Surgical Management of Hypothalamic Hamartomas with Epilepsy: A 5-year experience with 92 cases

Chunde Li, M.D., Shiqi Luo, M.D,
Department of Neurosurgery,
Beijing Tiantan hospital, Beijing, P.R. China.
Introduction

- Hypothalamic hamartomas (HHs) are rare congenital heterotopic lesions often associated with precocious puberty (PP) and gelastic seizures (GS) or Other Seizure Types. The incidence of HHs has been estimated ranges from 1/100,000 to 1/1,000,000. HHs also combined with: Cognitive impairment, Behavioral and psychiatric problems and other malformations: Pallister–Hall syndrome
Case 141, M, 2yr, GS after birth, PP 1yr
Case 10, F, 10yr, Hydrocephalus
Case 12, M, 4yr, Callosal agenesis
Case 54, M, 14yr, Arachnoid cysts
M, 9mo, HH with polydactyl, Pallister-Hall syndrome
Case 315, Mr. 3yr, HH with polydactyl, Pallister-Hall syndrome
Classification of hypothalamic hamartoma

- Several classification for HHs: Valdueza et al., Regiset al. and Delalande and Fohlen. The Delalande classification system(2003) are used widely.
Classification system for HH, proposed by Luo (in Chinese 2004; in English 2014) were used in Beijing Tiantan Hospital, China.

Classification of hypothalamic hamartoma and prognostic factors for surgical outcome

Figure 1. Classification of hypothalamic hamartoma (Types I-IV).
Type I: 35.9%
Type II: 12.1%
Type IV: 11.2%
<table>
<thead>
<tr>
<th>Type</th>
<th>PP</th>
<th>GS</th>
<th>EP</th>
<th>Asym</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I</td>
<td>82%</td>
<td>4%</td>
<td>10%</td>
<td>9%</td>
</tr>
<tr>
<td>Type II</td>
<td>46%</td>
<td>27%</td>
<td>50%</td>
<td>19%</td>
</tr>
<tr>
<td>Type III</td>
<td>43%</td>
<td>74%</td>
<td>92%</td>
<td>1%</td>
</tr>
<tr>
<td>Type IV</td>
<td>8%</td>
<td>92%</td>
<td>92%</td>
<td>4%</td>
</tr>
</tbody>
</table>
92 HHs in recent 5 years

- 92 patients (55M, 37F) underwent surgery between January 2011 and December 2015. All of them experienced several types of seizure (gelastic, tonic, partial, atonic, generalized tonic clonic)
- The average age was 6.8 years (range: 1.5 to 34 years)
- Type II: 8 ; Type III: 47 ; Type IV :37
- All patients were operated by microsurgical resection under direct vision
- Surgical approach selection:
  - Type II: Pterional approach or Orbitozygomatic approach
  - Type III & IV: Transcallosal interforniceal approach
- The average Follow-up time were 31 months (range from 4 to 63 months).
Orbitozygomatic approach

- M, 6y
- GS 2y and EP for 4 mo
- MRI: type II
- OP: Left orbitozygomatic approach, total removed
- Post OP: uneventful and seizure-free for 12 mo
Transcallosal interforniceal (TIF) approach (Type III and IV)
retraction of the columns of the fornix
Case 1: F, 6y

- Canadian ethnic Chinese, 2y GS, EP, Diagnosed HH in Toronto Sick Kids Hospital, AED, no effect. Cognition and movement retrograded and became mutism and living in wheelchair for 6 mo.
- Transcallosal Interforniceal Approach: 85%
- After op: EP reduced 95% and became normal girl
case 2: M, 6y

- GS and EP
- CT and MRI: type IV small HH
- OP: TIF
- seizure-free for 3 years
case 3, M, 4y11mo

- GS after birth, PP: 2.5y; EP: 4y9mo
- MRI: type II HH
- OP: TIF: 85%
- PostOP: temporary hypothalamus damage and seizure-free for 16mo
Reoperation case

- F, 13y
- 3mo: GS; 3y: EP
- 2009 (6y): other hospital: partial removed with no effect.
- 2015: TIF removed about 85%, infected after OP and VP for hydrocephalus 1 mo later. seizure-free for 10mo
- Pre-op
- Post-op
<table>
<thead>
<tr>
<th>Type</th>
<th>No</th>
<th>Approach</th>
<th>Resection extent achieved(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>PA</td>
<td>OA</td>
</tr>
<tr>
<td>II</td>
<td>8</td>
<td>5</td>
<td>1</td>
</tr>
<tr>
<td>III</td>
<td>47</td>
<td></td>
<td></td>
</tr>
<tr>
<td>IV</td>
<td>37</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>92</td>
<td>5</td>
<td>1</td>
</tr>
</tbody>
</table>
# Outcomes of seizure

