Idiopathic photosensitive occipital lobe epilepsy (IPOE)\textsuperscript{5,12,73,75,109,110} manifests with focal seizures of occipital lobe origin, which are elicited by photic stimuli.

**Clari fications on Classification**

Occipital seizures precipitated by photic stimuli were overshadowed in the 1989 ILAE classification by the prevailing view that photosensitive epilepsies are mainly generalised.\textsuperscript{4} The new ILAE diagnostic scheme\textsuperscript{5} recognised IPOE as a new syndrome of reflex epilepsy with age-related onset. This is also maintained in the new ILAE report but with less certainty; IPOE was rated 2 on a score of 1–3 (3 being the most clearly and reproducibly defined) indicating the certainty with which the ILAE Core Group believed that each syndrome represented a unique diagnostic entity.\textsuperscript{6}

The boundaries of this syndrome of IPOE are genuinely uncertain. OPSs may start in adulthood, be part of idiopathic childhood occipital epilepsy of Gastaut (ICOE-G),\textsuperscript{111} develop later in children with rolandic seizures or occur accidentally during IPS of normal individuals or those with migraine.\textsuperscript{12,109} Gastaut\textsuperscript{111} included IPOE in his syndrome:

- Seven of 63 patients had IPS evoked occipital spikes, ‘not seen in the resting EEG’ and ‘unrelated to eye opening and closing’.
- Seven other patients with typical occipital paroxysms had generalised PPRs, which sometimes were associated with myoclonus (see also case 11 in Gaustaut et al.\textsuperscript{111}).

Contrary to this is the view that ‘reflex triggering of seizures have not been reported’ in ICOE-G.\textsuperscript{75}

Depending on severity, there may be three significant groups of OPSs:\textsuperscript{12,109}

1. OPSs may occur in patients with a low occipital epileptogenic threshold to IPS that manifests with seizures only under extreme exposure to the offending stimulation. These are ‘seizures that do not require a diagnosis of epilepsy’.\textsuperscript{5} Accidental single isolated occipital seizures in normal young people\textsuperscript{112} or patients with migraine\textsuperscript{112} during IPS are most likely, due to a low threshold to such events, and may not happen again.

2. OPSs in patients with idiopathic occipital epileptogenicity, which probably constitute the major part of IPOE. Patients, usually children, have clinical PPRs elicited by various environmental light stimulation (video games are far more common than television).
3. OPSs occurring in patients, usually children, with idiopathic focal or generalised epilepsies other than IPOE. These are often demanding cases with regard to diagnosis and management.

4. OPSs with bizarre ictal symptomatology mimicking hysterical attacks or migraine are well reported.\textsuperscript{12} That these symptoms, even the very prolonged and unusual ones, are ictal has been documented with ictal EEGs.\textsuperscript{73}

Considering all of these, the data presented in this chapter may not accurately represent a single syndrome of IPOE.

**Demographic Data**\textsuperscript{109,113}

Onset of the first provoked seizure may range from 15 months to adulthood (peak 12–14 years). Whether females predominate\textsuperscript{110,113} is debatable.\textsuperscript{109} Prevalence is reported as low, at around 0.4\% of all epilepsies.\textsuperscript{109} However, OPSs reached epidemic proportions in Japan among children watching the animated cartoon television programme *Pokémon (Pocket Monsters)*.\textsuperscript{74}

**Clinical Manifestations**\textsuperscript{12,73,75,109,110}

Occipital seizures precipitated by photic stimuli are induced by video games and less often by television or other photic stimuli. These reflex seizures contain all the elements detailed in the spontaneous seizures of occipital lobe epilepsy.\textsuperscript{73,114-116} OPSs commonly manifest with visual hallucinations, blurring of vision or blindness, alone or in combination. Less often, these visual symptoms may follow other ictal occipital manifestations, such as deviation of the eyes and head, eyelid fluttering and orbital pain.

Visual symptoms may be the only ictal manifestations, usually lasting for seconds and frequently 1–3 min. When longer (5–15 min), other ictal manifestations also occur.\textsuperscript{73,114} Consciousness is not impaired during the phase of visual symptoms.

*Progression of visual seizures to other ictal symptoms:* Autonomic symptoms, such as those occurring in Panayiotopoulos syndrome (mainly retching and ictal vomiting), often follow the occipital symptoms and may end with secondarily GTCSs.\textsuperscript{73,75}

*Other type of seizures:* Patients with IPOE may have exclusively OPSs. Others may also have spontaneous visual or other types of seizures. These vary from eyelid fluttering, myoclonic jerks and absences to GTCSs that occur independently of the occipital seizures.\textsuperscript{109,113} In some cases, spontaneous secondarily GTCSs occur only during sleep.\textsuperscript{12} Rarely, patients with rolandic seizures may later develop OPSs.\textsuperscript{12,115}

*Post-ictal symptoms:* OPSs, like the spontaneous occipital seizures, are more likely than any other type of focal seizures to be followed by headache, nausea and vomiting. The headache is usually mild and diffuse, but may also be severe and throbbing, occurring 10–20 min after the end of the visual
hallucinations. Post-ictal headache may also be associated with vomiting, lasting for several hours. 73

Precipitating Factors
By definition, all patients with IPOE are sensitive to flickering lights. Depending on the severity of photosensitivity in some patients, seizures may be elicited by minimal photic provocation; in others, combined pattern and photic or prolonged exposure may be responsible, whereas in others still (probably most cases of IPOE) photic stimuli are effective only if combined with other precipitating factors such as excitement or frustration, fatigue and sleep deprivation. 11

Diagnostic Procedures
All except the EEG are normal.

