Complex partial seizures of childhood onset: a clinical and encephalographic study¹

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Electroclinical manifestations of 30 patients with complex partial seizures (CPS) of childhood onset (<16 yr) were evaluated. Only patients with both clinical CPS and focal specific epileptiform activity shown on the electroencephalogram were selected. Febrile convulsions were present in 9 patients, suggesting that they play a significant role in the pathogenesis of CPS. All patients had a period of unresponsiveness with a blank stare and amnesia; 21 had an aura, including affective symptoms in 20. Hallucinations (5) and illusions (1) were infrequent. Only 16 had epileptiform discharges in their initial waking record, whereas in 8 others, epileptiform discharges were activated during sleep. Foci were predominantly unilateral (28) and mostly temporal (26); only 2 were bi-temporal, in contrast to the reported incidence of 31%-42% in adults, suggesting that the natural evolution of unilateral foci is to become bilateral.

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There are few published studies on the electroclinical manifestations of complex partial seizures (CPS) of childhood onset. Most cases were selected primarily on the basis of the clinical diagnosis alone,¹⁻⁶ which introduces a certain bias into the analysis of the data. Thus patients with episodes simulating CPS but associated with generalized epileptiform activity, as well as those with nonepileptic "spells" associated with a normal electroencephalogram (EEG), may have been included in previous publications as well. Only three studies that we know of were based on both clinical and EEG criteria.⁷⁻⁹ We have therefore evaluated 30 patients with definite CPS who had both (*a*) clinical symptoms

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Table 1. Risk Factors*

	Febrile Convulsions	No Febrile Convulsions
Nonfebrile seizures	0	11†
Structural lesion	0	5
Neonatal illness	0	4
Trauma	0	1
Central nervous system infection	0	1
Family history	0	0
No factor	0	6

* In 9 patients with febrile convulsions and 21 patients without febrile convulsions

† Generalized in 7 and focal motor in 4

consistent with CPS starting before the age of 16 years and (b) focal specific epileptiform activity on EEG.

Materials and Methods

A total of 52 patients who had been followed up by the Pediatric Neurology Service of The Cleveland Clinic Foundation with a clinical diagnosis of CPS beginning before 16 years of age was considered for this study. All were interviewed and a history was obtained from both the patient and a parent. CPS was diagnosed according to the symptomatology set out in the classification proposed by the International League Against Epilepsy in 1981.10 The first EEG performed at our institution and at least one subsequent EEG from each patient were evaluated. In each case, electrodes were placed according to the International 10-20 System, and focal specific epileptiform activity was defined as spikes and sharp waves as set forth by the Committee on Terminology at the Eighth International Congress of Electroencephalography and Clinical Neurophysiology.¹¹ Only patients with focal sharp transients which two electroencephalographers (D.S.D., H.L.) agreed were clearly defined epileptogenic activity were included. In order to ensure that all subjects had definite CPS, patients with poorly defined sharp transients of doubtful epileptogenic significance, nonepileptogenic focal sharp transients in the temporal region (small sharp spikes, psychomotor variants, 14- and 6-Hz positive spikes, and/or wicket spikes), multifocal independent epileptiform discharges (including temporal involvement), and/or benign focal epileptiform discharges of childhood were excluded. We believe that the latter patients form a separate specific group with its own particular electroclinical manifestations and prognosis.¹²

Of the 52 patients tested, only 30 (17 males and 13 females) fulfilled our stringent criteria, having both clinical CPS and focal specific epileptiform activity on EEG. At the time of the study, these patients ranged in age from 6 years to 24 years and 6 months (average, 14 yrs and 11 mo). The age of onset of CPS was predominantly bimodal (birth to 5 yrs in 12 and 10 to 16 yrs in 13).

Results

Risk Factors (Table 1). Twenty patients had had a seizure (with or without fever) prior to the onset of CPS, whereas 9 had febrile convulsions (FC). Structural lesions were present in 5 patients. One child had a cyst in the right temporal lobe, diagnosed by computed tomography (CT) and manifested clinically by cranial asymmetry; the right temporal convexity was more prominent, though there was no focal neurologic abnormality. Two patients had an atrophic lesion in the left cerebral hemisphere as shown on the CT scan, manifested as right hemiatrophy in one and right hemiparesis with dysphasia in the other. Another child, who had tuberous sclerosis and macrocephaly, demonstrated calcified lesions in the left parietal lobe, right frontal lobe, and the periventricular region. The last patient had a small lesion in the right frontal lobe which was enhanced by contrast material and visualized on the CT scan. The carotid angiogram was normal, and there were no focal neurologic findings. The lesion was thought to be static.

