

# Adrenal conditions and surgery. UHL Children's Hospital guideline



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## Abbreviations:

Adrenal insufficiency – AI  
Hypothalamo-pituitary-adrenal – HPA

## 1. Introduction and who this guideline applies to

Individuals with adrenal insufficiency (AI) are unable to mount stress response during events that cause major physiological stress such as medical or surgical procedures, thus additional hydrocortisone is required.

In 2020, Woodcock, et al representing the Association of Anaesthetists, the Royal College of Physicians and the Society for Endocrinology UK published the guidelines on perioperative

management of glucocorticoids. In 2022, the British Society of Paediatric Endocrinology & Diabetes (BSPED) have produced a consensus document on this topic for children and adolescents. These guidelines have provided the framework in developing this current UHL guideline. This guideline applies to;

A) those children undergoing surgery but who are on steroid replacement treatment due to **Adrenal conditions** which include:

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

B) those children at risk of/confirmed to have **hypothalamo-pituitary-adrenal axis (HPA) suppression** (tertiary AI) from long-term steroid treatment such as:

- Patients on long term continuous systemic steroids (e.g. prednisolone) for underlying chronic medical conditions. Daily physiological glucocorticoid requirements in children are around 8mg/m<sup>2</sup>/day cortisol (hydrocortisone). Thus children taking any preparation of steroid equivalent preparation at doses higher than this on continuous basis for three weeks or longer should be considered to be at risk of HPA suppression and potential to have adrenal crisis. Refer to table 1 for steroid inflammatory equivalence dose.
- Weaning regime of long-term steroid treatment. The BNF for children (BNFc) recommends that gradual withdrawal of systemic corticosteroids should be considered in those whose disease is unlikely to relapse and have received more than 40mg prednisolone (or equivalent) daily for > 1 week or 2 mg/kg/day for 1 week or 1mg/kg/day for 1 month.
- The BTS/SIGN 2019 asthma guidelines state a dose of >800 micrograms Beclomethasone dipropionate daily or equivalent (Fluticasone > 400 microgram/day) puts a child at risk of clinical adrenal insufficiency.
- Abrupt cessation of long-term steroid treatment

\*Some of the patients in this group may already have been confirmed to have HPA axis suppression and may have been provided a BSPED steroid card which covers their emergency steroid regime which parents should be familiar with.

| <b>Table 1:<br/>Steroid inflammatory equivalence chart (equivalences relative to 5mg prednisolone)</b> |                                 |  |
|--|---------------------------------|--|
| <b>Steroid</b>   | <b>Route</b>                    | <b>Equivalent dose</b>                 |
| Hydrocortisone   | Enteral/ Intravenous            | 20 mg                                  |
| Prednisolone   | Enteral                         | 5 mg                                   |
| Methylprednisolone   | Intravenous                     | 4mg                                    |
| Dexamethasone  | Enteral/ Intravenous            | 750 microgram                          |
| Deflazacort  | Enteral                         | 6 mg                                   |
| Betamethasone  | Enteral                         | 750 microgram                          |
| Prednisolone enema   | rectal                          | 1 x 20mg enema ~ 9mg oral prednisolone |
| Triamcinolone  | Intra-articular / intramuscular | See notes below *                      |

**\*Triamcinolone withdrawal:** In patients who have received more than physiological doses of Kenalog (more than one injection during a three-week period), withdrawal should not be abrupt. The dose should be reduced and the dosage interval increased until a dose or not more than 40mg and a dosage interval of at least three weeks have been achieved. Clinical assessment of disease activity may be needed.

This group of patients can be discussed with the Paediatric Endocrine Consultant On-call for advice at the pre-assessment stage.

## **2. Types of surgery**

These children (both A and B listed above) may need minor or major surgery.

**Major Surgery** is defined as surgery lasting more than 90 minutes with variable recovery periods and expected delay in restarting oral intake.

