



**DMD
CARE UK**
Best care for all



i Information Leaflet

Adrenal insufficiency

**from long term use
of steroid in Duchenne
muscular dystrophy (DMD)**



**Duchenne
UK**



NorthStar
Clinical Network

dmdcareuk.org

Why is steroid medicine used in DMD?

Doctors and nurses prescribe steroid medicines for people with DMD as they improve muscle strength, protect the heart and lungs and reduce the chances of developing a severe curved spine (scoliosis).

There are two types of steroid prescribed for people with DMD: Prednisolone or Deflazacort. Steroid medicines are also sometimes prescribed to girls with DMD (manifesting carriers).

Steroid medicines are beneficial in DMD, but there can be side effects. One of the side effects when steroid medicines have been used for more than a month is that they cause the adrenal glands to stop producing their own steroids and become “sleepy” (**adrenal insufficiency**).

What are the adrenal glands and what do they do?

The adrenal glands are small triangular-shaped organs, that sit on top of our kidneys.

They make several important hormones, including cortisol, that control energy, well-being and regulate vital body functions, especially during times of stress, illness and accidents.

During minor illness in healthy people, the adrenal glands make approximately three times more cortisol than usual.

How can I find out more information?

This leaflet provides general information.

Your doctor or nurse will explain more in the clinic. There will be some differences in management plans between hospitals and plans may be personalised depending on your child’s situation.



In the event of an accident, an operation or severe illness, the adrenal glands make about five to ten times more cortisol to help the body react appropriately.

Why are the adrenal glands “sleepy” in people with DMD treated with steroids?

The steroid medicine that your child takes is an artificial version of cortisol.

To treat the muscle problem in DMD, it is given at a much greater dose than the amount normally produced by the adrenal glands. Once the body gets used to such high doses of steroid (after more than a month) the adrenal glands go to “sleep” and stop making cortisol. This is called **adrenal insufficiency**.

What do I need to do with steroid medicines during a mild-moderate illness?

For mild to moderate illness (e.g. flu-like illness or any acute illness with high fever), people with adrenal insufficiency need more steroid than usual. This is called a **stress dose**.

Your medical team will agree a stress dosing plan with you, and you should be given a copy of this to keep at home.

You should always tell your medical team after you give your child a stress dose of steroid.

What happens during a severe illness or when my child cannot take steroid medicines by mouth?

The situations when adrenal insufficiency can be a major problem for your child include:

- (a) Severe illness (especially with vomiting and/or diarrhoea or excessive sleepiness).
- (b) Major stress like a broken bone with a lot of pain.

In these situations, your child will need to be given steroid as an injection of hydrocortisone before attending the hospital. Without extra steroid, the body will not cope with the stress and it can be life threatening.

What do I do if my child needs a hydrocortisone injection?

Your child needs to be given an injection of steroid medicine (hydrocortisone) urgently, regardless of the type of steroid or dose pattern they usually receive.

Your nurse or doctor may have taught you how to give the injection, or told you to give it to the ambulance crew to administer. When you phone 999, let them know that your child has DMD, has adrenal insufficiency and is at risk of an **ADRENAL CRISIS**.

After being given a hydrocortisone injection, your child should attend hospital as soon as possible for a medical review.

What needs to happen if my child is having an operation?

If your child is having an operation (whether it is minor or major), the doctor should give an extra dose of steroid through a drip into the vein at the start of the operation. Steroid should be given through a drip until usual steroid medicine can be taken by mouth. Dental extractions under local anaesthetic do not usually require additional steroid, but your local team can give advice on this.

What happens if we plan to discontinue steroid treatment?

Adrenal insufficiency can be present in anyone who has taken steroid for more than a month.

Please do not stop taking steroid suddenly. If your child plans to stop taking steroid, the medicine should be gradually reduced under the guidance of the treating medical team. Once your child is off steroid treatment, blood tests will be arranged to check that the adrenal glands are making cortisol.

For people on long-term steroids, NorthStar clinicians will regularly check that an oral sick day steroid plan is in place, whether the family has access to home-dose hydrocortisone injection packs, and are trained on how/when to administer this. Ask your clinician if you are unsure about this or have any questions.



DMD CARE UK

Best care for all

DMD Care UK is a nationwide initiative to ensure every person living with Duchenne muscular dystrophy (DMD) in the UK has access to the best care.

This project is funded by Duchenne UK, Joining Jack and the Duchenne Research Fund. They work closely with the John Walton Muscular Dystrophy Research Centre in Newcastle and in collaboration with the North Star Network, funded by MDUK.

DMD Care UK has produced a series of information leaflets for DMD patients, families and other non-specialists on the recommended standards of care for DMD.

Find out more at dmdcareuk.org

Do you have questions or feedback about this leaflet? Get in touch with support@duchenneuk.org

