

Epilepsy and Epileptic Syndromes Cases Presented at Neuropediatric Clinic of "Mother Theresa" University Hospital Center, Tirana, 2012-2014

Afërdita Tako Kumaraku, Aida Bushati, Agim Gjokopulli, Renald Mecani, Valentina Tashko

"Mother Theresa" University Hospital Center Tirana, Albania

Abstract: Object of study: Identification of new epilepsies and epileptic syndromes cases presented at Neuropediatrics Clinic of "Mother Theresa" University Hospital Center, Tirana - 2012, classified according to ILAE 1985-89. Study Methodology: The study is prospective. The study includes all new epilepsy cases filled in the outpatient and inpatient clinic. The study has taken into account children who have had more than two episodes of unprovoked seizures. Every individual has undergone these examinations: Neurological examination, EEG Registration, fundoscopic examination, blood electrolytes, Children under 1-year-old with open fontanels were examined with Transfontanelar Ultrasound, A large part of them with CT- scan of the head. Results: 511 total cases, 232 females (45.4%) and 279 males (54.5%). Children 0-15 years included as follows: 73 children 0-1 years old, 165 children 1-4 years old, 179 children 5-10 years old and 94 children 11-15 years old. Partial epilepsy stood out in 298 children, benign-central-temporal forms in 78 children (15.2%), temporal epilepsy 41 children (8%), occipital epilepsy in 119 children (23.2%) and multifocal in 60 children (11.7%). 12 cases have resulted symptomatic with pathologic CT scan. 103 children (20.1%) with secondary generalized partial epilepsy; 29 have evolved from simple partial to secondary generalized, 74 from complex partial to secondary generalized. Generalized epilepsy, 67 children (13.1%), 16 atonic forms, 23 generalized tonic-clonic, 3 clonic, 11 tonic, 4 mioclonic, 5 absence, 4 West syndrome, 1 case with encephalopathy; 3 unclassified, 40 cases of febrile seizures. Conclusions: Out of 511 cases, 58.3% belong to partial forms, followed by secondary generalized partial forms 20.2%. Male cases stand out with 54.6% of total cases. Predominant age is 5-10 years old (35%). Febrile convulsions account for 7.8% of cases. Idiopathic forms are predominant. (partial and generalized 86.26% 76.71%).

Keywords: partial epilepsy, generalized, febrile convulsions

1. Purpose of the Study

The purpose of the study is to identify new cases of epilepsy and epileptic syndromes presented at the Neuropediatric Clinic of "Mother Theresa" University Hospital Center in Tirana, during 2012, classified according to ILAE 1985-89.

The objectives of this study are: To determine a therapeutic and diagnostic checklist for epilepsy cases. Promote new creative ideas for adequate logistics, which would help increase the level of diagnosis and documentation of epilepsy cases.

The methodology of the study:
The study was prospective.

All new epilepsy cases represented in hospital's and outpatient center's clinic were included. It took into account children who have had more than two episodes of epileptic seizures. Each individual underwent these examinations:

- Neurological examination,
- Registration of EEG,
- Fundoscopy
- Children under 1-year-old with open fontanels were examined with Transfontanelar ultrasound.
- Ultrasound.
- Electrolytes
- Head

A large number of the patients were examined with head CT.

This study used ILAE 1985-89 Classification.

International Classification of Epilepsy Types and Epileptic Syndromes (ILAE 1989)

1. Epilepsy and partial epileptic syndromes

- A. Idiopathic dependent on onset age.
 - Benign childhood Epilepsy with centro-temporal peaks (EPR)
 - Benign childhood Epilepsy with Occipital spike waves (EPO)
 - Primary reading Epilepsy
- B. Symptomatic
 - Frontal lobe Epilepsy
 - Parietal lobe Epilepsy
 - Temporal lobe Epilepsy
 - Occipital lobe Epilepsy
 - Partial continuous childhood epilepsy (S. Kojewnikoff)
 - Syndrome characterized by one explosive-specific cause

