Annals of Case Reports
Massager N, et al. Ann Case Rep: 8: 1245
www.doi.org/10.29011/2574-7754.101245
www.gavinpublishers.com

Case Report

Syncope due to Cervical Spine Hyperextension in Hypermobile Ehlers-Danlos Syndrome: Successful Treatment with Cranio-Cervical Stabilization: Case Report

Nicolas Massager[1,2*], Valérie De Decker[3], Chiara Mabiglia[4], Anthony Nguyen[1], Henri-Benjamin Pouleau[1], Alexandre Jodaïtis[1], Daniele Morelli[1]

[1]Department of Neurological Surgery, University Hospital Tivoli, avenue Max Buset 34, 7100 La Louvière, Belgium

[2]Faculty of Medicine, Université Libre de Bruxelles, Route de Lennik 808, 1070 Brussels, Belgium

[3]Rheumatology, Private Practice, Brussels, Belgium

[4]Department of Neuroradiology, University Hospital Erasme, route de Lennik 808, 1070 Brussels, Belgium

***Corresponding authors**: Nicolas Massager, University Hospital Tivoli, Avenue Max Buset 34, B-7100 La Louviere, Belgium

Citation: Massager N, De Decker V, Mabiglia C, Nguyen A, Pouleau HB, et al (2023) Syncope due to Cervical Spine Hyperextension in Hypermobile Ehlers-Danlos Syndrome: Successful Treatment with Cranio-Cervical Stabilization. Case Report. Ann Case Report. 8: 1245. DOI:10.29011/2574-7754.101245

Received: 27 March 2023, **Accepted:** 31 March 2023, **Published:** 03 April 2023

Abstract

Background: The hypermobile form of Ehlers-Danlos syndrome (h-EDS) may be associated with cranio-vertebral instability causing neurological symptoms. The underlying mechanism of these symptoms refers to brainstem compression in flexion. We report a unique case of h-EDS with cranio-cervical instability causing syncopal events in cervical extension successfully treated with surgical stabilization.

Case Presentation: A 25-year-old woman with h-EDS complained of syncopal events occurring systematically when she made cervical extension. Radiological evaluation confirmed cranio-cervical instability due to ligamentous hyper laxity. Radio metrics performed on dynamic cervical MRI in flexion and extension revealed pathological values in the range of motion of the cervical spine. Surgical treatment with occipito-axial stabilization was performed. No postoperative complication occurred. The patient did not experience any syncopal event after the surgery. The reduction in the range of motion was not felt to be disabling by the patient. Postoperative dynamic radiographs confirmed the stabilization of the occipitocervical junction.

Conclusions: Syncope occurring systematically due to cranio-cervical instability in extension may occur in patients with h-EDS. Successful treatment with surgical posterior stabilization of C0-C2 allows to eradicate the disabling symptoms of syncope.

Keywords: Craniocervical Instability; Hypermobile Ehlers-Danlos Syndrome; Syncope; Surgical Stabilization; Case Report

Abbreviations: h-EDS: Hypermobile Ehlers Danlos syndrome; CXA: clivo-axial angle; pBC2: Grabb-Oaks measurement (perpendicular Basion-C2); BAI: basion-axis interval

Introduction

Hypermobile Ehlers Danlos syndrome (h-EDS) is a hereditary connective tissue disorder characterized by joint hypermobility and tissue fragility [1], neurological and spinal manifestations have already been reported [2-4]. Clinical symptoms refer to craniocervical instability with ventral brainstem compression [3,5]. We report a unique case of h-EDS not associated with Chiari malformation that presents disabling neurological symptoms of syncope occurring systematically when the patient made cervical hyperextension.

Case Presentation

A 25-year-old woman with h-EDS was referred to our department for the diagnosis and treatment of repeated discomfort, weakness and syncope in a few seconds after that the patient positioned in cervical hyperextension. The patient complained of increasing episodes of such disabling and bothersome symptoms with time. The symptoms started with discomfort and resolved if the patient stopped cervical spine extension but developed to syncope if the patient maintain hyperextension for more than a couple of seconds. The neurological examination was normal, no sign of clinical deficit, and no cervical medullary syndrome.

