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)
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 [Immu 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 TCLG LL as a malignancy. The disease is infrequently symptomatic enough to be diagnosed, but when it is called TCLG LL 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.
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 (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 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.
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 (pluripotential 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.
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:

<table>
<thead>
<tr>
<th>Cell</th>
<th>Leukemia</th>
<th>Acute</th>
<th>Subacute</th>
<th>Chronic</th>
<th>Aleukemia</th>
<th>NOS*</th>
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<tbody>
<tr>
<td>Lymphocyte</td>
<td>Lymphocytic leukemia</td>
<td>98353</td>
<td>98003</td>
<td>98233</td>
<td>98203</td>
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<td>Monocyte</td>
<td>Monocytic leukemia</td>
<td>98913</td>
<td>98603</td>
<td>98603</td>
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<tr>
<td>Stem cell (primitive white cell)</td>
<td>Stem cell leukemia</td>
<td>98013</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Granulocyte (neutrophil)</td>
<td>Myelocytic/myelogenous leukemia</td>
<td>98613</td>
<td>98603</td>
<td>98633</td>
<td>98603</td>
<td>98603</td>
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<tr>
<td>Granulocyte (eosinophil)</td>
<td>Eosinophilic leukemia</td>
<td>99643</td>
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<tr>
<td>Granulocyte (basophil)</td>
<td>Basophilic leukemia</td>
<td>99703</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Megakaryocyte</td>
<td>Megakaryocytic leukemia</td>
<td>99403</td>
<td></td>
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<td></td>
</tr>
<tr>
<td>Erythroblast</td>
<td>Erythroblast leukemia</td>
<td></td>
<td></td>
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<td></td>
<td>98403</td>
</tr>
<tr>
<td>Plasma cell</td>
<td>Plasma cytic leukemia</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>97433</td>
</tr>
<tr>
<td>T-cell</td>
<td>Adult T-cell leukemia, HTLV-positive</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>98273</td>
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<tr>
<td>Unspecified</td>
<td>Leukemia, NOS</td>
<td>98013</td>
<td>98003</td>
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</tr>
</tbody>
</table>

*NOS— not otherwise specified
The following leukemia morphologies were added to ICD-O-2 effective with Jan. 1, 1998 diagnoses:

(Continued from page 5)

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

<table>
<thead>
<tr>
<th>ICD-O-2 Code</th>
<th>Term</th>
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</thead>
<tbody>
<tr>
<td>9821/3</td>
<td>Acute lymphoblastic leukemia, L1 type, NOS(*)</td>
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<td>Acute lymphoid leukemia, L1 type (*)</td>
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<tr>
<td></td>
<td>Acute lymphatic leukemia, L1 type (*)</td>
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<tr>
<td></td>
<td>Lymphoblastic leukemia, L1 type (*)</td>
</tr>
<tr>
<td>9826/3</td>
<td>FAB L1 (*)</td>
</tr>
<tr>
<td>9828/3</td>
<td>Acute lymphoblastic leukemia, L2 type, NOS</td>
</tr>
<tr>
<td></td>
<td>Acute lymphocytic leukemia, L2 type</td>
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<tr>
<td>9840/3</td>
<td>FAB M6 (*)</td>
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<tr>
<td>9861/3</td>
<td>Acute myeloid leukemia, NOS (*)</td>
</tr>
<tr>
<td></td>
<td>Acute myeloblastic leukemia, NOS (*)</td>
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<tr>
<td></td>
<td>Acute granulocytic leukemia, NOS (*)</td>
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<td>Acute myelogenous leukemia, NOS (*)</td>
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<td></td>
<td>Acute myelocytic leukemia, NOS (*)</td>
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<td>9866/3</td>
<td>FAB M3 (*)</td>
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<td>FAB M4 (*)</td>
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<td>9871/3</td>
<td>Acute monoblastic leukemia with eosinophils</td>
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<td></td>
<td>FAB M4E</td>
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<tr>
<td>9872/3</td>
<td>Acute myeloid leukemia, minimal differentiation</td>
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<td></td>
<td>Acute granulocytic leukemia, minimal differentiation</td>
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<td>Acute myelogenous leukemia, minimal differentiation</td>
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<td>Acute myelocytic leukemia, minimal differentiation</td>
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<tr>
<td></td>
<td>FAB M0</td>
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<tr>
<td>9873/3</td>
<td>Acute myeloid leukemia, without maturation</td>
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<tr>
<td></td>
<td>Acute myeloblastic leukemia, without maturation</td>
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<tr>
<td></td>
<td>Acute granulocytic leukemia, without maturation</td>
</tr>
<tr>
<td></td>
<td>Acute myelocytic leukemia, without maturation</td>
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<td>FAB M1</td>
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<th>ICD-O-2 Code</th>
<th>Term</th>
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<td>Acute myelocytic leukemia, with maturation</td>
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<td>FAB M5 (*)</td>
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<tr>
<td></td>
<td>FAB M5A (*)</td>
</tr>
<tr>
<td></td>
<td>FAB M5B (*)</td>
</tr>
<tr>
<td>9910/3</td>
<td>Megakaryoblastic leukemia, NOS (C42.1)</td>
</tr>
<tr>
<td></td>
<td>FAB M7</td>
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</tbody>
</table>

