Eating problems in the hypermobile Ehlers-Danlos syndrome (a.k.a. Joint hypermobility syndrome)

Problemas alimentarios en el síndrome de Ehlers-Danlos hipermovil (o síndrome de hiperlaxitud articular)

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No conflicts of interest to disclose
Sin conflictos de interes a declarar
Pas de conflit d’intérêt à déclarer
Clinical observations in the hypermobile Ehlers-Danlos syndrome (hEDS)

• A subgroup of patients with hEDS is underweight
• Suspicion or diagnosis of Anorexia Nervosa (AN)
• Eating-related complaints
**Clinical observations in hEDS:**

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- Suspicion or diagnosis of Anorexia Nervosa (AN)
- Eating-related complaints

**Research: very few studies...**

**Hasija et al. (2008):** Joint hypermobility is associated to malnutrition in children. Malnourished hypermobile children have more musculoskeletal symptoms than non-hypermobile children.

**Sanjay et al. (2013):** High percentage (57%) of healthy hypermobile children were underweight. Significant negative correlation between hypermobility and Body Mass Index (BMI).

**Study “P-SED” Hospital Hôtel Dieu Paris** (in preparation):

76 women with hEDS (mean age = 36.7) → 15.7% (n=13) with a BMI < 18.5 (underweight).

Recruitment of control group is currently underway, but underweight in French adults women: 1.8% - 11.4% (HAS, 2010).
Clinical observations in hEDS:

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Research: very few studies...

Zarate et al. 2010: Case report of an hEDS patient with eating difficulties and underweight suspected with AN even if she strongly denied this fact.

Bulbena et al. 2013, 2015:
- Hypermobile secondary school students reported anorexic experiences to a greater extent than those who were non-hypermobile.
- Model “Neuroconnective phenotype” for hypermobility + anxiety: Ergotropic behaviors, such as decreased appetite and weight but increased activity and over control (features linked to AN).

Goh et al. 2014:
- Hypermobility was significantly more common in patients with AN than in controls.
- A connective tissue disorder could cause an eating disorder.
Clinical observations in hEDS:
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Research: very few studies...

Berglund & Björck, 2012:
«The most statistically significant differences between the subjects with EDS and the comparison group were found for OHIP (Oral Health Impact Profile), items 3, 4, and 8»:

"I have had pain in the mouth",
"I have had discomfort when eating",
"I have been forced to interrupt meals"
<table>
<thead>
<tr>
<th>MUSCULOSKELETAL</th>
<th>NON-MUSCULOSKELETAL</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ankle sprain</td>
<td><strong>Cardiovascular:</strong> Low progressive aortic root dilatation, Pseudo-Raynaud's phenomenon, Mitral valve prolapse, Varicose veins.</td>
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<td>Arthralgias</td>
<td><strong>Dental:</strong> Dental neuralgia, Gingivitis, Temporomandibular joint pain, dental pains to cold/warm.</td>
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<td>Bursitis</td>
<td><strong>Gastrointestinal:</strong> Abdominal pain, Bloating, Bowel disturbance, Dysphagia, Food intolerances, Gastritis, Nausea, Reflux gastroesophageal.</td>
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<tr>
<td>Cramps</td>
<td><strong>Mucocutaneous:</strong> Gingival inflammation/recessions, Atrophic scars, Easy bruising, Hernias, Hyperextensible skin (mild), Hypoplastic lingual frenulum, Keratosis pilaris, Light blue sclerae, Resistance to local anaesthetic drugs, Soft skin texture. Striae rubrae and/or distensae in young age.</td>
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<tr>
<td>Carpal tunnel syndrome</td>
<td><strong>Neuropsychiatric:</strong> Anxiety, Cognitive impairment, Depression, Dysautonomia, Dysequisa, Enhanced interoception, Hyperesthesia, Hyperosmia, Hyperacusis, Fatigue, Headache, Poor sleep, Proprioception dysfunction, Somatosensory amplification.</td>
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<tr>
<td>Deviations of knee axes (genu varum and genu valgum)</td>
<td><strong>Ocular:</strong> Myopia, Palpebral ptosis, Strabismus.</td>
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<tr>
<td>Dislocation of joints</td>
<td><strong>Urogynaecological:</strong> Dysmenorrhea, Dyspareunia, Urinary stress incontinence, Meno/metrorrhagia, Vaginal and uterine prolapses.</td>
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<tr>
<td>Dystonia</td>
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<td>Epicondylitis</td>
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<td>Flat feet</td>
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<td>Fibromyalgia</td>
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<td>Hypotonia</td>
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<td>Mechanical back pain</td>
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<td>Mild scoliosis</td>
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<td>Osteoarthritis</td>
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<td>Recurrent luxations and subluxations</td>
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<td>Temporomandibular joint dysfunction</td>
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<td>Tendonitis</td>
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<td>Torticollis</td>
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<tr>
<td>Spinal anomalies</td>
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<tr>
<td><strong>GASTROINTESTINAL (GI) PROBLEMS</strong></td>
<td>dysphagia, gastroesophageal reflux, abdominal bloating and pain, constipation/diarrhea, irritable bowel syndrome, nausea, etc.</td>
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<tr>
<td><strong>TEMPOROMANDIBULAR DISTURBANCES</strong></td>
<td>dislocation, pain...</td>
</tr>
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<td>- Altered mastication patterns</td>
<td>(Rodke et al. 2014)</td>
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<td>- Avoidance hard foods</td>
<td>(Rodrigues et al. 2012)</td>
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<td>- Restriction of mandibular opening</td>
<td>(Myiers, 1985)</td>
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<td><strong>HYPERALGESIA, ENHANCED INTEROCEPTION, SOMATOSENSORY AMPLIFICATION</strong></td>
<td>influence the perception/tolerance to pain.</td>
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<td><strong>SMELL AND TASTE ABNORMALITIES</strong></td>
<td>hyperosmia, dysgeusia.</td>
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<td>- Decrease food acceptability. Nutritional problems and weight changes</td>
<td>(Mattes et al. 1990; Hamonet et al. 2014)</td>
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<tr>
<td><strong>DENTAL PROBLEMS AND ORAL MUCOSA FRAGILITY</strong></td>
<td>dental pain to cold/warm, caries, reduced tolerance to some food textures and temperatures</td>
</tr>
<tr>
<td><strong>FOOD ALLERGIES AND INTOLERANCES</strong></td>
<td>allergies (milk, egg, wheat and soy), and celiac disease</td>
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Illustrative case: Miss M, 18 years old, recently diagnosed with hEDS