The neurophysiological evaluation revealed no disturbance. The radiological examination included standard X-ray of the cervical spine in neutral position as well as in flexion and extension (Figure 1), and cervical MRI in neutral position and in flexion and extension (Figure 2). The patient declared that she could extend more intensively the cervical spine during both exams but avoided to do it to avoid having a syncopal event during the exam. Radio metrics were made on MRI and have included the clivo-axial angle (CXA), the Grabb-Oaks measurement (pBC2) and the basion-axis interval (BAI) [6,7]. The CXA was measured at 138° in flexion, so at the inferior limit of the normal range: no kyphotic instability. The pBC2 was of 9mm, also at the limit for brainstem compression in flexion. The BAI interval between BAI in flexion and BAI in extension revealed craniocervical instability (value in flexion >12mm and interval >4mm) [6,7].

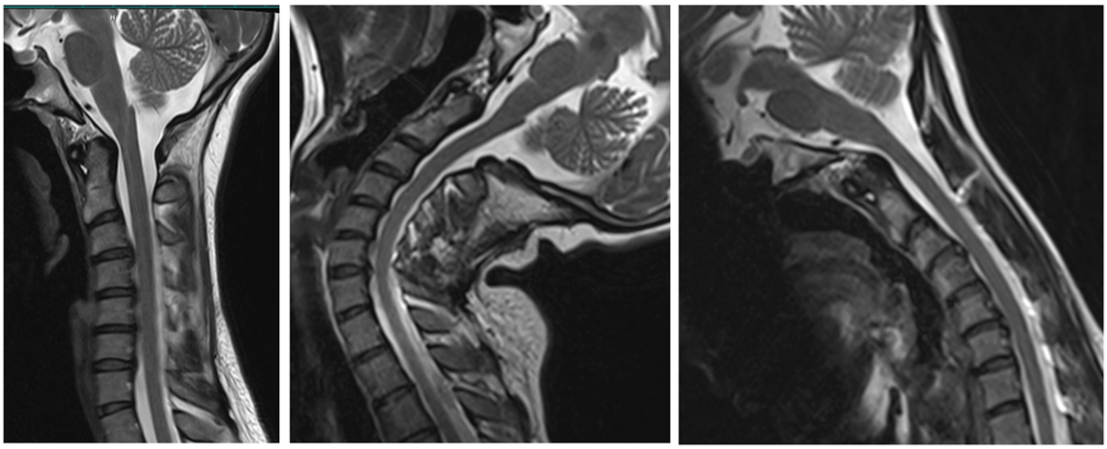

Figure 1: Dynamic MRI evaluation of the craniovertebral junction. Measurements of CXA, pBC0 and BAI were performed on these images in neutral position (left image), extension (middle image) and flexion (right image).

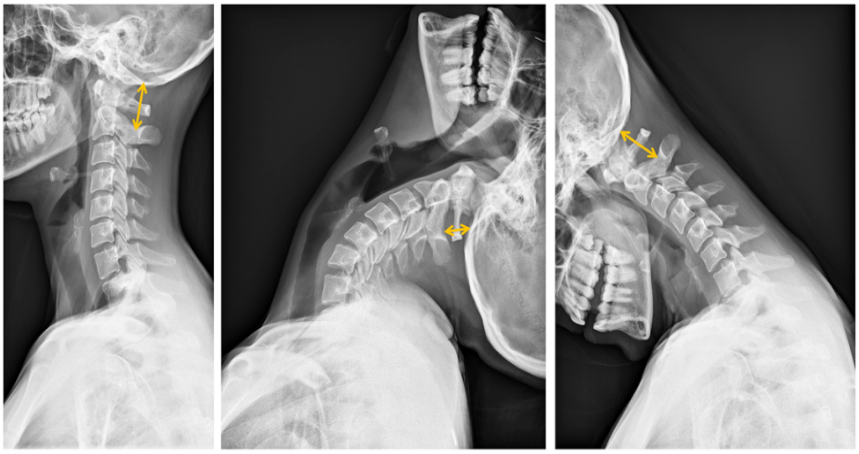

Figure 2: Dynamic X-ray in neutral position (left image), extension (middle image) and flexion (right image) showed marked preoperative occipito-axoidal instability of the craniovertebral junction.

