

Adrenal Insufficiency Related Adverse Events in Congenital Adrenal Hyperplasia



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BACKGROUND

Congenital adrenal hyperplasia (CAH) is a rare condition characterised by adrenal insufficiency (AI) and is associated with a life long risk of adrenal crises, a leading cause of mortality in CAH.

There is a paucity of information on the epidemiology of acute adverse events (adrenal crises and sick day episodes) in this population.

OBJECTIVE

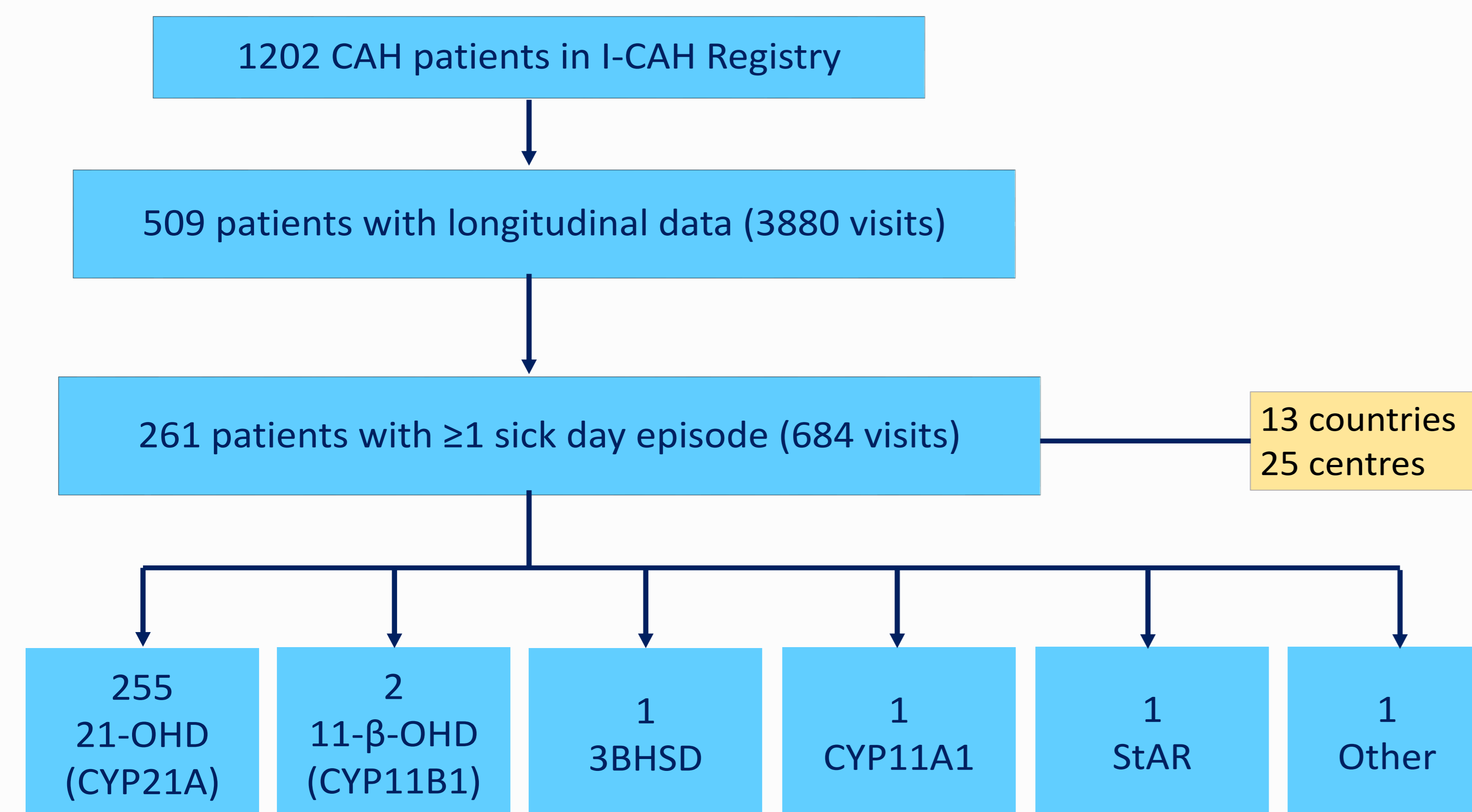
To investigate the frequency, aetiology and consequences of adverse events secondary to AI in CAH

METHOD

A longitudinal analysis of 261 patients with CAH in the International Congenital Adrenal Hyperplasia Registry (I-CAH registry, www.i-cah.org) which collects information on acute adverse events.

RESULTS

Figure 1. CAH patients and diagnostic categories



- Data for 261 patients with 1 or more sick day episodes (SDE) was divided into paediatrics, age <18y (n=215) and adults, age ≥18y (n=46)
- 1,034 SDE were evaluated. Of these, 920 SDE (608 visits) were documented in children and 226 SDE (76 visits) in adults.

