Congenital Adrenal Hyperplasia :Survey on the current practice in U.K

U Niranjan[1], A Natarajan [2]
Sheffield Children's Hospital NHS Trust[1], Doncaster Royal Infirmary[2]

Introduction

Congenital adrenal hyperplasia(CAH) varies considerably in clinical presentation and progression causing challenges in its management. The ultimate goal of treatment is to achieve normal growth and development while avoiding adrenal crisis and Hyperandrogenisation.

Aim

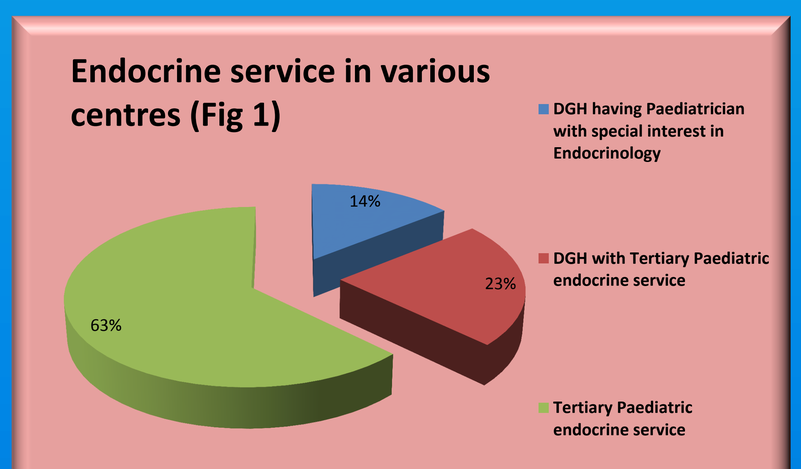
Our aim was to ascertain the current practice in the U.K on CAH management in children and compare these with the endocrine society recommendations.

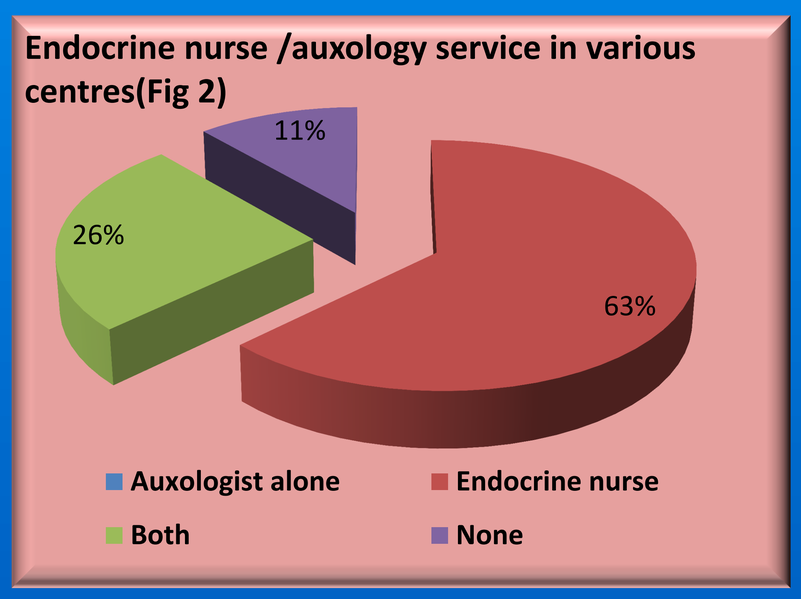
Methods

An online survey with ten questions was emailed to the British Society of Paediatric Endocrinology (BSPED) members asking for one response from each centre regarding CAH management.

Results

The survey was completed by 35centres: Response rate – 38% (35/92).
The Endocrine service provided by each centre varied (Fig 1).

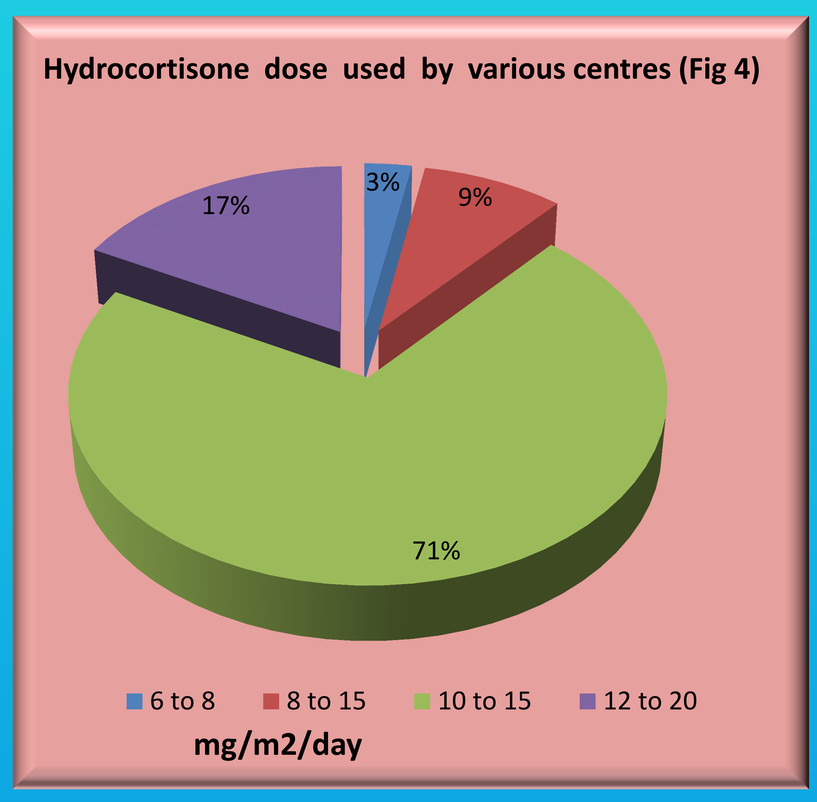




The number of patients per centre varied:
- Tertiary centres : 15 to 120 patients
- DGHs : 5 to 30 patients

