

Surgery for drug resistant partial epilepsy in children with focal cortical dysplasia: anatomical–clinical correlations and neurophysiological data in 10 patients

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Objective: To analyse a population of children with focal cortical dysplasia operated on for drug resistant partial epilepsy, with emphasis on clinical features, seizure semiology, interictal and ictal EEG and stereo EEG findings, histological and topographical characteristics of the lesions, extension and localisation of cerebral excision, and its postoperative effect on seizure frequency.

Methods: 10 patients were studied, aged between 26 months and 11 years (median 6 years). Magnetic resonance imaging (MRI) abnormalities were unilobar (temporal 3, frontal 2), bilobar (2), or multilobar (1); the two patients with negative MRI suffered from frontal seizures. Presurgical diagnostic steps varied in complexity and invasiveness depending on the anatomical/electrical/clinical features of each patient. In four patients they included only scalp video EEG monitoring, and in six, also invasive recordings using stereotactically implanted intracerebral electrodes. Surgery consisted of corticectomy plus lesionectomy in all cases.

Results: 70% of the patients were seizure-free after a minimum postoperative follow up of 25 months. These included three patients with temporal lesions and four of seven patients with other lobar or multilobar extratemporal localisation. One patient had improvement in seizure control. Outcome was poor in multilobar patients, but a class Ia outcome was obtained in one case after partial lesionectomy associated with bilobar corticectomy. All patients showed developmental improvement.

Conclusions: Analysis of the data in these patients allowed the production of an "anatomical-clinical concordance" list, which appeared to be correlated with the diagnostic steps performed. Carrying out a stereo EEG exploration in the most complex cases proved useful in defining the epileptogenic zone in extratemporal and multilobar epilepsies. Stereo EEG recordings facilitated a tailored resection of extralesional cortex.

Surgical treatment for drug resistant partial epilepsy may be an early option in childhood because it offers the possibility of achieving a definitive cure, preventing cognitive deterioration, and enhancing the overall quality of life and psychosocial adaptation. In childhood, a malformation of cortical development is found in 14% of cases of drug resistant partial epilepsy and it accounts for 18–26% of all histological diagnoses in paediatric epilepsy series.^{1–3} The epileptogenic role of cortical malformations and the possibility of good seizure control after their complete removal are well documented. Seizure-free outcome is said to be better with the complete excision of the cortical dysplasia than with incomplete excision.^{4–5} A positive outcome is likely if a discrete lesion is found and if it does not involve functionally eloquent cortex.⁴

Magnetic resonance imaging (MRI) techniques have substantially improved the evaluation of epileptic patients in recent years, particularly in children with malformations of cortical development.^{1–7} However, limits in delineating the precise extension of the malformation have been pointed out.^{8–11} As a result it has been suggested that neurophysiological techniques should be employed for a better definition of the epileptogenic zone.¹² In children particularly, chronic subdural electrodes,¹³ mixed techniques,^{3–5} and stereo electroencephalography (EEG)¹⁴ have been employed successfully.

In this study we present a population of children with focal cortical dysplasia operated on for drug resistant partial epilepsy, our aim being to describe the general clinical features, seizure semiology, interictal and ictal EEG,

and stereo EEG peculiarities. These data were correlated with histological and topographical characteristics of the lesions, principally with the extension and localisation of the cerebral excision and its postoperative effect on seizure frequency.

METHODS

We studied 10 consecutive patients under 16 years of age affected by severe drug resistant partial epilepsy and operated on in the Epilepsy Surgery Centre "C Munari" in Milan between May 1996 and September 2000. They were all identified as having a focal cortical malformation on the basis of histological examination.

The general characteristics of the patients are given in table 1. There were five girls and five boys, and their age at the time of surgery ranged from 26 months to 11 years (median 6 years 3 months). Six patients had either a family history of epilepsy or febrile convulsions. Eight patients had several seizures a day; one had daily seizures and one had weekly seizures. Only two had a seizure-free period of any length (three years in one and six months in the other), while two patients had experienced frequent epileptic activity.

Seven patients were right handed, one was left handed, and the remaining two were ambidextrous. In seven patients neurological examination was negative; one (case 8) had congenital hemiparesis and another (case 4) developed mild right hemiparesis with dysarthria after prolonged status epilepticus. A 2 year 7 month old girl (case 6) had choreic

