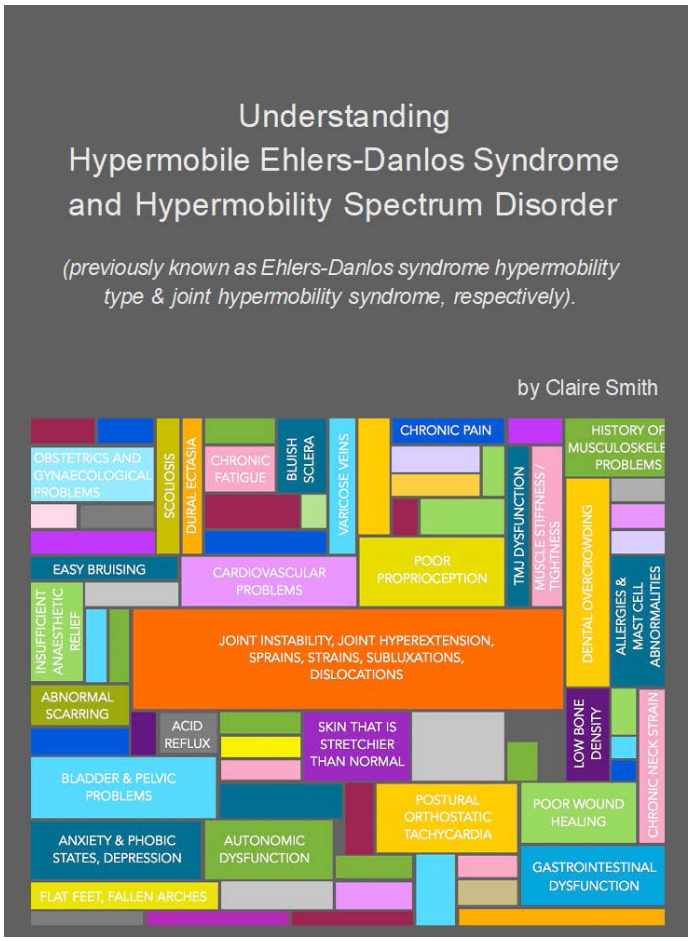
Pregnancy and childbirth & hEDS/HSD

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

Chapter 2
Part 2 - Area specific symptoms & comorbidities



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Pregnancy and childbirth & hEDS/HSD

Pregnancy and childbirth - introduction

HSD, and hEDS, are quite a broad diagnoses with varied severity and complexities seen between individuals. It is not surprising then, that the experience of childbirth within this group is also varied. Some pregnant women with hEDS/HSD experience more severe forms of the problems that many women (hypermobile or not) do during pregnancy; some experience these problems earlier in the pregnancy than would be expected in non-hypermobile women; some find the impact of any problems that occur may have a lasting or greater affect. Others, however, experience few problems (if any). In some cases, the effects of childbirth on the body remain unclear until average recovery periods have passed. Women's experiences are also affected by local standards of general, maternal and postnatal healthcare, personal circumstances and so on.

In 2016, an important paper was published from Sweden (Sundelin H.E. et al), which has helped to clarify concerns raised in the past. Dr Alan Hakim explains: '*The researchers identified 314 cases of pregnancy in women with either EDS or JHS* [HSD] *through the Swedish Patient Register and Medical Birth Register. The cases were compared with 1,247,864 controls* (pregnant women without a diagnosis of EDS/HSD). *The risk of complications in EDS/JHS* [EDS/HSD] *was assessed after adjusting for maternal age, smoking, number of pregnancies, and year of birth. Reassuringly, EDS/JHS* [EDS/HSD] *was not associated with any increased risk of preterm birth, the need for a caesarean section, stillbirth, complications in the infant at delivery (a low Apgar score), or the infant being small for gestational age or large for gestational age. At the same time similar observations have been published by Hugon-Rodin et al (2016). The obstetric outcomes in those participants with EDS-H/JHS* [hEDS/HSD] *were similar to those of the general French population for deliveries by caesarean section and premature births. However, they did find that the risks of spontaneous abortion (28%) and multiple spontaneous abortion (13%) in females with EDS-H/JHS* [hEDS/HSD] *were somewhat higher than is seen in the general population. General population studies show spontaneous abortion (loss of the fetus before 20 weeks gestation) occurs in 10-20% of women (Tulandi T. et al 2016).*'

Thankfully, awareness and understanding of hEDS/HSD among medical staff in the UK is on the rise and the age at which people are diagnosed has fallen dramatically over the last ten years alone. Naturally this improves the incidence of uneventful pregnancies (McLuckie F. 2015 HMSA).

Medication check

Very few medications have been found to be completely safe during pregnancy, so it is a good idea for all medicines to be reviewed with the pharmacist and/or primary care doctor, for advice on whether or not they can be discontinued altogether. Depending on the type of medications, it may be possible to stop some straight away, while others need to be weaned gradually. Where possible, medicines should be completely out of the system before the woman attempts to become pregnant.

