



Adrenal Hypoplasia Congenita

SURGERY GUIDELINES FOR ALL WITH ADRENAL INSUFFICIENCY

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The following guidelines are for **ALL** patients who are on replacement glucocorticoid therapy and take hydrocortisone or any other steroid.

The medical conditions include **ALL** the variations of congenital adrenal hyperplasia, salt losing, non-salt losing, LOCAH as well as adrenal hypoplasia congenita, hypopituitarism and Addison's.

It is ESSENTIAL to make the anaesthetist aware that the patient is cortisol deficient and is receiving glucocorticoid therapy.

A bolus dose of IV hydrocortisone is needed pre surgery and post-surgery.

For major surgery a **bolus dose** of IV hydrocortisone of 2mg/kg body weight needs to be given *pre* and *post*-surgery.

For minor surgery a **bolus dose** of IV hydrocortisone of 1mg/kg body weight must be given *pre* and *post*-surgery.

Elective Surgery

- Any patient who is cortisol deficient should not be starved for more than 6 hours.
- Blood pressure should be monitored and charted every 4 hours.

Pre-Surgery Morning List

1. The patient should preferably be first on the morning list.
2. Normal previous evening and morning doses of glucocorticoid should be given as usual.
3. **Hourly blood glucose concentrations** should be carefully monitored from 6 am.
4. At the same time a dextrose-saline infusion should be commenced and maintained until the patient is tolerating oral fluids.

Pre-Surgery Afternoon List

1. The patient should preferably be first on the afternoon list.
2. The patient should be given their usual morning glucocorticoid dose and lunchtime dose just before the start to be 'nil by mouth'.
3. **Hourly blood glucose concentrations** should be carefully monitored after 4 hours after start of starvation. At the same time a dextrose-saline infusion should be commenced and maintained until the patient is tolerating oral fluids.

At Induction

A bolus dose of IV hydrocortisone (2mg/kg body weight) must be given at induction

During Surgery

For operations expected to exceed 4 hours a further bolus of hydrocortisone during the procedure will be required. Alternatively, for prolonged procedures and when post-operative recovery is likely to be slow, consideration should be given to hydrocortisone infusion.

Post-Surgery

As the patient comes out of the anaesthetic, a post bolus IV dose of hydrocortisone (1mg/kg body weight) should be given for minor surgery and for major surgery (2mg/kg body weight).

1. Regular post-operative blood glucose measures should be undertaken at 2 hourly intervals until the patient is able to tolerate oral fluids.
2. Post-operative dosing schedule will be determined by the extent of the surgical procedure and should be conducted in liaison with the Endocrine Team.
3. It is important to remember that a bolus IV dose does not last longer in the body than 4 – 6 hours, and this must be remembered when considering the post dosing schedule.

Note:

Remember that hydrocortisone has marked mineralocorticoid activity and care must be exercised in situations where water retention would be a disadvantage e.g. cerebral oedema when betamethasone or dexamethasone should be used.

Fludrocortisone

If the patient is receiving fludrocortisone treatment then they should continue to take this as normal with no change to dose.

If nil by mouth then there is no problem omitting the fludrocortisone as the intravenous fluids will be sufficient to maintain sodium in the blood.

Emergency Surgery

A bolus dose of IV hydrocortisone 2mg/kg body weight MUST BE GIVEN pre surgery.

For post-surgery a bolus of IV hydrocortisone (2mg/kg body weight) for major surgery and for minor surgery (1mg/kg body weight) must be given.

The endocrine team should be informed and they will set out a post-operative dosing and monitoring schedule which will be determined by the extent of the surgical procedure.



Table: 1 Summary of Surgery Protocol

Stage	Minor surgery e.g. cystoscopy	Major surgery e.g.: genitoplasty
AT INDUCTION	Hydrocortisone IV 2mg/kg and repeated if procedure exceeds 4 hours	Hydrocortisone IV 2mg/kg
DURING SURGERY		Repeat IV hydrocortisone on 4 hourly basis or hydrocortisone infusion.
POST-OP	As the patient comes out of the anaesthetic give Hydrocortisone IV 1mg/kg Can use repeat IV regimen of hydrocortisone (2mg/kg) in lieu of routine medication until tolerating oral fluids. Then return to oral therapy which must include that day's fludrocortisone dose	Give the same dose of IV hydrocortisone (2mg/kg) at end of operations as at induction Can use repeat IV regimen of hydrocortisone. Alternatively, when post-operative recovery is likely to be slow consider hydrocortisone infusion. Continue IV administration until tolerating fluids. Then return to oral therapy at twice normal dose for 24 hours* then return to normal requirements. As soon as on oral therapy reintroduce fludrocortisone dose

*Post- operative dosing schedule will be determined by the extent of the surgical procedure and should be conducted in liaison with the Endocrine Team.

Fluids

100ml/kg/day if weight < 10 kg

80ml/kg/day if weight 10-30 kg

60ml/kg/day if weight > 30 kg

If salt losing congenital adrenal hyperplasia use: 5% glucose with 0.45% sodium chloride.

For other cases use: 4% glucose with 0.18% sodium chloride.

Hydrocortisone Infusion

Rates of hydrocortisone infusions will be dependent on individual weight, pubertal status and cortisol clearance rates. To achieve a cortisol concentration of 1000nmol/l (0.36mg/ml) as one might desire in a PICU patient with sepsis, suggested infusion rates are outlined in Table 2 below

Table 2: Infusion Rates

Situation	Cortisol clearance (ml/24hrs)	Infusion rate (mg/24hr)
Prepubertal: 10kg	74	25
10-20kg	147	50
20kg	294	100
Pubertal	430	155
Post pubertal	290	105

Infusion Rate (mg/24hrs) = clearance (ml/24hr) x stead