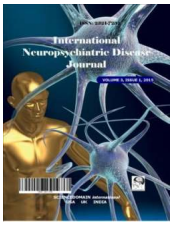

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# EEG Characterizations and Clinical Features in Sudanese Patients with Panayiotopoulos Syndrome

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***Authors' contributions***

*This work was carried out in collaboration between all authors. Author HAB wrote the protocol, performed the statistical analysis and managed the literature searches. Author MSE designed the study and managed the analyses of the study. Author SFA wrote the first draft of the manuscript. All authors read and approved the final manuscript.*

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## ABSTRACT

**Introduction:** Panayiotopoulos syndrome (PS) is an age-related, and a relatively frequent benign epileptic syndrome, characterised by predominantly autonomic symptoms and/or simple motor focal seizures followed, or not, by impairment of consciousness. Interictal electroencephalograph (EEG) shows occipital spikes, although multifocal spikes with high amplitude sharp-slow wave complexes at various locations can be present.PS mimics gastroenteritis, encephalitis, syncope, migraine, sleep disorders or metabolic diseases.
**Aim:** The aim was to characterise the EEG waves among Sudanese patients with PS, presented at the EEG unit of The National Ribat University, and El magzoub neurosciences center.
**Patients and Methods:** Out of 4319 abnormal EEGs and based on the classical EEG findings and clinical presentations, the EEG of patients suggestive of PS were identified.
**Results:** EEG findings of patients with PS in the study agree with classical international studies, as they were clustering of sharp and slow waves, sometimes associated with brain spiking, but usually shifting from one region to the other with dominant occipital paroxysms of sharp and slow waves.

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GIT upsets were the cardinal autonomic features; abdominal pain and vomiting were constant symptoms in most of the patients. Past history of febrile convulsions was positive in more than one-fourth of patients. All patients experienced febrile convulsions and this may be the main cause of seeking medical advice.

**Conclusion:** The results indicate that Panayiotopoulos syndrome in Sudanese children is not uncommon; moreover, the under diagnosis appears to be high. The classical autonomic manifestations and EEG features of Panayiotopoulos syndrome were well defined in Sudanese patients. EEG shows occipital or extra-occipital abnormalities.

*Keywords: PS (Panayiotopoulos syndrome); EEGs (electroencephalograph).*

## 1. INTRODUCTION

Epilepsy has been revised and redefined in 2005 theoretically as disorder of the brain characterized by an enduring predisposition to generate epileptic seizure, and practically has been defined by; At least two unprovoked (or reflex) seizures, occurring more than 24 hours apart, or one unprovoked (or reflex) seizure and the probability of further seizures similar to general recurrence risk (at least 60%) after two unprovoked seizures, occurring over the next 10 years, or when diagnosed epileptic syndrome [1]. In 2014 theses new definitions had been announced by Task Force of the Executive Committee of the International League Against Epilepsy (ILAE) [1].

PS was first described by Panayiotopoulos through a 30-year prospective study, and defined as autonomic seizures specific to childhood [2,3]. Despite its dramatic and lengthy manifestations, PS is often misdiagnosed. The main reason for this is that emetic and other autonomic manifestations are not recognised as seizure events. Our aim was to detect the prevalence of PS among patients with abnormal EEGs presented to our clinics.

### 1.2 Definition and Presentation

In a recent expert consensus PS was defined as "a benign age-related focal seizure disorder occurring in early and mid-childhood. It is characterised by seizures, often prolonged, with predominantly autonomic symptoms, and by an EEG that shows shifting and/or multiple foci, often with occipital predominance [3]. Autonomic seizures and autonomic status epilepticus with ictal emesis are the cardinal manifestations of PS. Autonomic seizures are epileptic attacks that start or entirely manifest with ictal autonomic disturbances that may be objective, subjective, or both that occur in children with normal development [4,5].

