Prospective Cognitive Assessment in Children with Craniopharyngioma at Diagnosis, after Conservative Surgery and Before and after Adjuvant Radiation.

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Background

With current management, 9 /10 children with craniopharyngioma will survive 10 years (1), but most suffer chronic neuroendocrine and cognitive morbidity. Whether this is disease and/or treatment-related is still debated in the absence of prospective longitudinal cognitive assessments addressing the contributions of each.

Aim

1. To prospectively evaluate the prevalence of cognitive dysfunction at presentation and after a conservative surgical and radiation treatment strategy, aimed at avoiding hypothalamic morbidity.
2. To determine aetiology, incidence and severity of any cognitive deficits by tumour grade and treatment type.

Methods

- Between 1.1.10 and 1.1.15, 20 children (13M, 7F) of median (range) age 7.6 [1.9-17.18] years were newly diagnosed with craniopharyngioma and had at least 6 months of follow-up data.
- 15/20 underwent Wechsler IQ assessments [full scale (FSIQ), verbal (VIQ), performance (PIQ), working memory (WMI) and processing speed index (PSI)] after conservative surgery and before adjuvant proton (n=9) or IMRT (n=6) radiation to any residual.
- 7 subjects had a repeat assessment 1-3 years later. These did not differ for baseline demographic, tumour grade or cognitive score from those who did not undergo reassessment (p>0.16).
- Tumours were graded for hypothalamic involvement (Paris 0-2) at diagnosis.
- Data was analysed with parametric statistics and is shown as means (SD) and 95% CI.

Results

Table 1 – Baseline Wechsler IQ by Paris Grading

<table>
<thead>
<tr>
<th>Paris Grading</th>
<th>Mean IQ Scores</th>
<th>Before radiotherapy</th>
<th>After radiotherapy</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>FSIQ</td>
<td>VIQ</td>
<td>PIQ</td>
</tr>
<tr>
<td>Grade 0</td>
<td>Mean</td>
<td>128</td>
<td>136</td>
</tr>
<tr>
<td>N=1</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Grade 1</td>
<td>Mean</td>
<td>92</td>
<td>95</td>
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<tr>
<td>N=1</td>
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</tbody>
</table>

Fig. 1 – Wechsler IQ assessments Before (1) and after (2) surgery and radiotherapy

Table and Figure

- Baseline IQ data was available for only 15/20 patients assessed aged 8.98 [2.80-17.50] years.
- At diagnosis, the majority (13/15) had Grade 2 tumours, limiting exploration of effects of hypothalamic invasion on cognition.
- The overall mean [95% CI] FSIQ was 104.25[95.78-112.71], no different from age and sex standardised norms, and without interdomain difference (VIQ, PIQ, PSI, WMI, p>0.05).
- In 7 Grade 2 patients, reassessed 1.82 (0.60-2.80) years after radiation, IQ scores were not significantly changed (paired p>0.05)

Conclusions

- Children with craniopharyngioma are of average intelligence at diagnosis, despite significant (Grade 2) hypothalamic invasion in the majority.
- At reassessment, scores remained unchanged by a modern conservative treatment strategy (surgery and radiation), but follow up is short.
- This limited prospective data with a modern treatment strategy is encouraging; proactive surveillance and support may further limit decline.
- Larger prospective studies needed to assess contributions of disease and treatment are underway internationally given the disease rarity.