Contribution to an adapted physiotherapy in the hyperlaxity syndromes.

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What is hyperlaxity ? (1)

- **Increased articular range of motion** (encountered in individuals of same age, sex and ethnic group)
- **Familial Simple Joint Hypermobility Syndrome** (5 to 10% of the Caucasian population)

What is hyperlaxity ? (2)

- **Hereditary dystrophies** with abnormal **collagen** structure or metabolism (EDS, MFS, Larsen syndrome, Desbuquois syndrome, OI, Cutis Laxa)

- **Acromegalia**, **hyperparathyroidism**, **chronic alcoholism** and of specific **training** (dance, gymnastic…)

Beighton scale

Beighton & Horan, 1969



Scale of Brighton (1)

Grahame et al, 1992
Grahame et al, 2000

- ***Major criteria:***
  - ✓ A Beighton score of 4 out of 9 or greater
  - ✓ Arthralgia for longer than 3 months in four or more joints



Scale of Brighton (2)

Grahame et al, 1992
Grahame et al, 2000

- ***Minor criteria:***
  - ✓ A Beighton score of 1, 2 or 3 out of 9
  - ✓ Arthralgia in one to three joints or back pain or spondylosis, spondylolysis, spondylolisthesis
  - ✓ Dislocation in more than one joint or in one joint on more than one occasion
  - ✓ Three or more soft tissue lesions (e.g. epicondylitis, tenosynovitis, bursitis)
  - ✓ Marfanoid habitus
  - ✓ Skin : striae or hyperextensibility or thin skin or abnormal scarring
  - ✓ Eye signs : drooping eyelids or myopia or antimongoloid slant
  - ✓ Varicose vein or hernia or uterine/rectal prolapse
  - ✓ Mitral valve prolapse

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Scale of Brighton (3)

Grahame et al, 1992
Grahame et al, 2000

→ **Hypermobility syndrome** if **:**

- **2 major criteria**
- **1 major criterion + 2 minor criteria**
- **4 minor criteria**

Advantages



Inconvenients

- **Less stable articulations**
- Recurrent **subluxations**
- Frequent **enthesopathies**
- **Abnormal fragility** to traumatisms
- **Peripheral nerve lesions**
- **Chronic pain** (shoulders, hands, hips, knees, rachis)

→ Many pharmacological and physical treatments are **unhelpful** !



Case 1 : MFS (1)

- **Girl**, **19** years
- **182** cm, **56** kg
- **MFS** detected at the age of 4 + family **antecedent**



Case 1 : MFS (2)

- **Symptomatology :**
  - ✓ **Marfanoid habitus**
  - ✓ **Scoliosis**
  - ✓ **Kyphosis**
  - ✓ **Arachnodactyly**
  - ✓ **Polyarthralgy** (wrists, shoulders)
  - ✓ **Dorso-lumbalgy**
  - ✓ **Subluxation of the ankles**
  - ✓ **Tendinitis of left cubital posterior**
  - ✓ **Mitral valve prolapse**
  - ✓ **Myopia**

Case 1 : MFS (3)

Case 1 : MFS (4)

- **Treatment :**
  - **Postural correction**
  - **Reinforcement concentric** of the **abdominal** and **paravertebral** muscles on the **convex side** of the scoliosis
  - **Proprioceptive exercises** avoiding carefully stretching even autostretching
  - **Home individualized exercises**

Case 1 : MFS (5)

- **Results :**
  - ✓ Rachis **less painful**
  - ✓ **Partial correction** of the cyphosis and the forwards bending of the shoulders
  - ✓ **Stronger** abdominal **muscles**
  - ✓ **Improvement** of the **quality** of life

Case 1 : MFS (5)

- MFS specificity of kinesitherapy :
  - **Strechings avoided**
  - **Adapted muscular training** (cardiac insufficiency)

Case 2 : EDS III (1)

- **Girl**, **16** years
- **170** cm, **55** kg
- Clinical hyperlaxity wich can correspond to **EDS III** (immuno-histochemical but not genetically) + family **antecedents** of hyperlaxity

Case 2 : EDS III (2)

- **Symptomatology :**
  - ✓ **Pains** in the right **elbow** and **wrist**
  - ✓ Repetitive **knees** and **ankle sprains** and **subluxations**
  - ✓ **Epistaxis**
  - ✓ **Myopia**

Case 2 : EDS III (3)

Case 2 : EDS III (4)

- **Treatment :**
  - Wrist and elbow **muscles reinforcement** and **proprioceptive training** in order to **restrict the articular range of motion**
  - **Isokinetic device** to protect the muscles for excessive load
  - **Orthesis** when playing tennis
  - **Home individualized exercises**



