ICD-10-CM
Specialty Code Set Training
Pediatrics
2014
Module 1
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Clinical Examples Used in this Book

AAPC believes it is important in training and testing to reflect as accurate a coding setting as possible to students and examinees. All examples and case studies used in our study guides and exams are actual, redacted office visit and procedure notes donated by AAPC members.

To preserve the real world quality of these notes for educational purposes, we have not re-written or edited the notes to the stringent grammatical or stylistic standards found in the text of our products. Some minor changes have been made for clarity or to correct spelling errors originally in the notes, but essentially they are as one would find them in a coding setting.
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Neoplasms in Pediatric Patients

Children can get cancer just the same as an adult, but childhood cancers differ in some ways. They can occur suddenly, without early symptoms and have a high rate of cure. According to the Centers for Disease Control and Prevention (CDC), leukemia is the most commonly diagnosed cancer and leading cause of cancer death in children 0-19 years of age. Central nervous system cancer was second. In this section, we will discuss leukemia, brain cancer, neuroblastoma, and lymphoma.

Leukemia

As stated above, leukemia is the most common type of childhood cancer, and is a cancer of the white blood cells. Blood cells from in the bone marrow, and white blood cells help the body fight infection. In a healthy person, bone marrow produces white blood cells, red blood cells, and platelets. In a patient with leukemia, the bone marrow produces abnormal white blood cells that grow faster than normal cells that do not stop growing. The leukemia cells can crowd out the normal blood cells over time, which can lead to anemia, bleeding, and infections. Leukemia cells can also spread to the lymph nodes and other organs, which cause swelling and pain. Treatment options include chemotherapy, radiation therapy, stem cell transplants, and biological therapy with the goal being to achieve remission.

The codes for leukemia are located in categories C90-C95 in ICD-10-CM. They are broken down by type, temporal parameters, and remission status. Acute leukemia is a fast growing type, while chronic leukemia grows slowly. Children usually have one of the acute leukemias.

According to the National Cancer Institute remission is a decrease in or disappearance of signs and symptoms of cancer. In partial remission, some, but not all, signs and symptoms of cancer have disappeared. In complete remission, all signs and symptoms of cancer have disappeared, although cancer still may be in the body.

Acute Lymphoblastic Leukemia (ALL)

ALL is the most common type of cancer in children, including all leukemias. According to the National Cancer Institute, ALL accounts for 23 percent of cancer diagnoses in children younger than 15 years. It may also be referred to as acute lymphoid leukemia or acute lymphocytic leukemia. In a healthy child, bone marrow makes immature stem cells that become mature blood cells over time. A blood stem cell may become either a myeloid or lymphoid stem cell.

Lymphoid stem cells become lymphoblast cells and one of three types of lymphocytes:

- B lymphocytes—make antibodies to help fight infection
- T lymphocytes—help B lymphocytes make antibodies
- Natural killer cells—attack cancer cells and viruses

What occurs in ALL is that too many stem cells become lymphoblasts, B lymphocytes, or T lymphocytes, which are the leukemia cells. They do not work like normal lymphocytes and cannot fight infection very well. The number of leukemia cells also increases in the bone marrow and blood, so there is less room for healthy white blood cells, red blood cells, and platelets.
The codes for ALL in ICD-10-CM are as follows:

- C91.00 Acute lymphoblastic leukemia not having achieved remission
- C91.01 Acute lymphoblastic leukemia, in remission
- C91.02 Acute lymphoblastic leukemia, in relapse

**EXAMPLE**
Paula is brought in by her mother for a visit. She has acute myeloid leukemia, currently receiving chemotherapy treatments. She is eating better, but still has a poor appetite and is fatigued.

C91.00 Acute lymphoblastic leukemia, not having achieved remission

**Acute Myeloblastic Leukemia (AML)**
AML is the second most common form of leukemia in children. According to the National Cancer Institute, AML accounts for 20 percent of childhood leukemias. It may also be referred to as acute myeloid leukemia, acute myelogenous leukemia, acute granulocytic leukemia, or acute nonlymphocytic leukemia. A blood stem cell may become either a myeloid or lymphoid stem cell.

