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The assessment of visual acuity in children with periventricular damage: A comparison of behavioural and electrophysiological techniques

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Abstract

It has been controversial whether electrophysiology offers better precision than behavioural techniques in measuring visual acuity in children with brain damage. We investigated the concordance between sweep VEPs and Acuity Cards (AC) in 29 children with periventricular leukomalacia (PVL), the most common type of brain damage in preterm infants. An overall good correlation was shown but with relatively better behavioural acuity values. VEP/AC ratio was significantly correlated to corpus callosum posterior thinning. We propose that this result reflects the efficacy of the compensatory mechanisms following early brain damage which may differentially affect the two methods.

Keywords: VEP; Children; Brain lesion; Acuity; Periventricular leukomalacia

1. Introduction

Periventricular leukomalacia (PVL) consists of hypoxic-ischemic damage of periventricular white matter typical of preterm infants, usually occurring at the beginning of the third trimester of gestation (Volpe, 2003). The lesion generally involves the cortico-spinal tract, and is therefore associated with a motor impairment primarily affecting the lower limbs (Bax, Tydeman, & Flodmark, 2006). In most cases however, it also involves the optic radiations due to their particular distribution around the posterior horns of the lateral ventricles, giving rise to different types of visual disorders (Cioni et al., 1997; Jacobson, Ek, Fernell, Flodmark, & Broberger, 1996; Uggetti et al., 1996; van den Hout et al., 2004). At present, PVL is considered to be the major cause of visual impairment in prematurely born children (Jacobson, Lundin, Flodmark, & Ellstrom, 1998). The most frequent disorders are restriction of the visual field, difficulties in oculomotor behaviour and reduction of resolution acuity, often associated with a visual crowding effect that is more pronounced at near than at distance (Cioni et al., 1997; Eken et al., 1996; Fazzi et al., 2004; Fedrizzi et al., 1998; Jacobson, Ygge, Flodmark, & Ek, 2002; Jacobson et al., 1996; Lanzi et al., 1998; Scher, Dobson, Carpenter, & Guthrie, 1989).

Different techniques have been used for the early assessment of visual acuity in infants with brain lesions, including PVL. In several studies behavioural methods have been applied, based on the spontaneous preference of the infant for a complex pattern, as in the acuity card (AC) paradigm (Brown & Yamamoto, 1986; Searle, Horne, & Bourne, 1989; Spierer, Royzman, & Kuint, 2004). Other authors have instead used electrophysiological techniques, such as different types of visual evoked potentials (VEP) (Good, 2001; Good & Hou, 2006; Lim et al., 2005; Westall,

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Ainsworth, & Buncic, 2000). Abnormal results are usually found with both approaches in a significant proportion of subjects with congenital brain damage (Cioni et al., 1997; Fazzi et al., 2004; Jacobson et al., 2002; Lanzi et al., 1998). However, when the two methods are used in the same population, the results are often conflicting, possibly due to the heterogeneity of the populations assessed or to the specific characteristics of the techniques used (Bane & Birch, 1992; Mackie et al., 1995; Orel-Bixler, Haegerstrom-Portnoy, & Hall, 1989).

Behavioural and VEP measures of visual acuity are in fact feasible and deemed reliable and valid in infants and children with PVL, and both measures may be used in clinical practice. However, different results from the two techniques can be found as VEP acuity mainly depends on the integrity of the pathway from the eye to the visual cortex, while behavioural responses may involve higher processing and rely on additional factors such as attention and ocular motor abilities which may be affected in children with neurological disorders (Lim et al., 2005). In the present study we assessed the concordance between grating acuity tested with sweep VEPs and ACs in a group of children with PVL, also exploring the possible bases of discordant results.

2. Patients and methods

Twenty-nine children (age range 9 months–13 years) with a neuroradiological diagnosis of PVL, referred to the Stella Maris Scientific Institute in Pisa between 2004 and 2006, were selected for this study. Parental informed written consent for the study was obtained in all cases. The research was approved by the Ethical Committee of the Stella Maris Foundation.

The type of cerebral palsy was defined according to the criteria of Hagberg et al. (Hagberg, Hagberg, & Olow, 1975). Subjects with severe ophthalmologic abnormalities (such as retinopathy of prematurity stage III or upwards, major refraction problems, optic nerve atrophy) were not included in the study. In all subjects, the following assessments were performed: (i) a behavioural and electrophysiological measure of visual acuity and (ii) a detailed evaluation of other aspects of visual function. In those subjects in which good quality MRI scans were available a quantitative/qualitative assessment of the characteristics of the lesion was also performed. Cognitive development was assessed by the Griffiths scales (Griffiths, 1984) in children below 3 years of age and by the Wechsler scales in the others (Wechsler, 1989, 1991).

