Reviews

Pathoanatomy, biomechanics, and treatment of upper cervical ligamentous instability: A literature review

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Keywords: Cervical spine injuries, finite element analysis, 3D reconstruction, minimally invasive, spine surgery

https://doi.org/10.52965/001c.37099

Orthopedic Reviews

Vol. 14, Issue 3, 2022

Background

Cervical spine instability broadly refers to compromise of the articular congruity. It can be stratified according to spinal level, functional compromise, and mechanism of instability. Conventional wisdom advocates for use of bracing and physical therapy with only a subset of patients proceeding to obtain surgical treatment.

Objective

The purpose of this review article is to summarize the current state of knowledge on upper cervical ligamentous instability.

Methods

The literature search was performed in Mendeley. Search fields were varied until redundant. All articles were screened by title and abstract and a preliminary decision to include an article was made. The full-text screening was performed on the selected articles. Any question regarding the inclusion of an article was discussed by 3 authors until an agreement was reached.

Results

Many articles report on the etiological factors including ligamentous laxity, traumatic injury, syndrome instability, iatrogenic instability, congenital, and inflammatory causes. A few recent studies elucidate new findings regarding pathoanatomy through the use of finite element analysis. A few articles demonstrate the diagnosis and show that radiographs alone have a low diagnostic rate and that functional MRI may be able to better quantify instability. Conservative treatment has been described, but there are no outcome studies in the literature. Surgical treatment has been described in many different populations with good radiologic and clinical outcomes. Recently the use of preoperative 3D CT reconstruction has been described with radiographic and immediate postoperative patient-reported outcomes.

Conclusion

The presentation of upper cervical spinal instability can be asymptomatic, symptoms of isolated instability, symptoms of nerve irritation, vertebrobasilar insufficiency, or severe neurologic compromise. 3D fine element analysis models and motion-capture systems have the potential to increase our understanding of the pathoanatomic cascade in both traumatic and non-traumatic cases of upper cervical spinal instability. A few modalities

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on the horizon could increase diagnostic potential. More efforts are needed regarding the use of fine element analysis in understanding the pathoanatomic cascade, the long-term outcomes of children over a spectrum of syndromic causes, and the potential of preoperative virtual simulation to improve surgical outcomes.

1. INTRODUCTION

Cervical spine instability broadly refers to articular compromise that renders joints of the cervical spine vulnerable to disruption. Cervical spine instability, or CSI, may be stratified according to spinal level, functional compromise, and mechanism of instability.[1,2] Cervical instability occurs due to laxity or rupture of the supporting ligaments of the cervical spine, which allows for excess motion and displacement of the facet joints under normal physiologic conditions. Considering the architecture and proximity to both the spinal cord and brainstem, cervical spine instability can produce irritation to nerves, possible structural issues, and debilitating pain.[3]

Conventional wisdom generally supports the use of bracing and physical therapy to treat these patients. However, there are other options available for patients in whom non-operative options fail. There is no consensus in the literature regarding, in which patients a higher-level treatment option may benefit.

The purpose of this review article is to summarize the current state of knowledge on upper cervical ligamentous instability. We discuss new 3D finite element model reconstructions of the cervical spine that shed light on the pathoanatomy of cervical spine instability, advancements in the diagnosis of traumatic cervical spine injuries, and appraise the current state of minimally invasive and surgical options for this clinical entity.

2. MATERIALS AND METHODS

SEARCH STRATEGY

The literature search was performed using Medical Search Headings (MeSH) in Mendeley version 1.19.8. Articles published between January 1975 to December 2021 were Search fields were varied until no new articles were collected at which point the search was considered exhaustive.

STUDY SCREENING AND SELECTION

All articles were screened by title and abstract. An initial decision to include a given article was made based on relevance of the information within the abstract as determined by our inclusion/exclusion criteria (**Table 1**). This constructed a list of preliminarily included articles. These articles then underwent a full-text screening process. Any question regarding the inclusion of an article was discussed by all authors until an agreement was reached. The bibliographies of these articles were also hand-searched to identify any missing articles.

3. RESULTS

3.1. ETIOLOGY

LIGAMENTOUS LAXITY

A subset of patients with upper cervical spinal instability are patients with chronic patholaxity. Laxity of various osteoligamentous structures of the cervical spine correlate can be seen with no specific inciting event. The resulting instability may present with anterior vertebral translation, hypermobility at the atlantoaxial junction, or rotatory instability.

