

Classical congenital adrenal hyperplasia: about 7 cases

N. Draoui(Dr), N.Zeryouh (Dr) I. Assarrar (Dr), S. Rouf (Pr), H. Latrech (Pr)

Department of Endocrinology-Diabetology and Nutrition, Mohammed VI University Hospital Center, Faculty of Medicine and Pharmacy, University of Mohammed Ist, Oujda, Morocco.

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Introduction:

Congenital adrenal hyperplasia (CAH) is a group of autosomal recessively inherited disorders of various enzymes participating in the adrenal steroidogenesis. 21-hydroxylase deficiency (21-OHD) is the most common type of CAH (90–95%). Less frequent types of CAH are 11 β -hydroxylase deficiency (up to 8% of cases), 17 α -hydroxylase deficiency, 3 β -hydroxysteroid dehydrogenase deficiency, P450 oxidoreductase deficiency and StAR deficiencies.

Materiel and methods:

This is a retrospective descriptive study of 10 patients with congenital adrenal hyperplasia, who are followed-up in the Department of Endocrinology-Diabetology-Nutrition of Mohammed VI University Hospital Center of Oujda in the eastern Morocco

Resultat:

In our study 7 patients had classical congenital adrenal hyperplasia; four of these female patients were born with atypical genitalia, three male patients had neonatal salt wasting. Five of these patients had 21-hydroxylase deficiency and 2 patients had 11 β -hydroxylase deficiency (figure1). Female patients with atypical genitalia benefited from genital surgery. All of our patients are treated with hormone replacement therapy.

Discussion-conclusion:

Female infants who have classic CAH born with different degrees of ambiguous genitalia, while male infants with classic CAH born with a normal appearing genitals, but both male and female infants with congenital adrenal hyperplasia can have various alterations in glucocorticoid, mineralocorticoid, and sex steroid production, which in some cases require hormone replacement therapy and this is the case for all our patients; the 21-hydroxylase deficiency form is the most common type of CAH which is coherent with our results, followed by the 11 β -hydroxylase deficiency

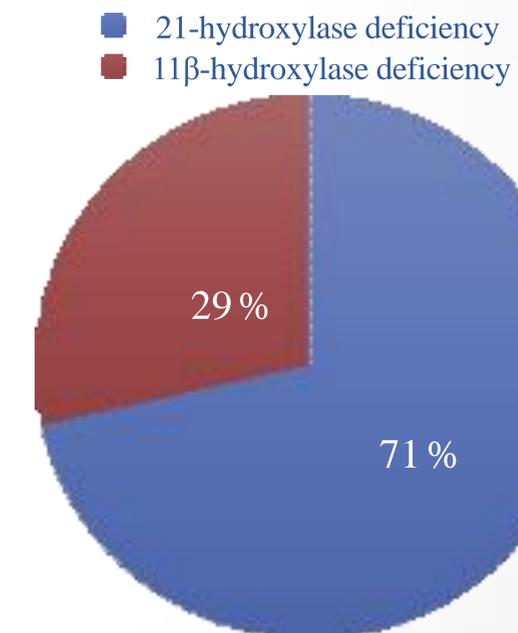


Figure 1 : the repartition of the different deficiencies