CONDITION

Clival chordoma

Learn about clival chordoma, a rare skull-base tumor, including its symptoms, diagnosis, treatment, possible complications, and long-term follow-up.

WHAT IT IS
Rare malignant skull-base tumor

BODY SYSTEM
Bones and nervous system

MAIN SYMPTOMS
Double vision, persistent headache, reduced vision, facial numbness

WHAT CAUSES IT
Usually unknown

HOW IT IS DIAGNOSED
Biopsy with tissue examination

MAIN TREATMENT
Surgery and radiation therapy

CAN IT BE CURED?
Sometimes, but recurrence remains possible

IS IT AN EMERGENCY?
Sometimes, with sudden vision loss

LONG-TERM OUTLOOK
Recurrence can occur years after treatment

Overview

A clival chordoma is a rare tumor that develops in the clivus , a bone near the center of the skull base. The clivus lies behind the nose and eyes, in front of the brainstem, and close to important nerves and blood vessels.

Chordomas arise from cells left over from the notochord , a structure involved in spinal development before birth. These cells usually disappear as the body develops, but remaining cells can later form a tumor.

Most chordomas grow slowly. However, they are considered malignant, or cancerous, because they can invade nearby bone and soft tissue, return after treatment, and sometimes spread to other parts of the body. At the skull base, growth may affect nerves responsible for eye movement, vision, facial sensation, swallowing, speech, and other functions.

Treatment is complex because the tumor may be close to the brainstem, carotid arteries, pituitary gland, and cranial nerves. Care at a center experienced in chordoma and skull-base tumors can provide coordinated input from surgeons, radiation oncologists, pathologists, radiologists, and other specialists.

Symptoms

Symptoms depend on the tumor's size, direction of growth, and effects on nearby structures. They often develop gradually and may resemble more common conditions.

Double vision is a frequent symptom. It may occur when the tumor affects a cranial nerve that moves the eye, particularly the sixth cranial nerve.

Other possible symptoms include:

These symptoms do not necessarily mean that someone has a clival chordoma. Eye, sinus, nerve, and pituitary disorders can cause similar problems. New or worsening symptoms should be discussed with a healthcare professional.

Seek emergency care for sudden vision loss, weakness on one side of the body, severe confusion, a seizure, fainting, or serious difficulty breathing or swallowing. After skull-base surgery, clear watery drainage from the nose may indicate leakage of fluid surrounding the brain. Contact the surgical team urgently, especially if drainage occurs with fever, severe headache, or neck stiffness.

- Persistent or worsening headache, often behind the eyes or deep in the head
- Blurred or reduced vision, abnormal eye movements, or a drooping eyelid
- Facial numbness, tingling, or pain
- Hearing changes, dizziness, or balance problems
- Nasal blockage or, less commonly, repeated nosebleeds
- Trouble swallowing, speech changes, or hoarseness
- Weakness of the tongue or shoulder
- Hormone-related symptoms if the pituitary gland is affected

Causes and risk factors

A clival chordoma develops when notochord-related cells acquire changes that allow abnormal growth. The cause in an individual is usually unknown.

Most chordomas are not inherited. They are not known to result from diet, stress, physical activity, or ordinary daily exposures, and there is no established screening test for people without symptoms or a recognized genetic risk.

Changes involving the TBXT gene, which directs production of the protein brachyury, are important in chordoma biology. Chordoma cells commonly produce brachyury, so pathologists may test for it when examining tumor tissue.

Very rarely, chordoma affects multiple members of a family. Some families have an inherited extra copy of the TBXT gene. People with a strong family history may be referred for genetic counseling, but genetic testing is not routinely needed for everyone with chordoma.

Chordoma can occur at any age, although it is more common in adults. Some benign notochord-related findings can resemble chordoma on imaging, making expert review important.

Diagnosis

Diagnosis usually begins with a neurological examination and imaging. The clinician may assess eye movement, vision, facial sensation, hearing, swallowing, strength, balance, and cranial nerve function.

Magnetic resonance imaging , or MRI, provides detailed images of the tumor and surrounding soft tissues. Contrast material may be given through a vein to show the tumor's boundaries and its relationship to the brainstem, nerves, blood vessels, and pituitary region.

A computed tomography, or CT, scan shows bone more clearly and can identify destruction or remodeling of the clivus. Blood vessel imaging may be needed if the tumor is close to the carotid arteries.

Other conditions in this area can resemble chordoma, including chondrosarcoma, benign notochordal lesions, pituitary tumors, plasmacytoma, and cancer that has spread from another organ.

A tissue sample is generally required to confirm the diagnosis. It may be obtained during a biopsy or surgery. The procedure should be planned by a skull-base team because the biopsy route and previous procedures can affect later treatment.