Table 1 Seizure semiology and findings on magnetic resonance imaging

Patients (age at surgery)	Age at seizure onset	Aura	Seizure characteristics	MRI findings
Case 1 (7 y 3 m)	20 m	Sensation in stomach	Psychomotor arrest, staring, R deviation of eye and trunk, facial expression of fear, oro-alimentary automatisms, vegetative signs, rigidity of both arms	L temporal pole atrophy with blurring
Case 2 (4 y)	1 y 10 m	No	Psychomotor arrest, staring, unresponsive, oro-alimentary automatisms. At the end she says "how disgusting" and tries to vomit Eye and head deviation to R, moan, apnoea, R blinking, limb rigidity	Slight cortical thickening with blurring of R temporal pole and T3–T5 gyri
Case 3 (9 y)	8 y 4 m	No	Continuous activity, L blinking, L face tonic, discrete R arm automatisms, frightened	Large white matter abnormality in R temporal pole, and posterior; Ammon horn cyst
Case 4 (3 y 8 m)	10 m	No	Fixed eyes, R face and arm clonic, R arm clonic	Cortico-subcortical signal alteration and cortical thickening in L frontal operculum and insula
Case 5 (6 y 6 m)	1 m	No, but he looks frightened	Epileptic status Psychomotor arrest, unresponsive, mydriasis, face flushing Staring, eye and head deviation to R, arm extension, frightened, apnoea, R arm and leg rigidity	L frontal lesion (polar)
Case 6 (2 y 7 m)	8 m	No	Spasms in clusters (upward eye rotation, flexion of head, and symmetrical abduction of arms)	Cortical thickening of L frontal lobe
Case 7 (2 y 2 m)	6 m	No	Spasms in clusters Psychomotor arrest and fixed gaze	Lesion in L uncus, hippocampus, and parahippocampal area and mesial temporo-occipital gyrus
Case 8 (6 y)	12 d	Probably yes: he looks for a support	Hypotonic drop, laughter Staring, tonic extension of L arm and leg, R arm dystonic with automatisms, R bicycling, oral automatism and flushing	Large and severe R cortical disorganisation in fronto-parieto-temporal lobes Surgical cavity (previously performed intervention)
Case 9 (11 y)	4 y 7 m	Internal tremor, not better specified	Psychomotor arrest, R head turning, R face clonic, responsive Fixed eyes, pallor, tachycardia, both arms and legs extension	Negative
Case 10 (7 y 3 m)	9 m	Cephalic sensation	Bilateral lip corner attraction Staring, abduction of L arm, clonic L arm, eye and sometimes head deviation to R, R arm gestural automatisms Several episodes of epileptic status Possible generalisation	Negative

d, day; L, left; m, month; MRI, magnetic resonance imaging; R, right; y, year.

movements. Two patients had normal psychomotor and cognitive development, while two others experienced cognitive deterioration after the seizure onset; three patients had specific cognitive deficits with a normal cognitive level; three had global psychomotor developmental delay, with a severe cognitive defect in one.

Presurgical evaluation

In all patients a detailed clinical history, a careful description of seizure semiology, and a neurological examination (including a neuropsychological developmental evaluation) were completed, and all previous EEG studies were collected for assessment.

MRI was done following a standardised protocol using a 1.5 T magnet (Philips ACS-NT) including T1 and T2 weighted sequences in the three spatial planes.¹⁵

Long term video EEG monitoring was done under intensive surveillance in nine of the patients (not case 4), allowing the frequent seizures to be recorded in all the cases.

All patients underwent stereotactic and stereoscopic angiography so as to acquire sufficient information for the surgical intervention and for safe intracerebral electrode implantation.

In six patients we undertook a video-stereo EEG investigation using stereotactically introduced intracerebral electrodes implanted under general anaesthesia. For each patient, 10 to

13 electrodes were implanted unilaterally: diameter 0.8 mm; 5 to 15 leads, each 2 mm long with an interelectrode distance of 1.5 mm. Video-stereo EEG monitoring was done under the same conditions as the video EEG. Low and high frequency electrical stimulation was applied to localise eloquent cortical areas (for example, motor, somatosensory, or visual areas) and to reproduce the patient's own seizures.^{16, 17} The stereo EEG study allowed us to determine the origin of ictal discharges and their spread. At the end of each stereo EEG monitoring, three cerebral zones were defined on the basis of the EEG features¹⁸:

- the epileptogenic zone—that is, the cortical region in which ictal discharges arise and initially spread;
- the lesional zone—that is, the regions in which absence of background rhythm, electrical depression, continuous slow waves (mainly delta, more or less monomorphic) are found;
- the irritative zone, in which it is possible to find spikes, spike-waves, and polyspike-waves.

Surgical procedure

Surgical intervention was undertaken on the basis of individual clinical, neuroradiological, and electrical data. The limits of the resection were tailored on the basis of the defined epileptogenic zone, taking into account the limits imposed by the functional anatomy. In the patients

Table 2 Scalp video EEG results and further choices according to the new classification of the patients (see text)

		Neurophysiological presurgical study: scalp EEG		
		Interictal	Ictal	Further step
Case 1	Video EEG: one seizure	Normal background activity L temporal theta diffused to frontal L temporal S-W	L ant and mid temporal rhythmic theta activity, temporal S-W diffused to central area	Surgical intervention
Case 2	Video EEG: two seizures	Normal background activity Monomorphic delta in mid-temporal lead S, poly S, S-W in R ant and mid-temporal lobe	Low voltage fast activity predominant in R f-t, rhythmic slow S at 1 Hz, predominant in R f-t	Surgical intervention
Case 3	Video EEG: one seizure	Normal background activity Spike activity in R f-c-t lobes	Flattening in R temporal, diffusing to f-c, recruiting S-W at 2 Hz in R f-t region	Surgical intervention
Case 4	Video EEG: no seizure	Slow L background activity Slow waves in L c-p-t S-W, poly S-W in L c-p, sometimes R	–	Surgical intervention
Case 5	Video EEG: seven seizures	Slow background activity Delta activity in L f-c S, poly S, poly S-W in L f-c	Irregular delta waves, flattening, low voltage recruiting fast activity in L ant frontal region	Stereo EEG: 12 ic electrodes in R f-t Seizures: 9 spontaneous, 19 with stimulation
CoA (case 6)	Video EEG: three series of spasms	Absence of background activity Bilateral f-c-t delta with diffusion Focal S and S-W on L f-c region with diffusion to R f-c 12 Hz rhythmic activity	Fast diffused flattening with rhythmic fast activity and low voltage fast activity in L frontal region	Stereo EEG: 11 ic electrodes in L f-c Seizures: 12 spontaneous, 4 with stimulation
Case 7	Video EEG: four series of spasms	Slow background activity Continuous delta activity in L t-o S, S-W, poly S-W in L f-c-t	Diffused low voltage fast activity, predominant in L post region	Stereo EEG: 12 ic electrodes in L t-o-p Seizures: 8 spontaneous, 5 with stimulation
Case 8	Video EEG: eight seizures	Slow background activity Continuous delta activity in R hemisphere S, S-W in R hemisphere	Low voltage fast activity in R f-c-t, high voltage slow waves in t-p, then recruiting activity	Stereo EEG: 11 ic electrodes in R c-p Seizures: 15 spontaneous, 4 with stimulation
Case 9	Video EEG: seven seizures	Slow L background activity Multifocal L slow S	Rhythmic slow waves in L f-c, diffuse flattening predominant in L f-c	Stereo EEG: 13 ic electrodes in L f-c Seizures: 3 spontaneous, 10 with stimulation
Case 10	Video EEG: six seizures + one status of minor motor episodes	Slow background activity Slow waves in R frontal diffusing to L ant	Diffused attenuation followed by low voltage fast activity in R f-c-t region, rapid S-W mainly in R ant frontal region	Stereo EEG: 10 ic electrodes in R f-t Seizures: 1 spontaneous + status of minor motor episodes

