

Steroid management during emergency in DMD



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SeCondary Osteoporosis & its Therapy Duchenne Musclerary Dystrophy

SCOT-DMD

**Muscular
Dystrophy UK**
Fighting muscle-wasting conditions



**CHIEF
SCIENTIST
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Scottish Muscle Network



NorthStar
Clinical Network

THE DIAGNOSIS AND MANAGEMENT OF DUCHENNE MUSCULAR DYSTROPHY

A GUIDE FOR FAMILIES

UNITED KINGDOM



Adrenal suppression and crisis are a potentially life-threatening complications of long-term steroid use (See Figure 5). It is important to know that you may be at risk of an adrenal crisis if your steroids are stopped suddenly or if doses are missed because of illness or other reasons for more than 24 hours. All patients taking chronic daily steroids should have a plan in place that outlines what to do in case of missed doses or during times of major illness or severe trauma, such as a car crash, or "stress doses" of steroids may be needed. Information about preventing, recognizing, and managing adrenal insufficiency, as well as when and how to use stress doses of steroids, are included in the 80 Norwich Revised Protocol, which can be found here: <https://www.norwichprotocol.org>

SYMPTOMS OF AN ADRENAL CRISIS

You should should receive information about the signs and symptoms of an adrenal crisis.

- Severe fatigue
- Nausea/vomiting
- Low blood sugar
- Low blood pressure
- Passing out.

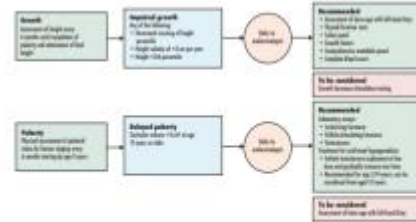


Figure 5. Assessment and Management of DMD and Fabry while taking Corticosteroids

8. BONE HEALTH (OSTEOPOROSIS) MANAGEMENT (Figure 6)

Bone health is important in both the ambulatory and non-ambulatory phases of Duchenne. People living with Duchenne at all ages have weak bones, especially if they are taking steroids. Steroids cause bones to have a lower bone mineral density, increasing the risk of fractures (broken bones) compared to the general population. Muscle weakness and decreased mobility are also risk factors for weak bones.

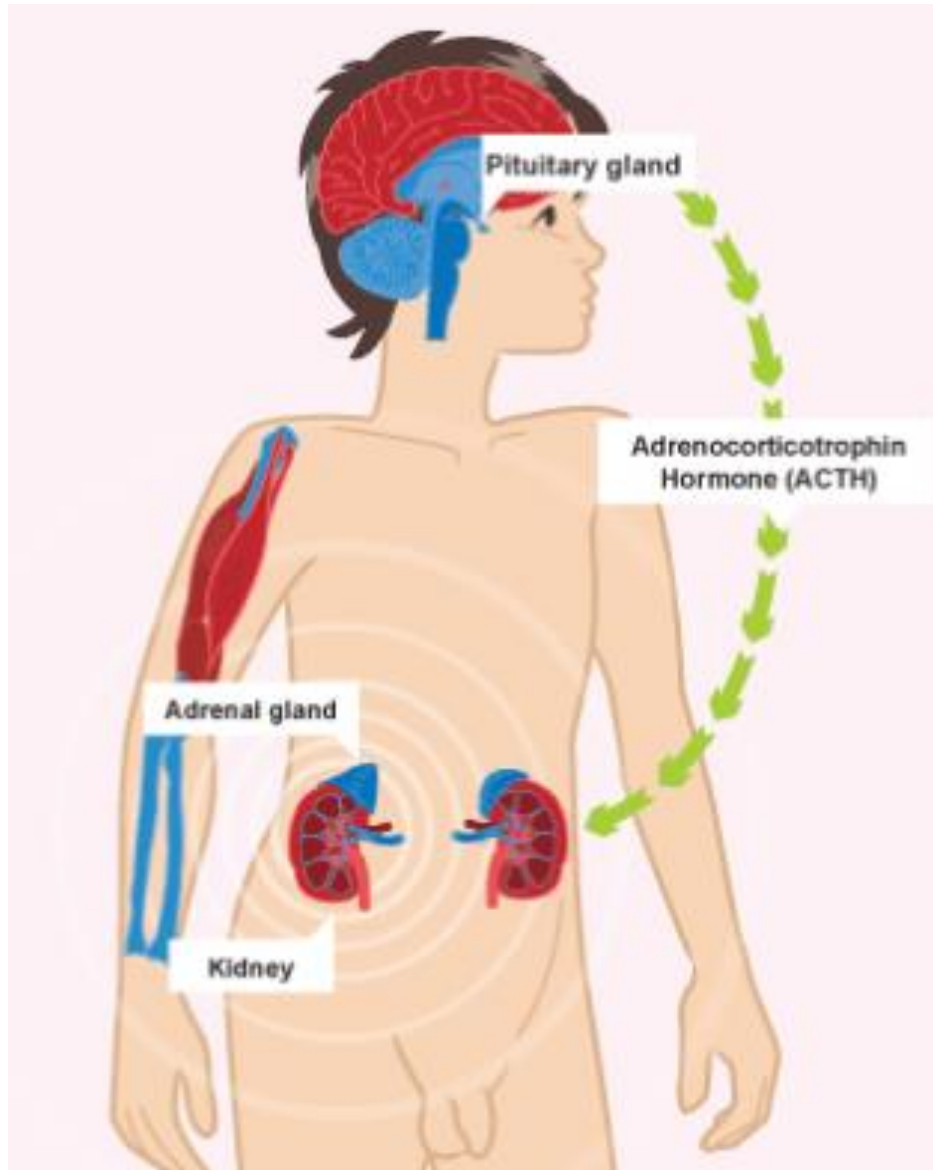
Dual energy X-ray absorptiometry (DEXA) is a non-invasive test that measures bone mineral density of the long bones (usually the leg or arm). Thinner bone is less healthy and is more susceptible to fractures. Measuring your bone mineral density is important in monitoring overall bone health. It is recommended you have DEXA scans at least every 2 to 3 years. People living with Duchenne, especially those taking steroids, are at risk for vertebral compression fractures. Vertebral compression fractures occur when vertebrae (the bones of the spine) sustain small fractures, causing them to become misshapen and to collapse on one another. These fractures, and result in vertebral collapse, can cause pain. Vertebral compression fractures can be seen on a lateral (side view) spinal X-ray, even without having back pain. Vertebral fractures may be treated with bisphosphonates, especially if pain is present. It is recommended you have lateral spine X-rays every 1 to 2 years or more frequently if you experience back pain.



