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# **Treating Wilms Tumor**

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

#### How is Wilms tumor treated?

Overall, about 9 of 10 children with Wilms tumors are cured. A great deal of progress has been made in treating this cancer in recent decades. Much of this progress is the result of children with Wilms tumors taking part in <u>clinical trials</u> of new treatments.

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.

Most children with Wilms tumors will get more than one type of treatment. The main types of treatment are:

- Surgery for Wilms Tumors
- Chemotherapy for Wilms Tumors
- Radiation Therapy for Wilms Tumors

#### **Common treatment approaches**

In the United States and Canada, surgery is the first treatment for most Wilms tumors. In Europe and some other parts of the world, doctors often prefer to give a short course of chemotherapy before the surgery. There seems to be no difference in the results

The first goal of treatment is to remove the main tumor in the kidney, even if the cancer has spread to other parts of the body.

Sometimes the tumor might be hard to remove because it is very large, it has grown into nearby blood vessels or other vital structures, or it's in both kidneys. For children with these tumors, doctors might use chemotherapy, radiation therapy, or a combination of these to try to shrink the tumor(s) before surgery.

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

Treatment of Wilms Tumor, By Type and Stage

#### Who treats Wilms tumors?

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 typically treated by a team of specialists. The doctors on this team often include:

- A **pediatric surgeon** or **pediatric urologist** (a surgeon who treats urinary system problems in children [and genital problems in boys])
- A **pediatric oncologist** (a doctor who uses chemotherapy and other medicines to treat children with cancer)
- A pediatric radiation oncologist (a 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, nurses, nurse practitioners (NPs), physician assistants (PAs), psychologists, social workers, rehabilitation specialists, and other health professionals.

- How to Find the Best Cancer Treatment for Your Child
- Navigating the Health Care System When Your Child Has Cancer

#### **Making treatment decisions**

Treatment for Wilms tumors can often be effective, but it can also cause serious side effects. It's important to discuss all treatment options and their possible side effects with your child's cancer care team so you can make an informed decision.

Treatment options depend on several factors, including:

- The type, stage, and histology of the Wilms tumor
- The child's age
- If the tumor cells have certain chromosome changes
- Possible side effects
- The child and family's preferences

Take time to learn about all of your child's treatment options. Be sure to ask questions about anything that is unclear. Also, talk to your child's cancer care team about the goals of each treatment and what you and your child can expect during treatment.

The treatment team will also help you manage side effects. They can help you work closely with nutritionists, psychologists, social workers, and other professionals to understand and deal with medical problems, stress, and other issues related to treatment.

If time allows, getting a second opinion from another doctor experienced with your child's type of cancer is often a good idea. This can give you more information and help you feel more confident about the treatment plan you choose. If you aren't sure where to go for a second opinion, ask your doctor for help.

- Questions to Ask Your Child's Doctor About Wilms Tumor
- How to Talk to Your Child's Cancer Care Team
- Seeking a Second Opinion

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

Today, most children and teens with cancer are treated at specialized children's cancer centers. These centers offer the most up-to-date-treatment by conducting clinical trials (studies of promising new therapies). Children's cancer centers often conduct many clinical trials at any one time, and in fact most children treated at these centers take part in a clinical trial as part of their treatment. Clinical trials are one way to get state-of-the art cancer treatment. Sometimes they may be the only way to get access to newer treatments (although there is no guarantee that newer treatments will be better). They are also the best way for doctors to learn better methods to treat these cancers. Still, they might not be right for everyone. If you would like to learn more about clinical trials that might be right for your child, start by asking the treatment team if your clinic or hospital conducts clinical trials.

Clinical Trials

#### Considering complementary and alternative methods

You may hear about alternative or complementary methods that your doctor hasn't mentioned to treat your child's tumor 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 your regular medical care. Alternative treatments are used instead of standard medical treatment. Although some of these methods might be helpful in relieving symptoms or helping people feel better, many have not been proven to work. Some might even be harmful. 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.

Complementary and Integrative Medicine

#### Preparing for treatment

Before treatment, the doctors and other members of the team will help you, as a parent, understand the tests that will need to be done. The team's social worker will also counsel you about the problems you and your child might have during and after treatments such as surgery, and might be able to help you find housing and financial aid if needed.

When Your Child Has Cancer

#### Help getting through cancer treatment

Your child's 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 can also be an important part of your care. These might include nursing or social work services, financial aid, nutritional advice, rehab, or spiritual help. For children and teens with cancer and their families, other specialists can be an important part of care as well. The American Cancer Society also has programs and services – including rides to treatment, lodging, 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.

