



EMERGENCY CARE FOR ADRENAL HYPOPLASIA CONGENITA

Please also read our Ambulance leaflet

PATIENT NAME

PATIENT HOME

ADDRESS

POST CODE

DATE OF BIRTH

HOSPITAL NUMBER

TO WHOM IT MAY CONCERN:

The above patient has a rare genetic disorder adrenal hypoplasia congenita (congenital adrenal hypoplasia) and is under the care of the endocrine team at:-.....

Please contact the endocrine team **AS SOON AS POSSIBLE** on telephone:-.....

He/she must be seen by a medical physician IMMEDIATELY. Time in a waiting room or triage situation is INAPPROPRIATE.

The patient is on replacement steroids and is at risk of a life threatening adrenal crisis if not treated quickly. A crisis will occur when there is an electrolyte imbalance with febrile illness, fluid depletion from vomiting and diarrhoea, burns, serious illness and injury. Signs of an impending crisis can include, weakness, dizziness, floppiness, failure to respond, nausea and vomiting, hypotension, hypoglycaemia, pallor, and clammy sweating.

1. INSERT IV CANNULA

2. URGENT BLOOD TESTS REQUIRED

- Basic metabolic panel including: - sodium, potassium, chloride, and bicarbonate, urea, glucose, creatinine and calcium.
- Cortisol to get idea of current status.
- Check capillary blood glucose level and perform any other appropriate tests (e.g. urine culture).

3. TREATMENT

STAT: Solu-Cortef injection IV or IM, if NOT already given by the parent or paramedic

Doses are as follows:-

Age range (years)	Dose (mg)
0 – 1	25
1 – 5	50
Over 5	100

4. IV FLUIDS

1. Commence IV fluids Infusion of 0.45% sodium chloride, 5% glucose at maintenance rate (extra if patient is dehydrated). Add potassium depending on electrolyte balance.
2. Commence hydrocortisone infusion in 50 ml 0.9% sodium chloride via syringe pump.
3. Monitor for at least twelve hours before discharge.
4. **IMPORTANT** If blood glucose is <2.5 mmol/l give bolus of 2ml/kg of 10% glucose.
5. If patient is drowsy, hypotensive and peripherally shut down with poor capillary return give:- 20ml/kg of 0.9% sodium chloride stat.

IF IN ANY DOUBT ABOUT THIS PATIENT'S MANAGEMENT, PLEASE CONTACT PATIENTS ENDOCRINE TEAM ON _____

Professor Peter Hindmarsh

Divisional Clinical Director for Paediatrics and Adolescents at UCLH

Professor Peter Hindmarsh Professor of Paediatric Endocrinology

Divisional Clinical Director for Paediatrics and Adolescents at UCLH

"The ideas expressed are independent of the authors' affiliations. Data provided is from current literature and should always be discussed with your endocrinologist first"

<http://www.cahisus.co.uk>