C. Cryptogenic (are distinct from symptomatic because their assumed organic etiology is not demonstrable)

2. Epilepsy and generalized epileptic syndromes

- A. Idiopathic dependent on appearance age. (in order of appearance)
 - Benign neonatal convulsions
 - Family benign neonatal convulsions

- Myoclonic benign childhood epilepsy
 - Childhood absence epilepsy type
 - Youth absence epilepsy type
 - Youth Myoclonic Epilepsy
 - Epilepsy with generalized tonic – clonic seizures mainly during waking up
 - Other generalized epilepsy not included in the above categories
 - Epileptic seizures caused by specific activator causes
- B. Cryptogenic and / or symptomatic (based on age of onset)
- West Syndrome
 - Lennox-Gastaut Syndrome
 - Myoclonic astatic epilepsy
 - Myoclonic absence epilepsy
- C. Symptomatic
- Specific etiology
 - o myoclonic encephalopathy
 - o Epileptogenic infantile encephalopathy with burst-suppression
 - o Any other generalized epilepsy not included in the above categories
 - Non-specific etiology
 - o Seizures that could complicate specific pathological conditions
3. Epilepsy and epileptic syndromes for which it is not possible to determine the partial or generalized origin.
- A. With both partial and generalized seizures
- Neonatal convulsions
 - Female childhood Myoclonic Epilepsy
 - Landau-Kleffner syndrome (Acquired epileptic aphasia)
 - Other undetermined Epilepsy cases, not classified above
- B. With unclear seizure definition, either partial or generalized
4. Special Syndrome
- Febrile convulsions
 - Isolated epileptic occasional seizures
 - Seizures caused only by visibly activating mental state (toxic or metabolic).

2. Results

There were represented 511 cases, of which 232 women (45.4%) and 279 men (54.5%). The study includes children from 0-15 years old, 73 children 0-1 years old, 165 children 1-4 years old, 179 children 5-10 years old and 94 children 11-15 years old. Most common were partial forms in 298 children, of whom 78 children (15.2%) had benign central-temporal forms, 41 children (8%) temporal, 119 children (23.2%) occipital, 60 children (11.7%) multifocal form. 12 cases have resulted in symptomatic with pathologic CT scan. 103 children (20.1%) with partial-secondary generalized. 29 of them evolved from simple partial to secondary generalized and 74 cases from partial complex to secondary generalized. 67 children (13.1%) were presented with generalized epilepsy, of whom 16 atonic forms, 23 generalized tonic-clonic forms, 3 clonic forms, 11 tonic forms, 4 myoclonic forms, 5 absence forms, 4 forms with West syndrome and 1 form with epileptogenic encephalopathy. Unclassified

were 3 children cases and 40 other cases of febrile convulsions, who started therapy after meeting 5 long-term treatment criteria of febrile convulsions.

3. Discussion

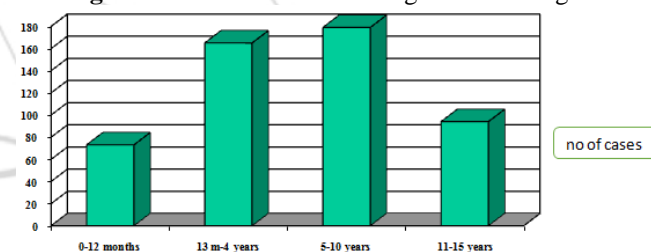
The incidence of epilepsy in the world is estimated to be about 20-50 cases per 100,000 inhabitants. It shows the number of new cases per 100,000 inhabitants at risk.

In various studies, age is taken in consideration sometimes in the moment of diagnosis and sometimes at the appearance of seizures. Our study used the age at the time of establishing the diagnosis, which in a majority of cases coincides with the onset of disease.

