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About Rhabdomyosarcoma

Overview and Types

If you or 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?](#)

Research and Statistics

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

- [Key Statistics for Rhabdomyosarcoma](#)
- [What's New in Rhabdomyosarcoma Research?](#)

What Is Rhabdomyosarcoma?

Cancer starts when cells in the body begin to grow out of control. Cells in nearly any part of the body can become cancer, and can spread to other areas of the body. To learn more about how cancers start and spread, see [What Is Cancer?](#)¹ For information about the differences between childhood cancers and adult cancers, see [Cancer in Children](#)².

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 type of sarcoma made up of cells that normally develop into skeletal (voluntary) muscles. These are muscles that we control to move parts of our body.

Well before birth, 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 very early forms of muscle 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 RMS 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:

- The 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 (ERMS)

ERMS usually affects children in their first 5 years of life, but it can occur at older ages as well.

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 (ARMS)

ARMS typically affects all age groups equally. It makes up a larger portion of RMS in older children, teens, and adults 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.

ARMS tends to grow faster than ERMS, and it usually requires more intense treatment. However, in some cases of ARMS, the cancer cells lack certain [gene changes](#)³, which makes these cancers act more like ERMS (and allows doctors to give less intense treatment).

Anaplastic rhabdomyosarcoma and undifferentiated sarcoma

Anaplastic rhabdomyosarcoma (also called *pleomorphic rhabdomyosarcoma*) is an uncommon type that occurs mainly in adults and 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 and teens, 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.

Hyperlinks

1. www.cancer.org/cancer/cancer-basics/what-is-cancer.html
2. www.cancer.org/cancer/cancer-in-children.html
3. www.cancer.org/cancer/rhabdomyosarcoma/causes-risks-prevention/what-causes.html

References

National Cancer Institute. Childhood Rhabdomyosarcoma Treatment (PDQ®). 2018. Accessed at www.cancer.gov/types/soft-tissue-sarcoma/hp/rhabdomyosarcoma-treatment-pdq on May 21, 2018.

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Wexler LH, Skapek SX, Helman LJ. Chapter 31: Rhabdomyosarcoma. In: Pizzo PA, Poplack DG, eds. *Principles and Practice of Pediatric Oncology*. 7th ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2016.

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Key Statistics for Rhabdomyosarcoma

About 400 to 500 new cases of rhabdomyosarcoma (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.

About 3% of all childhood cancers are RMS. 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 (chest or abdomen).

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 [Survival Rates for Rhabdomyosarcoma by Risk Group](#)¹.

Hyperlinks

1. www.cancer.org/cancer/rhabdomyosarcoma/detection-diagnosis-staging/staging-survival-rates.html

References

American Cancer Society. *Cancer Facts & Figures 2019*. Atlanta, Ga: American Cancer Society; 2019.

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What's New in Rhabdomyosarcoma Research?

The treatment of rhabdomyosarcoma (RMS) has come a long way in the past few

decades, and more patients are surviving RMS than ever before. 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 are helping 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](#)¹ (the *PAX/FOXO1* fusion gene) seen in ARMS. These cancers have been found to act more like embryonal rhabdomyosarcoma (ERMS), which generally needs less intensive treatment than ARMS. Because of this finding, doctors can now give patients with these cancers less intense treatments 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.

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 are described in [Radiation Therapy for Rhabdomyosarcoma](#)².

Other ways to give radiation are also being studied. For example, in **stereotactic body radiation therapy (SBRT)**, a special machine aims very thin beams of radiation at the tumor from many different angles, concentrating it on the tumor very precisely for short periods of time.

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 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 also studying adding **newer chemotherapy drugs** such as irinotecan and temozolomide to the standard [chemotherapy](#)³ regimens for patients who have a higher risk of the tumor recurring.

For patients at a high risk of tumor recurrence, doctors have looked at giving chemotherapy more frequently (such as giving it every 2 weeks instead of every 3 weeks). This concept is called **interval compression**. But so far, it's not clear whether this works better than giving it at standard intervals.

Newer targeted drugs and immunotherapy

[Drugs that target specific parts of cancer cells](#)⁴ (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-1R inhibitors, such as cixutumumab (IMC-1A2) and ganitumab (AMG479)
- Drugs that affect a tumor's ability to make new blood vessels, such as bevacizumab (Avastin), sorafenib (Nexavar), and regorafenib (Stivarga)
- 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 sonidegib (Odomzo)
- Drugs that target other cellular proteins, such as dasatinib (Sprycel)

Researchers are also testing [ways of boosting the body's own immune system](#)⁵ 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.

Hyperlinks

1. www.cancer.org/cancer/rhabdomyosarcoma/causes-risks-prevention/what-

- [causes.html](#)
2. www.cancer.org/cancer/rhabdomyosarcoma/treating/radiation-therapy.html
 3. www.cancer.org/cancer/rhabdomyosarcoma/treating/chemotherapy.html
 4. www.cancer.org/treatment/treatments-and-side-effects/treatment-types/targeted-therapy.html
 5. www.cancer.org/treatment/treatments-and-side-effects/treatment-types/immunotherapy.html

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