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## 1. Introduction

**This guideline applies to those children undergoing surgery but who are on Steroid replacement treatment due to Adrenal conditions which include:**

- Central Adrenal Insufficiency: Hypopituitarism (multiple pituitary hormone deficiencies)
- Primary Adrenal Insufficiency such as: Congenital Adrenal Hyperplasia (CAH), Addison's disease, adrenal hypoplasia congenita, previous history of adrenalectomy and other rare conditions (for example Smith-Lemli –Opitz on hydrocortisone replacement)

**In addition, consider using these guidelines for those children who are at risk of adrenal suppression and adrenal crisis due to long-term steroid treatment such as:**

- patient on long term ( 4 weeks or more continuous use) systemic steroids (e.g.: Prednisolone, Dexamethasone, Budesonide, above licensed dose of steroid inhalers)
- patients who are on a weaning regime of long-term steroid or have weaned off long-term steroid use in the preceding 3 months.
- Regular use of inhaled steroids doses above licensed dose (beclomethasone >800 micrograms/day or fluticasone >400 micrograms/day)

Please discuss with the Paediatric Endocrine Consultant On-call for advice in this group of patients.

## 2. Pre-admission

Children on long-term steroid replacement may need minor (short procedures of up to 30 minutes anaesthetic time) or major (surgery requiring prolonged anaesthetic time of more than 30 minutes or a procedure which is likely to cause post-operative nausea, vomiting or inability to feed adequately) surgery.

Girls with Congenital Adrenal Hyperplasia may require admission for surgery for the following indications:

A) Reconstructive genital surgery, which usually takes place initially at around 2 years of age with the potential for further corrective surgery in the teenage years. Such procedures should be regarded as **major surgery**.

B) Examination under anaesthetic (EUA) may also be required on some occasions. This is a brief procedure which would be regarded as **minor surgery**.

### **Planning admission:**

The following guidance should be followed in planning for such admissions on the surgical ward:

1. Notification of the date of admission to the named Paediatric Endocrine Consultant a few days prior to admission.
2. The patient should preferably be placed first on the surgical list in the morning. Prior liaison with the anaesthetist is essential.
3. Inform the family to ensure the child receives the normal hydrocortisone dose the evening before.
  - **If the child is on the morning list**, ensure the child has their normal morning dose of hydrocortisone and fludrocortisone with the last drink allowed.
  - **If the child is in the afternoon list**, the normal dose of morning hydrocortisone and fludrocortisone should be given and the afternoon dose of hydrocortisone should be given with the last drink allowed.
4. **Please check the up-to-date dose of the child's medication from the family and double check in medical notes/up-to-date clinic letter.** If any queries with the normal doses, please contact the endocrine secretary on 01162587737 so the record on clinical workstation or CITO can be checked for up to date doses.

### **2.1 On the day of the surgery**

#### **Preoperative:**

If there is a delay in going to theatre which involves fasting more than 6 hours, child should be commenced on full maintenance IV fluids (5% dextrose + 0.9 % Sodium Chloride). Check blood glucose when IV fluids commenced and hourly thereafter. Ensure child has received the normal dose of Hydrocortisone in the morning or afternoon depending on the time of the operation as stated in guidance point 3 above.

## Major Surgery

1. The anaesthetic team must administer a stress dose of IV hydrocortisone at induction in anaesthetic room pre-surgery.

### IV hydrocortisone bolus induction dose:

Under 1 year: 25 mg

1-5years: 50mg

Over 5 years: 100mg

2. Commence IV maintenance fluids (5% dextrose + 0.9 % Sodium Chloride) in theatre, if the child has not been on it already.

3. If procedure is expected to exceed 4 hours, a further bolus of IV hydrocortisone (dose as stated above) during the procedure will be required. This will be prescribed and administered by anaesthetic team.

## Postoperative management:

1. Post operatively:

- Continue IV fluids,
- Prescribe IV hydrocortisone at a dose of 100mg/m<sup>2</sup>/day (m<sup>2</sup> - [see Appendix for BSA calculation](#)) with total dose split into 4 equal divided dose given 6 hourly IV
- Check blood glucose 2 hourly until child is eating and drinking.

2. If there are concerns that the child is unstable or needs PICU care, please consider Adrenal crisis

– *follow the Adrenal Crisis management please follow the link:*

[Adrenal Crisis UHL Childrens Medical Guideline](#) or search via the UHL policy & guidelines library.

3. When the child is eating and drinking, stop the IV fluids and IV hydrocortisone and change to oral hydrocortisone which is 30 mg/m<sup>2</sup>/day in 4 equal divided doses (6 hourly). Restart their normal oral fludrocortisone dose that the child is on at home.

