



DMD CARE UK

BEST CARE FOR ALL



 Information Leaflet

**ADRENAL
INSUFFICIENCY**
**FROM LONG TERM USE
OF STEROID IN DUCHENNE
MUSCULAR DYSTROPHY (DMD)**



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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 steroids prescribed for people with DMD: Prednisolone or Deflazacort. Steroid medicines are also used infrequently in some girls with DMD (manifesting carriers with DMD).

Whilst steroid medicines are beneficial in DMD, 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 maybe personalized depending on your child’s situation.



During major illness (like 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 produced by the adrenal glands of a healthy person. Once the body gets used to such high doses of steroid after more than a month, the adrenal glands go to “sleep” (adrenal insufficiency) and stop making cortisol.

Do I need to do anything 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), more steroids are required in people with adrenal insufficiency.

It is likely that your child will be advised to have extra or increased steroid during illness (stress dose) and the specifics of this will be advised by the treating medical team.

There are many approaches to manage such a situation. It would be useful to inform your medical team if stress dose of steroid is given for mild to moderate illness.

What do I have to do with steroid medicines 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 is very sleepy).
- (b) Major stress like an acute fracture with a lot of pain.

In these situations, there is a need to give steroid as an injection before attending the hospital. Your child could become seriously unwell if not managed promptly with steroid.

What do I need to do in the situations described above?

Steroids need to be given quickly, regardless of the type of steroid/regimen that your child usually receives.

Without extra steroid, the body will not cope with the stress and it can be life threatening. Your child should be given an injection of steroid medicine (hydrocortisone) urgently.

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 as a result is at risk of an **ADRENAL CRISIS**. Anyone who has been given a hydrocortisone injection should attend hospital as soon as possible for a medical review.

Is there a need to do anything if my child is having an operation?

If your child is having an operation (whether it is a minor or major operation), 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 cover, 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 have 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 to check that the adrenal glands are making cortisol will be arranged.

DMD Care UK is a nationwide initiative to ensure every person living with Duchenne in the UK has access to the best care. DMD Care UK is funded by Duchenne UK and Joining Jack. They work closely with the John Walton Muscular Dystrophy Research Centre in Newcastle and in collaboration with the North Star Network, funded by MDUK. Find out more at dmdcareuk.org

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