



Expert Recommendations for the Treatment of Recurrent Chordoma



The Chordoma Foundation developed this booklet based on the article, "Best practices for the management of local-regional recurrent chordoma: a position paper by the Chordoma Global Consensus Group", which was written by an international group of chordoma experts with support from the Chordoma Foundation. The article was published in the medical journal *Annals of Oncology* in June 2017.

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Introduction

If you have been treated for chordoma in the past and are now being told your tumor is growing again, you probably have a number of questions. This booklet is intended to help answer those questions so you can make informed treatment decisions and obtain 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 60 doctors who specialize in caring for chordoma patients. The Chordoma Foundation and the European Society for Medical Oncology brought this group together to define evidence-based recommendations for treating chordoma.

The consensus group first met in December 2013 and developed guidelines for medical professionals on the diagnosis and treatment of primary chordomas, as well as chordoma tumors that are advanced or metastatic. The Chordoma Foundation made these recommendations available to patients and caregivers in our booklet, *Expert Recommendations for the Diagnosis and Treatment of Chordoma*. This booklet can be viewed and downloaded at chordoma.org/educational-materials.

The consensus group came together again in November 2015 to develop detailed recommendations for treating locally and regionally recurrent chordoma – a complex topic which required in-depth discussion and careful analysis of available evidence. The resulting guidelines were published in an open-access paper in the high-profile medical journal, *Annals of Oncology*, in June 2017. The full paper can be accessed at chordoma.org/recurrence-guidelines. As part of our commitment to helping patients and caregivers make the most informed treatment choices, we are now making the guidelines for recurrent chordoma available to you here in this booklet.

How to use this booklet

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

1. We recommend that you start by reading through the "Understanding recurrent chordoma" section, which contains important background information.
2. Flow charts throughout the booklet summarize the information in each section and walk you through steps you and your treatment team should take to determine whether you have a recurrence and what treatment options are available to you. **Please read all information carefully and discuss it with your doctors.**
3. Toward the end of the booklet you will find a section dedicated to comprehensive palliative and supportive care – specialty care that addresses pain management and any other side effects to help you live well and feel your best while dealing with chordoma. Palliative care is important and can be beneficial for all chordoma patients during diagnosis, treatment, and after treatment ends.
4. Text noted with the Chordoma Foundation logo  contains additional information, explanation, and tips from the Chordoma Foundation to help you understand and act on the expert recommendations.
5. Terms in **bold blue font** are included in the glossary (p. 32). 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.

Understanding recurrent chordoma

More than half of all chordoma tumors grow again after initial treatment. This is called a **recurrence**. It is not unusual for chordoma patients to have a recurrence several years after initial treatment. Many patients have more than one recurrence over time.

If the tumor grows back in the same place as the original tumor, this is known as a **local recurrence**. If it grows back in an area close to where the original tumor grew, this is called a **regional recurrence**. A regional recurrence is the result of tumor spreading to areas in direct physical contact with the site of the original tumor, such as nearby bones or muscles. When tumor spreads beyond the site of the original tumor to other areas of the body, this is called **metastasis** or **metastatic disease**.

There is no way to know for sure if a chordoma will recur because several factors affect your chance of having a recurrence. These include the size of your original tumor, how much of the tumor was removed during your initial surgery, your age, and the nature of your initial surgery and/or radiation.

Chordomas that recur are difficult to control and are rarely curable. However, there are certain circumstances in which a recurrent chordoma might be cured with appropriate treatment. And, even when a recurrence is not curable, there are treatment options that can help you live longer with good quality of life. Understanding when a cure is and is not possible — as explained later in this booklet — is critical in determining how to confront a recurrence. This knowledge can help you and your medical team pursue a treatment plan that offers the best outcome possible, while avoiding treatments that can expose you to unnecessary or unacceptable risks.

Finding the right medical team


When confronting a recurrence, selecting the right medical team is a critical first step. Depending on your situation, the medical center where you were treated initially may or may not be best suited to diagnose and treat a recurrence.

It is important to be evaluated and treated by a **multidisciplinary team** of specialists who have substantial experience treating chordoma. Teams with this experience are typically only found at larger hospitals, sometimes called **referral centers**, which see large numbers of patients.

Experts recommend your treatment team include the following specialists with experience diagnosing and treating chordoma:

- Pathologist
- Radiologist
- Radiation oncologist
- Surgeon
- Medical oncologist
- Palliative care specialist



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. 

Diagnosing a recurrence

In some cases, worsening or new symptoms are the first sign of a recurrence. Other times, new tumor growth is seen on routine follow-up scans. In either situation, your doctors will need to do further testing to learn more about what is happening.

Imaging

Your doctors will need to order imaging tests to determine if your chordoma has recurred. If it has, the imaging will provide more information about the new tumor growth, such as the size and exact location.

