

Surgical and Anesthetic Precautions: Hypermobility Spectrum Disorder (HSD) and Hypermobility 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

**Alan Spanos MD
Clinical Associate
UNC School of Medicine
Chapel Hill, NC
January 2018**

Surgical and Anesthetic Precautions: **Hypermobility Spectrum Disorder (HSD) and** **Hypermobility 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.

BEWARE THE UNCONSCIOUS PATIENT!	<p><i>In the unconscious HSD/hEDS patient, a little force may displace any joint.</i></p> <p><i>Treat unconscious HSD/hEDS patients with full spinal stabilization</i> as if they have a spinal injury. If you don't, then you may cause one!</p> <p><i>Use NO traction on limbs.</i></p> <p><i>Use extreme care with the chest:</i> the ribs easily dislocate front or back.</p>
BEWARE THE LARYNGOSCOPE!	<p><i>Use extreme gentleness, with minimal, if any, anterior traction on the laryngoscope. The jaw may dislocate</i> 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>
BEWARE NECK MOTION!	<p><i>Keep patient's head in neutral position throughout.</i> Movement of unstable subcranial joints may cause spinal cord damage during incautious patient handling during anesthesia. Consider a soft collar.</p>
LOCAL ANESTHESIA	<p>HSD/hEDS patients are often resistant to local anesthetics: <i>they may need much larger doses than other patients, and these may need to be repeated during a procedure.</i> Ropivacaine may work better than lidocaine or bupivacaine.</p>
SURGICAL TECHNIQUE	<p>Use minimal force when cutting or moving tissues. Cut blood vessels may contract poorly: electrocautery is appropriate. Tissue healing may be prolonged. Close layers without tension using slowly-absorbable or non-absorbable sutures. Reinforce them with steri-strips etc. as appropriate.</p>
BLEEDING & BRUISING	<p>These are due to fragile small blood vessels, not an intrinsic blood disorder, so elaborate clotting tests are rarely indicated. Be alert for slowly-accumulating, deep hematomas.</p>
POST-OPERATIVE PAIN	<p>Painful polyneuropathy is common in HSD/hEDS. Post-operative pain may be more severe and more prolonged than normal. Be liberal with analgesics.</p>
CARDIO-VASCULAR INSTABILITY	<p>HSD/hEDS patients are subject to hypotension and/or tachycardia due to low blood volume, and defective venoconstriction. Liberal IV fluids usually can address this.</p>
GI DYSFUNCTION	<p>Poor GI motility is routine in HSD/hEDS, worse after surgery. Minimize constipating agents, and use laxatives pre-emptively. Consider pro-motility agents.</p>
CARDIAC RESCUCITATION	<p>Some HSD/hEDS patients have loose costosternal joints, sometimes palpably displaced. For them, chest compressions could in theory be very dangerous, causing rib detachments, a flail chest and even heart or lung puncture by freed anterior ribs. There is no consensus on whether cardiac resuscitation should include chest compressions in patients with clear evidence of rib displacements.</p>

Alan Spanos, MD, (919) 967-2927, alan.spanos@yahoo.com.

This document is online at www.AlanSpanosMD.com. It was updated July 2018.

For more information, see the Ehlers Danlos Society at ehlers-danlos.com.