Vision Assessment of Nonverbal Patients

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ABSTRACT

Introduction: The most important aspect of a pediatric eye exam is the quantitative measurement of visual acuity, yet many children in a pediatric practice are nonverbal or preverbal.

Methods: The clinician has a number of options for assessing vision in these children, including the central, steady, maintained technique, behavioral measurements, and electrophysiological measurements. Each of these approaches has advantages and disadvantages.

Results: All techniques for acuity measurement have their pros and cons. A newer technique, the sweep visual evoked potential (sVEP), can be used to measure different types of visual acuity (grating, vernier, contrast sensitivity) and also does not require any verbal responses.

Conclusion: Since vernier acuity offers a better approximation to Snellen acuity, the sVEP holds promise as a useful tool in the pediatric office and lab.

INTRODUCTION

Often, in the practice of pediatrics, the clinician confronts a child with poor motor control yet who has the possibility of measurably and useful vision. A patient’s inability to attend to a task—for example, to look at a card or a computer monitor—makes measures of vision difficult. The number of these children is increasing in frequency due to increased survival rates of premature infants—i.e., those most likely to experience cerebral palsy and strabismus. Other times, the child is preverbal or nonverbal, yet at risk for vision impairment in one or both eyes.

The clinician’s armamentarium for quantitatively measuring a child’s vision is expanding. Visual acuity measures in children can be performed using a number of procedures, each with advantages and disadvantages. In this report, we describe these various procedures, with a special

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emphasis on the steady state visual evoked potentials (sVEP) as a tool for measuring vision in nonverbal patients. Common to all procedures is their dependence on a reasonably cooperative patient.

CENTRAL-STEADY-MAINTAINED (CSM) PROCEDURE

In the clinic, a behavioral technique that uses central fixation (visual axis looks directly at object of regard), steady fixation (presence or absence of nystagmus), and maintenance of fixation (in strabismic patient, fixation on target of interest is maintained under binocular viewing conditions, or is not maintained, indicating possible amblyopia).1 In the presence of strabismus, the CSM test is reliable and predictive of visual acuity.

The term steady, as used in the CSM procedure, indicates whether nystagmus is present. If present, visual acuity is usually reduced, depending on the cause of nystagmus. On the other hand, in situations where low vision is identified, the presence of nystagmus implies a likelihood of at least some pattern vision—i.e., the ability to see shapes.

The central/steady/maintained (CSM) procedure can be coupled with a vertical prism test to determine presence of amblyopia when no obvious strabismus is present. In this procedure, a vertical prism is placed in front of one of the eyes (e.g., 10° base down). Since the prism degrades visual acuity somewhat, the patient will choose to view a fixation target with the eye that is not behind the prism. Should the patient prefer fixation with the eye behind the prism, this suggests that the fellow eye has amblyopia.2,3

PSYCHOPHYSICAL MEASURES OF VISION

Behavioral techniques that rely on preferential looking are also useful in the clinic, and have proven of great value in clinical trials and studies of pediatric eye diseases. The prototype of these techniques is referred to as the Teller Acuity Card Procedure, a procedure used widely in clinical trials and which measures grating acuity thresholds.4–9 In short, the patient is positioned in an environment devoid of distractions, and is shown two cards, one to his/her left, and the other to his right. One of the cards is a blank, and the other has grating lines. Children will choose to look at the card with lines, and either turn their head, and or their eyes to the lined card. Cards with progressively increasing spatial frequency are shown to the child, until the card reaches the child’s grating acuity threshold, at which point no behavioral sign occurs. The spatial frequency of the card last seen is the child’s threshold for grating acuity.

This procedure for measuring grating acuity has much to recommend it. The test is inexpensive, portable, easy to administer, reliable, and well-validated in many populations of children.10–13 A drawback to grating acuity measures, and this is particularly true for amblyopia, is that grating acuity thresholds tend to overestimate acuity (i.e., underestimate the severity of amblyopia).14

Children taking the test, of course, have to be able to show the behavior indicating that they see the card. Thus the child with significant cerebral palsy, for example, will have difficulty demonstrating that he/she sees the card, even though vision may be present. Even so, grating acuity measures are comparable when performed behaviorally or electrophysiologically, even in children with cortical visual impairment.15

Behavioral measures of vernier acuity for use in preverbal children have also been developed.16,17 As described below, vernier thresholds are of some interest, because vernier acuity more accurately correlates with Snellen acuity. A variety of psychophysical measures of vernier acuity are
available to test adult observers. Generally, these tests require the patient to indicate, either verbally or by pushing a button, that the target’s offsets are visible.

