Treating Wilms Tumor

General treatment information

Overall, about 9 of 10 children with Wilms tumor are cured. A great deal of progress has been made in treating this disease. Much of this progress in the United States has been because of the work of the National Wilms Tumor Study Group (now part of the Children’s Oncology Group), which runs clinical trials of new treatments for children with Wilms tumor. Today, most children with this cancer are treated in a clinical trial to try to improve on what doctors believe is the best treatment. The goal of these studies is to find ways to cure as many children as possible while limiting side effects by giving as little treatment as needed.

Because Wilms tumors are rare, few doctors outside of those in children’s cancer centers have much experience in treating them. Children with Wilms tumors are treated with a team approach that includes the child’s pediatrician as well as specialists at a child’s cancer center. For Wilms tumors, the doctors on this team often include:

- A pediatric surgeon or pediatric urologist (doctor who treats urinary system problems in children [and genital problems in boys])
- A pediatric oncologist (doctor who uses chemotherapy and other medicines to treat childhood cancers)
- A pediatric radiation oncologist (doctor who uses radiation therapy to treat cancer in children)

Many other specialists may be involved in your child’s care as well, including other doctors, physician assistants (PAs), nurse practitioners (NPs), nurses, psychologists, social workers, rehabilitation specialists, and other health professionals. Having a child go through cancer treatment often means meeting lots of specialists and learning about parts of the medical system you probably haven’t been exposed to before. For more on this, see Children Diagnosed With Cancer: Understanding the Health Care System.

After your child’s tumor is found and its stage and histology are determined, the cancer
care team will discuss treatment options with you. It's important to discuss all of the options as well as their possible side effects with your child’s doctors so you can make an informed decision. (For a list of some questions to ask, see What should you ask your child’s doctor about Wilms tumor?)

If time allows, it can often be helpful to get a second opinion if you have questions about the recommended plan (or if you just want to confirm that it’s the best option). This can provide you with more information and help you feel more confident about the treatment plan you choose.

The main types of treatment for Wilms tumor are:

- Surgery
- Chemotherapy
- Radiation therapy

Most children will get more than one type of treatment.

In the United States, surgery is the first treatment for most Wilms tumors. In Europe, doctors often prefer to give a short course of chemotherapy before the surgery. There seems to be no difference in the results from these 2 approaches.

The first goal of treatment is to remove the primary (main) tumor, even if the cancer has spread to distant parts of the body. Sometimes the tumor might be hard to remove because it is very large, it has spread into nearby blood vessels or other vital structures, or it’s in both kidneys. For these children, doctors might use chemotherapy, radiation therapy, or a combination of the 2 to try to shrink the tumor(s) before surgery.

If any cancer is left after surgery, radiation therapy or more surgery may be needed.

**Thinking about taking part in a clinical trial**

Clinical trials are carefully controlled research studies that are done to get a closer look at promising new treatments or procedures. Clinical trials are one way to get state-of-the art cancer treatment. In some cases, they may be the only way to get access to newer treatments. They are also the best way for doctors to learn better methods to treat cancer. Still, they are not right for everyone.

If you would like to learn more about clinical trials that might be right for your child, start by asking the doctor if your clinic or hospital conducts clinical trials. You can also call our clinical trials matching service at 1-800-303-5691 for a list of studies that meet your child's medical needs, or see our Clinical Trials page.
Considering complementary and alternative methods

You may hear about alternative or complementary methods that your doctor hasn’t mentioned to treat your child's cancer or relieve symptoms. These methods can include vitamins, herbs, and special diets, or other methods such as acupuncture or massage, to name a few.

Complementary methods refer to treatments that are used along with regular medical care. Alternative treatments are used instead of a doctor’s medical treatment. Although some of these methods might be helpful in relieving symptoms or helping your child feel better, many have not been proven to work. Some might even be dangerous.

