
Ways of thinking

Bleeding Disorders: Classification

Blood Vessel Constriction ← **Platelet** Aggregation → **Coagulation** Cascade

Blood Vessel Disorders:

Hereditary
- H.H.telangiectasia
- Marfans sy.

Acquired
- Simple easy bruising
- Aging,Scurvy,
- Drugs - steroids
- Viral infections.

Platelet Disorders:

Function disorder
- Drugs – Aspirin
- Kidney failure: uremia.

Thrombocytopenia:
- Immune - ITP
- Drugs, viral Infection
- Aplastic anemia.
- Chemotherapy.

Coagulation Disorders:

Hereditary:
- Haemophilia A, B
- Von Willebrand's

Acquired:
- Liver disease.
- Drugs - Heparin.
- Inhibitors – immune.
- Blood Transfusion

DIC – Disseminated Intravascular Coagulation → All factor deficiency – Septicemia

Starship Paediatric Bleeding Questionnaire Scoring Key

Symptom	Score					
	-1	0	1	2	3	4
Epistaxis		No or trivial (<5)	>5 or more than 10 mins	Consultation only	Packing or cautery or antifibrinolytic	Blood transfusion or DDAVP or replacement therapy
Cutaneous		No or trivial (<1cm)	>1cm and no trauma	Consultation only		
Bleeding from Minor Wounds		No or trivial (<5)	>5 or more than 5 mins	Consultation only	Surgical haemostasis	Blood transfusion or DDAVP or replacement therapy
Oral Cavity		No	Referred at least once	Consultation only	Surgical haemostasis or antifibrinolytic	Blood transfusion or DDAVP or replacement therapy
Tooth Extraction	No bleeding in at least 2 extractions	None done or no bleeding in 1 extraction	Reported, no consultation	Consultation only	Resuturing or packing	
GI Bleeding		No	Associated with ulcer, portal hypertension, haemorrhoids, angiodysplasia	Spontaneous	Surgical haemostasis, blood transfusion, replacement therapy, DDAVP, antifibrinolytic	Blood transfusion or DDAVP or replacement therapy
Surgery	No bleeding in at least 2 surgeries	Non done or no bleeding in 1 surgery	Reported, no consultation	Consultation only	Surgical haemostasis or antifibrinolytic	Blood transfusion or DDAVP or replacement therapy
Menorrhagia		No	Consultation only	Antifibrinolytics, pill use	D& C, iron therapy, ablation	Blood transfusion or DDAVP or replacement therapy or hysterectomy
Post-Partum Haemorrhage	No bleeding in at least 2 deliveries	None or no bleeding after 1 baby	Consultation only	D&C, iron therapy, antifibrinolytics	Blood transfusion or DDAVP or replacement therapy	Hysterectomy
Muscle Haematomas		Never	Post trauma, no therapy	Spontaneous, no therapy	Spontaneous or traumatic requiring DDAVP or replacement therapy	Spontaneous or traumatic requiring surgical intervention or blood transfusion
Haemarthrosis		Never	Post trauma, no therapy	Spontaneous, no therapy	Spontaneous or traumatic requiring DDAVP or replacement therapy	Spontaneous or traumatic requiring surgical intervention or blood transfusion
CNS Bleeding		Never			Subdural, any intervention	Intracerebral, any intervention
Other		No	Reported	Consultation only	Surgical haemostasis, antifibrinolytic or iron therapy	Blood transfusion, replacement therapy or desmopressin

For Von Willebrand Disease and a Score $\geq$ 2

Sensitivity = 84%
Specificity = 75%
Positive Predictive Value 0.15
Negative Predictive Value 0.99

https://media.starship.org.nz/bleeding-questionnaire/bleedingquestionnaireandscoringdoc.pdf

A bleeding score of $\geq$ 2 has a likelihood ratio of 3.5 (2.1 – 5.4)

