

Diastematomyelia

A patient guide

In brief: Diastematomyelia, now more correctly termed split cord malformation, is a rare condition present from birth in which the spinal cord is divided lengthways into two halves over part of its length. The dividing structure is a bony spur or a fibrous band that runs in the cleft between the two halves. Although the condition is defined by this splitting, the dividing element frequently anchors the cord and so produces tethering, which is why diastematomyelia and tethered cord are so often mentioned together; it also carries a strong association with congenital scoliosis.

What diastematomyelia (split cord malformation) is

Diastematomyelia is a developmental anomaly of the spinal cord in which the cord is split longitudinally into two parts, known as hemicords, over a variable number of spinal segments. Each hemicord usually contains its own central canal and one set of nerve roots, one at the front and one at the back. In other words, each half behaves like half a cord rather than a complete duplicate cord. The split is created and held open by a median septum that lies within the cleft between the two hemicords.

The older literature used several overlapping names for cord-doubling anomalies, including diastematomyelia, diplomyelia and double cord. The neurosurgeon Dachling Pang brought these together under a single heading, split cord malformation, which is the term most spinal specialists now use. The word diastematomyelia remains in common use, particularly for the variant in which a bony spur divides the cord, and is the name many people first encounter.

The essential point that sets this condition apart from many other spinal anomalies is that it is a splitting lesion rather than simply a fixation lesion. The defining feature is the presence of two hemicords plus a median septum. This is conceptually different from a simple tethered cord, in which a single, intact cord is anchored at its lower end, and from open spinal defects such as myelomeningocele. The septum is the surgically and clinically distinctive feature, and its nature, whether bony or fibrous, determines the subtype.

Pang Type I and Type II

Under Pang's unified scheme, split cord malformation is divided into two anatomical subtypes that differ in how the two hemicords are housed and what separates them.

Type I describes two hemicords, each contained within its own separate dural tube, the dura being the tough membrane that surrounds the cord. The two tubes are separated by a rigid bony or osseo-cartilaginous spur, sometimes called a median spur or septum. This is the lesion classically called diastematomyelia. The rigid spur transfixes the cord and acts as the tethering element, and its firm, dura-sheathed nature is what distinguishes Type I both from Type II and from other spinal anomalies.

Type II describes two hemicords lying within a single, shared dural tube, separated by a non-rigid fibrous or fibrovascular band rather than a bony spur. This corresponds to the older term diplomyelia. The fibrous band can still tether the cord, but the absence of a rigid bony spur and of separate dural sleeves makes Type II anatomically distinct from Type I.

A small proportion of cases, of the order of five per cent, show features of both types at different levels in the same person. These composite or mixed forms are occasionally labelled informally, but there is no standardised name for them. Distinguishing Type I from Type II matters because it shapes the imaging approach and the surgical considerations, with Type I generally being the more technically demanding to address.

How it forms (embryology)

Split cord malformation is thought to arise from a single developmental error that occurs very early in pregnancy, at around the time the structures of the early spine and spinal cord are being laid down. Pang, together with Dias and Ahab-Barmada, proposed a unified theory of how all the cord-doubling anomalies come about, centred on an event during gastrulation, roughly the third week after conception.

In this account, an abnormal adhesion forms between two of the early tissue layers, the ectoderm and the endoderm, around the closure of a transient structure called the primitive neurenteric canal. This adhesion creates an accessory canal, and around it condenses a band of tissue described as an endomesenchymal tract. This tract passes through and bisects the developing notochord and neural plate, splitting them into two halves and so producing two hemineural plates that go on to form the two hemicords.

What happens to this tract then determines the subtype. If the tract persists and goes on to ossify, it becomes the bony spur enclosed by two separate dural tubes, giving Type I. If it remains as fibrous or meningeal tissue within a single dural tube, it becomes the fibrous band of Type II. This single mechanism also helps to explain why the condition so often sits alongside anomalies of the vertebral bones, overlying skin lesions and, occasionally, anomalies of the gut, since all of these structures are influenced by the same early developmental region.