ant, anterior; c, central; EEG, electroencephalogram; f, frontal; ic, intracerebral; L, left; o, occipital; p, parietal; post, posterior; R, right; S, spike; S-W, spike and waves; t, temporal.

explored by stereo EEG the cortical specimens were analysed in relation to the anatomical location of intracranial electrodes in order to establish the relations among the epileptogenic zone, the anatomical lesion, and the neuropathological data. Lesionectomy was considered complete when neuropathological study revealed no dysplastic tissue at the resection borders.

Postoperative seizure outcome was determined according to the classification proposed by Engel *et al.*¹⁹ Follow up ranged from 25 months to 5 years 10 months (mean 3 years 6 months).

A careful routine neuropathological study with immunocytochemical analysis was made on surgical specimens. On the basis of neuropathological investigation three types of dysplasia were recognised: architectural, cytoarchitectural, and Taylor's type of cortical dysplasia; the particular features of these are detailed elsewhere.²⁰

RESULTS

MRI was positive in eight patients, showing a lesion that involved the frontal lobe in two, the temporal lobe in three, two lobes (temporo-occipital and fronto-insular) in two, and three lobes (fronto-parieto-temporal) in one.

The correlative study between seizure characteristics and MRI findings (table 1) allowed us to draw up a list of the patients ordered by degree of anatomical-clinical concordance (from maximum to minimum): maximum concordance was

found with the presence of seizures characterised by epigastric sensation, followed by the development of a temporal seizure and evidence of a left temporal pole lesion (case 1); minimum concordance was established for polymorphic seizures, probably of frontal origin, with a negative MRI (case 10).

Long term scalp video EEG

Interictal and ictal scalp EEG features are detailed in table 2. The background activity was abnormal in seven patients. Interictal abnormalities were localised in two cases, frequently multilobar in four patients, and predominantly on the affected hemisphere with frequent contralateral involvement in four. Ictal phenomena were recorded in nine patients with a mean of four seizures per patient (range one to eight). Initial ictal discharges were well localised in four patients, while a diffuse ictal pattern predominant in a focal area was found in four; one patient (case 5) showed extended delta activity followed by a focalised pattern. The well localised ictal discharges obtained in the three patients (cases 1, 2, and 3) who had concordance of electroclinical and MRI evidence allowed surgical intervention after these diagnostic steps had been taken.

Case 4 was submitted to surgical intervention without the long term video recording of seizures because of the patient's young age and severe and rapid cognitive deterioration (mainly in language tasks), and the impossibility of complete

Table 3 Seizure outcome after surgical intervention tailored on the basis of scalp and stereo EEG explorations

Case	Stereo EEG		Surgery		Follow up	Engel class
	Interictal	Ictal (EZ)	Involved structures	Type		
Case 1			L temporal (pole, amygdala, hippocampus, T1 → T4)	Corticectomy + complete lesionectomy	2 y 11 m	Ia
Case 2			R temporal (pole; ant T1; ant-mid T2, T3, T4; mesial)	Corticectomy + complete lesionectomy	3 y 1 m	Ia
Case 3			R temporal (polar, basal and lateral; ant hippocampus)	Corticectomy + complete lesionectomy	5 y 4 m	Ia
Case 4			L triangular and opercular part of F3	Corticectomy + incomplete lesionectomy	2 y 11 m	IV
Case 5	Lesional zone: orbital and dorsal F2/F3 Irritative zone: orbital, F2, F3, ant and medium cingulate gyrus	Widespread frontal, mesial and lateral	L frontal (polar, orbital and dorsal area)	Corticectomy + complete lesionectomy	3 y 8 m	IV
Case 6	Lesional zone: gyrus rectus, int and lat F1, mid ant F2 Irritative zone: int and mid F2, orbital gyrus	Global explored zone except: precentral gyrus, posterior F1 and mesial frontal cortex	L frontal (lateral and mesial polar, basal and dorsal F1, F2, F3)	Corticectomy + complete lesionectomy	2 y 1 m	Ia
Case 7	Lesional zone: T2, T3, T4, mid and inf occipital gyrus Irritative zone: T1, T2, mid T4, fusiform gyrus, hippocampus	Lesion + cuneus and fusiform gyrus, T3, T4, posterior hippocampus	R temporo-occipital (mesial, basal and lateral)	Corticectomy + incomplete lesionectomy	3 y 7 m	Ia
Case 8	Lesional zone: widespread Irritative zone: widespread	Widespread	R fronto-parieto-temporal	Second intervention: incomplete lesionectomy	5 y 10 m	IId
Case 9	Lesional zone: orbital, dorsal F2, mesial F1 Irritative zone: F2-F3 sulcus, dorsal F2, precentral gyrus	F2-F3 sulcus, F2, anterior cingulate gyrus	L frontal (lateral)	Corticectomy	2 y 4 m	Ia
Case 10	Lesional and irritative zones: basal frontal cortex	R orbital and gyrus rectus minor motor status	R frontal (polar, basal, mesial and lateral)	Corticectomy	4 y 8 m	Ic

ant, anterior; EZ, epileptogenic zone; inf, inferior; int, internal; L, left; lat, lateral; R, right.

