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Defining the dose, type and timing of glucocorticoid and mineralocorticoid replacement in 256 children and adults with congenital adrenal hyperplasia (CAH) in the I-CAH registry

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Abstract

Objectives: Physiological replacement is important for optimal control of congenital adrenal hyperplasia (CAH). We examined glucocorticoid and mineralocorticoid replacement in children and adults with CAH.

Methods: Data were extracted in February 2017 for 22 centres in 14 countries from the international I-CAH registry (www.i-cah.org). 1501 events from 269 patients seen between 1987 and 2017 were analyzed.

Results: 256 patients had information on glucocorticoids (F 136, M 116, 4 sex not assigned; 0-1y n=130, 69F, 1-8y n=153 82F, 8-12y n=42 26F, 12-18y n=39 23F, 18-30y n=27 12F, 30-60y n=26 14F). The majority of pediatric patients were treated with hydrocortisone (HC) and adults with prednisolone (Pred) and some with cortisone acetate (CA) and dexamethasone (DEX); 0-1y: HC 92%, CA 8%, Dex 1%, 1-8y: HC 93%, CA 6%, Pred 1%, 8-12y: HC 83%, CA 7%, Dex 5%, Pred 5%, 12-18y: HC 69%, CA 3%, Dex 18%, Pred 10%, 18-30y: HC 33%, CA 4%, Dex 26%, Pred 37%, 30-60y: HC 31%, Dex 12%, Pred 54%. The HC-equivalent dose varied significantly between age groups, $p=0.02$ (mean \pm sd in mg/m²/day); 0-1y (15.3 \pm 8.3), 1-8y (13.6 \pm 12.3), 8-12y (15.2 \pm 5.9), 12-18y (15.7 \pm 6.8), 18-30y (16.0 \pm 5.1), 30-60y (12.2 \pm 5.8). Information on mineralocorticoids was available in 227 patients (F 119, M 105, 3 sex not assigned). Average fludrocortisone dose and frequency of administration was (mean \pm sd, frequency in % of patients); 0-1y (101.2 \pm 62.1mcg, od 59%/ bd 32%/ tds 11%), 1-8y (91.07 \pm 61.7mcg, od 70%/ bd 26%/ tds 4%), 8-12y (84.41 \pm 44.1mcg, od 82%/ bd 32%/ tds 3%), 12-18y (111.4 \pm 52.6mcg, od 81%/ bd 19%), 18-30y (134.5 \pm 68.2mcg, od 90%/ bd 10%), 30-60y (152.9 \pm 74.4mcg, od 71%/ bd 29%). Total fludrocortisone dose mcg/m²/day was significantly higher in children younger than 8y, $p<0.0001$ (mean \pm sd): 0-1y (274.2 \pm 181.7), 1-8y (146.7 \pm 129.7), 8-12y (63.3 \pm 34.3), 12-18y (66.43 \pm 34.3), 18-30y (77.1 \pm 37.1), 30-60y (74.8 \pm 37.9).

Conclusions: Data from a large international cohort of CAH patients confirm variations in the hormonal replacement regimens between pediatric and adult patients. Glucocorticoid doses were high in some age groups compared to recommendations in current guidelines.

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