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


1Great Ormond Street Hospital, London, 2University of Manchester, 3On behalf of the HPAT group

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 hypothalamic 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.

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

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

<table>
<thead>
<tr>
<th>Age group</th>
<th>Height SDS</th>
<th>Weight SDS</th>
<th>BMI SDS</th>
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<tbody>
<tr>
<td>At Presentation</td>
<td>8.11 (3.5 to 15.6)</td>
<td>-0.29 (4.5 to 1.0)</td>
<td>+0.42 (-3.6 to 2.3)</td>
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<tr>
<td>1.1 yr FU</td>
<td>10.5 (4.1 to 19.0)</td>
<td>0.1 (-3.4 to 1.4)</td>
<td>1.1 (-3.1 to 2.2)</td>
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<tr>
<td>Mann Whitney N16 pairs</td>
<td>ns</td>
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<tr>
<td>N21 at presentation</td>
<td>-1.05 (-2.02 to -0.82)</td>
<td>-0.46 (-1.35 to -0.44)</td>
<td>0.3 (-0.54 to 1.13)</td>
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Patients were significantly shorter at presentation.

iTPS

Aims

1. To longitudinally describe clinical, endocrinological 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.

Results

Distribution of total N21 cases by ultimate diagnosis (fig 1a) and by centre (fig 1b)

I case was lost to follow up abroad.
14 remained idiopathic 1.6 years later.
In remaining 7 diagnoses were made shortly after presentation:
1 disseminated 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.

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

Majority of idiopathic cases remained stable on follow up

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 year 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.