TRAUMA

CSI secondary to trauma ranges in severity from minor ligamentous injury to subjective subluxation events to frank vertebral dislocation. Atlantoaxial instability occurs after energetic impact and can presents as rotatory instability, C1-C2 horizontal displacement, C1-C2 distraction injury, and atlanto-dens instability. Occipitocervical dissociation results from complete or near complete disruption of occiput/C1 ligamentous structures and follows extreme forces of hyperflexion, hyperextension, or hyper-lateral flexion. Fractures of the atlas , axis, or odontoid can comitantly occur and can also contribute to upper cervical spinal instability.[4]

SYNDROMIC

Many syndromes are associated with CSI and include pseudoachondroplasia, Larsen syndrome, diastrophic dysplasia, 22q11.2 Deletion syndrome, Morquio syndrome, Goldenhar syndrome, spondyloepiphyseal dysplasia congenita, Kniest dysplasia, and fibrodysplasia ossificans progressive.[5]

IATROGENIC

Cervical spine instability may result from surgical intervention to treat a variety of conditions, including congenital deformities like Chiari II malformations.[6] Osteoradionecrosis may also cause cervical spine instability in patients receiving radiotherapy for head and neck cancer.[7]

CONGENITAL

Congenital upper cervical anomalies, including unilateral absence of C1, aplasia of the odontoid, and os odontoideum predispose patients to supra-axial CSI. Though the natural histories of C1 absence and odontoid aplasia are unknown, os odontoideum, a transverse gap separating the basal segment from the odontoid segment, may be caused by vascular or mechanical compromise resulting in segmental necrosis.[8]

Table 1. Our inclusion and exclusion criteria as applied during the title/abstract screening and full-text screening.

Inclusion Criteria	Exclusion Criteria
• Upper Cervical Spinal Instability • Use of conservative, minimally invasive, or surgical treatment options • Level III Evidence and higher	• Other upper cervical disease or diffuse spinal instability • Absence of a studied treatment option • Absence of clinical, functional, or radiographic outcomes • Expert Opinion or Case Reports

INFLAMMATORY

Chronic inflammatory diseases cause synovitis and ethisitis, resulting in progressive instability characterized by atlantoaxial subluxation, vertical subluxation, or subaxial subluxation. Rheumatoid arthritis and psoriatic arthritis both have predilection for the cervical spine, with various studies showing over a 40% incidence in CSI with each.[9, 10] Less commonly, Grisel Syndrome, a rare post-infectious inflammatory reaction, can cause atlantoaxial subluxation secondary to soft tissue inflammation.[11]

3.2. RISK FACTORS

Known risk factors for ligamentous laxity include diagnosis of chronic patholaxity, os odontoideium, Down's syndrome, connective tissue disorders. Os odontoideum results in a mobile dens and can cause the incompetence of the transverse ligament.[12] Down's syndrome results specifically in C1-C2 instability, in addition to decreased strength of the transverse and alar ligaments. These changes predispose Downs syndrome patients to upper CSI with potential for neurologic complications.[13] A significant risk factor for CSI in patients with Downs syndrome is age. Younger age correlates with an increase in the atlantoaxial gap and a higher degree of ligamentous laxity.[14] Connective tissue disorders like Marfan's Syndrome can also increase ligamentous laxity thereby decreasing the efficacy of upper cervical stabilizing ligaments, resulting in cervical instability.[11]

Risk of cervical spine instability secondary to trauma can be stratified according to mechanism of injury. The most common traumas associated with cervical injury are falls and motor vehicle accidents.[4] Thus, populations at increased risk thus follow a bimodal distribution with one peak in the 2nd and 3rd decade among men and another in elderly women. Additional risk of occipitoaxial dislocation following high energy impact exists among children due to their horizontal articular surfaces and ligament laxity.[1]

Multiple independent risk factor for cervical spine instability among patients with rheumatoid arthritis have been identified. These include male sex, positivity for rheumatoid factor and ACPA, erosive changes in the peripheral joints, and presence of osteoporosis.[9] Additionally, established mutilating changes, corticosteroid administration, and prior joint surgeries correlate with faster progression of CSI in RA patients.[10]

3.3. CLINICAL PRESENTATION

Presentation of cervical spine instability is highly variable and may depend on the location of instability (occiput-C2 vs C3-C7), the mechanism of instability, and the involvement of osseous, ligamentous, or vascular structures. It has previously been suggested that there exist two categories of cervical spine instability: radiographically appreciable CSI (RACSI) and clinical CSI (CCSI).

