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Association of Pediatric Hematology/Oncology Nurses

A Handbook for Families

Acute Myelogenous Leukemia

ONCOLOGY SERIES

Acute Myelogenous Leukemia

A HANDBOOK FOR FAMILIES

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ABOUT THIS COVER

This cover is designed for your child to color and personalize.

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WHAT IS LEUKEMIA?

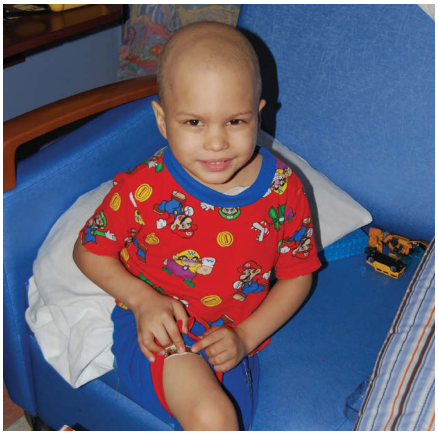
Leukemia is a cancer of the blood and bone marrow. The blood cells that are made in the bone marrow include red blood cells (RBCs), white blood cells (WBCs), and platelets. Each type of blood cell has its own job in the body. RBCs provide oxygen and energy to the body. WBCs are the infection-fighting cells. Platelets help blood clot (or thicken); the blood's ability to clot is important during certain situations, such as when you have a cut. In this way, platelets control bleeding and the appearance of bruises. Leukemia occurs as a result of abnormal growth of immature blood cells, called blast cells. These immature cells grow out of control, crowd out the normal cells (WBCs, RBCs, and platelets) in the bone marrow, and eventually spill out into the bloodstream. As a result, leukemia may be found in other parts of the body such as the lymph nodes, liver, spleen, central nervous system (the brain and spinal cord), testicles, skin, and other organs.

WHAT IS BONE MARROW?

Bone marrow is the spongy tissue located in the center of bones, which is where blood cells are developed. This process of blood cell development is called hematopoiesis. The process of hematopoiesis starts with a stem cell and results in the formation of RBCs, WBCs, and platelets. When these cells are fully developed, they enter the blood system and circulate throughout the body.

BONE MARROW CELLS

Red Blood Cells (RBCs)	Carry oxygen throughout the body
White Blood Cells (WBCs)	Help the body fight infection
Platelets	Prevent bleeding and promote clotting



WHAT IS ACUTE MYELOGENOUS LEUKEMIA?

Acute myelogenous leukemia (AML) is a rapidly progressing type of leukemia that develops from the myeloid cell line. AML results from the overproduction of very young, immature cells called leukemia blasts. These blasts (immature blood cells) cannot carry out the normal function of a healthy blood cell. Instead, they crowd within the bone marrow and decrease the production of normal bone marrow cells.

HOW ARE DIFFERENT TYPES OF AML CLASSIFIED?

AML used to be classified based on how the leukemia bone marrow cells looked under a microscope. In 2001, the World Health Organization (also known as WHO) introduced a new system for grouping the different types of AML based on gene changes in the leukemia cells. As specific changes have been identified through research, they have been grouped to help indicate how well the patient will respond to treatment. A 2008 update included more gene changes linked to AML. In 2016, the grouping system was updated to include leukemia biomarkers,

which are very important to the diagnosis, prognosis (predicted outcome), and treatment of leukemia. (A *biomarker* is a particle in the blood that can be measured to see how well the body responds to treatment.)

These groupings help guide clinicians and researchers on how best to treat the different types of AML. Some gene changes are linked with a higher risk for disease and may require more intensive treatment. Be sure to discuss your child's specific gene changes with the treatment team so you know the classification of your child's AML and how it impacts your child's treatment.

WHAT ARE THE SIGNS AND SYMPTOMS OF AML?

Signs and symptoms of AML vary in severity from patient to patient. Symptoms often result from overcrowding in the bone marrow by abnormal leukemia cells. When this happens, there is little room in the bone marrow for normal cells (RBCs, WBCs, and platelets) to develop. As a result, your child is at risk for anemia, infection, and bleeding. In addition, the leukemia cells circulate in the blood stream and settle into other parts of the body, causing infiltration (congestion) or swelling in these areas.

Some of the common signs and symptoms of AML include

- fatigue (becoming tired easily)
- shortness of breath during physical activity
- paleness
- easy bruising or bleeding
- mild fevers or night sweats
- recent infections
- bone and joint pain
- swollen gums
- weight loss or anorexia (loss of appetite)
- swollen lymph nodes
- swollen liver or spleen
- pinpoint red spots on the skin
- headaches.