ACUTE LYMPHO CYTIC LEUKEMIA

(FRENCH-AMERICAN-BRITISH CLASSIFICATION)

<table>
<thead>
<tr>
<th>STEM CELL</th>
</tr>
</thead>
<tbody>
<tr>
<td>L1 Lymphoblastic leukemia, child type (98353-- acute)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>LYMPHOID-COMMITTED CELL</th>
</tr>
</thead>
<tbody>
<tr>
<td>L2 Lymphoblastic leukemia, adult type (lymphoblastic, small-large)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>LYMPHOBLASTIC</th>
</tr>
</thead>
<tbody>
<tr>
<td>L3 Lymphoma - like leukemia (Burkitt)</td>
</tr>
</tbody>
</table>

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

Abstracting, Coding, & Staging Leukemia (Cont'd)

OTHER LYMPHOID MALIGNANCIES (CONT'D)

9731/3 Solitary plasmacytoma (malignancy of plasma cell/ B-lymphocyte)
Definition Single areas of invasion and destruction 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
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 save! 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
### 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 thrombocytopenia**
- **Definition**: overproduction of platelets/thrombocytes resulting in circulatory problems
- **ICD-9 code**: 238.7
- **Synonyms**: Essential thrombocytopenia, Essential hemorrhagic thrombocytopenia, Idiopathic hemorrhagic thrombocytopenia, Primary thrombocytopenia

**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)
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 sideroblasts 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: CMMML, 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

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.
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)

DIAG NO STIC STUDIES--PHYSICAL EXAM

Key information: lymph node enlargement, secondary masses, abdominal tenderness, organomegaly (hepatosplenomegaly, hepatomegaly, splenomegaly), bruises, petechiae

DIAG NO STIC 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

DIAG NO STIC 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)

DIAG NO STIC 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 Deoxynucleotidyl 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

DIAG NO STIC STUDIES--ENDOSCOPY

Endoscopic examinations are not useful for determining the extent of leukemia and blood diseases.

DIAG NO STIC STUDIES--OPERATIVE REPORT

Operative reports are not useful for determining the extent of leukemia and blood diseases.

DIAGNOSTIC STUDIES (3)

DIAG NO STIC 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 in the 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; leukopenia (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)
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 aspiration 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)**

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

**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 1, 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-0-3.

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...
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 the consultant reports the same tissue to be “acute myeloid leukemia, AMML(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 number. 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 22, 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)
Abstracting, Coding, & Staging Leukemia (Cont’d)

(Continued from page 13)

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

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%
Polythemia vera
10%
Essential thrombocytopenia
< 10%

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 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.

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

Chemotherapy for Acute Lymphoblastic Leukemia

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
Amsa crine
Mitoxantrone
Idrubicin (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, asparaginase, dexamethasone
Vincristine, prednisone, ant-hracycline, with or without asparaginase

Maintenance chemotherapy with combinations of the following:

6 mercaptopurine
Cyclophosphamide
Cytarabine
Prednisone
Vincentine
Carmustine
Daunorubicin
Doxorubicin
Teniposide

Chemotherapy for Chronic Lymphocytic Leukemia

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

Combinations

COP (cytosan, 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)
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
Systemic antibiotics--to prevent infection when patient is myelosuppressed

Semi-synthetic Penicillin

- Types of penicillins are: Amoxicillin, Amoxicillin, Ampicillin, Azlocillin, Bacampicillin, Carbenicillin, Cloxacillin, Cylcacin, Dicloxacillin, Methicillin, Moxolocillin, Naflcillin, Oxaclillin, Penicillin G, Penicillin V, Piperacillin, Ticarcillin, Ticarcillin

Aminoglycosides

- Types of aminoglycosides are: Amikacin, Gentamicin, Kanamycin, Neomycin, Netilmicin, Streptomycin, Tobramycin
- Brand names of aminoglycosides are: Amikin, Apogen, Gramycin, Kantiex, Klebcin, Nebcin, Neo-IM, Netromycin

Cephalosporin
- Type of cephalosporin are: Cefadroxil, Cefalothin, Cefazolin, Cefonicid, Cefoperazone, Ceforanide, Cefotaxime, Cefotetan, Cefoxitin, Cefazidime, Ceftizoxime, Ceftriaxone, Cefurozime, Cephalexin, Cephalothin, Cepharidine, Moxalactam
- Brand names of cephalosporin are: Ancef, Anspor, Ce-lor, Cefadyl, Cefobid, Cefotan, Duricef, Fortaz, Keflert, Keflex, Keftin, Neutrad, Kefuroz, Ketzol, Mandol, Mefozin, Monocid, Moxacin, Rocelfast, Seffin, Neutral, Tazidime, Ultracerv, Velosef, Zinacef

Immunoglobulin (intra venous Ig)

OTHER BLOOD DISEASES

MYELO FIBRO SIS
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...
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/LegalLetterInterpretingHIPAA.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.