

Ehlers-Danlos Syndrome and the Spine

A patient guide

In brief: Ehlers-Danlos syndrome (EDS) is a group of inherited disorders of connective tissue, the material that gives ligaments, tendons, skin, blood vessels and the lining of the nervous system their strength and resilience. Because the spine is held together and protected almost entirely by connective tissue, EDS has a particular relationship with it, and that relationship is the subject of this page rather than the systemic features of the condition as a whole. The fault lies chiefly in collagen, the principal structural protein of these tissues; when collagen is abnormal the ligaments that should restrain the joints of the spine become lax, the discs and their surrounding structures wear earlier, and the curves of the spine are less reliably maintained. From a spinal point of view this expresses itself in several recognisable patterns: instability where the skull meets the neck and at the first two vertebrae beneath it, early and often multilevel disc degeneration, scoliosis and kyphoscoliosis, an over-representation of conditions such as Chiari malformation, tethered cord and dural ectasia, a tendency to spondylolisthesis, and, when surgery is contemplated, a set of particular challenges arising from soft tissue that holds sutures and bone anchors poorly and heals less predictably. The hypermobile and classical types are those most often seen in this spinal context. The single most important idea is that EDS is a disorder of the connective tissue that builds the spine, so that the spine inherits the fragility, laxity and slow healing of that tissue.

What Ehlers-Danlos syndrome is, and why it concerns the spine

Ehlers-Danlos syndrome is not a single disease but a family of heritable connective tissue disorders. The 2017 international classification recognises thirteen distinct types, each defined by its clinical pattern and, where known, its underlying genetic fault. What unites them is an abnormality of connective tissue, and in most types that means an abnormality of collagen, the rope-like protein that gives ligaments, tendons, joint capsules, skin and the coverings of the nervous system their tensile strength. The shared clinical signature is a triad of joint hypermobility, skin that stretches more than usual, and tissue fragility.

The spine is, in essence, a stack of bones held in alignment by connective tissue. Ligaments restrain how far each vertebra can move on the next; the intervertebral discs, themselves largely collagen, both cushion and tether adjacent vertebrae; tough capsules surround the small facet joints; and a continuous sleeve of dura mater encloses the spinal cord and nerve roots. When the connective tissue from which all of this is made is abnormal, the consequences are felt throughout the spine. This is why a condition that is, on the surface, a matter of stretchy skin and bendy joints comes to occupy the attention of a spinal surgeon at all.

For the purposes of the spine, the hypermobile type (hEDS) and the classical type (cEDS) are the forms most frequently encountered. The hypermobile type is by far the most common and is recognised on clinical grounds, since no confirmatory genetic test is yet available for it. The classical type combines marked skin involvement with hypermobility and has identifiable genetic causes. The much rarer vascular type carries its own distinct and serious considerations relating to the fragility of blood vessels, which colours any discussion of surgery near the spine. This page describes how these connective tissue abnormalities manifest in the spine and what the published literature reports about them, with emphasis on the hypermobile and classical types.

Laxity, instability and the upper cervical spine

The region where the skull meets the spine is the most specialised and the most heavily scrutinised in the context of EDS. Here the head balances on the first cervical vertebra, the atlas, which in turn pivots on the second, the axis, and the whole arrangement is restrained not by interlocking bone but by an array of strong ligaments. In EDS these ligaments share in the general laxity of the connective tissue, and the literature describes a recognised

tendency toward craniocervical instability, in which the junction between skull and spine moves more than it should, and atlantoaxial instability, in which the first and second vertebrae move excessively on one another.

The clinical picture associated with this instability, as reported in the literature, includes severe headache and neck pain, and a constellation sometimes termed the cervical medullary syndrome, in which the lower brainstem and upper spinal cord are thought to be irritated or compressed by the abnormal movement. Lower cranial nerve disturbances and signs of myelopathy have also been described. A central and openly acknowledged difficulty in this field is distinguishing the generalised hypermobility that is inherent to EDS from genuinely pathological instability, since the two can look similar and the radiological thresholds are still debated.

