Upper Cervical Instability in Hypermobility Spectrum Disorder

DISCLAIMER: You cannot diagnose UCI based on the information here. These management recommendations are generally appropriate but might not be appropriate for you. Always check with your health care provider for proper diagnosis and management.

This information is based on the publication: Russek LN, Block NP, Byrne E, Chalela S, Chan C, Comerford M, Frost N, Hennessy S, McCarthy A, Nicholson LL, Parry J, Simmonds J, Stott PJ, Thomas L, Treleaven J, Wagner W and Hakim A (2023) Presentation and physical therapy management of upper cervical instability in patients with symptomatic generalized joint hypermobility: International expert consensus recommendations. Front. Med. 9:1072764. It is available at: https://www.frontiersin.org/articles/10.3389/fmed.2022.1072764/full. Case studies at https://www.frontiersin.org/articles/10.3389/fmed.2022.1072764/full#supplementary-material

It is important to realize that instability is not the same as hypermobility. Hypermobility is about having too much motion at joints. Instability is about not being able to control motion at the joints; instability means that the muscles and nerves are not able to sense where the joint is, or are unable to control movement at the joint. Proper training can often teach people with hypermobile joints how to maintain stability at those joints. Physical therapy can often help decrease cervical instability.

- **Determining you have upper cervical instability (UCI) requires the following 3 criteria be met:**
    1) **Symptoms of either musculoskeletal or neurological UCI (Table below).** Some people may have mostly musculoskeletal problems from instability, while others will have neurological symptoms; however, people often have some of both, and the proportion can change from day to day. Some symptoms are 'common but not diagnostic,' which means that people with UCI are very likely to have these symptoms, but the symptoms are common for other reasons, as well, so they don't prove UCI. However, if they are absent, UCI is less likely to be present.

Symptoms Suggestive of Musculoskeletal or Neurological Upper Cervical Instability

	Common	Highly suggestive
Musculoskeletal UCI		
· Heavy/bobble head, you feel like you need to support or brace your head to decrease symptoms		X
· Apprehension about initiation or maintenance of neck movement or travel in vehicle		X
· Lump in throat, trouble swallowing		X
· Consistent clicking or clunking in the neck associated with neck movement		X
· Cervical sensorimotor symptoms such as tinnitus, dizziness		X
· Suboccipital headaches; Yoke/coat-hanger distribution pain; Neck tension, muscle spasm	X	
· Brain fog	X	
· Inconsistent or poor response to treatment for the neck	X	
· Sleep disturbance, snoring, sleep apnea	X	
Neurological UCI		
· Seizure-like activity, diagnosis of 'non-epileptic seizures' or 'pseudo seizures'		X
· Passing out not associated with dysautonomia (are provoked by neck motion, or without POTS symptoms)		X
· Lump in throat, choking, trouble swallowing, voice changes		X
· Dysautonomia not responding to standard treatment	X	X
· 'Boat rocking' instability (not due to musculoskeletal issues); Ataxia: Poor coordination (not due to joint instability)		X
· Facial tingling/numbness; Pulling sensation in face, head, teeth, tongue (muscle contraction, not just pain)		X
· Vision changes- trouble with convergence, double vision, aura (teichopsia)		X
· Dystonia: involuntary muscle contractions causing involuntary movements or postures		X
· Intermittent dysesthesias (numbness or tingling) in the limbs, not associated with local issues		X
· Sleep disturbance, snoring, sleep apnea	X	
· Cognitive changes, more than brain fog		X

2) **Symptoms are altered by neck movement or position:**
   - If UCI symptoms (besides just pain) are increased with neck motion, forward head posture or activity (e.g. working at a computer), leaning forward. If you are afraid to tip your head back, as in washing your hair or going to the hairdresser. Also, if symptoms increase when you are upright with the neck unsupported, if symptoms feel better when wearing a neck brace. If you are afraid that someone touching your neck will aggravate UCI symptoms. If these symptoms do not change with neck position or movement, the symptoms are likely caused by something other than UCI.