Be sure to talk to your child's cancer care team about any method you are thinking about using. They can help you learn what is known (or not known) about the method, which can help you make an informed decision.

Help getting through cancer treatment

The cancer care team will be your first source of information and support, but there are other resources for help when you need it. Hospital- or clinic-based support services are an important part of your child’s care and your care, too. These might include nursing or social work services, financial aid, nutritional advice, rehab, or spiritual help.

The American Cancer Society also has programs and services – including rides to treatment, lodging, support groups, and more – to help you get through treatment. Call our National Cancer Information Center at 1-800-227-2345 and speak with one of our trained specialists on call 24 hours a day, every day.

The treatment information given here is not official policy of the American Cancer 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 him or her questions about your treatment options.

Surgery for Wilms Tumors

Surgery is the main treatment for nearly all children with Wilms tumor. It should be done by a surgeon who specializes in operating on children and has experience in treating these cancers.
Removing the tumor

The main goal of surgery is to remove the entire Wilms tumor in one piece to keep the cancer cells from possibly spreading in the abdomen (belly). Surgeons who operate on these tumors are careful to limit the chance of this type of cancer spread whenever possible. If the surgeon finds (either with imaging tests done before surgery, or when starting the operation) that the entire tumor can't be removed safely, other treatments may be used first. If these treatments shrink the tumor enough, surgery can then be done more safely.

Depending on the situation, different operations might be used.

**Radical nephrectomy:** This is the most common surgery for a Wilms tumor that's only in one kidney, as it provides the best chance of making sure all of the tumor is removed. During this operation, the surgeon makes an incision (cut – usually down the middle of the abdomen) and removes the cancer along with the whole kidney, the fatty tissue around the kidney, the ureter (tube that carries urine from the kidney to the bladder), and the attached adrenal gland that sits on top of the kidney. Most children do very well with only one kidney.

**Partial nephrectomy (nephron-sparing surgery):** In the small number of children who have Wilms tumors in both kidneys, the surgeon will try to save some normal kidney tissue, if possible. The surgeon may remove the kidney containing the most tumor with a radical nephrectomy. In the other kidney the surgeon may do a partial nephrectomy, removing just the tumor and a margin of normal kidney around it. Another option might be partial nephrectomies on both kidneys.

Sometimes, both kidneys need to be removed completely. The child would then need dialysis several times a week. In this procedure, a machine does the job of the kidneys by filtering waste products out of the blood. Once the child is healthy enough, a kidney transplant may be an option if a donor kidney becomes available.

**Assessing the extent of the disease (surgical exploration)**

Another main goal of surgery (radical or partial nephrectomy) is to determine the extent of the cancer and whether or not it can all be removed. Lymph nodes near the kidney will be removed during surgery to look for cancer cells in them. Cancer often spreads to lymph nodes (bean-sized collections of immune cells). Lymph node removal is known as a *regional lymphadenectomy*.

The other kidney and nearby organs such as the liver may also be looked at closely,
and any suspicious areas biopsied (samples taken to be checked for cancer under a microscope).

Knowing if a Wilms tumor has spread to the lymph nodes, the other kidney, or other nearby organs is important in determining its stage and further treatment options.

**Placing a central venous access device (port)**

Often, if the child is going to get chemotherapy, a surgeon will insert a small plastic tube called a catheter into a large blood vessel – usually under the collar bone. This tube may be called a venous access device, central venous catheter, or just a port. The tube might be put in during the surgery to remove the tumor, or as a separate operation (especially if chemo is going to be given before the surgery).

**Possible risks and side effects of surgery**

Surgery to remove a Wilms tumor is a serious operation, and surgeons are very careful to try to limit any problems either during or after surgery. Complications during surgery, such as bleeding, injuries to major blood vessels or other organs, or reactions to anesthesia, are rare, but they can happen.

Almost all children will have some pain for a while after the operation, although this can usually be helped with medicines if needed. Other problems after surgery are not common but can include internal bleeding, blood clots, infections, or problems with food moving through the intestines.

