Official Journal of the European Paediatric Neurology Society

Original article

Sweep visually evoked potentials and visual findings in children with West syndrome

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ARTICLE INFO

Article history:
Received 19 March 2013
Received in revised form
4 October 2013
Accepted 11 November 2013

Keywords:
Sweep visual evoked potentials
Childhood epilepsy
West syndrome
Grating visual acuity
Cortical visual impairment
Nystagmus
Strabismus

ABSTRACT

Background: West syndrome (WS) is a type of early childhood epilepsy characterized by progressive neurological development deterioration that includes vision.
Aim: To demonstrate the clinical importance of grating visual acuity thresholds (GVA) measurement by sweep visually evoked potentials technique (sweep-VEP) as a reliable tool for evaluation of the visual cortex status in WS children.
Methods: This is a retrospective study of the best-corrected binocular GVA and ophthalmological features of WS children referred for the Laboratory of Clinical Electrophysiology of Vision of UNIFESP from 1998 to 2012 (Committee on Ethics in Research of UNIFESP n° 0349/08). The GVA deficit was calculated by subtracting binocular GVA score (logMAR units) of each patient from the median values of age norms from our own lab and classified as mild (0.1–0.39 logMAR), moderate (0.40–0.80 logMAR) or severe (>0.81 logMAR). Associated ophthalmological features were also described.
Results: Data from 30 WS children (age from 6 to 108 months, median = 14.5 months, mean ± SD = 22.0 ± 22.1 months; 19 male) were analyzed. The majority presented severe GVA deficit (0.15–1.44 logMAR; mean ± SD = 0.82 ± 0.32 logMAR; median = 0.82 logMAR), poor visual behavior, high prevalence of strabismus and great variability in ocular positioning. The GVA deficit did not vary according to gender ($P = .8022$), WS type ($P = .908$), birth age ($P = .2881$), perinatal oxygenation ($P = .7692$), visual behavior ($P = .8789$), ocular motility ($P = .1821$), nystagmus ($P = .2868$), risk of drug-induced retinopathy ($P = .4632$) and participation in early visual stimulation therapy ($P = .9010$).
Conclusions: The sweep-VEP technique is a reliable tool to classify visual system impairment in WS children, in agreement with the poor visual behavior exhibited by them.

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http://dx.doi.org/10.1016/j.ejpn.2013.11.008

1. Introduction

West syndrome (WS) is a rare type of early childhood epilepsy and a potential cause of cortical visual impairment (CVI) in infants.[1–9] This syndrome is characterized by infantile spasmus associated to progressive deterioration of cognition, mental retardation and disruption of cortical bioelectrical activity, characterized by delta and theta rhythms of large amplitudes and multifocal spikes superimposed in all derivations on EEG records, a disorganized and chaotic pattern known as hypsarrhythmia.[10,11]

The first symptoms of WS are usually manifested during the first year of life, from the sixth to the ninth month of age, with an incidence of 3–6:10,000 births.[12] This age of onset parallels a critical period required to proper maturity of visual pathway, including both retina and the visual cortex, comprehended from birth to the third year of life.[13] At this age, about a third of WS infants is not suspect to have epilepsy when first seen by a medical practitioner[14] and visual disabilities are hardly recognized as well.[2]

The WS can be etiologically classified as cryptogenic/idiopathic or symptomatic.[14] The most part of the causes associated to symptomatic WS is also commonly associated to CVI, such as sequelae of hypoxic ischemic encephalopathy, cortical immaturity, neurometabolic and genetic diseases mainly tuberous sclerosis, and Aicardi's.[15] Congenital abnormalities[5,6] of the brain and occipital cortex such as tuberous sclerosis, hemimegalencephaly, lissencephaly, agenesis of the corpus callosum, focal displasias and arteriovenous malformation, Down syndrome[16] and "self-induction" by periorbital stimulation[17] can also lead to symptomatic WS. The prognosis of WS is devastating, with a neurologic outcome that varies with epilepsy control (cessation of spasms and hypsarrhythmia) by oral administration of anti-epilepsy drugs,[10,11,18] meanly valproic acid. However, despite the benefits related to seizure control, it is known that the most part of the "gold standard" anti-epilepsy drugs currently in use alters GABAergic system, leading to undesirable retina toxicity and disturbance of the visual cortex manifested as bilateral visual field defects and decrease in grating acuities measurements, as reported to the use of vigabatrin, lamotrigine and topiramate.[19–24] The adrenocorticotrophic hormone (ACTH), which is not toxic to the neurological tissues, has provided a better control of the brain activity; however, iatrogenic systemic arterial hypertension limits the use of such medication.[25]

Nowadays, refined psychophysical tests and electrophysiological methods allow objective evaluations of the function of the visual pathway of patients who were not able to verbalize the visual perception information, including both preverbal children and those mentally handicapped. One of these tools is the objective measurement of the grating visual acuity threshold (GVA) elicited by sweep visually evoked potentials (s-VEP).[13,26,27]

The aim of this study is to demonstrate the applicability of the s-VEP to emit a sensorial evaluation of the status of the visual cortex in WS based on GVA measurement and its deficit estimation (GVAD) in relation to a normal population. Such technique allowed us to characterize the potential visual loss associated to WS, improving the ophthalmological evaluation of this group of patients, in spite of any verbal information and minimal cooperation commonly provided by them.

2. Methods

2.1. Participants and sample selection

This is a retrospective study of the records from a group of thirty children with WS who were referred for GVA measurement by the s-VEP technique to the Laboratory of Clinical Electrophysiology of Vision at the Universidade Federal de São Paulo (UNIFESP) from July/1999 to October/2012. This study followed the tenets of the Declaration of Helsinki and the Institutional Review Board previously approval (Committee on Ethics in Research of UNIFESP n° 0349/08).

The study inclusion criteria were children, from zero to 120 months of age, both male and female with the diagnosis of WS well-established by a child neurologist according to the classification criteria of epileptic seizures and syndromes and assigned as cryptogenic (absence of any underlying cause) or symptomatic (when associated to other brain disease).[28,14]

Symptomatic patients were grouped according to a previous classification criteria[29] as follows: children presenting any abnormalities of the brain development such as schizencephaly, lissencephaly, microcephaly, agenesis of the corpus callosum, Aicardi's disease and other genetic syndromes were classified as brain abnormal developmental cause; children presenting normal brain development with a confirmed or presumed perinatal insult such as hypoxia, prematurity, hypoglycemia, fetal tocotrauma, hemorrhage and central nervous system infections were classified perinatal cause; children presenting normal brain development with a confirmed postnatal insult such as hemorrhage and central nervous system infections, vascular occlusions, hypoglycemia, trauma, shock and heart attack were classified as postnatal cause.

All neuroimaging features were described in Table 1.

Children who presented congenital or drug-induced cataract or any other abnormality of the ocular anterior segment (ocular trauma, leucoma, ptosis, aniridia, glaucoma, congenital infections or uveitis) were excluded. Fundus abnormalities were described in Table 2.

2.2. Ophthalmologic evaluation

Ophthalmologic evaluation consisted of age-appropriate visual assessment including visual tracking ability, ocular positioning, presence of strabismus and/or nystagmus, external examination of the eyes and fundoscopy. During the evaluation, all children were awake and alert.

Evaluation of ocular motility consisted by testing the child's ability to maintain fixation on an object or penlight and to pursuit a visual stimulus into different gaze positions (visual tracking). Such assessment had been performed both under binocular and monocular conditions.

Visual behavior was classified as "good" or "poor" according to the child's visual tracking ability (good) or disability

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