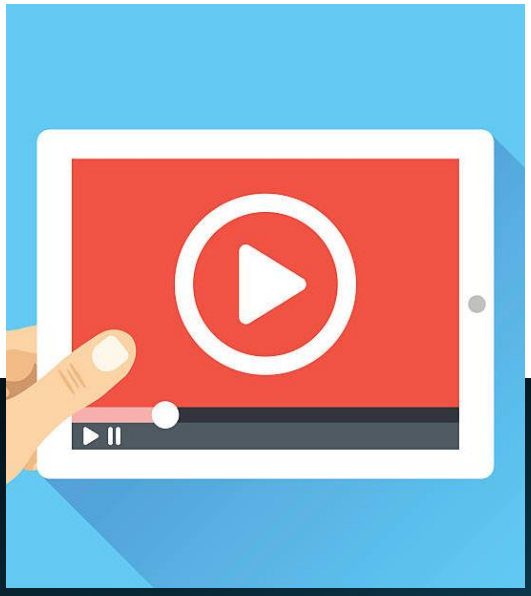
Heme/Lymph:
Myeloid Neoplasms



ICR Video Training Series | Iowa Cancer Registry

Presented by Melissa Riddle, ODS-C

March 2025

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1

The NORM:

The activities of the blood may be categorized as:

- Transportation
- Regulation
- Protection

These functional categories overlap and interact as the blood carries out its role in cellular functions.

2

2

Blood composition: Plasma

- About 55% of blood volume
- Watery fluid portion of blood (90% water)
- Transports nutrients & waste throughout the body
- Proteins, electrolytes, carbohydrates, minerals, and fats are dissolved in it.

3

3

Blood Composition : Formed Elements

RBC	WBC	Platelet
• Carry Oxygen • Carry CO2 • Gives color	• Fights infection • Excess weakens immune system	• Clotting

4

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Hemopoiesis

- The production of formed elements, or blood cells:
  - Before birth, hemopoiesis occurs primarily in liver and spleen, thymus, lymph nodes, bone marrow.
  - After birth, blood cells formed in bone marrow
- Blasts are new, immature blood cells
  - Blasts mature in the bone marrow or mature in other parts of the body
- Blood cell production: orderly, controlled
- Blood diseases develop when the body produces large numbers of abnormal blood cells

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5

Hemopoiesis

- Stem cell (“Master Cells” that generate other differentiated cell types. Each tissue within the body contains a unique type of stem cells that renew and replace that tissue)
  - "daughters" of stem cells becomes a precursor cell, either a lymphoid cell or a myeloid cell
  - mature into various blood cells

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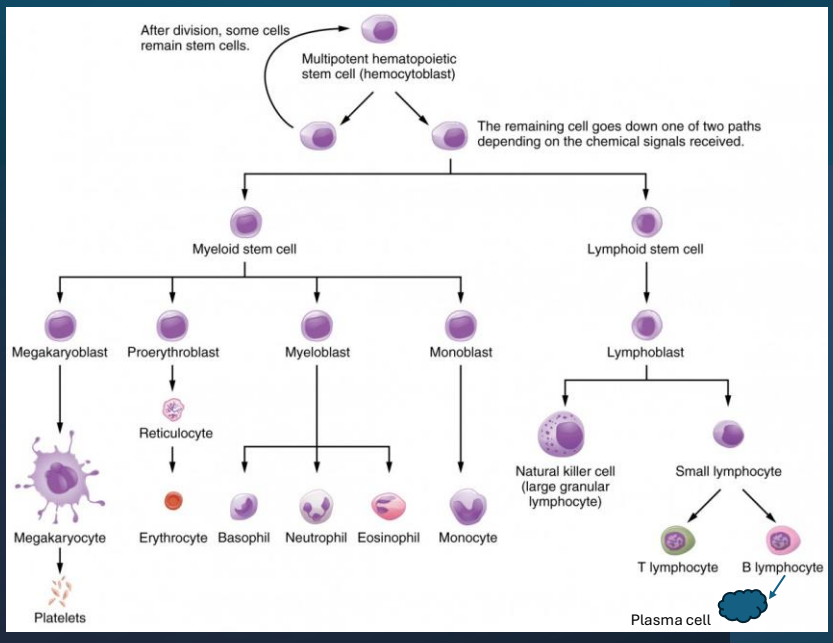

