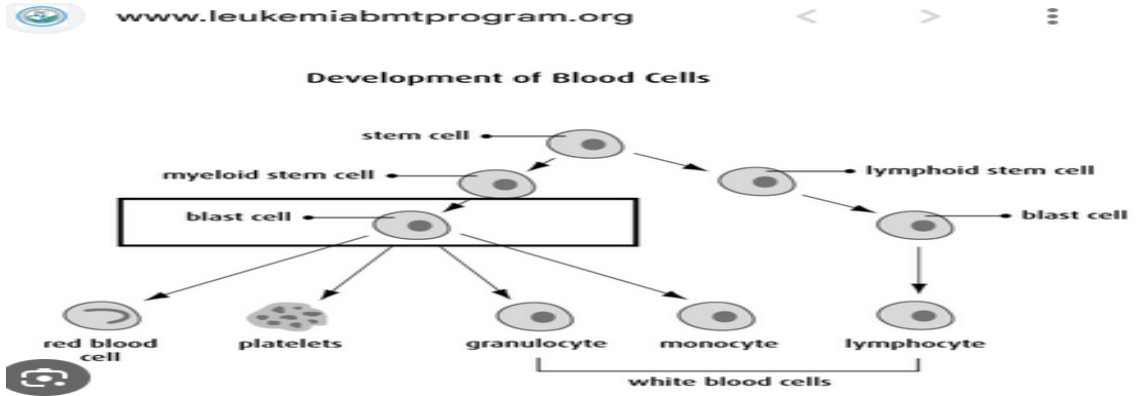


Bone marrow stem cells (too many BLASTS) fail to develop into healthy, mature blood cells, leading to:

LOW COUNTS (-penia) of functional RBC, WBC, or PLATELETS.

Leading to ANEMIA, INFECTIONS, and BLEEDING Problems.

And the signs and symptoms are related to anemia, infections and bleeding. But it can be no symptoms!



Myelo-Dysplastic Sx Reportability

Based on the implementation of the **International Classification of Diseases for Oncology, 3rd Edition (ICD-O-3)**, which went into effect for cases diagnosed in **2001 and later**, the behavior code for Myelodysplastic Syndromes (MDS) was updated!

from **/1 (borderline malignancy or low malignant potential)**

to **/3 (malignant primary site)**.

Section 30
Myelodysplastic Syndromes (MDS), Chronic Myeloproliferative Disorders (CMD), and Chronic Myelomonocytic Leukemia (CMML)

This section includes Myelodysplastic Syndromes (MDS), Chronic Myeloproliferative Disorders (CMD), and Chronic Myelomonocytic Leukemia (CMML). In the 3rd edition of the International Classification of Diseases for Oncology, the behavior code of myelodysplastic syndromes and chronic myeloproliferative disorders was changed from /1, borderline malignancy or low malignant potential, to /3, malignant primary site. These diagnoses became reportable for cases diagnosed in 2001+ when ICD-O-3 went into effect. CMML has been reportable for all years. CMML is included here and in the leukemia section.

Therefore, MDS cases before 2001 are NOT Reportable to FCDS.

All leukemias, myelodysplastic syndromes and chronic myeloproliferative diseases are assigned primary site bone marrow C421.
There are **NO** exceptions. This rule was implemented in ICD-O-2 in 1992.

MDS vs MYELOPROLIFERATIVE Neoplasms

MDS: Overproduction of **IMMATURE** and dysfunctional cells (**blasts**)

MPNs: Too many myeloid **MATURE** cells

Both can transform into Acute Myeloid Leukemia

-MYELODYSPLASTIC SYNDROMES (MDS); Usually Low counts (cyto-penias) of functional cells.

-MYELOPROLIFERATIVE Neoplasms (MPNs): cell counts OFTEN HIGH

- Polycythemia Vera (high number of **RBCs**)
  - hypercoagulability, collect **Phlebotomy ONLY** for **polycythemia vera** (2026) in OTHER THERAPY. Aspirin is not valid for PV.
- Essential Thrombocythemia (high number of **Platelets**)
  - Aspirin in the range of 70-100mgs (low dose) should be coded in OTHER THERAPY ONLY for this condition

Collect **phlebotomy ONLY** for polycythemia vera (9950/3).

Collect **blood-thinners** and/or **anti-clotting agents ONLY** for essential thrombocythemia (9962/3).

Aspirin is an ANTI-Platelets .if you see higher dosages like 160 mgs or higher dosages probably should not be coded

- Primary Myelofibrosis (also involves fibrosis)

Table B5: Myelodysplastic/myeloproliferative (MDS/MPN)

WHO Preferred Term	ICD-O
Chronic myelomonocytic leukemia • Myelodysplastic chronic myelomonocytic leukemia • Myeloproliferative chronic myelomonocytic leukemia	9945/3
Myelodysplastic/myeloproliferative neoplasm with neutrophilia	9876/3
Myelodysplastic/myeloproliferative neoplasm with *SF3B1* mutation and thrombocytosis (See also Table B4: Myelodysplastic neoplasms)	9982/3
Myelodysplastic/myeloproliferative neoplasm, NOS (unclassifiable) (See also Table B2: Myeloproliferative neoplasms (MPNs)	9975/3

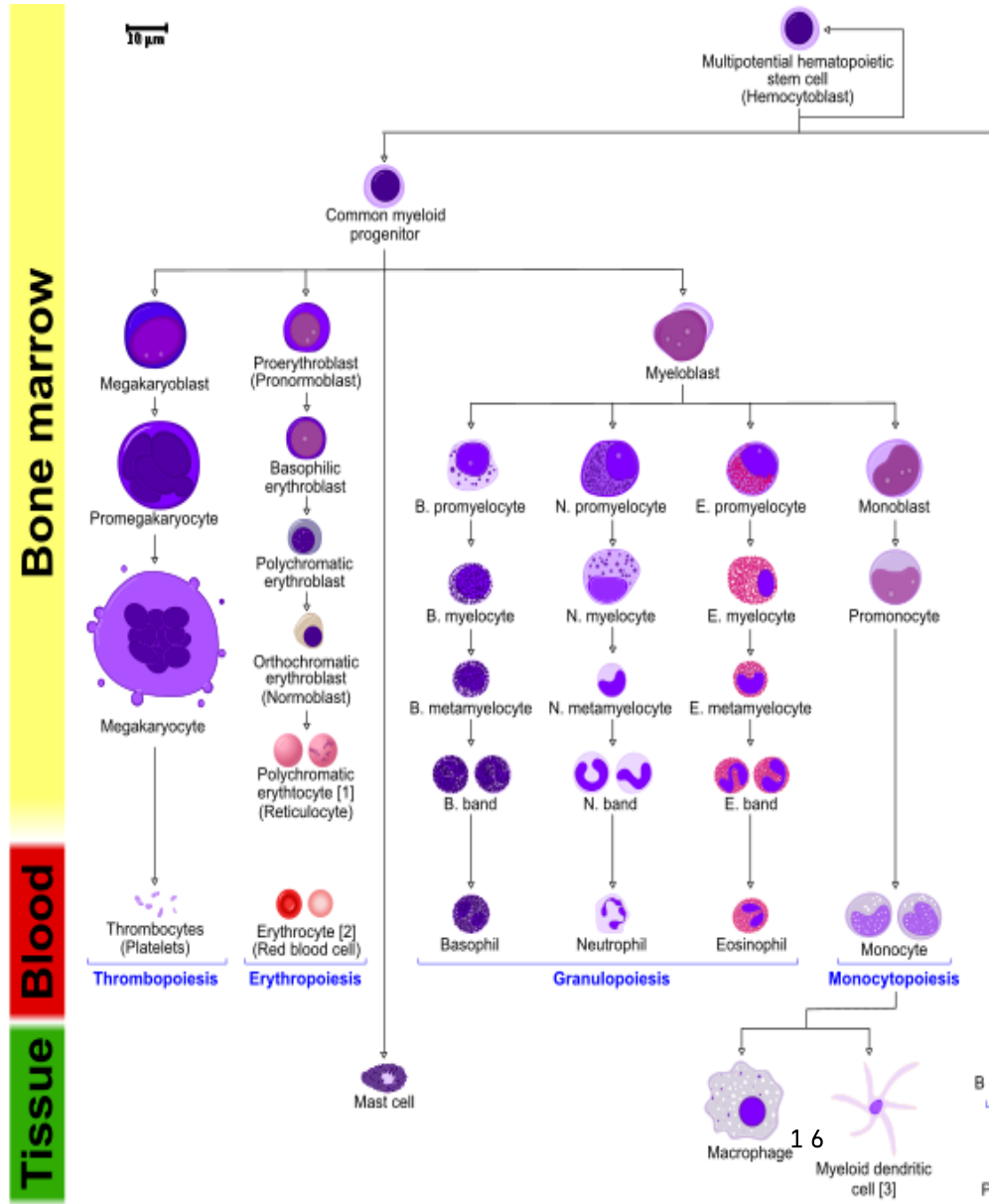
Overlapping of MDS and MPN

MDS : Overproduction of IMMATURE WBCs (blasts)

Table B4: Myelodysplastic neoplasms

WHO Preferred Term	ICD-O
Myelodysplastic neoplasms with defining genetic abnormalities	
Myelodysplastic neoplasm with low blasts and 5q deletion	9986/3
Myelodysplastic neoplasm with low blasts and *SF3B1* mutation	9982/3
Myelodysplastic neoplasm with biallelic *TP53* inactivation	9985/3
Myelodysplastic neoplasms defined morphologically	
Myelodysplastic neoplasm with low blasts	9985/3
• Myelodysplastic neoplasm with low blasts and single-lineage dysplasia	9980/3
• Myelodysplastic neoplasm with low blasts and multilineage dysplasia[†]	9985/3
Myelodysplastic neoplasm, hypoplastic	9985/3
Myelodysplastic neoplasm with increased blasts	9983/3
• Myelodysplastic neoplasm with increased blasts-1	9983/3
• Myelodysplastic neoplasm with increased blasts-2	9983/3
• Myelodysplastic neoplasm with increased blasts and fibrosis	9983/3
Myelodysplastic neoplasms of childhood	
Childhood myelodysplastic neoplasm with low blasts	9985/3
• Childhood myelodysplastic neoplasm with low blasts, hypocellular	
Childhood myelodysplastic neoplasm with increased blasts	9985/3

WHO Preferred Term	ICD-O
Other	
Myelodysplastic neoplasm with ring sideroblasts and multilineage dysplasia (MDS-RD-MLD) *(Note: This term is not included in the WHO 5th edition)*	9993/3
Myelodysplastic syndrome, NOS *(Note: This term is not included in the WHO 5th edition)*	9989/3



MPNs: Too many myeloid **MATURE** cells
LEUKEMIA: Too many **IMMATURE** cells

…Myeloproliferative Neoplasms

able B2: Myeloproliferative neoplasms (MPNs)

WHO Preferred Term	ICD-O
Chronic myeloid leukemia (CML)	9875/3
Chronic neutrophilic leukemia (CNL)	9963/3
Chronic eosinophilic leukemia (CEL)	9964/3
Polycythemia vera (PV)	9950/3
Essential thrombocythemia (ET)	9962/3
Primary myelofibrosis (PMF)	9961/3
• Primary myelofibrosis, prefibrotic	9961/3
• Primary myelofibrosis, fibrotic	9961/3
Juvenile myelomonocytic leukemia	9946/3
Myeloproliferative neoplasm, NOS (unclassifiable) (See also Table B5: Myelodysplastic/myeloproliferative (MDS/MPN)	9975/3

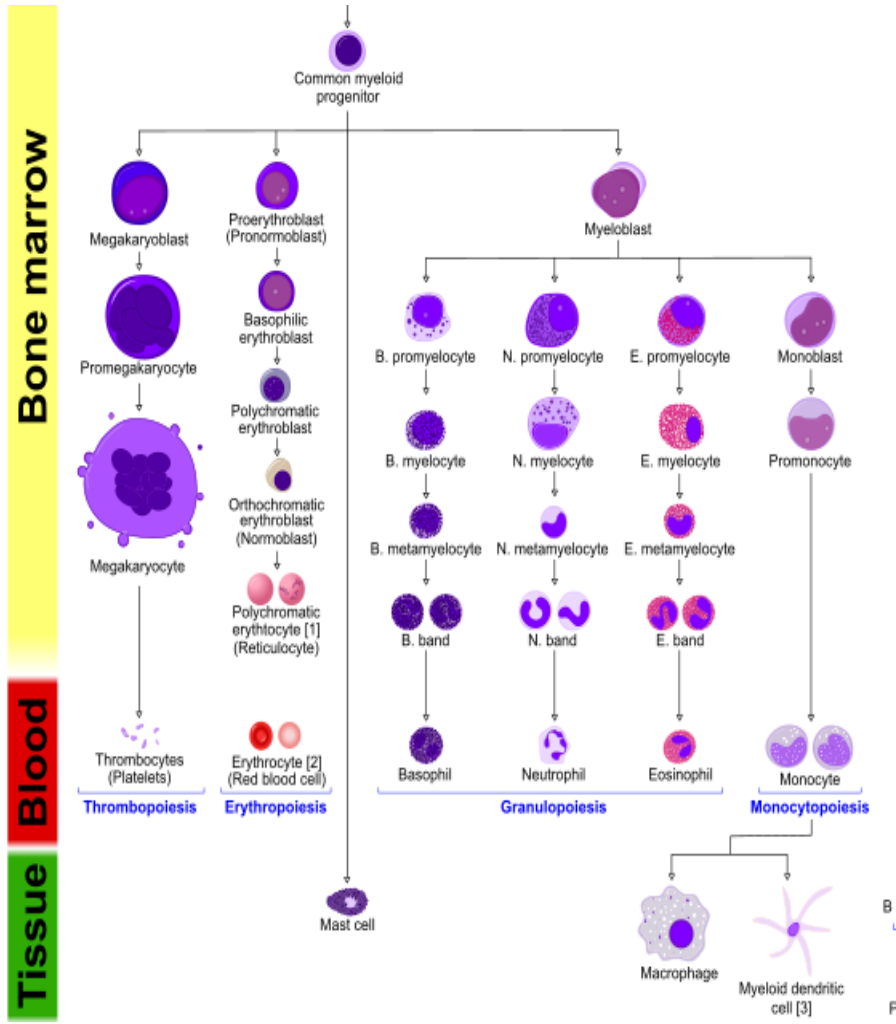
Hematopoietic and Lymphoid Neoplasm Coding Manual 75

Polycythemia : Abnormally high RBC count.

Polycythemia Vera:
Myeloproliferative Neoplasm that causes the faulty Bone Marrow to produce **too many RBCs**, often alongside high WBCs and Platelets. Usually, PV is driven by a JAK2 mutation.

The polycythemia with higher number of RBCs increases the blood viscosity (thickness), and there is higher risk of blood clots, strokes, or heart attacks.

Notice the **NEUTROPHILIC** and **EOSINOPHILIC**, and myelo**MONOCYTIC** Leukemias. They are all derived from the **Myeloid** LINE.



Myelofibrosis: Abnormal stem cells cause bone marrow scarring (fibrosis) replacing healthy bone marrow.
The abnormal stem cells overproduce abnormal blood cells that release excessive cytokines that stimulate intense scarring.

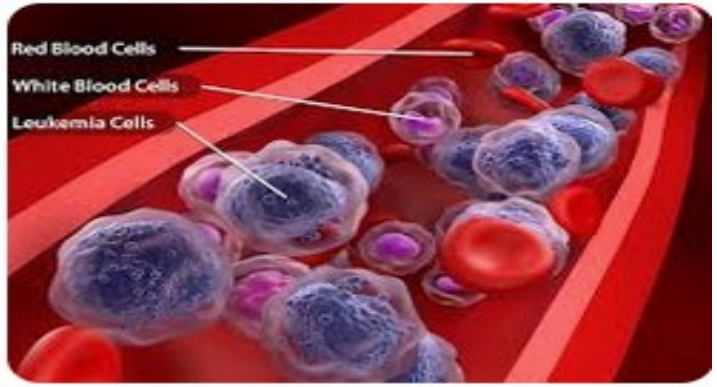
Leukemias (cancers of the blood) vs **Lymphomas**

Leukemias and Lymphomas

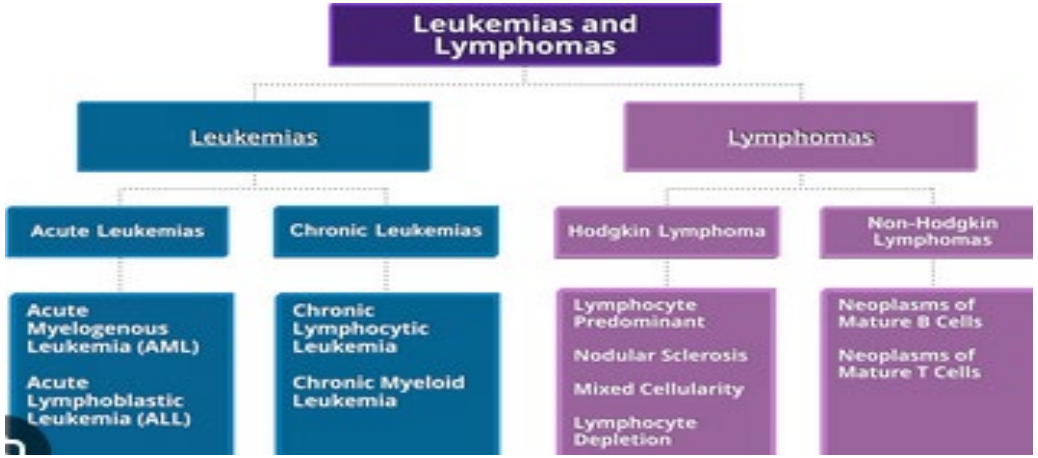
One of the differences between leukemia and lymphoma is that leukemia most commonly presents in the bone marrow and/or blood while lymphoma most commonly manifests in lymph nodes, lymphoid tissue, or lymphoid organs. When only the bone marrow is involved, the diagnosis is usually leukemia. Although rare, a lymphoma may present only in the bone marrow. (See PH rules, Modules 6 and 7, for instructions on coding primary site for lymphomas.)