<table>
<thead>
<tr>
<th>Type</th>
<th>Seizure-free</th>
<th>&gt;90% reduction</th>
<th>&gt;50% reduction</th>
<th>&lt;50% or no impro</th>
<th>Total No</th>
</tr>
</thead>
<tbody>
<tr>
<td>II</td>
<td>5 (62.5%)</td>
<td>1 (12.5%)</td>
<td>0</td>
<td>2 (25%)</td>
<td>8 (100%)</td>
</tr>
<tr>
<td>III</td>
<td>31 (67%)</td>
<td>9 (20%)</td>
<td>4 (8.7%)</td>
<td>2 (4.3%)</td>
<td>46 (100%)</td>
</tr>
<tr>
<td>IV</td>
<td>36 (97%)</td>
<td>0</td>
<td>1 (3%)</td>
<td>0</td>
<td>37 (100%)</td>
</tr>
</tbody>
</table>

1 child (Type III) die (1/47)
Complications

- One patient (Type III) died of the damage of hypothalamus (2011) (mortality: 1.1%)
- Permanent hypopituitarism: 1 patient
- Hydrocephalus: 2 cases (VP shunt)
- Transient diabetes insipidus and hyponatremia or hypernatremia: 31 (34%)
- Memory loss: 23 (25% transient), but only 5 (5.4%) permanent
- No hemiparesis and no hemorrhage
<table>
<thead>
<tr>
<th>Treatment modality</th>
<th>Seizure freedom</th>
<th>Indications</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Microsurgery</td>
<td>52-66%</td>
<td>Large HH with a significant intraventricular component located superior to the level of optic tracts</td>
<td>Immediate improvement of seizure activity. Factors favoring a transcallosal approach include younger age (interfrontal), smaller lesion, and less damage to optic tracts.</td>
</tr>
<tr>
<td>a. Transcallosal, interforniceal</td>
<td></td>
<td>HH with horizontal implantation plane, lateralized on to one side</td>
<td></td>
</tr>
<tr>
<td>b. Pterional/orbitozygomatic approach</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Endoscopic disconnection</td>
<td>49%</td>
<td>Small, intraventricular HH with a unilateral, vertical plane of attachment; the HH is approached through the contralateral foramen of Monro</td>
<td>Advantages: HH is approached without disturbing the trigone, surgical field is less cluttered. Stereotactic guidance.</td>
</tr>
<tr>
<td>(stereotactic guidance)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Radiosurgery</td>
<td>37% after a delay of 6-12 months</td>
<td>Small primary and secondary (residual) HH; preferred in adolescent and adults with intact memory and milder epilepsy as well as in select patients with bilateral mammillary body attachment</td>
<td>Latency period for epileptiform activity, potential injury to optic tract, memory is lower compared to microsurgery.</td>
</tr>
<tr>
<td>Radiofrequency ablation</td>
<td>Isolated case reports</td>
<td>Primary treatment of small HH and management of residual lesions</td>
<td>Minimally invasive, low morbidity. Disadvantages of tissue ablation and limited trajectory for larger lesions.</td>
</tr>
<tr>
<td>Combined modalities</td>
<td></td>
<td>Larger lesions depending on extent of resection or disconnection, seizure remission achieved with initial treatment</td>
<td>Individualization of approach, age, size, and anatomy experience.</td>
</tr>
</tbody>
</table>

**Comparison of various treatment modalities for hypothalamic hamartomas**

*Jayalakshmi S; et al (Annals of Indian Academy of Neurology, 2014, 17:43-7)*
Conclusion

1. Type IV (Delalande’s Type II): Transcallosal interforniceal approach is safe and can get the best result (total resection and seizure free 97%)

2. Type III (Delalande’s Type IV): TIF can also get subtotal resection in the middleline and improved the outcome (48% to 67%)

3. Type I (Delalande’s Type I) and Type II (Delalande’s Type III): Pterional or Orbitozygomatic approach is suitable

4. Microsurgical resection under direct vision can get very good results for experienced neurosurgeon
Thank you