CONDITION

Skull base chordoma

A skull base chordoma is a rare cancerous bone tumour that grows near the brain and important nerves.

WHAT IT IS
Rare cancerous bone tumour at the skull base

BODY SYSTEM
Bones and nervous system

MAIN SYMPTOMS
Headache, double vision, facial numbness, swallowing problems

WHAT CAUSES IT
Usually acquired cell changes; rarely inherited T-gene duplication

HOW IT IS DIAGNOSED
MRI and CT imaging, confirmed by tumour biopsy

MAIN TREATMENT
Surgery and high-dose focused radiation

CAN IT BE CURED?
Sometimes - treatment can provide lasting tumour control

IS IT AN EMERGENCY?
Sometimes - sudden vision loss or new one-sided weakness

LONG-TERM OUTLOOK
Often slow-growing, but recurrence can occur years later

Overview

A skull base chordoma is a rare cancerous tumour that begins in bone at the bottom of the skull. Most grow in a central skull base bone called the clivus, which sits behind the nose and near the brainstem.

Chordomas develop from small groups of cells left over from the notochord. The notochord is a structure that helps form the spine before birth and normally disappears as the body develops.

These tumours often grow slowly. However, they can press on or grow around important structures, including the nerves controlling eye movement, vision, facial sensation, hearing and swallowing. They may also be close to major blood vessels, the pituitary gland and the brainstem.

A skull base chordoma is considered malignant, meaning cancerous. It tends to cause problems by growing locally rather than by spreading quickly through the body. Treatment can be challenging because removing the whole tumour without harming nearby structures is not always possible.

Symptoms

Symptoms often develop gradually over months or longer. They depend on the tumour's size and which nerves or other structures it affects.

Possible symptoms include:

Pressure near the pituitary gland can sometimes affect hormone production. This may cause tiredness, changes in periods or sexual function, increased thirst, or other hormone-related symptoms.

These symptoms have many more common causes. Having one does not mean that you have a chordoma. New symptoms that persist or worsen should still be assessed.

- Ongoing or worsening headaches
- Double vision or difficulty moving one eye
- Blurred vision or loss of part of the visual field
- Drooping of an eyelid
- Facial pain, tingling or numbness
- Hearing changes or ringing in an ear
- Problems with balance or coordination
- A hoarse voice or difficulty speaking clearly
- Trouble swallowing
- Weakness of the tongue or shoulder
- Nasal blockage or, less commonly, nosebleeds

Causes and risk factors

A skull base chordoma forms when notochord cells develop changes that allow them to grow into a tumour. In most people, it is not clear why these changes happen.

A gene called TBXT , previously called the T gene , helps make a protein known as brachyury.

This protein is important during early development and is usually present in chordoma cells. Changes involving this gene appear to have a role in chordoma growth.

Most chordomas are not inherited. Very rarely, chordoma occurs in several members of a family because of an inherited duplication involving the TBXT gene. Genetic counselling may be appropriate when more than one close relative has had chordoma or when a specialist suspects a familial form.

There are no established lifestyle causes. Diet, stress, minor head injuries and ordinary infections are not known to cause skull base chordoma.

Diagnosis

Diagnosis usually starts with a medical history and a neurological examination. This includes checking eye movements, facial sensation, hearing, swallowing, strength and coordination.

Imaging helps show where the tumour is and how it affects nearby structures:

A biopsy is normally needed to confirm the diagnosis. During a biopsy, a small sample of the tumour is removed and examined by a pathologist. Special laboratory tests can identify brachyury and other features that help distinguish chordoma from tumours that may look similar on a scan, such as chondrosarcoma.

Because the biopsy route can affect later surgery and radiation planning, it is usually planned by the specialist skull base team that will manage the tumour.

Additional scans may be used to check for tumours elsewhere and to provide a baseline for future comparison. Hearing, vision, swallowing or pituitary hormone tests may also be needed.

- MRI uses magnets to create detailed images of the tumour, brain, nerves and other soft tissues.
- CT uses X-rays to show changes in the skull base bone and may help with surgical planning.

Treatment and management

Treatment is usually planned by a multidisciplinary team experienced in skull base tumours. The team may include skull base surgeons, neurosurgeons, ear, nose and throat surgeons, radiation oncologists, medical oncologists, radiologists, pathologists and rehabilitation specialists.

Surgery is often the first treatment when the tumour can be reached safely. The aim is to

remove as much tumour as possible while protecting the brainstem, nerves and major blood vessels.

Some chordomas can be approached through the nose using an endoscope, which is a narrow instrument with a camera. Others require an open operation or a combination of approaches. Complete removal is not always safe or possible, especially if the tumour surrounds nerves or blood vessels.

Focused radiation is commonly used after surgery to treat remaining tumour cells and lower the risk of local regrowth. It may also be the main treatment when surgery is not possible. Chordomas often require a high radiation dose, so precise techniques are important for limiting exposure to nearby healthy tissue.

Radiation options may include proton beam therapy, carbon ion therapy where available, or advanced photon treatments such as intensity-modulated or stereotactic radiation. The most suitable method is based on the tumour's location, previous treatment and access to specialist facilities. Proton therapy is useful in some cases because it can reduce the radiation dose beyond the target, but it is not automatically the best option for every tumour.

If a chordoma returns, management may involve another operation, further focused radiation or both. Reirradiation requires careful assessment because healthy tissues may already have received a substantial radiation dose.