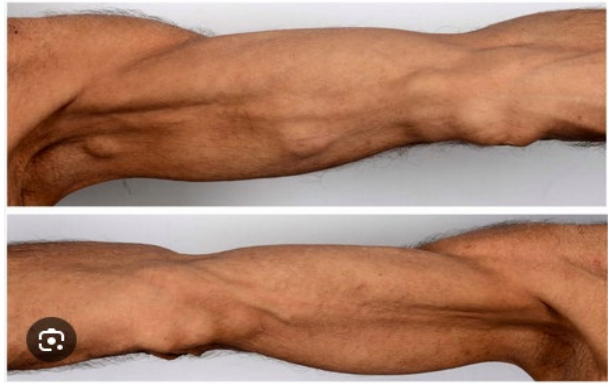
Both leukemia and lymphoma patients may have splenomegaly (enlargement of the spleen). Patients with leukemia may have leukemic infiltrate of the spleen. Splenomegaly does not mean that the leukemia originated in the spleen or that this neoplasm is lymphoma. The spleen filters and stores blood cells. The spleen involvement is usually secondary, much like metastases in solid tumors. The rare histologies that are primary in the spleen are identified in the Heme DB. The Primary Site will be listed as C422.

Hematopoietic and Lymphoid Neoplasm Coding Manual p12



WebMD
Acute Myeloid Leukemia (AML): Sy...





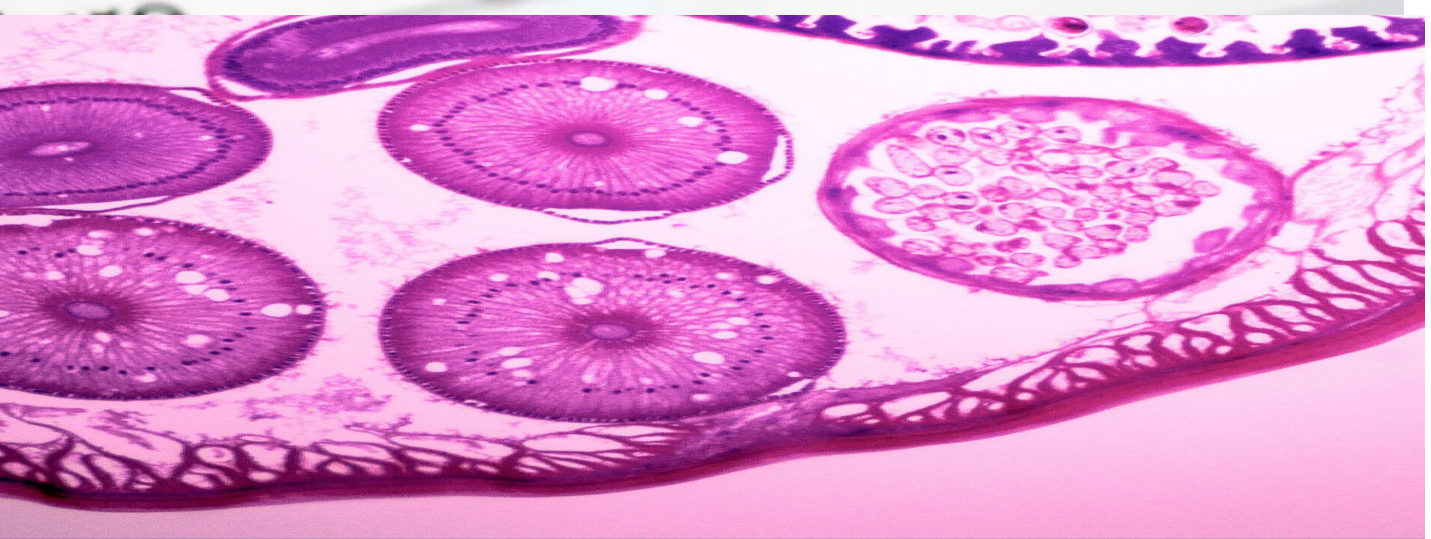
A rare involvement of epitrochlear lymph nodes in mantle cell lympho...

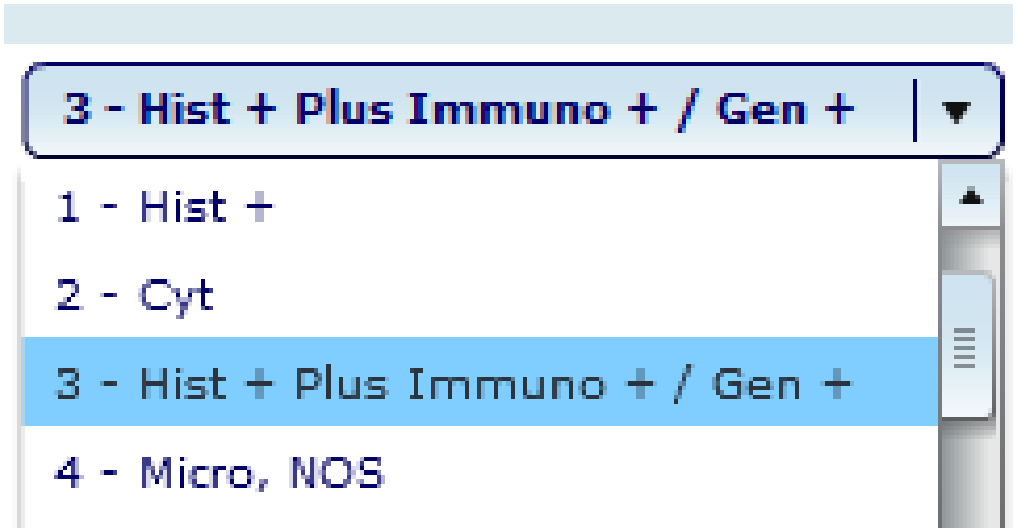
Visit >

Leukemia and lymphoma are terms that reflect the primary behavior and often the site of a neoplasm. Leukemias have cells circulating in the peripheral blood, which can originate in lymph nodes or the marrow. Lymphomas generally form solid masses in lymph nodes or organs containing lymphoid tissue; they may occasionally have circulating tumor cells as well.

Leukemia and lymphoma are terms that reflect the primary behavior and often the site of a neoplasm. Leukemias have cells circulating in the peripheral blood, which can originate in lymph nodes or the marrow. Lymphomas generally form solid masses in lymph nodes or organs containing lymphoid tissue; they may occasionally have circulating tumor cells as well.

Leukemias and lymphomas may also be defined as being chronic or acute. Chronic neoplasms are of longer duration and are slowly progressive while acute neoplasms are of shorter duration and rapidly progressing.





Diagnostic Confirmation

Leukemias

Diagnostic Process for Leukemia

For most patients, the first suspicion or presentation of a hematopoietic neoplasm will be symptoms such as unexplained weight loss, weakness, chronic fatigue, easy bruising, etc. When the physician suspects leukemia, he/she usually orders a complete blood count (CBC) and/or a peripheral blood smear. The CBC will identify abnormalities of the platelets, hemoglobin, white blood cells or red blood cells. When an abnormality is identified in the blood cell analysis, a bone marrow (BM) biopsy is usually the next procedure.

The CBC or bone marrow alone seldom provide a definitive diagnosis; however, the results usually provide one or more provisional diagnoses such as: myeloproliferative neoplasms, myeloid neoplasms, myelodysplastic/myeloproliferative neoplasms, myelodysplastic syndromes, or leukemia. These non-specific diagnoses are differential or provisional. More testing is needed to identify the specific hematopoietic or lymphoid neoplasm. Follow back on these cases should be done.

Diagnostic Confirmation

Include CBC and WBC in Leukemia text

CBC TEST NORMAL VALUES

WBC White blood cells	4,000–11,000/mcL
RBC Red blood cells	4,5–5.9 million/mcL

Text - Dx Procedures - Lab Tests

04-13-24 (MSMC) COMPLETE BLOOD CELL COUNT WITH DIFFERENTIAL (+): LEUKOCYTOSIS (WBC 27,170), MACROCYTIC ANEMIA (RBC 1,660,000, HGB 5.8, MCV 101.80), THROMBOCYTOPENIA (PLT 39,000). DIFFERENTIAL: NEUTROPENIA 1%, LYMPHOPENIA 7%, MONOCYTES 2%, BASTS 73% (HH). PATHOLOGIST COMMENTS: NUMEROUS BLASTS IDENTIFIED CONSISTENT WITH ACUTE LEUKEMIA. THE PATIENT LEFT AMA.

Code 1: Positive histology

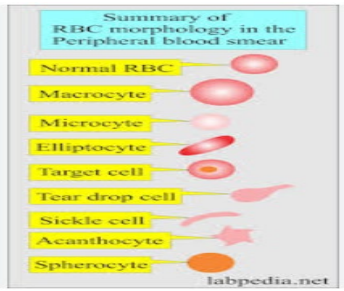
- **Leukemia** only (9800/3-9948/3)
  - Positive histology also includes complete blood count (CBC) or white blood count (WBC)
    - A registrar may **NOT** abstract a hematopoietic neoplasm based on a CBC or WBC with abnormal counts alone. **There must be a diagnosis of a reportable Heme neoplasm by the managing physician based on the CBC or WBC report.**
    - If immunophenotyping, genetic testing, or JAK2 is done and positive, see code 3
      - Code 1 is applicable if immunophenotyping, genetic testing, or JAK2 are done and not diagnostic, or if unknown if these were done

Dx Confirmation: 1 - Hist +

If we have a genetic test like a JAK1 and it is positive, but for the leukemia requires a JAK2 positive, then you code it to HISTOLOGY 1

CBC is counting the cells vs PBS (**Periphery Blood Smear**) is the visual detail the abnormal shapes and sizes, the immature cells.

A CBC (Complete Blood Count) gives *quantitative* numbers (how many of each cell), while a peripheral blood smear (PBS) provides *qualitative*, visual details (size, shape, color, internal structure) of those cells, often used to investigate abnormal CBC results or diagnose specific blood disorders like infections or anemias,



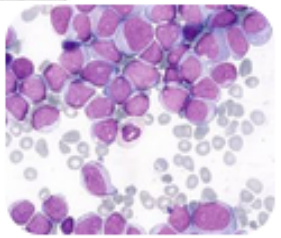
CBC – COMPLETE BLOOD COUNT

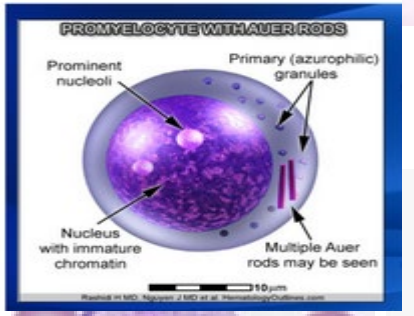
NORMAL LAB VALUES CHART

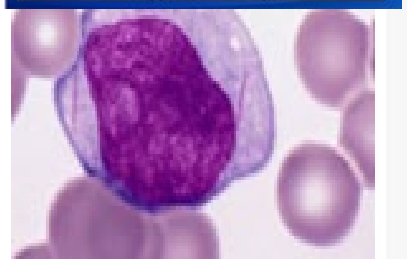
Test	Normal Range	Notes
Hemoglobin (Hb)	Male: 13–17 Female: 12–15 g/L	Carries oxygen
RBC Count	Male: 4.5–5.9 m/µL Female: 4.1–5.1 m/µL	Oxygen transport
WBC Count	4,000–11,000 /µL	Infection indicator
Platelets	1,5–4.5 lakh /µL (150,000–450,000 /µL)	Blood clotting
Hematocrit (Hct)	Male: 40–52% Female: 36–47%	% of RBCs in blood
MCV	80–100 fL	RBC size
MCH	27–33 pg	Hb per RBC
MCHC	32–36 g/dL	Hb concentration
RDW	11–15%	RBC size variation
Neutrophils	55–70%	Bacterial infection
Lymphocytes	20–40%	Viral infection
Monocytes	2–8%	Chronic infection
Eosinophils	1–4%	Allergy, parasites
Basophils	0.5–1%	Allergic response

Microbiology Research Group

Peripheral blood smear (PBS) results for leukemia often show **pancytopenia** (low red cells, platelets, and some white cells) or **high leukocytosis** (high white blood cells, often immature **blasts**) along with abnormal shapes and sizes (anisocytosis/ poikilocytosis), indicating immature white cells (blasts) crowding out normal cells, with specific findings like Auer rods pointing towards Acute Myeloid **AML**







Text - Dx Procedures - Lab Tests

04-13-24 (MSMC) COMPLETE BLOOD CELL COUNT WITH DIFFERENTIAL (+): LEUKOCYTOSIS (WBC 27,170), MACROCYTIC ANEMIA (RBC 1,660,000, HGB 5.8, MCV 101.80), THROMBOCYTOPENIA (PLT 39,000). DIFFERENTIAL: NEUTROPENIA 1%, LYMPHOPENIA 7%, MONOCYTES 2%, BASTS 73% (HH). PATHOLOGIST COMMENTS: NUMEROUS BLASTS IDENTIFIED CONSISTENT WITH ACUTE LEUKEMIA. THE PATIENT LEFT AMA.

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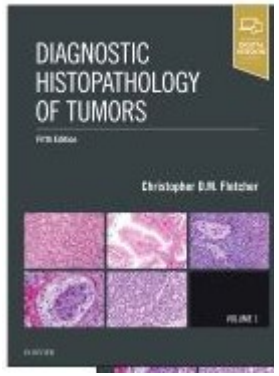
DIAGNOSTIC CONFIRMATION



Example 5 (Histologic confirmation plus genetic and immunophenotyping confirmation): Patient diagnosed with CLL by CBC and flow cytometry that was positive for both the genetic and CD antigens (immunophenotyping) for CLL per pathologist. A bone marrow biopsy was not performed. Since this is leukemia, the CBC is histologic confirmation, so this patient had histologic confirmation, genetic, and immunophenotyping positive for CLL. Code diagnostic confirmation 3, positive histology and positive genetic testing/immunophenotyping.

NOTICE:
"Since this is **leukemia**, the **CBC** is **HISTOLOGICAL** Confirmation"

Hematopoietic and Lymphoid Neoplasm Coding Manual 24

The CBC or bone marrow alone seldom provide a definitive diagnosis; however, the results usually provide one or more provisional diagnoses such as: myeloproliferative neoplasms, myeloid neoplasms, myelodysplastic/myeloproliferative neoplasms, myelodysplastic syndromes, or leukemia. These non-specific diagnoses are differential or provisional. More testing is needed to identify the specific hematopoietic or lymphoid neoplasm. Follow back on these cases should be done.

Code 1 Histology for FNA (for cancer registry purposes, FCDS Reporting)

Code 1 when the microscopic diagnosis is based on tissue specimens from biopsy, frozen section, surgery, autopsy or D&C or from aspiration of biopsy of bone marrow specimens. Code 1 is the preferred coding for Fine Needle Aspiration (FNA). Code 1 is also used for bone marrow biopsy, peripheral blood smears and other diagnostic methods for many leukemia cases (or Code 3). Leukemia can also be diagnosed with CBC or wbc PLUS OR MINUS Immunophenotyping, genetic testing, or JAK2 testing. Code 1 or Code 3 should be used depending on result of special testing.

FCDS Data Acquisition Manual 2025

Diagnostic Confirmation 3

Certain histologies will ALWAYS have a diagnostic confirmation code 3. These ICD-O codes/terms are defined by genetics, meaning the only way they can be diagnosed is through genetic testing.

- The following histologies are diagnosed based on immunophenotyping or genetics and therefore should only be diagnostic confirmation 3: 9806/3, 9807/3, 9808/3, 9809/3, 9812/3, 9813/3, 9814/3, 9815/3, 9816/3, 9817/3, 9818/3, 9819/3, 9865/3, 9866/3, 9869/3, 9871/3, 9877/3, 9878/3, 9879/3, 9896/3, 9897/3, 9911/3, 9912/3, 9965/3, 9966/3, 9967/3, 9968/3, 9986/3.

Hematopoietic and Lymphoid Neoplasm Coding Manual p12

3	9806/3	Preferred	Mixed phenotype acute leukemia with t(9;22)(q34;q11.2); BCR-ABL1
9	9807/3	Preferred	Mixed phenotype acute leukemia with t(v;11q23); MLL rearranged
0	9808/3	Preferred	Mixed phenotype acute leukemia, B/myeloid, NOS
1	9809/3	Preferred	Mixed phenotype acute leukemia, T/myeloid, NOS

2	9812/3	Preferred	B lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2); BCR-ABL1
3	9813/3	Preferred	B lymphoblastic leukemia/lymphoma with t(v;11q23); MLL rearranged
4	9814/3	Preferred	B lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22); TEL-AML1(ETV6-RUNX1)
5	9815/3	Preferred	B lymphoblastic leukemia/lymphoma with hyperdiploidy
6	9816/3	Preferred	B lymphoblastic leukemia/lymphoma with hypodiploidy(Hypodiploid ALL)
7	9817/3	Preferred	B lymphoblastic leukemia/lymphoma with t(5;14)(q31;q32); IL3-IGH
8	9818/3	Preferred	B lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3); E2A-PBX1(TCF3-PBX1)
9	9819/3	Preferred	B lymphoblastic leukemia/lymphoma, BCR-ABL1-like

Diagnosis Confirmation 3

Code 3: Positive histology PLUS positive immunophenotyping or genetic testing

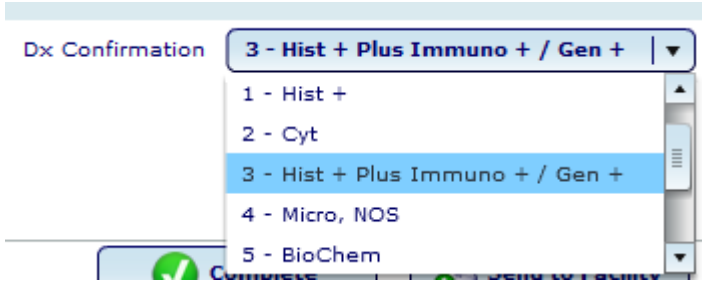
Code 3 can be used for cases diagnosed **2010+** with histologic confirmation (see code 1) **AND** immunophenotyping, genetic testing, or JAK2 confirmation

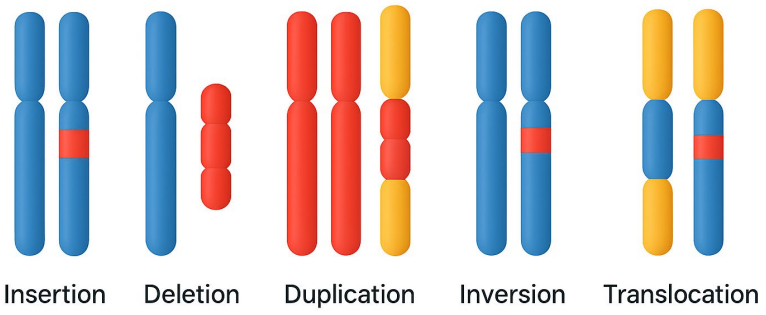
- Certain histologies will ALWAYS have a diagnostic confirmation code 3. These ICD-O codes/terms are defined by genetics, meaning the only way they can be diagnosed is through genetic testing.
  - The following histologies are diagnosed based on immunophenotyping or genetics and therefore should only be diagnostic confirmation 3: 9806/3, 9807/3, 9808/3, 9809/3, 9812/3, 9813/3, 9814/3, 9815/3, 9816/3, 9817/3, 9818/3, 9819/3, 9865/3, 9866/3, 9869/3, 9871/3, 9877/3, 9878/3, 9879/3, 9896/3, 9897/3, 9911/3, 9912/3, 9965/3, 9966/3, 9967/3, 9968/3, 9986/3.

