FUNCTIONAL HEMISPHERECTOMY VARIANT A REPORT OF TWENTY ONE CASES

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ABSTRACT

Objective and importance: *Although Rasmussen's hemispherectomy is a proven effective technique, the reduction in surgical time and in blood volume lost during its performance is crucial. This is achieved by means of the technical variant intended to disconnect instead of to remove the temporal lobe.*

Clinical presentation: *We present 21 children operated between 1997 and 2004 in whom a functional hemisfactory (FH) variant was performed. The clinical manifestations included the following crises: hemianopsia, versive crises, absences, CPC and tonic crises, with secondary generalization in some of the children.*

Intervention: *The proposed variant is a sort of combination of Rasmussen's hemispherectomy and Delalande's hemispherectomy, involving disconnection and isolation of the temporal lobe. This is achieved at cortical level by prolonging the incision of the occipital lobe disconnection up to the floor of the middle fossa. The incision of the mesial aspect of the occipital lobe by intraventricular route is extended towards the fornix to reach the free border of the cerebellar tentorium. The juncture of both incisions disconnects the neocortex from the temporal lobe, as well as the parahippocampal gyrus and the hippocampus itself. Since aprevious complete callosotomy, a temporal stem section and amigdala suction had been performed, the temporal lobe remains* in situ, *but non-functional.*

Conclusion: *It is the author' conviction that Rasmussen's Hemipherectomy is a valid procedure given its proven efficacy. We believe that the proposed technical variant is particularly useful for neurosurgeons that are in training in the field of epilepsy surgery, as a prior stage to the use of more restricted disconnecting techniques.*

Key words: *Functional hemispherectomy, Technical variant, Training Neurosurgeons*

INTRODUCTION

Among the major resective and hemispheric disconnecting procedures, regardless of the advent of new more sophisticated and less aggressive techniques[1,6,8,9] developed during the last decade to replace earlier ones, anatomical hemispherectomy[3,11] and Rasmussen's functional hemispherectomy[7,10] are still in regular use[5].

This is due to the fact that neurosurgeons performing epilepsy surgery must deal with the treatment of extensive hemispheric lesions or large epileptogenic areas that involve an entire hemisphere. Should the course of disease indicate with an acceptable degree of certainty that the patient under study possesses a "healthy" hemisphere, then quite obviously resection/disconnection of the pathological hemisphere is mandatory, not only to control seizures but also to ensure that the so-called healthy side fully recovers the functions inhibited or disturbed by the anomalous activity of the contralateral hemisphere.

In the opinion of the author, in these circumstances an ample exposure of the pathological hemisphere somehow guarantees the viability of the required disconnection. The new techniques

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collectively termed hemispherotomy[1], hemispheric deafferentations[8] or peri-insular disconnection[9] imply a certain degree of expertise. These issues, together with some intraoperative findings to be described more fully below, led to the development of the classic functional hemispherectomy variant presented herein.

Material and method

Of a series of 27 patients with epilepsy who underwent functional hemispherectomy (FH), we present 21 children operated between 1997 and 2004 in whom a Rasmussen FH Variant was performed. Eight of these children presented epilepsia partialis continua secondary to Rasmussen encephalitis. Five patients presented epilepsy secondary to secuelar lesions, one of them with severe epilepsy due to tumor removal and radiotherapy one year before and another with startle epilepsy[4]. Five patients presented hemimegaloencephaly. In two patients, epilepsy was associated with widespread focal cortical dysplasia and in one patient no pathological specimen was taken.

The clinical manifestations included the following crises: hemianopsia, versive crises, absences, CPC and tonic crises, with secondary generalization in some of the children.

In one of the patients who suffered from Rasmussen encephalitis, a hemispherotomy had been performed one year earlier.

Preoperative Video-telemetry was performed in all patients, recording not less than 10 epileptic events for each one.

All twenty one met the criteria of hemiplegia and hemianopsia before surgery.

Three examples are described, one with epilepsia partialis continua secondary to Rasmussen encephalitis, the second one with simple partial motor seizures on the left side due a secuelar post-tumoral removal epilepsy and another with complex partial seizures featuring a hypotonic component and lethargy and one with startle epilepsy.

CASE REPORTS

Patient 1

This girl was in good health up to age 4 years, when she presented left isolated focal simple clonic seizures. At age 5 years she developed complex partial and left hemi-generalized seizures. As from age 7 years, there was left epilepsia partialis continua and status epilepticus that required hospitalization on several occasions. Six months later she developed left hemiplegia, becoming unable to walk unaided.

