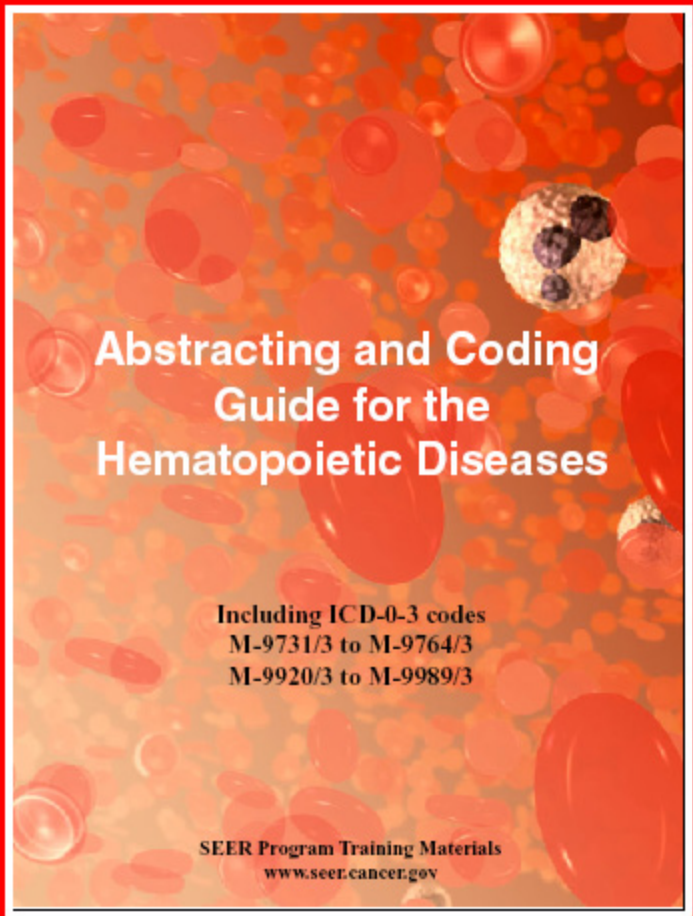

Abstracting and Coding Guide for the Hematopoietic Diseases

Including ICD-0-3 codes
M-9731/3 to M-9764/3
M-9920/3 to M-9989/3

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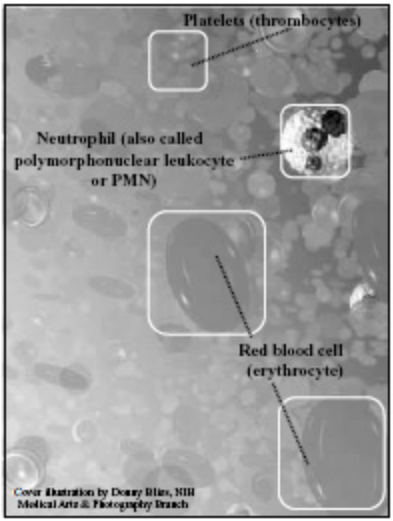
Platelets (thrombocytes)
Neutrophil (also called polymorphonuclear leukocyte or PMN)
Red blood cell (erythrocyte)
Cover illustration by Donny Bliss, NIH Medical Arts & Photography Branch

ABSTRACTING AND CODING

GUIDE FOR THE

HEMATOPOIETIC DISEASES

Including ICD-O-3 codes
M-9731/3 to M-9764/3
M-9920/3 to M-9989/3

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May, 2002

Please note
All of the materials in this document have been researched carefully and coded based on the best information and expertise available. The SEER Program is not responsible for errors in coding nor for misinterpretation of the information provided.

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CONTENTS

Disease profiles are listed in ICD-O-3 order. Only the preferred term is listed below. See Diagnosis Index for other terms.

Introduction iii

DISEASE PROFILES 1

* *indicates a new code or new diagnosis in ICD-O-3 that is reportable as of 01/01/01*

Plasma cell tumors

9731/3 Plasmacytoma 2
9732/3 Multiple myeloma 3
9733/3 Plasma cell leukemia* 4
9734/3 Plasmacytoma, extramedullary (not occurring in bone)* 5

Mast cell tumors

9740/1 Mastocytoma 6
9740/3 Mast cell sarcoma 7
9741/3 Malignant mastocytosis 8
9742/3 Mast cell leukemia***** 9

Neoplasms of histiocytes (macrophages) and accessory lymphoid (antigen presenting) cells

9750/3 Malignant histiocytosis 10
9751/1-9754/3 The Langerhans cell histiocytoses* 11
9755/3 Histiocytic sarcoma* 13
9756/3 Langerhans cell sarcoma* 14
9757/3 Interdigitating dendritic cell sarcoma* 15
9758/3 Follicular dendritic cell sarcoma***** 16

Immunoproliferative diseases

9760/3 Immunoproliferative disease 17
9761/3 Waldenstrom macroglobulinemia 18
9762/3 Heavy chain diseases 19
9764/3 Immunoproliferative small intestinal disease 20

Selected myeloid leukemias

*(**Note:** acute lymphoid and myeloid leukemias {M-9800 to M-9910} were reportable prior to 01/01/01 and are not covered in this document)*

9920/3 Therapy-related acute myeloid leukemia, NOS***** 21
9930/3 Myeloid sarcoma 22
9931/3 Acute panmyelosis with myelofibrosis (acute megakaryocytic leukemia) 23

Disease Profiles, continued

Other Leukemias

9940/3 Hairy cell leukemia 24
9945/3 Chronic myelomonocytic leukemia* 25
9946/3 Juvenile myelomonocytic leukemia* 26
9948/3 Aggressive NK-cell leukemia***** 27

Chronic myeloproliferative disorders

9950/3 Polycythemia vera* 28
9960/3 Chronic myeloproliferative disease* 30
9961/3 Myelosclerosis with myeloid metaplasia* 31
9962/3 Essential thrombocythemia* 32
9963/3 Chronic neutrophilic leukemia* 33
9964/3 Hypereosinophilic syndrome* 34

Myelodysplastic syndromes

9980/3 Refractory anemia* 38
9982/3 Refractory anemia with sideroblasts* 39
9983/3 Refractory anemia with excess blasts* 40
9984/3 Refractory anemia with excess blasts in transformation***** 41
9985/3 Refractory cytopenia with multilineage dysplasia* 42
9986/3 Myelodysplastic syndrome with 5q- syndrome* 43
9987/3 Therapy-related myelodysplastic syndrome, NOS* 44
9989/3 Myelodysplastic syndrome, NOS* 46

Appendix 1: Coding Treatment and Phenotype for the Hematopoietic Diseases 48

Appendix 2: Casefinding and Sequencing Guidelines for the Newly Reportable Hematopoietic Diseases 49

Resources 51

Index to ICD-9-CM Casefinding Codes Used in This Document 52

Diagnosis Index 53

List of Tables

B-Lymphocyte and Plasma Cell Neoplasms 1
Classification of Mastocytosis 9
Histiocyte Society Classification (1987) 12
Types of Langerhans Cell Histiocytosis 12
The Refractory Anemias 35
ICD-9-CM Casefinding Codes for Refractory Anemias 36
International Prognostic Scoring System for Myelodysplastic Diseases 37

INTRODUCTION

On January 1, 2001, cancer registrars in the United States began coding primary site and morphology terms using the *International Classification of Diseases for Oncology, Third Edition* (ICD-O-3). Over 200 new terms and synonyms were added in the third edition in the narrow range of codes from M-9590 to M-9989. This *Abstracting and Coding Guide for the Hematopoietic Diseases* was prepared in response to many questions about whether a variety of blood diseases are reportable to cancer registries.

One of the issues with the case reportability changes implemented with ICD-O-3 is that registrars are largely unfamiliar with many of these blood and bone marrow diseases and all the different names for individual diseases. Although this book does not include all of the lymphatic and hematopoietic diseases, it covers the majority of diseases that meet any of the following criteria:

1. New terms or codes in ICD-O-3
2. Rare or unusual blood or bone marrow diseases
3. Confusing terminology or many different names for the same condition
4. Coded in ICD-9-CM in non-specific "other" codes along with other non-reportable conditions
5. Generally have no TNM staging scheme

The following conditions are NOT covered in this fascicle:

- Hodgkin and non-Hodgkin lymphomas (M-9590 to M-9729)
- Lymphoid and myeloid leukemias (M-9820 to M-9910)
  Lymphomas and leukemias have been reportable for many years and abstracting and coding guidelines for these conditions are well-established.
- Certain non-reportable terms (M-9765/1 to M-9769/1; M-9831/1; M-9970/1; M-9975/1)

Structure of this document

The table format is the same for each disease. The sections of the table are:

Header preferred name of the disease and the title of the ICD-O-3 heading or group.

Preferred term preferred name of the disease (boldface term in ICD-O-3)

Synonyms other names for the same disease, gathered from reference books and Internet resources

ICD-9-CM the casefinding code for the disease (where the medical records coder should have coded the diagnosis)

ICD-O-3 the correct primary site (C_ _._), morphology (M-_ _ _ _) and behavior (/_) codes for this diagnosis

Definition a brief, mostly non-technical description of the disease

Treatment a list of methods used to treat the disease, in general order of common usage or effectiveness. Each mode of treatment is provided with a *suggested* code from the SEER Code Manual (third edition) and ROADS manual. The code definitions have been abbreviated due to limitations on space in the tables. If there is a question about any of the codes or abbreviations, always refer to the coding manuals. Wherever possible, treatment descriptions are as specific as possible, especially when they apply to particular situations such as limited extent of disease. These treatments are the most common for the disease; other treatments may be administered to the patient and should be included on the abstract. Ancillary drugs, such as colony stimulating factors which are not coded for lymphomas and leukemias, may be mentioned in the tables and are not to be coded for the newly reportable hematopoietic diseases.

Surgery procedures intended to cure or palliate the patient; excluding biopsies. The codes listed are the ones most likely to be appropriate for the disease. See the site-specific surgery codes in the SEER Code Manual or the ROADS manual for other choices.

Radiation generally coded to 1 beam radiation except as noted, although other codes such as 5, radiation NOS or 4, combination radiation may be appropriate

Chemotherapy wherever possible, one or more specific chemotherapy agents effective for the disease have been listed. Where specific agents are not listed, there may have been none mentioned in reference material or there may have been too many to list. Consult SEER Book 8, Antineoplastic Drugs for further information.

Hormone therapy wherever possible, one or more specific hormonal agents effective for the disease have been listed. In addition, if any hormone therapy agent is part of a multi-agent chemotherapy regimen it is coded in the hormone field. In the tables, hormones included in multi-agent regimens are identified by the phrase (when given as part of a chemotherapy regimen), and the phrase applies to any agent specifically listed. Hormonal agents used to enhance appetite or for other supportive measures are not coded. Consult SEER Book 8, Antineoplastic Drugs for further information.

Immunotherapy wherever possible, one or more specific biological response modifiers (immunotherapy) are listed. These may include but are not limited to monoclonal antibodies, vaccines, or immunotherapy agents. Consult SEER Book 8, Antineoplastic Drugs for further information.

Other therapy this treatment field includes other methods of treating the patient. Traditionally, this field is reserved for experimental and alternative treatments, but the newly reportable hematopoietic diseases are more chronic, and therefore have different approaches to treatment. Appendix A contains an excerpt from "Clarifications for Abstracting and Coding Hematopoietic Diseases" (May 22, 2001), available from www.seer.cancer.gov/Admin, describing the use of the "other therapy" field for coding newly reportable hematopoietic diseases. For other diseases such as leukemias and lymphomas that have been reportable for many years, treatments such as transfusions, phlebotomy, and supportive care should *not* be coded in the "Other Therapy" field.

EOD the number string in this field represents the 10 digits of the SEER Extent of Disease (EOD) field, which are largely the same as the 2003 Collaborative Staging System codes

Summary Stage 2000 the code and category of the stage for this disease (see references)

Other staging includes the names of other staging or classification systems a registrar might see in reference to the disease. In some instances, the full staging or classification scheme is included in this fascicle.

Notes other information about the disease, including further discussion of coding issues; characteristics of the disease, such as survival information and age at diagnosis; and relationship to other diseases.

Other Reference Material

Interspersed in some of the sections is further information about groups of diseases, such as the plasma cell tumors, the Langerhans cell histiocytoses, and the refractory anemias. A resources list has been included at the end of the fascicle to identify coding and disease references used in preparation of these guidelines. This information has been provided to enhance the cancer registrar's understanding of these diseases and is not intended to be a required part of the cancer abstract.

"Sarcoma" as the term is used in the hematopoietic diseases

A sarcoma is defined as a malignant neoplasm arising in tissue of mesodermal origin (as connective tissue, bone, cartilage, or striated muscle). The common use of the term is for malignancies that arise in the soft tissues and occasionally in solid organs. These are generally coded to the soft tissue or organ in which they arise. When used by a pathologist in the context of hematopoietic diseases, such as plasma cell tumors, mast cell tumors, or histiocytoses, sarcoma has another meaning: a deposit in an organ or tissue of neoplastic cells normally only found in the blood or bone marrow. Like their mesodermal counterparts, these conditions should be coded in ICD-O-3 to the site in which they arise, rather than blood or bone marrow. However, when the registrar is casefinding in ICD-9-CM coded disease indices, these sarcomas may inadvertently be coded incorrectly to soft tissue, ill-defined or unknown primary site. As such they will be identified from the disease index, but must be coded to the correct primary site in ICD-O-3.

Death Certificate Only cases

These tables have been created to abstract and code the disease history of *living* patients. Autopsy-diagnosed cases may follow the same guidelines, but cases identified by death certificate only will be uniformly coded to unknown (9, 99, 999, etc.) for all staging fields. Follow instructions in the SEER Program Code Manual or the ROADS manual for other fields on patients identified by death certificate only.

Plasma Cell Tumors

Plasma cell tumors that are reportable in ICD-O-3 are part of a series of diverse disorders of B lymphocytes and plasma cells collectively called plasma cell neoplasms. Not all plasma cell neoplasms are malignant, but the ones that are are reportable. All of the diseases are characterized by the proliferation of a type of B-cell lymphocyte (the plasma cell) which generates immunoglobulins that can be measured by immunoelectrophoresis in blood serum or urine.

B-Lymphocyte and Plasma Cell Neoplasms

Category	ICD-O-3	Report-able?	Diagnoses; comments
Monoclonal gammopathy of undetermined significance (MGUS)	9765/1	No	Associated with solid tumors of prostate, GI tract, kidney, breast, biliary tree; chronic inflammatory and infectious conditions; other diseases (myasthenia gravis, familial hypercholesterolemia, thyrotoxicosis, pernicious anemia, etc.)
Transient plasma cell dyscrasias		No	Inflammatory or non-clonal plasma cell reactions associated with viral infections, heart surgery, drug hypersensitivity
Malignant plasma cell diseases			
Plasmacytoma of bone (solitary myeloma)	9731/3	Yes	Solitary (localized) lesion
Multiple myeloma	9732/3	Yes	IgG, IgA, IgD, IgE, light chains; bone marrow involvement
Plasma cell leukemia	9733/3	Yes	Plasmacytic leukemia; bone marrow involvement
Plasmacytoma, extramedullary	9734/3	Yes	Solitary lesion(s) of soft tissues
Macroglobulinemia	9761/3	Yes	IgM spike; Waldenstrom macroglobulinemia
Heavy chain diseases	9762/3	Yes	IgG, IgA, IgM,, IgD heavy chain disease
Immunoproliferative small intestinal disease	9764/3	Yes	
Lymphoblastic lymphoma	9727/3 to 9729/3	Yes	Discussion of lymphomas is not included in this document
Primary amyloidosis	9769/1	No	Also called immunoglobulin deposition disease, systemic light chain disease, immune amyloidosis, immunocyte-associated amyloidosis
POEMS syndrome		No	POEMS = polyneuropathy, organo-megaly, endocrinopathy, monoclonal gammopathy, and skin changes

Adapted from *The Merck Manual of Diagnosis and Therapy*, Chapter 140 Plasma Cell Dyscrasias, Table 140-1, and from other Internet documents.

