

Benign Notochordal Cell Tumour (BNCT)

A patient guide

In brief: A benign notochordal cell tumour, often shortened to BNCT, is a small, harmless growth made up of the same primitive cells that, in the developing embryo, formed a rod-like structure called the notochord. The notochord is the forerunner of the spine, and although it disappears as the backbone forms, tiny clusters of its cells can remain trapped inside the bones of the spine and the base of the skull for life. A benign notochordal cell tumour is one of these clusters when it grows just large enough to be seen on a scan. It sits entirely within the bone, does not destroy the bone around it, produces no lump in the surrounding soft tissues, and almost always causes no symptoms whatsoever. The overwhelming majority are discovered by accident, as an incidental finding when a scan is performed for some entirely unrelated reason. The single most important idea in this whole subject is that a benign notochordal cell tumour is benign and inert, and that its main significance lies not in what it does, but in the fact that it must be told apart from its rare malignant relative, a tumour called chordoma. When the appearances on a scan are typical, the lesion is recognised, left undisturbed, and watched rather than treated.

What a benign notochordal cell tumour is, and where it comes from

To understand a benign notochordal cell tumour it helps to go back to the very earliest stages of development. Long before the bones of the spine appear, the embryo lays down a flexible rod that runs the length of the future backbone. This rod is the notochord, and it acts as a template around which the vertebrae and the discs between them are built. Once that scaffolding has done its job, the notochord is largely reabsorbed, leaving only the soft, jelly-like centre of each intervertebral disc, the nucleus pulposus, as its lasting remnant.

In a proportion of people, small islands of the original notochordal cells are left behind not in the discs but inside the bone itself, within the vertebral bodies and the bone at the base of the skull. These rests of cells are entirely normal in the sense that they are a common quirk of human development. When such a rest grows large enough to form a visible mass within the bone, it is termed a benign notochordal cell tumour. The same lesion has been described in the older literature under the name giant vertebral notochordal rest, and the two terms refer to essentially the same thing.

The word tumour here simply means a growth or swelling, and it should not be read as implying anything sinister. A benign notochordal cell tumour is, as its name states, benign. It does not spread, it does not invade, and it behaves as a quiet, static collection of cells sitting within the bone.

How common it is, and where it occurs

Benign notochordal cell tumours are far more common than most people would imagine, precisely because they so rarely announce themselves. Their true frequency has been revealed by autopsy studies, in which the spine and skull base of people who died of unrelated causes have been examined microscopically. Such studies have repeatedly found small notochordal lesions in a substantial minority of the population, with figures of up to around one in five reported in some cadaveric series. One frequently cited study found intraosseous notochordal lesions in a notable share of a hundred random autopsies, with the individual lesions ranging from a millimetre or two up to a little over a centimetre.

Because most of these lesions are tiny, only the larger ones are visible on imaging, and so the proportion picked up on scans during life is much lower than the autopsy figure, reported in one study at well under one per cent of those imaged. This gap between how often the lesions exist and how often they are seen is a recurring theme, and it means that benign notochordal cell tumours are almost certainly under-recognised rather than rare.

The lesions follow the distribution of the original notochord, and so they cluster in particular places. The bone at the base of the skull, the clivus, and the lowest part of the spine, the sacrum and coccyx, are the commonest sites, followed by the bones of the neck and the lower back. Autopsy work has described notochordal lesions in around a tenth of clivus specimens and a higher proportion of vertebral specimens. These are precisely the regions where the notochord ran during development, which is why the cells are found there.

The benign, intraosseous nature: what makes it harmless

The reassurance that surrounds a benign notochordal cell tumour rests on a small number of consistent features, all of which point to a lesion that is contained and inert.

The first is that it is intraosseous, meaning it lies entirely within the bone. It does not break out through the surface of the vertebra or the clivus, and it does not form a mass in the soft tissues around the bone. There is no associated lump pressing on nearby structures.

The second is that it does not destroy bone. A benign notochordal cell tumour does not eat away at or dissolve the bone in the way that an aggressive tumour does. On the contrary, the bone around it often appears slightly denser than normal, a feature described as mild sclerosis or osteosclerosis. The architecture of the bone is preserved rather than wrecked.

The third is that it has well-defined, well-corticated margins, meaning its edge is tidy and clearly bounded rather than ragged and infiltrating. It sits within the bone like a discrete, contained patch.

The fourth, and one of the most useful, is that it does not take up contrast dye on a scan. When a contrast agent is given during magnetic resonance imaging, areas with a rich or abnormal blood supply, such as many true tumours, light up. A benign notochordal cell tumour characteristically does not enhance, reflecting its quiet, non-aggressive nature.

