Neurosurgical Review (2026) 49:270
https://doi.org/10.1007/s10143-026-04195-z

METHODOLOGY

Surgical screening protocol for craniocervical instability secondary to ehlers-danlos syndrome and other connective tissue disorders: analysis of a 347 patient case series

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Received: 22 September 2025 / Revised: 10 January 2026 / Accepted: 20 February 2026
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Abstract

Background Evaluation of craniocervical instability (CCI) in individuals with connective tissue disorders (CTDs) remains controversial. We present a surgical screening protocol created and implemented at our institution for these individuals.

Methods We reviewed patients referred to our institution for CCI secondary to CTDs, between May 2018 and April 2022 using our two-stage protocol. Stage 1 included: history, clinical questionnaire, physical examination, non-invasive provocative testing, neuroimaging with morphometric analysis, Karnofsky Performance Scale. Items 1–5 were each scored on a 0 to 2 scale.

Individuals with a KPS ≤ 70 and an aggregate score ≥ 6 were recommended for Stage 2, which included additional neuroimaging, psychiatric evaluation, and a trial of intraoperative craniocervical traction (ICT) with clinical and morphometric scores. Individuals meeting criteria in both stages were considered surgical candidates. Postoperative outcomes were assessed.

Results Of the 347 individuals entering Stage 1, 190 progressed through Stage 2. Following advanced evaluation, 115 patients met full surgical qualification criteria, with 95 proceeding to craniocervical fusion (CCF) and reporting an 86.4% satisfaction rate on the North American Spine Society (NASS) satisfaction index. Twelve additional patients with borderline morphometric scores were offered surgery due to marked clinical improvement during ICT; however, this Subgroup demonstrated a lower NASS satisfaction rate (70%).

Conclusions We present a two-stage surgical evaluation protocol for CCI in patients with CTDs. ICT served as a critical diagnostic tool, clarifying the biomechanical contribution of CCI to symptomatology and aiding in the identification of appropriate surgical candidates. Further multicenter studies are warranted for validation.

Keywords Craniocervical Instability · Craniocervical Fusion · Intraoperative Craniocervical Traction · Connective Tissue Disorders · Ehlers-Danlos Syndrome

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Published online: 12 March 2026

Abbreviations

BAI	Basion Axial Interval
BDI	Basion Dens Interval
CXA	Clivo Axial Angle
CCF	Craniocervical Fusion
CCI	Craniocervical Instability
CCJ	Craniocervical Junction
CTA	Computed Tomography Angiography
CTD	Connective Tissue Disorders
EDS	Ehlers-Danlos Syndrome
ICT	Intraoperative Craniocervical Traction
KPS	Karnofsky Performance Scale
pB–C2	Perpendicular to the Basion to C2 Line (Grabb-Oakes measurement)
NASS	North American Spine Surgery (Satisfaction Index)

Introduction

The functional mobility of the craniocervical junction (CCJ) depends on its ligamentous structures [1]. Craniocervical Instability (CCI) may be congenital or acquired secondary to various pathologies, including trauma, neoplasms (e.g., chordoma), inflammatory diseases (e.g., rheumatoid arthritis, systemic lupus erythematosus), infection (e.g., Grisel's syndrome), and connective tissue disorders (CTDs) [2–6].

CCI in individuals with CTDs presents unique diagnostic and surgical challenges [5, 7–9], often manifesting with debilitating symptoms that appear disproportionate to findings on conventional neuroimaging and neurological examination—unlike other etiologies such as rheumatoid arthritis [10, 11].

A range of non-invasive diagnostic tools have been proposed to address these challenges, including morphometric analysis, dynamic imaging, trials of cervical collars or traction, cervicothoracic orthoses, and halo jackets [9, 12]. However, existing approaches described in the literature exhibit limitations related to standardization, reproducibility, and correlation with symptom reversibility, particularly in assessing vertical instability and cranial settling in individuals with connective tissue disorders. In response, we developed and evaluated a structured protocol designed to support clinical decision-making and guide management in this complex population.

Materials & methods

Patient selection and study design

From May 2018 to April 2022, we evaluated individuals with a genetic and/or clinical diagnosis of CTDs who were referred to our institution for assessment and management of suspected CCI [13]. All patients underwent MRI of the brain, cervical spine, and craniocervical junction as part of the initial evaluation. Individuals with radiologically obvious CCI pathologies or other structural craniocervical junction abnormalities on imaging were excluded from this study. Evaluation for CCI and potential surgical intervention was conducted using a two-stage protocol, as outlined below. This study is reported in accordance with the STROBE guidelines [14] and adheres to applicable elements of the PROCESS 2023 recommendations for surgical case series reporting [15].

Stage 1 (Initial screening)

Stage 1 (Initial Screening) consisted of six components: (1) a detailed history focused on features suggestive of CCI [12, 16]; (2) a standardized questionnaire assessing signs and symptoms of CCI and comorbidities commonly associated with CTDs [9, 16] (Supplementary Material 1); (3) a physical examination; (4) predefined, noninvasive provocative testing maneuvers designed to elicit or accentuate symptoms relevant to craniocervical instability, performed uniformly in all patients and graded using a standardized ordinal scoring system (0–2); (5) initial neuroanatomic imaging of the CCJ with morphometric analysis based on previously validated criteria in this population [8, 17–19]; and (6) assessment of functional status using the Karnofsky Performance Scale (KPS) [20]. With the exception of the KPS, each component was scored on a 0–2 ordinal scale, where 0 indicated no suspicion for CCI, 1 indicated intermediate suspicion (incomplete or partial findings), and 2 indicated strong suspicion, as summarized in Table 1.

Patients with a KPS score ≤ 70 (i.e., able to care for self but unable to carry on normal activity or perform active work, attributable to their chief complaints) and an aggregate screening score ≥ 6 were recommended to proceed to Stage 2 (Advanced Evaluation). Patients with an aggregate score < 6 and/or a KPS > 70 were advised to pursue conservative management (Fig. 1a).

Stage 1 Provocative testing

As part of Stage 1 screening, all patients underwent a standardized set of predefined provocative maneuvers intended to reproduce or accentuate symptoms associated with abnormal craniocervical biomechanics. These maneuvers were performed uniformly in all patients, independent of imaging findings. Provocative testing was scored on a 0–2 ordinal scale, with (0) assigned if 0–1 maneuvers were positive, (1) if 2–3 maneuvers were positive, and (2) if 4–6 maneuvers were positive.

Provocative testing scores were incorporated into the aggregate Stage 1 screening score and interpreted within the

Table 1 Stage 1 screening components and composite scoring framework (0–2 Scale)

Domain	Score = 0	Score = 1	Score = 2
History: features suggestive of craniocervical instability	No historical features suggestive of CCI *(e.g., no history of head or neck trauma, no prior head or neck surgery, no connective tissue disorder, no cervical manipulation, no prolonged diagnostic course)*	Limited or non-specific historical features suggestive of CCI *(e.g., single relevant historical feature such as prior head/neck trauma or connective tissue disorder, without a consistent pattern)*	Multiple characteristic historical features strongly suggestive of CCI *(e.g., combination of head or neck trauma, prior head or neck surgery, connective tissue disorder, history of cervical manipulation, and/or prolonged diagnostic odyssey with escalating care)*
Standardized questionnaire assessing symptoms relevant to CCI	Minimal or absent symptom burden *(e.g., isolated, intermittent, or non-specific symptoms)*	Moderate symptom burden *(e.g., multiple symptoms without clear localization)*	High symptom burden *(e.g., motion-aggravated headache, globus sensation, dysphagia for liquids, double vision, or other neurologic, autonomic, bulbar, vestibular, pain, or non-specific symptom clusters)*
Physical examination	Normal or non-localizing examination *(e.g., normal strength, coordination, gait, and bulbar function)*	Subtle, intermittent, or non-reproducible abnormalities *(e.g., equivocal weakness, mild imbalance, inconsistent bulbar findings)*	Clear focal or reproducible signs consistent with suspected pathology *(e.g., focal motor weakness, dyspnea attributable to neurologic dysfunction, ataxia, lower extremity spasticity, dysphagia for liquids)*
Provocative testing (Stage 1) *(six standardized, self-administered maneuvers performed under clinician instruction and observation; see Table Y for detailed description)*	0–1 provocative maneuvers positive	2–3 provocative maneuvers positive	4–6 provocative maneuvers positive
Imaging suspicion	No morphometric abnormality or incidental findings only *(e.g., basion–dens interval within normal range, no retroodontoid pannus, normal clivo-axial angle)*	Borderline or equivocal morphometric findings *(e.g., BDI at lower limit of normal, mild retroodontoid soft tissue, or borderline CXA)*	Definite morphometric abnormalities plausibly explaining symptoms *(e.g., BDI below normal reference range, thick retroodontoid pannus, and/or reduced clivo-axial angle)*
Karnofsky Performance Status (screening threshold)	KPS ≥ 70 (passes screening threshold)	N/A	KPS < 70 (fails threshold) exclusionary, not scored

Stage 1 screening incorporated clinical history, a structured intake questionnaire, physical examination, provocative testing, imaging review, and Karnofsky Performance Status (KPS). Stage 1 domains (except KPS) were scored using a 0–2 ordinal scale reflecting absent, incomplete/partial, or strong findings. KPS was applied as a binary screening threshold. Detailed descriptions of the standardized provocative testing maneuvers and their scoring are provided in Table 6. The full clinical intake questionnaire is provided in Supplementary Material 1

broader multimodal assessment framework rather than as stand-alone diagnostic criteria. The specific maneuvers and scoring rubric are summarized in Table 2.

