



Rhabdomyosarcoma and Soft Tissue Sarcoma

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A HANDBOOK FOR FAMILIES

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■ WHAT ARE RHABDOMYOSARCOMA AND SOFT TISSUE SARCOMA?

Rhabdomyosarcoma, also called rhabdo, is a cancer that develops from muscle cells (rhabdomyoblasts). It can arise from muscles in many different areas of the body, most commonly the head and neck, bladder, prostate gland, arms, legs, and vagina. Other, less common sites are the chest, abdomen, and genital and anal area. Rhabdomyosarcoma accounts for more than half of all soft tissue sarcomas diagnosed in children. The type of rhabdomyosarcoma is determined by looking at a piece of the tumor under the microscope. The most common types are embryonal and alveolar rhabdomyosarcoma.

The term *soft tissue sarcoma* is generally used to refer to a cancer that arises from soft tissue other than muscle, such as connective tissue, fibrous tissue, blood vessels, nerves, and joint membrane tissue. Soft tissues connect, support, and surround other body parts and organs. Soft tissue sarcoma is rare in infants and children.

■ WHAT ARE SOME OF THE SIGNS AND SYMPTOMS OF RHABDOMYOSARCOMA AND SOFT TISSUE SARCOMA?

The signs and symptoms of rhabdomyosarcoma and soft tissue sarcoma depend on the location of the tumor. For example, if the tumor is located in the head or neck, there may be swelling around the eye (proptosis), facial nerve weakness, ear drainage, headache, facial pain, a lump in the neck, or signs of sinusitis. If the tumor is located in the arm or leg, there may be a tender or enlarged area in or around the muscle. Tumors in the bladder may cause bloody urine (hematuria) or difficulty in urinating. Tissue may even be seen growing out of the urethra. Paratesticular tumors usually cause painless enlargement of the scrotum or groin area.



■ WHAT CAUSES RHABDOMYOSARCOMA AND SOFT TISSUE SARCOMA?

There is no answer at this time as to what causes pediatric rhabdomyosarcoma and soft tissue sarcoma. We do know, however, that the tumors are not contagious and cannot be caught from another person. No behaviors or lifestyle habits, including those practiced during pregnancy, have been associated with the development of these tumors. The cancer is not caused by the presence or absence of something (such as vegetables or vitamins) in the child's diet.

■ WHO GETS RHABDOMYOSARCOMA OR SOFT TISSUE SARCOMA?

Rhabdomyosarcoma accounts for approximately 4% of all childhood cancers diagnosed before the age of 15 years. Approximately 350 cases of rhabdomyosarcoma are diagnosed each year in the United States. The

majority of children are younger than nine years of age at the time of diagnosis. Tumors of the bladder and vagina occur more often in infants and young children; tumors of the trunk, arms, and legs are more likely to affect older children and adolescents. Rhabdomyosarcoma is slightly more common in males than in females. Soft tissue sarcoma is rare in children, accounting for 3% of all childhood cancers, but it can occur at any age. It may be associated with other hereditary conditions or with radiation exposure.

■ IS RHABDOMYOSARCOMA OR SOFT TISSUE SARCOMA INHERITED?

It does not appear that these tumors are inherited from a family member (genetic). However, chromosomal (genetic) abnormalities are present in these tumors. More research is needed in this area to develop improvements in early detection and treatment. Rhabdomyosarcoma may be associated with certain familial syndromes such as Li-Fraumeni syndrome, neurofibromatosis type I, Beckwith-Weidemann syndrome, cardiofaciocutaneous syndrome, and Costello syndrome.

■ WHAT IS METASTASIS?

Metastasis refers to the spread of a tumor from its original location (primary site) to other parts of the body. Rhabdomyosarcoma and soft tissue sarcoma may metastasize, or spread, to the lymph nodes, lungs, bone, bone marrow (the inside of bones where blood cells are made), and brain.

■ WHAT IS STAGING?

Staging is the process of determining the location and amount of the disease at the time of diagnosis. Staging for rhabdomyosarcoma and soft tissue sarcoma is based on the location of the tumor, whether there is lymph node involvement or metastasis, the type of tumor cell, and, if a surgical resection has been done, the amount of tumor remaining after surgery. The method of treatment proposed for your child will depend upon the stage of the disease at the time of diagnosis. Several staging systems are used for soft tissue sarcoma, but no one system applies to all types of this cancer. Your doctor can discuss the staging system for your child's soft tissue sarcoma. The clinical group staging system most commonly used for rhabdomyosarcoma is based on the extent of the disease and the amount of tumor remaining after surgery.