RACSI often reflects disruption of osseo-ligamentous structures resulting in a marked decrease in passive stability.[15] This category of instability can be further stratified according to location of disruption. Patients with RCSI of the upper cervical spine at the levels of C0, C1, and C2 commonly present with nerve irritation and vertebrobasilar artery insufficiency, manifesting as vertigo, tinnitus, dizziness, facial pain, arm pain, and migraine headaches.[16] It is important to note the difference in clinical presentation of lower cervical spina instability, which more often presents with muscle spasm, crepitation, and paresthesia with concomitant chronic neck pain.[16]

CCSI describes cervical spine instability that may or may not appear radiologically with corresponding symptomology. CCSI can be described as hypermobility. Symptoms include posture intolerance, easy cervical trunk fatigue, frequent need for self-manipulation, feeling of instability, and sharp pain.[15]

3.4. ANATOMY AND PATHOANATOMY

CURRENT UNDERSTANDING

The cervical spine is the most mobile portion of the spine, and therefore is dependent on ligamentous tissue for stability. The upper cervical spine consists of the occiput, the atlas, and the axis (C0, C1, and C2 respectively). The upper cervical spine is responsible for 50% of total neck flexion and extension, as well as 50% of total neck rotation.[17] Articular facets of the upper cervical spine have very little inherent stability and thus rely on capsuloligamentous attachments for stability and function.[8,18] Primary stabilizers of the cervical spine including the transverse, alar and capsular ligaments.[19] The transverse ligament helps restrict neck flexion, the alar ligaments limit axial rotation, and capsular ligaments maintain stability during neck flexion.[16]

The occipitocervical junction is stabilized by two groups of ligaments: the first group (the articular capsule ligaments, the anterior and posterior atlantooccipital ligaments, and the lateral atlantooccipital ligaments) anchors the cranium to the atlas while the second group (the apical dental ligament, the alar ligaments, the tectorial membrane, and the ligamentum nuchae) provides stability across the craniocervical junction.[1] The atlantoaxial joint is primarily stabilized by the transverse ligaments which secure the odontoid to the anterior arch of C1, with secondary support provided by the alar and apical ligaments.[13] Chronic instability can lead to a permanent lengthening of a ligament over time, which causes abnormal joint loads, chronic subluxation, and worsening instability.

FINITE ELEMENT ANALYSIS IN ELUCIDATING THE PATHOANATOMY OF THE UCSI

Finite element model analysis refers to concept by which given system is broken up into a large number of individual segments. By studying the kinematics/kinetics of each individual segment, information about the mechanical environment of the object of the whole can be deduced. In the recent years, there have been a few finite element model analysis that have demonstrated interesting additions to our knowledge base regarding the pathomechanics of the unstable upper cervical spine. Their results are summarized in **Table 2**.

3.5. DIAGNOSIS

The diagnosis of cervical instability is typically a clinical diagnosis based on history and physical exam. A history of inflammatory disease, connective tissue disorders, or trauma can heighten clinical suspicion.[20] Symptoms of cervical instability can be variable, and include but are not limited to neck pain, limited mobility, torticollis, and neurological symptoms.[21] A thorough neurological examination may indicate the level of instability and aid in monitoring the progress of patients during treatment. Physical exam maneuvers such as the Sharp-Purser test, side-bending test, passive upper cervical flexion test, and lateral stability test which can also be used for assessment of upper cervical spine instability.[22]

There is currently no standardized radiographic study that can consistently diagnosis cervical instability, and there is not a definitive correlation between clinical symptoms and degree of instability visualized on x-ray.[23] A recent cross-sectional study found that flexion and extension radiographs have very little utility in diagnosis of upper cervical ligamentous injuries. Oh and colleagues found no true positive in their population consisting of 4 cases of known traumatic UCSI. CT scan has replaced plain radiographs as the standard of care for screening of cervical spine injuries; however, cannot provide direct visualization of ligamentous injury.[24] Multidetector computed tomography remains to be the gold standard to assess the cervical spine in a traumatic setting.[25,26]

MRI is not frequently sought out in cases of traumatic cervical spine injury especially in the setting of a normal CT. A recent systematic review found that decision to obtain an MRI is largely based on clinical suspicion as opposed to craniometric measures from radiography or CT.[27] However, CT scan alone has the potential to miss important findings that can result in missed or delayed surgical treatment for patients.[26] A review found that 10% of their traumatic UCSI patients had positive MRI findings and 52% of these patients required subsequent surgical treatment.[26] Functional MRI has been posed as an imaging modality that can quantify the degree of subluxation through an arc of movement.[16]

