

Acute Adrenal Insufficiency

Management Guidelines
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Possible Presentations

- The most common is the child with known adrenal insufficiency who has an intercurrent illness.
- New presentation of adrenal insufficiency; think of this possibility with any severely ill child with **severe dehydration or shock**.
- Neonatal “collapse” in male at 1-3 weeks of age. (CAH)

Causes

- **Primary** adrenal diseases[↑] (ACTH levels) eg. Addison’s disease, congenital adrenal hyperplasia, adrenal aplasia/hypoplasia, adrenoleukodystrophy, adrenal destruction.
- **Secondary** adrenal insufficiency[↓] (ACTH levels) e.g. Pituitary disorders, hypothalamic disorders.
- **Withdrawal from** pharmacological doses of **corticosteroids**.

Clinical features

- Muscle weakness, lethargy, vomiting (*due to cortisol deficiency, electrolyte disturbances*).
- Weight loss (*due to anorexia, volume depletion*).
- Depression, anorexia (*due to cortisol deficiency*).
- Pigmentation (*in primary adrenal insufficiency, due to ACTH excess*).
- Hypoglycaemia (confusion, coma) (*due to decreased hepatic glucose production*).
- Dehydration, hypotension, shock (*due to loss of vasomotor tone and volume depletion*).

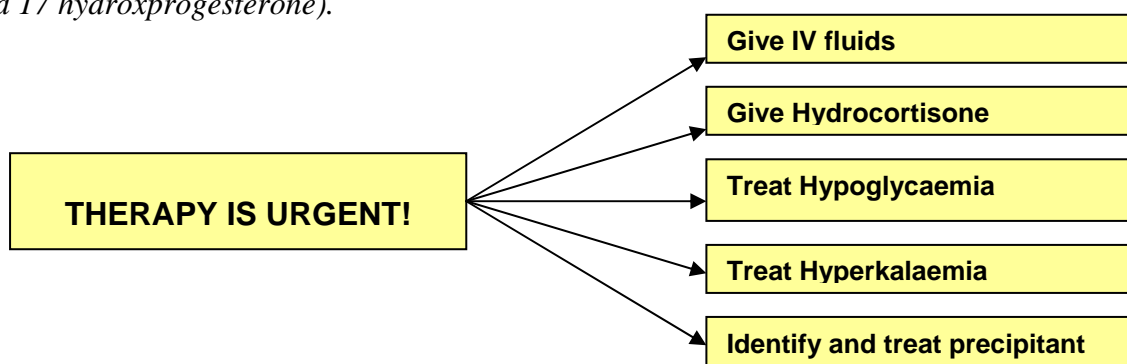
Biochemical features

- Electrolyte disturbances (low Na⁺, high K⁺) due to mineralocorticoid (Aldosterone) deficiency*.
- Elevated serum urea and creatinine due to associated dehydration*.
- Elevated plasma renin activity (as an index of volume depletion)*.
- Elevated plasma ACTH*.
- Low cortisol before and after adrenal stimulation eg. By Synacthen.
- Hypoglycaemia due to lack of glucocorticoid.
- *Only in primary adrenal deficiency: in hypothalamic-pituitary disorders mineralocorticoid function is unaffected.*

Investigations

- Immediate blood glucose using a dextrostix.
- Serum glucose, urea, sodium and potassium.
- Arterial, venous or capillary acid-base.

Where the underlying diagnosis is not known, collect at least 2 mL of clotted blood for later analysis (cortisol and 17 hydroxprogesterone).



Management

- a) Management of children with minor intercurrent illness who are able to tolerate oral medication.
- b) Management of more seriously unwell children requiring IV therapy.
- c) Management of patients who present requiring surgery under anaesthesia.

a) MANAGEMENT OF CHILDREN WITH MINOR INTERCURRENT ILLNESS WHO ARE ABLE TO TOLERATE ORAL MEDICATION.

Children with adrenal insufficiency or at risk (ie on steroids) **MUST BE GIVEN** increased doses of replacement hydrocortisone during illness or stress. Parents will often have these guidelines and may have tried these strategies prior to presenting to hospital:

- If moderately unwell and/or temperature is 38-39° C **give three times** their usual dose of hydrocortisone.
- If more unwell and/or temperature > 39° C **give four times** their usual dose of hydrocortisone.
- If a dose is vomited, give the increased dose again in ½ hour. If this is vomited then immediately give parenteral steroids (see below).
- If child has gastroenteritis or any cause of diarrhoea: **give four times** their usual dose of hydrocortisone. Consider parenteral treatment if any doubt about absorption.
- Child should wear *Medic Alert* identification.

