Treating Rhabdomyosarcoma

If you or your child has been diagnosed with rhabdomyosarcoma (RMS), your treatment team will discuss the options with you. It's important to weigh the benefits of each treatment option against the possible risks and side effects.

How is rhabdomyosarcoma treated?

- Clinical Trials

Society and is not intended as medical advice to replace the expertise and judgment of your cancer care team. It is intended to help you and your family make informed decisions, together with your doctor. Your doctor may have reasons for suggesting a treatment plan different from these general treatment options. Don't hesitate to ask your cancer care team any questions you may have about your treatment options.

Surgery for Rhabdomyosarcoma

Surgery is an important part of treatment for most rhabdomyosarcomas. Most people with RMS will get two types of surgery:

- The biopsy to diagnose the cancer
- The surgical treatment to remove the tumor(s)

Biopsy surgery

If RMS is suspected, a biopsy is needed to know for sure. The type of biopsy needed will depend on the results of imaging tests, the location and size of the tumor, the patient's age and health, and the expertise of the doctor. How the biopsy is done can affect later treatment, so **it's important that the biopsy is done by a doctor who is experienced in diagnosing and treating RMS**. See Tests for Rhabdomyosarcoma[1] to learn more about biopsies.

Surgery to remove the tumor

Unless it is clear that the cancer has spread to distant parts of the body, **surgery is usually the first step in treating RMS**. Complete resection (removal) of the main tumor, along with some surrounding normal tissue, is the goal whenever possible. If there are cancer cells at the edges (margins) of the removed specimen (meaning that some cancer cells may have been left behind), the surgeon may operate again to try to remove the remaining cancer.

In some cases, surgery may be done even if it's clear that all of the cancer can't be removed, because it may still help other treatments (chemotherapy and radiation) to

work better.

During surgery, nearby lymph nodes might be biopsied to determine if the cancer has spread to these areas, especially if:

- The main tumor is near the testicles in a boy who is 10 years of age or older
- The main tumor is on an arm or leg

Some types of surgery might need to be done by special surgeons. For example, removing tumors in the head and neck area may require surgical teams with ENT (ear, nose, and throat) surgeons, plastic surgeons, maxillofacial surgeons, and neurosurgeons.

If a tumor is large or is in a spot where removing it completely would severely affect the patient’s appearance or cause other problems, then surgery may be delayed until after chemotherapy and possibly radiation therapy to try to shrink it, or surgery might not be done at all (and radiation will be used instead).

What to expect with surgery

The type and extent of surgery can vary a great deal based on the location and size of the tumor. RMS can appear in many parts of the body, so it’s not possible to describe here all of the different types of operations that might be done. The surgical team will discuss the planned surgery with you, but make sure you ask questions if there are any parts of it that aren’t clear to you.

If the diagnosis of RMS wasn't confirmed by a biopsy before the main operation, the surgeon may first take only a small sample of the tumor. The sample is checked right away to see if it is cancer or not. If it can be determined that it is cancer while the surgery is still going on, the surgeon may try to remove the entire tumor and also remove some of the nearby lymph nodes to check for spread of the cancer. If the surgeon suspects the disease has spread to another part of the body, a piece of the possible metastatic tumor may be

reduce the risk of problems such as infections.

Possible risks and side effects

Short-term risks and side effects: Depending on where the tumor is, surgery for RMS can be a long and complex operation. Serious short-term side effects are not common, but they can include reactions to anesthesia, excess bleeding, blood clots, and infections. Pain is common after the operation, and the patient might need strong pain medicines for a while after surgery as the site heals.

Long-term side effects: The long-term side effects of surgery depend mainly on where the tumor is and what type of operation is done. Physical changes after surgery can range from little more than a scar to changes in appearance or in how some parts of the body function, which may require physical rehabilitation.

More information about Surgery

For more general information about surgery as a treatment for cancer, see Cancer Surgery[4].

To learn about some of the side effects listed here and how to manage them, see Managing Cancer-related Side Effects[5].

Hyperlinks

1. www.cancer.org/cancer/types/rhabdomyosarcoma/detection-diagnosis-staging/how-diagnosed.html
2. www.cancer.org/cancer/types/rhabdomyosarcoma/detection-diagnosis-staging/how-diagnosed.html
3. www.cancer.org/cancer/managing-cancer/making-treatment-decisions/tubes-lines-ports-catheters.html
4. www.cancer.org/cancer/managing-cancer/treatment-types/surgery.html
5. www.cancer.org/cancer/managing-cancer/side-effects.html

References

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

treatment-pdq on June 4, 2018.