- Finding Help and Support When Your Child Has Cancer
- Programs & Services

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 your cancer care team any questions you may have about your treatment options.

# **Surgery for Wilms Tumors**

Surgery is the main treatment for nearly all children with Wilms tumors. It's important that it is done by a surgeon who specializes in operating on children and has experience in treating these cancers.

- Removing the tumor
- Assessing the extent of the disease (surgical exploration)
- Placing a central venous catheter (port)
- Possible risks and side effects of surgery to remove a Wilms tumor
- More information about surgery

# Removing the tumor

The main goal of surgery is to remove the entire Wilms tumor in one piece, if possible. This is important because Wilms tumors tend to be fragile. Cutting into them makes it more likely that some cancer cells might spread inside 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 <u>imaging tests</u><sup>1</sup> done before surgery, or when starting the operation) that the entire tumor can't be removed safely, surgery might not be done right away. Instead, 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

A radical nephrectomy removes the entire kidney and some nearby structures.

This is the most common surgery for a Wilms tumor that's only in one kidney, because it provides the best chance of making sure all of the tumor is removed. Wilms tumors have often grown large by the time they are found. Sometimes, they are even larger than the kidney itself.

During this operation, the surgeon makes an incision (cut), usually down the middle of the belly. Then the surgeon removes the cancer along with the whole kidney, the adrenal gland that sits on top of the kidney, the surrounding fatty tissue, and the ureter (tube that carries urine from the kidney to the bladder).

Most children do very well afterward with only one kidney.

While it's not common, if there are larger tumors in both kidneys, both kidneys might need to be removed completely. The child would then need **dialysis** several times a week. During dialysis, a machine does the job of the kidneys by filtering waste products out of the blood.

Once the child is healthy enough, and if a donor kidney becomes available, a **kidney transplant** may be an option.

## Partial nephrectomy (nephron-sparing surgery)

A partial nephrectomy removes only part of the kidney(s).

For the small number of children who have Wilms tumors in both kidneys, this surgery might be done to try to save some normal kidney tissue. The surgeon may do a radical nephrectomy to remove the kidney containing the most tumor, and then a partial nephrectomy on the other kidney, removing just the tumor and a margin of normal kidney around it. Another option might be to do partial nephrectomies on both kidneys.

# Assessing the extent of the disease (surgical exploration)

During surgery to remove the tumor (either radical or partial nephrectomy), another main goal is to determine the extent of the cancer and if it can all be removed.

<u>Lymph nodes</u><sup>2</sup> (bean-sized collections of immune cells) near the kidney will be removed during surgery to look for cancer cells in them. When cancer cells from a Wilms tumor spread, they often go to the nearby lymph nodes first. 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 <u>biopsied</u><sup>3</sup> (samples taken to be checked for cancer under a microscope).

It's important for the cancer care team to know if a Wilms tumor has spread to the lymph nodes, the other kidney, or other nearby organs. This helps them determine the <u>stage of</u>

the Wilms tumor<sup>4</sup> and further treatment options.

# Placing a central venous catheter (port)

If your child is going to get chemotherapy for their Wilms tumor, a surgeon will often insert a small tube into a large blood vessel, usually under the collar bone. This tube is called a **central venous catheter**<sup>5</sup>, **venous access device**, or **port**.

It might be placed during the surgery to remove the tumor, or as a separate operation (especially if chemo is going to be given before the surgery).

One end of the catheter stays outside the body or just under the skin. This can be used to give chemo or take blood samples without the need for more needle sticks into veins. The catheter can stay in place for months.

# Possible risks and side effects of surgery to remove a Wilms tumor

Surgery to remove a Wilms tumor is a major operation, and surgeons are very careful to try to limit any problems either during or after surgery.

#### **During surgery**

Complications during surgery are rare, but they can happen. This includes bleeding, injuries to major blood vessels or other organs, or reactions to anesthesia.

# After surgery

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 these problems can include internal bleeding, blood clots, infections, or problems with food moving through the intestines.

# **Kidney function after surgery**

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. 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 they may eventually need a kidney transplant.

# More information about surgery

For more general information about surgery as a treatment for cancer, see <u>Cancer</u> <u>Surgery</u><sup>6</sup>.

To learn about some of the side effects listed here and how to manage them, see Managing Cancer-related Side Effects<sup>7</sup>.

