

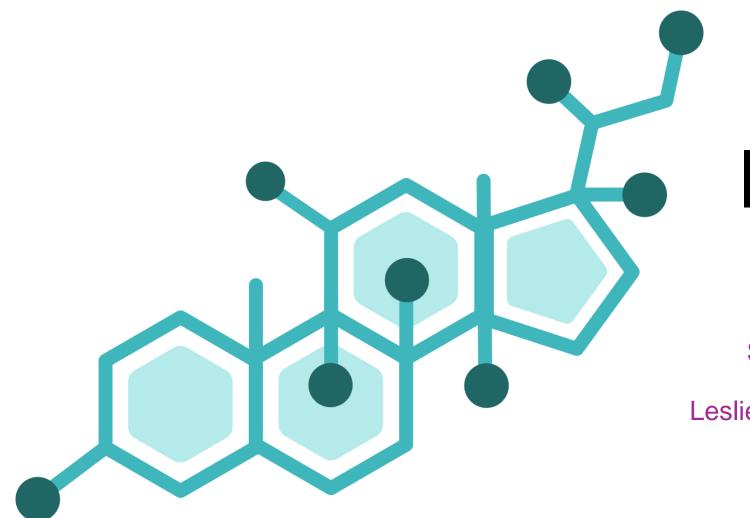
## **Adrenal Insufficiency**

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&

Leslie Pitts, MSN, CRNP, CPNP-AC, CDCES June 20, 2024





# Adrenal Insufficiency Training

Sarah Sparks, MSN, CRNP, FNP-C &
Leslie Pitts, MSN, CRNP, CPNP-AC, CDCES

# Objectives After completion of this training, the learner will be able to:

Describe adrenal gland function.

4

Recall adrenal pathophysiology and pharmacology.

Manage adrenal insufficiency appropriately.

Dose and administer Solu-Cortef emergency injection for adrenal crisis events.

# Adrenal Insufficiency School Training

- Alabama Senate Bill 52
- Amended the Alabama Safe Schools Act.
- Mandates adrenal insufficiency and adrenal crisis training for school staff.
- Authorizes trained school personnel to administer injectable medication to students with adrenal crisis.
- Alabama Health Services Safe Schools Adrenal Insufficiency Curriculum



# The Adrenal Glands



The adrenal glands are small organs that rest on the upper poles of the kidneys.

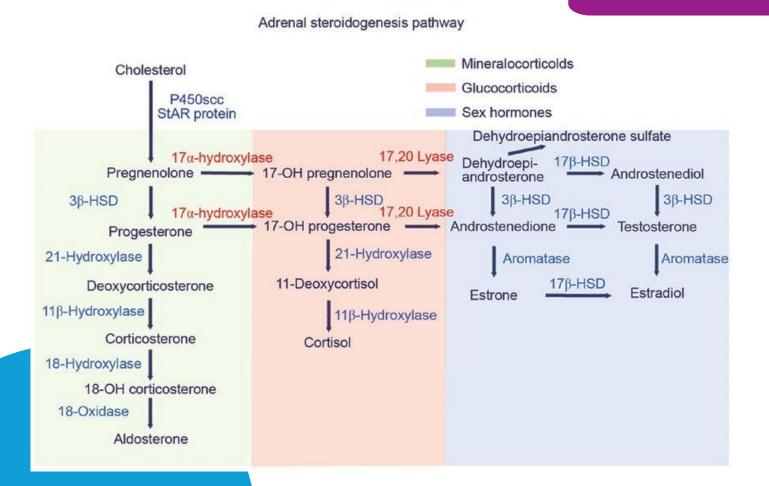
Each adrenal gland is composed of two distinct parts:

- •The Adrenal Cortex
- oThe Adrenal Medulla

# The Adrenal Cortex

### The three main adrenal hormones are:

- Mineralocorticoids = Aldosterone
- Glucocorticoids = Cortisol
- Adrenal Androgen = Dehydroepiandrosterone (DHEA)



All adrenocortical hormones are steroid compounds derived from cholesterol.