2.1. Assessment of visual acuity

Visual acuity was assessed by means of the Teller Acuity Cards (TAC) (Teller, McDonald, Preston, Sebris, & Dobson, 1986) and the sweep VEP (Hamer, Norcia, Tyler, & Hsu-Winges, 1989; Norcia & Tyler, 1985a, 1985b; Norcia, Tyler, Piecuch, Clyman, & Grobstein, 1987). The assessments were performed within the same session in random order.

2.1.1. Teller acuity cards

This test is based on a preferential looking paradigm, i.e. the spontaneous behavioural reaction of newborns and infants consisting of gazing or turning of the head toward the most salient of two or more alternative visual stimuli (McDonald et al., 1985). The stimulus consists of a series of stripes (grating) displayed on cards, and the acuity value is defined by the smallest stripe width that consistently elicits a preferential looking response. The spatial frequency of the cards ranges from 0.32 to 38 cycles/cm with a difference of ½ an octave between one card and the next one. The viewing distance was 57 or 84 cm, according to the age of the patient, and mean luminance ranged from 12 to 16 cd/m^2. Acuity values are expressed in minutes of arc (or cycles per degree) and can be compared to normative data reported in the literature (Courage & Adams, 1990; Dobson & Teller, 1978; Hertz, 1987; Hertz & Rosenberg, 1992; Mohn, van Hof-van Duin, Fetter, de Groot, & Hage, 1988; van Hof-van Duin & Mohn, 1986).

2.1.2. Sweep visual evoked potentials

Stimulus generation and signal analysis were performed using the Power Diva system (Digital Instrumentation for Visual Assessment, Smith-Kettlewell Eye Research Institute, San Francisco, CA) on separate computers (PowerMacintosh G3 Apple) (Good, 2001; Norcia & Tyler, 1985a; Norcia & Tyler, 1985b). Three active electrodes in a row were placed over the occipital pole at O1, Oz, and O2, each of them three cm apart from the other, with reference and ground electrodes placed on Cz and Pz, respectively, based on the International 10–20 system. Electrode impedance was between 3 and 10 kΩ and the EEG were amplified at a gain of 20,000 or 50,000 depending on the age of the subject. (Grass Model 12 amplifiers with analogue filter setting of 0.3 to 100 Hz. Measured at −6 dB points.) Stimuli were displayed on a CRT monitor (Mitsubishi Diamond-Pro 17″). The grating acuity stimulus consisted of a sine-wave vertical grating that alternated with a matched space-averaged luminance field displayed at 80% contrast (average luminance, 43 cd/m^2) at a rate of 3.76 Hz on–off modulation. The spatial frequency range was varied from 2 to 16\20 cycles/deg according to the age and the visual disability of the subject. The grating was swept from high to low spatial frequency during a 10-s trial. The viewing distance was 80 cm. We recorded 4–10 trials for each subject. Raw scalp potential recordings for each 10-s trial were digitalized to 16 bits precision and partitioned into 10 sequential epochs of 1 s duration (bins). For each bin, a recursive, least square algorithm was used to generate a series of complex-valued spectral coefficients representing the amplitude and phase of response components tuned to various harmonics of the stimulus frequency. We focused on the 1st and the 3rd harmonic for analysis. These spectral coefficients for each bin were averaged together across trials for each subject, channel and harmonic. Statistical significance was assessed based on a T^2_{circ} statistic, a phase sensitive, variance-normalized measure of mean amplitude. Response thresholds were calculated by regression of amplitudes from the bins where the response decreased linearly to the point of stimulus invisibility. The range of bins eligible for regression depended on the statistical significance and phase consistency of the response according to a robust algorithm (Norcia & Tyler, 1985b) that specifies four inclusion criteria: (1) response probability in each bin $\leqslant 0.16$, (2) phase consistency between consecutive bins between 80° and −100°, (3) at least a sequence of two consecutive bins showing responses of $P \leqslant 0.077$ and (4) any given bin should differ from both the preceding and the following by a factor of at least 0.3. This procedure, that matches exactly that used by Good and Hou (2006) with CVI patients, minimizes the artifacts arising from the VEP signal. Once the range was determined, the threshold value was established by extrapolating the regression line to 0 response amplitude. Brain electrical activity was recorded with Grass gold-cup electrodes placed on the scalp with a conductive gel. During a recording session, subjects sat alone or on a parent's lap in front of the monitor and an operator attracted their attention with a small toy on the center of the monitor. When the subjects were not attending to the stimuli recordings and sweeps (but not the stimulus appearance) were interrupted. When the stimulus resumed, its values were set as they were 0.5 s before the interruption. We did not use the first and last bin for analysis.

