Speech and Cognition After Hemispherectomy for Hemimegalencephaly: A Report from the Global Pediatric Epilepsy Surgery Registry

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Hemimegalencephaly (HME): rare congenital malformation of the brain characterised by the over-growth of one hemisphere

Rationale
The cause of HME is unknown, but HME is associated with severe seizures, contralateral motor deficit, and global developmental delay. Multiple antiseizure drugs are often required, but when seizures don’t respond to medications, hemispherectomy is advocated as the best choice of treatment for children. Hemispherectomy (total or partial removal of an affected cerebral hemisphere) is used for children with HME and hemimegalencephaly, and there is some evidence of improved cognitive and behavioral outcomes.

Objective
1. Describe the cognitive and language outcomes by articulating sounds, and reading ability.

Measures
Parental- and child-proxy instruments (Upper Extremity; Cognitive Function; Anger; Mobility; and Peer Relations). The questionnaires and Patient Reported Outcomes Measurement Information System (PROMIS) short-form measures were completed through the study website using REDCap electronic data capture tools through a series of steps.

Results
Cognitive and language skills at follow-up

Participant Characteristics

<table>
<thead>
<tr>
<th>Number of participants</th>
<th>45 (18 female, 27 male)</th>
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</thead>
<tbody>
<tr>
<td>Age at seizure onset</td>
<td>6.5 months (5.0-8.0)</td>
</tr>
<tr>
<td>Age of surgical candidacy evaluation</td>
<td>6.5 months (5.0-8.0)</td>
</tr>
<tr>
<td>Time from seizure onset to hemispherectomy</td>
<td>4.5 months (3.0-5.5)</td>
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<tr>
<td>Age at first hemispherectomy</td>
<td>6.0 years (5.5-6.5)</td>
</tr>
<tr>
<td>Years follow-up</td>
<td>8.0 years (6.5-9.0)</td>
</tr>
<tr>
<td>Age at follow-up</td>
<td>8.0 years (6.5-9.0)</td>
</tr>
</tbody>
</table>

Statistical Analyses

Analyses including a total of 45 children were conducted using IBM-SPSS (IBM, Inc. Corp., Cary, NC, USA). Descriptive statistics computed for the sample included means and standard deviations (SD) for continuous variables, frequencies and percentages for categorical variables. The association between each cognitive/language outcome with the following clinical characteristics was evaluated using a multivariable regression model: age at surgery, age at last resective surgery, side of hemispheric resection, and seizure-free status at follow-up (none vs. at least one seizure per year).

Language milestones attained

- Bobbled
  - 0-1 months
  - 1-2 months
  - 2-3 months
  - 3-4 months
  - 4-5 months
- First words
  - 6 months
  - 7-8 months
  - 8-9 months
  - 9-10 months
  - 10-11 months
  - 11-12 months
- First sentences (1 words)
  - 12-13 months
  - 13-14 months
  - 14-15 months
  - 15-16 months
  - 16-17 months
  - ≥18 months

Spearman correlations

<table>
<thead>
<tr>
<th>Cognition</th>
<th>Language</th>
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<tbody>
<tr>
<td>Age at surgical candidacy</td>
<td>0.38 (p &lt; 0.05)</td>
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<tr>
<td>Age at last resective surgery</td>
<td>0.32 (p &lt; 0.05)</td>
</tr>
<tr>
<td>Seizure-free status at follow-up</td>
<td>0.08</td>
</tr>
<tr>
<td>p-value</td>
<td>0.015</td>
</tr>
<tr>
<td>95% confidence interval</td>
<td>(-0.01, 0.59)</td>
</tr>
</tbody>
</table>

Discussion

Highlights

- Children who had undergone a right hemisphere resection had better cognition and reading skills.

- The registry enrolls parents/guardian volunteers of children with epilepsy who have had epilepsy surgery, are taking epilepsy surgery, are unknown or not known in the clinical questionnaires. Patients are recruited via email and social media. Information on current and potential volunteers is collected through the study website. The epilepsy surgery data capture tool is through a series of steps.

- The study explores clinical and demographic predictors of superior outcomes, including the proposed reasons for superior outcomes, and long-term outcomes.

- The registry was developed to provide parents/guardian volunteers of children with epilepsy who have had epilepsy surgery, are taking epilepsy surgery, are unknown or not known in the clinical questionnaires. Patients are recruited via email and social media. Information on current and potential volunteers is collected through the study website.

References