High-Risk groups for Adrenal Crisis are:

1. **Primary adrenal deficiency:**
   a. Congenital Adrenal Hyperplasia
   b. Addison’s disease or adrenal hypoplasia

2. **Secondary (pituitary) ACTH deficient conditions:**
   a. Hypopituitarism (congenital/idiopathic)
   b. Septo-optic dysplasia or midline CNS abnormalities

3. **Recent treatment with high dose synthetic steroids for prolonged periods**

All children at risk of adrenal insufficiency should be discussed with and admitted under the Paediatric Endocrinology Team with appropriate steroid replacement.

**Background**

The classic triad for adrenal crisis: ↓ serum Na, ↑ serum K, and ↓ serum Glucose

**Symptoms seen in Adrenal Crisis:** (not all have to be present)
- Dehydration, shock
- GI symptoms (Vomiting, Diarrhea, Abdominal Pain, Constipation)
- Syncope
- Fever
- Tachycardia
- Hypoglycemia
- Hypotension (postural drop)
- Acidosis
- Lethargy, weakness

**Investigations**

- Blood tests:
  - Electrolytes (particularly Na, K, glucose)
  - Calcium
  - FBC
  - +/-cap gas.
STRESS STEROID MANAGEMENT FOR CHILDREN WITH PRIMARY OR SECONDARY ADRENAL FAILURE

Clinical tests:
- Monitor vital signs (BP, Pulse, Temperature)
- Tachycardia with postural drop

Principles of Management

1. **Increased Cortisol is needed in times of stress**
   In normal subjects increases approximately 5+ fold during any major physical stress such as illness, anesthesia and surgery.

2. **Goal: to replace appropriate cortisol response in children who are adrenally insufficient**

3. **The preferred route is IV (or IM); hydrocortisone is the steroid of choice.**
   - A continuous IV infusion is ideal
   - Intermittent boluses 6 x per day (4 hourly) are another option

4. **High doses (stress) of hydrocortisone also provide full mineralocorticoid effect.**
   The exception is dexamethasone: has no significant mineralocorticoid effect.

5. **Stress doses of hydrocortisone can be reduced rapidly**
   - Back to simple replacement doses as the patient recovers.
   - “Tapering” the dose is not usually needed in this setting.

6. **Don't forget to treat the underlying cause of the stress**
   Consider occult infection/appendicitis/UTI etc.

Management Details

**A - Severely Unwell:**

Unstable / Crisis **OR** Unwell (vomiting, diarrhoea, drowsy), unable to tolerate oral hydrocortisone

- Bolus IV fluids: 10-30 ml/kg 0.9% saline, then maintenance 0.9% saline + 5% dextrose.

- Start IV steroids
  - Bolus - Hydrocortisone: 75-100 mg/m² IV (or IM) - then
  - Infusion - Hydrocortisone 55-100mg/m²/day until stable.

Body surface area: \( \sqrt{ \text{height (cm) x weight (kg)/3600} } \) or use **BSA calculator**.

Table 1 gives rapid estimates of hydrocortisone doses.
**STRESS STEROID MANAGEMENT FOR CHILDREN WITH PRIMARY OR SECONDARY ADRENAL FAILURE**

Table 1: Rapid Estimates of Hydrocortisone Dose if Weight or BSA not available

<table>
<thead>
<tr>
<th>Age</th>
<th>Initial bolus dose of hydrocortisone succinate (IM or IV):</th>
<th>Hydrocortisone Infusion (50 mg hydrocortisone in 50 ml 0.9% saline)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-6 months</td>
<td>12.5 mg</td>
<td>1ml/hr : infants</td>
</tr>
<tr>
<td>6mo -5 years</td>
<td>25 mg</td>
<td>2 ml/hr: pre-schoolers</td>
</tr>
<tr>
<td>5 -10 years</td>
<td>50 mg</td>
<td>3 ml/hr: older children</td>
</tr>
<tr>
<td>&gt;10 years</td>
<td>100 mg</td>
<td></td>
</tr>
</tbody>
</table>

Presumed adrenal insufficient patient undergoing CNS tumour resection:

Dexamethasone given for raised ICP is sufficient (see Table 3 in the Appendix for potency compared to hydrocortisone).
Review once off steroid in view of possible ACTH deficiency from surgery

B – Moderately Unwell:

Stable with fever, reduced activity:
If able to tolerate oral hydrocortisone: give 50 mg/m²/day
e.g.: Give 5x (five times) the patient’s normal total daily dose hydrocortisone, as 4-5 divided doses (breakfast, lunch, afternoon tea, dinner, bedtime)

Gastroenteritis: Stable with mild diarrhea:
If able to tolerate oral fluids start oral hydrocortisone 50mg/m²/day as above
If unable to tolerate oral fluids give IV/IM hydrocortisone (see ‘A’ above)

C – Mildly Unwell:

Respiratory, ear infection, no fever, vomiting or diarrhoea, and looks well
Continue normal maintenance doses of steroid.
No Fever >38 degrees, not on antibiotics.

