

FIGURE 1

Flow chart describing the screening process for determining that patients have upper cervical instability (UCI) associated with symptomatic generalized joint hypermobility (S-GJH), irritability and identifying yellow flags and red flags. The screening steps can be implemented in any order, using the check boxes on the right to keep track of decisions made at each step. See text and tables for more detailed discussion of each step in the process.

Common Upper Cervical Instability (UCI) **Symptoms and mechanical responses** of symptoms in patients with symptomatic generalised hypermobility

TABLE 2 Symptoms suggestive of musculoskeletal or neurological upper cervical instability.

	Common	Highly suggestive
Musculoskeletal UCI		
• Heavy/bobble head, patient feels like they need to support or brace their 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	X	
• Yoke/coat-hanger distribution pain	X	
• 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		
• Report of seizure-like activity, diagnosis of “non-epileptic seizures” or “pseudo seizures”		X
• Drop attacks not associated with dysautonomia (e.g., provoked by neck motion, or without dizziness common in POTS)		X
• Lump in throat, choking, trouble swallowing, voice changes		X
• Symptoms of dysautonomia (especially if not responding to standard treatment), persistent anxiety, functional GI dysfunction, poor temperature regulation, heat intolerance, presyncope,	X	X
• “Boat rocking” instability (not due to musculoskeletal issues)		X
• Ataxia: Poor coordination (not due to joint instability)		X
• Facial tingling/numbness		X
• 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 in the limbs, not associated with local issues		X
• Sleep disturbance, snoring, sleep apnea	X	
• Cognitive changes		X

TABLE 3 Symptoms of upper cervical instability (UCI) are altered by neck movement and/or position.

- Increased symptoms with neck motion into, or when held in, flexion, extension, and/or rotation, especially increased neurological symptoms
- Apprehension about neck extension (e.g., washing hair, going to the hairdresser)
- Increased symptoms when leaning forward, looking down
- Increased symptoms with forward head posture, e.g., using computer keyboard
- Increased symptoms when upright with neck unsupported
- Decreased symptoms when in neutral or wearing a neck brace
- Apprehension, anxiety, or fear of manual exam to the neck

Assessing and categorising irritability of Upper Cervical Instability (UCI) in patients with symptomatic generalised hypermobility to guide assessment and management

TABLE 4 Symptoms are mechanically irritable.

A. Condition is severe:

- Poor tolerance to any time vertical
- Bed bound due to cervical symptoms
- Need to use a walker or wheelchair due to moderate or intermittently severe problems with coordination and balance rather than pain or weakness, or restricted to bed due to cervical symptoms
- Extreme cervical spine guarding with fear of movement secondary to severe reactivity
- Choking, trouble swallowing, and voice changes
- Profound visual disturbances
- Severe nausea with any neck movement
- Functional outcome measure relevant to UCI classified as Severe