**Minor Surgery** is defined as a procedure lasting less than 90 minutes and the patient is expected to be eating and drinking by the next meal. This may include procedures such as MRI scans, endoscopy, dental extractions under general anaesthetic or other day case procedures. If procedure exceeds four hours or if the child is unstable, major surgery guidance has to be followed.

**Minor surgery 'without general anaesthesia'** includes those procedures that may require local anaesthesia (such as a skin biopsy, minor dental procedure like tooth extraction/filling) or procedures using non-anaesthetic sedation (e.g. chloral hydrate)

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

- a) Reconstructive genital surgery should be regarded as **major surgery**.
- b) Examination under anaesthetic (EUA) should be regarded as **minor surgery**.

## **3. Pre-admission planning:**

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. Ensure the correct oral (sick day dose) of Hydrocortisone for the child is available and recorded in the medical notes/drug chart prior to admission. This information may be available in the clinic letter or can be clarified with the named consultant for the child.
3. The patients should preferably be placed first on the surgical list in the morning. Prior liaison with the anesthetic is essential.
4. 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.

5. Please check the up-to-date dose of the child's medication from the family and the double check in medical notes/up-to-date clinic letter on DIT3. If any queries with the normal doses, please contact the endocrine secretary on 0116 258 7737 or the named Consultant Paediatric Endocrinologist for the child so the record on the clinical workstation or CITO can be checked for up to date doses and this information is recorded in the pre-admission records.

#### **4. On the day of the surgery**

##### **Pre-operative:**

- Ensure the child has received the normal dose of Hydrocortisone in the morning or afternoon depending on the time of the operation.
  - **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.
- The period of fasting should not be more than 6 hours without IV fluid replacement
- 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% glucose + 0.9 % Sodium Chloride)
- Children are at particular risk of hypoglycaemia. If they are fasting and not on IV saline and dextrose, they should have regular capillary blood glucose every 2 hours until IV fluids are commenced.
- Check blood glucose when IV fluids commenced and hourly thereafter.

##### **4.1: Major Surgery:**

Major Surgery is defined as surgery lasting more than 90 minutes with variable recovery periods and expected delay in restarting oral intake

**Table 2: Recommended doses for peri- and post-operative glucocorticoid cover in children with AI undergoing major surgery utilising either a continuous infusion or intermittent bolus in children and neonates**

| <b>Major Surgery: Intravenous Infusion (IVI) hydrocortisone doses</b>  |   |  |  |
|--|---|--|--|
| <b>Induction</b>   | IV bolus of hydrocortisone 2mg/kg (max 100mg)<br>(premature infants and neonates < 28 days corrected gestational age: 4mg/kg) |  |  |
| <b>Intraoperative</b>  | <b>IV hydrocortisone infusion as below</b>  |  |  |
| <b>Weight</b>  | <b>Total dose in 24 hours</b>   | <b>Infusion rate (50mg hydrocortisone in 50ml 0.9% sodium chloride*)</b>                           | <b>Additional considerations</b>   |
| ≤10kg  | 25 mg   | 1 ml/hr  | * Could consider more concentrated infusion in those needing fluid restriction (e.g. 100mg hydrocortisone in 50mls 0.9% saline).<br>* The hydrocortisone infusion can be run alongside 0.9% sodium chloride, 5% glucose and PlasmaLyte solutions |
| 10.1 to 20kg   | 50 mg   | 2 ml/hr  |  |
| 20.1 to 40kg   | 100 mg  | 4 ml/hr  |  |
| 40.1 to 70kg   | 150 mg  | 6 ml/hr  |  |
| Over 70kg  | 200 mg  | 8 ml/hr  |  |
| <b>Post-operative</b>  | Continue hydrocortisone infusion<br>Change to oral sick day steroids when stable and tolerating oral fluids / diet            |  |  |
| <b>Major Surgery: <b>Child</b> (over 28 days corrected gestational age) Intravenous Hydrocortisone Bolus doses</b>                               |   |  |  |
|  | <b>Hydrocortisone bolus dose</b>  | <b>Frequency</b>   | <b>Additional considerations</b>   |
| <b>Induction</b>   | <b>2mg / kg (max 100mg)</b>   |  | Consider infusion for prolonged procedures   |
| <b>Intraoperative</b>  | <b>2mg / kg (max 100mg)</b>   | Given at 6 hours IV  | 4 hourly if unstable   |
| <b>Post-operative</b>  | <b>1mg / kg (max 50mg)</b>  | Every 6 hours IV<br>Change to oral sick day steroids when stable and tolerating oral fluids / diet | In severe obesity consider substituting 50 mg hydrocortisone with 100 mg hydrocortisone  |
| <b>Major Surgery: <b>Premature infants and neonates</b> (less than 28 days corrected gestational age) Intravenous Hydrocortisone Bolus doses</b> |   |  |  |
|  | <b>Hydrocortisone bolus dose</b>  | <b>Frequency</b>   | <b>Additional considerations</b>   |
| <b>Induction</b>   | <b>4mg / kg</b>   |  | Consider infusion for prolonged procedures<br>Infants have a larger surface area thus use of 4mg/kg at induction   |
| <b>Intraoperative</b>  | <b>2mg / kg</b>   | Given at 6 hours IV  | 4mg/kg if unstable or consider 4 hourly doses  |
| <b>Post-operative</b>  | <b>2mg / kg</b>   | Every 6 hours IV<br>Change to oral sick day steroids when stable and tolerating oral feeds         | The oral dose can be given IV if not tolerating feeds  |