Okcu MF, Hicks J. Rhabdomyosarcoma in childhood and adolescence: Treatment. UpToDate. Accessed at www.uptodate.com/contents/rhabdomyosarcoma-in-childhood-adolescence-and-adulthood-treatment on June 4, 2018.

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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Chemotherapy for Rhabdomyosarcoma

Chemotherapy (chemo) is the use of drugs to treat cancer. Chemo is *systemic* therapy, meaning that the drugs enter the bloodstream and go throughout the body to destroy cancer cells. This makes chemo useful for killing cancer cells that have spread to other parts of the body, even if they can't be seen.

Chemo is an important part of treatment for rhabdomyosarcoma (RMS). Even if it appears that all of the cancer was removed by surgery, without chemo it is likely to come back.

After surgery, any tiny deposits of RMS that are still in the body can often be destroyed by chemo. If larger areas of tumor remain after surgery (or if surgery couldn't be done for some reason), chemo (along with radiation) can often shrink these areas. In some cases it may shrink the tumor enough that surgery can remove the remaining tumor completely.

Chemo drugs used to treat rhabdomyosarcoma

Doctors give chemo in cycles, which is usually treatment on 1 or 2 days in a row, followed by days off to give the body time to recover. For RMS, chemo is typically given

once a week for the first few months, and then less often. The total length of treatment usually ranges from 6 months to a year.

Some drugs can be taken by mouth, but most are given IV (injected into a vein).

A combination of chemo drugs is used to treat patients with RMS. Which drugs are used will often depend on which risk group[1] the patient is in.

For people in the **low-risk group**, the main combinations of drugs used are:

- **VA:** vincristine and dactinomycin (also known as *actinomycin-D*)
- **VAC:** vincristine, dactinomycin, and cyclophosphamide

For the **intermediate-risk group**, the most common regimens are:

- **VAC:** vincristine, dactinomycin, and cyclophosphamide
- **VAC/VI:** vincristine, dactinomycin, and cyclophosphamide, alternating with vincristine and irinotecan

Doctors are also studying whether adding the targeted drug[2] temsirolimus to the VAC/VI regimen might help it work better.

For people in the **high-risk group** (which includes those with metastatic disease), the **VAC** regimen is the most common one used. Because these cancers can be hard to treat, doctors have also studied the use of more intense chemo that includes several other drugs (such as doxorubicin, ifosfamide, and etoposide). Another approach that has been studied is to give higher doses of chemo, sometimes followed by a stem cell transplant. But so far it's not clear that either of these approaches is any better than standard chemo, and they can cause more side effects.

Most doctors recommend that people in the high-risk group be treated in a clinical trial[3] testing new drugs and drug combinations. It is hoped that newer drugs will help people in the high-risk group live longer.

Possible side effects

Chemo drugs can affect cells other than cancer cells, which can lead to side effects. The side effects depend on the type and doses of drugs, and the length of time they are given.

Children tend to have less severe side effects from chemo than adults and often recover from side effects more quickly. This is why doctors can often give them higher doses of chemo to kill the tumor.

General side effects: Side effects common to many chemo drugs include:

- Hair loss
- Mouth sores
- Loss of appetite
- Nausea and vomiting
- Diarrhea
- Increased chance of infections (from having too few white blood cells)
- Easy bruising or bleeding (from having too few blood platelets)
- Fatigue (from having too few red blood cells)

Most of these side effects tend to go away once treatment is finished. There are often ways to lessen these side effects. For example, drugs can be given to help prevent or reduce nausea and vomiting. Be sure to ask your doctor or nurse about medicines to help reduce side effects, and report any side effects your child has so they can be managed effectively.

Side effects of certain drugs: General sidl

Treatment for Rhabdomyosarcoma?[8]

More information about chemotherapy

For more general information about how chemotherapy is used to treat cancer, see Chemotherapy[9].

To learn about some of the side effects listed here and how to manage them, see Managing Cancer-related Side Effects[10].