Stages of Rhabdomyosarcoma

STAGE I

The tumor is located in the eye, head, or neck, or near the sex organs or bladder, and has not spread outside this area.

STAGE II

The tumor is located in only one area (other than the areas in stage I), is smaller than 2 inches (5 centimeters) across, and has not spread to the lymph nodes.

STAGE III

The tumor is located in only one area (other than the areas in stage I), is larger than 2 inches (5 centimeters) across, and may have spread to lymph nodes near the tumor.

STAGE IV

The cancer has spread from the primary site and is found in more than one other place at the time of diagnosis.

■ WHAT TESTS AND PROCEDURES WILL MY CHILD NEED?

To diagnose rhabdomyosarcoma or soft tissue sarcoma and to determine the extent of your child's disease, a number of tests and procedures are necessary, including:

Tumor Biopsy

A tumor biopsy is necessary to make a diagnosis and to determine what treatment should be chosen. The biopsy is usually done through an incision made by the surgeon in the operating room while the child is under anesthesia. Some medical centers do the biopsy in the radiology department, where the biopsy sample is obtained through a needle guided into the tumor using imaging studies. Your child's comfort and anxiety level will always be considered in planning procedures that involve needles.

Bone Marrow Aspiration and Biopsy

Bone marrow aspiration and biopsy are necessary to determine whether there are tumor cells in the bone marrow (the blood-producing factory of the body). The test involves inserting a special needle into one of the bones (usually the back part of the hip bone) and drawing some bone marrow into a syringe. It is usually necessary to obtain bone marrow samples from both hip bones. This test also involves taking a small piece of the hip bone (biopsy) to examine for tumor cells. This is the only way to determine whether or not tumor cells are in the bone marrow. This test helps the physician determine the stage of the disease and choose the proper treatment. Medication can be used to help the child remain still and to minimize any discomfort.

Blood Tests

Blood tests are frequently done to monitor the child's blood cell counts, electrolytes and body salts, and organ function. A complete blood count (CBC) is useful in detecting a drop in the number of red blood cells (oxygen carriers), white blood cells (infection fighters), or platelets (cells that help blood clot properly). Blood chemistries such as blood urea nitrogen (BUN) and creatinine monitor changes in kidney function. Other blood chemistries, like hepatic function panel and bilirubin, detect changes in liver function.

These tests are done at the time of diagnosis, and throughout therapy to monitor the child's response to treatment and to monitor possible side effects of therapy.



Lumbar Puncture

A lumbar puncture is a procedure done to obtain a small amount of cerebral spinal fluid, which is the fluid that surrounds and protects our brain and spinal cord. The fluid is tested for the presence of tumor cells. Children who have rhabdomyosarcoma located in the head and near the brain, may require this test at diagnosis. Most children are sedated for the procedure. A needle is inserted between the lower back bones (vertebrae) to collect the fluid. The spinal column is entered below the spinal cord to avoid injury to the cord. Spinal fluid is made continually by our bodies, so the small amount removed is quickly replaced.

MRI

An MRI (magnetic resonance imaging) is a test that gives very exact pictures of organs and tumors inside the body. The child is asked to lie on a table, which is then moved into a tube-like machine that surrounds the child with a magnetic field. The test is painless, but the machine makes a loud noise that may be scary for the child. The child must not move at all during the test, so a sedative may be given.

CT or CAT Scan

A CT (computerized tomography) or CAT (computerized axial tomography) scan is a computer-assisted X-ray that shows very precise pictures of internal organs and tumors. The procedure is painless, but the child must be able to lie absolutely still during the scan. Younger children may require sedation to help them lie still. The child may be asked not to eat or drink for several hours before the exam. It may be necessary for the child to drink a liquid containing a flavorless dye that makes the pictures clearer. Sometimes a small amount of dye is injected into a vein (IV). In general, no side effects from either type of dye occur, although allergic reactions are possible.

