Achieving a Consensus on Managing Idiopathic Thickening of the Pituitary Stalk (iTPS) through a National Multidisciplinary (MDT) Forum, Meeting Virtually

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Background

A virtual national multidisciplinary forum was set up in 2010 with the aim of providing a platform for wider discussion and improving management of rare childhood hypothalamo pituitary axis tumours (HPAT). Idiopathic thickening of the pituitary stalk (iTPS) is one such condition where single centre experience is limited, referral practice and management varies, and national guidance is currently lacking.

Aims

1. To longitudinally describe clinical, endocrinology and radiological outcomes of a case series with iTPS
2. To examine whether HPAT discussion changed management, and whether there was a centre based difference in management.
3. To examine if there was a correlation between pituitary volume, endocrinopathy and ultimate diagnosis.

Methods,patients and statistics

- Longitudinal retrospective case note review of investigations, time to endocrine replacement and auxological parameters at diagnosis and last follow up.
- 21 cases (11male,10female) with iTPS discussed at HPAT from 5 centres, between 1.1.12 and 21.10.15.
- MRI 3D volume was analysed with ITK-SNAP v 3.2.0 (www.itksnap.org) software with centralised radiology review.
- The correlation between pituitary volume and numbers of anterior pituitary deficits -termed the Endocrine Morbidity Score (EMS) and the difference between paired data was assessed by Spearman's rank correlation coefficient and Mann Whitney nonparametric statistics respectively using SPSS software.
- Data are presented as medians and ranges

Results

Distribution of total N21cases by ultimate diagnosis (fig 1a) and by centre (fig1b)

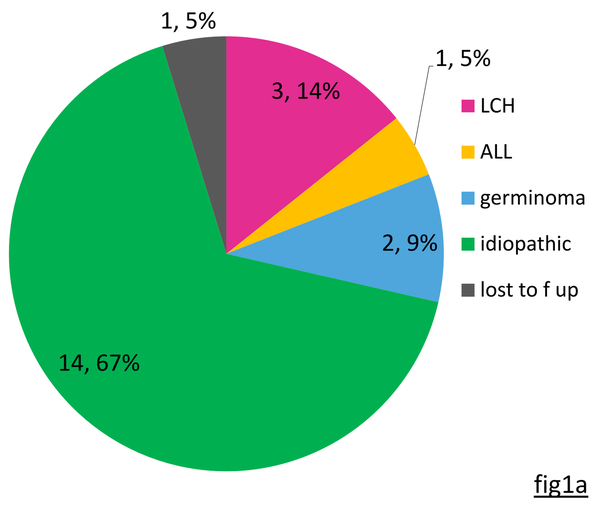


fig1a

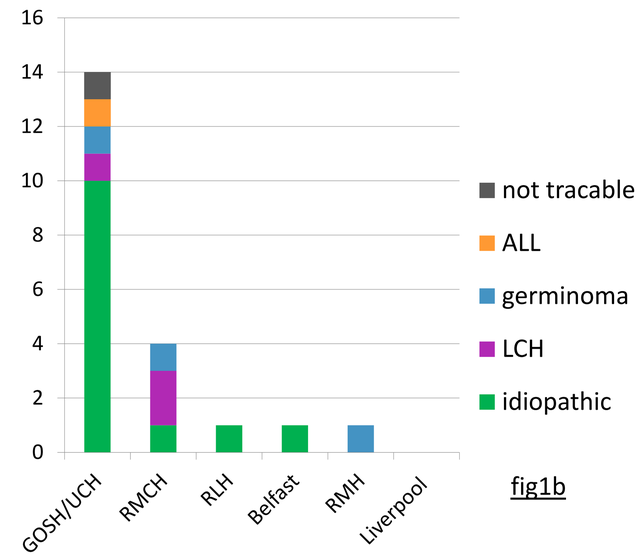


fig1b

I case was lost to follow up abroad .
14 remained idiopathic 1.6 years later.
In remaining 7 diagnoses were made shortly after presentation :
1disseminated relapsed ALL (GOSH)
1 presumed (unbiopsied) expanding germinoma (marker neg-GOSH)
1 biopsy proven germinoma(by pituitary biopsy,causing panhypopituitarism-RMH)
3 biopsy proven LCH ,2 (GOSH and RMCH) biopsies of coincident skeletal lesions at presentation and one transcranial biopsy (new ACTHd in 6 months-RMCH)
The majority of cases (66.7%) were GOSH with wide diversity across UK.