Figure A: BEIGHTON SCORE – Assessment tool for hypermobility

1

2

3

4

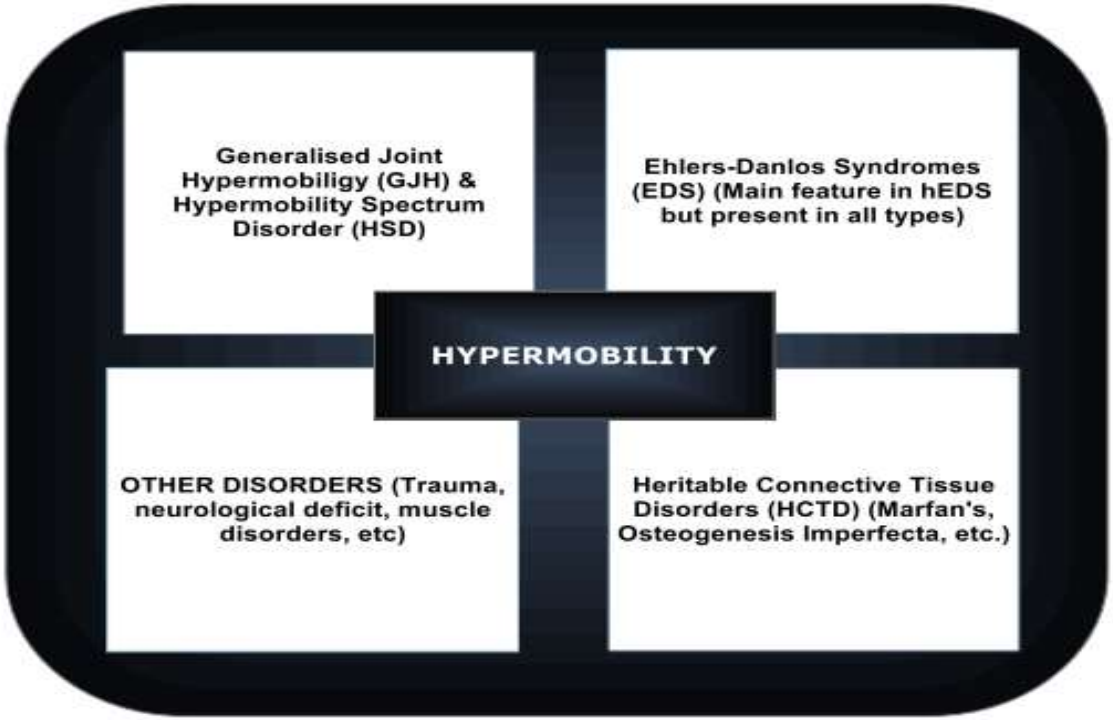
5

1 point for each side for 1-4 and 1 point for 5. Total 9. If ≥ 4/9, hypermobility is present(1).

- Studies show over 60% of adolescent girls and 35% boys have a Beighton score ≥ 4/9, 26% and 11.5% when defined as ≥ 6/9
- Often hypermobility causes no functional problems or pain and can be advantageous for certain activities e.g. sport or music
- Studies show over 30% of school age children complain of regular musculoskeletal pain
- The clinical challenge is to distinguish between those children within the normal spectrum of hypermobility and those with suspected EDS

Understanding HSD/EDS

Hypermobility spectrum disorders (HSD) are a group of conditions related to **Joint Hypermobility (JH).** HSD are diagnosed only after other possible conditions have been excluded, such as **Ehlers Danlos Syndrome(s) (EDS)** including **Hypermobile EDS (hEDS)** and the rarer EDS forms. Individuals with Joint Hypermobility in **5 or more joints** (who do not meet the criteria for EDS) are described as having **Generalised Joint Hypermobility (GJH)** and may still have significant effects on their health.



New Zealand Hypermobility and Ehlers-Danlos Syndromes Guideline 2019

Pediatric joint hypermobility: a diagnostic framework and narrative review

Table 1 Diagnostic framework for pediatric joint hypermobility in the presence of skin abnormalities, musculoskeletal complications, and/or core comorbid conditions

From: Pediatric joint hypermobility: a diagnostic framework and narrative review

	Generalized joint hypermobility	Skin and tissue abnormalities	Musculoskeletal complications	Core comorbidities
Asymptomatic				
Pediatric generalized joint hypermobility	Present	Absent	Absent	Absent
Pediatric generalized joint hypermobility with skin involvement	Present	Present	Absent	Absent
Symptomatic conditions				
Pediatric generalized joint hypermobility with core comorbidities	Present	Absent	Absent	Present
Pediatric generalized joint hypermobility with core comorbidities and with skin involvement	Present	Present	Absent	Present
Pediatric hypermobility spectrum disorder, musculoskeletal subtype	Present	Absent	Present	Absent
Pediatric hypermobility spectrum disorder, musculoskeletal subtype with skin involvement	Present	Present	Present	Absent
Pediatric hypermobility spectrum disorder, systemic subtype	Present	Absent	Present	Present
Pediatric hypermobility spectrum disorder, systemic subtype with skin involvement	Present	Present	Present	Present

Paediatric Working Group
The International Consortium on Ehlers-Danlos Syndromes & Related Disorders

Diagnostic Criteria for Paediatric Joint Hypermobility
This diagnostic checklist is to support doctors to diagnose paediatric joint hypermobility and hypermobility spectrum disorder

Distributed by The Ehlers Danlos Society.