Because of very disabling symptoms of brainstem compression/traction (syncope occurring systematically) and measurements of craniocervical instability at the radiological evaluation, an occipitocervical stabilization was proposed to the patient, who accepted it after extensive information on the benefits and risks of such procedure. The patient was operated on, and a C0-C2 arthrodesis was performed under radiological guidance. Care was taken to preserve ligaments attached to the spinous process and the lamina of C2. No sub occipital decompression was made, since no Chiari was present and symptoms occurred only in hyperextension.

The patient presented no postoperative complication and was discharged the day after surgery. She performed postoperative cervical physiotherapy and was controlled clinically and radiologically at 1, 3 and 6 months after surgery. The patient had a significate reduction of the amplitude of cervical flexion and extension (Figure 3) as well as lateral flexion (Figure 4), but this was clearly considered for the patient as more comfortable than the preoperative hyper laxity related to the h-EDS. The patient did not experience any episodes of weakness and syncope after the surgery, since these symptoms occurred preoperatively several times per day.

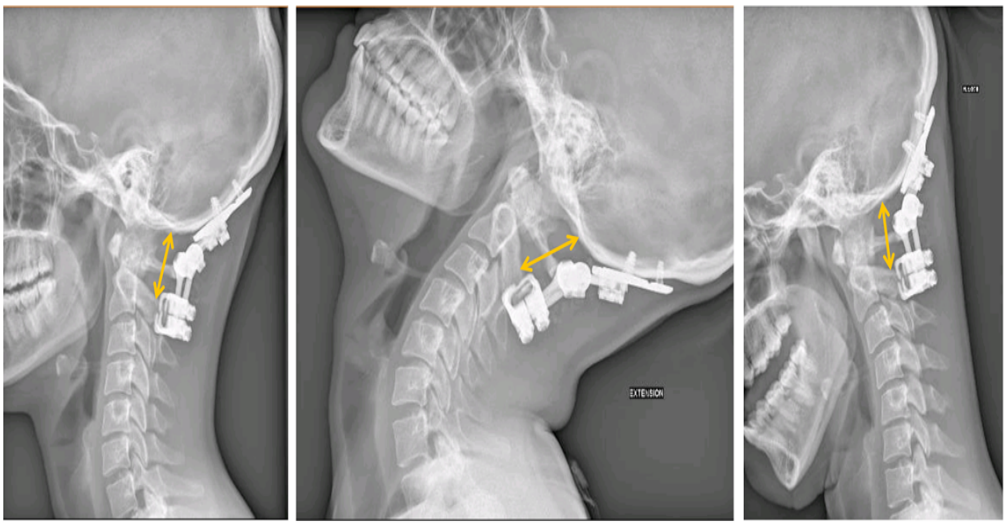

Figure 3: Postoperative dynamic X-ray ray in neutral position (left image), in extension (middle image) and in flexion (right image) demonstrated stabilization of the cranio-cervical junction.