D - Elective surgical procedures:

Simple elective surgery
Can be managed by increased oral hydrocortisone for 24-48 hours. (2-3x)
Often an oral dose of hydrocortisone at the onset of the ‘Nil by mouth’ period can suffice: discuss with anaesthetist regarding when Nil by mouth period starts
For more acute surgery or prolonged illness/surgery

Table 2: Peri-operative Stress Steroids

<table>
<thead>
<tr>
<th>Weight (kg)</th>
<th>Single dose Hydrocortisone pre-operatively PO - prior to being NBM</th>
<th>Rate of infusion intra- and post-operatively: 50 mg hydrocortisone in 50 ml 0.9% saline (ml/hr = mg/hr)</th>
</tr>
</thead>
<tbody>
<tr>
<td>3-10</td>
<td>25 mg</td>
<td>1</td>
</tr>
<tr>
<td>10-20</td>
<td>50 mg</td>
<td>2</td>
</tr>
<tr>
<td>&gt;20</td>
<td>100 mg</td>
<td>3</td>
</tr>
</tbody>
</table>

- Maintenance IV fluid rate of 0.9% NaCl with 5% dextrose.
- If weight is unknown dosing can be based on age:
  En route to surgery, prior to anaesthesia:
  - 0-3 yrs: Hydrocortisone 25 mg IV bolus, then 50 mg/m² (~25 mg) infusion for remaining 24 hrs
  - 3-12 yrs: Hydrocortisone 50 mg IV bolus, then 50 mg/m² (~50-60 mg) infusion for remaining 24 hrs
  - 12 yrs: Hydrocortisone 100 mg IV bolus, then 50 mg/m² (~100 mg) infusion for remaining 24 hrs

- Once stable: switch to maintenance oral hydrocortisone (~10mg/m²/day).

Examples

1. 12 year old male with CAH, acute vomiting, abdominal pain, temperature of 38.5, hypotensive:
   - Administer 100 mg hydrocortisone IM/IV followed by a continuous infusion of 100mg/m²/day until clinically stable.

2. 9 month old male with congenital hypopituitarism with upper respiratory infection and fever of 38.3°C. Usual medication hydrocortisone 2 mg am, 1.25 mg midday, and 1.25 mg at night. So total is 5 mg/day
   - Give 5 mg hydrocortisone x 5 times per day until fever resolves, then go back to maintenance dose.
   - See GP to investigate cause of fever.
STRESS STEROID MANAGEMENT FOR CHILDREN WITH PRIMARY OR SECONDARY ADRENAL FAILURE

References & Further Reading

Pediatric Endocrinology, Sperling, 3rd ed. 2008

Practical Endocrinology and Diabetes in Children 2nd ed. 2006

Pediatric Practice: Endocrinology, Michael Kappy, David Allen, Mitchell Geffner, 2010,

Australasia Paediatric Endocrine Group – Adrenal Gland

Appendix – Steroid Potency

Table 3: Steroid Potency

<table>
<thead>
<tr>
<th></th>
<th>Glucocorticoid effect in relation to hydrocortisone/cortisol = 1.0</th>
<th>Sodium Retention Effect Equivalent to 0.1 mg Florinef PO</th>
<th>Stress dose for individual glucocorticoid preparations equivalent to 100mg Cortisol PO</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cortisol (Hydrocortisone)</td>
<td>1</td>
<td>20</td>
<td>100 mg</td>
</tr>
<tr>
<td>Cortisone</td>
<td>0.8</td>
<td>20</td>
<td>125 mg</td>
</tr>
<tr>
<td>Prednisone</td>
<td>4 – 6</td>
<td>50</td>
<td>25 mg</td>
</tr>
<tr>
<td>Prednisolone</td>
<td>6 – 8</td>
<td>50</td>
<td>20 mg</td>
</tr>
<tr>
<td>Methylprednisolone (Solumedrol)</td>
<td>6 – 8</td>
<td>0</td>
<td>15-20 mg</td>
</tr>
<tr>
<td>Dexamethasone</td>
<td>80 to 120</td>
<td>0</td>
<td>1.5 mg</td>
</tr>
<tr>
<td>9a-Fluorocortisone (Florinef)</td>
<td>~None; but 15x mineralocorticoid effect</td>
<td>0.1</td>
<td>N/A</td>
</tr>
</tbody>
</table>