Adrenal crisis and sick day episodes among CAH patients: Preliminary report based on International CAH (I-CAH) registry

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Background

Congenital adrenal hyperplasia (CAH) is a rare condition that is associated with life long risk of adrenal crisis. This is a life-threatening event and account for increased mortality particularly in children (1,2). Management of CAH demands a fine balance between excess glucocorticoid leading to adverse effects and too little glucocorticoid risking adrenal crises (3). Frequent occurrence of sick day episodes warrants dose adjustment and education regarding adrenal crisis. In a condition such as CAH it is difficult to collect sufficient data from small cohorts at a single centre level to establish clinical significance. To address this issue we have used the I-CAH registry to investigate the frequency of sick day episodes and adrenal crisis.

Methodology

- The I-CAH registry (http://www.icah.org) is a part of the I-DSD registry(http://www.i-dsd.org), which has national ethics approval as a pseudoanonymized registry in the UK, and funded by MRCUK-G1100236, EUFP7-201444 and EUFP7-281654, for which patients provide consent for curation of routine clinical data.

- We have built a longitudinal module in the registry to collect prospective data on number and duration of sick day episodes, history of adrenal crisis and change in medications since last visit.

- This is a preliminary analysis of data collected through the longitudinal module. Data were searched using search criteria congenital adrenal hyperplasia and disorders of androgen excess.

- Number of sick day episodes and information on the management of individual episode were gathered from following tables in the longitudinal module.

- Further details on current medication and change of medication were gathered using below table.

Results

- Amongst the 2160 cases, in the I-DSD/CAH Registry, there were 600 records of CAH (n=559 21-OHD, n=16 11-β hydroxylase deficiency, n=10 other, n=15 not specified) and 389 cases (n=377 21-OHD, n=12 11-β hydroxylase deficiency) were analysed based on the consent level (shared internationally and within EU).
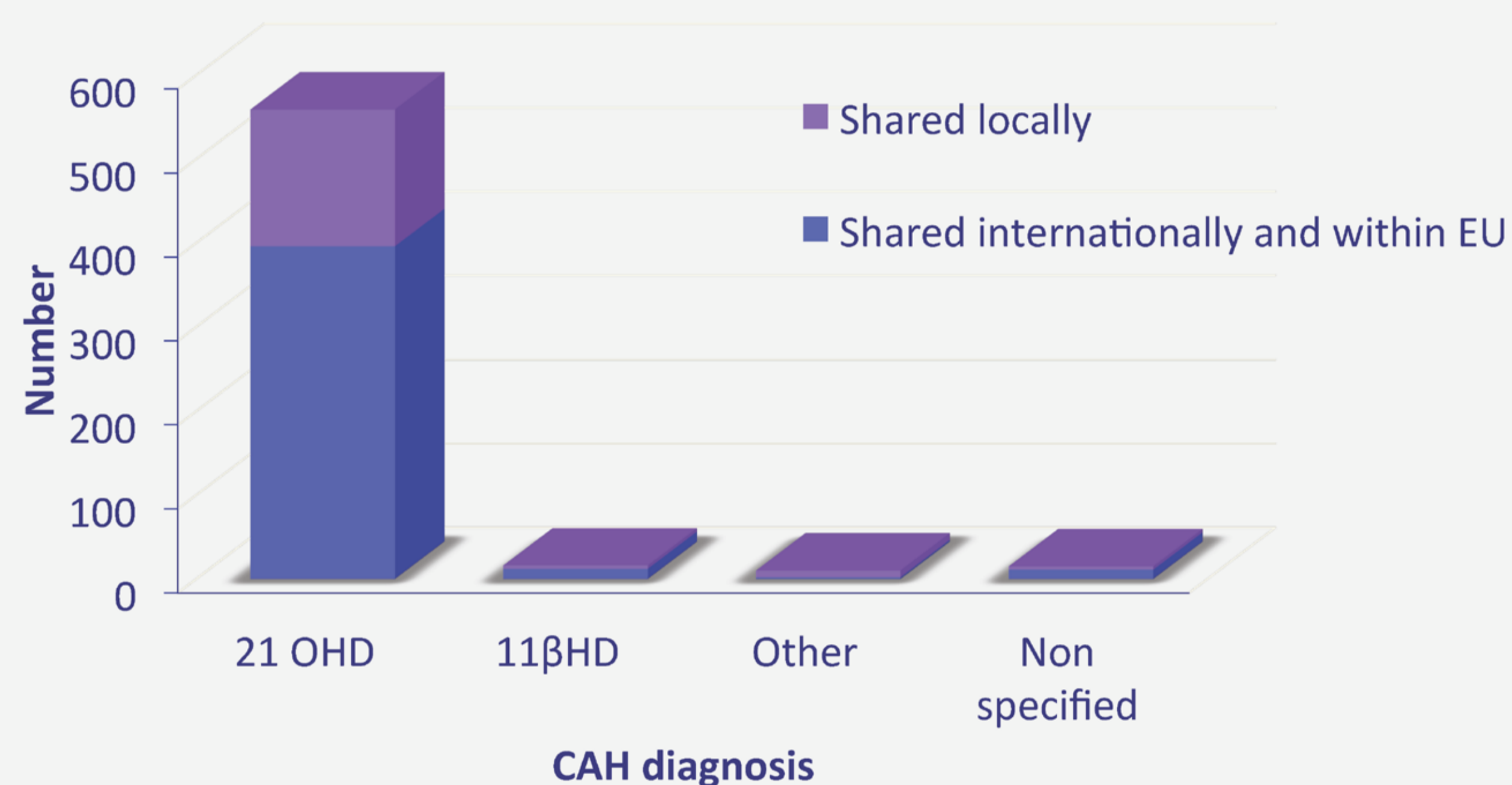
Graph 1 Number of CAH cases recorded in I-CAH registry based on consent level

