Managing children with Thickened Pituitary Stalk (TPS) and/or Idiopathic Central Diabetes Insipidus (ICDI): A Single Centre Experience on 63 children

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BACKGROUND

Children with TPS and/or ICDI present to different (endocrine, oncology, ophthalmology) specialists. Their rarity, absence of agreed radiological criteria or consensus guidance, make their management (to exclude an occult malignancy) problematic. Biopsy is too dangerous and cases may remain undiagnosed or evolve over decades.

OBJECTIVE

1. to longitudinally characterize a large childhood cohort presenting with TPS and/or ICDI
2. to assess clinical, visual and endocrine correlates over time

PATIENTS AND METHODS

SEARCH CRITERIA:

We searched the terms “thickened pituitary stalk” or “idiopathic diabetes insipidus” in electronic radiology and clinical document libraries at our split-site centre (UCLH/GOSH) over the last 30 years.

RESULTS

Patients with TPS were older at presentation than those with ICDI and TPS+ICDI (p<0.04) (Table 1).

- Tissue histology was available only in 10 to 37.5% of patients. A “watch and wait strategy” was adopted in the majority of TPS patients and in half of TPS+ICDI patients (Table 1).

- TPS+ICDI patients were more likely (38.5%) than ICDI (5.6%) and TPS (none) to have histiocytosis. Tumours were identified in 26.9% TPS+ICDI and 27.9% ICDI, 1.0±1.4 and 1.9±2.4 years later respectively, but not in TPS, 80% TPS cases remained unexplained (vs 61.1% ICDI and 34.6% TPS+ICDI) at a shorter follow-up (Figure 1).

- Patients’ main presenting features are shown in Figure 2.

- Multiple anterior pituitary deficits evolved with time across groups (GHd, 45-58%, TSHd 19-30%, ACTHD 13-21%, GnRHd 7-17%) but visual deficits, present in 8-23% at presentation, increased only in TPS+ICDI (7.6 to 34.6%) (Table 2).

CONCLUSIONS

Longitudinal endocrine and visual assessment of all patients with TPS and ICDI is important. ICDI is a negative prognostic factor for malignant disease, whilst the combination with TPS is more often associated with histiocytosis. TPS alone is unlikely to lead to malignancy but should be prioritized for endocrine follow-up.

**TABLE 1**

<table>
<thead>
<tr>
<th></th>
<th>TPS</th>
<th>ICDI</th>
<th>TPS+ICDI</th>
<th>p Value</th>
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</thead>
<tbody>
<tr>
<td>N</td>
<td>10</td>
<td>18</td>
<td>26</td>
<td></td>
</tr>
<tr>
<td>M/F</td>
<td>4/6</td>
<td>7/11</td>
<td>10/11</td>
<td>ns</td>
</tr>
<tr>
<td>Age at presentation (mean±SE) (years)</td>
<td>9.8±4.9</td>
<td>5.5±4.4</td>
<td>6.2±3.4</td>
<td>&lt;0.04</td>
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<tr>
<td>Age at last follow-up (mean±SE) (years)</td>
<td>13.15±5.9</td>
<td>11.42±6.6</td>
<td>13.82±5.0</td>
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<tr>
<td>Length of follow-up (mean±SE) (years)</td>
<td>2.5±1.6</td>
<td>5.2±5.1</td>
<td>5.8±3.9</td>
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<tr>
<td>Tissue histology available (biopsy or surgery) (%)</td>
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<td>22.2</td>
<td>37.5</td>
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<td>Watch and wait strategy (%)</td>
<td>90</td>
<td>72.2</td>
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**TABLE 2**

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<td>Visual deficits at presentation (%)</td>
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<td>7.6</td>
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<tr>
<td>Visual deficits at last follow-up (%)</td>
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<td>22.3</td>
<td>34.6</td>
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**Endocrinology at endocrine assessment**

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<tr>
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<th>GHD (%)</th>
<th>ACTHD (%)</th>
<th>TSHD (%)</th>
<th>DI (%)</th>
<th>GnRHd (%)</th>
<th>Hyperprolactinemia</th>
<th>Hyperprolactinemia</th>
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<tbody>
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<td>GHD (%)</td>
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<td>33.3</td>
<td>41.7</td>
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<td>11.1</td>
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<td>ACTHD (%)</td>
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<td>DI (%)</td>
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<td>GnRHd (%)</td>
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**Endocrinology at last follow-up**

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<th>DI (%)</th>
<th>GnRHd (%)</th>
<th>Hyperprolactinemia</th>
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<tr>
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<tr>
<td>GnRHd (%)</td>
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<td>29.2</td>
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<tr>
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