## Postoperative management:

1. Post operatively:
  - Continue IV fluids,
  - Continue IV hydrocortisone regime used during surgery, either the IV infusion (table 2.1) or 6 hourly IV bolus (table 2.2a or 2.2b) until child is eating and drinking and stable.
  - Check blood glucose 2 hourly until child is eating and drinking.
  - A more detailed assessment of fluids is required if intravenous fluids are necessary for more than 48 hours
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 sick day dose of hydrocortisone' which is 30mg/m<sup>2</sup>/day in 4 equal divided doses (6 hourly). The child's sick day dose regime should be available in pre-assessment record or BSPED AI card which parents may carry or copy available on CITO (careplan tab). If this is not available or unclear, please discuss with the child's named Consultant or the on call Paediatric Endocrinologist who can be contacted via Medirota or switchboard.

Please refer to **Appendix 1** for a guide to the dose based on body surface area and weight. Information on Alkindi which is an alternate oral preparation of hydrocortisone is provided in **Appendix 2**.

4. When oral hydrocortisone is prescribed, restart their normal once daily oral fludrocortisone dose at the normal time the child is on at home.
5. Change 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.
6. In those children with suspected Adrenal suppression due to long-term steroid treatment for other medical conditions (group B in section 1), prescribe hydrocortisone dose of 30mg/m<sup>2</sup>/day divided in four equal doses 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 their regimen and for advice on restarting their 'normal steroid treatment' if necessary.
7. A guide to when to give 'oral sick day steroid' is highlighted in **Appendix 3**.

## 4.2: Minor surgery requiring anaesthesia:

Minor surgery is defined as a procedure lasting less than 90 minutes and the patient is expected to be eating and drinking by the next meal. This may include procedures such as MRI scans, endoscopy, dental extractions under general anaesthetic or other day case procedures. **If procedure exceeds four hours or if the child is unstable, major surgery guidance has to be followed.**

**Table 3: Recommended dose for peri-operative glucocorticoid cover in minor surgery requiring general anaesthesia**

| Hydrocortisone dose for minor procedures requiring general anaesthesia |   |  |
|--|---|--|
|  | Hydrocortisone bolus dose                   | Post-operative   |
| <b>Induction</b>   | 2mg /kg (max 100mg)<br>(4mg/kg in neonates) | Oral sick day steroid doses for 24 hours – same as point 3 in postop management of major surgery in page 6 |

### Postoperative management:

1. On return from theatre, prescribe 'oral sick day dose' of oral hydrocortisone which is 30mg/m<sup>2</sup>/day in 4 equal divided doses (6 hourly). The actual dose details should be available in pre-assessment record or BSPED AI card which parents may carry or copy available in CITO (careplan tab). If this is unclear or unavailable, please discuss with the child's named Consultant or the on call Paediatric Endocrinologist who can be contacted via Medirota switchboard.