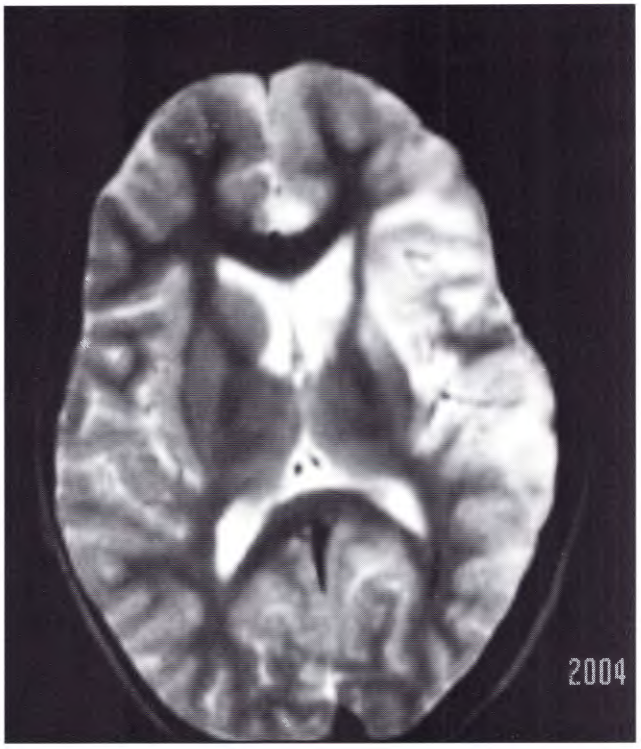

Fig. 1. MRI: Typical image of a Rasmussen encephalitis 3 years progress of epilepsia partialis continua.

At age 7 years an MRI showed signs of fronto-temporo-occipital hemiatrophy, with right supratentorial ventriculomegalia (Fig. 1). A Wada test disclosed left cerebral dominance for speech. The patient underwent a right functional hemispherectomy in November 1996. This patient was the first one in whom was applied the proposed variant of functional hemispherectomy.

Three months postsurgery she recovered unaided walking. Fifteen years old to date, she has remained seizure-free and without AEDs. Engel class IA. Hemiparesis has improved together with quality of life.

Patient 2

This girl, currently 8 years and 7 months of age, had a diagnosis of CNS tumor (ependymoblastoma) when she was 2 years and 10 months. Surgical resection was then carried out and she received adjuvant chemotherapy and skull and rachis radiotherapy. At 5 years of age she presented precocious puberty. At age 6 she developed brief, daily or weekly, simple partial motor seizures on

the left side. Initially, she was put on phenobarbital, then phenobarbital plus carbamazepine. At age 6 years there was recurrence of daily partial motor seizures, with or without secondary generalization. She received several anticonvulsivant schedules without response.

EEG studies showed spikes in the right hemisphere mainly on the frontal area. MRI demonstrated global ventricular dilatation mainly on the right. There was cortical hemispheric atrophy mostly on the same side with secondary lesion in both frontal lobes. Lastly, there was bilateral periventricular hyperintensity. A right functional hemishperectomy variant was performed in January 1999. At 5 years post-op, the patient is seizure free. Engel class IA.

Patient 3

This girl of 9 years of age, was healthy up to 8 months of age when she developed meningitis due to *Haemophyllus influenzae*. At 9 months of age, a brain CT scan showed a left hypodense fronto-temporo-parietal lesion plus subdural effusions. At age 1 years, an EEG disclosed a markedly asymmetrical focus of sharp waves, of greater amplitude and lower frequency, in the left anterior temporal region. Clinical findings included brief atonic seizures triggered by sound or somatosensitive stimuli (startle). Episodes could not be controlled with AEDs. She presented severe maturation delay, with loss of speech. Gait, drawing, scribbling and sphincter control were maintained, but right hemiparesis developed. At age 2 years, she developed partial seizures with cephalic version, eye gyration, and right tonic-clonic crises. At age 6 years, there were up to five episodes per day. Multiple therapeutic schedules failed to relieve symptoms. A brain MRI showed a wide left T_1 weighted hypointense fronto-temporo-parietal lesion. A left functional hemispherectomy variant was performed in January 1999.

