

## **Surgical and Anesthetic Precautions: Hypermobility Spectrum Disorder (HSD) and Hypermobile Ehlers Danlos Syndrome (hEDS)**

*The instructions below, and the handout on the following page, are constructed from an informal consensus by national experts on hypermobile Ehlers Danlos Syndrome (hEDS), and hypermobility spectrum disorder (HSD). It is unclear whether there is any real distinction between the conditions given these two labels. For patients having surgery who may have one of these conditions, but haven't yet had a formal diagnosis, I recommend that the same precautions be followed. They will do no harm if the patient does not have HSD/hEDS. If they do have HSD/hEDS, then the procedure will be substantially safer if these precautions are followed.*

### **For the patient: how to use this document:**

1. Make 3 copies of the following page.
2. Give one to the doctor who will do the procedure, and *make sure they read it.*
3. Give one copy to the person who will be giving you the anesthetic (anesthesiologist or nurse anesthetist). *Make sure they read it.*
4. If allowed, have one copy *pinned on front of your hospital gown* before the procedure. If it's not allowed, then keep that copy with your personal belongings in hospital as you may need it later.

This should ensure that all the people who need to know about your condition, will do what they need to, to keep you safe during the procedure.

The next page has my phone number and email address on it. You, or your medical providers, are welcome to call me if you have questions about how to manage your HSD/hEDS during or after your procedure.

Keep this file in your computer so you can use it in future – any time you are to have surgery, or just anesthesia for medical tests.

*Alan Spanos*

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For a detailed review, see Chopra P, Bluestein L: Perioperative Care in Patients with Ehlers Danlos Syndromes, open access at [https://www.scirp.org/pdf/ojanes\\_2019123013435518.pdf](https://www.scirp.org/pdf/ojanes_2019123013435518.pdf).

Also see <https://www.theilcfoundation.org/emergency-anesthetic-precautions>.

## Surgical and Anesthetic Precautions: Hypermobility Spectrum Disorder (HSD) and Hypermobile Ehlers Danlos Syndrome (hEDS)

The main feature of HSD/hEDS is **laxity of connective tissue**, including skin, ligaments, blood vessels and nerves. This can cause **potentially fatal problems** for these patients when unconscious, and/or having surgery.

<b>BEWARE THE UNCONSCIOUS PATIENT!</b>	<p><b><i>In the unconscious HSD/hEDS patient, a little force may displace any joint.</i></b></p> <p><b><i>Treat unconscious HSD/hEDS patients with full spinal stabilization</i></b> as if they have a spinal injury. If you don't, then you may cause one!</p> <p><b><i>Use NO traction on limbs.</i></b></p> <p><b><i>Use extreme care with the chest:</i></b> the ribs easily dislocate front or back.</p>
<b>BEWARE THE LARYNGOSCOPE!</b>	<p><b><i>Use extreme gentleness, with minimal, if any, anterior traction on the laryngoscope. The jaw may dislocate</i></b> on one or both sides. Manipulation of the laryngoscope can also damage the cricopharyngeal muscle and its nerves, the esophagus and the cervical spine.</p>
<b>BEWARE NECK MOTION!</b>	<p><b><i>Keep patient's head in neutral position throughout.</i></b> Movement of unstable subcranial joints may cause spinal cord damage during incautious patient handling during anesthesia. Consider a soft collar.</p>
<b>LOCAL ANESTHESIA</b>	<p>HSD/hEDS patients are often resistant to local anesthetics: <b><i>they may need much larger doses than other patients, and these may need to be repeated during a procedure.</i></b> If the patient says they can still feel the area, believe them!</p>
<b>SURGICAL TECHNIQUE</b>	<p>Use about half the normal force when cutting or moving tissues. Cut blood vessels may contract poorly: <b>electrocautery is appropriate.</b> Tissue healing may take at least twice as long as usual. <b>Close layers without tension using slowly-absorbable or non-absorbable sutures.</b> Reinforce them with steri-strips etc. as appropriate.</p>
<b>BLEEDING &amp; BRUISING</b>	<p>These are due to fragile small blood vessels, not an intrinsic blood disorder, so <b>clotting tests are rarely indicated.</b> Be alert for slowly-accumulating deep hematomas.</p>
<b>POST-OPERATIVE PAIN</b>	<p>Painful polyneuropathy is common in HSD/hEDS. Post-operative pain may be more severe and more prolonged than normal. <b>Be liberal with analgesics.</b></p>
<b>CARDIO-VASCULAR INSTABILITY</b>	<p>HSD/hEDS patients are subject to hypotension and/or tachycardia due to low blood volume, and defective venoconstriction. <b>Liberal IV fluids usually can address this.</b></p>
<b>GI DYSFUNCTION</b>	<p>Poor GI motility is routine in HSD/hEDS, and worse after surgery. <b>Minimize constipating agents, and use laxatives pre-emptively.</b> Pro-motility agents may be needed.</p>
<b>CARDIAC RESCUCITATION</b>	<p>Some HSD/hEDS patients have tenderness of the costosternal junctions, due to <b>recurrent subluxations of the costosternal joints.</b> If these patients need cardiac resuscitation, then <b>chest compression should perhaps be avoided,</b> since it could produce a flail chest and even cardiac rupture from the free end of a detached rib.</p>

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