2.2. Assessment of other aspects of visual function

A full ophthalmological assessment was performed in all subjects exploring in particular the presence of ocular abnormalities, optic atrophy and refractive errors.

Oculomotor behaviour was assessed by means of the observation of fixation, following, and presence of abnormal eye movements, such as spontaneous nystagmus. Strabismus was tested by examining symmetry of the corneal light reflex and by the cover test.

Visual field size was assessed using kinetic perimetry (Mohn & van Hof-van Duin, 1983). During central fixation of a centrally positioned white ball, an identical target was moved from the periphery towards the fixation point along one arc of the perimeter. Eye and head movements towards the peripheral ball were used to estimate the outline of the visual field. Normative data for fullterm and preterm infants are available (van Hof-van Duin, Heersema, Groenendaal, Baerts, & Fetter, 1992).

On the basis of the results of the individual aspects explored, a global vision score was obtained according to the classification system of Randò et al. (Rando et al., 2004).

2.3. Assessment of brain damage

MRI were scored according to the following criteria:

(1) *Global lesion score.* The characteristics of the lesion were scored by means of the classification system of Cioni et al. who was found to be significantly associated with visual impairment in children with PVL (Cioni, Bartalena, Biagioni, Boldrini, & Canapicchi, 1992). The score is based on the following items: size of lateral ventricles, evidence and extension of white matter damage and of white matter loss, thinning of the corpus callosum, evidence and size of cystic areas, size of the subarachnoid space and abnormalities of cortical grey matter. Each item was scored on a three grade scale, a score of 3 indicating the most severe MRI abnormalities. The scores of the seven items were summed to obtain a total score for each infant. The total scores were also classified on a three grade scale (Table 1).

(2) *Global atrophy index.* An indirect index of global white matter damage was obtained as the ratio of the sum of ventricular body and extracerebral space to the whole brain surface (Fig. 1a), a quantitative index easily measurable from MRI scans of children with PVL (criteria modified Ito et al., 1996). The measure was performed on MRI axial scans at a level just above the foramen of Monro. A lower value indicates a greater atrophy.

(3) *Corpus callosum index.* An indirect index of posterior white matter damage was obtained as the ratio between the thickness of the genu and the thickness of the posterior part of the body (Fig. 1b) (modified from (Iai, Tanabe, Goto, Sugita, & Niimi, 1994). The measure was performed at midsagittal MRI scanned level and was previously found to be easily measurable from MRI scans of children with PVL and correlated with the extent of motor impairment (Iai et al., 1994). A higher value indicates a greater posterior thinning of the corpus callosum.

(4) The assessment of global brain damage was performed in 25/29 subjects who had good quality MRI scans. Quantitative measures were only possible in 21/25 due to the availability of the required sections.

2.4. Statistical analysis

The correlation between TAC and sVEP was calculated by means of the Pearson's parametric test for bivariate correlation while the comparison between the mean values at TAC and sVEP was done by means of the Student's *t*-test. Multiple linear regression analyses were performed to investigate the possible influence of different factors on TAC and VEP development, correlating these values and the index between the VEP/Teller ratio (which reflects the concordance of the two measures) to the anatomical indexes, gestational age at birth and age at test, the global vision score and subscores. A p value below .05 was taken as significant.