ELECTROPHYSIOLOGY MEASURES OF VISION (VEP)

A third technique to measure acuity thresholds is an electrophysiological one. Here a visual stimulus is presented either in the form of a flash, or in a continuous fashion, over a period of a number of seconds. Traditional, transient (flash) VEP techniques are not well suited for measuring sensory thresholds.\textsuperscript{18, 19} In order to measure a threshold, one must present a substantial number of patterns that vary in their visibility. The accuracy of the threshold estimate depends in large measure on how many patterns are presented in the near-threshold region and on the signal to noise ratio (SNR) of the response. Traditional VEP techniques\textsuperscript{20, 21} have rather poor SNR and thus clinical acuity techniques based on them must rely on a relatively small number of patterns that are often presented well above threshold. Steady-state VEPs can be recorded at much higher SNR\textsuperscript{19}

Steady-state VEPs are generated by periodically modulating the contrast (or displacement) of the visual stimulus. The periodic stimulation forces the visual response to be periodic as well. In fact, the response to a periodic stimulation is confined to frequencies that are exact integer multiples of the known stimulation frequency. By forcing the response to occur at a small number of exactly known frequencies, one can use filtering techniques that are very selective against EEG noise. The high SNR afforded by steady-state techniques allows one to spend less time obtaining an accurate response estimate for a given stimulus condition. The time savings is significant enough in practice to allow the experimenter to measure the response to 10 or 20 different stimulus values presented over a period of 5–20 seconds.\textsuperscript{19, 22}

In the swept-parameter technique shown below (Figure 1), the stimulus is varied continuously from well above threshold to well below the expected threshold (grating and vernier examples), or well below threshold to well above threshold, (demonstrated in the contrast sensitivity part of the figure). The function that relates response amplitude to stimulus strength is

![Figure 1: Stimulus for recording vernier-related, grating, and contrast sensitivity VEP activity. For vernier acuity, the stimulus is alternated between two states. In the first state, the screen is covered with a bar grating. In the second state, portions of the screen are offset (several rows of offsets are used, only one is illustrated). The VEP response is larger in response to the transition between alignment and misalignment than it is for the return to alignment after misalignment. This response asymmetry produces odd harmonic power in the steady-state evoked response.](image)
used to estimate sensory threshold by extrapolation to zero-amplitude (Figure 2).

The sVEP can be used to measure vernier, contrast, luminance, and many other visual functions and activities, including more complex visual tasks such as contour integration. One of these vision functions, vernier acuity, is the ability to detect fine offsets. Since this ability to see offsets in lines exceeds predicted acuity, cortical factors likely are recruited for detection of offsets. Vernier acuity thresholds are particularly interesting, because these measures probably offer a better correlation with Snellen acuity than do other measures such as grating acuity.\textsuperscript{23, 24} If, indeed, vernier acuity thresholds do approximate Snellen acuity measures, then vernier acuity measurements could be particularly useful for the diagnosis and management of pediatric eye disorders, such as amblyopia.

Evidence is mounting that vernier acuity threshold when measured electrophysiologically corresponds to vernier acuity as measured psychophysically. For one, sVEP thresholds for vernier acuity occur in the hyperacuity range, when measured in the first harmonic. Vernier acuity thresholds are substantially worsened when offsets fail to align. A robust response is achieved in the second harmonic, though, and probably measures detection of stimulus motion. Comparisons of sVEP thresholds to psychophysical thresholds recorded from an identical stimulus in adult observers indicate that sVEP vernier threshold measures are comparable to psychophysical measures.

sVEP measures provide more information than simple threshold determinations. The signal amplitude can be evaluated and may indicate conditions in which numbers of neurons, or synapses are reduced, or cortical excitation/inhibition is altered. The most striking example of this comes from studies of children with cortical visual impairment (CVI), where reduced visual acuity is seen clinically, behaviorally, and electrophysiologically.\textsuperscript{15} sVEP signal amplitudes in children with CVI are reduced significantly. Conversely, we have seen that signal amplitudes are substantially increased in formerly pre-term but healthy infants, when compared to age-matched and healthy full-term infants.\textsuperscript{25}

Other attributes of the sVEP also lend themselves to analysis. For example, phase
shifts are monitored during the course of sVEP measurements, and could have implications for evaluating brain function during visual activities. Spectral edge can be evaluated (the point in temporal frequency below which the majority of the EEG signal occurs). Spectral edge findings could correlate with white matter disease. It is also possible to sweep other more complex tasks, such as the ability to detect contour in noise, and to measure stereopsis.

Nevertheless, there are disadvantages to the sVEP. The equipment is complicated and requires know-how to maintain and use. It is barely transportable, and therefore requires that patients come to a lab for measurements. The equipment often requires real-time support when in use. Grating acuity measured electrophysiologically overestimates acuity and underestimates amblyopia, similar to behavioral measures of grating acuity.

Questions as to whether the sVEP really measures vision, or whether it measures an electrophysiological epiphenomenon persist. sVEP acuity thresholds are usually lower (better) than are behaviorally measured thresholds, at any given age up to maturity of the visual function. The presence of an EEG signal induced by a visual stimulus does not necessarily indicate that the signal detects vision. However, experiments comparing electrophysiological measures to psychophysical measures in adults suggest that sVEP measures correlate with behavioral measures. Experiments must utilize adult subjects, and therefore indicate correlations in mature visual systems. It is reasonable to extrapolate to infants and children (and their less mature neurological system), but it will be difficult to correlate these two measures in nonverbal patients.

The clinician can tailor the approach to nonverbal patients accordingly. In clinical settings, the central, steady, maintained approach has much to recommend it. In cases where acuity problems are suspected but not confirmed by the CSM technique, a more advanced approach to assessment may be appropriate. Behavioral and electrophysiology techniques can help the clinician refine acuity measures, thereby offering children more targeted and precise treatment.

REFERENCES

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