Neonatal illness, defined as significant illness in the neonatal period which necessitated hospitalization, was present in 4 patients: this included respiratory distress with cardiac arrest and seizures in the first three days of life, neonatal seizures, aspiration with an episode of hypoglycemia, and severe jaundice requiring exchange transfusion. Two patients had suffered trauma, defined as an injury resulting in a concussion or skull fracture. Central nervous system infection occurred in one patient who had had measles encephalitis at the age of four years.

No patient had a parent or sibling with a history of seizures, and only 6 had no history of a potential risk factor.

In order to study the significance of FC as a risk factor, the relative frequency of all risk factors was evaluated in the 9 patients with FC and in the 21 patients without them. None of the patients in the FC group had a second risk factor.

In the group without FC, there were 11 patients with an afebrile seizure preceding CPS, 5 with a structural lesion, 4 with significant neonatal illness, and 1 with measles encephalitis. Only 6 had no risk factor.

Type of first seizure. Ten patients had a CPS as the first seizure; but most had either an FC (9), an afebrile generalized seizure (7), or an afebrile focal motor seizure (4) prior to CPS.

Clinical Manifestations (Table 2). All 30 patients had a blank stare, a period of unresponsiveness, and amnesia. Approximately two thirds had an aura and automatisms. Postictally, 19 were lethargic or slept and 7 were confused. Affective or autonomic symptoms were frequently part of the aura, occurring in 20 of 21 patients. These were usually manifested as a nonspecific "strange" feeling, abdominal sensation (usually in the epigastrium), or lightheadedness; hallucinations, illusions, and cognitive symptoms were infrequent but *never* represented the sole clinical expression of CPS.

Frequency of CPS. At the time of the study, 16 patients had one or more seizures per week; only 4 had less than one per year.

Psychiatric Problems. Behavioral problems were reported by parents of 8 patients, and included hyperactive behavior, temper tantrums, and disciplinary problems at home and in school. Mental retardation was mild in 4 and moderate in 2. Four children had received special education, and 2 had been diagnosed as having a psychotic illness.

Handedness. Eight patients were left handed and 22 were right handed.

EEG findings. Only 16 patients had specific focal epileptiform activity (spikes or sharp waves) on the initial EEG while awake, while 8 others had epileptiform discharges only when asleep, for a total of 24. Of the first group of 16 patients,

Table 2. Clinical manifestations of CPS

	No. of Cases
Blank stare	30
Unresponsive	30
Amnesia	30
Aura	21
Affective or autonomic	20
Hallucination	5
Illusion	1
Cognitive	1
Automatisms	20
Postictal lethargy/sleep	19
Postictal confusion	7

14 showed an increase in epileptiform activity with sleep. Foci of activity were temporal in 28 cases, with unilateral foci in 26 cases (13 in the left hemisphere and 13 in the right), and bilateral foci in 2. Extratemporal foci occurred in 2 patients, each of whom had a unilateral frontal focus. The incidence of left- and right-sided foci was approximately equal among right- and lefthanded patients (*Table 3*).

Discussion

Gastaut and Broughton¹³ indicated that CPS "consist of such complex and essentially psychic symptoms as mental confusion, behavioral automatisms, illusions, hallucinations, changes in affect and memory or ideational disturbances." Daly¹⁴ points out that all of these symptoms represent "alterations in the content of consciousness which, as the seizure evolves, may lead to an alteration in the level of consciousness manifested by unresponsiveness, confusion, and amnesia." Over the past 80 years, various authors have included patients with CPS when discussing uncinate epileptic fits,¹⁵ psychomotor epilepsy,¹⁶ temporal epilepsy,^{17,18} and limbic epilepsy.³ Although these reports generally refer to patients with CPS, other patients may also be included in these categories. In this paper, we use the term CPS to encompass those seizures listed above.

In most previous studies of CPS of childhood onset, patients were primarily selected based on a clinical diagnosis alone. Thus in the early studies of Glaser and Golub¹ and Glaser and Dixon,² a significant number of patients (34/110) had generalized epileptiform discharges. Even in a later study,³ a significant number of patients with generalized discharges on EEG were included. This group obviously included patients with generalized absence seizures (petit mal), introducing a significant bias into the analysis of data. Although Ounsted et al⁸ selected patients for their study of childhood-onset CPS on the basis of EEG and clinical criteria, there is some question regarding the appropriate diagnosis of some pa-

 Table 3. Relationship of favored hand and side of epileptogenic focus

	Right Handed	Left Handed	Total
Left-sided focus	9	4	13
Right-sided focus	11	4	15
Bilateral foci	2	0	2
Totaļ	22	8	30

tients in the group. Discussing the promising long-term outcome in approximately one third of Ounsted's patients, who were seizure-free and not on medication, Lindsay et al¹⁹ offered the explanation that this group may have included patients with benign focal epileptiform discharges of childhood, as they were not specifically excluded. Aird and Crowther⁹ and Bray⁷ based their patient selection on clinical criteria and focal EEG abnormalities; however, neither specified whether patients with benign focal epileptiform discharges or other non-epileptogenic sharp transients were excluded. In selecting our material, we took care to exclude such cases, including small sharp spikes, 14- and 6-Hz positive spikes, psychomotor variant, and wicket spikes.