### 1.3 Importance of EEG in Diagnosis of Ps

The EEG is the most useful diagnostic test. The results of all other investigations are normal. [6]. The EEG in PS commonly (90%) reveals transient focal EEG abnormalities of sharp waves that occur in children with or without epileptic seizures and disappear in the late teenage multifocal functional spikes that are accentuated by sleep, [6-11]. In PS, functional spikes appear in many brain locations, often shifting from one to another region in series of EEGs. Occipital spikes predominate, in two-thirds of the patients [12]. Results of a single routine EEG may be normal in 10% of patients, which should prompt a request for a sleep EEG. The frequency, location, and persistence of spikes do not determine clinical manifestations, duration, severity and frequency of seizures, or prognosis [12]. The dipole analysis and magneto-encephalography showed the multifocal nature of epileptogenicity in PS [13,14]. Ictal video-EEG has unequivocally documented the epileptic nature of the autonomic manifestations in PS [15-17]. These may start long after the onset of the electrical ictal discharge and manifest with tachycardia, breathing irregularities, coughing, or emesis, which would be impossible to consider as seizure events without EEG [15-17]. Other recognisable conventional seizure symptoms such as convulsions appear later in the ictal phase or may not appear at all. The electrical onset of the ictal EEG paroxysms is more often posterior than anterior, with right or left lateralization [15-17]. It is not surprising if we expect that the PS is easy to be missed as a non-specific GIT diseases which are common during childhood, thus we re-examined the EEGs of any patients presented to the EEG Unit of National Ribat University and El magzoub Neurosciences Center with autonomic features and had abnormal EEGs.

