

Hydromyelia

A patient guide

In brief: Hydromyelia describes a widening of the central canal, the tiny fluid-filled channel that runs down the middle of the spinal cord. Because this channel is lined by a delicate cell layer called ependyma, hydromyelia is, in its purest form, a developmental variant rather than a disease, and a small amount of it is frequently discovered by chance on a scan performed for another reason. It is closely related to, but distinct from, syringomyelia, and the two are often grouped together under the umbrella term syringohydromyelia.

What hydromyelia is (the central canal)

The spinal cord is the long bundle of nerve tissue that carries signals between the brain and the body. Running down its very centre, along its whole length, is a microscopic channel called the central canal. In the developing fetus this canal is a clear, fluid-filled tube lined by a specialised layer of cells known as ependyma, the same type of lining found in the fluid-filled spaces of the brain. The central canal is part of the cerebrospinal fluid system, the gently circulating watery cushion that surrounds and protects the brain and spinal cord.

Hydromyelia is the term used when this central canal becomes wider than expected and holds more fluid than usual. The word itself combines "hydro", meaning water or fluid, with "myelia", referring to the spinal cord, so it literally means fluid within the cord, specifically within the natural canal at its centre. The defining feature of true hydromyelia is that the widened space is lined, at least in part, by ependyma and is continuous with the central canal. In other words, it is an enlargement of a structure that already exists, rather than a new cavity carved out of the cord tissue.

An important point that shapes how hydromyelia is understood is what happens to the central canal as a person grows up. In most adults the central canal does not remain an open, fluid-filled tube. Detailed studies of human spinal cords have shown that, beyond around eighteen years of age, the canal is largely closed off or occluded along most of the cord in the majority of people. Instead of a clear lumen, it is replaced by accumulations of ependymal cells and supporting tissue. This near-complete closure of the canal in adulthood appears to be a feature distinctive to humans among mammals.

The clinical consequence of this is significant. When a clearly visible, fluid-filled central canal is seen on the scan of an adult, it often represents the persistence of a configuration that is normal in fetal life and early childhood but that usually regresses with age. Viewed this way, an isolated, small hydromyelia is reasonably understood as a developmental variant, a leftover of how the cord formed before birth, rather than evidence of an active problem. A particular focal widening of the canal near the lower tip of the cord, called the ventriculus terminalis or terminal ventricle, is normal in early childhood and is a recognised incidental finding when it persists into later life.

How it differs from syringomyelia, and the term syringohydromyelia

Hydromyelia and syringomyelia are frequently confused, and on a routine scan they can look very similar, yet there is a genuine distinction between them that is worth understanding.

Hydromyelia, as described above, is a widening of the natural central canal of the cord. The space is lined, fully or partly, by ependyma, and it is in continuity with the canal. It is often developmental and is frequently small.

Syringomyelia describes something different: a fluid-filled cavity, known as a syrinx, that lies within the substance of the spinal cord itself, beyond the confines of the central canal. Classically this cavity is not lined by ependyma but is bordered by a different, glial tissue, and it dissects into the cord tissue. Crucially, syringomyelia is usually secondary, meaning it develops as a consequence of an underlying structural cause, such as a Chiari

malformation at the base of the skull, a spinal cord tumour, scarring of the lining membranes after injury or inflammation (arachnoiditis), or a tethered spinal cord. Because of this, syringomyelia is more often associated with neurological symptoms and is more likely to enlarge over time than an isolated central-canal widening.

Several factors blur the line between the two. They appear alike on standard MRI. As a widened central canal enlarges, its ependymal lining can be disrupted, so that the appearance and behaviour begin to overlap with a true syrinx. Many cavities have mixed features that do not fall neatly into one category. For these reasons, clinicians and radiologists very commonly use the combined term syringohydromyelia to cover the whole spectrum of fluid-filled cavities and canal dilatations within the cord, without committing to a precise distinction that cannot always be made on imaging alone. Other phrases used for the same finding include "dilated central canal" and "slit-like syrinx".

Some authors have argued that, despite the overlap, hydromyelia can and should be separated from true syringomyelia, because the two groups tend to differ in their clinical picture and natural course. In this view, people with pure hydromyelia typically have no neurological deficit and, if they have any symptoms at all, these are often non-specific or musculoskeletal and unrelated to the imaging finding. By contrast, true syringomyelia is more likely to be linked to identifiable neurological signs and a progressive course driven by its underlying cause.

Why a small central-canal dilation is often a benign incidental finding

One of the most reassuring aspects of hydromyelia is that a small, isolated widening of the central canal is frequently a benign, incidental discovery. The term "incidental" means it was found by chance, on a scan performed for some unrelated reason, rather than because it was causing any problem.

