PAEDIATRIC ENDOCRINE EMERGENCY PROTOCOLS

MANAGEMENT OF CHILDREN AND YOUNG PEOPLE WITH ADRENAL INSUFFICIENCY WHEN HAVING MAJOR SURGERY OR MINOR PROCEDURES

SETTING	Emergency Departments, Children's Assessment Units, Paediatric Inpatient Wards, Operating Theatres
FOR STAFF	All staff working in Child Health within the South and West Wales region. Paediatric Anaesthetists, General Paediatricians, Emergency Medicine Clinicians, surgeons and theatre staff involved in perioperative care of children up to the age of 18 years
PATIENTS	Children or young people up to the age of 18 having surgery, who are diagnosed with adrenal insufficiency.

These guidelines have been produced to guide clinical decision making for the medical and nursing staff. **They do not replace the clinical judgement of a senior clinician**. Each case should be fully assessed individually with respect to the particular needs and circumstances of that patient.

Key Points

- Children and young people with Adrenal Insufficiency have a daily requirement for steroids at a replacement dose, but at times of physiological stress e.g. surgery, will require additional exogenous steroids.
- It is essential these patients receive increased doses of steroids at induction and post operatively, as without this they are at risk of a life-threatening adrenal crisis.
- This guideline aims to ensure that patients with adrenal insufficiency undergoing major surgery or minor procedures are adequately supplemented with steroids during the peri-operative period.

Contents

If there are concerns the child is unstable, please consider adrenal crisis. See separate guidance:

Management Of An Adrenal Crisis in Children And Young People

The ensure you are using the most up to date version of this document, please click on the link below and confirm the version number:
Paediatric Endocrine Clinical Guidelines (sharepoint.com)

Definitions

Adrenal Insufficiency (AI)

Inadequate production of glucocorticoid +/- mineralocorticoid by the patient's adrenal gland due to a problem at the level of the adrenal gland (primary), or the pituitary gland (secondary)

There are two major categories of adrenal insufficiency.

- Primary adrenal insufficiency is due the adrenal gland being unable to produce cortisol +/- aldosterone (e.g. CAH, Addison's)
- Secondary adrenal insufficiency is due to deficient adrenocorticotropin hormone secretion by the pituitary gland (e.g. hypopituitarism, exogenous steroid use)

Major surgery

Major surgery is defined as any procedure lasting >90 minutes with variable recovery periods and an expected delay in restarting oral intake

Minor procedures

Minor Procedures are defined as any procedure lasting <90 minutes and the patient is expected to be eating and drinking by the next meal (e.g. MRI scans, endoscopies, dental extractions under general anaesthetic or sedation). If the procedure exceeds >90 minutes management should proceed as per major surgery, with a further bolus of IV hydrocortisone given 4-6 hours after the initial dose.

Major Surgery: Peri and Post-Operative Management

Major surgery is defined as any procedure lasting >90 minutes with variable recovery periods and an expected delay in restarting oral intake**.**

Please inform paediatric endocrine team of the date of planned major surgery.

Day before surgery

- Advise usual oral steroid treatment.

Morning of Surgery

- All children should be placed first on the morning list, if possible, to minimise dehydration and fasting time
- Advise parents that usual oral hydrocortisone dose should be given at 6.00am with clear fluids (For those children also taking fludrocortisone this can also be taken then).
- Record blood pressure
- Insert IV cannula
- Monitor Capillary Blood Glucose (CBG) hourly once fasting time exceeds 4 hours
- If there is a delay of more than 2 hours in taking the child to theatre, IV maintenance fluids (0.9% sodium chloride plus 5% glucose or Plasma-Lyte 148 & Glucose 5%) should be started.
- If the child is due any further doses of hydrocortisone routinely while they are in hospital and NBM for the procedure, these should be given intravenously at the patient's usual dose.
- Anaesthetist to prescribe IV hydrocortisone for the procedure (see below)

Induction, Intraoperatively and Post- Operatively: Management of Steroids

In all cases, an initial bolus of hydrocortisone is given at induction, followed by **either hydrocortisone infusion or regular bolus doses (as preferred).** If the child is seriously unwell, septic or haemodynamically unstable then consider an IV hydrocortisone infusion, for example children admitted to PICU post-operatively.

Major Surgery: Continuous intravenous infusion (IVI) hydrocortisone doses			
Induction	IV bolus of hydrocortisone 2mg/kg (max 100mg) (premature infants and neonates <28 days corrected gestational age: 4mg/kg)		
Intraoperative	IV Hydrocortisone infusion as below		
Weight	**Total dose in 24 hours**	**Infusion rate (50mg hydrocortisone in 50ml 0.9% sodium chloride)**	**Additional Considerations**
Less than 10kg	25mg	1ml/hr	- Consider more concentrated infusion in those needing fluid restriction (e.g. 100mg hydrocortisone in 50mls 0.9% sodium chloride). - The hydrocortisone infusion can be run alongside 0.9% sodium chloride, 5% glucose and Plasma-Lyte solutions.
10.1 to 20kg	50mg	2 ml/hr	
20.1 to 40kg	100mg	4 ml/hr	
40.1 to 70kg	150mg	6ml/hr	
Over 70kg	200mg	8 ml/hr	
Post-Operative	Continue hydrocortisone infusion Change to oral sick day hydrocortisone when stable and tolerating oral fluids / diet		