To address that difficulty the literature has gathered around a small number of measurements taken on imaging, often performed with the neck in different positions to reveal movement. Among the most frequently cited are the clivo-axial angle, which describes the angle between the base of the skull and the upper spine and whose normal values are generally given as greater than around one hundred and thirty-five degrees; the Grabb-Mapstone-Oakes measurement; the Harris measurement; and the degree of rotation of the atlas on the axis. Systematic reviews note that there is as yet no full consensus on which parameters or thresholds best define instability, which is one reason this remains an area of careful and individualised assessment rather than fixed rules.

Chiari malformation, basilar invagination and the complex craniocervical junction

Closely tied to instability at the top of the spine is a group of associated structural conditions that the literature reports more often in EDS than in the general population. Chiari malformation type I, in which the lower part of the cerebellum sits lower than usual and crowds the space at the bottom of the skull, has been described as a comorbid condition in EDS. Basilar invagination, where the top of the spine settles upward into the base of the skull, is also recognised in this setting.

The literature increasingly describes a convergence of these features in some people with connective tissue disorders, sometimes referred to as a complex craniocervical junction, in which Chiari-type crowding, basilar invagination and craniocervical instability coexist. The point repeatedly emphasised is that the underlying mechanical instability, rather than the Chiari descent in isolation, is often the dominant feature in this group. This distinction matters because, as the surgical literature notes, operations directed purely at the Chiari descent address only part of the picture when instability is the driving abnormality. Authors writing on the subject stress that fusion of the skull to the upper spine is rarely the appropriate response to a Chiari malformation on its own and is reserved, in the published series, for those in whom mechanical instability and deformity of the brainstem or upper cord are the primary problem.

Early and multilevel disc degeneration

Beyond the craniocervical junction, one of the most consistently reported spinal features of EDS is degeneration of the intervertebral discs that begins earlier in life and tends to involve several levels. The disc is itself a connective tissue structure, with an outer ring of collagen fibres enclosing a softer core, and when that collagen is abnormal the disc is less able to resist the repeated loads of daily life. The literature on the hypermobile and classical types describes early discopathy and degenerative spondylotic change as a well-established association.

The mechanism described is twofold. The discs themselves are intrinsically weaker, and at the same time the lax ligaments allow segments of the spine to move more than they should, so that the discs and facet joints are subjected to greater and less well-controlled motion. In the neck and upper back this combination of instability and disc wear is reported to contribute to a loss of the normal forward curve of the neck and an increasing forward stoop, which over time can place the spinal cord under strain. In the lower back the same processes are linked in the literature to chronic pain and reduced function, with studies attributing much of the back pain in hypermobility to the extra work the paraspinal muscles must perform to compensate for ligaments that no longer restrain the segments reliably. The literature is careful to frame this lower back pain as multifactorial rather than purely structural.

Scoliosis, kyphosis and spinal deformity

Connective tissue not only holds the spine together but helps it hold its shape, and so it is unsurprising that the literature reports an increased frequency of spinal deformity in EDS. Scoliosis, a sideways curvature of the spine, and kyphoscoliosis, in which a sideways curve is combined with excessive forward bending, are both described. Estimates of how common scoliosis is in hypermobile EDS vary, with some series suggesting it affects roughly a third of those with the condition, though figures differ according to how it is defined and which population is studied.

The explanation offered is mechanical and developmental. The lax connective tissue around the growing or mature spine is less able to maintain alignment, so the spine is more inclined to drift away from a straight, balanced shape. There is also a distinct kyphoscoliotic type of EDS, one of the thirteen recognised forms, in which severe, often early curvature is a defining feature rather than an occasional association. In the more common hypermobile and classical types, deformity tends to be one element within a broader spinal picture rather than the whole of it. The literature treats the presence and progression of curves as something that depends heavily on the individual, the type of EDS and the stage of life.

Dural ectasia, Tarlov cysts and the membranes of the spine

The dura mater, the tough membrane that sheathes the spinal cord and nerve roots, is itself a connective tissue structure and is therefore subject to the same fragility seen elsewhere in EDS. The literature describes dural ectasia, a ballooning or widening of this membrane, most often in the lower spine where the pressure of cerebrospinal fluid is greatest. As the weakened dura stretches, it can erode subtly into the surrounding bone over time. Symptoms attributed to dural ectasia in the literature include low back pain, headache, and disturbances of sensation, sometimes with the characteristic feature of being eased by lying down.