Most children do well when only one kidney is removed. But if there are tumors in both kidneys, another concern is the loss of kidney function. In these cases, doctors must balance between making sure the tumors are removed completely and removing only as much of the kidney(s) as is needed. Children who have all or parts of both kidneys removed may need dialysis, and may eventually need a kidney transplant.

* References

See all references for Wilms Tumor

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Chemotherapy (chemo) uses anti-cancer drugs that are given into a vein or by mouth (in pill form). These drugs enter the blood and reach all areas of the body, which makes this treatment useful for cancer that has spread or might have spread beyond the kidney.

Most children with Wilms tumors will get chemo at some point during their treatment. In the United States, chemo is usually given after surgery. Sometimes it may be needed before surgery to shrink a tumor to make the operation possible. In Europe, chemo is given before surgery and continued afterward. In both cases, the type and amount of chemo depend on the stage and histology of the cancer.

A combination of chemo drugs is used to treat children with Wilms tumors. The chemo drugs used most often are actinomycin D (dactinomycin) and vincristine. For tumors at more advanced stages, those with unfavorable histology, or tumors that recur (come back) after treatment, other drugs such as doxorubicin (Adriamycin), cyclophosphamide, etoposide, irinotecan, and/or carboplatin may also be used.

These drugs are injected into a vein or into a venous access device. Different drugs, doses, and lengths of treatment are used, depending on the type and stage of the Wilms tumor and the child’s age. Most often, the drugs are given once a week for at least several months. They are usually given by a nurse in the doctor’s office or in the outpatient section of the hospital. Some children with Wilms tumors might need to stay in the hospital while they are getting chemo, but usually this is not needed.

Possible side effects of chemotherapy

Chemo drugs attack cells that are dividing quickly, which is why they often work against cancer cells. But other cells in the body, such as those in the bone marrow (where new blood cells are made), the lining of the mouth and intestines, and the hair follicles, also divide quickly. These cells are also likely to be affected by chemo, which can lead to side effects.

The side effects of chemo depend on the types and doses of drugs used, and the length of treatment. Possible short-term side effects can include:

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

Your child’s doctor and treating team will watch closely for any side effects that develop. 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 child’s doctor or nurse about medicines to help reduce side effects, and let them know if your child has side effects so they can be managed.

Along with the effects listed above, some drugs can have specific side effects. For example:

- Vincristine can damage nerves. Some patients may have tingling, numbness, weakness, or pain, particularly in the hands and feet.
- Doxorubicin can damage the heart. The risk of this happening goes up as the total amount of the drug given goes up. Doctors try to limit this risk as much as possible by not giving more than the recommended doses and by checking the heart with a test called an echocardiogram (an ultrasound of the heart) during treatment.
- Cyclophosphamide can damage the bladder, which can cause blood in the urine. The risk of this can be lowered by giving the drug with plenty of fluids and with a drug called mesna, which helps protect the bladder.

**Lab tests to check for chemo side effects**

Before each chemo session, your child’s doctor will do blood tests to see how well the liver, kidneys, and bone marrow are working. If there are problems, chemo might need to be delayed or the doses reduced.

**Long-term side effects of chemo**

Possible long-term effects of treatment are one of the major challenges children might face after cancer treatment.

For example, if your child is given doxorubicin (Adriamycin), there is a chance it could damage the heart. Your child’s doctor will carefully watch the doses used and will check your child’s heart function with imaging tests.

Some chemo drugs can increase the risk of developing a second type of cancer (such as leukemia) years after the Wilms tumor is cured. But this small increase in risk has to
be weighed against the importance of chemo in treating Wilms tumor. Some drugs might also affect fertility (the ability to have children) years later.


