All About Rhabdomyosarcoma

What is a sarcoma?

Sarcoma is a term that refers to any cancer of the bone, muscle, or other connective tissue, such as cartilage and tendons. Sarcomas can happen in children and adults and both males and females. There are many different types of sarcomas, depending on where the cancer cells grow and how they look under a microscope. Each type acts differently which decides how they are treated. Some sarcomas start from stem cells within the connective tissues, and these are more common in children than adults.

What is a rhabdomyosarcoma?

The name itself comes from a combination of 3 smaller words:

- *Rhabdo* means "rod-shaped."
- *Myo* is muscle.
- *Sarcoma* is the type of cancer, as described above.

Rhabdomyosarcoma cells tend to look rod-shaped under a microscope, and they have many features of muscle cells. During normal fetal development, cells called rhabdomyoblasts "grow up" to become the skeletal muscles of the body. When these cells multiply abnormally within a child’s body, a rhabdomyosarcoma happens. Rhabdomyosarcomas can occur anywhere in the body, but often in the head, neck, bladder, vagina, extremities (arms and legs) and the trunk.

Who gets rhabdomyosarcomas, and why?

Rhabdomyosarcoma is the most common type of soft tissue sarcoma found in children. It is still a rare cancer overall, making up about 3% of all childhood cancers. About 400-500 new cases of rhabdomyosarcoma are diagnosed in the United States every year.

There is no race or area of the world that has been linked with higher rates of rhabdomyosarcoma. It is slightly more common in boys than girls.

Some changes in DNA passed down from your parents (inherited) can increase the risk of rhabdomyosarcoma. It is known that rhabdomyosarcomas are associated with conditions such as Li-Fraumeni syndrome, Costello syndrome, and Noonan syndrome. The exact cause of these gene mutations is not known.

Some children who develop rhabdomyosarcomas also have congenital problems of various organ systems (abnormal development of heart, gut, brain, etc). The risk of rhabdomyosarcoma may be higher for children who also have certain rare genetic disorders, such as neurofibromatosis type 1. Unlike many adult cancers, there are no known environmental conditions that increase the chance of a person developing rhabdomyosarcoma. No connection has ever been found between rhabdomyosarcoma and exposure to toxic substances, environmental pollution, radiation (eg: x-rays during pregnancy), or physical injury (trauma). Not even tobacco smoke has been linked with development of this or any other childhood cancers.

Are all rhabdomyosarcomas the same?

No, there are different types of rhabdomyosarcomas. The classification is based on what the cells look like under a microscope, genetic mutations, and clinical behavior. The 2 major types of rhabdomyosarcomas are described below:

- **Embryonal, or "translocation negative" rhabdomyosarcoma:** This type tends to occur in the head and neck, bladder,
vagina in girls, and around the prostate and testes in boys. This type usually affects infants and young children. The cells have an immature appearance, meaning they look like developing muscle cells.

- **Alveolar, or "translocation positive" rhabdomyosarcoma:** This type of rhabdomyosarcoma is found more often in large muscles of the trunk, arms, and legs, and often affects older children or teenagers. It is called alveolar because the cancer cells form little hollow spaces, or "alveoli." Although alveolar rhabdomyosarcomas were originally named this way because of their appearance under the microscope, researchers have now found that nearly all of them bear specific genetic mutations.

### How are rhabdomyosarcomas found?

The answer to this question depends on the location of the tumor. As mentioned earlier, rhabdomyosarcomas can arise from any area in the body, but the most common sites are the head and neck area, the genitourinary organs (like the kidneys, bladder, fallopian tubes, and penis), and the extremities. Tumors in the head and/or neck region can cause headaches, nausea, vomiting, visual problems (double vision), facial drooping, and airway obstruction. Tumors in the GU area can cause blockage of the bladder or bowels, which can be a medical emergency depending on how badly it is obstructed. Growths on the extremities can be painful and limit the use and motion of the affected arm or leg. Sometimes, the rhabdomyosarcoma is not diagnosed until after the tumor cells have spread to other parts of the body. The most common areas they spread to are the lungs, bones, bone marrow, and lymph nodes.