- Histologies that are NEVER confirmation code 3
  The following histologies should never be assigned diagnostic confirmation 3 since they are non-specific codes and neither genetic testing or immunophenotyping are listed as Definitive Diagnostic Methods for these histologies in the Hematopoietic database: 9590/3, 9655/3, 9800/3, 9820/3, 9860/3, 9863/3, 9975/3, 9980/3, 9982/3, 9989/3, 9991/3.
  - If there is immunophenotyping or genetics available, then a more specific histology code may be able to be assigned

9590/3 Malignant lymphoma, NOS
9655/3 Classical Hodgkin lymphoma, lymphocyte depletion, reticular
9800/3 Leukemia, NOS
9820/3 Lymphoid Leukemia, NOS
9860/3 Myeloid Leukemia, NOS
9863/3 Chronic Myeloid Leukemia, NOS
9975/3 Myelodysplastic/myeloproliferative neoplasm, NOS, unclassifiable (MPN/MDS-U)
9980/3 Myelodysplastic neoplasm with low blasts and single-lineage dysplasia (MDS-LB-SLD)
9982/3 Myelodysplastic/myeloproliferative neoplasm with ring sideroblasts and single lineage dysplasia
9989/3 MDS, NOS
9991/3 Refractory neutropenia

CODE 3

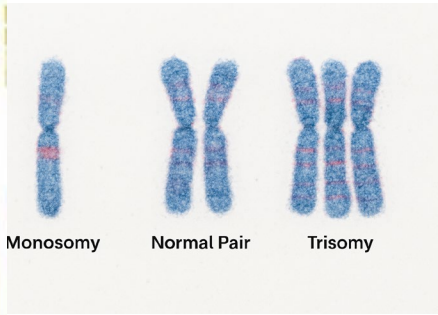


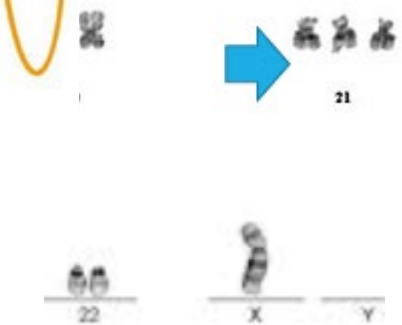


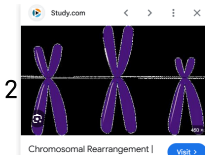
Selected Types of Abnormalities/Mutations

Mutation Type	Abbreviation(s)	Description	Nomenclature Example(s)
Insertion *	ins	Addition of DNA into a gene.	ins(18;5)(q21.1;q31.2)
Deletion	del	Removal of DNA; may occur in one or more base pairs, entire gene(s), or chromosome arm (p or q).	del(5q); del(6q21)
Duplication *	dup	DNA abnormally copied one or more times.	dup(21); FLT3-ITD (Where ITD = internal tandem duplication)
Inversion	inv	Rearrangement within a single chromosome in which a chromosome segment undergoes breakage and rearrangement within itself.	inv(16); inv(3); inv(16)(p13.1;q22); inv(3)(q21;q26.2) (Sometimes described as a translocation between a single chromosome: t(16;16)(p13.1;q22))
Translocation	t(x;x) **	Rearrangement between two chromosomes in which a chromosome segment breaks off and attaches to a different chromosome.	t(9;22); t(8;21); t(9;22)(q34;q11.2); t(8;21)(q22;q22)
Trisomy	(XY, +x) **	An extra copy (three total copies) of the specified chromosome.	47(XY,+8); Trisomy 21; Gain of chromosome 9 (Sometimes these are referred to as just "Trisomy" or "Gain of" abnormalities without abbreviation or specific karyotype notation.)
Monosomy	(XY,-x) **	The presence of only one chromosome from the specified chromosome pair.	45(XY,-16); Monosomy 7; Loss of chromosome 5 (Sometimes these are referred to as just "Monosomy" or "Loss of" abnormalities without abbreviation or specific karyotype notation.)

* Uncommon as a sole genetic/molecular abnormality documented in heme/lymphoid neoplasms.





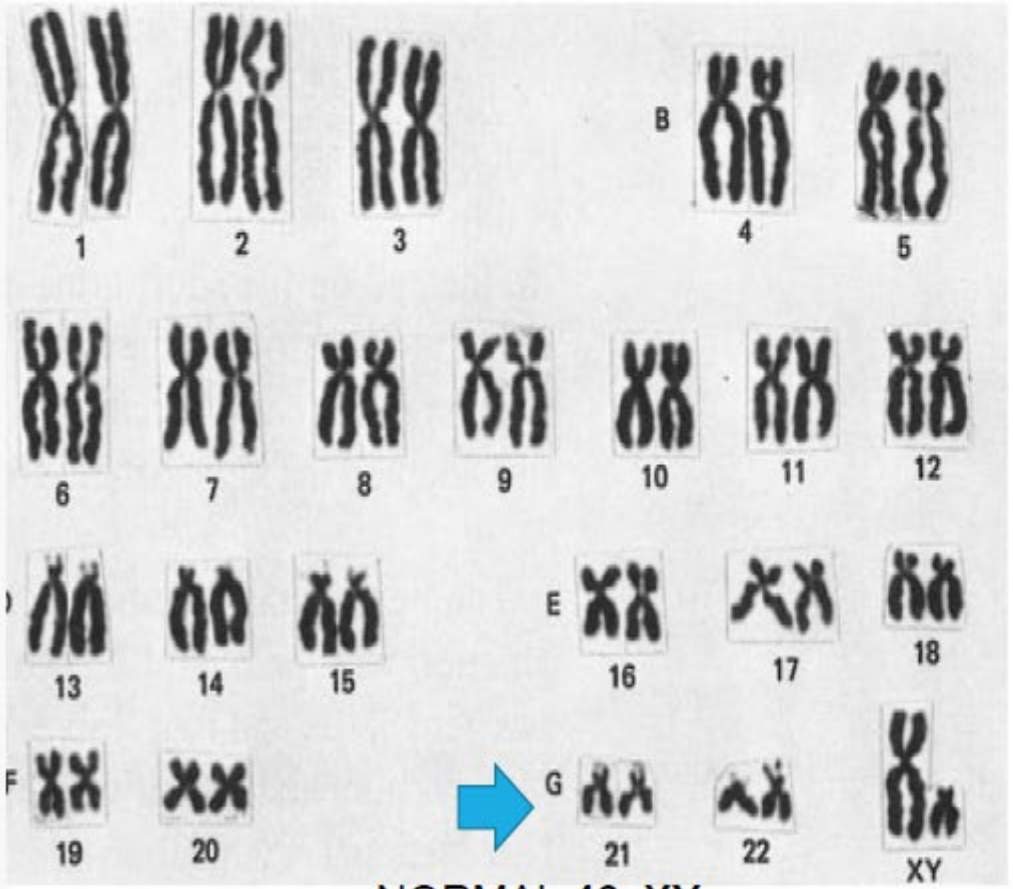


Trisomy	(XY, +x) **	An extra copy (three total copies) of the specified chromosome.	47(XY,+8); Trisomy 21; Gain of chromosome 9 (Sometimes these are referred to as just "Trisomy" or "Gain of" abnormalities without abbreviation or specific karyotype notation.)

9898/3	Preferred	Myeloid leukemia associated with Down syndrome

Normal Karyotype

A karyotype is a picture of the 22 pairs of autosomes and the pair of sex chromosomes that have been isolated from a cell in metaphase. (23 mother + 23 father = 46XX, 46XY)



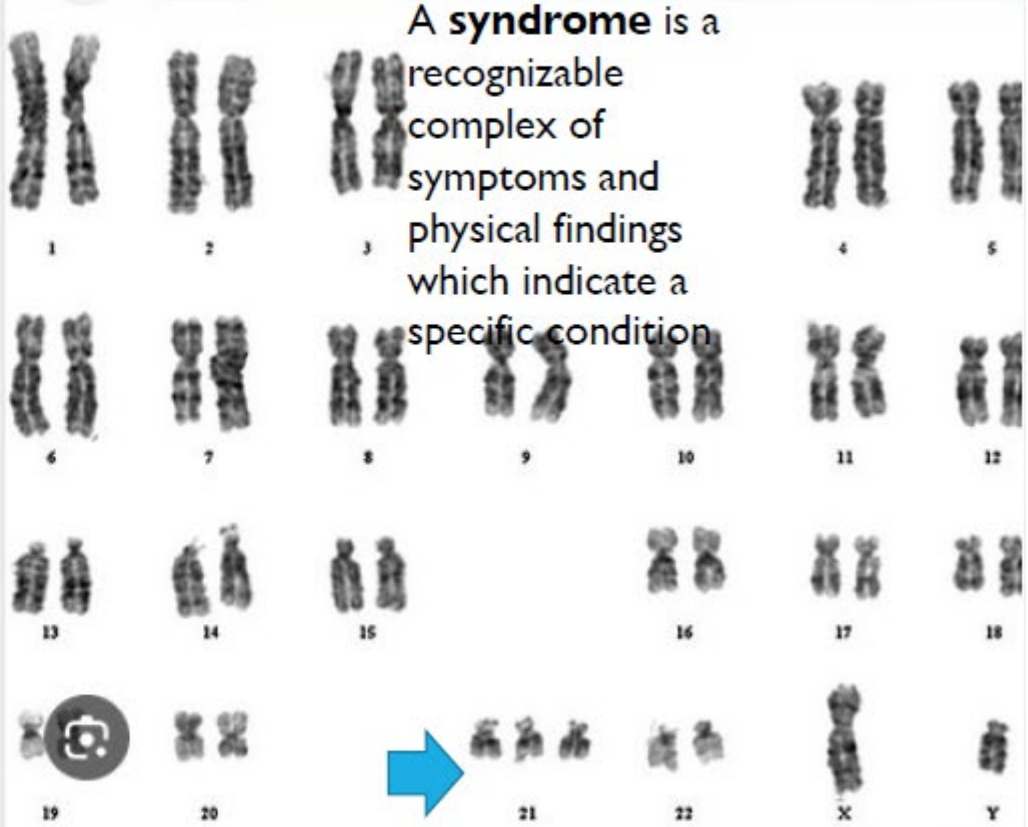
NORMAL **46, XY**

ABNORMAL KARYOTYPE

(47, XY, +21, Trisomy 21) **DOWN SYNDROME.**
Increased risk ALL in children and Alzheimer's.

A **syndrome** is a recognizable complex of symptoms and physical findings which indicate a specific condition

RUNX1 gene/transcription factor in chromosome 21 linked to Leukemias



Karyotype of a patient with trisomy 21

Visit

Children with Down's syndrome are at an increased risk of developing any type of acute leukemia. In particular, they are 150 times more likely to develop acute myeloid leukemia (AML) and are at a 33 times greater risk of developing acute lymphoblastic leukemia (ALL).

AML

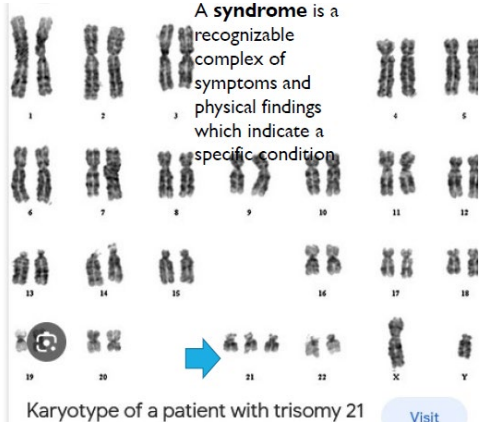
Histology 9861 Behavior 3 - Malignant

9898/3	Preferred	Myeloid leukemia associated with Down syndrome

Trisomy 21 and Down syndrome are the same

They have an EXTRA CHROMOSOME in the pair 21

Prevalence: 1 in every 700 live births, and they can live up to their 60s.



Text - Dx Procedures - Physical Exam - PE

12/05/2023,ORLANDO HEALTH- SEVEN YO W/F W/ HX OF ALL DX IN 10/2020 S/P CHEMO.44", 43 LBS. PMH: TRISOMY 21. PT PRESENTS TO ORLANDO HEALTH ARNOLD PALMER CHILDREN'S HOSP WITH RT CHEEK RASH

Text - Dx Procedures - Physical Exam - PE

18 MONTH OLD FEMALE WITH A HISTORY OF TRISOMY 21, TRANSIENT ABNORMAL MYELOPOIESIS OF DOWN SYNDROME, PULMONARY HYPERTENSION, AND S/P AV CANAL REPAIR WHO PRESENTED FOR 3 DAYS OF FEVER AND INCREASED WOB. LABS IN THE ED REMARKABLE FOR A WBC OF 28.2, HEMOGLOBIN OF 5.6, AND PLATELETS OF 22. THESE LABS ARE SUSPICIOUS FOR LEUKEMIC PROCESS, BMBX CONFIRMED AML.

9898/1	Preferred	Transient abnormal myelopoiesis

Text - Dx Procedures - Physical Exam - PE

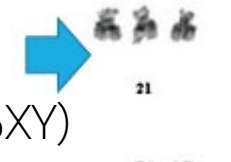
01/04/23(APH)- 7 YO W/NON-HISPANIC MINOR FEMALE PRESENTS FOR PED SURGICAL GASTROCUTANEOUS FISTULA, PMHX OF TRISOMY 21 AND AML LEUKEMIA, LIVES WITH GRANDMOTHER AND BROTHER, WEIGHT 37 LB, HEIGHT 42 IN, NEG TOBACCO, UNSURE OF FMHX OF CANCER, NEG ETOH USAGE

Beware NOT to code 9898/3 to ALL (ONLY to Myeloid, and NOT Lymphoid)

Other Trisomies:
Trisomy 18 (Edward Sx) Prevalence 1 in 5,000 births. High rate of in-utero lethality
Trisomy 13 (Patau Sx) 1 in 16,000 births Most die within a year of life.

Down's Syndrome and Leukemias

Trisomy 21 extra chromosome
47 XX +21
47 XY +21
(Normal 46XX or 46XY)

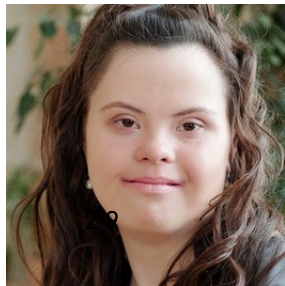


Down syndrome increases the risk of developing Leukemia 10 – 30-fold increase

- Early childhood: for AML <5 years old
- Any Age: ALL (Acute Lymphoblastic Leukemia) Although peak 2-4 y/ old

9898/3	Preferred	Myeloid leukemia associated with Down syndrome

Myeloid Leukemia and Myeloid sarcoma are the same primary



Abstractor Notes

(This code is effective for cases diagnosed 2010 and later. For cases diagnosed prior to 2010 see code 9860/3.)

Myeloid leukemia associated with Down Syndrome (ML-DS) is part of the Secondary myeloid lineage table in the WHO 5th edition of Hematolymphoid Tumors. (See Appendix B in the Hematopoietic Manual, Table B7)

This disease is unique to children with Down's Syndrome (DS). Blood and bone marrow are the principle sites of involvement. Extramedullary involvement, mainly of spleen and liver, is almost always present.

If this leukemia and myeloid sarcoma (9930/3) occur during the same clinical workup, this is one primary, the leukemia.

If the myeloid sarcoma occurs after the diagnosis of the leukemia, that is a manifestation of the leukemia and is the same primary.

See Multiple Primary Rule M3

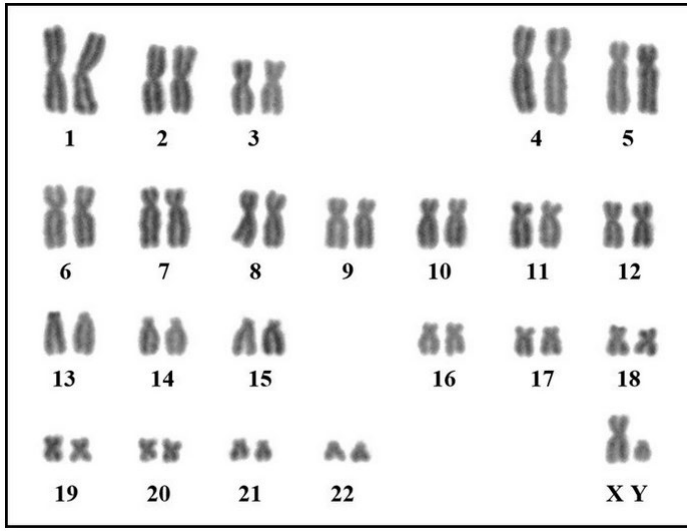
Diagnostic Confirmation

This histology can be determined by positive histology (including peripheral blood) with or without genetics and/or immunophenotyping. Review the Definitive Diagnostic Methods, Immunophenotyping and Genetics Data sections below, and the instructions in the Hematopoietic Manual for further guidance on assigning Diagnostic confirmation.

Trisomies and Multiple Myeloma

47 chromosomes
Trisomies in chromosomes 3,5,7,9,11,12, 15,19, and 21.
IMPROVED SURVIVAL in Trisomy 3 and 5
POOR PROGNOSIS in Trisomy 21 (Down Syndrome)

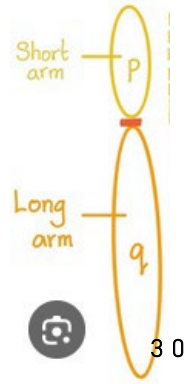
Hyper**di**ploidy with 48-78 chromosomes instead of normal 46 chromosomes have a better prognosis that hypodiploid cases (< 46 chromosomes 46XY, 46XX which are normal karyotype).