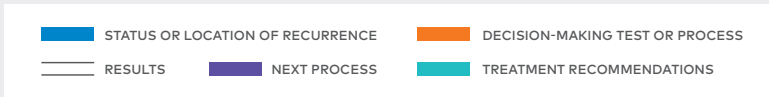
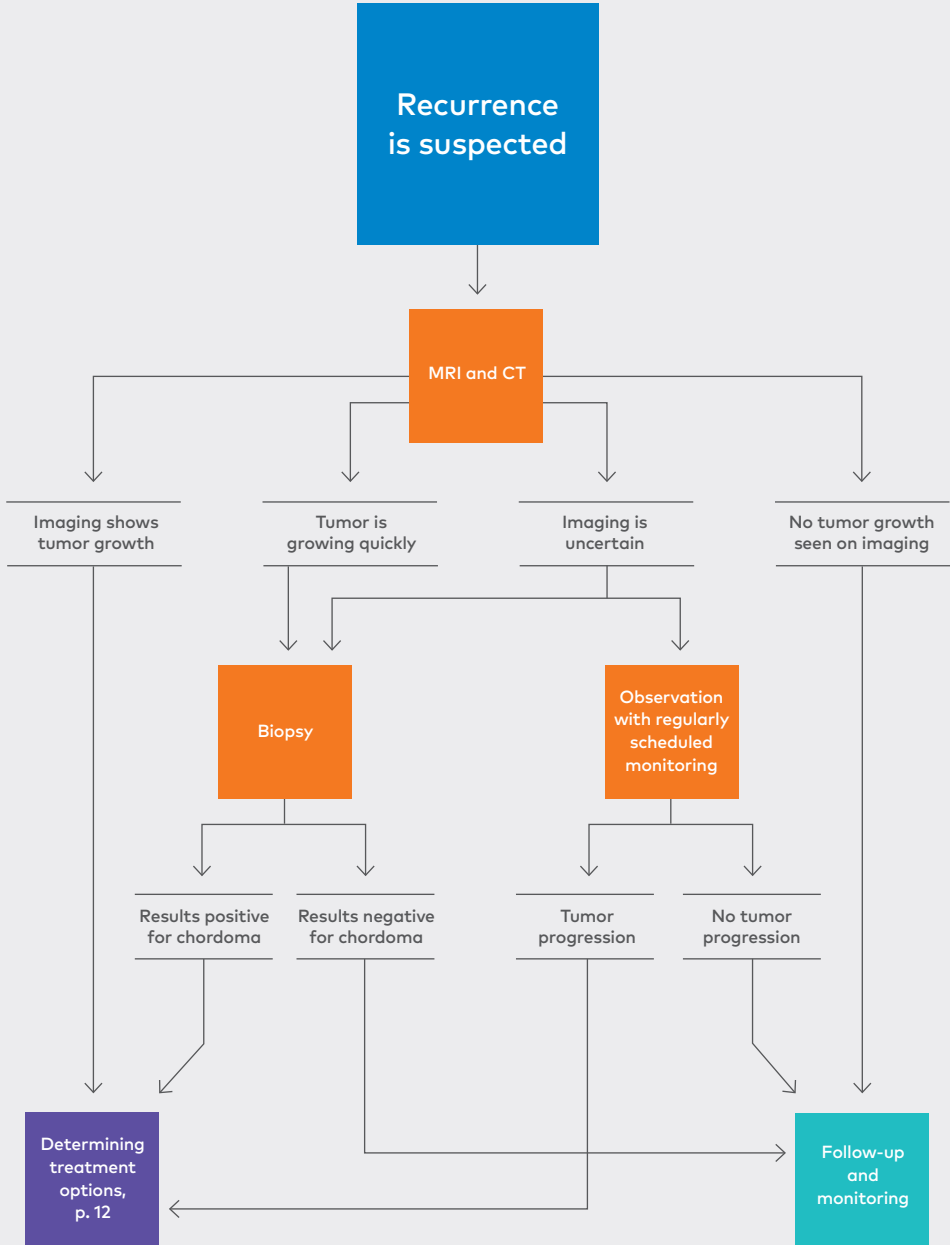
The first imaging test that should be done is **magnetic resonance imaging**, or **MRI**. Your MRI should be done with **contrast**. Contrast agents are special liquids injected into a vein which help organs and tissues appear more clearly on MRI images. **Computed tomography**, or **CT**, scans may also be ordered.

Biopsy

If your doctors cannot determine from imaging tests whether your tumor has recurred, a **biopsy** may be recommended, if it can be done safely. A biopsy may also be performed if your tumor is growing unusually fast or if your doctors think you may have developed a new form of cancer that is not chordoma. A **core-needle biopsy** is generally recommended and should be performed by doctors who have experience with chordoma. The tissue sample should be examined by a pathologist with experience diagnosing chordoma.

If your tumor is growing quickly, the tissue sample should be analyzed to determine if the tumor has become **poorly differentiated** or **dedifferentiated**, both of which may require different treatment approaches. Poorly differentiated chordomas often lose expression of a protein called **INI-1**, and sometimes dedifferentiated and **conventional chordoma** can as well. Loss of the INI-1 protein can be determined with a simple test performed by a pathologist on a sample of tumor tissue.

If your doctors are unsure whether your tumor has recurred and you are not experiencing any symptoms, it may be possible to wait and do imaging of the area again after a period of observation rather than performing a biopsy.



Determining treatment options

There are a number of factors that will affect what treatment options are available to you. Each patient's situation is unique and must be evaluated individually. Decisions about treatment should balance side effects, quality of life, and how well the treatment is expected to work. This will help ensure that you get the best treatment for your tumor with as little impact on your quality of life as possible.

Curative treatments versus treatments to delay tumor progression

When a recurrence is confirmed, it is important for your doctors to perform a full evaluation to help determine your options for treatment and whether the goal of these treatments will be to cure the disease or to delay tumor progression as long as possible. You and your doctors should consider a number of factors when making these determinations, including:

- where the tumor has recurred
- what treatments you have had in the past
- what side effects might be caused by the treatment
- what risks would be acceptable to you

Factors such as your age, other health conditions you have, and your ability to care for yourself and perform daily activities are also important for you and your treatment team to discuss.

A cure for recurrent chordoma – meaning that the tumor is permanently controlled – is only possible in a small number of cases. However, it is important to recognize when the potential for a cure exists, and use this knowledge to help guide treatment decisions. The two most important



factors that determine whether the goal of treatment is to cure the disease, which is also called **curative intent**, or whether the goal is to delay tumor progression as long as possible are:

1. the extent of the recurrence, and
2. whether high-dose radiation is possible.