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Hematopoietic Cell Types

Cell Type	Formation	Function	Types
Red blood cells (erythrocyte/RBC)	Made in Bone Marrow Seen in peripheral blood	Contains hemoglobin (protein) Transport oxygen from lungs through body	
White blood cells (leukocyte/WBC)	Made in Bone Marrow Found in blood & lymph tissue	Immune system Helps fight infection	Granulocyte • Neutrophils • Eosinophils • Basophils Monocytes Lymphocytes • B cell • T cell
Plasma cells (plasmacyte)	Develop from activated B cells	Make large amounts of specific antibody	
Platelets (thrombocyte)	Breaking off of a large cell in bone marrow Found in peripheral blood and spleen	Form blood clots Slow/stop bleeding Help wounds heal	

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Assigning Primary Sites

Draw blood with a small needle stick in the arm?

Code C42.1

Suck out bone marrow with a giant needle stick in the hip?

Code C42.1

Cut out or biopsy a hard, palpable, or shotty lymph node?

Code C770-C779

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Assigning Histology

- Histology code is based **SOLELY** on the pathologist's diagnosis
  - Managing physician statement or treating diagnosis
- Do **NOT** go through the path report looking at genetics or immunophenotyping to determine histology
  - The purpose of immunophenotyping or genetics information in the Heme DB is to help with determining **Diagnostic Confirmation**
    - Do not use it to determine your histology code alone or as the only source
    - The pathologist or managing physician must make the diagnosis

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Assigning Histology

- Do not try to make a diagnosis based on minor or major criteria that is included in the Heme DB
  - This information is used by pathologists to determine histology
  - It was included as additional information
  - This will be removed from the Heme DB for the next major update

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Schemas

Schema	Histology	Schema Discrim	Notes
HemeRetic 00830	9591	1, 2	
	9724, 9727, 9740-9742, 9749, 9751, 9755-97599762-9809, 9811-9820, 9831-9920, 9930, 9931-9993		
	9724, 9727, 9740-9742, 9762-9765, 9767-9809, 9831-9920, 9931-9993		C700-C729, C751-C753
	9749, 9766		**2018-2022** C700-C729, C751-C753
Plasma Cell Disorders 00822	9671, 9734 9731,9734, 9761		
	9671		**2018-2022** C700-C729, C751-C753
Plasma Cell Myeloma 00821	9732		

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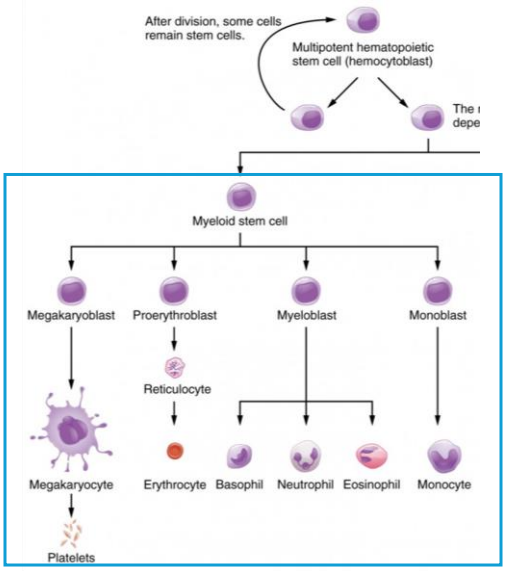
Myeloid Neoplasms

- WHO lists major subgroups:
  - Acute myeloid leukemia AML
  - Myelodysplastic syndromes (MDS)
  - Myeloproliferative neoplasms (MPN)
  - Myeloid and lymphoid neoplasm with eosinophilia and abn of PDGFRA, PDGFRB or FGFR1



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Myeloproliferative Neoplasm (MPN)

- **MPN unclassifiable** (MPN, U) – 9975 (2010+)
  - Studies performed at 6-12mo can often provide sufficient information on a more precise classification
    - If a more specific MPN is diagnosed change the histology to the more specific
    - Specific types:
      - Chronic eosinophilic leukemia, NOS 9964
      - Chronic myelogenous leukemia BCS ABL1 positive 9875
      - Chronic neutrophilic leukemia 9963
      - Essential thrombocythemia 9962
      - Polycythemia vera 9950
      - Primary myelofibrosis 9961

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Myelodysplastic Syndrome (MDS)

