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Treating Wilms Tumor

If your child has been diagnosed with a Wilms tumor, your child's 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 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 disease 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, 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 approaches.

The first goal of treatment is to remove the primary (main) tumor, even if the cancer has

spread to other parts of the body. Sometimes the tumor might be hard to remove because it's very large, it has spread 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 by Type and Stage of Wilms Tumor

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

Surgery for Wilms Tumors

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.

More information about Surgery

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

To learn about some of the side effects listed here and how to manage them, see Managing Cancer-related Side Effects⁷.

Hyperlinks

- 1. <u>www.cancer.org/cancer/wilms-tumor/detection-diagnosis-staging/how-diagnosed.html</u>
- 2. <u>www.cancer.org/treatment/understanding-your-diagnosis/lymph-nodes-and-cancer.html</u>
- 3. <u>www.cancer.org/treatment/understanding-your-diagnosis/tests/testing-biopsy-and-cytology-specimens-for-cancer.html</u>
- 4. www.cancer.org/cancer/wilms-tumor/detection-diagnosis-staging/staging.html
- 5. <u>www.cancer.org/treatment/treatments-and-side-effects/planning-managing/tubes-lines-ports-catheters.html</u>
- 6. <u>www.cancer.org/treatment/treatments-and-side-effects/treatment-types/surgery.html</u>
- 7. <u>www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects.html</u>

References

Chintagumpala M, Muscal JA. Treatment and prognosis of Wilms tumor. UpToDate. Accessed at www.uptodate.com/contents/presentation-diagnosis-and-staging-of-wilms-tumor on September 4, 2018.

Fernandez CV, Geller JI, Ehrlich PF, et al. Chapter 29: Renal Tumors. 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 Wilms Tumors

Chemotherapy (chemo) is the use 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 has spread or might have spread beyond the kidney.

Doxorubicin (Adriamycin)

treatment.

More information about chemotherapy

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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.

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.

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.

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 include:

- 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 include:

- 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.
- Radiation to the chest area can affect the heart and lungs. This doesn't usually cause problems right away, but in some children it might lead to heart or lung problems as they get older.
- In girls, radiation to the abdomen (belly) 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 <u>Living as a Wilms Tumor Survivor</u>⁵ for more on the possible long-term effects of treatment.

More information about radiation therapy

To learn more about how radiation is used to treat cancer, see Radiation Therapy⁶.

To learn about some of the side effects listed here and how to manage them, see Managing Cancer-related Side Effects⁷.

Hyperlinks

- 1. www.cancer.org/cancer/wilms-tumor/detection-diagnosis-staging/staging.html
- 2. www.cancer.org/cancer/wilms-tumor/about/what-is-wilms-tumor.html
- 3. www.cancer.org/treatment/understanding-your-diagnosis/tests/ct-scan-for-cancer.html
- 4. www.cancer.org/treatment/understanding-your-diagnosis/tests/mri-for-cancer.html
- 5. www.cancer.org/cancer/wilms-tumor/after-treatment/follow-up.html
- 6. <u>www.cancer.org/treatment/treatments-and-side-effects/treatment-types/radiation.html</u>
- 7. www.cancer.org/treatment/treatments-and-side-effects/physical-side-effects.html

and then give chemotherapy (and possibly radiation therapy) afterward. In Europe, 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, because the true extent of the tumor often can't be determined by <u>imaging tests</u>⁵ 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 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, and surgery has completely removed the tumor along with the entire kidney, nearby structures, and some nearby <u>lymph nodes</u>⁶.

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.

Stage II

These 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. When this is finished, chemo (doxorubicin, actinomycin D, and vincristine) is given for about 6 months.

Anaplastic 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

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.

Anaplastic 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 biopsy⁸ 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.

<u>Biopsies</u>⁹ (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 anaplastic), 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 <u>stem cell transplant</u>¹⁰ (sometimes called a *bone marrow transplant*) might also be an option in this situation, although this is still being studied.

Hyperlinks

- 1. www.cancer.org/treatment/treatments-and-side-effects/clinical-trials.html
- 2. childrensoncologygroup.org/
- 3. www.cancer.org/cancer/wilms-tumor/detection-diagnosis-staging/staging.html

- 4. www.cancer.org/cancer/wilms-tumor/about/what-is-wilms-tumor.html
- 5. <u>www.cancer.org/treatment/understanding-your-diagnosis/tests.html</u>
- 6. <u>www.cancer.org/treatment/understanding-your-diagnosis/lymph-nodes-and-cancer.html</u>
- 7. <u>www.cancer.org/treatment/understanding-your-diagnosis/tests/testing-biopsy-and-cytology-specimens-for-cancer.html</u>
- 8. <u>www.cancer.org/treatment/understanding-your-diagnosis/tests/testing-biopsy-and-cytology-specimens-for-cancer.html</u>
- 9. <u>www.cancer.org/treatment/understanding-your-diagnosis/tests/testing-biopsy-and-cytology-specimens-for-cancer.html</u>
- 10. <u>www.cancer.org/treatment/treatments-and-side-effects/treatment-types/stem-cell-transplant.html</u>

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