FLORIDA CANCER DATA SYSTEM

MARCH 2004 MONTHLY MEMO

WHAT'S NEW:

The Following newsletters and reports are currently available from the FCDS website:

- THE JANUARY/FEBRUARY 2004 MONTHLY MEMO
- DATA ACQUISITION MANUAL ERRATA (2/24/2004)
- DATA ACQUISITION MANUAL REVISED SECTION II pgs. 51-96
- COC CONVERSION ERRATA
- NAACCR 2004 Implementation Guidelines: Collaborative Staging and Benign/Borderline Intracranial and CNS Tumors
- FCDS TELECONFERENCE SERIES: NEW REPORTING REQUIREMENTS FOR 2004

Abstracting, Coding, & Staging Leukemia

SEER Training Website: http://training.seer.cancer.gov/ss_module08_lymph_leuk/leuk_unit01_sec01_intro.html

INTRODUCTION

Leukemias are cancers of the blood-forming tissues. White blood cells may be produced in excessive amounts and are unable to work properly which weakens the immune system.

The blood is made up of fluid called plasma and three types of cells and each type has special functions. White blood cells (also called WBCs or leukocytes) help the body fight infections and other diseases. Red blood cells (also called RBCs or erythrocytes) carry oxygen from the lungs to the body's tissues and take carbon dioxide from the tissues back to the lungs. The red blood cells give blood its color. Platelets (also called thrombocytes) help form blood clots that control bleeding.

Blood cells are formed in the bone marrow, the soft, spongy center of bones. New (immature) blood cells are called blasts. Some blasts stay in the marrow to mature. Some travel to other parts of the body to mature.

Normally, blood cells are produced in an orderly, controlled way, as the body needs them. This process helps keep us healthy. When leukemia develops, the body produces large numbers of abnormal blood cells. In most types of leukemia, the abnormal cells are white blood cells. The leukemia cells usually look different from normal blood cells, and they do not function properly.

Each year, leukemia is diagnosed in about 29,000 adults and 2,000 children in the United States.

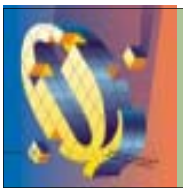
(Continued on page 3)

On the Web:

- **NAACCR Narrative Newsletter, Winter 2004 Edition** http://www.naaccr.org/filesystem/pdf/NAACCRNewsletterWinter2004.pdf
- **Cure: Cancer Updates, Research & Education** http://www.curetoday.com/
- **HIPAA Legislation** https://fcds.med.miami.edu/inc/links.shtml#priv

NATIONAL HEALTH INFORMATION PRIVACY AND SECURITY WEEK, APRIL 11-17, 2004

Mark your calendars for *National Health Information Privacy and Security Week, April 11 through April 17, 2004.* The week is designed to raise awareness among healthcare professionals, their employers, and the public of the importance of protecting the privacy, confidentiality, and security of personal health information. During the week, AHIMA and its members will work to educate and inform these groups of their rights and responsibilities related to the use and disclosure of personal health information.



FCDS Q & A:

LEUKEMIA

Seer Inquiry System: http://www.seer.cancer.gov/cgi-bin/seerinquiry/index.pl

References
SEER Program Code Man, 3rd Ed ;pgs 102, 104 Brief

Question
Grade, Differentiation--Lymphoma/Leukemia: What code is used to represent this field for a lymph node biopsy that reveals "well differentiated lymphocytic lymphoma" and a bone marrow biopsy that reveals "chronic lymphocytic leukemia/well differentiated lymphocytic lymphoma"?

Answer
Code the Grade, Differentiation field to 1 [Grade 1] for both of these cases because there is no mention of T-cell, B-cell, null cell, or NK cell involvement. Both cases have a pathologic description of well differentiated, not the descriptors "high grade," "low grade," or "intermediate grade" which must be ignored when coding grade for lymphomas.

For lymphomas, you cannot code the descriptions "high grade," "low grade," and "intermediate grade" in the Grade, Differentiation field because these terms refer to categories in the Working Formulation and not to histologic grade. However, you can code terms such as "well differentiated", "moderately differentiated" and "poorly differentiated" for lymphoma histologies.

References
Agents Not Listed in SEER Bk 8 ;pgs 4 (May 2002) Brief

Question
First Course of Cancer-Directed Therapy--All Sites: How do we code retinoic acid?

Answer
The code for retinoic acid depends upon the primary site and histology of the tumor. Code retinoic acid (also called Vitamin A, tretinoin, ATRA, all-transretinoic acid or Vesanoid) in the Immunotherapy field as 01 [Immuno administered as first course therapy] for acute promyelocytic leukemia. This drug is given to patients as an alternative to chemotherapy.

For all other sites/histologies, code retinoic acid in the Other Cancer-Directed Therapy Field. Use code 2 [Other experimental cancer-directed therapy] or 3 [Double-blind clinical trial, code not yet broken] if the drug is given as part of a protocol. If the drug isn't being given as part of a protocol or you don't know whether it is part of a protocol, use code 1 [Other cancer-directed therapy].

References
ICD-O-3 ;pgs 102, 147, 156... Brief

Question
Reportability/Behavior Code--Bone Marrow: Is T-cell large granular lymphocytic leukemia SEER reportable? Pages 102, 147, 156, 160-162 and 167 of the ICD-O-3 list it as 9831/1, but on page 17 this is listed as 9831/3.

Answer
T-cell large granular lymphocytic leukemia [9831] is a very indolent form of leukemia. It was assigned a behavior code of 1 by the editors of ICD-O-3 (as noted on pages 102, 147, 156 160-162, and 167 of the ICD-O-3 manual). The table on page 17 is the World Health Organization list of hematopoietic and lymphoid tumors. WHO recognizes TCLGLL as a malignancy. The disease is infrequently symptomatic enough to be diagnosed, but when it is called TCLGLL or one of the other terms included under histology 9831/1, it should be reported to SEER as a malignancy with a behavior code of /3.

(Continued from page 1)

In both men and women, leukemia incidence is highest among whites and lowest among Chinese, Japanese, and Koreans. The incidence in men is about 50% higher than in women for all racial/ethnic groups except Vietnamese, among whom the male rates are only slightly higher. Ethnic differences in the incidence rates are small in the youngest adult age group (30-54 years), but become more evident in each of the older age groups. It is found that childhood leukemia rates are highest among Filipinos, followed by white Hispanics, non-Hispanic whites and blacks.

ANATOMY

INTRODUCTION TO BLOOD

Blood is one of the connective tissues. As a connective tissue, it consists of cells and cell fragments (formed elements) suspended in an intercellular matrix (plasma). Blood is the only liquid tissue in the body that measures about 5 liters in the adult human and accounts for 8 percent of the body weight.

The body consists of metabolically active cells that need a continuous supply of nutrients and oxygen. Metabolic waste products need to be removed from the cells to maintain a stable cellular environment. Blood is the primary transport medium that is responsible for meeting these cellular demands.

The activities of the blood may be categorized as transportation, regulation, and protection. These functional categories overlap and interact as the blood carries out its role in providing suitable conditions for cellular functions.

The transport functions include:

- carrying oxygen and nutrients to the cells.
- transporting carbon dioxide and nitrogenous wastes from the tissues to the lungs and kidneys where these wastes can be removed from the body.
- Carrying hormones from the endocrine glands to the target tissues.