Taken together, these features describe a lesion that is contained within bone, leaves the bone intact, has a clean edge and shows no aggressive activity. This combination is what underlies the description of these lesions as benign.

How a benign notochordal cell tumour appears on scans

The imaging appearance of a benign notochordal cell tumour is well characterised, and recognising it is central to the whole approach to these lesions.

On magnetic resonance imaging, the workhorse of spinal imaging, a benign notochordal cell tumour typically appears as a rounded, well-defined area within the bone. It shows low signal, appearing dark, on the sequence known as T1-weighted imaging, and high signal, appearing bright, on the T2-weighted sequence. It does not enhance after contrast and shows no restriction on the diffusion sequences that can flag more aggressive lesions. Importantly, there is no breach of the bone surface and no extension into the surrounding soft tissues.

On computed tomography, the CT scan, the lesion is usually seen as an area of mild increased bone density, the slight sclerosis already described, again without any destruction of the bone or any soft-tissue mass.

Some benign notochordal cell tumours produce few or no changes at all on plain X-rays and on the CT scan, being apparent mainly on MRI. The pattern of dark-on-T1, bright-on-T2, non-enhancing, contained, sclerotic and without soft-tissue extension is the signature that allows a benign notochordal cell tumour to be recognised with confidence in the typical case.

Distinguishing it from chordoma: the heart of the matter

The reason benign notochordal cell tumours attract attention out of all proportion to the trouble they cause is their relationship to chordoma. A chordoma is a rare, slow-growing but genuinely malignant bone tumour that also arises from notochordal cells and occurs in the very same locations, the sacrum, the clivus and the spine. Because

the two share an origin and a habitat, telling them apart is the central task whenever a notochordal lesion is found.

Fortunately, in most cases the distinction can be made on imaging, and the contrasts are fairly clear. A benign notochordal cell tumour stays within the bone, leaves the bone intact or slightly denser, has a tidy margin, does not enhance with contrast and forms no soft-tissue mass. A chordoma, by contrast, tends to destroy bone, producing areas where the bone has been dissolved away, the appearance described as osteolysis; it enhances variably after contrast, reflecting its blood supply; and it characteristically extends beyond the bone to form a soft-tissue mass that may press on adjacent structures. In published comparisons, benign notochordal cell tumours have consistently shown mild sclerosis without bone destruction and no enhancement, while chordomas have shown bone destruction and enhancement.

Under the microscope, the two also differ, although the picture can be subtle. A benign notochordal cell tumour is made of bland, uniform, vacuolated cells resembling fat cells, with small regular nuclei, no dividing cells of note and, crucially, no myxoid, jelly-like background matrix between the cells and no aggressive growth. A chordoma typically shows that myxoid matrix, a lobulated growth pattern, more variation in the cells and signs of growth into surrounding tissue. Both lesions, however, share the same cellular markers, including a protein called brachyury that is specific to notochordal cells and is present in both, which is one reason a small biopsy sample alone can sometimes be misleading.

The debated link: can a chordoma arise from a benign notochordal cell tumour?

One of the most actively discussed questions in this field is whether benign notochordal cell tumours are simply harmless leftovers, or whether some of them represent the soil from which a chordoma may, very occasionally, grow. The evidence is genuinely mixed, and the matter is unresolved.

Several observations have fuelled the idea of a link. Chordomas and benign notochordal cell tumours arise from the same cells, occur in the same places and share the same molecular markers. There are reported cases in which a chordoma was found arising alongside, or apparently within, a pre-existing benign notochordal cell tumour, and cases in which a single lesion contained both benign notochordal areas and a chordoma component. Chordomas are rare before middle age, whereas benign notochordal tissue is found in the bones of people of all ages, a pattern some authors interpret as consistent with a slow progression over time. The fact that the two can be genuinely difficult to tell apart on a small biopsy has itself been taken as further evidence of a relationship.

The current understanding is that some notochordal lesions appear to sit on a spectrum, ranging from entirely benign rests through benign notochordal cell tumours to overtly malignant chordoma, and that, under certain circumstances, a benign notochordal cell tumour might in rare instances be a precursor of chordoma. At the same time, the great majority of benign notochordal cell tumours never behave in this way and remain harmless throughout life. The existence of a possible link is one of the main reasons that a newly found notochordal lesion is taken seriously and assessed carefully, and that lesions with typical benign features are kept under periodic review rather than simply forgotten.

Why typical lesions are usually not biopsied

It might seem natural to take a sample, a biopsy, of any newly discovered lesion in the bone. With benign notochordal cell tumours, however, the prevailing view in the literature is that when the imaging features are typical, biopsy is generally neither necessary nor advisable, and that careful imaging surveillance is the more appropriate first step.

