About Rhabdomyosarcoma

Overview and Types

If your child has been diagnosed with rhabdomyosarcoma or you are worried about it, you likely have a lot of questions. Learning some basics is a good place to start.

- What Is Rhabdomyosarcoma?
- What Are the Differences Between Cancers in Adults and Children?

Research and Statistics

See the latest estimates for new cases of rhabdomyosarcoma in the US and what research is currently being done.

- What Are the Key Statistics About Rhabdomyosarcoma?
- What’s New in Rhabdomyosarcoma Research and Treatment?

What Are the Differences Between Cancers in Adults and Children?

Cancers that develop in children are often different from the types that develop in adults. Childhood cancers are often the result of DNA changes in cells that take place very early in life, sometimes even before birth. Unlike many cancers in adults, childhood cancers are not strongly linked to lifestyle or environmental risk factors.

There are exceptions, but childhood cancers tend to respond better to treatments such as chemotherapy. Children’s bodies also tend to tolerate chemotherapy better than adults’ bodies do. But cancer treatments such as chemotherapy and radiation therapy can have long-term side effects, so children who have had cancer need careful attention for the rest of their lives.
Since the 1960s, most children and teens with cancer have been treated at specialized centers designed for them. These centers offer the advantage of being treated by a team of specialists who know the differences between adult and childhood cancers, as well as the unique needs of children with cancer and their families. This team usually includes pediatric oncologists (childhood cancer doctors), surgeons, radiation oncologists, pathologists, pediatric oncology nurses, and nurse practitioners.

These centers also have psychologists, social workers, child life specialists, nutritionists, rehabilitation and physical therapists, and educators who can support and educate the entire family.

Most children with cancer in the United States are treated at a center that is a member of the Children’s Oncology Group (COG). All of these centers are associated with a university or children’s hospital. As we have learned more about treating childhood cancer, it has become even more important that treatment be given by experts in this area.

When a child or teen is diagnosed with cancer, it affects every family member and nearly every aspect of the family’s life. You can read more about coping with these changes in our document Children Diagnosed With Cancer: Dealing With Diagnosis.

- References

  See all references for Rhabdomyosarcoma

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What Is Rhabdomyosarcoma?

Sarcomas are cancers that develop from connective tissues in the body, such as muscles, fat, bones, the linings of joints, or blood vessels. There are many types of sarcomas. Rhabdomyosarcoma (RMS) is a cancer made up of cells that normally develop into skeletal muscles. The body has 3 main types of muscles.

- **Skeletal** (voluntary) muscles are muscles that we control to move parts of our body.
- **Smooth** muscle is the main type of muscle in internal organs (except for the heart).
For example, smooth muscles in the stomach and intestines push food along as it is digested. We do not control this movement.

• **Cardiac** muscle is the main muscle type in the heart.

About 7 weeks into the development of an embryo, cells called *rhabdomyoblasts* (which will eventually form skeletal muscles) begin to form. These are the cells that can develop into RMS. Because this is a cancer of embryonal cells, it is much more common in children, although it does sometimes occur in adults.

We might think of our skeletal muscles as being mainly in our arms and legs, but these skeletal muscle cancers can start nearly anywhere in the body, even in some parts of the body that don’t normally have skeletal muscle.

Common sites of RMS include:

• Head and neck (such as near the eye, inside the nasal sinuses or throat, or near the spine in the neck)
• Urinary and reproductive organs (bladder, prostate gland, or any of the female organs)
• Arms and legs
• Trunk (chest and abdomen)

**Types of rhabdomyosarcoma**

There are 2 main types of RMS, along with some less common types.

**Embryonal rhabdomyosarcoma**

Embryonal rhabdomyosarcoma (ERMS) usually affects children in their first 5 years of life, but it is the most common type of RMS at all ages.

The cells of ERMS look like the developing muscle cells of a 6- to 8-week-old embryo. ERMS tends to occur in the head and neck area, bladder, vagina, or in or around the prostate and testicles.

Two subtypes of ERMS, botryoid and spindle cell rhabdomyosarcomas, tend to have a better prognosis (outlook) than the more common conventional form of ERMS.

**Alveolar rhabdomyosarcoma**
Alveolar rhabdomyosarcoma (ARMS) typically affects all age groups equally. It makes up a larger portion of RMS in older children and teens than in younger children (because ERMS is less common at older ages).

ARMS most often occurs in large muscles of the trunk, arms, and legs. The cells of ARMS look like the normal muscle cells seen in a 10-week-old fetus.

ARMS tends to grow faster than ERMS and usually requires more intense treatment.

**Anaplastic rhabdomyosarcoma and undifferentiated sarcoma**

Anaplastic rhabdomyosarcoma (formerly called pleomorphic rhabdomyosarcoma) is an uncommon type that occurs in adults but is very rare in children.

Some doctors also group undifferentiated sarcomas with the rhabdomyosarcomas. Using lab tests, doctors can tell that these cancers are sarcomas, but the cells don’t have any features that help classify them further.

Both of these uncommon cancers tend to grow quickly and usually require intensive treatment.

**Rhabdomyosarcoma in adults**

Most rhabdomyosarcomas develop in children, but they can also occur in adults. Adults are more likely to have faster-growing types of RMS and to have them in parts of the body that are harder to treat. Because of this, RMS in adults is often harder to treat effectively.

