Cervical Spine Instability

A Patient Guide

What's Cervical Spine Instability?

Cervical spine instability means one or more vertebrae can move beyond their normal range of motion. Instability can be caused by mechanical problems or ligament laxity in any part of the cervical spine.^[1]

The cervical spine is a complex structure that consists of ligaments, tendons, discs, capsules, and deep muscles that predominantly hold the spine in place and surface muscles that predominantly enable movement. All these structures can experience problems and consequently cause issues.^[2]

Craniocervical instability (CCI) refers to instability in the joint between the skull and the first vertebra, also called the atlas.^[3] On the other hand, **atlantoaxial instability (AAI)** describes instability between the 1st cervical vertebra (C1) and the 2nd cervical vertebra (C2), which is also called the axis.^[4] Since the main rotation of the head takes place in this area, problems frequently occur at this level.^[5] The atlas and axis differ from the other bony structures in the cervical spine because there are no discs between the skull, atlas and axis. The atlas sits on top of the axis by encircling the dens of the axis. This complex area is only held together by ligaments, tends, and capsules.

Any part of the cervical spine can be unstable. Most commonly, literature distinguishes between the **upper cervical spine** - the area between the skull and the axis - and the **lower cervical spine** - all other cervical vertebrae below. Imaging reports will describe C1, C2, etc., up to C7. The letter refers to the area of the spine (C stands for cervical), and the vertebrae are counted from the top, each receiving a number. So C1 would be the atlas and C2 the axis. The cervical spine has a huge weight to carry and is extremely flexible at the same time, so it is predisposed to injuries. At the same time, healing is complicated because ligaments and capsules are poorly supplied with blood.^[6]

What could cause Cervical Spine Instability?

There are various causes of cervical spine instability. Some are traumatic or sudden and others are gradual or insidious. Instability can develop after a whiplash injury, most often due to car accidents (MVA or motor vehicle accident).^[7] However, iatrogenic injuries (caused by medical treatment) are documented as well, for example, as a complication after chiropractic maneuvers^[8] or hyperextension of the neck during surgery.^[9] Any traumatic event to the head and/or neck, especially one that causes a quick twisting of the head^[10], can lead to instability. Chronic poor or suboptimal positioning^[11], especially from looking down at computer screens, tablets, and phones, may contribute to instability as well.

Sometimes, an underlying condition leads to instabilities:

- Genetic connective tissue disorders such as the Ehlers-Danlos syndromes or Marfan syndrome^[12]
- Other genetic conditions such as Down syndrome^[13] or Hurler syndrome^[14]
- Diseases that lead to systemic inflammation, such as rheumatoid arthritis^[15]
- Chronic infections such as Lyme disease^[16] and tuberculosis^[17]
- Malignant diseases such as cancer^[18]
- Chiari malformation surgery sometimes leads to CCI

The areas most commonly affected by instability are the ones with high flexibility or junctions between one area of high flexibility and another very inflexible one. Injuries that affect only ligaments or capsules are especially hard to identify. Subluxations of the facet joints can occur with or without bony injury and are even harder to diagnose.

What symptoms occur?

Depending on the area and severity, the symptoms^[20] ^[21] ^[22] ^[23] can vary from mild to lifethreatening.^[24] ^[25] We can distinguish between more common but unspecific symptoms and rarer but more specific symptoms.^[26]

Common & unspecific symptoms:

- Headaches and neck pain
- Muscle spasm
- Brain Fog
- Sleep apnea
- Dysautonomia symptoms, for example, nausea, diarrhea/gastrointestinal disorder, nervousness, hot flashes, and feeling cold at the same time

More specific symptoms:

- Heavy head; it feels like you cannot hold your head up
- Trouble swallowing; feeling of a lump in the throat; changes in voice quality
- Cracking, popping noise in the affected area
- Dizziness
- Trouble hearing, muffled hearing, but also hypersensitivity to sounds, tinnitus
- Central sleep apnea (comes from the brain stem)
- Drop attacks
- Balance issues, disturbances of fine motor skills (difficulty grasping glasses or small objects, dropping objects), poor coordination
- Tingling and numbness of face, arms, and legs
- Paralysis, numbness, pins and needle sensation, tingling
- Hypersensitivity syndromes, including light and sound sensitivity
- Visual disturbances of all kinds, trouble focusing, strabismus
- Involuntary twitching and dystonia
- Cognitive, mental, and personality changes (depression, depersonalization, memory loss)

