

Congenital adrenal hyperplasia or CAH are any of several types of genetic diseases (autosomal recessive) that can result in excessive or deficient production of sex steroids or cortisol. This can have a profound effect on the reactions of the body to stress.

Thanks to Katherine Remick, MD for developing the original educational module.



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## Objectives



At the end of this educational module,  
the viewer will be able to:

- Define adrenal crisis and congenital adrenal hyperplasia
- List the causes of adrenal crisis
- Describe the appropriate field treatment for a Congenital Adrenal Hyperplasia patient in the prehospital setting
- Review state and local scope of practice

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## Anatomy Review

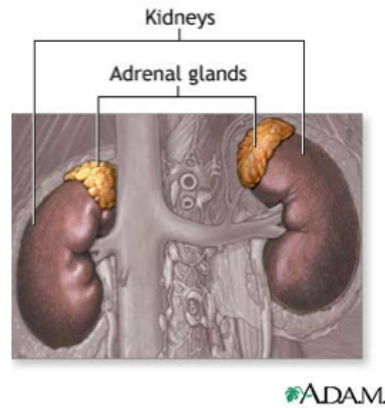
Adrenal glands- sit on top  
of both kidneys

Outer portion- cortex

Inner portion- medulla

Cortex produces:

- Cortisol
- Aldosterone
- Testosterone



The adrenal glands sit on top of both kidneys. The outer portion is the cortex and the inner portion is the medulla. The cortex produces hormones including cortisol, aldosterone, and testosterone.



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## Definitions

- Adrenal crisis is a life threatening condition that occurs when there is not enough cortisol produced
- Congenital adrenal hyperplasia (CAH) is an autosomal recessive genetic disorder which results in improper glucocorticoid hormone synthesis
- CAH patients have an enzyme deficiency, most common is 21-hydroxylase, which inhibits the conversion of cholesterol into cortisol

Adrenal crisis is a life threatening condition that occurs when there is not enough cortisol produced

Congenital adrenal hyperplasia (CAH) is an autosomal recessive genetic disorder which results in improper glucocorticoid hormone synthesis

CAH patients have an enzyme deficiency, most common is 21-hydroxylase, which inhibits the conversion of cholesterol into cortisol therefore these patients are at particular risk for having an adrenal crisis.