resection of the lesion owing to anatomical limitations (insula). The other six patients underwent a stereo EEG exploration to define the epileptic zone (table 2).

Stereo EEG exploration

On average, 11 intracerebral electrodes per patient (range 10 to 13) were implanted under stereotactic conditions. Chronic video-stereo EEG monitoring allowed us to record at least one spontaneous seizure per patient (range 1 to 15, mean 8). Moreover, several seizures were induced by electrical stimulation in five patients. All stereo EEG features are detailed in table 3.

In the four patients with an MRI evident lesion, the stereo EEG defined lesional zone roughly coincided with neuroimaging data, while in general the irritative zone revealed a larger cerebral involvement.

Of the remaining two with negative MRI, in one case (case 8), the lesional and irritative zones were coincident and located in the basal frontal cortex; in the other (case 9), the presence of a specific poly-spike activity cluster in a limited region (fig 1C) allowed us to make a diagnosis of Taylor's type cortical dysplasia, subsequently confirmed by histological diagnosis.²⁰

The epileptic zone was greater than the neuroradiological lesion in two cases and was widespread in case 8, in which the MRI showed an extended right hemispheric lesion. The stereo EEG recordings identified the epileptogenic zone in two patients without a neuroradiological lesion.

Surgery

The surgical intervention (table 3) consisted of a corticectomy in two patients with frontal lobe seizures without an MRI lesion, and a lesionectomy plus a tailored corticectomy in the others.

In these latter, the lesionectomy was complete in five patients and incomplete in three (cases 4, 7, and 8). No patient had complications following surgery. One patient (case 7), in whom the resection included all the occipital lobe and the occipito-temporal and occipito-parietal junctions, had a homonymous hemianopia; however, her behaviour shows no difference from before the intervention.

Seizure outcome

After a mean (SD) follow up of 43 (15) months, seven patients (70% of cases) are seizure-free (six in class Ia and one in class Ic). The age of seizure onset, the duration of the epilepsy, and the seizure frequency before the intervention did not show any relation to postoperative seizure-free outcome. Among the patients in whom the seizures persisted after surgery, one patient (case 8) showed noticeable improvement, with one seizure per night. In the other two (cases 4 and 5) seizures are unchanged. Case 4, despite the poor outcome, showed a dramatic improvement in speech function. Postoperative seizure-free outcome was obtained in the two patients in whom the MRI was negative and in five of the eight cases with MRI lesions, including case 7, in whom two lobes were involved.