- **MDS unclassifiable** (NOS) – 9989
  - Generic disease description
  - Previously known as "preleukemia"
  - Provisional diagnosis and further diagnostic procedures are performed and identify a more specific MDS
    - Specific types:
      - MDS with excess blasts 9983
      - MDS with multilineage dysplasia 9985
      - MDS with ring sideroblasts and single lineage dysplasia 9982
      - MDS with sinle lineage dysplasia 9980
      - Refractory cytopenia of childhood 9985
      - Refractory neutropenia 9991
      - Refractory thrombocytopenia 9992
      - MDS with ring sideroblasts and multilineage dysplasia 9993

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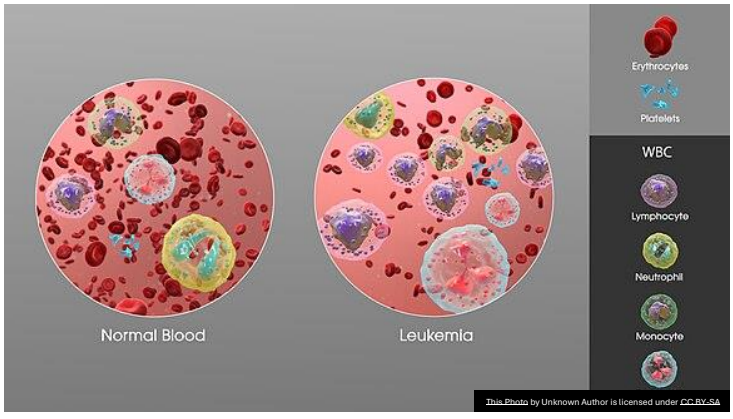
Leukemia

- Usually involves white blood cells (WBC)
  - Bone marrow produces and excessive amount of abnormal WBC which don't function properly
- Symptoms:
  - Fever/chills
  - Persistent fatigue/weakness
  - Frequent or severe infections
  - Unplanned weight loss
  - Swollen LN, liver or spleen
  - Easy bleeding or bruising – recurrent nose bleeds
  - Petechiae skin (tiny red spots)
  - Excessive sweating – night sweats
  - Bone pain or tenderness



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Myeloid Leukemia

- Affects the myeloid cells typically responsible for developing various types of mature blood cells like
  - Red blood cells
  - White blood cells
  - Platelets
- Chronic (CML) - abnormal growth of WBCs
  - Most have a gene mutation – Philadelphia chromosome (BCR-ABL1)
- Acute (AML) – starts in bone marrow or other types of blood-forming cells
  - Symptoms come on quickly over days to weeks and disease progresses quickly

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Leukemic Cutis

- Rare infiltration of neoplastic leukocytes into the epidermis, dermis, or subcutis from an existing leukemia that results in clinically identifiable cutaneous lesions
  - An advanced phase of the leukemia having poor prognosis
  - Most often diagnosed via skin biopsy – punch, shave, etc.; utilizing IHC/biomarker testing
- Commonly associated with CMML and AML
  - Code to the specific type of systemic leukemia
- If only leukemic cutis is known code to **C421, 9800/3**
  - Follow back to determine the specific systemic leukemia histology

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Polycythemia

- Increase in red blood cells
  - Measure of hematocrit levels
- Alternate names:
  - Erythrocytosis
  - Familial polycythemia
    - Occurs in families – genetic changes
  - Idiopathic polycythemia
  - Secondary polycythemia
- **Polycythemia Vera 9950 – Reportable**
  - Must state PV

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Ancillary Studies

Immunophenotyping:

- Technique used to study the protein expressed by the cells
  - IHC stains
  - Flow cytometry analysis (fresh tissue)

Cytogenetics:

- Study of chromosomes and how genetics play a role in development and progression of certain diseases
  - FISH

Molecular diagnostics:

- Collection of techniques to analyze biological markers in the genome and proteome, how their cells express their genes as proteins
  - PCR (common), southern blot, gene sequencing

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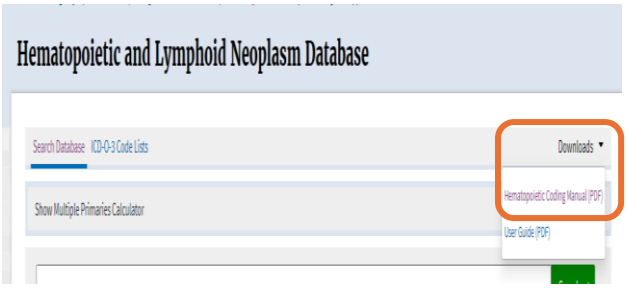
Heme-Lymph Manual



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

2. Hematopoietic & Lymphoid Neoplasm Coding Manual (PDF, 1.0 MB)
   a. Reportability instructions and rules for determining the number of primaries, the primary site and histology, and the cell lineage or phenotype
   b. The introduction to the manual has an updated Steps in Priority Order for using the Hematopoietic and Lymphoid Neoplasm Coding Manual & Database.