Cutaneous markers and the scoliosis association

One of the features that makes diastematomyelia clinically recognisable is that it frequently announces itself on the surface of the back. Skin markings, known as cutaneous stigmata, overlie the lesion in a large proportion of cases, with figures across series ranging widely but commonly cited around forty to two-thirds. The single most frequent marking is a patch of excess hair over the midline of the spine, known as hypertrichosis and sometimes described as a faun tail because of its appearance. This hairy patch is reported in a high proportion of adults and a smaller proportion of children, and the faun tail is particularly linked with Type I. Other recognised markings include a midline dimple, a dermal sinus, a small fatty lump or lipoma, and a birthmark such as a capillary haemangioma or vascular naevus.

The other major association, and the one that places the condition firmly within the orthopaedic and spinal surgeon's field, is with congenital scoliosis, a sideways curvature of the spine arising from differences in the way the spinal bones formed before birth. Split cord malformation is found in a meaningful minority of congenital scoliosis cases, with figures generally quoted in the range of around five to eighteen per cent. Looking the other way, scoliosis or kyphoscoliosis affects more than half of people with split cord malformation. Vertebral anomalies such as hemivertebrae, butterfly vertebrae, failures of segmentation, a widened distance between the pedicles and spina bifida occulta tend to cluster at or close to the level of the septum. Because of this strong link, a congenital curve or a midline skin marking is one of the common reasons the underlying split cord is first looked for.

How it relates to tethered cord

The relationship between diastematomyelia and tethered cord is a frequent source of confusion, and clarifying it is central to understanding the condition. The two are conceptually separate but clinically overlapping.

A simple tethered cord involves a single, intact spinal cord that is abnormally anchored at its lower end, for example by a tight or fatty filum terminale, the fine strand at the bottom of the cord. Split cord malformation, by contrast, involves a cord that has been divided into two hemicords by a median septum. The defining abnormality is the splitting, not the anchoring.

However, the dividing septum itself very commonly causes tethering. In Type I the rigid bony spur transfixes the cord, and in Type II the fibrous band passes through it; in both cases the septum pins the cord in place. As a result, split cord malformation frequently presents in the same way as tethered cord syndrome, with progressive changes in the legs, sensation, bladder and bowel function, and pain. It is also often accompanied by other tethering

features such as a thickened filum, a low-lying conus, a lipoma, a dermoid or epidermoid, or a dermal sinus. So while the two conditions are distinct, the septum is a tethering structure, and this is why diastematomyelia and tethered cord are so closely linked in practice.

How it tends to present

The way diastematomyelia comes to attention varies considerably with age, and a proportion of people have no symptoms at all and are found incidentally during imaging carried out for another reason.

In children, the condition is often suspected because of a midline skin marking over the spine, the discovery of a congenital scoliosis, a foot deformity such as a high-arched or cavus foot, a disturbance of walking or gait, back or leg pain, or changes in bladder function. Differences between the two legs, such as a difference in length or in calf size, and a dropped foot are among the neuro-orthopaedic signs that can be seen.

In adults, presentation tends to be later and is often dominated by pain or by a slowly progressive neurological change. Symptoms may be brought on or worsened by a period of growth, by an injury, or by physical activity. Across all ages, the common thread is that the symptoms reflect either the tethering and stretching of the divided cord or the associated anomalies, and they may evolve gradually over time. The pattern of presentation is one reason whole-spine imaging is valued, since the condition can affect more than one level and is frequently accompanied by other findings.

How it is assessed (MRI/CT)

Imaging is central to identifying diastematomyelia, characterising its subtype and planning any intervention, and two modalities play complementary roles.

Magnetic resonance imaging, or MRI, of the whole spine is the primary investigation. It demonstrates the two hemicords, shows the level and extent of the cleft, locates the lower end of the cord known as the conus, and reveals associated abnormalities such as a syrinx, lipoma or dermoid. It also helps to separate Type I from Type II by showing whether the hemicords lie in one dural tube or two.

Computed tomography, or CT, best characterises the bony spur of Type I and the surrounding vertebral anomalies, and it is important for surgical planning where a bony septum is present. CT shows the rigid, ossified nature of the spur and its relationship to the surrounding bone in a way that complements the soft-tissue detail of MRI.