Bone Scan

A bone scan may be done to determine whether the tumor has spread to any of the bones. It involves injecting a small amount of isotope (radioactive marker) into a vein, allowing 2–3 hours for the isotope to distribute itself throughout the body, and then having the child lie still for scans (pictures) to be taken of the entire body. This isotope is picked up by tumor cells so that the tumor's location in the body can be seen. The scanning process is painless, but some children need sedation to help them lie still.

X ray

A complete skeletal X ray may be necessary to confirm the findings found on other scans. A chest X ray may be necessary to determine whether the tumor has spread to the lungs. Efforts will be made to minimize your child's exposure to X rays.

Ultrasound

The ultrasound test uses high-frequency sound waves to look at internal body organs or tumors. It can help detect tumors in the lymph nodes, abdomen, or scrotum. It is painless and involves no radiation. If the abdomen is being scanned, the child is not allowed to eat or drink anything for about 4 hours before the test, but can resume a normal diet after the scan.

Central Venous Access Device

A central venous access device (CVAD), sometimes called a central line, is a permanent intravenous (IV) tube that may be used during your child's therapy. The tube allows fluids, medications, and blood products to be given and blood to be removed without painful needle sticks. It can be used to administer medications, chemotherapy, blood products, and nutritional support when needed, as well as to draw blood for testing. It is inserted by a surgeon while the child is under anesthesia. You and your child's doctor will decide whether your child needs a CVAD.

■ HOW CAN RHABDOMYOSARCOMA AND SOFT TISSUE SARCOMA BE TREATED?

Three types of therapy are commonly used to treat rhabdomyosarcoma and soft tissue sarcoma: surgery, chemotherapy, and radiation. The type of therapy chosen depends upon the type of tumor and the extent of the disease. A combination of treatments is often used. The order of treatments also depends on the child's presenting signs and symptoms. Surgery and radiation may be delayed to allow for chemotherapy to shrink the tumor, or they may occur as part of the initial therapy. Novel targeted agents, such as antibody therapy, are also currently being evaluated for the treatment of certain types of rhabdomyosarcoma. Your child's doctor will talk with you about the best treatment for your child, and decisions about treatment will be made by your family and the doctor.

Surgery

Surgery to remove the bulk of the tumor often can be done at the time of diagnosis. For some soft tissue sarcomas, surgery may be the only treatment required. For very large tumors, chemotherapy or radiation treatments may be given first to shrink the tumor. This is done to minimize the risk of surgical injury to the normal tissues surrounding the tumor. A "second-look" surgery may be done after the child has received a certain amount of treatment. This type of surgery can determine the child's response to the treatment received so far and can help the healthcare team plan any additional therapy.

Chemotherapy

Chemotherapy involves giving medicines that will help kill the cancer cells, shrink the tumor, and prevent the tumor from spreading to new places. Several chemotherapy medications are known to be effective in the treatment of rhabdomyosarcoma and soft tissue sarcoma, but no single chemotherapy medicine can control this disease by itself. As a result, chemotherapy medications are usually given in combinations, through a vein. Treatment usually lasts several days and often is given in the hospital. Your child may receive some chemotherapy medicines that can be given in a clinic or at home. When chemotherapy is needed, close monitoring of the child is a priority. Your child's doctor and nurse will explain the possible side effects of the specific chemotherapy treatment recommended for your child.



Radiation

Radiation therapy is a special kind of X-ray treatment used in rhabdomyosarcoma and some soft tissue sarcomas. Radiation treatment is very precise and is given in specially-measured amounts by radiation therapy experts. If radiation is necessary for your child, the radiation oncologist will discuss with you exactly how the radiation will be given and how long the treatments will last. In general, children experience very few side effects while they are receiving radiation therapy. Some children become more tired than usual or have decreased appetites. There can be delayed side effects, depending on which areas of the body receive the radiation treatment. The possible side effects will be discussed with you in detail by the radiation oncologist.

■ HOW LONG WILL MY CHILD'S THERAPY LAST?

The length of therapy depends upon the type and stage of your child's tumor at the time of diagnosis. Usually the treatment is given over several months, and it could last as long as a year. Follow-up blood tests and check-ups between chemotherapy treatments are usually done in the clinic or doctor's office.

■ WHAT NEW METHODS OF TREATMENT EXIST?

