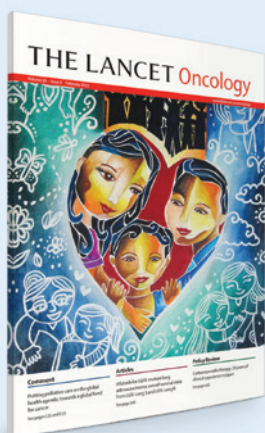




Expert Recommendations for the Diagnosis and Treatment of Chordoma

The Chordoma Foundation developed this booklet based on the article "Building a global consensus approach to chordoma: a position paper from the medical and patient community," which was written by an international group of chordoma experts and patient advocates. The article was published in the medical journal *The Lancet Oncology* in February 2015.



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Hans Keulen

JULY 28, 1957 — OCTOBER 29, 2015



This patient booklet is dedicated to the memory of Chordoma Foundation Board member and European Liaison, Hans Keulen. Hans served the chordoma community tirelessly, promoting research and assisting numerous European patients. We are grateful to Hans for organizing the chordoma physician community and spearheading the development of these guidelines. He inspired us with his optimism, his cheer, and his passion for improving patient care. He is deeply missed, and we will continue to persevere in his honor to find effective treatments for this disease, to educate patients about their choices, and to support them in making the best possible care decisions.

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Introduction

What should I do? If you've been diagnosed with chordoma, you've probably found yourself asking this question. It's an important question to ask, because, when it comes to treating chordoma, what you do — or don't do — can have a major impact on your life. This booklet is intended to help you answer that question so that you can make informed treatment decisions and get the best care possible.

The treatment recommendations you will find here were developed by the Chordoma Global Consensus Group – a multidisciplinary, international group of more than 40 doctors who specialize in caring for chordoma patients. The Chordoma Foundation and the European Society for Medical Oncology first convened this group in 2013 to define evidence-based recommendations for diagnosing and treating primary chordoma.


The group's goal was to develop a reference that could help doctors across the world provide better and more consistent care to their chordoma patients. The resulting guidelines were published in 2015 in the medical journal *The Lancet Oncology*. As part of our commitment to helping patients and caregivers make the most informed treatment choices, we have made those same recommendations available to you here in this booklet.

The consensus group came together again in November 2015 to develop detailed recommendations for treating locally and regionally recurrent chordoma. The resulting guidelines were published in an open-access paper in the medical journal, *Annals of Oncology*, in June 2017. The Chordoma Foundation's booklet with those recommendations can be viewed and downloaded at chordoma.org/educational-materials.



How to use this booklet

The following pages are a faithful summary of the information and recommendations presented in *The Lancet Oncology* paper. Some things to note as you read through the booklet:

1. If you are newly diagnosed, please take time to read through the "Understanding chordoma" section for important background information about the disease.
2. If you are preparing for any stage of treatment, you will also find sections containing information and expert recommendations on surgery, radiation, and drug therapy. **Please read this information carefully and discuss it with your caregivers and doctors.**
3. Text noted with the Chordoma Foundation logo  contains additional information, explanations, and tips from the Chordoma Foundation to help you understand and act on the expert recommendations.
4. Terms in **bold blue font** are included in the glossary (p. 31). The Chordoma Foundation put together the glossary as a supplement to the publication.

If you have any questions as you read through this booklet, please contact a Chordoma Foundation Patient Navigator at chordoma.org/request-help or by calling (888) 502-6109.

Finding the right medical team

If you or someone you know might have chordoma, the most important thing to do is to find a medical center with a team of experts who have experience caring for chordoma patients.



Experts recommend that you find a medical center experienced in:

- Sarcoma or bone pathology
- Radiology
- Orthopedic or neurosurgical spine surgery (for patients with spine tumors)
- Skull base neurosurgery (for patients with skull base tumors)
- Radiation oncology
- Medical oncology
- Palliative care

Chordoma is a rare disease, and it affects very important and complex parts of the body. For these reasons, appropriate diagnosis and treatment of chordoma requires very specialized care provided by multiple types of doctors. This team approach involving multiple specialists is called **multidisciplinary care**. It is typically only found at larger hospitals called **referral centers**, which see large numbers of patients, and is not available at most local hospitals.


All members of the care team should have substantial experience treating tumors of the skull base and spine including chordoma.

It is also recommended that your doctors discuss your case in a multidisciplinary **tumor board**. This is a regular meeting where different specialists come together to review each patient's situation and to develop the best treatment plan. As a patient, you benefit from the knowledge and experience of many experts instead of just one or two. This is very important for treating a complicated disease like chordoma.



