

Steroid dependence (PIP)

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The following guidance is taken from the Partners In Paediatrics (PIP)

Key amendments to this document

Date	Amendment	Approved by:
9 th March 2024	Document reviewed and amended: change to age - BCH like cut off to be 1 yr. 1-5 years = 50mg	Paediatric Governance Meeting
27 th August 2024	Document had minor amendment- Diane Cluley was removed as co-owner, retired 3 years ago	Dr James West

Steroid dependence 2018–20

STEROID DEPENDENCE

Hypothalamic-pituitary-adrenal axis impairment

RECOGNITION AND ASSESSMENT

Definition

- Children with the following conditions are corticosteroid-dependent with a depressed or absent pituitary-adrenal axis:
 - hypopituitarism
 - adrenal insufficiency
 - congenital adrenal hyperplasia
 - growth hormone insufficiency
 - prolonged oral corticosteroid use >2 months

Corticosteroid-dependent children cannot mount an appropriate adrenal response when shocked or stressed

- Corticosteroid-dependent children are encountered in a number of ways:
 - at presentation and first diagnosis
 - for elective surgical and investigative procedures
 - for emergency surgery or when acutely unwell
 - with hyponatraemia, hyperkalaemia +/- hypoglycaemia and hypotension

MANAGEMENT

Dose guidance for hydrocortisone IV whilst nil by mouth:

Age	Single stress dose	Continuous infusion dose	6-hrly bolus dose
<2 yr	25 mg	25 mg/day	6 mg
2–5 yr	50 mg	50 mg/day	12.5 mg
>5 yr	100 mg	100 mg/day	25 mg

- Continuous hydrocortisone infusion – avoids peaks and troughs (should be first line of treatment)
- Dilute required amount of hydrocortisone in sodium chloride 0.9% 50 mL and infuse over 24 hr

Minor surgery, or general anaesthesia/sedation for imaging or other minor procedure, or mild systemic illness

- Give single stress dose of hydrocortisone IV at induction pre-surgery
- On return from theatre give stress dose hydrocortisone i.e. 30 mg/m²/day oral
- divide dose into 4 equal doses 6-hrly, for 1 day only
- If unable to tolerate oral fluids 4 hr after theatre, commence IV maintenance fluids and give hydrocortisone IV see **Dose guidance for hydrocortisone IV whilst nil-by-mouth**
- Change to 30 mg/m²/day oral in 4 divided doses for 1 day once tolerating oral fluids
- Patient to carry steroid card^[1]
- Discuss any concerns with consultant endocrinologist

Major surgery

- Check pre-operative endocrine management discussion has taken place
- Give single stress dose of hydrocortisone IV at induction in anaesthetic room pre-surgery, followed by either continuous infusion or 6-hrly divided doses as above
- Commence maintenance fluids of glucose 5% and sodium chloride 0.9% in theatre and continue until child is eating and drinking post-operatively – maintain blood sugar >4 mmol/L

- Continue hydrocortisone IV in above doses until child is eating and drinking, then change to oral stress dose, equal to 30 mg/m²/day
- divide dose into 4 equal 6-hrly oral doses ^[17]_[SEP]
- Recommend reduction of hydrocortisone to usual oral supplementation doses 2 days after discharge
- Continue usual medications, e.g. fludrocortisone, levothyroxine, desmopressin

Acute illness

- During illness, corticosteroid-dependent children can usually be managed at home
- if able to take hydrocortisone orally, give stress dose of hydrocortisone 30 mg/m²/day as 4 divided doses 6 hrly for 2–3 days
- if unable to take oral corticosteroids (e.g. vomiting or acute collapse), parents to administer hydrocortisone IM:
 - aged <2 yr: 25 mg
 - aged 2–5 yr: 50 mg
 - aged >5 yr: 100 mg
- If hydrocortisone IM required, hospital assessment necessary
- If hydrocortisone IM not available and child too unwell to take oral corticosteroids call 999
- Continue usual dose of other medication e.g. fludrocortisone, levothyroxine, desmopressin, growth hormone etc.

Management of unwell corticosteroid-dependent children requiring hospital assessment

- Resuscitate (ABC)
- Monitor BP and GCS
- Obtain IV access
- Take blood for glucose, FBC, blood culture, U&E, bicarbonate and blood gas
- If blood glucose <4 mmol/L: give bolus of glucose 10% 2 mL/kg and monitor blood glucose

First line treatment

- Give hydrocortisone IV as single stat dose, followed by either continuous infusion or 6-hrly divided doses to avoid peaks and troughs see **Dose guidance for IV hydrocortisone whilst nil-by-mouth**
- Maintain blood sugar >4 mmol/L
- If shock give sodium chloride 0.9% 20 mL/kg
- Commence IV maintenance with sodium chloride 0.9% and glucose 5% at maintenance rate (extra if dehydrated)
- add potassium depending on electrolyte result
- Severely ill: commence hydrocortisone infusion (see **Dose guidance for hydrocortisone IV whilst nil-by-mouth**)
- When oral fluids tolerated change to hydrocortisone 30 mg/m²/day oral 6-hrly in 4 divided doses, and continue for 2–3 days after recovery from acute episode
- Discuss any concerns with on-call consultant endocrinologist