Hyperlinks

1. www.cancer.org/cancer/types/rhabdomyosarcoma/detection-diagnosis-staging/staging.html
2. www.cancer.org/cancer/managing-cancer/treatment-types/targeted-therapy.html
3. www.cancer.org/cancer/managing-cancer/making-treatment-decisions/clinical-trials.html
4. www.cancer.org/cancer/managing-cancer/side-effects/pain/peripheral-neuropathy.html

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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Radiation Therapy for Rhabdomyosarcoma

Radiation therapy uses high-energy radiation to kill cancer cells. It is often an effective way to kill cancer cells that can't be removed with surgery. When radiation therapy is

How radiation therapy is done

This type of treatment is given by a doctor called a **radiation oncologist**. Before

studies suggest that this may be a good way to preserve the function of these organs in many children.

Other newer techniques, such as stereotactic radiotherapy and proton beam radiotherapy, are discussed briefly in What's New in Rhabdomyosarcoma Research?[3]

Possible side effects

The side effects of radiation therapy depend on where the radiation is aimed, the dose of radiation, and the person's age. (Young children are much more likely to be affected by radiation.) Some side effects are likely to last a short time, while others might last longer.

Short-term side effects can include:

- Fatigue
- Increased risk of infections
- Effects on the skin in areas that receive radiation, ranging from hair loss and mild sunburn-like changes to more severe skin reactions
- Nausea, vomiting, and diarrhea (from radiation to the abdomen or pelvis)
- Damage to the bladder, which might cause urinary problems (from radiation to the abdomen or pelvis)
- Mouth sores and loss of appetite (from radiation to the head and neck area)

Long-term side effects can be more serious, especially in growing children, so doctors try to limit them as much as possible.

Small children's brains are very sensitive to radiation, so doctors try to avoid using radiation to the head whenever possible. If it is needed, it is aimed very carefully to tryfrom radiation to

To limit the risk of serious long-term effects from radiation, doctors use the lowest dose of radiation therapy that is still effective.

More information about radiation therapy

To learn more about how radiation is used to treat cancer, see Radiation Therapy[5].

To learn about some of the side effects listed here and how to manage them, see Managing Cancer-related Side Effects[6].

Hyperlinks

High-Dose Chemotherapy and Stem Cell Transplant for Rhabdomyosarcoma

- More information about stem cell transplant

A stem cell transplant (sometimes referred to as a *bone marrow transplant*) makes it possible to use much higher doses of chemotherapy (chemo) than would normally be possible. Chemo drugs kill rapidly dividing normal cells (such as those in the bone marrow, where new blood cells are made) as well as cancer cells. Higher doses of these drugs might be more effective in treating some cancers, but they can't be given because the severe damage to the bone marrow would cause life-threatening shortages of blood cells.

A stem cell transplant can get around this problem by taking out and saving some of the patient's own blood-forming stem cells (either from the blood or bone marrow) before high-dose chemo and then putting them back into the blood after chemo is over. The stem cells then travel to the bone marrow, which lets the normal marrow regrow.

Stem cell transplants are used to treat some aggressive childhood cancers, but so far it's not clear if they can help rhabdomyosarcoma patients. Because of the severe side effects they can cause, most doctors recommend they be used only as part of a clinical trial[1] at this time.

More information about stem cell transplant

To learn more about stem cell transplants, including how they are done and their potential side effects, see Stem Cell Transplant for Cancer[2].

For more general information about side effects and how to manage them, see Managing Cancer-related Side Effects[3].

Hyperlinks

1. www.cancer.org/cancer/managing-cancer/making-treatment-decisions/clinical-trials.html
2. www.cancer.org/cancer/managing-cancer/treatment-types/stem-cell-

transplant.html
3. www.cancer.org/cancer/managing-cancer/side-effects.html

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National Cancer Institute. Childhood Rhabdomyosarcoma Treatment (PDQ®). 2018. Accessed at www.cancer.gov/types/soft-tissue-sarcoma/hp/rhabdomyosarcoma-treatment-pdq on June 4, 2018.

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Rhabdomyosarcoma That Progresses or Recurs After Initial Treatment

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can be done. If radiation therapy wasn't part of the initial treatment, it may be used as well.

In rare cases, surgery may be used for cancers that recur in other parts of the body, such as if there is a small recurrence in a lung. Radiation therapy might be another option here as well.

Most often, chemotherapy is the best option if the cancer has spread to other parts of the body. This might include some of the drugs listed in Chemotherapy for Rhabdomyosarcoma, as well as newer drugs now being studied.

Because these tumors are often hard to treat, clinical trials[1] of newer treatments may be a good option in many cases.

Hyperlinks

1. www.cancer.org/cancer/managing-cancer/making-treatment-decisions/clinical-trials.html

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