WHAT CAUSES AML?

We do not know what causes AML, but we do know that some things may increase the risk of developing AML. These include

- previous exposure to certain chemotherapy agents (alkylating agents or topoisomerase inhibitors) and radiation therapy
- certain genetic disorders, including Down syndrome, Fanconi anemia, myelodysplastic syndrome, monosomy 7, Shwachman-Diamond syndrome, neurofibromatosis type 1, Bloom syndrome, and severe congenital neutropenia
- exposure to tobacco smoke.

AML is not contagious and cannot be passed from one person to another.

HOW MANY CHILDREN DEVELOP AML?

Each year in the United States, there are approximately 4,900 new cases of all types of leukemia diagnosed in people age 20 years and younger. Of these children, adolescents, and young adults who have leukemia, approximately 15%–20%—or about 1 in 5—will be diagnosed with AML. This is roughly 730 cases of AML each year in people age 20 years and younger.

WHERE IS AML FOUND IN THE BODY?

AML begins in the bone marrow and travels into the peripheral blood (circulating bloodstream). In some children, it also can be found in the cerebral spinal fluid (the fluid that surrounds the brain and spinal cord). AML may develop under the surface of the skin and appear as subcutaneous nodules (or raised lesions on the skin). In rare instances, the leukemia cells clump together, forming a mass or tumor. These tumors are referred to as chloromas.

WHAT TESTS AND PROCEDURES WILL MY CHILD NEED?

Blood Tests

Blood tests are done to monitor your child's blood cells and body chemistries. The complete blood count (known as a CBC) is used to detect anemia (low hemoglobin or hematocrit, a measure of low RBCs), detect changes in WBCs (infection-fighting cells), and detect changes in platelets (cells that help the blood clot). The percentage of leukemia blasts or immature WBCs also is assessed. A chemistry panel will look at your child's electrolytes (salt and sugar levels) and liver and kidney function.

Bone Marrow Aspiration and Biopsy

Bone marrow aspiration and biopsy are the primary tests used in the diagnosis of leukemia. These tests determine whether leukemia cells are present in the bone marrow (the blood-producing factory of the body). Bone marrow aspiration involves inserting a special needle into one of the bones (usually the front or back part of the hip bone) and drawing some bone marrow into a syringe. It sometimes is necessary to obtain bone marrow from both the right and left hip bones. Another part of this test involves taking a small piece of the bone marrow to examine for leukemia cells. This is referred to as a bone marrow biopsy. Bone marrow aspiration and biopsy are the only ways to positively determine whether leukemia cells are in the bone marrow. This test helps the physician determine the type of leukemia and choose proper treatment for your child. Sedation medication or sometimes general anesthesia will be used to help your child remain still and to prevent or minimize discomfort during the procedure. Your child may experience some mild discomfort after the procedure, which usually is relieved with a mild pain reliever such as acetaminophen. However, discuss the use of any medication with your child's physician or healthcare provider prior to use.

Cytogenetic Analysis

Cytogenetic analysis is the process of analyzing changes in the chromosomes of leukemia cells. During this analysis, defects in the number and structure of these chromosomes (chromosomal abnormalities) are carefully assessed. It is important to note cytogenetic analysis for leukemia analyzes only the abnormalities of the chromosomes in the leukemia cells, not other cells in the body. Abnormalities in the chromosomal number and structure of leukemia cells do not imply that your child has a genetic problem. Cytogenetic analysis of these leukemia cells is important in determining the best treatment for your child's leukemia.

Lumbar Puncture

The central nervous system (brain, spinal cord, and cerebrospinal fluid [CSF]) is a hiding place for leukemia cells. To determine whether leukemia has spread into the central nervous system, your child must undergo a lumbar puncture, also referred to as a spinal tap. In a lumbar puncture, a small needle is inserted into the back between the lower spinal bones (vertebrae) to obtain a sample of CSF. The fluid is sent to a laboratory and examined for any evidence of leukemia. The majority of children require sedation to remain still during this procedure. Most of the time, the lumbar puncture is performed at the same time as the bone marrow aspiration and biopsy. The results of the lumbar puncture will help physicians determine the type of treatment your child will receive.

Venous Access Device

A *venous access device* (VAD), sometimes called a central line, is an intravenous (IV) catheter that may be used for the duration of your child's therapy. It is inserted during surgery when your child is under heavy sedation or general anesthesia. It can be used to administer medication, chemotherapy (cancer-fighting drugs), blood products, and nutritional support. It also may be used to draw blood for testing purposes. The VAD may remain in your child for the duration of treatment and will be removed surgically when it is no longer needed.