B. Condition is easily flared:

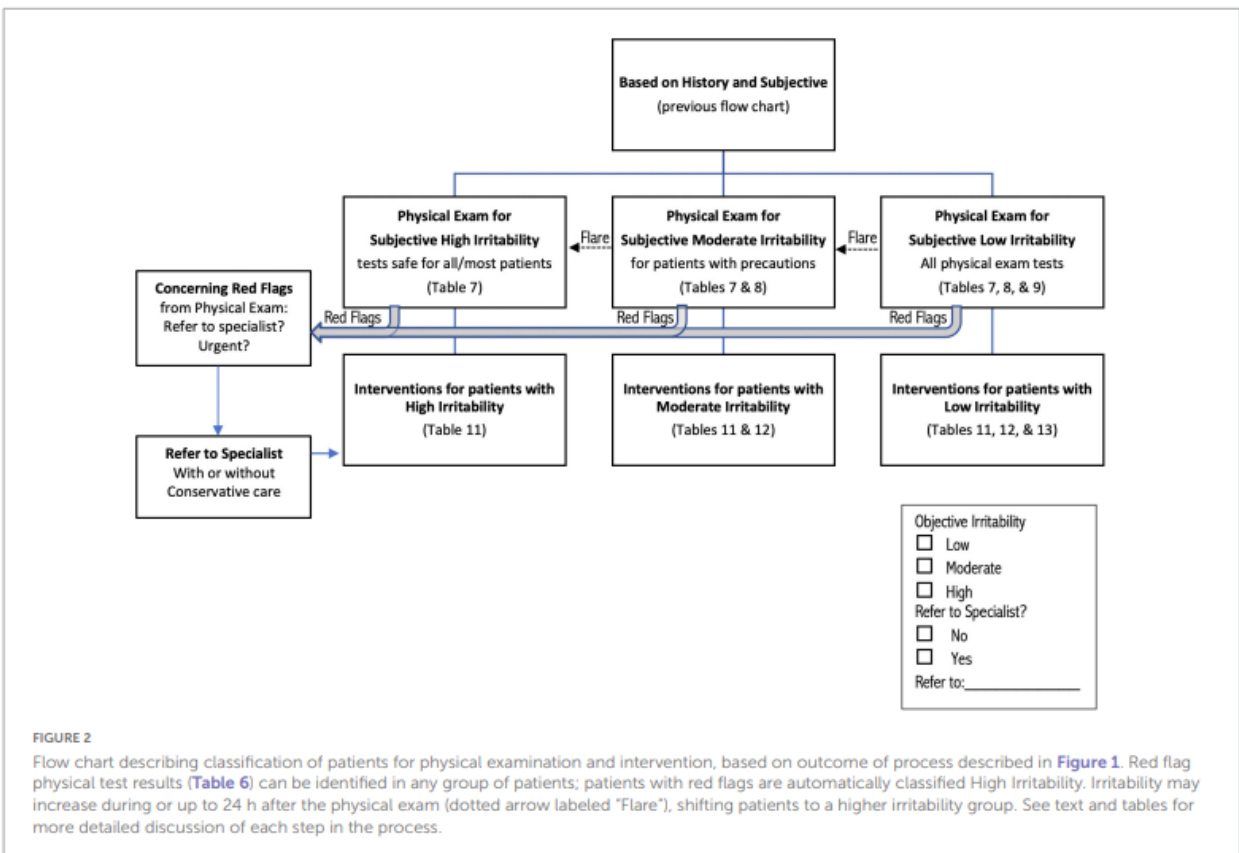
- UCI flares are disproportionate compared to provoking insult or activity. e.g., aggravated by minor rapid/unexpected movements/perturbations, traveling in car/bus, prolonged postures.
- Presyncope, syncope, drop attacks or seizure-like episodes with neck extension or rotation.
- History of excessive provocation associated with previous conservative care including hands-on manual therapy or exercise.

C. Prolonged time to calm after flare:

- Provoked UCI symptoms take excessive time to settle to pre-flare state: e.g., more than 24 h for pain or more than several hours for neurological symptoms
- Pt regularly needs to resort to wearing a cervical collar or bedrest to ease symptoms after a flare
- Inability to tolerate being upright for > 24 h after flare

Grading mechanical irritability

- Low irritability:
 - A, B, and C are all typically absent, or
 - B or C might be occasionally present at a low level.
- Moderate irritability:
 - A, B, or C are intermittently present, or
 - A or B or C is frequently present, but not all three consistently.
- High irritability:
 - A, B, and C are all frequently present.



Screening red and yellow flags and differential diagnoses in Upper Cervical Instability (UCI) in patients with symptomatic generalised hypermobility

TABLE 5 Some recommended yellow flag assessment tools.

- OSPRO-YF 10 or OSPRO-YF 17: Multidimensional assessment tools for identifying YF in physical/physiotherapy patients (30)
- Anxiety and depression questions from the spider impact scale developed specifically for patients with HSD/hEDS (31, 32)
- Tampa scale of kinesiophobia (TSK) any version (anxiety and fear of movement) (33)
- Fear avoidance beliefs questionnaire (FABQ) (anxiety and fear of movement) (34)
- Orebro musculoskeletal pain questionnaires (multidimensional) (35)

TABLE 6 Red flags (36, 42, 43).