Other symptoms:

- Pain all over the body
- Vertigo
- Burning sensation along the spine (neuropathic pain), but also along the nerves of the spine (hands, face)
- Muscle loss and weakness
- Weakness all over the body
- Tremor

Possible consequences of CCI and AAI could be:

- Myelopathy
- Brain stem compression
- Jugular Vein compression
- Intracranial hypertension
- Vagus Nerve dysfunction/degeneration
- Vertebrobasilar invagination
- CSF flow changes

Severe CCI/AAI are rare while mild forms of CCI/AAI but also lower cervical spine instability, particularly in the EDS community, are quite common.^[27]

How to get diagnosed?

Diagnostic tests can be done in a variety of positions. The body can be positioned supine (laying flat on your back) or upright (vertical / weight-bearing). Instabilities are often missed on static (still) imaging. Dynamic imaging (flexion, extension and/or rotation) often detects instabilities due to (isolated) ligamentous injury or laxity that are hard to diagnose and often missed on static imaging.^[28] The movement of the neck can either be done by the patient or with the assistance of another person. Some people experience a flare of their symptoms after dynamic imaging. Neurosurgeons (and other clinicians treating these complex conditions) often have very specific preferences for the imaging views they want to see before making a diagnosis and/or treating these challenging problems.

Diagnostic tests:

- Upright dynamic MRI^[29]: This MRI is done while sitting (weight-bearing) in an open system with the head in maximum flexion and extension (and in some countries such as Germany, also in rotation and bent to both sides). Advantages: not static, upright, weight-bearing, shows connective tissue in motion, allows better assessment of upper and lower cervical spine stability or instability, no radiation; disadvantages: low-resolution imaging due to (unavoidable) motion artifact.
- Flexion-Extension X-ray^[30]: Traditional x-ray imaging with some of the same advantages as above. Advantages: accessible and low cost; Disadvantages: radiation exposure, no visualization of connective tissue or soft tissues. Traditional x-ray imaging detects primarily bony abnormalities and has many other diagnostic limitations.
- 3D CT Scan with Rotation^[31]: A CT scan with rotational views allows for assessment of rotation between C1 and C2, which is mainly limited by the ligamenta alaria.
 Rotational CT is the preferred method to evaluate C1/2 rotational instability.
 Advantages: best assessment of C1/2 rotational instability. disadvantages: high radiation, no visualization of soft tissues.
- DMX (Digital Motion X-ray)^[32]: DMX is real-time imaging mostly used by chiropractors in the US. The cervical spine is visualized during movement with continuous radiation. Advantages: dynamic imaging may show instabilities that might otherwise undetected^[33]; disadvantages: not available outside of the US, significant radiation exposure.

Additional Diagnostic:

Those tests are not standard diagnostic tests and are only done in special circumstances. Varying countries also have different evaluation protocols for cervical spine instabilities.

- Detailed neurologic physical examination^[34]: Testing should include the following: gait evaluation, cranial nerve exam, coordination tests (eg: looking for Dysdiadochokinesia), testing muscular strength and sensation, observation of any abnormal movements (eg: dystonia), and reflex testing (eg: Hoffmann sign, Babinski reflex, deep tendon reflexes, and clonus). This physical examination should be part of a comprehensive evaluation of a patient presenting with clinical concern for cervical instability. These tests must be performed by a qualified clinician avoiding maneuvers that may increase irritability. Some clinicians will prescribe a neck brace trial as part of the diagnostic evaluation. Keeping a journal during this time is extremely important in order to correlate symptoms.
- **Otoneurology**^[35]: Otoneurology evaluates the nerves connecting the brain, eyes, and ears. The tests may be exhausting and increase the patient's symptoms.
- **Positron-Emission-Tomography (PET)**^[36]: PET is an imaging method that evaluates the biochemical and physiological function of a particular organ (eg: the brain).
- Autonomic nervous system diagnostics^[37]: ANS testing is used to show disturbances of the autonomic nervous system, which occur with many different neurological disorders, for example, Parkinson's, Multiple Sclerosis, or cervical spine instabilities. Testing can include the tilt table test, sweat tests, and heart rate variability under different circumstances.
- Ultrasound is an imaging technique where sound waves are bounced off of structures in the body (eg: the neck, including the jugular vein and vagus nerve).
 There is no radiation and a variety of views can be obtained to get a more complete picture including cross-sectional areas.^[38] ^[39] ^[40]