Important points to consider

- For any tumor location, the decision to have surgery alone, surgery plus radiation, or radiation alone should be made together with your doctors, based on your individual situation. There is currently no published data to support the same recommendations for all patients.
- If you previously had high-dose radiation near the location of your recurrence, your doctors should take particular caution when planning for surgery because there is a higher risk of complications and wound healing issues with tissue that has been treated with radiation.
- Treatments for recurrent chordoma may cause serious side effects, so it is important to carefully consider and discuss with your treatment team the risks and benefits of any treatment option, along with how the treatment will impact your overall health and quality of life.

EXTENT OF RECURRENCE

In order for your doctors to determine exactly where the tumor has recurred and to what degree, you will need to have more extensive imaging scans. A CT scan of your whole body should be done, as well as an MRI of your entire spine, to determine whether the tumor has spread. Your doctors should also compare your new MRI and CT images to those that were taken after your last treatments. This will help them determine how much of what is seen in the new images is recurrent tumor, and what might be **treatment sequelae** (changes that are a result of treatment).

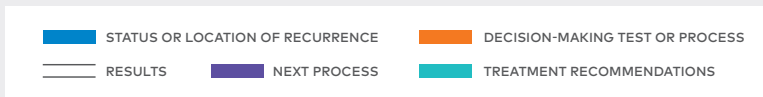
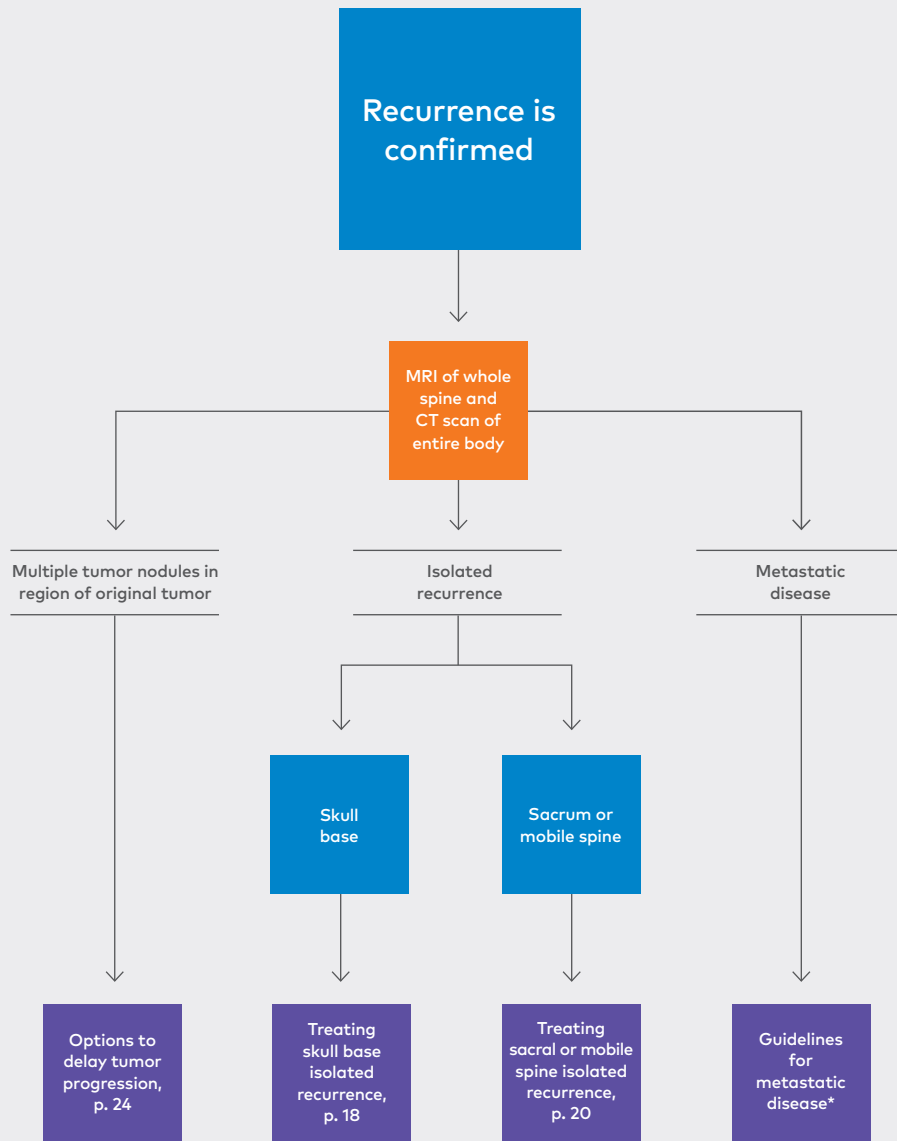
The results of this pre-treatment imaging will show your doctors whether you have:

- an **isolated recurrence**, which is a single tumor at or near the site of the original tumor;
- a **multifocal recurrence**, which is multiple tumors at or near the site of the original tumor;
- or **metastatic disease**, which is one or more tumors in other parts of the body in addition to the recurrent tumor.

An isolated recurrence may be curable under certain conditions. In this case, high-dose radiation and surgery should be considered first to give you the best chance for a cure.

Multifocal recurrences are very unlikely to be cured and currently there is no known cure for metastatic disease. However, multiple treatment options are available to control or temporarily stop tumor growth, as well as to alleviate symptoms. These include **debulking** surgery, low-dose radiation including **stereotactic body radiation therapy (SBRT)** and **stereotactic radiosurgery (SRS)**, **ablative therapy**, and **systemic therapy** (see "Options to delay tumor progression" beginning on p. 24). In many cases, these therapies can control the disease for multiple years while allowing patients to maintain their quality of life.

If the tumor does not appear to be growing and you are not experiencing symptoms, your doctors might recommend a period of observation followed by more imaging to determine what treatment is necessary.



