Three Year Experience of a National Interdisciplinary Initiative to Enhance the Management of Hypothalamic Pituitary Axis Tumours (HPATs) Using Multi-site Videoconferencing *on Behalf of the UK HPAT Interest Group.*

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Background

- Childhood HPATs are complex and very rare, so experience in any single centre is limited
- Their high survival carries with it high neuroendocrine morbidity
- As there is currently no evidence-base, treatment is individualised on a case basis according to consensus (2005) guidance (ref 1).

<u>Hypothesis</u> - Centralised care or wider multi-professional decision making may improve neuroendocrine and visual outcomes.

- In 2011, a 6-month national HPAT advisory group pilot, was set up with a view to acquiring funding for centralising care from the UK Department of Health.
- Though centralisation was not ultimately funded, the pilot was well received.
- The Northern (Liverpool/Manchester) and Southern (London) 3-centre HPAT meetings subsequently continued independently, adding more national and international centres (Ireland and Australia) to their virtual meetings.

Aims

1. **To facilitate multi-professional dialogue across centres nationally (including adult pituitary specialists) in a videoconference format.**
2. **To enhance diagnostic and treatment decision-making through education and sharing audits, knowledge and experience.**

Methods

- Initially we undertook regular, minuted monthly videoconference meetings between as many as 25 professionals across 3 sites. Multi-professional attendance at one meeting is shown on the right . Over the next few years, more centres and specialties.

- Despite significant high level IT input at all 3 sites and a (20k) IP based system at GOS, facilitating the provision of a live video conference with simultaneous high quality imaging initially proved challenging. These issues were subsequently resolved with time, with localized Cisco MCU in conjunction with ISDN Gateway.