Stage 2 (Advanced evaluation)

Stage 2 (Advanced Evaluation) involved a comprehensive diagnostic workup to evaluate surgical candidacy (Fig. 1b).

This flowchart illustrates the Advanced Evaluation phase of our protocol, detailing the clinical components assessed and the specific selection criteria for surgical qualification.

Values indicate the number of subjects meeting selection criteria at each stage of the process.

The bottom section of the flowchart provides the distribution of postoperative outcomes, based on the North American Spine Society (NASS) score.

This process included CT angiography of the CCJ, medical and cardiac clearances, and double psychiatric evaluation and clearance as described below.

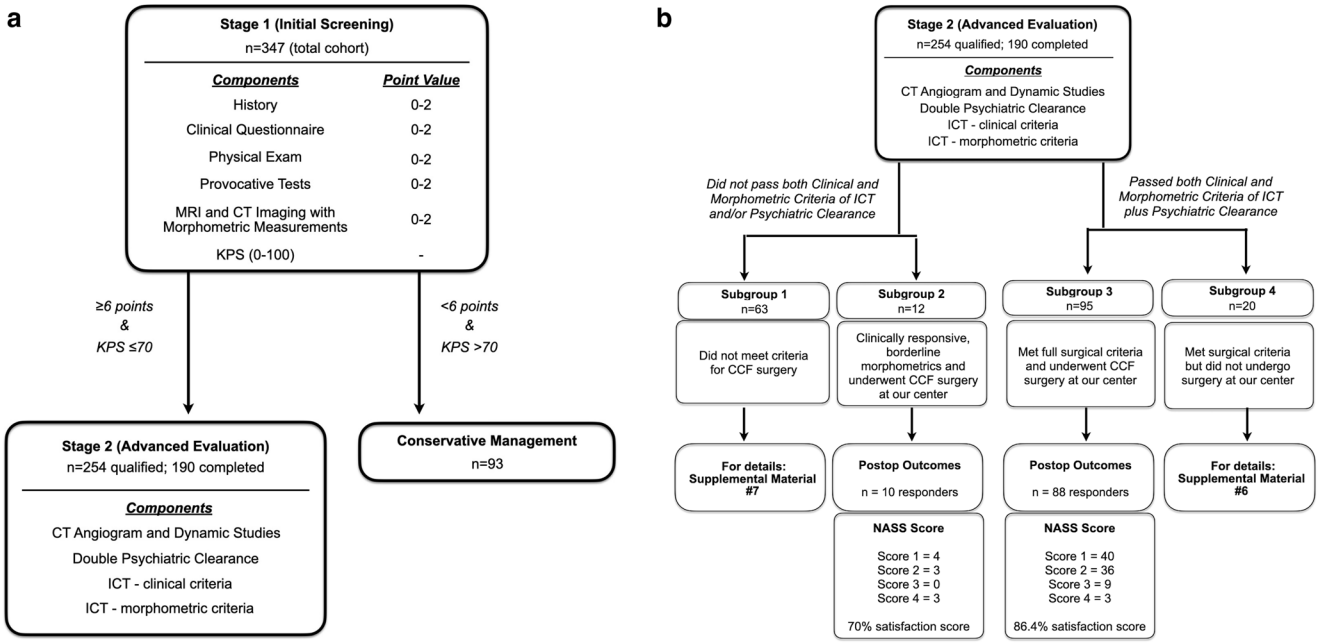


Fig. 1 **a** Stage 1 (Initial Screening): components, scoring criteria, and patient attrition. This flowchart illustrates the Initial Screening phase of our diagnostic protocol. It details the specific clinical and radiological components assessed, the standardized scoring patterns applied to each diagnostic item, the selection criteria, and the numeric breakdown of the patient cohort. Values indicate the number of subjects meeting the inclusion thresholds at each step of the preliminary evaluation. **b** Stage 2 (Advanced Evaluation): diagnostic components, surgical selection, and postoperative outcomes

Following completion of the above screening steps, candidates underwent a trial of ICT, which was evaluated using a defined scoring system (outlined below). As part of the ICT protocol, fluoroscopic imaging was performed intraoperatively in the seated position prior to the application of traction forces to assess cervical alignment under flexion–extension conditions in conjunction with contemporaneous clinical assessment.

Individuals who met criteria in both Stage 1 and Stage 2 and demonstrated positive scores on both components of the ICT assessment were deemed surgical candidates for CCF.

Diagnostic intake tool

As part of Stage 1 screening, individuals completed a diagnostic intake tool developed internally for patients with CTDs (Supplementary Material 1).

Psychiatric clearance

All individuals underwent psychiatric evaluation by two independent trained professionals.

—an outside mental health specialist local to the individual and a neuropsychologist at our institution. These evaluations were essential not only to identify established psychiatric comorbidities that can be associated with CTDs—such as anxiety disorders, depression, personality disorders, obsessive–compulsive disorder, and mood disorders—but also to further assess the patient's subjective symptom burden. These evaluations aimed to distinguish between symptoms arising from underlying pathophysiology and those potentially influenced or amplified by psychological factors. Particular attention was given to evaluating how psychological factors may influence the perception of pain and other symptoms, as well as the patient's ability to cope with the postoperative recovery process. Findings from both evaluations were integrated into the multidisciplinary surgical planning to ensure appropriate support and risk stratification.

Additional details can be found in Supplementary Material 2.

Imaging

All imaging studies were reviewed using a commercial electronic platform (*OsiriX;* Pixmeo, Switzerland) [21]. Stage 1 MRI was systematically reviewed for posterior fossa and craniocervical junction anatomy, including assessment for Chiari malformation, which was recorded as a comorbidity rather than a defining diagnostic criterion.

Table 2 Stage 1 provocative testing maneuvers and scoring framework

Domain	Provocative maneuver	Patient instruction	Positive response criterion
Axial loading	Downward axial compression	Sit upright. Place both palms flat on the top of the head and gently push the head straight downward toward the chest without flexion or extension. Pressure is increased gradually as tolerated	Marked worsening of the patient's primary symptoms
Axial unloading	Upward axial distraction	Sit upright. Place both hands beneath the jaw with palms facing upward, fingers contacting the submandibular and mastoid regions. Gently lift the head straight upward toward the ceiling without tilting	Marked improvement of the patient's primary symptoms
Rebound response	Post-distraction rebound	After release of axial distraction, symptoms are reassessed at rest	Strong rebound or exacerbation of symptoms following release
Intensity check (worsening)	Severity confirmation – worsening	If symptoms worsened during testing, patient is asked to characterize intensity	Worsening described as extremely strong or intense
Intensity check (improvement)	Severity confirmation – improvement	If symptoms improved during testing, patient is asked to characterize intensity	Improvement described as extremely strong or intense
Dynamic loading	Vehicular vibration history	Patient reports symptom behavior when riding in a car on a bumpy road	Marked worsening of symptoms with vibration or jolting

Six standardized provocative maneuvers were incorporated into the Stage 1 screening framework. Responses were scored using an ordinal scale based on the number of maneuvers eliciting symptom reproduction or modulation: 0 if 0–1 maneuvers were positive, 1 if 2–3 maneuvers were positive, and 2 if 4–6 maneuvers were positive. Testing was self-administered by the patient under clinician instruction and observation and was intended to assess directional symptom response rather than symptom severity. Provocative testing contributed to Stage 1 screening and did not serve as a standalone diagnostic criterion

ICT protocol

Following informed consent, Gardner-Wells tongs were applied in the operating room, and the patient was placed in a sitting position after a baseline neurological exam. Lateral-view digital fluoroscopic imaging of the CCJ was obtained in neutral, flexion, and extension positions. After baseline documentation of presenting signs and symptoms, the tongs were connected to a rope-and-pulley system, and an initial vertical traction force of 20 pounds was applied. Traction was gradually increased every 10 min as tolerated, up to a maximum of 35 pounds (or 40 pounds in individuals with a strong athletic build). At each increment, patients were asked to report any change—positive, negative, or none—in each of their pre-identified symptoms and/or signs, expressed subjectively as a percentage relative to baseline (e.g., "10% worse," "90% better," or "no change"). Fifteen minutes after reaching the final traction level, follow-up digital fluoroscopic images of the CCJ were obtained, along with a repeat neurological examination. An example of the clinical and morphometric data obtained during ICT is presented (Table 3).

