

Recurrent, unexplained seizures

Intense craving for salt

Acute abdominal pain

Electrolyte imbalances

Definitive diagnosis is based on measurements of functional cortisol reserve. The fasting serum cortisol and urinary 17-hydroxycorticosteroid levels are low and fail to rise, and plasma **adrenocorticotrophic hormone (ACTH)** levels are elevated with corticotropin (ACTH) stimulation, the definitive test for the disease.

Therapeutic Management

Treatment involves replacement of **glucocorticoids (cortisol)** and **mineralocorticoids (aldosterone)**. Some children are able to be maintained solely on oral supplements of cortisol (cortisone or hydrocortisone preparations) with a liberal intake of salt. During stressful situations (such as fever, infection, emotional upset, or surgery), the dosage must be tripled to accommodate the body's increased need for glucocorticoids. Failure to meet this requirement will precipitate an acute crisis. Overdosage produces appearance of cushingoid signs.

Children with more severe states of chronic adrenal insufficiency require mineralocorticoid replacement to maintain fluid and electrolyte balance. Other forms of therapy include monthly injections of desoxycorticosterone acetate or implantation of desoxycorticosterone acetate pellets subcutaneously every 9 to 12 months.

Nursing Care Management

After the disorder is diagnosed, parents need guidance concerning drug therapy. They must be aware of the continuous need for cortisol replacement. Sudden termination of the drug because of inadequate supplies or inability to ingest the oral form because of vomiting, places the child in danger of an acute adrenal crisis. Parents should always have a spare supply of medication. Ideally, families will have a prefilled syringe of hydrocortisone and have