Epilepsy & Behavior Case Reports 2 (2014) 49–53

Contents lists available at ScienceDirect

Epilepsy & Behavior Case Reports

journal homepage: www.elsevier.com/locate/ebcr

Case Report

Noninvasive treatment alternative for intractable startle epilepsy

Sylvia Klinkenberg [a,e,*], Sander Ubbink [b], Johannes Vles [a,e,f], Anton de Louw [f], Mariette Debeij van Hall [f], Dyon Scheijen [c], Jan Brokx [d,e]

[a] Department of Neurology, Maastricht University Medical Center, Maastricht, The Netherlands
[b] Department of Otorhinolaryngology/Head and Neck Surgery, University Medical Center Groningen, The Netherlands
[c] Department of Audiology, Adelante Zorggroep, Hoensbroek, The Netherlands
[d] Department of Otorhinolaryngology/Head and Neck Surgery, Maastricht University Medical Center, Maastricht, The Netherlands
[e] School for Mental Health & Neuroscience, University Maastricht, The Netherlands
[f] Epilepsy Center Kempenhaeghe, Heeze, The Netherlands

ARTICLE INFO

Article history:
Received 24 January 2014
Received in revised form 12 February 2014
Accepted 13 February 2014
Available online 1 April 2014

Keywords:
Intractable epilepsy
Startle-provoked seizures
Sound generators
Retrainers
Behavioral therapy

ABSTRACT

We describe a treatment alternative for intractable, startle-provoked, epileptic seizures in four children aged between 8 and 14. Three of the four children had symptomatic localization-related epilepsy. They all suffered from intractable epilepsy precipitated by sudden sounds. The fact that seizures tended to occur with high frequency – more than one seizure a day – had a clear impact on daily life. Clinical seizure pattern demonstrated asymmetric tonic posturing in all four children. Three children experienced several seizure types including focal seizure onset. All children had focal neurological signs or learning disabilities or a combination of both. Our noninvasive treatment method using psychoeducational counseling and sound generators was applied in four children, resulting in a seizure frequency reduction of ≥50% in two of them.

1. Introduction

Startle-provoked epileptic seizures (SPES) are not a uniform epileptic entity. The majority of children with SPES are known to have a congenital brain condition or an acquired brain injury which occurred early in life [1–5]. Seizures tend to be highly refractory, and impact on daily life and psychomotor development over time is high. Several authors report a favorable outcome of epilepsy surgery, although numbers are small [2,3,6,7].

Unexpected sounds are the stimuli most frequently found to precipitate SPES, although somatosensory and visual stimuli have also been reported. Background sounds are known to reduce startle responses in a positive way. In the case of a weak prepulse — a soft sound ahead of the startle inducing sound, reduction of startle response and perceived stimulus is reported [8]. In the audiological field, psychobehavioral therapies are regularly used to reduce symptoms caused by an oversensitivity to sounds, for instance, in the conditions tinnitus or hyperacusis. Most treatments are based on the tinnitus-retraining therapy introduced by Jastreboff [9]. The usage of sound generators (tinnitus masker) or hearing aids is part of this therapy. The effect of background sounds on startles and the use of sound generators in psychobehavioral therapy in the audiological field were the bases for a treatment alternative for SPES. In patients with epilepsy, Brown and Fenwick have already described a positive effect of behavioral therapy on seizure activity [10]. We report four children with SPES. One of the boys with SPES reported an improvement in seizure frequency during holidays, especially when staying at the beach, where he heard the sound of waves breaking continuously. This was the basis for considering a treatment alternative for SPES. We combined both methods (relaxation and distraction) and can now report nonpharmaceutical seizure-management in 4 children with intractable SPES predominantly triggered by auditory stimuli. We used a noninvasive method consisting of a combination of psychoeducational counseling and sound generators.

2. Case reports

Four children, all male, aged between 8 and 14, suffered from intractable epilepsy precipitated by sudden sounds.

2.1. Patient 1

Patient 1 is male, firstborn of healthy parents; pregnancy and delivery were uneventful. Neonatal period was complicated by a group B meningococcal sepsis. Seizures occurred from the age of fourteen months,

Abbreviations: SPES, startle-provoked epileptic seizures; ASR, acoustic startle reflex; MR, magnetic resonance.

* Corresponding author at: Department of Neurology, Maastricht University Medical Center, P.O. Box 5800, 6202 AZ Maastricht, The Netherlands. Tel.: +31 43 3875058; fax: +31 43 3877055.
E-mail address: s.klinkenberg@mumc.nl (S. Klinkenberg).

http://dx.doi.org/10.1016/j.ebcr.2014.02.002

and he developed an epileptic encephalopathy. He suffers from myoclonic seizures of the left arm, atonic seizures, and tonic seizures. The latter were provoked by sudden sounds or unexpected visual stimuli. Neurological examination revealed no focal abnormalities; there is a psychomotor developmental delay with severe learning disabilities. Extended diagnostic work-up, including magnetic resonance (MR) imaging and MR spectography, and neurometabolic and genetic evaluation were all normal. Three seizure types have coexisted since the onset of epilepsy. He was not a suitable candidate for epilepsy surgery. At the age of 5 years, a vagal nerve stimulator was implanted, and a revision took place after two years because of dysfunction and increase in seizure frequency. Ketogenic diet had no effect. At age 10, his epilepsy was still refractory, with seizure frequency varying from several seizures a day to 30 seizures a day, depending on triggering factors. He used lamotrigine and clorazepam at the time of referral to the audiological team. Treatment alternative with sound generators was initiated after counseling both the boy and his parents. During a follow-up period of a year at the child neurology outpatient clinic, his parents reported that he continuously removed his retrainers. As there was no change in seizure frequency, they stopped the sound generator therapy.

