CONDITION

Sacrococcygeal chordoma

Sacrococcygeal chordoma is a rare cancer that grows in the bones at the base of the spine.

WHAT IT IS
Rare malignant bone tumor of the sacrum and tailbone

BODY SYSTEM
Bones, nerves, bowel, and bladder

MAIN SYMPTOMS
Sacral pain, leg symptoms, numbness, bowel or bladder changes

WHAT CAUSES IT
Notochord remnants; cancerous change usually unexplained

HOW IT IS DIAGNOSED
MRI, CT, and planned needle biopsy

MAIN TREATMENT
Specialist surgery and high-dose precision radiation

CAN IT BE CURED?
Sometimes - complete local treatment may cure

IS IT AN EMERGENCY?
Sometimes - sudden bladder, bowel, or leg nerve symptoms

LONG-TERM OUTLOOK
Slow-growing cancer with substantial long-term recurrence risk

Overview

Sacrococcygeal chordoma is a rare type of bone cancer. It develops in the sacrum, the large triangular bone at the bottom of the spine, or near the coccyx, also called the tailbone.

A chordoma begins from remnants of the notochord. The notochord is a structure that helps form the spine before birth. Small groups of its cells can remain in the spine into adulthood. In rare cases, these cells become cancerous.

Chordomas often grow slowly. However, they can grow into nearby bone, muscles, nerves, or pelvic organs. A tumor in the sacrum may affect the nerves that control the legs, bladder, bowel, and sexual function. This is why specialist assessment and long-term follow-up matter, even when the tumor has been removed.

Sacrococcygeal chordoma is different from chordoma at the base of the skull or in the mobile

spine. The overall principles of care are similar, but surgery and possible complications depend greatly on the tumor's location.

Symptoms

Symptoms often develop gradually. Because lower back and tailbone pain are common for many other reasons, diagnosis may take time.

Pain is the most frequent symptom. It may feel deep in the lower back, sacrum, buttocks, or tailbone. Sitting may make it worse. Some people have pain that travels down a leg because the tumor is pressing on a nerve.

Other possible symptoms include:

These symptoms do not necessarily mean that you have a chordoma. More common conditions can cause similar problems. Persistent, unexplained symptoms still deserve medical assessment.

- Numbness or tingling in the buttocks, groin, or legs
- Leg weakness or difficulty walking
- Constipation, rectal pressure, or difficulty passing stool
- Difficulty starting urination or fully emptying the bladder
- Loss of bladder or bowel control
- Changes in sexual function
- A lump near the lower spine, although many tumors cannot be felt

Causes and risk factors

Sacrococcygeal chordoma comes from notochord cells left over from early development. It is usually not possible to explain why these cells later form a tumor.

Chordoma is not known to result from an injury, sitting for long periods, diet, exercise habits, or something you did. There are no established lifestyle causes.

Most chordomas are not inherited. Very rarely, chordoma occurs in more than one member of a family. Some familial cases involve an extra copy of the TBXT gene, also called the brachyury gene. This gene helps control notochord development.

A genetic assessment may be considered when several close relatives have had chordoma or when another feature suggests an inherited form. Routine genetic testing is not needed for

everyone with this condition.

Chordoma can occur at different ages but is more often diagnosed in adults. Sacral chordoma is more common in men than in women.

Diagnosis

Diagnosis usually begins with imaging. Magnetic resonance imaging, or MRI, shows the tumor and its relationship to nerves, muscles, and pelvic organs. Computed tomography, or CT, gives more detail about damage to the bone.

Additional scans may check whether the tumor has spread. These may include CT imaging of the chest, abdomen, and pelvis. The exact scans used depend on the clinical situation.

A biopsy is needed to confirm the diagnosis. During a biopsy, a specialist removes a small sample of tissue so that a pathologist can examine the cells under a microscope.

Biopsy planning is especially important with a suspected chordoma. The needle path may need to be removed during later surgery. Whenever possible, the biopsy should therefore be arranged by the specialist bone tumor team that would manage the operation. An unplanned operation or biopsy can make later treatment more difficult.

The pathology report distinguishes chordoma from other tumors that can occur in the sacrum. Tumor tissue may also undergo molecular testing, particularly if the chordoma cannot be removed, has returned, or has spread.

There is no blood test that can confirm sacrococcygeal chordoma.

Treatment and management

Treatment should be planned by a multidisciplinary sarcoma team. This usually includes surgeons, radiation oncologists, medical oncologists, radiologists, pathologists, rehabilitation professionals, and specialist nurses.

For a localized sacral chordoma, surgery is often the main treatment when the tumor can be removed safely. The aim is to take out the tumor in one piece with a border of tissue around it. This is called an en bloc resection with clear margins .

The operation may involve removing part of the sacrum. Which nerves can be preserved depends on the tumor's height and spread. Before surgery, the team should explain possible effects on walking, bladder and bowel control, sexual function, wound healing, and daily independence.

Radiation therapy uses focused energy to damage cancer cells. Chordoma is less sensitive to standard doses of radiation than many other cancers, so treatment often requires a high dose delivered with great precision. Proton therapy, carbon-ion therapy, or advanced photon techniques may be considered. Access to these methods differs between treatment centers.

Radiation may be used before or after surgery, particularly when the tumor is close to the surgical edge or cannot be removed with clear margins. It may also be the main local treatment when surgery is not possible or would cause unacceptable harm.

If chordoma returns or spreads, treatment may include further surgery, radiation, a targeted medicine, or a clinical trial. Targeted medicines act on specific signals used by tumor cells. No single drug works for everyone, and traditional chemotherapy generally has limited activity against chordoma.