Do **NOT** use STORE or STR for Heme/Lymph cases
Use the Heme/Lymph Manual and DB

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Diagnostic Confirmation

Description	Code
Positive Histology • Peripheral blood smear; Microscopic tissue	1
Positive Cytology – rare	2
Positive Histology (1) + Positive Immunophenotyping AND/OR Positive Genetic studies	3
Positive microscopic confirmation, unknown type (not 1-3)	4
Positive lab/tumor marker	5
Direct visualization without microscopic confirmation	6
Radiology/Imaging confirmation	7
Clinical diagnosis only (other than 5-7)	8
Unknown whether microscopic confirm or not	9

Leukemia **ONLY:** CBC, WBC

Includes peripheral blood smear follow by flow cyto

+Immunophenotype/Genetic study w/OUT micro confirmation

Physician statement

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Transformation

- A disease where histology actually changes.
  - Chronic phase leukemia progressing (transforming) to acute phase
    - More aggressive
  - **Rules M8-13** cover transformations

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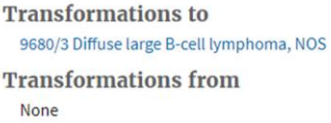
Transforms to…

- Chronic **transforms to** an acute/more severe neoplasm
  - Heme DB shows the acute neoplasm in the **transforms to** section.
- Example: CLL/SLL 9823/3
  - ➢**Transforms to** section shows 9823/3 **transforms to** 9680/3
  - ➢This means the CLL/SLL is a chronic neoplasm and the DLBCL is an acute neoplasm



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Transforms from…

- Acute neoplasms may have multiple histologies listed in the **transforms from** field.
  - Histologies listed in the **transformations from** field are chronic.
- Example: Plasma cell myeloma 9732/3 (acute)
  - ➢**Transforms from** lists 9731/3 and 9734/3 (chronic)

Transformations to

None

Transformations from

9731/3 Solitary plasmacytoma of bone
9734/3 Extraosseous plasmacytoma

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Ambiguous Terms

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"Consistent With"

- "*Consistent with*" is historically and currently considered ambiguous terminology
  - Becoming the standard of reporting Heme diagnoses
- For Heme Neoplasms **ONLY**
  - "Consistent with" is a definitive diagnosis
  - This is **NOT** an ambiguous term

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Ambiguous Terms

DO use	**DON'T** use
Ambiguous Terms for Reportability and Casefinding • Heme Manual, page 27, #4	Ambiguous Terms to code specific histology • Heme Manual, pg 41, #3 • **Example**: Myeloproliferative disease, probably polycythemia vera; code MPN, NOS. [9975/3]

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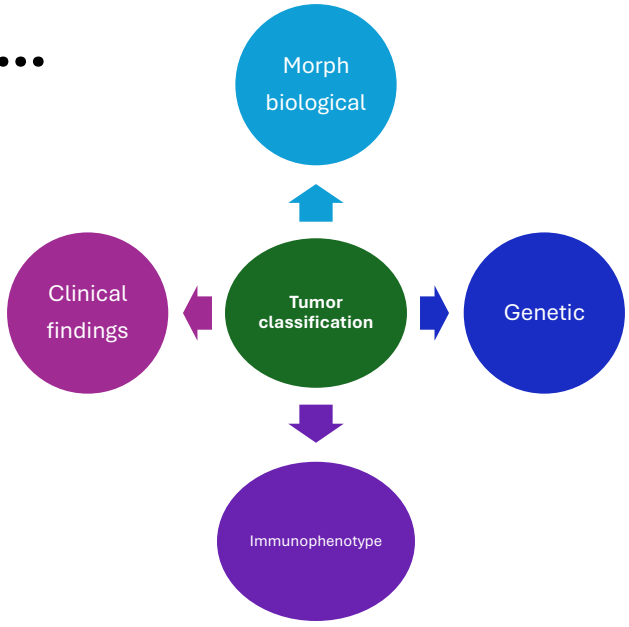
Remember...