# **Hyperlinks**

- 1. <a href="www.cancer.org/cancer/types/wilms-tumor/detection-diagnosis-staging/how-diagnosed.html">www.cancer.org/cancer/types/wilms-tumor/detection-diagnosis-staging/how-diagnosed.html</a>
- 2. www.cancer.org/cancer/diagnosis-staging/lymph-nodes-and-cancer.html
- 3. www.cancer.org/cancer/diagnosis-staging/tests/biopsy-and-cytology-tests.html
- 4. <u>www.cancer.org/cancer/types/wilms-tumor/detection-diagnosis-staging/staging.html</u>
- 5. <u>www.cancer.org/cancer/managing-cancer/making-treatment-decisions/tubes-lines-ports-catheters.html</u>
- 6. www.cancer.org/cancer/managing-cancer/treatment-types/surgery.html
- 7. www.cancer.org/cancer/managing-cancer/side-effects.html

#### References

Fernandez CV, Geller JI, Ehrlich PF, et al. Chapter 24: Renal Tumors. In: Blaney SM, Adamson PC, Helman LJ, eds. *Pizzo and Poplack'omT5 ET BT 1 0 7p5 ET BT 1 0 7p5 ET BT 1 0 7p5* 

Smith V, Chintagumpala M. Treatment and prognosis of Wilms tumor. UpToDate. 2024. Accessed at https://www.uptodate.com/contents/treatment-and-prognosis-of-wilms-tumor on November 22, 2024.

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# **Chemotherapy for Wilms Tumors**

Most children with Wilms tumors get chemotherapy as part of their treatment.

Chemotherapy (chemo) is the use of certain types of drugs to treat cancer. These drugs enter the blood and reach all areas of the body, which makes this treatment useful for cancer that might have spread beyond the kidney.

- When might chemotherapy be used for Wilms tumors?
- Which chemo drugs are used to treat Wilms tumors?
- How is chemo given for a Wilms tumor?
- Possible side effects of chemo
- More information about chemotherapy

# When might chemotherapy be used for Wilms tumors?

Most children with Wilms tumors will get chemo at some point during their treatment. (Some children with <u>very low risk tumors</u><sup>1</sup> might not need it.)

In the United States and Canada, chemo is usually given after surgery for a Wilms tumor. Sometimes it may be needed before surgery, to shrink a tumor and make the operation possible.

In Europe and some other parts of the world, chemo is often given before surgery and

# Which chemo drugs are used to treat Wilms tumors?

A combination of chemo drugs is used to treat children with Wilms tumors. The chemo drugs used most often are:

- Actinomycin D (dactinomycin)
- Vincristine

For tumors at more advanced stages, tumors with anaplastic histology, or tumors that recur (come back) after treatment, other chemo drugs might also be used, such as:

- Doxorubicin (Adriamycin)
- Cyclophosphamide
- Etoposide
- Irinotecan
- Carboplatin
- Ifosfamide
- Topotecan

# How is chemo given for a Wilms tumor?

Chemo drugs for Wilms tumors are infused into the blood. This is done either through a vein (IV) or through a <u>central venous catheter</u><sup>3</sup> (a thin tube inserted into a large blood vessel during surgery).

Different drugs, doses, and lengths of treatment might be 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.

Chemo is usually given by a nurse in a clinic or in the outpatient section of the hospital. Some children with Wilms tumors might need to stay in the hospital while getting chemo, but usually this is not needed.

#### Possible side effects of chemo

Chemo drugs can affect cells other than cancer cells, which can lead to side effects.

The side effects of chemo depend on which drugs are given, the doses used, and the

length of treatment

#### Short-term side effect

Possible short-term side

- Hair loss
- Mouth sores
- Loss of appetite
- Nausea and vomiting
- Diarrhea or constipation
- Increased chance of infe
- Easy bruising or bleeding
- Fatigue or extreme tire to be few red blood cells)

Your child's doctor and treatile to closely for any side effects that develop. There are often ways to lessen or example, drugs can be given to help prevent or reduce nausea and

Ask your child's cancer care to about medicines to be no reduce side effects. Be to let them key wif your child ha any side effects, so to be managed.

# Drug-specific side effects

Along with the effects listed above, some chemo drugs used to treat Wilms tumors can have specific side effects. For example:

Vincristine can damage nerves. Some patients may have tingling, numbness.

Wierekistise, or pain, particularly in the hands and feet. (This is called may h3 Tms (VinlfF4cristine)

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

Before each chemo session, the cancer care team will get <u>blood tests</u><sup>4</sup> to check blood cell levels and see how well your child's liver and kidneys are working. If there are problems, chemo might need to be delayed or the doses reduced.

# Long-term side effects of chemo

Some chemo drugs have possible long-term effects. This is one of the major challenges children might face after cancer treatment.