Please refer to **Appendix 1** for a guide to the dose based on body surface area and weight.

Information on Alkindi which is an alternate oral preparation of hydrocortisone is provided in **Appendix 2**.

2. Restart their '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. Advise the parents to continue the 'sick day dose' of oral hydrocortisone as in step 2 above for 48 hours and then reduce to their 'normal' dose.
4. If child is unable to tolerate oral fluids by 4 hours after returning from theatre, commence IV maintenance fluids (5% glucose + 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 'sick day dose' of oral hydrocortisone and fludrocortisone as in 1, above when oral fluids tolerated.
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 given every 6 hours 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.

### 4.3: Minor procedures not requiring general anaesthesia:

**Table 4: Recommended dose for peri-operative glucocorticoid cover in minor surgery not requiring general anaesthesia**

| <b>Hydrocortisone advice for minor procedures NOT requiring general anaesthesia</b>   |   |
|---|---|
| <b>Medical procedures (local anaesthetic or sedation)</b>   | <b>Oral hydrocortisone dose</b>   |
| Minor procedure – local anaesthetic (e.g. skin biopsy)  | Give oral sick steroid dose prior to procedure.                             |
| Minor dental procedures e.g. filling, tooth extraction  | Continue for up to 24 hours if in pain or unwell                            |
| MRI scans (using sedation)<br>Non-anaesthetic sedation (e.g. chloral hydrate) does not merit use of IV hydrocortisone. Sick day dosing with oral hydrocortisone is sufficient | Give oral sick day steroid dose prior to procedure and continue for the day |

### 5. Helpful Contact numbers:

- Endocrine Secretary : 0116 258 7737
- Paediatric Endocrine Specialist Nurses\*: 0116 258 5326 (office), Mobile: 07921545455, or 07929776711

\*Both nurses work part-time so please do not leave urgent voice mail messages

- Oncall Paediatric Endocrine Consultant – Working hours – refer to Medirota for COW Endocrine Consultant and their contact mobile number or via switchboard

Out of hours – East Midlands Paediatric Endocrine Consultant oncall: via switchboard

- For non urgent message: team can be contacted via email [paediatricendocrineteam@uhl-tr.nhs.uk](mailto:paediatricendocrineteam@uhl-tr.nhs.uk)

### 6. Education and training

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

### 7. Monitoring Compliance

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



## 8. Supporting References

1. Woodcock T, Barker P, Daniel S, Fletcher S, Wass JAH, Tomlinson JW, Misra U, Dattani M, Arlt W, Vercueil A. Guidelines for the management of glucocorticoids during the peri-operative period for patients with adrenal insufficiency: Guidelines from the Association of Anaesthetists, the Royal College of Physicians and the Society for Endocrinology UK. *Anaesthesia*. 2020 May; 75 (5): 654-663. Doi: 10.1111/anae.14963. Epub 202 Feb 3. Erratum in: *Anaesthesia*. 2020 Sep; 75 (9): 1252. PMID: 32017012.
2. Emergency and peri-operative management of adrenal insufficiency in children and young people. BSPED consensus guidelines. Developed by the Paediatric Adrenal Insufficiency Group On Behalf of the British Society of Paediatric Endocrinology & Diabetes 2022 . For further information scan QR code below or visit [BSPED |BSPED Adrenal Insufficiency Consensus Guidelines \(https://www.bsped.org.uk/adrenal-insufficiency\)](https://www.bsped.org.uk/adrenal-insufficiency)