# Glucocorticoids

- Approximately 95% of glucocorticoid activity comes from cortisol.
- In normal situations, CRH, ACTH, and cortisol secretory rates demonstrate a circadian rhythm.

  - Peak- early morning (6-8 AM)Trough evening (10 PM 2 AM)
- Stressors also increase cortisol secretion.

# Cortisol

Increases and sustains blood glucose by stimulating gluconeogenesis in the liver and decreasing glucose use in the body.

Anti-inflammatory effects

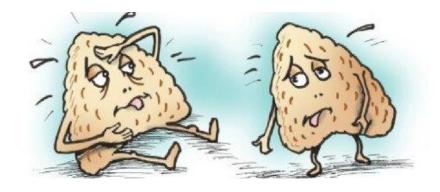
Decreases eosinophils and lymphocytes



# Glucocorticoids

### Deficiency

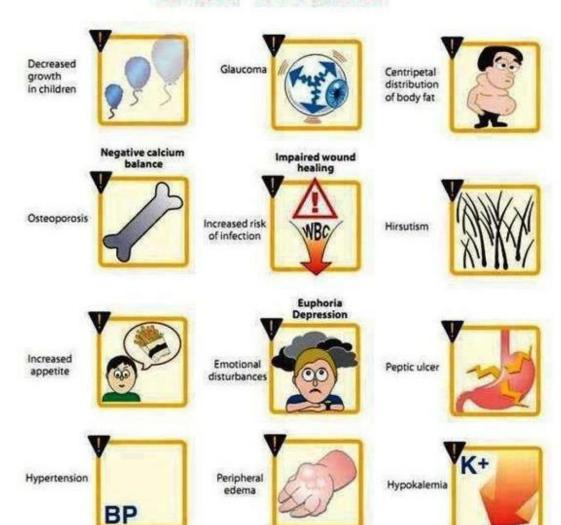
- Hypoglycemia
- Nausea, Vomiting
- Fatigue
- Muscle Weakness



### **Excess**

- Hypertension
- Weight Gain
- Poor Growth
- Stria
- Sleep Disturbance
- Anxiety
- Menstrual Irregularity

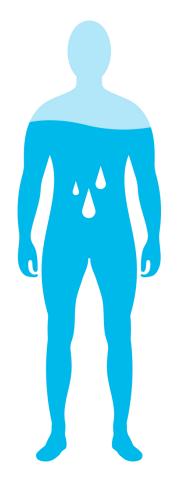
# CORTICOSTEROIDS Side Effects



# Steroid Risks

- Poor Growth
- Increased Weight Gain
- Decreased Bone Mineral Density
- Hypertension
- Adrenal Suppression

# Mineralocorticoids



Aldosterone accounts for 90% of mineralocorticoid activity

Aldosterone promotes sodium reabsorption and potassium excretion

Aldosterone also affects sodium absorption in the intestine, especially the colon.

Without aldosterone, the kidney loses excessive amounts of sodium and water.