Pregnancy

During pregnancy, there are changes in the blood level concentration of various hormones such as oestrogen and progesterone, necessary to enable the fetus to grow in a proper well-balanced environment and to prepare the mother for the birth. Along with weight gain, a female's body is out of balance with their shape taking a different form. Significant musculoskeletal strain can occur as a result of the increase in weight, borne primarily by the lower back and pelvis (Taylor D. et al 1981). At the same time, female hormones and relaxin work to increase joint laxity (Ainsworth S.R. & Aulincino P. 1993). For some females, this may cause pre-existing problems to worsen and may trigger new joint pains and instability. Commonly described symptoms include pelvic girdle pain and sacroiliac joint dysfunction (see page 95 of this book for more information on both) caused by increased instability of the joints, and low back pain caused by increased laxity in the spinal joints and weight gain. Everyone's experience is different however; some find pregnancy debilitating, experiencing symptoms that make walking or weight-bearing difficult or impossible; others report feeling much better during pregnancy, even experiencing a reduction in symptoms (Molloholli M. 2011 / Tinkle B. 2010/ & Hakim A.J. 2013h). The reason for this improvement in some women may be due to hormonal changes that cause

positive psychological mood changes and decreased sensitivity of the muscle pain receptors.

Management

Pacing of activities and getting proper rest to reduce the chance of symptom flare-ups are recommended (Hakim A. J. 2013h). Activities that cause pain should be limited or avoided, but overall, it's important to remain as active as possible. If able, exercise such as walking, swimming or exercise in water is usually recommended (Hakim A. J. 2013h).
Dr Brad Tinkle recommends hypermobile women begin pelvic floor exercises while they are pregnant (Tinkle B. 2010). Physical rehabilitation with support from a therapist may also be required; early and throughout pregnancy. (Hakim A. J. 2013h). If lower back pain or pelvic instability occur, use of a pelvic belt / sacroiliac support may be beneficial.

Informing appropriate medical staff

The midwife allocated will go through the mother-to-be's medical history and concerns at the first meeting. Ensuring the healthcare team are aware of potential problems from the start can help to reduce the risk of them arising, and speed up any necessary interventions (should they occur) so that complications or difficulties for both mother and child may be minimised (Hakim A. J. 2013h). It is advisable to take some information on hEDS/HSD along for reference (see hypermobility.org), as midwives don't know every medical condition. In some cases, referral may be made to an obstetrician who can asses the level of medicalised care needed, and will discuss any concerns there may be regarding birth positioning, anaesthesia, natural delivery versus caesarean etc.

At the time of writing, there are no formal obstetric management guidelines for patients with hEDS/HSD. Instead, management and birth plans should be made on a case-by-case basis, taking into account the diagnosis and severity of EDS, to optimise maternal and neonatal outcomes. '*Careful, collaborative antenatal planning should help reduce risks. Clear documentation of risks, together with birth and care plans, should be used to alert staff on duty when a woman with EDS-H/JHS* [hEDS/HSD] *presents in labour, thus reducing the incidence of complications*' (Molloholli M., Specialty Registrar in Obstetrics and Gynaecology,Horton General Hospital, Oxford Radcliffe Hospitals NHS Trust).

Childbirth complications

Studies to determine how often women with EDS experience obstetric and gynaecologic issues compared with the general population are scarce, with those that are available concentrating mainly on the vascular type.

As discussed in the comments by Dr Hakim on page 101 (based on two recent studies by Sundelin H.E. et al 2016, and Hugon-Rodin et al 2016), maternal and fetal outcomes in those with EDS hypermobile type are, reassuringly, generally good. However, additional risks that should be taken into account when considering delivery are:

- A higher risk of unusually rapid delivery (Charvet P.Y. et al 1991, Lind J. & Wallenburg H.C. 2002 / Sorokin Y. et al 1994 / Castori M.2012 / Hakim A.J. 2016).
- Any perineal trauma caused by tearing or episiotomy may heal more slowly than 'normal' or healing may be impaired (Hakim A. J. 2013h & 2016 HMSA).
- Any surgery required will need to take the possible effect on healing into account. (Molloholli M.2011.)

Earlier research (preceding the 2016 studies by Sundelin and by Hugon-Rodin), such as that by Hermanns-Lê T. et al 2014, had reported complications including increased rates of wound ruptures along surgical incisions and delayed wound healing, the uterus failing to contract after the delivery of the baby, pelvic prolapse, deep venous thrombosis and coccyx dislocation. The chance of perineal trauma either by spontaneous delivery or instrumental delivery, and large tears and tissue disruption leading to fistulas and sphincter dysfunction are described by Molloholli M. (2011). According to Bird H. 2007; Kochharn P.K. 2011, and Hermanns-Lê T. et al 2014 there is an increased risk of postpartum haemorrhage, and any factors likely to cause obstetric haemorrhage (placental abruption or placenta praevia) may be exacerbated by the laxity of the connective tissues.