Treatment

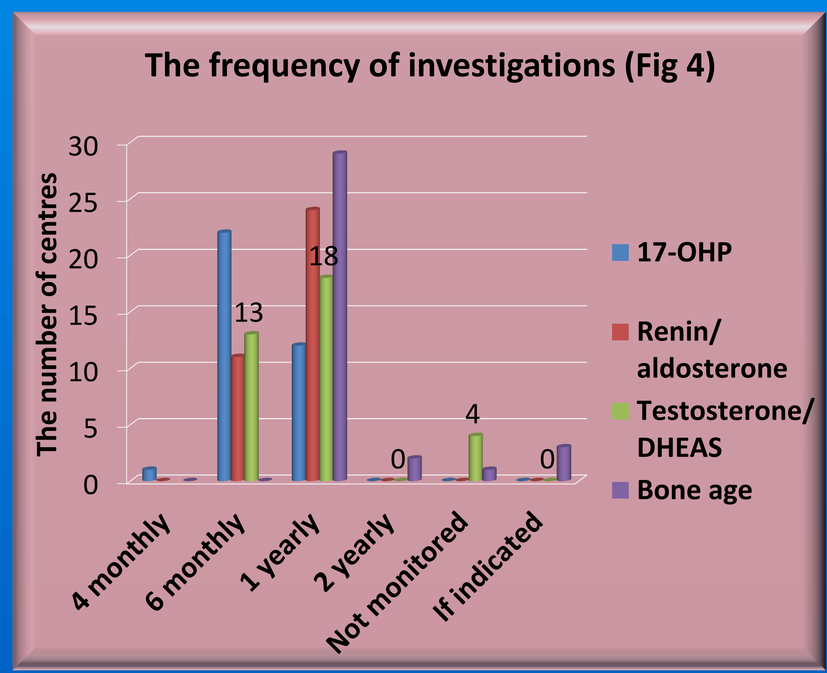
The hydrocortisone dose used by each centre varied with 71% (25/35) of centres using 10-15mg/m2/day(Fig 3).The fludrocortisone dose ranged from 50-300mcg/day with 60% (21/35)using 50-150mcg/day.



Monitoring

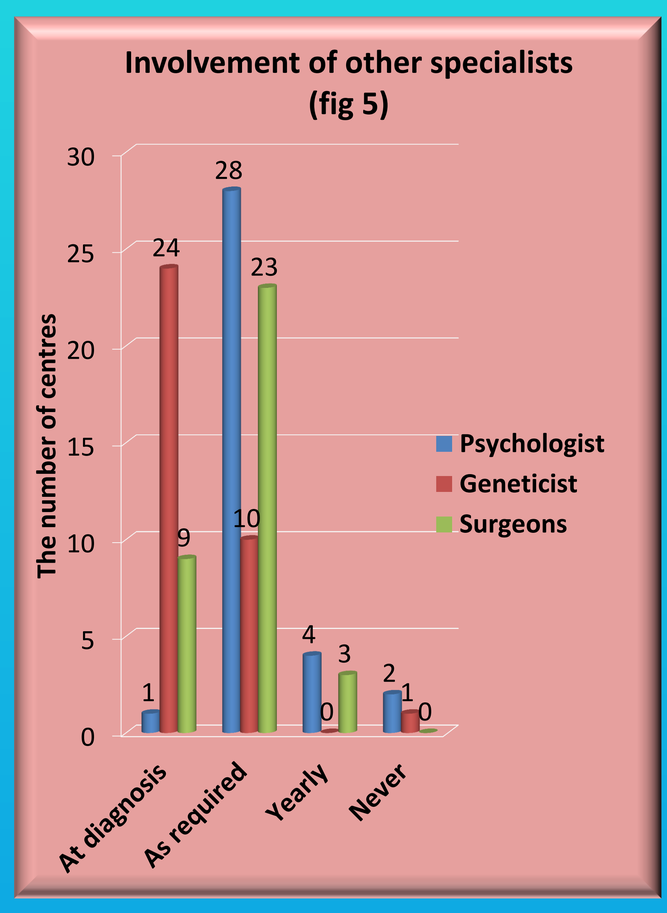
The frequency of clinical reviews was contentious and centres felt it varied depending on the child's age and clinical status. Reviews were done :
- 3-4 monthly in 68% (24/35)
- 6 monthly in 31% (11/35) centres

The frequency of investigations varied significantly between centres (Fig 4)



Involvement of other Specialists

Genetic counselling was provided at diagnosis in 69% of centres while surgical (66%) and psychology (80%) input were primarily on an 'as required' basis (Fig 5).



Conclusion

Our survey highlights the diversity in U.K in managing children with CAH as compared with the recommendations made by the endocrine society guidelines. It demonstrates inconsistent involvement of other specialists which is an essential part of this multifaceted condition.

This is compounded by the complexity of individual cases and wide variation in the progression of CAH during various stages of life.

References

1)Speiser, P.W. et al., 2010. A Summary of the Endocrine Society Clinical Practice Guidelines on Congenital Adrenal Hyperplasia due to Steroid 21-Hydroxylase Deficiency. *International journal of pediatric endocrinology*, 2010, p.494173.

2)Trapp, C.M., Speiser, P.W. & Oberfield, S.E., 2011. Congenital adrenal hyperplasia: an update in children. *Current opinion in endocrinology, diabetes, and obesity*, 18(3), pp.166–70.