*See *Expert Recommendations for the Diagnosis and Treatment of Chordoma* booklet

POSSIBILITY OF HIGH-DOSE RADIATION


For patients with isolated recurrences, high-dose radiation given with or without surgery provides the best chance of achieving a cure or long-term disease control. A dose of at least 74 GyE to the entire tumor is required, so if you have an isolated recurrence an important first step is for your doctors to determine whether this amount of radiation can be safely delivered.

There is a lifetime maximum amount of radiation that healthy tissues like bone, nerves, and arteries can receive without being harmed. If you have never had radiation, you can very likely receive high-dose radiation to treat new tumor growth.

If you have had radiation in the past and the recurrence is in an area that was radiated, it will likely not be safe for you to have high-dose radiation again.

Palliative and supportive care

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. See the “Comprehensive palliative and supportive care” section on p. 28 for more information. 

However, if the recurrence is outside or at the edge of the area that was previously radiated, it may be possible to have additional high-dose radiation. Every case is unique, so it is important for a radiation oncologist with experience treating chordoma to review your previous radiation plans in relation to the current tumor growth.

The treatment recommendations for isolated recurrences differ based on the location of the tumor. The next sections discuss in detail the pathways that determine the recommendations for each location.

For patients with multifocal recurrences, metastatic disease, or if radiation cannot be given at a dose of at least 74 GyE, a lower dose of radiation may still be able to help control growth of the tumor or alleviate symptoms, but it will not be curative (see “Options to delay tumor growth” beginning on p. 24).


TESTS TO GET BEFORE TREATMENT

No matter the extent of your recurrence, your doctors should perform a complete physical and neurological assessment to help prepare for treatment. This includes discussing any symptoms you are experiencing and how fast they are progressing. When evaluating your pain levels, your doctors should note what pain is related to previous treatments and what is related to the new tumor growth. If you have a skull base chordoma, you might also need evaluation of your vision, hearing, and endocrine system function. These tests will help your doctors determine the effects of the new tumor growth and consider this information in treatment planning.



Did you know?

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. 

Treating skull base isolated recurrence

As discussed in the previous section, your doctors should first determine whether you are able to have high-dose radiation treatments, based on what radiation treatments you have had in the past. It is important for a radiation oncologist who has experience treating skull base chordoma to examine your past radiation treatment plans in relation to the current tumor growth to make this determination.

If your doctors determine that you are able to have high-dose radiation, surgery should also be considered in order to help increase the effectiveness of radiation. If surgery is not possible, high-dose radiation alone can be considered. There is very little data comparing the effectiveness of surgery plus radiation versus radiation alone for recurrent skull base tumors. These options should be discussed in detail with your doctors.

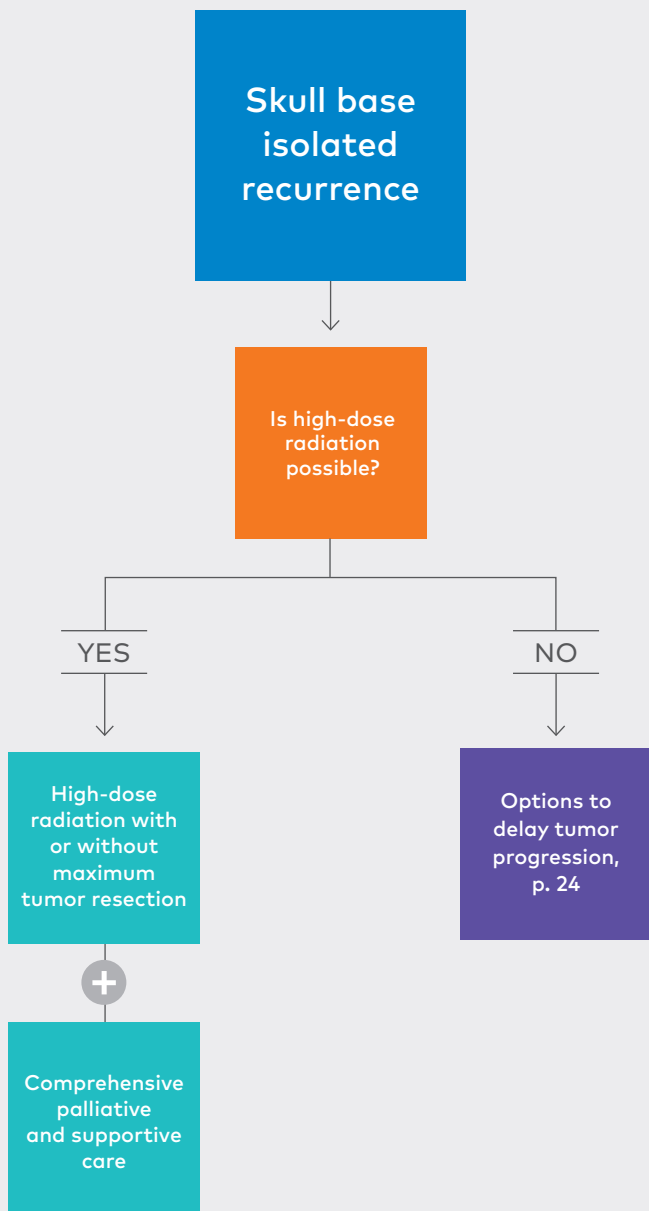
If your doctors determine that you are unable to have high-dose radiation and your tumor is progressing or you are experiencing symptoms, it is recommended that you consider other treatment options such as debulking surgery, low-dose radiation, or systemic therapy. More information on these options begins on p. 24 of this booklet.






A comprehensive palliative and supportive care plan should be part of your overall treatment plan to help address any side effects or other quality of life concerns and provide support for you and your family members. See p. 28 for more information on palliative and supportive care.

If your tumor is slow-growing or you are not experiencing symptoms, your doctors might recommend a period of observation before treatment.