- References

See all references for Wilms Tumor

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Radiation Therapy for Wilms Tumor

Radiation therapy uses high-energy rays or particles to kill cancer cells. It is usually part of treatment for more advanced Wilms tumors (stages III, IV, and V) and for some earlier stage tumors with unfavorable histology.

The type of radiation used for Wilms tumors, known as external beam radiation therapy, focuses radiation from outside the body on the cancer. The total dose of radiation is divided into fractions, usually given 5 days a week for a couple of weeks.

Before treatments start, the radiation team takes careful measurements with imaging tests such as CT or MRI scans to determine the correct angles for aiming the radiation beams and the proper dose of radiation. Your child may be fitted with a plastic mold that looks like a body cast to keep him or her in the same position during each treatment so that the radiation can be aimed more accurately.

Each treatment is much like getting an x-ray, although the dose of radiation is much higher. For each session, your child lies on a special table while a machine delivers the radiation from precise angles. Each session lasts about 15 to 30 minutes, with most of the time being spent making sure the radiation is aimed correctly. The actual treatment time is much shorter. The treatment is not painful, but some younger children may be given medicine to make them drowsy or asleep before each treatment to help make sure they stay still.
Newer radiation techniques

Some techniques help doctors aim the treatment at the tumor more accurately while reducing the radiation exposure to nearby healthy tissues. These techniques may help increase the success rate and reduce side effects.

Three-dimensional conformal radiation therapy (3D-CRT): 3D-CRT uses the results of imaging tests such as MRI and special computers to precisely map the location of the tumor. Several radiation beams are then shaped and aimed at the tumor from different directions. Each beam alone is fairly weak, which makes it less likely to damage normal body tissues, but the beams converge at the tumor to give a higher dose of radiation there.

Intensity modulated radiation therapy (IMRT): IMRT is an advanced form of 3D therapy. Along with shaping the beams and aiming them at the tumor from several angles, the intensity (strength) of the beams can be adjusted to limit the dose reaching the nearby normal tissues. This lets the doctor deliver a higher dose to the tumor. Many major hospitals and cancer centers now use IMRT.

Possible side effects of radiation therapy

Radiation is often an important part of treatment, but young children’s bodies are very sensitive to it, so doctors try to use as little as possible to help avoid or limit any problems. Radiation therapy can cause both short-term and long-term side effects, which depend on the dose of radiation and where it’s aimed.

Possible short-term effects:

- Effects on skin areas that receive radiation can range from mild sunburn-like changes and hair loss to more severe skin reactions.
- Radiation to the abdomen (belly) can cause nausea or diarrhea.
- Radiation therapy can make a child tired, especially after several days or weeks of treatment.

Possible long-term effects:

- Radiation can slow the growth of normal body tissues (such as bones) that get radiation, especially in younger children. In the past this led to problems such as short bones or a curving of the spine, but this is less likely with the lower doses of radiation used today.
- Radiation that reaches the chest area can affect the heart and lungs. This does not
usually cause problems right away, but in some children it might lead to heart or lung problems as they get older.

- In girls, radiation may damage the ovaries. This might lead to abnormal menstrual cycles or problems getting pregnant or having children later on.
- Radiation slightly increases the risk of developing a second cancer in the area, usually many years after it is given. This doesn’t happen often with Wilms tumors because the amount of radiation used is low.

See the section What happens after treatment for Wilms tumor? for more on the possible long-term effects of treatment.

- References
  See all references for Wilms Tumor

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Treatment by Type and Stage of Wilms Tumor

In the United States, most children with Wilms tumors are treated in clinical trials developed by the Children’s Oncology Group. The goal of these studies is to cure as many children as possible while limiting side effects by giving as little treatment as is necessary. This is done by comparing the current best treatment with one the doctors think might be better. Because of this, treatment may differ slightly from what is described here.