3. BNF hydrocortisone dose: <https://bnfc.nice.org.uk/drugs/hydrocortisone/>

## 9. Keywords

Adrenal Insufficiency, Alkindi, Endocrine, Hydrocortisone, Major surgery, Minor surgery

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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><br>Dr S. Shenoy Consultant Paediatrician, Endocrinology   | <b>Executive Lead</b><br>Chief Medical Officer |
| <b>Details of Changes made during review: September 2022</b> <ul style="list-style-type: none"><li>• Dose of hydrocortisone for major and minor surgery was amended and updated</li><li>• Section on minor procedure without anaesthesia was added</li><li>• Tertiary AI group definition was expanded to include all potential patient groups</li><li>• Dose guide for oral sick day dose of hydrocortisone was provided as a supplementary page to make it convenient for the surgical team</li><li>• Information on newer oral preparation Alkindi was included as a supplementary page as some patients are on this preparation when they are admitted.</li><li>• Entire guidelines has been brought in line with the BSPED consensus document published in 2022</li></ul> |  |

## Appendix 1: Pre-calculated oral hydrocortisone sick day stress doses

A guide to the oral sick day steroid dose which provides the equivalent dose to 30mg/m<sup>2</sup>/day is given in **Appendix 2**. However, the actual dose may vary depending on the strength and preparation of the available hydrocortisone medication. Hydrocortisone tablet is available as Alkindi capsule (**Appendix 3**) which comes in strengths of 0.5mg/1mg/2mg strength and preferable to use in neonates and children under the age of 5 years where smaller doses are required.

Hence, a pragmatic approach for convenience may be to divide the total daily sick day steroid dose according to the strength of the hydrocortisone preparation prescribed with dose rounded up to the nearest suitable dose of ease of administration.

| Weight (kg) | BNFc surface area | Total daily sick day steroid dose (mg) equivalent to 30/m <sup>2</sup> /day | Sick day hydrocortisone: dose | Frequency |
|-------------|-------------------|---|-------------------------------|-----------|
| 1           | 0.1               | 3   | <b>0.8</b>                    | 4 x a day |
| 2           | 0.16              | 5   | <b>1.2</b>                    | 4 x a day |
| 3           | 0.21              | 6   | <b>1.5</b>                    | 4 x a day |
| 4           | 0.26              | 8   | <b>2</b>                      | 4 x a day |
| 5           | 0.3               | 9   | <b>2.5</b>                    | 4 x a day |
| 6           | 0.34              | 10  | <b>2.5</b>                    | 4 x a day |
| 7           | 0.38              | 11  | <b>3</b>                      | 4 x a day |
| 8           | 0.42              | 13  | <b>3</b>                      | 4 x a day |
| 9           | 0.46              | 14  | <b>3.5</b>                    | 4 x a day |
| 10          | 0.49              | 15  | <b>4</b>                      | 4 x a day |
| 15          | 0.65              | 20  | <b>5</b>                      | 4 x a day |
| 20          | 0.79              | 24  | <b>6</b>                      | 4 x a day |
| 25          | 0.92              | 28  | <b>7.5</b>                    | 4 x a day |
| 30          | 1.1               | 33  | <b>7.5</b>                    | 4 x a day |
| 35          | 1.2               | 36  | <b>10</b>                     | 4 x a day |
| 40          | 1.3               | 39  | <b>10</b>                     | 4 x a day |
| 45          | 1.4               | 42  | <b>10</b>                     | 4 x a day |
| 50          | 1.5               | 45  | <b>10</b>                     | 4 x a day |
| 55          | 1.6               | 48  | <b>12.5</b>                   | 4 x a day |
| 60          | 1.7               | 51  | <b>12.5</b>                   | 4 x a day |
| 65          | 1.8               | 54  | <b>12.5</b>                   | 4 x a day |
| 70          | 1.9               | 57  | <b>15</b>                     | 4 x a day |
| 75          | 1.9               | 57  | <b>15</b>                     | 4 x a day |
| 80          | 2.1               | 63  | <b>15</b>                     | 4 x a day |
| 90          | 2.2               | 66  | <b>15</b>                     | 4 x a day |

# What is Alkindi®?



Alkindi contains a medicine called hydrocortisone. Hydrocortisone belongs to a group of medicines known as corticosteroids. Hydrocortisone is a synthetic version of the hormone cortisol. Cortisol is made naturally by the adrenal glands in the body. Alkindi is used when the body is not making enough cortisol, because part of the adrenal gland is not working (adrenal insufficiency, often caused by an inherited condition called congenital adrenal hyperplasia).