Myeloid stem cells become one of three types of white blood cells:

- Red blood cells—carry oxygen and other substances to all tissues of the body
- White blood cells—fight disease and infection
- Platelets—form blood clots to stop bleeding

In children with AML, there is an overproduction of immature white blood cells (myeloblasts). Just as in ALL, the abnormal leukemia cells build up and crowd out the normal cells. There are eight subtypes of AML. Most physicians use two main systems to classify them: the French, American, British (FAB) system and the World Health Organization (WHO) classification system. The FAB system classifies the subtypes based on how much the leukemic cells have matured and the type of blood cell the cancer developed from. Examples of FAB subtypes include M0 (myeloblastic) and M4 (myelomonocytic). The WHO system classifies the subtypes by broad groupings based on expected outcomes. Examples of WHO subtypes include AML with recurrent genetic abnormalities and therapy-related AML.

The codes for AML in ICD-10-CM are as follows:

- C92.00 Acute myeloblastic leukemia, not having achieved remission
- C92.01 Acute myeloblastic leukemia, in remission
- C92.02 Acute myeloblastic leukemia, in relapse
EXAMPLE
Jack presents for a check-up. He has acute myelogenous leukemia that is in remission. He is doing well and his bone marrow results indicate less than 4 percent blast cells and his blood cell counts remain within normal limits. He had chemotherapy as part of his treatment regimen.

- C92.01 Acute myeloblastic leukemia, in remission
- Z92.21 Personal history of antineoplastic chemotherapy

Juvenile Myelomonocytic Leukemia (JMML)
JMML is a rare form of leukemia usually affecting children under the age of 2 years. It may also be referred to as juvenile chronic myeloid leukemia or chronic myelomonocytic leukemia in infancy. It causes a more severe disruption in blood counts early and is not as responsive to treatment. Hematopoietic stem cell transplantation (SCT) is the only curative approach for this cancer.

The codes for JMML in ICD-10-CM are as follows:
- C93.30 Juvenile myelomonocytic leukemia, not having achieved remission
- C93.31 Juvenile myelomonocytic leukemia, in remission
- C93.32 Juvenile myelomonocytic leukemia, in relapse

EXAMPLE
Mindy is brought in to be seen for her JMML in remission three months after her first stem cell transplant. Unfortunately, her leukemia has relapsed. A second stem cell transplant is discussed.

- C93.32 Juvenile myelomonocytic leukemia, in relapse
- Z94.84 Stem cell transplant status

Central Nervous System Tumors
According to the American Cancer Society, approximately 24 percent of childhood cancers are central nervous system tumors. To better understand central nervous system tumors, it is important to understand the structure of the central nervous system (CNS). The CNS is made up of the brain and spinal cord. The main areas of the brain are the cerebrum, cerebellum, and brain stem. Most childhood tumors are located in the cerebellum and brain stem.
The cerebrum is the largest portion of the brain; it is split into two hemispheres with four lobes: the frontal lobe, the temporal lobe, the parietal lobe, and the occipital lobe.

The cerebellum is located at the back of the brain and helps coordinate movement. Tumors in this area of the brain can cause problems including difficulty with coordination and movements in the extremities, and changes in speech rhythm.

The brain stem is the lower part of the brain that connects the brain to the spinal cord and helps control breathing and the heartbeat. Most of the cranial nerves start in the brain stem. It is divided into three main parts: the midbrain, pons, and medulla oblongata. Tumors in this area of the brain can cause weakness, stiff muscles, and problems with sensation, facial movements, hearing, and swallowing.

The cranial nerves emerge directly out of the base of the brain. They perform various functions, such as bringing information to the brain from the sense organs and control muscles.