Most of the advances in the treatment of childhood cancer have been made through a process known as clinical trials. In clinical trials, the best known (standard) treatment for a particular cancer is compared with a new (experimental) treatment that is believed to be at least as good as, and possibly better than, the standard treatment. Clinical trials allow doctors to determine whether promising new treatments are safe and effective.

Participation in clinical trials is voluntary. On one hand, because clinical trials involve research into new treatment plans, all risks cannot be known ahead of time, and unknown side effects may occur. On the other hand, 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 as well as the potential benefits with your child's doctor and treatment team.



More information about clinical trials is available in the free booklet Taking Part in Cancer Treatment Research Studies (National Cancer Institute Publication No. 105). To obtain this booklet, as well as a great deal of useful information about childhood cancer, call 800.4CANCER (800.422.6237). The booklet can also be downloaded from the National Cancer Institute website at www.cancer.gov/clinicaltrials/resources/taking-part-treatment-trials. You can also find general information on clinical trials at www.cancer.gov/clinicaltrials/resources/what-to-know-about-trials.

■ 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 no doubt confirm the reality of your child's situation.

Many family members feel that they are somehow responsible for their child's disease, or they feel guilty that they were not able to detect it sooner. Remember that this disease is often not noticeable until it is quite advanced, so there may have been no way to detect it in the early stages.

In addition to shock and guilt, you and your family will probably feel anger and sadness. 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 venting your feelings can help you cope with this situation. Remember that your child will benefit if family members continue to show that they care through support and communication.

■ HOW CAN I HELP MY CHILD?

As a parent, you will often notice changes in your child during the treatment. These changes or symptoms can make you feel even more helpless. It is always 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 in body appearance are temporary. They often bother adults much more than they bother the child or their siblings and friends. All of your feelings about what your child is going through during treatment must be balanced by remembering that treatment provides an opportunity to cure the disease and to have your child go on to live a full and meaningful life.

It is important to reinforce to your child that nothing he or she did or said caused this disease. Telling your child that your angry or sad feelings are directed at the cancer and not at him or her will help preserve honesty and closeness in your relationship. Like you, your child will need someone with whom he or she can share feelings. Don't hesitate to ask your child to express his or her 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 your child's new needs, it is important to remember that he or she still has all the rights of any growing, developing person. Do not avoid using direct terms and explanations with your child. Children tolerate treatment better if they understand it and are allowed to be active decision makers whenever possible. The same is true for parents.



■ IS MY CHILD'S DIET IMPORTANT DURING TREATMENT?

Yes. We know from research that well-nourished children tolerate therapy better and have fewer treatment delays as a result of illness. It may be difficult for your child to resume normal eating habits during therapy, so you will need to be flexible and creative. Often numerous small meals are more easily tolerated than three large ones. Children usually are more interested in eating foods that they help prepare. It is important to include your child in the social activity of family meals even if full meals aren't eaten. Remember, nobody wins food fights—it is best not to force your child to eat.



Make sure that foods high in protein and carbohydrates are readily available. Multivitamins, medicines, and herbs should be discussed with your healthcare team before you give them to your child, because a negative interaction between them and the chemotherapy is possible. A dietitian trained in children's energy needs can offer you guidance. The medical staff can intervene 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 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 his or her schoolwork. Talk to staff at your child's school about arranging services, including help from a home tutor, until he or she is able to return to school. You should also discuss school attendance with your child's doctor. Many pediatric hospitals have programs that enable children to attend school while they are hospitalized.



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

■ PROGNOSIS STATEMENT

Prognosis is a word that doctors use to describe what they believe the outcome will be for a patient, including the chances of recovery. Your child's cancer prognosis is as unique as he or she is. There are many factors that affect survival outcomes. A child's age at diagnosis, the extent of disease, genetic factors, infections, nutrition, delays in the timing of treatment, and how well your child's body can tolerate chemotherapy and radiation all play an important role in the child's long-term outcome. Because general statements about survival rates are just numbers, please talk to your medical team about your child's individual prognosis and plan of care.

■ ONLINE RESOURCES

CureSearch. www.curesearch.org.

National Cancer Institute. (2009). Childhood Rhabdomyosarcoma (PDQ®): Treatment. In PDQ® Cancer Information Summaries [Online]. Available at <http://cancernet.nci.nih.gov/cancertopics/pdq/treatment/childrhabdomyosarcoma>.

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