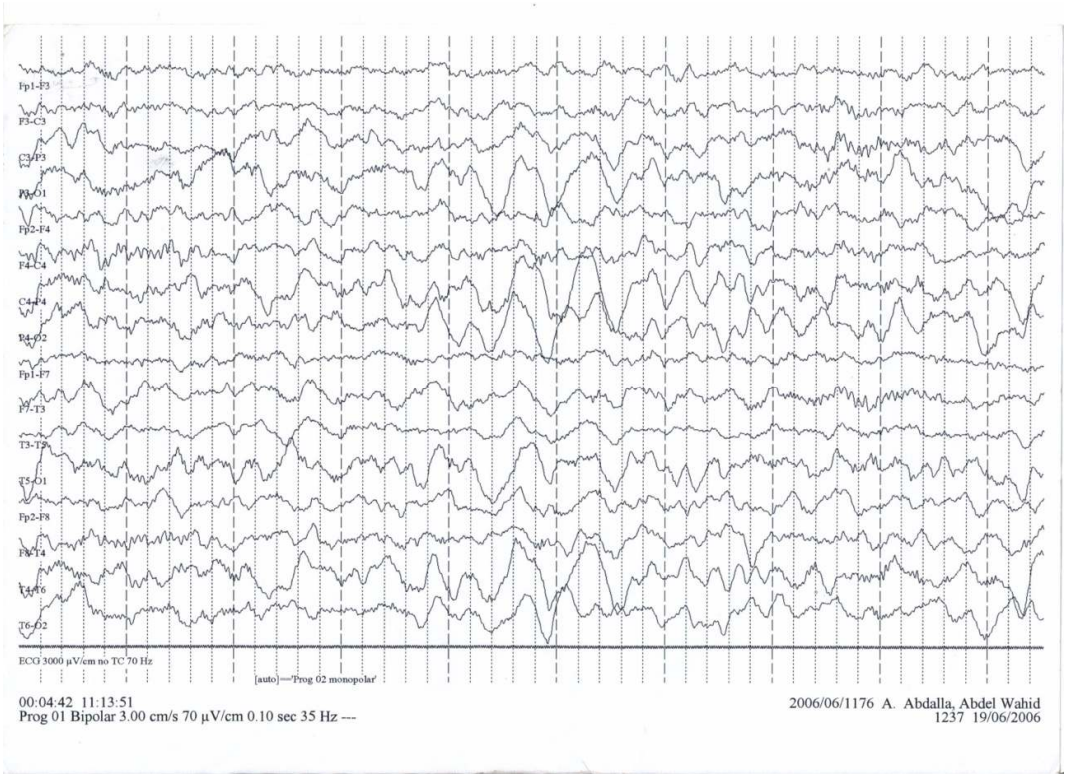


**Fig. 1. Transient paroxysms of sharp and slow waves with Occipital predominance in a Sudanese child with PS**

## 2. MATERIALS AND METHODS

It is a retrospective and cross-sectional study from March 2003 to May 2014 (eleven years) carried on the EEG Unit, of the Ribat University and El magzoub Neurosciences Center located in Khartoum, that usually received EEG requests for patients from all over Sudan. The study included7273 (4319 had abnormal EEGs) patients. The EEGs interpretation was done by a team consisted of the three authors (an experienced neurologist and neurophysiologist, PhD, and MSs candidates). The EEG machines that have been used in both centres, throughout the research period, were of the same model, they were Medtronic.

### 2.1 Inclusion Criteria

The population of the study included all Sudanese patients presented to the EEG Unit of the Ribat University and El magzoub Neurosciences, and had abnormal EEG, history of convulsions and or frequent GIT symptoms as nausea and vomiting.

### 2.2 Exclusion Criteria

We excluded from the study any patient fulfilled the inclusion criteria, but his referring record, or clinical examination revealed evidence of neurological deficit and /or had abnormal MRI.

## 3. RESULTS

During the period of the study, the two centres received 7273 patients, their EEGs were re-examined; 4319 were found to have abnormal EEGs, and 2954 had normal EEGs. Based on the definitive, “classic” characteristic of Panayiotopoulos Syndromes’ EEG findings, forty-five patients with PS have been diagnosed (Fig. 1), representing an overall prevalence of 1.04%, among all abnormal EEGs in the study, as shown in (Table 1). The females Sudanese patients with PS were 26 (57.8%) while the males were 19 (42.2%), illustrated by Fig. 2.

Table 3 showed that; the mean age for Sudanese patients with PS was almost 8 years old, 70% of them their ages were between 5-12 years, lessened to 8.8% for patients who their

aged less than 12 years. All the different EEGs which have been characterized in PS were seen in our study. The classical occipital paroxysms of sharp and slow waves were detected in all our patients. Clusters of epileptogenic sharp and slow waves with occasional spikes shifting from one region to the other characterized many EEGs. In many patients these occipital sharp and slow waves were concomitant with secondary generalised seizure discharges of polyspikes clusters (Table 4). All the patients were referred for EEG because of convulsions. History showed that all of them experienced nausea and vomiting, while 80% had bouts of diarrhoea. Abdominal pain was one of the cardinal features in 73.3% of patients. Past history of febrile convulsions was positive in 26.7% of all patients (Table 5).

**Table 1. Distribution of patients according to EEG findings presented to both centres**

| The EEG center | The abnormal EEGs | The normal EEGs |
|---|---|---|
| The Ribat University | 2063 (58.56%) | 1460 (41.44%) |
| El magzoub Neurosciences | 2256 (60.2%) | 1494 (38.8%) |
| Total | 4319 | 2954 |

**Table 2. Prevalence of PS in patients with abnormal EEGs**

| Center | Patients with abnormal EEGs | Patients with PS |
|---|---|---|
| National Ribat University | 2063 | 20 (1%) |
| El magzoub Neurosciences Center | 2256 | 25(1.1%) |
| Total | 4319 | 45 (1.04%) |

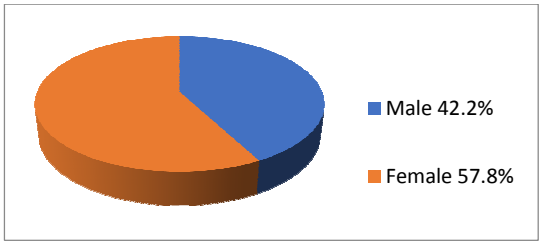


**Fig. 2. Gender distribution of patients with PS**

**Table 3. The age (mean, number and percentage) distribution among Sudanese patients with PS**

| Age in years | Number | Percentage (%) |
|---|---|---|
| 1-4 | 7 | 15.6 |
| 5-8 | 20 | 44.4 |
| 9-12 | 14 | 31.1 |
| More than 12 | 4 | 8.9 |
| Total | 45 | 100.0 |