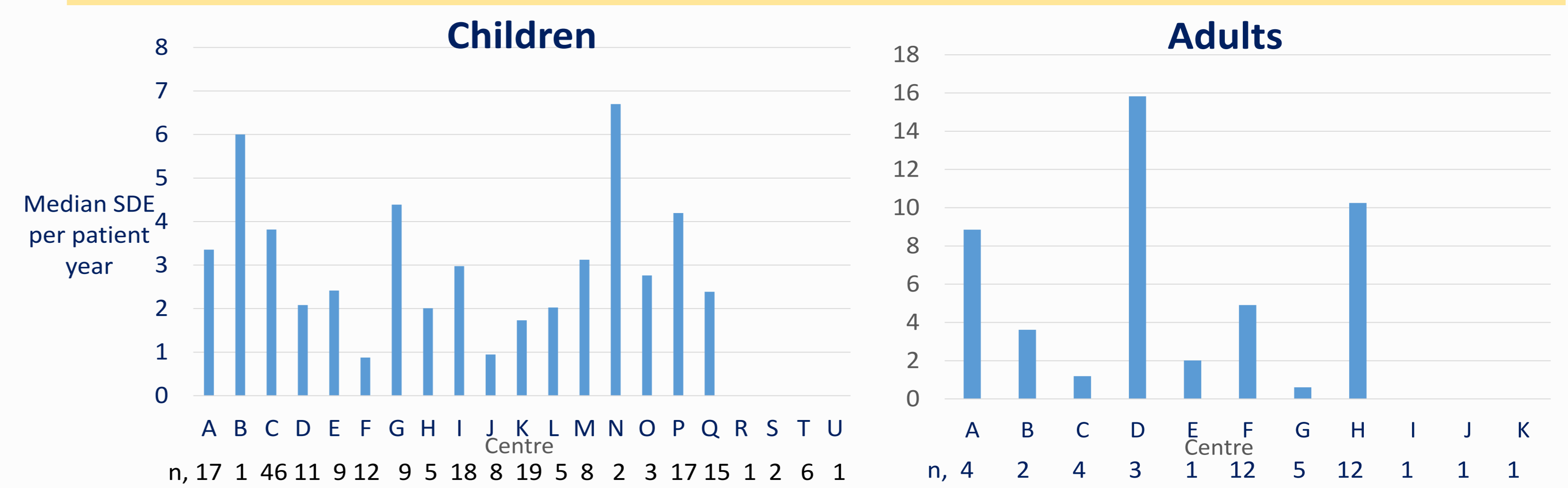
Table 1. Classification of 21-OHD phenotype in children and adults

| CAH type* | Patients aged <18y, n (%) | Patients aged ≥18y, n (%) |
|-----------|---------------------------|---------------------------|
| CYP21A | 211 (98.1%) | 44 (95.6%) |
| - SW | 198 (93.8%) | 40 (90.9%) |
| - SV | 13 (6.2%) | 0 |
| - NC | 0 | 4 (9.1%) |

