



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

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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 or someone you know has chordoma, you've probably found yourself asking this question. And 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 publication is intended to help you answer that question so that you can make informed medical decisions and get the best care possible for yourself or your loved one.

This booklet contains recommendations developed by a group of over 40 leading doctors who specialize in caring for chordoma patients. The Chordoma Foundation, along with the European Society for Medical Oncology, convened this group to reach agreement on the best ways to treat chordoma based on all available evidence. Without such agreement among experts, many chordoma patients were not getting the most current and appropriate treatment, and therefore were not being given the best chance to beat their disease. Thus, 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 expert recommendations were published in 2015 in the high-profile medical journal *The Lancet Oncology*.

But recommendations created for doctors are not enough. The Chordoma Foundation believes that it is also very important to make these recommendations available to patients and caregivers in a format that is understandable and usable. That is why we created this booklet. Our hope is that it arms you with information and concepts that help you win your fight against chordoma. Should you have any questions as you read through this information, please contact support@chordoma.org and a patient navigator will gladly assist you.



How to Use This Booklet

The following pages are a faithful summary of the information and recommendations presented in *The Lancet Oncology* paper. Text shaded in a light blue box and/or noted with a Chordoma Foundation logo contains additional information, explanations, and tips from the Chordoma Foundation to help you understand and act on the expert recommendations.

If you are newly diagnosed, please take time to read through the “Understanding Chordoma” section for important background information about the disease.

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.

Terms in **bold blue font** are included in the glossary. The Chordoma Foundation put together the glossary (on p. 28) as a supplement to the publication.

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 in caring for chordoma patients.

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.

Depending on the stage of your disease, you may need to see different types of doctors. A Chordoma Foundation patient navigator can help you find a multidisciplinary referral center that has experience with chordoma and direct you to the right point of contact based on your needs. Please contact support@chordoma.org for assistance. 

Experts recommend that you find a medical center proficient 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



Chordoma is not always easy to diagnose and can be confused with other diseases. Getting the correct diagnosis can affect the treatment that you have. 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. 

All members of the care team should have substantial experience in 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.

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 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. In about 30 to 40 percent of patients the tumor eventually spreads, or metastasizes, to other parts of the body. The most common places for chordomas to metastasize are the lungs, liver, bones, or lymph nodes.

Locations of Chordoma

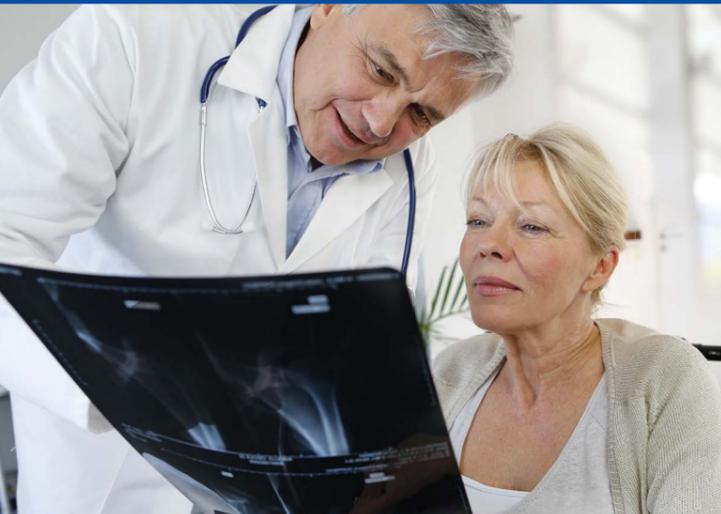
About half of all chordomas form at the bottom of the spine, in bones called the sacrum. About 30 percent form within the center of the head in an area called the skull base — usually in a bone called the **clivus**. The remaining 20 percent of chordomas form in the spine at the level of the neck, chest, 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 known, but researchers are working to figure it out.