To date, she experiences no startle epilepsy, the main objective of her surgical treatment. The latest Video-EEG performed in July 1999 showed that the epileptogenic focus is currently in the right hemisphere, contralateral to the operated side. Up to date she is in Engel class IIA. (Table 1).

Table 1. Demographic and clinical features of 21 epileptic cases undergoing hemispherectomy variant technique

Pt	Sex	Age at onset	Presenting Symptom	Ct/MRI Findings	Age at surgery	Follow-up	J.Engel Follow-up Clasification (> 1yr post-op)
1 (A. Ll.)	F	4 yr 6 mo	EPC	Rt. cerebral hemiatrophy	8 yr 7 mo	7 yr 8 mo	IA, without AEDs
2 (R.A.)	M	11 yr	EPC	Lf. cerebral hemiatrophy	14 yr	7 yr 4 mo	IA, with AED monotherapy
3 (M.F.)	M	10 yr	EPC	Rt. cerebral hemiatrophy	16 yr	7 yr 4 mo	IA, without AEDs
4 (M.D.)	F	6 yr	EPC	Rt. cerebral hemiatrophy	6 yr 8 mo	1 yr 2 mo	IA, without AEDs
5(O. Ch.)	F	2 yr10 mo	MPS	Rt. cortical hemispheric atrophy	6 yr 7 mo	5 yr yr 6 mo	IA, without AEDs
6 (E.V.)	F	2 yr 6 mo	Startle Epilepsy	Lf. fronto-temporo-parietal atrophy	9 yr	5 yr 6 mo	IA, without AEDs
7 (A.M.)	F	2 yr 6 mo	EPC	Lf. cerebral hemiatrophy	4 yr 6 mo	4 yr 9 mo	IA, without AEDs
8 (J.S.)	F	2 yr 6 mo	MPS	Lf. fronto-parietal atrophy	13 yr 10 mo	3 yr 11 mo	IA, without AEDs
9 (M.T.)	F	6 yr 11 mo	MPS	Rt. fronto-parieto-temporal atrophy	13 yr 10 mo	3 yr 11 mo	IA, without AEDs
10(C.L.)	F	1 mo	MPS	Rt. cerebral hemiatrophy	1 yr	3 yr 8 mo	IA
11 (J.V.)	M	12 yr 8 mo	EPC	Rt. hemiatrophy	14 yr	3 yr 2 mo	IA

Foundations of the technique

The basic concept arose from an observation when operating the first patients cases 1, 2 and 3. After resection of the central area, it was clearly visualized that the frontal and temporal cortical veins passed from a congestion stage to another of evident "arterialization". That is to say, an inversion in cerebral blood flow was unequivocal. (Fig. 2).

This event led to unavoidable temporal cortical vein clotting, opting for henceforth a disconnection and subpial "emptying" of the temporal lobe. Thus, new hemodynamic alterations were prevented. Lobe emptying was carried out from the posterior section (when disconnecting it from the occipital lobe) toward its tip, including mesial structures. (Fig. 3, A and B).

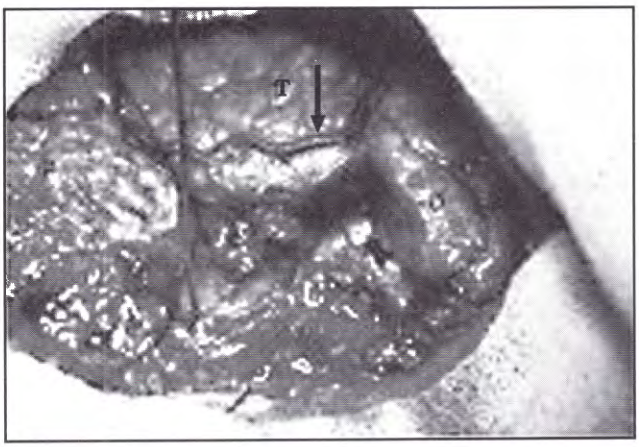

Fig. 2. Central area resection is observed. F: frontal lobe; T: temporal lobe; O: occipital lobe. The thin arrow shows the arterialization of a cortical vein and a significant increase in its size. Thick arrow: temporal lobe disconnection.

In other cases, similar hemodynamic changes were observed, attributable to unavoidable clotting of cortical drainage veins. In both cases, no emptying but temporal lobe disconnection was carried out[1] (Fig. 4, A and B).