Locate a doctor

The Chordoma Foundation Doctor Directory can help you find doctors around the world who have experience treating chordoma. Visit the Doctor Directory at **chordoma.org/doctor-directory**.


You can also contact a Chordoma Foundation Patient Navigator for assistance at **chordoma.org/request-help** or by calling (888) 502-6109. 

Understanding chordoma

Chordoma is a **rare bone cancer** that is diagnosed in just 1 in 1 million people per year. At any given time, fewer than 1 in 100,000 people are living with chordoma.

The basics

Chordoma is part of a group of malignant bone and soft tissue tumors called **sarcomas**. It is diagnosed most often in people in their 50s and 60s, but it can occur at any age. About twice as many men are diagnosed with chordoma as women. Chordoma can run in families, but this is very rare.

Chordoma very rarely occurs in children, adolescents, and young adults under the age of 35. To learn more, visit chordoma.org/pediatric-aya. 

Chordoma tumors usually grow slowly. A tumor might cause symptoms for years before doctors find it. A chordoma can come back, or recur, after treatment — usually in the same place as the first tumor. This is called a **local recurrence**. Spread of the tumor to other parts of the body — called **metastatic** disease — occurs in about 30 to 40 percent of patients. The most common places for chordomas to metastasize are the lungs, liver, bones, or lymph nodes.

Locations of chordoma

About half of all chordomas form at the bottom of the spine, in bones called the **sacrum**. About 30 percent form within the center of the head in an area called the skull base — usually in a bone called the **clivus**. The remaining 20 percent of chordomas form in the spine at the level of the neck, chest, or lower back, also called the **mobile spine**. Very rarely, chordomas can start in more than one place along the spine.

Causes of chordoma

Chordoma tumors develop from cells of a tissue called the **notochord**, a structure in an embryo that helps in the development of the spine. The notochord disappears when the fetus is about 8 weeks old, but some notochord cells remain behind in the bones of the spine and skull base. Very rarely, these cells turn into cancer called chordoma. What causes notochord cells to become cancerous in some people is still not fully understood, but researchers are working to figure it out.

Types of chordoma

There are three types of chordoma, which are classified based on how they look under a microscope. **Conventional** chordoma is the most common form and now includes chondroid chordoma, which was previously a separate type. **Dedifferentiated** and **poorly differentiated** chordoma are very rare types that are more aggressive and typically grow faster than the conventional type. There is some evidence that these two types are more prevalent in younger patients (under the age of 35).¹

¹WHO Classification of Tumours Editorial Board. WHO classification of tumours of soft tissue and bone, 5th ed. IARC Press, 2020

Chordoma is not always easy to diagnose and can be confused with other diseases. Getting the correct diagnosis can affect the treatment you receive. For this reason, it is very important for your diagnosis to be made by doctors who have experience diagnosing and treating chordoma patients. Getting a second opinion to confirm the diagnosis can be helpful before making treatment decisions. If you have not yet had treatment, ask whether any additional tests are needed to rule out other possible tumor types before going forward with treatment. 🔄





Diagnosing chordoma

It is important to go to a referral center as soon as chordoma is suspected, even before you know for sure that you have chordoma.

Imaging

Chordoma tumors are typically detected through imaging tests, which show organs and other structures inside the body, including tumors. The way the tumor looks on imaging tests can tell a radiologist whether the tumor might be chordoma.

When a chordoma is suspected, you will need an **MRI**, also called **magnetic resonance imaging**, to help doctors make a diagnosis and plan for treatment. This is the best way to see a chordoma and how it is affecting the tissue around it, such as muscles, nerves, and blood vessels. No matter where the tumor is located, an MRI of the entire spine should be performed to see if the tumor may have spread to or developed in other areas of the spine. Chordoma is best seen on an MRI with a setting called T2 weighted imaging.

A **CT (computed tomography)** scan is another form of imaging test that is recommended in addition to MRI if it is not certain whether the tumor is chordoma. CT scans of the chest, abdomen, and pelvis are recommended.

Imaging tests should be interpreted by a radiologist who has experience diagnosing bone tumors.

Biopsy


Imaging studies can show the possibility of a chordoma, but a definitive diagnosis can only be made by a pathologist who examines a sample of tumor tissue under a microscope. For this reason, your medical team may consider taking a small sample of tissue from the tumor, called a **biopsy**, prior to surgery. However, biopsies are not recommended if the tumor cannot be reached safely or when there is a high risk of spreading tumor cells. For sacral and mobile spine tumors, a **trocar CT-guided biopsy** is recommended and should be done from the back.