Plasmacytoma Plasma Cell Tumors
9731/3

Preferred Term	Plasmacytoma, NOS	
Synonyms	Plasmacytoma of bone; osseous plasmacytoma Plasma cell tumor Isolated plasmacytoma of bone; localized plasmacytoma Solitary myeloma; mono-ostotic myeloma Solitary plasmacytoma	
ICD-9-CM	238.6 Neoplasm of uncertain behavior of plasma cells 203.8 Other immunoproliferative neoplasms	
ICD-O-3	C40._ or C41._ [bone] M-9731/3	
Definition	Solitary lytic lesion of plasma cells found on skeletal survey in an otherwise asymptomatic patient; bone marrow examination from an uninvolved site contains less than 5% plasma cells. Usually there are no serum abnormalities and no Bence Jones protein in urine. *(see notes)*	
Treatment	Radiation to lesion	Radiation 1
	Surgery (laminectomy or curettage)	Surgery of primary site (bone): 10 (local tumor destruction or excision), 20 (partial resection), 30 (radical resection with limb salvage)
	Chemotherapy (if symptomatic or if M-protein increases--usually reserved for multiple myeloma)	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
EOD	999 10 9 99 99	
Summary Stage 2000	1 Localized (if multifocal or disseminated, see multiple myeloma M9732/3)	
Other staging	Durie/Salmon (stages I-III)	
Notes	• Part of the spectrum of plasma cell neoplasms--see second note under plasma cell leukemia, M-9733/3. • Rare (about 5% of all plasma cell neoplasms). • About 25% of patients have a serum and/or urine M-protein; this should disappear following adequate irradiation of the lytic lesion. • If a magnetic resonance imaging (MRI) reveals unsuspected bony lesions which were undetected on standard radiographs, the diagnosis is multiple myeloma (M-9732/3). • Most common sites: spine and long bones of arms and legs • Cure rate at 10 years: 35%. • Recurrence rate: up to 25%. 55 to 70% develop multiple myeloma within two years. • In the Kiel classification (now obsolete), solitary plasmacytoma is considered a form of lymphoplasmacytic lymphoma.	

Multiple myeloma Plasma Cell Tumors
9732/3

Preferred Term	Multiple myeloma	
Synonyms	Myeloma, NOS *Subtypes:* IgG, IgA, IgD or IgE myeloma Myelomatosis Plasma cell myeloma; medullary plasmacytoma; plasma cell sarcoma Kahler's disease [and other eponymic names]	
ICD-9-CM	203.0 Multiple myeloma	
ICD-O-3	C42.1 M-9732/3	
Definition	Neoplastic production of excess plasma cells in the bone marrow which in turn produce immunoglobulins (antibodies) and light chains. Cytokines elaborated by the myeloma cells create lytic lesions that damage bones.	
Treatment	Chemotherapy (thalidomide, melphalan, doxil {doxorubicin}, vincristine, BiCNU, adriamycin, carmustine, cyclophos-phamide, etoposide; proteosome inhibitors)	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	Prednisone, decadron (when given as part of a chemotherapy regimen)	Hormone therapy 1
	Radiation to painful areas	Radiation 1
	Interferon alfa	Immunotherapy 1
	Bisphosphonates	Other therapy 1
	Supportive care (to prevent problems such as kidney damage and infection)	Do not code; record in remarks
	Blood or marrow stem cell transplantation	Immunotherapy 3 (allogenic bone marrow transplantation) or 5 (stem cell); also 2 (autologous) or 4 (NOS)
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	Durie/Salmon (stages I-III)	
Notes	• Multiple myeloma is part of a spectrum of diseases labeled plasma cell neoplasms--see second set of notes under plasma cell leukemia, M-9733/3. • Multiple myeloma comprises 94% of symptomatic plasma cell neoplasias. • Highly treatable but rarely curable. • Median survival: untreated, 7 months; with chemotherapy, 24-30 months for stage III; 35-40 months for stage II; 40-46 months for stage I. • The disease may be referred to as IgG, IgA, IgD or IgE myeloma. IgM myeloma is very rarely seen.	

Plasma cell leukemia Plasma Cell Tumors
9733/3

Preferred Term	Plasma cell leukemia	
Synonyms	PCL Plasmacytic leukemia Primary PCL *(for secondary PCL, see notes below)*	
ICD-9-CM	203.1 Plasma cell leukemia	
ICD-O-3	C42.1 M-9733/3	
Definition	Circulating peripheral blood plasma cells exceeding 20% of peripheral blood white cells. May be associated with lymphadenopathy and organomegaly, less frequently with bone pain and osteolytic lesions.	
Treatment	Chemotherapy (melphalan, doxil {doxorubicin}, vincristine, cyclophosphamide, etoposide)	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	Prednisone, decadron, dexamethasone (when given as part of a chemotherapy regimen)	Hormone therapy 1
	Patients under age 60: Peripheral stem cell or allogenic bone marrow transplantation	Immunotherapy 3 (allogenic bone marrow transplantation) or 5 (stem cell); also 2 (autologous) or 4 (NOS)
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	Durie/Salmon (stages I-III)	
Notes	• Rare; occurs in about 2% of plasma cell neoplasias. • Aggressive disease with short survival. Renal failure is common. • Median survival (treated): 2 to 7 months. • Occurs more frequently in conjunction with IgD and IgE myelomas and light-chain only myeloma. • Called primary PCL when diagnosed in the leukemic phase [usually a new primary]. • Called secondary PCL when there is leukemic transformation of a previously recognized multiple myeloma [record as transformation of previously reported multiple myeloma--no new abstract].	
	• The spectrum of plasma cell tumors (from best prognosis to worst) Solitary plasmacytoma of bone Extramedullary plasmacytoma Multiple myeloma Plasma cell leukemia	

Plasmacytoma, extramedullary Plasma Cell Tumors
9734/3

Preferred Term	Plasmacytoma, extramedullary (*not occurring in bone)*	
Synonyms	EMP Soft tissue plasmacytoma	
ICD-9-CM	238.6 Neoplasm of uncertain behavior of plasma cells 203.8 Other immunoproliferative neoplasms	
ICD-O-3	C_ _._ (code to site of origin) M-9734/3	
Definition	Isolated plasma cell tumor of soft tissues, most commonly occurring in the tonsils and/or nasopharynx (Waldeyer ring), paranasal sinuses or lung. Negative skeletal x-rays and bone marrow biopsy. About 25% of patients have serum and/or urine M-protein which should disappear following adequate irradiation.	
Treatment	Radiation therapy to isolated lesion (with fields that cover the regional lymph nodes)	Radiation 1
	Surgical resection (in selected cases where the lesion can be removed easily, such as the tonsil)	Surgery of primary site: *See site-specific surgery code for organ of origin.*
	Chemotherapy (if symptomatic or if M-protein increases--usually reserved for multiple myeloma)	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
EOD	Single/solitary/unifocal/isolated disease: 999 10 9 99 99 All others: 999 80 9 99 99	
Summary Stage 2000	Single/solitary/unifocal/isolated disease: 1 Localized All others: 7 Distant	
Other staging	Durie/Salmon (stages I-III)	
Notes	• Part of the spectrum of plasma cell neoplasms--see second note under plasma cell leukemia, M-9733/3. • Extramedullary plasmacytomas comprise about 3% of all plasma cell neoplasms. • Median survival (treated): 8.5 years. About 10-20% of solitary extramedullary plasmacytomas will progress to multiple myeloma. • May recur locally, but less likely to progress to multiple myeloma than solitary plasmacytomas of bone.	

Mastocytoma (*see also malignant mastocytoma M-9740/3*) Mast Cell Tumors
9740/1

Preferred Term	Mastocytoma	
Synonyms	Mast cell tumor, NOS Mast cell disease Mast cell proliferative disease Extracutaneous mastocytoma Solitary mastocytoma of skin Urticaria pigmentosa (non-neoplastic)	
ICD-9-CM	238.5 Neoplasm of uncertain behavior of histiocytic and mast cells ***Note:*** *If behavior code is /1, this diagnosis is not reportable*.	
ICD-O-3	C_ _._ (code to site of origin) M-9740/1	
Definition	Isolated or localized deposits of excess mast cells in the skin or other organs that produce large amounts of histamines.	
Treatment (record only if diagnosis is reportable-by-agreement)	Childhood mastocytoma usually clears up on its own.	Record as "no treatment" in remarks field
	Urticaria pigmentosa (present alone without systemic disease) clears or improves as adolescence approaches.	Record as "no treatment" in remarks field
	For adults, corticosteroids	Hormone therapy 1
	For adults, PUVA (psoralen and ultraviolet light)	Other therapy 1
EOD	999 99 9 99 99 (unstaged if behavior code is /1)	
Summary Stage 2000	9 Unstaged if behavior code is /1	
Other staging	Metcalfe (1991), categories I - IV (*see page 9)*	
Notes	• 75% of cases occur during infancy or early childhood. • Mast cells are a type of immunologic cell that remains in body tissues rather than in circulating blood like white blood cells (lymphocytes). • Mastocytoma is not reportable as a malignancy if coded to behavior /1. • Extracutaneous mastocytoma is very rare and has a non-destructive growth pattern, no skin lesions, and no evidence of systemic mastocytosis; most reported cases have been localized to lung. • Mastocytoma and urticaria pigmentosa rarely, if ever, develop into systemic mastocytosis, and both spontaneously improve over time. • Prognosis is excellent if onset of disease is before age 10; late onset disease tends to be persistent, is associated more often with systemic disease, and carries a higher risk of malignant transformation.	

Mast cell sarcoma Mast Cell Tumors
9740/3

Preferred Term	Mast cell sarcoma
Synonyms	MCS Malignant mast cell tumor Malignant mastocytoma
ICD-9-CM	202.6 Malignant mast cell tumors
ICD-O-3	C_ _._ (code to site of origin) M-9740/3
Definition	Localized but destructive tumor growth pattern of highly atypical, immature mast cells.
Treatment	Because mast cell sarcoma is so rare, very little information comparing the results of different treatment strategies is available.
EOD	Single/solitary/unifocal/isolated disease: 999 10 9 99 99 All others: 999 80 9 99 99
Summary Stage 2000	Single/solitary/unifocal/isolated disease: 1 Localized All others: 7 Distant
Other staging	Metcalfe (1991), categories I - IV *(see page 9)*
Notes	• Very rare. • Distant spread is possible, as is a leukemic phase. • Similar to extracutaneous mastocytoma (M-9740/1) except that growth pattern is destructive.

Malignant mastocytosis Mast Cell Tumors
9741/3

Preferred Term	Malignant mastocytosis	
Synonyms	Systemic tissue mast cell disease (SMCD) Systemic mastocytosis Systemic mastocytosis with associated clonal, hematological non-mast-cell lineage disease (SM-AHNMD) Indolent systemic mastocytosis (ISM) (M-9741/1) Aggressive systemic mastocytosis (ASM)	
ICD-9-CM	202.6 Malignant mast cell tumors	
ICD-O-3	C_ _._ (code to site of origin) M-9741/3	
Definition	Accumulation of increased numbers of mast cells in the tissues other than skin, such as the liver, spleen, bone marrow, stomach, and small intestine.	
Treatment	There is no effective treatment for aggressive systemic mastocytosis. Most treatment descriptions are based on single case reports. Systemic mastocytosis is only symptomatically treated.	
	Antihistamines, drugs to reduce stomach acid, migraine treatment, cromolyn (a mast cell stabilizer) for bowel symptoms, ketotifen (oral mast cell stabilizer (for symptom relief)	Do not code; record in remarks
	Methylprednisolone (low dose)	Hormone therapy 1
	Interferon alpha, cyclosporin	Immunotherapy 1
EOD s	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	Metcalfe (1991), categories I - IV *(see page 9)*	
Notes	• There is no known treatment that decreases the number of mast cells within tissue. • Some clinicians restrict the term systemic mastocytosis to cases with skin lesions, and apply the term malignant mastocytosis to those without. Other authors use the term benign systemic mastocytosis for those cases with limited lesions, mild symptoms, and prolonged course, and the term malignant systemic mastocytosis for those with tumor mass formation, leukemia, or bone marrow failure. • Systemic mastocytosis may be associated with cutaneous mastocytosis.	

Mast cell leukemia Mast Cell Tumors
9742/3

Preferred Term	Mast cell leukemia
Synonyms	MCL
ICD-9-CM	207.8 Other specified leukemia
ICD-O-3	C42.1 M-9742/3
Definition	Presence of systemic mastocytosis in addition to large numbers of mast cells in the blood and/or the bone marrow.
Treatment	Because mast cell leukemia is so rare, very little information comparing the results of different treatment strategies is available.
EOD	999 80 9 99 99
Summary Stage 2000	7 Distant
Other staging	Metcalfe (1991), categories I - IV *(see below)*
Notes	• Use this code on the rare occasion when mast cell leukemia is the first diagnosis made. If systemic mastocytosis (M-9741/3) is the first diagnosis, record mast cell leukemia as progression of disease--no new abstract. • Mast cell leukemia invariably arises in patients with a history of systemic mastocytosis. About 15% of patients with the malignant form of systemic mastocytosis will eventually progress to mast cell leukemia.

Classification of Mastocytosis (Adapted from Metcalfe DD. Classification and diagnosis of mastocytosis: current status. *J Invest Dermatol* 1991:96:2S-4S.)

I. Indolent mastocytosis (M-9741/1)
  A. Skin only
    1. Solitary mastocytoma (M-9740/1)
    2. Urticaria pigmentosa (non-neoplastic)
    3. Diffuse cutaneous mastocytosis (M-9741/3)*
    4. Telangiectasia macularis eruptiva perstans (non-neoplastic)
  B. Systemic (with or without urticaria pigmentosa) (M-9741/3)*
    1. Bone marrow
    2. Gastrointestinal system
    3. Other organs
II. Mastocytosis with associated hematologic disorder (with or without urticaria pigmentosa) (M-9741/3)*
  A. Dysmyelopoietic disorders*
  B. Myelopoietic disorders*
  C. Acute nonlymphatic leukemia*
  D. Malignant lymphoma*
  E. Chronic neutropenia
III. Mast cell leukemia (M-9742/3)*
IV. Lymphadenopathic mastocytosis with eosinophilia (with or without urticaria pigmentosa: aggressive mastocytosis) (M-9741/3)*

* indicates a reportable diagnosis

Malignant histiocytosis Neoplasms of Histiocytes (Macrophages) and Accessory Immune Cells
9750/3

Preferred Term	Malignant histiocytosis
Synonyms	MH Systemic histiocytosis Histiocytic medullary reticulosis [obs]
ICD-9-CM	202.3 Malignant histiocytosis
ICD-O-3	C42.1 M-9750/3
Definition	General term for several types of progressive systemic invasive proliferation of atypical histiocytes. Pancytopenia in terminal phase.
Treatment	Histiocytosis is so rare that there is little research into its cause and treatment.
EOD	999 80 9 99 99
Summary Stage 2000	7 Distant
Other staging	Histiocyte Society classification (classes 1-4) *(see page 12)*
Notes	• Extremely rare • Symptoms include fever, hepato- and splenomegaly, and constitutional symptoms. • Unlike most other cancers, MH sometimes goes into remission without treatment. • Three types of histiocytosis (not all are malignant): 1. Lipid histiocytosis, also called Niemann-Pick disease (an inherited disorder of fat metabolisms--not malignant). 2. Sinus histiocytosis with massive lymphadenopathy, also called Rosai-Dorfman disease (a rare benign histiocytic proliferative disorder of the lymph nodes in which the distended sinuses are completely, or nearly completely, filled by histiocytes). 3. Histiocytosis X (now preferably called Langerhans cell histiocytosis), a generic term embracing eosinophilic granuloma, Letterer-Siwe disease, and Hand-Schuller-Christian disease, and indicating a shared common origin for the three entities. They are all characterized by a proliferation of Langerhans cells and are best thought of as mono-ostotic, polyostotic or disseminated forms of the same disease. • Only disseminated Langerhans cell histiocytosis (Letterer-Siwe disease) (M-9754/3) is reportable.

