

Q+A from 10th Jan 2023: Clinician focus

Bone and Endocrine, Dr Jarod Wong

Question 1: Are you aware of difficulties having the recommended home-hydrocortisone kit provided. Should they be provided by the GP or the neuromuscular specialist?

There are regional differences in who is responsible for provision of kits. In some areas this has to be provided on an on-going basis by the hospital but in others, this can be on repeat prescription with the GP. It is important that the hydrocortisone is replaced when used or if it becomes out of date; and we hope that this is an item that can be incorporated into the NorthStar clinic review proforma.

Training of families to give emergency hydrocortisone injections at home should be strongly encouraged; Some families will never feel confident in doing the injections themselves and others are very capable of doing so, after training. The stress dose survey showed that families who have been trained are more confident and less anxious, highlighting the importance of training where possible. Staff-time to deliver the initial training of injection and updating of training is an issue in some sites. DMD Care UK are looking into a solution for this issue in terms of education and training in this important area by the development of centralised educational resources.

Supplying the hydrocortisone packs to families (without training) for emergency staff (ambulance or A&E staff) to administer is a potential solution. Note that

there are examples where families have had packs and prescription letters to hand to paramedics or A&E doctors, but they still had not been administered. We are working to raise awareness amongst clinical teams, alongside empowering patients and their families to be confident in advocating for themselves in an emergency.

‘Who does what’ (neuromuscular vs endocrine) needs to be decided locally, depending on the resources available. Group or online training (including hydrocortisone injections) to families is an option, and has been conducted in some sites

Question 2: Do families worry about having the kits at home?

Patient organisations are not aware of families not wanting to be provided with hydrocortisone kits. Ideally, they do want training in their use. The main issues that families raise is that some sites give hydrocortisone kits and others do not – this causes frustration. The 2018 international standards of care and DMD Care UK endocrine and bone working group guidance recommends that they should be at least *offered* by all sites.

Adrenal insufficiency information (eg DMD Care UK adrenal insufficiency leaflet) should also be made available to families. Doing this raises the issue of adrenal crisis with families and what to be aware of so that there is greater understanding of the issue.

Some clinicians did report that some families do not want packs at home; and some do not want to use them themselves but would hand them to emergency medical staff. This is felt to be an uncommon situation and should be judged individually, on clinical merits.

Question 3: There is a request from families (via the DMD Care UK survey) for long-term steroid users to be seen regularly in an endocrine clinic. How are we approaching this?

A large number (approximately 50-60%) of boys with DMD on long term steroids have active bone/endocrine issues. Sites need to consider this between teams. Some options include

- 1- Reviewing all boys on steroids in the endocrine clinic “early” on (eg first few years of steroids) and then again in 2-3 years’ time, unless clinical issues arise meanwhile. This model is undertaken by other specialties, e.g. respiratory in some sites.
- 2- Regular online MDT discussions between neuromuscular and endocrine/bone teams could also be a model to triage patients that need to be seen in the endocrine clinic.

Question 4: Is the steroid stress dosing webinar still available?

Yes – on the DMD Care UK website: <https://dmdcareuk.org/webinars-%26-videos>