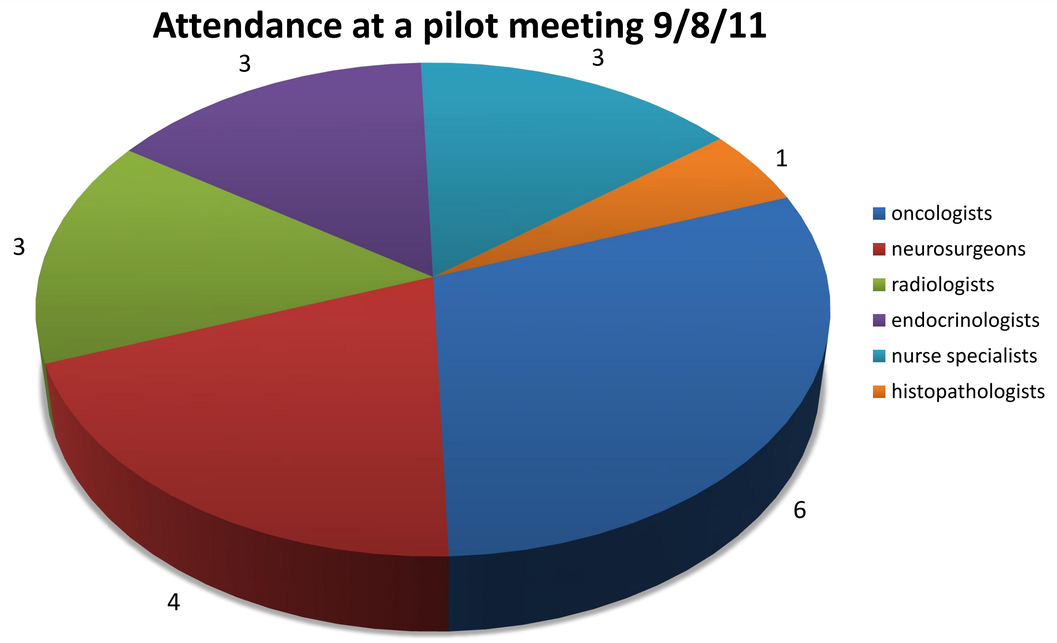
Summary of Meetings – Table 1

	Meetings	Number of new cases	Number of reviews from previous meetings	Types of new tumours discussed
Pilot study	7 meetings	9 new	None	4 craniopharyngiomas 3 supersellar lesions / gliomas 2 others
Jan → Dec 2012	12 meetings	33 new	14 reviews 7 reviewed multiple times	9 prolactinoma 7 craniopharyngioma 4 suprasellar lesion 2 thickened pituitary stalks 11 others
Feb → Sept 2013	8 meetings	25 new	16 reviews 9 reviewed multiple times	6 craniopharyngiomas 6 thickened pituitary stalk 4 supersellar lesions / gliomas 2 prolactionomas 7 others
Overall	27 meetings	67 new cases	30 reviews 16 reviewed multiple times	17 craniopharyngiomas 11 prolactinomas 11 supersellar lesions/gliomas 8 thickened pituitary stalks 20 others

There have been 3 lectures given so far

1. **Molecular biology of craniopharyngiomas**
2. **Genetics of pituitary tumours, with diagnosis of prolactinomas**
3. **Trans-sphenoidal endoscopic surgical review of a patient**

- Audits on craniopharyngiomas undertaken at all 3 sites, one published (2) with 1 on prolactinomas.
- 2 further audits from GOSH (thickened pituitary , optic gliomas) are posters at this meeting