The Langerhans cell histiocytoses Neoplasms of Histiocytes (Macrophages)
9751, 9752, 9753, 9754 and Accessory Immune Cells

Preferred Term	Langerhans cell histiocytosis	
Synonyms	**Note:** Langerhans cell histiocytosis (LCH) has many names and four different ICD-O-3 codes depending on the amount of involvement. *See the table on page 12 for codes, synonyms, and reportability.* **Note**: Only disseminated or generalized LCH is reportable in ICD-O-3.	
ICD-9-CM	202.3 Malignant histiocytosis 202.5 Letterer-Siwe disease	
ICD-O-3	C42.1 M-9754/3 (disseminated Langerhans cell histiocytosis) For other diagnoses, code to site of origin *(see page 12)*. M-975_/_	
Definition	Neoplastic proliferation of Langerhans cells in any of several organs or systems	
Treatment	Chemotherapy (cladribine, 2-CDA, velban, methotrexate, vinblastine, cytoxan, etoposide, vincristine, VP-16, aracytine {cytarabine}, 6-mercaptopurine); topical nitrogen mustard for skin involvement.	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	Prednisone (when given as part of a chemotherapy regimen); corticosteroids	Hormone therapy 1
	Cyclosporin, Interferon-alfa	Immunotherapy 1
	Skin involvement: PUVA (a type of ultraviolet light treatment)	Other therapy 1
	Isolated bone lesion: surgical curettage	Surg Primary site (bone) 10
	Radiation therapy (for bone lesions)	Radiation 1
	Supportive care for adverse effects of the disease (antibiotics, ventilatory support, physical therapy, selenium-based shampoo)	Do not code; record in remarks
EOD	Single/solitary/unifocal/isolated disease (9751, 9752--if reportable): 999 10 9 99 99 All others (9753--if reportable; 9754/3): 999 80 9 99 99	
Summary Stage 2000	Single/solitary/unifocal/isolated disease (9751, 9752--if reportable): 1 Localized All others (9753--if reportable; 9754/3): 7 Distant	
Other staging	Lahey score; Histiocyte Society classification (classes 1-4) *(see next page*)	
Notes	• Most often affects children under the age of two years. • Radiation is contraindicated in children because of the risk of secondary cancer. • More common in northern Europeans and rare in blacks. • LCH may in some instances regress on its own without treatment. • Associated with malignant lymphomas. • Survival rates: unifocal disease--95% overall; systemic--75%.	

Histiocyte Society Classification (1987)	
Class	**Diagnoses**
1	Langerhans cell histiocytosis *(see table below)*
2	Hemophagocytic lymphohistiocytosis Infection-associated hemophagocytic syndrome
3	Acute myelogenous leukemia FAB M5 (M-9891/3) Malignant histiocytosis (M-9750/3) True histiocytic lymphoma (M-9755/3)
4	Sinus histiocytosis with massive lymphadenopathy Xanthogranuloma Reticulohistiocytoma

Note: Terms in classes 2 and 4 are NOT reportable.

Types of Langerhans Cell Histiocytosis (Class 1 above)				
Name/Diagnosis	**ICD-O-3 code**	**Organ systems involved**	**Number of lesions**	**Report-able?**
Langerhans cell histiocytosis, NOS Langerhans cell granulomatosis Histiocytosis X, NOS Langerhans cell granulomatosis Type II histiocytosis	9751/1	Not stated	Not stated	No
Langerhans cell histiocytosis, unifocal Langerhans cell granulomatosis, unifocal Langerhans cell histiocytosis, mono-ostotic Eosinophilic granuloma (solitary)	9752/1	Single (usually bone)	Single	No
Langerhans cell histiocytosis, multifocal Langerhans cell histiocytosis, poly-ostotic Hand-Schuller-Christian disease [obs] Eosinophilic granuloma, multifocal	9753/1	Single (usually bone)	Multiple	No
Langerhans cell histiocytosis, disseminated Langerhans cell histiocytosis, generalized Letterer-Siwe disease Acute progressive histiocytosis X Nonlipid reticuloendotheliosis [obs] Progressive disseminated Langerhans cell histiocytosis Acute differentiated progressive histiocytosis Acute infantile reticuloendotheliosis Acute reticulosis of infancy	9754/3	Multiple (bones, skin, liver, spleen, lymph nodes)	Multiple	**Yes**

Note: Other terms used to describe syndromes considered to be Langerhans cell histiocytosis (LCH) include reticuloendotheliosis, Hashimoto-Pritzker syndrome, self-healing histiocytosis, and pure cutaneous histiocytosis (none of which are reportable).

Histiocytic sarcoma Neoplasms of Histiocytes (Macrophages) and Accessory Immune Cells
9755/3

Preferred Term	Histiocytic sarcoma	
Synonyms	True histiocytic lymphoma Monocytic sarcoma True histiocytic sarcoma	
ICD-9-CM	200.0 Lymphosarcoma and reticulosarcoma *(see notes below)*	
ICD-O-3	C_ _._ (code to site of origin) *(see below)* M-9755/3	
Definition	Tumor mass of macrophage/mature tissue histiocytic cells occurring in any of several sites. Most common sites: lymph nodes (one third) skin (one third), intestinal tract and other extranodal sites. Symptoms in addition to a mass include fever and weight loss, hepatosplenomegaly and symptoms specific to the involved site.	
Treatment	Therapy is modeled after the treatment of comparably staged diffuse large cell lymphomas, but the optimal approach remains to be defined.	
	Chemotherapy (cyclophosphamide, doxorubicin, vincristine, procarbazine, fludarabine, mitoxantrone)	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	Radiation therapy	Radiation 1
	Prednisone, dexamethasone (when given as part of a chemotherapy regimen)	Hormone therapy 1
	Anti-CD20 monoclonal antibody (rituximab)	Immunotherapy 1
	Autologous or allogeneic bone marrow or peripheral stem cell transplantation (under clinical evaluation)	Immunotherapy 3 (allogenic bone marrow transplantation); also 2 (autologous), 4 (NOS) or 5 (stem cell)
EOD	Single/solitary/unifocal/isolated disease: 999 10 9 99 99 All others: 999 80 9 99 99	
Summary Stage 2000	Single/solitary/unifocal/isolated disease: 1 Localized All others: 7 Distant	
Other staging	Histiocyte Society classification (classes 1-4) *(see previous page*) Sometimes staged according to Ann Arbor staging for lymphomas.	
Notes	• Rare neoplasm. • Aggressive, with poor response to therapy. • Affects all ages. • A previous name for histiocytic sarcoma, true histiocytic lymphoma, is now considered incorrect. Most previously-termed histiocytic lymphomas are now thought to be lymphocytic lymphomas. True malignancies of histiocytes are rare. • Histiocytic sarcoma is more closely related to malignant histiocytosis (M-9750/3) than to Langerhans cell histiocytoses (M-9751 to M-9754, M-9756).	

Langerhans cell sarcoma Neoplasms of Histiocytes (Macrophages) and Accessory Immune Cells
9756/3

Preferred Term	Langerhans cell sarcoma	
Synonyms	None; part of Langerhans cell histiocytosis category of diagnoses	
ICD-9-CM	202.9 Other and unspecified malignant neoplasms of lymphoid and histiocytic tissue (may also be incorrectly coded to 157.4 Malignant neoplasm of islets of Langerhans)	
ICD-O-3	C_ _._ (code to site of origin) *(see below)* M-9756/3	
Definition	Tumor mass of Langerhans cell histiocytes occurring in multiple organs including lymph nodes, spleen, liver, lung, and bone.	
Treatment	Chemotherapy (vinblastine, VP-16, 6-mercaptopurine)	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	Steroids (when given as part of a chemotherapy regimen)	Hormone therapy 1
EOD	Single/solitary/unifocal/isolated disease: 999 10 9 99 99 All others: 999 80 9 99 99	
Summary Stage 2000	Single/solitary/unifocal/isolated disease: 1 Localized All others: 7 Distant	
Other staging	Histiocytosis grouping system (classes 1-4) *(see page 12)*	
Notes	• Considered a higher grade variant of Langerhans cell histiocytosis (M-975_) • Exceedingly rare. • Occurs in all age ranges. • Very aggressive; 4-year survival rate of 60–80%.	

Interdigitating dendritic cell sarcoma Neoplasms of Histiocytes (Macrophages)
9757/3 and Accessory Immune Cells

Preferred Term	Interdigitating dendritic cell sarcoma	
Synonyms	IDCS Dendritic cell sarcoma, NOS [non-specific for follicular or interdigitating] Interdigitating reticulum cell sarcoma Reticulum cell sarcoma [non-specific for follicular or interdigitating] ***(Note:** not the same as the obsolete term for malignant lymphoma)* Interdigitating cell sarcoma Interdigitating dendritic cell tumor (M-9757/1); IDC tumor Paracortical interdigitating cell sarcoma	
ICD-9-CM	202.9 Other and unspecified malignant neoplasms of lymphoid and histiocytic tissue (may also be incorrectly coded to sarcoma of a specified site)	
ICD-O-3	C_ _._ (code to site of origin) *(see notes below)* M-9757/3	
Definition	Rare sarcoma arising from the interdigitating reticulum cells in the T-cell area of the lymph node and forming a node-based tumor.	
Treatment	The following treatments are based on limited case reports.	
	Surgical excision of the mass lesion	Surgery of primary site *(see site-specific scheme)*
	Multiagent chemotherapy (bleomycin; cyclophosphamide; carmustine; cisplatin; cytosine arabinoside; cytarabine hydrochloride; doxorubicin; daunorubicin; etoposide; ifosfamide; methotrexate; mechlorethamine hydrochloride; mercaptopurine; mitoxantrone; melphalan; procarbazine; vincristine; vindesine).	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	Radiation therapy	Radiation 1
	Asparaginase	Immunotherapy 1
	Cortisone; dexamethasone; prednisone; predonine (when given as part of a chemotherapy regimen)	Hormone therapy 1
EOD	Single/solitary/unifocal/isolated: 999 10 9 99 99 Multifocal or disseminated: 999 80 9 99 99	
Summary Stage 2000	Single/solitary/unifocal/isolated: 1 Localized Multifocal or disseminated: 7 Distant	
Other staging	None	
Notes	• Extremely rare disease of adults; only about 50 cases in world literature. • Aggressive neoplasm, unresponsive to conventional therapy. • *See also follicular dendritic cell sarcoma (M-9758/3)* • Interdigitating (mature) DCs are found in the secondary lymphoid organs (lymph nodes, spleen, tonsils) and have captured antigens in the periphery and migrated to the secondary lymphoid organs to present antigens to T cells.	

Follicular dendritic cell sarcoma Neoplasms of Histiocytes (Macrophages)
9758/3 and Accessory Immune Cells

Preferred Term	Follicular dendritic cell sarcoma	
Synonyms	FDC sarcoma Follicular dendritic cell tumor Reticulum cell sarcoma [non-specific for follicular or interdigitating subtype] *(**Note:** not the same as the obsolete term for malignant lymphoma)* Dendritic reticulum cell sarcoma [non-specific for follicular or interdigitating subtype] Sarcoma of follicular dendritic cells	
ICD-9-CM	202.9 Other and unspecified malignant neoplasms of lymphoid and histiocytic tissue (may also be incorrectly coded to sarcoma of a specified site)	
ICD-O-3	C_ _._ (code to site of origin) *(see notes below)* M-9758/3	
Definition	Rare sarcomas arising from the follicular reticulum cells in the B-cell area of the lymph node and forming a node-based tumor; can also arise in extranodal sites such as spleen, tonsil, oral cavity, gastrointestinal tract, liver, soft tissue, and skin.	
Treatment	The following treatments are based on limited case reports.	
	Complete excision of lesion	Surgery of primary site (see site-specific scheme)
	For intra-abdominal tumors, chemotherapy	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	For intra-abdominal tumors, radiation therapy	Radiation 1
EOD	Single/solitary/unifocal/isolated: 999 10 9 99 99 Multifocal or disseminated: 999 80 9 99 99	
Summary Stage 2000	Single/solitary/unifocal/isolated: 1 Localized Multifocal or disseminated: 7 Distant	
Other staging	None	
Notes	• Follicular dendritic cell tumor (FDT) is a rare neoplasm usually occurring in the laterocervical lymph nodes, but presentations elsewhere are also well documented. • First recognized as a distinct subtype of dendritic cell sarcoma in 1986, about 50 cases reported in world literature. • Dendritic cells are covered with long membrane extensions (that make them look like dendrites in nervous tissues ... hence the name). Dendritic cells are sometimes called reticulum cells or veiled cells. • Follicular DCs are found in the secondary lymphoid organs (lymph nodes, spleen, tonsils) and have captured antigens in the periphery and migrated to the secondary lymphoid organs to present antigens to B cells. • *See also interdigitating dendritic cell sarcoma (M-9757/3)*	

Immunoproliferative disease Immunoproliferative Diseases
9760/3

Preferred Term	Immunoproliferative disease, NOS
Synonyms	Immunoproliferative disorder Immunoproliferative neoplasm Immunoproliferative neoplasia
ICD-9-CM	203.8 Other immunoproliferative neoplasms
ICD-O-3	C42.1 (usually) M-9760/3
Definition	Category of disorders characterized by abnormal proliferation of primary cells of the immune system (lymphocytes) or by excessive production of immunoglobulins.
Treatment	There is no information available on treatment for this diagnosis. Wherever possible, code to a specific diagnosis, such as multiple myeloma, and refer to that page of guidelines in this text.
EOD	999 80 9 99 99
Summary Stage 2000	7 Distant
Other staging	None
Notes	The immunoproliferative diseases are a very broad category of neoplasms of the lymphoid cells that include T- and B-cell lymphomas, Hodgkin lymphoma, mycosis fungoides and Sezary syndrome, plasmacytic tumors, acute and chronic lymphocytic leukemia, Waldenstrom macroglobulinemia, heavy chain disease, immunoproliferative small intestinal disease and a number of other unspecified lymphocytic or plasmacytic neoplasms. Wherever possible, code to a specific diagnosis rather than to "immunoproliferative diseases, NOS."

Waldenstrom macroglobulinemia Immunoproliferative Diseases
9761/3

Preferred Term	Waldenstrom macroglobulinemia	
Synonyms	WM Macroglobulinemia; primary macroglobulinemia; idiopathic macroglobulinemia; macroglobulinemia of Waldenstrom	
ICD-9-CM	273.3 Macroglobulinemia	
ICD-O-3	C42.0 M-9761/3 (**Note:** Waldenstrom macroglobulinemia is always coded to blood) *(See also note 4 below.)*	
Definition	Myeloproliferative disorder of B-lymphocytes producing abnormal plasma cells that secrete excess IgM causing hyperviscosity (thickening) of blood.	
Treatment *WM is not a newly reportable disease	Systemic chemotherapy (leukeran {chlorambucil}, alkeran, cytoxan {cyclophosphamide}, melphalan, leustatin {cladribine}, fludarabine)	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	Interferon alpha; gene therapy	Immunotherapy 1
	Stem cell transplantation	Immunotherapy 5
	Corticosteroids, prednisone	Hormone therapy 1
	Plasmapheresis or plasma exchange	Do not code; record in remarks*
	Packed red cell transfusions, antibiotics, or platelet transfusions (supportive care)	Do not code; record in remarks*
	Radiation to reduce size of spleen	Radiation 1
	Splenectomy	Surgery of other sites 1
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	None	
Notes	• Disease may present with involvement of bone marrow, but should be coded to C42.0, blood. • Relatively indolent; median survival: 5 years. • Median age at diagnosis: 65. • Symptoms of hyperviscosity include weakness, fatigue, drowsiness, paleness, easy bleeding, fever, weight loss, dizziness and headaches. • Cross-referenced in ICD-O-3 to M-9671/3, lymphoplasmacytic lymphoma, when diagnosed in sites other than blood and bone marrow. • Synonyms for M-9671/3 include well-differentiated plasmacytoid lymphocytic lymphoma, lymphoplasmacytic immunocytoma, plasmacytic-lymphocytic lymphoma, and plasmacytoid small lymphocytic lymphoma.	

Heavy chain disease Immunoproliferative Diseases
9762/3

Preferred Term	Heavy chain disease	
Synonyms	Heavy chain disease (unspecified/NOS) HCD ***Subtypes*** **Alpha:** α [Greek symbol alpha]-heavy chain disease**;** IgA heavy chain disease**;** alpha chain disease **Mu** [new in ICD-O-3]: IgM heavy chain disease; mu-chain disease; μ [Greek symbol mu]-heavy chain disease**;** **Gamma:** gamma-HCD; Franklin disease; IgG heavy chain disease; γ [Greek symbol gamma]-heavy chain disease; gamma chain disease **Delta:** IgD heavy chain disease; δ [Greek symbol delta]-heavy chain disease [not listed in ICD-O-3 but coded here]	
ICD-9-CM	273.2 Other paraproteinemias	
ICD-O-3	C42.1 M-9762/3	
Definition	Lymphoplasmacytic proliferative disorders characterized by the uncontrolled production of abnormal immunoglobulin heavy chains. Specific subtypes are identified by serum protein electrophoresis.	
Treatment	Asymptomatic patients: no treatment	Do not code; record in remarks
	Symptomatic patients: chemotherapy agents effective in treating multiple myeloma (cyclophosphamide (cytoxan), vincristine, chlorambucil, doxorubicin).	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	Broad spectrum antibiotics	Do not code; record in remarks
	Corticosteroids, prednisone	Hormone therapy 1
	Radiotherapy	Radiation 1
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	None	
Notes	• **Alpha (IgA):** most common HCD; usually appears between ages 10 and 30 years; mainly involves gastrointestinal tract; geographically concentrated in the Middle East; closely related to Mediterranean lymphoma or immunoproliferative small intestinal disease (see M-9764/3); rare hepatosplenomegaly and lymphadenopathy. • **Gamma (IgG)**: only about 100 cases in world literature; median survival: 1 year; median age 61; hepatosplenomegaly and lymphadenopathy common. • **Mu (IgM):** rare; median age 48; associated with CLL. • **Delta (IgD):** only one case reported in world literature.	