Description of the technique

Temporal lobe isolation is achieved as follows:

a) Forwardly, by a intra-ventricular route, disconnecting the anterior white commissure (AWC), extending the anterior callosotomy up to the tip (lamina terminalis), gyrus rectus and fronto-orbital region as well.

b) Backwardly, by disconnecting the fornix, and by gradually disconnecting the mesial surface of the occipital lobe. As such surgical stage, at choroidal plexus entry level to the sphenoidal horn

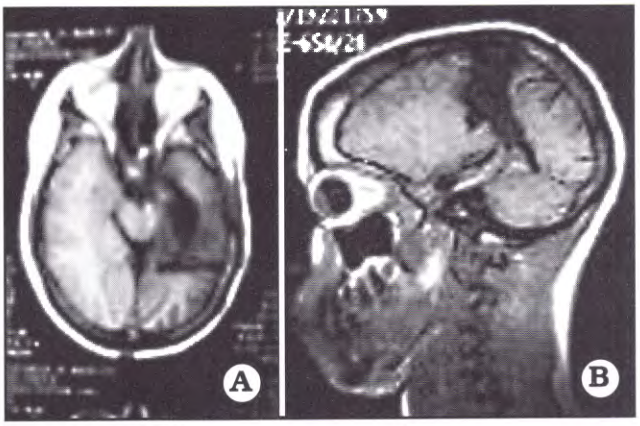

Fig. 4. A. MRI: axial view showing temporal lobe disconnection, without resection. B. MRI: sagittal view showing central area resection with temporal lobe disconnection.

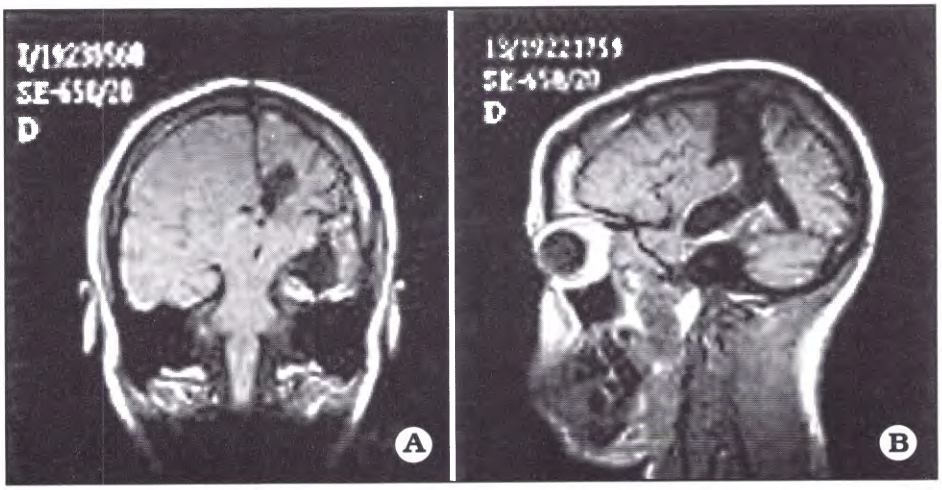

Fig. 3. A. MRI: coronal view showing temporal lobe emptying, as well as central area resection. B. MRI: sagittal view corresponding to the same patient.

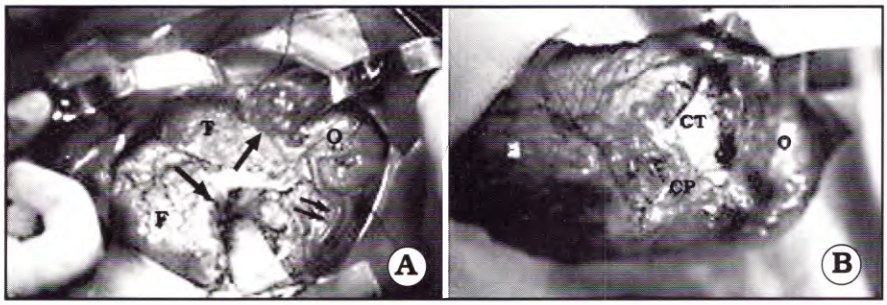

Fig. 5. A. Central area resection (thin arrow) and temporal lobe disconnection (thick arrow) from behind the insula are observed. F: frontal lobe; T: temporal lobe; O: occipital lobe. Two arrows: arterialized vein. B. F: frontal lobe; T: temporal lobe; CP: choroidal plexus; CT: cerebellar tentorium. The arrow shows the posterior disconnection of the temporal lobe.