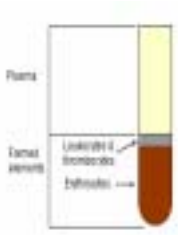
The regulation functions include:

- Helping regulate body temperature by removing heat from active areas, such as skeletal muscles, and transporting it to other regions or to the skin where it can be dissipated.
- Playing a significant role in fluid and electrolyte balance because the salts and plasma proteins contribute to the osmotic pressure.
- Functioning in pH regulation through the action of buffers in the blood.

The protection functions include:

- Preventing fluid loss through hemorrhage when blood vessels are damaged due to its clotting mechanisms.
- Helping (phagocytic white-blood cells) to protect the body against microorganisms that cause disease by engulfing and destroying the agent.
- Protecting (antibodies in the plasma) protect against disease by their reactions with offending agents.

COMPOSITION OF THE BLOOD



When a sample of blood is spun in a centrifuge, the cells and cell fragments are separated from the liquid intercellular matrix. Because the formed elements are heavier than the liquid matrix, they are packed in the bottom of the tube by the centrifugal force. The light yellow colored liquid on the top is the plasma, which accounts for about 55 percent of the blood volume and red blood cells is called the hematocrit, or packed cell volume (PCV). The white blood cells and platelets form a thin white layer, called the "buffy coat," between plasma and red blood cells.

Plasma

The watery fluid portion of blood (90 percent water) in which the corpuscular elements are suspended. It transports nutrients as well as wastes throughout the body. Various compounds, including proteins, electrolytes, carbohydrates, minerals, and fats, are dissolved in it.

Formed Elements

The formed elements are cells and cell fragments suspended in the plasma. The three classes of formed elements are the erythrocytes (red blood cells), leukocytes (white blood cells), and the thrombocytes (platelets).

Erythrocytes (red blood cells)

Erythrocytes, or red blood cells, are the most numerous of the formed elements. Erythrocytes are tiny biconcave disks, thin in the middle and thicker around the periphery. The shape provides a combination of flexibility for moving through tiny capillaries with a maximum surface area for the diffusion of gases. The primary function of erythrocytes is to transport oxygen and, to a lesser extent, carbon dioxide.

Leukocytes (white blood cells)

Leukocytes or white blood cells are generally larger than erythrocytes, but they are fewer in number. Even though they are considered to be blood cells, leukocytes do most of their work in the tissues. They use the blood as a transport medium. Some are phagocytic, others produce antibodies, some secrete histamine and heparin, and others neutralize histamine. Leukocytes are able to move through the capillary walls into the tissue spaces, a process called diapedesis. In the tissue spaces they provide a defense against organisms that cause disease and either promote or inhibit inflammatory responses.

(Continued on page 4)

(Continued from page 3)

There are two main groups of leukocytes in the blood. The cells that develop granules in the cytoplasm are called granulocytes and those that do not have granules are called agranulocytes. Neutrophils, eosinophils, and basophils are granulocytes. Monocytes and lymphocytes are agranulocytes.

Neutrophils, the most numerous leukocytes, are phagocytic and have light-colored granules. Eosinophils have granules and help counteract the effects of histamine. Basophils secrete histamine and heparin and have blue granules. In the tissues, they are called mast cells. Lymphocytes are agranulocytes that have a special role in immune processes. Some attack bacteria directly; others produce antibodies.

Thrombocytes (platelets)

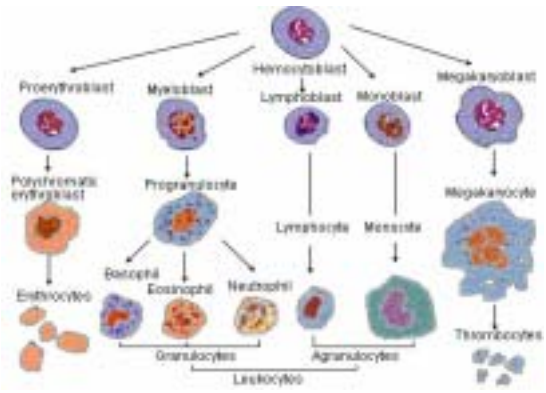
Thrombocytes, or platelets, are not complete cells, but are small fragments of very large cells called megakaryocytes. Megakaryocytes develop from hemocytoblasts in the red bone marrow. Thrombocytes become sticky and clump together to form platelet plugs that close breaks and tears in blood vessels. They also initiate the formation of blood clots.

BLOOD CELL LINEAGE

The production of formed elements, or blood cells, is called hemopoiesis. Before birth, hemopoiesis occurs primarily in the liver and spleen, but some cells develop in the thymus, lymph nodes, and red bone marrow. After birth, most production is limited to red bone marrow in specific regions, but some white blood cells are produced in lymphoid tissue.

All types of formed elements develop from a single cell type -- stem cell (pleuripotential cells or hemocytoblasts). Seven different cell lines, each controlled by a specific growth factor, develop from the hemocytoblast. When a stem cell divides, one of the "daughters" remains a stem cell and the other becomes a precursor cell, either a lymphoid cell or a myeloid cell. These cells continue to mature into various blood cells.

A leukemia can develop at any point in cell differentiation. The illustration below shows the development of the formed elements of the blood.



ABSTRACTING, CODING, & STAGING

Leukemia and other Blood Diseases

Including lymphocytic and myeloid leukemias, myelodysplastic syndromes (refractory anemias), polycythemia vera, essential thrombocythemia, myelofibrosis, agnogenic myeloid metaplasia, and Waldenstrom macroglobulinemia.

RELATED ADJECTIVES AND EQUIVALENT TERMS	
Leukemia = leuko-	Chronic (C) a severe stage of chronic leukemia is "blast crisis" Acute (A) Lymphocytic (L) = lymphoblastic, lyphatic lymphoid Myelogenous (M) = non-lymphocytic (NL), myelocytic (M), myeloblastic (M), granulocytic (G), monoblastic (M)
	Examples
	Chronic lymphocytic leukemia (CLL) Acute myelogenous leukemia (AML) = Acute granulocytic leukemia (AGL)

RELATED ADJECTIVES AND EQUIVALENT TERMS	
Myelo- = bone marrow	Examples Myelofibrosis= fibrosis of the bone marrow Myelodysplasia= abnormal production of cells in the bone marrow Myeloproliferative= excessive proliferation (reproduction) of cells in the bone marrow
Key Words	-blast an early or immature form of the cell -cyte a mature or later form of the cell phenotype cell line (myeloid or lymphoid, T-cell or B-cell

ICD-O Codes -- Primary Site	
C42.1	Bone marrow

All leukemias, myeloproliferative and lymphoproliferative disorders and myelodysplastic syndromes are coded to C42.1, bone marrow, because blood cells are generated in the bone marrow.

(Continued on page 5)

(Continued from page 4)

MORPHOLOGY AND GRADE

MAJOR CATEGORIES OF LEUKEMIAS

If the diagnostic term in the pathology report is not in the list below, be sure to consult your ICD-O manual.

The major categories of leukemias are grouped by cell type--malignancies of lymphocytes and granulocytes or monocytes (also called non-lymphocytic leukemias). Within each category, the behavior of the leukemia is subdivided into acute, subacute, and aleukemia, chronic, and not otherwise specified.