Types of Chordoma

There are four types of chordoma, which are classified based on how they look under a microscope. All behave similarly and are treated in the same ways except for **dedifferentiated** chordoma, which occurs in only about 5 percent of patients. Dedifferentiated chordomas are more aggressive and generally grow faster than the other types of chordoma.



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 **magnetic resonance imaging**, also called **MRI**, 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. CT scans of the chest, abdomen, and pelvis are also recommended.

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. See p. 11 for recommendations about having a biopsy.

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. 

Another imaging test called **computed tomography**, also called **CT** or CAT scan is recommended in addition to MRI if it is not certain whether the tumor is chordoma.

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 **trocac CT-guided biopsy** is recommended and should be done from the back.

Trocac 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 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. Please contact support@chordoma.org for assistance. 

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.

Pathology

Tissue samples should be evaluated by a pathologist who has experience diagnosing bone tumors. Your pathologist may test your tumor tissue for the

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 from time to time 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 CT and MRI. 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.

presence of a protein called **brachyury**. Nearly all chordomas have high levels of brachyury, which makes it helpful for diagnosis.

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 is a good idea 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.



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. Your doctors can help you understand the various treatment options that are available to you, and create an individualized treatment plan based on your choices.

The following sections contain important information about surgery and radiation to consider when planning your treatment. 

Tests to Get Before Treatment

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

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 "Tests to Get After Treatment" on p. 24.)

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. 21) 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.

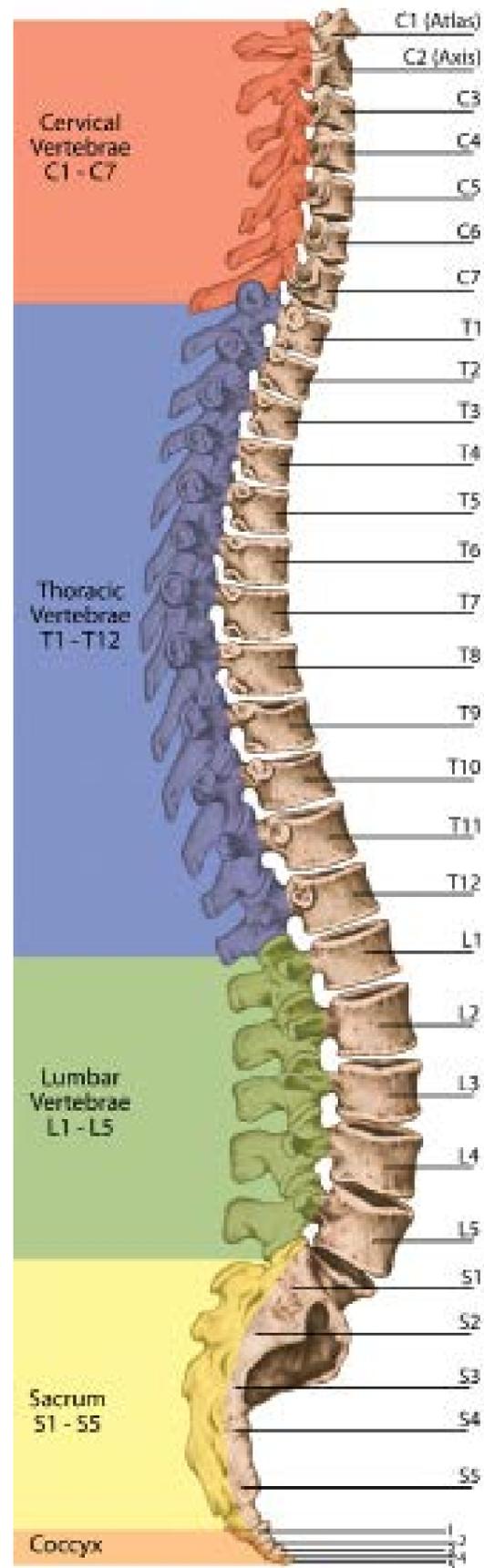


The way surgery is performed can have a big effect on your outcome. Here are three things that all chordoma patients should know before having surgery:

- 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 should be comfortable with the treatment plan agreed on by you and your medical team.