The ICT set-up and testing are illustrated in Fig. 2 and Supplementary Material 3.

Morphometric measurements

Biomechanical assessment of CCI was performed using four established morphometric parameters: the basion-dens interval (BDI), basion-axial interval (BAI), Grabb-Oakes measurement (pB-C2 distance), and the clivo-axial angle (CXA) [8, 17–19]. Technical details are provided in Supplementary Material 4.

Static measurements were obtained from MRI in all patients as part of Stage 1 imaging and compared against established normative values, including reference ranges derived from upright and dynamic craniocervical imaging studies [22]. MRI included evaluation of the brain, cervical spine, and craniocervical junction. Dynamic measurements—acquired both at pre-traction and at peak traction—were obtained during ICT using calibrated digital fluoroscopy (*Iso-C,* Siemens, Germany). Calibration of the fluoroscopic imaging during ICT was performed against a known object placed in the midsagittal plane.

The difference between each morphometric value at peak traction and its corresponding pre-traction measurement was defined as the translational value. These translational changes were evaluated against established normal reference ranges and predefined surgical thresholds (Table 4).

Scoring of the ICT test

The ICT Clinical Score was considered positive if most or all of the patient's chief CCI-related signs and symptoms improved by ≥ 75% at peak traction compared with the pre-traction baseline. The ICT Morphometric Score was considered positive when one or more of the following criteria were met: (1) a ≥ 2 mm increase in the translational BDI, and/or (2) a ≥ 4 mm change in the BAI or pB–C2, observed on flexion/extension images or during traction. Patients with both a positive ICT Clinical Score and a positive ICT Morphometric Score met our criteria for CCF surgery.

CCF Procedure

Occipital condyle fixation was selected instead of a traditional occipital plate construct based on biomechanical, anatomical, and patient-specific considerations relevant to CTDs. This approach allows a shorter construct with a reduced lever arm, minimizing bending moments at the craniovertebral junction while providing rigid occipito-cervical stabilization and reducing subcutaneous hardware burden in patients with skin fragility.

The occipital condyles provide dense cortical bone with more reliable screw purchase than the thin and variable

Table 3 Example of Data Collected During Intraoperative Craniocervical Traction (ICT)

	Improvement (%) with Traction Applied			
Symptoms	*20 lbs*	*30 lbs*	*35 lbs*	*35 lbs (delay)*
Suboccipital Headache	35%	80%	90%	95%
Neck Pain	55%	70%	80%	90%
Weakness, Bilateral Upper Extremity	40%	80%	90%	95%
Numbness, Bilateral Upper and Lower Extremity	40%	85%	90%	95%
Hyperreflexia, Bilateral Lower Extremity	42%	75%	90%	100%
Fatigue	30%	80%	90%	95%
Brain Fog	25%	85%	90%	95%
Photophobia	10%	60%	80%	95%
Phonophobia	15%	60%	80%	95%
Shortness of Breath	40%	75%	90%	100%
Double Vision	30%	80%	90%	95%
Dysphagia	25%	85%	90%	95%
Morphometrics		Pre-traction		Peak-traction
BDI (mm)		2.9		8.5
BAI (mm)		5.2		7.9
pB-C2 (mm)		3.9		8.3
CXA (degrees)		156		155

Illustrative patient case showing percentage improvement in pre-identified signs and symptoms at incremental traction weights, as reported by the patient, alongside changes in neurological examination findings. Corresponding morphometric measurements of the craniocervical junction are shown for both pre-traction and peak-traction states

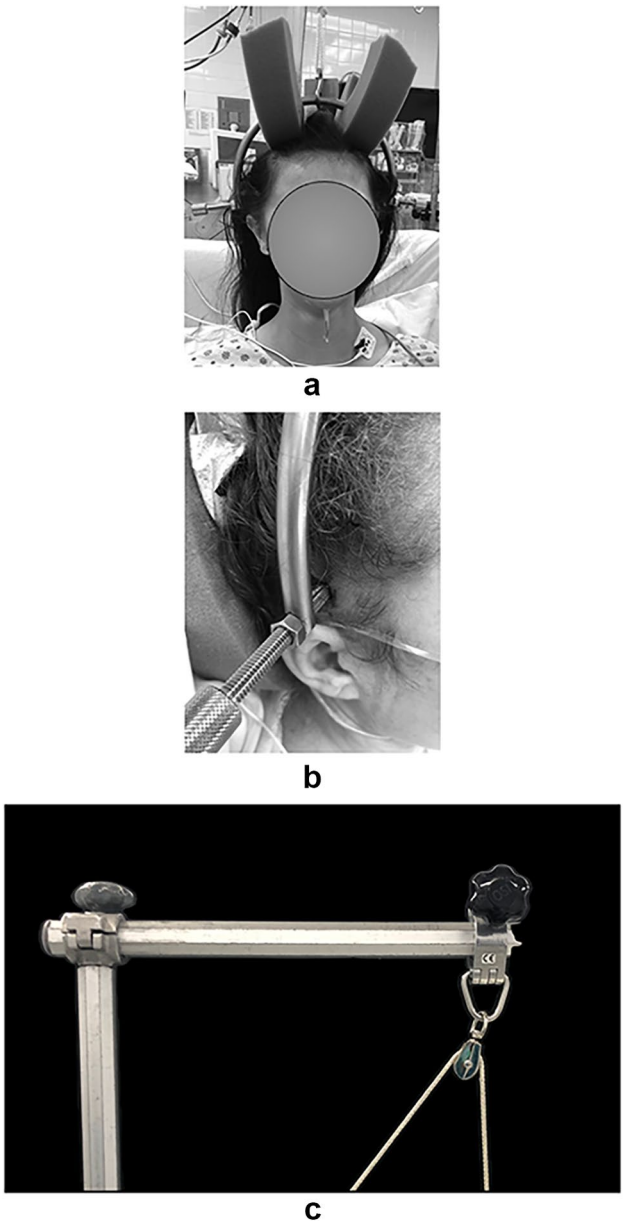


Fig. 2 **a** Intraoperative Cervical Traction (ICT). Frontal view of the ICT setup, showing Gardner-Wells tongs secured with protective padding. **b** Intraoperative Cervical Traction (ICT). Lateral view of the Gardner-Wells tongs showing the cranial pin insertion site. **c** Intraoperative Cervical Traction (ICT). Configuration of the pulley and rope system utilized for vertical traction

squamous occiput. Based on over 20 years of experience performing CCF in CTD patients, we observed frequent hardware-related issues with traditional occipital plate constructs, including prominence, painful hardware, next level disease and metal fatigue, which motivated adoption of condylar fixation in this population. The safety and feasibility of this technique have been demonstrated in a published 250-patient series with acceptable complication rates and durable fixation [23], and prior reports in EDS-associated CCI have specifically compared occipital condyle–based constructs with traditional occipital fixation [24]. While occipital plates remain commonly used, condylar fixation offered specific advantages in our CTD patients and has therefore been routinely employed in our practice.

Postoperative monitoring and outcome questionnaire

Patients were routinely evaluated at 6 and 12 months postoperatively, with additional follow-up conducted on a PRN basis thereafter. A custom-designed postoperative outcome questionnaire was distributed as a one-time, simultaneous mailing in March 2023 to all individuals who had undergone CCF. The complete questionnaire is provided in Supplementary Material 5.

For the purposes of the present analysis, outcomes were limited to patient-reported changes in preoperative signs and symptoms and postoperative satisfaction as measured by the North American Spine Society (NASS) Patient Satisfaction Index [25]. The NASS Patient Satisfaction Index is a validated 4-point instrument; scores of 1 ("treatment met expectations") and 2 ("did not improve as much as hoped but would undergo the same treatment again") were categorized as satisfied outcomes, whereas scores of 3 ("treatment helped but would not undergo the same treatment again") and 4 ("same or worse than before treatment") were categorized as dissatisfied outcomes. Comprehensive evaluation of the full dataset will be presented in a subsequent publication focused on long-term CCF outcomes.