Goals of surgery for skull base isolated recurrence

In general, the goal of surgery for recurrent skull base tumors is complete resection. However, this is often not possible for recurrent skull base tumors. In these cases, debulking surgery can be considered. See p. 24 for more information on debulking surgery.



 STATUS OR LOCATION OF RECURRENCE	 DECISION-MAKING TEST OR PROCESS	
 RESULTS	 NEXT PROCESS	 TREATMENT RECOMMENDATIONS

Treating sacral or mobile spine isolated recurrence

For sacral and mobile spine (cervical, thoracic, and lumbar) recurrences, your doctors should first consider:

- whether your tumor ruptured during previous surgery or was taken out in more than one piece
- what radiation treatments you have received

This will help determine whether surgery alone, surgery plus radiation, or radiation alone is the best option for you. It is important for a radiation oncologist who has experience with chordoma to examine any past radiation treatment plans in relation to your current tumor growth to make this determination.

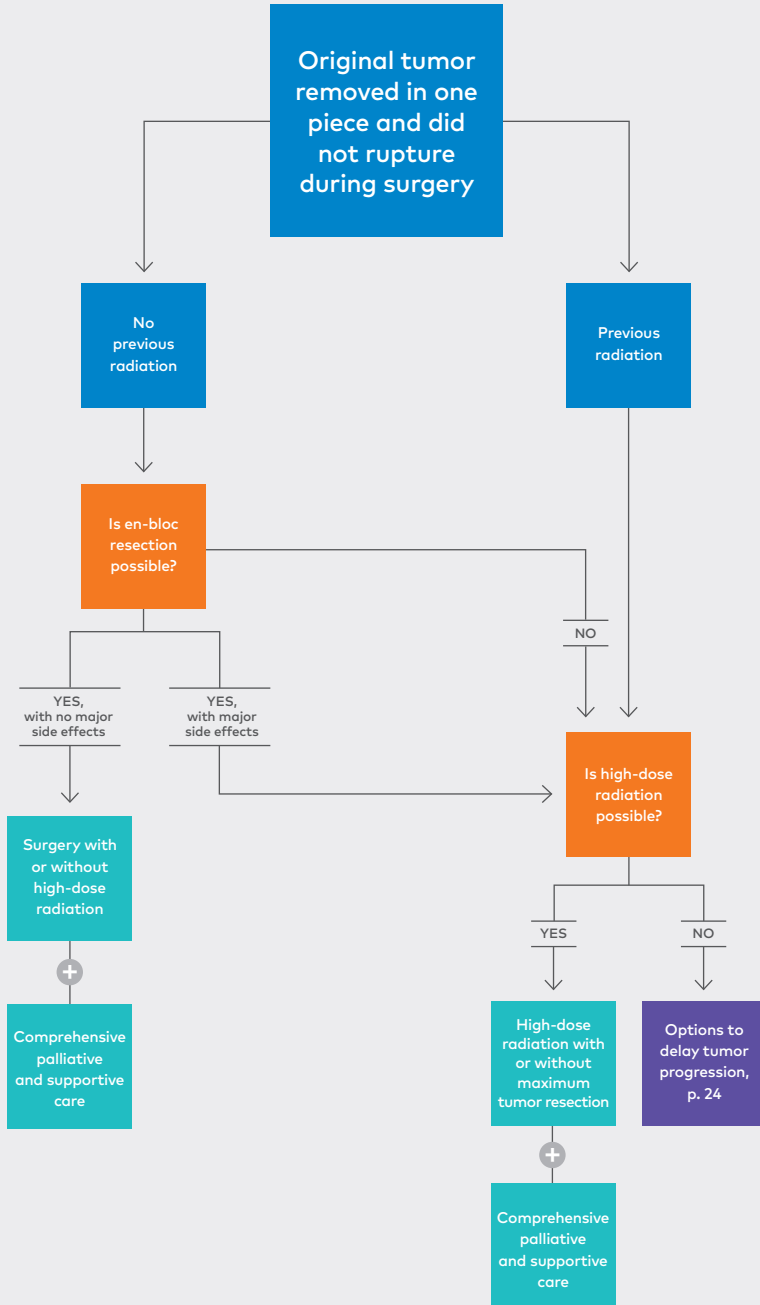
If your original tumor was completely removed in one piece (en-bloc resection) and:

You **have not** had radiation, surgery to remove the entire tumor should be considered first.

- If en-bloc resection is possible and the risks of side effects are acceptable, surgery is recommended. High-dose radiation may also be recommended following surgery.
- If en-bloc resection is not possible or the risks of side effects are not acceptable, high-dose radiation alone should be considered.
 - » If high-dose radiation is not possible, other treatment options should be considered (see "Options to delay tumor progression" on p. 24).

You **have** had radiation, your doctors will need to determine whether you can have further high-dose radiation.

- If high-dose radiation is possible, this is the recommended treatment. Surgery to remove as much tumor as possible should also be considered.
- If high-dose radiation is not possible, other treatment options should be considered (see "Options to delay tumor progression" on p. 24).



If your original tumor ruptured during previous surgery or was taken out in more than one piece (not en-bloc), your doctors will need to determine whether you can receive high-dose radiation.

- If high-dose radiation is possible, the recommended treatment is high-dose radiation alone, without surgery.
- If high-dose radiation is not possible, other treatment options should be considered (see "Options to delay tumor progression" on p. 24).