### Reporting of side effects

If your child gets any side effects, talk to your doctor or pharmacist, this includes side effects not listed in the Alkindi package insert. You can also report side effects directly to United Kingdom Yellow Card Scheme. Website: [www.mhra.gov.uk/yellowcard](http://www.mhra.gov.uk/yellowcard) or search for MHRA Yellow Card in the Google Play or Apple App store. By reporting side effects, you can help provide more information on the safety of this medicine.

### Important

Alkindi packaging contains a patient information leaflet about taking Alkindi. Please read the leaflet carefully, if you have any questions please ask your doctor or pharmacist.

### Notes

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Scan to view the dosing guide video



Diurnal Europe BV, Van Heuven Goedhartlaan 935 A,  
1105 BD Amstelveen, The Netherlands.  
Tel: +31 (0)20 6615 072.  
Email: [info@diurnal.co.uk](mailto:info@diurnal.co.uk)







Date of preparation June 2022

Inf EU-GB-0177

## Dosing Guide

Information for use only with patients who have been prescribed Alkindi® (hydrocortisone granules in capsules for opening)

### How to open the Alkindi capsule and give the granules

|   |  |
|---|--|
| <p>1. Hold capsule so that the text is at the top and tap the capsule to make sure the granules are at the bottom</p>  | <p>4. Pour all granules out of capsule</p> <p>Either pour all the granules directly onto the child's tongue</p>    |
| <p>2. Gently squeeze the bottom of the capsule</p>    | <p>OR pour all the granules directly onto a spoon and place them in the mouth</p>    |
| <p>3. Twist off the top of the capsule</p>   | <p>OR for children who are able to take soft food, sprinkle the granules onto a spoonful of cold or room temperature soft food (such as yoghurt or fruit puree) and give immediately</p>  |

Whichever method is used, tap the capsule to ensure all the granules are removed.

If you give the granules directly into the mouth, give a drink (e.g. water, milk, breast-milk or formula-milk) immediately after administration to help ensure all granules are swallowed.

If you give the granules sprinkled onto a spoonful of soft food, administer immediately (within 5 minutes) and do not store for future use.

**DO NOT add the granules to liquid before administration** as this can result in less than the full dose being given, and might also dissolve the taste masking of the granules allowing the bitter taste of hydrocortisone to become apparent.

**Alkindi granules come in a capsule that must be opened before use, discard the empty capsule after use out of reach of children. Do NOT swallow the capsule – small children may choke.**