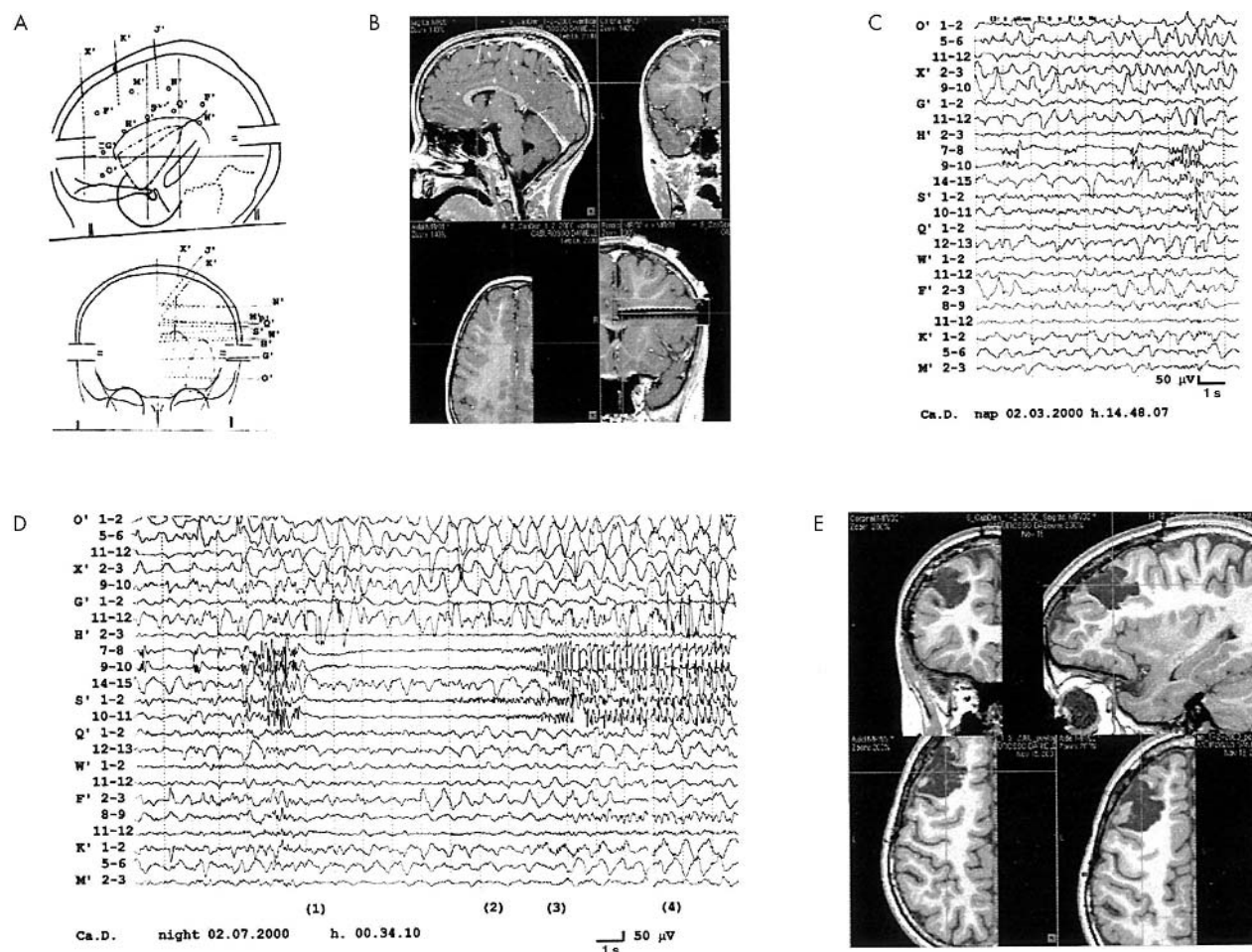


Figure 1 Case 9: stereotactic scheme (A); magnetic resonance images (MRI) with the trajectory of electrode H' (B); interictal trace (C); spontaneous seizure (D); and postoperative MRI (E). (A) The stereotactic scheme shows, in lateral and coronal views, the location of the 13 intracerebral electrodes, all implanted in the left fronto-central region except for two in the parietal lobe. (B) T1 weighted MRI sequences showing the trajectory of the electrode H': the slice at the bottom shows the real trajectory acquired with the implanted electrodes, while the upper slice makes it possible to appreciate the structures explored as it shows the trajectory without the artefacts caused by the metallic materials. In particular, the mesial leads of this electrode explore the frontal cingulate gyrus, and the lateral leads the lateral cortex of F3; the white square on the trajectory of the electrode identifies the leads from 7 to 10, where the activity of the F2-F3 sulcus is recorded. (C) Nap trace with complete montage in which it is possible to see three burst of ingradating fast activity well localised in the intermediate leads of electrode H'; after slowing, the last burst involves the external leads of two adjacent electrodes S' and F', both exploring the frontal convexity. The particular interictal pattern identified on the F2-F3 sulcus allowed us to hypothesise the presence of focal cortical dysplasia, which was confirmed by histological examination. (D) Spontaneous seizure recorded during nocturnal sleep. Clinically, the patient starts groaning at (1) orienting the head to the left; (2) still groaning, he bends moving the left arm apparently trying to grasp the father; (3) the patients looks at the observer without answering his questions; (4) dystonic posturing of the right arm. The seizure will end almost 20 seconds later. Electrically, three seconds before the clinical onset one of the typical burst of fast activity begins in the intermediate leads of H', followed by a spike activity of greater amplitude which involves also the external leads of S' and F' and the mesial leads of H' and S'; a few milliseconds before the clinical onset the ictal pattern is characterised by low voltage fast discharge involving all the leads mentioned; this discharge ingrades until (3) after which a rhythmic S-W discharge continues until the end of the fit. (E) Postoperative MRI: the image shows the limited surgical intervention which consisted of excision of the medial and posterior part of F2 and the F2-F3 sulcus.

All the patients operated on for temporal epilepsy had a good outcome and are in class Ia. Seizure outcome has been favourable in three of four patients with frontal epilepsy and in one of the three patients with multilobar involvement. The three with temporal epilepsy and lesions, who were operated on without stereo EEG, became seizure-free. Four of the six patients in whom intracerebral electrodes were implanted had a good outcome, including case 10, who had two seizures in the first year and has now been seizure-free for 4.8 years.

Four of the five patients submitted to a complete lesionectomy are seizure-free. Three patients had an incomplete lesionectomy: in cases 4 and 8 the limits were represented by the anatomical area involved: the insula in case 4 and the central region in case 8; in case 7, incomplete lesion resection was undertaken on the basis of stereo EEG

recording which showed that a part of the lesion was not epileptogenic. Among these three patients, only the last is seizure-free.

Cognitive development in all the children improved globally, even in those patients with persistence of seizures. We noticed in particular better attention than before the intervention, a richer vocabulary, and more fluent language.

DISCUSSION

This study involved a small number of children with drug resistant partial epilepsy, associated with histologically proven focal cortical dysplasia. The number of patients is not substantially different from that in other published studies of children with focal cortical dysplasia,² in surveys including both adults and children with focal cortical