This benign character flows directly from the anatomy already described. Because the central canal normally closes in adulthood, a small persistent fluid channel reflects a fetal configuration that simply did not fully regress. Studies of routine spinal MRI have found that a visible central canal is, in fact, uncommon overall, seen in roughly one and a half per cent of cervical and thoracic studies in one large series. Where it is seen, it is typically two to four millimetres across, thin and slit-like or filiform, smooth in outline, more often in the thoracic region, and, importantly, clinically silent and unrelated to the symptoms that prompted the scan. In such cases it is interpreted as a normal variant or a persistent central canal.

In children, and in young people being screened for spinal curvature, an incidental syrinx or hydromyelia is found in a meaningful minority, the great majority of whom have no symptoms from it. The natural history of this isolated form is generally favourable. There are documented cases of an asymptomatic, localised widening of the canal remaining stable over more than two decades, without the development of spinal signs or symptoms. Progression is the exception rather than the rule when no causative lesion is present.

The features that point towards a benign, incidental hydromyelia can be summarised as: a small or slit-like cavity; a smooth outline; no swelling or signal change within the surrounding cord tissue; no associated condition such as a Chiari malformation, tumour, tethering or arachnoiditis; and no neurological symptoms. Features that instead raise the possibility of a significant, true syringomyelia needing closer investigation include a larger or expanding cavity, dissection of fluid to the side of the canal, signal change within the cord, neurological signs, or an identified structural cause.

Causes and associations (Chiari, dysraphism, tethered cord)

While isolated hydromyelia is often developmental and without an identifiable cause, widening of the central canal can also occur in association with a number of underlying conditions. Recognising these associations is central to distinguishing a harmless variant from a finding that warrants attention.

The most important association is the Chiari malformation, particularly types I and II. In a Chiari malformation, the lower part of the brain, the hindbrain, sits lower than normal and herniates through the opening at the base of the skull, the foramen magnum. This obstructs the normal flow and pulsation of cerebrospinal fluid at that level and disturbs the pressure relationship between the head and the spine. Over time these disturbed dynamics can drive widening of the central canal and cavitation within the cord. In children with spina bifida and a Chiari II malformation, hydromyelia is commonly seen on imaging, although only a proportion of these children ever develop symptoms from it.

A tethered spinal cord and spinal dysraphism form another group of associations. Dysraphism is the umbrella term for conditions in which the spine and spinal cord do not form completely before birth. When the lower end of the cord is abnormally anchored or tethered in place, the resulting mechanical strain and altered fluid dynamics can contribute to canal widening or syrinx formation.

A number of theories have been proposed to explain how disturbed fluid flow translates into cord cavitation. The classical hydrodynamic or "water-hammer" theory described pressure waves of fluid being driven into the central canal. Later refinements emphasised the dissociation of pressure between the skull and the spine. More recent thinking frames the process as an accumulation of fluid filtered from blood vessels into the cord tissue when normal fluid circulation is obstructed or the cord is tethered, sometimes preceded by a reversible, swollen state of the cord known as the "presyrinx". These mechanistic ideas share a common theme: the cavity tends to arise from a disturbance of fluid circulation, which is why attention is directed towards the underlying cause.

How it tends to present (often none)

In a great many cases, hydromyelia produces no symptoms at all and is discovered purely by chance. This is especially true of the small, isolated, slit-like canal widenings that represent a persistent fetal configuration. Such findings are frequently silent, are stable over long periods, and are unrelated to whatever prompted the scan in the first place.

Where symptoms are attributed to a fluid cavity within the cord, they relate to the parts of the cord affected and tend to belong to the syringomyelia end of the spectrum rather than to pure hydromyelia. Cavities that expand within the cord can, in principle, interfere with the nerve pathways that carry sensation and movement. Classic features described in true syringomyelia include a characteristic pattern of reduced sensation to pain and temperature across the shoulders and arms while the sense of light touch is preserved, wasting and weakness of the hand and arm muscles, and, particularly in growing children, a progressive curvature of the spine. Changes in bladder or bowel function are also recognised in more advanced cases.

It is important to keep this in perspective. These progressive features are characteristic of symptomatic syringomyelia with an underlying cause, not of the small incidental canal dilatation that constitutes most isolated hydromyelia. People found to have pure hydromyelia very often have a normal neurological examination, and any aches or pains they report are frequently of musculoskeletal origin and not explained by the canal finding itself.

How it is assessed (MRI)

Magnetic resonance imaging, or MRI, is the investigation of choice for hydromyelia and for any fluid cavity within the spinal cord. MRI uses a strong magnetic field and radio waves, with no x-rays, to produce detailed images of the soft tissues of the cord. It serves two purposes that are equally important.

The first purpose is to characterise the cavity itself. Images taken in different planes allow the widened canal or cavity to be measured and described: its size and shape, whether it appears confined to the central canal or extends into the cord tissue, and whether there is any swelling or signal change in the surrounding cord that might suggest

it is active rather than quiescent.