Virtually any blood component can become malignant:

Cell	Leukemia	Acute	Subacute	Chronic	Aleukemia	NOS*
Lymphocyte	Lymphocytic leukemia	98353	98003	98233	98203	98203
Monocyte	Monocytic leukemia	98913	98603	98603	98603	98603
Stem cell (primitive white cell)	Stem cell leukemia	98013				
Granulocyte (neutrophil)	Myelocytic/ myelogenous leukemia	98613	98603	98633	98603	98603
Granulocyte (eosinophil	Eosinophilic leukemia			99643		99643
Granulocyte (basophil)	Basophilic leukemia	98703				98703
Megakaryo-cyte	Megakaryo-cytic leukemia	99103				
Erythroblast	Erythro-leukemia					98403
Plasma cell	Plasmacytic leukemia					97333
T-cell	Adult T-cell leukemia, HTLV-positive					98273
Unspecified	Leukemia, NOS	98013	98003	98003	98003	98003

*NOS— not otherwise specified

(Continued on page 6)

(Continued from page 5)

The following leukemia morphologies were added to ICD-O-2 effective with Jan. 1, 1998 diagnoses:

(*) new term(s) leukemia morphologies were added to ICD-O-2 effective with Jan. 1 1998 diagnoses:

ICD-O-2 Code	Term
9821/3	Acute lymphoblastic leukemia, L1 type, NOS(*) Acute lymphocytic leukemia, L1 type (*) Acute lymphoid leukemia, L1 type (*) Acute lymphatic leukemia, L1 type (*) Lymphablastic leukemia, L1 type (*) FAB L1 (*)
9826/3	FAB L3 (*)
9828/3	Acute lymphoblastic leukemia, L2 type, NOS Acute lymphocytic leukemia, L2 type Acute
9840/3	FAB M6 (*)
9861/3	Acute myeloid leukemia, NOS (*) Acute myeloblastic leukemia, NOS (*) Acute granulocytic leukemia, NOS (*) Acute myelogenous leukemia, NOS (*) Acute myelocytic leukemia, NOS (*)
9866/3	FAB M3 (*)
9867/3	Acute myelomonocytic leukemia, NOS(*) FAB M4 (*)
9871/3	Acute melomonocytic leukemia with eosinophils FAB M4E
9872/3	Acute myloid leukemia, minimal differentiation Acute myeloblastic leukemia, minimal differentiation Acute granulocytic leukemia, minimal differentiation Acute myelogenous leukemia, minimal differentiation Acute myelocytic leukemia, minimal differentiation FAB M0
9873/3	Acute myeloid leukemia, without maturation Acute myeloblastic leukemia, without maturation Acute granulocytic leukemia, without maturation Acute myelocytic leukemia, without maturation FAB M1
9874/4	Acute myeloid leukemia, with maturation Acute myeloblastic leukemia, with maturation Acute granulocytic leukemia, with maturation Acute myelogenous leukemia, with maturation Acute myelocytic leukemia, with maturation FAB M2
9891	FAB M5 (*) FAB M5A (*) FAB M5B (*)
9910/3	Megakaryoblastic leukemia,. NOS (C42.1) FAB M7

ACUTE LYMPHOCYTIC LEUKEMIA

ACUTE LYMPHOCYTIC LEUKEMIA

(FRENCH-AMERICAN-BRITISH CLASSIFICATION)

STEM CELL	L1 Lymphoblastic leukemia, child type (98353- - acute) lymphoblastic, small
LYMPHOID-COMMITTED CELL	L2 Lymphoblastic leukemia, adult type (lymphoblastic, small-large)
LYMPHOBLASTIC	L3 Lymphoma - like leukemia (Burkitt)

Lymphocytic leukemias can be subdivided by type of cell surface antigens: 95% are B-cell and 5% are T-cell type.

ACUTE MYELOGENOUS (NON-LYMPHOBLASTIC) LEUKEMIA

(FAB CLASSIFICATION)

Lymphocytic leukemias can be subdivided by type of cell surface antigens: 95% are B-cell and 5% are T-cell type.

(Continued on page 7)

(Continued from page 6)

ACUTE MYELOGENOUS (NON-LYMPHOBLASTIC) LEUKEMIA (CONT'D) (FAB CLASSIFICATION)

Lymphocytic leukemias can be subdivided by type of cell surface antigens: 95% are B-cell and 5% are T-cell type.

M0 Undifferentiated leukemia (98013) stem cells predominate or cell type unidentified

M1 Myeloblastic leukemia without maturation (98723- -acute)
immature white blood cells predominate

M2 Myeloblastic leukemia with maturation (98723- -acute) with partial differentiation

M3 Promyelocytic leukemia (98663- -acute)
Promyelocytes predominate subdivided M3a without eosinophilia
M3b with eosinophilia (98603)

M4 Combination myeloblastic - monoblastic leukemia (98723- - acute; 98683- -chronic) each component constitutes greater than 20% of the blasts in the bone marrow subdivided
M4 acute myelomonocytic leukemia

M4EO acute myelomonocytic leukemia with eosinophilia

M5 Monoblastic leukemia (98603- -monocytic, NOS; 98913- -acute; 98603- -subacute; 98603- -chronic; 98603 aleukemic)
monoblasts predominate
subdivided M5a acute monocytic leukemia without
differentiation (98913- -monoblastic)
M5b acute monocytic leukemia with differentiation (promonocytic)

M6 Erythroleukemia (98403) immature red and white cells predominate

M7 Megakayrocytic leukemia (99103- -acute)

OTHER LYMPHOID MALIGNANCIES

9732/3 Multiple myeloma (malignancy of plasma cell/B-lymphocyte)

Definition	Single area of invasion and destruction of bone by proliferating plasma cells
ICD-9 code	203.0
Synonyms	Kahler disease Myelomatosis Plasma cell myeloma

OTHER LYMPHOID MALIGNANCIES (CONT'D)

9731/3 Solitary plasmacytoma (malignancy of plasma cell/B-lymphocyte)

Definition	Single areas of invasion and desruction of bone by proliferating plasma cells
ICD-9 code	238.6
Synonyms	Solitary myeloma Plasma cell tumor

9733/3 Plasma cell leukemia (malignancy of plasma cell/B-lymphocyte)

Definition	presence of abnormal level of plasma cells in circulating blood
ICD-9 code	203.1
Synonyms	Plasmacytic leukemia

9765/1 Monoclonal gammopathy of unknown significance (MGUS) (not considered a malignancy in ICD-0-3)

Definition	unexplained production of a single gamma globulin (rather than all types)
ICD-9 code	273.1

9761/3 Waldenstrom's macroglobulinemia

Definition	rare malignancy involving excess B-lymphocytes that secrete immunoglobulins (code to C42.0); similar to myeloma but without bone damage resulting in circulatory problems because blood is too thick to flow properly
ICD-9 code	273.3
Synonyms	Waldenstrom syndrome Primary macroglobulinemia

CHRONIC MYELOPROLIFERATIVE DISORDER

(In ICD-O-3, behavior will change from /1 to /3 and these cases will be reportable)

9950/3 Polycythemia vera (PV, P.vera)

Definition	overproduction of erythrocytes/red blood cells
ICD-9 code	238.4
Synonyms	Polycythemia rubra vera Proliferative polycythemia Chronic erythremia (ICD-9 207.1) Primary polycythemia Splenomegalic polycythemia Vasquez-Osler disease; Osler-Vasquez disease

(Continued on page 9)

EDUCATION AND TRAINING

March-April 2004

FCDS 2004 EDUCATIONAL TELEPHONE CONFERENCE SERIES

COLLABORATIVE STAGING PART I

Date: March 24, 2004
Time: 2PM - 4PM
Dial In Number: 888-476-3762
Participant code: 359957

COLLABORATIVE STAGING PART II

Date: April 14, 2004
Time: 2PM-4PM
Dial In Number: 888-422-7137
Participant code: 175525

NCRA 30TH ANNUAL EDUCATIONAL CONFERENCE

"On The Trail to New Horizons: Celebrating 30 years of Pioneering the Way for Cancer Data Research"

Date: April 20-23, 2004
Location: Portland, OR

Register before March 19th to $ave! The Conference will offer approximately 19 CE's towards your CTR credential!