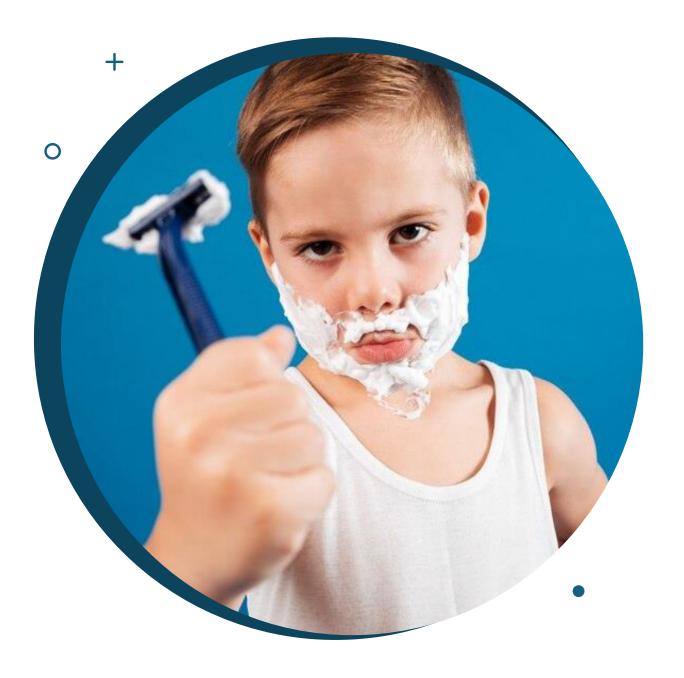
# Mineralocorticoids

### **Deficiency**

- Weight Loss
- Fatigue
- Nausea, Vomiting, Anorexia
- Salt-Craving
- Hypotension
- Hyperkalemia, Hyponatremia
- Metabolic Acidosis with Normal Anion Gap

### **Excess**

- Hypertension
- Elevated Sodium
- Low Potassium
- High Calcium
- Fatigue
- Headache
- Muscle Weakness



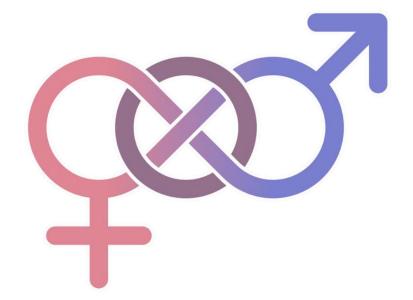
# Adrenal Androgens

- The adrenal cortex continually secretes male sex steroid hormones:
  - DHEA
  - DHEA-Sulfate (DHEAS)
  - Androstenedione
  - 11-Hydroxyandrostenedione
- These hormones are converted to testosterone.
- Play a role in the onset of puberty.

# Adrenal Androgens

### Deficiency

- Decreased Pubic and Axillary Hair
- Under-virilized 46,XY Male



### **Excess**

- Premature Adrenarche
- Acne
- Body Odor
- Advanced Bone Age
- Precocious Puberty
- Virilization of 46,XX female

# Adrenal Medulla

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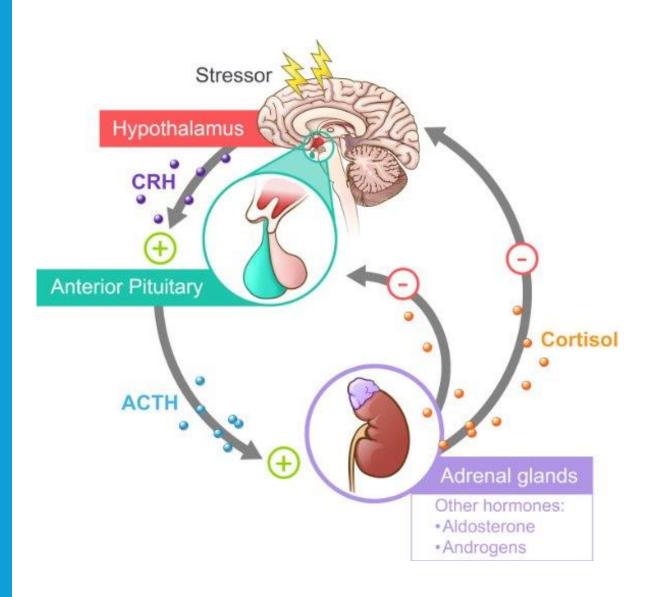
Epinephrine - 80%