HOW IS AML TREATED?

Treatment for AML is determined according to the risk assignment for your child's disease. Risk assignment is based on the type of AML your child has, any risk factors identified during their initial bone marrow evaluation (cytogenetic analysis results), and their response to the first round of chemotherapy. Some children may have the opportunity to take part in a clinical trial that is testing newer drugs to treat AML.

Chemotherapy

All children with AML are treated with chemotherapy. The chemotherapy drugs used to treat AML typically are administered intravenously (through the VAD). Depending on the drug used, it may be given rapidly or over a prolonged period of time. Children with AML are hospitalized to receive their medications and frequently remain hospitalized for several weeks. The chemotherapy used to treat AML is very strong and significantly lowers blood counts (the number of RBCs, WBCs, or platelets), putting your child at risk for infections, bleeding, and other complications. Following the intense chemotherapy, your child likely will remain in the hospital until their blood counts begin to recover. In an effort to prevent serious infections, your child likely will be on medications to help prevent infections from bacteria and fungus. If your child has a fever or any concern for infection, they will require evaluation by the medical team and hospitalization for observation and management of any possible infections.

Intrathecal Chemotherapy

All children with AML are treated with *intrathecal chemotherapy*, which is chemotherapy given directly into the spinal fluid by performing a lumbar puncture (spinal tap) and slowly injecting the drug into the spinal fluid. Most children require sedation to remain still during lumbar punctures. Even if no leukemia cells are detected in the spinal fluid, intrathecal chemotherapy is given at specific intervals during treatment to prevent the leukemia cells from spreading to the spinal fluid. For children with AML cells in their cerebrospinal fluid at diagnosis, intrathecal chemotherapy is given more frequently to treat the leukemia.

Monoclonal Antibody Therapy

Monoclonal antibodies are a new class of drugs for the treatment of AML. They have shown promising results in adult studies and in preliminary children's studies. These antibodies are used to deliver drugs and toxins to cancer cells. For patients with AML, the antibody attaches to a protein found on the surface of many AML cells. This class of drugs is sometimes referred to as "magic bullets." These drugs target and deliver most of the chemotherapy to the leukemia cells, sparing the normal healthy cells from the effects of treatment. Currently, these drugs are used in combination with chemotherapy to improve the long-term survival of children with AML.

Hematopoietic Stem Cell Transplant

Children who have been diagnosed with high-risk AML and have a sibling with the same human leukocyte antigen (HLA) tissue type may receive a hematopoietic stem cell transplant (HSCT) following induction chemotherapy. *High-risk AML* is defined as AML that does not achieve remission after induction chemotherapy, has high-risk cytogenetic features, or has relapsed.

HLA type refers to the unique set of proteins present on the surface of the WBCs. There is a 25% chance that two full siblings will share the same HLA type. All children who are to receive HSCT must have HLA typing completed to determine the most suitable donor. *HLA typing* is a test to determine how closely the tissue of one person matches the tissue of another person. The tissue type is obtained either through a blood test or by swabbing the inside of the mouth.

The healthcare team may decide to perform HLA typing on a child with AML, their parents, and any full siblings soon after diagnosis. For patients who require a transplant and do not have a suitable sibling match, the healthcare team may decide to search for unrelated donors through a registry such as the National Marrow Donor Program. All patients who undergo HSCT will first receive intensive chemotherapy to achieve remission.

WHAT ARE THE SIDE EFFECTS OF TREATMENT?

Chemotherapy

In addition to killing AML cells, cancer chemotherapy also damages normal tissue and causes side effects. Side effects from chemotherapy often begin soon after chemotherapy is initiated and continue for 2–4 weeks. Common side effects include nausea, vomiting, hair loss, fatigue, and diarrhea; these can occur regardless of the route of chemotherapy administration (oral, IV, or intrathecal). The most common serious side effect from chemotherapy is a drop in

normal blood counts (the number of RBCs, WBCs, and platelets), because many chemotherapy drugs used to treat AML can cause anemia (low RBC count), neutropenia (low WBC count), and thrombocytopenia (low platelet count). AML therapy often causes significant neutropenia, which places a child at risk for developing life-threatening infections that can last for several weeks. Depending on which blood cells have lowered counts, your child may experience fatigue, headaches, bruising, and difficulty breathing. Some side effects may last longer than a few weeks or may not occur until years after therapy. Your child's healthcare team will continue to see your child after treatment has been completed. Certain tests may be performed to look for any late effects of treatment.

Monoclonal Antibody Therapy

Common side effects of monoclonal antibody therapy include nausea, vomiting, headache, loss of appetite, fever and chills (most common with the first dose), fatigue, and weakness.