For further information about the NCRA Annual Educational Conference, please visit the NCRA website at http://www.ncra-usa.org/index.html

NCRA PRE CONFERENCE WORKSHOPS

NCRA Short Course, 2004
Central Cancer Registries:
Design, Management, and Use

Monday, April 19th, 1 p.m. - 5:00 p.m.
Continuing, Tuesday, April 20th 8:00 a.m. - 5:00 p.m.

(Important Note: This workshop requires a minimum registration in order to be offered. NCRA will notify all registrants when the minimum has been met. Otherwise, NCRA reserves the right to cancel this workshop with a full refund by March 30th if the minimum has not been met. Registrants are encouraged to plan accordingly.)

More Survey Savvy:
Creating Best Practices and Implementing Them.
Brought to you by the Commission on Cancer.

Monday, April 19th, 1 p.m. - 5:00 p.m.
Tuesday, April 20th 8:00 a.m. - 5:00 p.m.

(Registration $150 per person includes all workshop materials and Saturday continental breakfast, box lunch)

Registration is limited to 150 people.

(Important Note: This workshop requires a minimum registration in order to be offered. NCRA will notify all registrants when the minimum has been met. Otherwise, NCRA reserves the right to cancel this workshop with a full refund by March 30th if the minimum has not been met. Registrants are encouraged to plan accordingly.)

Upcoming Training, Workshops, & Seminars 2004

NAACCR ANNUAL CONFERENCE
"New Frontiers in Cancer Surveillance"

Date:
June 8-9, 2004

Location:
Salt Lake City, Utah

For further information about the NAACCR Annual Conference visit the NAACCR website at
http://www.naaccr.org

FLORIDA CANCER DATA SYSTEM ANNUAL MEETING

Date:
July 27-28, 2004

Location:
Embassy Suites Hotel
USF/Busch Gardens
Tampa, FL

FLORIDA CANCER REGISTRARS ASSOCIATION ANNUAL MEETING

Date:
July 29-30, 2004

Location:
Embassy Suites Hotel
USF/Busch Gardens
Tampa, FL

(Continued from page 7)

CHRONIC MYELOPROLIFERATIVE DISORDER (CONT'D)

9960/3 Chronic myeloproliferative disease, NOS

Definition	overproduction of one or more type of blood cell
ICD-9 code	238.7
Synonyms	Chronic myeloproliferative disorder

9961/3 Myelosclerosis with myeloid metaplasia

Definition	condition where bone marrow is initially overactive but then develops fibrosis or scar tissue; term used when myelofibrosis/myelosclerosis is the first disease identified
ICD-9 code	238.7
Synonyms	Myelofibrosis as a result of myeloproliferative disease Megakaryocytic myelosclerosis Myelofibrosis (idiopathic) with myeloid metaplasia (AMM) Agnogenic myeloid metaplasia (term commonly used in US; not in ICD-0-3) Primary myelofibrosis (PMF) See also acute myelofibrosis (9931/3)

9962/3 Essential thrombocythemia

Definition	overproduction of platelets/thrombocytes resulting in circulatory problems
ICD-9 code	238.7
Synonyms	Essential thrombocythemia Essential hemorrhagic thrombocythemia Idiopathic hemorrhagic thrombocythemia Primary thrombocythernia

9863/3 Chronic myelogenous leukemia

MYELODYSPLASTIC SYNDROMES

9989/3 MYELODYSPLASTIC SYNDROMES (MDS)

Definition	disruption of production of blood cells--white, red, platelets; development of poor quality blood cells resulting in low levels of mature, functional cells; <30% blasts in marrow
ICD-9 code	238.7 unless indicated otherwise below
Synonyms	Pre-leukemia; preleukemic disorders Smoldering leukemia; oligoblastic leukemia; subacute myeloid leukemia de novo myelodysplastic syndrome - develops with no known cause Secondary myelodysplastic syndrome - develops following chemotherapy or radiation therapy for another disease; has poorer prognosis than de novo MDS Dysmyelopoietic syndrome; hemopoietic dysplasia

(Continued on page 10)

(Continued from page 9)

FAB CLASSIFICATION OF REFRACTORY ANEMIAS

9980/3 Refractory anemia, NOS (20-30% of patients)

Definition	Presence of megaloblastoid erythroid hyperplasia in marrow and macrocytic anemia with reticulocytopenia in blood
ICD-9 code	284.9
Synonyms	Refractory anemia with multilineage dysplasia Refractory anemia without sideroblast RA

9982/3 Refractory anemia with sideroblasts (2-5%)

Definition	same as RA, but with at least 15% of marrow red cell precursors being ringed sideroblasts (characteristic ring-shaped deposits of iron in red blood cell)
ICD-9 code	285.0
Synonym	Refractory anemia with ringed sideroblasts RARS

9983/3 Refractory anemia with excess blasts (40%)

Definition	5-20% of marrow is myeloid blasts, 1-5% in circulating blood
ICD-9 code	285.0
Synonyms	RAEB

9984/3 Refractory anemia with excess blasts in transformation (25%)

Definition	20-30% of marrow cells are blasts and > 5% blasts in circulating blood; most progress to acute leukemia
ICD-9 code	285.0
Synonyms	RAEB-T

9945/3 Chronic myelomonocytic leukemia (15-20%)

Definition	Increased monocytes in blood; marrow may or may not contain increased number of blasts
ICD-9 code	205.1
Synonyms	CMML CMMoL

9945/3 Chronic myelomonocytic leukemia in transformation

Definition	increased biologic activity indicating that chronic leukemia is evolving into acute leukemia
ICD-9 code	205.1

FAB CLASSIFICATION OF REFRACTORY ANEMIAS (CONT'D)

9895/3 Acute myeloid leukemia arising from myelodysplastic syndrome

Definition	a new ICD-0-3 category identifying cases of acute myeloid leukemia that had a preexisting myelodysplastic syndrome
ICD-9 code	205.0
Synonyms	Secondary acute myeloid leukemia

Non-reportable conditions

Pancytopenia	low levels of all types of blood cells
Aplastic anemia	complete failure of production of all types of blood cells, as from high doses of chemotherapy or radiotherapy
Fanconi Anemia	Rare familial disorder with symptoms of severe aplastic anemia, hypoplasia of bone marrow and other symptoms
Myelofibrosis, NOS	Filling of the bone marrow with fibrous tissue (SNOMED code)
Secondary...	For example, secondary polycythemia, secondary myelofibrosis: conditions resulting from another disease (post-polycythemic myeloid metaplasia, postpolycythernic splenomegaly

EXTENT OF DISEASE EVALUATION

COMMON METASTATIC SITES

Spread	Primary Site/Mets
Lymphatic Spread	Rare; leukemia is considered systemic at the time of diagnosis.
Hematogenous Spread	Leukemia may invade many visceral sites, including skin, breast, eye, spleen, lymph nodes

(Continued on page 11)

(Continued from page 10)

DEFINITIONS

Key words/involvement: terms which indicate possible involvement by tumor. Common terms are provided, but the list is not all-inclusive.