The bimodal age of onset of CPS in our patients is similar to that encountered by Aird and Crowther,⁹ whose subjects were also younger than 16; they did not have a satisfactory explanation for this phenomenon. We evaluated all clinical and EEG characteristics in the two extreme groups (birth to 5 yrs and 10 to 16 yrs) and also found no differentiating features.

The high proportion of FC in our series (9/30)differs significantly from the reported incidence of 1.9%-3.6% in the normal population.²⁰ This either supports the hypothesis that FC are important in the pathogenesis of CPS²¹ or indicates that patients with epileptogenic foci involving the temporal lobe have a marked tendency toward FC at a younger age. The fact that FC always preceded CPS might favor the first explanation, while the absence of any other predisposing factor in all 9 patients with FC also suggests that they constitute a pathogenetic factor.

A family history of seizures was found in 60% of cases of CPS by Bray,⁷ and Ounsted et al⁸ found a prevalence of seizure disorders in the siblings of 15% of his probands. They suggested that a child can be genetically predisposed toward development of FC, which if prolonged would result in sclerosis of Ammon's horn and formation of an epileptogenic focus. Our study does not support a genetic predisposition to FC, as none of our patients had a first-degree relative with a history of seizures. In addition to the 9 patients with FC predating CPS, a significant number of patients had had prior afebrile seizures, which were generalized in 7 and focal in 4: thus we speculate that an afebrile seizure similar to an FC may act as an independent predisposing factor for CPS, though the possibility that seizures may be triggered by the same epileptogenic focus that eventually causes CPS should also be considered. Five patients had a structural lesion as shown on CT, underlining the importance of investigating any focal seizure disorder. Clinical findings may have suggested that the CT scan would be abnormal in 4 patients, namely, 2 with focal neurologic deficits, 1 with cranial asymmetry, and 1 with tuberous sclerosis. Therefore, 1 patient had a structural lesion demonstrated on CT without accompanying clinical signs.

All 30 of our patients had CPS characterized by unresponsiveness, a blank stare, and amnesia. Not all seizures were manifested as unresponsiveness; occasionally, only an aura was present, and hallucinations, illusions, and cognitive symptoms were infrequent, suggesting that if the patient and/or relatives describe symptoms compatible with an aura with no alteration of consciousness, the diagnosis of CPS should be doubted. Most of our patients (16/30) had one or more seizures per week. Only 4 were well controlled on medication, which reduced seizures to one or less per year, suggesting that CPS is generally difficult to control. However, our population may be biased in that our patients were referred because their seizures were difficult to control. Ten patients who had CPS clinically but normal EEGs were excluded from our study. Six of these patients had infrequent seizures. This suggests that some CPS may be easily controlled, and in those patients, the epileptiform activity is totally suppressed by anticonvulsant medication. In support of this, we offer a patient who had CPS clinically, but had had three normal EEGs since 1974. During her fourth EEG, performed after partial drug withdrawal, three episodes of CPS (clinical and EEG) originating from the left temporal region were recorded. Her interictal record demonstrated frequent sharp waves from the left mesial temporal region.

Lindsay et al²² commented that overt psychosis developed in 9 of their 87 patients, as also occurred in two out of 30 cases in our series. Seven of Lindsay's patients had a left-sided focus and 2 had bilateral foci; in our series, 1 patient had a left-sided focus and 1 had bi-temporal foci. No psychotic patients had only right-sided foci in either study. While we expected that the 8 lefthanded patients would have a high incidence of left-sided foci, this was not supported by our results.

Sleep is known to be an extremely good activator of epileptiform activity on an EEG.²³ It proved to be very useful in our patients, 8 of

whom experienced epileptiform activity only during sleep. Our incidence of extratemporal epileptogenic foci is similar to the 8% reported by Gastaut.²⁴ However, it is significant that only 2 patients had bi-temporal discharges, in contrast to the 31%-41% reported previously in adults.^{17,25,26} This suggests two possibilities: either the natural evolution of unilateral foci is to become bilateral or else CPS has a different pathogenic mechanism in adults than it does in children.

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