As with chemotherapy, the most serious common side effect of monoclonal antibody therapy is the lowering of blood counts (the number of RBCs, WBCs, or platelets). When chemotherapy and monoclonal antibodies are used together, the side effects may be more severe. Veno-occlusive disease (VOD) of the liver is a possible side effect when a patient is receiving monoclonal antibody therapy. VOD causes an increase in bilirubin (which causes jaundice, a temporary yellowing of the skin), an enlarged and painful liver, and retention of fluid (resulting in weight gain). Less common side effects include rash, hives, irregular heartbeat during infusion, dizziness or fainting, anxiety, and difficulty sleeping. A rare but serious side effect is an allergic reaction during the infusion.

WHAT IS A CLINICAL TRIAL?

Clinical trials allow healthcare providers to determine whether promising new treatments are safe and effective. Most of the advances in the treatment of childhood cancer have been made through the use of clinical trials. During clinical trials, the best-known standard treatment for a particular cancer is compared with a new experimental treatment. The experimental treatment is believed to be at least as good as, and possibly better than, the standard treatment.

Participation in clinical trials is voluntary. Because clinical trials involve research into new treatment plans, all risks cannot be known ahead of time and unknown side effects may occur. However, children who participate in clinical trials can be among the first to benefit from new treatment approaches. Before making a decision about your child's participation in a clinical trial, you should discuss the risks and the potential benefits with your child's doctor and treatment team.

WHAT IS REMISSION?

Remission, in patients with AML, means standard testing techniques cannot find leukemia cells in the bone marrow after chemotherapy. The amount of leukemia cells left in the bone marrow after the first cycle, or induction cycle, of chemotherapy is important in determining the next path of treatment. There are two forms of remission:

1. Remission (or pathologic complete remission) is defined as having less than 5% leukemia cells when the bone marrow is studied under a microscope, blood cell counts have returned to normal, and there are no other signs or symptoms of leukemia.
2. Complete molecular remission is defined as 0% leukemia cells in the bone marrow when using more sensitive tests, such as polymerase chain reaction (PCR).

Minimal residual disease (MRD) is the name given to small numbers of leukemia cells that remain during treatment or after treatment when the patient has no symptoms or signs of disease and appears to be in remission. MRD is the most accurate predictor of relapse, so the goal is to achieve complete molecular remission.

WHAT IS REFRACTORY AML?

Refractory AML occurs when the disease fails to go into remission after completion of induction chemotherapy. Children with refractory AML require a more aggressive treatment to achieve remission.

WHAT IS RELAPSE?

Relapse occurs when the AML cells return in the bone marrow after remission has been achieved. If your child's AML relapses, your healthcare team will meet with you to discuss further treatment options.

WHAT FACTORS AFFECT PROGNOSIS AND TREATMENT?

About 67% of children and adolescents younger than 19 years who are diagnosed with AML are cured, which means they do not have any signs of cancer for 5 years after completing therapy. Cure rates for children with refractory or relapsed AML are much lower. How well a child recovers from AML depends on several factors:

- the child's age at diagnosis
- race or ethnicity
- obesity
- whether AML occurred after a previous cancer treatment
- the type of AML
- certain chromosome or gene changes in the leukemia cells
- if the child has Down syndrome
- if the leukemia is in the brain and spinal cord
- if the leukemia went into remission after the first cycle of chemotherapy
- if AML has returned, the length of time from remission to relapsed disease.

HOW LONG DO THERAPIES LAST?

Treatment for AML generally lasts 6–8 months. If your child undergoes a bone marrow transplant, recovery time may be longer than if he or she were treated only with chemotherapy. Many children will experience treatment delays due to prolonged thrombocytopenia, neutropenia, or serious side effects of treatment. As a result, it often is difficult to predict exactly when treatment will end.

WHAT NEW METHODS OF TREATMENT ARE AVAILABLE?

Researchers are looking for better ways to treat AML. New drugs to treat AML are being researched, including more monoclonal antibodies, new chemotherapy drugs, and other types of immunotherapy. Your healthcare team will be your best resource for more information regarding new therapies being developed that may be used for your child.

WHAT ARE THE POTENTIAL LATE SIDE EFFECTS?

AML therapy may result in chronic side effects that can occur years after completing treatment. Possible side effects may include damage to the heart muscle and development of new cancer caused by the chemotherapy (secondary malignancy). Another possible side effect from treatment is infertility or sterility (the inability to conceive or father a child). Other late effects specific to certain types of treatment are possible and will be explained by the healthcare team.

HOW CAN I WORK WITH MY CHILD'S HEALTHCARE TEAM?

