British Columbia's Children's Hospital
Reference Care Plan

Adrenal Insufficiency

Date Initiated: 2010/08/15
Date Revised: 2015/11/08

Patient Population:

Children with adrenal insufficiency are unable to make the life sustaining hormone cortisol. At home, cortisol replacement is managed by the parents. However, the child may present to the hospital for many different reasons: intercurrent illness, surgery, chemotherapy or diagnostic procedures. Acute adrenal crisis can occur in a patient with adrenal insufficiency if cortisol replacement and or stress dosing is not administered as directed. Some children with adrenal insufficiency also require aldosterone replacement (Florinef ®) to manage fluid and electrolyte balance.

Definitions:

- **Adrenal Crisis:** Acute adrenal crisis is a life-threatening condition that occurs when the body is severely cortisol deficient.
- **Primary Adrenal Insufficiency:** is caused by inability of the adrenal glands to produce cortisol e.g. removal of adrenal glands, undeveloped adrenal glands, congenital adrenal hyperplasia (CAH), Addison disease, or adrenoleukodystrophy.
- **Secondary Adrenal Insufficiency:** is caused by the inability of the pituitary gland to signal the adrenal glands to produce cortisol e.g. hypopituitarism or suppression of hypothalamic-pituitary-adrenal axis from long-term steroid use (more than 2 weeks).
- **Cortisol Replacement:** Hydrocortisone, prednisone or prednisolone are commonly used for cortisol replacement.
- **Stress Dosing:** Patients with adrenal insufficiency require stress/illness dosing of their cortisol replacement during times of physical stress e.g. illness with fever, surgical procedures or acute trauma.

Problem/Potential Problem	Objectives	Anticipatory/Therapeutic Nursing Interventions	Evidence-base/Rationale
1. Potential for adrenal crisis due to: • **Interruption of routine cortisol replacement and absorption by fasting (NPO), vomiting, or diarrhea** • **Increased cortisol needs from physical stress of illness, fever, surgery or diagnostic procedure.**	Child will maintain adequate cortisol replacement.	a) Ensure cortisol replacement is given **on time**. b) Be aware that during times of physical stress (fever, trauma, procedures, surgery, infections), cortisol replacement should be double or triple the normal daily dose. c) Ensure cortisol replacement is given by an alternate route (IV or IM) if child is NPO or has vomiting or diarrhea. Call physician for orders. d) Monitor blood glucose levels if ordered.	**Delayed or inadequate administration of cortisol replacement could precipitate adrenal crisis.** **A child on cortisol replacement should NEVER miss a dose.**

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2. Potential for altered fluid and electrolyte balance due to low cortisol and aldosterone levels.	Child's serum sodium and glucose levels will remain within normal limits.	a) Ensure aldosterone replacement (Florinef®) is given if ordered. b) Provide additional salt as ordered.	**Aldosterone replacement (Florinef®) is required to maintain sodium balance.**
3. Anxiety and loss of control, ineffective coping related to: • **hospital environment** • **medical/surgical procedures** • **illness** • **pain** • **new diagnosis** • **developmental stage** • **past experience**	Refer to *Psychosocial Care Reference Care Plan*.	a) Refer to *Psychosocial Care Reference Care Plan*. b) Listen to parent's concerns and suggestions as they have likely received previous education on their child's condition and management.	**Parents may recognize subtle changes in their child's behaviour/health condition before these changes are noticed by staff. Prompt initiation of stress dosing will help avoid an adrenal crisis.**

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4. Ability to care for child at home on discharge from hospital	Family and primary caregivers will demonstrate ability to provide care for the patient in the home/ community as evidenced by: a) Ability to administer prescribed medication. b) Ability to problem solve an illness episode and to determine when to give emergency injection or illness dose at home. c) Has medication including Solu-Cortef and injection supplies. d) Ability to perform IM injection e) Knows how and when to contact endocrinologist on call.	a) Endocrine nurse clinician (local 7927) to provide teaching on illness management and emergency treatment. b) The following handouts may be given to the family by the endocrine Nurse Clinician: • Congenital Adrenal Hyperplasia • Hypopituitarism • Info on Addison disease • *Management of Hydrocortisone Replacement* • *Hydrocortisone Injection Instructions* • *Salt Replacement for Mineralocorticoid Deficiency* (as needed) • *School Letter for Cortisol Dependent Children* • *Immunizations for Children Who are Cortisol-Dependent* • *Influenza Vaccine for Children who are Cortisol-Dependent* c) Collaborate with discharge planning nurse/ nurse coordinator, social worker and community liaison nurse to prepare for transition to home/community	**Parents/care-givers must be able to recognize illness and understand the treatment of the illness.** **Illness that is not recognized or treated appropriately with cortisol could lead to potentially life- threatening adrenal crisis.**

References:

- Jung C, Inder W. Management of adrenal insufficiency during the stress of medical illness and surgery. *Medical Journal of Australia* 2008;188(7):409–413.
- Shulman D, Palmert M, Kemp S. Adrenal insufficiency: Still a cause of morbidity and death in childhood. *Pediatrics* 2007;119(2):485–493.