Scottish Muscle Network Guidance

The Management of Skeletal Health in Boys with Duchenne Muscular Dystrophy

This guideline is based on consensus between paediatricians managing children with neuromuscular disorders and paediatric endocrinologists with interests in bone disorders.

Aim

The aim of this guideline is to maintain health, mobility and function as much as possible, by promoting bone health, and treating those with pathological fractures.

BONE HEALTH MONITORING

1) **Before starting steroids and annually**

- **Blood Test:**
  Bone profile (Calcium, Phosphate), Alkaline phosphatase, 25 hydroxy vitamin D and Parathyroid hormones (PTH)

- **DXA scan**

- **Lateral thoracolumbar spine x-ray:**
  Performed when starting steroids.

  Consider repeating at regular intervals (e.g., every two years).

  Perform if there is back pain, absent growth, loss of height regardless of previous investigations.

  In centres with the new iDXA scanner, assessment of vertebral morphology using DXA images (DXA lateral vertebral morphometry) may be equivalent to spine x-rays for detection of vertebral compression abnormalities.

- **Vitamin D supplement:**
  Supplement with vitamin D 600-1200IU daily (Depending on local availability).
  Alternatively, if there is poor adherence, consider a stoss dose of vitamin D 150,000 IU six monthly.

- **Diet:**
  Give healthy diet/calcium intake advice leaflet
2) **Growth and pubertal monitoring**

- **Growth:**
  Height and weight every clinic

- **Bone age:**
  Perform in all boys at least once after the age of 13 years.

- **Examination of testicular volume:**
  Regular assessment from the age of 13 years is recommended or jointly in liaison with paediatric endocrinologist.

### MANAGEMENT OF VITAMIN D LEVELS

**(a) Vitamin D < 25 nmol/L**

**Up to 12 years:**

Vitamin D 3000 IU daily (12 weeks), or Vitamin D 150,000 IU stoss.

Consider Sandocal 400 1 tablet b.d. for 4 weeks if serum calcium is low.

**> 12 years:**

Vitamin D 6000 IU daily (12 weeks), or Vitamin D 300,000 IU stoss.

Consider Sandocal 400 2 tablets b.d. for 4 weeks if serum calcium is low.

Repeat vitamin D levels after 3-6 months is recommended

**(b) Vitamin D < 50 nmol/L but ≥ 25 nmol/L**

Increase maintenance Vitamin D supplement by 200-400IU for 12 weeks

Consider stoss dose maintenance therapy if poor adherence.

### Common vitamin D preparations available

<table>
<thead>
<tr>
<th>Preparation</th>
<th>Form</th>
<th>Concentration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abidec (multivits)</td>
<td>liquid</td>
<td>400u/0.6ml</td>
</tr>
<tr>
<td>Dalivit (multivits)</td>
<td>liquid</td>
<td>400u/0.6ml</td>
</tr>
<tr>
<td>Desunin</td>
<td>tablet</td>
<td>800u</td>
</tr>
<tr>
<td>Fultium D3</td>
<td>capsule</td>
<td>800u</td>
</tr>
<tr>
<td>Fultium D3*</td>
<td>drops</td>
<td>2,740u/ml (200u = 3drops)</td>
</tr>
<tr>
<td>Invita D3*</td>
<td>drops</td>
<td>2,400u/ml (67u = 1drop)</td>
</tr>
<tr>
<td>Thorens*</td>
<td>liquid</td>
<td>10,000u/ml</td>
</tr>
<tr>
<td>Invita D3*</td>
<td>liquid</td>
<td>25,000u/ml</td>
</tr>
<tr>
<td>Fultium D3</td>
<td>capsule</td>
<td>20,000u</td>
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</table>
*In infants, children and adolescents Fultium-D₃ Drops, Invita D3 drops/liquid, and Thorens liquid can be mixed with a small amount of children’s foods, yogurt, milk, cheese or other dairy products. The drops/liquid must not be mixed into a bottle of milk or container of soft foods in case the child does not consume the whole portion and consequently does not receive the full dose. Also, it can be mixed with a spoonful or a small amount of cold or lukewarm food immediately before use.

DISCUSS OR REFER TO PAEDIATRIC ENDOCRINOLOGIST WITH AN INTEREST WITH BONE HEALTH IF:

1) **Pathological fracture(s)** (e.g., painful vertebral fracture, painful hip/femur fractures, asymptomatic vertebral fractures, ≥ 3 long bone fractures)

2) **“Abnormal” DXA**

3) **Persistently low vitamin D despite treatment regimen**

4) **Pubertal delay or bone age delay > 3 years (aged 13 years or older)**

5) **Poor growth and/or short stature**

This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined on the basis of all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to guideline recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guideline or any local guidelines derived from it should be fully documented in the patient’s case notes at the time the relevant decision is taken.