Case 2 : EDS III (5)

- **Results :**
  - ✓ **Decrease** in **pain**
  - ✓ **Increase** in the **stability** of her right upper limb even when she plays tennis
  - ✓ **Isokinetic evaluation** objectivizes an **improvement of maximal moment** of 20 to 25 % in all trained muscles

Case 2 : EDS III (6)



Case 2 : EDS III (7)

- EDS specificity of kinesitherapy :
  - To **limit the articular amplitude** → semiflexible **orthosis**
  - **Stretchings avoided**
  - **Concentric** muscular **reinforcement**
  - **Isokinetic** device



Case 3 : OI (1)

- **Girl**, **13** years
- **163** cm, **62** kg
- Clinical diagnosis of **OI** at the age of 11 (no genetic confirmation), family **antecedents** of multiples fractures and osteoporosis



Case 3 : OI (2)

- **Symptomatology :**
  - ✓ Multiples **fractures** (forearm x2, left foot, right scaphoid)
  - ✓ Multiples **luxations** (patella, mandible, left wrist)
  - ✓ Repetitive **sprains** of the **wrists** and **ankles**
  - ✓ **Osteoporosis**
  - ✓ **Strabismus**
  - ✓ **Epistaxis**
  - ✓ **Heamatoma**
  - ✓ **Slow cicatrisation**

Case 3 : OI (3)

Case 3 : OI (4)

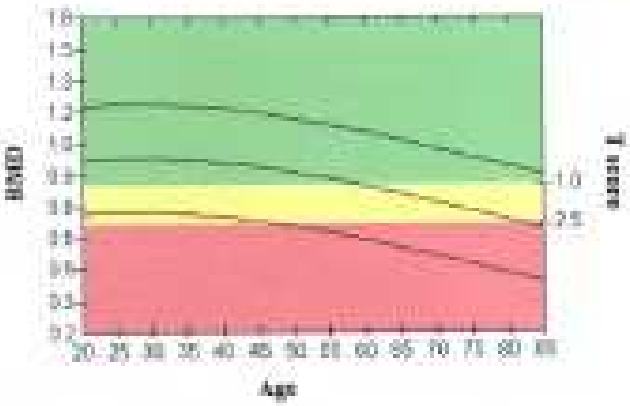
DXA Scan Information:

Scan:	[illegible]
Scan Mode:	Fast Performance
Analysis:	[illegible]
Operator:	RMS
Model:	Hologic QDR-4000 (S/N 15332)
Comment:	

Results Summary:

Total[...] BMD:			0,637 g/cm²		T score:	-2,8
Peak reference:			65%		Z score:	NA
Age matched:			NA			

Region:	Area [cm²]	BMC [g]	BMD [g/cm²]	T score	%PR	Z score	%AM
Neck:	4,86	3,02	0,620	-2,7	69%	0,0	NA
Troch:	7,88	3,82	0,485	-2,6	67%	0,0	NA
Inter:	17,59	12,48	0,709	-1,1	82%	0,0	NA
Total:	30,33	19,32	0,637	-2,8	65%	0,0	NA
Ward's:	1,09	0,68	0,626	-1,5	78%	0,0	NA



Fracture Risk	WHO Classification*
Not increased	Normal
Increased	Osteopenia
High	Osteoporosis

* WHO 1994



Case 3 : OI (5)

- **Treatment :**
  - Mandible **splint**
  - **Proprioceptive reeducation** on instable Freeman plate
  - **Muscular active and activo-passive reinforcement** in physiotherapy
  - **Learned** successfully **to avoid and/or reduce** the **subluxations** of the wrist, mandible and patella
  - **Home individualized exercises**



Case 3 : OI (6)

- **Results :**
  - **Reduction** in the **subluxation**
  - **Reduction** in the **painful** sprain



Case 3 : OI (7)

- OI specificity of kinesitherapy :
  - **Prudence to avoid any fracture**
  - **Avoiding important resistances**
  - **Isokinetism**
  - **Exercises of proprioception**
  - **Osteodensitometric examination**

Guidelines for kinesitherapy in hyperlaxity syndromes



- **Avoid** the **stretchings**
- **Osteodensitometry** before any mobilisation
- **Proprioceptive training** to an adequate limited articular range among patients prone to frequent luxations/subluxations
- Each treatment must be **adapted to individual patient** (cardiac problems, sport practice…)
- **Home individualized exercises**

Thank you
for your attention.

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