**This document focuses on RMS in children, but most of the information here (including much of the treatment information) applies to RMS in adults as well.**

- References
- See all references for Rhabdomyosarcoma

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What Are the Key Statistics About Rhabdomyosarcoma?

About 3% of all childhood cancers are rhabdomyosarcoma (RMS). About 350 new cases of RMS occur each year in the United States. The number of new cases has not changed much over the past few decades.

Most rhabdomyosarcomas are diagnosed in children and teens, with more than half of them in children younger than 10 years old. These tumors are usually embryonal rhabdomyosarcomas (ERMS) and tend to develop in the head and neck area or in the genital and urinary tracts. Alveolar rhabdomyosarcoma (ARMS) affects all age groups and is found more often in the arms, legs, or trunk.

RMS is slightly more common in boys than in girls. No particular race or ethnic group seems to have an unusually high rate of RMS.

The prognosis (outlook) for people with RMS depends on many factors, including the type of RMS, the location and size of the tumor, the results of surgery, and whether the cancer has metastasized (spread). Children aged 1 to 9 tend to have a better outlook than infants or older children or adults. Statistics related to survival are discussed in the section Survival Rates for Rhabdomyosarcoma by Risk Group.

- References
See all references for Rhabdomyosarcoma

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What’s New in Rhabdomyosarcoma Research and Treatment?

The treatment of rhabdomyosarcoma (RMS) has come a long way in the past few decades, largely due to the work of the Intergroup Rhabdomyosarcoma Study Group.
(now known as the Soft Tissue Sarcoma Committee of the Children’s Oncology Group). However, more work needs to be done. Research on RMS is being done at many medical centers, university hospitals, and other institutions across the world.

**Better classification of rhabdomyosarcomas**

Newer molecular techniques may help doctors better categorize RMS and predict which patients will respond best to certain treatments. For example, rather than just looking at the cancer cells under a microscope, researchers have begun to use special genetic tests to help classify RMS.

About 1 out of 4 cancers that doctors would usually classify as alveolar rhabdomyosarcoma (ARMS) have been found to lack the typical gene change (the \textit{PAX/FOXO1} fusion gene) seen in ARMS. Some early studies have shown that these cancers seem to act more like embryonal rhabdomyosarcoma (ERMS) than ARMS. ERMS generally requires less intensive treatment than ARMS. If this finding is confirmed in other studies, it may allow doctors to use less intensive treatments on these cancers and still achieve the same results.

**Improving standard treatments**

A major goal of current research is to treat all patients more effectively, while reducing the need for intensive treatments (and their side effects) when possible. For example, researchers are studying whether children who have a low risk of the tumor recurring can be treated without using potentially harmful treatments such as radiation therapy and the chemotherapy drug cyclophosphamide.

Because children’s bodies are very sensitive to radiation, doctors are looking for ways to limit the doses as much as possible. Newer radiation therapy techniques allow doctors to aim the radiation more precisely, limiting the amount that reaches normal body tissues. Some of these techniques were described in the section Radiation Therapy for Rhabdomyosarcoma.

Other ways to give radiation are also being studied. For example, in \textit{stereotactic radiation therapy}, a special machine aims high doses of radiation at the tumor from many different angles, concentrating it on the tumor very precisely for short periods of time.

\textit{Proton beam radiation} is another newer approach. Standard radiation beams give off the same amount of radiation at all points as they pass through the body. Proton beam
radiation uses radioactive particles that travel only a certain distance before releasing most of their energy. Doctors can use this property to limit the radiation reaching normal body tissues. This new approach seems promising, but it’s not yet clear if it’s better than other newer forms of radiation therapy. It’s also available in only a limited number of centers around the country at this time.

Doctors are studying adding newer chemotherapy drugs such as irinotecan and temozolomide to the standard chemotherapy regimens in those who have a higher risk of the tumor recurring.

For patients at a high risk of tumor recurrence, doctors are looking at maximizing the early treatment with drugs such as cyclophosphamide and ifosfamide by giving them more frequently (a concept called *interval compression*).

### Newer treatment approaches

Drugs that target specific parts of cancer cells (as opposed to just attacking fast-growing cells, as chemotherapy drugs do) are now being studied for use in RMS. Some of these drugs are already being used to treat certain adult cancers. Examples of newer targeted drugs being studied for use against RMS include:

- IGF-1 receptor inhibitors, such as cixutumumab (IMC-1A2)
- Drugs that affect a tumor’s ability to make new blood vessels, such as bevacizumab (Avastin) and sorafenib (Nexavar)
- Drugs that target the mTOR protein, such as temsirolimus (Torisel) and everolimus (Afinitor)
- Drugs that target the ALK protein, such as crizotinib (Xalkori)
- Drugs that target the cell’s hedgehog pathway, such as LDE225
- Dasatinib (Sprycel)

Researchers are also testing other new ways to treat RMS. For example, some researchers are looking at exposing some of the body’s immune system cells, called dendritic cells, to the abnormal PAX-FOXO1 protein that is found in many ARMS cells. The hope is that the dendritic cells will then cause the immune system to attack these cells, no matter where they are in the body.

Eventually, a combination of these approaches may prove to be the best way to treat RMS.

- References
- See all references for Rhabdomyosarcoma