Electroencephalography 12,73,75,109,110
By definition, all these patients are photosensitive and IPS elicits abnormal EEG paroxysms of spikes or polyspikes, which may be entirely confined to the occipital regions, or PPRs of generalised spike–wave discharges (GSWD), which predominate in the posterior regions (Figs. 2.1 and 2.2). Spontaneous, mainly posterior, spikes often appear in the resting EEG. Centrottemporal spikes may coexist.

Occipital spikes and other posterior abnormalities induced by IPS are considered to be of much lower epileptogenic capacity than generalised PPRs. They may occur in 50% of patients who do not have seizures. 13 Occipital spikes precede generalised PPRs in 90% of photosensitive patients when light and pattern are combined during IPS. 13,117,118

Ictal EEGs have documented the occipital origin and spreading of the discharges to the temporal regions. 75

In my experience with video-EEG recordings, most patients with IPOE also have other types of seizure induced by IPS such as eyelid, limb, body or finger myoclonic jerks, eyelid flickering or brief absences that are sometimes mild and may escape detection without video-EEG and if cognition is not tested (Fig. 2.1). 12

Visual evoked potentials are always of abnormally high amplitude, 75 as indeed they are in any type of photosensitive epilepsy (Fig. 1.3).

Aetiology
By definition, idiopathic photosensitive occipital lobe epilepsy is idiopathic with genetic influences. Some patients have a family history of IPOE or IGE. Overlapping with JME has been reported. 113 Symptomatic occipital photosensitivity 119 is not part of IPOE.
Differential Diagnosis

The differential diagnosis of IPOE includes migraine (rarely an actual problem if symptoms are appropriately analysed), ICOE-G, IGE (probably of management importance) and non-epileptic paroxysmal events (sometimes very difficult to differentiate).
The differential diagnosis of visual seizures from all types of migraine with visual aura has been detailed elsewhere. Some seizures of IPOE may be prolonged, also progressing from visual symptoms to nausea and vomiting with altered consciousness.\textsuperscript{73,120} The spread of the discharge from the occipital cortex can be slow, and responsiveness may be maintained while the patient is vomiting.\textsuperscript{73,120} These seizures may be erroneously diagnosed as migraine proper.

In children and adolescents, the differentiation of IPOE from ICOE-G may not be needed if they are the same syndrome.

Differentiation of IPOE from generalised photosensitive epilepsies should rely on clinical criteria. Occipital spikes often precede generalised discharges in photosensitive epilepsies.

In adults we have reported occipital photosensitivity in adult patients aged about 30 who presented with a late-onset first GTCS (often preceded by visual symptoms).\textsuperscript{121} Of 1,550 patients with seizures, three women and two men (0.3\%) had EEG occipital photosensitivity and onset of solitary (three patients) or infrequent seizures in adulthood (median age 31 years, range 26–35 years).

Fig. 2.2 (a) Sample from an EEG of a man who had his first seizure at the age of 35 years while in a lift cradle at work. His vision became blurred, he felt dizzy and, within 2 min, he had a GTCS. No further seizures occurred in the next 6 months of follow-up. MRI was normal. (b) Sample from an EEG of a woman who had her first seizure at the age of 31 years. There was a cluster of precipitating factors; she had consumed a few alcoholic drinks, was sleep deprived, 4 months pregnant and dancing exposed to flickering discotheque lights until the early hours of the next day. She first experienced whirling lights in front of her eyes, visual perception became disturbed and within 1 min she had a GTCS. She was well during the next 4 months of follow-up. MRI was normal.
All five of these patients had generalised convulsions, which were preceded by blurring of vision or elementary visual hallucinations in four cases. Precipitation by lights, alone or in combination with other factors, was apparent in only two patients. Seizures were diurnal in all but one patient. According to the inclusion criteria, all patients had EEG occipital spikes elicited by IPS (Fig. 2.2). Neurological and intellectual states, as well as brain imaging, were normal.

**Prognosis**

Prognosis varies significantly among affected individuals. This depends on the severity of photosensitivity and exposure to the offending visual stimuli. There are rare case reports of normal young people or patients with migraine having an occipital seizure during IPS. Some patients may have only one or two occipital seizures in their life despite exposure to precipitating factors and no drug treatment. Others, particularly those who also have spontaneous seizures, may need medication for 1–3 years, together with strict avoidance of or cautious exposure to insulting stimuli. However, other patients may have frequent spontaneous and elicited occipital fits alone or in combination with other types of seizures, which include myoclonic jerks, often of the eyelids, infrequent absences or GTCSs.

**Management**

Advice about avoidance of precipitating factors is essential and is similar to that given to patients with any type of photosensitivity. Particular emphasis is needed about video games and television. Commercially available blue lenses, named Z1, that are highly effective in controlling PPRs may be of value.

The effectiveness of valproate has not been tested in OPSs as it has in generalised photosensitive seizures. Patients with IPOE who are resistant to valproate became seizure free with add-on carbamazepine. Levetiracetam or clobazam are possible alternatives.
Reflex seizures and related epileptic syndromes
Panayiotopoulos, C.P.
2012, XVII, 62 p. 12 illus. in color., Softcover