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Language recovery after left hemispherotomy for Rasmussen encephalitis

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ABSTRACT

Purpose: Hemispherotomy (H) is the gold standard treatment to cure epilepsy in Rasmussen encephalitis (RE). Linguistic prognosis after surgery remains the main issue when the dominant hemisphere is involved. The topic of the present research is to specify the long-term linguistic profile of the right hemisphere after left dominant H for RE.

Methods: We followed 6 children 8.4 to 14.6 years of age who underwent left H for RE. Preoperatively, four children experienced aphasia, but for two, worsening occurred after surgery. Age at H ranged from 4.1 to 8.4 years. The mean duration of epilepsy was 1.2 years and 5.6 years for follow-up. Neuropsychological evaluation included longitudinal follow-up of intellectual efficiency measurement and a long-term outcome of language using various components of receptive and expressive oral speech with computerized tasks.

Key findings: Preoperatively, verbal comprehension index (VCI) was dramatically decreased in 4/6 patients, and performance reasoning index (PRI) was low in 5/6 participants, demonstrating a global impact of RE itself. Postoperatively, all children recovered sufficiently to attend a regular VCI (above 70) in a mean of 5 years after H, and 5/6 recovered normal or adapted school. There was a dissociation in favor of VCI, while PRI decreased in 5/6 patients. We found a specific linguistic profile for these children recovering language in the right hemisphere: normal verbal comprehension, and weakness of grammatical judgment, word repetition, statement production, semantic verbal fluency and metaphonological abilities. Language recovery scores were statistically correlated with those of Working Memory Index.

Significance: This study emphasizes for the first time the ability of the right hemisphere to functionally reorganize language over a long period of time following left H for RE. Syntactic abilities and phonology remain low and support the hypothesis of an early left hemispheric specialization. Nevertheless, lexico-semantic processes recover in the right hemisphere that could reflect a pre-existing potential of both hemispheres. Our results support a decision to proceed to H in classical left RE disease until the late childhood even if there is no complete aphasia before surgery. These data should be taken in account in the overall postoperative follow-up and rehabilitation strategy.

1. Introduction

Rasmussen encephalitis (RE) is a rare and severe epileptic and inflammatory unilateral encephalopathy which occurs in previously healthy children, and it is associated with progressive motor and cognitive deficits, associated with language disturbances if the disease affects the left language dominant hemisphere [1]. The gold standard treatment of RE is hemispherec/rotomy (H) [2–4]. Seizure control is obtained in over 90% of cases, with a very meaningful improvement of autonomy and quality of life, even if motor and neurovisual sequelae are unchanged and functionally significant [5]. When RE affects the left hemisphere, the surgical decision still remains uncertain and often hesitant because of the threat of aphasia, although definitive aphasia has not been well demonstrated. On the other hand, postponing or avoiding H runs the risk of persistence of severe epilepsy, including episodes of status epilepticus, with slow and insidious but inexorable deterioration of cognition [6–8]. Among the main issues regarding the management of RE, that of the timing of left H and the potential for language recovery remain a matter of debate.

Long-term postoperative follow-up after left H shows that the right hemisphere is able to subserve functional speech. However, while specific linguistic abilities develop optimally, others do not [9–14]. Nevertheless, data regarding substitution abilities of the right hemisphere for speech in children with ongoing left RE are sparse [15–18], and postoperative long-term outcome data are inexistant.

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The aim of this study was to determine the linguistic profile of 6 patients with RE who underwent left dominant H [4]. All of them had longitudinal follow-up of intellectual quotient and speech skills. Moreover, a detailed investigation of speech was performed 5.6 years postoperatively that not only permits the evaluation of abilities but also limits the abilities of the right hemisphere to take over speech after aphasia occurring in childhood. We analyzed features (age at onset of epilepsy and of H, duration of epilepsy, preoperative language, and postoperative follow-up) considered as predictive of language recovery.

2. Material and methods

2.1. Participants (Table 1)

Six subjects participated in this research. All had RE involving the left, language-dominant and underwent left H [4] at our institution. These 6 subjects (6 females) were prospectively followed up in our department between 2003 and 2008 with preoperative and postoperative neuropsychological evaluation. During the same period, 98 children underwent H including 16 for RE (of which 6 were from non-French-speaking descent and 4 were French-speaking but operated on the nondominant hemisphere). The medical variables are given in Table 1. The mean age at epilepsy onset was 4.8 years (SD: 1.6), and children were operated on at a mean age of 6.1 years (SD: 1.9). Before the onset of seizures, all the children were completely healthy. Preoperatively, all patients had focal seizures, four of them experienced epilepsia partialis continua (EPC), and one had secondarily generalized seizures. The mean duration of epilepsy was 14 months (SD: 5 months), which was relatively short and homogenous among this population. Patients experienced speech regression in relation to the increase of seizure frequency and progressive hemisphere atrophy due to RE that worsened before H (4/6) or after left H (2/6).