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

Your Spine



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. 21). 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 depending on the location of the tumor and which nerves are involved. 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.

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 to surgery	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



Did you know?

Tumor tissue removed during surgery is critical for research to identify new ways to treat chordoma. When you have surgery, you can help to advance research by contributing extra tissue from your tumor to the Chordoma Foundation Biobank. The purpose of the Biobank is to make tumor samples available to qualified researchers interested in studying chordoma.

To learn more, visit www.chordoma.org/biobank. If you are interested in participating in the Biobank call (877) 230-0164 or email biobank@chordoma.org before you have surgery. 

LUMBAR AND THORACIC TUMORS

The principles of surgery for tumors in this area are generally the same as for sacral tumors. The goal of surgery is to achieve a wide resection, completely removing the tumor in one piece (**en-bloc**) with wide margins, whenever possible. When the tumor is removed, the surgeon should plan to remove the biopsy area 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. Radiation therapy alone rather than surgery should be considered if the tumor cannot be safely removed, or if the potential side effects of surgery are unacceptable to 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.

A wide resection is ideal, whenever possible. However, the location of chordomas near important structures sometimes makes this difficult or impossible.

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.

Any time you are considering treatment it is a good idea to talk with your medical team about the role that radiation therapy should play in your treatment. 

TYPES OF RADIATION

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

Chordomas are generally treated with a beam of radiation that is delivered to the tumor from a source outside of the body. This is called **external beam radiation**. Radiation with beams of charged particles, called **particle therapy**, is generally recommended for treating chordoma because it can be focused most precisely. 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 known whether there is any difference in effectiveness between protons and carbon ions. In some cases, highly focused **photon radiation** can be a suitable alternative to particle therapy as long as a high enough dose can be delivered to the target area without damaging healthy tissue. Sometimes it may be helpful to combine photon radiation and particle therapy. 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**.

Another method of delivering radiation, called **brachytherapy**, involves inserting a small amount of radioactive material inside the body during surgery. This method is used infrequently, 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.

Tests to Get After Treatment

For the first 5 years after your treatment you need an MRI every 6 months. The MRI should look at the area of the original tumor, as well as any areas where it could spread. If no chordoma is present after 5 years, you need an MRI of the area where the tumor was at least once a year for 15 years.

The National Comprehensive Cancer Network guidelines for the treatment of bone tumors also recommends chest imaging every 6 months for 5 years, and then annually thereafter, to see if chordoma has spread to the lungs. Some experts believe you also need an MRI of the whole spine to check for a recurrence of the tumor. It is important to talk with your doctors about what monitoring you need after treatment.

Treatment of Local Recurrence

It is common for chordomas to come back or re-grow after initial treatment.

If your chordoma comes back in the same place as the original tumor it is called a **local recurrence**. When this happens, it is usually no longer possible to be cured; however, additional treatment may be possible that can control the tumor for long periods of time. Treatment options may include surgery, radiation therapy, and sometimes **drug therapy** (see the drug therapy section on p. 26).

Currently, there is not general agreement about the best way to treat recurrent chordoma, but the Chordoma Foundation is working with experts to gather information and develop recommendations that will be available in the future. Talk with your medical team about current treatment options available to you and any possible side effects of those

treatments. It is important to balance the potential benefit of these treatments with the effects that they have on your quality of life.

Treatment of Advanced Disease

Chordomas are considered advanced when a local recurrence can no longer be stopped with surgery or radiation, or when the tumor has spread to other parts of the body.

Cancer that has spread to other parts of the body is called **metastatic**. When chordoma metastasizes it can no longer be cured, and treatment is meant to prolong life and manage symptoms. Treatment for metastatic chordoma can include surgery, radiation, or, in some cases, a procedure called **radiofrequency ablation**, which uses radio waves to heat and destroy the tumor. Additionally, **drug therapy** can slow the progression of advanced or metastatic chordoma.