Table 1
Global lesion score (41)

#	Ventricle size	WM abn. SI	WM reduction	Cysts	Subarach. space	Corpus callosum	Cortical GM	Total score	Global grade
1	3	2	2	2	3	2	1	15	2
2	3	3	3	3	3	3	3	21	3
3	2	2	2	2	2	2	1	13	2
4	2	3	3	1	2	2	1	14	2
5	na	na	na	na	na	na	na	na	na
6	1	2	2	1	1	1	1	9	1
7	2	2	2	1	2	2	1	12	2
8	1	2	2	1	1	2	1	10	1
9	2	2	2	2	2	1	1	12	2
10	3	2	3	2	3	2	1	16	2
11	2	2	2	2	1	1	1	11	1
12	3	2	2	1	1	1	1	11	1
13	2	2	2	1	2	2	1	12	2
14	na	na	na	na	na	na	na	na	na
15	1	2	2	1	1	1	1	9	1
16	2	2	2	1	1	1	1	10	1
17	2	2	2	3	2	2	1	14	2
18	2	2	2	1	1	1	1	10	1
19	2	2	2	1	2	2	1	12	2
20	2	2	2	1	1	1	1	10	1
21	3	3	3	2	3	3	1	18	3
22	2	2	2	3	2	1	1	13	2
23	2	2	2	1	2	2	1	12	2
24	3	2	2	1	3	1	1	13	2
25	2	1	1	1	1	1	3	10	1
26	na	na	na	na	na	na	na	na	na
27	na	na	na	na	na	na	na	na	na
28	1	2	2	1	1	1	1	9	1
29	2	3	2	1	2	1	3	14	2

WM, white matter; abn, abnormal; SI, signal intensity; subarach, subarachnoidal; GM, gray matter; na, not available.

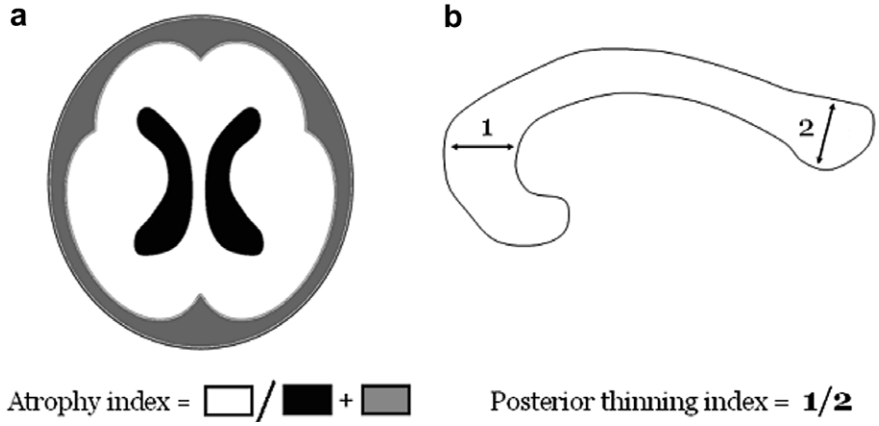

Fig. 1. Representation of global atrophy index (a) and corpus callosum posterior thinning indexes (b). (a) Black, ventricular body; gray, extracerebral space; white, whole brain surface. (b) 1, genu; 2, splenium.

3. Results

The clinical description of our cohort is reported in Table 2. Mean gestational age at birth was 32 weeks (range 25–40). At least one of the aspects of visual function assessed was abnormal in all subjects but one. Strabismus was present in 24 children (21 esotropia and 3 exotropia), oculomotor disorders in 21 and visual field reduction in 15. Ten subjects had an intelligence quotient (IQ) in the normal range, 12 presented a mild retardation and the remaining 7 a severe retardation. All subjects showed a cerebral palsy, which was a hemiplegia in one child, a diplegia in 15 and a tetraplegia in the remaining 13.

3.1. Visual acuity

To define normal and abnormal acuity, normative values from Courage and Adams (1990) were used. When visual acuity was assessed with the TAC, six subjects showed abnormal values (<2 SD), 12 showed borderline values (between 1 and 2 SD) while the remaining 11 were normal. With sweep VEPs, worse results were generally obtained: 11 subjects showed abnormal visual acuity, 10 borderline and only 7 showed normal values. In the remaining subject sweep VEPs were recorded twice but no reliable results were obtained due to the presence of excessive artefacts.

A highly significant correlation was found between TAC and sweep VEP, with a p value $< .001$ at Pearson test (Fig. 2). However, the difference between the mean acuity values obtained with TAC and sweep VEPs was statistically significant (Student's t-test; p value $= .011$). In 8/29 subjects the value of acuity measured by TAC was more than 50% higher than the measure obtained by sweep VEPs, and in two of the eight it was more than double. Conversely, only 2/29 subjects showed an acuity on sweep VEPs between 50% and 100% higher than the value obtained on the TAC.