3.6. CONSERVATIVE TREATMENT

Conservative treatment cervical instability is generally indicated in patients without severe pain, neurologic deficits, or vertical translocation with compromise of the vertebral artery.[28] This generally includes pain control, immobilization with bracing, and subsequent activity modification to avoid motions which may worsen instability.[29] Immobilization can be done via a halo vest. The halo vest was originally designed to treat patients with severe scoliosis or neck muscle paralysis due to poliomyelitis, however since then it has been widely used to immobilize the neck in pre- or postoperative settings for cervical spine injuries or deformities and upper cervical disorders. Braces are typically used for a minimum of three months until reassessment of the neurological condition with repeat imaging.[29] Formal physical therapy can also aid in treatment.[30] However, there are virtually no studies regarding the outcomes of patients having received conservative treatment for diagnosed UCSI.

3.7. MINIMALLY INVASIVE TREATMENTS

The results of our systematic review demonstrated that minimally invasive treatment options for upper cervical spinal instability are not well described, with no articles describing the use of steroid injections, pulsed radiofrequency, peripheral nerve stimulation, or transcutaneous electrical nerve stimulation in the management of these patients. All literature searches pertaining to minimally invasive treatments resulted in endoscopic surgical results or smaller incision surgeries which are discussed in surgical treatments.

3.8. SURGICAL TREATMENT

Minimally invasive treatment options for cervical instability such as atlantoaxial fusion can significantly decrease muscle injury, minimize blood loss, reduce the duration of hospital stay, which leads to improved functional outcomes compared with conventional procedures.[31,32]

BACKGROUND

While it is agreed that occipito-cervical fusion should only be used for cases of instability of the cranio-cervical junction that result in neurological deficits or severe potential neurological damage, there is no uniformity of opinion for specific criteria to characterize "instability" beyond neurological sequelae.[33]

The integrities of the transverse ligament, anterior longitudinal ligament, tectorial membrane, and alar ligaments are the most significant contributors to the stability of the

Table 2. A summary of the recent finite element model analyses regarding the pathoanatomy of cervical spine instability.

Lead Author	Title	Year	Journal	Institution	Level of Evidence	Pathology Studied	Results	Conclusions
M. Beauséjour,	Contribution of injured posterior ligamentous complex and intervertebral disc on post-traumatic instability at the cervical spine	2020	Computer Methods in Biomechanics and Biomedical Engineering		Non-clinical study (Finite Element Analysis)	Traumatic UCSI	Posterior ligamentous complex (PLC)removal had little impact at C2-C3 but increased local range of motion (ROM) at the injured level by 77.2% and 190.7% at C4-C5 and C6-C7, respectively. Complete IVD rupture had the largest impact on C2-C3, increasing C2-C3 ROM by 181% and creating a large antero-posterior displacement of the C2-C3 segment.	The PLC may not play as critical of a role in stabilized the posterior upper cervical spine, and the IVD plays a larger role. Finite element analysis is an apt tool by which to understand the pathomechanics of the cervical spine.
Ivancic P	Cervical spine instability following axial compression injury: A biomechanical study	2014	*Orthopaedics and Traumatology: Surgery and Research*		IV	Traumatic UCSI	The sagittal instability parameters indicated extension-compression injuries at the upper and middle cervical spine and flexion-compression injuries at the lower cervical spine. Increases in extension RoM were 14.9° at the upper cervical spine and 24.9° (P< 0.05) at the middle cervical spine and in flexion RoM at C7/T1 were 25.6° RoM and NZ increases in axial rotation and lateral bending were nearly symmetric among left and right.	Head-first collisions may result in biomechanical instability by different mechanisms in the upper cervical spine as compared to the middle cervical spine.
Wang X Feng M Hu Y	Establishment and Finite Element Analysis of a Three-dimensional Dynamic Model of Upper Cervical Spine Instability	*2019*	*Orthopaedic Surgery*		Non-clinical study (Finite Element Analysis)	Traumatic UCSI	After the upper cervical spine instability, the pressure of the alar ligament during the upper cervical spine extension was increased from 2.85 to 8.12 MPa. The pressure of the flavum ligament was increased during the upper-cervical spine flexion, from 0.90 to 1.21 MPa. The pressure of the odontoid ligament was reduced during the upper cervical spine flexion and extension, from 10.46 to 6.67 MPa and 25.66 to 16.35 MPa, respectively. The pressure of the anterior longitudinal ligament and cruciate ligament was increased. The pressure of the anterior longitudinal ligament was increased during flexion and extension, from 7.70 to 10.10 MPa and 10.45	Finite-element analysis has the capacity to increase our understanding regarding ligament stress in upper cervical spinal instability.