Trocar CT-guided biopsy uses a CT scan to precisely direct the biopsy needle to the correct location. The biopsy needle is enclosed in a tube to keep tumor cells from spreading along the path of the needle — this is often called “seeding.” Check with your doctors to make sure they will use this method if a biopsy is planned.

If you have a biopsy before surgery, it is recommended that your surgeon take out the tissue around the area of the biopsy during surgery in order to remove any chordoma cells that might have spread when the biopsy disturbed the tumor.


If you've been told you might have chordoma

You should avoid having a biopsy or surgery to confirm the diagnosis of chordoma outside of a referral center because, if not done properly, these procedures can cause the chordoma to spread.

If you cannot travel to see a specialist, have your imaging tests sent to a referral center for a second opinion before you get treatment. 

Pathology


Tissue samples should be evaluated by a pathologist who has experience diagnosing bone tumors. Your pathologist may test your tumor tissue for the presence of a protein called **brachyury**. Nearly all chordomas have high levels of brachyury, which makes it helpful for diagnosis.

Poorly differentiated chordoma often loses expression of a protein called INI1, and sometimes dedifferentiated and conventional chordoma can as well. The Chordoma Foundation Medical Advisory Board suggests that chordoma patients under 35 years of age, or whose tumors are growing unusually fast, have their tumors tested for loss of INI1. This simple testing can be done by a pathologist on a sample of tumor tissue. 



If you have already had initial treatment outside of a referral center

Regardless of what initial treatment you may have had, it is still very important to be evaluated at a referral center as soon as possible. In particular, it is a good idea to have a sample of your tumor sent to a referral center where an expert pathologist can confirm the diagnosis.

A Chordoma Foundation Patient Navigator can help you find a center that has experience with chordoma. Contact us at **chordoma.org/request-help** or by calling (888) 502-6109. 

Could my tumor be something else?

Diseases that can be mistaken for chordoma include:

Benign notochordal cell tumors (BNCT)

These benign spine tumors can be seen on an MRI or CT scan and can sometimes look like chordoma. However, BNCT stay confined within the bone and do not spread into other tissues like chordomas can. If you have a suspected BNCT, you should have an MRI or CT scan periodically to look for changes. Images should be reviewed by a radiologist with expertise in bone tumors.

Chondrosarcoma

This type of bone cancer looks very similar to chordoma on MRI and CT scans. A specific type of MRI called diffusion MRI, or D-MRI, may help doctors tell the difference. Sometimes it is only possible to know a tumor is not chondrosarcoma after having a biopsy.

Giant cell tumor of the bone (GCTB)

These tumors look somewhat different on imaging tests than chordoma, and tend to be located in the upper part of the sacrum.

Sacral schwannoma

These tumors damage the bone differently than chordomas do, look different on imaging tests, and do not spread to nearby muscles or joints.

Other tumors of the spine and skull base


These include other bone cancers such as Ewing sarcoma and osteosarcoma, as well as a type of nervous system tumor called a myxopapillary ependymoma. Lymphoma, a cancer of the body's immune system, and multiple myeloma, a blood cancer, can also cause tumors in these areas.

Metastasis (spread) of another cancer


Sometimes cancers in other places in the body can spread to the bones in the spine or skull base.

Initial treatment

After you are diagnosed with chordoma you will most likely need to have surgery, radiation, or both. These treatment methods have the potential to cure some chordoma patients if done properly.

The first treatment you have makes a big difference for your quality of life and the chances of the tumor coming back. Therefore, it is important to carefully consider your options and make an informed decision about your treatment. 

In most cases, surgery is recommended as the main treatment for chordoma. Radiation therapy is generally recommended after surgery to kill any remaining tumor cells. At times radiation is given before surgery to reduce the risk of the tumor spreading during surgery. If your tumor is located where surgeons cannot reach it or if the side effects of surgery are very serious and unacceptable to you, radiation may be given as the only treatment instead of surgery.

Doctors do not always agree on whether patients whose tumors can be removed by surgery should choose radiation therapy instead of surgery. Therefore, before having treatment you should talk with your doctors about all of your options and understand the risks and benefits of each type of treatment. It can be helpful to get multiple opinions from doctors who have experience treating chordoma patients. 

Because chordoma is a complex disease to treat, it is important to have a medical team that includes multiple specialists who work together to coordinate your care. A doctor specializing in radiation therapy for cancer, called a radiation oncologist, will need to be part of your medical team even before surgery to plan with your surgeons for any radiation treatment you will receive before or after surgery. Additionally, supportive care to deal with the symptoms of the disease and side effects of treatment should be considered from the beginning. See “Managing side effects” on p. 28 for more information.