Norepinephrine - 20%

Minimal amounts of Dopamine

### Function:

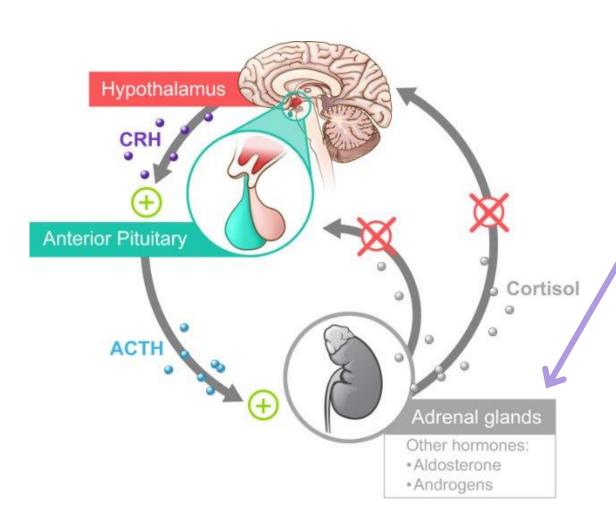
- Increase cardiac output and vascular resistance
- Physiologic stress response



# HPA Axis and Glucocorticoid Regulation



# Types of Adrenal Insufficiency



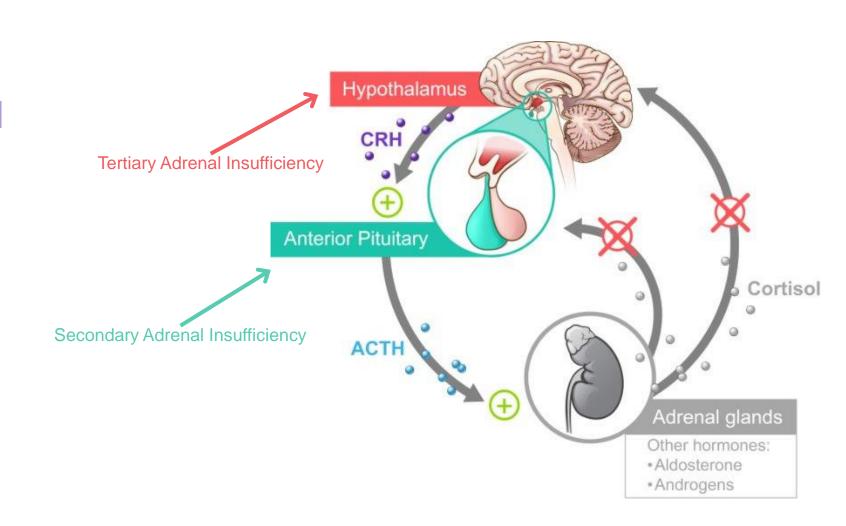
### **Primary Adrenal Insufficiency**

- Affects 10-15 per 100K individuals
- Causes include:
  - Impaired steroid synthesis (CAH)
  - Adrenal destruction/dysfunction (ALD, Addison's disease)

# Types of Adrenal Insufficiency

# Central Adrenal Insufficiency

- Secondary and tertiary AI
- 150-280 per 1M individuals
- Congenital
- Genetic
- Acquired
  - Sellar mass
  - Infiltrative
  - Autoimmune
  - Traumatic brain injury



# TREATMENT OF ADRENAL INSUFFICIENCY

- Treatment of adrenal insufficiency includes daily oral hydrocortisone replacement to mimic the body's natural stress response. This is called a "maintenance dose" and is divided every 8 hours.
- During times of fever, vomiting/diarrhea, trauma (i.e., broken bone), or surgery, children with adrenal insufficiency will need a "stress dose." This is typically double or triple the maintenance dose, depending on the severity.
- Patients who are also deficient in aldosterone will require replacement with daily oral fludrocortisone. The fludrocortisone dose does not change with stress dosing.