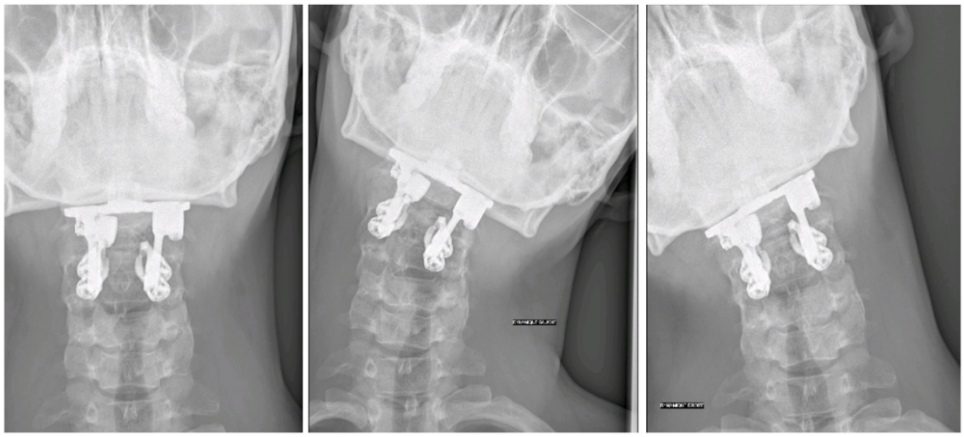

Figure 4: Dynamic anteroposterior radiograph of the cranio-cervical spine after surgery showing the limitation in lateral hypermobility induced by C0-C2 arthrodesis.

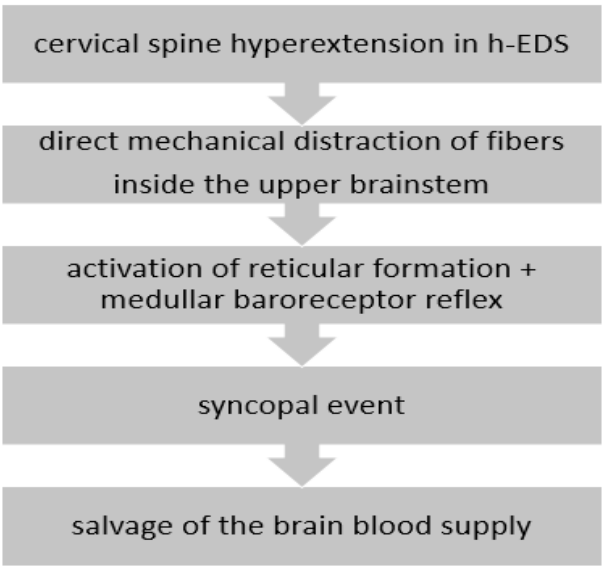

Figure 5: Proposed pathophysiological mechanism of the syncope in our patient.

Discussion and Conclusions

Patients with h-EDS may present some neurological and spinal manifestations of their connective disease, especially when Chiari malformation type I is associated with the disease [6,7]. However, even in patients with h-EDS without Chiari-I, some neurological symptoms may occur [2-4]. Neck pain and sub occipital headache are the most common findings. Dizziness, nausea, presyncopal or syncopal events have also been reported [7]. Although already described, syncope is an extremely rare symptom associated with h-EDS. To our knowledge, syncopal events precipitated by cervical extension in h-EDS patients have never been reported so far. The common symptoms associated with h-EDS are cervical pain or headache, are generally related to atlanto-occipital and atlanto-axial instability, and have been attributed to compression of the brainstem by hyper flexion [3,5]. No patient with symptoms related to occipito-cervical hyperextension in h-EDS has been reported in the literature. We have no definite explanation of why our patient experienced this exceptional presentation: no clear medical history or prior trauma has occurred in her past. However, our patient is an avid swimmer since childhood, and the repeated efforts of flexion-extension of the cervical spine during years during swimming in a patient with h-EDS could perhaps represent a contributing factor of the problem. Generally, patients with h-EDS are forbidden since infancy to play sports with excessive spinal movements. The exact pathophysiological mechanism of syncopal events in our patient are not known. No change in cardiac rhythm or blood pressure has been recorded during the syncopal episodes. Therefore, we hypothesized that the syncope of our patient is not from a classical vasovagal type [8]. We postulated that the initiation of the syncope starts in the brainstem itself, caused by distraction of fibres of the upper brainstem, inducing consequently a dysfunction of the medullar baroreceptor reflex and the midbrain reticular activating system [8-10]. The final purpose of the syncope is to salvage the brain's blood supply. The Figure 5 shows our understanding of the mechanism of the syncope in cervical spine hyperextension in h-EDS. We present an h-EDS patient with presyncopal and syncopal events occurring systematically and caused by cervical hyperextension. The preoperative radiological evaluation has included craniovertebral junction abnormality by a dynamic MRI evaluation, which has already shown its usefulness on this topic [11]. This exam has shown pathological values of BAI [7]. Surgical cranio-cervical stabilization has completely resolved these symptoms. Stabilization of the C0-C2 interval was confirmed on postoperative dynamic X-ray. Cranio-cervical stabilization, either by Trans articular screws or by posterior clamps, is the most common surgical procedure for symptomatic flexion-related h-EDS, sometimes associated with occipital decompression. The patient will be followed radiologically in the long-term since there is a possibility that, taking the hypermobility of the patient in consideration, accelerated degeneration of the cervical spine may occur below the arthrodesis, especially at the C2-C3 level. This case presentation highlights the importance of making a detailed analysis of the possible origins of symptoms in patients with symptomatic h-EDS, in relation with their hyper laxity of the cervical spine. The major role of dynamic radiological imaging, including MRI, is crucial in the search for the appropriate therapy.