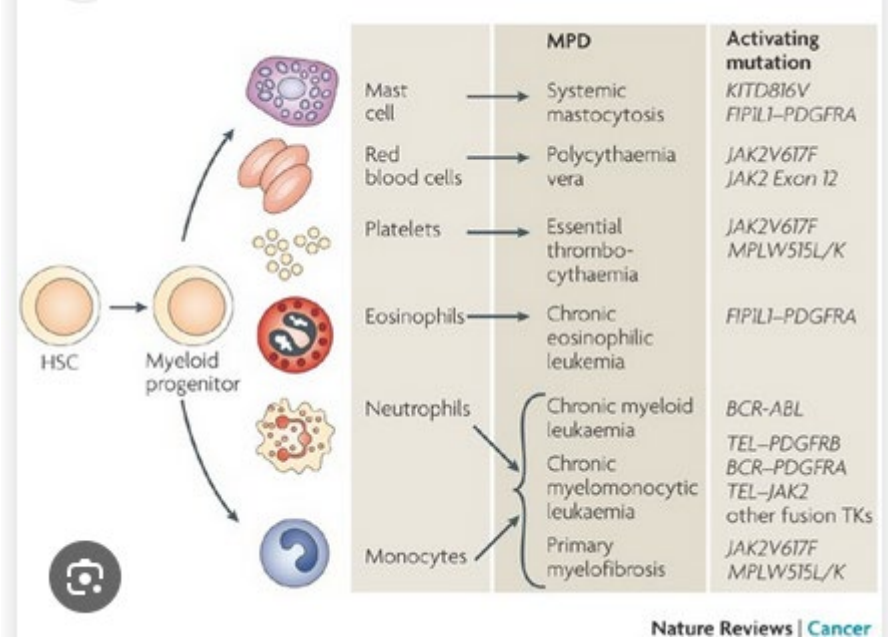
Human normal karyotype from normal metaphase cell of this study (control group).

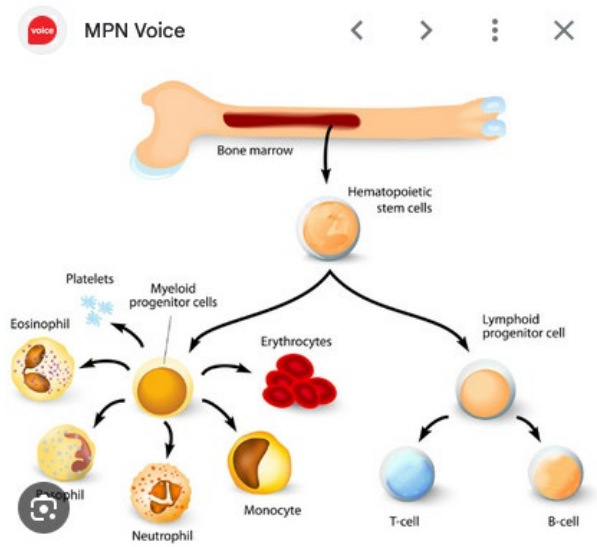
Normal karyotype is 46XX (female) or 46XY (male)

Trisomy 12 and CLL in 15-20% of cases



JAK2 Mutation





JAK2 protein regulates **blood cell production**. When it is mutated, becomes **overactive** and signals cells to grow and divide more, leading to **excess of cells**. Controls how many blood cells the stem cells make. Janus kinase 2 is a gene and protein that regulates **cell growth**, especially blood cell production in the bone marrow.

It can be mutated in Leukemias/Lymphomas, MDS and Myeloproliferative Neoplasms as Polycythemia Vera...

Here where the JAK2 INHIBITORS WORK! Ruxolitinib Pacritinib Fedratinib Memolotinib

Note 2: Common histologies for JAK2

- Record JAK2 for any hematopoietic neoplasm. It is most commonly used for the following histologies listed below. Nearly all people with polycythemia vera, and about half of those with primary myelofibrosis and essential thrombocythemia, have the mutation.
  - Polycythemia Vera (9950/3)
  - Primary myelofibrosis (9961/3)
  - Essential Thrombocytopenia (9962/3)
  - Chronic myelomonocytic leukemia (9945/3)

"JAK-STAT pathway" controls genes for **cell proliferation**, **immune** responses, and inflammation.

9715/3: Anaplastic large cell lymphoma, ALK-negative / Breast implant-associated anaplastic large cell lymphoma

JAK1 mutation

Immune dysregulation/chronic autoinflammation

Genetics Data

DUSP22 locus at 6p25.3
JAK1 mutations
STAT3 mutations
TR genes are clonally rearra

Signs and Symptoms

Axillary lymphadenopathy (breast)
Drenching night sweats
Fatigue
Fever (for no known reason)
Pain in the chest, abdomen, or bones (for no known reason)
Peripheral and abdominal lymphadenopathy
Peri-implant effusion (breast)

JAK3 mutation

9719/3: Extranodal NK/T cell lymphoma, nasal type

Genetics Data

Epigenetic regulators (BCOR, KMT2D, ARID1A, EP300)
Recurrent mutations JAK/STAT pathway genes (STAT3, *JAK3*, STAT5B)
Tumor suppressor genes (TP53, MGA), and RNA helicase (DDX3X)
6q21-25 chromosomal aberration (harbors suppressor genes PRDM1, PTPRK, HACE1, and FOXO3)

9701/3: Sezary syndrome

Cutaneous (skin) lymphoma which presents with generalized skin lesions. Can also present in leukemic form in the peripheral blood or bone marrow. Involvement of any organ can be seen at advanced stages. Assign C449 if specific primary site cannot be determined.

Genetics Data

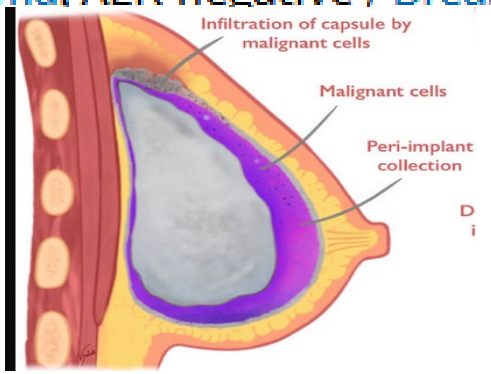
Chromatin modifiers (ARID1A, TRRAP, DNMT3A, TET2)
Driver mutations are also found in DNA damage response pathways (TP53, POT1, ATM), JAK/STAT signalling (STAT5B; *JAK3*),
Recurrent gain of function mutations (PLCG1, CARD11, CD28, CARMIL2[RLTPR])
UV radiation induced mutations

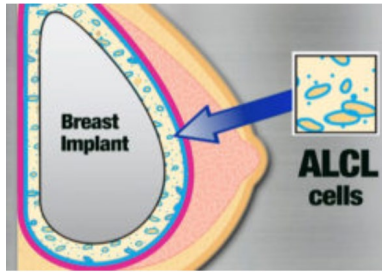
BIA-ALCL: Breast Implant Associated –Anaplastic Large Cell Lymphoma

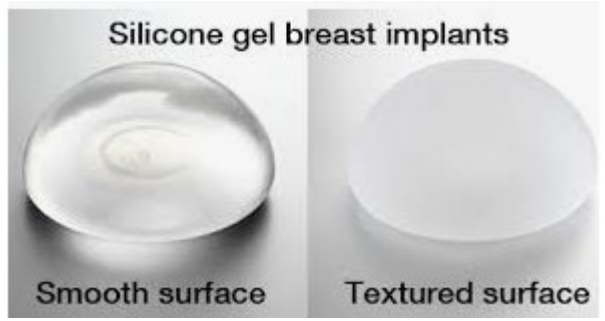
9715/3: Anaplastic large cell lymphoma, ALK-negative / Breast implant-associated anaplastic large cell lymphoma

JAK1 mutation

9715/3







1rs case reported in 1997
Approx 55,000 Breast implants are placed in the USA yearly for cosmetic and RECONSTRUCTIVE indications.

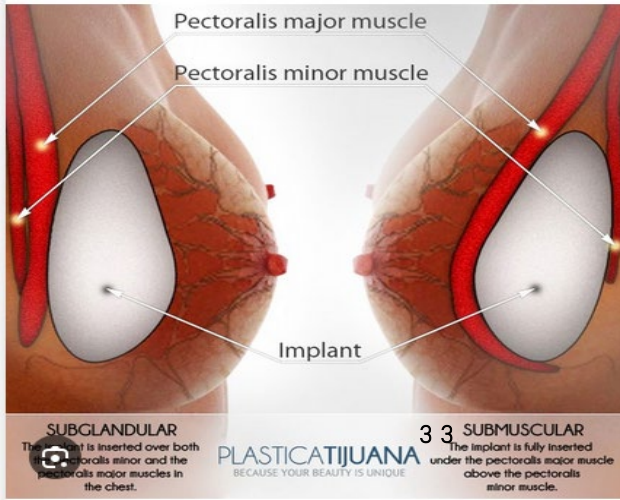
SIGNS/SYMPTOMS B-symptoms, pain, mass, rash, localized effusion not explained by trauma or infection…
Usually, cancer start showing 8-10 years after the Breast implants. **Textured** implants give more trouble than **Smooth** implants.

- IN SITU: Periprosthetic **effusion** called **seroma (FLUID** accumulates!**)** when it is in-situ.
- INFILTRATIVE disease: may be a breast mass or can include LN involvement without the breast mass.

DIAGNOSIS Mammography not very useful compared with an Ultrasound. CT scan, MRI, PET scan

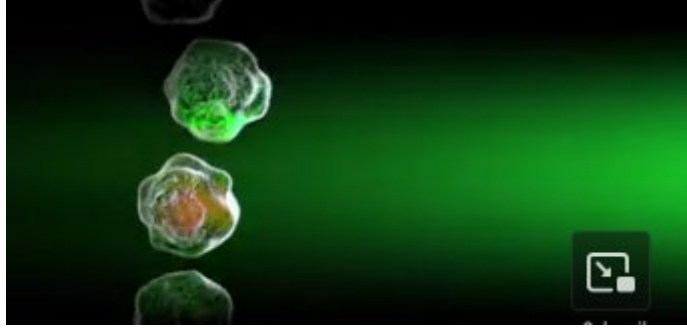
TREATMENT
-SURGERY Removal of the breast implant and its surrounding capsule (total capsulectomy)
Is often curable, especially when caught early.
-CHEMOTHERAPY / RADIATION
May be added if metastasis to lymph nodes or beyond

PROGNOSIS
Survival rate is up to 93% at 3 years and 85% overall survival rate at 5 years.
Disease-related mortality for INFILTRATIVE disease may be as high as 40% in 2 years.



https://pmc.ncbi.nlm.nih.gov/articles/PMC6605706/

IMMUNOPHENOTYPING code 3



- **Immunohistochemistry** (**IHC**) (The Tissue Method)For solid tumors like breast, lung, lymphomas.

9727/3: Blastic plasmacytoid dendritic cell lymphoma

- **Flow Cytometry** (The Liquid Method) For blood, BM, cytology

Think of **flow cytometry** as a high-speed cell sorter that uses lasers to identify what kind of cell is passing through, and immunophenotyping as the “fluorescent name tag" that tells the pathologist which cells are cancer cells.

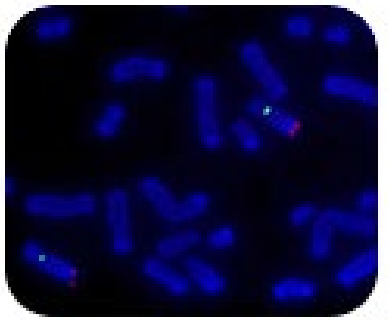
It helps to identify the specific type of leukemia, MDS, MPN...

Note that just because a marker may be listed in the immunophenotyping or genetics fields, it does not mean that one marker is confirming the diagnosis. For many of the neoplasms, especially the B-cell lymphomas, there is a wide range of immunophenotyping results that are shared. Ge

Immunophenotyping

CD4+ (expression/positive)
CD34- (no expression/negative)
CD56+ (expression/positive)
CD303+ (expression/positive)
TCF4+ (expression/positive)
TCL1A+ (expression/positive)
CD304+ (expression/positive)
CD3- (no expression/negative)
CD14- (no expression/negative)
CD19- (no expression/negative)

Diagnosis Confirmation 3



3 - Hist + Plus Immuno + / Gen +

FISH panel test Code 3

FISH (Fluorescence In Situ Hybridization) panel is a molecular GENETIC test which targets DNA/RNA (genes, chromosomes)
Using a fluorescent probe, it identifies GENETIC abnormalities such as gene amplifications, deletions, translocations.

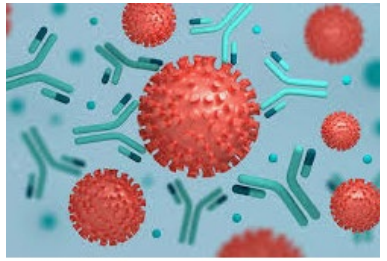
Example: Fish panel test for B-ALL positive for:
t(1;19)(q23;p13) translocation, resulting in the *TCF3-PBX1* fusion gene appearing as an unbalanced translocation (der(19)t(1;19)).

t = Translocation. This means a piece of one chromosome has broken off and attached to another chromosome.

(1;19) = The translocation involves **Chromosome 1** and **Chromosome 19**. Pieces of these two chromosomes are exchanged.

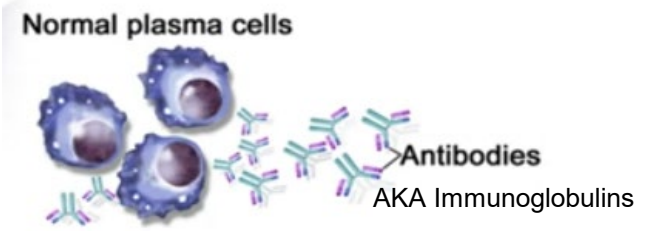
Location: (q23;p13)
q23 Refers to the q arm (long arm) of chromosome 1, specifically region 2, band 3
p13) Refers to the p arm (short arm) of chromosome 19, specifically region 1, band 3

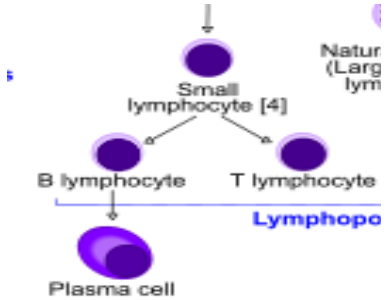
Diagnosis Confirmation 5: + lab test/ marker study for Multiple Myeloma

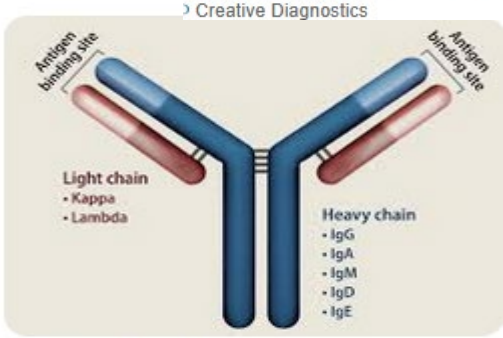


Creative Diagnostics

LYMPHOCYTES B differentiate into PLASMA cells

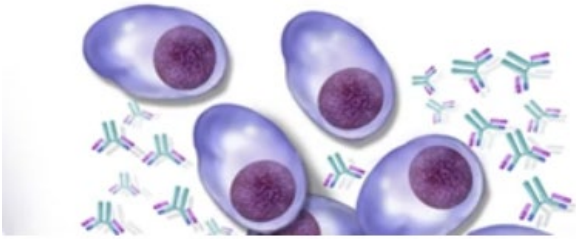






ResearchGate

normal immunoglobulin



gy#pathologylab#biochemist

Code 5: Positive laboratory test/marker study

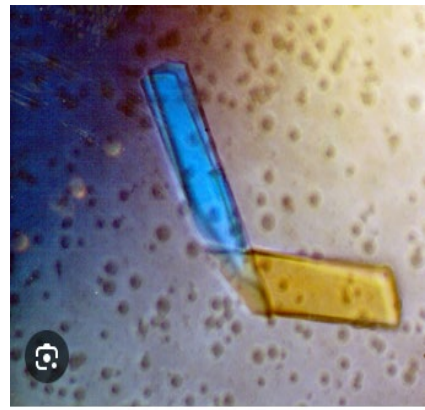
Assign code 5 when the diagnosis of cancer is based on laboratory tests, tumor marker studies, genetics or immunophenotyping that are diagnostic for that specific cancer as listed under Definitive Diagnostic Methods in the Hematopoietic Database.

- Common laboratory test include Bence-Jones protein or SPEP for multiple myeloma
- Common genetic marker JAK2 is used for Myeloproliferative neoplasms (Table B2: Myeloproliferative neoplasms (MPNs)

Hematopoietic and Lymphoid Neoplasm Coding Manual 25

Do not use this code when there is a histological confirmation, which includes a peripheral blood smear is done (which qualifies for a code 1), or a peripheral blood smear followed by flow cytometry (which may qualify for a code 3). Flow cytometry studies are normally done based on an abnormal peripheral blood smear. If unable to find documentation that a peripheral blood smear was done first, assume that it was and code 3

- ***Example 1:*** CT scan consistent with plasma cell myeloma (9732/3). Twenty-four-hour urine protein elevated with the presence of Bence-Jones kappa. Assign code 5 because the diagnosis is based on the positive Bence-Jones and there is no histologic confirmation in this case. Bence-Jones protein is a lab test listed in the Heme DB as one of the definitive diagnostic methods for plasma cell myeloma.