You should talk with your medical team about all of these options and what treatments are best for your situation.



Quality of Life

Advanced disease and side effects from surgery can cause pain and reduce your quality of life. If you are dealing with pain or other quality of life concerns, palliative care or supportive care specialists may be able to provide treatment options to help with your specific symptoms. Most cancer centers have doctors, as well as nurses and social workers, who can talk with you about supportive care options.

Drug Therapy for Advanced or Metastatic Chordoma

Drug therapy, or **systemic therapy**, is the use of drugs that spread throughout the body to kill cancer cells. This can include drugs that act directly on the tumor as well as drugs that cause the immune system to attack tumors. Systemic therapy is typically prescribed by a type of doctor called a medical oncologist, and sometimes by a neuro-oncologist.

Traditional **chemotherapy**, which kills fast growing cells, generally does not work well on chordoma and is not usually used to treat it. Instead, doctors often prescribe a type of drug called a **targeted therapy** that works by blocking a specific protein (the “target”) in the tumor. Some targeted therapies that have helped chordoma patients improve temporarily include:

- **PDGFR** inhibitors such as imatinib and sunitinib
- **EGFR** inhibitors such as erlotinib, gefitinib, and cetuximab

One study showed that a type of drug called an mTOR inhibitor, when combined with imatinib, was more effective than imatinib alone.

These drugs may be prescribed to treat chordoma even though they have not been approved by government agencies for this use. This is called **off-label** use and it is legal. However, in some countries the cost of drugs used off-label is not always covered by insurance or healthcare systems.

One way to find out more about which targeted therapies might work best for you is to have molecular profiling tests done on your tumor tissue. Every cancer tumor has genetic mutations, and these profiling tests provide you and your doctor with more information about the mutations in your individual tumor. A Chordoma Foundation patient navigator is available to talk with you about these tests by emailing support@chordoma.org.

Researchers are working to understand more about chordoma and to identify additional targeted therapies that could be effective. A summary of the latest information about chordoma which could help your oncologist choose a targeted therapy can be found at www.chordoma.org/targets. 



Research and New Treatments

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

Though there is much about the treatment of chordoma that experts agree on, many questions still remain about how best to treat chordoma patients in certain situations, particularly local recurrences. The Chordoma Foundation is working with doctors and researchers to conduct studies to help answer these questions.

As researchers learn more about chordoma, evidence is regularly emerging about new treatment approaches that could possibly help chordoma patients. To know whether those treatments are safe and effective, they need to be carefully tested in chordoma patients through research studies called **clinical trials**. The Chordoma Foundation is working with doctors around the world to start more clinical trials for chordoma.

Participating in a clinical trial can give you access to promising new treatments that could be more effective than other treatments available to you. For a list of clinical trials open to chordoma patients see www.chordoma.org/clinical-trials.

Cancer experts encourage patients to participate in clinical trials whenever possible. Clinical trials are especially important for patients with rare diseases like chordoma because they provide care in a way that is very structured and closely monitored. If you are interested in searching for a clinical trial that is right for you and your stage of treatment, talk with your doctor or contact a Chordoma Foundation patient navigator at support@chordoma.org. 

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 gene that is present at high levels in nearly all chordoma tumors.

carbon ion therapy - A type of **particle therapy** (see p. 29) that uses beams of carbon ions to kill cancer cells.

chemotherapy - See **systemic therapy** on p. 30.

clinical trials - Research studies that are done to test whether a treatment is safe and effective for patients with a specific disease.

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

computed tomography (CT) scan - A type of imaging scan that is used to help diagnose chordoma. 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.

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.

diffusion MRI (D-MRI) - A type of **MRI** (see p. 29) which can help doctors tell the difference between chordoma and chondrosarcoma, in order to make a correct diagnosis.

drug therapy - See **systemic therapy** on p. 30.