Other words/no involvement: other terms seen in reports which indicate an abnormality but do not indicate a neoplastic process. Common terms are provided, but the list is not all-inclusive.

Key information: information to look for in the report of the study. Key information helps define the extent of disease.

DIAGNOSTIC STUDIES (1)

DIAGNOSTIC STUDIES--PHYSICAL EXAM
Key information: lymph node enlargement, secondary masses; abdominal tenderness, organomegaly (hepatosplenomegaly, hepatomegaly, splenomegaly), bruises, petechiae

DIAGNOSTIC STUDIES--LABORATORY STUDIES

CBC (Complete Blood Count) with Platelets
Histochemistry of Bone Marrow pathologic examination of bone marrow biopsy specimen using special staining techniques to determine subclassification of the blood disease. Histochemistry of the bone marrow is a laboratory study performed in addition to the standard histologic analysis of the specimen.

Liver Function Tests (LFT)
Renal Function
Chemistry Screening Panel
Cerebrospinal Fluid Examination

DIAGNOSTIC STUDIES--IMAGING

Key information: involvement of visceral organs, spread to adjacent tissues or organs; lymph node enlargement; sites of distant organs or lymph nodes involved

Chest X-ray
Imaging, Bone
Imaging, Liver/spleen

DIAGNOSTIC STUDIES (2)

DIAGNOSTIC STUDIES--TUMOR MARKERS
Key information: to differentiate types of leukemia and help determine prognosis

DNA Studies

OTHER TUMOR MARKERS

Ferritin--elevated levels are present in lymphoproliferative diseases; nonspecific to leukemia; also a marker for Hodgkin lymphoma or head and neck cancer

TDT (Terminal Deoxynucleotidal Transferase)--differentiates acute lymphocytic leukemia from acute non-lymphocytic leukemia; also useful in differentiating lymphomas; TDT levels are absent in patients in remission

6-2 Microglobulin--Also called Beta 2-M. Elevated levels are present in lymphoproliferative disorders; non-specific to chronic lymphocytic leukemia.

Philadelphia Chromosome (Ph1)--presence of abnormal chromosome in bone marrow confirms diagnosis of chronic myelogenous leukemia; absence of Ph1 chromosome does not rule out CML

DIAGNOSTIC STUDIES--ENDOSCOPIES
Endoscopic examinations are not useful for determining the extent of leukemia and blood diseases.

DIAGNOSTIC STUDIES--OPERATIVE REPORT
Operative reports are not useful for determining the extent of leukemia and blood diseases.

DIAGNOSTIC STUDIES (3)

DIAGNOSTIC STUDIES--PATHOLOGY

Key information: cell type, percent of cells in blast phase

CYTOLOGY REPORTS: pleural effusion (thoracentesis) or ascites (paracentesis)

BONE MARROW BIOPSY Also called bone marrow aspiration. Aspiration of bone marrow cells to determine involvement by tumor. This procedure is used to diagnose leukemia. Bilateral bone marrow biopsies and aspirations should be done for accurate assessment of blast content.

Key words
Aleukemic--the presence of malignant cells in the bone marrow with a normal or leukopenic count in the circulating blood

Auer rods--abnormal cytoplasmic granules which distinguish AML from ALL.

Blasts--cells in an immature stage of cellular development; also called stem cells

Blast crisis--for chronic leukemia, increased blast cells inthe peripheral blood accompanied by progressive splenomegaly and fever

-cytosis -- abnormal increase in the number of cells; leukocytosis (increased white cells); thrombocytosis (increased thrombocytes or platelets)

Leukostasis--leukemic involvement of the lungs or central nervous system characterized by thickening or "sludging" of blood vessels due to the increased number of cells in the blood

-penia -- abnormal decrease in the number of cells; leukocytopenia (decreased white cells); thrombocytopenia (decreased platelets); neutropenia (decreased granulocytes); pancytopenia (decrease in all types of cells)

Richter's syndrome--large cell lymphoma which develops after treatment for CLL treatment-induced acute leukemia--a second, acute leukemia that develops after chemotherapy for chronic leukemia

OTHER BLOOD DISEASE

EXTENT OF DISEASE EVALUATION--OTHER BLOOD DISEASES

MYELOFIBROSIS

CBC and platelets
Bone marrow biopsy and aspiration (see above). The aspiration looks at the cells;

(Continued on page 12)

(Continued from page 11)

the biopsy looks at the structure of the bone marrow to detect the scar tissue or fibrosis.

MYELODYSPLASTIC SYNDROMES

CBC and platelets
Bone marrow aspiration and biopsy
Chromosomal analysis of bone marrow

ESSENTIAL THROMBOCYTHEMIA AND POLYCYTHEMIA VERA

CBC and platelets
Bone marrow aspiration and biopsy

WALDENSTROM'S MACROGLOBULINEMIA

CBC
Bone marrow aspriation and biopsy
Immunoelectrophoresis

STAGING

RELATIONSHIP TO TNM STAGING: Anatomic staging is not applicable to the leukemias.

RELATIONSHIP TO SUMMARY STAGING: All leukemias are considered disseminated at time of diagnosis.

PROGNOSTIC AND THERAPEUTIC STAGINGS

CHRONIC LYMPHOCYTIC LEUKEMIA (RAI STAGING)

Stage	Description
Stage 0	Lymphocytosis greater than 5,000 cell/mm and greater than 40% of cells in the bone marrow
Stage 1	Lymphocytosis with large lymph nodes
Stage 2	Lymphocytosis with enlargement of spleen and/or liver
Stage 3	Lymphocytosis and marrow replacement resulting in anemia
Stage 4	Lymphocytosis and low platelet due to marrow replacement

INTERNATIONAL WORKSHOP ON CLL CLINICAL STAGING

Clinical stage A

No anemia or thrombocytopenia, < 3 areas of lymphadenopathy
(Rai Stage 0, I, II)

Clinical stage B

No anemia or thrombocytopenia, > or = 3 areas of lymphadenopathy (Rai Stage I - II)

Clinical stage C

Anemia and/or thrombocytopenia (Rai Stage III - IV)

There are no staging systems for other blood diseases. The acute leukemias and myelodysplastic syndromes are described and classified histologically by the French-American-British (FAB) classification.

An International Prognostic Scoring System has been developed for MDS, based on the bone marrow blast percentage, number of peripheral blood cytopenias, and cytogenetic subgroup, which stratifies cases into four risk groups. In addition, other prognostic classification systems for MDS include the FAB system (described above), the Bournemouth score, the Sanz score, and the Lille score.

ABSTRACTING KEYS

All users of this module are encouraged to download and read the very useful material contained in the "ABSTRACTING AND CODING GUIDE FOR THE HEMATOPOIETIC DISEASES" publication available on the SEER Training website at *http://training.seer.cancer.gov/ss_module08_lymph_leuk/pdfs/Abst%20Coding%20Guide%20Heme%20Diseases.pdf*.