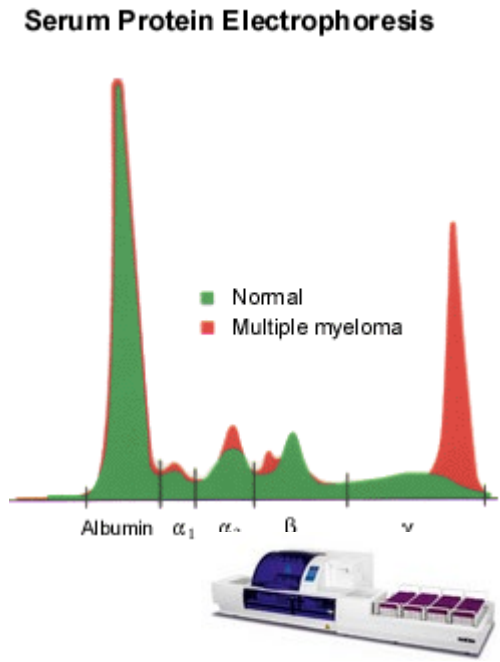
3 6

Bence Jones protein - Wikipedia

Bence Jones proteins are part of immunoglobulins (light chains kappa and lambda). They are produced in excess by neoplastic plasma cells in Multiple Myeloma, they are not complete antibodies but rather fragments that circulate in the blood and are excreted in the urine

Dx Confirmation 5: SPEP/UPEP/Bence-Jones proteins

Dx Confirmation | 5 - BioChem



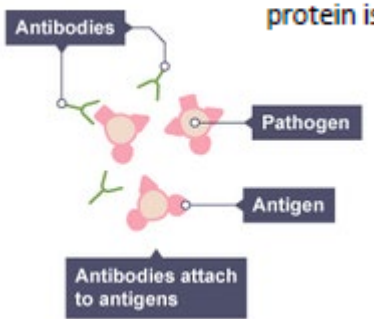
SPEP (**Serum Protein Electrophoresis**) in **Multiple Myeloma**

Multiple Myeloma cause excessive proliferation of ABNORMAL PLASMA cells secreting abnormal immunoglobulins. A **M spike** is an abnormal antibody protein

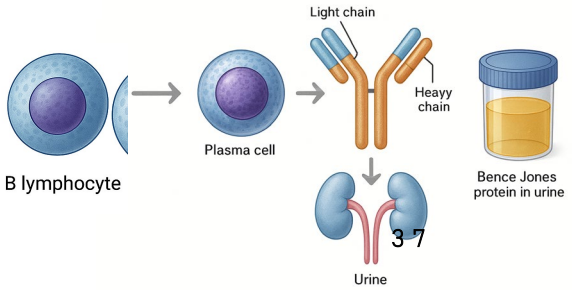
MM secrete abnormal cells from the lymphoid lineage, more specific from the B lymphocytes they secrete abnormal PLASMA cells which secrete gamma globulins or antibodies.

Urine Protein Electrophoresis (**U**PEP or Urine Protein ElectroPhoresis)
Both **SPEP** or **UPEP** give a **M-spike**
A sample of peripheral blood or a 24 hrs. urine sample is collected to detect **Monoclonal** (**M** proteins).
Then Bence-Jones proteins (light chains) can be detected within the same sample of urine specially if followed by immunofixation electrophoresis.

Example 1: CT scan consistent with plasma cell myeloma (9732/3). Twenty-four-hour urine protein elevated with the presence of Bence-Jones kappa. Assign code 5 because the diagnosis is based on the positive Bence-Jones and there is no histologic confirmation in this case. Bence-Jones protein is a lab test listed in the Heme DB as one of the definitive diagnostic methods for plasma cell myeloma.



Code	Description
5	Positive laboratory test/marker study



Codes for Hematopoietic and Lymphoid Neoplasms (9590/3-9993/3)

Microscopically Confirmed

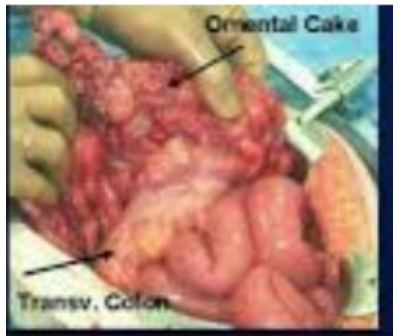
Code	Description
1	Positive histology Includes: peripheral blood smear only
2	Positive cytology
3	Positive histology PLUS: • Positive immunophenotyping AND/OR • Positive genetic studies Includes: peripheral blood smear followed by flow cytometry *(Effective for cases diagnosed 1/1/2010 and later)*
4	Positive microscopic confirmation, method not specified

Not Microscopically Confirmed

Code	Description
5	Positive laboratory test/marker study *Note 1*: Includes cases with positive immunophenotyping or genetic studies and *no* histological confirmation *Note 2:* This does not include cases where a peripheral blood smear is done (code 1) and peripheral blood smear followed by flow cytometry (code 3)
6	Direct visualization without microscopic confirmation
7	Radiology and other imaging techniques without microscopic confirmation
8	Clinical diagnosis only (other than 5, 6 or 7)

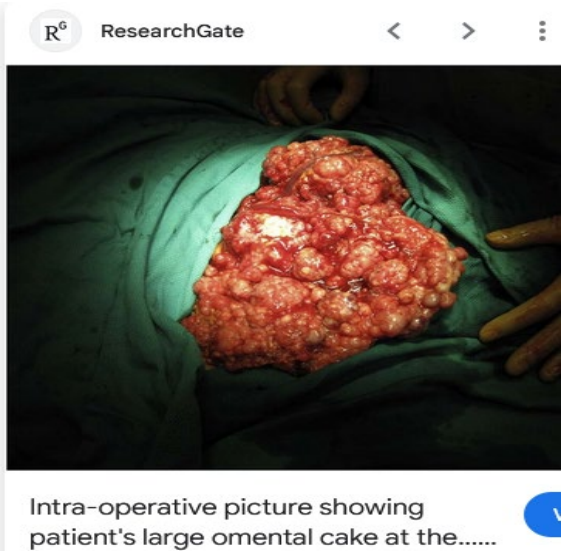
Confirmation Unknown

Code	Description
9	Unknown whether or not microscopically confirmed; death certificate only



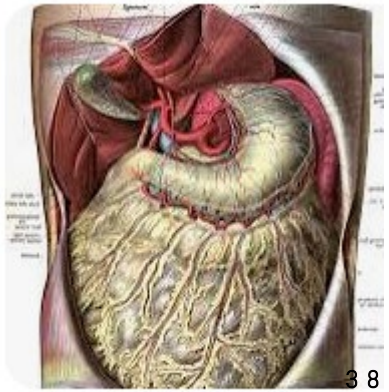
6 Direct Visualization

An example: Taking a patient to the Operating Room for a car accident for internal bleeding and having an Incidental having the patient omental caking all over his/her body. All invaded by cancer! Physician states is cancer in the operative report.
Other Example: The diagnosis is determined by Gross autopsy findings (no tissue or cytologic confirmation)

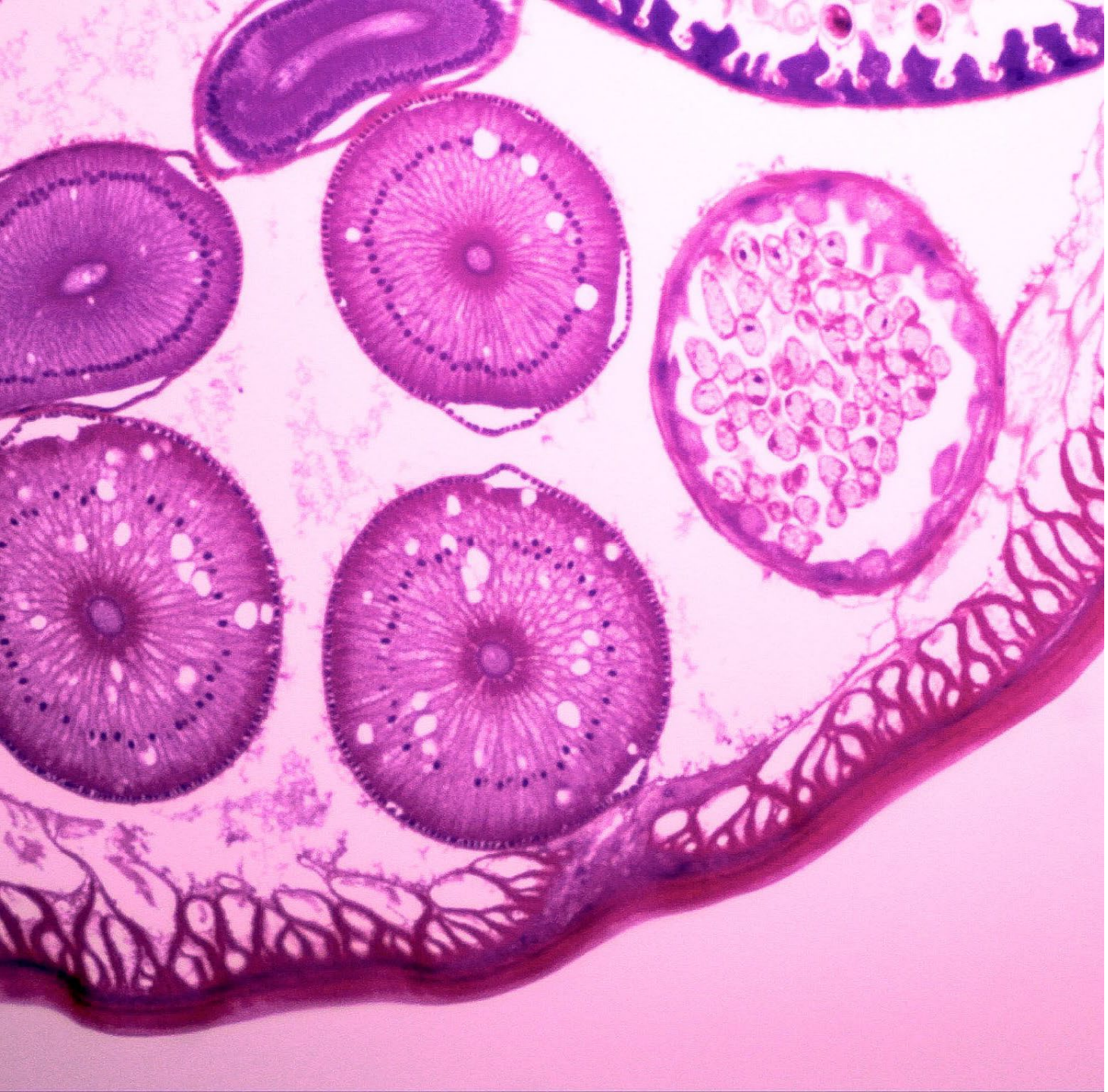


Omental caking: The Omentum becomes thick, dense and irregular with appearance compared to a "cake"

Omentum: a fatty, apron like fold of the peritoneum (the membrane lining the abdominal cavity) that hangs from the stomach and cover the intestines.



Greater omentum - Wikipedia



MORE ABOUT LEUKEMIAS, LYMPHOMAS, HISTIOCYTOSIS...

Hodgkin Lymphoma
Accounts for
4.54 % of ALL NEW CASES
2.05 % of ALL DEATHS
From blood cancers in the USA

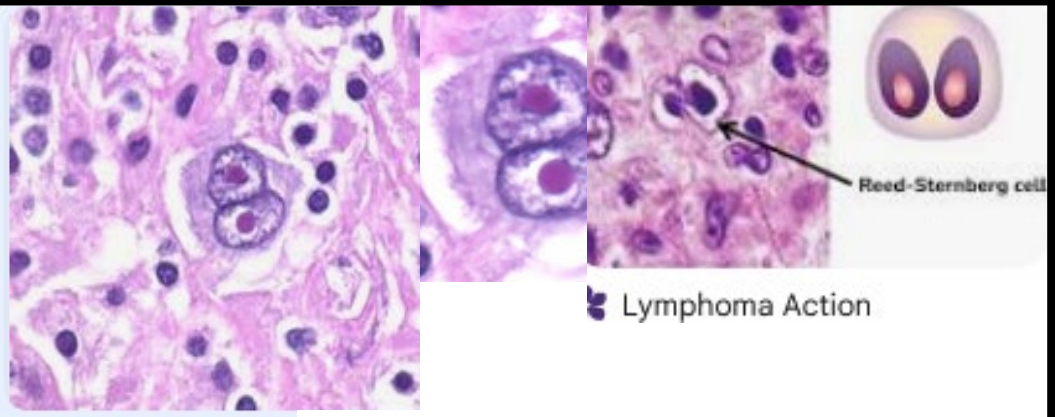
LYMPHOMAS

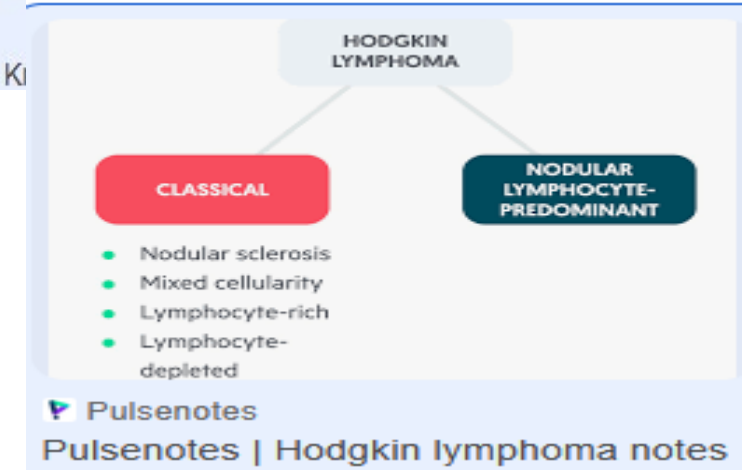
NON-HODGKIN LYMPHOMA
IS ESTIMATED TO ACCOUNT FOR
41% OF ALL NEW CASES AND 32% OF ALL DEATHS
FROM BLOOD CANCERS IN 2025 IN THE UNITED STATES

Source: (6).

© 2025 American Association for Cancer Research®. *AACR Cancer Progress Report 2025.* 2502013-W57.

Classical HODGKIN Lymphoma





10%

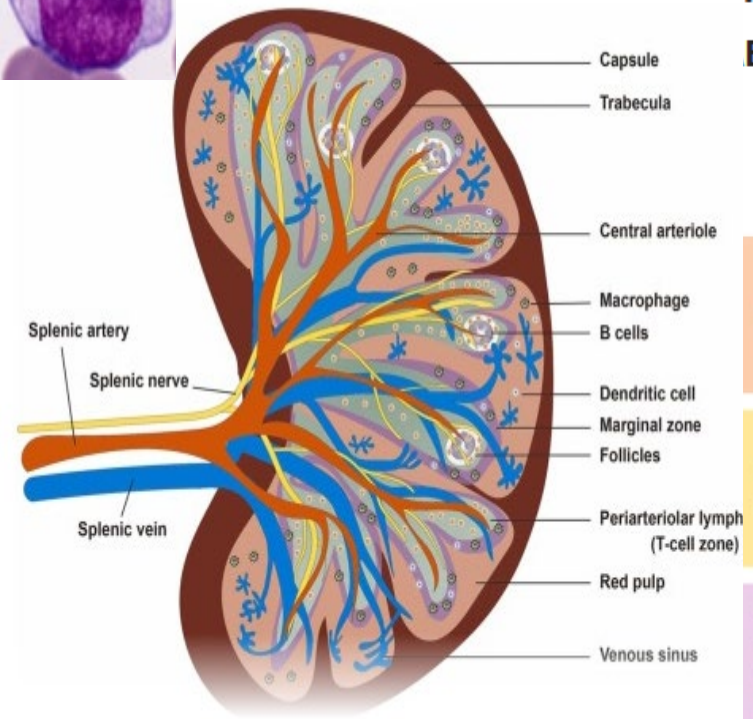
NON-HODGKIN Lymphomas

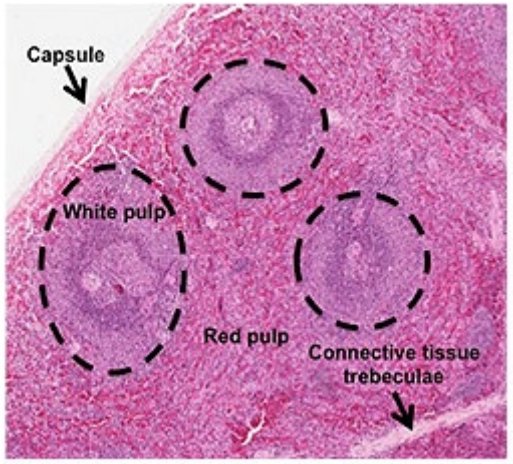
All the other lymphomas including:

- Skin lymphomas (Mycosis Fungoides…)
- Ocular lymphomas
- T cell lymphomas (Peripheral T-cell, Extranodal NK …)
- B cell lymphomas (DLBC, Follicular, CLL/SLL , Mantle, Marginal, MALT…)
-

Lymphoplasmacytic Lymphoma (Also Called Waldenstrom's Macroglobulinemia or Immunocytoma)