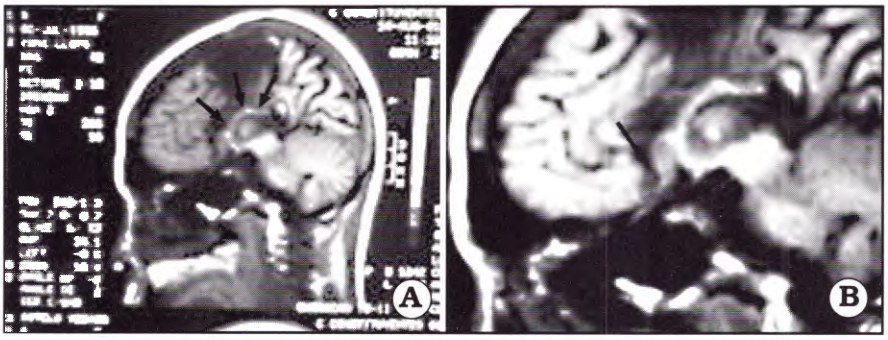

Fig. 6. A. B. Parasagital corpus collosum (three arrows) and gyrus rectus (one arrow) section.

of the lateral ventricle, the ependymum is aspirated in the direction towards the quadrigeminal cistern. Proceeding with temporal parenchyma disconnection, though sparing the basal pia mater (attached to the cerebellar tentoriun), disconnection of the temporal cortex is completed. This incision is the prolongation in cephalo-caudal direction of the occipital lobe disconnection, and passes the insula from behind. (Fig. 5, A and B).

Through a periventricular route the temporal lobe stem section as well as the amigdala suction, are performed. This surgical step is crucial keeping in mind the important thalamic and hypothalamic amigdala connections. (Gloor 591 – 721)

c) Since a previous complete collosotomy has been performed, the temporal lobe remains *in situ*, vascularized, but dysfunctionalized. The only certainty accepted interhemispheric connections are through the corpus callosum and the posterior hippocampal commissure, as recently described by Gloor in 1997[2] (Fig. 6 A and B).

RESULTS

According to the Engel classification, 14 patient are in class I, one in class IV, five have not been classified because of a follow-up of less than one year and one patient operated on in two-step, died 7 days of the second surgery, due to a coagulation disorder secondary to congenital factor-7 deficiency.

COMMENTS AND CONCLUSIONS

As stated above, it is the authors' opinion that this technical variant achieves the same objective as Rasmussen's functional hemispherectomy, reducing surgical time as well as blood loss. This becomes crucial when it is kept in mind that, in most cases, these procedures are indicated in pediatric patients in whom volemias are obviously lower than those of the adolescent and adult population.

We believe that the proposed variant is particularly useful for neurosurgeons starting the practice of Epilepsy Surgery, as a stage prior to the use "more modern techniques". The duration of such "prior" stage will depend essentially on the frequency with which this surgical modality is undertaken.

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RESUMEN

Objetivo e importancia. *Si bien la hemisferectomía de Rasmussen es una técnica de probada eficacia, la reducción en el tiempo quirúrgico y en la pérdida sanguínea es crucial. Esto es logrado por medio de una variante técnica de desconexión en lugar de remoción del lóbulo temporal.*

Presentación clínica. *Se presentan 21 niños operados entre 1991 y 2004 en quienes se efectuó una variante de la hemisferectomía funcional (HF).*

Intervención. *La variante propuesta es una combinación de la hemisferectomía de Rasmussen y la hemisferotomía de Delalande efectuándose una desconexión y aislamiento del lóbulo temporal. Para ello, a nivel central se prolonga la incisión de la desconexión del lóbulo temporal hasta el piso de la fosa media la incisión de la cara medial del lóbulo occipital se extiende hacia el fornix para alcanzar el borde libre del tentorium. La unión del ambas incisiones desconecta el neocórtex del lóbulo temporal.*

Conclusión. *Es opinión del autor que la hemisferectomía de Rasmussen es un procedimiento de probada eficacia. La variante técnica propuesta es particularmente útil para los neurocirujanos en formación en la cirugía de la epilepsia.*

Palabras clave: *hemisferectomía funcional, neurocirujanos en formación, variante técnica*

NOTA: Este trabajo ha sido presentado para su publicación en la revista Child¨s Nevous System con una casuística menor.