Immunoproliferative small intestinal disease Immunoproliferative Diseases
9764/3

Preferred Term	Immunoproliferative small intestinal disease	
Synonyms	IPSID Mediterranean lymphoma Mediterranean abdominal lymphoma	
ICD-9-CM	200.8 Lymphoma, other named variants 203.8 Other immunoproliferative neoplasms	
ICD-O-3	C17._ M-9764/3 C17.0 Duodenum, C17.1 Jejunum, C17.2 Ileum	
Definition	IgA lymphoproliferative disorder of the small bowel resulting in diarrhea, malabsorption, and eventually a B cell lymphoma.	
Treatment	Cytotoxic and antibiotic agents (*see notes below*)	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	Hormones (prednisone if used in combination with chemotherapy)	Hormone therapy 1
EOD	Single/solitary/unifocal/isolated disease: 999 10 9 99 99 Multifocal, involving adjacent tissues, or disseminated: 999 80 9 99 99	
Summary Stage 2000	Single/solitary/unifocal/isolated disease: 1 Localized Multifocal, involving adjacent tissues, or disseminated: 7 Distant	
Other staging	Galen system, Khojasteh system and Salem system	
Notes	• Three stages/phases of IPSID: 1. Early Stage: tetracycline for 6 months; usually a good outcome. 2. Prelymphomatous Stage: cyclophosphamide +/- prednisone and tetracycline; variable but usually a poor outcome. 3. Lymphomas: cyclophosphamide, doxorubicin, teniposide, vinblastine, prednisone +/- bleomycin • Endemic in the Mediterranean basin, Mideast, Far East, Africa; sporadic in Europe, North & South America, immigrants from developing countries, native Indians. • Associated with alpha heavy chain disease (M-9762/3) and MALT lymphoma (M-9699/3) • Most common in duodenum and proximal jejunum, diffusely involving the part of the small intestine that is affected.	

Therapy-related acute myeloid leukemia Myeloid Leukemias
9920/3

Preferred Term	Therapy-related acute myeloid leukemia (unspecified/NOS)	
Synonyms	Therapy-related acute myeloid leukemia, alkylating agent related Therapy-related acute myeloid leukemia, epipodophyllotoxin-related Topoisomerase II inhibitor-related acute myeloid leukemia Secondary acute myeloid leukemia due to prior chemotherapy	
ICD-9-CM	205.8 Other myeloid leukemia	
ICD-O-3	C42.1 M-9920/3	
Definition	Development of acute myelogenous leukemia as a result of treatment for a previous malignancy with ionizing radiation or certain chemotherapy agents.	
Treatment	Generally refractory to treatments for acute myeloid leukemia.	
	Erythropoietin, colony stimulating factor	Ancillary drugs–do not code; record in remarks
	Supportive care	Do not code; record in remarks
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	None	
Notes	• Therapy-related leukemia is estimated to be 10-20% of all acute myelogenous leukemias diagnosed. Incidence is increasing. • Median survival: 9.7 months in one large Japanese study, 7.5 months in a US study. • Most commonly occurs after combination chemotherapy and radiation therapy for Hodgkin lymphoma, breast cancer, and chronic lymphocytic leukemia. • The majority of secondary leukemias resulting from the use of cytotoxic drugs can be divided into two well-defined groups depending on whether the patient has received: 1) alkylating agents or 2) drugs binding to the enzyme DNA-topoisomerase II (epipodophyllotoxins and anthracyclins). • Alkylating agent related: due to cytoxan, myleran, or leukeran chemotherapy; also MOPP regimen. TRL develops 5 to 7 years after administration of alkylating agent chemotherapy. • Epipodophyllotoxin related: due to VP-16, Etoposide, Vepesid, teniposide (Vumon) or epipodophyllotoxin chemotherapy. TRL develops 6 months to 5 years after epipodophyllotoxin therapy. • Anthracyclin related: due to any of the -rubicin drugs, such as doxorubicin • **Note**: therapy-related leukemia will almost always be a subsequent primary. • Very similar to post-MDS leukemias; characterized frequently by a preleukemic phase, trilineage dysplasia, frequent cytogenetic abnormalities involving chromosomes 5 and 7 and a poor prognosis.	

Myeloid sarcoma Myeloid Leukemias
9930/3

Preferred Term	Myeloid sarcoma	
Synonyms	Chloroma Granulocytic sarcoma Extramedullary myeloid tumor	
ICD-9-CM	205.3 Myeloid sarcoma	
ICD-O-3	C_ _._ (Code to site of mass. *See definition below).* M-9930/3 **Note:** ICD-O-3 rule E applies.	
Definition	Tumor mass of myeloid cells occurring in an extramedullary site or bone. Most common sites: skull, paranasal sinuses, sternum, ribs, vertebrae and pelvis; less commonly lymph nodes and skin.	
Treatment	Radiation therapy to site of mass	Radiation 1
	Systemic chemotherapy	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
EOD	Single/solitary/unifocal/isolated disease: 999 10 9 99 99 All others: 999 80 9 99 99	
Summary Stage 2000	Single/solitary/unifocal/isolated disease: 1 Localized All others: 7 Distant	
Other staging	WHO classification	
Notes	• Cross-referenced to M-9861/3, acute myeloid leukemia. • Myeloid sarcoma may be an isolated occurrence or in association with acute myeloid leukemia, chronic myeloid leukemia, chronic idiopathic myelofibrosis, hypereosinophilic syndrome, or polycythemia vera. It may occur simultaneously with an acute myeloid leukemia, be the first evidence of a relapse after acute myeloid leukemia treatment, or precede the conventional diagnosis of a myeloid leukemia by months or years. It is generally diagnosed in two patient groups, those under 35 and those over 65 years old. • If diagnosed simultaneously with another hematopoietic disease, do not consider the myeloid sarcoma as a separate primary. • Although myeloid sarcomas are radiosensitive, systemic chemotherapy is warranted in most cases. However, specific details for therapy and prognosis are dependent on the underlying myeloid leukemia subtype.	

Acute panmyelosis with myelofibrosis Myeloid Leukemias
9931/3

Preferred Term	Acute panmyelosis with myelofibrosis	
Synonyms	Acute megakaryocytic leukemia Acute myelofibrosis Malignant myelofibrosis Acute myelosclerosis, NOS Acute myelodysplasia with myelofibrosis Malignant myelosclerosis [obs] Acute panmyelosis, NOS	
ICD-9-CM	238.7 Neoplasm of uncertain behavior of other lymphatic and hematopoietic tissues	
ICD-O-3	C42.1 M-9931/3	
Definition	Very rare form of acute myelogenous leukemia. Proliferative process involving granulocytes, erythrocytes and megakaryocytes with associated fibrosis of the marrow.	
Treatment	Chemotherapy (poor response)	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	None	
Notes	• Acute, severe syndrome with short survival. • Also called megakaryoblastic leukemia (M7 ANLL) with prominent fibrosis. • Acute panmyelosis with myelofibrosis is ***not*** the same as myelofibrosis (idiopathic, primary or NOS) or agnogenic myeloid metaplasia (M-9961/3).	

Hairy cell leukemia Other Leukemias
9940/3

Preferred Term	Hairy cell leukemia	
Synonyms	Leukemic reticuloendotheliosis Hairy cell leukemic variant	
ICD-9-CM	202.4 Leukemic reticuloendotheliosis	
ICD-O-3	C42.1 M-9940/3	
Definition	Easily controlled chronic disorder of lymphocytes. The disease is called hairy cell leukemia because the cancer cells look "hairy" when examined under a microscope.	
Treatment	If asymptomatic, no treatment	Do not code; record in remarks
	Cladribine (leustatin) (first line therapy 2-chlorodeoxyadenosine, 2-CdA) Chlorambucil, vincristine, cytoxan and fludarabine are rarely used.	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	Interferon alfa, pentostatin, rituxan (rituximab--monoclonal antibody)	Immunotherapy 1
	Prednisone (when given as part of a chemotherapy regimen)	Hormone therapy 1
	Splenectomy (for severe thrombocytopenia)	Surgery of other sites 1
	BL22 (recombinant immunotoxin) (under clinical evaluation)	Immunotherapy 1
	Allogeneic bone marrow transplantation (rare instances--refractory cases)	Immunotherapy 3 (allogenic bone marrow transplantation); also 2 (autologous) or 4 (NOS) or 5 (stem cell)
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	None	
Notes	• HCL is a chronic (slow progressing) lymphocytic leukemia, first reported in 1958, that does not develop into acute (rapidly progressing) leukemia. • The decision to treat is based on cytopenias (especially if symptomatic), increasing splenomegaly, indications that the disease is progressing, or the presence of other, usually infectious complications.	

Chronic myelomonocytic leukemia Other Leukemias
9945/3

Preferred Term	Chronic myelomonocytic leukemia, NOS	
Synonyms	CMML CMML type I; CMML-1 [blasts <5% in blood; < 10% in marrow] CMML type II; CMML-2 [blasts 5-19% in blood; 10-19% in marrow] Chronic myelomonocytic leukemia in transformation [obs] Subacute myelomonocytic leukemia Chronic myelomonocytic syndrome	
ICD-9-CM	205.1 Chronic myeloid leukemia	
ICD-O-3	C42.1 M-9945/3	
Definition	Bone marrow production of abnormal cells that results in excess of monocytes as well as erythrocytes, megakaryocytes, and myelocytes. 5 to 20% blasts in bone marrow. Some proliferative features in marrow.	
Treatment	Supportive care	Do not code; record in remarks
	Allogenic peripheral stem cell or bone marrow transplantation	Immunotherapy 5 (stem cell) or 3 (allogenic bone marrow transplantation); also 2 (autologous) or 4 (NOS)
	Interferon alfa	Immunotherapy 1
	Red cell transfusions	Other Therapy 1; record in remarks
	Deferoxamine (iron chelating agent)	Other Therapy 1; record in remarks
	Erythropoietin, colony stimulating factor	Ancillary drugs–do not code; record in remarks
	Hormones (ineffective)	Hormone therapy 1
	Cytotoxic agents: high dose cytarabine; busulfan; hydroxyurea; others	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	French-American-British (FAB) classification; Bournemouth score; Sanz score, Lille score; International Prognostic Scoring System for MDS (*see page 37*)	
Notes	• Occurs predominantly in older patients. • Symptoms include mild anemia and thrombocytopenia, hepatosplenomegaly • Transformation to acute leukemia in 30% of patients. • Median survival: 14 to 18 months. • Part of former FAB classification of myelodysplastic syndromes. • In World Health Organization classification of hematopoietic diseases, CMML was moved to the Myelodysplastic Syndromes/Myeloproliferative Syndromes (MDS/MPS) category.	

Juvenile myelomonocytic leukemia Other Leukemias
9946/3

Preferred Term	Juvenile myelomonocytic leukemia	
Synonyms	JMML Juvenile chronic myelomonocytic leukemia (JCML) Juvenile chronic myeloid leukemia Juvenile myelomonocytic leukemia syndrome	
ICD-9-CM	205.9 (not specified as acute or chronic) unspecified myeloid leukemia	
ICD-O-3	C42.1 M-9946/3	
Definition	Clonal (stem cell) hematopoietic disorder of childhood characterized by proliferation of granulocytes and monocytes.	
Treatment	Bone marrow or stem cell transplantation	Immunotherapy 5 (stem cell) or 3 (allogenic bone marrow transplantation) (under clinical evaluation); also 2 (autologous) or 4 (NOS)
	Cytosine arabinoside and mitoxantrone (in clinical trials)	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	None	
Notes	• Disease of children up to age 14; 75% occur in children under 3. • Has features of both myeloproliferative and myelodysplastic disorders. • Generally poor prognosis even with intensive chemotherapy or bone marrow transplantation. • New diagnosis in WHO classification	

Aggressive NK-cell leukemia Other Leukemias
9948/3

Preferred Term	Aggressive NK-cell leukemia	
Synonyms	Large granular lymphocyte leukemia, NK-cell type, aggressive [REAL classification] Aggressive NK-cell leukemia-lymphoma	
ICD-9-CM	Not indexed in ICD-9-CM; may be coded to 204.8, Other lymphoid leukemia or 207.8 Other specified leukemia	
ICD-O-3	C42.1 M-9948/3	
Definition	Systemic proliferation of NK (natural killer) cells with aggressive clinical course.	
Treatment	Cladribine (leustatin) (first line therapy--2-chlorodeoxyadenosine, 2-CdA)	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	See notes below.	
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	None	
Notes	• Very rare; related to T-cell large granular lymphocytic leukemia (M-9831/1) but highly aggressive. • Most patients die within two months. • Strong association with Epstein-Barr virus (EBV). • More prevalent among Asians than whites. • Age range: teenagers and young adults. • No 'typical' treatment. Therapy often includes blood transfusions to treat anemia and antibiotics to treat infections. Chemotherapy is sometimes used. • New diagnosis in WHO classification • For analysis this disease should be included with acute lymphcytic leukemia.	

Polycythemia vera Myeloproliferative Disorders
9950/3

Preferred Term	Polycythemia vera	
Synonyms	Polycythemia rubra vera; P. vera Chronic erythremia [obs] Proliferative polycythemia Spent-phase polycythemia Primary polycythemia Primary erythremia Splenomegalic polycythemia Vaquez-Osler disease; Osler-Vaquez disease; Osler's disease	
ICD-9-CM	238.4 Polycythemia vera 207.1 Chronic erythremia (Hilmeyer-Schoner disease)	
ICD-O-3	C42.1 M-9950/3	
Definition	Overproduction of all blood cell lines, especially erythrocytes/red blood cells and platelets causing viscosity (thickening) of circulating blood.	
Treatment	Phlebotomy	Other therapy 1 (symptom relief); record in remarks
	Radioisotope Phosphorous-32	Radiation 3
	Alkylating agent therapy	Chemotherapy 2
	Hydroxyurea	Chemotherapy 2
	Interferon alfa	Immunotherapy 1
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	None	
Notes	• Chronic disease with a propensity to convert to AML. P. vera can develop a "spent phase" late in its course that resembles agnogenic myeloid metaplasia with cytopenia and marrow hypoplasia and fibrosis. • Secondary polycythemia is not a reportable disease. • Do not accession polycythemia, NOS. There are two categories of polycythemia, primary and secondary. Only primary polycythemia is reportable. Secondary polycythemia has a number of causes, such as smoking and extensive periods at high altitudes. The nonreportable secondary polycythemia is too common to make a blanket statement that any polycythemia (NOS) is reportable. • ***See also discussion on next page.***	

Polycythemia notes, continued	• Polycythemia is a condition characterized by an increase in the production of red blood cells. Polycythemia can be broken down into two categories (primary polycythemia and secondary polycythemia). ***Primary Polycythemia (also called Polycythemia Vera)*** The cause for the increased red blood cell production here is an overproduction of red blood cells by the bone marrow. Too many cells are packed into normal plasma volume. This is a chronic, progressive disease most common in middle-aged men. There is an increased number of red blood cells, an overgrowth of these blood cells in the bone marrow and commonly an enlarged spleen. Symptoms include headache, difficulty concentrating, pain in the fingers and toes. There is a danger of the formation of blood clots or hemorrhage. The treatment for polycythemia vera is periodic removal of some blood (phlebotomy) and radiation and/or chemotherapy with anti-metabolite drugs such as hydroxyurea. ***Secondary Polycythemia (also known as erythrocytosis)*** Here the cause for the increased red blood cells is the body's response to a perceived need for increased red blood cells. Prolonged lack of oxygen at high altitudes, chronic lung problems such as emphysema or cardiac insufficiency or other heart disease are conditions which commonly cause secondary polycythemia. Here, the treatment is directed towards the underlying medical cause for the increased red blood cell production. Another type of polycythemia is identified as the presence of a high concentration of red blood cells due to decreased plasma volume. In ***relative polycythemia***, some patients appear to have an excess of RBCs due to a loss of volume in the liquid portion of the blood, the plasma. This may be due to dehydration, diuretics (substances causing an increased loss of water through the urine, such as caffeine), burns, stress, or high blood pressure.