*N.B. Increase **only** the dose of hydrocortisone (not fludrocortisone).*

When the stress of illness is over, revert to the previous hydrocortisone dose without tapering.

b) MANAGEMENT OF MORE SERIOUSLY ILL PATIENTS WHO REQUIRE IV THERAPY.

Susceptible patients who present with vomiting but with are not otherwise unwell should be considered to have incipient adrenal crisis. To attempt to prevent this from developing further:

- Administer IM or IV hydrocortisone 2 mg/kg.
- Give trial of oral fluids and observe for 4-6 hours before considering discharge.
- Discuss with Emergency or Endocrine consultant before discharge.

For all other children

1. Give intravenous fluids:

Shock or severe dehydration:

- Normal saline 20 mL/kg IV bolus
- Repeat until circulation is restored
- Give remaining deficit plus maintenance as normal saline with 5% dextrose * over 24 hr.
- Check electrolytes and glucose frequently

Moderate dehydration

- Normal saline 20 mL/kg IV bolus even if NOT clinically shocked
- Give remaining deficit plus maintenance as normal saline with 5% dextrose* over 24 hr

Mild or no dehydration:

- No bolus
- Give 1.5 x maintenance fluid volume administered evenly over 24 hours

*Additional dextrose may be required to ensure euglycaemia

2. Give hydrocortisone:

Administer hydrocortisone intravenously. If IV access is difficult, give IM while establishing intravenous line. Dose given is according to age.

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| ▪ Neonate: | 25 mg stat and then 10-25 mg, 4 hourly |
| ▪ 1 month – 1 year: | 25 mg stat, then 25 mg, 4 hourly |
| ▪ Toddlers (1-3 years): | 25-50 mg stat then 25-50 mg, 4 hourly |
| ▪ Children (4-12 years): | 50-75 mg stat, then 50-75 mg, 4 hourly |
| ▪ Adolescents and adults: | 100-150 mg stat, then 100 mg, 4 hourly |

When stable, reduce IV hydrocortisone dose, then switch to triple dose oral hydrocortisone therapy, gradually reducing to maintenance levels (10-15 mg/m²/day).

For patients with mineralocorticoid deficiency start fludrocortisone at maintenance doses (usually 0.1 mg daily) as soon as the patient is able to tolerate oral fluids.

3. Treat hypoglycaemia

Hypoglycaemia is common in infants and small children.

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| ▪ Treat with IV bolus of 2-4 mL/kg of 10% dextrose |
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Maintenance fluids should contain between 5 and 10% dextrose.

4. Treat hyperkalaemia (see **Hyperkalaemia** protocol for details)

Hyperkalaemia usually normalises with fluid and electrolyte replacement.

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| <p>□ K⁺ > 6 mmol/L</p> <p>▪ Perform an ECG and apply cardiac monitor as arrhythmias and cardiac arrest may occur</p> |
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| <p>□ K⁺ > 7 mmol/L <u>and</u> hyperkalaemic ECG changes (eg peaked T waves, wide QRS)</p> <p>▪ Give nebulised salbutamol 5mg</p> <p>▪ Give 10% calcium gluconate 0.5 mL/kg (max. 20 mL) IV over 3-5 mins</p> <p>▪ Commence infusion of insulin 0.1 units/kg/hour IV together with an infusion of 50% dextrose 2 mL/kg/hr</p> |
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| <p>□ K⁺ > 7 mmol/L with normal ECG</p> <p>▪ Give sodium bicarbonate 1-2 mmol/kg IV over 20 minutes, with an infusion of 10% dextrose at 5 mL/kg/hour</p> |
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5. Identify and treat potential precipitating causes such as sepsis.

6. Admit to appropriate inpatient facility.

c) MANAGEMENT OF PATIENTS WHO PRESENT REQUIRING SURGERY UNDER ANAESTHESIA

Children with / or at risk of adrenal insufficiency need glucocorticoid cover for surgery which needs to be discussed with anaesthetic staff to prevent intra or post operative adrenal crisis.

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| <p>□ For most minor surgery</p> <p>▪ At induction: single dose of IV hydrocortisone as recommended for acute adrenal insufficiency. (see above)</p> <p>▪ Then three to four times the usual oral dose for 24 hours.</p> |
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| <p>□ For major surgery</p> <p>▪ At induction: give the IV hydrocortisone dose recommended for acute adrenal insufficiency. (see above)</p> <p>▪ Then 100 mg/m²/day IV divided into 4-6 doses until the major post-operative stress is resolved.</p> |
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