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## Introduction and Objectives

Despite existing guidelines, there is no unified approach to glucocorticoid (GC) and mineralocorticoid replacement in congenital adrenal hyperplasia (CAH). We used data from the I-CAH Registry to identify geographical and temporal variations in the treatment with glucocorticoids and mineralocorticoids of children and adults with CAH.

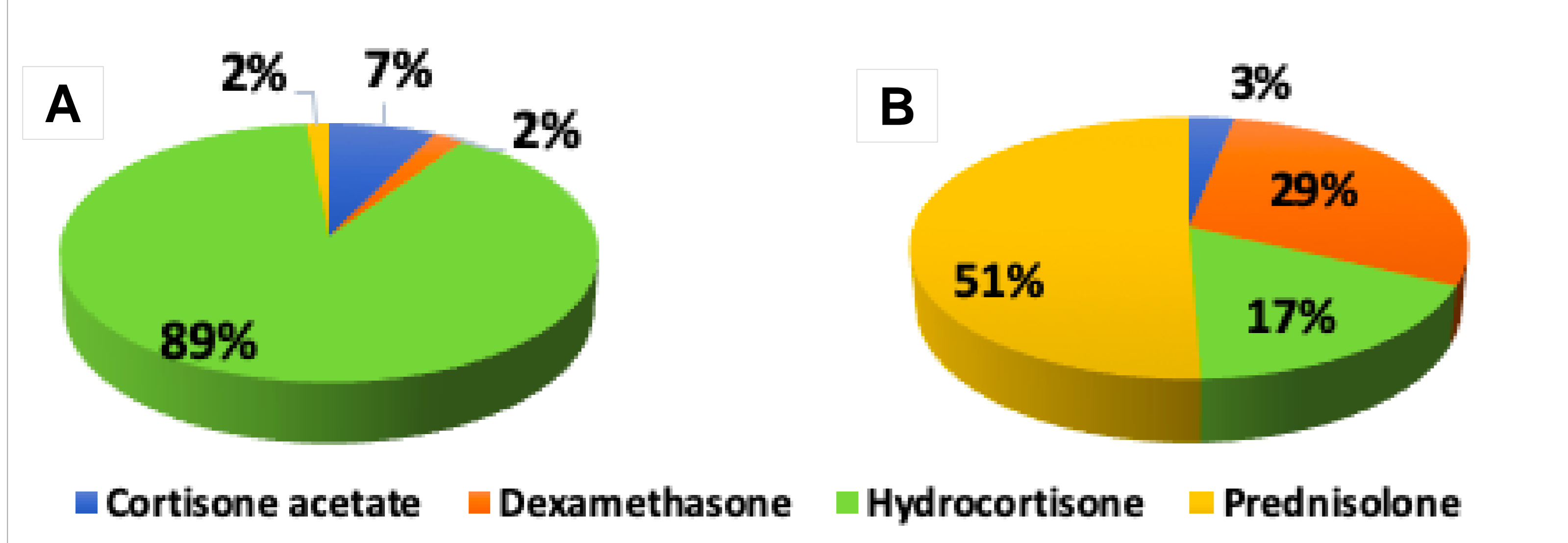
## Conclusion

There are international variations in hormone replacement therapy, with a tendency for high doses in younger patients. Further evidence regarding related health outcomes is needed to help improve the medical management of patients with CAH.

