Details of study ID 201808_SA

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Office for Rare Conditions, University of Glasgow, Royal Hospital for Children, Glasgow, UK. |
| Coinvestigators: | Xanthippi Tseretopoulou, Jillian Bryce, Faisal Ahmed and the participating centres (https://home.i-dsd.org/i-cah-acute-adrenal-insufficiency-project/) |
| Date of Approval: | August 2018 |
| Name of study: | Acute Adrenal Insufficiency Related Adverse Events in Children with Congenital Adrenal Hyperplasia (CAH) |

**Summary of the proposed work performed with the registry data:**

**Background:** CAH is the most common cause of primary adrenal insufficiency during childhood and patients are at risk of life-threatening adrenal crises. Previous studies have shown that children are at greater risk of illness sequelae. Previous work in 2019 concluded that the real-world data that are collected within the I-CAH Registry show wide variability in the reported occurrence of adrenal insufficiency-related AE.


The project is being continued to investigate whether the level of variation has changed over the period of the last 3 years, both across centres who participated in the 2019 exercise but also by accumulating data from new to the Registry centres.

**Methodology:** Retrospective data collection from the I-CAH Registry. Centres will be approached to enter/update data in the Registry and respond to a survey about the management of SDE in their centre.

**Primary Objective:** To understand the current international benchmark of AI-related AE and the change in this benchmark.

**Secondary Objective:** To investigate any association between the available resources and the management of SDE, with the SDE rate per centre.

**Inclusion criteria:** All patients with the diagnosis of classic 21-hydroxylase deficiency CAH in the I-CAH Registry with age <18 years at the time of clinic visits with a minimum of 2 clinical encounters 6 months apart for the period 01/08/2019-31/07/2022 (minimum 6 encounters).

**Exclusion criteria:**
- Age>18 years on 31/7/22
- All other forms of CAH

The data that will be collected and analysed will be in the following fields:
### Core Data
**Centre:** centre name, country  
**Record:** register ID record, country of usual residence

### CAH Assessment Data
**CAH Longitudinal data:** date of the assessment, Weight, Height  
**Current medication:** glucocorticoids, fludrocortisone, fludrocortisone frequency, current GC replacement, salt replacement  
**Adverse events:** oral steroids, adrenal crisis, number of days, HC injection, emergency management and predisposing condition. If no data exist in the Registry for each visit per patient, we will assume that the patient had no adverse events for the period that preceded that visit.

### Lay summary for the public (for the I-DSD/CAH websites and other publicity materials) (maximum 50 words)
CAH is a serious condition and is associated with life-threatening adrenal crises and sick day episodes. We want to explore the variation of the frequency (incidence) of these episodes among centres that treat children with CAH and see whether this shows any change over time.

### Publication Plan for authorship in outputs (refer to guidance)
All contributors who participate in the project shall be included as authors if their participation is in accordance with the I-DSD/I-CAH publication plan.  
The project group who will be closely involved in the work (project design, data extraction, data analysis, writing the paper) will be listed first.  
The senior authors will be involved in designing the project and data analysis, acting as guarantors and taking responsibility for the work will be listed last authors. All other contributors will be listed in alphabetical order between these two groups of authors. More than two authors from any one contributing centre will need careful justification.