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

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.

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.

Glossary of Terms (continued)

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** on p. 30 and **carbon ion therapy** on p. 28.

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.

proton therapy, proton beam therapy - A type of **particle therapy** that uses beams of protons to kill cancer cells.

radiofrequency ablation - A procedure that uses a needle to deliver energy to the tumor, causing it to heat up, and killing the cancer cells within it.

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.

systemic therapy - The use of drugs that spread through the body to kill cancer cells. Also called **chemotherapy**, or **drug therapy**. Chemotherapy typically refers to drugs that kill fast growing cells.

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.

trocarn 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.

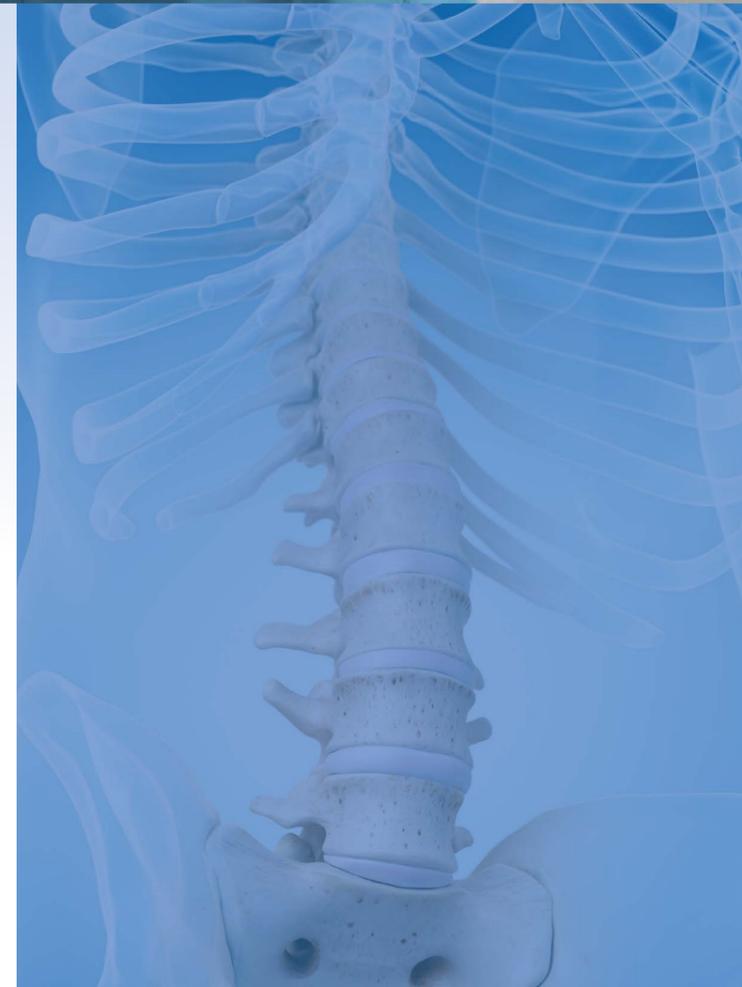


Learn more about chordoma

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

Get help from a patient navigator at the Chordoma Foundation by emailing support@chordoma.org or calling (888) 502-6109.

Connect with others through the Chordoma Foundation peer support program by emailing peersupport@chordoma.org.





Important note about this publication: *The content of “Expert Recommendations for the Diagnosis and Treatment of Chordoma” was developed based on the treatment guidelines paper published in The Lancet Oncology in February 2015, following the first meeting of a global consensus group of chordoma experts (see inside front cover for full article citation and complete list of consensus group members). The consensus group members and the Chordoma Foundation Medical Advisory Board have reviewed this guide for accuracy of its content. This information is not meant to take the place of medical advice. You should always talk with your doctors about treatment decisions. If you have questions about the information contained in this document, please contact a Chordoma Foundation patient navigator at support@chordoma.org.*