## Results

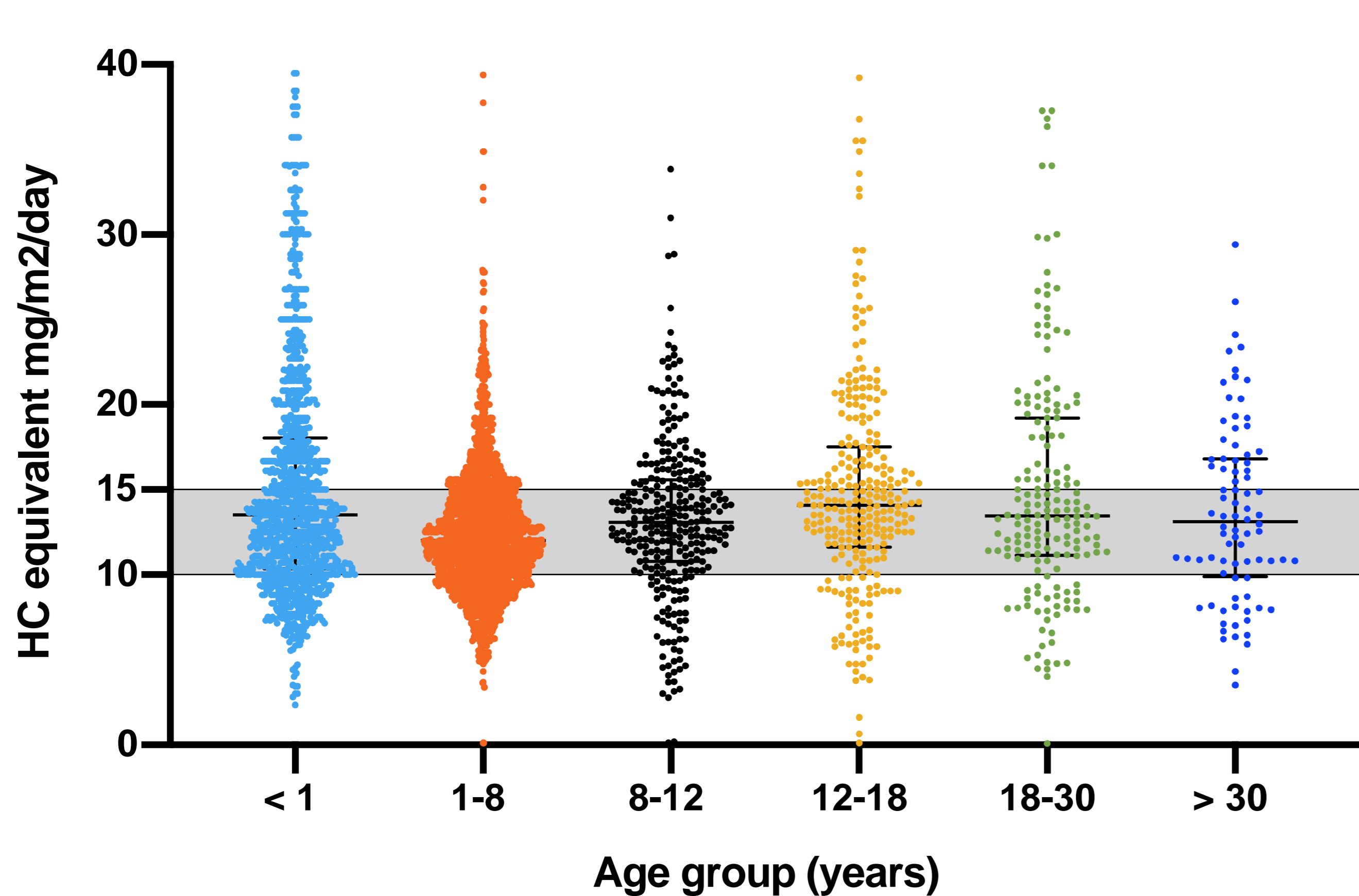
The most frequently used glucocorticoids were hydrocortisone in children (88%), prednisolone (51%) and dexamethasone (28%) in adults.

Figure 1. Types of glucocorticoids used in children (A) and adults (B)