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First Course Treatment

- Varies by the type of hematopoietic neoplasm
- Multiple Primary Rules: **M10-M13**
  - Transformation rules that are affected by treatment
    - Refers to the patient receiving at least one form of cancer-directed treatment such as surgery or systemic therapy
    - It does **NOT** include passive treatment like supportive care or observation

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Other Treatment

- Do **NOT** collect blood transfusions as treatment
- Phlebotomy is treatment for <u>polycythemia vera</u> **ONLY**
- Blood-thinners and/or anti-clotting agents for <u>essential thrombocythemia</u> **ONLY**

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Treatment

- Leukemias
  - Chemo
  - Bone marrow transplants
  - Biological Response Modifiers
  - Hormones
- Other
  - Do not collect blood transfusions (whole blood or platelets) as treatment. Blood transfusions are widely used to treat anemia, and it is not possible to collect this procedure in a meaningful way.

Donor Leukocyte Infusion

- **Donor Leukocyte infusions (DLI)**:
  - AKA: Buffy coat infusion
  - Following a Hematopoietic Stem-Cell Transplant (HSCT)
  - Treatment of heme neoplasms, specifically leukemias, is increasing
- Code as **immunotherapy** when reportable heme neoplasm is treated with donor leukocyte infusion, even if not listed in treatment section of Heme DB for that specific neoplasm

Chemotherapy

- Gleevec (Imatinib) for CML
- Revlimid, Velcade for myeloma
- Campath is monoclonal antibody against the antigen CD52 found on T and B cell lymphocytes in CLL.
- Mylotarg approved for AML
- Bexxar and Zevalin to treat relapsed B-cell NHL.

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External Beam Radiation

- Used most often before transplant
- Usually given with chemotherapy
- Radioimmunotherapy, radioactive molecule attached
- Given alone or with chemo for stem cell transplant

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Hemat Transplant/Endo Procedure

- **Conditioning** = High doses of chemo with or without radiation given to destroy normal BM cells
- **Rescue** = Stored stem cells given back to patient through infusion (actual BMT or PBSCT)
- **Harvest** = collection of stem cells from either the bone marrow or bloodstream
- **Bone Marrow Transplant** = stem cells taken from bone marrow
- **Stem Cell Transplant/Peripheral Blood Stem Cell Transplant** = stem cells taken from the bloodstream

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Hemato Transplant and Endocrine Proc

- **Bone Marrow Transplant** (BMT) **10**
  - Allogenic – From a donor **12**
  - Autologous – From patient **11**
  - Syngeneic – From patient's twin
    - *Coded as Allogenic*
- Induction/Conditioning, remission, and maintenance drugs are recorded.
- **Code 88** when:
  - Oncology referral for heme transplant
  - Only a harvest was performed not the rescue

40

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Hemato Transplant/Endo Procedure

- **Stem Cell Transplant/PBSCT 20**
  - Replinish supply of health blood-forming cells
    - BMT
    - PSCT
    - Umbilical cord blood transplant
- **Code 88** when:
  - Only a harvest performed, unknown if rescue done
  - Oncology referral for stem cell transplant or other hematopoietic transplant

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Immunotherapy

- Patients' immune system fights the cancer.
- Includes antibodies, vaccines, and monoclonal antibodies (MoAbs).
- Remember SEER*Rx

