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**Proceedings Paper:**

Daniel, E., Sandrk, M., Blankenstein, O. et al. (2017) 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. In: Hormone Research in Paediatrics. 10th International Meeting of Pediatric Endocrinology, 14-17 Sep 2017, Washington D.C.. 10th Individual Abstracts for International Meeting of Pediatric Endocrinology: Free Communication and Poster Sessions, Abstracts, 88, S1. Karger Publishers, p. 358. ISSN: 1663-2818. EISSN: 1663-2826.

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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

## Authors

Eleni Daniel<sup>1</sup>, Marija Sandrk<sup>2</sup>, Oliver Blankenstein<sup>3</sup>, Uta Neumann<sup>3</sup>, Hedi Claahsen-van der Grinten<sup>4</sup>, Annelieke van der Linde<sup>4</sup>, Feyza Darendeliler<sup>5</sup>, Sukran Pyrazoglu<sup>5</sup>, Berenice B Mendonca<sup>6</sup>, Tania S S Bachega<sup>6</sup>, Mirela C Miranda<sup>6</sup>, Carlo Acerini<sup>7</sup>, Tulay Guran<sup>8</sup>, Ana Vieites<sup>9</sup>, Niels H Birkebaek<sup>10</sup>, Martine Cools<sup>11</sup>, Tatjana Milenkovic<sup>12</sup>, Walter Bonfig<sup>13</sup>, Jeremy William Tomlinson<sup>14</sup>, Faisal Ahmed<sup>15</sup>, Heba Elsedfy<sup>16</sup>, Antonio Balsamo<sup>17</sup>, Sabine E Hannema<sup>18</sup>, Claire Higham<sup>19</sup>, Navoda Atapattu<sup>20</sup>, Corina Lichiardopol<sup>21</sup>, Ruth E Krone<sup>22</sup>, Klaus Mohnike<sup>23</sup>, Richard J Ross<sup>1</sup>, Nils Krone<sup>2</sup>

## Affiliations

1. Department of Oncology and Metabolism, University of Sheffield, Sheffield, United Kingdom
2. Academic Unit of Child Health, University of Sheffield, Sheffield, United Kingdom
3. Institute for Experimental Pediatric Endocrinology, Charite-Universitätsmedizin Berlin, Berlin, Germany
4. Department of Pediatric Endocrinology, Radboud University Medical centre, Nijmegen, the Netherlands
5. Pediatric Endocrinology Unit, Istanbul Faculty of Medicine, Istanbul, Turkey
6. Department of Internal Medicine, University of Sao Paulo, Sao Paulo, Brazil
7. Department of Pediatrics, University of Cambridge and Addenbrooke's Hospital, Cambridge, United Kingdom
8. Pediatric Endocrinology and Diabetes, Marmara University, Istanbul, Turkey
9. Centro de Investigaciones Endocrinológicas (CEDIE-CONICET), Hospital de Niños Ricardo Gutiérrez, Buenos Aires, Argentina
10. Department of Pediatrics, Aarhus University Hospital, Aarhus, Denmark
11. Pediatric Endocrinology, Pediatric and Genetics Research Unit, University Hospital Ghent, Ghent University, Ghent, Belgium
12. Department of Endocrinology, Institute for Mother and Child Healthcare of Serbia "Dr Vukan Čupić" Belgrade, Serbia
13. Department of Pediatrics, Technical University of Munich, Munich
14. Oxford Centre for Diabetes, Endocrinology & Metabolism, Churchill Hospital, Oxford, United Kingdom
15. Pediatric Endocrinology, University of Glasgow, Glasgow, United Kingdom
16. Pediatric Endocrinology, Ain Shams University, Cairo, Egypt
17. Medical and Surgical Sciences, Pediatric Unit, Center for Rare Endocrine Diseases (CARENDO BO), S.Orsola-Malpighi University Hospital, Bologna, Italy
18. Department of Pediatric Endocrinology, Sophia Children's Hospital, Erasmus Medical Centre, Rotterdam, Netherlands
19. Department of Endocrinology, Christie Hospital NHS Foundation Trust, Manchester, United Kingdom
20. Pediatric Endocrinology, Lady Ridgeway Hospital, Colombo, Sri Lanka
21. Department of Endocrinology, University of Medicine and Pharmacy Craiova, Craiova, Romania

22. Department of Endocrinology and Diabetes, Birmingham Women's and Children's Hospital, Birmingham, United Kingdom  
23. Department of Pediatrics, Otto-von-Guericke University, Magdeburg, Germany

## 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](http://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/m<sup>2</sup>/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/m<sup>2</sup>/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.

**Acknowledgements:** Funded by MRCUK-G1100236, EUFP7-201444, EUFP7-281654.  
**Conflicts:** RJR is a Director of Diurnal Plc.