B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma: special coding circumstances

This 7 word phrase is the World Health Organization's formal name for a single disease entity that has different presentations. The diseased cells look the same under the microscope; only the background tissue is different–in blood or bone marrow it has traditionally been called chronic lymphocytic leukemia (9823/3), in lymph nodes the name has traditionally been small lymphocytic lymphoma (9670/3). The two names had to be split to be compatible with ICD-10. This diagnosis is listed at 9823/3 and cross-referenced to 9670/3 in ICD-O-3.

To choose the correct code:

If this complete diagnostic term is diagnosed in blood or marrow, code as leukemia (9823/3, CLL).
If this complete diagnostic term is diagnosed in tissue or lymph nodes, code as lymphoma (9670/3, SLL).
If this complete diagnostic term is diagnosed in both blood or bone marrow and any other tissue or lymph nodes, code as a lymphoma (9670/3).

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

(Continued on page 13)

(Continued from page 12)

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. 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.

"Coding to the higher morphology code"

The general ICD-O-3 guideline to use the numerically higher morphology code if the diagnosis of a single tumor includes two modifying adjectives with different code numbers (Rule K) does not apply to the hematopoietic diseases (M-9590-9989) in general. For the hematopoietic diseases, code to the more specific morphology, if that can be determined, which may not be the numerically higher code number. For example, if the facility pathology report states "acute myelogenous leukemia" (M-9861/3) and a consultant reports the same tissue to be "acute myeloid leukemia, AML1(CBF-alpha/ETO)" (M-9896/3), code the case to M-9896/3 because it is more specific, not because it is a numerically higher code. The primary term for M-9861/3 includes the term NOS (not otherwise specified) and many synonyms; thus it can be considered a non-specific diagnosis. On the other hand, the primary term for M-9896/3 does not include the term NOS and may therefore be considered more specific. When in doubt which code to use, consult a medical advisor or pathologist.

Complete remission (CR) is the key to determining prognosis for leukemia. A complete remission is defined as:

Bone marrow containing less than 5% blasts
Normalization of erythrocyte, granulocyte, and platelet counts
Resolution of organomegaly
Return to normal performance status

A patient is considered free of clinical evidence of disease when first remission is accomplished. Treatment of any relapses should be considered as subsequent therapy. The date of the relapse should be considered the date of first recurrence.

Tumor size is not relevant for leukemia; the "Size of Tumor" field should be coded 999.

Leukemia can involve organs and body tissues, not just the blood and bone marrow. If the patient has a known leukemia and develops what appears to be a second primary, the histology of the new site should be carefully checked to assure that it is indeed a new primary and not a manifestation of the leukemia in a solid organ.

Radiation therapy to the brain or central nervous system should be coded for leukemia cases, regardless of whether metastases are known at the time of treatment.

If a patient had CNS radiation and surgery, record the appropriate code for the sequence of treatment.

Understanding Cytogenetic and Molecular Terminology
The WHO classification of leukemias includes cytogenetic qualifiers for disease terms because of a decision in 1994 that it was important to define and name leukemias by the chromosomal changes in cancerous cells. To understand the diagnostic terminology, remember that normal human cells have 23 pairs of autosomal chromosomes, numbered 1 to 23, and two sex chromosomes, labeled "X" or "Y." Many things can happen to these chromosomes as they split and come together, including breakage, transference, and complete loss of portions of the chromosome. In a cytogenetic description, t means a translocation or a reciprocal exchange of genetic material between two chromosomes. The letter q represents the long arm of chromosome, and the letter p represents the short arm of the chromosome.

A diagnostic term such as "acute myelogenous leukemia t(15;17)(q22;q11-12)" would be read as a translocation of material from the long arm of chromosome 15 in region 22 which has been swapped with the material on the long arm of chromosome 17 in the region between 11 and 12. Each cytogenetic abnormality is unique. When coding leukemias, be careful not to confuse the various translocations.

A disease may have both cytogenetic and molecular markers. Both are listed with the code in ICD-O-3. Again, when coding leukemias, be careful not to confuse the various molecular markers.

Abbreviation Full name of marker

ABL Abelson murine leukemia oncogene
BCR breakpoint cluster region
CBF core binding factor
ETO eight twenty one (8;21)
MYH11 myosin, heavy polypeptide 11
PML promyelocytic leukemia
RARA retinoic acid receptor, alpha

SEER PROGRAM CODING GUIDELINES for Acute Leukemia

1. Code the FAB category, if there is one in the diagnosis. There may be times when the term "FAB" is not part of the statement. The "FAB" is implied if the leukemia is described as "L" or "M" with a number, such as L2 or M5. Use the guidelines for ambiguous terminology in the SEER Program Code Manual, third edition, if the diagnosis uses a term such as "consistent with" or "probable."

2. All of the following are equivalent terms:
Granulocytic
Myeloblastic
Myelocytic

(Continued on page 14)

(Continued from page 13)

Myelogenous
Myeloid
Non-lymphocytic
The following are equivalent terms:
Lymphoblastic
Lymphocytic
Lymphoid
Lymphatic

3. If the diagnostic statement lists a specific acute leukemia cell type, code that term.

4. If the diagnostic statement lists more than one FAB classification, for example "M1 or M2" or "M4 vs M5a" without further comment, revert to the NOS code, because even the pathologist cannot make a decision on the subtype.

5. The terms "maturation" and "differentiation" are synonymous.

6. All leukemias are coded to primary site C42.1, Bone marrow, except myeloid sarcoma and leukemic reticuloendotheliosis. These are coded to the site specified in the pathology report.

7. Acute non-lymphocytic leukemia is a term that is not in ICD-O-3. It is a non-specific term similar to "non-small cell" carcinoma. In the absence of a more specific pathologic description, code acute non-lymphocytic leukemia as 9861/3, Acute Myelogenous Leukemia, NOS.

Likelihood of progression to acute leukemia

Refractory anemia rare
Refractory anemia with sideroblasts rare
Refractory anemia with excess blasts 40%
Refractory anemia with excess blasts in transformation 60-75%
Chronic myelomonocytic leukemia 30%
Polycythemia vera < 10%
Essential thrombocythemia < 10%

TREATMENT

SURGERY AND RADIATION THERAPY

LEUKEMIA

Within the categories of leukemia (ALL, AML, CLL, CML), treatment is similar, but treatment strategies do vary by category. Successful treatment consists of ablation of leukemia in the bone marrow and treatment or prevention of systemic disease, including infiltration of visceral sites.

CLL usually has a protracted, indolent course, and therefore is treated conservatively (treatment is deferred until the patient becomes symptomatic). Stage 0 cases are usually not treated.

Splenectomy or splenic radiation is a consideration for splenomegaly due to infiltration by CLL.

Key words

Prophylaxis (prophylactic)--administration of treatment in the absence of clinical symptoms to prevent the worsening of the condition; for example prophylactic CNS radiation to prevent CNS involvement, antibiotic prophylaxis to prevent infection
Sanctuary-site disease--leukemia cells present in visceral organs, such as brain, central nervous system, or testes

SURGERY

Cancer-directed surgery is generally not performed for the treatment of leukemia. Surgery for leukemia is included with surgical treatment for "all other sites."

RADIATION THERAPY

Radiation therapy to the central nervous system is prophylactic treatment to prevent or delay the occurrence of metastases from some varieties of leukemia. CNS radiation is not indicated for AML, but is important for ALL.