Case summaries Table 2

Pts	Diagnostic age Weigh+ Height SDS	Position	Baseline function At diagnosis	Diagnosis and initial treatment/plan	Data between diagnosis and HPAT meeting	Relapse management	(Age) and Discussion at HPAT Meeting	Outcomes of HPAT Meeting	Post HPAT meeting Notes
AL	3.6	Suprasellar /pituitary	GH deficiency Poor visual attention, nystagmus and full optic discs.	Surgical decompression, proton beam radiotherapy	Patient discussed soon after diagnosis	None	**(3.7) Review scans, decide between radiotherapy and/or surgery. If surgery, what approach?**	**Opted for Surgical decompression transsphenoidally followed by proton beam radiotherapy**	Ocular symptoms remain and are unexplained, Proton therapy was a success and growth hormone treatment should be started
HA	4.9 W 1.25 H -0.95	Suprasellar	GH deficiency Mild Visual impairment	Surgical decompression	None of note	2x Surgical decompression + reservoir	**(5.6) Should surgery be attempted alongside radiotherapy?**	**Surgery opted for before radiotherapy. cyst had re-expanded – before proton beam was considered, the cyst should be deflated with a reservoir**	Patient is Hypopituitary, and is gaining excessive weight. However, vision is excellent. W 1.53 H -1.93
LE	17.2	Suprasellar	cortisol, T4, fT4, and 25OHvitamin D deficiency, low insulin Visual disturbance (L)	Ommaya reservior insertion, cyst drainage, tap placement.	GH, TSH and ACTH deficiency. Vision improved	None	**(17.4) Surgical clearance or radiotherapy?**	**Decided that surgical clearance was too difficult, therefore patient would need NHS conventional radiotherapy, or private proton beam.**	Patient opted to go private for proton beam radiotherapy, on treatment for deficiencies, but headaches remain
DA	2.5 W-0.13 -	Suprasellar	No visual impairment TSH deficiency Normal otherwise	Surgical debulking on 3 occasions No radiotherapy	Panhhypopituitary after debulking (3rd) developed DI, with triphasic response.	None	**(3.3) Leaving or removing residual tumour would not change field of the radiotherapy, so surgical debulking again prior to radiotherapy or not?**	**No to surgical debulking – straight for proton beam radiotherapy. Removing residual tumour would add risk of surgical and endocrine complication with little benefit.**	She now has ACTH deficiency, and Visual impairment in right eye – homonymous hemianopia Lost to follow up – not sure if proton beam occurred.
CC	11. W -0.99 H -1.81	Suprasellar	Very mild visual Impairment GH and IGF-1 Deficiency	Surgical decompression	None of note	None	**(11.1) Surgery or proton beam?**	**No to surgery – straight for proton beam radiotherapy. Resection would not alter the subsequent radiation field, + risk of coning**	Visual impairment, doing well at school. March 2014 MRI shows craniopharyngioma is smaller. W -0.11 H -1.84
AM	4.1 W -0.80 H -1.64	Suprasellar	GH, IGF1, ACTH and TSH Deficiencies S&L and visual impairment (R-blind)	Surgical resection and radiotherapy.	None of note	2x Surgical decompression	**(4.7) What to do with residual – surgery, radiotherapy or nothing?**	**No to surgery or radiotherapy – watch and wait approach with annual scans**	IGF Deficiency, completely panhypopituitary and stable Has made language improvement, looking for visual support in school, still needs 1:1 support W -1.44 H -2.03
JP	1.9 W 1.46 H 1.64	Pituitary	GH, ACTH, TSH deficiencies, visual impairment (L)	Surgical decompression and resection	None of note	1x Surgical decompression	**(2.0) Discussed twice – review scans – both meetings discussed scans – should patient receive proton beam therapy despite age?**	**Decided to delay radiotherapy, and continue 3 monthly MRI scans to monitor potential progress/recurrence.**	Asymptomatic, stable MRI, no recurrence
NP	5.6 W 3.26 H 2.49	Suprasellar	GH and TSH deficiency Low working IQ	Surgical Resection and decompression 5 ops in total	None of note	2x Surgical	**(18.8) Query Surveillance scanning and need for surgical follow up regarding in situ VP shunt?**	**Stop further surveillance scanning, no need for neurosurgical follow up.**	W – 0.45 H 3.27
DG	7.9 W 3.63 H 0.3	Suprasellar	Patient registered blind and obese	Cyst drainage + VP shunt, radiotherapy (also emergency cyst aspiration)	GH, IGF, TSH, LH,FSH deficiencies Weight gain	None	**(14.3) Continue surveillance scanning?**	**Stop further surveillance scanning Scans stable over past 3 years with suprasellar component showing no change.**	-
RP	7.7 W 2.14 H 0.78	Pituitary	GH, ACTH, PRL and gonadotrophin deficiencies, IQ-85	Surgical resection and local radiotherapy	Escalating hypothalamic obesity, panhypopituitary with DI	4x Surgical debulking	**(15.0) Discussed twice – 1 – compare scans from UCH and GOSH – are new symptoms are in keeping with growing mass? 2 – should surgery be an option?**	**Surgery declined for now. Suprasellar mass is in keeping with known craniopharyngiomana – patient should lose weight**	Patient very disabled

Results

- In 27 meetings spanning 2.5 years.
- 67 HPAT clinical cases (including quality imaging) were discussed in relation to formulating management plans (Table 1)
- 16 discussed on multiple occasions.
- Table 2 shows type of cases discussed, questions posed & decision outcomes in **10** craniopharyngioma cases (most common tumour type discussed).
- In the first **7** cases, we focused on acute management decisions.
- The first 4 of these proceeded to surgery (3) or proton beam radiation (1).
- The following 3 cases were managed conservatively.
- All are in remission and doing well.
- The last 3 cases were brought to meetings late in their management, to demonstrate high morbidity, discuss future management, education and acquisition of long-term outcome data.
- Currently we have :
- 7 participating centres.
- 3-4 cases discussed per month.
- Alternating with themed meetings.
- Aiming to collect outcome data.

Conclusion

- **A national, regular, multidisciplinary consultation for discussing rare HPATs is feasible and welcomed, facilitating dialogue amongst a wide specialist professional grouping and influencing management.**
- **With appropriate funding, such collaborative experience with outcome data collection, regular on-going audits, and an educational programme should enhance the management of this rare patient group, resulting in better outcomes and shaping the national standard of care.**

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