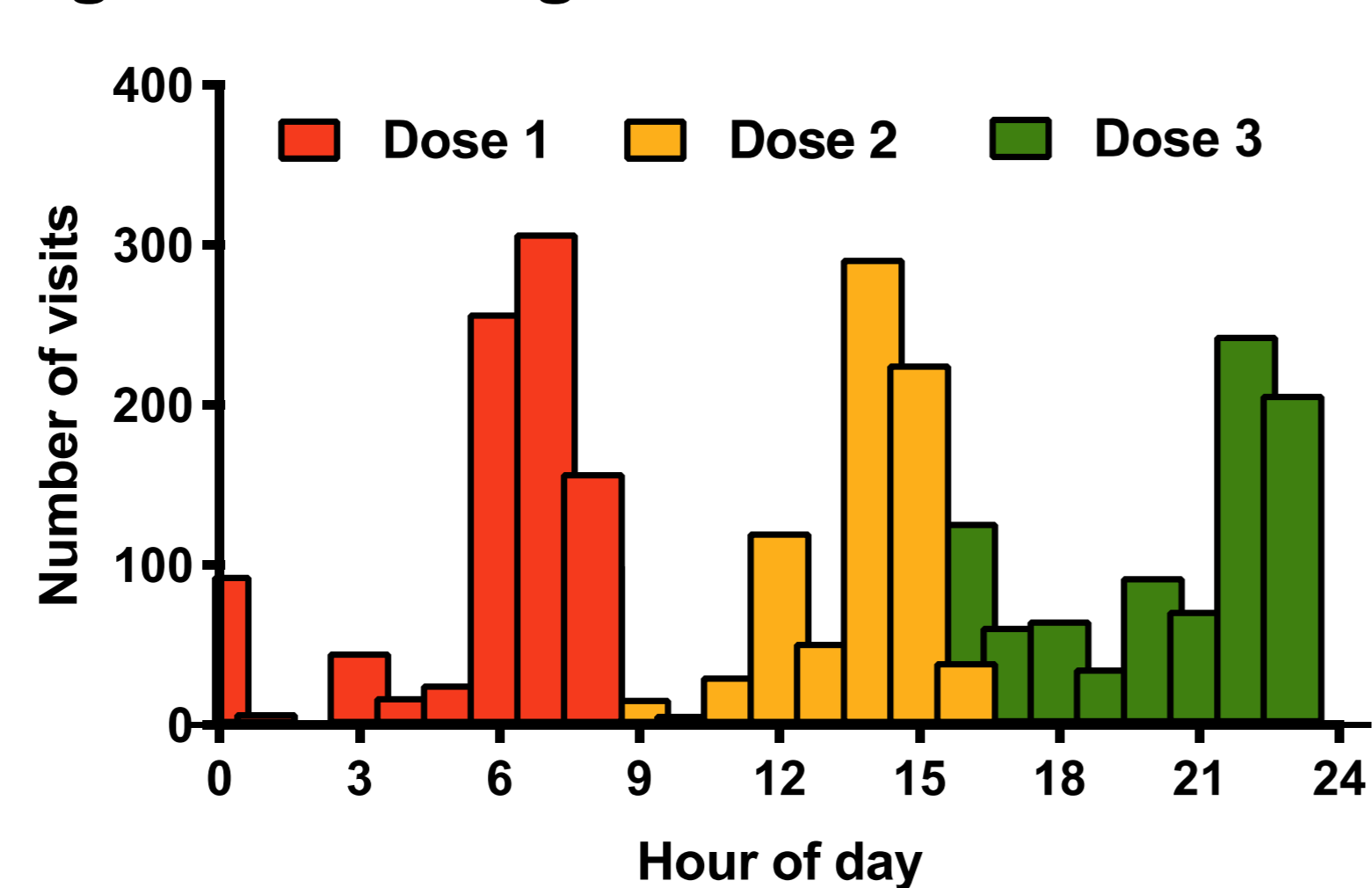
Glucocorticoid doses varied significantly between age groups, frequently exceeding the recommended range of 10-15 hydrocortisone (HC) equivalent mg/m<sup>2</sup>/day.