#### For example:

- If your child is given doxorubicin, there is a chance it could damage their heart. Your child's cancer care team will carefully watch the doses used. They will also check your child's heart function with imaging tests.
- Some chemo drugs can increase the risk of developing a <u>second type of cancer</u><sup>5</sup> (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 the Wilms tumor.
- Some drugs might also affect fertility<sup>6</sup> (the ability to have children) years later.

See <u>Living as a Wilms Tumor Survivor</u><sup>7</sup> for more on the possible long-term effects of treatment.

# More information about chemotherapy

For more general information about how chemotherapy is used to treat cancer, see <u>Chemotherapy</u><sup>8</sup>.

To learn about some of the side effects listed here and how to manage them, see Managing Cancer-related Side Effects<sup>9</sup>.

# **Hyperlinks**

1. <u>www.cancer.org/cancer/types/wilms-tumor/detection-diagnosis-staging/staging.html</u>

- 2. <u>www.cancer.org/cancer/types/wilms-tumor/detection-diagnosis-staging/staging.html</u>
- 3. <u>www.cancer.org/cancer/managing-cancer/making-treatment-decisions/tubes-lines-ports-catheters.html</u>
- 4. <u>www.cancer.org/cancer/diagnosis-staging/tests/understanding-your-lab-test-results.html</u>

# **Radiation Therapy for Wilms Tumors**

Radiation therapy uses high-energy rays or particles to kill cancer cells. It is often part of treatment for certain Wilms tumors.

- When might radiation therapy be used for Wilms tumors?
- How is radiation therapy given for a Wilms tumor?
- Types of radiation therapy
- Possible side effects of radiation therapy for Wilms tumors
- More information about radiation therapy

# When might radiation therapy be used for Wilms tumors?

Radiation is often part of treatment for more advanced Wilms tumors (stages III, IV, and V). It is also part of treatment for some earlier stage tumors with anaplastic histology. (See Wilms Tumor Staging<sup>1</sup> for an explanation of histology and stages.)

It might be used:

- After surgery to try to make sure all of the cancer is gone
- Before surgery to try to shrink the tumor to make it easier to remove
- Instead of surgery if it can't be done for some reason

For more on this, see Treatment by Type and Stage of Wilms Tumor.

# How is radiation therapy given for a Wilms tumor?

The type of radiation used for Wilms tumors is called **external beam radiation therapy**. For this treatment, radiation is aimed at the cancer from a machine outside the body.

Before treatments start, the radiation team will take careful measurements with imaging tests such as <u>CT</u><sup>2</sup> or <u>MRI</u><sup>3</sup> scans. This helps them determine the proper dose of radiation and the correct angles for aiming the radiation beams.

This planning session is called **simulation**. Your child may be fitted with a plastic mold that looks like a body cast. The mold keeps them in the same position during each treatment so that the radiation can be aimed more accurately.

Radiation is usually given 5 days a week for several weeks. Each session lasts about

15 to 30 minutes, with most of the time spent making sure the radiation is aimed correctly. The actual treatment time is much shorter.

Radiation treatment is much like getting an x-ray, although the dose of radiation is much stronger. For each session, your child lies on a special table while a machine delivers the radiation from precise angles.

The treatment is not painful, but some younger children may be given medicine beforehand to make them drowsy or fall asleep so they stay still.

# Types of radiation therapy

Modern radiation therapy techniques help doctors aim the treatment at the tumor more accurately than in the past. These techniques may help increase success rates and reduce side effects when treating Wilms tumors.

## 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.

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 doctors deliver a higher dose to the tumor.

Many major hospitals and cancer centers now use IMRT.

#### **Proton beam therapy**

This treatment uses protons to kill cancer cells, instead of x-rays or other types of radiation.

Protons are parts of atoms that travel a certain distance before releasing most of their

energy. They cause little damage to the tissues they pass through. This is different from x-rays, which give off the same amount of energy as they pass through normal tissue both before and after reaching the tumor.

This property of protons allows doctors to give higher doses of radiation to the tumor while doing less damage to the normal tissue around it.

Proton therapy can be helpful in treating tumors when it's very important to limit the radiation that reaches nearby structures. This type of radiation therapy requires very specialized equipment, so only a limited number of centers in the United States offer it at this time.

# Possible side effects of radiation therapy for Wilms tumors

Radiation therapy is often an important part of treatment for Wilms tumors, but young children's bodies are very sensitive to it. Doctors try to use the lowest dose of radiation and aim it as precisely as possible. They also try to shield some parts of the body from the radiation, to help avoid or limit any problems.

Still, radiation can cause both short-term and long-term side effects. These side effects depend on the dose of radiation and where it's aimed.