### **Corticosteroid Comparison Chart**

		Potency relative to Hydrocortisone		Half-Life	
	Equivalent Glucocorticoid Dose (mg)	Anti- Inflammatory	Mineral- Corticoid	Plasma (minutes)	Duration of Action (hours)
Short Acting					
Hydrocortisone (Cortef, Cortisol)	20	1	1	90	8-12
Cortisone Acetate	25	0.8	0.8	30	8-12
Intermediate Acting	Intermediate Acting				
Prednisone	5	4	0.8	60	12-36
Prednisolone	5	4	0.8	200	12-36
Triamcinolone	4	5	0	300	12-36
Methylprednisolone	4	5	0.5	180	12-36
Long Acting					
Dexamethasone	0.75	30	0	200	36-54
Betamethasone	.6	30	0	300	36-54
Mineralcorticoid					
Fludrocortisone	0	15	150	240	24-36
Aldosterone	0	0	400+	20	

Reference: Adrenal Cortical Steroids. In Drug Facts and Comparisons. 5<sup>th</sup> ed. St. Louis, Facts and Comparisons, Inc.: 122-128, 1997

### When?

Illness
Fever > 101
Mild to Moderate Trauma
Seizure
Surgery

# Oral Stress Dosing

### How?

Double or triple the maintenance dose.

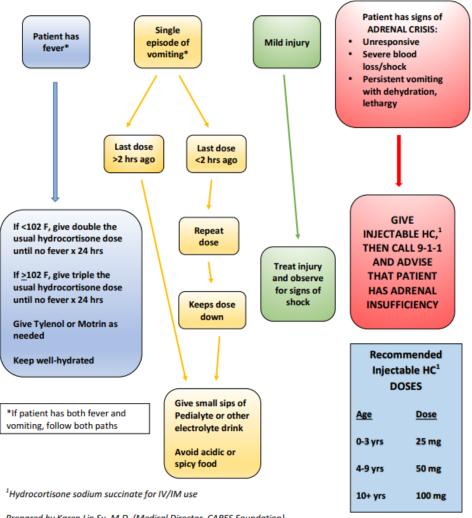
If on stress dosing for > 3 days will need to gradually taper dosing back to maintenance.

# OF ORAL STRESS DOSING

	Normal Dose	Double Dose	Triple Dose
Morning	1 pill	2 pills	3 pills
Afternoon	½ pill	1 pill	1½ pills
Bedtime	½ pill	1 pill	1½ pill

	Normal Dose	Double Dose	Triple Dose
Morning	2 mL	4 mL	6 mL
Afternoon	2 mL	4 mL	6 mL
Bedtime	1 ½ mL	3 mL	4 ½ mL

### **Stress Dose Guidelines for CAH**



Prepared by Karen Lin Su, M.D. (Medical Director, CARES Foundation)

Disclaimer: This document is intended for informational purposes only and should not be used in place of medical advice from the patient's physician or other health-care provider.

# Emergency Medical Services

Alabama EMS Adrenal Insufficiency Protocol

- Current EMS protocols of many states do not :
  - Address adrenal insufficiency
  - Allow emergency medical response personnel to treat individuals with medical ID that say "Adrenal Insufficiency"
  - Administer patient-carried medication used to treat adrenal crisis

Treat underlying cause of crisis per appropriate protocol (i.e. sepsis, fracture, burn)

Cardiac monitor

Check blood glucose, if hypoglycemic treat per hypoglycemia protocol.

Establish IV

If patient has personal medications, administer per accompanying instructions. If patient does not have personal medications or dosing instructions, utilize the following:

Hydrocortisone Sodium Succinate
100mg IM or IV
2 mg/kg IM or IV, 100mg max

Methylprednisolone 125mg IM or IV 2 mg/kg IM or IV, 125mg max

> <u>Dexamethasone</u> 5mg IM or IV

# RESOURCES







# My Adrenal Insufficiency Action Plan

My Name:

My Medications:



Normal Day

Fever or Sick or Hurt

I take my **regular** doses

Morning:

Afternoon:

Bedtime:



DO NOT FEEL GOOD I take my **stress** doses

Morning:

Afternoon:

Bedtime:

Call the Endocrine doctor if you do this for more than 3 days

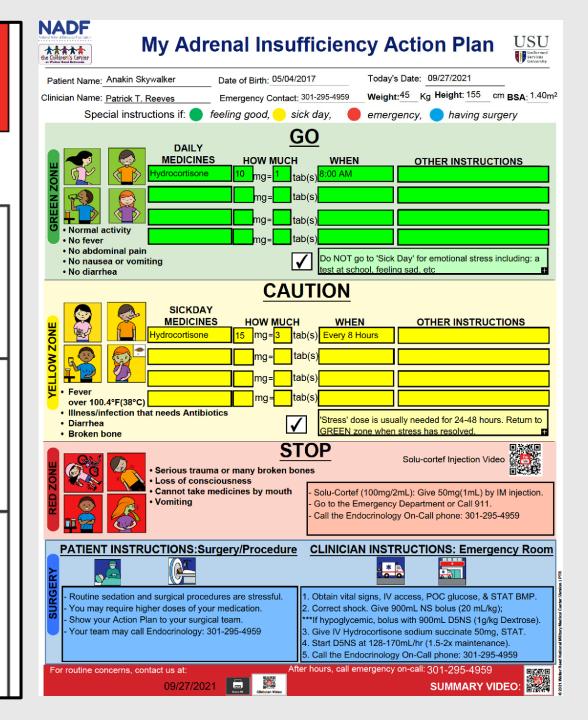


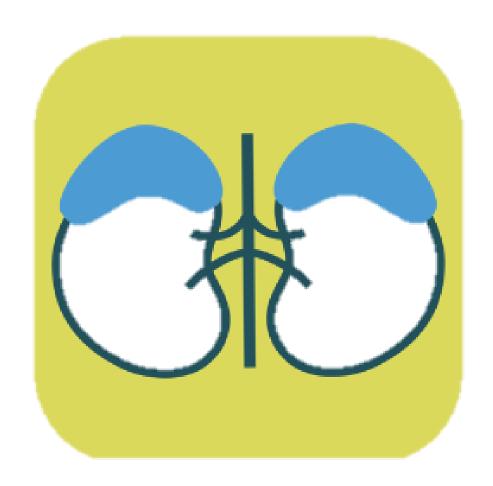
I FEEL BAD

Vomiting, Diarrhea, Fainting Unable to keep pills down I take my Emergency Shot

Solu-Cortef Dose:

Inject Solu-Cortef.
Call (205) 638-9100 and ask for the
"Endocrine Doctor On Call"
Tell them you had to use Solu-Cortef.
Go to the Emergency Room.
Tell them you have "Adrenal
Insufficiency" and you used Solu-Cortef.





# Preventing Adrenal Crisis Emergencies with the PACE App

- Apple or Android: Go to the app store and search: "PACE by ChaiCore"
- Available in English or Spanish
- The application is free and does not require cellular data or wireless internet.
- No data is collected by this application
- PACE by ChaiCore is a grantfunded research application studying efficacy of Adrenal Crisis content for informing patients about their disease.





This app is part of a research study that you have consented to participate in. It should not be downloaded on devices that do not belong to you.

The information is intended for general patient education. No personal information is relayed to the developers.

This is not intended to replace medical advice or recommendations from you or your child's provider. Please tap agree to begin using.

version 1.4

AGREE

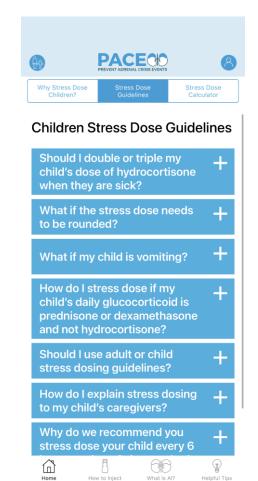






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How to Inject







Stress Dose Guidelines

Stress Dose



Why do children with AI have to stress dose?