Chronic myeloproliferative disease Myeloproliferative Disorders
9960/3

Preferred Term	Chronic myeloproliferative disease	
Synonyms	Chronic myeloproliferative disorder (CMD)	
ICD-9-CM	238.7 Neoplasms of uncertain behavior of other lymphatic and hematopoietic tissues	
ICD-O-3	C42.1 M-9960/3	
Definition	Overproduction of one or more types of blood cells.	
Treatment	Close observation	Code as "no treatment;" record in remarks
	Chemotherapy	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	Irradation to painful areas	Radiation 1
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	None	
Notes	• Least specific of the myeloproliferative disorder diseases (which include polycythemia vera, chronic myelofibrosis with myeloid metaplasia, and essential thrombocythemia). Thought to result from an abnormality of a pluripotent hematopoietic stem cell.	
	• Do not accession myeloproliferative disease/myeloproliferative disorder, NOS. Myeloproliferative disorder/disease is coded to M-9975/1 and is more of a general category of bone marrow overproduction problems than a specific diagnosis. Both diagnoses are coded as 238.7 but only some of the cases would be considered reportable. Of these, the diagnoses ***chronic*** myeloproliferative disease/disorder and perhaps the myeloproliferative disorder/disease cases which, based on the complete medical record, would be coded as M-9975/3. Each case should be evaluated for reportability. **Note:** the clinician uses the term "chronic" to indicate a sustained level of a specific element such as blast cells in the blood, rather than long-term duration of the disease.	

Myelosclerosis with myeloid metaplasia Myeloproliferative Disorders
9961/3

Preferred Term	Myelosclerosis with myeloid metaplasia	
Synonyms	**Note:** the terms myelosclerosis and myelofibrosis are interchangeable. Myeloid metaplasia, NOS Myelofibrosis with myeloid metaplasia (MMM); myeloid metaplasia with myelofibrosis Primary myelofibrosis (PMF) Myelofibrosis as a result of myeloproliferative disease Megakaryocytic myelosclerosis Megakaryocytic myeloid metaplasia; idiopathic myeloid metaplasia Agnogenic myeloid metaplasia (AMM) Chronic idiopathic myelofibrosis; idiopathic myelofibrosis Myelofibrosis-osteosclerosis Chronic granulocytic-megakaryocytic myelosclerosis	
ICD-9-CM	238.7 Neoplasms of uncertain behavior of other lymphatic and hematopoietic tissues (myeloid metaplasia terms) 289.8 Other specified diseases of blood and blood-forming organs (myelofibrosis and myelosclerosis terms)	
ICD-O-3	C42.1 M-9961/3	
Definition	Overproduction of multiple cell lines resulting in bone marrow failure.	
Treatment	Interferon alfa	Immunotherapy 1
	Splenectomy	All other sites: Surgery of other sites 1
	Splenic irradiation	Radiation 1
	Hydroxyurea	Chemotherapy 2
	Allogenic peripheral stem cell or bone marrow transplantation	Immunotherapy 5 (stem cell) or 3 (allogenic bone marrow transplantation); also 2 (autologous) or 4 (NOS)
	Thalidomide (anti-angiogenesis agent)	Other therapy 1
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	None	
Notes	• Common symptoms: splenomegaly (with pain, early satiety, anemia, fatigue and other symptoms), immature granulocytes or erythrocytes in peripheral blood, fibrosis of marrow. Spleen and liver are often involved. • Most patients are over age 60 at diagnosis • Median survival: 3.5 to 5.5 years. • Myelofibrosis and agnogenic myeloid metaplasia are coded to 289.8 rather than 238.7. Why ICD-9-CM splits these diagnoses into two codes when they are all synonyms for the same disease process is unknown.	

Essential thrombocythemia Myeloproliferative Disorders
9962/3

Preferred Term	Essential thrombocythemia	
Synonyms	Idiopathic thrombocythemia Essential hemorrhagic thrombocythemia Idiopathic hemorrhagic thrombocythemia Primary thrombocythemia Thrombocythemia vera Essential thrombocytosis	
ICD-9-CM	238.7 Neoplasms of uncertain behavior of other lymphatic and hematopoietic tissues	
ICD-O-3	C42.1 M-9962/3	
Definition	Overproduction of thrombocytes/platelets	
Treatment	Anagrelide (platelet inhibitor)	Other therapy 1
	Hydroxyurea	Chemotherapy 2
	Aspirin (blood thinner)	Other therapy 1
	Interferon alfa	Immunotherapy 1
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	None	
Notes	• Chronic disease with a small propensity (10%) to convert to AML. • Essential thrombocythemia can develop a "spent phase" late in its course that resembles agnogenic myeloid metaplasia with cytopenia and marrow hypoplasia and fibrosis. • Long median survival	
	• Do not accession thrombocytopenia; this is not the same condition as thrombocythemia.	

Chronic neutrophilic leukemia Chronic Myeloproliferative Disorders
9963/3

Preferred Term	Chronic neutrophilic leukemia
Synonyms	None
ICD-9-CM	205.1 Chronic myeloid leukemia
ICD-O-3	C42.1 M-9963/3
Definition	Rare myeloproliferative disease characterized by sustained peripheral blood neutrophilia and hypercellular bone marrow.
Treatment	No information found.
EOD	999 80 9 99 99
Summary Stage 2000	7 Distant
Other staging	None
Notes	• Fewer than 100 cases reported in international literature, mostly reporting on cytogenetics and/or association with other hematopoietic diseases. • Sometimes categorized as an atypical chronic myeloproliferative disease (in contrast to "classic" myeloproliferative diseases like polycythemia vera).

Hypereosinophilic syndrome Chronic Myeloproliferative Disorders
9964/3

Preferred Term	Hypereosinophilic syndrome	
Synonyms	HES Chronic eosinophilic leukemia Chronic eosinophilic syndrome Idiopathic hypereosinophilic syndrome (IHS, IHES)	
ICD-9-CM	205.1 Chronic myeloid leukemia 288.3 Eosinophilia (hypereosinophilic syndrome)	
ICD-O-3	C42.1 M-9964/3	
Definition	Persistent high eosinophil count when all known causes of a raised eosinophil count (such as parasite infection) have been excluded. Rare variant of acute myeloid leukemia in which blasts and immature eosinophils proliferate. CNS involvement appears to be common.	
Treatment	If no organ damage is identified, no initial treatment	Do not code; record in remarks
	Supportive care to suppress organ damage	Do not code; record in remarks
	Corticosteroids (first line therapy) (prednisone, prednisolone)	Hormone therapy 1
	Chemotherapy (hydroxyurea, cytoxan, vincristine, myleran, methotrexate, leukeran, etoposide)	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	Interferon alfa, cyclosporine	Immunotherapy 1
	Splenectomy for palliation	All other sites scheme: Surgery of other sites 1
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	None	
Notes	• Do not accession Eosinophilia, NOS. Eosinophilia is sometimes thought of as a hypersensitivity illness or an allergic reaction producing excess eosinophils. This type is generally reversible. Only hypereosinophilic syndrome (as a myeloproliferative disorder) and chronic eosinophilic leukemia are considered neoplastic (uncontrolled production of excess eosinophils) and reportable. • Eosinophilic leukemia should be distinguished from CML with large numbers of eosinophils. • The “syndrome” includes gradual onset of cardiopulmonary symptoms accompanied by fever, sweating, fatigue, and weight loss, often with hepatosplenomegaly, lymphadenopathy, and congestive heart failure. Tissue infiltration by eosinophils leads to tissue damage in heart, lungs, CNS, skin, and GI tract. Rapid intervention for cardiac disease is essential.	

THE REFRACTORY ANEMIAS
(WHO Classification: Myelodysplastic Syndromes)

The following diagnoses are collectively termed myelodysplastic syndromes and are generally treated the same way.

ICD-O-3	Diagnosis	Blood findings	Bone marrow findings
9980/3	Refractory anemia (5-10% of MDS cases)	Anemia; no or rare blasts	<5% blasts; <15% ringed sideroblasts
9982/3	Refractory anemia with ringed sideroblasts (10-12% of MDS cases)	Anemia; no blasts	<5% blasts; 15% or more ringed sideroblasts
9983/3	Refractory anemia with excess blasts (40% of MDS cases)	Cytopenias type 1: <5% blasts, no Auer rods type 2: 5-19% blasts; with or without Auer rods	Uni- or multilineage dysplasia type 1: 5-9% blasts; no Auer rods type 2: 10-19% blasts; with or without Auer rods
9984/3	Refractory anemia with excess blasts in transformation [obs] (RAEB-T)	This diagnosis is not part of the WHO classification, which eliminated the RAEB-T category and lowered the blast threshold for the diagnosis of AML from 30 percent to 20 percent. The term was previously used in the FAB classification.	
9985/3	Refractory cytopenia with multilineage dysplasia (24% of MDS cases)	Bi- or pancytopenia; no or rare blasts; no Auer rods	<5% blasts; <15% ringed sideroblasts; no Auer rods; dysplasia in 10% or more of cells in two or more myeloid cell lines
9986/3	Myelodysplastic syndrome with 5q deletion (5q-) syndrome	Anemia; <5% blasts; usually normal or increased platelet count	<5% blasts; isolated 5q deletion cytogenetic abnormality; no Auer rods
9989/3	Myelodysplastic syndrome, NOS	Cytopenias; no or rare blasts; no Auer rods	Unilineage dysplasia (one myeloid cell line); <5% blasts; no Auer rods

Adapted from: Jaffe et al.: *World Health Organization Classification of Tumours: Pathology and Genetics, Tumors of Haematopoietic and Lymphoid Tissues*

ICD-9-CM Casefinding Codes for Refractory Anemias

ICD-9-CM code 284.9 lists a mix of aplastic anemia and early refractory anemia. Aplastic anemia and refractory anemia are not synonymous. Most cases of aplastic anemia may be autoimmune or may follow ingestion of a toxin or hepatitis. Refractory anemia, as used in the MDS context, is a clonal process.

Do **not** accession the following non-clonal, non-malignant terms:

aplastic anemia, unspecified
idiopathic aplastic anemia, NOS
medullary hypoplasia

ICD-9-CM code 285.0 lists a number of sideroblastic anemias (SA). The table below indicates that most of them are not malignant but are either inherited or secondary to other causes. ICD-O-3 is only interested in acquired primary [idiopathic] sideroblastic anemia. Great care must be taken to rule out non-primary and inherited anemias.

Do **not** accession the following diagnoses coded to 285.0:

hypochromic with iron-loading anemia
sideroachrestic anemia
sex-linked hypochromic SA
pyridoxine-responsive (hypochromic) anemia
secondary SA
drug-induced SA
chronic inflammatory SA
congenital SA
hereditary SA
vitamin B6-responsive SA
SA secondary to disease

Classification of Sideroblastic Anemias

1. Inherited
   - Sex-linked
   - Pyridoxine-responsive
   - Hypochromic anemia with iron loading

2. Acquired
   - A. Primary (also known as idiopathic)
     - Refractory anemias (with excess sideroblasts)—the myelodysplastic syndrome family (these are the only sideroblastic anemias that are reportable)
   - B. Secondary
     - Drug induced
     - Chronic inflammatory

International Prognostic Scoring System for Myelodysplastic Diseases
An estimate of survival rate and risk of progressing to acute myelogenous leukemia.
Score the three variables and add the values together for a total score. Then proceed to the next table.

Prognostic Variable	Score Value				
	0	**0.5**	**1.0**	**1.5**	**2.0**
Bone marrow blasts (%)	< 5%	5-10%		11-20%	21-30%
Karyotype[a]	Good	Intermediate	Poor		
Cytopenia[b] (lineages affected)	0 or 1	2 or 3			

a: Good = normal, -Y, del(5q), del (20q); intermediate = all other abnormalities;
poor = complex (3 abnormalities) or chromosome 7 abnormalities.
b: Cytopenias defined as Hb <100 g/L, platelet count < 100,000/L, absolute neutrophil count <1500/L.

Risk Group Scores	Total Score	Median Survival	% converting to AML
Low	0	68 mo	7
Intermediate-1	0.5-1.0	42 mo	10
Intermediate-2	1.5-2.0	42 mo	40
High	$ 2.5	5 mo	54

This page adapted from *Harrison's Online*, Chapter 109, page 14; www.harrisonsonline.com.

Refractory anemia Myelodysplastic syndromes
9980/3

Preferred Term	Refractory anemia	
Synonyms	RA Refractory anemia without sideroblasts Aregenerative anemia; nonregenerative anemia Primary refractory anemia	
ICD-9-CM	284.9 Aplastic anemia, unspecified	
ICD-O-3	C42.1 M-9980/3	
Definition	Bone marrow production of abnormal erythrocytes that result in megaloblastoid erythroid hyperplasia in the marrow and macrocytic anemia with reticulocytopenia in circulating blood.	
Treatment	Supportive care	Do not code; record in remarks
	Red cell transfusions	Other therapy 1; record in remarks
	Erythropoietin, colony stimulating factor, amifostine, antithymocyte globulin (Atgam)	Ancillary drugs–do not code; record in remarks
	Cytotoxic agents (occasionally beneficial) (cytarabine, topotecan, melphalan, azacytidine, decitabine)	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	Allogenic peripheral stem cell or bone marrow transplantation (under clinical evaluation)	Immunotherapy 5 (stem cell) or 3 (allogenic bone marrow transplantation); also 2 (autologous) or 4 (NOS)
	Hormones (ineffective)	Hormone therapy 1
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	French-American-British classification; Bournemouth score; Sanz score, Lille score; International Prognostic Scoring System for MDS *(see page 37)*	
Notes *(see also pages 35-37)*	• Occurs predominantly in older patients (over age 60). • Symptoms include anemia, bleeding, easy bruisability, fatigue. • Patients with RA or RARS who have intermediate-2 or higher-risk disease should probably be treated in the same manner as patients with RAEB. • Transformation to acute leukemia is uncommon (10%). • Median survival: 5 years.	

Refractory anemia with sideroblasts Myelodysplastic syndromes
9982/3

Preferred Term	Refractory anemia with sideroblasts	
Synonyms	RAS Refractory anemia with ringed sideroblasts (RARS) Sideroblastic anemia; primary sideroblastic anemia; pure sideroblastic anemia Refractory anemia with hemochromatosis Acquired idiopathic sideroblastic anemia (AISA)	
ICD-9-CM	285.0 Sideroblastic anemia	
ICD-O-3	C42.1 M-9982/3	
Definition	Bone marrow production of abnormal cells that results in megaloblastoid erythroid hyperplasia in the marrow and macrocytic anemia with reticulocytopenia in circulating blood (same as RA); 15% of marrow red cell precursors are ringed sideroblasts.	
Treatment	Supportive care	Do not code; record in remarks
	Red cell transfusions	Other therapy 1; record in remarks
	Erythropoietin, colony stimulating factor, amifostine, antithymocyte globulin (Atgam)	Ancillary drugs–do not code; record in remarks
	Cytotoxic agents (occasionally beneficial) (cytarabine, topotecan, melphalan, azacytidine, decitabine)	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	Allogenic peripheral stem cell or bone marrow transplantation (under clinical evaluation)	Immunotherapy 5 (stem cell) or 3 (allogenic bone marrow transplantation); also 2 (autologous) or 4 (NOS)
	Hormones (ineffective)	Hormone therapy 1
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	French-American-British classification; Bournemouth score; Sanz score, Lille score; International Prognostic Scoring System for MDS *(see page 37)*	
Notes *(see also pages 35-37)*	• Larger number of red cell precursors unable to use iron to make the hemoglobin to carry oxygen around blood stream. Unused iron is deposited in characteristic rings within cell. • Occurs predominantly in older patients. • Symptoms include anemia, bleeding, easy bruisability, fatigue; progressive iron overload. • Transformation to acute leukemia is uncommon (10%). • Median survival: 6 years. • Patients with RARS who have intermediate-2 or higher-risk disease should probably be treated in the same manner as patients with RAEB.	