Figure 2. Glucocorticoid doses used in different age groups



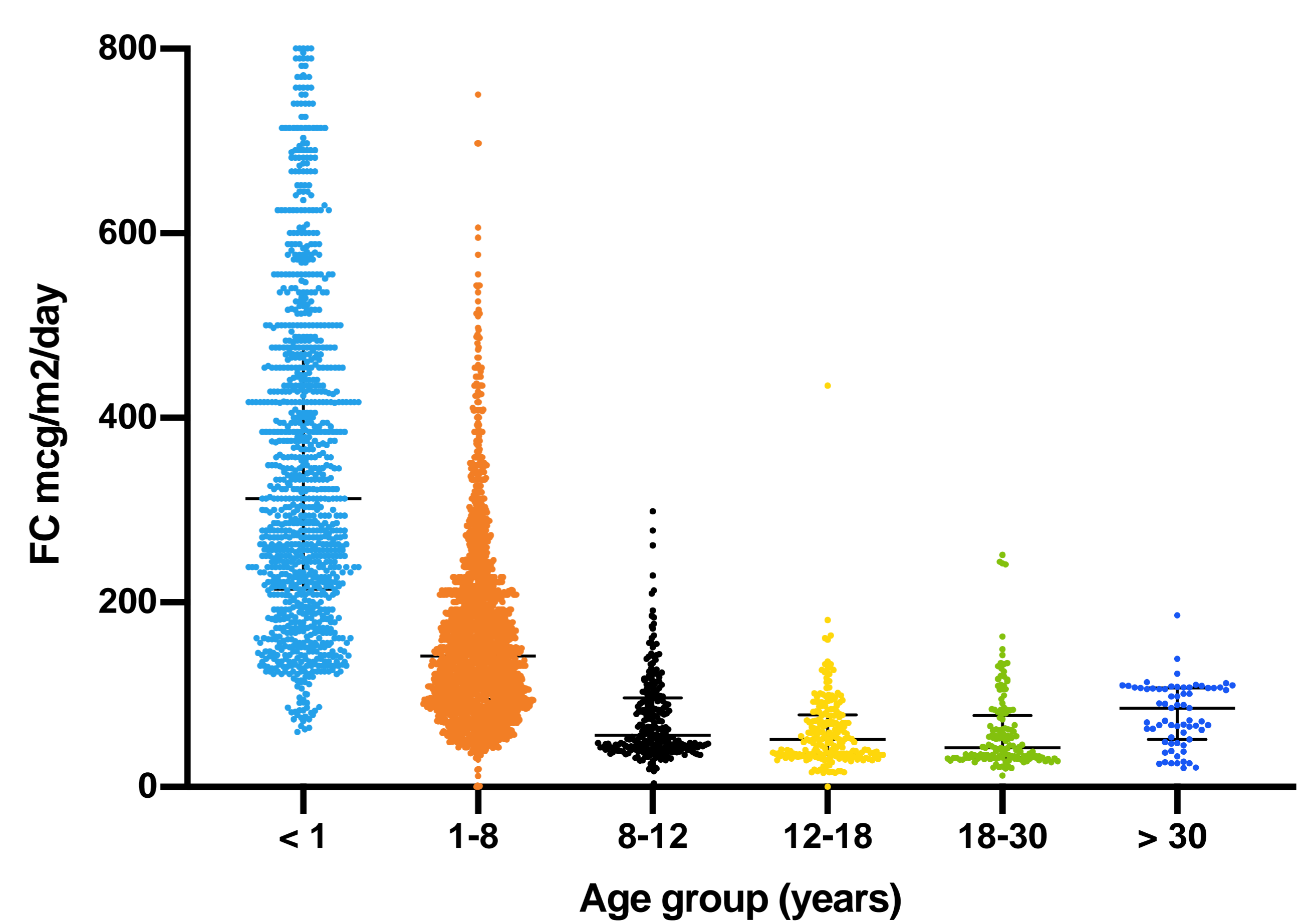
Most children received three GC doses per day (74%); adults frequently received one (49%) or two (34%) daily doses. There was large variability in timing of doses throughout the day, especially in children.

Figure 3. Timing of GC doses in children



Fludrocortisone (FC) was used in 80.9% of patients, with significant variability of relative doses (mcg/m<sup>2</sup>/day) among age groups.

Figure 4. Fludrocortisone doses for different age groups



We found large variations in hormone replacement regimens among different countries, in particular with regards to doses of GC used in children.

Figure 5. GC doses in children from different countries (Analysis limited to countries with over 50 doses recorded)

