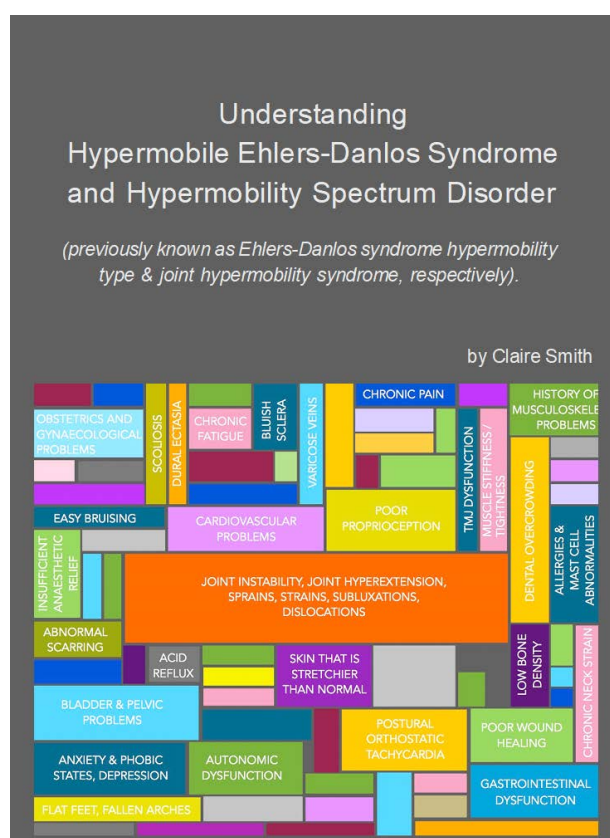


Craniocervical instability and hEDS

The following extract is taken from:
Understanding hypermobile Ehlers-Danlos Syndrome and Hypermobility Spectrum Disorder

Chapter 2 Part 2 - Area specific symptoms & comorbidities



© Claire Smith
Redcliff-House Publications

Craniocervical instability (also known as also known as the syndrome of occipitoatlantoaxial hypermobility) and hypermobile Ehlers-Danlos syndrome

The craniocervical junction (the interface between the skull and cervical spine) comprise a joint(s) which seems to be susceptible to the same strain and injuries as seen in other joints in Ehlers-Danlos syndrome (Tinkle B. et al 2017). The lack of structural stability at the craniocervical junction may lead to a deformation of the brainstem, upper spinal cord, and cerebellum. Studies about the prevalence as well as the symptoms, imaging, and management are scarce, but in some individuals it is thought craniocervical instability can result in one or more of the following: Nerve dysfunction, compression of the brain stem (loose ligaments can misalign the proper angle of the odontoid bone causing it to push backwards, compressing the brainstem), cranial settling (the skull sinks downward onto the spine), and Chiari malformation (Gazit Y. et al 2016 / The Pain Relief Foundation - Prof P. Eldridge et al / Milhorat T.H. 2007)

© **Claire Smith**

Author; Editor for the Hypermobility Syndromes Association; International Consortium for EDS and Associated Disorders Expert Patient (UK)

CCI and hEDS - extract taken from: Understanding Hypermobile Ehlers-Danlos Syndrome and Hypermobility Spectrum Disorder.

First published May 2017. Book reviewed in full by Dr A. Hakim: April 2017. Version 1.1 – Planned Extract
Date of Review: April 2020.

For more information on this publication, please visit:
redcliffhousepublications.co.uk

References:

Tinkle B; Castori M; Burgland B; Grahame R.; Kazkaz; Levy H 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 Part C Semin Med Genet* 9999C:1-22

Gazit Y; Jacob G; Grahame R. 2016 - Guest Editor: Balbir -Gurman A. - Ehlers-Danlos Syndrome—Hypermobility Type: A Much Neglected Multisystemic Disorder. *Rambam Maimonides Med J.* 2016 Oct; 7(4): e0034. Published online 2016 Oct 31. doi: 10.5041/RMMJ.10261. PMID: PMC5101008

