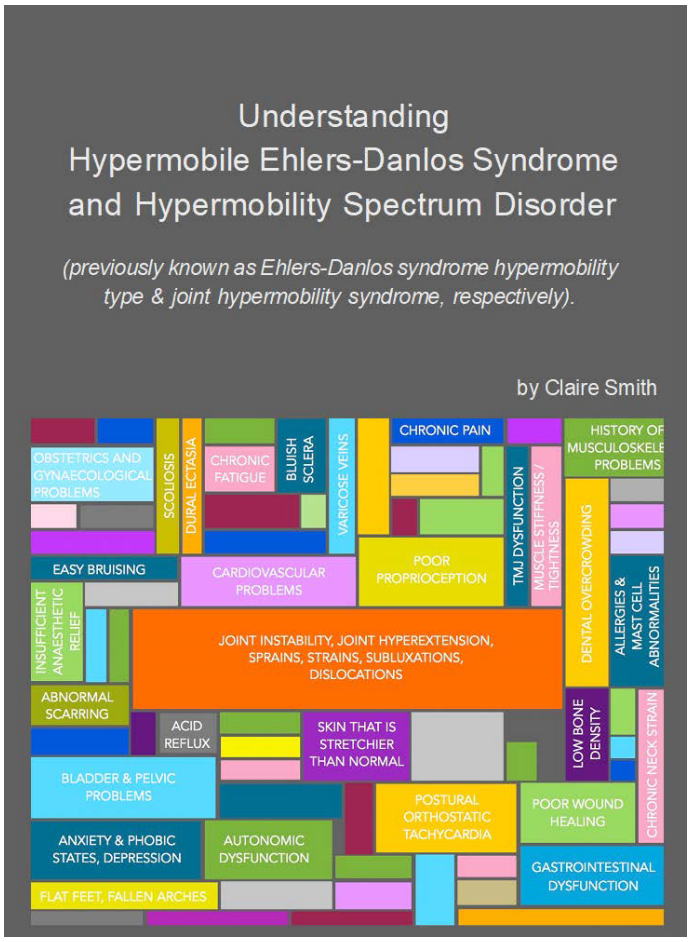
Surgical considerations - hEDS/HSD

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

Chapter 3
Diagnosis and Management



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Surgical considerations

'Many patients with hEDS/HSD are able to undergo the full range of surgical operations which may be offered to the general population, as long as their surgeon has been made familiar with their disorder, understands any possible complications this may cause, and is prepared to make any necessary alterations to their operating technique.' (Hakim A. 2015 - HMSA LWW Guide).

Each person and their symptoms / pre-existing and comorbid conditions are different. For that reason, it is very important that full details of medical history are made clear to all of the medical team involved in an individuals care, and appropriate measures to avoid complications, put in place.

When planning surgery, particular consideration should be given to:

- Instability of the cervical spine and/or temporomandibular joint. In the latter, intubation may need to be carried out more carefully due to temporomandibular joint dysfunction and the possibility of a slightly more fragile mucosal lining of the mouth (Castori M. 2012).
- The need for surgical and nursing staff to be prepared to take extra precautions when moving or positioning a patient with hEDS/HSD, in order to prevent injury, subluxation, or dislocation (Johnston B.A. 2006)
- Delays in wound healing are experienced by some with hEDS/HSD, and should be allowed for when planning for the removal of stitches and post operative movement. Experts recommend leaving sutures in place for longer than normal and/or brief immobilisation (Beighton P. 1969 / Shirley E.D. et al 2012 / Burcharth J. 2012 / Tinkle B. et al 2017); multilayered closure techniques with sufficient numbers of sutures (prefer nylon sutures over staples), deep stitches, and the support of wound closure strips (Weinberg J.et al 1999/ Tinkle B. et al 2017).
- The effects of muscle deconditioning may impact on recovery times and rehabilitation should be individualised (Castori M. 2012). For example, post-operative exercises designed to restore range of motion may need to be less aggressive than standard protocols, because the stiffness does not typically develop post-operatively in those with EDS, and there is a need to avoid stretching of repaired or reconstructed tissue (Shirley E.D. et al 2012)
- In patients who also have the common comorbidity, cardiovascular dysautonomia, it may be considered prudent to carry out cardiac evaluation with a possible echocardiogram before surgery. The anaesthetist involved in the patient's care should be made aware of the patient's cardiovascular dysautonomia diagnosis to allow them to plan for any increased risk of hemodynamic changes, such as a lowering in blood pressure, whilst under anaesthesia (Castori M 2012).
- Post operative orthostatic headache may occur in some patients as a result of spinal anaesthesia (Tinkle B. et al 2017).

In the case of minor surgeries (those which don't involve anaesthesia or assisted breathing), for example: dental procedures, tissue biopsies, mole removal etc., consideration should be given to the frequently reported resistance to topical EMLA cream and intradermal (situated or applied within the layers of the skin) lidocaine injections in hEDS/HSD patients (Arendt-Nielsen L. 1990 / Hakim A.J. 2005). According to Castori (2012), a double dose of anesthetic by intradermal injection may be an effective first choice option.

Although evidence is lacking, local anaesthetic resistance is also reported in those with hEDS/HSD in respect of epidural analgesia. This may result in insufficient or ineffective analgesia during instrumental or caesarean deliveries or other surgical procedures (Castori M. 2012, Hakim A.J. 2005).

If a patient's medical history shows potential for local anaesthetic resistance, then alternative methods of analgesia, for example, patient controlled analgesia (PCA), may be set up early, or in addition to other prescribed pain relief to enhance their action (Kochharn P.K. 2011).

NB/ Anesthetic issues are also discussed on pages 25, 65 and 104 of this book.

Extract © Claire Smith
Author; Editor for the Hypermobility Syndromes Association; International Consortium for EDS and Associated Disorders Expert Patient (UK).

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