Your child's care requires a team approach. As a parent, you know your child better than anybody else and that makes you an important member of the team caring for your child. It is important that you talk openly with your child's healthcare team. Be sure to ask questions if there is anything that you do not understand or are concerned about. You may find it helpful to write down your questions when you think of them. Here are some examples of questions to ask:

- What kind of cancer does my child have?
- Has the cancer spread beyond the primary site?
- What treatment choices are available?
- What treatment do you recommend and why?
- What risks or side effects does the recommended treatment have?
- What should we do to prepare for treatment?
- What is my child's outlook for survival?
- What are the chances of a recurrence?

Use this space to write down additional questions.

ARE MY FEELINGS NORMAL, AND WHAT CAN I DO ABOUT THEM?

Hearing that your child has cancer can be shocking and overwhelming. At first you may not believe it, or you may hope that the diagnosis is wrong. However, the changes you see in your child and the experience of being in the hospital and beginning treatment will confirm the reality of your child's situation.

Many family members feel they are responsible for their child's disease or feel guilty that they were not able to detect it sooner. Remember that we do not know exactly what causes AML and that both the symptoms of AML and their severity vary from patient to patient.

In addition to shock and guilt, you and your family will probably feel angry and sad. Even the youngest family members are likely to be affected. These feelings are normal, and each family member will express them in different ways and at different times. It can be very difficult to feel so many strong emotions at once. Talking honestly with one another about feelings, reactions, and questions will help everyone in the family.

It may seem difficult to talk to friends, family, or even medical staff, but expressing your feelings can help you cope with this situation. Remember that your child will benefit if family members continue to show their caring through support and communication.

HOW CAN I HELP MY CHILD?

As a parent, you likely will notice changes in your child during the treatment. These changes or symptoms can make you feel even more helpless. It is essential to remember that, in spite of changes on the outside, your child is still the same person on the inside. Hair loss and other changes to the body are temporary. They often bother adults much more than they bother the child or the child's siblings and friends. All of the feelings about what your child is going through during treatment must be balanced by remembering that treatment provides an opportunity to cure the disease so your child can go on to live a full and meaningful life.

It is important to reinforce to your child that nothing they did or said caused this disease. Telling your child that your angry or sad feelings are directed at cancer and not at them preserves honesty and closeness in your relationship. Like you, your child will need someone with

whom they can share feelings. Don't hesitate to ask your child to express their feelings, and don't be afraid to explain what is happening and why.

In spite of the disease, your child is still growing and learning. All children—sick and well—need love, attention, discipline, limits, and the opportunity to learn new skills and try new activities. As you begin to learn about the new special needs of your child, it is important to remember that he or she still has all the needs and rights of any growing, developing child.

When talking to your child about their illness, do not avoid using direct terms and explanations. Children tolerate treatment better if they understand it and are allowed to be an active decision maker whenever possible. The same is true for parents.

IS MY CHILD'S DIET IMPORTANT DURING THE TREATMENT?

Yes. We know from research that well-nourished children tolerate therapy better and have fewer treatment delays due to illness. It may be difficult for your child to resume normal eating habits during therapy, so you will need to be flexible and creative. Offering smaller meals more often may be more successful than offering larger meals three times a day. Children usually are more interested in eating food that they help prepare. It is important to include your child in the social activity of family meals even if full meals are not eaten. Remember, nobody wins food fights—it's best not to force your child to eat.

Make sure that foods high in protein and carbohydrates are readily available to your child. It may be necessary throughout your child's treatment to provide supplemental nutritional support. Multivitamins, medicines, and herbs should be discussed with your healthcare team before you give them to your child because an interaction between them and the chemotherapy is possible. A dietician trained in children's calorie and energy needs can offer you guidance. The medical staff can help if there is a nutritional problem.

CAN MY CHILD ATTEND SCHOOL DURING TREATMENT?

Your child's ability to attend school will depend on the intensity of the therapy and on the response to treatment. Some children tolerate chemotherapy and radiation better than others. Your child may not be able to attend school for extended periods because of treatment or hospitalization. However, it is important that your child keep up with their schoolwork. Talk to the staff at your child's school about arranging additional services, such as help from a home tutor, until your child is able to return to school. You also should discuss school attendance with your child's doctor. Many pediatric hospitals have programs that allow children to attend school while they are hospitalized.

School is important because it helps children maintain social contact with their peers. Your child's time with friends will be an important part of recovery and will ease the adjustment when they return to school. It is important for your child to return to school as soon as they are medically able to do so.

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IMPORTANT PHONE NUMBERS

NOTES

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