Physical therapists: They can use various manual testing techniques to evaluate for instability. Two tests that have been described as very useful include the sharp purser test^[41] to examine the ligamentum transversum and the passive intervertebral movement test (PIM test)^[42] to evaluate the ligamentum alaria and transversum, as well as the membrana tectoria and posterior atlantooccipitalis. A recent publication suggests using a modified sharp purser without provocation, alar ligament test, cervical distraction, as well as mobility and provocation tests.^[43] It is extremely important to note that these tests should only be performed by qualified individuals and only after performing a comprehensive assessment to determine irritability. Some patients (especially ones with moderate irritability) will experience a severe flare of their symptoms following these maneuvers requiring a considerable period of time to recover (if they recover at all). Severely unstable or irritable patients may experience a neurologic emergency if inappropriate maneuvers are performed.

Angles that can be assessed on imaging by a qualified provider^[44]:

- Bulls angle
- Powers ratio
- Basion dens interval (BDI)
- Basion Axial interval (BAI)
- Harris measurement
- McGregor's line for basilar invagination
- Grabb Oaks line for brainstem compression
- Wackenheim line
- Clivo axial angle (CXA)

Important notes:

- These tests are challenging to perform and interpret. They are only as accurate as the examiner is competent: Meaning if these tests are not performed by qualified professionals knowledgeable about assessing cervical spine instability, the findings are useless.^[45]
- Get more than one opinion when possible and preferably from an expert in the field (if there is none in your country, some experts offer online consultations).
- The severity of the symptoms does not always correlate with the severity of the imaging and vice versa.^[46] Imaging findings must always be correlated with the clinical presentation.
- Measurements:^{[47] [48]} CXA: 145 to 160 degrees is normal, below 135 is an indication for surgery, Horizontal Harris: more than 12 mm indicates instability, Grabb, Mapstone, Oaks: more than 9 mm indicates brain stem compression.
- It's important to know that no formal studies have been done to evaluate normal versus pathological angles. Your clinical team will evaluate your imaging findings in the context of your physical exam and make recommendations accordingly.

How to treat Cervical Spine Instability? Non-invasive Treatments

In general:

- No treatment works for everyone!
- High-velocity chiropractic maneuvers are mostly contraindicated for all people with instabilities.^[49] So are traction devices in many settings!^[50]
- Often, reducing muscle spasms around the neck worsens the symptoms. The body creates muscle spasms to compensate for instability.
- Always listen to your body and stop if something doesn't feel right.
- You will need a very well-educated and experienced physical therapist who always listens to your symptoms and knows how to handle them.
- It may help to start very slowly in a prone position (depending on the severity of your symptoms) and only with the weight of the body and then gradually increase exercises as tolerated. Some therapists will start exercises with a neck brace on.

Non-invasive therapies:

- Neurophysiological concepts such as PNF or Vojta therapy^[51] may have a positive effect on certain neurological conditions, for example, stroke and spinal cord damage.
- Wearing a neck brace,^[52] ^[53] ^[54] especially when the symptoms are severe, may help control symptoms and prevent further damage but should be done under the guidance of your clinical team. Braces should not be worn full-time due to the possibility of increasing muscle weakness and increasing instability.
- Physical therapy such as isometric exercises, muscle building, hydrotherapy, and Brunkow therapy^[55] aimed at strengthening the core muscles are beneficial for the cervical spine as well. Manual therapy, such as myofascial release or trigger point release, if tolerated, may be helpful as well as muscle energy techniques.^[56]
- Taping^{[57] [58]} might reduce pain, can passively stabilize the spine, and increase blood flow.
- Pilates^{[59] [60]} strengthens core muscles and musculature throughout the body positively affecting the cervical spine when applied appropriately.
- Med-X training^[6] might be too much strain for severely affected individuals, but it could increase muscle strength for others.
- Osteopathy^[62] may be helpful when applied by a qualified provider knowledgeable about these conditions.
- Craniosacral therapy is generally very gentle and may help reduce pain in some patients.
- Nutritional supplements may improve a variety of symptoms including fatigue, muscle strength, and correct micronutrient deficiencies (especially in those with gastrointestinal problems).^[63]
- Patients have reported benefits with regenerative therapies such as prolotherapy with or without PRP^{[64] [65]}, and cell based injections but more research is desperately needed.