Each patient's situation is different, so you should talk with your doctors about the course of treatment that makes the most sense for you. They can help you understand the various treatment options that are available to you, and create an individualized treatment plan based on your choices. 🗣️

Tests to get before treatment

Surgery is the most common initial treatment for chordoma. Before you have surgery for chordoma, you will need MRI and CT scans to help your surgeon plan.

If you have a skull base or cervical spine chordoma, a type of imaging test called **angiography** should be done to show the location of blood vessels that need to be protected during surgery. You will also need an examination that measures the function of your cranial nerves, visual acuity (how sharp your vision is), visual field, hearing, and pituitary gland function. Completing this examination prior to surgery will allow your doctors to know what has changed if you experience side effects.


After surgery and before radiation treatment, an MRI and possibly also a CT will be done to see if any of the tumor is still present. You also need regular MRI scans for several years after treatment, whether you have surgery, radiation therapy, or both. (See "Follow-up after treatment" on p. 28.)

Surgery

Surgery to remove the tumor is generally recommended when it can be performed without causing unacceptable side effects or causing the tumor to spread.

SKULL BASE AND CERVICAL TUMORS

Surgery should be performed in a medical center with substantial experience in skull base and upper cervical spine surgery. If you have a skull base tumor, your surgeons should be trained in surgical approaches that access the skull base from the front (nose or mouth) and side of the head. Skull base operations are usually done by a team that includes a neurosurgeon and an ear, nose, and throat (ENT) surgeon. ENT surgeons are also called otolaryngologists.

The location and size of your tumor will determine the surgical approach that is best for you. Sometimes it may be necessary to do surgery from multiple directions to safely remove different parts of a skull base tumor. Ask your surgeon about different surgical approaches that are available for you, and discuss the risks and benefits of each. 

The goal of surgery for tumors in these areas is to remove all visible tumor tissue, whenever possible. Because chordomas in the skull base and cervical spine often touch important nerves and blood vessels, it is usually not possible to remove these tumors in a single piece or to achieve a wide resection (see "Margins for chordoma surgery" on p. 24) without causing serious harm. For this reason, even if all visible tumor is removed, microscopic chordoma cells are likely to be left behind after surgery. Radiation therapy is generally recommended after surgery to prevent these remaining cells from re-growing. If the entire tumor cannot be removed, your surgeon should remove as much of the tumor as possible, especially from around the brainstem and optic nerve, so that later radiation therapy can be more effective.

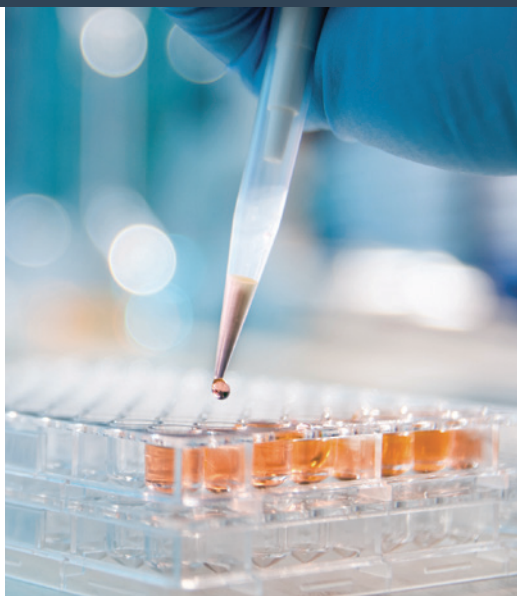
Surgery in the skull base and cervical spine can cause damage to the brainstem and cranial nerves, which control important functions like speech and swallowing. To reduce the risk of serious nerve injury, **neurophysiological monitoring** is recommended during surgery.

LUMBAR AND THORACIC TUMORS

The goal of surgery for tumors in this area is to completely remove the tumor in one piece (**en-bloc**) with wide margins. Removing the tumor in more than one piece should be avoided if at all possible. The surgeon must be very careful to avoid disturbing or spilling the contents of the tumor during surgery because this can cause it to re-grow or spread. The tissue that was touched by the biopsy needle should be removed during surgery as well.

An en-bloc resection might not be possible if the tumor has extended into the neck, chest, or behind the abdomen. In this case there may be tumor tissue left behind, and radiation following surgery should be considered. Sometimes radiation may be recommended before as well as after surgery, especially when an incomplete resection is likely. If your tumor is located where surgeons cannot reach it or if the side effects of surgery are very serious and unacceptable to you, radiation may be recommended as the only treatment instead of surgery.