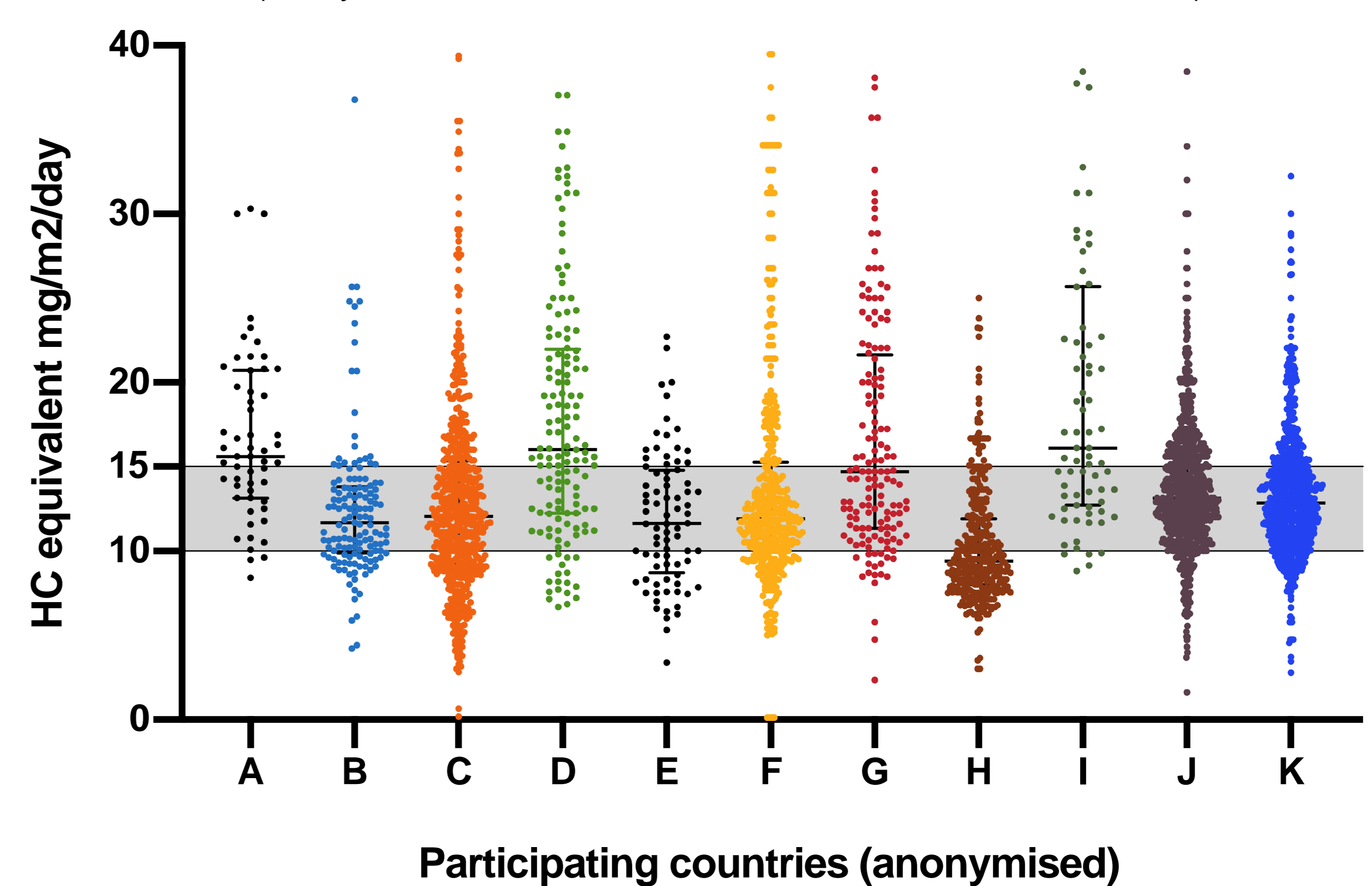


Table. Gluco- and mineralocorticoid doses (median with interquartile range) used in children from different countries (anonymized). The analysis only included countries with over 50 doses recorded.

Country	A	B	C	D	E	F	G	H	I	J	K
HC equivalent (mg/m <sup>2</sup> /day)	15.6 (13.1-20.7)	11.6 (9.8-13.8)	12.0 (9.3-15.3)	16.0 (12.1-21.9)	11.6 (8.7-14.7)	11.9 (10-15.2)	14.7 (11.3-21.6)	9.4 (8.0-11.9)	16.1 (12.7-25.6)	13.1 (11.3-15.6)	12.8 (10.9-15.0)
FC (mcg/m <sup>2</sup> /day)	140 (69-231)	107 (81-212)	151 (48-441)	250 (173-340)	175 (100-256)	170 (115-294)	169 (115-238)	169 (125-263)	110 (86-172)	161 (87-256)	147 (95-240)

## Methods

We analysed data from 618 patients (350 females), 4866 patient visits (31 centres from 16 countries) between 1982 and 2018 with regards to the type, dose and timing of glucocorticoid and mineralocorticoid replacement. Hydrocortisone (HC) conversion: 20mg hydrocortisone = 4mg prednisolone = 750µg dexamethasone = 25mg cortisone acetate.