There are 12 pairs of cranial nerves:

| CN I—Olfactory | CN VII—Facial |
| CN II—Optic | CN VIII—Vestibulocochlear |
| CN III—Oculomotor | CN IX—Glossopharyngeal |
| CN IV—Trochlear | CN X—Vagus |
| CN V—Trigeminal | CN XI—Accessory |
| CN VI - Abducens | CN XII - Hypoglossal |

The most common cranial nerve tumors in childhood are optic gliomas, which are tumors of the optic nerve (CN II).
From an ICD-10-CM perspective, follow the guideline regarding code look up. When a histologic term is documented, that term should be referenced first, rather going straight to the Neoplasm Table. A specific code may be listed, or it may indicate the appropriate column in the Neoplasm Table that should be referenced. For example, when the term Astrocytoma is referenced in the Alphabetic Index, it lists multiple types and mostly sends the user to Neoplasm, malignant, by site. But, under the same term (astrocytoma) when the subterm subependymal is referenced, it lists code D43.2 as a default and also sends the user to Neoplasm, uncertain behavior, by site. Without proper look-up, an incorrect code may be reported.

In the Neoplasm Table, the following is seen for cerebellum, brain stem, and optic nerve neoplasms:

<table>
<thead>
<tr>
<th>Neoplasm-brain NEC</th>
<th>Malignant Primary</th>
<th>Malignant Secondary</th>
<th>Ca In situ</th>
<th>Benign</th>
<th>Uncertain Behavior</th>
<th>Unspecified Behavior</th>
</tr>
</thead>
<tbody>
<tr>
<td>cerebellum NOS</td>
<td>C71.6</td>
<td>C79.31</td>
<td>-----</td>
<td>D33.1</td>
<td>D43.1</td>
<td>D49.6</td>
</tr>
<tr>
<td>medulla oblongata</td>
<td>C71.7</td>
<td>C79.31</td>
<td>-----</td>
<td>D33.1</td>
<td>D43.1</td>
<td>D49.6</td>
</tr>
<tr>
<td>midbrain</td>
<td>C71.7</td>
<td>C79.31</td>
<td>-----</td>
<td>D33.1</td>
<td>D43.1</td>
<td>D49.6</td>
</tr>
<tr>
<td>pons</td>
<td>C71.7</td>
<td>C79.31</td>
<td>-----</td>
<td>D33.1</td>
<td>D43.1</td>
<td>D49.6</td>
</tr>
<tr>
<td>stem</td>
<td>C71.7</td>
<td>C79.31</td>
<td>-----</td>
<td>D33.1</td>
<td>D43.1</td>
<td>D49.6</td>
</tr>
<tr>
<td>nerve optic</td>
<td>C72.3-</td>
<td>C79.49</td>
<td>-----</td>
<td>D33.3</td>
<td>D43.3</td>
<td>D49.7</td>
</tr>
</tbody>
</table>

Following are the codes with descriptors:

- C71.6 Malignant neoplasm of cerebellum
- C71.7 Malignant neoplasm of brain stem
- C72.30 Malignant neoplasm of unspecified optic nerve
- C72.31 Malignant neoplasm of right optic nerve
- C72.32 Malignant neoplasm of left optic nerve
- C79.31 Secondary malignant neoplasm of brain
- C79.49 Secondary malignant neoplasm of other parts of nervous system
- D33.1 Benign neoplasm of brain, infratentorial
- D33.3 Benign neoplasm of cranial nerves
- D43.1 Neoplasm of uncertain behavior of brain, infratentorial
- D49.6 Neoplasm of unspecified behavior of brain
- D49.7 Neoplasm of unspecified behavior of endocrine glands and other parts of nervous system
EXAMPLE
A four-year-old girl is brought in by her mother for complaints of decreased vision and proptosis of her right eye. She also has been experiencing depth perception issues. A CT scan confirms an optic glioma.

C72.31 Malignant neoplasm of right optic nerve

EXAMPLE
A seven-year-old boy is seen for his medulloblastoma of the cerebellum. His parents present with him to discuss treatment options of radiation therapy and chemotherapy, and ventriculoperitoneal shunt placement.

C71.6 Malignant neoplasm of cerebellum

Bone Cancers
Bone cancer is an uncommon type of cancer as it begins in the bones and does not spread to the bones from another site. It can occur in any bone in the body, but most commonly are seen in the long bones of the body in the extremities. Some bone cancers primarily affect children, others affect mostly adults.