Table 4 Normal values for morphometric measurements

Normal Values for Morphometric Measurements		
Morphometric	*Normal Range*	
CXA	136—165°	
pB-C2 distance	6—8.5 mm	
BAI	0—12 mm	
BDI (Harris)	3.2—11.8 mm	
BDI (Gonzalez)	4.7—9 mm	
Normal Values for Translational Morphometric Measurements		
Morphometric	*Normal Range*	*Surgical threshold*
CXA	N/A	135° or less
pB-C2 distance	1—2 mm	≥4 mm
BAI	1—2 mm	≥4 mm
BDI	1—1.5 mm	≥2 mm

Statistical analysis

We retrospectively collected data from the electronic medical record and performed analyses using Microsoft Excel and GraphPad Prism. Individuals with incomplete medical records were excluded. Morphometric variables measured at pre- and peak-traction were compared using two-tailed *t* tests. Changes in patient-reported signs and symptoms between pre-traction, peak-traction, and post-surgical states were evaluated using McNemar's test, with calculations performed in R (RStudio, Posit PBC). Statistical significance was defined as a p-value < 0.05.

Results

Enrollment in stage 1

From May 2018 to April 2022, 347 individuals with a clinical and/or genetic diagnosis of a CTD were referred to our institution for evaluation of CCI (mean age 37 ± 12 years; 87.3% female). All individuals were enrolled in Stage 1 of the protocol (flowchart; see Fig. 1a).

Chiari I malformation was present as a comorbidity in 33.2% of patients. Ehlers–Danlos syndrome (EDS) was the most common CTD diagnosis (75.3%), although many individuals exhibited mixed CTD phenotypes. Regardless of the specific CTD subtype, CCI emerged as a final common pathway. Across CTD subtypes, patients were evaluated using a uniform screening framework for suspected CCI.

Enrollment in stage 2

Of the individuals who entered Stage 1, 254 met criteria to proceed to Stage 2, and 190 completed the advanced evaluation (Fig. 1b).

Surgical qualification

The 190 individuals who completed Stage 2 were categorized into four subgroups based on whether they met surgical criteria and elected to proceed with CCF surgery. Subgroup 1 (n = 63) did not meet criteria for CCF surgery. Subgroup 2 (n = 12) had borderline morphometric scores on ICT but were offered surgery based on remarkable improvements in signs and/or symptoms during ICT and proceeded with CCF surgery at our center. Subgroup 3 (n = 95) met full surgical criteria and underwent CCF surgery at our center. Subgroup 4 (n = 20) met surgical criteria but did not undergo

surgery at our institution (for reasons which are summarized in Supplementary Material 6).

Stage 2 (Advanced Evaluation) procedures and subgroup allocation are summarized in a flowchart (Fig. 1b).

Subgroup 1 was further analyzed to identify which components of the advanced evaluation led to disqualification from surgery (Supplementary Material 7).

Exceptions to surgical qualification were made for Subgroup 2 based on specific clinical considerations. Although these 12 individuals narrowly missed the ICT morphometric threshold for BDI by fractions of a millimeter, 6 exhibited marked improvement in neurological deficits at peak traction, while the remaining 6 experienced striking relief of their baseline chief complaints during traction.

Signs and symptoms

Table 5 summarizes the most common presenting signs and symptoms reported at Stage 1 enrollment, grouped by neurologic relevance, among patients who advanced to Stage 2 and for the subset who subsequently met surgical eligibility. Patients who advanced to Stage 2 demonstrated a higher prevalence of objective neurological signs and symptoms consistent with cervicomedullary syndrome compared with the overall cohort, with further enrichment among patients who met the strict surgical qualification criteria. In contrast, nonspecific symptoms were prevalent across all groups and did not demonstrate a similar stepwise concentration (Table 5).

Figure 3a illustrates common pre-traction signs and symptoms in individuals who completed Stage 2 (Advanced Evaluation) that demonstrated the greatest improvement during ICT at peak traction, with the relief threshold defined as 75% or greater for each sign or symptom (Fig. 3a).

Each boxplot displays the interquartile range (25th–75th percentile) with the median indicated by a horizontal line; whiskers represent the minimum and maximum values. All comparisons between pre-traction and peak-traction values were statistically significant (p-value < 0.001).

Morphometric analysis

Key morphometric parameters—including basion-dens interval (BDI), basion-axial interval (BAI), pB–C2 distance, and clivo-axial angle (CXA)—were measured both pre-traction and at peak traction. All four morphometric parameters showed statistically significant changes from pre-traction to peak traction during ICT ($p < 0.001$ for all; Fig. 3b).

Complications of the ICT procedure

No patients who underwent ICT experienced direct complications from the application of tongs or weights, such as skull fractures, dislocations, hemorrhages, brain injuries, or cerebrospinal fluid leaks.

Two patients with previously documented severe CCI who qualified for CCF at the conclusion of the Stage 2 screening process, developed intense and persistent rebound symptoms following completion of ICT. After a period of observation, both proceeded to CCF surgery during the same hospitalization, with resolution of rebound symptoms and their chief complaints postoperatively.

Outcomes of CCF

Of the 107 surgical patients (Subgroups 2 and 3), 98 completed the NASS Patient Satisfaction Index via a one-time postoperative questionnaire, administered at a mean of 2.3 ± 1 years after surgery. Satisfaction (NASS score 1 or 2) was reported by 70% of Subgroup 2 and 86.4% of Subgroup 3, indicating that surgery met expectations or provided sufficient benefit to warrant the same procedure again (Fig. 1b).

The same questionnaire also documented changes in preoperative signs and symptoms. At follow-up, a substantial proportion of patients in both Subgroup 2 and Subgroup 3 reported $\geq 75\%$ improvement in many of their preoperative symptoms, with outcomes presented separately for each subgroup (Fig. 4).

Representative postoperative radiographs demonstrating craniocervical fusion using occipital condyle–based fixation are shown in Fig. 5.

Postoperative complications

Postoperative surgical complications following CCF are summarized as follows. Among the 107 surgical patients (12 from Subgroup 2 and 95 from Subgroup 3), complications were uncommon. One patient (0.93%) developed a postoperative infection related to needle aspiration of a seroma performed by an outside provider, which required surgical debridement and hardware revision. One patient (0.93%) developed painful profile due to a side connector to the C1 lateral mass screw causing a C2 amputation neuroma, managed with partial unilateral hardware removal and neuroma excision. One patient (0.93%) developed adjacent-level subaxial instability one year postoperatively, necessitating extension of the fusion construct.

Five patients (4.7%) experienced mast cell activation syndrome (MCAS)–related reactions to a bone matrix substitute, resulting in secondary wound dehiscence; all were managed with removal and reinsertion of the graft material, after which

Table 5 Signs and symptoms

Sign/Symptom	Overall Cohort (n = 347)	Qualified for Stage 2 (n = 254)	Met surgical criteria (n = 115)
Category 1: Objective neurological signs			
Observed visual disturbance/nystagmus	273 (78.7%)	243 (95.7%)	111 (96.5%)
Motor weakness	249 (71.8%)	219 (86.2%)	105 (91.3%)
Dyspnea (when clinically attributable to brainstem dysfunction)	210 (60.5%)	186 (73.2%)	93 80.9%)
Motor coordination dysfunction/ataxia	186 (53.6%)	168 (66.1%)	76 (66.1%)
Lower extremity spasticity	165 (47.6%)	148 (58.3%)	69 (60.0%)
Objective proprioceptive dysfunction	135 (38.9%)	120 (47.2%)	59 (51.3%)
Dysphagia for liquids	115 (33.1%)	101 (39.8%)	51 (44.3%)
Category 2: Symptoms suggestive of cervicomedullary syndrome (CMS)			
Headache	292 (84.1%)	252 (99.2%)	115 (100.0%)
Hypoesthesia/paresthesia	284 (81.8%)	248 (97.6%)	110 (95.7%)
Photophobia	273 (78.7%)	230 (90.6%)	105 (91.3%)
Syncope/Presyncope	260 (74.9%)	227 (89.4%)	105 (91.3%)
Nausea	193 (55.6%)	164 (64.6%)	75 (65.2%)
Globus sensation (lump in throat)	158 (45.5%)	137 (53.9%)	61 (53.0%)
Dysphonia	140 (40.3%)	125 (49.2%)	54 (47.0%)
Category 3: Nonspecific symptoms			
Cognitive complaints ("brain fog")	299 (86.2%)	251 (98.8%)	112 (97.4%)
Palpitations	260 (74.9%)	217 (85.4%)	101 (87.8%)
Muscle spasms	249 (71.8%)	207 (81.5%)	99 (86.1%)
Tremors	167 (48.1%)	145 (57.1%)	70 (60.9%)
Thermosensory dysfunction	107 (30.8%)	87 (34.3%)	39 (33.9%)