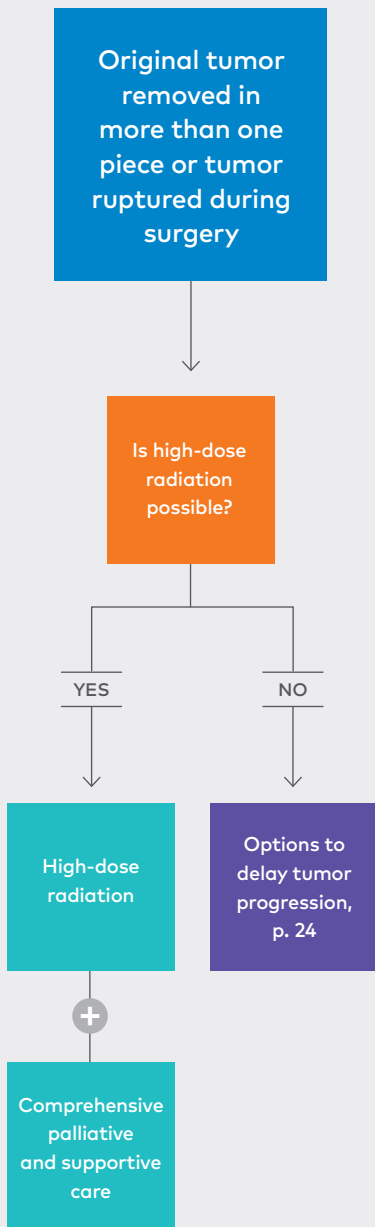
A comprehensive palliative and supportive care plan should be part of your overall treatment plan, to help address any side effects or other quality of life concerns and provide support for you and your family members. See p. 28 for more information on palliative and supportive care.






If your doctors are unsure of the best treatment option for you, a period of observation is recommended if your disease is stable, your tumor is slow-growing, or you do not have noticeable symptoms.

Goals of surgery for sacral and mobile spine isolated recurrence

In general, the goal of surgery for sacral or mobile spine tumors is to remove the tumor in one piece (en-bloc resection) with surgical **margins** of at least 1mm of healthy tissue. It is very important that all attempts are made to limit the risk of rupturing the tumor during surgery, which can cause tumor cells to spread.

Recurrences within the chest cavity, abdomen, and pelvis cannot typically be removed in one piece. The goal of surgery in these cases is to remove as much of the tumor as possible to increase the effectiveness of later radiation.



 STATUS OR LOCATION OF RECURRENCE	 DECISION-MAKING TEST OR PROCESS
 RESULTS	 NEXT PROCESS
	 TREATMENT RECOMMENDATIONS

Options to delay tumor progression

When curative treatment is not possible, there are a number of treatment options to consider with your doctors to help manage your disease. With the right care, your tumor may be controlled for many years while also having good quality of life.

Debulking surgery

In a debulking surgery some of the tumor is removed in order to help relieve or avoid symptoms caused by compression of important structures such as nerves, spinal cord, or the brainstem. It can also be used to separate healthy tissues from your tumor so that you can more safely undergo radiation if necessary. Debulking surgery is recommended only in certain cases, because it is not curative and the chance of serious side effects increases with each surgery.

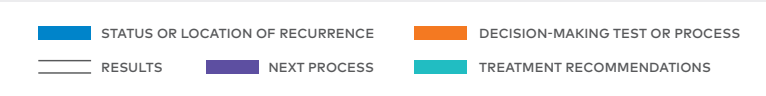
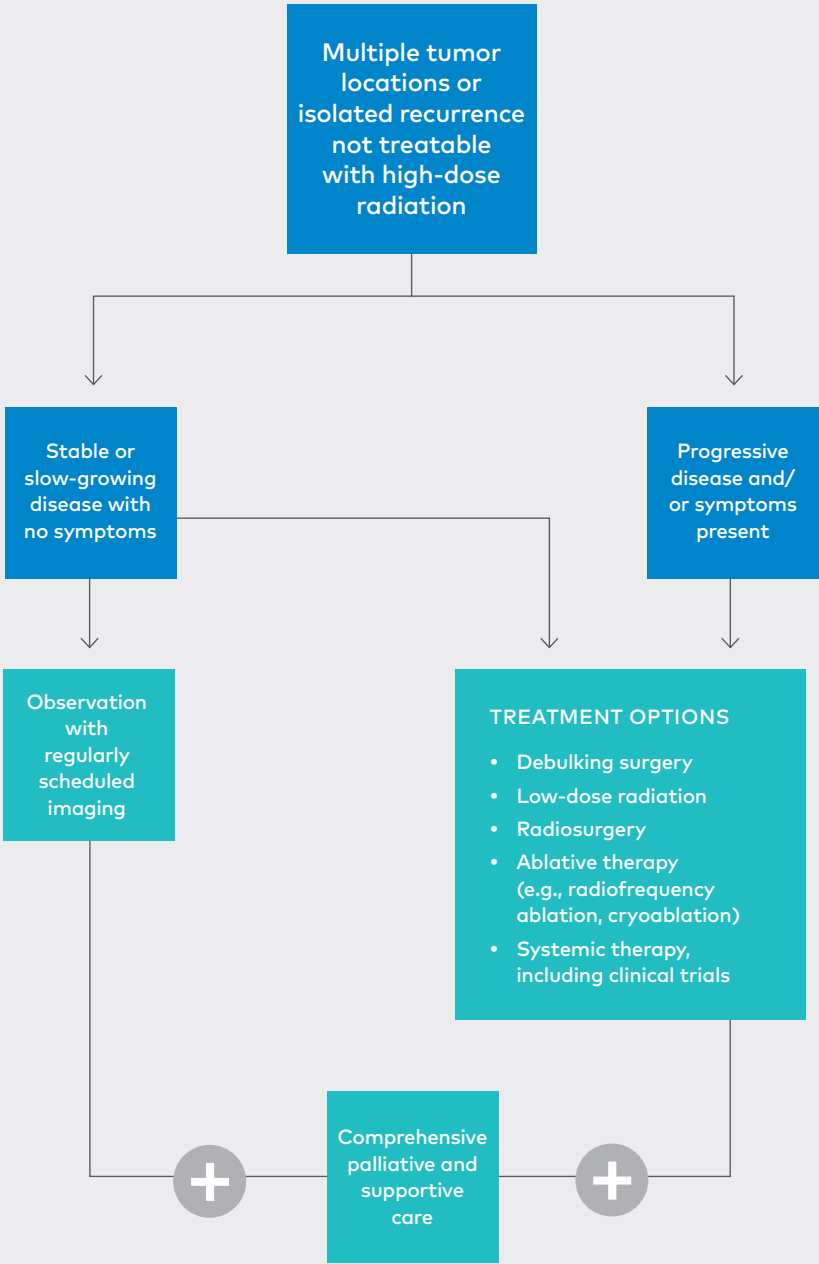
Low-dose radiation


Low-dose radiation may be given alone or following a debulking surgery to slow tumor progression or relieve symptoms. Your doctors should consider all previous radiation treatments and the location of the recurrent tumor to determine whether radiation is safe for you.