Milhorat TH; Bolognese PA; Nishikawa M, McDonnell NB; Francomano CA. - Syndrome of occipitoatlantoaxial hypermobility,

cranial settling, and chiari malformation type 1 in patients with hereditary disorders of connective tissue. *J Neurosurg Spine* 2007; 7(6):601-9

The Pain Relief Foundation: - Prof Eldridge P; Dr Nash T; Dr Sharma M; Dr Wiles J; Dr Wells J; Lawton W; Gee D; Daley J. - <http://www.thepainrelieffoundation.com/craniocervical-instability/>

Resources:

The Pain Relief Foundation -including:

What is craniocervical Instability?; What is the link to Ehlers-Danlos syndrome?; How does craniocervical Instability occur?; What are the symptoms of craniocervical Instability?; How is craniocervical instability diagnosed? <http://www.thepainrelieffoundation.com/craniocervical-instability/>

Julier K. for Ehlers-Danlos Support UK, and ITV News 2017 - Evidence building for EDS patients with CCI needs to start somewhere

©2017 C.E. Smith

Permission has been granted for the following extracts from this publication to be reproduced by the Royal College of General Practitioners (RCGP):

Assessment wheel diagram; Craniocervical instability and hEDS/HSD; Fibromyalgia and hEDS/HSD; Initial diagnosis by clinical assessment; Mast cell activation syndrome and hEDS/HSD; Pregnancy and childbirth; Surgery.

This permission has been granted on the understanding that the extracts will form part of the reference resources available on the RCGP online toolkit for Ehlers-Danlos syndrome and Hypermobility Spectrum Disorder, helping to improve awareness and understanding of these conditions and their comorbidities.

This book and the individual contributions contained in it are protected under copyright by the author and publisher (other than as may be noted herein). The front cover, including image are designed and owned by the author and subject to copyright, all rights reserved.

Except as expressly stated above, no part of this publication may be reproduced or transmitted in any form or by any means, electronic or mechanical, including photocopying, recording, or any information storage and retrieval system, without the permission in writing from the source /publisher - Redcliff-house Publications (redcliffhouse@hotmail.com)

International Standard Book Number: 978-1-9997300-0-0

Printed in the UK

First printing 5 June 2017

Use of a term in this book should not be regarded as affecting the validity of any trademark or service mark.

Notices:

Every effort has been made to make this book as complete and accurate as possible, but no warranty or fitness is implied. To the fullest extent of the law, neither the Publisher nor the author, contributors, or editors, assume liability or responsibility for errors or omissions, nor any liability or responsibility for any injury and / or loss or damages to persons, entity, or property as a matter of negligence, products liability or otherwise, or from any use or misuse / misapplication or operation of any methods, products, instructions, or ideas contained in the material herein.

Knowledge and best practice in this field are constantly changing. As new research and experience broaden our understanding, changes in research methods, professional practices, or medical treatment may become necessary.

Readers are urged to take appropriately qualified medical advice in all cases. The information in this book is intended to be useful to the general reader, but should not be used as a means of self-diagnosis, or for the prescription of medication. It is the responsibility of practitioners, relying on their own experience and knowledge of their patients, to make diagnoses, to determine dosages and the best treatment for each individual patient, and to take all appropriate safety precautions.

Practitioners and researchers must check clinical procedures and always rely on their own experience and knowledge in evaluating and using any information, methods, compounds, or experiments described herein. In using such information or methods they should be mindful of their own safety and the safety of others, including parties for whom they have a professional responsibility.

With respect to any drug or pharmaceutical products identified, the publisher and the author make no representation, express or implied, that any drug dosages in the book are correct. Lay readers are advised to check product information and to always seek advice from their general practitioner or healthcare provider. Practitioners are advised to check the most current information provided (i) on procedures featured or (ii) by the manufacturer of each product to be administered, to verify the recommended dose or formula, the methods and duration of administration and contraindications.

The book from where the following extracts have been taken was produced in association with the Hypermobility Syndromes Association (HMSA) registered charity number: England and Wales (1011063) and in Scotland (SC037916), holding NHS England Information Standard accreditation. Having been through the HMSAs information standards process, there is an excellent level of medical accuracy and sound management advice to be found in this easily accessible book.