A 7-year update report of a national, interdisciplinary endeavour to improve outcomes for children and young people under 19years of age with Hypothalamic Pituitary Axis Tumours (HPAT)

using multi-site video conferencing

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Background

Paediatric HPAT, though generally benign, incur significant neuroendocrine morbidity. Their management is unclear and the paediatric neuro-oncology or adult forum at which they are discussed lack pituitary or age-specific expertise respectively. The UK National HPAT Interest Group has pioneered a monthly, interdisciplinary decision-making forum, using multi-site video conferencing, to garner necessary experience and evidence of outcomes to assist worldwide referrers in managing these rare and often aggressive pituitary tumours. It delivers educational lectures, promotes research, and publishes evidence-based (AGREE11) national guidance on idiopathic Thickened Pituitary Stalk (TPS), Craniopharyngioma, Pituitary Adenomas, and continuously audits evidence since 2010.

Patients

The forum held 256 case-discussions over 7 years, involving 182 patients (46 required multiple reviews). Referrals were received from 8 London Hospitals, 7 UK paediatric oncology centres, and occasionally from European and International centres, including Hong-Kong, Canada and Australia.

Diagnoses and neuro-endocrine morbidities

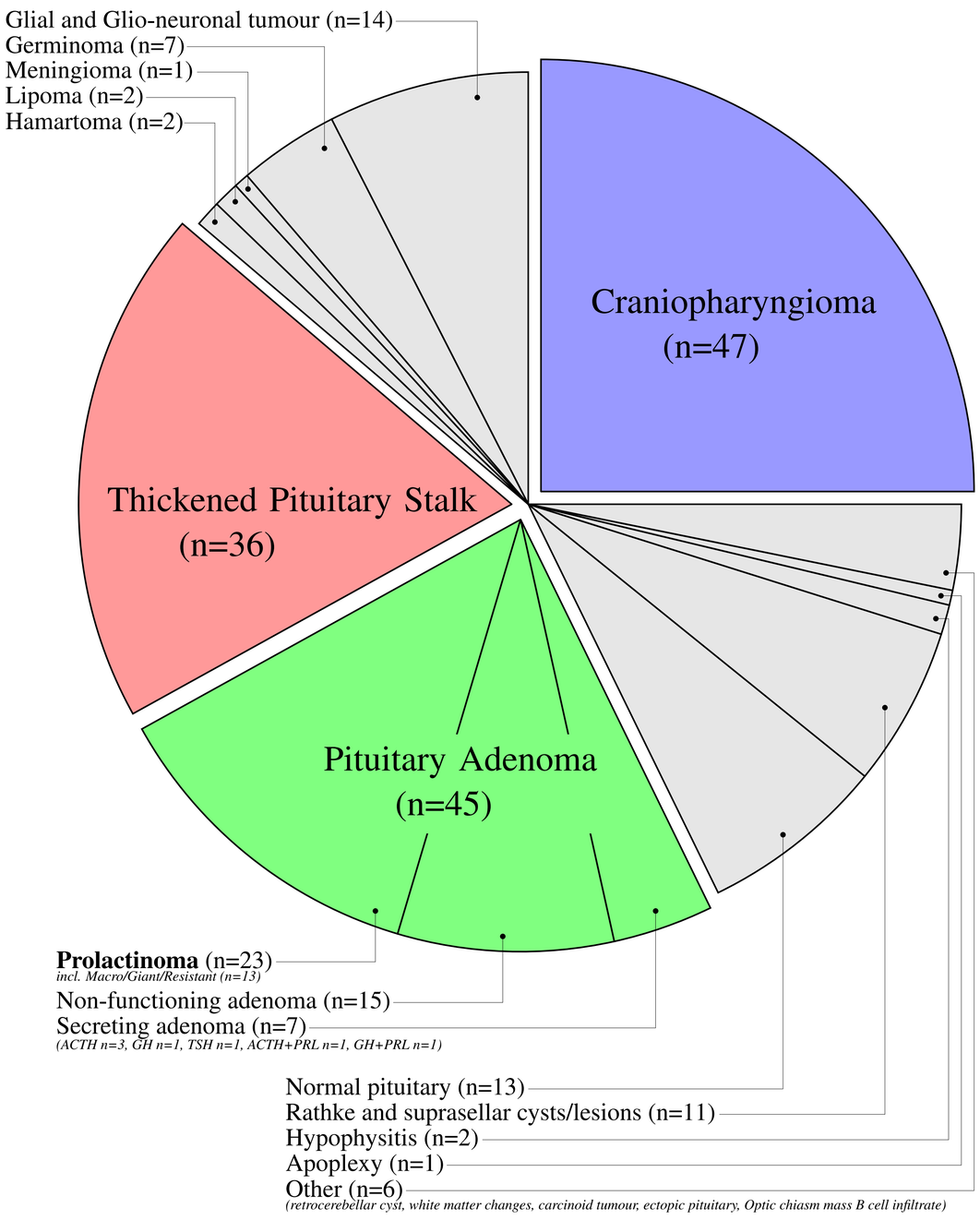

Fig 1: Range of diagnoses (n=186)

Seizure	3 (6.5%)	1 (2.7%)	0 (0%)
Raised intracranial pressure	21 (45.6%)	1 (2.7%)	4 (8.9%)
Headache	10 (21.7%)	4 (11.1%)	21 (46.7%)
Behaviour change	3 (6.5%)	1 (2.7%)	6 (13.3%)
Visual field defect, loss of visual acuity, nystagmus or subjective visual disturbance	26 (56.5%)	4 (11.1%)	13 (28.8%)