### How are rhabdomyosarcomas evaluated?

The workup of these tumors typically includes the following studies:

- CT and MRI scans of affected area(s).
- Chest x-ray or CT scan of chest.
- Bone scan.
- Bone marrow biopsy.
- For genitourinary location:
  - Cystoscopy (scope to look inside the bladder).
  - Barium enema and rectal ultrasound.

### How are rhabdomyosarcomas staged?

The staging system for rhabdomyosarcoma is very complicated. All rhabdomyosarcomas are assigned a group, a stage, and a risk category. In order for stage to be assigned, the disease must be determined to be in either a "favorable" or an "unfavorable" site. You provider will be able to discuss with you the specific stage, group and risk category of the rhabdomyosarcoma.

### How are rhabdomyosarcomas treated?

Treatment of these tumors calls for a multidisciplinary approach, with input from providers who use surgery, chemotherapy, and radiation therapy to treat cancer. Rhabdomyosarcomas behave very differently, depending on their type and location. Because of this, each case is treated differently.

Some of the most commonly used chemotherapies are: vincristine, dactinomycin, cyclophosphamide, etoposide, ifosfamide, epirubicin, and doxorubicin. Even when a rhabdomyosarcoma is completely removed, the child remains at high risk for metastatic disease (cancer that has spread to a different area). For this reason, chemotherapy is used for all patients to treat any microscopic cells that may be present in lymph nodes or the bloodstream. With use of aggressive chemotherapy, the risk of metastatic disease is reduced.

In addition to chemotherapy, patients require surgery, radiation, or a combination of these two for a phase of treatment called "local control." Although all rhabdomyosarcomas are biopsied surgically at the time of diagnosis, surgery is often needed to remove all of some of the tumor. Some tumors cannot be removed safely. If the tumor can be removed completely, often only surgery and chemotherapy are needed. If the surgeon or pathologist are concerned that microscopic cells may have been left...
behind after surgery, either at the tumor margin or in the lymph nodes, radiation after surgery will likely be recommended. If the tumor cannot be removed at all, higher-dose radiation is used.

Delivery of radiation may be done with x-rays, often using a technique called intensity modulated radiation treatment (IMRT), or with proton therapy. Proton therapy is available at some specialized centers around the world, and may offer advantages during treatment of rhabdomyosarcomas through reduction of radiation to normal, growing tissues.

**Clinical Trials**

There are clinical research trials for most types of cancer, and every stage of the disease. Clinical trials are designed to determine the value of specific treatments. Trials are often designed to treat a certain stage of cancer, either as the first form of treatment offered, or as an option for treatment after other treatments have failed to work. They can be used to evaluate medications or treatments to prevent cancer, detect it earlier, or help manage side effects. Clinical trials are extremely important in furthering our knowledge of this disease. It is through clinical trials that we know what we do today, and many exciting new therapies are currently being tested. Talk to your provider about participating in clinical trials in your area. You can also explore currently open clinical trials using the OncoLink Clinical Trials Matching Service.

**Follow-up Care and Survivorship**

After treatment for childhood cancer, you will be followed closely to monitor for the cancer coming back, to manage ongoing side effects, and to transition to survivorship. At first, you will have frequent appointments with providers and have ongoing tests to monitor your health. As time goes on, these visits and testing will become less frequent. The oncology team will discuss the plan for follow-up.

What can you do to live a healthy life after treatment? There is no supplement or specific food you can eat to assure good health, but there are things you can do to live healthier, prevent other diseases, detect cancers early and work with the social and emotional issues, including insurance, employment, relationships, sexual functioning, and fertility, that a prior cancer diagnosis sometimes brings with it. Your oncology team is there to support you and can help you find support resources.

It is important to have a plan for who will provide follow-up care (an oncologist, survivorship doctor or primary care doctor). Talk with the team about developing a survivorship care plan or develop one using the Smart ALACC tool. Your child may benefit from being seen in a survivorship clinic. At a survivorship clinic, a provider can review your child’s history and provide recommendations. You can contact cancer centers in your area to see if they have a survivor’s clinic or search for a clinic on OncoLink's survivorship clinic list.

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