*SW, salt-wasting; SV, simple-virilising; NC, non-classic CAH

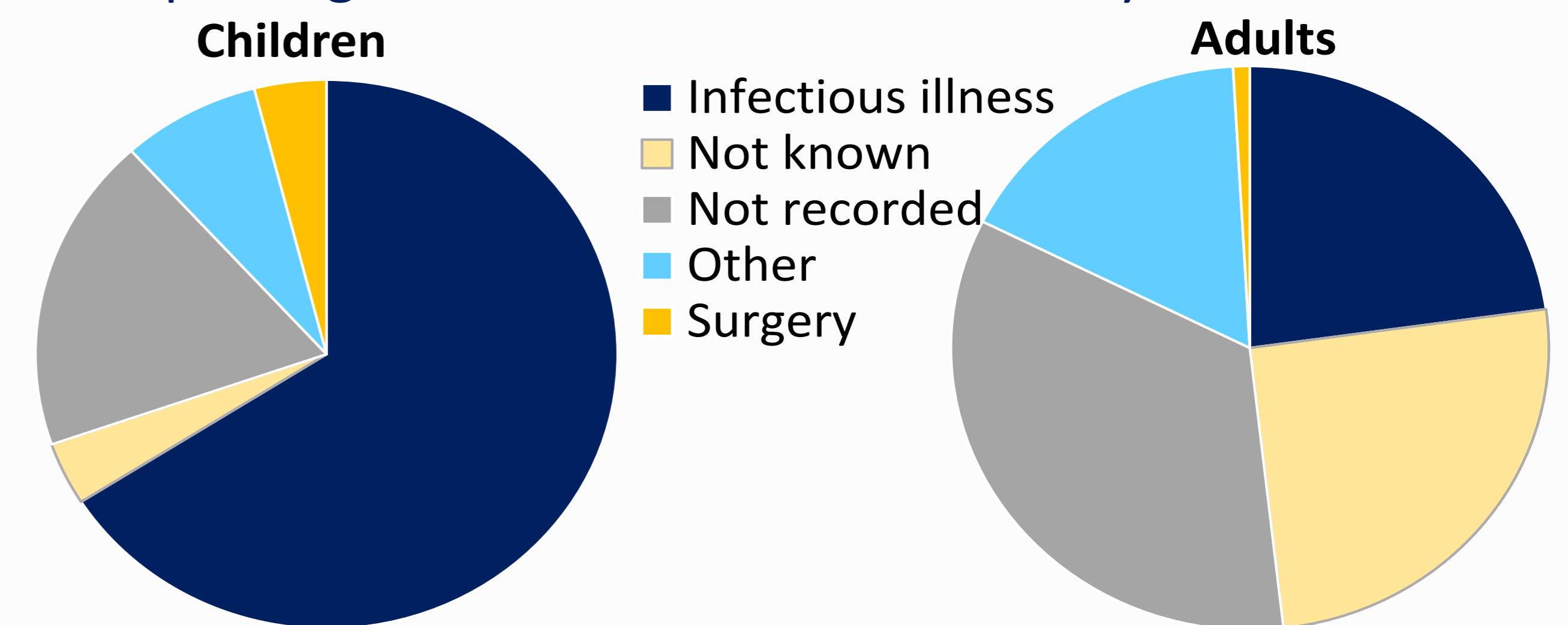
RESULTS

- The median number of SDE for all centres per patient per year was 3.0 (IQR 1.7-4.7) for children and 3.9 (1.8-10.2) for adults (P=0.26)



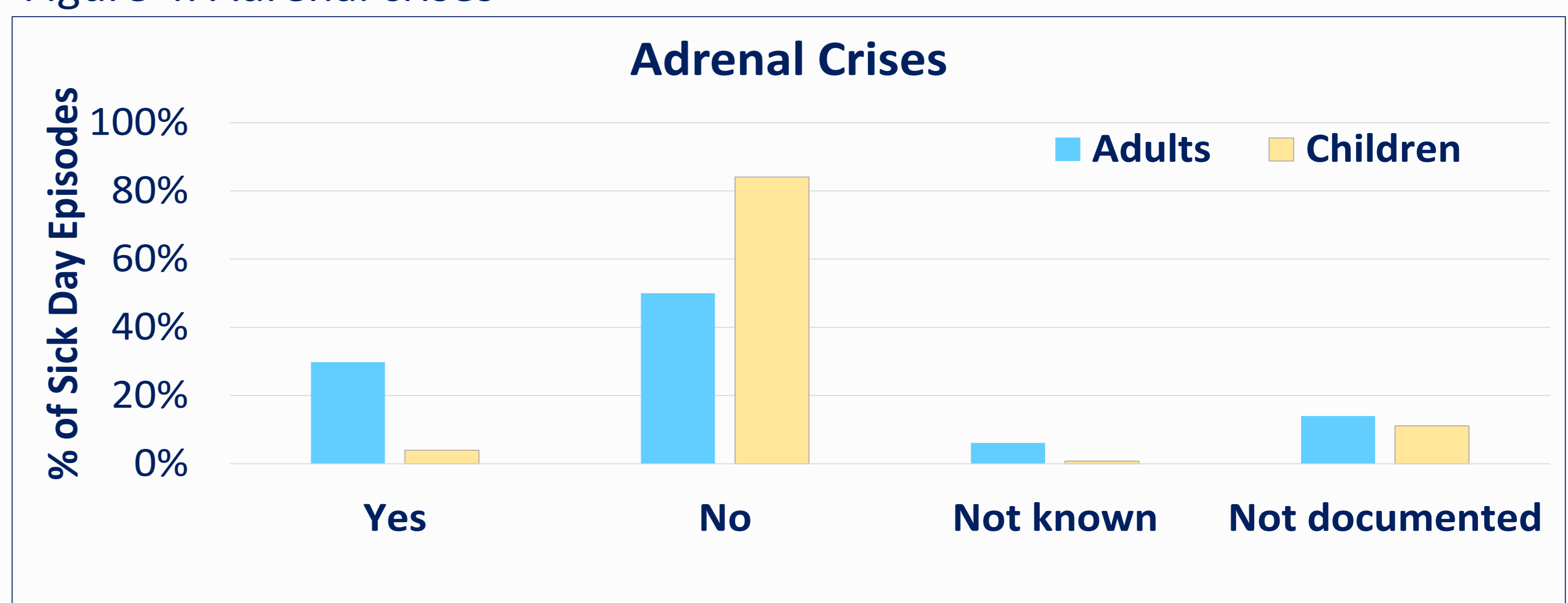
- Children had longer sick day episodes compared with adults (3 days (IQR 2.0-5.0) versus 2 days (1.0-3.0), P<0.05 and infectious illness was the most frequent precipitating event (Figure 3).

Figure 3. Precipitating events associated with sick days and adrenal crises



- In children, younger age and a low hydrocortisone (HC) dose were associated with a greater number of SDE (P<0.01)
- An adrenal crisis (AC) was reported in 4% (37/920) and 30% (34/114) of SDEs (Figure 4) in children and adults, respectively (P<0.05)

Figure 4. Adrenal crises



- In children and adults, female sex was a predictor of hospital admission, P<0.05

CONCLUSIONS

- The I-CAH registry is a valuable tool for evaluating adverse events in patients with CAH.
- Preliminary data suggest that patients experience frequent SDE
- Young children on lower HC doses present with a greater number and longer duration of SDE
- Adults are more likely to experience AC.