Involvement of single lymph node chains with CLL can be treated with radiation therapy.

CHEMO AND OTHER THERAPIES

DRUGS COMMONLY USED FOR TREATING THE LEUKEMIAS

Chemotherapy for Acute Myeloid Leukemia
Daunorubicin and cytarabine
Daunorubicin, cytarabine and thioguanine
Cytarabine and idarubicin
Cytarabine and mitoxantrone
Cytarabine or methotrexate intrathecally for known CNS infiltration

Other drugs
Etoposide
Amsacrine
Mitoxantrone
Idarubicin (new)
Homoharringtonine

Chemotherapy for Acute Lymphocytic Leukemia

High-dose methotrexate or intrathecal methotrexate for CNS prophylaxis
The average length of treatment is between one and one-half and three years for ALL
Vincristine, prednisone, doxorubicin or daunorubicin
Vincristine, prednisone, asparaginase, daunorubicin
Vincristine, prednisone, asparaginase, doxorubicin
Vincristine, prednisone, doxorubicin, intrathecal methotrexate
Methotrexate, vincristine, asapraginase, dexamethasone
Vincristine, prednisone, anthracycline, with or without asparaginase

Maintenance chemotherapy with combinations of the following:

6 mercaptopurine
Cyclophosphamide
Cyatarabine
Prednisone
Vincristine
Carmustine
Daunorubicin
Doxorubicin
Teniposide

Chemotherapy for Chronic Lymphocytic Leukemia

Chlorambucil (oral) with or without prednisone
Cytoxan (oral) with or without prednisone
Fludarabine (under clinical evaluation for refractory CLL)

Combinations
COP (cytoxan, vincristine, prednisone)

Key words

Induction--administering chemotherapy to obtain remission
Nadir--the lowest number of stem cells in the bone marrow; the point in chemotherapy administration when the patient is at greatest risk of developing infection; also called myelosuppression
Remission--the clinical disappearance of leukemia, characterized by a normal peripheral blood count, less than 5%

(Continued on page 15)

(Continued from page 14)
blasts in
the bone marrow, no clinical signs or symptoms of leukemia,
and no evidence of CNS or visceral infiltration by leukemia
Consolidation--administering high doses of
chemotherapy for a
short term to maintain remission
Maintenance--administering relatively low doses of chemotherapy
for a long term to maintain remission

Hormones

Prednisone in combination with alkylating agents

Biological Response Modifiers (under clinical evaluation)

Bone Marrow Transplantation (autogenous = from the patient; allogenic = from another patient); also called ABMT
Interleukin-2 (under clinical evaluation)
Colony stimulating factors (CSF) (under clinical evaluation)

SUPPORTIVE CARE

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; cardiovascular 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 thin the blood 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.

Leukemia patients are susceptible to infections (herpes zoster, pneumocystis carinii, and candidal albicans) because they have few bone marrow reserves to fight infection.

Transfusion with fresh frozen plasma, cryoprecipitate and/or platelets
Isolation techniques
White blood cell transfusions

Systemic antibiotics--to prevent infection when patient is myelosuppressed

Semi-synthetic Penicillin

Types of penicillins are: Amdinocillin, Amoxicillin, Ampicillin, Azlocillin, Bacampicillin, Carbenicillin, Cloxacillin, Cyclacillin, Dicloxacillin, Methicillin, Mexlocillin, Nafcillin, Oxacillin, Penicillin G, Penicillin V, Piperacillin, Ticarcillin
Brand names of penicillins are: Amcill, Amoxil, Augmentin, Azlin, Bactocill, Beepen-VK, Bicillin L-A, Cloxapen, Coactin, Crysticillin, Duracillin, Dycill, Dynapen, Geocillin, Geopen, Ledercillin, Mezlin, Nafcil, Nallpen, NaMPICIL, Omnipen, Pathocil, Penapar, Pentids, Pen Vee K, Permapen, Pfizerpen, Pipracil, Polycillin, Polymox, Principen, Prostaphlin, Robicillin, Spectrobid, Staphcillin, Sumox, Supen, Tegopen, Ticar, Timentin, Totacillin, Trimox, Unipen, Utimox, V-Cillin K, Wycillin, Wymox

Aminoglycosides
Types of aminoglycosides are: Amikacin, Gentamincin, Kanamycin, Neomycin, Netilmicin, Streptomycin, Tobramycin
Brand names of aminoglycosides are: Amikin, Apogen, Garamycin, Kantrex, Klebcil, Nebcin, Neo-IM, Netromycin

Cephalosporin
Type of cephalosporin are: Cefaclor, Cefadroxil, Cefamandole, Cefazolin, Cefonicid, Cefoperazone, Ceforanide, Cefotaxime, Cefotetan, Cefoxitin, Ceftazidime, Ceftizoxime, Ceftriaxone, Cefurozime, Cephalexin, Cephalothin, Cephapirin, Cephradine, Moxalactam.
Brand names of cephalosporin are: Ancef, Anspor, Ceclor, Cefadyl, Cefobid, Cefotan, Duricef, Fortaz, Kefletl, Keflex, Keflin, Neutral, Kefuroz, Kefzol, Mandol, Mefozin, Monocid, Moxam, Rocefphin, Seffin Neutral, Tazidime, Ultracef, Velosef, Zinacef

Immunoglobulin (intravenous Ig)

OTHER BLOOD DISEASES

MYELOFIBROSIS
Patients with early myelofibrosis are treated symptomatically to keep the patient comfortable and minimize side-effects of more aggressive treatment. Patients with severe anemia require blood transfusions every one to three months. Hydroxyurea may be given to reduce painful

(Continued on page 16)

(Continued from page 15)

splenomegaly, or the spleen may be surgically removed. Erythropoietin or androgens may stimulate red blood cell production. Sometimes chemotherapy and radiation therapy are used. A small proportion of myelofibrosis patients may develop acute myelogenous leukemia, which is treated with chemotherapy (see above).

MYELODYSPLASTIC SYNDROMES

The primary treatment of the myelodysplastic syndromes is supportive therapy with blood transfusions of specific types of cells (rather than whole blood) and antibiotic treatment of any infections that may develop. Patients with fewer than 5% blasts have a long chronic phase during which they receive many red blood cell transfusions. Patients with more aggressive disease will progress more quickly to acute leukemia which is treated with chemotherapy (see above). Bone marrow transplantation is a treatment option for younger patients (less than age 55). Certain categories of cases are treatable with either low-dose or intensive (high dose) chemotherapy. Hematopoietic growth factors are an immunotherapy under clinical investigation. Other types of supportive care include vitamin therapy, steroids, and immunosuppressive agents.

POLYCYTHEMIA VERA

Because this is a disease of over-production of red cells, it is treated by blood removal (phlebotomy or venisection). If cells other than red cells are also increased, radioactive phosphorus or chemotherapy may be used to suppress the abnormal and excessive cell production.

WALDENSTROM'S MACROGLOBULINEMIA

Supportive care is the standard treatment. Stable disease may need no treatment. Plasmapheresis (removal of the IgM antibody from the blood) may be effective for some patients. Other patients may respond to chemotherapy with chlorambucil.