After surgery to remove the tumor, plastic and reconstructive surgery will likely be required to repair or replace bone or tissue lost during surgery. This should be planned at the time of initial surgery to reduce complications. Metal implants used to stabilize the spine can interfere with radiation, so a radiation oncologist should be consulted when surgery is planned if stabilization is required.



Donating tumor tissue

Tumor tissue removed during surgery is critical for research to identify new ways to treat chordoma. Patients who have surgery in the United States can help advance research by donating tumor tissue to the Chordoma Foundation Biobank.

To learn more, visit **chordoma.org/tumor-donation**. If you are interested in helping to advance research, call (877) 230-0164 or email **tumordonation@chordoma.org** before you have surgery. 🌐

The way surgery is performed can have a big effect on your outcome.

Here are three things that all chordoma patients should know before having surgery:

- Any tumor cells left behind after surgery can re-grow. Therefore, **the entire tumor should be removed whenever possible**, ideally with wide margins of healthy tissue surrounding the tumor. Completely removing the tumor lowers the risk of the tumor coming back after treatment, and improves the odds of survival.
- For most patients, radiation therapy is recommended after surgery. However, **the way surgery is done can affect what radiation therapy you can have**. For example, if part of your spine has to be removed during surgery, it may need to be replaced with metal implants, which can interfere with radiation. Therefore, plans for surgery should be made with the input of a radiation oncologist.
- Because chordomas tend to be located near important structures, **surgery can cause serious side effects that can affect your quality of life**. Before surgery, you should ask your surgeon about the risks of surgery and what to expect afterwards. If you are not comfortable with the likely side effects of surgery, ask about what other options you have. You and your medical team should agree on a treatment plan that you are comfortable with.

Additional considerations for surgery depend on the location of the tumor.

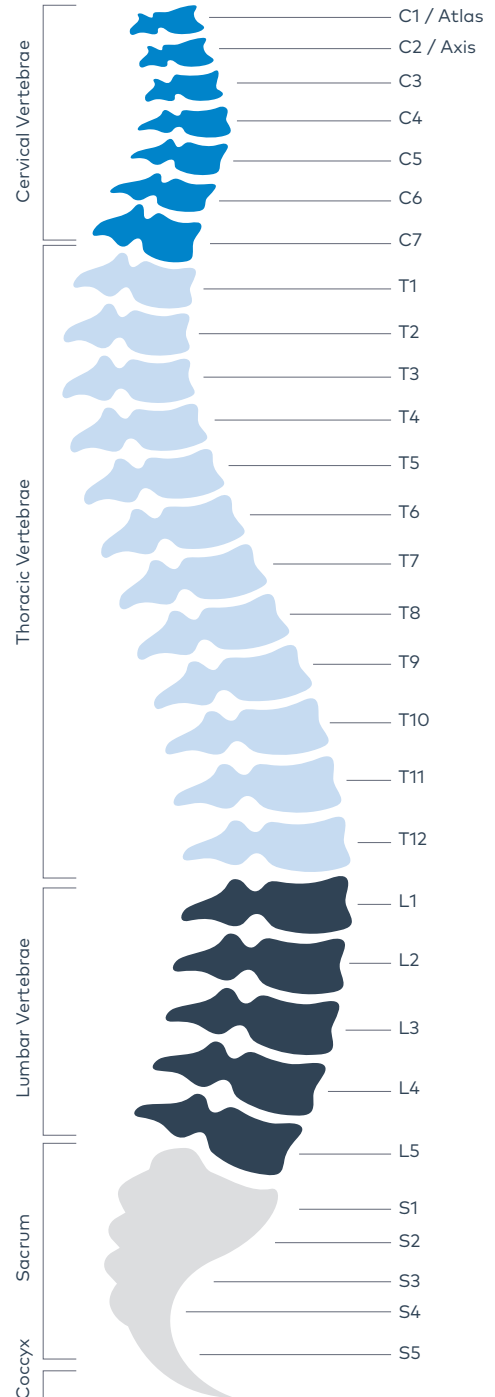


SACRAL TUMORS

The goal of surgery for sacral chordomas is to completely remove the tumor in one piece (**en-bloc**) with wide margins of normal tissue surrounding it. Intralesional resection should be avoided if at all possible (see "Margins for chordoma surgery" on pg. 24). The surgeon must be very careful to avoid disturbing or spilling contents of the tumor during surgery because this can cause it to re-grow or spread. Additionally, if a biopsy was performed, your surgeon should plan to take out the tissue that was touched by the biopsy needle to remove any tumor cells that may have been left behind.