The table summarizes the most common presenting signs and symptoms, categorized into three groups: Objective neurological signs (Category 1); Symptoms suggestive of cervicomedullary syndrome (Category 2); and Nonspecific symptoms (Category 3). Data are expressed for three distinct cohorts: the Overall Cohort (n = 347), the subcohort which qualified for Stage 2 (n = 254), and the subcohort which met the full surgical criteria (n = 115; comprising Subgroup 3 and 4). The data are expressed as the number of patients, and corresponding percentage within each specific cohort

use of the substitute was discontinued. Two patients (1.87%) developed postoperative seromas at 5–6 weeks, one of which was associated with medication-related side effects and were treated with seroma evacuation and medication adjustment. One patient (0.93%) experienced primary wound dehiscence requiring hospital readmission within 30 days with matrix removal and reinsertion. No cases of implant failure or pseudoarthrosis were observed in this cohort.

Postoperative surgical complications following CCF are summarized in Table 6.

Surgical non-responders

Among the 107 surgically treated patients (from Subgroups 2 and 3), six individuals (5.6%) reported no improvement or a worsening of symptoms at follow-up, corresponding to a NASS Patient Satisfaction Index score of 4. Three of these patients were from Subgroup 2 and three were from Subgroup 3.

Retrospective data analysis showed that these six non-responders (NASS score = 4) had the minimum qualifying score required to proceed from Stage 1 to Stage 2. In contrast, a representative sample of surgical responders with favorable outcomes (NASS scores 1–2) demonstrated higher aggregate Stage 1 clinical scores (8 points).

In retrospect, the clinical exam component of the Stage 1 score in two of the six non-responders was probably elevated due to co-existing comorbidities (specifically a syringomyelia cavity in one patient and an occult tethered cord in another) rather than as a product of craniocervical instability (CCI) itself.

Baseline functional severity did not distinguish responders from non-responders, as the mean entry KPS was 60–65 in both groups. Importantly, all non-responders exhibited objective neurological signs consistent with craniocervical instability (including findings such as nystagmus, motor weakness, and dyspnea), indicating that surgical candidacy was not driven by nonspecific symptoms alone. However, the six non-responders (NASS score = 4) demonstrated a relatively higher burden of nonspecific symptoms, as defined by Table 5.

With respect to Stage 2 evaluation, all three non-responders in Subgroup 3 met morphometric criteria during ICT, demonstrating translational changes that exceeded predefined thresholds. In the three patients from Subgroup 2, the ICT morphometric score was a fraction of a mm short of the threshold, and surgical qualification was driven primarily by marked clinical improvement.

Discussion

We developed and implemented a two-stage surgical screening and evaluation protocol for individuals with suspected CCI secondary to CTDs. This protocol evolved over years

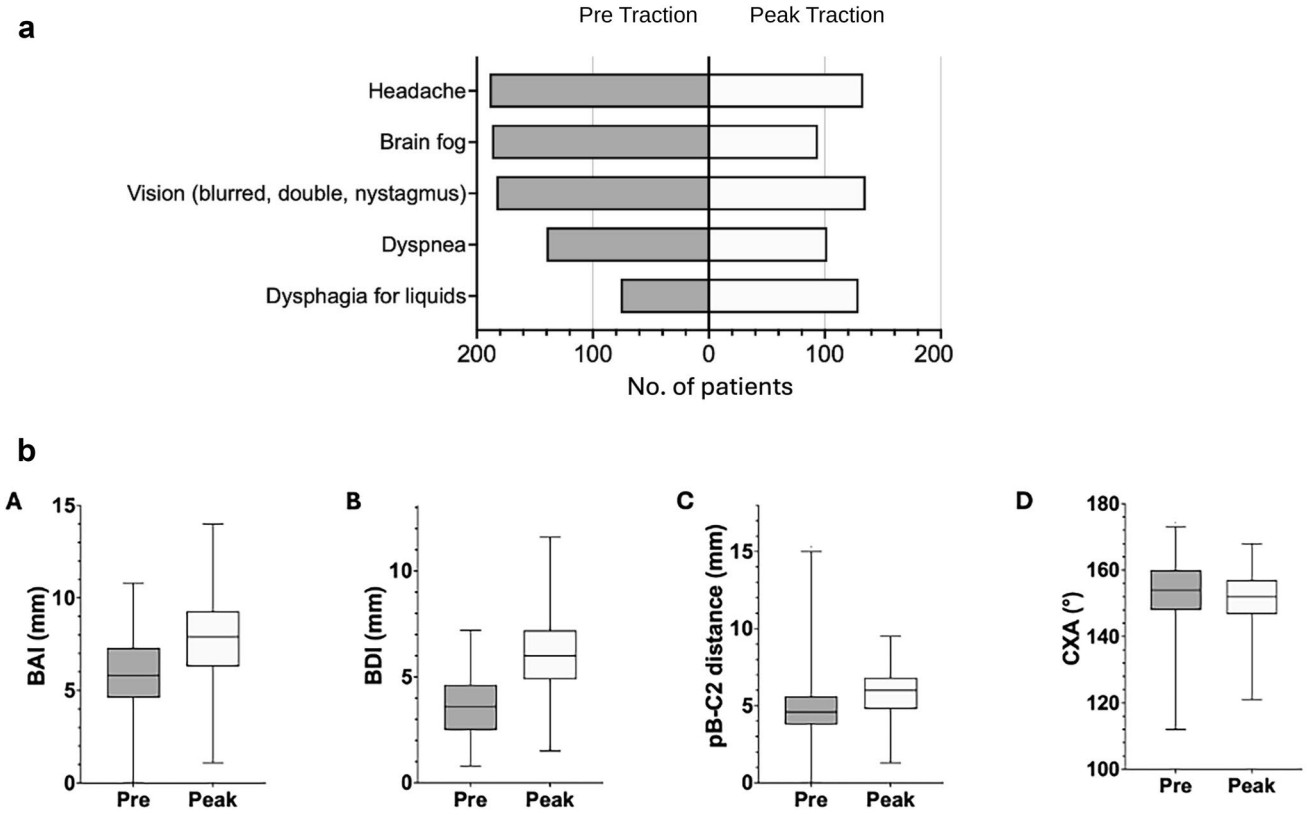


Fig. 3 **a** Mirrored histogram of the most common signs and symptoms responsive to Intraoperative Craniocervical Traction (ICT) at Peak-Traction, in (Stage 2, Advanced Evaluation, n = 190). Left panel: Number of patients reporting each sign or symptom at the pre-traction baseline**.** Right panel: Number of patients experiencing ≥ 75% improvement at peak traction, for each sign/symptom. **b** Morphometric changes observed during ICT. Comparisons are shown between pre-traction (Pre) and peak-traction (Peak) values for each morphometric parameter obtained during ICT (n = 190): (A) Basion–Axial Interval (BAI), measured in mm; (B**)** Basion–Dens Interval (BDI), measured in mm; (C) Grabb–Oakes measurement (pB–C2 distance), measured in mm; (D) Clivo–Axial Angle (CXA), measured in degrees

of clinical experience with this complex patient population and was designed to integrate multiple layers of evaluation—beginning with initial screening and followed by advanced assessment.

Our goal was to create a stringent, multidimensional framework that improves upon prior screening methods by synthesizing previously disparate clinical, functional, physiologic and radiographic data into a cohesive profile to guide decision-making. No single component of the protocol was considered sufficient in isolation. Rather, the strength of the approach lies in the integration of complementary assessments. Within this framework, ICT offered particularly valuable diagnostic and prognostic insight. ICT served as a dynamic adjunct to the broader assessment, revealing functional biomechanical instability that may be overlooked by static imaging modalities and routine clinical examination.