- Jaw pain or dysfunction of the temporomandibular joint is extremely common in those with symptomatic joint hypermobility or hereditary disorders of connective tissue (HDCT) like EDS (often referred to as TMJ but the appropriate term is TMD). TMD may greatly impact neck pain and other symptoms related to cervical instability. Addressing TMD may help alleviate symptoms of cervical instability.^[66]
- Relaxation and breathing techniques are very useful.^[67]
- Improve posture and activities of everyday life (eg: avoid carrying heavy bags, carrying bags on one side, etc).
- Use orthotics (OTC or custom) and braces for unstable joints as needed.^[68]

Invasive Treatments:

- Anterior (from the front) and posterior (from the back) surgery use different hardware and have different risks, indications and benefits.
- Anterior surgery is used most frequently for problems in the lower cervical spine. The surgeries of CO, C1, and C2 are usually done posteriorly using a combination of plates and screws (due to limited access from an anterior approach).^[69]
- Some surgeons perform minimally invasive C0-C2 fusion using only condylar screws.
- All surgeons develop and refine their techniques over the course of their career.
- Ask lots of questions (before any surgery) including:
 - Why do you propose this particular technique?
 - What are your outcomes for my specific problem?
 - How many surgeries have you performed (total and for my planned procedure)?
 - What will happen if you encounter an unexpected finding during surgery?
 - Who is the ideal candidate for my procedure?
 - Who is not a candidate for my procedure?
 - Why do you think I am a good candidate for this procedure?
 - What reservations do you have about performing this procedure on me?
 - What is most challenging from a technical standpoint for this procedure?
 - What do you anticipate being most challenging in my specific case?

Important Notes for Surgery:

- Worldwide, only a few neurosurgeons have expertise in CCI surgery for people with connective tissue disorders. It's important to see someone who is well-versed with the complications of these conditions and understands all comorbidities of EDS as well. You'll need a multi- and interdisciplinary team that understands EDS and its comorbid conditions pre- and post-op.
- The better your muscles work before surgery, the "easier" the recovery after.^[7]
- Surgery is always the last option when all conservative treatment options fail.
 Surgery always has risks. Surgery in those with EDS should always be undertaken after careful consideration of the alternatives and risks.
- Surgery is not a cure; outcomes differ widely. Symptoms can improve but they can also worsen. Surgery of the upper cervical spine is complicated and risky due to its precarious location. Recovery can take months to years.
- Fusion of one segment of the spine in people increases the load in the neighboring segments^[72]. Especially in people with connective tissue disorders, the risk of needing further surgery (eg: fusions) is an important consideration.
- Surgery for CCI in EDS patients is controversial. There is a tremendous lack of data and research. There are no long-term outcome studies after CCI surgery with EDS and these are desperately needed.

Prevention: [73] [74] [75] [76] [77] [78]

Especially in people with connective tissue disorders like EDS, preventing any condition is far preferred over treating that same condition. Cranial instability is especially important to prevent since it can be particularly difficult to treat.