4. Reduce the oral hydrocortisone to their 'normal' oral doses at least 2 days after major surgery provided the child has remained well and eating/drinking. *Some children may need a longer course if they remain unwell and so please contact the Endocrine Consultant for advice if there are concerns.*

5. In those children with suspected Adrenal suppression due to long-term steroid treatment for other medical conditions, prescribe hydrocortisone dose of 30mg/m<sup>2</sup>/day divided in four equal doses (m<sup>2</sup> - see [Appendix for BSA calculation](#)) for a 3-5 days course and then stop. Restart their 'normal' steroid treatment. *Contact the Endocrine Consultant on call for advice on the course of length for this regimen and for advice on restarting their 'normal steroid treatment' if necessary.*

## Minor surgery

1. The anaesthetic team will administer stress dose of IV hydrocortisone at induction pre surgery (doses in the table [above](#) in the major surgery section).
2. On return from theatre, give oral hydrocortisone which is 30 mg/m<sup>2</sup>/day in 4 equal divided doses (6 hourly). Restart the normal dose of fludrocortisone, which the child is on at home, the next day or in the post-op period if that day's dose of fludrocortisone was not taken pre-op.
3. If child is unable to tolerate oral fluids 4 hours after theatre, commence IV maintenance fluids (5% dextrose + 0.9 % Sodium Chloride) and IV hydrocortisone as per the 'major surgery postoperative management' stated above. Check blood glucose every 2 hours whilst on IV fluids.

Change over to oral hydrocortisone and fludrocortisone as in 2, above when oral fluids tolerated as stated in the major surgery section.

4. Advise the parents to continue the dose of oral hydrocortisone as in step 2 above for 48 hours and then reduce to their 'normal' dose.
5. In those children with suspected Adrenal suppression due to long-term steroid treatment for other medical conditions, prescribe hydrocortisone dose of 30mg/m<sup>2</sup>/day divided in four equal doses (m<sup>2</sup> - see [Appendix for BSA calculation](#)) for 48 hours and then stop and restart their 'normal' steroid treatment. Contact the Endocrine Consultant on call for advice on restarting their 'normal steroid treatment' if necessary.

### 2.2 Helpful Contact numbers:

- Endocrine Secretary : 01162587737 (tel)
- Pauline Jones, Paediatric Endocrine Specialist Nurse : 01162585326 (works part-time – please do not leave urgent messages out-of-hours)
- Out-of- hours - East Midlands Paediatric Endocrine Consultant on call: via Switchboard

## 3. Education and Training

No new training or education is required to implement this guideline.

## 4. Monitoring Compliance

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements
Appropriate dose of hydrocortisone pre, during and postop care	Medical notes	Dr Shenoy - Consultant Paediatrician	5 yearly	Audit meetings
Appropriateness of swap over to oral hydrocortisone	Medical notes	Dr Shenoy – Consultant Paediatrician	5 yearly	Audit meetings

## 5. Supporting References

- i) <https://www.bsped.org.uk/media/1823/bsped-adrenal-insufficiency-card-v31.pdf>
- ii) Spieser, et al. Congenital Adrenal Hyperplasia Due to Steroid 21-Hydroxylase Deficiency: An Endocrine Society Clinical Practice Guideline. J Clin Endocrinol Metab, November 2018, 103(11):4043–4088
- i) Treatment of classic congenital adrenal hyperplasia due to 21-hydroxylase deficiency in infants and children. UpToDate, July 2020.

## 6. Key Words

Congenital Adrenal Hyperplasia, Hydrocortisone, Adrenal Insufficiency,

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**The Trust recognises the diversity of the local community it serves. Our aim therefore is to provide a safe environment free from discrimination and treat all individuals fairly with dignity and appropriately according to their needs. As part of its development, this policy and its impact on equality have been reviewed and no detriment was identified.**

Contact & review details	
<b>Guideline Lead (Name and Title)</b> S. Shenoy Consultant Paediatrician	<b>Executive Lead</b> Chief Medical Officer
<b>Details of Changes made during review: September 2020</b> <ul style="list-style-type: none"><li>• Specified the parameter of 'long term' systemic steroids and expanded the list of steroids to consider</li><li>• Added for use in patients that have weaned off systemic steroids in the 3 months preceding the surgery</li><li>• Added that the guideline should be considered for use in patients on <i>regular</i> high doses of inhaled steroids</li><li>• CITO added to be checked when confirming up-to date individual dosing</li><li>• Added that it is the responsibility of anaesthetist to administer hydrocortisone prior to both minor and major surgery</li><li>• Dosing of IV hydrocortisone added to guideline</li><li>• Frequency of hydrocortisone administered to children with suspected adrenal suppression changed from TDS to QDS</li><li>• Removed the advice to give double the normal dose of hydrocortisone on return from theatre following minor surgery, changed to give oral hydrocortisone which is 30 mg/m<sup>2</sup>/day in 4 equal divided doses</li><li>• Updated contact numbers</li><li>• Updated references</li></ul>	

## Appendix 1. Body surface area in children

# BODY SURFACE AREA IN CHILDREN

## Body-weight under 40kg

Body-weight (kg)	Surface area (m <sup>2</sup> )
1	0.10
1.5	0.13
2	0.16
2.5	0.19
3	0.21
3.5	0.24
4	0.26
4.5	0.28
5	0.30
5.5	0.32
6	0.34
6.5	0.36
7	0.38
7.5	0.40
8	0.42
8.5	0.44
9	0.46
9.5	0.47
10	0.49
11	0.53
12	0.56
13	0.59
14	0.62
15	0.65
16	0.68

Body-weight (kg)	Surface area (m <sup>2</sup> )
17	0.71
18	0.74
19	0.77
20	0.79
21	0.82
22	0.85
23	0.87
24	0.90
25	0.92
26	0.95
27	0.97
28	1.0
29	1.0
30	1.1
31	1.1
32	1.1
33	1.1
34	1.1
35	1.2
36	1.2
37	1.2
38	1.2
39	1.3
40	1.3