Refractory anemia with excess blasts Myelodysplastic syndromes
9983/3

Preferred Term	Refractory anemia with excess blasts	
Synonyms	RAEB RAEB I; RAEB-1 [defined as 5-9% blasts in marrow] RAEB II; RAEB-2 [defined as 10-19% blasts in marrow]	
ICD-9-CM	285.0 Sideroblastic anemia	
ICD-O-3	C42.1 M-9983/3	
Definition	Abnormal production of erythrocytes, megakaryocytes and myelocytes. 5 to 20% myeloid blasts in bone marrow; subdivided type I, 5-9% and type II, 10-19%); 1 to 5% blasts in circulating blood.	
Treatment	Supportive care	Do not code; record in remarks
	Red cell transfusions	Other therapy 1; record in remarks
	Erythropoietin, colony stimulating factor; desferrioxamine (iron chelating agent	Ancillary drugs–do not code; record in remarks
	Cytotoxic agents (occasionally beneficial) (cytarabine, topotecan, melphalan)	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	Allogenic peripheral stem cell or bone marrow transplantation (under clinical evaluation)	Immunotherapy 5 (stem cell) or 3 (allogenic bone marrow transplantation); also 2 (autologous) or 4 (NOS)
	Hormones (ineffective)	Hormone therapy 1
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	French-American-British classification; Bournemouth score; Sanz score, Lille score; International Prognostic Scoring System for MDS *(see page 37)*	
Notes *(see also pages 35-37)*	• Occurs predominantly in patients over age 50. • Symptoms include anemia, bleeding, easy bruisability, fatigue. • Transformation to acute leukemia: 25% in RAEB I; 33% in RAEB II. • Median survival: RAEB I, 18 months; RAEB II, 10 months. • Patients with RAEB are usually in the intermediate-2 and high-risk groups and should be considered for treatment options with the intent to cure.	

Refractory anemia with excess blasts in transformation Myelodysplastic syndromes
9984/3

Preferred Term	Refractory anemia with excess blasts in transformation [obs]	
Synonyms	RAEB-T, RAEB(t), RAEBIT	
ICD-9-CM	285.0 Sideroblastic anemia	
ICD-O-3	C42.1 M-9984/3	
Definition	Panmyelosis with 20 to 30% blasts in bone marrow, > 5% blasts in circulating blood.	
Treatment	Supportive care	Do not code; record in remarks
	Red cell transfusions	Other therapy 1; record in remarks
	Erythropoietin, colony stimulating factor; desferrioxamine (iron chelating agent	Ancillary drugs–do not code; record in remarks
	Cytotoxic agents (occasionally beneficial) (cytarabine, topotecan, melphalan)	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	Allogenic peripheral stem cell or bone marrow transplantation (under clinical evaluation)	Immunotherapy 5 (stem cell) or 3 (allogenic bone marrow transplantation); also 2 (autologous) or 4 (NOS)
	Hormones (ineffective)	Hormone therapy 1
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	French-American-British classification; Bournemouth score; Sanz score, Lille score; International Prognostic Scoring System for MDS *(see page 37)*	
Notes *(see also pages 35-37)*	• Occurs predominantly in older patients. • Symptoms include anemia, bleeding, easy bruisability, fatigue. • Transformation to acute leukemia in 60 to 75% of patients. • Median survival: 6 months or less. • Patients with RAEB-T are usually in the intermediate-2 and high-risk groups and should be considered for treatment options with the intent to cure.	
	• RAEB-T was a category in the former FAB classification with a criterion of 20-30% blasts in the bone marrow. In the World Health Organization classification of hematopoietic diseases, the minimum percentage of blasts for acute myeloid leukemia was lowered to 20%, and cases formerly meeting RAEB-T criteria are now classified as acute myeloid leukemia (AML).	

Refractory cytopenia with multilineage dysplasia Myelodysplastic syndromes
9985/3

Preferred Term	Refractory cytopenia with multilineage dysplasia	
Synonyms	RCMD Myelodysplastic syndrome, unclassified	
ICD-9-CM	238.7 Neoplasm of uncertain behavior of other lymphatic and hematopoietic tissues	
ICD-O-3	C42.1 M-9985/3	
Definition	Bi- or pancytopenia with bone marrow dysplasia of more than one cell line.	
Treatment	Supportive care	Do not code; record in remarks
	Red cell transfusions	Other therapy 1; record in remarks
	Erythropoietin, colony stimulating factor; desferrioxamine (iron chelating agent)	Ancillary drugs–do not code; record in remarks
	Hormones (ineffective)	Hormone therapy 1
	Cytotoxic agents (occasionally beneficial) (cytarabine, topotecan, melphalan)	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	Allogenic peripheral stem cell or bone marrow transplantation (under clinical evaluation)	Immunotherapy 5 (stem cell) or 3 (allogenic bone marrow transplantation); also 2 (autologous) or 4 (NOS)
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	International Prognostic Scoring System for MDS *(see page 37)*	
Notes *(see also pages 35-37)*	• Related to M-9895/3, acute myeloid leukemia with multilineage dysplasia • 24% of MDS cases in WHO classification. • Occurs predominantly in older patients. • Symptoms include significant neutropenia and thrombocytopenia with <1% blasts in blood and <5% blasts in marrow. Chromosomal abnormalities similar to RAEB. • Transformation to acute leukemia occurs in 11% of cases. • Median survival: 33 months	
	• RCMD is a new diagnosis listed in the World Health Organization classification of hematopoietic diseases.	

Myelodysplastic syndrome with 5q- syndrome Myelodysplastic syndromes
9986/3

Preferred Term	Myelodysplastic syndrome (MDS) with 5q- syndrome	
Synonyms	MDS with 5q deletion syndrome MDS with chromosome 5 abnormality MDS associated with isolated del(5q) chromosome abnormality	
ICD-9-CM	238.7 Neoplasm of uncertain behavior of other lymphatic and hematopoietic tissues	
ICD-O-3	C42.1 M-9986/3	
Definition	Bone marrow dysplasia showing refractory macrocytic anemia with oval macrocytes and monolobulated megakaryocytes. 75% of patients have increased cellularity of marrow. One third of patients have chromosomal abnormality that consists of deletion of the long arm of chromosome 5 (5q). Marrow blasts < 5%.	
Treatment	Supportive care	Do not code; record in remarks
	Red cell transfusions	Other therapy 1; record in remarks
	Erythropoietin, colony stimulating factor	Ancillary drugs–do not code; record in remarks
	Hormones (ineffective)	Hormone therapy 1
	Cytotoxic agents (occasionally beneficial) (cytarabine, topotecan, melphalan)	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	Allogenic peripheral stem cell or bone marrow transplantation (under clinical evaluation)	Immunotherapy 5 (stem cell) or 3 (allogenic bone marrow transplantation); also 2 (autologous) or 4 (NOS)
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	International Prognostic Scoring System for MDS *(see page 37)*	
Notes *(see also pages 35-37)*	• More common in women (2:1 ratio). • Occurs predominantly in older patients. • Symptoms include transfusion-dependent refractory anemia with iron overload. • Better prognosis than other types of MDS • Transformation to acute leukemia in 10% of patients.	
	• MDS with 5q- syndrome is a new diagnosis in the World Health Organization classification of hematopoietic diseases.	

Therapy-related myelodysplastic syndrome Myelodysplastic Syndromes
9987/3

Preferred Term	Therapy-related myelodysplastic syndrome (MDS)	
Synonyms	Therapy-related myelodysplastic syndrome (unspecified/NOS) Secondary myelodysplastic syndrome (secondary MDS) Therapy-related myelodysplastic syndrome, alkylating agent related Therapy-related myelodysplastic syndrome, epipodophyllotoxin related	
ICD-9-CM	238.7 Neoplasm of uncertain behavior of other lymphatic and hematopoietic tissues	
ICD-O-3	C42.1 M-9987/3	
Definition	Bone marrow failure that develops after treatment with chemotherapy or radiation therapy. There is no predominant cell type that is dysplastic, and the marrow aspirate is often inadequate to review for dysplasia, as the marrow is often hypocellular with fibrosis.	
Treatment	Allogenic peripheral stem cell or bone marrow transplantation (under clinical evaluation)	Immunotherapy 5 (stem cell) or 3 (allogenic bone marrow transplantation); also 2 (autologous) or 4 (NOS)
	Erythropoietin, colony stimulating factor	Ancillary drugs–do not code; record in remarks
	Supportive care	Do not code; record in remarks
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	International Prognostic Scoring System for MDS *(see page 37)*	
Notes *(see also pages 35-37)*	• Only about 10% of all MDS patients have secondary MDS. • Prognosis unfavorable; median survival: 10-12 months. • Alkylating agent-related (85% of cases): due to cytoxan, myleran, or leukeran chemotherapy; also MOPP regimen. Occasionally nitrosourea associated. • Epipodophyllotoxin related (< 15% of cases): due to VP-16, etoposide, vepesid, teniposide (vumon) or epipodophyllotoxin chemotherapy • Poorer prognosis than other types of MDS due to resistance to therapy • Patients convert to AML within a short period. *continued on next page*	

Therapy-related myelodysplastic syndrome notes, *continued*	• Therapy-related MDS is a new diagnosis listed in the World Health Organization classification of hematopoietic diseases. • Therapy-related myelodysplasia is a recognized long-term complication of cancer chemotherapy and radiation therapy. Therapy-related MDS has been reported most frequently after treatment for Hodgkin lymphoma, non-Hodgkin lymphoma, multiple myeloma, ovarian cancer, testicular cancer, breast cancer, and polycythemia rubra vera. Therapy-related MDS usually develops 3-7 years after exposure to chemotherapy and is most frequently related to complete or partial loss of chromosome 7. • Approximately 80% of cases of AML occurring after exposure to antineoplastic drugs are preceded by MDS.

Myelodysplastic syndrome, NOS Myelodysplastic Syndromes
9989/3

Preferred Term	Myelodysplastic syndrome, NOS	
Synonyms	MDS Myelodysplastic syndrome, unclassifiable Older terminology: preleukemia, preleukemic syndrome, smoldering leukemia, refractory dysmyelopoietic anemia, subacute myelogenous leukemia Myelodysplasia, NOS *See notes regarding this term on page 47.*	
ICD-9-CM	238.7 Neoplasm of uncertain behavior of other lymphatic and hematopoietic tissues	
ICD-O-3	C42.1 M-9989/3	
Definition	A myelodysplastic syndrome that lacks findings appropriate for classification as any of the refractory anemias. Blasts in the blood and marrow are not increased. *See notes regarding myelodysplasia, NOS on page 47.*	
Treatment	Allogenic peripheral stem cell or bone marrow transplantation (under clinical evaluation)	Immunotherapy 5 (stem cell) or 3 (allogenic bone marrow transplantation); also 2 (autologous) or 4 (NOS)
	Erythropoietin, colony stimulating factor	Ancillary drugs–do not code; record in remarks
	Supportive care	Do not code; record in remarks
	Cytotoxic agents (occasionally beneficial) (cytarabine, topotecan, melphalan)	Chemotherapy 1 (NOS), 2 (single agent), or 3 (multiple agents)
	Red cell transfusions	Other therapy 1; record in remarks
EOD	999 80 9 99 99	
Summary Stage 2000	7 Distant	
Other staging	French-American-British classification; Bournemouth score; Sanz score, Lille score; International Prognostic Scoring System for MDS *(see page 37)*	
Notes *(see also pages 35-37)*	• Occurs predominantly in older patients. • Percent transforming to acute leukemia: unknown. • Median survival: 6 months or less. *continued on next page*	

Myelodysplastic syndrome notes, continued	Myelodysplasia (NOS) is a term with two meanings: the bone marrow malfunction and malignancy and also disorders of spinal cord development (such as spina bifida). The term is sometimes used as a synonym for myelodysplastic syndrome, NOS (M-9989/3). Make sure that the diagnosis refers to the hematopoietic disease. Then determine whether the physician is using the term generically to describe bone marrow malfunction (such as thrombocytopenia or pancytopenia) or referring to myelodysplasia as part of a neoplastic syndrome (with reference to refractory anemia or some other reportable term). Myelodysplasia as a spinal cord disorder or describing a category of bone marrow failure (with reference to the –penias) is not reportable.

APPENDIX 1
Coding Treatment and Phenotype for Hematopoietic Diseases

from: "Clarifications for Abstracting and Coding Hematopoietic Diseases" (May 22, 2001), SEER Program, National Cancer Institute. Available from www.seer.cancer.gov/Admin

TREATMENT

For many of the newly reportable hematopoietic diseases, the principal treatment is either supportive care, observation, or another type of treatment that does not meet the usual definition that treatment "modifies, controls, removes or destroys proliferating cancer tissue." Such treatments include phlebotomy, transfusions, aspirin, supportive care and observation. In order to document that patients with hematopoietic diseases did have some medical treatment, SEER and the Commission on Cancer have agreed to record these treatments as "Other Treatment" (code 1) for the hematopoietic diseases ONLY. A complete description of the treatment plan should be recorded in the text field for "Other Treatment" on the abstract.

- Transfusions may include whole blood, RBCs, platelets, plateletpheresis, fresh frozen plasma (FFP), and cryoprecipitate.
- Phlebotomy may be called blood removal, blood letting, or venisection.
- Aspirin (also known as ASA or acetylsalicylic acid and many brand names) is used as a treatment for essential thrombocythemia. To determine whether aspirin is administered for pain, cardiovascular protection or thinning of platelets in the blood, use the following general guideline: pain control: 325-1000 mg every 3-4 hours; cardio-vascular protection: starts at about 160 mg/day; aspirin treatment for essential thrombocythemia is low dose (70-100 mg/day). Record ONLY aspirin therapy intended to prevent platelet aggregation for symptomatic control of thrombocythemia.

Standard cancer treatments such as chemotherapy, radiation (including P32 for polycythemia) and surgery (such as splenectomy for myelofibrosis) should be recorded in the appropriate data fields.

CODING PHENOTYPE

Assigning 6th digit immunophenotype Sixth digit codes for T-cell, B-cell, and NK-cell phenotyping of lymphomas and leukemias should be based on the diagnosis as specifically stated in the pathology report. Sixth digit phenotype codes should not be used when T- or B- cell is implied from the boldface header in the morphology numeric list. In other words, if no T- or B-cell designation is provided in the pathology or laboratory report, do NOT code the T- or B- cell designation based on the boldface header in ICD-O-3. For example, a diffuse large B-cell lymphoma would be coded to 9680/36; a diffuse centroblastic lymphoma would be coded to 9680/39. When cases are analyzed, they can be grouped by cell line as stated in the category headings in the lymphoma and leukemia sections of the morphology numeric list.

APPENDIX 2
Guidelines for Reporting and Sequencing the Hematopoietic Diseases

Definition of NRHD: Newly Reportable Hematopoietic Disease

- Any of the myeloproliferative or myelodysplastic diseases that changed from /1 borderline to /3 malignant in the ICD-O-3. Diagnoses marked with an asterisk (*) in the Table of Contents would be considered NRHD.

Reporting Guidelines

1. Accession and abstract only ***new*** NRHD diagnoses.
   - If disease was known prior to 2001, do not accession.

     Example: Patient with primary myelosclerosis diagnosed in May 2000 and currently receiving monthly blood transfusions.
     *This case would **not** be new and reportable because the diagnosis was known prior to 01/01/2001.*

2. Active treatment does not affect new reportability
   - Disregard NRHD diagnosed prior to 01/01/2001 undergoing active treatment.

     Example: Patient with idiopathic thrombocythemia diagnosed in 11/1999 is admitted for regular 8 week cycles chemotherapy through 2000 and into 2001.
     *This is **not** a new and reportable case even though the patient is receiving active treatment in 2001 because the diagnosis was known prior to 01/01/2001.*

3. Compare diagnoses to check for transition to another hematopoietic disease.
   - To determine whether patient has a single primary or more than one, use the ICD-O-3 hematopoietic diseases multiple primaries foldout table available from SEER.
     - Code the first diagnosis in ICD-O-3 even if diagnosed prior to 2001.
     - Code the second diagnosis in ICD-O-3.
     - Find the row containing the first diagnosis.
     - Find the column containing the second diagnosis.
     - Note the symbol in the intersection cell:
       If S, the second diagnosis is presumably the same primary as the first diagnosis
       If D, the second diagnosis is a different disease process than the first diagnosis and a subsequent abstract should be prepared.

     Examples:
     First diagnosis: Hodgkin lymphoma. Second diagnosis: multiple myeloma.
     These are different primaries. Prepare 2 abstracts.
     First diagnosis: Diffuse large B-cell lymphoma. Second diagnosis: follicular lymphoma, grade 2.
     These are the same disease process. Do not prepare a second abstract.

