

# Faecal Incontinence: An Uncommon Manifestation of Panayiotopoulos Syndrome

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## Abstract

Panayiotopoulos Syndrome (PS) is a benign, common, age-dependent syndrome, with susceptibility to idiopathic vegetative seizures and autonomic status epilepticus. It is formally recognized by the International League against Epilepsy (ILAE). It is one of the three focal epileptic child benign syndromes next to rolandic epilepsy and idiopathic occipital epilepsy in children. It is characterized by its relative mildness and confusing manifestations. It is frequently misdiagnosed as non-epileptic disorders.

**Keywords:** Child • Faecal incontinence • Epilepsy • Panayiotopoulos syndrome

## Introduction

Two children of 12 and 9 years, from a non-consanguineous marriage, with specific no family or personal history, they have been having, since the age of five and six respectively, episodes of involuntary faecal incontinence followed by brief impairment of consciousness (2-3 minutes) and deviation of the eyes; while awake and asleep in both cases. These episodes occur at the frequency of twice per three months and of once every six months respectively [1-3].

## Case Report

Neurological examination was normal, so were the blood tests. The Electroencephalograph (EEG) in case 1, showed bilateral paroxysmal spikes predominant in temporal and occipital areas becoming generalized with hyperventilation (Figures 1-3); in case 2 EEG showed paroxysmal spikes in the right hemisphere. Brain and medullar Magnetic Resonance Imaging (MRI) had no defects in both cases. The epileptic origin was retained. The first patient was put on carbamazepine (Tegretol LP 200 mg at a dose of 1 tablet × 2/day) which decreased the frequency of incontinence episodes. The decision was not to treat the second child on account of the remote seizure frequency, lack of social impact of the disease.

## Discussion

The PS was first described in 1973 by Panayiotopoulos from a prospective study spreading over 30 years. The prevalence of PS in the general population is 2-3/1000. The age of onset varies from 1 to 14 years with a peak at 3-6 years [4] as it is in our cases. There is no apparent sex prevalence. Clinically, the

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cardinal manifestations of PS are vegetative seizures and autonomic status epilepticus. In approximately half of the cases, the seizures last from 1 to 30 minutes with an average of 9 minutes. Two-thirds of the seizures occur during sleep, the same child may have nocturnal and diurnal seizures, as seen in our patients [5]. The seizure commonly begins with vegetative events (81%), mainly represented by emetic triad (72%) that may be incomplete, made of nausea, retching and vomiting. Pupillary abnormalities including mydriasis and rarely miosis are possible. Other autonomic manifestations can be documented, namely, hypersalivation (6%), changes in thermoregulation as subjective and/or objective, critical or post critical rise in temperature, abnormal intestinal motility (3%) and irregularities in breathing and heart rates. Urinary incontinence is generally associated with impaired consciousness. Our patients have paroxysmal episodes of faecal incontinence; this event is rarely reported. The EEG is the most useful investigation. It shows great variability of focal spikes. All regions of the brain may be affected. The posterior regions, however, are predominant with two thirds of children who have at least an EEG with occipital paroxysms or occipital spikes (Figures 1 and 2), as seen in our first patient's EEG. In 40%, EEG abnormalities can be unilateral (Figure 3) and mainly on the right side (70%), like our second case.

The pathophysiology is not well known: an inherent autonomic instability may be responding by vagosympathetic access to a cortical hyperexcitability. Faecal incontinence was related to seizure activity in the fronto-mesial regions involved in the control of the pelvic floor muscles. Despite the high incidence of autonomic status epilepticus, the PS remains a remarkably benign affection, hence, prophylactic treatment with an antiepileptic medication is not recommended even in patients who have long-lasting or recurring seizures. The anti-epileptic treatment is desirable if a child has multiple recurrences or if the parents insist [4]. Most authors advocate carbamazepine as a first line treatment. Others argue for the efficacy of levetiracetam. Treatment was initiated in the first patient because of the recurrence of seizures, psychosocial impact and the insistence of the parent's contrast of the second case. The aim of our study is to evoke PS in front of repetitive autonomic manifestations in children presented with faecal incontinence, mainly if it is associated with impairment of consciousness.



Figure 1. Focal paroxysmal spikes.



Figure 2. Focal paroxysmal spikes predominant in temporal and occipital areas.