Why Stress Dose Children?

Stress Dose Guidelines

What is the child's daily dose of hydrocortisone?



SUBMIT















### **Helpful Tips**

- · Always carry a day's supply of medicine
- · Send sick day rules to school or college nurse or clinic. Suggest a standard triple dose for the nurse to give the student until parents or caregivers get there
- Tell other providers, like a dentist or surgeon, about the person's adrenal insufficiency diagnosis
- · Drink liquids containing sugar and salt to prevent dehydration when sick (drinks like Gatorade, Pedialyte, Sprite, or 7-Up)
- · Eat a carb and protein snack before exercising. Drink water and bring a salty snack (pretzels, nuts) to eat during exercise
- · Tell emergency personnel if you or a loved one has adrenal insufficiency and is steroid dependent











#### What is AI?

What are the adrenal glands?

What is adrenal insufficiency

What causes AI?

What are symptoms of not having enough cortisol and aldosterone?

What happens if a person with











- 1. Wash your hands.
- 2. Assemble your equipment.























### **Adult Stress Dose Guidelines**

Why do adults with AI have to stress dose?



What dose of hydrocortisone do I use when I am sick?



What if I vomit?



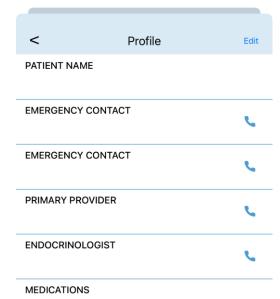












# Getting Ready for School/Camp: Parent Tips



- Contact your local CARES Support Group Leader to talk with other parents of children with CAH
- Get Medical Alert Identification
- Build a team
- Give yourself time
- Keep records/make lots of copies
- Bring a friend
- Get feedback

### 504 Plan

- A child with classical CAH is at risk of adrenal crisis and requires appropriate monitoring and care to avoid serious illness and possible death
- Though CAH is not thought of as a *disability,* children with classical CAH are at serious risk for an adrenal crisis without proper medication, monitoring of health status, access to water, and appropriate emergency medical response
- Parents can advocate for 504 plans for their child to maintain his/her health and welfare



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### **Classroom Environment**

Acceptable caregiver/student ratio and group size: # of teachers per # o children with a group size of
Seat student near the teacher to permit discreet monitoring of health
Develop individualized rules for the student to the meet student's needs
Evaluate the classroom structure against the student's needs
Seat student near a positive role model
Additional accommodations:



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Behaviors	
	Modify school rules that may discriminate against the student (e.g. water bottle in the classroom; bathroom breaks; trips to nurse's office to take medications, etc.)
	Arrange for the student to leave the classroom voluntarily to go to the nurse's office or designated "safe place" when needing medication, not feeling well, or to change clothes
	Develop a system or a code to communicate key information between student and teacher (e.g. permission to take a bathroom break, go to the nurse's office, are you feeling okay, in need of help, etc.)
	Teacher awareness/monitoring of behavior changes that relate to medication or overall health
	Develop/Use self-monitoring strategies
	Implement a classroom behavior management system
	Additional accommodations:



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Personal (	Care
	Provide access to water in classroom at all times
	Permit student unrestricted bathroom access
	Permit student to use bathroom in nurse's office or other designated "safe place"
	Change clothes/diapers in nurse's office or other designated "safe place"
Exercise a	and Physical Activity
	Full participation in physical education classes and team sports without restrictions
	Participation in physical education classes and team sports with limitations due to temperature or humidity  Specify:
	Provide PE instructors and sports coaches with training in monitoring and treatment of child
	Emergency Response Kit and water always to be available at the site of physical education class and team sports practices and games
	Additional accommodations:



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### Field Trips and Extra-Curricular Activities