Ehlers-Danlos National Foundation state: '*In connection with natural delivery, women with EDS have experienced incontinence, weak pelvic floor, prolapse of the uterus, sprained joints of the pelvis, separation of the symphysis pubis (the joint between the two pubic bones in the frontal lower part of the pelvis) and rupture of the rectal musculature*' (12/EDNF.org). In medical terms, these disorders may be considered less severe complications of birth than those seen in other forms of EDS (e.g. vascular type), but they can have a profound effect on life and confidence. Also see pages 95, 101, and 105-109 of this book).

Deciding whether to opt for a vaginal delivery or a caesarean section can be difficult. There is no absolute indication for caesarean section and, indeed, possible delays in wound healing with such a procedure may need to be taken into account (Kochharn P.K. 2011). Castori (2012 & 2012b) does state that in order to minimise the risk of pelvic prolapses, caesarean section should be considered the first choice when a vaginal delivery without episiotomy cannot be anticipated. Over all, however, the benefits and risks are decided through clinical judgement (Hakim A. J. 2013h & 12/EDNF.org).

Positioning

Hypermobile women with unstable hip/pelvic, knee or spinal joints are vulnerable to injury if placed in inappropriate positions during labour or operative delivery (Molloholli M. 2011). When regional or general anaesthesia is effective in eliminating pain, great attention to positioning is required in order to avoid inadvertent joint dislocations or subluxations (Molloholli M.2011 / Kochharn P.K. 2011).

> *'Positioning during delivery should be carefully thought through to make this as comfortable and physically safe as possible. It is worth discussing this with the midwife and trying out positioning for a normal delivery, noting what works. Try to ensure the hips/legs are supported and those with pelvic girdle pain will benefit from delivery positions that do not involve lying on their backs.'*
> (Hakim A. HMSA Website - Jan 2016)

Anaesthesia

Due to what is hypothesized to be a *resistance* to anaesthesia or a *faster rate of anesthesia absorption* through the extracellular matrix of the connective tissue, in some cases those with hEDS/HSD may gain little or no benefit from epidural anaesthesia or from other local painkillers given before the repair of an episiotomy or tear (Hakim A.J. 2013h / Tinkle B. T.2010 / Hakim A. et al 2005 / Kochharn P.K. 2011). The subject of anesthesia is also discussed in Chapter 1 (page 25), Chapter 2 (page 65), and Chapter 3 (page 165).

Dysautonomia and childbirth

Although not specific to hEDS/HSD, a study of women with dysautonomia by Dr. Blair Grubb & Dr Kanjwal et al, found that about 30% of women felt their symptoms worsened after giving birth, while 70% of women reported that their symptoms remained stable (Kanjwal K.K. et al 2009). A later study by the Mayo Clinic found no differences in the autonomic function tests scores in women with a form of dysautonomia called postural orthostatic tachycardia (POTs) who had children, compared to women with POTs who did not (Kimpinski K. 2010). For more information on dysautonomia and POTs in relation to hEDS/HSD, see page 69.

As previously discussed, the effects of taking most medicines, including those for dysautonomia, on the unborn baby, are often unknown and should be discussed with the GP, ideally before conception. Existing dysautonomia should also be discussed with the anaesthetist prior to the birth, as it may trigger exaggerated hypotension during regional or general anaesthesia (Kochharn P.K. 2011 / Jones T.L 2008).

Postpartum and beyond

For those experiencing pain, muscle weakness or subluxations/dislocations etc., the work involved in nursing, lifting, carrying and general handling of an infant can prove difficult, and it may be necessary to enlist some help in the weeks postpartum (Molloholli M.2011 / Hakim A. J. 2013h). Sleep deprivation and physical and emotional stress can cause flare-ups of pain, fatigue and anxiety. These can be hard to manage, especially as women who usually take pain or anxiety mediations may not be able to do so. Women should be encouraged to raise any concerns with their healthcare team. Implementing self management techniques such as; pacing and resting, considering posture when lifting, carrying, pushing prams or buggies, and changing nappies can all reduce fatigue and strain on muscles and joints.

The infant

New mothers, who are themselves affected by hEDS/HSD, are often, understandably, watchful for signs that their child may have inherited the same disorder. According to Dr Alan Hakim, even though hypermobility has a high inheritance, it does not mean the child will necessarily develop any symptoms, either in childhood or later on in life. It is important to note that infants and children are generally more hypermobile and may lose this to some degree as they grow. Even if they remain hypermobile, it does not necessarily mean that they will develop symptoms. The important thing is that, if they do develop any problems, their doctor appreciates that hEDS/HSD is present in the family and recognises that this may explain their symptoms.

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