Controversies of CCI in patients with CTDs

In our experience, individuals with CTDs may present across a wide clinical spectrum, ranging from high-performance athletes to individuals who are severely debilitated or bedridden [12, 26, 27]. Paradoxically, some asymptomatic individuals may exhibit marked hypermobility of the CCJ, while others with profound neurological compromise may show minimal or no abnormalities on standard neuroanatomic imaging. While CCI is increasingly recognized as a potential cause of neurological dysfunction in individuals with CTDs, variability in clinical presentation, perplexing imaging findings, and uncertainty surrounding optimal management continue to pose challenges. Additionally, several of these individuals may present with Complex Chiari I malformation [7, 12, 28, 29], further complicating diagnosis and treatment. This heterogeneity has contributed to

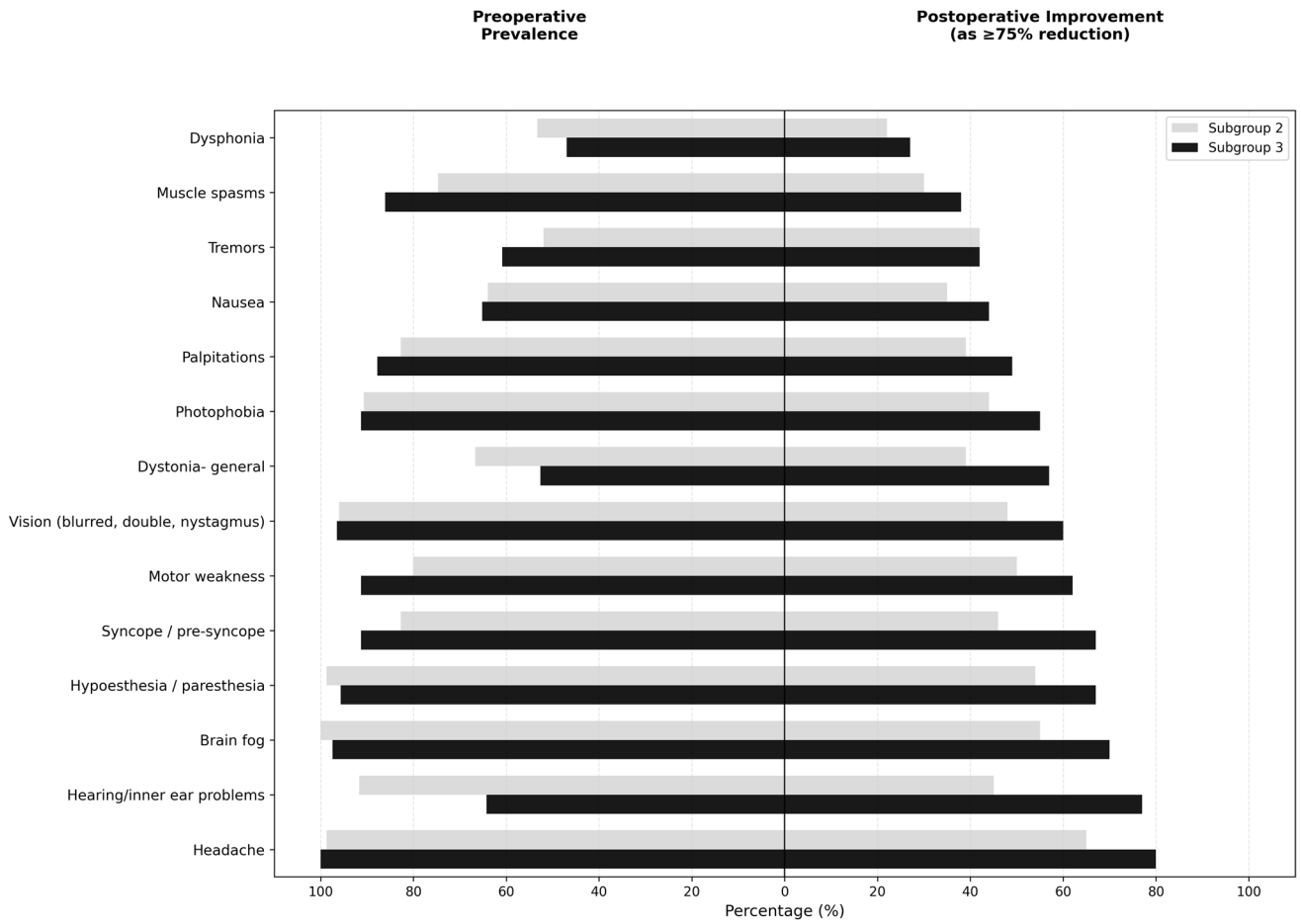


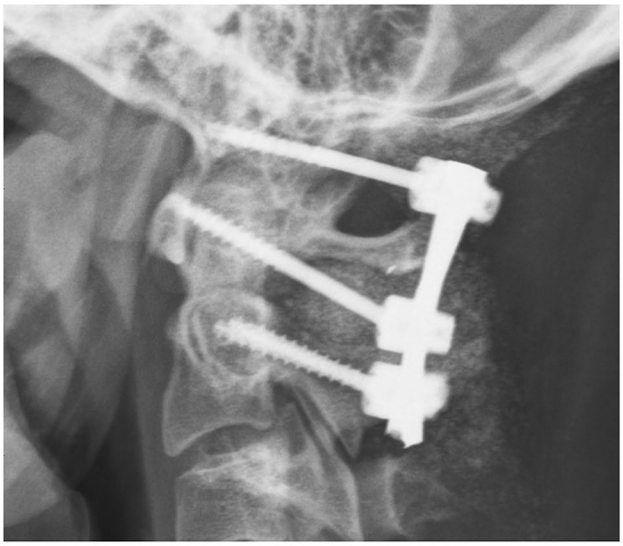
Fig. 4 Preoperative prevalence and postoperative improvement of most common signs and symptoms, stratified by surgical subgroup (Subgroup 2 and Subgroup 3), after Craniocervical Fusion (CCF). 107 patients underwent surgery; 98 completed the postoperative survey. The left side of the histogram indicates the percentage of individuals in each subgroup reporting a given sign or symptom preoperatively The right side of the histogram represents the percentage of those same individuals who reported a ≥ 75% reduction in the corresponding sign or symptom at postoperative follow-up. Light gray bars correspond to Subgroup 2. Dark bars represent Subgroup 3. Data were collected via a structured, symptom-based outcome questionnaire at a mean of 2.3 years after surgery (SD 1.0 year)

ongoing controversy regarding when CCJ hypermobility should be considered pathological and when surgical intervention is warranted.

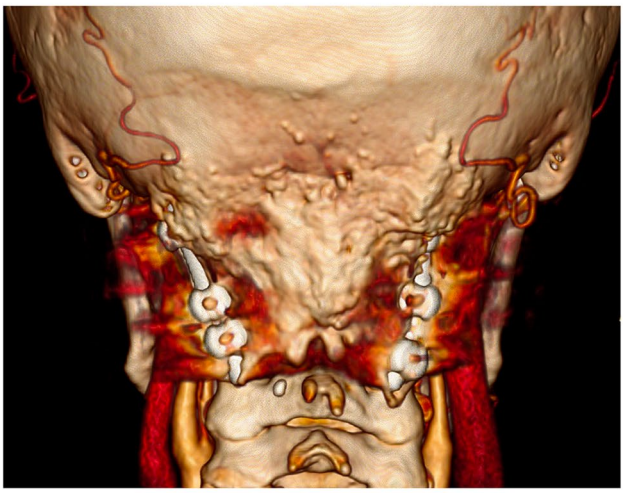
Consistent with the classification proposed by Goel and colleagues, most patients in our cohort correspond to Goel's Type III atlantoaxial instability. This category is characterized by clinically meaningful instability that lacks radiographic evidence on static or dynamic imaging in flexion/extension, yet exhibits pathological biomechanics during intraoperative manipulation [30, 31]. In CTD populations, intrinsic ligamentous laxity further complicates the interpretation of dynamic studies, such that hypermobility—when assessed in isolation—should not be equated with pathological instability. Prior work by Goel and colleagues has been seminal in emphasizing the role of axial atlantoaxial instability and C1-C2 facet alignment in neurological dysfunction at the craniovertebral junction [30, 31]. In our hypermobile CTD cohort, however, classical patterns of fixed facetal malalignment were uncommon, highlighting important phenotypic differences in the biomechanical mechanisms driving craniocervical instability.

In a recent review article, Lohkamp et al. recommended that surgical intervention for suspected CCI in individuals with CTDs should be considered only when both clear radiographic evidence of instability and concordant clinical symptoms are present [32]. It was also highlighted that a 4–6 week trial of hard collar cervical immobilization may represent a valuable adjunctive strategy for surgical selection in this population.

