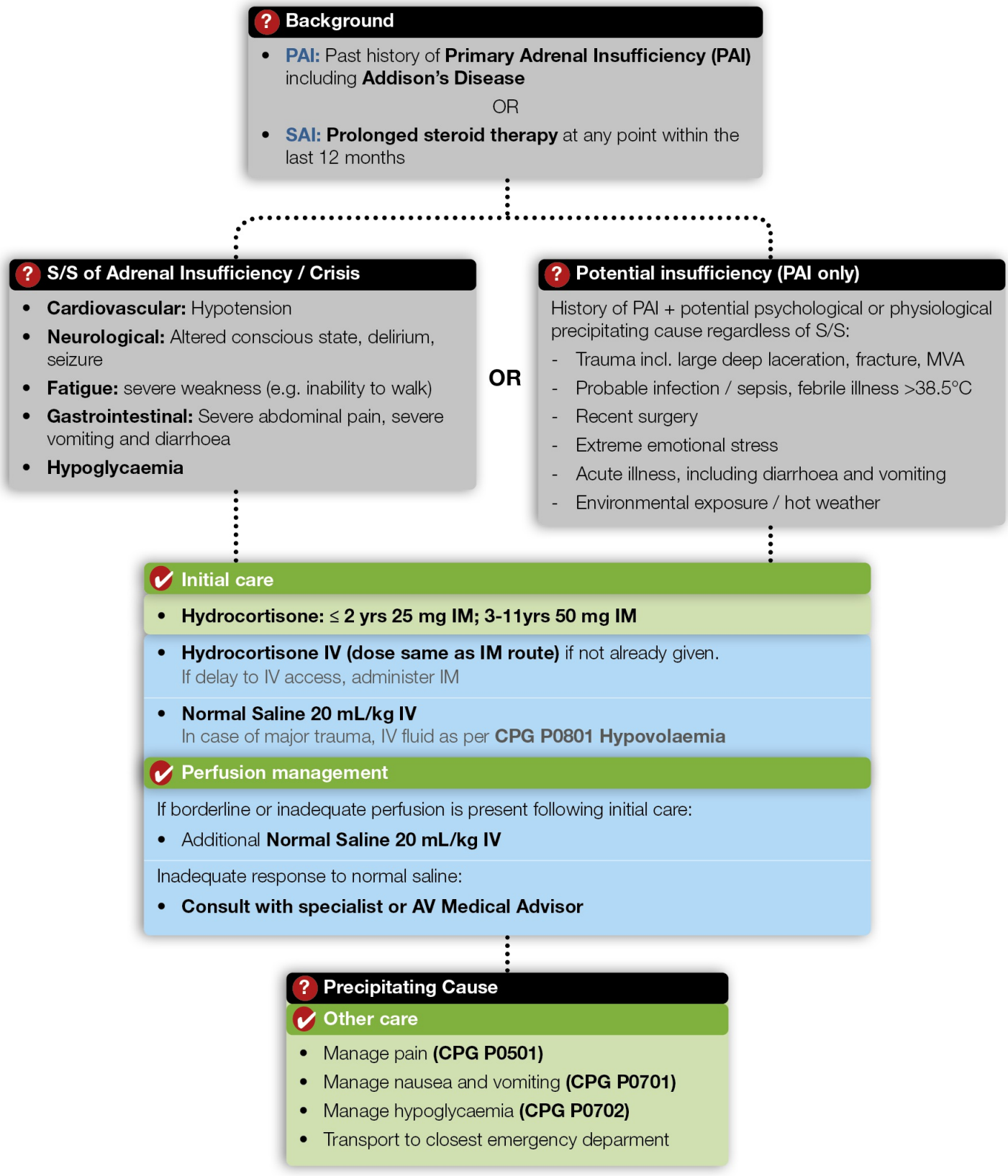
Adrenal Insufficiency CPG P0714

Flowchart



Care Objectives

- Prioritise corticosteroid therapy
- Support perfusion with IV fluid
- Transport to closest emergency department

Intended patient group

- Aged < 12 with suspected adrenal insufficiency

General Notes

Adrenal insufficiency

- An endocrine disorder characterised by inadequate production of the adrenal hormones cortisol +/- aldosterone leading to impaired regulation of glucose levels and cardiovascular function.
- Can lead to an **adrenal crisis**, a severe life-threatening form of adrenal insufficiency.