- Avoid collision sports: Avoidance of any sport that could cause harm to the cervical spine (eg: boxing, football, soccer, etc) should be considered even if a person is completely asymptomatic. Choose activities that train the core muscles and proprioception and don't strain joints (eg: biking, kayaking, paddleboarding, swimming, and walking). Use the swimming stroke that feels best on your body. Avoid risky activities especially ones where falls are likely.
- Avoid strain on the neck: Try not to stare down at a phone or laptop put those at eye level, always look straight in the mirror or at the person in front of you, and avoid too much head rotation. Don't carry too much weight on your back or in your hands. Also, distribute weight equally on both sides. Be mindful when driving a car.
- Use assistive devices: For instance, you can use wedges for sitting, prism glasses, use a headset when on the phone for an extended period of time; use chairs or similar if you want to reach something higher up. Do not hyperextend the neck to look up or drink from a glass.
- Good posture when sitting, standing, or lying down: With desk jobs, contract your abdominal muscles when looking at a computer screen. Standing desks and ergonomic chairs (eg: rocker bottom kneeling chairs) can be helpful. Use pillows to support posture, such as neck pillows and body pillows for sleeping, u-shaped pregnancy pillows to prop up hips and shoulders, and/or braces that support good posture. Sleep on your back or side, but not on your belly! Change your position frequently throughout the day.
- Work to improve your proprioception (knowing where your body is in space without looking). Many of us are unaware of putting our joints in vulnerable positions due to poor proprioception.

- Caution with surgery and anesthesia: Your head and neck may be repositioned under general anesthesia so be sure your care team is aware of any concerns you have. This is especially important since you will be unable to tell them at the time that a given position is uncomfortable for you. Ask if your body will be repositioned under anesthesia as this is a time of vulnerability as well. When in doubt, mention any concern of joint instability or hypermobility to your care team.
- Caution with dental appointments: Dentist appointments cause strain on the neck and jaw due to positioning and having the mouth open for a prolonged period of time.
- Manage stress with relaxation techniques and build strength in your neck. Use heat or cold as needed. Don't clench your teeth. Address jaw issues immediately because they can contribute to neck problems.
- Pace yourself. Avoid the boom and bust cycle. It is easy to overdo it when you feel good because you feel so behind but avoid that temptation. Be patient with yourself and rest when needed. Work on getting good quality sleep when it is dark and being active (outside with fresh air if possible during daylight) to optimize the circadian rhythm.

Further reading:

We recommend reading the following articles/books for a great summary of symptoms and management of upper cervical spine instability:

Russek, L.N., Block, N.P., Byrne, E., Chalela, S., Chan, C., Comerford, M., Frost, N., Hennessy, S., McCarthy, A., Nicholson, L.L. and Parry, J., 2023. Presentation and physical therapy management of upper cervical instability in patients with symptomatic generalized joint hypermobility: International expert consensus recommendations. Frontiers in Medicine, 9, p.4020. Jovin, D., Barnum, R., Atwal, P., Bluestein, L., Chopra, P., Dempsey, T., Dorff, S., Herman, K., Hamilton, M., Koby, M., Klinge, P., Maitland, A., Maxwell, A. J., Mitakides, J., Pocinki, A. G., Rosenthal, L., Saperstein, D., Schofield, J., Tishler, J., Block, E., Block, N. 2020. Disjointed, Navigating the Diagnosis and Management of hypermobile Ehlers-Danlos Syndrome and Hypermobility Spectrum Disorders. Hidden Stripes Publications, Inc.

Specifically, Chapter 15: Imaging and Cervical Instability, by Dr. Miles Koby.

Henderson Sr, F.C., Austin, C., Benzel, E., Bolognese, P., Ellenbogen, R., Francomano, C.A., Ireton, C., Klinge, P., Koby, M., Long, D. and Patel, S., 2017, March. Neurological and spinal manifestations of the Ehlers–Danlos syndromes. In American Journal of Medical Genetics Part C: Seminars in Medical Genetics (Vol. 175, No. 1, pp. 195-211).

Websites: https://bobbyjonescsf.org/

Image: Cervical Vertebra: Image by OpenClipart-Vectors from Pixabay

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Holding your head up shouldn't be this hard.

This patient guide was written by patients for patients and reviewed by medical professionals. Craniocervical instability is a challenging condition in terms of diagnostics but also management where patients might feel left alone with their symptoms. EDS Awareness/Chronic Pain Partners and the media team have worked to provide the best current information to guide you in determining what is best for you. As always, it is not intended as medical advice and patients should be evaluated by trained medical professionals with expertise in this niche area of specialty. We do recognize there are few providers and neurosurgeons who treat CCI. Many are simply overwhelmed and overworked with waiting lists too long to help many people in difficult situations in need of urgent care. We hope the future of CCI examinations, testing and treatment will advance to meet the demanding needs of today's patients.

Stay informed on EDS-related information at www.edsawareness.com.