Declarations: Ethics, consent and permission

Ethics approval: The present article was approved by the local Institutional Review Board of the Ethical Committee of the University Hospital Tivoli (Comité d'Ethique du CHU Tivoli, avenue Max Buset 34, 7100 La Louvière, Belgique).

Consent to participate: Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Availability of data and supporting materials: Not applicable.

Funding: We declare no source of funding for the present publication.

Competing interests: We declare no competing interest, financial and non-financial.

Acknowledgements: None.

Author's contribution: Not applicable

References

1. Tinkle B, Castori M, Berglund B, Cohen H, Grahame R, et al (2017) Hypermobile Ehlers-Danlos syndrome (a.k.a. Ehlers-Danlos syndrome type III and Ehlers-Danlos syndrome hypermobility type): clinical description and natural history. Am J Med Genet 175C: 48-69.
2. Castori M, Voermans NC (2014) Neurological manifestations of Ehlers-Danlos syndrome(s): a review. Iran J Neurol 13: 190-208.
3. Henderson FC, Austin C, Benzel E, Bolognese P, Ellenbogen R, et al (2017) Neurological and spinal manifestations of the Ehlers-Danlos syndromes. Am J Med Gen 175: 195-211.
4. Savasta S, Merli P, Ruggieri M, Bianchi L, Sparta MV (2011) Ehlers-Danlos syndrome and neurological features: a review. Childs nerv Syst 27: 365-371.
5. Halko GJ, Cobb R, Abeles M (1995) Patients with type IV Ehlers-Danlos syndrome may be predisposed to atlantoaxial subluxation. J Rheumatol 22: 2152-2155.
6. Henderson FC Sr, Henderson FC Jr, Wilson WA IV, Mark AS, Koby M (2018) Utility of the clivo-axial angle in assessing brainstem deformity: pilot study and literature review. Neurosurg Rev 41: 149-163.
7. Henderson FC Sr, Francomano CA, Koby M, Tuchman K, Adcock J, et al (2019) Cervical medullary syndrome secondary to craniocervical

instability and ventral brainstem compression in hereditary hypermobility connective tissue disorders: 5-year follow-up after craniocervical reduction, fusion, and stabilization. Neurosurg Rev 42: 915-936.

8. Wieshmann U, Meierkord H (1995) Cough syncope with hypertension caused by brainstem compression? Eur J Neurol 2: 498-500.

9. Habek M, Crnosija L, Gabelic T, Adamec I, Skoric MK (2017) The role of brainstem in syncope development. Clin Neurophysiol 128: e230.

10. Weig SG, Buckthal PE, Choi SK, Zellem RT (1991) Recurrent syncope as the presenting symptom of Arnold-Chiari malformation. Neurology 41: 1673-1674.

11. Gupta V, Khandelwal N, Mathuria SN, Singh P, Pathak A, et al (2007) Dynamic magnetic resonance imaging evaluation of craniovertebral junction abnormalities. J Comput Assist Tomogr 31: 354-359.