- History and symptoms**
- Seizure-like activity, pseudo-seizures
 - Rapidly progressing neurological signs with decreasing functional status
 - Drop attacks or syncope not associated with orthostatic intolerance (e.g., HR and BP changes)
 - Altered consciousness or memory, severe or frequent changes in cognitive status
 - Increased bowel/bladder control dysfunction
 - Headache worse with Valsalva maneuver
 - Need to use a walker or wheelchair due to moderate or intermittently severe problems with coordination and balance rather than pain or weakness
 - Symptoms significantly increased after MVA, whiplash, trauma
- Physical examination**
- Abnormal central nervous system reflexes: Babinski, Hoffmann, clonus, hypertonia
 - Abnormal cranial nerve findings: Altered visual field, eye movement, unequal pupil size, amblyopia (lazy eye), facial sensory loss
 - Observed speech or swallowing dysfunction, choking, tongue dysfunction, sleep apnea (lower cranial nerves)
 - Abnormal vertebrasilar insufficiency tests with auditory and vision changes, evidence of vertigo, presyncope or syncope
 - Ataxia, gross neurogenic gait abnormalities, inability to perform tandem gait, Romberg sign present
 - Dysdiadochokinesia: e.g., rapidly alternating pronation/supination, grip release test, fast finger or foot tapping
 - Dystonia, myoclonic jerking
 - FASTER Indications of stroke: Face, Arms, Stability (standing), Talking, Eyes. R is for React.

TABLE 10 Differential diagnoses that should be considered (1, 6, 39, 40).

- Chiari malformation
- Migraine/headache
- Intracranial hypotension (cerebrospinal fluid leak)
- Idiopathic intracranial hypertension
- Tethered cord
- Eagle syndrome
- Dysautonomia unrelated to cervical instability
- Functional neurologic disorder
- Functional movement disorder
- Tarlov cysts

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Physical examination of Upper Cervical Instability (UCI) in patients with symptomatic generalised hypermobility according to irritability grading

TABLE 7 Physical test and findings, safe for all patients.

	Contributing*	Common*	Diagnostic*
Observation based tests			
Posture, full body, sitting and/or standing, and segmental alignment	X	X	
Breathing pattern (chest vs. diaphragmatic, excessive accessory muscle use)	X	X	
Significant muscle guarding or reluctance to move neck		X	
Observe gait for ataxia, gross, and fine motor dyscoordination not due to other joint hypermobility			X
Observe for cranial nerve VII dysfunction: Lip drooping, unequal smile, eyelid twitching			X
Observe for dystonia, myoclonic jerking			X
Neurological tests			
Cranial nerve III, IV, VI tests: Oculomotor nerve/eye movement			X
Reflex tests not involving neck: e.g., Hoffmann, Babinski, clonus			X
Cranial nerve X, XII tests: Uvula, tongue (avoid gag)			X
Dysidiadochokinesia: e.g., rapidly alternating pronation/supination, fast finger or foot tapping			X
Testing of hand dexterity (need to distinguish from finger hypermobility). E.g., grip release test			X
Other tests			
Palpation for muscle spasm, especially suboccipitals, sternocleidomastoid, levator scapulae, upper trapezius		X	
Use of a rigid cervical brace for several weeks decreases signs and symptoms			X

TABLE 8 Physical tests and findings for moderate and low irritability patients only.