After surgery to remove the tumor, plastic and reconstructive surgery will likely be required to repair or replace tissue lost during surgery. This should be planned at the time of initial surgery to reduce complications.

In some cases, surgery for sacral tumors can cause serious side effects including loss of bowel and bladder control, sexual function impairment, and movement problems. Surgeons can usually predict how severe these side effects will be



The table below shows the **recommended primary treatment** for chordoma in each part of the sacrum and likely side effects from surgery.

SACRAL VERTEBRA	RECOMMENDED TREATMENT	SIDE EFFECTS OF SURGERY
S1	Radiation is an advisable alternative	Side effects are very serious
S2	Dependent on patient preferences and quality of life considerations	Serious side effects are likely
S3	Surgery	If S2 nerve roots are not damaged, about 40 percent of people recover from any side effects
S4 or below	Surgery	Most important functions can be preserved

depending on the location of the tumor and which nerves are involved (see chart above). Due to the risks of surgery, radiation may be an alternative to surgery for some patients.

However, radiation alone is less likely to successfully control the tumor than surgery and radiation together. Additionally, the high dose radiation needed for treatment can also cause serious side effects later. Talk with your doctors about your options to decide what treatment plan is best for you.

Margins for chordoma surgery

You will probably hear the term **surgical margins** or just **margins** when your surgeon plans your treatment. Margins are the healthy tissue surrounding the tumor that is removed along with the tumor. Surgeons take out this healthy tissue to attempt to prevent microscopic cancer cells from being left behind.

The size of the margin can determine how likely the tumor is to come back after surgery. Levels of surgical margins described by the consensus group for chordoma are:

- **Wide resection:** The entire tumor is removed, with at least 1 millimeter of healthy tissue around the tumor.
- **Marginal resection:** Less than 1 millimeter of healthy tissue around the tumor, but no visible tumor tissue left behind.
- **Intralesional resection:** Visible tumor tissue is left behind, or tumor cells have spilled into the surrounding area during surgery.

Additionally, an **en-bloc resection** means that the entire tumor was removed in one piece. An en-bloc, wide resection is the best outcome, whenever possible. However, the location of chordomas near important structures sometimes makes this difficult or impossible. For skull base tumors, a **gross total resection**, which is removing the entire tumor in more than one piece, is usually the goal.

Radiation therapy

The purpose of radiation therapy is to kill or stop the growth of tumor cells within the body. Not all radiation is the same, and the way it is given makes a big difference. Below are important things to know about how radiation should be used for the treatment of chordoma.

ROLE OF RADIATION


Radiation can serve two different roles in the treatment of chordoma:

- **To reduce the risk of recurrence after surgery:** Radiation is usually recommended after surgery to kill any remaining tumor cells that are left behind during surgery. Sometimes part of the radiation is also given before or during surgery.
- **The main treatment instead of surgery:** Radiation is sometimes recommended as the primary treatment following a biopsy if the tumor cannot be removed with surgery or if the risks of surgery are unacceptable to you.

DOSE OF RADIATION

The most important thing to know about radiation is that high doses are required to control chordoma. Specifically, a dose of at least 74 GyE (Gray Equivalents) is recommended. This dose should be given to any visible tumor as well as any areas where your doctors believe there may be microscopic tumor remaining after surgery. Even if the tumor was completely removed in one piece (en-bloc) there could still be microscopic tumor cells nearby, and these can grow into tumors if not radiated. When an en-bloc resection is achieved the dose of radiation to the areas surrounding where the tumor was can be limited to 70 GyE.

Radiation is typically given in small doses (1.8-2 GyE per dose) during multiple sessions over several weeks. The dose of radiation delivered during each session is called a **fraction**. The radiation from each fraction accumulates over time until the total intended dose is reached.

Sometimes larger fractions will be given over a smaller number of sessions. This is called **hypofractionation**. When hypofractionation is used the total amount of radiation given is less than when standard fractionation is used, but the effect will be the same. 

The amount of radiation required to treat chordoma is higher than what healthy tissue can handle. For this reason, the radiation dose must be focused on the tumor while avoiding important nearby structures such as the brain, brainstem, nerves, or spinal cord. Radiation that is highly focused is called **conformal radiation**. Your radiation oncologist should plan radiation therapy to deliver the necessary dose to the tumor without causing harm to surrounding tissues.

TYPES OF RADIATION

Several different types of radiation and delivery methods can be used to treat chordoma.

External radiation

Chordomas are generally treated with a beam of radiation that is delivered to the tumor from a source outside the body. This is called **external beam radiation**.