2.2. Patient 2

Patient 2 is the third son of healthy parents, born after an uneventful pregnancy and delivery. At age three months, he suffered from pneumococcal meningitis. He developed a spastic right-sided hemiplegia and localization-related complex partial seizures over a period of two years. He became seizure-free and did not use antiepileptic drugs for several years until he developed startle seizures at age 10. A sudden sound resulted in a tonic seizure, sometimes accompanied by urinary or fecal incontinence. Seizure frequency was variable, depending on triggering factors such as a pencil falling in the classroom or a school bell ringing. He had at least 2–3 seizures a day; they did not respond to antiepileptic drugs. He attends a normal school. His MR imaging showed an encephalomalacia of the left hemisphere. Epilepsy surgery was considered, but his parents preferred the noninvasive treatment alternative with sound generators. He and his parents were counseled by our audiological team, and therapy with sound generators was initiated with considerable success. Over a period of a few years, seizure frequency was reduced to one per month. He was even able to watch fireworks and to set one off himself when wearing his sound generators.

2.3. Patient 3

Patient 3 was born of healthy parents; pregnancy and delivery were uneventful. He is known to have Von Willebrand's disease. He experienced a traumatic head injury due to a fall from a trampoline at the age of four years and 6 months complicated by intraparenchymal bleeding and subdural hematoma with mass effect requiring evacuation twice. Magnetic resonance imaging reveals a cystic encephalomalacy of the right cerebral hemisphere and postcontusional abnormalities of the left frontal lobe. He has a left-sided hemiparesis, neglect, and hemianopia. He has learning disabilities and a behavioral disorder. He suffers from simple partial seizures with a tingling sensation of the left side of his face and startle-provoked tonic seizures with posturing of left arm and deviation of head and eyes to the left. The startle-provoked seizures increased in frequency from the age of 7 and were refractory. He uses oxcarbazepine and clobazam. During the work-up for epilepsy surgery, treatment alternative with sound generators was tried and proved effective but not curative. His seizure frequency decreased from several seizures a day prior to the sound generator therapy to one seizure a day during the follow-up period of six months at the child neurology outpatient clinic. He subsequently underwent a temporoparietooccipital disconnection in the right hemisphere at age nine.

2.4. Patient 4

Patient 4 is a boy, born at term, of healthy parents; pregnancy was uneventful but delivery was complicated by peripartal asphyxia. He suffers from infantile encephalopathy with learning disabilities, a predominantly left-sided tetraplegia, and localization-related epilepsy. Magnetic resonance imaging reveals ischemic lesions in the right hemisphere and the left frontal lobe. From the age of eight, he has suffered from complex partial seizures of the left part of the body; initially, he responded well to antiepileptic drugs. After two years, the seizures became startle-provoked; unexpected sounds gave rise to increase in seizure frequency and seizure severity. Tonic seizures occurred at least 3 times a day, sometimes in a cluster of more than an hour despite the use of levetiracetam, lamotrigine, and clobazam. He was referred to our audiological team in parallel with the work-up for epilepsy surgery. Treatment alternative with sound generators had no effect on seizure frequency. Functional right-sided hemispherectomy was performed at age fourteen.

In two boys (patients 1 and 2), seizures could also be triggered by unexpected visual stimuli. Seizures tend to occur with high frequency, more than one seizure a day, exerting a considerable impact on daily life and psychomotor development. Clinical seizure pattern involved asymmetric tonic posturing in all four children. Three children demonstrated several seizure types including focal seizure onset. All children had focal neurological signs or learning disabilities or a combination of both. Auditory examination revealed normal auditory functions in all four. (For a more detailed description of case history, see Table 1.)

2.5. Intervention

All four children and their parents were counseled by the same team of audiologists and followed a tailor-made treatment protocol for each child adapted from ‘the specialised care treatment protocol’ of Cima et al. [11]. After a simple, understandable explanation of the auditory system, the purpose of the sound generators was explained.

The sound generator is a Beltone TBR (tinnitus breaker) 62D, non-occluding open fit. This small instrument does not block the external auditory canal. A sound generator produces a preset, broadband, background sound just audible in silence. This background sound does not disturb conversation, nor does it attract the child's attention continuously. The sound generators are fitted in both ears.

The sound produced by the sound generators does not prevent or mask startle-inducing sounds. During several visits, the frequency spectrum of the noise produced by the sound generators is adjusted to create a sound in which the child recognizes a known pleasant sound (e.g., whispering creek or rustling wind through the trees). The intensity of the sound can be heard when it is focussed on in silence, but the aim is for the child to become accustomed to it. The sound generators should create a relaxing mood and offer a feeling of control over the auditory environment.

2.6. Summary

Seizure frequency reduction of 50% or more occurred after appropriate fitting and counseling by the audiologists in two out of four children. One of the nonresponders repeatedly removed the sound generators. Two boys, one of whom experienced no effect at all from the intervention, successfully underwent epilepsy surgery.

3. Discussion

Lesions responsible for SPES are highly diverse including epileptogenic lesions of supplementary motor and sensorimotor areas and primary sensorimotor, premotor, and perisylvian cortices [1–4]. Emotional and psychodynamic factors also play a role in the pathogenesis of SPES. In all four children, seizures were precipitated by an acoustic