Sequencing Newly Reportable Hematopoietic Diseases

1. For the newly reportable hematopoietic diseases, sequence numbering applies as of 01/01/2001.

   Example: Colon cancer diagnosed 1998, refractory anemia diagnosed 02/20/2001.
   Colon cancer originally sequence 00 becomes sequence 01 and refractory anemia is sequence 02.

2. A first primary (any site) is sequence 00.

   Example: No cancers known prior to 02/2001 diagnosis of refractory anemia.
   Refractory anemia is sequence 00.
   Then, in 07/2001, that same patient's bone marrow shows acute myeloid leukemia.
   According to the hematopoietic diseases multiple primaries table, this is the same disease process. Sequence remains 00 and histology remains coded as refractory anemia. (Do make a note in a text field that the disease has undergone a transformation to leukemia.)

3. Any subsequent reportable primary adds to sequence.

   Example: Chronic myelomonocytic leukemia diagnosed 05/2001. Prostate cancer diagnosed 09/2001.
   CMML originally sequence 00 becomes sequence 01 and prostate cancer is sequence 02.

4. If the NRHD was not sequenced before, do not count it now.

   Example: Polycythemia vera diagnosed 1999 (stable), breast cancer diagnosed 10/19/2001.
   Disregard polycythemia since it was not reportable in 1999. Breast cancer is sequence 00.

   Example: Agnogenic myeloid metaplasia diagnosed 2000. Bone marrow biopsy 02/2001 shows primary myelofibrosis.
   Primary myelofibrosis is another name for agnogenic myeloid metaplasia--they have the same morphology code. Since the AMM was diagnosed prior to 2001, this case is not added to the registry.

   Exception to sequencing guideline #4:
   If a hematopoietic disease diagnosed before 01/01/2001 transforms into another disease (in other words, can now be coded with a different code number) after 01/01/2001, enter the case into the registry under the name of the disease diagnosed after 01/01/2001 with the 2001 or later diagnosis date, *even though the Hematopoietic Diseases multiple primaries table says they are the same disease process.* (This exception allows the case to be included in the registry for incidence purposes. After a few years, it will not be necessary to apply the exception because most hematopoietic diseases will be diagnosed after 01/01/2001.)

   Example: Myelodysplastic syndrome diagnosed 1998 and followed with periodic bone marrow biopsies. Bone marrow 03/2001 shows refractory anemia with excess blasts.
   Myelodysplastic syndrome (9989/3) was not reportable. The recent diagnosis of RAEB has a different number (9983/3) even though it is part of the same disease process. Therefore, it is now reportable. Accession the case as refractory anemia with excess blasts with a 2001 diagnosis date and note in remarks that the myelodysplastic syndrome was diagnosed previously.

RESOURCES

Coding Resources

International Classification of Diseases for Oncology, Third Edition. Geneva, World Health Organization, 2000.

International Statistical Classification of Diseases, Injuries, and Causes of Death. Ninth Revision. Clinical Modification. Washington, DC, US Department of Health and Human Services, 1979 (DHHS No. (PHS) 80-1260).

SEER Summary Staging Manual 2000: Codes and Coding Instructions. Bethesda, MD, National Cancer Institute, NIH Pub. No. 01-4969.

SEER Extent of Disease: Codes and Coding Instructions, Third Edition. Bethesda, MD, National Cancer Institute, 1998 NIH Pub. No. 98.

ICD-9-CM Internet site: http://www.e-mds.com/icd9 (e-MDs coding resource)

Disease Resources

World Health Organization Classification of Tumours: Pathology and Genetics, Tumors of Haematopoietic and Lymphoid Tissues, ES Jaffe, NL Harris, H Stein and JW Vardiman. Lyon: IARC Press, 2001.

Consultations with pathologists: James Vardiman, MD (see above); Timothy Cote, MD, CTR; Marshall Lichtman, MD, and K. Shanmugaratnam, MD

Internet Sites

National Cancer Institute CancerNet information service
http://www.cancer.gov/cancer_information/cancer_type/

State of the Science Leukemia Conference (10/30-31/00)
http://www.conference-cast.com/webtie/sots/leukemia2/lectures.htm

American Society of Hematology educational materials
http://www.hematology.org/education/educationbook.cfm

Frontiers in Bioscience
http://www.bioscience.org/atlases/tumpath/hematop/resource.htm

Leukaemia Research Fund
http://dspace.dial.pipex.com/lrf-/diseases/index.htm

Emedicine--instant access to the minds of medicine
http://www.emedicine.com/

The Histiocyte Society
http://www.histio.org/society/

Medline Plus health information
http://www.nlm.nih.gov/medlineplus/ency/article/000068.htm

Lymphoma Information Network
http://www.lymphomainfo.net/nhl/classify.html

Merck Manual Online
http://www.merck.com/pubs/mmanual_home/

Various other internet resources, including web sites for specific diseases and other online references

INDEX TO ICD-9-CM CODES USED IN THIS GUIDE

200.0 Lymphosarcoma and reticulosarcoma, 13
200.8 Lymphoma, other named variants, 20
202.3 Malignant histiocytosis 10, 11
202.4 Leukemic reticuloendotheliosis, 24
202.5 Letterer-Siwe disease, 11
202.6 Malignant mast cell tumors, 7, 8
202.9 Other and unspecified malignant neoplasms of lymphoid and histiocytic tissue, 14, 15, 16
203.0 Multiple myeloma, 3
203.1 Plasma cell leukemia, 4
203.8 Other immunoproliferative neoplasms, 17, 20
204.8 Other lymphoid leukemia, 27
205.1 Chronic myeloid leukemia, 25, 33, 34
205.3 Myeloid sarcoma, 22
205.8 Other myeloid leukemia, 21
205.9 Unspecified myeloid leukemia (not specified as acute or chronic), 26
207.1 Chronic erythremia (Hilmeyer-Schoner disease), 28
207.8 Other specified leukemia, 9, 27
238.4 Polycythemia vera, 28
238.5 Neoplasm of uncertain behavior of histiocytic and mast cells, 6
238.6 Neoplasm of uncertain behavior of plasma cells, 2, 5
238.7 Neoplasm of uncertain behavior of other lymphatic and hematopoietic tissues, 23, 30, 31, 32, 42, 43, 44, 46
273.2 Other paraproteinemias, 19
273.3 Macroglobulinemia, 18
284.9 Aplastic anemia, unspecified, 38
285.0 Sideroblastic anemia, 39, 40, 41
288.3 Eosinophilia (hypereosinophilic syndrome), 34
289.8 Other specified diseases of blood and blood-forming organs, 31

DIAGNOSIS INDEX

indicates a diagnosis that is NOT reportable
Italicized page number: a reference to the diagnosis

α-heavy chain disease, 19
γ-heavy chain disease, 19
δ-heavy chain disease, 19
μ-heavy chain disease, 19
5q deletion syndrome, myelodysplastic syndrome with, *35*, 43
5q- syndrome, myelodysplastic syndrome with, *35*, 43

Abnormality, chromosome 5, myelodysplastic syndrome with, 43
Abnormality, isolated del(5q) chromosome, myelodysplastic syndrome associated with, 43
Acquired idiopathic sideroblastic anemia, *36*, 39
Acquired primary sideroblastic anemia, *36*
Acute
- differentiated progressive histiocytosis, 12
- infantile reticuloendotheliosis, 12
- lymphocytic leukemia, *17*
- myelodysplasia with myelofibrosis, 23
- myelofibrosis, 23
- myelogenous leukemia, *21*
- myelogenous leukemia FAB M5, 12
- myeloid leukemia, *22*, *41*
  - secondary, due to prior chemotherapy, 21
  - therapy-related, 21
  - therapy-related alkylating agent-related, 21
  - therapy-related epipodophyllotoxin-related, 21
  - topoisomerase II inhibitor-related, 21
  - with multilineage dysplasia, *42*
- myelosclerosis, NOS, 23
- panmyelosis, NOS, 23
- panmyelosis with myelofibrosis, 23
- progressive histiocytosis X, 12
- reticulosis of infancy, 12

Aggressive
- large granular lymphocyte leukemia, NK-cell type, 27
- mastocytosis, *9*
- NK-cell leukemia, 27
- NK-cell leukemia-lymphoma, 27
- systemic mastocytosis, 8

Agnogenic myeloid metaplasia, *23*, *28*, 31, *32*
AISA, 39
Alkylating agent-related therapy-related acute myeloid leukemia, 21
Alkylating agent-related therapy-related myelodysplastic syndrome, 44
Alpha chain disease, 19
Alpha heavy chain disease, *20*
AMM, 31
Amyloidosis, primary , *1*

Anemia
- aplastic *see Aplastic anemia*
- aregenerative, 38
- dysmyelopoietic, refractory, 46
- hypochromic with iron loading , *36*
- nonregenerative, 38
- pyridoxine-responsive hypochromic , *36*
- refractory *see Refractory anemia*
- sideroachrestic , *36*
- sideroblastic *see Sideroblastic anemia*

ANLL M7 with prominent fibrosis, *23*
Aplastic anemia, idiopathic, NOS , *36*
Aplastic anemia, unspecified , *36*
Aregenerative anemia, 38
ASM, 8
Atypical chronic myeloproliferative disease, *33*

B-cell lymphoma, *17*
Blasts, excess, in transformation, refractory anemia with [obs], *35*, 41
Blasts, excess, refractory anemia with, *35*, 40
Breast cancer, *21*, *45*

Cancer
- breast, *21, 45*
- ovarian, *45*
- testicular, *45*

Chloroma, 22
Chromosome 5 abnormality, myelodysplastic syndrome with , 43
Chronic
- eosinophilic leukemia, 34
- eosinophilic syndrome, 34
- erythremia [obs], 28
- granulocytic-megakaryocytic myelosclerosis, 31
- idiopathic myelofibrosis, *22*, 31
- inflammatory sideroblastic anemia , *36*
- lymphocytic leukemia, *17*, *19*, *21*, *24*
- myeloid leukemia, *22*, *34*
- myeloid leukemia, juvenile, 26
- myelomonocytic leukemia
  - NOS, 25
  - in transformation [obs], 25
  - juvenile, 26
- myelomonocytic syndrome, 25
- myeloproliferative disease, 30
- myeloproliferative disease, atypical, *33*
- myeloproliferative disorder, 30
- neutrophilic leukemia, 33

CLL, *19*
CMD, 30
CML, *34*

CMML, 25
CMML type I, 25
CMML type II, 25
CMML-1 , 25
CMML-2 , 25
Congenital sideroblastic anemia , *36*
Cutaneous histiocytosis, pure , *12*
Cutaneous mastocytosis, diffuse, *8*, *9*
Cytopenia, refractory, with multilineage dysplasia, *35*, 42

Delta heavy chain disease, 19
Dendritic cell
sarcoma
NOS, 15
follicular, *15,* 16
interdigitating, 15, *16*
tumor, follicular, 16
tumor, interdigitating , 15
Dendritic reticulum cell sarcoma, 16
Differentiated progressive histiocytosis, acute, 12
Diffuse cutaneous mastocytosis, *9*
Disease
alpha-chain, 19
alpha heavy chain, *20*
atypical chronic myeloproliferative, *33*
chronic myeloproliferative, 30
Franklin, 19
gamma-chain, 19
Hand-Schuller-Christian [obs] , *10*, 12
heavy chain, *1, 17*, 19
immunoproliferative, NOS, 17
Kahler's, 3
Letterer-Siwe, *10*, 12
mast cell , 6
mast cell proliferative , 6
mu-chain, 19
myeloproliferative, *30*
myeloproliferative, atypical chronic, *33*
myeloproliferative, myelofibrosis as a result of, 31
Niemann-Pick , *10*
Osler's, 28
Osler-Vaquez, 28
Rosai-Dorfman , *10*
small intestinal, immunoproliferative, *1, 17*, *19*, 20
systemic tissue mast cell, 8
Vaquez-Osler, 28
Disorder
chronic myeloproliferative, 30
immunoproliferative, 17
myeloproliferative, *30*
Disseminated Langerhans cell histiocytosis, *10*, 12
Disseminated Langerhans cell histiocytosis, progressive, 12
Drug-induced sideroblastic anemia , *36*
Dyscrasias, transient plasma cell , *1*
Dysmyelopoietic anemia, refractory, 46
Dysplasia, multilineage, refractory cytopenia with, *35*, 42
Dysplasia, multilineage, acute myeloid leukemia with, *42*

EMP, 5
Eosinophilia, NOS , *34*
Eosinophilic
granuloma , *10*
multifocal , 12
solitary , 12
leukemia, chronic, 34
syndrome, chronic, 34
Epipodophyllotoxin-related therapy-related acute myeloid leukemia, 21
Epipodophyllotoxin-related therapy-related myelodysplastic syndrome, 44
Erythremia, chronic [obs], 28
Erythremia, primary, 28
Essential
hemorrhagic thrombocythemia, 32
thrombocythemia, 32
thrombocytosis, 32
Excess blasts, refractory anemia with, *35*, 40
Excess blasts in transformation, refractory anemia with [obs], *35*, 41
Extracutaneous mastocytoma , 6, *7*
Extramedullary myeloid tumor, 22
Extramedullary plasmacytoma (not occurring in bone), *1, 4, 5*

FDC sarcoma, 16
Follicular dendritic cell sarcoma, *15,* 16
Follicular dendritic cell tumor, 16
Franklin disease, 19
Fungoides, mycosis, *17*

Gamma-chain disease, 19
Gamma heavy chain disease, 19
Gamma-HCD, 19
Gammopathy, monoclonal, of undetermined significance , *1*
Generalized Langerhans cell histiocytosis, 12
Granular lymphocytic leukemia, T-cell large , *27*
Granulocytic sarcoma, 22
Granulocytic-megakaryocytic myelosclerosis, chronic, 31
Granuloma, eosinophilic, multifocal , 12
Granuloma, eosinophilic, solitary , 12
Granulomatosis, Langerhans cell , 12
Granulomatosis, Langerhans cell, unifocal , 12

Hairy cell leukemia, 24
Hairy cell leukemic variant, 24
Hand-Schuller-Christian disease [obs] , *10*, 12
Hashimoto-Pritzker syndrome , *12*
HCD, 19
Heavy chain disease, *1, 17*, 19
α-, 19
δ-, 19
γ-, 19
μ-, 19
alpha, 19, *20*
delta, 19
gamma, 19
IgA, 19
IgD, 19
IgG, 19
IgM, 19
mu, 19
Hemochromatosis, refractory anemia with, 39
Hemophagocytic syndrome, infection associated , 12
Hemophagocytic lymphohistiocytosis , 12
Hemorrhagic thrombocythemia, essential, 32
Hemorrhagic thrombocythemia, idiopathic, 32
Hereditary sideroblastic anemia , *36*
HES, 34
Histiocytic
lymphoma, true, 12, 13
medullary reticulosis [obs], 10
sarcoma, 13
sarcoma, true, 13
Histiocytosis
acute differentiated progressive, 12
Langerhans cell, *10*, 11, 12, *13, 14*
NOS , 12
disseminated, *10*, 12
generalized, 12
mono-ostotic, 12
multifocal , 12
poly-ostotic , 12
progressive disseminated, 12
unifocal , 12
lipid **,** *10*
malignant, 10, 12, *13*
pure cutaneous , *12*
self-healing , *12*
sinus, with massive lymphadenopathy , *10*, 12
systemic, 10
type II , 12
X, NOS , *10*, 12
X, acute progressive, 12
Hodgkin lymphoma, *17*, *21*, *45*
Hypereosinophilic syndrome, *22*, 34
Hypereosinophilic syndrome, idiopathic, 34
Hypochromic with iron loading anemia , *36*
Hypoplasia, medullary , *36*
IDC tumor , 15
IDCS, 15
Idiopathic
aplastic anemia, NOS , *36*
hemorrhagic thrombocythemia, 32
hypereosinophilic syndrome, 34
macroglobulinemia, 18
myelofibrosis, *23*, 31
myelofibrosis, chronic, *22*, 31
myeloid metaplasia, 31
thrombocythemia, 32
IgA heavy chain disease, 19
IgD heavy chain disease, 19
IgG heavy chain disease, 19
IgM heavy chain disease, 19
IHES, 34
IHS, 34
Immunocytoma, lymphoplasmacytic, *18*
Immunoproliferative
disease, NOS, 17
disorder, 17
neoplasia, 17
neoplasm, 17
small intestinal disease, *1, 17*, *19*, 20
Indolent mastocytosis , *9*
Indolent systemic mastocytosis , 8
Infantile reticuloendotheliosis, acute, 12
Infection-associated hemophagocytic syndrome , 12
Interdigitating
cell sarcoma, 15
cell sarcoma, paracortical, 15
dendritic cell sarcoma, 15, *16*
dendritic cell tumor , 15
reticulum cell sarcoma, 15
IPSID, 20
ISM , 8
Isolated del(5q) chromosome abnormality, myelodysplastic syndrome associated with, 43
Isolated plasmacytoma of bone, 2