Osteosarcoma
According to the American Cancer Society, osteosarcoma (also called osteogenic sarcoma) is the most common type of bone cancer in children and adolescents. Bones are made up of four main kinds of cells:

- **Osteoblasts**—responsible for making new bone, rebuild existing bones when they break
- **Osteocyte**—star-shaped cells usually found in compact bone, and are old osteoblasts that have stopped making new bone
- **Lining cells**—flat bone cells that cover the outside surface of all bones and are formed from old osteoblasts that have stopped making new bone
- **Osteoclasts**—break down and reabsorb existing bone

Like osteoblasts in normal bone, osteoblasts in cancerous bone also produce bone matrix, except that it is not as strong. St. Jude Children's Research Hospital provides the following information on osteosarcoma:

- It is slightly more common in boys.
- It is most common between the ages of 10–20, and rarely diagnosed in children before age 5.
- It occurs most often in the long bones on either side of the knee and the upper arm, commonly found at the metaphysis.
- About 15–20 percent of the patients have metastatic disease at the time of diagnosis.
- Presence of metastasis impacts survival rates (localized osteosarcoma—75 percent, patients with metastasis—30 percent).
According to the guidelines (I.C.2) when a histologic term is documented, that term should be referenced first, rather than going immediately to the Neoplasm Table, to determine which column in the Neoplasm Table is appropriate. When the term *Osteosarcoma* is referenced in the Alphabetic Index, it sends the user to *Neoplasm, bone, malignant*.

**EXAMPLE**

15-year-old David is brought in with complaints of pain, swelling, and decreased motion in his left knee. He has been having trouble with it for a few months, but just told his parents about it as pain has begun to wake him up at night. An X-ray showed an abnormality and he was sent for bone scan, then biopsy that confirmed osteosarcoma of the distal femur.

*C40.22 Malignant neoplasm of long bones of left lower limb*

**Ewing Sarcoma**

Ewing sarcoma is the second most common malignant bone tumor in children. They occur most often in the early teenage years. It is named after Dr. James Ewing who noted a difference in the cells from osteosarcoma. They are most often located in the lower extremities, pelvis, chest wall, and upper extremities. As opposed to osteosarcoma that usually occurs at the ends of the long bones, Ewing sarcoma of bone mainly occurs in the middle of the long bones.

There are three main types that are considered the Ewing sarcoma family of tumors:

- **Ewing sarcoma of bone**: Ewing sarcoma that starts in the bone is the most common tumor of the Ewing family of tumors.
- **Extraosseous Ewing tumor (EOE)**: sarcoma that starts in soft tissues around bones, but look and act like Ewing sarcomas in the bones. They are also called extraskeletal Ewing sarcoma.
- **Peripheral primitive neuroectodermal tumor (PPNET)**: rare form that starts in the bone or soft tissue. PPNETs that start in the chest wall are called Askin tumors.

The codes selected will depend on the type of Ewing sarcoma. When the term *Tumor* with the subterm *Ewing's* is referenced in the Alphabetic Index, it sends the user to *Neoplasm, bone, malignant, by site*. But if the term *Tumor* with the subterm *Askin's* is referenced in the Alphabetic Index, it sends the user to *Neoplasm, connective tissue, malignant*. It is important for the provider to be specific as to the site of the sarcoma, and for the coder to be familiar with the terminology used for Ewing sarcoma.

**EXAMPLE**

After suffering a fracture with no preceding trauma, Michael was found to have Ewing sarcoma of the right femur which had spread to the inguinal lymph nodes. His parents are bringing him today to discuss treatment and obtain a referral to Oncology.