3) **Symptoms are mechanically irritable:**
   - Your UCI symptoms are easily aggravated: it is aggravated by minor things, or the flare is out of proportion to what caused the flare. For example, being a passenger in a car for 10 minutes doesn't aggravate a healthy neck; if that aggravates your neck, it is 'irritable'.
   - Once aggravated, your UCI symptoms take a long time to calm down (several hours for neurological symptoms, 24 hours for pain. Or if you are unable to be upright (out of bed) for more than 24 hours after a flare because of UCI symptoms. Flares that last a long time suggest the neck is 'irritable'.
   - UCI symptoms are severe: e.g., bed-bound, need to use a wheelchair or walker due to coordination problems not associated with the legs, choking or trouble swallowing, serious visual changes, severe nausea with neck movement (not vestibular). If you experience these problems, talk to your MD.

- **<u>If you think you have UCI, how would it be diagnosed?</u>**
  a. There are some physical tests that can determine whether neurological structures in the neck are functioning properly. These tests include reflexes, cranial nerve tests, and coordination tests. There are also some physical tests to determine whether there is too much motion at the joints; however, as noted above, having too much motion doesn't always mean the joint is unstable; also, some instability tests might cause serious flares. Finally, there are some physical tests that identify risk factors that can increase instability, such as poor posture, poor body awareness or poor motor control.
  b. An MRI test done with the neck in full flexion and extension can see whether the vertebrae move too much, as well as whether something is pressing on the spinal cord. Some experts feel the MRI needs to be done upright instead of lying down, but upright MRI are not always available. Flexion and extension x-rays might be helpful, but are not as accurate as MRI.

- **<u>If you have UCI, what can you do about it?</u>**
  a. Functional training: how to protect your neck during daily activities:

<u>Examples of Functional Training for Hypermobile People with Irritable UCI</u>:

- Use a sitting hip hinge to bend forward for cleaning teeth, reaching at the table, leaning forward for eating foods, etc. In severely unstable UCI and intolerant to upright sitting use wedge lying for eating. A hard cervical collar may be needed to avoid excessive cervical-spine movement.
- Lie down for things like putting in contacts, to prevent you from moving your neck in bad ways.
- Turning in bed: using one sided bridge and reach with top arm to log roll to side lying from supine lying.
- Look straight into the mirror for positional cueing: hair brushing, drying with blow dryer. Never bend into flexion/extension/side/rotate to dry hair. A hard cervical collar may help avoid excessive cervical spine movement.
- Belly button rule: Always face people you are talking with or directions you need to look. Cueing is your nose needs to stay in alignment with the belly button, turn your feet or body to keep alignment.
- Toileting: Avoid straining, use a squatty potty if constipated. Use long reacher for buttock care to avoid twisting.
- Dressing: Dressing stick for donning shirts, lying down for pants, socks, slip on shoes or elastic shoelaces, long shoehorn, etc. to minimize bending over to dress.
- Sexual activity: Person who has instability should be on the bottom position. The person without instability should be the more active partner. Clearance from MD is recommended.
- Cooking: Pre-packaged foods requiring little prep time, microwave meals, etc.
- Electronics usage (e.g., phone): side-lying, supports for holding devices, holding phones at eye level, avoid looking down at devices ('text-neck').

  b. Be mindful of your body position sitting, standing, sleeping. Make sure you are in good posture sitting, standing, sleeping. Use good body mechanics and ergonomics when using computers/technology. (see picture, next page)

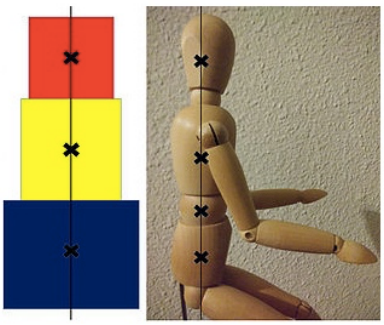


Figure 1. Stacked, aligned posture

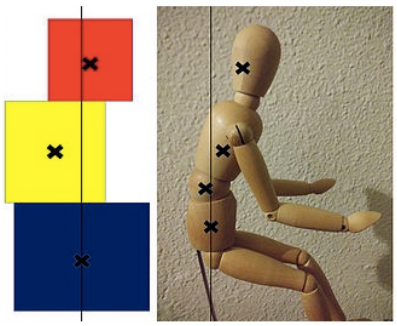
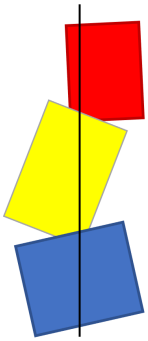


