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Management of salt-wasting congenital adrenal hyperplasia in pediatric patients

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Description

Congenital Adrenal Hyperplasia (CAH) is a group of autosomal recessive disorders characterized by enzymatic deficiencies in the cortisol biosynthesis pathway. Among the various forms of CAH, salt-wasting CAH is one of the most severe, posing significant challenges in the management of pediatric patients. Diagnosis of salt-wasting CAH typically occurs in the neonatal period or early infancy through newborn screening or clinical presentation with adrenal crisis. Newborn screening involves measuring 17-hydroxyprogesterone (17-OHP) levels, with elevated levels prompting confirmatory testing such as ACTH stimulation test and genetic analysis. Early diagnosis is crucial for initiating prompt treatment and preventing life-threatening adrenal crises.

Adrenal crises are medical emergencies characterized by severe adrenal insufficiency, electrolyte imbalances and hypoglycemia. Prompt recognition and management are essential to prevent morbidity and mortality in pediatric patients with salt-wasting CAH. Acute management involves the administration of intravenous fluids, glucocorticoids (hydrocortisone) and correction of electrolyte abnormalities, particularly hyponatremia and hyperkalemia. Close monitoring of vital signs, electrolytes and blood glucose levels is imperative during and after treatment.

The foundation for long-term management for salt-wasting CAH in pediatric patients is hormone replacement therapy to replace deficient cortisol and mineralocorticoid hormones. The goals of therapy include achieving normal growth and development, maintaining electrolyte balance and preventing adrenal crises. Hormone replacement therapy typically consists of oral glucocorticoids (e.g., hydrocortisone, prednisone) and mineralocorticoids (e.g., fludrocortisone).

Glucocorticoids replace deficient cortisol production, suppressing adrenal androgen excess and reducing ACTH levels. Hydrocortisone is the preferred glucocorticoid due to its pharmacokinetic profile resembling endogenous cortisol secretion. The dosage is individualized based on age, weight, and clinical response, with divided doses mimicking the diurnal cortisol rhythm. Fludrocortisone replaces deficient aldosterone production, promoting sodium retention and potassium excretion to prevent salt wasting and electrolyte imbalances. The initial dosage is titrated to achieve normal serum electrolyte levels, with close monitoring of blood pressure and electrolytes to prevent over- or under-replacement.

Some patients with salt-wasting CAH may require additional sodium supplementation during periods of illness or high ambient temperatures to prevent dehydration and electrolyte disturbances. Regular monitoring and follow-up are essential components of managing salt-wasting CAH in pediatric patients to

assess treatment efficacy, adjust medication dosages and monitor for complications. Regular assessment of height, weight and developmental milestones to ensure appropriate growth and development. Periodic measurement of serum sodium, potassium and chloride levels to monitor for electrolyte imbalances. Regular blood pressure measurements to detect and manage hypertension or hypotension. Monitoring blood glucose levels to detect hypoglycemia or hyperglycemia associated with glucocorticoid therapy. Periodic measurement of 17-OHP levels and ACTH stimulation tests to assess adrenal suppression and treatment adequacy.

With appropriate management, the long-term prognosis for pediatric patients with salt-wasting CAH is generally favorable. However, long-term glucocorticoid and mineralocorticoid therapy may be associated with complications. Despite treatment, patients remain at risk of adrenal crises, emphasizing the importance of adherence to medication regimens and recognition of stress situations. Prolonged glucocorticoid therapy may impair linear growth, necessitating close monitoring and adjustments to minimize growth retardation.

Chronic glucocorticoid therapy increases the risk of osteoporosis and fractures, highlighting the importance of calcium and vitamin D supplementation and regular bone density assessments. Glucocorticoid-induced obesity, insulin resistance and dyslipidemia may develop over time, necessitating lifestyle modifications and monitoring of metabolic parameters.

Conclusion

In conclusion, management of salt-wasting congenital adrenal hyperplasia in pediatric patients requires a multidisciplinary approach involving pediatric endocrinologists, nurses, dietitians and caregivers. Early diagnosis, prompt recognition and aggressive management of adrenal crises are important to prevent morbidity and mortality. Chronic hormone replacement therapy with glucocorticoids and mineralocorticoids aims to achieve normal growth, maintain electrolyte balance and prevent long-term complications. Regular monitoring and follow-up are essential to assess treatment efficacy, adjust medication dosages and monitor for complications, ultimately optimizing long-term outcomes and quality of life for pediatric patients with salt-wasting CAH.