TREAT-NMD website



ADRENAL SUPPRESSION FROM LONG TERM USE OF STEROIDS



The pituitary gland the master regulator of hormone control in the body

Adrenal glands make steroid hormones

Steroid medicines (eg Prednisolone or Deflazacort) are much higher than the amount produced by the body

This leads to switching off of the master regulator (ie ACTH)

During times of mild illness, a healthy adrenal gland makes about twice the amount of steroid to cope with illness

During times of major illness, a healthy adrenal gland makes up to 10-20 times the amount of steroid to cope with illness

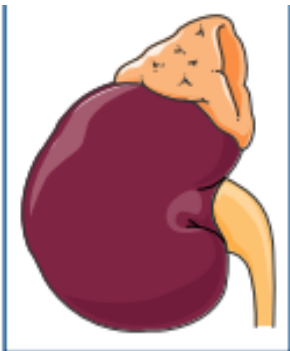
Prescribed doses of Prednisolone or Deflazacort is usually about 4-5 times more than what the adrenal gland would normally produce (ie if able to keep steroid down enough to cope with mild illness)

All patients who take steroids for longer than 6 months have adrenal suppression (sleepy adrenal glands)

Information about

Adrenal Suppression from Long Term Use of Steroid in Duchenne Muscular Dystrophy (DMD)

Access to hydrocortisone as injections to be administered at home during severe illness (when unable to take steroids by mouth)



Intramuscular injection video

Here you will see a video demonstrating an intramuscular injection of hydrocortisone



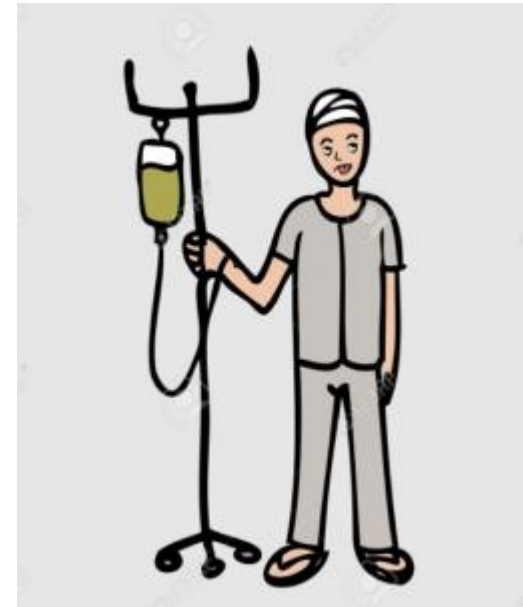
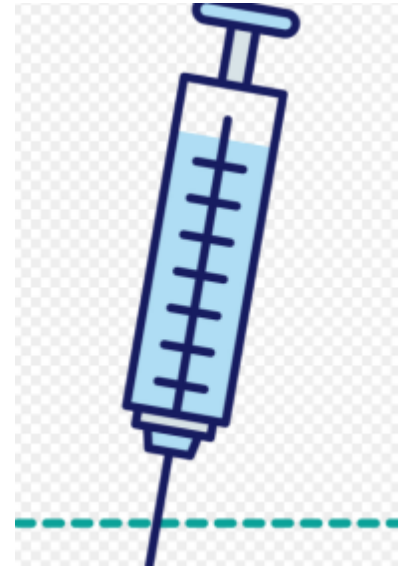
<http://www.smn.scot.nhs.uk/patients-and-families/dmd/>

Have emergency plans in place (steroid card, hospital alerts)

If serious vomiting illness or unable to tolerate oral steroids (eg major operations)

- Need plan for extra steroid to be given either as an injection or as a drip

Not to stop steroid abruptly



1- Adrenal suppression (for those on steroids)

Need emergency steroid plan in place

2- Growth

Regular height monitoring

Growth hormone not routinely recommended

3- Puberty

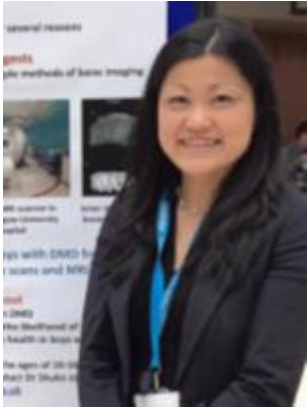
Assessment of puberty

Testosterone from 14 years (if no signs of puberty)

4- Bone

Routine spine imaging

Consideration of bisphosphonate if presence of vertebral fracture



S Joseph



J Dunne



M Di Marco



C Duncanson



I Horrocks



S Shepherd and students



Research radiographers & MRI Physics





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