Table 1: Distribution of cases in group ages

	0-12 Months	13 Months-4 Years	5-10 Years	11-15 Years
No. of cases	73	165	179	94
Cases in[%]	14.3%	32.3%	35%	18.4%

Figure 1: Distribution according to different ages



The most affected age is 5-10 year-old (35% of cases), followed by 1-4 Years old (32.3% of cases).

Table 2: Distribution according to gender

	Female	Male	Total
No. of cases	232	279	511
	45.4%	54.6%	100%

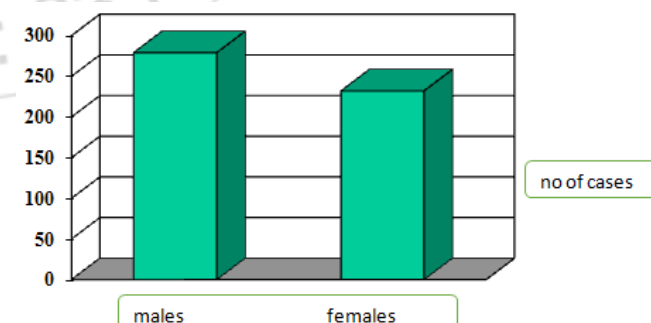


Figure 2: Distribution according to gender

Male sex is observed to predominate in most studies in the world, with an emphasis in 5-10 years old. This is due to a greater exposure of boys to cranial trauma, acceptance of disease and cultural characteristics with a greater attention towards boys than girls (Leviton and Cowan 1982). The same is noticed in our study as well, with predominance of males over females (54.6% versus 45.4%). This difference was not noticeable when studying the prevalence of

epilepsy. This prevalence is described in the world as 1.5-33.7 sick people per 1000 inhabitants. The large difference in the figures indicates that a large number after seeing the doctor for the first time and treated, then they neglect periodical follow up checkup which leads to reduced no. of cases. This is why in developed countries with a high awareness about the disease the prevalence is higher, indicator for a careful follow-up.

Table 3: Classification of cases

	<i>Partial</i>	<i>Generalized</i>	<i>Secondary generalized</i>	<i>Unclassified</i>	<i>Febrile seizures</i>	<i>Total</i>
No. of cases	298	67	103	3	40	511
Cases in [%]	58.3	13.1	20.2	0.58	7.8	100