Following H, all six subjects became seizure-free (Engel Class I) and could be weaned from antiepileptic medication. All had right hemianopsia and hemiparesis predominating in the upper limb, permitting autonomous walking. All children and their parents gave informed consent to contribute to the research which was approved by local ethics committee. Two mothers were classified as level 1 socioprofessional category (low level), three mothers to level 2, and one mother to level 3 (higher level).

2.2. Neuropsychological assessment (Table 2)

For the six children, we collected data on preoperative intellectual efficiency and language evolution when the child was first referred to our institution.

When indication of H was confirmed, a preoperative cognitive assessment was made using the Wechsler scale and a qualitative evaluation of language at our institution. This cognitive follow-up was then redone twice (12 to 18 months/2 to 3 years) postoperatively. Finally, all six children underwent a third extensive long-term evaluation following left H with a comprehensive battery of language tests including lexical, phonological, and syntactical processes. Neuropsychological testing consisted of measurement of intellectual efficiency with the WISC-IV scale [19] and detailed examination of oral speech using the Phonemic Discrimination Scale (EDP 48) [20], the Computerized Oral Speech Battery (BILO) [21], and complementary verbal tasks (verbal fluency and metaphonology). This evaluation was performed 5.6 years (mean) following surgery.

The EDP 48 evaluates auditory phonemic discrimination abilities for opposition consonants. This scale was used in order to disclose auditory discrimination troubles that could have interfered with the interpretation of findings concerning language features, as seen in children with febrile seizures [20].

The BILO allows evaluating the various components of receptive and expressive oral speech, taking into account specificities of speech according to age. One of the major advantages of this battery is that being computerized, it takes into account the time required to perform various tasks. Six tasks were performed in order to evaluate lexicon (Oral Comprehension, Lexical Judgment, and Lexical Production), phonology (Word Repetition), and morphosyntax (Grammatical Judgment and Statement Production).

Two metaphonological tasks (suppression of phoneme and phoneme fusion task) evaluated the level of phonological consciousness and were drawn from the dyslexia evaluation battery called ODEDYS [22].

Verbal fluencies (VFs) assess evaluating the access to the lexicon based on a given criterion, whether semantic (SemVF), categorical, phonological (PhonVF), or literal. The test used [23] comprises categorical (animals) verbal fluency task and a phonological (letter m) verbal fluency task. For each, the subject has to produce maximum words in 1 min.

2.3. Analysis methods

This study started with a descriptive investigation of the course of individual intellectual and speech performances.

For the analysis of the third and extensive long-term outcome, the z-test or typicality test was used to compare the observed means with normal and determine whether the observed performances significantly differed from the normal. Given the small number of subjects, nonparametric tests were applied. The Wilcoxon test for paired samples was used to compare the performances among each other and identify the strong and weak points for each studied domain. Nonparametric correlations (Rho of de Spearman) were performed to examine the potential link between the various areas.

3. Results

3.1. Evolution of the intellectual profile (Fig. 1)

3.1.1. Preoperative evolution of the intellectual profile

Given that the children were healthy before seizure onset, there was massive impact of RE on neuropsychological functioning. Three children (#1 — PL, #2 — ML, and #3 — IK) had intellectual efficiency measurement early after the onset of seizures, two (#2 — ML and #3 — IK) of

Table 1
Medico-surgical features (in years).

Subjects	Sex	Age at onset of epilepsy	Duration of epilepsy	Age at surgery	Postoperative follow-up	Age at evaluation	Schooling
1 — PR	F	4.1	1.3	5.4	3	8.4	Normal
2 — ML	F	4	1.1	5.1	3.6	8.7	Adapted
3 — IK	F	2.6	0.8	3.4	8.3	11.7	Adapted
4 — OG	F	5.1	1.1	6.2	6.3	12.5	Specialized
5— LF	F	6.1	2	8.1	6	14.1	Adapted
6 — PRe	F	7.2	1.2	8.4	6.2	14.6	Adapted
Mean (SD)		4.8 (1.6)	1.2 (0.4)	6.1 (1.9)	5.6 (1.9)	11.7 (2.6)	