#### Possible short-term effects

- Effects on areas of skin that get 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 to the kidney area can damage the kidneys. This is more likely to be a concern in children who need treatment in both kidneys.
- 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.

National Cancer Institute. Wilms Tumor and Other Childhood Kidney Tumors Treatment (PDQ®)—Health Professional Version. 2024. Accessed at https://www.cancer.gov/types/kidney/hp/wilms-treatment-pdq on November 22, 2024.

National Comprehensive Cancer Network. NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®): Wilms Tumor (Nephroblastoma). V2.2024. Accessed at https://www.nccn.org on November 22, 2024.

Smith V, Chintagumpala M. Treatment and prognosis of Wilms tumor. UpToDate. 2024. Accessed at https://www.uptodate.com/contents/treatment-and-prognosis-of-wilms-tumor on November 22, 2024.

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# Treatment of Wilms Tumor, By Type and Stage

Treatment for Wilms tumor is based mainly on the cancer's stage and its histology, as described in detail below.

In the United States, most children with Wilms tumors are treated in <u>clinical trials</u><sup>1</sup> developed by the <u>Children's Oncology Group</u><sup>2</sup>.

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.

- How type, stage, and risk group guide treatment for Wilms tumor
- Treating stage I Wilms tumors
- Treating stage II Wilms tumors
- Treating stage III Wilms tumors
- Treating stage IV Wilms tumors
- Treating stage V Wilms tumors
- Treating recurrent Wilms tumors

# How type, stage, and risk group guide treatment for Wilms tumor

Treatment for <u>Wilms tumor</u><sup>3</sup> is based mainly on the **stage** of the cancer and whether its **histology** is favorable or anaplastic. (See <u>Wilms Tumor Staging</u><sup>4</sup> for an explanation of staging and histology.)

Other factors can influence treatment as well, including:

- The child's age
- If the tumor cells have certain chromosome changes
- The size (weight) of the main tumor

These factors are combined to assign the child to a <u>risk group</u><sup>5</sup>, which is used to help guide treatment.

In the United States and Canada, doctors prefer to use surgery as the first treatment for Wilms tumor in most cases. Chemotherapy (and possibly radiation therapy) are then given afterward.

In Europe and some other parts of the world, doctors prefer to start the chemotherapy before surgery. The results from these approaches 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. This is because the true extent of the tumor often can't be determined by <u>imaging tests</u><sup>6</sup> alone. The doctors use what they learn during surgery to guide further treatment.

But sometimes it's clear that the cancer has already spread beyond the kidney, based on imaging tests even before surgery is done. This can affect the order in which treatments are given, as well as the extent of surgery.

# **Treating stage I Wilms tumors**

Stage I Wilms tumors are only in the kidney, and surgery has completely removed the tumor along with the entire kidney, nearby structures, and some nearby <u>lymph nodes</u><sup>7</sup>.

# Favorable histology

**Children younger than 2 years with small tumors** (weighing less than 550 grams) may not need further treatment after surgery. 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 for several months, with the drugs actinomycin D and vincristine.

If the tumor cells have certain chromosome changes, the drug doxorubicin (Adriamycin) may be given as well.

#### **Anaplastic histology**

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

# **Treating stage II Wilms tumors**

Stage II Wilms tumors have grown outside the kidney into nearby tissues, but surgery has 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.

#### Anaplastic histology, with focal (only a little) anaplasia

When the child recovers from surgery, radiation therapy is given over several weeks. After this is finished, chemo (doxorubicin, actinomycin D, and vincristine) is given for about 6 months.

## Anaplastic histology, with diffuse (widespread) anaplasia

After surgery, children with these tumors get radiation over several weeks. This is followed by a more intense type of chemo using the drugs vincristine, doxorubicin,

etoposide, cyclophosphamide, and carboplatin. Mesna (a drug that helps protect the bladder from the effects of cyclophosphamide) is given along with this, for about 6 months.

# **Treating stage III Wilms tumors**

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.

#### Anaplastic 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.

#### Anaplastic 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.

#### Tumors that are very large and/or are in places that make them hard to remove

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 children with these tumors, a small <u>biopsy</u><sup>8</sup> 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,

- 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 harder to treat. Children with these tumors are usually treated with aggressive chemo, such as the ICE regimen (ifosfamide, carboplatin, and etoposide) or with other regimens being studied in clinical trials.

Very high-dose chemo followed by a <u>stem cell transplant</u><sup>11</sup> (sometimes called a **bone marrow transplant**) might also be an option in this situation, although this is still being studied.

Adamson PC, Helman LJ, eds. *Pizzo and Poplack's Principles and Practice of Pediatric Oncology*. 8th ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2021.

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