Consistent with this approach, patients presenting with isolated, nonspecific symptoms in the absence of objective neurological findings or concordant evidence of biomechanical instability were not advanced to surgical intervention in our protocol. Importantly, the stratified symptom analysis



a



b

Fig. 5 **a** Postoperative radiological results. Lateral radiograph of one of our CCF constructs, featuring condylar screws, C1 lateral mass screws, and C2 pedicle screws. **b** Postoperative radiological results. Three-dimensional CT reconstruction illustrating the postoperative anatomy at the 1-year follow-up

presented in Table 5 empirically supports this screening approach. Progression from Stage 1 to Stage 2, and ultimately to surgical eligibility, was associated with increasing concentration of objective neurological signs and anatomically localizing features, whereas nonspecific symptoms remained prevalent across all groups and did not uniquely distinguish surgical candidates. This stepwise enrichment pattern suggests that escalation within the diagnostic pathway was driven by neurologically meaningful findings rather than symptom burden alone, reinforcing the construct validity of the staged screening framework.

Table 6 Surgical Complications and Management from the 107-patient surgical series

Complication	n (%)	Description	Management
Infection	1 (0.93%)	Iatrogenic infection following needle aspiration of a small seroma by an outside provider	Debridement and hardware revision with debridement
Painful Profile	1 (0.93%)	A side connector to the C1 lateral mass screw caused a C2 amputation neuroma	Partial (one-sided) hardware removal (C1 lateral mass screw and connector). Excision of the C2 neuroma
Next Level Disease	1 (0.93%)	Sub-axial instability (1 year later)	Extension of the fusion construct to T3
MCAS Reaction	5 (4.7%)	Mast cell activation syndrome reaction to a bone matrix substitute (Stryker) with secondary wound dehiscence	Bone matrix removal and reinsertion. The bone matrix substitute was abandoned, and the old bone matrix mix re-adopted
Wound Dehiscence with 30-day readmission	1 (0.93%)	Primary wound breakdown with readmission within 30 days	Matrix removal and reinsertion
Seroma	2 (1.87%)	Postoperative seroma formation at 5–6 weeks; linked to the side effects to a medication in one case	Seroma evacuation. The responsible medication was removed in one case
Implant Failure	0 (0.0%)	N/A	N/A
Pseudoarthrosis	0 (0.0%)	N/A	N/A

107 total patients. 12 patients from subgroup 2. 95 patients from subgroup 3

Critical analysis of our longitudinal experience with dynamic imaging modalities and immobilization trials

In the process of seeking a reliable surgical screening protocol for this challenging population, we evaluated various diagnostic tools, starting back in 1998. This longitudinal experience allowed for a critical appraisal of these modalities, revealing significant advantages alongside intrinsic flaws that negatively affected their diagnostic reliability in CTD patients.

When paired with conventional supine MRI, upright MRI obtained in the neutral position proved useful for visualizing position-dependent shifts within the craniocervical junction (CCJ) and adjacent central nervous system structures. However, the relatively low magnetic field strength of

most upright MRI systems (typically 0.6 T or less) resulted in suboptimal spatial and contrast resolution, limiting image definition and precluding precise and reproducible morphometric measurements. These technical constraints significantly reduced confidence in quantitative assessments derived from these studies.

Additional reliability concerns emerged when flexion–extension maneuvers were incorporated into neuroimaging protocols (MRI, CT, and plain radiography). The effective fulcrum of flexion is highly variable and may occur in the lower cervical spine, mid-cervical region, or at the CCJ itself. Among these, the so-called "military position" most effectively stresses the CCJ; however, achieving this posture consistently for the prolonged duration required during CT or MRI acquisition is a difficult task. The experience and proficiency of technicians with dynamic spine imaging protocols varied widely, with the occasional cervical collar inadvertently left in place during the study. Flexion–extension studies in upright MRI are particularly prone to motion artifacts, whereas supine MRI or CT studies rely on positioning aids such as sandbags, the size, placement, and consistency of which introduce additional elements of variability to the process, thus impacting reliability.

CTD patients are hypermobile by definition, raising the issue of whether CCJ morphometrics should be interpreted using CTD-specific normative ranges, analogous to the disease-specific criteria established for atlanto-axial interval measurements in patients with Down syndrome. Many asymptomatic CTD individuals can achieve extreme ranges of CCJ motion without triggering any symptoms at all. Consequently, excessive reliance on flexion–extension imaging in these cases would predictably generate false positives in asymptomatic individuals who are merely hypermobile. Conversely, patients with severe, symptomatic craniocervical instability (CCI) who often avoid wide movements and provocative maneuvers because of the risk of prolonged and debilitating symptom flares, would be at risk of being considered false negatives, if dynamic imaging is regarded as the critical key to both diagnosis and surgical qualification.

Digital motion X-ray (DMX), or dynamic digital fluoroscopy, whether single- or biplanar, can be useful in demonstrating overt dynamic subluxations, particularly in Goel Type I and Type II instability. Since the design of this study excluded any radiologically self-evident CCJ pathology, our patient population consists predominantly of Goel Type III instability, which appeared as a false negative on DMX.

Rotational CT with three-dimensional reconstructions raised similar concerns. Conventional normal reference values for rotation were frequently exceeded by CTD patients, often without any associated clinical deterioration. In our experience, isolated rotational instability has been encountered almost exclusively in cases of Bow Hunter syndrome, which generally follows a relatively straightforward diagnostic and surgical decision-making pathway. In all other scenarios, rotational CCJ instability in CTD patients occurred within a broader pattern of biomechanical compromise, in which horizontal and/or vertical vectors played a more dominant role. Accordingly, rotational CT did not prove useful as a foundational diagnostic test for our purpose.

We previously employed 2–4-week cervical collar immobilization trials but abandoned this approach due to multiple confounding factors. Collar fitting was inconsistent in patients with micrognathia or variant neck morphology. Patient compliance was often poor secondary to temporomandibular joint pathology, mast cell-mediated dermatological reactions, and sensory integration challenges. In many cases, prolonged collar use induced severe cervical muscle deconditioning, later leading to worsening the underlying CCI.

Non-invasive cervical traction is a provocative test conceptually similar to ICT. However, micrognathia, temporomandibular joint pathology, inconsistent device fit, limited control of traction vectors, and the absence of feasible standardization progressively restricted its role in our protocol. Ultimately, non-invasive traction was relegated to an adjunctive screening tool during Stage 1 evaluation, reserved for cases requiring additional confirmation before advancement to Stage 2.

We also explored the use of halo vests approximately two decades ago. Prolonged immobilization often resulted in more severe deconditioning than observed after cervical collar trials. In addition, CTD-related immunologic and inflammatory vulnerabilities predisposed patients to frequent pin-site infections, while osteopenia commonly led to pin loosening and the need for repeated pin rotations. Poor soft-tissue healing occasionally resulted in cosmetically unfavorable frontal scarring. Finally, halo fixation offered limited flexibility in reaching positions associated with maximal symptomatic improvement, particularly when compared with traction-based techniques.

In contrast, ICT became our tool of choice for Stage 2, permitting graded, reproducible, and standardized axial traction under direct clinical observation, in a controlled environment. ICT provided complete control over application points, magnitude, and direction of the vectors in action, combined with calibrated imaging and systematic morphometric measurement. Patients consistently reported that clinical improvement at peak ICT traction exceeded—often by orders of magnitude—any benefit observed during non-invasive cervical traction, collar immobilization, or halo vest trials. This observation suggested that ICT more accurately reproduced the biomechanical correction

ultimately achievable by craniocervical fusion. In addition, ICT gave us a valuable insight into the dynamics of the vertical component of CCI, which appears to be an anatomically frequent and clinically relevant feature in this patient population.

The Role of ICT

In 2004, we incorporated the ICT technique, commonly used in trauma settings, into our surgical screening protocol and paired it with morphometric analysis. This adaptation combined the provocative diagnostic utility of ICT with objective, real-time measurements, allowing us to correlate morphometric changes with both patient-reported symptomatic improvements and neurologic examination findings. This approach enabled us to distinguish non-pathologic CCJ hypermobility (i.e., hypermobility without symptom provocation or relief during controlled traction) from clinically significant, pathologic CCI. It also provided objective criteria to guide surgical qualification. Importantly, ICT testing was used to correlate positional alignment changes with symptom response under controlled conditions, rather than as a stand-alone diagnostic test for craniocervical instability. Morphometric data obtained at peak traction further informed intraoperative positioning during CCF, improving surgical precision (Fig. 5). Additionally, this protocol enhanced our understanding of vertical instability—i.e., cranial settling—as a critical component of CCI in this patient population. In this cohort, craniocervical fusion was performed using occipital condyle–based fixation, informed by prior work demonstrating the safety, durability, and biomechanical advantages of this technique in a large CTD population, including a previously reported series of 250 patients [23].

