Ehlers-Danlos Syndromes
New Zealand

Health Passport

First Name/s	
Last Name	
Date of Birth	
GP/GP Practice	
NHI Number	

Please ensure I take this with me when I leave

Fold

Only include this page if you have a known vascular concern

Red Flags / Emergency

Allergy	Reaction

vEDS – VASCULAR EDS

or known vascular concerns relating to Ehlers-Danlos Syndrome

Arterial rupture or unusual bleeding in a child or young adult. Aorta, other large vessels and small vessels can be involved

All EDS – ANAESTHETICS AND SURGERY

• The use of intranasal DDAVP (desmopressin) will dramatically reduce the risk of bleeding both pre- operatively and in the presence of an acute bleed/haemorrhage (regardless of whether the patient is clinically vascular EDS or not)

• Local anaesthetic – local, regional or epidural has less effect and slower onset in EDS

- May need more anaesthetic and longer wait before beginning procedure

• Surgical issues

- Prone to bleeding
- Problems with tourniquet – compartment syndrome
- Potential for tissue damage and subluxation with positioning issues

Cite : New Zealand Hypermobility and Ehlers-Danlos Syndromes Guideline 2019

Red Flags / Emergency

Allergy	Reaction

All EDS/HSD – ANAESTHETICS AND SURGERY

• The use of intranasal DDAVP (desmopressin) will dramatically reduce the risk of bleeding both pre- operatively and in the presence of an acute bleed/haemorrhage (regardless of whether the patient is clinically vascular EDS or not)

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Acute Management

Intolerances	Reaction

ACUTE EMERGENCIES

• Vascular rupture – appropriate vascular surgery or interventional radiology referral
• Dislocations – appropriate orthopaedic referral
• Acute pain – usual principles
• Bleeding - DDAVP (Desmopressin) intra-nasally is recommended for acute haemorrhage to help stop bleeding

INJURIES – INSTABILITY, SUBLUXATIONS, DISLOCATIONS, TENDON & LIGAMENT

• Each individual injury episode (persisting subluxation or actual dislocation, tendon or ligament strain or tear) should be treated as a new trauma on its own merits and not just passed off as “part of your condition”

More information on page 7+

MY EMERGENCY CONTACT

Name_____

Number_____

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List of medications

Medication	Dose

List of previous surgical procedures

Surgery/Procedure	Year

List of diagnosed conditions

Diagnosis	Diagnosed (Year)

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What are Ehlers-Danlos Syndromes and Hypermobility Spectrum Disorders

"When you cannot connect the issues, think connective tissues"

Ehlers-Danlos syndromes (EDS) are a group of 13 inherited connective tissue disorders.

Each person with an EDS syndrome will have their own unique issues. This often leads to confusion from medical professional, but its important to note that not one person will have the exact same issues as another with an EDS syndrome.

- Varied in how they affect the body and in their genetic causes;
- Characterised by, but not limited to joint hypermobility, skin hyperextensibility and tissue fragility.
- Can cause symptoms throughout the body, requiring medical attention and validation.

Hypermobility spectrum disorder (HSD) is condition that involves joint hypermobility.

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- Can cause symptoms throughout the body, requiring medical attention and validation;
- Diagnosed after all other conditions that cause joint hypermobility, including all EDS types, have been excluded.

A medical diagnosis can help with validation, but also gives patients a pathway of care to proactively looking after themselves, and also helps other medical professionals with this pathway.

About Connective Tissue

Connective tissue is the material in the body that binds the body together, supports, and separates different tissues and organs. Found between other tissues everywhere in the body, it provides strength, structure and flexibility, and helps perform general functions, as well as specialized services. A Connective tissue disorder disrupts these most fundamental processes and how structures of the body hold together, so resulting problems can be widespread, in a wide range of severities and affect areas that might seem to be otherwise unrelated.

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PAIN – INITIAL (ACUTE)

Pain has no proven pathology in EDS, rather pain is believed to be largely caused by repeated cycles of differing types of trauma injuries. Undiagnosed and untreated injuries lead to further musculoskeletal dysfunction and pain as poor compensating strategies are employed to cope with the acute unresolved injury pain. Usually the pain an EDS patient is suffering is not benign and the underlying cause should be diagnosed and treated where possible.

• Usual principles for initial management of acute or chronic pain
  - Usually start with paracetamol and work up as needed
  - There are no specific analgesics proven to have significant advantage in Hypermobility Spectrum Disorder/EDS
• Bracing or splinting can be helpful in the short term
• Hot or cold packs, warm bath, etc.

INJURIES – INITIAL EMERGENCY AND ACUTE (SHORT TERM) STRATEGIES INSTABILITY, SUBLUXATIONS, DISLOCATIONS, TENDON & LIGAMENT

• Each individual injury episode (persisting subluxation or actual dislocation, tendon or ligament strain or tear) should be treated as a new trauma on its own merits and not just passed off as "part of your condition"

• Dislocations cause pain and overstretching of the muscles which in turn cause muscle spasms. Immediate assessment and treatment of a dislocation is important because once muscle spasm occurs, it is very difficult to reduce the joint without anaesthesia. Immediate diagnosis and intervention is important to improve patient outcomes.

• If the joint does not spontaneously reduce, reductions should only be attempted by clinicians experienced in reduction techniques. Treatment is usually closed reduction, as soon as possible, to decrease potential complications, which may include soft tissue injury, articular surface injury, and neurovascular compromise.

• Dislocation - initial reduction
  - Because those with EDS may have severe pain, instability or injury elsewhere in the same region, some typical traction and twisting movements used for reduction may need to be modified to prevent injury elsewhere from the procedure itself, e.g. with shoulder dislocation pulling from the wrist or forearm may injure wrist or elbow. May need to modify hand position and grip. The force needed to reduce may be less than that needed for non-EDS.
  Some patients can "spontaneously reduce"
  - Protect skin – it may be fragile. Use padding if needed.
  - Ligament and tendon injuries may take longer to heal and may recur after relatively less trauma after the first episode
• Subluxations may respond well to gentle manipulation rather than actual reduction
• Splinting and bracing are important after reduction.

SURGERY AND ANAESTHESIA

Clinicians planning surgery or anaesthesia in a patient with EDS: Wiesmann et al: Recommendations for anaesthesia and perioperative management in patients with Ehlers-Danlos Syndrome(s)

• Surgical complications may be increased due to slow healing and potential for bleeding. Appropriate strategies should be planned and discussed in EDS context
• DDAVP (Desmopressin) intra-nasally pre-operatively will reduce the risk of a life-threatening haemorrhage
• Recurrence of prolapse, hernias, etc. after surgery may occur because of the inherently abnormal ligaments
• Some issues with anaesthesia:
  - Unstable neck may be an issue with positioning
  - Slow and suboptimal response to local anaesthetic including epidurals
  - Tourniquet can cause bruising and compartment syndrome
  - Positioning can cause unexpected subluxations including temporomandibular joint during anaesthesia

Ehlers-Danlos Syndromes
New Zealand

More information can be found here:

https://ehlers-danlos.org.nz

Our clinical pathways document can be found here:
https://ehlers-danlos.org.nz/eds-clinical-pathway