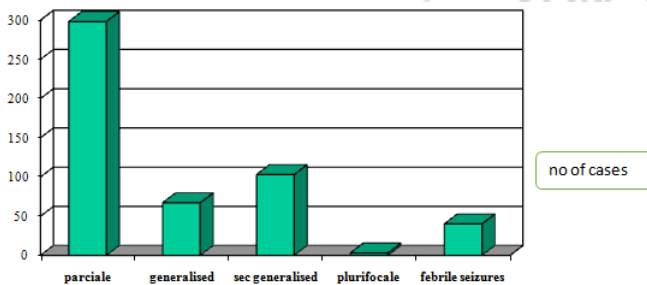


Figure 3: Classification of cases

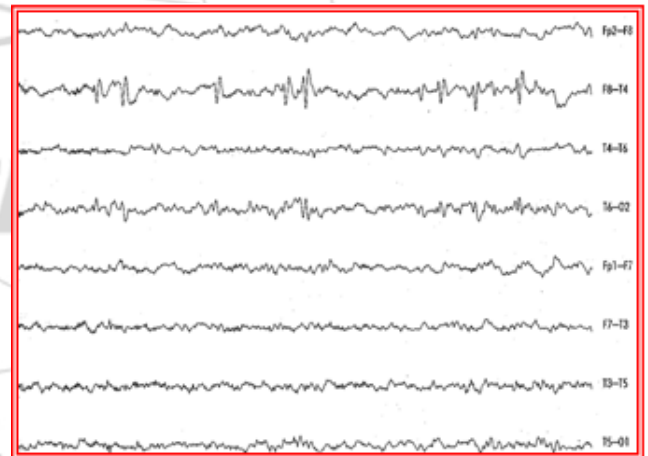
To classify epilepsy cases are used several Classifications: (ICES) International Classification of Epileptic Seizures which is easy to understand and to implement. It is primarily based on clinical epilepsy data, and less in ictal EEG and interictal data. Seizures are classified into simple partial where consciousness is preserved and complex where it is not. In this classification conscience is "the level of consciousness and / or the patient's response to external applied stimuli". Consciousness is defined "patient's contact with the event during and after the period in question" while the response / responsiveness "patient's ability to execute simple commands or voluntary movements". However, this is not a complete classification because for instance does not allow absence to be classified as primary generalized seizure alongside tonic-clonic or atonic and myoclonic. Therefore, this classification system is involved from ILAE 1989 in which are integrated epileptic syndromes and unclassified forms. But often in clinics, (as mentioned in the theoretical epilepsy treatment) are used mergers of different classifications, those best suited to the clinical aspect, EEG and etiology.

Benign partial epilepsy (EPB) is a very common form of childhood epilepsy. EPB appears in a limited time of childhood and the most common age of onset is 5-11 years. Genetic factors are predominant; it was observed that 35-59% of these children's relatives had epilepsy. Type the seizure may be simple partial motor, sensor-

autonomous or complex type, and sometimes secondary generalized.

EPB with central-temporal spikes is also called Rolandic form and is the most common of childhood. The literature states about 25% of cases, our study concluded 26.2% of cases. This form can occur during sleep and being awake, but 75% of cases have seizures while being asleep. When the patient is awake develops the so-called oro-pharyngeal seizure: partial seizure consists in increased salivation, suffocating voices from mouth and throat, trembling of the lips, oral cavity paresthesia with the feeling of dryness, inability to swallow, jaw's contraction and sense of suffocation. The seizure rarely affects only half of the face and is called "hemifacial hysteria". Sometimes facial seizure is associated with the same side hand, shoulder or leg shaking or secondary generalized. Consciousness may be maintained at all times. In other cases, various degrees of consciousness disturbance can be observed.

Sleep seizures often occur as soon as children fall asleep, but sometimes while still awake. The seizure begins as partial secondary generalized or generalized. These seizures have increased salivation and muffled sounds. Their frequency is different. Most of children have only one such seizure, which disappears when therapy is started. Some patients may have seizures during the first year, which slowly decrease and increase.



Benign Partial Epilepsy with Rolandic spikes

EEG states are slow multiphasic hyper-voltage spikes, located in Rolandic area, often negative. They increase during sleep and have a maximum of occurrence in stage 3 and 4 of NREM sleep.

Table 4: Partial Epilepsy

	<i>T-C</i>	<i>Temporal</i>	<i>Occipital</i>	<i>Multifocal</i>	<i>Total</i>
No. of cases	78	41	119	60	298
Cases in %	26.2%	13.75%	39.93%	20.13%	100%

With EEG could be found sharp waves, slow spiking or spikes with partial placement or multifocal sometimes secondary generalized. Based on their placement, some forms could be distinguished. They are represented in table no.5.

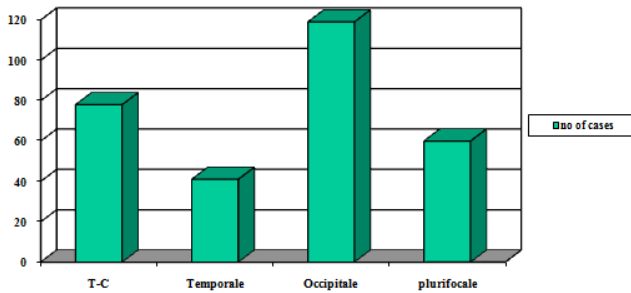


Figure 4: Partial Epilepsy

EPB with occipital paroxysm (EPBPO) is a common form of epilepsy. In our study occupies 39.33% of the cases. It is more pronounced in boys than in girls. The crisis can be simple partial or secondary generalized. They have visual or hearing aura. Seizures can be hemi-clonic or tonic-clonic and accompanied by dysphasia and headache, usually not associated with loss of consciousness, but may have different degrees of obscuring it.