Full participation in field trips and extra-curricular activities without restriction
Teachers/coaches for all field trips and extra-curricular activities will be trained in monitoring and treatment of student
School nurse or other personnel trained in monitoring and treatment of student will accompan student on all field trips
Parent/Guardian will be permitted to accompany student on field trips and during extra- curricular activities without restriction
Emergency Response Kit and water always to be available at the site of field trips and extra- curricular activities
Additional accommodations:



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#### Communication

□ Arrange "check-in/check-out" procedure to ensure parent←→teacher communication
 □ Keep student's medical information confidential, except to the extent that the student/parent/guardian decides to openly communicate about it with others
 □ Provide reasonable notice to parent/guardian when there will be change to the regular schedule such as a substitute teacher or field trips
 □ Provide each substitute teacher/caregiver with written instructions regarding the monitoring of student's health, a list of signs and symptoms of adrenal crisis and appropriate response as well as agreed upon accommodations.
 □ Attach photo of student to medical and accommodations information sheet(s) to facilitate instant recognition of student
 □ Student to wear medical alert bracelet to assist emergency personnel in responding properly
 □ Additional accommodations:



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#### **Emergency Evacuation and Shelter In-Place** Parent/Guardian will supply school/camp with a Medical Supply Kit for the child In the event of emergency evacuation or shelter-in-place situation designated staff will ensure child's Medical Supply Kit is kept with the student and will provide monitoring and medications as needed Three days medication at stress-dose levels will be kept on school grounds at all times for use in case of emergency evacuation or shelter in-place Additional accommodations: **Additional Considerations** Alert bus driver In-service training of teacher(s) on child's disability/condition Provide group/individual counseling Provide social skill group experiences Develop intervention strategies for transitional periods (e.g. cafeteria, physical education, etc.) Arrange for provision of at-home services in case of extended absence Additional accommodations:

# Causes of Adrenal Crisis

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Unknown adrenal insufficiency

Untreated adrenal insufficiency

Undertreated adrenal insufficiency

Abrupt discontinuation of long-term steroids

### Physiological Stress

- Vomiting
- Diarrhea
- Viral or Bacterial Infection
- ∘Trauma
- Broken Bones
- Seizures
- Surgery

# Adrenal Crisis Symptoms

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Nausea/ Hypotension Dehydration Shock Vomiting Acute Hypoglycemia Hyperkalemia **Abdominal Pain** Hyponatremia Hypercalcemia Anxiety Weakness Fatigue

## What does adrenal crisis look like?



# Adrenal Emergency Medication Intramuscular Solu-Cortef

(hydrocortisone sodium succinate)

Solu-Cortef injections are given for two basic reasons:

- Unable to absorb the oral dose (vomiting, repeated diarrhea)
- Decreased level of consciousness
- Prescription:
  - Act-O-Vial 100mg/2ml.
  - DOSE: 25mg, 50mg, or 100mg
  - Frequency: As needed

# Ensuring the Effectiveness and Safety of Solu-Cortef



- Storage and Stability
  - Room temperature (59-77°F) before reconstitution.
  - Protect from light and moisture to maintain stability. Do not freeze.
  - Check the expiration date printed on the vial or packaging.
     Do not use the medication past this date as it may not be effective or safe.
  - Once reconstituted, Solu-Cortef solution should be used immediately.
  - If not used immediately, Solu-Cortef should be stored in the refrigerator (36-46°F) and used within 3 days (72 hours). After this period, it should be discarded.
- Additional Notes
  - Visual Inspection: Before administration, inspect the reconstituted solution for any particulate matter or discoloration. If either is present, do not use the solution.
  - Handling: Always use aseptic techniques when handling and reconstituting the medicine to avoid contamination.

# Administering Solu-Cortef



# Future of Solu-Cortef Emergency Administration

#### CrossJet's Zeneo®

• Prefilled, automatic, disposable needle-free jet injector



### **SOLUtion Pre-filled Syringe**



SOLUtion Medical TwistJect™





Questions

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