Biomechanics of the CCJ and cranial settling

Cranial settling has been described in pathologies which disrupt the anatomic integrity of the bony elements of the CCJ, such as neoplasms, trauma, rheumatoid arthritis, and infections [5, 33]. Historically, CTDs have not been considered causes of cranial settling. However, our findings using ICT suggest that ligamentous compromise of the CCJ in CTDs—typically in the absence of overt bony pathology—can indeed result in vertical instability or cranial settling. Importantly, surgical correction of this instability was associated with improvement and/or resolution of related neurological signs and symptoms in a majority of treated patients. These observations further suggest that dynamic cranial settling, with consequent brainstem and/or neurovascular compression, may contribute to the frequent discordance observed between static neuroanatomic imaging findings and the severity of clinical presentation in this patient population.

Consistent with prior literature demonstrating poor correlation between static craniocervical morphometrics and symptom severity in connective tissue disorder populations, surgical decision-making in this cohort was based on concordance between dynamic morphometric change during ICT and real-time symptom response, rather than static imaging abnormalities alone [12, 16].

Interpretation of surgical non-response

Among the 107 surgically treated patients which came from Subgroups 2 and 3, six individuals (5.6%) reported either no postoperative improvement or symptomatic worsening at follow-up, corresponding to a NASS Patient Satisfaction Index score of 4. Three of these patients were from Subgroup 2 and three from Subgroup 3. Detailed retrospective analysis of these cases is presented in the Results section.

The three cases from Subgroup 2 had morphometric measurement just fractions of mm short of the qualifying threshold and were offered surgery due to marked clinical improvement occurring during ICT. In retrospect, this clinical response carried disproportionate weight in the final surgical decision. By the same token, lack of symptomatic improvement despite meeting morphometric thresholds in asymptomatic subjects who only have CCJ hypermobility (and not symptomatic instability) reinforces the tenet that biomechanical correction alone does not uniformly translate into clinical benefit in patients with multisystem connective tissue disorders. The combination of an ICT clinical score along with an ICT morphometric score is aimed at protecting against these risks. This dichotomy underscores the rationale for our dual ICT scoring system, which requires concordance between morphometric improvement and real-time symptom response. This approach aligns with prior literature demonstrating poor correlation between static craniocervical morphometrics and symptom severity in CTD populations [12, 16]. Our surgical decision-making was based on dynamic concordance rather than static imaging abnormalities alone.

An important lesson learned after the analysis of this small group of non-responders (NASS score = 4) stems from the marked difference in the rates found between Subgroups. The 3/10 proportion of non-responders encountered in Subgroup 2 serves as a cautionary note against making exceptions to our internal rule, which requires both clinical and morphometric ICT scores to be positive before conferring surgical qualification. Conversely, the 3/88 proportion in Subgroup 3 provides encouraging feedback for the continued use of the two-stage protocol and the dual ICT scoring system for both diagnosis and surgical selection.

These observations do not undermine the protocol's validity but rather define its discrimination limits near the eligibility threshold. They highlight the need for continued refinement of weighting strategies in borderline cases and support future prospective investigation aimed at improving specificity without inappropriately restricting access for well-selected candidates.

Limitations of this study

This study reflects the experience of a single surgeon at a single institution specialized in the evaluation and treatment of patients with connective tissue disorders (CTDs) and craniocervical instability (CCI). As such, inter- and intra-observer variability may arise if this protocol is implemented at other centers. The study is further limited by its retrospective design, reliance on patient-reported outcomes, and lack of a sham-controlled comparator; therefore, placebo and expectancy effects cannot be excluded.

Although the morphometric measurements and neurological examination findings obtained during ICT are objective, patient-reported symptoms and perceived symptom improvement remain inherently subjective. To mitigate potential confounding, patients underwent two independent psychiatric evaluations to assess for major psychiatric disorders and coping capacity.

During invasive cervical traction (ICT), symptom response was assessed using structured, symptom-specific elicitation combined with contemporaneous neurological examination, rather than global or anecdotal impressions. In addition to tracking pre-identified symptoms, standardized functional assessments were applied uniformly, irrespective of patients' reported complaints. For example, all patients underwent a swallowing assessment before and during traction, in which they were asked to drink water under direct observation. Objective signs of dysphagia—such as coughing or choking during swallowing—were recorded and compared between pre-traction and peak-traction states. These observed changes during traction were detected through structured clinician observation, including swallowing function, respiratory pattern, and congruence between verbal response and body language, reducing the likelihood that symptom modulation during ICT reflected expectancy or suggestion alone.

Nevertheless, we recognize that placebo effects cannot be fully excluded in a retrospective surgical series, particularly for subjective symptom domains. To address this limitation more directly, a prospective randomized crossover study comparing surgical and conservative management is currently underway at our institution.

Conclusions

The evaluation and management of CCI secondary to CTDs remains a subject of ongoing controversy, with debate surrounding diagnostic protocols and surgical selection criteria fueling understandable skepticism. In this study, we present our institutional experience using a structured surgical screening protocol specifically developed to assess CCI in individuals with CTDs, with the goal of introducing greater diagnostic clarity and rigor to clinical decision-making. Among all components of the protocol, ICT proved to be the most revealing, providing dynamic biomechanical insight not readily captured by static imaging or conventional assessments alone.

While this represents a preliminary, single-surgeon, single-center experience, we strongly believe that further multicenter studies are warranted. We are encouraged that the initial CCF outcome data reported here support the clinical utility of our screening protocol. Looking ahead, we anticipate that future studies—particularly those evaluating surgical outcomes in patients selected using this protocol—will help resolve ongoing controversies in the field. In parallel, we are currently initiating a randomized crossover study at our institution comparing surgical and conservative treatment strategies in this population, which we believe will offer critical insight into optimal care pathways moving forward.

Supplementary Information The online version contains supplementary material available at https://doi.org/10.1007/s10143-026-04195-z.

Acknowledgements This work was supported with a gift provided by the Canerector Foundation. We would like to acknowledge the patients who trusted us with their care. We would also like to acknowledge Dr. Thomas H. Milhorat for his mentorship, Dr. Roger W. Kula for his example.

Authors' contributions Paolo A Bolognese—conceptualization, drafting original manuscript, writing, revising the manuscript, data collection, data analysis/interpretation, patient care, supervision, resources Allison R Bloom—conceptualization, revising the manuscript John B Biggins—conceptualization, data collection, data analysis/interpretation, revising the manuscript Andrew Brodbelt—conceptualization, data analysis/interpretation, revising the manuscript Misao Nishikawa—conceptualization, data analysis/interpretation, revising manuscript Mansoor Foroughi—conceptualization, revising the manuscript Ilene S Ruhoy—conceptualization, drafting the original manuscript, data analysis/interpretation, data collection, revising the manuscript Randall Dass—conceptualization, data analysis/interpretation, revising the manuscript Kamil Rohana—conceptualization, data collection, data analysis/interpretation, revising the manuscript Jeffrey D Wood–data collection, data analysis/interpretation, drafting original manuscript, revising the manuscript David Putrino—resources and supervision, revising the manuscript Veit Rohde—revising the manuscript Christoph Bettag—revising the manuscript Shawn Belverud—revising the manuscript Travis Caton—conceptualization, data analysis/interpretation, revising the manuscript Tanvir Choudhri—supervision, revising the manuscript.

Funding This work was supported by a generous Gift provided by the Canerector Foundation.

Data availability The raw data have been attached in the SUPPLEMENTS section and titled: SUPPLEMENTARY MATERIAL 10—Raw data of the ICT patients.

Declarations

Ethics approval Research Reporting Guidelines
This study was conducted in accordance with the Declaration of Helsinki and adhered to the STROBE and PROCESS 2023 reporting guidelines.
IRB approval
The retrospective record review pertinent to this study was approved by the Internal Review Board of the two Institutions where the procedures took place: (IRB #13-655B: North Shore University Hospital – Northwell, Manhasset, NY; WIRB #1276110 and WIRB #1262008: Mount Sinai South Nassau – Oceanside, NY).

Consent to participate Standard informed consent was given by the patients for all surgical procedures.

Consent to publish The authors affirm that human research participants provided informed consent for publication of the images in Figs. 2a, and 2b.

Competing interests The authors declare no competing interests.

Clinical trial number Not applicable.

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