3.2. Correlation between visual acuity and clinical features

TACs were significantly correlated with all the visual subscores, except for strabismus and visual fields, with gestational age at birth ($p = .021$) and with chronological age ($p = .037$). A significant correlation was also observed with several aspects of brain damage such as the thinning of corpus callosum ($p = .022$), the subscore of global lesion classification assessing the size of cystic area ($p = .029$), and the atrophy index ($p = .025$).

Sweep VEPs were significantly correlated with all the visual subscores, except for strabismus and visual fields, with thinning of corpus callosum ($p = .025$) and size of lateral ventricles ($p = .019$).

3.3. Correlation between VEP/TAC ratio and clinical features

When VEP/TAC ratio was tested for simple linear correlations against markers of brain damage, the strongest correlation was found with the corpus callosum posterior thinning index ($r = -.63$; $p = .004$) (Fig. 3). A statistically significant correlation was also observed with the global atrophy index ($r = -.46$; $p = .05$). In contrast, VEP/TAC ratio did not correlate with global lesion score ($r = .13$; $p = .55$), gestational age at birth ($r = -.32$; $p = .13$), age at test ($r = -.24$; $p = .24$) and visual score ($r = -.24$, $p = .23$). Multiple linear regression analyses were performed to assess the independent effects of markers of lesion on VEP/TAC ratio. These markers included global lesion score, corpus callosum posterior thinning index and global atrophy index. The adjusted R2 of the model was 0.53 ($p = .005$). After adjusting for the other lesion indexes, a statistically significant correlation was only observed with the corpus callosum posterior thinning index ($p = .001$), while significant correlations were not found for the atrophy index ($p = .36$) or for the global lesion score ($p = .69$). As TAC were significantly correlated to age at

Table 2
Clinical data

N.	Assessment Age (ms)	GA	Weight	Neurological Diagnosis	Cognitive level	Strabismus	Ocular Motricity	*Visual Field*	*TAC*	Sweep VEP	TAC cycles/deg	Sweep VEP
1	9	29	1315	T	Mild MR	EI	Ab	R	●	●	6.5	6.64
2	11	26	750	T	Severe MR	EI	Ab	N	●	●	6.5	6.81
3	15	40	2550	T	Mild MR	EI	Ab	N	○	○	11.4	11.2
4	16	28	1210	D	Normal	E	Ab	N	○	○	13	12.58
5	19	28	1070	D	Normal	E	N	N	◐	◐	10.3	19.89
6	20	29	1380	D	Normal	EI	N	N	◐	◐	13	12.3
7	20	28	1020	T	Mild MR	EI	Ab	R	○	◐	12	7.59
8	24	31	1500	T	Severe MR	EI	Ab	R	◐	○	9.8	0
9	27	37	2500	T	Normal	EI	Ab	R	◐	◐	11.4	10.31
10	32	32	1080	T	Severe MR	XI	Ab	R	○	○	18.2	15.63
11	33	28	750	D	Mild MR	EI	Ab	N	●	●	9.1	9.47
12	35	36	1250	D	Mild MR	EI	N	R	◐	●	13	11.33
13	36	32	1300	D	Mild MR	EI	Ab	R	◐	●	19	10.83
14	37	40	3070	T	Severe MR	XI	Ab	R	●	●	0.8	0
15	40	33	1700	D	Normal	EI	Ab	R	○	○	16	16.29
16	44	40	1050	D	Normal	\	N	N	○	○	26	20
17	45	32	1650	T	Mild MR	EI	Ab	R	◐	◐	11.4	10.98
18	48	37	1120	T	Normal	EI	Ab	N	◐	◐	13	16.43
19	49	33	2480	D	Mild MR	EI	Ab	R	○	–	20	na
20	58	30	1050	D	Normal	EI	N	R	○	◐	26	11.06
21	61	25	630	T	Severe MR	EI	Ab	R	◐	◐	0	0
22	67	32	1250	D	Normal	\	N	N	◐	◐	16	9.23
23	78	27	1200	T	Severe MR	EI	Ab	R	●	◐	4.2	7.64
24	90	30	950	T	Mild MR	EI	Ab	R	◐	●	13	8.57
25	139	26	1390	D	Mild MR	\	Ab	N	●	●	9.1	9.03
26	147	32	1230	D	Mild MR	\	Ab	N	◐	●	15.8	9.53
27	155	40	2100	H	Normal	\	N	N	○	●	26	14.64
28	156	39	3010	D	Severe MR	EI	N	N	○	●	24	9.63
29	156	33	1400	D	Mild MR	XI	Ab	N	○	○	19	17.07

GA = gestational age; **MR** = mental retardation; **E** = esotropia; **T** = Tetraplegia; **D** = Diplegia; **H** = Hemiplegia; **E(I)** = Intermittent Esotropia; **X** = exotropia; **X(I)** = intermittent exotropia; **N** = normal; **Ab** = abnormal; **R** = reduction ● = abnormal; ◐ = visual acuity between 1 and 2 SD; ○ = normal; na = not available.