Table 1 Auxology – at diagnosis and follow-up N=16 Median (range)

	Age years	Height SDS	Weight SDS	BMI SDS
At Presentation N21	8.11 (3.5 to15.6)	-0.29 (-4.5 to 1.0)	-0.29 (-3.6 to 2.3)	+0.42 (-1.6 to 2.6)
At 1.8 yr FU (0.1to 7.1) N16	10.5 (4.1 to19.0)	-0.5 (-3.4 to 1.7)	0.1 (-3.4 to 1.4)	1.1 (-3.1 to 2.2)
Mann Whitney N16 pairs		ns	ns	ns
N21 at presentation Mean and 95% CI		-1.05 (-2.02 to -0.82)	-0.46 (-1.35 to 0.44)	0.3 (-0.54 to 1.13)

Patients were significantly shorter at presentation

Initial and follow up endocrine morbidities in patients with a diagnosis fig(3a) and idiopathic cases fig (3b.)

fig3a

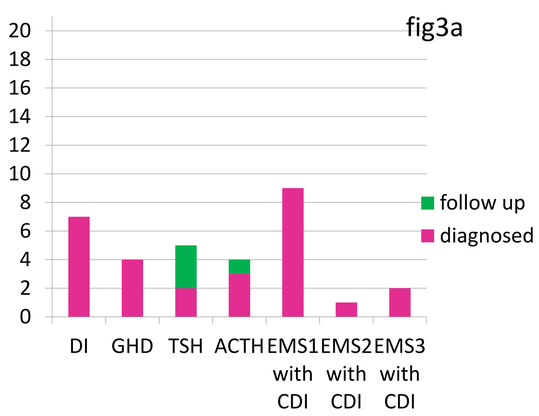
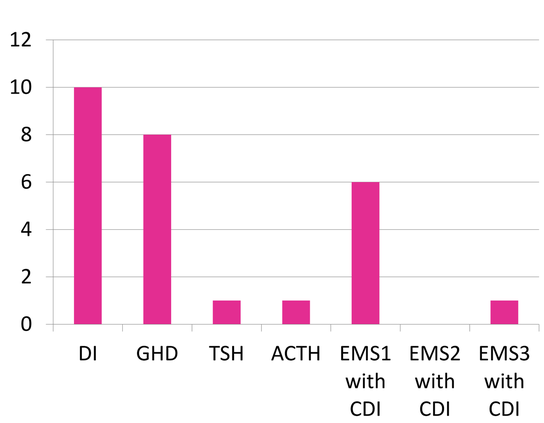