- **Particle therapy** uses beams of charged particles and is generally recommended for treating chordoma because it can be focused very precisely on the tumor. Two different types of particles are commonly used: protons and carbon ions. These are usually referred to as **proton therapy** or **proton beam therapy**, and **carbon ion therapy**. It is not yet known whether there is any difference in effectiveness between protons and carbon ions. Proton and carbon ion treatment centers are very expensive to build, so they do not exist at every medical institution.
- **Photon radiation** can be a suitable alternative to particle therapy in some cases, as long as a high enough dose can be delivered without damaging healthy tissue.

Sometimes it may be helpful to combine particle therapy and photon radiation. For all types of external beam radiation, imaging is needed every day of treatment to make sure that the radiation is going to exactly the right place. This technique is called **image guidance**.

Internal radiation

Another method of delivering radiation, called **brachytherapy**, involves inserting a small amount of radioactive material inside the body during surgery. This method is not used very often for chordoma, but can sometimes be helpful for delivering a high enough dose of radiation to



the area near the brainstem or spinal cord. When used, it is usually given in combination with external beam radiation.


What matters most about radiation is that a high enough dose is delivered to the area that needs it while delivering a safe, lower dose to important nearby structures. Whether the right dose can be delivered with a particular type of radiation depends on a number of factors, including the shape of the area being radiated and the location of important structures that must be avoided. In general, the more precisely the radiation can be focused (the more conformal it is) the better.

It is important to have a detailed discussion with your radiation oncologist to understand the type of radiation therapy that is best for you and the short-term and long-term side effects you can expect.

Follow-up after treatment


After you complete treatment, you will need to maintain a consistent schedule of follow-up imaging to check whether the tumor has returned or spread to other areas.

You will need an MRI every 6 months for the first 5 years after treatment, then annually for at least 15 years. The MRI should look at the area of the original tumor as well as any areas where it could spread, such as the rest of the spine, the lungs, and the abdomen. CT scans may also be used for chest imaging.

It is important for a chordoma expert who knows your case to review your follow-up scans and compare them to your previous ones. Depending on your situation and the treatment you've had, follow-up visits are typically handled by either your surgeon or radiation oncologist. 

Managing side effects

Palliative care, also called supportive care, can improve the quality of life and well-being of patients dealing with a serious illness by preventing and treating symptoms of the disease or the side effects of its treatment. Palliative care is often confused with hospice care or end of life care, *but they are not the same*. Hospice care is a type of palliative care that is intended for the end of life period — generally for patients expected to live for less than six months — while other aspects of palliative care can benefit patients at any stage of a life-threatening or chronic illness.

Chordoma experts recommend that palliative care be part of the care plan for chordoma patients from the time of diagnosis, through all stages of treatment, as well as after treatment is complete. No matter what treatment you have for your recurrence, palliative care can help address pain, mobility and functional issues, mental and emotional health, nutrition, and many other concerns to help you live well while managing your chordoma. 



As doctors and researchers learn more about chordoma, evidence is regularly emerging about new treatment approaches that could possibly help chordoma patients.

Additionally, the Chordoma Foundation is initiating and supporting research to identify new, more effective treatments for chordoma.

To stay up to date on the progress that is being made, please visit **chordoma.org** and sign up to receive our emails.

Chordoma Foundation Patient Navigators are also available to provide more information, help answer your questions, and connect you with others for support. Contact a Patient Navigator at **chordoma.org/request-help** or by calling (888) 502-6109.

Learn more

Visit the Chordoma Foundation at **chordoma.org** for more information on chordoma, including research updates, the latest news on treatments, and ways to get involved.

Get help from a Chordoma Foundation Patient Navigator at **chordoma.org/request-help** or by calling (888) 502-6109.

Connect with other patients and caregivers through the Chordoma Connections online community at **community.chordoma.org**.

Important note about this booklet

The content herein was developed by the Chordoma Foundation in consultation with members of the Chordoma Global Consensus Group (see inside front cover for complete list of consensus group members). This information is not meant to take the place of medical advice. You should always talk with your doctors about treatment decisions.