IMPLICATION OF HIPAA FOR CENTRAL CANCER REGISTRIES

The following letter was mailed to all Tumor Registrars and Administrators of all Hospitals, Ambulatory Surgical and Radiation Therapy Centers, and Path Labs in Florida. The documents mentioned in the letter may be found at these websites:

http://naaccr.org/Training/files/AcademicLetterInterpretingHIPAA.pdf

http://naaccr.org/Training/files/LegalLetterInterpretingHIPAA.pdf

http://naaccr.org/Training/files/FAQsRegardingHIPAAandCancerRegistries.pdf

Dear Tumor Registrar/Administrator:

The Health Insurance Portability and Accountability act of 1996 (HIPAA) became law April 14, 2001. While most organizations have two full years – until April 14, 2003 – to comply, questions regarding how this new law impacts cancer reporting have arisen.

The North American Association of Central Cancer Registries (NAACCR) has provided materials that address these questions. As you will see, HIPAA regulations only minimally impact current state cancer reporting procedures. Specifically,

> **HIPAA allows for the reporting of identifiable cancer data to public health entities. Because the Florida Cancer Data System falls under the definition of a public health entity, HIPAA allows your facility to continuing to report data to us in compliance with state law. Written informed consent from each cancer patient reported to public health entities is not required under HIPAA; rather hospitals must simply document that reporting has occurred.**

Enclosed please find a copy of a letter from the NAACCR legal counsel, an academic interpretation of HIPAA from Professor James G. Hodge, Jr., J.D., LL.M., of the Georgetown University Law Center, and a list of frequently asked questions and answers.

We hope this material is beneficial in your understanding the HIPAA requirements regarding cancer incidence reporting.

Sincerely,

Jill A. MacKinnon
Administrative Director, FCDS

Dr. Youjie Huang, DOH

DEADLINES AND REMINDERS

COMPLETENESS REPORT

11,163 new cases were added to the FCDS Masterfile in January 2004 and 13,057 were added in February 2004.

As of February 29, 36% of the 2003 Cancer Admissions has been reported to FCDS. 67% is expected.

REMINDER

75% of the 2003 Cancer Admissions are due by March 31, 2004.

NEW PROCEDURE FOR ACTIVE CTR'S APPLYING FOR THE FIRST TIME FOR AN FCDS ABSTRACTOR CODE

As of March 1st, 2004, any active CTR applying for the first time for an FCDS Abstractor Code will no longer be required to submit 25 cases. They must complete and submit the following documentation to FCDS:

a) A photocopy of their most current CTR Certificate indicating active certification from NCRA.
b) A completed FCDS Cancer Abstractor Code Request Form (can be downloaded from the FCDS Web site under Downloads: 2003 FCDS DAM).
c) A signed and completed copy of the CTR attestation (can be downloaded from the FCDS Web site under What's New).

All non CTR's applying for permission to submit cancer cases to FCDS must continue to submit 25 abstracts in hard copy form for review and approval to obtain an FCDS Cancer Abstractor Code.

ABSTRACTOR CODE RENEWAL DEADLINE

FCDS Cancer Abstractor Codes expire on June 30th, 2004. You must fill out a new form to continue submitting work to FCDS. Completed forms should be sent to your Field Coordinator during the month of June, 2004.

On July 1, during the database conversion work, (v.10 to v.10.1) we will take all the requests mailed in, and renew the expiration date to June 30, 2005.

The renewal form is located on the FCDS IDEA page of our website: http://fcds.med.miami.edu/inc/idea.shtml.

Please note: This is the 3 letter/number code (ex: 3GV) coded on the abstract. This is not the FCDS IDEA user id (ex: mrudolph). The Path and Radiation users don't need the 3 letter code.

ANNUAL MAIL FILE REVIEW

The Annual Mail File Review forms were mailed to all FCDS mail recipients on February 20, 2004. Please be sure to review the document. In an effort to efficiently correspond with you, please make any and all corrections to the information provided directly on the form itself and return it to FCDS by March 14, 2004. You may fax the form to 305-243-4871. Please feel free to contact FCDS anytime throughout the year to inform us of any changes.

RADIATION THERAPY CENTERS CANCER CASE IDENTIFICATION PROGRAM

Beginning with the 2003 patient encounters, Radiation Therapy Centers are responsible for identifying and reporting all of their cancer cases to the Florida Cancer Data System using the FCDS-IDEA Single Entry or the File Upload modules. The deadline to submit the cases is June 30, 2004. Please log on to the FCDS website fcds.med.miami.edu or call Betty Hallo at (305) 243-2627 for additional information.

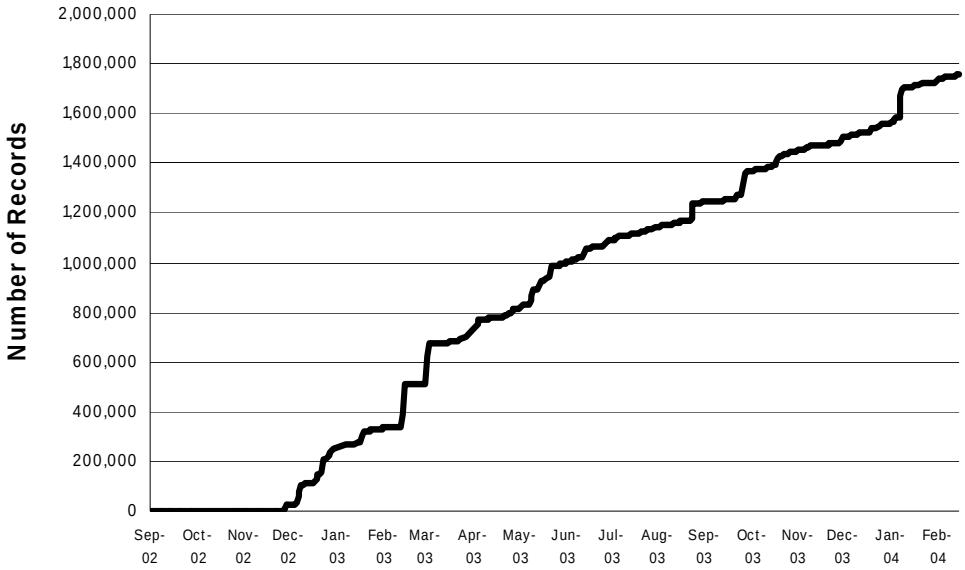
PATH LAB REPORTING

Every anatomic pathology laboratory that reads biopsy and surgical resection specimens collected from patient encounters, within the state of Florida MUST electronically submit the specified data for every malignant cancer case. Specimens read between July 1, 2003 and December 31, 2003 must be submitted to FCDS on or before June 30, 2004.

FLORIDA CANCER DATA SYSTEM

Path Reporting

Cumulative Path Data Received



FCDS Florida Cancer Data System
A Joint Project of the Sylvester Comprehensive Cancer Center and the Florida Department of Health

PROJECT DIRECTOR:
Lora Fleming, MD, PhD

ADMINISTRATIVE DIRECTOR:
Jill Mackinnon, CTR

EDITORIAL STAFF:
Mark Rudolph, M.S.
Mayra Alvarez, RHIT, CTR
Melissa K. Williams

CONTRIBUTORS:
Betty Fernandez
Megsys Herna, CTR
Stuart Herna, CTR

FCDS Florida Cancer Data System
A Joint Project of the Sylvester Comprehensive Cancer Center and the Florida Department of Health

P. O. BOX 016960 (D4-11)
MIAMI, FL 33101

662826