*The mean age of onset = 7.93 years*

**Table 4. The characterizations and frequency of different EEG findings in Sudanese patients with PS**

| EEG findings | Numbers of patients | Percentage |
|---|---|---|
| Occipital paroxysms of sharp and slow waves | 45 | 100% |
| Clusters of epileptogenic sharp and slow waves with occasional spikes shifting from one region to the other, with evident occipital sharp and slow waves | 18 | 40% |
| Occipital sharp and slow waves with secondary generalised seizure discharges | 34 | 75% |
| Clusters of polyspikes with dominant occipital sharp and slow waves | 7 | 15.6% |
| Normal alpha background | 11 | 25% |
| Response to hyperventilation | 35 | 77.8% |

## 4. DISCUSSION

We showed that PS is seen in Sudanese children since we found EEGs with features of PS in 45(1.04%) of children with clinical features and abnormal EEGs. The classical features were seen in all the 45(100% of the patients). This is not surprising because it is known that the EEG of 90% of patients with PS shows sharp waves that appear in childhood with or without epileptic seizures and disappear in the late teenage (6-8). The multifocal epileptogenesis consists of paroxysms of transient polyspikes and slow waves in the EEG were detected in all our patients, with occipital predominance, this multifocal epileptogenesis accorded with

Panayiotopoulos and Sugita [15,18] observations and the occipital predominance partially agreed with Covanis findings, as he observed in two-thirds of his patients with PS [19]. Furthermore, extra-occipital epileptogenic spikes shifting from one region to another have been observed in EEGs of many Sudanese patients. This multifocality may be the explanation for the successions of different autonomic symptoms and their synchronicity may be the trigger for convulsions. The GIT symptoms particularly nausea and vomiting were the cardinal autonomic manifestations in all Sudanese patients and all of them developed convulsions which were the actual cause for their seeking medical care. In general, some of the patients with PS have normal EEGs [2], but it wasn't the case in our study as the abnormality in the EEG was the essential including criterion. In the same line with Panyiotopolous and Covanis outcomes [20,21], the extra-occipital epileptogenic spikes shifting from one region to another have been observed in EEGs of many Sudanese patients. Lada et al found nausea and vomiting were the main autonomic symptoms 80% and 86% respectively, consistent with our finding, but Linda observed convulsions only in 50% of patients [22]. It is worthy of note that all our patients presented after they had convulsions with a consequent request of EEG investigation. This means that the 45 cases we found may be the tip of the iceberg. The inference is that there are some unrecognized cases with a high level of misdiagnosis. We therefore recommend that all Sudanese children with the classical clinical features of nausea and vomiting especially when persistent should have an EEG carried out even when they don't have convulsions.

**Table 5. The clinical features of PS among Sudanese patients**

| Clinical feature | Number | Percentage |
|---|---|---|
| Convulsions | 45 | 100% |
| Nausea | 45 | 100% |
| Vomiting | 45 | 100% |
| Abdominal pain | 33 | 73.3% |
| Diarrhoea | 36 | 80% |
| History of febrile convulsions | 12 | 26.7% |

## 5. CONCLUSION

PS is not uncommon in Sudanese children. All our patients had the classical EEG features of occipital and extra-occipital shifting slow spikes There is therefore a need for a high index of suspicion in all Sudanese children with the clinical features in order to increase the diagnosis.

## CONSENT

It is not applicable. The consent is not applicable as the research is a retrospective study.

## ETHICAL APPROVAL

The ethical approval was obtained from the ethical and clinical research committee – faculty of medicine-National Ribat University.

## COMPETING INTERESTS

Authors have declared that no competing interests exist.

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