Treatment for Wilms tumor is based mainly on the stage of the cancer and whether its histology (appearance under the microscope) is favorable or unfavorable (anaplastic). In the United States, doctors prefer to use surgery as the first treatment in most cases, and then give chemotherapy (and possibly radiation therapy) afterward. In Europe, doctors prefer to start the chemotherapy before surgery. The results seem to be about the same.

Most often, the stage and histology of the cancer are actually determined when surgery
is done to remove the cancer, because the true extent of the tumor often can’t be determined by imaging tests alone. The findings from surgery are then used to guide further treatment. But sometimes it’s clear that the cancer has already spread beyond the kidney even before surgery is done, based on imaging tests. This can affect the order in which treatments are given, as well as the extent of surgery.

Stage I

These tumors are only in the kidney. Surgery has completely removed the tumor along with the entire kidney, nearby structures, and some nearby lymph nodes.

Favorable histology: Children younger than 2 years with small tumors (weighing less than 550 grams) may not need further treatment, such as chemo. But they need to be watched closely because the chance the cancer will come back is slightly higher than if they also got chemo. If the cancer does come back, the chemo drugs actinomycin D (dactinomycin) and vincristine (and possibly more surgery) are very likely to be effective at this point.

For children older than 2 and for those of any age who have larger tumors, surgery is usually followed by chemo with actinomycin D and vincristine. If the tumor cells have certain chromosome changes, the drug doxorubicin (Adriamycin) may be given as well. The chemo is given for several months.

Unfavorable histology: For children of any age who have tumors with unfavorable histology, surgery is usually followed by radiation therapy to the area of the tumor, along with chemo with actinomycin D, vincristine, and doxorubicin (Adriamycin) for several months.

Stage II

These tumors have grown outside the kidney into nearby tissues, but surgery removed all visible signs of cancer.

Favorable histology: After surgery, standard treatment is chemo with actinomycin D and vincristine. If the tumor cells have certain chromosome changes, the drug doxorubicin (Adriamycin) may be given as well. The chemo is given for several months.

Unfavorable histology with focal (only a little) anaplasia: When the child recovers from surgery, radiation therapy is given over several weeks. When this is finished, chemo (doxorubicin, actinomycin D, and vincristine) is given for about 6 months.
Unfavorable histology with diffuse (widespread) anaplasia: After surgery, these children get radiation over several weeks. This is followed by a more intense type of chemo using the drugs vincristine, doxorubicin, etoposide, cyclophosphamide, and carboplatin, along with mesna (a drug that helps protect the bladder from the effects of cyclophosphamide), which is given for about 6 months.

Stage III

Surgery cannot remove these tumors completely because of their size or location or for other reasons. In some cases, surgery may be postponed until other treatments are able to shrink the tumor first (see below).

Favorable histology: Treatment is usually surgery if it can be done, followed by radiation therapy over several days. This is followed by chemo with 3 drugs (actinomycin D, vincristine, and doxorubicin). If the tumor cells have certain chromosome changes, the drugs cyclophosphamide and etoposide may be given as well. Chemo is given for about 6 months.

Unfavorable histology with focal (only a little) anaplasia: Treatment starts with surgery if it can be done, followed by radiation therapy over several weeks. This is followed by chemo, usually with 3 drugs (actinomycin D, vincristine, and doxorubicin) for about 6 months.

Unfavorable histology with diffuse (widespread) anaplasia: Treatment starts with surgery if it can be done, followed by radiation therapy over several weeks. This is followed by chemo, usually with the drugs vincristine, doxorubicin, etoposide, cyclophosphamide, and carboplatin, along with mesna (a drug that helps protect the bladder from the effects of cyclophosphamide). Chemo lasts about 6 months.

In some instances the tumor may be very large or may have grown into nearby blood vessels or other structures so that it can’t be removed safely. In these children, a small biopsy sample is taken from the tumor to be sure that it’s a Wilms tumor and to determine its histology. Then chemo is started. Usually the tumor will shrink enough within several weeks so that surgery can be done. If not, then radiation therapy might be given as well. Chemo will be started again after surgery. If radiation was not given before surgery, it’s given after surgery.