https://seer.cancer.gov/seertools/seerrx/

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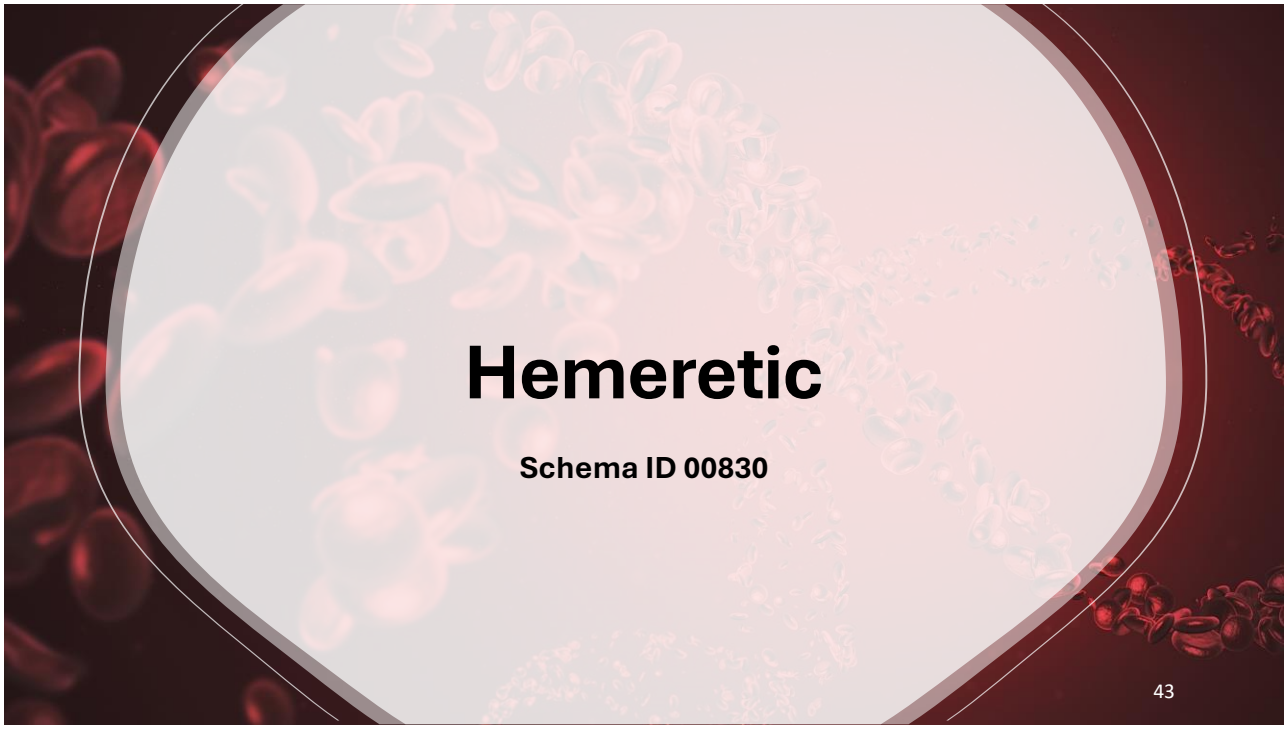

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Heme Retic Schema | Coding

Schema ID 00830	Always…
Tumor Size Clinical	999
Tumor Size Path	999
Tumor Size Summary	999
Reg Nodes Pos/Exam	99/99
LVI	8
EOD Pri Tumor	__ __ __
EOD Reg Nodes	888
EOD Mets	88
SS18	__
Grade Clinical/Path/yc/yp	8

100 Local
700 Systemic (see codes given)
999 Unknown

1 Local
7 Distant
9 Unkn if ext or mets

44

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Heme Retic Schema | SSDI - JAK2

Description: Janus Kinase 2 (JAK2, JAK 2) is a gene mutation that increases susceptibility to several myeloproliferative neoplasms (MPNs).

Note 1: Phys statement of Jak2 can be used when no other info.

Note 2: Looking for gene mutation. JAK2 done on whole blood

Note 3: Record JAK2 for any heme neoplasm.

Most used for the following histologies:

- Polycythemia Vera (9950/3)
- Primary myelofibrosis (9961/3)
- Essential Thrombocytopenia (9962/3)
- Chronic myelomonocytic leukemia (9945/3)

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JAK2

Code	#3862 JAK2
0	JAK2 stated as neg
1	JAK2 pos for mutation V617F WITH or WITHOUT other mutations
2	JAK2 pos for exon 12 mutation
3	JAK2 pos for other specified mutation
4	JAK2 pos for more than one mutation other than V617F
5	JAK2 pos NOS, specified mutation not stated
7	Test ordered, results not in chart
9	Not documented in med record, not assess, unknown if assessed