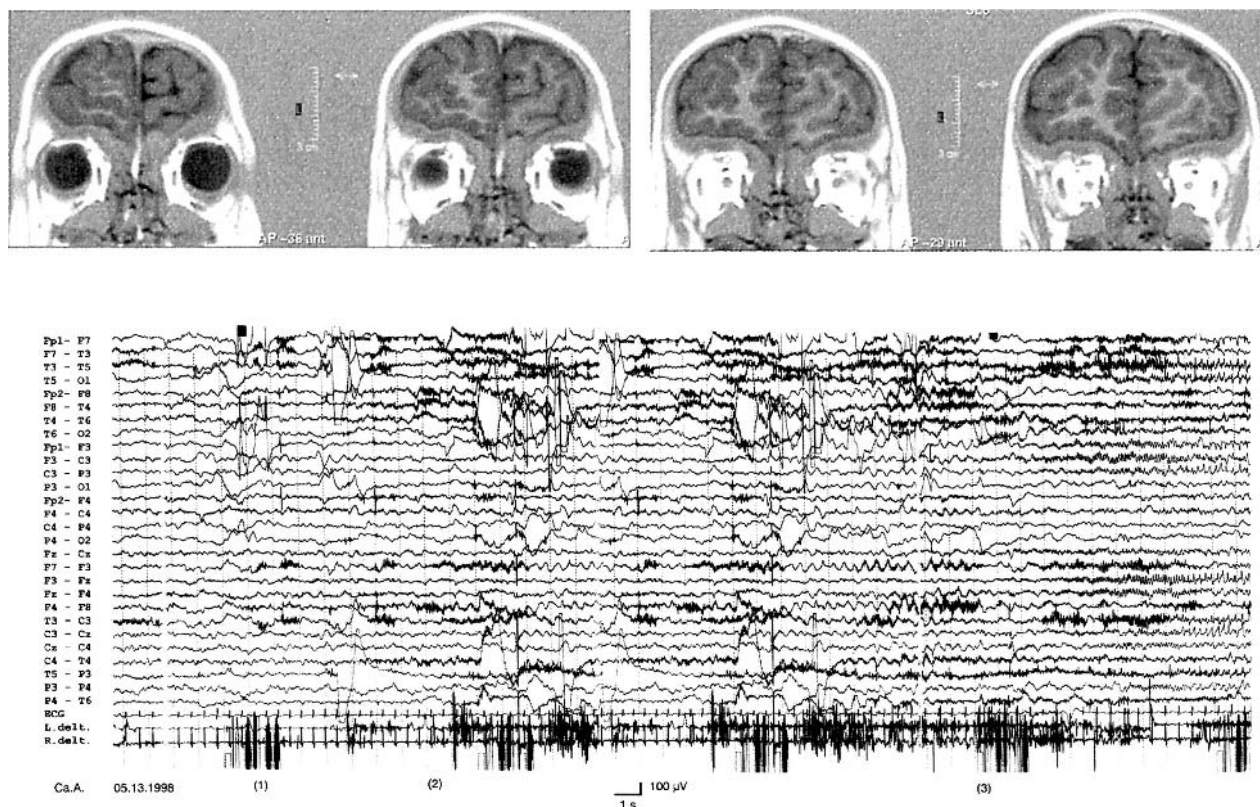


Figure 2 Case 5: presurgical magnetic resonance imaging (MRI) and a spontaneous seizure recorded during sleep on video EEG monitoring. Top: MRI (coronal views in a T1 weighted sequence) showing an abnormality of gyration in the left frontal pole. Clinically the seizure arises one minute after spontaneous awakening: the patient moans, presenting a little jerk of the left arm and extending both limbs at (1); at (2) a slow head and eyes deviation to the right is present together with asymmetrical limb flexion, and at (3) he looks like frightened; almost 15 seconds later the seizure stops without speech disturbance. Bottom: The EEG shows a localised flattening in the left fronto-central leads followed by a rhythmic recruiting delta activity in both fronto-temporal regions; 12 seconds later a fast recruiting activity at 18 Hz characterises the trace of the left fronto-precentral regions; this activity is also evident in posterior temporal and central regions.

dysplasia,^{5 14 21} or in other studies on resective surgery in children with mixed types of lesion, including focal cortical dysplasia.^{3 22 23}

Though published reports do not always give a detailed description of the localisation and extension of cortical dysplasia in children, the prominence of parietal and central localisation is described in studies by Chassoux *et al*,¹⁴ and of multilobar lesions in studies by Hong *et al*.²¹ Other studies, on the other hand, have shown a frontal predominance^{2 24 25} or a heterogeneous distribution.⁵ In our survey, unilobar focal cortical dysplasias are located most often in the frontal lobe (four cases, including the two patients in whom MRI was negative), but the temporal lobe was involved in three cases, thus suggesting that this location could be relatively common in children. In fact, cases with temporal lobe involvement are reported more often in adults.^{9 26} One possible explanation for this is the lesser impact on children's psychosocial development of temporal lobe epilepsy compared with extratemporal forms, probably linked to a lower frequency of seizures and milder seizure semiology.

As the validation of presurgical strategies and the surgery itself is strongly dependent on the seizure outcome following surgery, in this study we have presented only patients in whom an operation had already been done and whose postoperative follow up had been of substantial duration (at least two years). The overall result in our population is 70% freedom from seizures. This is not very different from that of Chassoux's study, in which (basing the delimitation of epileptogenic zone on stereo EEG recordings) a seizure-free

outcome was achieved in 64% of adults and children combined. In other series of children operated on for focal cortical dysplasia the percentage of totally cured patients varies from the 30% of the 20 patients of Polkey,² to the 57% of 14 patients of Edwards,⁵ 52% of Wyllie's cases,³ and 41.6% of Hong's.²¹

As in another series of patients with focal cortical dysplasia,⁵ the general characteristics of onset, duration, and frequency of seizures did not seem to influence the surgical outcome in our group of patients. This is the reason why we attempted to identify and analyse other variables that might play a role in postoperative outcome. In our practice we usually have at our disposal at the time of the patient's first clinical evaluation at least two main types of data: detailed ictal semiology and MRI features. For this reason we decided to organise the study in relation to these two variables.