Table 1: Neuro-ophtalmological deficits at presentation for Craniopharyngiomas (n=46), TPS (n=36) and Pituitary Adenomas (n=45)

	Panhypopituitarism - at diagnosis	2	(4.4%)
	- post treatment and median follow-up of 7.5yr	36	(80%)
	- recovery of adrenal reserve in ACTH suppression	6	(13.3%)
	Isolated pituitary hormone deficiency	10	(22.2%)
	Hypothalamic dysfunction	4	(8.8%)
	Growth failure (suspected or confirmed GHD)	23	(51.1%)
	Pubertal delay/Central precocious puberty	3+1	(8.8%)
	Diabetes insipidus - at diagnosis	26	(72.2%)
	- at median follow-up of 3 yrs (n=18)	1	new case
	Growth Hormone Deficiency - at diagnosis	12	(33.3%)
	- at median follow-up of 3 yrs (n=18)	5	new cases
	Pubertal delay or arrest +/- amenorrhoea	14	(60%)
	Precocious puberty	1	(4.3%)
	Galactorrhoea	6	(26%)

Table 2: Endocrinopathies at diagnosis for Craniopharyngiomas (n=46), TPS (n=36) and Prolactinomas (n=23)

An expert forum to address HPAT specificities

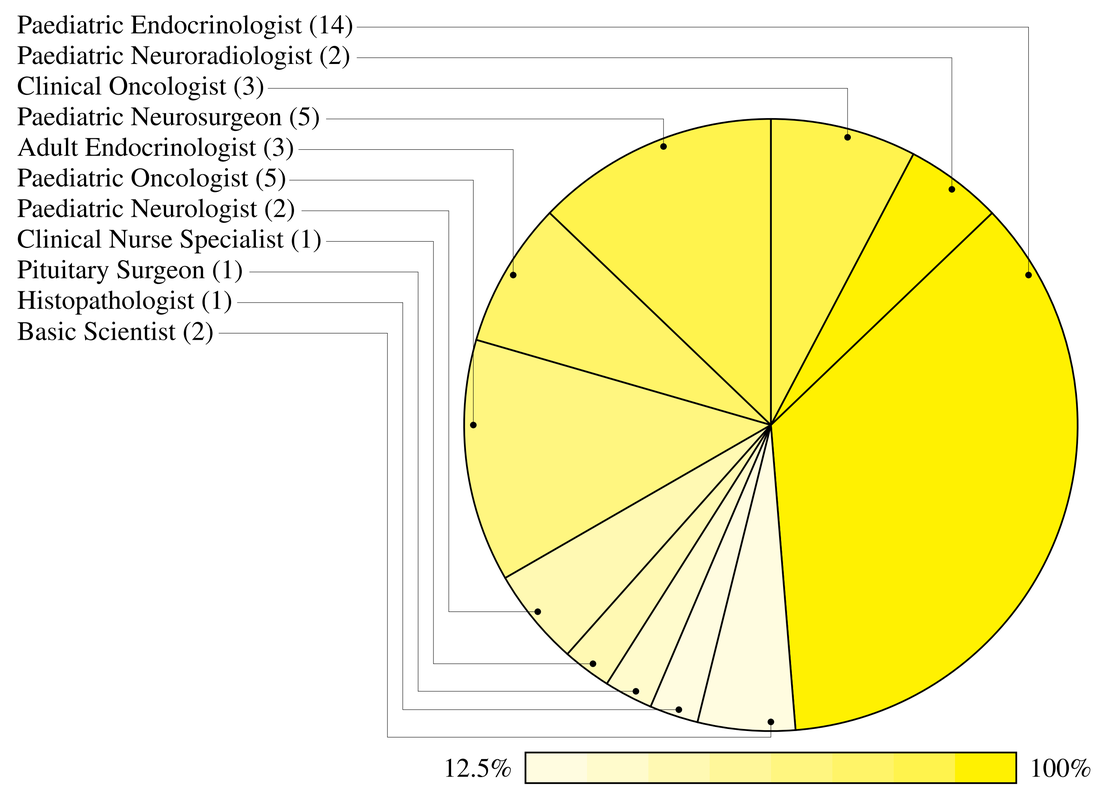

Fig 2: Specialty representation and attendance rate at 8 meetings from January 2017 to January 2018

Summary of clinical guidance requested by referring institutions

Investigation strategies were sought for 44.5% patients. Recommendations by the HPAT forum included dynamic endocrine (21%) and genetic testing (eg: Familial Isolated pituitary Adenoma, BRAF mutations) and guidance on biopsy (8%) in idiopathic TPS.

Medical treatment advice helped optimizing Cabergoline dosing regimens and initiating Growth Hormone replacement in 7.6% and 6% cases respectively.

The forum favored **Radiation** therapy for 9.8% of the cases and advised against it in 3.8%.

Complex **Surgical** options were discussed in 19.2% cases and consensus reached. For example, advice not to debulk an invasive Craniopharyngioma was upheld by the neuro-oncology MDT and further surgery cancelled, in order to prevent significant hypothalamic damage.

Extra-Regional transfer of care was facilitated in 3 cases (1 Cushing disease, 2 Craniopharyngioma) when specialist endoscopic transsphenoidal surgery was unavailable at the referring centre.

Follow-up plans including neuro-imaging surveillance timing were provided in 35% cases and discharge facilitated in 4%.

Conclusion

A supra-regional childhood HPAT discussion forum is clearly welcome and continues to expand: it facilitates patient referral and outcome audit, and centralizes clinical decision-making. Our evidence regarding benefit suggests that there is a demand for this expert advisory body to be formally recognized as a model of care.

Disclosure: The authors declare no conflict of interest.

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