Whole-spine imaging is generally favoured rather than imaging a single region, because split cord malformation may occur at more than one level and is frequently accompanied by other anomalies along the length of the spine. A congenital scoliosis or a midline cutaneous marking is one of the recognised prompts for obtaining such imaging.

How it is distinguished from related conditions

Diastematomyelia sits within a family of conditions known as spinal dysraphisms, and distinguishing it from its neighbours rests on a small number of defining features.

It differs from open spinal dysraphism, such as myelomeningocele, in which neural tissue is exposed at birth, because in diastematomyelia the skin is typically intact and the hallmark is the divided cord with its median septum rather than an open defect. It differs from the other closed or occult dysraphisms, including lipomyelomeningocele, dermal sinus and tight filum terminale, by the presence of the two hemicords and the dividing septum; those other conditions involve a single cord. Importantly, however, diastematomyelia can co-exist with any of these, so finding one does not exclude the others.

The feature that most reliably marks out Type I is the rigid bony or osseo-cartilaginous spur, sheathed in dura and separating two dural tubes. This element is found in no other dysraphism, and its presence is what separates Type I both from Type II and from the wider group of spinal anomalies. The fundamental distinction throughout is

between a splitting lesion, which diastematomyelia is, and a purely fixation lesion such as a simple tethered cord, even though the two overlap because the septum so often tethers the cord.

What the evidence shows about management (septum resection/untethering, especially before scoliosis correction)

The following describes themes drawn from the surgical literature and is presented in general, impersonal terms; it is not guidance for any individual situation.

Where split cord malformation is causing symptoms, the surgical literature describes an approach directed at eliminating the tethering septum. In Type I this involves excising the bony spur together with the intervening dural sleeve, and in Type II it involves dividing the fibrous band, in each case combined with releasing any associated tethering feature such as a thickened filum, lipoma or dermoid. Type I procedures are generally reported as technically harder, with a slightly higher rate of complications than Type II. Modern series that use intra-operative neurophysiological monitoring, which tracks nerve function during the operation, report neurological improvement or stabilisation in the majority of cases, with low rates of lasting deterioration.

For people with split cord malformation who have no symptoms, the literature is divided. Some authors describe a case for early, preventive surgery, particularly in Type I, on the grounds that deterioration can occur during growth, that surgery becomes more difficult later, and that an established fixed deficit rarely recovers. Others favour careful observation, weighing the risk of operating against the risk of waiting. Natural-history data indicate a genuine risk of progressive deficit during growth in untreated cases, which is the consideration at the heart of this debate.

The question of timing in relation to scoliosis surgery has been particularly discussed. The classical teaching held that the bony septum should be resected and the cord released before any correction of the spinal deformity, because correcting and lengthening a spine whose cord is transfixated by a spur was thought to risk neurological injury. More recent series have challenged a single universal rule. Several studies report that people who are neurologically intact or stable can undergo deformity correction without first removing the septum, safely and effectively, when this is done carefully and with monitoring. A range of strategies is described, including staged and single-stage operations, the use of halo or skull-femoral traction, and, for a wide bony spur, a technique of vertebral column resection carried out through the septum. The overall picture from the literature is that whether and when to deal with the septum relative to scoliosis correction is individualised, depends heavily on neurological status and on the anatomy of the septum, and remains an area of ongoing study and debate.

Outlook

The outlook in split cord malformation depends on the subtype, on whether symptoms are present, on the level and extent of the lesion, and on the associated anomalies, so any general statement carries wide variation. A recurring theme in the literature is that established neurological deficits, once fixed, tend not to recover, whereas surgery undertaken in the context of stable or progressive symptoms is most often reported as producing improvement or, at the least, halting further decline. Series using modern monitoring describe low rates of permanent worsening following surgery.

Because the condition is associated with congenital scoliosis and a range of other spinal anomalies, the longer-term picture is frequently shaped as much by the deformity and the associated findings as by the split cord itself. Long-term follow-up is a consistent feature of the published series, reflecting the possibility of gradual change over time, particularly during periods of growth. Taken together, the evidence presents diastematomyelia as a condition in which careful characterisation of the anatomy and of any change over time underpins the way it is understood and managed.