We therefore ordered the patients in an "anatomical-clinical concordance" list, which strongly influenced the subsequent diagnostic steps and seemed to be related to the surgical outcome. For example, in the three patients with temporal lobe seizures the temporal focal cortical dysplasia was considered the main pathogenic factor; video EEG recordings corroborated this, showing interictal and ictal activities that were well localised over the affected temporal lobe. These three patients are in class Ia, in agreement with many reports of a better seizure-free outcome in patients with temporal lesions than with extratemporal lesions (where surgery was done without an initial invasive study).^{3 5 27-29}

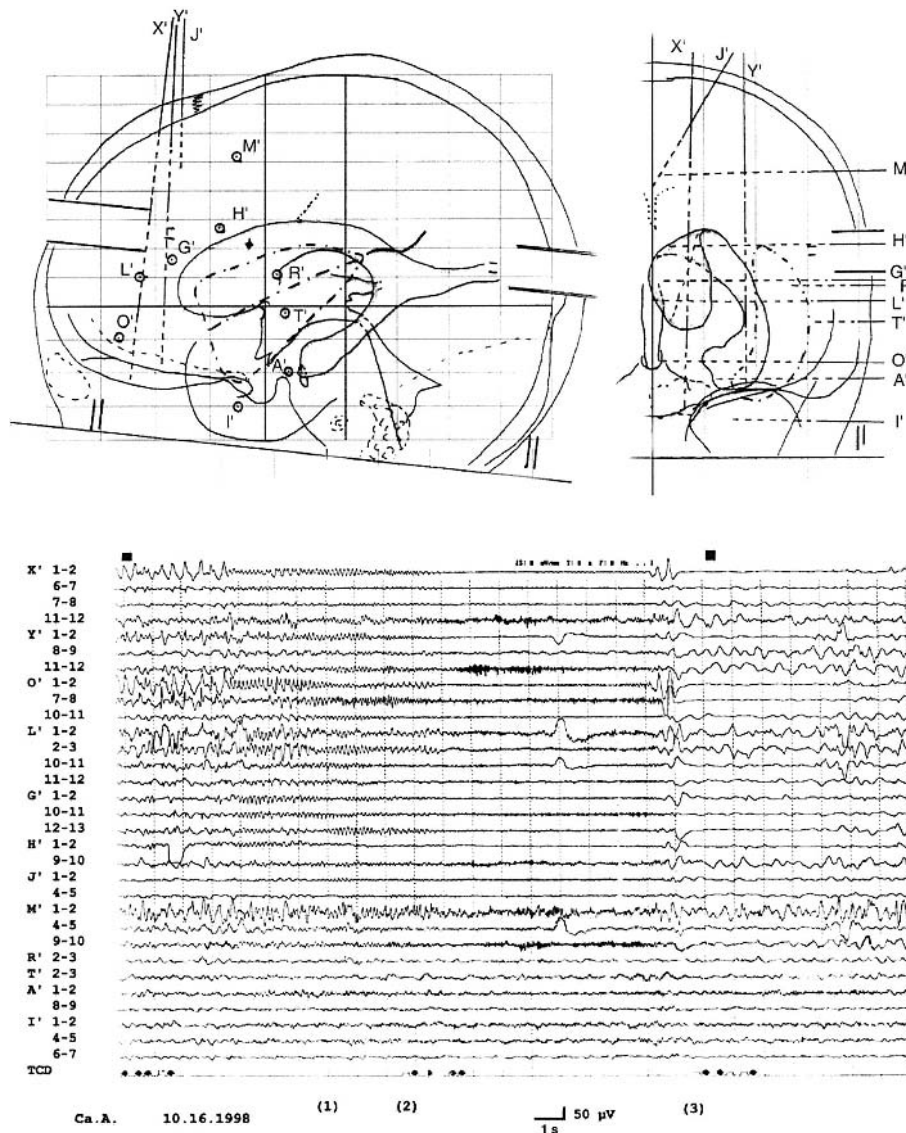


Figure 3 Case 5: stereotactic scheme and spontaneous seizure recorded with depth electrodes. Lateral (upper left) and frontal (upper right) views of the stereotactic scheme show the location of the 12 intracerebral electrodes. Clinically at (1) the patient glances to the left, at (2) he speaks with his mother but, immediately after, he bends his head low and right with staring, (3) he begins to play. Three seconds before (1), the EEG shows the abrupt beginning of a spindle activity at 6 Hz involving (mainly in the mesial leads) an extensive frontal-basal and anterior region; at (1) a rhythmic sharp activity at 12–14 Hz is evident in the orbital gyrus (O'7-8); at (2) a low voltage fast activity begins in the same region and rapidly involves almost all the frontal lobe and the temporal pole, showing a greater tonicity in the orbital area (X'1-2 and 6-7, Y'1-2, O'1-2 and 7-8) and external F2 (L'11-12, probably within the lesion). This activity ends after 7 s, leaving postictal depression in the orbital region and postictal delta activity in the other leads.

On the other hand, a poor degree of anatomical-clinical concordance forced us to consider exploration with intracerebral electrodes in patients with extratemporal epilepsy associated with a lesion on MRI. In these cases, interictal scalp EEG features were not well localised and ictal video EEG monitoring showed at least a bilobar ictal onset discharge. These features precluded a precise definition of the cortical resection but allowed a more detailed implantation strategy. Particularly in the two patients with negative MRI, ictal video EEG recordings represented a very useful source of information for an accurate planning of the stereo EEG exploration.