### Appendix 3: Patient Information: Sick days: When to give additional steroids

| Situation   | Change to usual hydrocortisone dose   | Length of change  | When to get help?  |
|---|---|---|--|
| <b>Minor illness</b>  |   |   |  |
| Mild cold / runny nose with no fever.<br>Minor playground bumps and bruises   | No change   |   |  |
| <b>Moderate or severe illness</b>   |   |   |  |
| Fever, flu, infection, childhood illnesses (usually not well not enough to go to school)                              | Sick day doses required   | For as long as the illness lasts  | Contact GP or medical team if not improving after 24-48 hours  |
| Vomiting or diarrhoea   | Sick day doses required   |   |  |
|   | If sick day dose tolerated (kept down for at least 30 minutes with no diarrhoea or continuous vomiting), then continue oral sick day dosing                   |   |  |
|   | If sick day dose not tolerated, give IM hydrocortisone injection  | If an IM injection of hydrocortisone is required then dial 999 and inform them that the patient is having an adrenal crisis |  |
| Drowsy and unresponsive   | Give IM hydrocortisone injection  |   |  |
| Major trauma or severe shock (e.g. suspected fracture, road traffic accident, head injury with loss of consciousness) | Give IM hydrocortisone injection  |   |  |
| <b>Other (discuss with medical team)</b>  |   |   |  |
| Routine or travel vaccinations  | Consider 1 or 2 doses of sick day steroids. Continue if necessary   |   |  |
| COVID-19 vaccine  | Sick day doses required   | Sick day steroids for 48 hours  |  |
| Long haul flight over 12 hours  | Give usual morning dose at 6 to 8 hourly intervals  |   |  |
| Child or centre specific recommendations  |   |   |  |
| <b>Surgical and dental procedures</b>   |   |   |  |
| Minor surgery (e.g. dental extraction under local anaesthetic)  | Sick day dose prior to procedure  | Return to usual dose immediately afterwards<br>Continue for 24 hrs if in pain/unwell  | Inform medical staff including dentist and anaesthetist that you/your child have adrenal insufficiency and take steroids |
| Major surgery (e.g. operation requiring general anaesthetic)  | Sick day steroids (oral or IV) on day of procedure even when fasting.<br>Intravenous hydrocortisone will be given on induction as per local hospital protocol | As per local policy or contact treatment centre for advice  |  |

## Appendix 4: BSPED adrenal insufficiency (AI) card



### British Society for Paediatric Endocrinology and Diabetes: Adrenal Insufficiency Card

#### PAEDIATRIC STEROID CARE PLAN FOR SICK DAYS AND EMERGENCIES

#### IMPORTANT MEDICAL INFORMATION FOR PARENTS/CARERS & HEALTHCARE STAFF

**This patient has adrenal insufficiency and is steroid dependent. Steroids should never be missed. Any stress situation, such as illness or surgery will require additional treatment**

|                                       |   |
|---------------------------------------|---|
| Name                                  | Address                                   |
| DOB                                   |   |
| NHS / CHI Number                      |   |
| GP Contact Details                    | Next of Kin Contact Details               |
| Paediatric Lead Consultant & Hospital | Paediatric Endocrine Team Contact Details |
| Diagnosis                             | Date of issue                             |

Print double sided and 'flip pages short edge'.

BSPED AI Card.v1.4 Nov 2022

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| Sick days: When to give additional steroids   |  |   |  |
|---|--|---|--|
| Situation   | Change to usual steroid dose   | Length of change  | When to get help?  |
| <b>Minor illness</b>  |  |   |  |
| Mild cold / runny nose with no fever. Minor playground bumps and bruises  | No change  |   |  |
| <b>Moderate or severe illness</b>   |  |   |  |
| Fever, flu, infection, childhood illnesses (usually not well enough to go to school)                                    | Sick day doses required  | For as long as the illness lasts                                | Contact GP or medical team if not improving after 24-48 hours  |
| Vomiting or diarrhoea   | Sick day doses required  |   |  |
|   | If sick day dose tolerated (kept down for at least 30 minutes with no frequent diarrhoea or vomiting), then continue oral sick day dosing          |   |  |
|   | <b>If sick day dose not tolerated, give IM hydrocortisone injection</b>  |   | If an IM injection of hydrocortisone is required, then dial 999 and inform them that the patient is having an adrenal crisis |
| Drowsy and unresponsive   | <b>Give IM hydrocortisone injection</b>  |   |  |
| Major trauma or severe shock (e.g., suspected fracture, road traffic accident, head injury with loss of consciousness). | <b>Give IM hydrocortisone injection</b>  |   |  |
| <b>Other (discuss with medical team)</b>  |  |   |  |
| Routine or travel vaccinations  | Consider 1 or 2 doses of sick day steroids. Continue if symptomatic  |   |  |
| Long haul flight  | Give usual morning dose at 6 to 8 hourly intervals   |   |  |
| Child or centre specific recommendations  |  |   |  |
| <b>Surgical and dental procedures</b>   |  |   |  |
| Minor surgery (e.g., dental extraction under local anaesthetic)   | Sick day dose prior to procedure<br>Return to usual dose immediately afterwards  | Continue sick day doses for up to 24 hours if in pain or unwell | Inform medical staff including dentist and anaesthetist that you/your child have adrenal insufficiency and take steroids     |
| Major surgery (e.g., operation requiring general anaesthetic)   | Sick day steroids (oral or IV) on day of procedure even when fasting. Intravenous hydrocortisone given on induction as per local hospital protocol | As per local policy or contact treatment centre for advice      |  |