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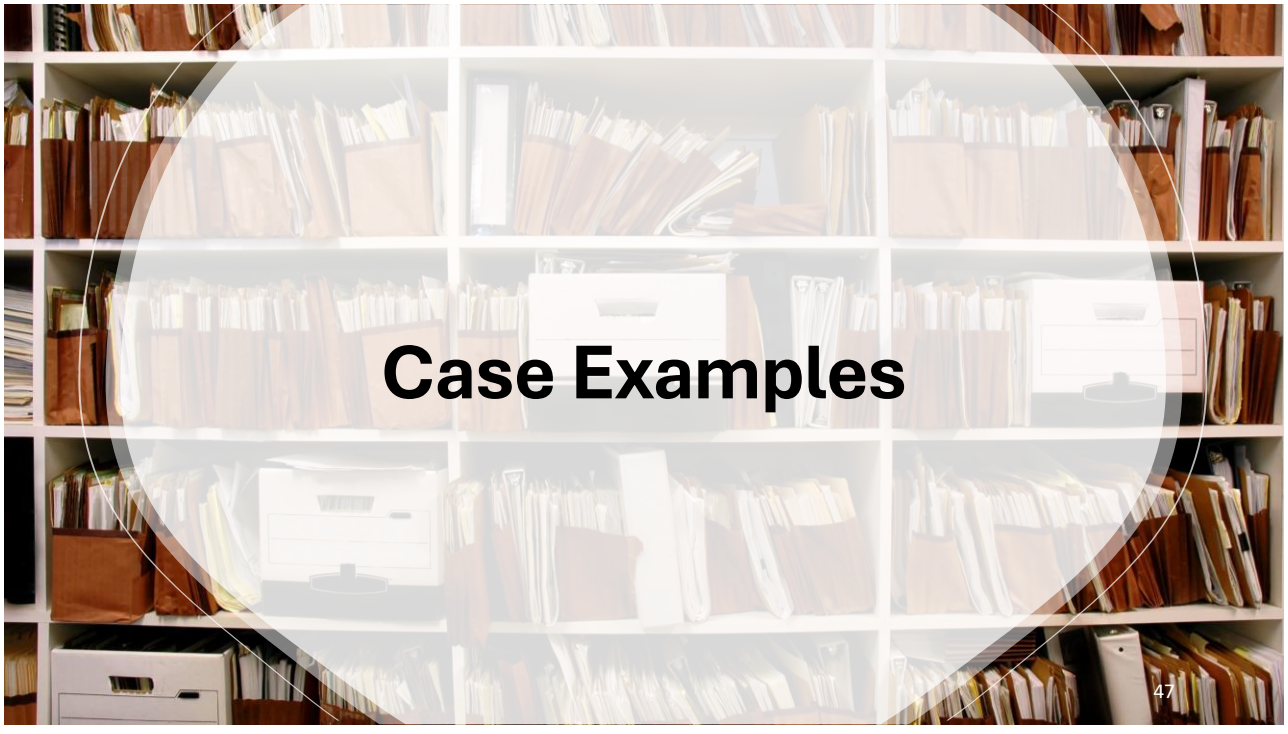

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Case Scenario 1

- 2022 Acute Myeloid Leukemia diagnosed and is in your registry database
- Presents back to your facility in 2024 with a new diagnosis of Myeloid Sarcoma.

How many primaries?

Is the 2024 case a new primary?

48

48

Case Scenario 2

- Bone marrow biopsy: high grade myeloid stem cell neoplasm, 17% blasts by differential count; this could be classified as MDS w/ increased blasts (MDS-IB2) per the WHO 5th edition classification, or MDS/AML per ICC
- FISH and cytogenetics reveal loss of 7q but no other AML related genetic abnormalities
- Physician confirms MDS/AML

What is the appropriate histology code?

A. MDS w/ increased blasts 9983

B. AML 9861

50

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Case Scenario 3

1/12/2024 Bone marrow biopsy: Myelodysplastic syndrome, mutated RUNX1

Oncology note: Patient has MDS with 5q deletion and acute myelogenous leukemia with mutated RUNX1

How many primaries?

Is this case reportable?

Primary Site(s):

Histology(ies):

52

52

1/12/24 Bone marrow biopsy: MDS; FISH shows deletion of long arm chromosome 5 (5q del)

1/25/24 Treated with Lenalidomide

7/23/24 Bone marrow biopsy: Acute myelogenous leukemia; t(8;21)(q22;q22.1) resulting in RUNX1-RUNX1T1

Physician states patient has MDS 5q del and AML with mutated RUNX1

How many primaries?

Primary Site(s):

Histology(ies):

Case Scenario 4

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SEER*Educate

Training | Coding CEs

- **Dx 2018-2024**
  - Heme Series 1-6
    - 5 cases each series

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Questions

Contact Info

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