Types

Primary Adrenal Insufficiency (PAI)

- Includes Addison's Disease and Congenital Adrenal Hyperplasia (CAH)
- Due to adrenal gland destruction or impairment
- Often triggered by physiological or psychological stress

Secondary Adrenal Insufficiency (SAI)

- Occurs where pituitary or hypothalamic impairment reduces production of adrenocorticotropic hormone (ACTH) in turn reducing steroid hormone secretion from adrenal glands
- Can occur following prolonged (glucocorticoid) steroid therapy. If therapy is suddenly ceased or the dose is inadequate for the amount of stress present, the adrenal glands may not be able to produce sufficient cortisol to meet demand, precipitating an adrenal crisis.

Prolonged steroid therapy

- Illnesses managed with steroid therapy include: asthma, inflammatory bowel diseases, rheumatic diseases, vasculitis and organ transplantation.
- The potency, dose, and duration of steroid therapy can vary. As a guide, patients likely to have SAI in this setting will have prescribed steroid therapy for greater than 4 weeks.
- As SAI is also capable of precipitating life-threatening adrenal crisis it must be viewed as comparable in severity to PAI.

Assessment

The assessment items below are of particular relevance in adrenal insufficiency.

- **History:** PAI, corticosteroid pharmacotherapy, physiological or psychological stress
- **ECG:** Due to risk of hyperkalaemia
- **BGL:** Due to risk of hypoglycaemia

Symptoms of adrenal insufficiency may be mild to severe. Early signs may include mood swings, irritability, joint pain, fatigue, difficulty to rouse, and abdominal cramping.

Unstable PAI children are dynamic and have the potential to deteriorate quickly. Do not leave unwell PAI children at home or refer to GP. Consider early the potential need for MICA assistance especially where long transport times are required.

Gender identity

In a small proportion of children, the mechanisms of PAI (especially CAH) can result in undifferentiated development of sexual organs during in-utero growth. Children may be born with atypical genitalia, and this may or may not have been surgically corrected at some point. Be mindful of gender cues in conversation as a patient may be referred to as they/them.

Management

All patients

- Patients with a history of PAI must be considered for treatment with hydrocortisone where any physiological or psychological stressor is considered moderate or severe in order to avoid potential adrenal crisis.
- Signs and symptoms of adrenal insufficiency are not required to manage this patient group.
- Parent / carers of children with PAI are generally well educated about the illness and can often identify symptoms of adrenal insufficiency. Accordingly the parent / carer may have initiated their ‘sick day management plan’ including administration of their own IM injection of hydrocortisone. Alternatively the parent / carer may request Paramedics administer hydrocortisone. Review the patient's care plan as a part of your assessment.
- The patient with a history of PAI may have very poor veins for IV access. Do not delay hydrocortisone while trying to gain IV access. Use the IM route.

Extended travel time > 1 hour

- Consult with the patient’s endocrine specialist or receiving hospital to establish ongoing IV fluid management plan and any other care priorities.

Hydrocortisone is unlikely to cause harm but has the potential to be life-saving.
If there is any doubt, initiate Hydrocortisone and IV fluids.

Related Resources

- Video: Adrenal crisis - When to give an emergency injection (Consumer Information)
- RCH Adrenal Insufficiency Guideline
- https://av-digital-cpg.web.app/assets/pdf/MAC/MAC Paper - Adrenal Insufficiency CPG - MAC Sept 2021.pdf