Standard chemotherapy usually has limited benefit for chordoma. Targeted medicines may slow selected advanced or inoperable tumours, but they are not generally curative. Clinical trials may offer access to treatments under study.

Supportive care can include pain management, swallowing or speech therapy, vision support, physiotherapy, psychological support and treatment for pituitary hormone problems.

Complications

The tumour itself can damage nearby cranial nerves, which are the nerves connecting the brain to the eyes, face, ears, tongue and throat. This may cause lasting double vision, facial numbness, hearing loss or swallowing difficulty.

Pressure on the brainstem or blockage of normal fluid circulation around the brain can cause more serious neurological problems. These are less common but require prompt assessment.

Possible complications of surgery include bleeding, infection, nerve injury and leakage of cerebrospinal fluid, the clear fluid surrounding the brain. Risks differ according to the tumour's location and the surgical approach.

Radiation can cause tiredness and temporary irritation of nearby tissues. Delayed effects can include hormone problems, hearing or vision changes, damage to bone or soft tissue, and injury to nerves or blood vessels. Treatment teams plan radiation carefully to reduce these risks.

Chordoma can return at or near the original site, sometimes years after treatment. Spread to the lungs, bones, liver or other areas is possible, particularly with recurrent or advanced chordoma, but local recurrence is usually the main concern.

Prevention

There is no known way to prevent most skull base chordomas. They are not linked to a preventable behaviour or exposure, and routine screening is not recommended for the general population.

People from the very small number of families with inherited chordoma risk may be offered genetic counselling. A genetics specialist can explain testing and whether monitoring may be appropriate for relatives.

Living with it and what to expect

Recovery after skull base treatment may take weeks or months. Fatigue, headaches, nasal symptoms or changes in appetite can occur during early recovery. Rehabilitation may be needed if treatment has affected vision, balance, speech or swallowing.

Some nerve problems improve as swelling settles, while others may be long-lasting. Prism glasses, eye treatments, hearing devices, swallowing therapy and other forms of rehabilitation can help people adapt.

Follow-up normally includes regular MRI scans and neurological examinations. The timing is based on the original tumour, the amount removed, radiation treatment and previous scan findings. Long-term surveillance is important because chordoma can return after a long period of stability.

Many people resume work, education, family life and usual activities, although adjustments may be needed. The long-term course is not the same for everyone. Smaller tumours that can be treated effectively may remain controlled for many years. Tumours that cannot be fully treated, return repeatedly or spread elsewhere are harder to control.

Emotional effects may continue after physical recovery. Anxiety before scans is common, particularly when monitoring continues for years. A specialist nurse, counsellor, support group or mental health professional can help if worry is interfering with sleep or daily life.

When to get medical help

Arrange a routine medical assessment if you develop persistent headaches, double vision, facial numbness, swallowing difficulty or other unexplained neurological symptoms. These symptoms are usually caused by something other than chordoma, but ongoing or worsening changes should be checked.

If you have been treated for chordoma, contact your specialist team promptly about new symptoms, a steady worsening of existing symptoms, clear watery drainage from the nose, or possible treatment side effects. Do not wait for the next routine scan.

Seek urgent or emergency help for:

- Sudden loss of vision
- New weakness or numbness affecting one side of the body
- A first seizure or a seizure lasting longer than usual
- Severe headache with repeated vomiting, confusion or unusual drowsiness
- Sudden difficulty speaking, breathing or swallowing saliva
- Fever, neck stiffness or marked illness after skull base surgery
- Clear fluid from the nose after surgery, especially with headache or fever

Common questions

Is a skull base chordoma cancer?

Yes. A chordoma is a malignant bone tumour. It often grows more slowly than many other cancers, but it can invade nearby tissues, return after treatment and occasionally spread to distant organs.

Can a skull base chordoma be completely removed?

Sometimes. Complete removal may be possible when the tumour is well placed and separate from major nerves and blood vessels. In other cases, leaving a small amount behind is safer than causing serious neurological injury. Focused radiation can then treat the remaining tumour.

Does chordoma spread to other parts of the body?

It can, but local growth and recurrence are usually the main concerns. Distant spread is more likely in advanced or repeatedly recurrent chordoma. Follow-up may include imaging beyond the skull base when clinically appropriate.

Is proton therapy always needed?

No. Proton therapy can deliver a high dose while limiting radiation beyond the tumour, which is useful near sensitive structures. Advanced photon radiation and, in some centres, carbon ion therapy may also be considered. Previous treatment, tumour shape and specialist expertise all affect the choice.

Is skull base chordoma hereditary?

Usually not. Most people have no family history. Rare families carry an inherited TBXT gene duplication that increases chordoma risk. Genetic assessment may be considered when several relatives have had chordoma.

How long can someone live with a skull base chordoma?

There is no single answer. Outlook is shaped by whether the tumour can be controlled locally, its size and position, response to treatment, recurrence and any spread elsewhere. Because late recurrence can happen, long-term follow-up matters even after successful treatment.

Sources

- National Cancer Institute. Chordoma Treatment (PDQ®)-Patient Version
- Stacchiotti S, Sommer J, Chordoma Global Consensus Group. Chordoma: a position paper from the medical and patient community. The Lancet Oncology . 2015;16(2):e71-e83 71190-8)
- 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
- National Organization for Rare Disorders. Chordoma