

Primary adrenal insufficiency: About a pediatric series

INTRODUCTION

Adrenal insufficiency in children is rare and potentially serious because of the risk of acute adrenal insufficiency, which can occur at any time. This complication is lethal in the absence of prompt and appropriate treatment. etiologies are dominated by the genetic causes that are far the most frequent

Objective:

Report diagnostic circumstances ,phenotypic forms and causes of adrenal insufficiency in children

Population, Methodology

This is a retrospective study of n cases of children and adolescents with adrenal insufficiency hospitalized in the department of endocrinology between January 1988 and December 2014 we evaluated the clinical, biological, etiological and evolutive features of adrenal insufficiency in children and adolescents

Results

54 cases were reported (44% boys, 56% girls) The average age was 6 years ± 1.2 (2-18)
The consultation Reason: AIS: 59%, adrenal insufficiency: 20%, Congenital adrenal hyperplasia: 5% melanoderma

A familial form was observed in 14% (10% PEA, 4% HCS)
Clinical feature was typical 42% (emaciation, asthenia, anorexia, hypotension melanoderma, digestive disorders).
Paucisymptomatic in 58%
Biologically, the Serum potassium was high in 44.44% serum Na was low in 42.59% . glucose was low in 12.94% and symptomatic in 50%
The etiologies were HCS 73% (21 OH 67% 11 OH 3% B OL 2% 17 β OH 1%) IS autoimmune 22.6% Allgrove sd 1.4%, tuberculosis 1.8%, adrenoleukodystrophy 1.8%
Replacement therapy has helped improve symptoms in all cases
The evolution was complicated iterative decompensation of 45% and difficulties of balancing (overdose 30%)



DISCUSSION

The existence of even isolated melanoderma should suggest adrenal insufficiency and make the necessary investigations in order to avoid serious complications. An etiological exploration is essential for proper therapeutic management.

Furthermore, the bases of the treatment based on the substitution .. glucocorticoid and mineralocorticoid if necessary, and .on patient education actively involved in prevention acute accidents.

Conclusion

Adrenal insufficiency is rare in children. Genetic diseases dominated by HCS are the most common causes in infants and very young children. Dehydration with a salt-losing syndrome should suggest this etiology. In the older child, the autoimmune origin is predominant. It can be isolated or associated with a polyendocrinopathy that it will identify early. Replacement therapy started quickly should be reviewed and monitored to prevent under and overdoses