EEG is characterized by well-structured basis activity, with sharp spikes and partial waves.

EPB with affective symptoms (BPEAS)

The child has a terrorized expression. He searches insistently for his parent, or runs away somewhere covering his face with his hands. They can experience automatisms such as swallowing, cough and usually lasts 1-2 min. It is not associated with tonic-clonic or tonic seizures. In general, they have good prognosis and respond very well to CBZ therapy.

Teenage EPB accounts for 24% of epilepsy cases which appear from 12-18 years old, and never extend over 20 year-olds. It appears more in boys (71%) than in girls. Usually the first seizure is followed within 36 hours by more seizures, which are never repeated, without neurological deficits. They appear during the day and while falling asleep, and can be secondary generalized. Therapy with anticonvulsants is not started in these cases. Postictal EEG is usually normal or with small atypical focal abnormalities.

Table 5: Generalized epilepsy

	No. of cases	Cases in %
Atonic	16	23.88%
Tonic-clonic	23	34.32%
Tonic	11	16.41%
Clonic	3	4.47%
Myoclonic	4	5.97%
Absence	5	7.46%
Flexion spasm	4	5.97%
Encephalopathy	1	1.49%

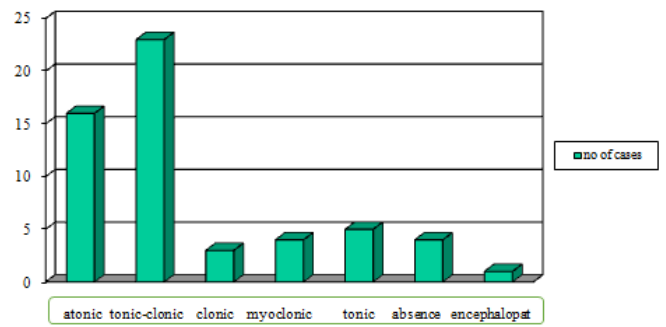


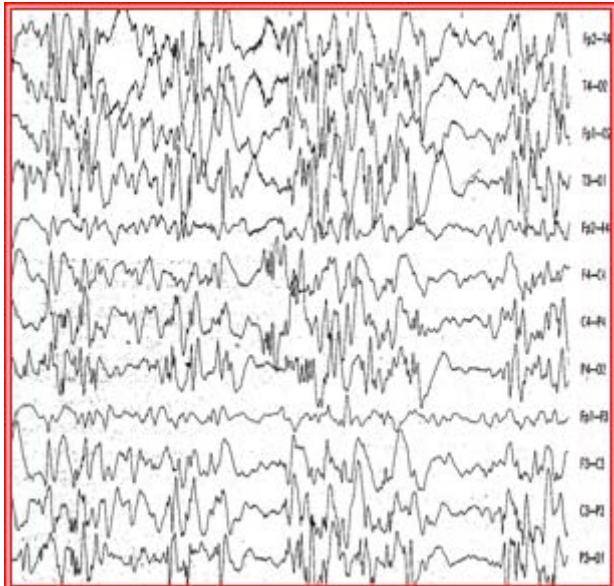
Figure 5: Generalized epilepsies

Generalized epilepsies occupy 33.2% of cases, where 13.1% are primarily generalized. Tissot (1770), was probably the first to describe absence in children. Gibbs et al. 1935 connected spike waves 3 cycles / sec with epilepsy. Terms such as petit mal, petit mal variant, minor motor convulsions, absence epilepsy, myoclonic atstatic epilepsy, pyknolepsy, pyknolepticpetit mal are used to describe the myoclonic crisis, tonic and atonic that sometimes accompany absence. The exact identification of the seizure is very important for laboratory examinations, the most appropriate therapy and prognosis. As described in the literature, the greatest number of primary generalized epilepsies constitute by tonic-clonic (34.32%) and those atonic (23.88%).