Stage IV

These tumors have already spread to distant parts of the body at the time of diagnosis. As with stage III tumors, surgery to remove the tumor might be the first treatment, but it
might need to be delayed until other treatments can shrink the tumor (see below).

**Favorable histology:** Surgery to remove the tumor is the first treatment if it can be done, followed by radiation therapy. The entire abdomen will be treated if there is still some cancer left after surgery. If the cancer has spread to the lungs, low doses of radiation might also be given to that area. This is followed by chemo, usually with 3 drugs (actinomycin D, vincristine, and doxorubicin) for about 6 months. If the tumor cells have certain chromosome changes, the drugs cyclophosphamide and etoposide may be given as well.

**Unfavorable histology:** Treatment might start with surgery if it can be done, followed by radiation therapy. The entire abdomen will be treated if there is still some cancer left after surgery. Low doses of radiation will also be given to the lungs if the cancer has spread there. This is followed by chemo with the drugs vincristine, doxorubicin, etoposide, cyclophosphamide, and carboplatin, along with mesna given for about 6 months. If the tumor cells have diffuse (widespread) anaplasia, some doctors might try the chemo drugs irinotecan and vincristine first instead (although this is not yet a commonly used treatment). The treatment would then be adjusted if the tumor shrinks in response to these drugs.

If the tumor is too large or has grown too much to be removed safely with surgery first, a small biopsy sample may be taken from the tumor to be sure that it’s a Wilms tumor and to determine its histology. Chemo and/or radiation therapy may then be used to shrink the tumor. Surgery might be an option at this point. This would be followed by more chemo and radiation therapy if it wasn’t given already.

For stage IV cancers that have spread to the liver, surgery may be an option to remove any liver tumors that still remain after chemo and radiation therapy.

**Stage V**

Treatment for children with tumors in both kidneys is unique for each child, although it typically includes surgery, chemo, and radiation therapy at some point.

**Biopsies** (tissue samples) of tumors in both kidneys and of nearby lymph nodes may be taken first, although not all doctors feel this is needed because when both kidneys have tumors, the chance that they are Wilms tumors is very high.

Chemo is typically given first to try to shrink the tumors. The drugs used will depend on the extent and histology (if known) of the tumors. After about 6 weeks of chemo, surgery (partial nephrectomy) may be done to remove the tumors if enough normal kidney
tissue can be left behind. If the tumors haven’t shrunk enough, treatment may include more chemo or radiation therapy for about another 6 weeks. Surgery (either partial or radical nephrectomy) may then be done. This is followed by more chemo, possibly along with radiation therapy if it hasn’t been given already.

If not enough functioning kidney tissue is left after surgery, a child may need dialysis, a procedure where a special machine filters waste products out of the blood several times a week. If there is no evidence of any cancer after a year or two, a donor kidney transplant may be done.

**Recurrent Wilms tumor**

The prognosis and treatment for children with Wilms tumor that recurs (comes back after treatment) depends on their prior treatment, the cancer’s histology (favorable or unfavorable), and where it recurs. The outlook is generally better for recurrent Wilms tumors with the following features:

- Favorable histology
- Initial diagnosis of stage I or II
- Initial chemo with vincristine and actinomycin D only
- No previous radiation therapy

The usual treatment for these children is surgery to remove the recurrent cancer (if possible), radiation therapy (if not already given to the area), and chemo, often with drugs different from those used during first treatment.

Recurrent Wilms tumors that do not have the features above are much harder to treat. These children are usually treated with aggressive chemo, such as the ICE regimen (ifosfamide, carboplatin, and etoposide) or others being studied in clinical trials. Very high-dose chemo followed by a stem cell transplant (sometimes called a bone marrow transplant) might also be an option in this situation.

- **References**
  See all references for Wilms Tumor