Major Surgery: Child (>28days corrected gestational age) with intravenous hydrocortisone boluses			
	Hydrocortisone bolus dose	**Frequency**	**Additional considerations**
Induction	2mg/kg (max 100mg)		- Consider neonatal doses for infants who are significantly small for gestational age or with growth faltering
Intraoperative	2mg/kg (max 100mg)	Given at 6 hours IV	- Consider infusion for prolonged procedures - 4 hourly if unstable
Post- operative	1 mg/kg (max 50mg)	Every 6 hours IV Change to oral sick day hydrocortisone when stable and tolerating oral fluids / diet	- In severe obesity consider substituting 50 mg hydrocortisone with 100 mg hydrocortisone

Major surgery: premature infants and neonates (<28 days corrected gestational age) with intravenous hydrocortisone boluses			
	Hydrocortisone bolus dose	**Frequency**	**Additional considerations**
Induction	4mg/kg		
Intraoperative	2mg/kg	Given at 6 hours IV	- Consider infusion for prolonged procedures - 4mg/kg if unstable or consider 4 hourly doses
Post-operative	2mg/kg	Every 6 hours IV Change to oral sick day steroids when stable and tolerating oral feeds	- The oral dose can be given IV if not tolerating feeds

Minor Procedures: Peri and Post-operative Management

Minor procedures are defined as any procedure lasting <90 minutes and the patient is expected to be eating and drinking by the next meal (e.g. MRI scans, endoscopies, dental extractions under general anaesthetic or sedation). If the procedure exceeds >90 minutes management should proceed as per major surgery, with a further bolus of IV hydrocortisone given 4-6 hours after the initial dose.

Day before surgery/procedure

- Advise usual oral steroid treatment and diet

Morning of Surgery

- All children should be placed first on the morning list, if possible, to minimise dehydration and fasting time.
- Take oral hydrocortisone dose at 6.00am with clear fluids (For those children also taking fludrocortisone this can also be taken then).
- Record blood pressure
- Insert IV cannula
- Monitor Capillary Blood Glucose (CBG) hourly once fasting time exceeds 4 hours
- If there is a delay of more than 2 hours in taking the child to theatre, IV maintenance fluids (Plasma-Lyte 148 & Glucose 5%) should be started.
- If the child is due any further doses of hydrocortisone routinely while they are in hospital and NBM for the procedure, these should be given intravenously at the usual dose.
- Anaesthetist to prescribe hydrocortisone as below for procedure.

Induction, Intraoperatively and Post- Operatively

Hydrocortisone dose for minor procedures requiring general anaesthesia		
	Hydrocortisone bolus dose	Post-Operative
Induction	2mg/kg (max 100mg) * (4mg/kg in neonates)	Oral sick day steroid doses for 24 hours
** It would seem prudent to use the neonatal dosing for infants who are significantly small for gestational age or failing to thrive and as such, whilst not neonates, are a neonatal size*		

Hydrocortisone advice for minor procedures NOT requiring general anaesthesia	
Medical procedure- local anaesthetic (e.g. skin biopsy) Minor dental procedures e.g. filling, tooth extraction	Give oral sick day steroid dose prior to procedure. Continue for up to 24 hours if in pain or unwell
MRI scans (using sedation) 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

Oral Sick Day Rules

A total daily hydrocortisone dose of around 30/mg/m2/day given as four evenly spaced doses is recommended for illness. A guide to the doses is provided below; however, the actual dose may vary depending on the strength and preparation of the available hydrocortisone medication.

Weight (kg)	BNFc surface area	Total daily sick day steroid dose (mg) (30mg/m2/day)	Sick day hydrocortisone: Dose	Frequency
1	0.1	3	**0.8**	4 x a day
2	0.16	5	**1.2**	4x a day
3	0.21	6	**1.5**	4 x a day
4	0.26	8	**2**	4 x a day
5	0.3	9	**2.5**	4 x a day
6	0.34	10	**2.5**	4 x a day
7	0.38	11	**3**	4 x a day
8	0.42	13	**3**	4 x a day
9	0.46	14	**3.5**	4 x a day
10	0.49	15	**4**	4 x a day
15	0.65	20	**5**	4 x a day
20	0.79	24	**6**	4 x a day
25	0.92	28	**7.5**	4 x a day
30	1.1	33	**7.5**	4 x a day
35	1.2	36	**10**	4 x a day
40	1.3	39	**10**	4 x a day
45	1.4	42	**10**	4 x a day
50	1.5	45	**10**	4 x a day
55	1.6	48	**12.5**	4 x a day
60	1.7	51	**12.5**	4 x a day
65	1.8	54	**12.5**	4 x a day
70	1.9	57	**15**	4 x a day
75	1.9	57	**15**	4 x a day
80	2.1	63	**15**	4 x a day
90	2.2	66	**15**	4 x a day

References

1. www.bsped.org.uk. (n.d.). BSPED |BSPED Adrenal Insufficiency Consensus Guidelines. [online] Available at: https://www.bsped.org.uk/adrenal-insufficiency. [Accessed 5 Dec. 2022].

2. Kumaran, A. and Watts, G. (2019) Guideline for steroid replacement in children with adrenal insufficiency or at risk of adrenal suppression who require surgery or a procedure requiring sedation or general anaesthetic, PAEDIATRIC INNOVATION, EDUCATION & RESEARCH NETWORK. Available at: https://www.piernetwork.org/steroid-replacement-adrenal-insufficiency-surgery.html. (Accessed: December 5, 2022).

3. Woodcock, T. et al. (2020) "Guidelines for the management of glucocorticoids during the Peri-operative period for patients with adrenal insufficiency," Anaesthesia, 75(5), pp. 654–663. Available at: https://doi.org/10.1111/anae. 14963.