	Contributing*	Common*	Diagnostic*
Other motion and control			
Thoracic range of motion, range, and quality	X	X	
Scapular muscle strength and motor control	X	X	
Excessive use of temporomandibular muscles to provide cervical stabilization (secondary finding)		X	
Neck motion and control			
Cervical range of motion: Overall, looking for apprehension, range, and quality		X	X
Deep neck flexor recruitment efficiency	X	X	
Cervical stabilizer motor control inhibition and inefficient recruitment (e.g., craniocervical flexion test, suboccipital extensor test)	X	X	
Sensorimotor tests: Eye-head coordination, trunk-head coordination, smooth pursuit visual tracking	X	X	
Cervical proprioception: Joint position error	X	X	
Other tests			
Neurodynamic tests may be cautiously performed, eliminating or caution with neck motion	X	X	
Orthostatic intolerance: NASA lean test or stand test		X	X
Structural tests			
Cervical axial load in supine			X
Alignment of C1 (manual assessment)			X

TABLE 9 Physical tests and findings only for low Irritability patients.

	Contributing*	Common*	Diagnostic*
Ligamentous testing			
Abnormal passive accessory intervertebral movements (PAIVMs) or passive physiological intervertebral movements (PPIVMs) at OA and AA (if trained)	X	X	
Alar ligament test			X
Modified sharp-purser cervical instability relocation test (NOT the provocation test)			X
Cervical distraction in supine		X	X
Mobility tests			
Isolated AA ROM		X	X
Neurodynamic tests with neck motion		X	X
Provocation tests			
Craniocervical flexion test provocation of UCI symptoms.		X	
Vertebrobasilar insufficiency positional test			X

*Contributing factors = not diagnostic but providing information about potential causes.

*Common = findings that are likely to be fairly common, but not necessarily diagnostic.

*Diagnostic = findings that are likely to be less common, but more diagnostic.

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Interventions in Upper Cervical Instability (UCI) in patients with symptomatic generalised hypermobility according to irritability

TABLE 11 Interventions for all patients, and interventions to avoid in high irritability patients.

<p>General education</p> <ul style="list-style-type: none"> o About S-GJH and UCI o "Safety netting": recognizing signs and symptoms that trigger emergency or urgent follow-up or referral; self-care in these situations (e.g., wear cervical brace) <p>Posture and body mechanics education</p> <ul style="list-style-type: none"> o Sitting, standing, and sleeping posture, positioning, and body support o Body awareness and mindfulness in various positions (sitting, standing, lying down) o Avoiding or limiting neck motion if small range motion is safe o Functional training for posture and joint protection during essential ADLs such as bathing, brushing teeth, brushing hair, washing hair, sleeping postures, putting in contacts, eating, etc. o Body mechanics, ergonomics, joint protection, activity pacing o Orthotics and braces, as needed throughout the lower extremities and lumbar spine, to provide stable base for cervical spine o Importance of shoe-wear support for spinal alignment <p>Pain science and pain self care</p> <ul style="list-style-type: none"> o Relaxation, autonomic nervous system balancing (not requiring neck movement) o Breathing, e.g., diaphragmatic or slow breathing o Pain neuroscience education, addressing catastrophization, mindful use of language to enhance feelings of safety o Self-care "toolbox": e.g., pain management strategies (e.g., heat, ice, transcutaneous electroneural stimulation, topical analgesics, relaxation, positive thinking, etc.) <p>Neck bracing (if appropriate)</p> <ul style="list-style-type: none"> o Education about use of neck brace: how to put on, how often to use, when to use (e.g., during ADLs, flares, car travel) o Custom fitting of rigid or soft cervical brace <p>Manual therapy</p> <p>Some high irritability patients will not tolerate manual therapy, even remote from the neck, and it should be discontinued if not tolerated.</p> <ul style="list-style-type: none"> o Cautious myofascial release, trigger point release or neuromuscular inhibition techniques in the thoracic and lumbar spine, scapulae, lower and upper extremities. o Cautious myofascial release, trigger point release or neuromuscular inhibition in the upper trapezius, levator scapulae, and sternocleidomastoid ONLY by clinicians with S-GJH/UCI expertise <p>Motor control</p> <p>Some High Irritability patients will not tolerate motor control training, even remote from the neck, and this should be discontinued if not tolerated. These should be done with neck, torso and limbs suitably supported, generally in neutral position.</p> <ul style="list-style-type: none"> o Eye movement muscle energy techniques o Pelvic and lumbar stability training; finding pelvic neutral. Ensure that the cervical spine is optimally aligned and supported o Motor control training of the cervical spine, near mid-line o Supine with head supported, scapular recruitment in neutral "safe zone," side lying supported head and arm <p>Aerobic exercise</p> <ul style="list-style-type: none"> • e.g., Recumbent bike, pedal exerciser (if there is no indication of neural tension/tethered cord) <p>Interventions to AVOID with high irritability patients</p> <ul style="list-style-type: none"> • Exercises involving moderate to large neck movements, such as cervical range of motion <ul style="list-style-type: none"> o Some patients will not tolerate any neck movement, even chin tucks o Isometrics with more than minimal force • Cervical axial loading (weight on head) or distraction (manual or mechanical) • Only therapists with S-GJH/UCI expertise should perform any manual therapy to the cervical spine, and some patients may not tolerate any manual therapy, even by experts • Positioning that creates neural tension (e.g., pelvic tilt in some people) or isometric load (e.g., quadruped) to the cervical spine