Patient name: _____ DOB: _____ DOV: _____ Evaluator: _____

Children From 5 Years Of Age Until Biological Maturity

L☐R☐ L☐R☐ L☐R☐ L☐R☐ ☐

Beighton Score: ____/9
Must be a minimum of 6

Skin and Tissue Abnormalities

- ☐ Unusually Soft Skin – unusually soft and/or velvety skin
- ☐ Mild Skin extensibility
- ☐ Unexplained striae distensae or rubae at the back, groin, thighs, breasts and/or abdomen without a history of significant gain or loss of body fat or weight
- ☐ Atrophic scarring involving at least 1 site and without the formation of truly papyraceous and/or haemosideric scars as seen in classical EDS
- ☐ Bilateral piezogenic papules in the heel
- ☐ Recurrent hernia, or hernia in more than one site (excludes congenital umbilical hernia)

Score: ____/6
Must be a minimum of 3

Musculoskeletal Complications

- ☐ Episodic Activity related pain not meeting the chronic pain frequency and duration criteria
- ☐ Recurrent joint dislocations, or recurrent subluxations in the absence of trauma, and/or frank joint subluxation on physical exam in more than one joint (excludes radial head <2yrs)
- ☐ Soft tissue injuries – One major (needing surgical repair) and/or current multiple minor tendon, and/or ligament tears

Score: ____/3
Must be a minimum of 2

Co-Morbidities

- ☐ Chronic primary pain
- ☐ Chronic fatigue
- ☐ Functional GI disorders
- ☐ Functional bladder disorders
- ☐ Primary dysautonomia
- ☐ Anxiety

Any number causing distressor disability?
Y / N

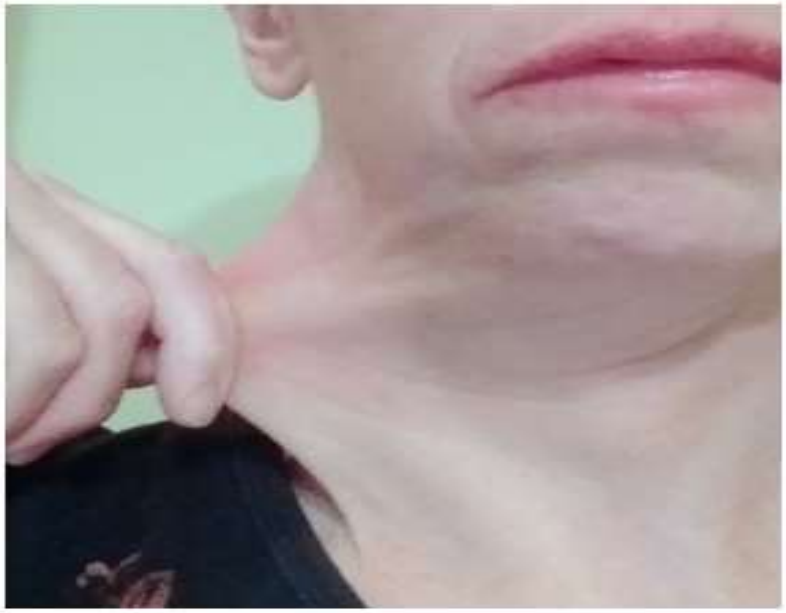
Prerequisites:

1. This framework can only be used after exclusion of other Ehlers Danlos subtypes, heritable disorders of connective tissue, syndromic conditions, chromosomal microdeletions, skeletal dysplasia's, or neuromuscular disorders. From biological maturity or the 18th birthday, whichever is earlier, the 2017 Adult criteria should be used.

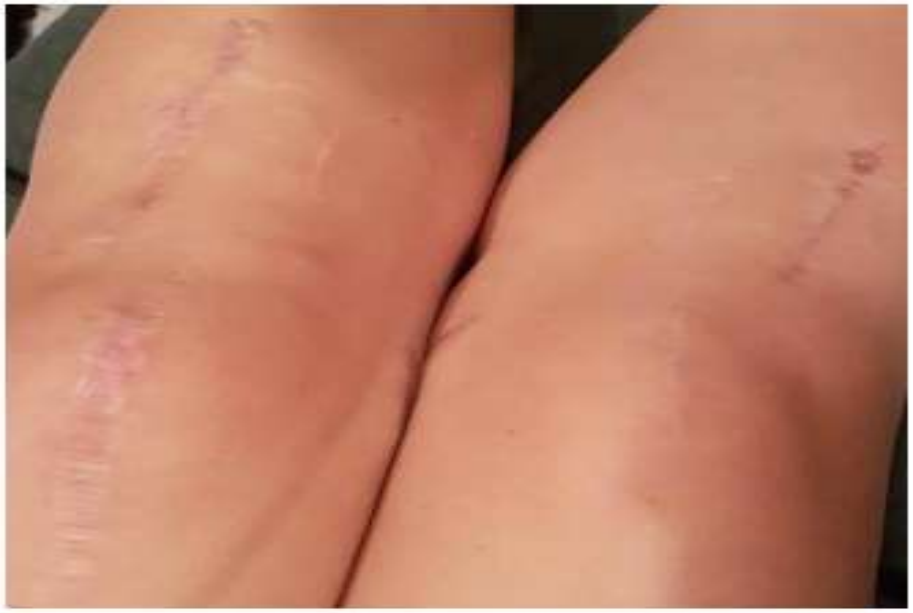
2. No genetic cause for hEDS has been identified at the time of publication of the checklist. In the future disease-causing genetic mutations may be identified in hEDS. In that scenario, if a child has a biological parent with an hEDS diagnosis and a confirmed disease-causing genetic mutation and the child also has the same mutation with GJH then the hEDS diagnosis should be used.