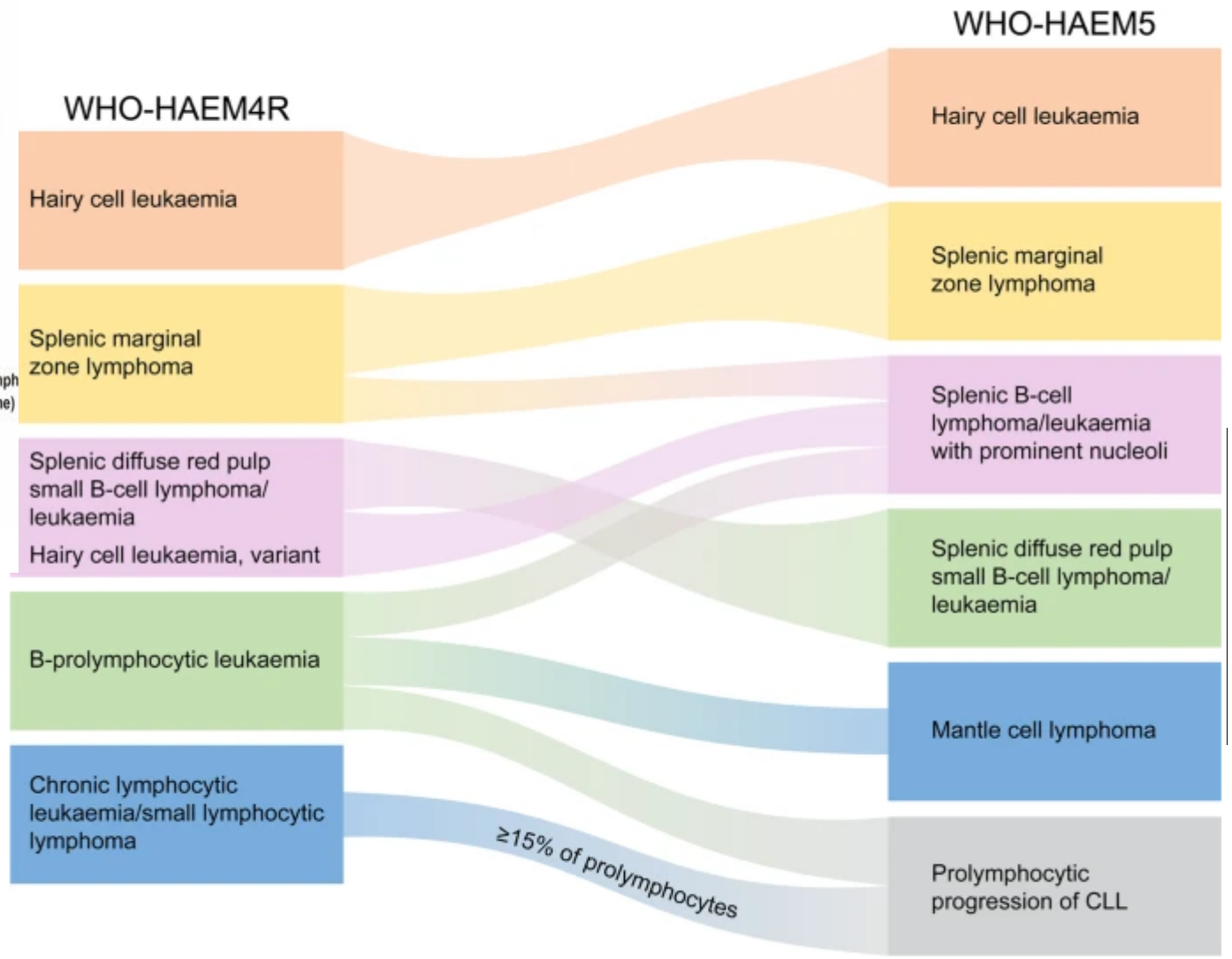
90%



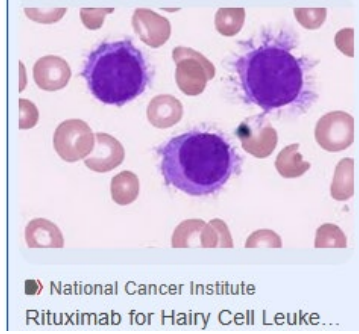


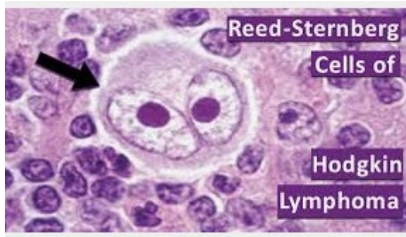
Spleen red and white pulp

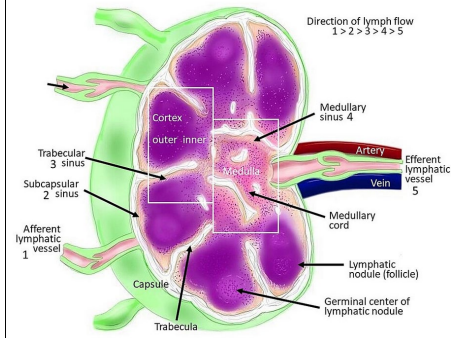
Fig. 1: Summary of the relationship between splenic B-cell lymphoma entities as named and defined in the revised 4th edition of the WHO classification (WHO-HAEM4R) and in the present 5th edition (WHO-HAEM5).

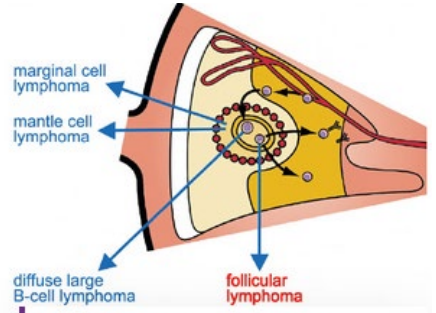


Some cases previously classified as B-prolymphocytic leukaemia do represent (blastoid) mantle cell lymphoma (as was already indicated in WHO-HAEM4R) or prolymphocytic progression of CLL. Cases









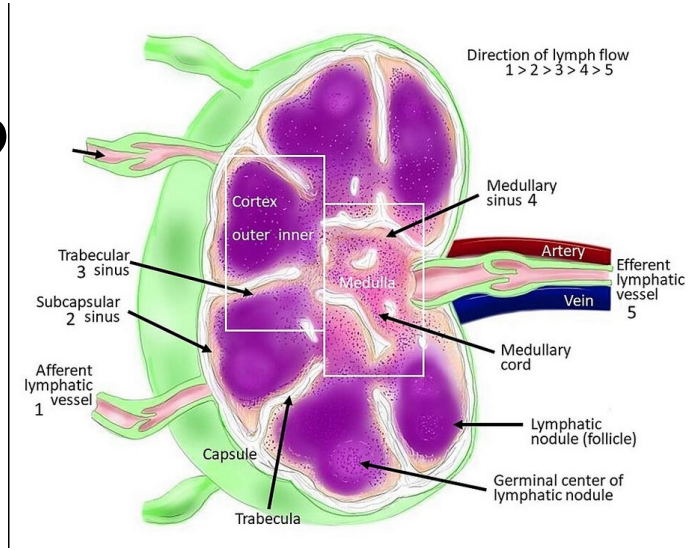
A section of a lymph node showing the origin of follicular lymphoma

Lymphomas

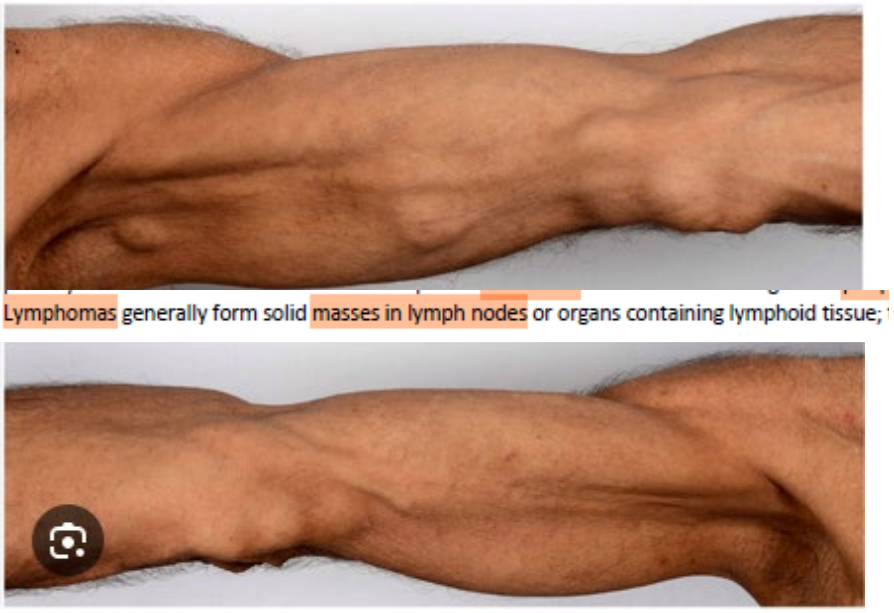
Where do they get their name from?

Lymphomas generally form solid masses in Lymph Nodes or organs containing lymphoid tissue.

- MANTLE Cell Lymphoma: Refers to the “mantle zone” of the lymph node, where the malignant B cells are found.

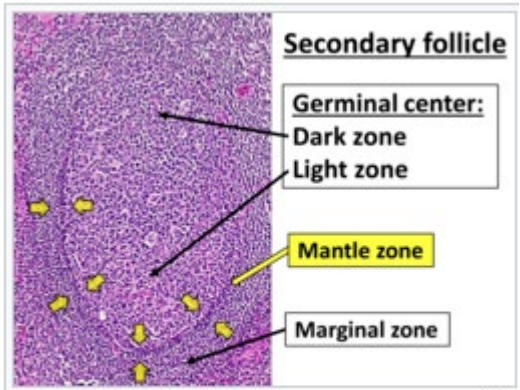


BMJ Case Reports

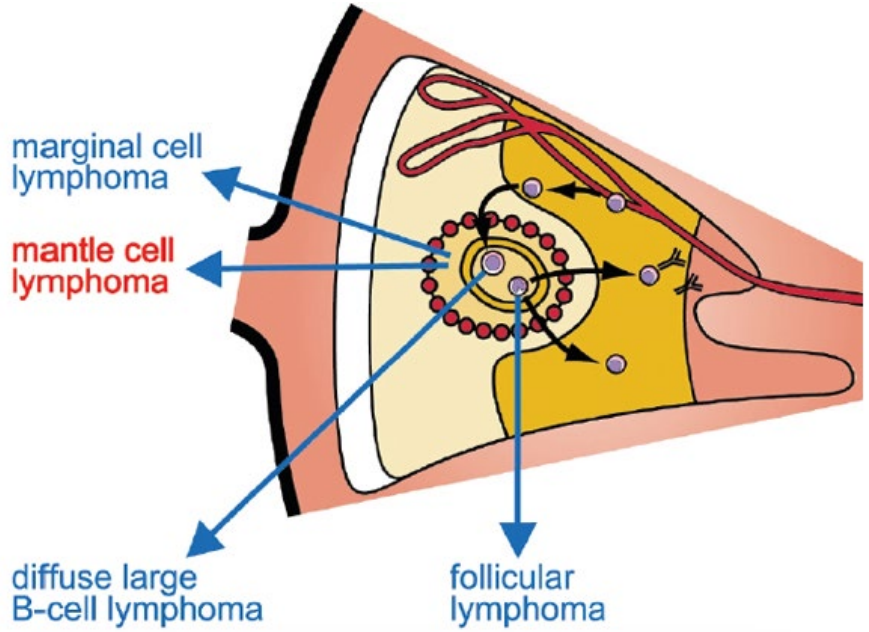


A rare involvement of epitrochlear lymph nodes in mantle cell lympho...

Visit >



Histology of a normal lymphoid follicle, with yellow arrows pointing at mantle zone.

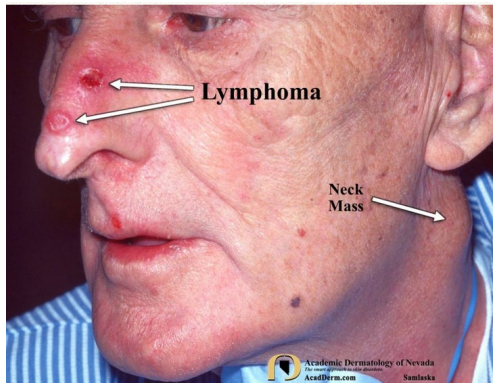


A section of a lymph node showing the origin of mantle cell lymphoma

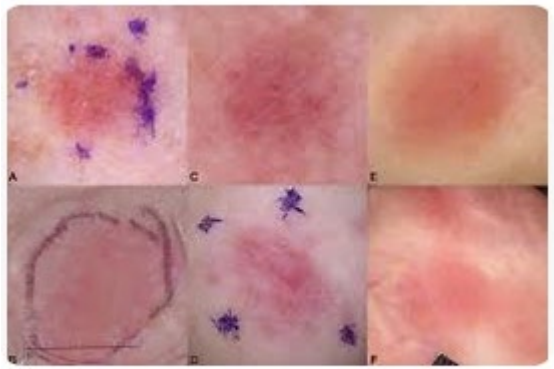
SKIN Lymphomas

- **Cutaneous T-cell Lymphoma (CTCL) - Most Common**
  - **Mycosis fungoides (MF):** The most common type, often presenting as red, scaly patches or plaques.
  - **Sézary syndrome:** A more advanced form of CTCL characterized by leukemia (blood involvement) and erythroderma (red, itchy skin over the whole body).
  - **Primary cutaneous CD30+ lymphoproliferative disorders:** Includes primary cutaneous anaplastic large cell lymphoma (often presents as a single tumor) and lymphomatoid papulosis.
  - **Subcutaneous panniculitis-like T-cell lymphoma:** Affects the fat layer under the skin.
  - **Primary cutaneous peripheral T-cell lymphoma (rare subtypes):** Includes gamma-delta T-cell lymphoma and CD8+ aggressive cytotoxic T-cell lymphoma.
- **Cutaneous B-cell Lymphoma (CBCL) - 20-25% of cases**
  - **Primary cutaneous follicle-center lymphoma:** Often found on the head, neck, or torso; generally slow-growing.
  - **Primary cutaneous marginal zone lymphoma (PCMZL):** Indolent (slow-growing) type often found on arms, legs, and trunk.
  - **Primary cutaneous diffuse large B-cell lymphoma, leg type:** A more aggressive form, frequently found on the lower legs.
- **Other/Rare Types**
  - **Blastic plasmacytoid dendritic cell neoplasm (BPDCN):** A rare, aggressive type of skin lymphoma.
  - **Extranodal NK/T-cell lymphoma, nasal type:** A rare T-cell lymphoma that can

Leukemia Foundation. American Cancer Society. Dartmouth Cancer Center



Lymphoma Type	Appearance on skin	Common Body Areas	Growth Rate	Prognosis / 5-yr Survival	Prevalence in Australia
Mycosis Fungoides (T-cell)	Red, scaly or patchy skin, which may appear like eczema	Trunk, buttocks	Slow growing	Good in early stages	Most common cutaneous lymphoma
Sézary Syndrome (T-cell)	Very itchy skin, red scaly skin with possible nail and hair changes	Whole body	Aggressive	Poorer than MF	Rare subtype (2-5% of CTCL cases)
Primary Cutaneous Follicle Centre Lymphoma (B-cell)	Solitary or groups of papules, nodules or plaques	Head, neck and trunk	Indolent	Excellent at around 90-95%	Relatively common
Primary Cutaneous Marginal Zone Lymphoma (B-cell)	Single or many red-purple papules or nodules	Trunk, arms, legs	Indolent but relapse is possible	Excellent at around 90-98%	Rare
Primary Cutaneous Diffuse Large B-Cell Lymphoma, Leg-type (B-cell)	Ulcerated, painful, rapidly growing tumours	Usually legs	Aggressive	Poor (less than 50%)	Rare



Journal of the American Academy of Derma...
cutaneous lymphoma clinic ...

https://sundoctors.com.au/blog/skin-lymphomas/

Leukemia CUTIS (CMML and AML)

The New England Journal of Medicine
Leukemia Cutis | New England Journal...

While lymphomas usually are from the lymph nodes, we may have skin lymphomas.
But do not confuse skin lymphomas with Leukemia cutis
Cutis from Latin ***cutis***, means "skin"

Leukemia cutis
Leukemia cutis is a rare manifestation of leukemia characterized by infiltration of malignant leukocytes into the skin (epidermis, dermis, or subcutis), producing visible, clinically recognizable lesions. Leukemia cutis may precede, follow, or occur concurrently with the diagnosis of systemic leukemia. It is an advanced phase of the leukemia having a poor prognosis that also strongly correlates with additional sites of extramedullary involvement. This can alter the appropriate treatment regimen for a patient. It is a type of "metastasis" or spread of the leukemia cells.

The "conventional" definition for leukemia cutis is the infiltration of skin from a bone marrow primary. It is most often diagnosed via skin biopsy—punch, shave, etc., utilizing IHC/biomarker testing and is commonly associated with chronic myelomonocytic leukemia (CMML) and acute myeloid leukemia (AML). As such, it a reportable condition especially when preceding a confirmed systemic leukemia diagnosis.

In rare cases, the leukemia cutis is diagnosed and there is no history of AML, and bone marrow biopsy is either not done or comes back negative. In this situation, the diagnosis date would be the date of the positive leukemia cutis skin bx—punch, shave, etc. The case should be coded to C421; 9800/3 Leukemia NOS until the official systemic leukemia diagnosis is rendered. If possible, follow back should be conducted to determine the specific systemic leukemia histology (CMML; AML) and the treatment received. If the leukemia cutis follows or occurs concurrently with the diagnosis of a systemic leukemia, it is NOT a separate primary but merely an advanced stage of the systemic leukemia diagnosis

Lymphomas

Code C778 for lymphoma (Multiple LNs regions)

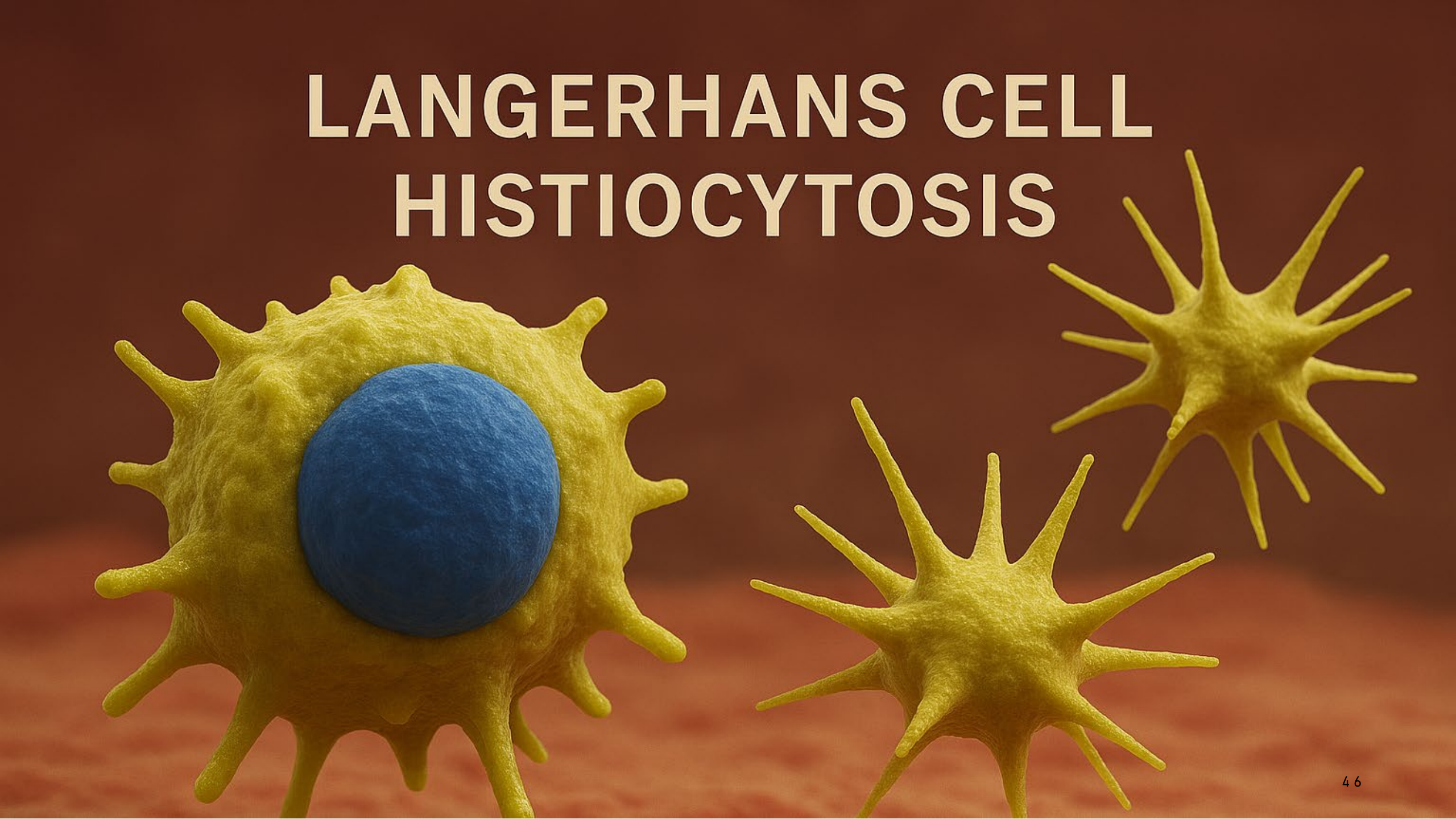
- Code C778 when there are MULTIPLE LNs/Lymph Node CHAINS involved.
- Code C779 when there is lymph node involvement, but unknown if multiple lymph node chains involved