Diagnostic criteria (Brighton et Villefranche criteria):
• Beighton score for hypermobility 7/9 (mother and sister also present hypermobility)
• Chronic pain (arthralgias, myalgias)
• Thin and mildly hyperextensible skin
• Recurrent dislocations (including the temporomandibular joint)

Other manifestations of the hEDS spectrum:
• Easy bruising, dysautonomia, chronic fatigue, sleep problems, respiratory problems, food intolerances, GI problems (constipation, abdominal pain, bloating, dysphagia, gastroesophageal reflux, nauseas).

“\textit{I can’t even swallow my own saliva}”
“\textit{I feel when small pieces of food go down my throat}”
“\textit{Chewing hurts my jaw and face}”.
Illustrative case: Miss M, 18 years old, recently diagnosed with hEDS

Current situation:

- Miss M lives alone. Her level of autonomy fluctuates. High level of school absenteeism.
- She often uses a stick to walk.
- She regularly consumes cannabis to relieve pain.
- BMI = 15.8 «severely underweight». Her entourage suspected AN. She deny want to lose weight.
- high food selectivity, eating avoidance
  - she eats very slowly and sparingly
  - eats small quantities
  - cuts food into tiny pieces...

Related to hEDS manifestations
Cas clinique: Exploration du trouble alimentaire comorbide au SED-H (suspicion d'AM):

- Contour Drawing Rating Scale (body image assessment) (Thompson et al. 2009)
  - Proper perception of her body image.
  - Dissatisfied with her body but with a positive magnitude (i.e. she wants to increase her body size)

Clinical interview:

- Intense fears and irrational thoughts about food
  - “Most foods and textures are dangerous to me”
  - “If I eat a hamburger my jaw gets dislocated”
  - “I’m afraid my stomach can crack”

- Focalization on food
  - She thinks about meals several hours per day. Every meal and shopping at the supermarket last for hours.

- Eating problems lead to avoidance behaviors in the social sphere
  - She is ashamed of her "slow and complicated" way to eat.
  - The fear to be nauseous, to vomit or bloating in front of others makes she avoid restaurants and family meals.

- Meals are no longer associated with a source of pleasure.

Anorexia nervosa?

- Mini-DSM-IV (Sheehan et al. 1980)
  - AN (-)
  - Social Phobia (specific) (+)
Diagram illustrating possible relationships between some features or common co-occurring problems in hEDS might contributing to eating difficulty and weight loss.

- Food allergies and intolerances
- Gastrointestinal symptoms
- Enhanced interoception
- Hyperalgesia
- Somatosensory amplification
- Temporomandibular disturbances
- Dental problems
- Oral mucosa fragility
- Smell and taste abnormalities
- Propioception impairment
- Visible signs of illness (ex. orthosis)

Difficulty/painful eating and/or digestion
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Fear of pain

Eating avoidance and/or
Food selectivity and/or
Dysfunctional eating behaviors (e.g. slower, in smaller pieces...)


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**Distorted body schema/image**
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  - Visible signs of illness (ex. orthosis)

- **Worsening hEDS physical symptoms**
  - Difficulty/painful eating and/or digestion
  - Fear of pain

- **Eating avoidance and/or**
  - Food selectivity and/or
  - Dysfunctional eating behaviors (e.g. slower, in smaller pieces...)

- **Weight loss and/or**
  - Nutritional problems and/or
  - Eating disorders (e.g. anorexia nervosa)

- **Distorted body schema/image**

_Baeza-Velasco, Van den Bossche, Grossin & Hamonet, 2016_
Conclusion

• Features and common co-occurring problems of hEDS may favor difficulty eating, significant weight loss and even eating disorders such as AN with consequent poor nutrition.

• The relationship between eating problems and hEDS merits more clinical and research attention.
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<th>Gracias</th>
<th>Merci</th>
<th>Thanks</th>
<th>Obrigado</th>
<th>Grazie</th>
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<td><a href="mailto:carolina.baeza-velasco@parisdescartes.fr">carolina.baeza-velasco@parisdescartes.fr</a></td>
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