TABLE 12 Interventions for patients with moderate irritability.

<ul style="list-style-type: none"> • All interventions discussed in Table 11 <p>Education</p> <ul style="list-style-type: none"> • Functional training, as described in Table 11 plus: Meal preparation, positional training for ADL and IADLs, standing, pivoting, stand pivoting, squatting, half-kneeling, pushing/pulling light objects, rotational upright core training, sweeping, shopping, light housework, carrying, driving, and lifting. <p>Motor control and strength training</p> <ul style="list-style-type: none"> • Proprioception, motor control, and strengthening exercises for: <ul style="list-style-type: none"> o Lower extremities, including knee, foot, ankle o Shoulders and scapulae o Thoracic spine o Continue and progress for pelvis and lumbar spine • Proprioception, motor control, and stabilization training for the cervical spine through available pain-free range. This may include using the head laser, starting by maintaining the head stable while moving the arms or legs, walking, and gradually progressing to small, controlled neck movements • Gentle axial loading of the cervical spine (e.g., up to 1 pound/450 grams) if tolerated • Low load cervical isometrics, with cuing to deactivate superficial muscles <p>Manual therapy:</p> <ul style="list-style-type: none"> • Manual therapy for 1st rib, thoracic spine, acromioclavicular and sternoclavicular joints • Soft tissue techniques for cervical muscles in spasm, physiological quieting • Gentle manual techniques for C1 and C2 (if the therapist is trained) • AVOID aggressive soft tissue or joint-based manual therapy to the cervical spine <p>Aerobic exercise:</p> <ul style="list-style-type: none"> • E.g., Recumbent bike/peddler (if no neural tension signs); walking

TABLE 13 Interventions for patients with low irritability.

<ul style="list-style-type: none"> • All interventions discussed in Tables 11, 12 <p>Education</p> <ul style="list-style-type: none"> • Functional training, as described in Tables 11, 12, plus: Occupation related functional training, i.e., prolonged desk work, phone, heavier household chores, gardening, etc. Sports specific training with precautions such as avoiding contact sports such as football or modification to sports such as no "heading" the ball <p>Manual therapy:</p> <ul style="list-style-type: none"> • Additional muscle energy techniques in the cervical spine <p>Motor control and strength training</p> <ul style="list-style-type: none"> • Proprioception and motor control using larger cervical ranges. Cervical axial loading may decrease symptoms during proprioceptive training • Trunk-head coordination, eye-head coordination, eye-balance exercises • Resistance training for the cervical spine • Return to function/sport exercises, if appropriate, which may include more aggressive exercise, if tolerated. These may include perturbation, unpredictable challenges, and more endurance exercise for the neck <p>Aerobic exercise:</p> <ul style="list-style-type: none"> • E.g., Walking, recumbent or upright bike. Some patients may tolerate running, swimming, aerobics with or without precautions
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