A pathologist examines the tissue and may test for brachyury. Additional molecular testing may be considered for advanced disease or when evaluating clinical trials. After diagnosis, imaging may be used to check the lungs, bones, liver, lymph nodes, or other areas, although many patients have disease limited to the skull base.

Treatment and management

Treatment depends on the tumor's location, size, symptoms, previous therapy, the patient's health, and its relationship to critical structures. Surgery and radiation therapy are the main treatments.

Surgery

The goal is to remove as much tumor as possible without causing unacceptable harm. Complete removal with a margin of healthy tissue is often difficult because the clivus is surrounded by vital nerves, arteries, and the brainstem.

Many clival chordomas can be reached through the nose with an endoscopic endonasal procedure , using a camera and narrow instruments. Other tumors require an open skull-base approach, a combination of approaches, or occasionally more than one operation.

Removing every visible part is not always safe. Leaving tissue attached to a carotid artery, cranial nerve, or the brainstem may preserve function and prevent serious complications. The surgical team should explain the expected balance between tumor control and neurological risk.

Radiation therapy

Radiation therapy is commonly considered after surgery, even when no obvious tumor remains on imaging. It may also be used when surgery is not possible or would carry excessive risk.

Chordoma often requires a high radiation dose for durable control. Treatment must be carefully shaped to protect nearby structures. Proton therapy may reduce exposure beyond the target in some patients. Other options include specialized photon radiation and, at certain centers, carbon-ion therapy.

No single technology is best for everyone. The choice depends on the tumor's anatomy, previous radiation, available expertise, and treatment plan. A radiation oncologist experienced in skull-base chordoma should compare the options.

Radiation is usually delivered over multiple sessions. Shorter, highly focused treatment may be suitable for selected cases, such as a small recurrence, but it is not interchangeable with a full initial course.

Recurrent or advanced disease

Chordoma may return in the original area years after treatment. Options can include repeat surgery, carefully selected re-irradiation, focused radiation, or systemic therapy.

No standard drug treatment reliably cures chordoma, and traditional chemotherapy usually has limited activity. Targeted medicines, immune-based treatments, or clinical trials may be

considered for unresectable, recurrent, or metastatic disease. Molecular findings can sometimes help guide these choices.

Supportive care may include pain management, rehabilitation, nutrition support, speech or swallowing therapy, vision services, hormone replacement, and emotional support.

Complications

Complications may result from the tumor, surgery, radiation, or recurrence.

Tumor growth can cause double vision, facial numbness, hearing loss, swallowing problems, or weakness of the tongue or shoulder. Effects on the pituitary gland may disrupt hormones involved in thyroid function, stress response, reproduction, growth, and water balance.

Possible surgical complications include bleeding, infection, stroke, nerve injury, and leakage of cerebrospinal fluid , the fluid surrounding the brain and spinal cord. Skull-base reconstruction techniques help reduce leakage risk after endonasal surgery.

Radiation may cause fatigue, skin or nasal irritation, hair loss in the treated area, or temporary worsening of symptoms. Delayed effects can include hormone deficiencies, hearing or vision problems, tissue injury, cognitive changes, or damage to nearby nerves and blood vessels. Risk depends on the area treated, dose, technique, and previous therapy.

Local recurrence remains possible after extensive treatment. Chordoma can also spread to distant sites, although local skull-base growth is often the main concern. Recurrence may develop after a long symptom-free period.

Follow up

Follow-up generally includes clinical visits and repeat MRI of the skull base. CT may be added when bone detail is needed. Chest or other body imaging may be recommended based on the tumor's features, previous findings, and treatment history.

There is no single schedule for everyone. Timing depends on the extent of surgery, pathology results, radiation, residual disease, and time since treatment. Because recurrence can occur years later, monitoring usually continues long term.

Follow-up may also include eye examinations, hearing tests, pituitary hormone blood tests, and assessments of swallowing, speech, balance, or nerve function.

Contact the care team between appointments about persistent new headaches, double vision, facial numbness, hearing changes, swallowing problems, unexplained changes in weight or

energy, increased thirst and urination, or new pain elsewhere.

Sudden vision loss, stroke-like symptoms, seizures, severe breathing or swallowing difficulty, or confusion require immediate medical care.

Sources

- National Cancer Institute. Chordoma . About Rare Brain and Spine Tumors. National Cancer Institute, Center for Cancer Research.
- National Cancer Institute. Childhood Chordoma Treatment (PDQ®): Health Professional Version . PDQ Cancer Information Summaries. National Cancer Institute.
- 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.
- 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.
- WHO Classification of Tumours Editorial Board. Central Nervous System Tumours . 5th ed. International Agency for Research on Cancer; 2021.