Common Error to code 779

Rule PH21: Multiple lymph node regions

1. Code the primary site to lymph nodes of multiple regions (C778) when multiple lymph node regions are involved.
   a. If a physician documents that the lymphoma originated in a specific lymph node region, code that as the primary site (this is very rare).
   b. If imaging, or the physician states "lymph nodes above and below the diaphragm," code to C778.
   Note: Multiple lymph nodes regions involved can be on the same side or both sides of the diaphragm

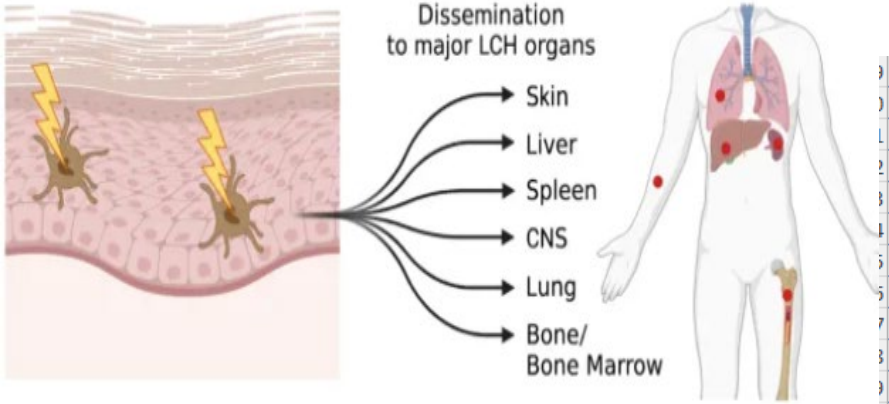
LANGERHANS CELL HISTIOCYTOSIS



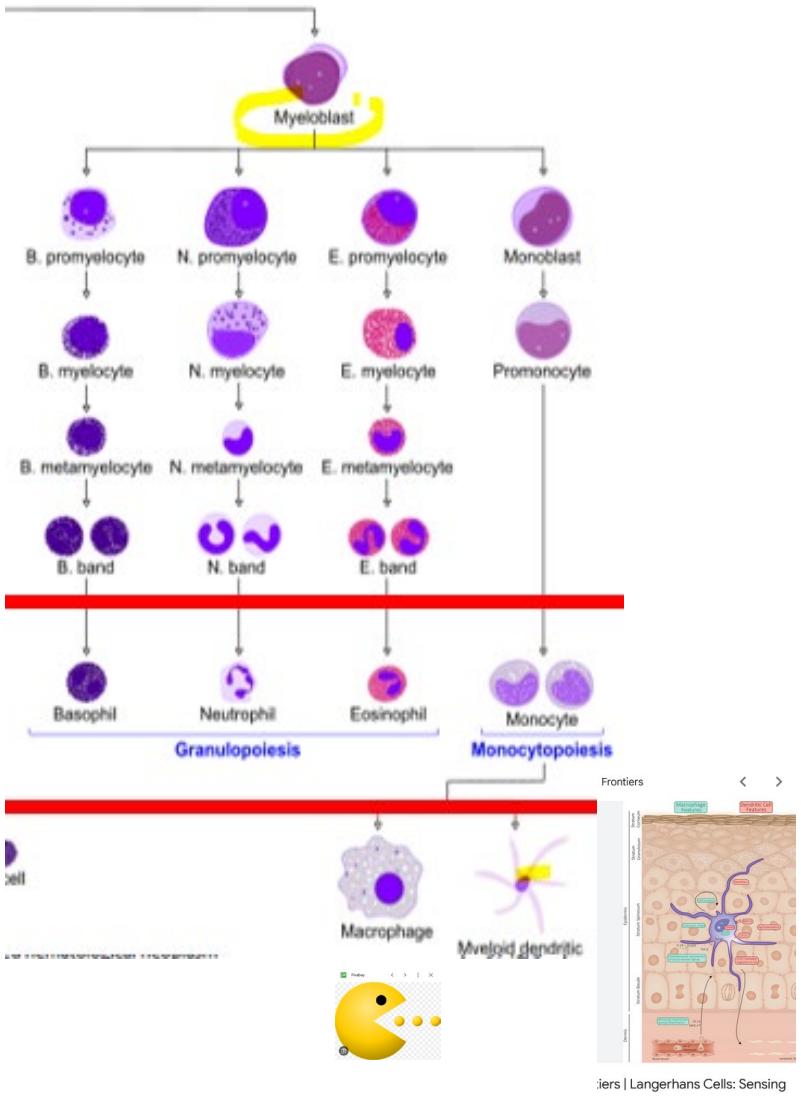
Langerhans cell Histiocytosis (is not lymphoma or leukemia)

- Histiocytosis was first described in the mid to late 1800s.
- Through the years, it has been known by various names:
  - Eosinophilic granuloma
  - Letterer-Siwe disease (LSD)
    (Infants < 2 years)
  - Hashimoto-Pritzker disease
  - Hand-Schüller-Christian syndrome and histiocytosis X .
    (children, young adults)

In 1987, the name Langerhans cell histiocytosis (LCH) was introduced.
A Langerhans cell is a tissue –resident **macrophage**/specialized **dendritic cell** of the skin and mucosa.



9751/1	Preferred	Langerhans cell histiocytosis, NOS
9751/1	Related	Langerhans cell histiocytosis, monostotic
9751/1	Related	Langerhans cell histiocytosis, polystotic
9751/3	Preferred	Langerhans cell histiocytosis, disseminated
9751/3	Synonym	Langerhans cell granulomatosis
9751/3	Related	Eosinophilic granuloma
9751/3	Related	Acute progressive histiocytosis X
9751/3	Related	Histiocytosis X, NOS
9751/3	Related	Hand-Schuller-Christian disease
9751/3	Related	Letterer-Siwe disease
9751/3	Related	Nonlipid reticuloendotheliosis



The term **histiocytosis** is not more closely named after macrophages and dendritic cells primarily because it is an antiquated, umbrella term rooted in 20th-century pathology that was coined *before* the precise functional distinctions between macrophages and dendritic cells were understood. Merriam-Webster +2

"Histiocyte" literally means "tissue cell" (from Greek *histos*, "tissue" + *cyte*, "cell"), a term originally used to describe any large, mononuclear phagocytic cell residing in tissue, regardless of its specific immune

Table B11: Langerhans cell and other dendritic cell neoplasms

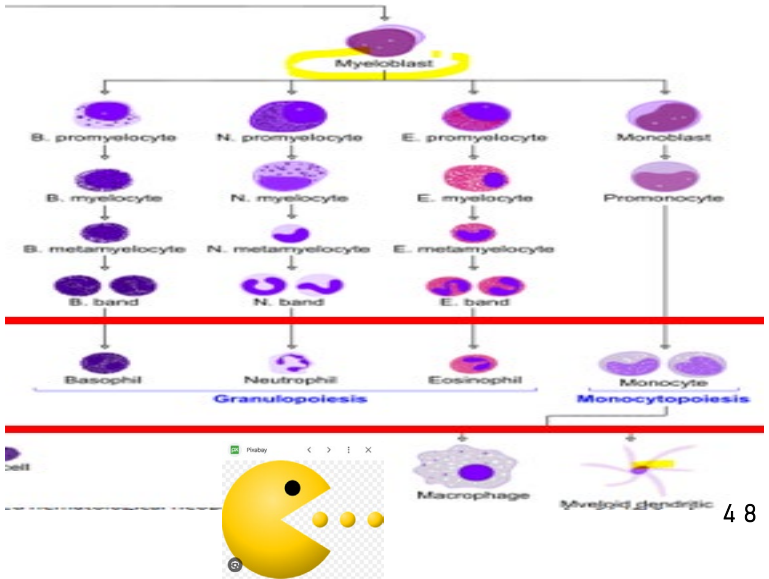
WHO Preferred Term	ICD-O
Langerhans cell neoplasms	
Langerhans cell histiocytosis	9751/1
Langerhans cell histiocytosis, disseminated	9751/3
Langerhans cell sarcoma • Primary Langerhans cell sarcoma • Secondary Langerhans cell sarcoma (following or associated with another hematological neoplasm)	9756/3
Other dendritic cell neoplasms	
Indeterminate dendritic cell tumor	9757/3
Interdigitating dendritic cell tumor	9757/3

Table B12: Histiocyte/macrophage neoplasms

WHO Preferred Term	ICD-O
Histiocytic neoplasms	
Juvenile xanthogranuloma (JXG)	9749/1
Erdheim-Chester disease (ECD)	9749/3
Rosai-Dorfman disease (RDD)	9749/3
ALK-positive histiocytosis	9750/3
Histiocytic sarcoma (HS)	9755/3

These are the tables added relevant to Histiocytosis.

Notice: dendritic cells and macrophage terminology.



Langerhans cell Histiocytosis (LCH) Reportability

9751/3 Langerhans cell Histiocytosis Reportable prior to 2021

9751/1 Langerhans cell Histiocytosis, NOS for 2021+ "Non-Reportable"

2021+ Only LCH **Disseminated/MULTISYSTEM** is Reportable 9751/3

All other forms of LCH are not reportable, except: **Brain/CNS 9751/1**

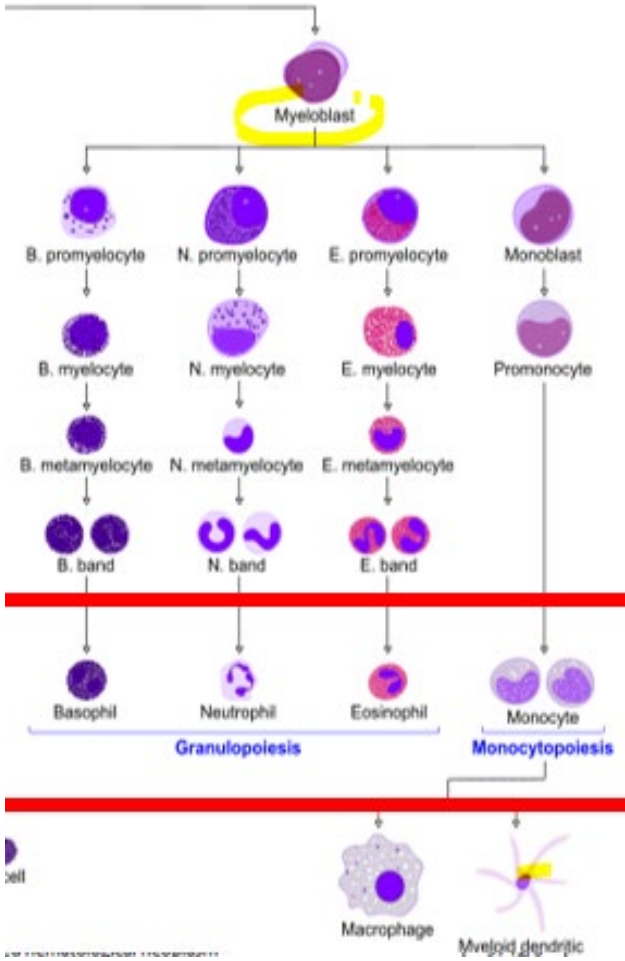
Langerhans cell histiocytosis presents in several different forms

1. DISSEMINATED/MULTISYSTEM: Occurs in infants who present with fever, cytopenia, skin and bone lesions, and hepatosplenomegaly. Pulmonary disease in childhood is clinically variable. The skin, bone, and marrow are preferential sites of involvement. The liver and spleen are also common sites; however, they are typically metastatic and not the primary site. Involvement of bone marrow, liver, and lung are regarded as high-risk factors. (*9751/3*)

2. SOLITARY/UNIFOCAL form: Usually occurs in older children or adults who most commonly present with a lytic bone lesion eroding the cortex. Solitary lesions may occur at other sites as mass lesions or enlarged lymph nodes. Other sites include adjacent soft tissue (skull, femur, vertebra, pelvic bones, and ribs) and less commonly lymph nodes, skin and lung. (Only reportable if benign Brain and CNS, See 9751/1)

3. MULTIFOCAL/UNISYSTEM multifocal disease-Usually occurs in young children who present with multiple or sequential destructive bone lesions often associated with adjacent soft tissue masses. Skull and mandibular involvement are common. Diabetes insipidus follows cranial involvement. (Only reportable if benign Brain and CNS, See 9751/1)

Self-healing Langerhans cell histiocytosis (LCH) of the skin means that the disease regressed without treatment. This is a known phenomenon and is non reportable.



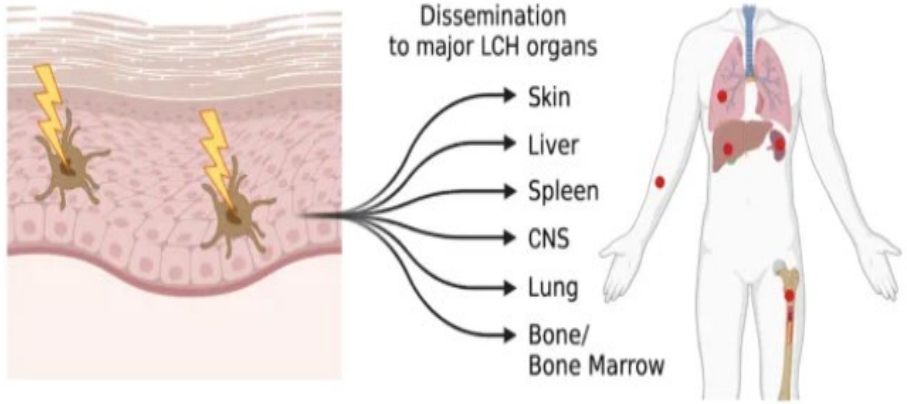
Langerhans cells in Histiocytosis are pathogenic , immature **myeloid** derived **dendritic cells**

Langerhans Cell Histiocytosis (LCH) Neoplasm

Comparison Chart: LCH Classification

	Multifocal Unisystem LCH	Disseminated/Multisystem LCH
Organs Affected	Multiple sites within **one** system (e.g., only bone).	**Two or more** organ systems.
Common Sites	Bone (skull, mandible), skin, lymph nodes.	Skin, bone, liver, spleen, bone marrow, lung.
High-Risk Sites	No (bone/skin/pituitary).	Yes (Liver, spleen, marrow).
Typical Patient	Children 2–10 years.	Infants/young children (<2 years).
Clinical Examples	Hand-Schüller-Christian disease.	Letterer-Siwe disease.
Prognosis	Generally favorable (can spontaneously remit).	Poor; high mortality if untreated.
Behavior (/3)	Historically considered benign (formerly).	Classified as Malignant (9751/3).

Hand Schuller Christian disease can be disseminated/multi systemic involving the skull/bones, lymph nodes, liver, skin, pituitary gland.



Remember: 2021 +

- Only **Disseminated**/**Multisystem** IS REPORTABLE!
- Also, Brain/CNS is reportable.

9751/1	Preferred	Langerhans cell histiocytosis, NOS
9751/1	Related	Langerhans cell histiocytosis, monostotic
9751/1	Related	Langerhans cell histiocytosis, polystotic
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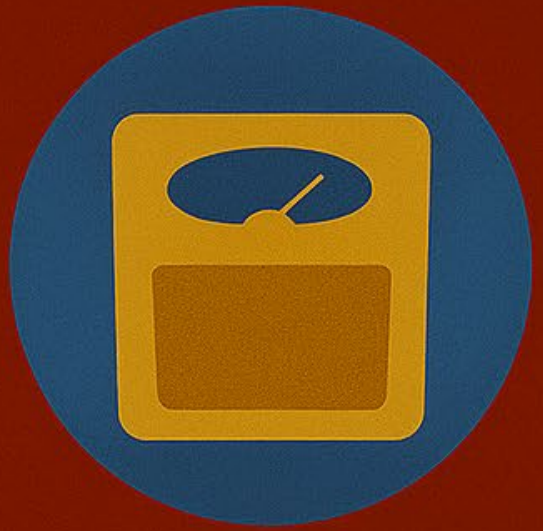
9751/3	Synonym	Langerhans cell granulomatosis
9751/3	Related	Eosinophilic granuloma
9751/3	Related	Acute progressive histiocytosis X
9751/3	Related	Histiocytosis X, NOS
9751/3	Related	Hand-Schuller-Christian disease
9751/3	Related	Letterer-Siwe disease
9751/3	Related	Nonlipid reticuloendotheliosis

CLINICAL PRESENTATION

Fever

Drenching night sweats



Weighing Skein

Rash or kinn lnode

HEMATO Clinical Presentation

B symptoms (fever, drenching night sweats, weight loss)

They are termed "**B symptoms**" because of the **Ann Arbor staging** system used for **lymphomas**. They represent specific **systemic**, "constitutional" **symptoms** that help define the stage of the disease (suffix B), indicating a more advanced or aggressive prognosis compared to "A" (absence of these symptoms).