There is evidence for the value of invasive recording to obtain a better delineation of the epileptogenic zone in cases where extratemporal epilepsy is suspected, with or particularly without positive MRI findings. Some investigators have suggested the use of acute electrocorticography to record and evaluate interictal epileptiform discharges as a

measure of the limits of the resection.^{8,21} Other studies have used chronic invasive exploration (including subdural strips, subdural grids, and depth electrodes) to provide more information about the parts of the cortex functionally linked to the dysplasia. The patients in the studies by Edwards and Wyllie were submitted to an operation after ictal EEG surface recording and invasive study (subdural and epidural grids) in the majority of cases. A few of Edwards' cases³ and 20% of Polkey's² were studied with chronic intracerebral electrode implantation; in Hong's study, chronic subdural electrodes were used.²¹ Only one study described the results obtained after stereo EEG recording.¹⁴

In our group of patients the extent of cortical excision was defined mainly on the basis of ictal clinical and electrographic data recorded during the chronic stereo EEG procedure—always considering the localising indications provided by the MRI study. As a general statement, in our cases intralesional electrodes were often the sites of the ictal

onset discharges, but ictal patterns might vary from one seizure to another, and very often lesional and non-lesional electrodes were involved simultaneously, as described in a previous study in non-paediatric patients.³⁰ Tailoring surgical excision on this basis led to a seizure-free outcome of four of the six patients explored.

Stereo EEG recordings played an important role in clarifying the relations between the dysplastic area and the apparently normal cortex in the organisation of the epileptogenic network. Although recent reports on histological specimens prove that cortical malformation is intrinsically epileptic,^{31, 32} some clinical and EEG findings^{2, 8-11} suggest that the cortical region in which ictal discharges arise and primitively spread is not strictly coincident with the dysplasia, or that it is larger than the lesion.

We also found that the onset of spontaneous and induced seizures involves electrodes inserted outside the dysplastic lesion; these data were confirmed by histological examination, in which a part of the surgical specimen was histologically normal. Our cases confirm on the one hand that the epileptogenic zone may involve histologically proven normal tissue, and on the other, that incomplete lesion resection with preservation of part of the malformed tissue (proven to be non-epileptogenic) could give a good outcome. An example of this possibility is given by our case 7, cured after incomplete lesionectomy of a bilobar focal cortical dysplasia, and also by four of 10 patients in Chassoux's study who were seizure-free despite an incomplete lesionectomy. Dysplastic cortex can, in fact, participate in the organisation of function along with normal cortex,³³ which is supported by recent data acquired using functional MRI,³⁴ and defining the actual extension of the epileptogenic area by ictal invasive recordings can help to spare part of the dysplastic cortex. Nevertheless, the possibility of curing patients with an incomplete lesionectomy remains the exception rather than the rule, and we agree with those investigators who maintain that a complete or major excision of both the MRI visible lesion and the cortical areas displaying ictal activity leads to the best results.^{4, 5, 14}

Concerning the localisation of the epileptogenic zone, we emphasise the role of stereo EEG in rendering seizure-free three of four patients with frontal lobe epilepsy, among whom there were two with negative MRI. Considering that surgical results in frontal lobe epilepsy are less favourable than in temporal lobe epilepsy, and that surgical series have reported a seizure-free outcome in about 60–65% of cases,³⁵⁻³⁷ our 75% of cured patients with a frontal excision should be regarded as an encouraging result. This finding could suggest that there may be a correlation between the presence of focal cortical dysplasia and a good outcome; in fact the three cited series of patients with frontal lobe epilepsy are composed of subjects with various types of lesion and also of patients without any lesion. However, the possibility of exploring cerebral areas infolded in a sulcus when using intracerebral electrodes (fig 1) could have contributed to a better delineation of the epileptogenic zone. Our case 5 shows the difficulty in finding a technique that can guarantee a completely successful surgical outcome, despite good location of the epileptogenic lesion and corticectomy and lesionectomy (figs 2 and 3). According to Sisodiya¹² and Jayakar *et al*,¹³ some cases are not seizure-free despite complete resection, probably because of the presence in these cases of more than one epileptogenic focus, which may become evident after the excision of the first focus. As the great majority of patients show cognitive developmental delay—resulting from the lesion itself, the repetitive seizures, and the need for powerful drugs—the aim of surgery in these children should not only be relief from severe epilepsy but also improvement in developmental progress.³⁸ We believe our data can help in

confirming this possibility, considering that our treated patients showed clinically evident psychosocial and cognitive improvement.

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NEUROLOGICAL PICTURE

Knitting artifact

Artifacts are recorded signals that are non-cerebral in origin¹ and may be either physiological, due to body activities, or non-physiological, which arise from either external electrical interference or internal electrical malfunctioning of the recording system.

We describe here the first known case of artifact generation resulting from a patient knitting during a video EEG recording. In the figure, apparent spikes or sharply contoured slow waves are seen phase reversing at T3 and T4. On video, these changes are seen to coincide with the patient transferring a stitch

from one knitting needle to the other. These sharp transients were also seen on separate occasions at Cz, P3, T4, and other derivations.

No eye blinks or gross movements of the body by the patient were seen during these changes. One hypothesis might be that the wool being knitted by the patient causes a build up of static electricity either on the knitting needles or on the wool itself. During the transfer of the stitch from one needle to the other, the tapping of the needles together may cause the release of this static electricity and allow it to be recorded on the EEG.

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EEG showing knitting artifact, representing as spikes with a maximum at T3 and T5. AC27 and AC28 represent the left and right sphenoidal electrodes, respectively. AC23 and AC24 are the left upper canthus and right lower canthus electrodes, respectively, for EOG recording. The AC25–AC26 derivation is ECG recording.