Supportive care is part of treatment at every stage. It can include pain management, physiotherapy, mobility aids, bowel or bladder support, wound care, help with sexual health, and emotional support.

Complications

A sacrococcygeal chordoma can damage nearby structures as it grows. Pressure on sacral nerves may cause chronic pain, weakness, numbness, constipation, or problems controlling the bladder and bowel.

Treatment can also have lasting effects. Sacral surgery may change walking, sitting comfort, pelvic stability, bladder or bowel function, and sexual function. Large operations can involve a long recovery and a risk of wound complications. Radiation can affect nearby skin, bone, nerves, and pelvic organs.

Chordoma can return in or near the original area, including years after treatment. Local recurrence may be difficult to treat because of scar tissue and the tumor's closeness to nerves and organs.

Some chordomas spread to distant parts of the body. Possible sites include the lungs, other bones, liver, and lymph nodes. The risk is generally greater when the tumor has returned locally or is more biologically aggressive.

Prevention

There is no known way to prevent sacrococcygeal chordoma. It develops from embryonic notochord remnants and is not linked to a preventable habit or exposure.

There is also no routine screening program for people without symptoms. Relatives of someone with chordoma do not usually need screening because most cases are not inherited.

If chordoma has affected several close family members, a genetics service can review the family history and discuss whether testing or research-based surveillance is appropriate.

Living with it and what to expect

Chordoma often grows over years rather than weeks, but slow growth does not make it harmless. Its location can allow it to cause significant problems before it spreads elsewhere.

Treatment and recovery can take months. After major sacral surgery, rehabilitation may focus on walking, strength, sitting, bladder and bowel routines, and adjusting to changes in sensation. Some people return to many of their usual activities, while others need long-term practical support or mobility equipment.

Regular imaging is needed after treatment because chordoma can return late. Follow-up commonly continues for many years and may be lifelong. The type and timing of scans are based on the original tumor, treatment, surgical margins, symptoms, and previous scan results.

The outlook is different for each person. Important factors include whether the tumor is localized, whether it can be completely treated, its position around the sacral nerves, and whether it has returned or spread. A localized tumor removed with clear margins offers the best chance of long-term control, but recurrence remains possible.

Living with uncertainty can be difficult. A specialist nurse, rehabilitation team, counselor, or chordoma support organization may help with practical changes, relationships, employment, and worries about future scans.

When to get medical help

Arrange a routine medical appointment if you have persistent or worsening pain around the sacrum or tailbone, especially when it occurs with leg symptoms, a lump, or unexplained changes in bowel or bladder function. These symptoms commonly have causes other than chordoma, but they should be assessed.

If you have been treated for chordoma, contact your specialist team about new pain, numbness, weakness, bladder or bowel changes, an unexplained lump, or a clear decline in mobility. Do not wait for the next scheduled scan to report a new symptom.

Seek urgent medical assessment for:

These can be signs of serious pressure on the lower spinal nerves. Go to an emergency department or call local emergency services if symptoms are sudden, severe, or progressing quickly.

After surgery, promptly report fever, increasing wound redness, drainage, wound opening, or rapidly worsening pain to the surgical team.

- New numbness around the genitals, inner thighs, or buttocks
- New difficulty starting urination or an inability to pass urine
- New loss of bladder or bowel control
- Rapidly worsening leg weakness or difficulty walking

Common questions

Is sacrococcygeal chordoma cancer?

Yes. It is a malignant bone tumor, meaning it can invade nearby tissue and may spread. It often grows more slowly than many other cancers, but it still requires specialist cancer care.

Can sacrococcygeal chordoma be cured?

Sometimes. Cure is most possible when a localized tumor can be completely treated with surgery, often together with carefully planned radiation. Even after successful treatment, long-term scans are needed because chordoma can return years later.

How fast does a sacral chordoma grow?

Many chordomas grow slowly over months or years. Growth rate is not the same for every tumor, and symptoms can change faster if the tumor begins pressing on a nerve or pelvic organ.

Will I need surgery?

Surgery is often recommended for a removable, localized sacral chordoma, but it is not suitable in every case. Tumor size, location, previous treatment, likely nerve damage, and your general health all affect the plan. Precision radiation may be used with surgery or as the main local treatment.

Can chordoma come back after treatment?

Yes. Chordoma has a substantial risk of local recurrence, including many years after treatment. This is why follow-up continues long term. A recurrence may be treated with another operation, radiation, medicines, or a clinical trial, depending on its location and extent.

Is chordoma inherited?

Usually not. Most people with chordoma have no family history of it. Rare inherited cases have been linked to changes involving the TBXT gene. Genetics advice may be useful when more than one close relative has had chordoma.

Sources

- National Cancer Institute. Chordoma: Diagnosis and Treatment
- Chordoma Foundation. Diagnosis and treatment guidelines
- Strauss SJ, Frezza AM, Abecassis N, et al. "Bone sarcomas: ESMO-EURACAN-GENTURIS Clinical Practice Guideline for diagnosis, treatment and follow-up." Annals of Oncology . 2021;32(12):1520-1536.
- Stacchiotti S, Sommer J, Chordoma Global Consensus Group. "Building a global consensus approach to chordoma: a position paper from the medical and patient community." The Lancet Oncology . 2015;16(2):e71-e83.
- Stacchiotti S, Gronchi A, Fossati P, et al. "Best practices for the management of local-regional recurrent chordoma: a position paper by the Chordoma Global Consensus Group." Annals of Oncology . 2017;28(6):1230-1242.