Lead Author	Title	Year	Journal	Institution	Level of Evidence	Pathology Studied	Results	Conclusions
							to 13.75 MPa, respectively. The pressure of the cruciate ligament was increased during flexion and extension, from 2.29 to 4.34 MPa and 2.32 to 4.40 MPa, respectively. During upper cervical spine flexion, the angle of the atlanto-occipital joint was increased from 3.49° to 5.51°, and the angle of the atlanto-axial joint was increased from 8.84° to 13.70°. During upper cervical spine extension, the angle of the atlanto-occipital joint was increased from 11.16° to 12.96°, and the angle of the atlanto-axial joint was increased from 14.20° to 17.20°.	
Liang L	Establishment and evaluation of a cadaveric model of chronic strain-induced upper cervical spine instability based on fascia-bone theory	*2020*	*Chinese Journal of Tissue Engineering Research*			Chronic Patholaxity UCSI	(1) During anterior flexion, the range of motion of the atlantoaxial joint (C1-2) and the entire upper cervical vertebra (C0-2) of the specimens after modeling was significantly larger than that before modeling ($P < 0.05$). During posterior extension, the range of motion of the atlantooccipital joint (C0-1) and the entire upper cervical vertebra (C0-2) of the specimens after modeling was significantly larger than that before modeling ($P < 0.05$). During both flexion and extension, the range of motion of the atlantoaxial joint (C1-2) and the entire upper cervical vertebra (C0-2) of the specimens after modeling was significantly larger than that of the pre-modeling specimen ($P < 0.05$). (2) During lateral flexion, the range of motion of the atlantooccipital joint (C0-1), the atlantoaxial joint (C1-2), and the entire upper cervical vertebra (C0-2) of the specimens after modeling was increased compared with that before modeling. However, there was no significant difference ($P > 0.05$). (3) During right rotation, the range of motion of the whole upper cervical spine (C0-2) of the specimens after modeling was significantly increased compared with that before modeling ($P < 0.05$). During both left and right rotation, the range of motion of the atlantoaxial joint (C1-2) and the whole upper cervical spine (C0-2) of the specimens was significantly larger than that of the specimens before modeling ($P < 0.05$).	

upper cervical spine when injured.[34] Providing criteria for stability that indicates specific intervention will improve primary treatment success and optimize healing time.[35] Today, typical occipito-cervical fusion includes a posterior approach and involves use of rigid plate implants, wiring, and bone graft materials to achieve a solid spinal fusion.[36]

CHILDREN

In children, nontraumatic upper cervical spine instability results from abnormal development of bone or ligamentous structures, or connective tissue disorders.[37] This can ultimately compromise the spinal cord during movement of the cervical spine. In children, surgical fixation is hindered by fragile posterior structures. There has been a push for the implementation of modern screw fixations and treatment protocols in children 6 years and older.[38] Good outcomes have been shown in a few pediatric syndromes for upper cervical spine instability including Morquio syndrome,[39] sponyloepiphyseal dysplasia,[38] and other skeletal dysplasias.[38]

TRAUMATIC

Upper cervical spine trauma often involves craniocervical dissociation and atlantoaxial instability.[40] Atlantoaxial transpedicular screw fixation has been established as a safe and effective treatment for upper cervical spine trauma, achieving full atlantoaxial rotational function for a high percentage of patients.[41,42]

The authors of a prospective controlled trial report similar function and radiographic outcome, decreased donor site morbidity in the setting of monosegemental anterior discectomy and interbody fusion when comparing Syncage-C filled with autologous cancellous bone graft as compared to tricortical iliac crest autograft. However, operation time and hospital stay were significantly shorter in the in the Syncage-C filled group. Importantly, functional and radiographic outcomes were good in both groups.