There are several reasons for this stance. The imaging signature of a typical benign notochordal cell tumour is distinctive enough that the diagnosis can usually be made with confidence from the scans alone. A biopsy of such a lesion can be actively misleading, because the benign notochordal cells share their cellular markers, including brachyury, with chordoma, so a small fragment can be misinterpreted and a harmless lesion mistaken for a malignant one. There are documented cases in which an unnecessary biopsy of a typical benign notochordal cell

tumour led to a misdiagnosis of chordoma and, in turn, to unnecessary surgery. A biopsy also carries its own small risks, and in regions such as the clivus and sacrum these are not negligible.

For all these reasons, the literature describes a general principle that, where a lesion has the typical features of a benign notochordal cell tumour, well-corticated margins, no enhancement, no soft-tissue extension, no bone destruction and no progression on scans, it is reasonable to observe it with interval imaging rather than to biopsy or remove it. Biopsy is reserved for lesions that show atypical or worrying features, or where the relative risks of leaving the diagnosis uncertain outweigh the risks of the procedure. These are matters of careful judgement, often reached through discussion among several specialists rather than by any single fixed rule.

What the evidence shows about management and follow-up (impersonal)

It is important to be clear at the outset that the evidence base for managing benign notochordal cell tumours is built largely on autopsy studies, retrospective case series, individual case reports and narrative reviews. There are no large randomised trials, and the diagnostic criteria themselves are acknowledged in the literature to be partly subjective and usually settled by multidisciplinary consensus. The descriptions that follow report what the literature observes, not guidance for any individual, and decisions in practice are made on a case-by-case basis by the treating team.

****Surveillance**** is the course described for the typical, benign-appearing lesion. A widely cited set of features used to support this approach includes the absence of any related neurological problem, well-corticated bony margins, a size below roughly thirty-five millimetres, no enhancement with contrast, no soft-tissue extension, no penetration of the lining of the spinal canal, and no progression on scans. Where a lesion meets these criteria, the literature describes it as reasonable to follow it with interval imaging, typically magnetic resonance imaging, in the first instance rather than to intervene.

****Reassurance from follow-up series**** supports this conservative stance. In published series of benign-appearing notochordal lesions followed over time, the lesions have generally remained stable. A retrospective series of incidental benign-appearing notochordal lesions of the clivus, for example, reported that all the lesions remained stable over a median follow-up of close to three years, even though a minority showed minimal contrast uptake or minor bone changes that might at first seem concerning. Such findings underpin a watch, wait and rescan approach for these incidental lesions.

****Specialist and multidisciplinary input**** is a recurring theme. Because benign notochordal cell tumours sit so close to chordoma, and because the diagnosis rests on the careful interpretation of imaging features, the literature describes these lesions as being best assessed by clinicians familiar with notochordal tumours, frequently within a multidisciplinary team that may include specialist radiologists, spinal surgeons, neurosurgeons, pathologists and oncologists. This is particularly so for lesions in the clivus or sacrum, or for any lesion with atypical features.

****Intervention**** is not the path for the typical benign notochordal cell tumour. Surgery and other active treatments belong to the management of chordoma, where aggressive lesions are treated with surgical removal, often followed by specialised radiotherapy such as proton beam therapy. Distinguishing the benign lesion that should be watched from the malignant one that needs treatment is, again, the whole purpose of the careful assessment.

In summary, the evidence points towards recognition of the typical lesion on imaging, avoidance of unnecessary biopsy, surveillance with interval scans, assessment by clinicians experienced with notochordal tumours, and reservation of active treatment for the rare situation in which the picture is, or becomes, that of a chordoma, all within a framework of individualised, multidisciplinary decision-making rather than fixed rules.

Outlook

The outlook with a benign notochordal cell tumour is, for the overwhelming majority of people, entirely reassuring. By its nature the lesion is benign, contained within the bone, non-destructive and usually silent, and the

published follow-up series describe these lesions as remaining stable over years of observation. Most people who are found to have one will never experience any symptom from it, and the lesion will simply sit quietly in the bone as it has, in all likelihood, done for decades already.

The element of caution that surrounds these lesions arises almost entirely from their kinship with chordoma. Because a small number of notochordal lesions may sit on a spectrum that ends in malignancy, and because the benign and malignant forms can resemble one another, a newly discovered lesion is assessed carefully, typical lesions are watched rather than disturbed, and atypical or changing lesions are investigated more thoroughly. This is best understood not as alarm but as sensible vigilance.

Across the whole field, the consistent themes are that benign notochordal cell tumours are common, harmless and usually incidental; that their imaging signature allows them to be recognised and distinguished from chordoma in most cases; that typical lesions are watched rather than biopsied or removed; and that any individual situation is a matter for assessment by clinicians experienced in these tumours rather than something that can be read from a label alone.