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| Steroid Treatment Regimen   |                          |  |
|---|--------------------------|--|
| Medication (and preparation)<br><i>e.g., Hydrocortisone (10mg tablet)</i> | Time<br><i>e.g., 6am</i> | Dose<br><i>e.g., 5mg (half a tablet)</i> |
|   |                          |  |
|   |                          |  |
|   |                          |  |
|   |                          |  |
|   |                          |  |

| Oral sick day steroid treatment regimen in the event of an illness<br>(See back page for more information on when this is required) |      |      |
|---|------|------|
| Medication (and preparation)  | Time | Dose |
|   |      |      |
|   |      |      |
|   |      |      |
|   |      |      |
| If patient takes fludrocortisone, no change in dose is necessary  |      |      |

| Emergency Steroid Injection   |                                  |                                       |
|---|----------------------------------|---------------------------------------|
| If they show no sign of improvement, have persistent vomiting or diarrhoea, become drowsy or unresponsive, or has had a major accident or injury give <b>intramuscular (IM) injection of hydrocortisone immediately</b> |                                  |                                       |
| Age less than 1-year:<br><b>25mg</b>  | Age 1 to 5 years:<br><b>50mg</b> | Age 6 years and over:<br><b>100mg</b> |
| Dial 999 stating they are having an ADRENAL CRISIS; they should be taken to the nearest A&E without delay   |                                  |                                       |

**Important information for medical emergency teams**

If this patient is unwell and brought to hospital, the following management should be instituted promptly:

- Administer Emergency IM/IV hydrocortisone**  
 Age less than 1-year 25mg  
 Age 1 to 5 years 50mg  
 Age 6 years and over 100mg  
 (Check if this has been given by parent, caregiver, or ambulance crew)
- Check **blood glucose**. Give 2ml/kg 10% dextrose IV if glucoses <3mmol/L
- If **circulation** is compromised, give 10 ml/kg 0.9% saline bolus
- Obtain U&E & **start IV fluids** 0.9% Saline/5% dextrose at a maintenance rate
- Ongoing hydrocortisone doses:**
  - Continue with IV hydrocortisone 2mg/kg (max 100mg) every 4-6 hours (or hydrocortisone infusion if very unwell).
  - Once stable, the IV bolus dose of hydrocortisone is 1mg/kg (max 50mg) every 6 hours.
  - For neonates (< 28 days) the IV bolus dose of hydrocortisone is 4mg/kg every 4-6 hours. Once stable the IV bolus dose is 2mg/kg every 6 hours.
  - Please refer to BSPED guidance for full details.
- Once tolerating oral fluids, swap to oral sick day dosing until back to normal self (usually 2-3 days of sick day dosing).

Important: Please observe until patient is tolerating oral steroids at sick day dosing. Contact your acute paediatric or paediatric endocrine team if admission is required.

If the patient needs a **general anaesthetic or surgery**, please contact the paediatric endocrine team for a perioperative plan. The recommended doses for hydrocortisone can also be found on the BSPED website below.

For further information scan the QR code or visit **website** to see BSPED guidance:  
<https://www.bsped.org.uk/adrenal-insufficiency>



<https://www.bsped.org.uk/clinical-resources/guidelines/>