Figure 2. Postural misalignment



This picture of posture as stacked blocks shows how poor posture stresses muscles and joints, and why it is important to correct posture from the pelvis or feet up. Without a stable base (feet if standing, pelvis if sitting), the neck can't be stable. Picture from http://www.voxhumanajournal.com/huffington2018.html

c. Use orthotics or braces (if appropriate) for the feet, legs, and low back to improve alignment. The neck can only be stable on a stable base, from the feet up.

a. Calm a sensitive nervous system. Many people with UCI also have a sensitive central nervous system, especially if symptoms have been present a long time or have been frightening. Calm the nervous system through relaxation training, slow diaphragmatic breathing, cognitive behavioral approaches (e.g., avoiding negative thinking or catastrophizing). If hands on treatment (manual therapy or massage) or exercise only help for a few hours or make you worse, you probably have sensitive nerves.

b. Create and use a self-care 'toolbox' to manage pain: heat, ice, transcutaneous electroneural stimulation (TENS), topical analgesics, relaxation, positive thinking, etc.

c. Use a rigid neck brace, if/when necessary. Commonly used braces include the Aspen™, Miami-J™. For milder UCI, a posture reminder such as the BACK Neck Brace™ might help. We think most people should use a neck brace only when needed (e.g., during a flare or car travel), but people with severe instability might need it all the time. It is important that neck muscles do not become weaker. Braces should be fit by a trained professional.

d. If UCI is severe, manual therapy is probably not appropriate, but very gentle manual therapy to address muscle spasm might be helpful. High speed manipulation is not safe.

e. If UCI is severe, begin 'exercise' away from the neck. For example, start with pelvic and lumbar motor control training lying down with the head supported. For mild-moderate UCI, you might be able to do body awareness and motor control training of the neck, shoulders or shoulder blades early on. Avoid chin tucks. Focus on body awareness and proprioceptive training. Avoid large neck motions and neck resistance until you are doing quite well. Ideally, exercise should not increase pain. However, slight increases in discomfort of no more than 2 units (on 0-10 scale) for 2 hours are sometimes acceptable. Talk to your healthcare provider.

f. Aerobic exercise, if tolerated. For severe instability, you probably need a non-impact activity, such as a recumbent bike/peddler. Once instability is mild-moderate, you may tolerate walking. Be sure to keep good postural alignment. Again, this activity should not increase UCI symptoms much, if at all.

g. Do NOT have neck traction. Although it may feel good at the time, experts believe it is not good for UCI.

"**Red Flags**" are indications that there might be something that requires urgent medical attention. Since HSD is a complicated condition, many of these 'Red Flags' have exceptions, as noted below.

Red Flag: Talk to your healthcare provider	**Might not be *as much* of a concern if…**
Seizure-like activity, pseudo-seizures	Always a concern if not yet diagnosed
Rapidly progressing neurological signs (physical abnormalities, not just symptoms) with decreasing functional status	Always a concern if not yet diagnosed
Fainting not associated with orthostatic intolerance	Fainting due to POTS, diagnosed narcolepsy
Altered consciousness or memory, severe changes in cognitive status	Brain fog aggravated by stress and pain
Increased bowel/bladder control dysfunction	Known incontinence or prolapse (e.g. post-partum or long-term)
Headache worse with Valsalva maneuver (holding breath and blowing)	You've had headaches like this for a long time
Need to use a walker/WC due to severe/variable coordination problems	Need due to pain or weakness in legs, or fatigue