It may also be possible for you to have stereotactic radiosurgery (SRS) or stereotactic body radiation therapy (SBRT), which are both types of **hypofractionated radiation** (radiation given in a small number of doses). When hypofractionation is used, the total amount of radiation given is less but the effect is thought to be the same as standard fractionation.

Ablative therapy

There is some evidence to suggest that ablative therapies like **cryoablation**, **radiofrequency ablation (RFA)**, or **high-intensity focused ultrasound (HIFU)** can be used to help manage symptoms caused by recurrent tumors. Cryoablation destroys cancer cells with extreme cold, while RFA uses heat. Both are delivered through small probes inserted directly into the tumor. HIFU destroys cancer cells with high-frequency sound waves delivered from



Systemic therapies that have been approved to treat other cancers can be prescribed to chordoma patients off-label by a medical oncologist. Off-label prescribing is allowed as long as there is a good reason and a low risk of harm. However, in some countries the cost of drugs used off-label is not always covered by insurance or healthcare systems, and therefore could be very expensive for the patient. It is important to talk with your doctors about the options in this section. More information about systemic therapies that may be off-label options for chordoma patients can be found at [chordoma.org/systemic-therapy](https://www.chordoma.org/systemic-therapy). 


outside of the body. More research is needed to determine how effective these procedures are for treating chordoma, but they are options you can discuss further with your doctors.

Systemic therapy

There are no drugs currently approved by any government regulatory agencies for the treatment of chordoma. However, evidence has shown that certain types of systemic therapies such as **targeted therapy** and **immunotherapy** that are commonly used to treat other cancers may also help patients with recurrent chordoma.

Conventional chemotherapy is typically not effective in treating chordoma. However, some patients with poorly differentiated or dedifferentiated chordoma have benefitted from sarcoma chemotherapy regimens.

How do I decide?

Every patient's situation is unique. If your tumor cannot be treated with high-dose radiation, it is important to discuss all of the options in this section in detail with your medical team and your family, weighing the risks and benefits of each option for your situation. The treatment options listed here involve various types of doctors, so it is important to consult with a team of doctors who specialize in these treatments and have experience treating chordoma patients. For some patients, just one type of treatment might be the best decision, while for others a combination of treatments might provide the best option. 

The Chordoma Foundation's Medical Advisory Board (MAB) recommends that patients with recurrent chordoma who cannot be treated with surgery or high-dose radiation consult with their medical team — including an experienced chordoma medical oncologist — about whether participating in a **clinical trial** might be right for them. Each patient's situation is unique, but in general, the MAB recommends patients pursue systemic therapy treatment options in the following order of priority:

1. Chordoma-specific clinical trials


Start by considering trials designed specifically for chordoma patients, or those that are enrolling a group of chordoma patients to a specific arm of the trial. These trials are likely to have strong scientific justification and to be conducted by teams with significant experience caring for chordoma patients.

2. Other relevant clinical trials recommended by an experienced physician

If you are not eligible for any chordoma-specific trials, ask your oncologist about other clinical trials that might be options for you.

3. Off-label therapy with evidence of clinical benefit for chordoma patients

If you are not eligible for any clinical trials, consult with your oncologist about off-label use of approved drugs that have been used to treat chordoma patients.

The Chordoma Foundation has developed resources to assist patients who are considering systemic therapies. A list of chordoma-specific trials as well as chordoma-relevant trials recommended by the MAB are available at chordoma.org/clinical-trials and more information on systemic therapies that may be off-label options for chordoma patients can be found at chordoma.org/systemic-therapy. For more information contact a Chordoma Foundation Patient Navigator at chordoma.org/request-help. 

Comprehensive palliative and supportive care

Palliative care is an important part of cancer care. Sometimes called supportive care, it is recommended for all cancer patients from the time of diagnosis, through all stages of treatment, and after treatment is complete to address symptoms of the disease itself or side effects of its treatment.

The intent of palliative medicine is not to treat or cure your disease, but to ease symptoms and side effects. You and your care team will determine which palliative care treatments are right for you based on your symptoms and other individual needs.

In addition to managing physical symptoms and side effects, palliative care also includes other sources of support to meet your emotional, spiritual, practical, and social needs, and to help you and your family plan for the future.

Physical medicine and rehabilitation, anesthesiology, psychology, social work, radiology, and pain medicine are just some of the specialties that can provide palliative care to chordoma patients. A comprehensive palliative care plan should include:


- **Pain management**, including careful assessment of the cause of your pain. For instance, it is important to determine whether the pain is the result of your original tumor, past treatments, or the new tumor growth. Most chordoma patients experience both **somatic pain** and **neuropathic pain**, which should be correctly diagnosed and treated using pain management guidelines and specialized pain management techniques as needed. Management of pain should be a critical part of every chordoma patient's care plan.
- **Management of other symptoms** such as nausea and vomiting, fatigue, breathing or swallowing difficulties, mobility issues, and loss of bowel, bladder, or sexual function.