*C40.21 Malignant neoplasm of long bones of right lower limb*

*C77.4 Secondary and unspecified malignant neoplasm of inguinal and lower limb lymph nodes*
Lymphoma in Childhood

Lymphoma refers to cancer of the lymphatic system, which carries lymph. Lymph is a colorless, watery fluid that contains lymphocytes (white blood cells) and travels through the lymph system. The lymphatic system includes:

- **Lymph glands (also called nodes)** — filter lymph and store white blood cells; found throughout the body
- **The thymus** — located in the chest behind the sternum; lymphocytes grow and multiply in this organ
- **The spleen** — located on the left side of the abdomen near the stomach; makes lymphocytes, filters blood, stores blood cells, and destroys old blood cells
- **The tonsils and adenoids** — located in the back of the throat; makes lymphocytes
- **The bone marrow** — makes white blood cells, red blood cells, and platelets
- **The lymph vessels** — network of tubes that collect lymph from different parts of the body and return it to the bloodstream

Lymphomas are divided into two general categories: non-follicular, which include Hodgkin’s lymphoma and non-Hodgkin’s lymphoma; and follicular. Follicular lymphoma is rarely seen in the pediatric population, so it will not be covered.

An important note to remember is that other types of cancer may spread to lymph tissue such as the lymph nodes. If the cancer starts somewhere else and spreads to the lymph tissue, they are not lymphomas, but metastatic cancer of the primary site.

**Hodgkin’s Lymphoma (HL)**

Hodgkin’s lymphoma involves a type of cell known as a Reed-Sternberg cell, which is an abnormal lymphocyte. The Reed-Sternberg cells divide again and again, and don’t die when they should. They are much larger than normal cells. There are two types of Hodgkin’s lymphoma: classical and nodular lymphocyte-predominant.

Classical Hodgkin’s lymphoma is the typical type of Hodgkin’s lymphoma, accounting for 95 percent of all cases. It is divided into four subtypes that are based on how the cancer cells look microscopically:

- **Nodular sclerosis classical Hodgkin’s lymphoma** (C81.1-) — This subtype is the most common, accounting for about 60–80 percent of cases. It mainly occurs in children and tends to start in the neck or chest.
- **Mixed cellular classical Hodgkin’s lymphoma** (C81.2-) — This subtype is the second most common, accounting for 15–30 percent of cases. It most often occurs in the upper half of the body.
- **Lymphocyte-rich classical Hodgkin’s lymphoma** (C81.4-) — This subtype accounts for about 5 percent of the cases, usually occurring in the upper half of the body.
- **Lymphocyte-depleted classical Hodgkin’s lymphoma** (C81.3-) — This subtype accounts for less than 1 percent of cases and is seen mainly in older people.
Nodular lymphocyte predominant Hodgkin lymphoma (C81.0-) is rare and not normally seen in children. It is a slow-growing form that is diagnosed by the presence of a lymphocyte-predominant (LP) cell (sometimes called a popcorn cell due to its appearance) that is unique to this type of lymphoma.

The codes for Hodgkin’s lymphoma are located in category C81 and are broken down by type and affected lymph nodes. Examples include:

- C81.01 Nodular lymphocyte predominant Hodgkin lymphoma, lymph nodes of head, face, and neck
- C81.12 Nodular sclerosis classical Hodgkin lymphoma, intrathoracic lymph nodes
- C81.23 Mixed cellularity classical Hodgkin lymphoma, intra-abdominal lymph nodes
- C81.34 Lymphocyte-depleted classical Hodgkin lymphoma, lymph nodes of axilla and upper limb
- C8.45 Lymphocyte-rich classical Hodgkin lymphoma, lymph nodes of inguinal region and lower limb

There are also codes in each subcategory that contain a selection for multiple sites and unspecified code.

**EXAMPLE**

Charlotte is seen in the clinic. She has nodular sclerosis Hodgkin’s of the mediastinal lymph nodes.

- C81.12 Nodular sclerosis classical Hodgkin lymphoma, intrathoracic lymph nodes

**Non-Hodgkin’s Lymphoma**

Just as with Hodgkin’s lymphoma, non-Hodgkin’s lymphoma is classified by how the cells look microscopically. From a coding standpoint, the codes are broken down by type and lymph nodes affected. There are four major types of childhood non-Hodgkin’s lymphoma:

- **Diffuse large B-cell lymphoma** (DLBCL) (C83.3-) — The most common type of non-Hodgkin’s lymphoma in the U.S. (according to the American Cancer Society). It is an aggressive lymphoma with several subtypes, including anaplastic, CD30-positive, and plasmablastic, all located under the listed subcategory.

