VIII MISCELLANEOUS OBSERVATIONS

History of a photosensitive patient

Richard, a 12-year-old boy of normal intelligence, is the oldest in a family of two. Pregnancy, delivery and early development were normal. No serious illnesses or traumata occurred.

His mother, who was 23 years old at the time of his birth, had suffered from headache attacks provoked by emotions and bright lighting since her (early) childhood. EEG recording revealed epileptiform activity for which she was treated with phenobarbitone and phenytoin. However, no consistent reduction of her headache complaints was achieved on this medication-scheme and treatment with antiepileptic drugs was stopped. Recent EEG examination at our department did not reveal epileptiform activity or photosensitivity at that time.

Richard, too, suffered from headache attacks with nausea and vomiting monthly since the age of eight years. Like his mother, these attacks were provoked by emotions and bright lights. EEG examination at the age of eight years revealed spontaneous, generalized epileptiform activity and photosensitivity. Treatment with a combination of phenobarbitone and phenytoin did not influence his headache.

At the age of 12, after a period of relative strain at school and at home, he had a partial complex seizure when watching colour TV at a distance of approximately one meter from the set. Disco-lighting and flashing lights in a pop-music program on TV made him feel awkward and irritated his eyes. Physical examination, including testing of visual acuity and stereopsis, did not reveal any abnormality.

EEG examination, carried out while receiving phenobarbitone 60 mg and phenytoin 90 mg daily, showed various abnormalities. Asymmetrical occurrence of alpha-rhythm both in terms of amplitude and of frequency was found with the left parieto-occipital region showing better developed alpha activity than the right one. Spontaneous generalized spike waves with a left temporo-parietal onset and a duration of 4 sec were recorded with accompanying clinical signs of loss consciousness with eye-blinking.

IPS yielded a symmetrical response and a CPPR at the following frequency ranges: 8-50 Hz at eye closure, 10-50 Hz with eyes closed and 6-50 Hz with eyes open. Discrete eye blinking was observed during the provoked generalized discharges. Richard was also sensitive for small patterns presented at a visual angle of 10 degrees while stimulation of the right visual half-field was more effective than the left in provoking such discharges. Generalized epileptiform discharges could be provoked at a distance of two meters or less from a black-and-white TV.

After these examinations he was treated with valproate monotherapy (1200 mg daily): headache attacks and epileptic seizures disappeared and EEG examinations showed no more generalized epileptiform activity but only occasionally bitemporal sharp waves. His photosensitivity was reduced to the extent that he did not meet our criteria any longer; only during eye
closure and with eyes closed could some reaction be evoked with flash frequencies in the range of 18-30 Hz. Richard's pattern-sensitivity was likewise reduced (he was only sensitive for patterns at a visual angle threshold of 48") while his TV-sensitivity disappeared. He remained seizure- and headache-free for two years upon which time it was decided that the medication should be reduced. Nevertheless his headache recurred at the daily valproate dosage of 500 mg while, first shortly after, he suffered from a tonic clonic convulsion when viewing a colour TV at a distance of approximately two meters. Compared to the last EEG examination with valproate 1200 mg daily, his reaction on IPS was much stronger and his TV-sensitivity was, again, very marked. The valproate dosage was increased to 1000 mg daily, whereupon the epileptic attacks, headache complaints and visual sensitivity were strongly suppressed for another two years.

History of a self-inducing photosensitive patient

Johanna, a 13-year-old girl of normal intelligence receiving no medication is the oldest of two siblings. Pregnancy, delivery and early development were normal. There was no history of serious diseases or traumas.

Her father, who was 29 years old at the time of her birth, suffered from partial epilepsy from a right fronto-temporal focus. He presented with tonic clonic convulsions at the age of 13. Until recently the tonic clonic convulsions occurred at a frequency of 1-2 yearly despite anticonvulsant treatment. He proved to be photosensitive to stroboscopic flashes of 10-30 Hz, while no pattern-sensitivity was found. The paternal grandmother also suffered from epilepsy (diagnosis and photosensitivity unknown).

Johanna started eye-blinking with upward deviation of the eye-balls at the age of 12, especially when talking. Eye-blinking increased since that time and she now blinks nearly continuously. Sunlight and artificial lighting have a provocative effect on her eye-blinking behaviour according to her mother. She sometimes does not answer a question or lets things drop from her hands while eye-blinking. Furthermore, she herself noticed she sometimes looses her memory for a short period of time. Although she does not complain about her eye-blinking behaviour or about any visual stimulus, in daily life or upon examination, she nevertheless wants to get rid of this behaviour because friends at school tease her by calling her "flashing beacon".

Physical examination, including visual acuity and stereopsis, did not reveal any abnormality. EEG examination showed a normal background activity without spontaneous epileptiform activity. IPS yielded a symmetrical photic driving response and symmetrical CPPR’s at the following frequency ranges: 6-40 Hz during eye closure, 10-40 Hz with eyes closed and 10-30 Hz with eyes open.

A pattern viewed at a visual angle of 13° composed of vertically-oriented stripes provoked generalized epileptiform activity. TV-sensitivity was not found. Self-induction was registered about once every minute during the two hour telemetered EEG and closed circuit TV-monitoring. Slow eye closures with upward rotation of the eye-balls and additional eye-blinking at a frequency of about 10 Hz elicited generalized spike-wave complexes with a duration of 1-3 seconds in 50 percent of occurrence of these movements. These epileptiform discharges were often accompanied by slow, irregular eye-blinks, unlike the eye-blinks which preceded them. As additional evidence of the phenomenon of self-induction, the discharges could be evoked by her on request.