- **Psychological support** to help you cope with the emotional difficulties of chordoma. Anxiety and depression are common for many patients dealing with cancer and should be addressed. This can include family-oriented support to help you and your family find ways that you can best support each other.
- **Assistance considering end-of-life decisions**, such as the creation of **advanced medical directives**, as well as determining what type of home or other care setting will be best for you and your family that will ensure continuity of care.

What is palliative care?

According to the World Health Organization (WHO), "palliative care is an approach that improves the quality of life of patients and their families facing the problems associated with life-threatening illness, through the prevention and relief of suffering by means of early identification and impeccable assessment and treatment of pain and other problems, physical, psychosocial, and spiritual."


A Chordoma Foundation Patient Navigator can assist you in learning more about palliative care options and how to access them. Contact a Patient Navigator at chordoma.org/request-help or by calling (888) 502-6109. 



Follow-up after treatment

You should have an MRI every three to six months for the next three years following any treatment you have for your recurrence. Beyond that, there is not enough data for recommendations of an optimal follow-up schedule for recurrent chordoma. Your doctors will use their best judgment based on the state of your disease, general health, ongoing treatments, and other factors to decide on follow-up care. You should be vigilant about caring for yourself and checking in with your doctors.

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.

In addition, Chordoma Foundation Patient Navigators are 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

ablative therapy

A treatment that delivers extreme heat or cold to a tumor using small needles or probes inserted directly into the tumor. These therapies include cryoablation, high-intensity focused ultrasound, and radiofrequency ablation.

advanced medical directive

A legal document that specifies what actions should or should not be taken for an individual's health if they are no longer able to make decisions for themselves because of illness or incapacitation.

biopsy

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

chemotherapy

A type of systemic therapy that is designed to kill rapidly dividing cancer cells.

clinical trial

Research studies involving human patients that are done to test whether drugs are safe, and how well they will work to treat a specific disease.

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.

contrast

A dye or other substance that is injected into a vein to help areas of the body show up more clearly on scans like MRIs and CTs.

conventional chordoma

The most common type of chordoma, also called classic or classical chordoma. This type is typically slow growing.

core-needle biopsy

A type of biopsy performed with a wide needle. Also called a core biopsy.

cryoablation

A type of ablative therapy that uses a needle to deliver extreme cold to a tumor in order to kill cancer cells.

curative intent

Treatments that are given for the purpose of curing a disease or providing long-term survival.

debulking

Surgical removal of part of a tumor.

dedifferentiated

A type of chordoma that is more aggressive and usually grows faster than conventional chordoma.

Dedifferentiated chordomas occur in only 5 percent of patients.

en-bloc resection

Surgical removal of an entire tumor in one piece.

high-intensity focused ultrasound, or HIFU

A type of ablative therapy that destroys cancer cells with high-frequency sound waves delivered from outside of the body.

hospice

A specific type of supportive care that is provided to patients who are near the end of life and have stopped treatments meant to cure or control their disease. The main goal is to help patients feel as comfortable as possible, and to support both patients and family members through end of life. If a treatment option becomes available, patients can be taken out of hospice care and receive that treatment.

hypofractionated radiation

Radiation treatment in which the total dose of radiation is divided into a small number of large doses and given over a short period of time, usually 1-5 days.

immunotherapy

A type of systemic therapy that is designed to stimulate the immune system to help the body fight disease.

INI-1

A protein that regulates the growth of certain tumor cells. INI-1 is lost in many poorly differentiated chordoma tumors, and, very rarely, in some conventional and dedifferentiated chordomas.

isolated recurrence

A single recurrent tumor at or near the site of the original tumor.

local recurrence

Tumor that has grown back in the same location after treatment.

magnetic resonance imaging, or MRI

A type of imaging scan that uses magnetic fields to help visualize and diagnose cancer, and is used after treatment ends to check for recurrence or metastasis.

margins, or 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. Wider margins provide a lower chance of recurrence.

metastasis, metastatic disease

The process of cancer cells spreading to other parts of the body. These tumors are called metastases, or metastatic disease.

multidisciplinary team

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

multifocal recurrence

Multiple tumors at or near the site of the original tumor.

neuropathic pain

Chronic pain caused by nerve damage.

off-label

The practice of prescribing drugs that are not approved by government agencies to treat a particular disease. Doctors are allowed to prescribe drugs off-label at their discretion as long there is good reason and it would not put a patient at undue risk of harm.

palliative care

Care given to improve the quality of life of patients who have a serious or life-threatening disease. Also called supportive care.

poorly differentiated

A type of chordoma that is more aggressive and usually grows faster than the conventional type. It is more common in children and young adults, and is often characterized by loss of the INI-1 protein.

radiofrequency ablation, or RFA

A type of ablative therapy that uses a needle to deliver heat and electrical energy to a tumor in order to kill cancer cells.

recurrence

Tumor that has grown back after initial treatment.

regional recurrence

Tumor that has grown back in the area adjacent to where the primary tumor was located.

referral center

A hospital, treatment center, or network of treatment centers where doctors have significant expertise in particular diseases.

somatic pain

Pain that results from the activation of sensory receptors even in the absence of injury or damage. It is one of the most common types of pain experienced by cancer patients.

stereotactic body radiation therapy, or SBRT

A type of external radiation therapy that uses special equipment to position a patient and precisely deliver radiation to tumors in all parts of the body except the brain, over a small number of treatments.

stereotactic radiosurgery, or SRS

A type of external radiation therapy that uses special equipment to position the patient and precisely deliver radiation to tumors in or near the brain, over a small number of treatments.

supportive care

See palliative care.

systemic therapy

The use of drugs that spread through the body to kill cancer cells. It includes chemotherapy, targeted therapy, and immunotherapy.

targeted therapy

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

treatment sequelae

A chronic condition or injury resulting from treatment for a disease.



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