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[1]. 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[2]. **Hemispherectomy** (total or partial removal of an affected cerebral hemisphere) **is an effective treatment option**, resulting in seizure control in 60-85% of cases, and there is some evidence of improved cognitive and behavioral outcomes[1,2].

Past studies of cognitive and language outcomes after a hemispherectomy among children with HME have been limited by:

- small sample sizes
- poor reporting or operationalization of cognitive outcomes

One study reports that better developmental and language outcomes are associated with seizure control, and a shorter duration between seizure-onset and hemispherectomy[5].

Objective

1) Delineate the **cognitive and language outcomes** of hemispherectomy for children with HME

2) Identify the characteristics that were associated with **optimal outcomes**

Measures

Parents used Likert-type scales to report on their children's cognitive and language skills. Specifically, parents reported the age at which their child reached various language milestones, and their children's current cognitive ability, use of speech (described to parents as the ability to express thoughts and feelings by articulating sounds), and reading ability.

Global Pediatric Epilepsy Surgery Registry

World's first patient-driven, web-based registry allowing parents and guardians of children with epilepsy to report on outcomes after epilepsy surgery

The registry recruits parents/legal guardians of children with epilepsy who have had epilepsy surgery, are being evaluated for epilepsy surgery, declined surgery, or were not found to be surgical candidates. Parents are recruited via email and social media. Informed consent and baseline surveys in English are completed through the study website using REDCap electronic data capture tools through a series of questionnaires and Patient Reported Outcomes Measurement Information System (PROMIS) short-form parent proxy instruments (Upper Extremity; Cognitive Function; Anger; Mobility; and Peer Relations). The surveys collect information on demographics, seizure onset and characteristics, etiology, surgical evaluation, surgery, seizure outcomes, comorbidities, development, and quality of life. Annually, parents will be asked to update responses to the surveys until the child reaches age 18. The goal of the registry is to have 250 participants by the end of 2020. It has no end date.

Participant Characteristics

Number of participants: 45
(18 female, 27 male)

Age at seizure onset: 2.6 months
(SD 6.5, range: -2.5 months — 33.5 months)

Age at surgical candidacy evaluation: 6.5 months
(SD 12.3, range: -0.5 months — 5.1 years)

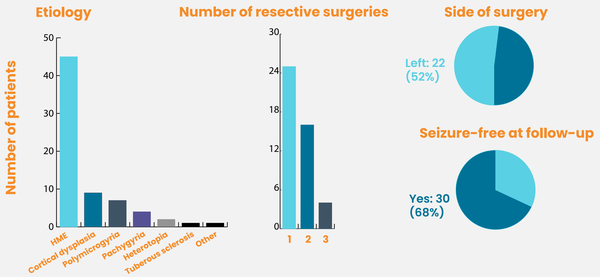
Time from seizure onset to hemispherectomy: 8.3 months
(SD 8.4, range: 0.6 months — 5.5 months)

Age at first hemispherectomy: 0.9 years
(SD 1.1, range: 1.5 months — 5.5 years)

Age at last resective surgery: 2.1 years
(SD 2.2, range: 1.5 months — 8.1 years)

Years follow-up: 6.6 years
(SD 4.5, range: 2.5 months — 17.4 years)

Age at follow-up: 8.7 years
(SD 4.8 , range: 12.0 months — 18.0 years)



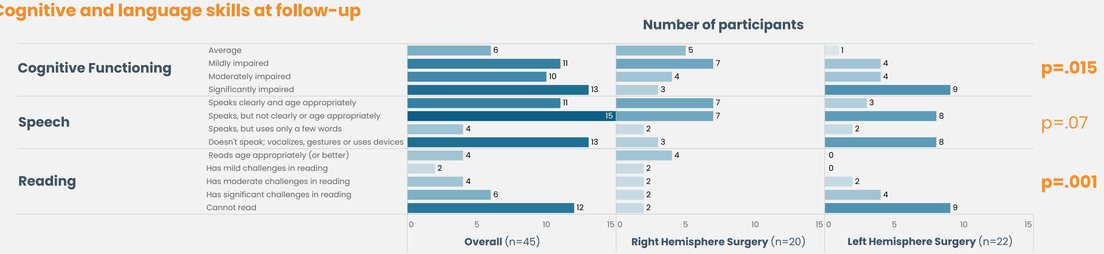
Statistical Analyses

Analyses were conducted using SAS 9.4 (SAS Institute Inc. Cary, NC, USA). Descriptive statistics computed for the sample included means and standard deviations (SD) for continuous variables, and frequencies and percentages for categorical variables. The association between each cognitive/language outcome with the following clinical characteristics was evaluated: side of surgical resection, age of seizure-onset, age at first hemispherectomy, age at follow-up, and seizure status at follow-up (none vs. at least one per year).