- Data were entered from 13 countries.
- There were 191 adult patients (>16 years) and 240 children (<16 years).
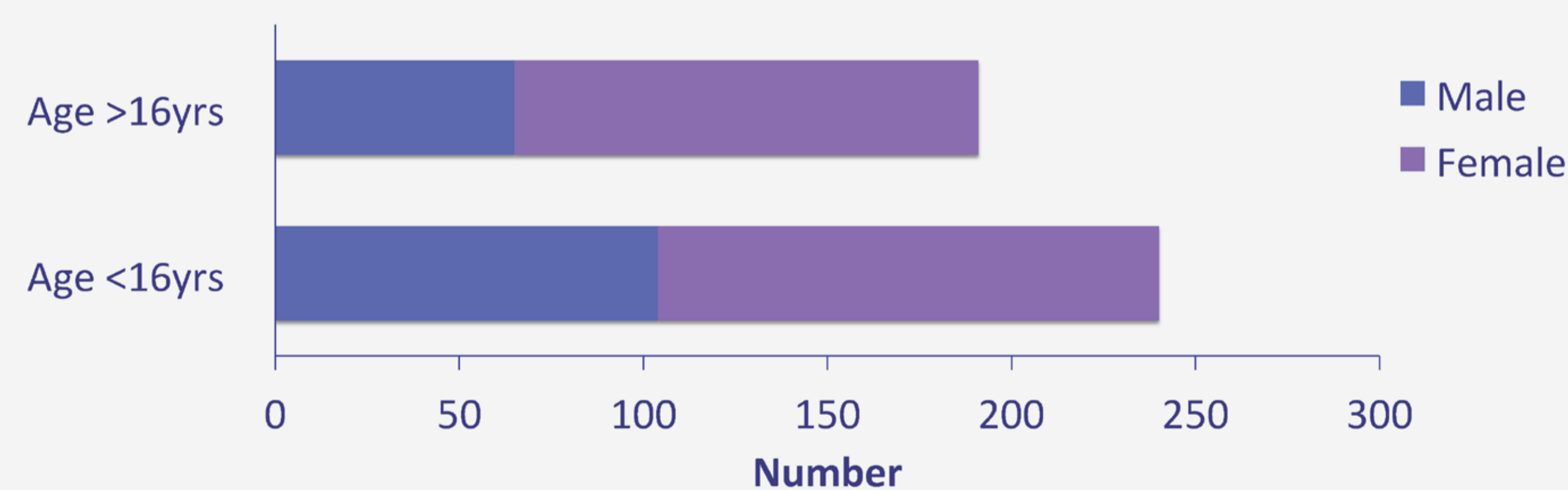
Graph 2 Age distribution of CAH patients in the I-CAH registry

- There were 113 patients with longitudinal data with 448 CAH visit, and the recorded number of sick day episodes was 42. Duration of Sick day episodes ranged between 1-29 days and mean duration of an episode was 2.3 days. There were no reported adrenal crises. Dose of medication was changed based on the sick day episodes.

Conclusions

- The I-CAH registry provides a tool to look at adverse events in patients with CAH. Preliminary data suggest that patients experience frequent episodes where they evoke the sick day rules and appropriate dose adjustments are made based on the sick day episodes.

References
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