- **Lymphoblastic lymphoma** (C83.5-) — An aggressive lymphoma in which too many lymphoblasts are in the lymph nodes and thymus gland. More common in teens and young adults and more often affecting males than females.

- **Anaplastic large cell lymphoma** (ALCL) (C84.6-, C84.7-) — An aggressive lymphoma, usually the T-cell type, that express a marker called CD30. ALCL is divided into two groups, depending on the presence of anaplastic lymphoma kinase (ALK), an abnormal protein found on the cells’ surface. Subcategory C84.6 denotes ALK-positive, which has the abnormal protein, and subcategory C84.7 denotes ALK-negative, which do not have the abnormal protein.
• **B-cell non-Hodgkin’s lymphoma** (Burkitt and Burkitt-like) (C83.7)—An aggressive lymphoma that is broken into three main types: sporadic, seen throughout the world; endemic, which occurs in Africa, and immunodeficiency-related, which is most often seen in AIDS patients.

**EXAMPLE**

Patrick is brought in for evaluation for fever, night sweats, and weight loss. A biopsy confirms diffuse large B-cell lymphoma of the spleen.

C83.37 Diffuse large B-cell lymphoma, spleen

**Sequencing of Neoplasms**

Sequencing of neoplasms is dependent upon treatment. According to the guidelines (I.C.2.a, I.C.2.b, and I.C.2.c) the order of the codes is dependent upon treatment delivery.

If treatment is directed at the malignancy, the malignancy is the first-listed code.

**EXAMPLE**

Jack presents for evaluation for his cancer of the brain stem to discuss possible CyberKnife treatment.

C71.7 Malignant neoplasm of brain stem

If the patient presents for administration of chemotherapy, radiation therapy, or immunotherapy, the appropriate code from Z51 is the first-listed code.

**EXAMPLE**

Christian presents for his induction chemotherapy treatment. He has acute lymphoblastic leukemia.

Z51.11 Encounter for antineoplastic chemotherapy

C91.00 Acute lymphoblastic leukemia, not having achieved remission

If the patient presents with anemia associated with an adverse effect of the administration of chemotherapy or immunotherapy and the only treatment is for the anemia, the anemia code is sequenced first, followed by the appropriate codes for the neoplasm and the adverse effect. When the admission/encounter is for management of an anemia associated with an adverse effect of radiotherapy, the anemia code should be sequenced first, followed by the appropriate neoplasm code and code Y84.2 *Radiological procedure and radiotherapy as the cause of abnormal reaction of the patient, or of later complication, without mention of misadventure at the time of the procedure.*

**EXAMPLE**

Eight-year-old Paula is admitted for a blood transfusion due to anemia caused by her chemotherapy treatment. She has mixed cellular Hodgkin’s lymphoma of the abdomen.

D64.81 Anemia due to antineoplastic chemotherapy

T45.1X5A Adverse effect of antineoplastic and immunosuppressive drugs

C81.23 Mixed cellular classical Hodgkin lymphoma, intra-abdominal lymph nodes
If the patient presents with anemia associated with a malignancy, even if the treatment is solely directed at the anemia, the malignancy is listed first, followed by the code D63.0 *Anemia in neoplastic disease*. When code D63.0 is referenced in the Tabular Index there is an instructional note under the code that indicates to code first the neoplasm.

**EXAMPLE**

Avery has acute myeloblastic leukemia which has caused her to become anemic. She is being admitted for blood transfusion.

- C92.00 Acute myeloblastic leukemia, not having achieved remission
- D63.0 Anemia in neoplastic disease

If the patient presents for management of dehydration due to the malignancy and only the dehydration is being treated (intravenous rehydration), the dehydration is sequenced first, followed by the codes for the malignancy.

**EXAMPLE**

Matthew has cancer of the brain stem. He has trouble swallowing, so he has not been drinking as he should and has become dehydrated. He is admitted for intravenous rehydration.

- E86.0 Dehydration
- C71.7 Malignant neoplasm of brain stem