Examination

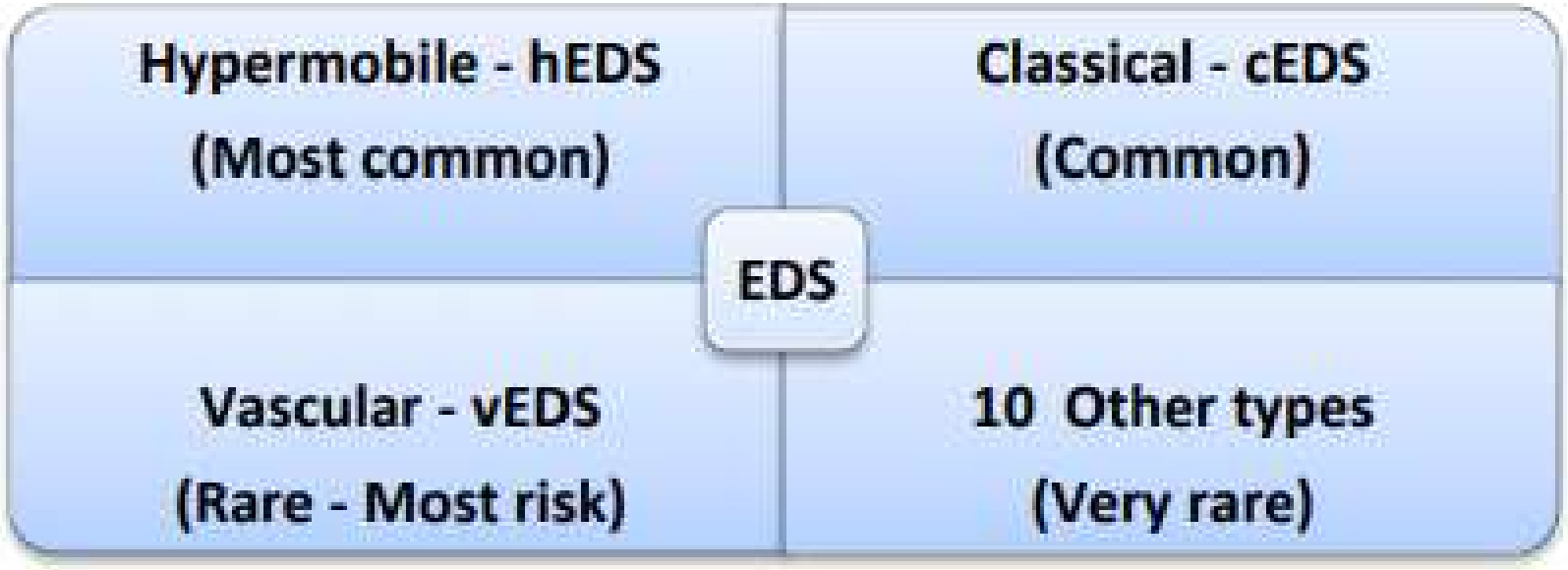
Figure B:
SKIN - HYPEREXTENSIBILITY



SKIN - SCARRING



Diagnosis - EDS



New Zealand Hypermobility and Ehlers-Danlos Syndromes Guideline 2019

cEDS

cEDS – CLASSICAL EDS (1,13)

- Relatively common
- **Major criteria**
  - Skin features - hyperextensible skin, atrophic scarring (esp. knees & elbows)
  - Generalised Joint Hypermobility
- **Minor criteria**
  - Easy bruising
  - Soft, doughy skin
  - Skin fragility
  - Molluscoid pseudotumours
  - Subcutaneous spheroids
  - Hernia or history of
  - Epicanthal folds
  - Complications of GJH
  - Family history of 1st degree relative

To diagnose cEDS:

- Criterion 1 – Skin features

Plus

- Criterion 2 – GJH &/or at least 3 minor criteria

Diagnostic confirmation with genetic testing is possible