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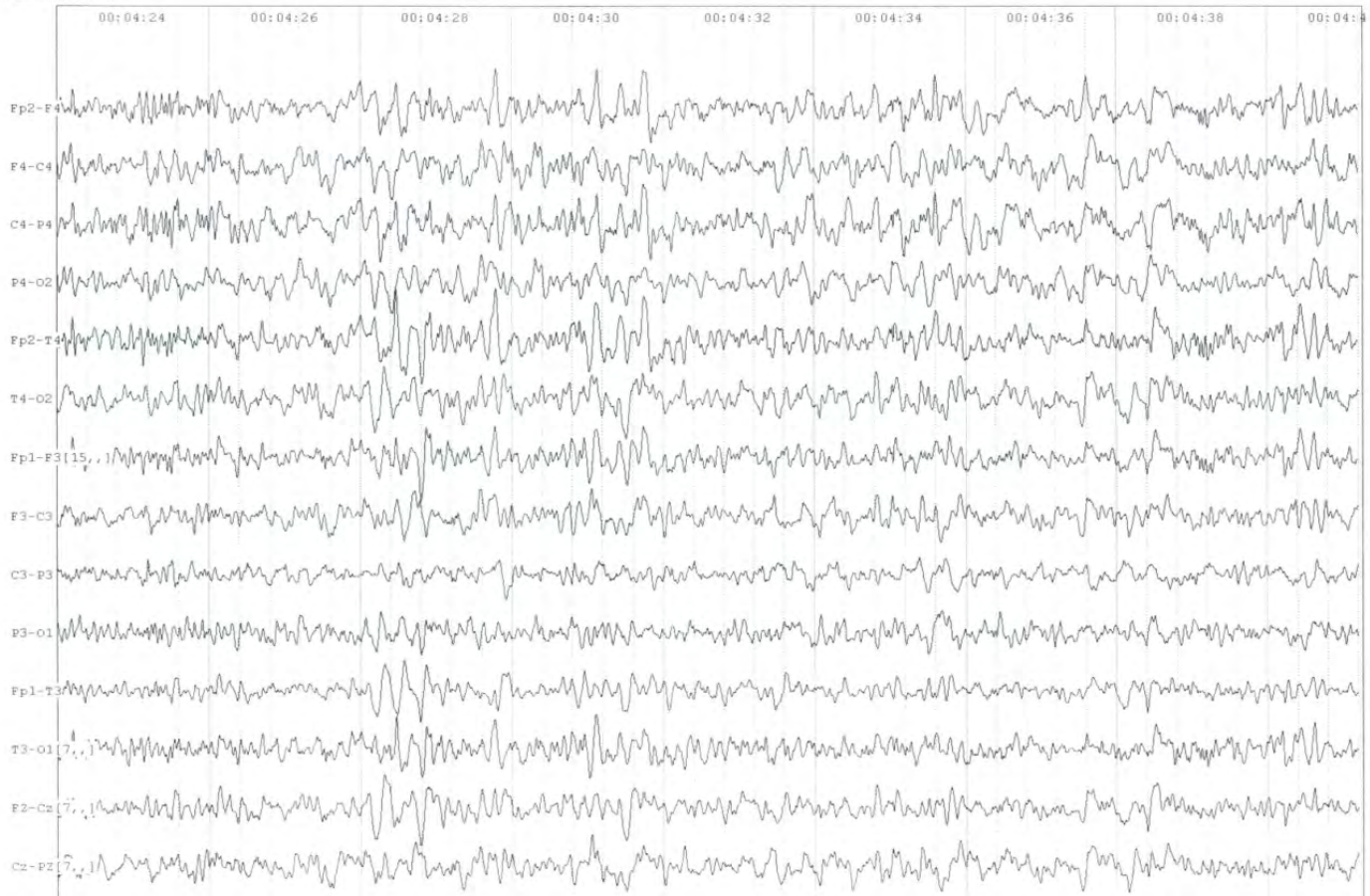


Figure 3. Paroxysmal spikes in the right hemisphere.

## Conclusion

The autonomic manifestations, intercritical EEG abnormalities and response to antiepileptic medication allowed us to rectify the diagnosis. Thus, to prevent an escalation of abusive or invasive complementary examinations, an EEG must be requested in children presented with autonomic features including faecal incontinence, as it must be considered foremost as an epileptic disorder.

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