Principles and Practice of Clinical Electrophysiology of Vision

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Visually Evoked Cortical Potentials in Cortical Blindness

Emiko Adachi-Usami

"Cortical blindness" is bilateral visual loss due to dysfunction of both occipital lobes. It is diagnosed on the basis of behavioral observations that reflect problems in seeing, even though the patients can hardly describe their visual loss. Therefore, laboratory tests such as computed tomography, magnetic resonance imaging, electroencephalography, and visual evoked cortical potentials (VECP) must be relied on to provide the diagnosis of cortical blindness.

Among such objective tests, the VECP has raised the hope that it could be used to quantify functional visual loss because correspondences between subjective visual functions such as visual acuity, color vision, and central visual field defects and the VECP have been reported to occur. However, the results appearing in the literature are still in conflict. In the present chapter, the VECP and cortical blindness will be described.

GENERAL CLINICAL VISUAL SIGNS

In textbooks, visual acuity loss in cortical blindness is described as being total in both eyes. However, when we carefully read published case reports, descriptions of visual acuity even during the recovery stage do not sufficiently clarify whether the patients are still totally blind or not because expressions are used such as "light perception" and "counting fingers," which depend on the patients' behavior. Nonetheless, visual agnosia is a characteristic sign of cortical blindness. As a result, the definitive patterns of visual dysfunction such as color sense, binocularity, spatial sense, and macular sparing are obscure. On the other hand, pupillary light reflexes and ocular movements generally remain normal.

CAUSES OF CORTICAL BLINDNESS

The most common cause of cortical blindness is generalized cerebral hypoxia at the striate, parietal, and premotor regions, as well as vascular lesions of the striate cortex. Cerebral hypoxia can be caused by intoxication with carbon monoxide or nitrogen oxide and by inflammation such as meningitis, encephalitis, vascular occlusion, trauma, and so on. It occurs secondarily to transtentorial herniation, hemodialysis, hypoglycemia, and congenital malformations.

In any case, hypoxia is the final result.

VISUAL EVOKED CORTICAL POTENTIALS IN CORTICAL BLINDNESS

The VECP is generally considered to originate from central retinogeniculocalcarine pathways. However, the involvement of extrageniculate pathways cannot be completely ruled out. Therefore, VECPs in cortical blindness have received considerable attention. However, there is still no agreement about the VECP findings.

In the majority of published papers, VECP studies were done with flash stimulation. With the recent advances of technique in recording VECPs, it is generally said that the evaluation of flash VECPs is not as reliable as that of pattern VECPs. For example, Hess et al.[11] found in four patients with acute occipital blindness that no pattern VECP could be obtained, but a flash VECP was recorded. They concluded that the flash method was not appropriate for differentiating occipital blindness from psychogenic visual disorders.

Nevertheless, flash VECP is still being used effectively for patients who cannot fixate on the stimulus field such as in cortical blindness, infants, mentally retarded children, and unconscious patients.

The studies described below are concerned mainly with flash VECPs; their results are classified simply as normal, abnormal, and recovering.

Works Reporting Normal Visual Evoked Cortical Potentials

Spehlmann et al.[18] reported a 66-year-old patient with cortical blindness caused by numerous bilateral cerebral infarcts; no light perception was reported, but the patient showed flash VECPs of normal amplitude on repeated examinations.

Frank and Torres[10] recorded flash VECPs in 30 children with cortical blindness and found no significant differences between the patients and age-matched children with central nervous system diseases but without blindness. Only 1 patient with encephalopathy and increased intracranial pressure showed no response. As described above, Hess et al.[11] found normal flash VECPs and an absence of pattern VECPs. Normal flash *and* pattern VECPs were reported by Celesia et al.[6] in a 72-year-old patient who had infarction in bilateral areas 17 and part of area 18. They concluded that VECPs are mediated by extrageniculocalcarine pathways.

Newton et al.[16] reported a 16-month-old child with cortical blindness following *Haemophilus influenzae* meningitis. The flash VECP was normal, as were the fundi. Using both flash and pattern stimuli, Celesia et al.[7] found that VECPs were preserved, and positron-emission tomography showed a functioning island of occipital cortex that most likely represented the generator of the VECP.

These reports may support the evidence that extrageniculate pathways are also involved in the generation of flash VECPs. However, as Hoyt[12] pointed out, although the second visual system may be capable of mediating VECPs in some cases, it does not seem to be capable of sustaining any kind of cognitive vision.

Works Reporting Abnormal Visual Evoked Cortical Potentials

Because of interindividual variations of flash VECP waves and poor cooperation or fixation of the patient, it is hard to make a definite diagnosis of an abnormal response. Careful studies that demonstrate the abnormality of VECPs have been reported by a number of authors.

Kooi and Sharbrough[13] reported a case with post-traumatic cortical blindness whose flash VECPs were abnormal, with none of the normal initial five waves being identifiable, while the vertex potential was recordable.

Regan et al.[17] followed an infant for 15 months whose cortical blindness had presumably begun at the age of 3.5 months. VECPs recorded at 4 months were monophasic, and the latency was prolonged; the VECP waves grew progressively more complex with age. However, recovery could be anticipated from the VECP development.

Chisholm[8] reported a case of cortical blindness due to bilateral occipital infarction and found VECPs to be absent.

Aldrich et al.[2] found that flash or pattern VECPs recorded during blindness were abnormal in 15 of 19 patients but were not correlated with visual loss.

Works Reporting the Recovery of Visual Evoked Cortical Potentials in Accordance With Visual Improvements

Several authors reported that VECP improvement paralleled vision recovery. Barnet et al.,[3] in six clinically blind patients, observed that flash VECPs were depressed in three of them and that in two others the VECPs were preserved several days before visual improvement became evident. Duchowny et al.[9] reported that changes in short-latency VECP components were correlated with visual ability. However, up to the present, there is no irrefutable evidence that short-latency components are related to the striate cortex.

A recent work by Makino et al.[14] described in a follow-up study that the flash VECP configuration became normal with the passage of time. Miyata et al.[15] studied a case of transient cortical blindness caused by recurrent hepatic encephalopathy and found prolonged latency and a reduced amplitude of the second wave when the patient lost vision completely but a return to normal values after treatment.

Two other papers[1, 4] pointed out that either flash or pattern VECPs were present in normal configurations when the patient could see well and that they were nonrecordable when the patient claimed no vision. The configuration of the VECPs might be a criterion for evaluating the abnormality of VECPs, even though it is nonspecific.

Recently, Bodis-Wollner and Mylin,[5] using VECPs of monocular and dynamic random-dot pattern stimuli, found that the recovery of binocular vision was delayed in comparison to the recovery of monocular vision. They concluded that it was not due to simple acuity impairment or convergence deficiency and thus provided evidence for the vulnerability of postsynaptic cortical mechanisms of human binocular vision.

CONCLUSION

As mentioned above, the VECP findings in cortical blindness are still controversial. There are two reasons for this. One is that the patient's visual loss cannot be quantitatively determined by subjective testing of visual acuity, binocular vision, color sense, and spatial vision because of the uncertainty of the patient's responses. It is therefore hard to make a comparison between the VECP results and the subjective and clinical visual signs.

Another reason is that the pathological lesions that cause cortical blindness are not often localized in the striate cortex or extrastriate cortex but spread widely throughout the parietal and temporal regions.

In any case, the theme of the relationship between cortical blindness and VECPs is fascinating, at least from the point of view of study of the origin of VECPs and the hope of differentiating cortical blindness from psychogenic visual disturbances.

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