Glossary of terms

angiography

An imaging test that is done before surgery to show the location of important blood vessels.

biopsy

A procedure that uses a needle to remove a small tissue sample from the tumor to be tested in order to make a diagnosis.

brachytherapy

A type of radiation therapy in which a small amount of radioactive material is placed into the body to kill cancer cells.

brachyury

A protein that is present at high levels in nearly all chordoma cells.

carbon ion therapy

A type of **particle therapy** that uses beams of carbon ions to kill cancer cells.

clivus

The surface of a bone at the base of the skull. It is surrounded by the brainstem and both carotid arteries.

computed tomography, or CT, scan

A type of imaging scan that uses x-rays to help visualize structures within the body such as a tumor. CT scans can also be used to help guide the needle during a biopsy. They are sometimes referred to as "CAT" scans.

conformal radiation

Types of radiation that can focus the beams of radiation on the tumor, while minimizing the amount of radiation that reaches the surrounding healthy tissue.

conventional

The most common type of chordoma. It is typically slow growing. Chondroid chordoma, previously a separate type, is now considered conventional chordoma.

dedifferentiated

A type of chordoma that is more aggressive and usually grows faster than conventional chordomas. Dedifferentiated chordomas occur in only 5 percent of patients.

en-bloc

During surgery, removal of the tumor in one piece without cutting it into smaller pieces.

external beam radiation

Radiation that is delivered from outside the body.

fraction

The dose of radiation delivered during a session of a radiation therapy.

hypofractionation

A radiation treatment technique that gives larger doses of radiation over a smaller number of sessions. The total amount of radiation given is less than when standard fractionation is used, but the effect is the same.

image guidance

The use of frequent imaging, such as MRI or CT, during radiation treatments to help direct the radiation to the right place.

IN11

A protein that regulates the growth of certain tumor cells. IN11 is lost in most poorly differentiated chordoma tumors.

local recurrence

Re-growth of the tumor in the same location after treatment.

magnetic resonance imaging (MRI)

A type of imaging scan that is used initially to help diagnose chordoma, as well as during follow up to check for recurrence or metastasis.

margins, surgical margins

The healthy tissue surrounding the tumor that is taken out along with the tumor to make sure that no cancer cells are left behind. The wider the tumor-free margins the lower the chances of recurrence.

metastatic

When cancer has spread to other parts of the body it is called metastatic. The process of spreading is called **metastasis**. The tumors that occur beyond the site of the original tumor are called **metastases**.

mobile spine

The parts of the spine not including the sacrum. These include the cervical spine (neck), thoracic spine (upper back), and lumbar spine (lower back).

multidisciplinary care

Treatment that involves a team of physicians from the various disciplines. In the case of chordoma, these disciplines include sarcoma or bone pathology, radiology, spine surgery or skull base surgery, otolaryngology, radiation oncology, medical oncology, and palliative care.

neurophysiological monitoring

The use of devices during surgery to monitor the functioning of neural structures such as the spinal cord, nerves, and brain. This is done to guide the surgeon during the operation, and to reduce the risk of damage to the patient's nervous system.

notochord

The tissue in a fetus that acts as the building blocks for the spine. The notochord disappears when the fetus is about 8 weeks old, but some notochord cells are left behind in the bones of the skull and spine.

off-label

The practice of prescribing drug treatments that are not approved by government agencies to treat a particular disease. Doctors are allowed to prescribe drugs off-label if they believe it is in the best interest of the patient.

particle therapy

A type of **external beam radiation** that uses beams of protons, neutrons, or positive ions for the treatment of cancer. See also **proton therapy** and **carbon ion therapy**.

PDGFR

A protein in some cancer cells that causes them to grow uncontrollably. This protein can be blocked with certain targeted drug therapies.

photon therapy

A type of **external beam radiation** that uses x-rays to kill cancer cells.

poorly differentiated

A type of chordoma that is more aggressive and usually grows faster than conventional chordoma. It is more common in children and young adults, and typically loses expression of the INI1 protein.

proton therapy, proton beam therapy

A type of **particle therapy** that uses beams of protons to kill cancer cells.

referral center

A hospital, treatment center, or network of treatment centers where doctors have expertise in particular diseases. Patients are referred to a center based on their diagnosis.

sacrum

The lowest section of the spine, consisting of five sacral bones and the coccyx.

sarcoma

Cancer of bone and connective tissue such as cartilage, fat, muscle, and blood vessels. Chordoma is a type of sarcoma.

systemic therapy

The use of drugs that spread through the body to kill cancer cells. Also called drug therapy. Chemotherapy is a type of systemic therapy that kills fast growing cells and is usually not effective for chordoma.

targeted therapy

A type of systemic therapy that works by blocking a specific gene or protein (the "target") in a patient's specific tumor cells.

trocac CT-guided biopsy

A type of biopsy that uses a CT scanner to guide the placement of the biopsy needle. This is the type of biopsy that is recommended for chordoma to reduce the chance of spreading tumor cells.

tumor board

A hospital's tumor board is a group of different types of specialists who meet regularly to review each patient's situation and make treatment recommendations.



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