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Early hemispherectomy in catastrophic epilepsy A neuro-cognitive and epileptic long-term follow-up

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Summary The authors report their experience about a neuro-cognitive and epileptic long-term follow-up of children with catastrophic epilepsy treated with hemispherectomy in the first 5 years of life.

Nineteen children with resistant epilepsy that significantly interfered with their neuro-cognitive development underwent hemispherectomy within 5 years of life (mean: 2 years, 3 months; range: 5 months to 5 years). All patients were assessed before surgery and after, at least at the end of the follow-up (mean: 6 years and 6 months; range: 2–11 years and 2 months) with a full clinical examination including motor ability and functional status evaluation as well as behaviour observation, neuroimaging and an ictal/interictal prolonged scalp video-EEG.

A seizure-free outcome was obtained in 73.7% of patients. Gross motility generally improved and cognitive competence did not worsen, with an evident progress in two cases.

Consistently with previous reports, evolution was worse in cortical dysplasia than in progressive or acquired vascular cerebropathies. The excellent epileptic outcome and the lack of developmental deterioration in comparison with other more aged series seem to suggest a possible better evolution in earlier surgery treatment. To confirm this suggestion, however, further experience with larger series is needed.

Introduction

Hemispherectomy has been successfully used to treat various types of medically intractable

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hemispheric epileptogenic conditions, mostly in children and adolescents.[1–5] Indeed, the procedure has proved to provide excellent seizure control and improvement in the quality of life in a high percentage of the cases.[2,6–13]

In more recent times, increased interest has been paid towards the possible advantages of an early referral for elective surgery in infants and very young children. Several factors account for the progressively diminishing reluctance for an earlier treatment. First, the cumulated experiences of the deleterious effects of the so-called catastrophic epilepsies. Then, the conviction that a precocious removal or disconnection of the epileptogenic hemisphere may favour the compensatory reorganization of the healthy brain besides avoiding the interference of seizures with the early neurological development.[2,4,14] Further contributing factors should be identified in (1) the earlier recognition of an organic lesion and, consequently, the possibility of an early surgical indication, which is nowadays allowed by the modern diagnostic tools for anatomical and functional neuroimaging, (2) the reduced complication rate of this kind of operation, which results from the introduction of less heavy technical surgical variants, and (3) the progress in intraoperative anesthesiology and postoperative intensive care.

In the absence of randomized studies on early versus late surgery, which can not be proposed for ethical reasons, and in the light of the difficulty to compare relatively small and un-homogeneous clinical series, we considered that the report of our own experience, concerning 19 young children surgically treated in the first five years of life and fully investigated preoperatively and followed postoperatively for a relatively long period of time, could contribute to provide further insight into the outcome of young children undergoing hemispherectomy, with particular regard to the timing of the surgical treatment and the aetiologies of the epilepsy.

Patients and methods

Among the patients treated with hemispherectomy within 5 years of age in the Child Neurosurgery Unit of our Hospital, from 1980 to December 2003, we enrolled in the study only 19 thoroughly studied children, six of which had been reported in a previous paper.[10]

This study was retrospective for the patients who underwent hemispherectomy before December 1996, and prospective for those treated between January 1997 and December 2003.

The patients were examined for the surgical candidature after a non-effective drug treatment that made epilepsy to be considered refractory: epilepsy was considered drug resistant when at least three drugs at the maximal dosage were used without seizure control.

All patients were assessed in the Child Neurology Unit before and after surgery using a standard procedure as follows:

- full clinical examination with a detailed anamnestic history;
- ictal/interictal prolonged scalp video-EEG examination;
- neuroimaging (mainly, MRI);
- evaluation of motor abilities;
- neuropsychological assessment (general intelligence and specific abilities);
- behaviour observation;
- evaluation of functional status.

Generally, after surgery this procedure was performed at outcome; in some cases there was a serial assessment.

EEG and seizure evaluations

The pre-surgery EEG was aimed at evaluating type, localisation, extent, frequency and amplitude of interictal epileptiform discharges in both the affected and healthy hemispheres; the presence of independent, asymmetric or synchronic discharges was also considered. Moreover, ictal patterns were analyzed, with particular attention to the focal location of seizure onset and the type of extension.

Epilepsy was classified according to the International League Against Epilepsy (ILAE) classification (1989).[15]

Seizure outcome was assessed using the Engel's scale. Engel's classification[16] includes four classes of worsening epileptic outcome, ranging from a ''seizure-free'' condition (class I) to a ''not worthwhile improvement'' (class IV). Furthermore, every class is subdivided into sub-categories with different degrees of outcome severity (increasing from a to d).

Neuroimaging

All the patients studied from 1980 to 1992 (# 1, 2, 11, 14 and 15) underwent brain CT scan examination before and after surgery, using uninter-leaved 5 mm thick axial slices.

All the patients evaluated from 1992 to 1998 (# 4, 5, 6, 8, 9, 12, and 13) were examined using a 0.5 T