Related to this is the Tarlov cyst, a fluid-filled outpouching that forms at the root of a spinal nerve, attributed in EDS to weakening of the delicate connective tissue sheaths around the nerve roots. Such cysts are frequently incidental and silent, though the literature records that they can occasionally produce symptoms. The membranous fragility of EDS is also linked in the literature to a tendency toward spontaneous leaks of cerebrospinal fluid, where the dura gives way and fluid escapes, producing a characteristic low-pressure headache. These membranous features are part of what makes the spine in EDS distinctive, since they reflect fragility not of the bones or ligaments but of the coverings of the nervous system itself.

Tethered cord, spondylolisthesis and other associations

The literature describes several further spinal associations in EDS, reported with varying degrees of certainty. Tethered cord syndrome, in which the lower end of the spinal cord is held abnormally low and under tension, has been reported in association with EDS, though the authors of the larger reviews are candid that the epidemiological evidence linking the two is still developing rather than firmly established.

Spondylolisthesis, the forward slippage of one vertebra upon the one below, is another recognised association, and case reports document high-grade slips in EDS that can be accompanied by neurological symptoms. The mechanism is consistent with the rest of the picture, since the ligaments and supporting structures that would ordinarily resist such slippage share in the general laxity. Taken together with the instability, disc degeneration, deformity and membranous features already described, these associations underline the recurring theme that runs through the whole subject: the spine in EDS reflects, at every level from the skull to the sacrum, the qualities of the connective tissue from which it is built.

Particular challenges when surgery is considered

The connective tissue abnormality that underlies EDS does not stop at the spine but extends to every tissue a surgeon must handle, and the literature is consistent in reporting that spinal surgery in EDS carries particular challenges. These are described here as part of understanding the condition, not as advice for any individual.

The first challenge is the tissue itself. Skin, fascia and the deeper soft tissues are more fragile and hold sutures less reliably, so wounds are reported to heal more slowly and less predictably. Large database studies have found that people with EDS undergoing spinal surgery have markedly higher rates of wound-related complications, including a substantially raised risk of surgical site infection, a pattern attributed to the role of abnormal collagen in healing. The second challenge concerns the anchoring of metalwork to bone. Screws and other fixation rely on a firm grip within bone and on the surrounding ligaments to hold a construct steady; where the connective tissue is lax and, in some individuals, the bone quality less robust, the literature reports higher rates of hardware loosening, instrumentation failure and failure of the bone to fuse, or pseudarthrosis. Series of deformity correction in EDS document reoperations for instrumentation failure and non-union.

A third challenge, particularly emphasised for the vascular type but relevant to caution generally, is the fragility of blood vessels and the tendency to prolonged bleeding, which has been reported to complicate operations near the spine. The combined effect of these factors is that perioperative complication rates after spinal surgery in EDS are reported in some series to be considerably higher than in comparable patients without the condition, and the literature repeatedly counsels caution, careful patient selection and individualised planning, particularly where the precise type of EDS is uncertain. The literature also notes that EDS is associated with higher rates of adjacent segment disease after lumbar fusion, again attributed to the underlying connective tissue abnormality.

How the spine in EDS is assessed and understood

Bringing these threads together, the assessment of the spine in EDS as described in the literature rests on relating the clinical picture to careful imaging. The diagnosis of EDS itself is made on clinical grounds, supported where relevant by genetic testing and by clinical scoring of joint hypermobility such as the Beighton score, which gives a numerical measure of how mobile the joints are.

Where instability of the upper cervical spine is in question, the literature describes the use of imaging performed in different head positions to reveal abnormal movement, together with the specific measurements already mentioned, such as the clivo-axial angle. Magnetic resonance imaging is used to look at the spinal cord, the discs, the dura and any associated Chiari descent or cyst formation, while computed tomography shows the bony detail of the craniocervical junction. The recurring message in the systematic reviews is one of incomplete consensus and genuine difficulty in separating constitutional hypermobility from pathological instability, which is why this field is characterised by individualised, expert assessment rather than fixed numerical rules. The literature describes a condition best understood not as any single spinal problem but as a connective tissue disorder whose effects are written across the whole spine, and whose management, where it is considered at all, is approached with particular care.