Treatment with valproate 1200 mg daily (incidentally with bloodlevel 56 mg/l only, which
raises the question of compliance) reduced her IPS-sensitivity range and pattern-sensitivity only slightly while having no effect on her self-induction behaviour or rate during the following five years.

EEG examination carried out while on phenobarbitone 100 mg daily confirmed her TV-sensitivity at a distance of about two meters or less; no spontaneous EA was recorded.

Miscellaneous histories of photosensitive, non-self-inducing patients:

Ursula, a 12-year-old girl of normal intelligence with primary generalized epilepsy, had a history of febrile convulsions and was successfully treated with phenobarbitone and later valproate monotherapy. At the age of ten, the valproate medication was reduced and stopped after which she had complained of myoclonic movements in the right side of her face, occurring only in the morning upon awakening. After a period of nightsleep deprivation and strong emotional involvement, she also suffered from a tonic clonic convulsion on awakening. Extensive questioning about specific circumstances on which she awoke each morning revealed that she laid underneath an attic-window. No more myoclonic or tonic clonic seizures occurred after repositioning her bed to another corner of the attic.

She was sensitive to IPS in all three eye conditions (eye closure and eyes closed in the range 15-30 Hz, eyes open in the range 15-20 Hz) and the provoked discharges were accompanied by myoclonic jerking.

Anna, a 23-year-old woman of normal intelligence had a history of tonic clonic convulsions starting at the age of seven years induced by TV-viewing only. She experienced that these seizures only occurred within a viewing distance of 1.5 meters from a black-and-white TV set at high image contrast. She achieved better seizure control by avoiding watching TV (100% seizure-free intervals) than by taking phenobarbitone, yet watching TV.

Miscellaneous histories of self-inducing, photosensitive patients:

René, a 23-year-old man of normal intelligence, started having partial complex seizures at 10 years of age (diagnosis: partial epilepsy of right frontal focal origin). At about the same time he demonstrated self-inducing behaviour with slow eye-closures and blinking, especially when starting a conversation. He became aware of such behaviour only at the end of attending primary school, he said. When people nag him with his behaviour, he simply explains to them that he has suffered permanent neuronal damage during birth (this cannot be confirmed by physicians). Therefore, he maintains that he suffers from so-called “light-reflexes” and is being attracted to sunlight or bright artificial lighting. Sunlight, especially during the summer, increases the self-induction rate more than artificial light.

Notwithstanding his knowledge about the influence of sunlight, he never wears sunglasses. There has been some diminishing in the rate of self-induction with advancing age, according to René. He also says that this behaviour can better be controlled now he is fully aware of what he is suffering from. Self-induction seems to exert some sort of tension-releasing effect on him. In his own room at home he has a disco-lighting installation, which he switches on if he feels that he needs to relax.

During the EEG investigation, carried out when he was on valproate 300 mg daily, he proved to be (only) sensitive to IPS during eye closure at 18 Hz but had a high rate of self-induction of EA (at least once every 20 seconds). He refused any further EEG investigations after this examin-
ation and stopped consulting his neurologist altogether until the time of writing (six years later).

Annelies, a 17-year-old girl of normal intelligence, with a primary generalized epilepsy (tonic clonic convulsions, absences and myoclonic movements over the whole body) started having seizures at the age of four years. At that time she was compulsively attracted to the TV-set, a floor-lamp, sunlight and venetian blinds. Her parents found her often with her nose pressed on the TV-set while the set was turned on blinking and staring and sometimes with head nodding; she seemed to be in a trance. When she took antiepileptic drugs (valproate and ethosuximide) the self-induction behaviour diminished remarkably.

At the age of 17, she now admits to self-induce only if she is upset; in normal circumstances she feels too ashamed to self-induce.

EEG examination carried out while receiving valproate 900 mg and ethosuximide 750 mg daily, showed moderate sensitivity to IPS in all three eye conditions, accompanied by myoclonic movements of her whole body (eye closure in the range of 15-20 Hz, eyes closed and open in the range of 15-25 Hz), pattern and TV. Self-induction was not registered.

Johan, a 25-year-old man with a partial epilepsy (probably caused by a right temporal focus), had his first seizure in front of a TV at the age of nine years and was treated with high dosages of various antiepileptic drugs. His frequent eye-blinks in that period were not recognized as self-induction behaviour. There were many psychosocial problems since his childhood. He had used soft drugs and stimulating drugs since his late tens. The obvious self-induction behaviour with slow eye closures and eye-blinking noted during the taking of the clinical history and physical examination was something he refused to talk about: “it was nobody’s business”.

EEG examination carried out while receiving carbamazepine 400 mg and valproate 2000 mg daily, revealed moderate sensitivity to IPS in all eye conditions (eye closure in the 15-25 Hz range, eyes closed in the 15-30 Hz range, eyes open in the 15-18 Hz range) with self-induction of epileptiform activity approximately once every minute.

Henk, a 33-year-old hospitalised male of subnormal intelligence suffering from secondary generalized epilepsy started having seizures at the age of three years. At the age of nine years it was noticed that he was compulsively attracted to sunlight and artificial lights. When walking in the sunshine, he turns his face towards the sun and blinks fast followed often by myoclonic movements of his body at which time he stops walking shortly. Sometimes the myoclonic seizures lead to a tonic clonic convolution. When keeping an appointment with the dentist, he is administered diazepam 10 mg in advance as a safety precaution, to prevent him from compulsive attraction to the dentist’s artificial lighting.

EEG examination carried out while receiving valproate 2700 mg and carbamazepine 1200 mg daily, showed sensitivity to IPS only at 18 Hz during eye closure and at the 15-20 Hz range with eyes open. His self-induction rate (eye blinking and slow eye closures) was about once every five minutes.