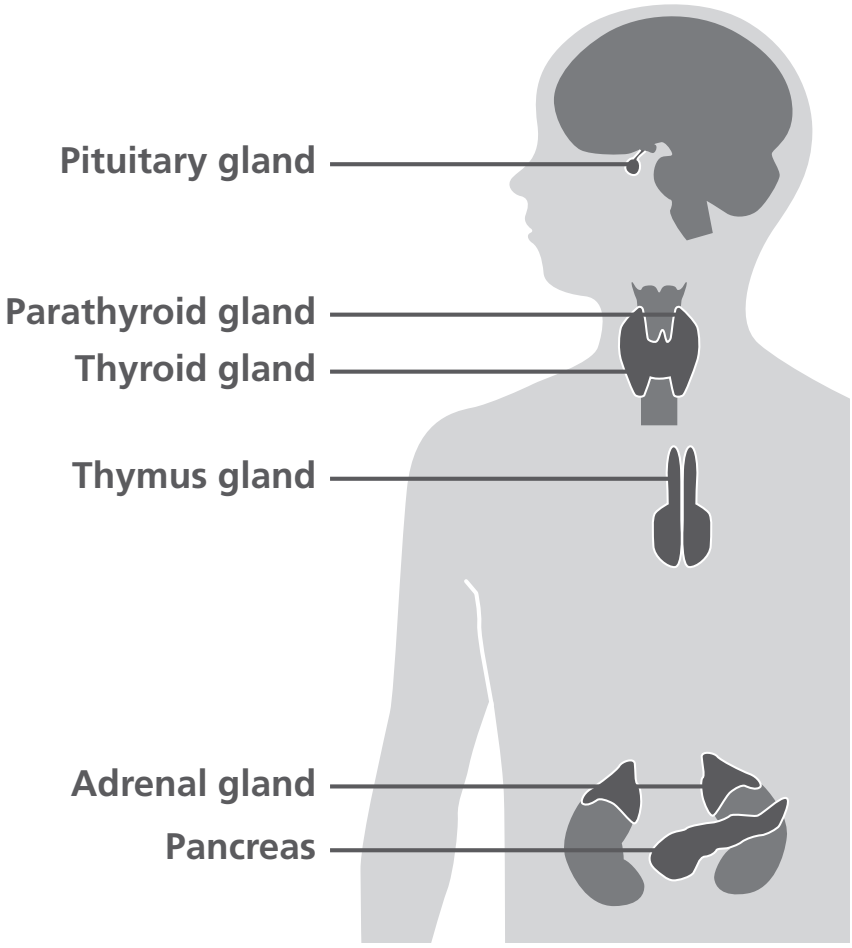
Great Ormond Street Hospital for Children NHS Foundation Trust: Information for Families and Professionals

Emergency steroid management plan for boys with Duchenne muscular dystrophy (DMD)

Many boys with Duchenne muscular dystrophy (DMD) are being treated with steroid medication – either prednisolone or deflazacort. When these medications have been used on a long term basis, boys can develop adrenal insufficiency. Adrenal insufficiency occurs when steroid medication stops the body's natural production of steroid hormones called corticotrophin-releasing hormones and adrenocorticotrophin-releasing hormones from the hypothalamus and pituitary glands. This then reduces release of natural steroid hormones from the adrenal gland. This is called adrenal suppression.



Adrenal suppression means that when your child

- Is Ill
- Is having an operation
- Has a serious accident

They may not be able to mount the usual 'stress' response in the body by increasing the amount of steroid hormones circulating in the blood. This can result in your child becoming very unwell, very quickly. This effect can continue for up to a year after stopping steroid medication.

If at any stage, you become concerned about your child, please use your fast track access to your local children's ward or take your child to the nearest Accident and Emergency Department. Always state that your child is on long-term steroid medication and take this information sheet with you to show the medical and nursing teams.

 Ref: 2014F1574

What to do if your child becomes unwell

Trivial illness, such as a mild cold

- No extra steroid required.

Significant illness, temperature 38°C or above but steroids can still be taken by mouth

- Increase your child's steroid medication to twice daily doses 12 hours apart instead of once daily
- When your child is well again, go back to the once daily dose of steroid medication

Diarrhoea and vomiting (D&V) illness and is unable to take the dose at anytime

- Use your fast track access to your local children's ward or take your child to the nearest Accident and Emergency Department straight away
- Your child will need a steroid injection

Planned or emergency operations or serious accidents

- Your child will need extra steroid medication to be given.
- Always tell doctors that your child is taking long term steroid medication.

Information for medical professionals

Signs and symptoms of acute adrenal insufficiency:

- Nausea and vomiting
- Tiredness, weakness, lassitude
- Signs of dehydration
- Tachycardia, poor pulse volume
- Hypotension
- Signs and symptoms of hypoglycaemia

Emergency management

Emergency Management dose:

0-1yrs – Efcortisol® 25mg IM in an emergency and 1/3 tube of Glucogel®

1-5yrs – Efcortisol® 50mg IM in an emergency and 1/3 tube of Glucogel®

5yrs+ - Efcortisol® 100mg IM in an emergency and 1/3 tube of Glucogel®

Doses thereafter should be 2mg/kg IV of hydrocortisone 6 hourly whatever age group.

Hypoglycaemia should be treated with 2ml/kg of 10% dextrose (if needed in hospital after Glucogel® administration)

The steroid regimen will need to be reviewed on a regular basis. Children typically will be able to return to oral medication when they are recovering.

This information sheet was compiled with reference to:
- GOSH clinical guidelines – glucocorticoid treatment 2012
- Action Duchenne – Steroid treatment guideline in Red Emergency Folder
- Children's Hospital at Westmead guideline on steroid management in DMD by Helen Young 2010
- Reviewed by Endocrine team Dr Caroline Brain and Prof Dettani at GOSH

Compiled by the Dubowitz Neuromuscular Centre
in collaboration with the Child and Family Information Group

Great Ormond Street Hospital for Children NHS Foundation Trust
Great Ormond Street
London WC1N 3JH

www.gosh.nhs.uk

 Ref: 2014F1574