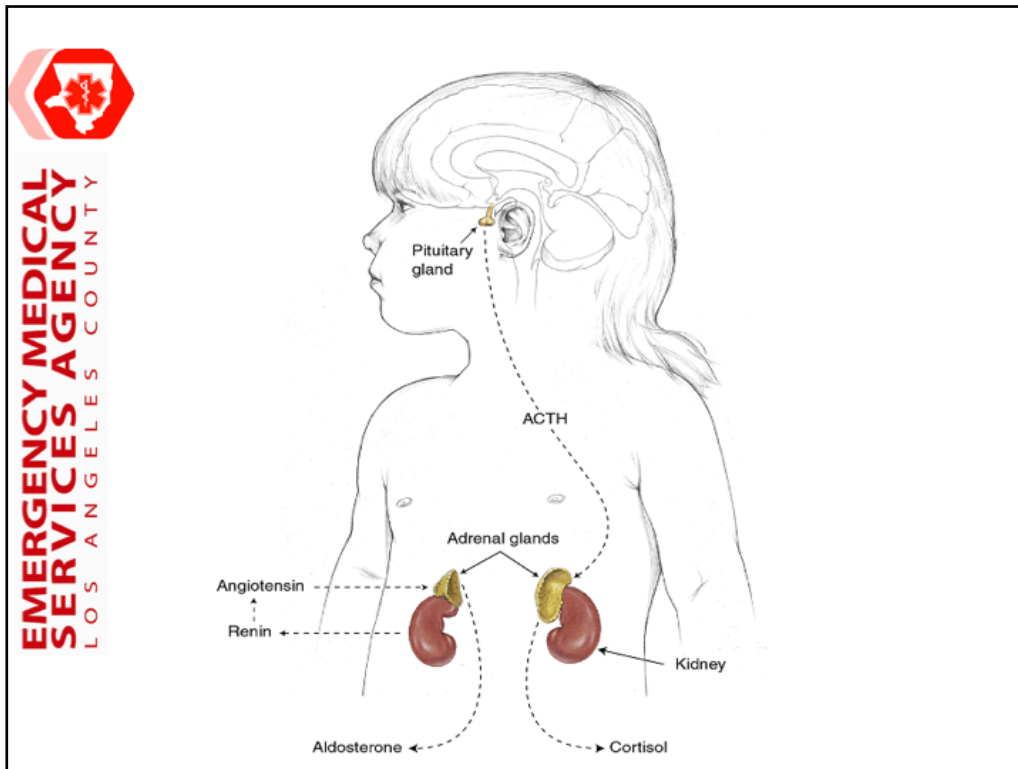


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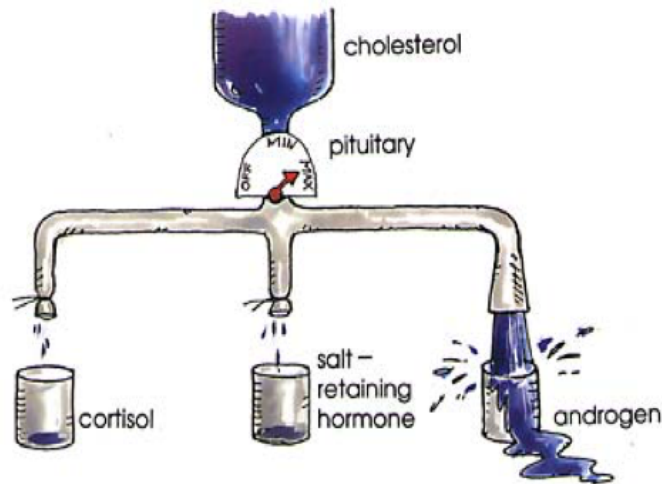
## Causes of Adrenal Crisis

- Improper cortisol synthesis from Congenital Adrenal Hyperplasia (CAH)
- Dysfunction of the adrenal cortex (Addison's disease)
- Disease of the pituitary (Hypopituitarism)
- Resistance to Adrenocorticotrophic hormone (ACTH)

Causes of adrenal crisis include deficiency anywhere along the pathway to cortisol production and release by the adrenal glands. CAH is one cause. Patients with dysfunction of the adrenal glands themselves (known as Addison's disease), those with disease of the pituitary, which releases adrenocorticotrophic hormone stimulating the adrenals to release cortisol, and those with resistance to the activity of ACTH also are at risk for adrenal crisis.



ACTH is a hormone produced in the pituitary gland that stimulates the adrenal glands to release cortisol. Abnormalities anywhere along this pathway can result in lack of a normal response to medical illness.



Patients with congenital adrenal hyperplasia lack enzymes that result in normal production of mineralocorticoids – this causes decrease in cortisol which the body needs as a thermoregulatory hormone and aldosterone which can lead to loss of salt and dehydration. The result is the overproduction also of androgens, which can result in virilization or masculinization of babies.



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## Precipitating Factors of Adrenal Crisis

- Infection
- Physical Stress
- Dehydration
- Surgery
- Sudden discontinuation of medication
- Injury to the pituitary or adrenal gland

Any of the following stresses to the normal physiology of a patient with CAH can result in life threatening consequences:

Infection

Physical Stress

Dehydration

Surgery

Sudden discontinuation of medication

Injury to the pituitary or adrenal gland





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## Signs and Symptoms of Adrenal Crisis

- Nausea
- Fever
- Pallor
- Confusion
- Weakness
- Tachycardia
- Tachypnea
- Hypotension/shock

*If left untreated, may result in death*

Signs and symptoms are not specific for adrenal crisis – so a high level of suspicion for this condition has to be maintained for a patient with CAH. These symptoms may include:

Nausea

Fever

Pallor

Confusion

Weakness

Tachycardia

Tachypnea

Hypotension/shock



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## Case Study

- Paramedics respond to the home of a 2 year-old boy
- Teenage sister states the boy began to have fever, vomiting and diarrhea several hours ago
- She states the child has congenital adrenal hyperplasia
- He is wearing a medical alert bracelet that states “adrenal insufficiency”

Let's illustrate what we are discussing by presenting a case:

Paramedics respond to the home of a 2 year-old boy

Teenage sister states the boy began to have fever, vomiting and diarrhea several hours ago

She states the child has congenital adrenal hyperplasia

He is wearing a medical alert bracelet that states “adrenal insufficiency”



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## Assessment

- The boy's vital signs:
  - HR 180, RR-30,
  - Cap refill-delayed 3 secs
  - LOC- Responds to verbal by moving head and crying
  - Skins- Hot to touch

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## Case Continues

- Sister states she tried to give the child a double dose (stress dose) of hydrocortisone (Solu-Cortef®) as prescribed by the physician, but the child immediately vomited it up.
- She attempted to contact her parents but they did not answer her phone call.
- Her mother usually gives him an IM injection of hydrocortisone (Solu-Cortef®) when he cannot tolerate medication by mouth.
- She has his injection kit next to her.



Sister states she tried to give the child a double dose (stress dose) of hydrocortisone (Solu-Cortef®) by mouth as prescribed by the physician, but the child immediately vomited it up.

She attempted to contact her parents but they did not answer her phone call.

Her mother usually gives him an IM injection of hydrocortisone (Solu-Cortef®) when he cannot tolerate medication by mouth.

She has his injection kit next to her.




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## Questions?

- What should the paramedic do?
- What can the paramedics do?

*What should the paramedic do?*

*What can the paramedics do?*

	<h2>California Scope of Practice</h2>	
<b>EMERGENCY MEDICAL SERVICES AGENCY</b> LOS ANGELES COUNTY	<u><b>CALIFORNIA EMT-I REGULATIONS - LOS ANGELES COUNTY POLICIES</b></u>	<u><b>CALIFORNIA PARAMEDIC REGULATION -LOS ANGELES COUNTY POLICIES</b></u>
	<p>Basic Scope - all individuals certified in California must be trained and tested in these practices:</p> <p><i>Assist patients with the administration of physician prescribed devices, including but not limited to:</i></p> <p><i>patient operated medication pumps, sublingual nitroglycerin, and <u>self-administered emergency medications</u>, including epinephrine devices.</i></p>	<p>Basic Scope - all individuals licensed in California must be trained and tested in these practices:</p> <p><i>Administer approved medications by the following routes: intravenous, <u>intramuscular</u>, subcutaneous, inhalation, transcutaneous, rectal, sublingual, endotracheal, oral or topical</i></p>

Per State of California Title 22, Division 9, Prehospital Emergency Medical Services, Chapter 2, EMT 100063: Scope of Practice of EMT – (16) “Assist patients with the administration of physician prescribed devices including but not limited to, patient operated medication pumps, sublingual nitroglycerin, and self-administered emergency medications, including epinephrine devices”

Paramedics may perform any activity in the scope of practice of an EMT – therefore can assist patients in delivering normally self-administered medications – such as hydrocortisone in this case.



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## Key Points

- A paramedic operates under both EMT and paramedic Scope of Practice
- A paramedic can assist with the emergency administration of hydrocortisone (Solu-Cortef®) IM which falls under both EMT and paramedic scope of practice
- Contact the base hospital with any questions
- These patients must be transported to the hospital for further evaluation/treatment

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## Summary

- Under both EMT and Paramedic regulations, emergency medications prescribed by a physician can be given with the assistance of parents/caregivers
- **These medications should be given as soon as possible and can be life saving!**

Under both EMT and Paramedic regulations, emergency medication can be given with the assistance of parents/caregivers

Call the base hospital for any questions/direction

**These medications should be given as soon as possible and can be life saving**