It is worth mentioning that among the various forms of generalized epilepsies, there are no sharp boundaries. For example, very often absence is associated with clonic, tonic and atonic components. In our study this stands in 7.46 % of primary generalized epilepsy cases, while in literature it is described about 13% (Bloom et al. 1978; Cowan et al 1989). I believe that this discordance exists because not all cases with epilepsy come and register at the University Hospital Center.



EEG: Absence Seizure



WEST Syndrome

Infantile spasms or West Syndrome is an important form of primary generalized epilepsy, which consists of infantile spasms, hypsarrhythmia and mental retardation. The spasms are mainly resistant to conventional treatment with anti epileptic drugs. It was described for the first

time about 150 years ago by West, while the treatment with corticosteroids was described 45 years ago. The spasms are in clusters. Approximately 42% of patients have spasms in flexion and extension, only 34% have only in flexion and 22% in extension. Mainly occur at the stage of falling asleep and waking up. In our study we had 4 of these (5.97%). They are mentioned because of the reserved prognosis, degree of disability, as well as side effects of therapy.

Juvenile Myoclonic Epilepsy

Another form is the Myoclonic epilepsy or Lennox-Gastaut, whose characteristics are: short frequent seizures, convulsions which are often resistant to therapy, critical condition, usually associated with mental retardation. To establish such a diagnosis, it takes time, because the process of establishing all the elements to meet the criteria for this diagnosis requires time (because in the beginning it can be confused with benign childhood myoclonic epilepsy). In our study we had 4 cases that met all the diagnostic criteria.

Table 6: Febrile seizures

Type	Represented in EEG	No. of cases	Cases in[%]
Partial	T-C	14	35
	Temporal	8	20
	Occipital	10	25
	Multifocal	2	5
Generalized		6	15
Total		40	100

Febrile seizures are the most frequent cause of seizures during childhood. They are present up to the age of 6 years old. The risk of febrile crisis in population is 2.7% to 3.3%. The risk for repeated crisis after the first febrile crisis is 29% to 35%. Increased risk for febrile crisis is associated with positive family history to febrile or a febrile crisis, temperature lower than 40° C during the first febrile crisis. The risk of epilepsy after the first febrile crisis is 1-2.4%. The risk for epilepsy after a complex febrile crisis is 4.1% to 6%. Also important is the duration of the febrile crisis. In our clinic were presented 40 children (7.8% of cases). Considering the incidence and its prognosis, we think that the number recommended to conduct detailed examination is high taking in consideration the protocol of treatment and diagnosis of CF. In such cases, detailed information should be provided by the primary doctors.

- Predominant age is 5-10 years (35%).
- Febrile convulsions account for 7.8% of cases.
- Idiopathic epileptic forms are predominant (partial 86.26% and generalized 76.71%).

References

- [1] Epilepsy in CHILDHOOD and adolescence; sec. Edition, Appelton R., J. Gidds 1998
- [2] Epilepsy; Kruja J; 2001
- [3] Epileptic seizures, pathophysiology and clinical semiology; Luders H. O.; Noachtar S., 2000
- [4] Pediatric epilepsy, Sillanpa M, Johannessen S, Blennow G, M Dam; 1990
- [5] Disease of the nervous system in CHILDHOOD, Jean Aicardi; 1994
- [6] Evidence-base study on Children epileptic seizures, Archives of CHILDHOOD; March 2004
- [7] CME lectures, January 2004, James Holmes

4. Conclusions

- Partial epileptic forms have predominated from 511 reported cases in our clinic during 2002 with 58.3%, followed by partial secondary generalized epilepsies 20.2%.
- It was noticed a slight predominance in males with 54.6% of cases.