NONTRAUMATIC

Occipitocervical fixation in patients with non-neoplastic disease, predominantly rheumatoid arthritis, and neoplastic diseases results in overall improved condition. An efficient technique reported in the literature for non-traumatic upper cervical spine fixation was the placement of an intraoperatively contoured Luque rectangle wired from the occiput to appropriate cervical spine levels.[43] For nontraumatic cervical spine fixation, rigid techniques seem to lead to a better imagiological improvement when compared to wiring ones.[44]

NOVEL APPROACHES

In contrast to the classic posterior surgical approach, anterior transarticular screw though previously shown to have academic promise, has been neglected within the literature in recent years.[45] A recent study in the early 2000's provided a surgical technique and also reported favorable preliminary outcomes and encouraging in a series of 42 patients.[45]

NOVEL TECHNIQUES

Generally, there has been in a shift in the literature towards transpedicular screws atleast within certain realms of cervical spinal instability. Translaminar screws are typically used when pedicle screw placement is contraindicated or not possible. However, recent data suggest similar biomechanical stability and fusion rates of translaminar screws to other more common posterior fixation procedures. In addition, translaminar screw placement is technically less demanding and reduces the risk of vertebral artery injury.[46]

A technique that has been described is addition of an atlantoaxial pedicle screw to posterior fusion. Atlantoaxial screws are especially suitable for the treatment fracture instability as compared to additive stability provided by wire or lamina folders. A recent study found reliable atlantoaxial stability, maintenance a normal biomechanical line of the upper cervical spine, satisfactory bony fusion.[47] Old odontoid nonunion and both anterior- and posterior-destroyed atlantoaxial stabilities are also indications for atlantoaxial posterior pedicle screw fixation.[47]

Lastly, the use of preoperative 3D CT reconstruction to allow for patient-specific instrumentation is an important recent novel addition to the literature. Though the results of our literature search only revealed one included article on this topic, it is important to discuss how patient specific instrumentation may affect future clinical and research efforts. A recent study used preoperative virtual stimulation reduction to allows for adjustment of the clivus-axial angle (CAA), to guide intraoperative reduction, and to evaluate possible screw trajectories. This study from a high degree of success as measured by target and postoperative CAA values, complication rate, and post operative neck disability index.[48]

4. DISCUSSION

There is a wide range of etiologies that can lead to upper cervical spinal instability. Known risk factors including chronic patholaxity, down's syndrome, connective tissue disorders, rheumatoid arthritis, and os odontoideum. The clinical spectrum can range from asymptomatic, symptoms of isolated instability, symptoms of nerve irritation, vertebrobasilar insufficiency, or severe neurologic compromise. 3D fine element analysis models and motion-capture systems have the potential to increase our understanding regarding the pathoanatomic cascade in both traumatic and non-traumatic cases of upper cervical spinal instability.

Diagnosis is often missed given the low reported sensitivities for radiography and computed tomography in detecting subtle displacements in the cervical spine. MRI remains an option but is not cost-efficient and thus there is large resistance into incorporation in the evaluation of the UCSI patient. Functional MRI and other forms of dynamic imaging hold the possibility of increasing our diagnostic capabilities and thus result in earlier surgical treatment;

however, clinical evidence to support their integration into evaluation protocols is lacking.

Reliable radiographic fusion and stable neural outcomes have been described in certain subsets of the adult population and in a handful of pediatric syndromes. Preoperative virtual simulation through three-dimensional computed tomography reconstruction allows for personalization of instrument placement and shows early promise. More efforts are needed regarding the use of fine element analysis in understanding the pathoanatomic cascade, the long-term outcomes of children over a spectrum of syndromic causes, and on the potential of preoperative virtual simulation to improve surgical outcomes.

AUTHOR ROLES

Study concept and design: N. V.; I. U.; O.V.; Acquisition of data: N. V.; I. U.; O.V.; Analysis and Interpretation of data: N. V.; H.T.; H.K.; V.A.; I.U.; O.V.; Drafting of the Manuscript: N. V.; H.T.; H.K.; V.A.; I.U.; O.V.; critical revision of the manuscript for important intellectual content: N. V.; H.T.; H.K.; V.A.; I.U.; O.V.; Statistical Analysis: none; Administrative, technical, and material support N. V.; I. U.; O.V.; Study supervision: N. V.; I. U.; O.V.;.

DISCLOSURES STATEMENT

We have no disclosures or potential conflicts of interest.

FURTHER INFORMATION

This manuscript did not receive any funding and has not been presented at any conferences.

Submitted: January 05, 2022 EDT, Accepted: March 28, 2022 EDT

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