- A = Asymptomatic: The suffix "A" is used if the patient does not have these specific systemic symptoms.
- B = Symptoms Present: The suffix "B" is added to the stage (e.g. Stage IIB) if the patient presents with any of the symptoms : **Fever** above 38 C (100.4 F), **Drenching Night sweats**, **weight loss** (more than 10% of body weight within 6 months).
- When the B symptoms are present means it is **systemic disease**.

Other symptoms:

LEUKEMIAS: petechiae (red spots), bleeding, infection…

LYMPHOMAS: Painless swelling of lymph nodes, itching, night sweats, dermatitis in skin lymphomas,

MDS: Low blood cells counts (cytopenias) leading to anemia, infection, and bleeding…

MPN: Overproduction of cells, splenomegaly, itching, thrombosis, headache… (**Polycythemia Vera**: severe itching especially after warm showers, vision changes, numbness, tingling, burning, flushing, neurological, risk of blood clots provoking a stroke or heart attack due to high blood viscosity…)

MDS/MPN: Features of both

MULTIPLE MYELOMA: CRAB features: Ca+ elevation (hypercalcemia), Renal failure, Anemia, Bone pain (primarily back/hips).

LANGERHANS CELL HISTIOCYTOSIS: Sings and symptoms depending the site involved. Bone or abdominal pain, skin rash, sores, flaky scalp often mistaken for dandruff, loose teeth, swollen gums, lung symptoms…

TREATMENT

LEUKEMIAS

TREATMENT for Leukemias is in 3 phases

There are three main phases for treatment of Hematopoietic neoplasms, specifically leukemias. These phases may differ based on the histology, age of the patient, risk factors and comorbidities.

1. **Induction**
   The goal of induction therapy is to get the leukemia to go into remission. This is achieved by finding no leukemia cells in a bone marrow biopsy. Induction is not a cure as there may still be leukemia cells in the body. Induction chemotherapy usually last for a month. Several different treatment regimens may be used during this phase to achieve remission.

   1 month

2. **Consolidation/Intensification**
   If remission is achieved through the induction phase, another phase of chemotherapy is given. This phase usually includes the same drugs that were given for the induction phase. This phase usually lasts for several months. Drugs are given in high doses, and the treatment is fairly intense. During this phase patients may also have a bone marrow or stem cell transplant

   Several months

3. **Maintenance**
   Maintenance therapy can last up to two years.

   UP to 2 years

TREATMENT

First course for HEMATOLYMPHOID

FIRST COURSE of therapy ENDs when the treatment PLAN is COMPLETED or REMISSION is **achieved**!
No matter how long it takes to complete the plan

EXAMPLE:

-Patient had INDUCTION therapy but did **not** achieve complete **remission**.
Disease progressed and the patient received autologous **stem cell transplant**.
This is still first course treatment since the patient had not achieved remission.

TREATMENT: PSORALEN + Ultraviolet A = **PUVA**

PUVA (Code as Other therapy)

Subcategory
code to Other Therapy when used in combination with *Ultraviolet* A light (PUVA therapy)
Photosensitizer

NSC Number
45923
045923

Primary Site
None

Histology
None

Remarks
A photoactive agent (photosensitizer). FDA approved use as palliation in cutaneous T-cell lymphoma.

Coding
This drug should be coded

Name	Category	Primary Site	Code?
Ultraviolet A	Other therapy		Yes
Beta Carotene	Ancillary Agent		No
Psoralen	Ancillary Agent		Yes

To treat CUTANEOUS T-cell Lymphomas:
- Mycosis Fungoides
- Sezary Syndrome
- Cutaneous anaplastic large cell lymphoma

Psoralen is a natural photoactive compound found in plants (like citrus, figs, cloves…) that acts as a **photosensitizer**.
It is administrated orally or topically and upon exposure to UVA light damages the DNA of the cancer cells causing them to die.

Subcategory
code to Other Therapy when used in combination with Ultraviolet A light (PUVA therapy)
Photosensitizer

Treatment…

Collect **phlebotomy ONLY** for polycythemia vera (9950/3). produces too many red blood cells (RBCs

Collect **blood-thinners** and/or **anti-clotting agents ONLY** for essential thrombocythemia (9962/3).

high platelet count

Aspirin is antiPLATELET

Name	Category	Primary Site	Code?
Aspirin	Other therapy	Essential Thrombocythemia	Yes

Remarks

Aspirin is approved treatment for Essential Thrombocythemia (ET) ONLY. The therapeutic dose for essential thrombocythemia is in the range of 70-100 mg/day (this is considered low-dose).

Aspirin for cardiovascular protection and pain control is in the range of 160 mg/day and higher.

Hydroxyurea in both conditions act as a cytoreductive agent, reducing elevated blood cell counts to prevent clotting and thrombosis.

Treatment

VITAMIN A

Retinoids, particularly ATRA (All-Trans Retinoic Acid) are able to treat leukemia and lymphoma primarily by **forcing immature**, rapidly dividing **cancer** cells to **mature** (**differentiation**) and **cancer cells** to undergo programmed cell death (apoptosis).
Also, Retinoids as ATRA **can break down some abnormal fusion proteins** (PML/RARa) formed during abnormal chromosome exchange as translocation t(15;17)

TREATMENT

Vitamin A code in OTHER THERAPY

ATRA-IV (Vesanoid) vs ISOTRETINOIN vs Ali tretinoin

Remarks

Atra is currently in clinical trials for Head & Necks cancers. It is under investigation as treatment of Kaposi Sarcoma. DO NOT CODE *ATRA* has treatment for these sites/histologies until FDA approval. Code as either 2, 3, or 6 in Other Therapy field.

ATRA has received FDA approval for treatment of Acute Promyelocytic Leukemia (APL). *ATRA* should be coded "1" in Other Therapy field.

Liposomal all-trans retinoic acid. Antigenics, Inc.

Coding

This drug should be coded

In Leukemia, FORCES immature, cancerous blood cells to MATURE into normal, functional WBCs,.
APL (Acute Promyelocytic Leukemia)

Name	Category	Primary Site	Code?
ATRA-IV	Differentiation inducing agent	Lymphoma, Acute promyelocytic leukemia	Yes

Name	Category	Primary Site	Code?
Ali*tretinoin*	Differentiation inducing agent	Breast, Kaposi sarcoma, leukemia, sarcoma, skin cancer	No
Accutane	Differentiation inducing agent	Prostate cancer	No

TREATMENT

TRETINOIN =ATRA = VESANOID

Remarks

Tretinoin is FDA approved for treatment of acute promyelocytic leukemia (APL). It is a retinoid/differentiation agent and at this time the precise mechanism of action has not been fully elucidated so it cannot be assigned to chemotherapy, hormone or BRM/ immunotherapy. Code this agent as treatment in the "Other Therapy" data field (using code 1).

Phase II (head & neck)
Administered topically, *tretinoin* stimulates epithelial turnover and reduces epithelial cell cohesiveness. Under investigation for treatment of Kaposi sarcoma lesions. FDA approved therapy of acute promyelocytic leukemia. NCI.Code this agent as treatment in the "Other Therapy" data field (using code 1).

Name	Category	Primary Site	Code?
Tretinoin	Differentiation inducing agent	Acute Promyelocytic leukemia (APL), AIDS, head & neck cancer, Opportunistic infection	See Remarks
Ali*tretinoin*	Differentiation inducing agent	Breast, Kaposi sarcoma, leukemia, sarcoma, skin cancer	No
Accutane	Differentiation inducing agent	Prostate cancer	No

Name

Tretinoin

Alternate Names

Aberel
Airol
All-trans-retinoic acid
Avita
Dermairol
Eudyna
Renova
Retin-A
Retinoic acid
Retisol-A
Stieva-A
Vesanoid
Vitamin A Acid
Vitinoin

Abbreviations

ATRA
Vit A

TREATMENT

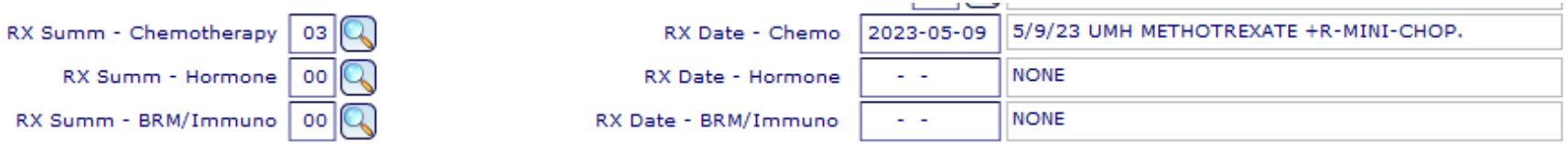
ATO (Arsenic trioxide)

Trisenox is FDA approved use on **Acute Promyelocytic Leukemia** and **Multiple myeloma**
It should be coded as Chemotherapy

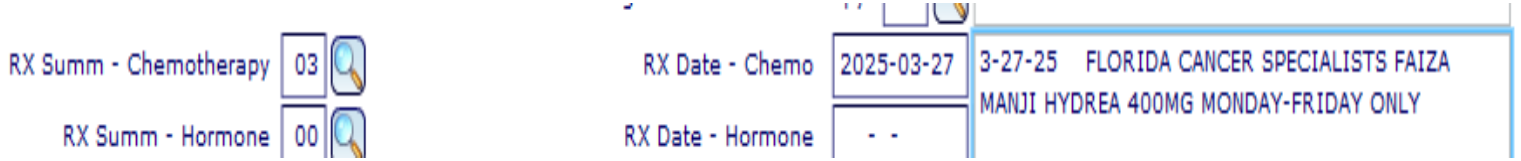
COMMON ERRORS IN **TREATMENT** CODING TEXT"

Do **NOT** enter the Protocol ACRONYM **ONLY** when coding.

- Please specify individual agents in each section (Chemo, Hormone, Immunotherapy).



- Very often 03 is coded in Chemotherapy for multiple medication.
  And only 1 chemotherapy agent is written in text!
  Most of the time when we point out the discrepancy (especially in NO hematic cancer), there are more chemo agents to be added.



- Name ONLY chemotherapy agents in CHEMOTHERAPY. Name ONLY hormone agents in HORMONE text. Name ONLY IMMUNOTHERAPY agents in corresponding text section.

HEMATOPOIETIC MULTIPLE PRIMARY CALCULATOR

NEVER start with the Multiple Primary Calculator

Multiple Primaries Calculator

⚠ **Do not start with the Multiple Primaries Calculator. Rule M15 is not a default and should only be used if none of the earlier rules (M1–M14) apply.**

1. Start with the database first and gather as much information on your histology as you can.
2. Determine if you have a histology that transforms to another histology, or a histology that has another histology transforming to it. If you have either one of these, rules M8-M13 would apply.
   - **Do NOT apply Rule M15 to histologies that have transformation information.** See Transformations: Chronic and Acute Neoplasms.
3. Carefully go through all the rules (M1-M14) in chronological order to determine which one is applicable. STOP at the FIRST applicable rule
   - Some registrars are getting to rule M15 when one of the other rules from M1-M14 actually apply, so read the rules carefully.

⚠ **Do NOT compare the number of primaries determined from Rule M1-M14 with the Multiple Primaries Calculator.**
- **It is only appropriate to use the Multiple Primaries Calculator when the applicable M rule specifically instructs one to use it**

- **DATABASE** first for Histology

IMPORTANT NOTE: ALWAYS REVIEW TRANSFORMATION INFORMATION BEFORE GOING TO THE M RULES.

When determining if you have multiple primaries, always check the "Transformations to" and the "Transformation from" fields in the hematopoietic database. The acute neoplasm is listed in the "Transformations to" section; whereas histologies listed in the "Transformations from" section are the chronic neoplasms.

If the "transformation to" AND "transformation from" fields are BOTH "none," then the neoplasm is NOT a chronic or an acute neoplasm per our rules AND Rules M8-M13 will not apply. Physicians may still document a transformation though.

Rules M8-M13
These rules are only for the histologies that have transformations listed in the Heme DB.

- Then Hemato **MANUAL** for rules M1-M14
- Then Rule M15 **MULTIPLE** PRIMARY **CALCULATOR**

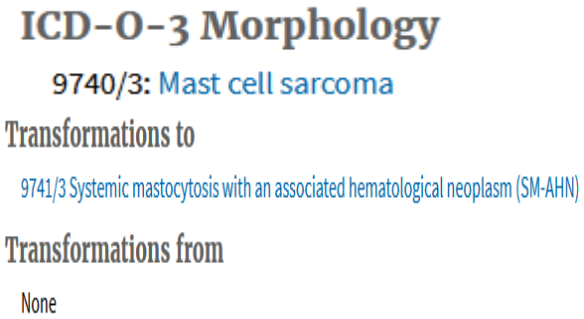
Instructions for Multiple Primaries

- **First: DATABASE** for Histology and look if there is **TRANSFORMATION** involved (**M8-M13**).
- **Second**: Hemato **MANUAL** for rules **M1-M14** and **STOP** at the first rule that applies.
- **Last**: Then Rule **M15:** **MULTIPLE** PRIMARY **CALCULATOR**

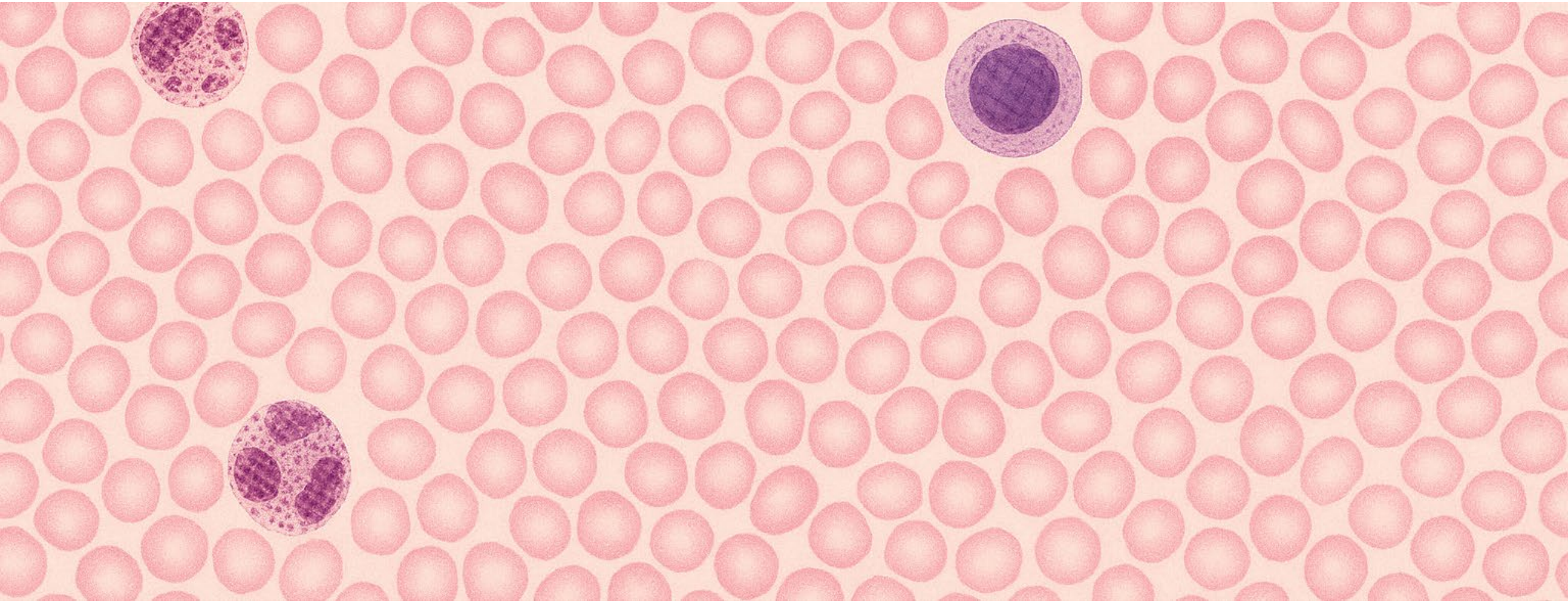
Summary of the Multiple Primary Rules

Use this as a guide only, do not use this summary to determine the appropriate M rule.

Multiple Primary Rule	Covers	Comments
M1 (Single primary)	Limited information, death certificate only cases	
M2 (Single primary)	Same histology, same primary	Exception for 9699/3, need to review
M3 (Single primary)	Myeloid sarcoma and Acute Myeloid leukemia or Mast cell sarcoma and Mast cell leukemia	Histologies: 9740, 9742, 9930, and all AML histologies
M4 (Single primary)	Two or more NHL's diagnosed in the same biopsy	See PH11 or PH15 for histology
M5 (Single primary)	Hodgkin and non-Hodgkin in the same biopsy	See PH14 for histology
M6 (Multiple primaries)	Hodgkin and non-Hodgkin in different biopsies	
M7 (Single primary)	More specific histology and NOS histology	*Example:* Follicular lymphoma (specific histology) and NHL, NOS (NOS histology) Review same primaries section in the Hematopoietic database
M8 (Single primary)	Chronic and acute neoplasm diagnosed at the same time and there is one biopsy	Histologies must have information in the Transformation to or Transformation from field in the database
M9 (Single primary)	Chronic and acute neoplasm diagnosed at the same time and there is no documentation of the biopsy(ies)	Histologies must have information in the Transformation to or Transformation from field in the database
M10 (Multiple primaries)	Chronic and acute neoplasm diagnosed at different times	Histologies must have information in the Transformation to or Transformation from field in the database
M11 (Multiple primaries)	Chronic and acute neoplasm diagnosed at the same time and there are two biopsies	Histologies must have information in the Transformation to or Transformation from field in the database
M12 (Single primary)	Acute neoplasm reverts to chronic neoplasm, no (or unknown), treatment given for acute neoplasm	Histologies must have information in the Transformation to or Transformation from field in the database
M13 (Multiple primaries)	Acute neoplasm reverts to chronic neoplasm and treatment given for the acute neoplasm	Histologies must have information in the Transformation to or Transformation from field in the database
M14 (Single Primary)	Post transplant lymphoproliferative disorder (9971/3) with an accompanying hematopoietic neoplasm	See PH1 for coding histology
M15 (Multiple primaries calculator)		See Multiple Primaries Ca



Hematopoietic and Lymphoid systems



… TO BE CONTINUED!

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