JCML, 26
JMML, 26
Juvenile
chronic myeloid leukemia, 26
chronic myelomonocytic leukemia, 26
myelomonocytic leukemia, 26
myelomonocytic leukemia syndrome, 26

Kahler's disease, 3

Langerhans cell
granulomatosis , 12
granulomatosis, unifocal , 12
histiocytosis, *10*, 11, 12, *13*, *14*
NOS , 12
disseminated, *10*, 12
generalized, 12
mono-ostotic , 12
multifocal , 12
poly-ostotic , 12
progressive disseminated, 12
unifocal , 12
sarcoma, 14
Large granular lymphocyte leukemia, NK-cell type, aggressive, 27
Large granular lymphocytic leukemia, T-cell , *27*
LCH, 11
Letterer-Siwe disease, 12
Leukemia
acute lymphocytic, *17*
acute myelogenous, *21*
acute myelogenous, FAB M5, 12
acute myeloid *see Acute myeloid leukemia*
acute non-lymphocytic (ANLL) M7 with prominent fibrosis, *23*
chronic eosinophilic, 34
chronic lymphocytic, *17*, *19*, *21*, *24*
chronic myeloid, *22, 34*
chronic myeloid, juvenile, 26
chronic myelomonocytic
NOS, 25
in transformation [obs], 25
juvenile, 26
chronic neutrophilic, 33
hairy cell, 24
juvenile myelomonocytic, 26
juvenile myelomonocytic syndrome, 26
large granular lymphocyte, NK-cell type, aggressive, 27
NK-cell, aggressive, 27
mast cell, 9
megakaryoblastic, *23*
myelogenous, subacute, 46
myelomonocytic, subacute, 25
plasma cell, *1*, 4
primary, 4
secondary , 4
plasmacytic, *1*, 4
smoldering, 46
T-cell large granular lymphocytic , *27*
Leukemia-lymphoma, NK-cell, aggressive, 27
Leukemic reticuloendotheliosis, 24
Leukemic variant, hairy cell, 24
Light chain only myeloma, *4*
Lipid histiocytosis , *9*
Localized plasmacytoma, 2
Lymphadenopathic mastocytosis with eosinophilia, *9*
Lymphoblastic lymphoma, *1*
Lymphocytic
leukemia, acute, *17*
leukemia, chronic, *17, 19, 21*, *24*
leukemia, T-cell large granular , *27*
lymphoma, well-differentiated plasmacytoid, *18*
lymphoma, plasmacytoid small, *18*
Lymphohistiocytosis, hemophagocytic , 12
Lymphoma
B-cell, *17*
Hodgkin, *17*, *21*, *45*
lymphoblastic, *1*
lymphocytic well-differentiated plasmacytoid, *18*
lymphoplasmacytic, *2*, *18*
MALT, *20*
Mediterranean, *19*, 20
Mediterranean abdominal, 20
non-Hodgkin, *45*
plasmacytic-lymphocytic, *18*
small lymphocytic, plasmacytoid, *18*
T-cell, *17*
true histiocytic, 12, 13
well-differentiated plasmacytoid lymphocytic, *18*
Lymphoplasmacytic immunocytoma, *18*
Lymphoplasmacytic lymphoma, *2*, *18*

Macroglobulinemia, *1*, 18
idiopathic, 18
primary, 18
Waldenstrom, *17*, 18
Malignant
histiocytosis, 10, 12, *13*
mast cell tumor, 7
mastocytoma, 7
mastocytosis, 8
myelofibrosis, 23
myelosclerosis [obs], 23
MALT lymphoma, *20*
Massive lymphadenopathy with sinus histiocytosis , *10*, 12
Mast cell
disease , 6
disease, systemic tissue, 8
leukemia, 9
proliferative disease , 6
sarcoma, 7
tumor
NOS , 6
malignant, 7
Mastocytoma , 6
extracutaneous , 6, *7*
malignant, 7
solitary, of skin , 6, *9*

Mastocytosis, *9*
aggressive, *9*
aggressive systemic, 8
diffuse cutaneous, 9
indolent , *9*
indolent systemic , 8
lymphadenopathic, with eosinophilia, *9*
malignant, 8
systemic, 8, *9*
with associated clonal, hematological non-mast-cell lineage disease, 8
with associated hematologic disorder, *9*
MCL, 9
MCS, 7
MDS, 46 *see also Myelodysplastic syndrome*
associated with isolated del(5q) chromosome abnormality, 43
with 5q deletion syndrome, *35*, 43
with chromosome 5 abnormality, 43
Mediterranean abdominal lymphoma, 20
Mediterranean lymphoma, *19*, 20
Medullary
hypoplasia , *36*
plasmacytoma, 3
reticulosis, histiocytic [obs], 10
Megakaryoblastic leukemia (M7 ANLL) with prominent fibrosis, *23*
Megakaryocytic myeloid metaplasia, 31
Megakaryocytic myelosclerosis, 31
Megakaryocytic-granulocytic myelosclerosis, chronic, 31
Metaplasia
agnogenic myeloid, *23*, *28*, 31, *32*
idiopathic myeloid, 31
megakaryocytic myeloid, 31
myeloid
NOS, 31
with myelofibrosis, 31
with myelosclerosis, 31
MGUS , *1*
MH, 10
MMM, 31
Monoclonal gammopathy of undetermined significance , *1*
Mono-ostotic Langerhans cell histiocytosis , 12
Mono-ostotic myeloma, 2
Monocytic sarcoma, 13
Mu chain disease, 19
Multifocal eosinophilic granuloma , 12
Multifocal Langerhans cell histiocytosis , 12
Multilineage dysplasia, acute myeloid leukemia with, *42*
Multilineage dysplasia, refractory cytopenia with, *35*, 42
Multiple myeloma, *1*, 3, *4*, *5*, *45*
Mycosis fungoides, *17*
Myelodysplasia, NOS, 46, *47*
Myelodysplasia, acute, with myelofibrosis, 23
Myelodysplastic syndrome
NOS, *35*, 46
associated with isolated del(5q) chromosome abnormality, 43
secondary, 44
therapy-related, 44
alkylating agent related, 44
epipodophyllotoxin-related, 44
unclassifiable, 46
unclassified, 42
with 5q- syndrome, *35*, 43
with 5q deletion syndrome, *35*, 43
with chromosome 5 abnormality, 43
Myelofibrosis, *23*
acute, 23
as a result of myeloproliferative disease, 31
chronic idiopathic, *22*, 31
idiopathic, *23*, 31
malignant, 23
primary, *23*, 31
with acute myelodysplasia, 23
with acute panmyelosis, 23
with myeloid metaplasia, 31
Myelofibrosis-osteosclerosis, 31
Myelogenous leukemia, acute, *21*
Myelogenous leukemia, subacute, 46
Myeloid
leukemia, acute, *22*, *41*
leukemia, acute, with multilineage dysplasia, *42*
leukemia, chronic, *22*, *34*
leukemia, chronic, juvenile, 26
metaplasia, megakaryocytic, 31
sarcoma, 22
metaplasia
NOS, 31
agnogenic, *23*, *28*, 31, *32*
idiopathic, 31
with myelofibrosis, 31
with myelosclerosis, 31
tumor, extramedullary, 22
Myeloma
NOS, 3
IgA, 3
IgD, 3
IgE, 3
IgG, 3
light chain only, *4*
mono-ostotic, 2
multiple, *1*, 3, *4*, *5*, *45*
plasma cell, 3
solitary, *1*, 2
Myelomatosis, 3

Myelomonocytic leukemia
chronic
NOS, 25
juvenile, 26
in transformation [obs], 25
juvenile, 26
subacute, 25
syndrome, juvenile, 26
Myelomonocytic syndrome, chronic, 25
Myeloproliferative
disease, *30*
atypical chronic, *33*
chronic, 30
myelofibrosis as a result of, 31
disorder, *30*
disorder, chronic, 30
Myelosclerosis
acute, NOS, 23
chronic granulocytic-megakaryocytic, 31
malignant [obs], 23
megakaryocytic, 31
with myeloid metaplasia, 31

Neoplasia, immunoproliferative, 17
Neoplasm, immunoproliferative, 17
Neutrophilic leukemia, chronic, 33
Niemann-Pick disease , *10*
NK-cell
leukemia, aggressive, 27
leukemia-lymphoma, aggressive, 27
type large granular lymphocyte leukemia, aggressive, 27
Non-Hodgkin lymphoma, *45*
Nonlipid reticuloendotheliosis [obs], 12
Nonregenerative anemia, 38

Osler's disease, 28
Osler-Vaquez disease, 28
Osseous plasmacytoma, 2
Osteosclerosis-myelofibrosis, 31
Ovarian cancer, *45*

P. vera, 28
Pancytopenia , 47
Panmyelosis, acute, NOS, 23
Panmyelosis, acute, with myelofibrosis, 23
Paracortical interdigitating cell sarcoma, 15
PCL, 4
Pigmentosa, urticaria , 6, *9*
Plasma cell
dyscrasia, transient , *1*
leukemia, *1,* 4
myeloma, 3
sarcoma, 3
tumor, 2
Plasmacytic leukemia, 4
Plasmacytic tumor, *17*
Plasmacytic-lymphocytic lymphoma, *18*
Plasmacytoid lymphocytic lymphoma, well-differentiated, *18*
Plasmacytoid small lymphocytic lymphoma, *18*
Plasmacytoma
NOS, 2
extramedullary (not occurring in bone), *1, 4,* 5
localized, 2
medullary, 3
of bone, 2
of bone, isolated, *1,* 2
osseous, 2
soft tissue, 5
solitary, 2, *4*
PMF, 31
POEMS syndrome , *1*
Poly-ostotic Langerhans cell histiocytosis , 12
Polycythemia
NOS , *28*
primary, 28
proliferative, 28
relative , 29
rubra vera, 28, *45*
secondary , *28*, 29
spent phase, 28
splenomegalic, 28
vera, *22*, 28
Polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy (POEMS) syndrome , *1*
Preleukemia, 46
Preleukemic syndrome, 46
Primary
amyloidosis , *1*
erythremia, 28
macroglobulinemia, 18
myelofibrosis, *23,* 31
polycythemia, 28
refractory anemia, 38
sideroblastic anemia, 39
thrombocythemia, 32
Progressive
disseminated Langerhans cell histiocytosis, 12
histiocytosis, acute differentiated, 12
histiocytosis X, acute, 12
Proliferative polycythemia, 28
Pure cutaneous histiocytosis , *12*
Pure sideroblastic anemia, 39
Pyridoxine-responsive (hypochromic) anemia , *36*

RA, 38
RAEB, 40
RAEB I, 40
RAEB II, 40
RAEB-T, 41
RAEBIT, 41
RAEB(t), 41
RARS, 39

RAS, 39
RCMD, 42
Refractory
anemia, *35*, *36*, 38
primary, 38
with excess blasts, *35*, 40
with excess blasts in transformation [obs], *35*, 41
with hemochromatosis, 39
with ringed sideroblasts, *35*, 39
with sideroblasts, 39
without sideroblasts, 38
cytopenia with multilineage dysplasia, *35*, 42
dysmyelopoietic anemia, 46
Relative polycythemia , 29
Reticuloendotheliosis, 12
acute infantile, 12
leukemic, 24
nonlipid [obs], 12
systemic aleukemic, 12
Reticulohistiocytoma , 12
Reticulosis, acute, of infancy, 12
Reticulosis, histiocytic medullary [obs], 10
Reticulum cell sarcoma, 15, 16
dendritic, 16
interdigitating, 15
Ringed sideroblasts, refractory anemia with, *35*, 39
Rosai-Dorfman disease , *10*
Rubra vera, polycythemia, 28, *45*

Sarcoma
dendritic cell
NOS, 15
follicular, *15*, 16
interdigitating, 15, *16*
FDC, 16
granulocytic, 21
histiocytic, 13
interdigitating cell, 15, 16
interdigitating cell, paracortical, 15
Langerhans cell, 14
mast cell, 7
monocytic, 13
myeloid, 22
of follicular dendritic cells, 16
reticulum cell, 16
dendritic, 16
interdigitating, 15
true histiocytic, 13

Secondary
acute myeloid leukemia due to prior chemotherapy, 21
MDS, 44
myelodysplastic syndrome, 44
polycythemia , *28*, 29
sideroblastic anemia , *36*
Sex-linked hypochromic sideroblastic anemia , *36*
Sezary syndrome, *17*
Self-healing histiocytosis , *12*
Sideroachrestic anemia , *36*
Sideroblastic anemia, 39
acquired idiopathic, *36*, 39
acquired primary, *36*
chronic inflammatory , *36*
congenital , *36*
drug induced , *36*
hereditary , *36*
hypochromic with iron loading , *36*
hypochromic pyridoxine-responsive , *36*
primary, 39
pure, 39
secondary , *36*
secondary to disease , *36*
sex-linked hypochromic , *36*
vitamin B6-responsive , *36*
Sideroblasts
refractory anemia with, *35*, 39
refractory anemia without, 38
ringed, refractory anemia with, *35*, 39
Sinus histiocytosis with massive lymphadenopathy , *10*, 12
SM-AHNMD, 8
Small intestinal disease, immunoproliferative, *1, 17*, *19*, 20
Small lymphocytic lymphoma, plasmacytoid, *18*
SMCD, 8
Smoldering leukemia, 46
Soft tissue plasmacytoma, 5
Solitary
eosinophilic granuloma , 12
mastocytoma of skin , 6, *9*
myeloma, *1,* 2
plasmacytoma, 2, *4*
Spent-phase polycythemia, 28
Splenomegalic polycythemia, 28
Subacute myelogenous leukemia, 46
Subacute myelomonocytic leukemia, 25
Syndrome
5q-, myelodysplastic syndrome with, *35*, 43
5q deletion, myelodysplastic syndrome with, *35*, 43
chronic eosinophilic, 34
chronic myelomonocytic, 25
Hashimoto-Pritzker , *12*
hemophagocytic, infection-associated , 12
hypereoosinophilic, *22*, 34
hypereosinophilic, idiopathic, 34
juvenile myelomonocytic leukemia, 26
myelodysplastic *see Myelodysplastic syndrome*
myelodysplastic, unclassified, 42
POEMS , *1*
preleukemic, 46
Sezary, *17*

Systemic
histiocytosis, 10
mastocytosis, 8, *9*
aggressive, 8
indolent , 8
with associated clonal, hematological non-mast-cell lineage disease, 8
tissue mast cell disease, 8

T-cell large granular lymphocytic leukemia , *27*
T-cell lymphoma, *17*
Telangiectasia macularis eruptiva perstans , *9*
Testicular cancer, *45*
Therapy-related
acute myeloid leukemia, 21
alkylating agent-related, 21
epipodophyllotoxin-related, 21
myelodysplastic syndrome, 44
alkylating agent related, 44
epipodophyllotoxin-related, 44
Thrombocythemia
essential, 32
essential hemorrhagic, 32
idiopathic, 32
idiopathic hemorrhagic, 32
primary, 32
spent phase, 32
vera, 32
Thrombocytopenia , 32
Thrombocytosis, essential, 32
Topoisomerase II inhibitor related acute myeloid leukemia, 21
Transient plasma cell dyscrasias , *1*
True histiocytic lymphoma, 12, 13
True histiocytic sarcoma, 13
Tumor
dendritic cell, follicular, 16
dendritic cell, interdigitating , 15
extramedullary myeloid, 22
mast cell, NOS , 6
mast cell, malignant, 7
IDC , 15
plasma cell, 2
plasmacytic, *17*
Type II histiocytosis , 12

Unclassifiable myelodysplastic syndrome, 46
Unclassified myelodysplastic syndrome, 42
Undetermined significance, monoclonal gammopathy of , *1*
Unifocal Langerhans cell granulomatosis , 12
Unifocal Langerhans cell histiocytosis , 12
Unspecified aplastic anemia , *36*
Urticaria pigmentosa , 6, *9*

Vaquez-Osler disease, 28
Vera, polycythemia, *22*, 28
Vitamin B6-responsive sideroblastic anemia , *36*

Waldenstrom macroglobulinemia, *17*, 18
WM, 18

Xanthogranuloma , 12

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