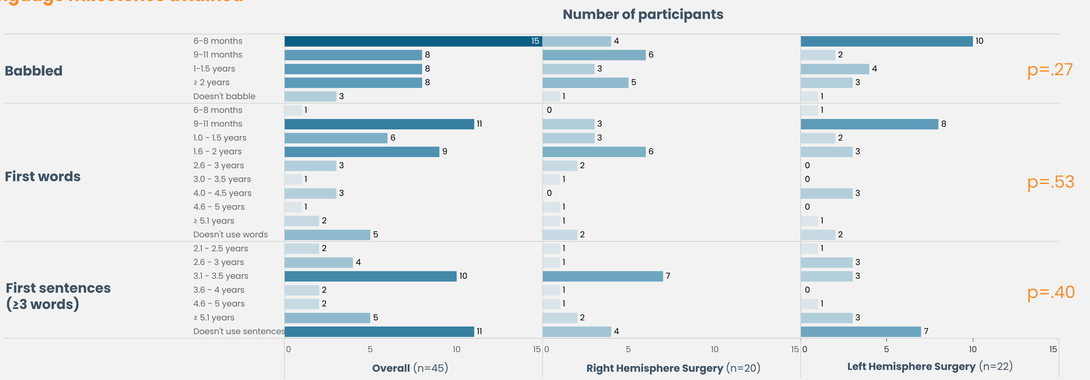
Given that the cognitive/language outcomes were ordinal, the exact Cochran-Armitage test for trend was used when the clinical variable was binary, and the Spearman correlation was used when the clinical variable was continuous. The Spearman correlation coefficient is interpretable as very weak (.00-.19), weak (.20-.39), moderate (.40-.59), strong (.60-.79), or very strong (.80-1.0).

Results

Cognitive and language skills at follow-up



Language milestones attained



Spearman correlations

			Age at seizure onset	Age at first hemispherectomy	Time from seizure onset to hemispherectomy	Age at follow-up
Cognitive and language outcomes at follow-up	Cognition	correlation coefficient	0.38	0.17	0.08	-0.2
		95% confidence interval	(0.07, 0.62)	(-0.15, 0.46)	(-0.24, 0.38)	(-0.48, 0.12)
		p-value	**p= .016**	p= .30	p= .64	p= .23
	Current Speech	correlation coefficient	0.37	0.25	0.12	0.26
		95% confidence interval	(0.08, 0.61)	(-0.05, 0.52)	(0.41, -0.19)	(-0.05, 0.52)
		p-value	**p= .015**	p= .11	p= .44	p= .09
	Current Reading	correlation coefficient	0.29	0.11	-0.01	0.14
		95% confidence interval	(-0.09, 0.60)	(-0.27, 0.47)	(-0.38, 0.37)	(-0.24, 0.49)
		p-value	p= .13	p= .57	p= .98	p= .47
Attainment of language milestones	Babble	correlation coefficient	0.31	-0.1	-0.24	0.03
		95% confidence interval	(0.01, 0.57)	(-0.40, 0.21)	(-0.51, 0.07)	(-0.28, 0.33)
		p-value	**p= .046**	p= .53	p= .12	p= .86
	First words	correlation coefficient	0.28	0.04	-0.08	-0.23
		95% confidence interval	(-0.03, 0.54)	(-0.27, 0.34)	(-0.38, 0.23)	(-0.50, 0.08)
		p-value	p= .07	p= .81	p= .63	p= .15
	First sentences	correlation coefficient	0.32	0.36	0.25	0.05
		95% confidence interval	(-0.01, 0.59)	(0.04, 0.62)	(-0.08, 0.54)	(-0.28, 0.37)
		p-value	p= .06	**p= .029**	p= .14	p= .76

Highlight

Side of hemispheric resection was associated with cognition and language outcomes at follow-up:

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

References

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