The second purpose, which is at least as important, is to look for an underlying cause. This is why imaging is often extended beyond the immediate area. The brain and the junction between the skull and the spine are examined for a Chiari malformation or for a build-up of fluid within the brain. The cord is examined for any tumour. The fluid spaces around the cord are examined for signs of scarring or tethering. Where a tumour or an inflammatory cause is suspected, an injection of contrast dye may be used to make such lesions more conspicuous.

In addition to imaging, a careful clinical neurological examination, and in some settings specialised electrical tests of nerve pathway function, can help to separate a clinically silent hydromyelia from a symptomatic syringomyelia. A persistent terminal ventricle at the lower tip of the cord has a typical appearance, following the signal of cerebrospinal fluid and not taking up contrast, and is usually recognised as an incidental finding.

How it is distinguished from related conditions

Distinguishing hydromyelia from related conditions rests on combining the appearance of the cavity with the search for an underlying cause and the clinical picture.

The principal distinction is from true syringomyelia. A finding that is small, slit-like, smooth, confined to the central canal, accompanied by no cord signal change, and unaccompanied by any structural cause or neurological deficit sits towards the hydromyelia end of the spectrum. A finding that is larger or expansile, dissects into the cord tissue beyond the canal, is accompanied by cord signal change or neurological signs, or is associated with an identified cause such as a Chiari malformation, sits towards the syringomyelia end and prompts closer investigation. The combined term syringohydromyelia is used when this distinction cannot be confidently made.

The persistent terminal ventricle is distinguished by its characteristic location at the conus, the lower tapering end of the cord, its smooth fluid-signal appearance, and its lack of contrast enhancement. A presyrinx state, in which the cord is swollen with excess fluid but has not yet cavitated, is distinguished by signal change without a discrete cavity, and is recognised because it may be reversible if normal fluid flow is restored. Other cystic lesions of the cord, and tumour-associated cavities, are distinguished chiefly by their imaging features and by the presence of an associated mass or abnormal enhancement.

What the evidence shows about management (surveillance for isolated forms; cause-directed for symptomatic secondary syrinx)

The published evidence supports an approach that is firmly shaped by whether the finding is an isolated, asymptomatic canal dilatation or a symptomatic cavity with an underlying cause. The summary that follows describes what the literature shows and is provided for information only; it is not advice for any individual.

For isolated or idiopathic hydromyelia, that is a small canal dilatation with no identified cause and no symptoms, the literature supports observation. This commonly takes the form of monitoring, often with a follow-up MRI at an interval to confirm that the finding is stable. Long-term series document that such isolated dilatations frequently remain unchanged over many years, and routine surgery is not generally indicated for them.

For symptomatic, progressive syringomyelia with an identified cause, the weight of evidence supports treating the cause rather than the cavity. Where the cavity is associated with a Chiari malformation, surgery to decompress the craniovertebral junction and restore normal fluid flow is the relevant approach. Where the cord is tethered, untethering is the relevant intervention. Where there is arachnoid scarring, surgery to free the scarred membranes is directed at the obstruction. The unifying principle is that addressing the disturbance of cerebrospinal fluid flow allows many cavities to stabilise or shrink, and many people to improve or stabilise. Procedures that drain the cavity directly, by inserting a small tube to divert fluid, carry a higher rate of long-term failure and are generally

reserved for selected or difficult cases that have not responded to cause-directed surgery.

In children with a Chiari II malformation and spina bifida, in whom hydromyelia is commonly seen, intervention is reserved for those who develop symptoms, and the type of treatment is tailored to whether the clinical picture points to the hindbrain or to a tethered cord.

The single most important point that emerges from the evidence is that the behaviour of a canal dilatation or syrinx is governed largely by whether an underlying cause is present. An isolated dilatation tends to stay quiet; a cavity tied to a structural cause is the one more likely to enlarge and to cause symptoms.

Outlook

The outlook for hydromyelia is, for most people who have it, reassuring, and it depends very largely on which part of the spectrum a particular finding occupies.

A small, isolated, incidental widening of the central canal, the kind found by chance on a scan and unaccompanied by symptoms or by any underlying condition, has a favourable natural history. The evidence describes such findings remaining stable over many years, in some documented cases over more than two decades, without progressing or producing neurological problems. For findings of this character, the typical course is one of monitoring rather than treatment, and the finding often proves to be a quiet, lifelong companion of no practical consequence.

Where a fluid cavity is symptomatic and is driven by an underlying cause, the outlook is shaped by that cause and by how it is managed. Surgery directed at restoring normal cerebrospinal fluid flow, by decompressing a Chiari malformation, untethering a tethered cord, or freeing arachnoid scarring, allows many cavities to stabilise or shrink and many people to improve or remain stable over the longer term. The recognition of reversible precursor states, such as the presyrinx, underlines the value of identifying and addressing the cause.

The overarching message of the literature is one of careful distinction: most isolated hydromyelia is a benign developmental variant requiring nothing more than reassurance and, sometimes, interval monitoring, while the smaller group of cavities tied to a structural cause is the group that benefits from closer assessment and, where appropriate, cause-directed treatment.