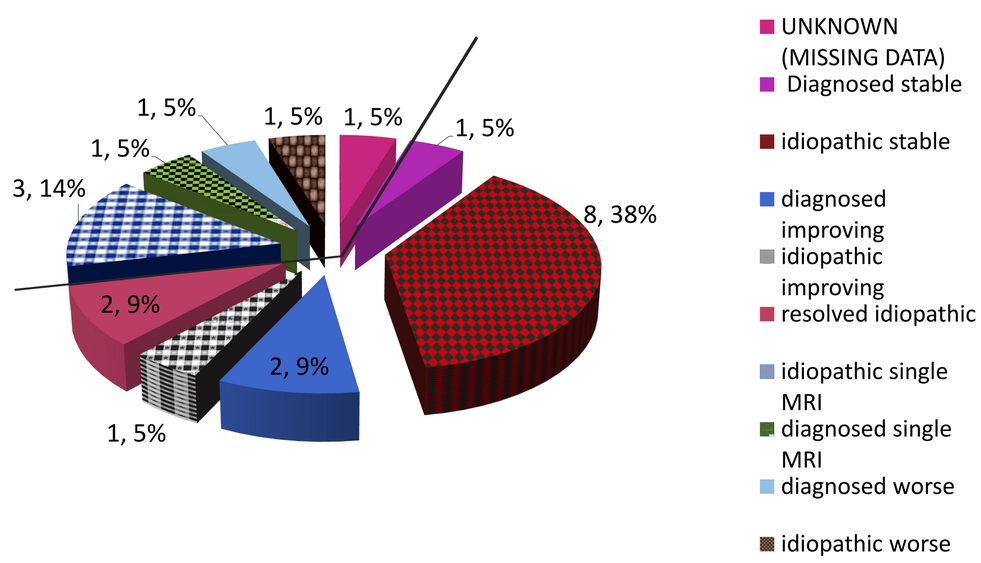
fig3b



17/21 patients had presenting CDI in both diagnosed and idiopathic groups.(4/14 in iTPS had no CDI over time)
Only 95% patients had dynamic pituitary tests at presentation
Patients with an eventual diagnosis tended to have a greater EMS at diagnosis and with time than those that remained idiopathic .
2 patients scanned for acute psychoses had no endocrine morbidity until 15month follow up

MDT discussion significantly influenced initial management in 40%.In the remainder MDT agreed with local plan(30%)or cases were presented for interest (30%)

Fig.5MRI outcome on follow up in all the cases(n=21)(median 1.8years)



Majority of idiopathic cases remained stable on follow up

Investigations done in total and in idiopathic cases fig 2

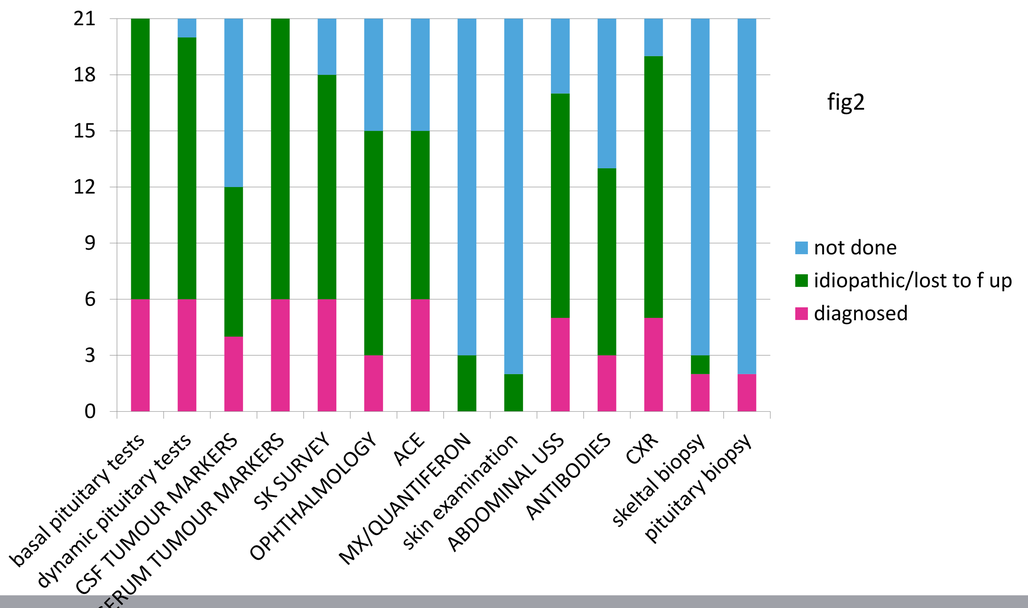


fig2

Summary

CDI is common at presentation, but occult GHD is also frequent.Patients were significantly shorter at presentation. Those with disease diagnosed within 2 years may evolve EMS more quickly. All GHD cases in iTPS (57%) were diagnosed on initial testing .
2. There was no correlation between pituitary volume and EMS or other identifying features
3. About 70% cases remained idiopathic and stable at 2 yr follow up, with resolution in MR appearances in 25% of these despite ongoing CDI and GHD.
4. There was a centre based difference in investigations, especially CNS tumour markers and pituitary biopsy.