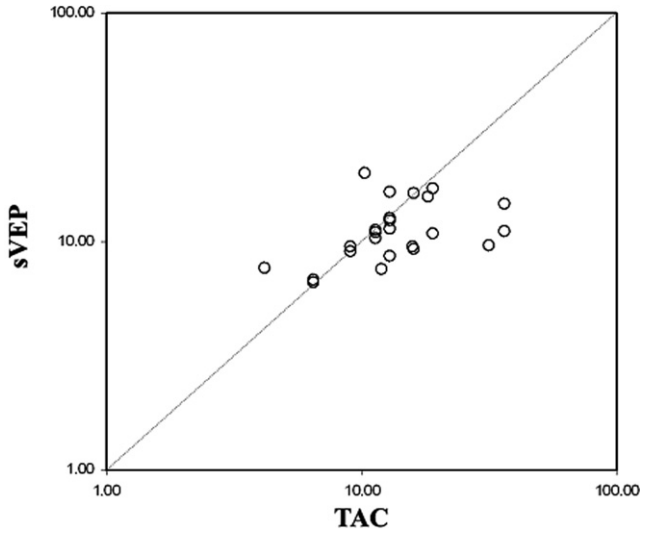

Fig. 2. Correlation between TAC and sweep VEP values.

test, a second multiple regression analysis was performed adjusting for the age at test, reaching analogous results.

4. Discussion

The main purpose of the study was to explore the degree of concordance between electrophysiological and behavioural measures of visual acuity in children with PVL. These two methodologies are commonly used in young infants or non cooperative patients as they are based on passive responses or spontaneous reactions. In the present study we used the sweep VEP technique, an electrophysiological method particularly useful in paediatric population because of the short time of execution, and the Teller ACs, one of the most widely used behavioural techniques in children with brain damage and cerebral visual impairment.

In accordance with previous reports comparing electrophysiological and behavioural measures of visual acuity in healthy infants and clinical populations (Arai, Katsumi, Paranhos, Lopes De Faria, & Hirose, 1997; Bane & Birch, 1992; Katsumi, Mehta, Larson-Park, Skladzien, & Hirose, 1994; Mackie et al., 1995; Orel-Bixler et al., 1989; Wiener, Wellish, Nelson, & Kupersmith, 1985), we found significant correlation coefficients between the two methods in PVL children. However, when analysing the difference of the means for the two groups of measures, Teller acuities were significantly higher than VEP acuities, in contrast with most previous studies in children with other types of congenital brain damage. In several cohorts of children with CVI, often secondary to hypoxic-ischaemic encephalopathy at term, visual acuities were reported to be higher when assessed with VEPs, than with behavioural tests (Good, 2001; Lim et al., 2005). A similar pattern was also observed in children with multiple neurological deficits by Orel-Bixler and co-workers, who found a better VEP acuity in 12/41 assessed patients as opposed to a better preferential looking (PL) acuity in only 2/41 (Orel-Bixler et al., 1989). This pattern has been interpreted as a result of the interrelation between the type of damage and the specific mechanisms underlying visual responses explored by the two techniques, that are thought to involve the central nervous system at different levels. VEPs are time-locked responses to visual stimuli assessing pathway conduction and cortical activity of the primary visual cortex, and are thus mainly dependent on the integrity of central visual structures (Hoyt, 1984; Smith, 1984). Preferential looking techniques are based on behavioural responses with gaze/attention shift and target exploration, and thus also require processing in higher visual associative areas or attentional networks. Consistent with this hypothesis is the finding of a relative prevalence of VEP over PL acuity in normal developing infants, as a result of the higher complexity of the behavioural task, with PL acuity reaching VEP values only around 14 months post-term (Riddell et al., 1997). The different rate of maturation of the two responses has been interpreted as related to the process of myelination, which is faster in the optic radiations compared to extrastriatal visual pathways (Barkovich, Kjos, Jackson, & Norman, 1988).

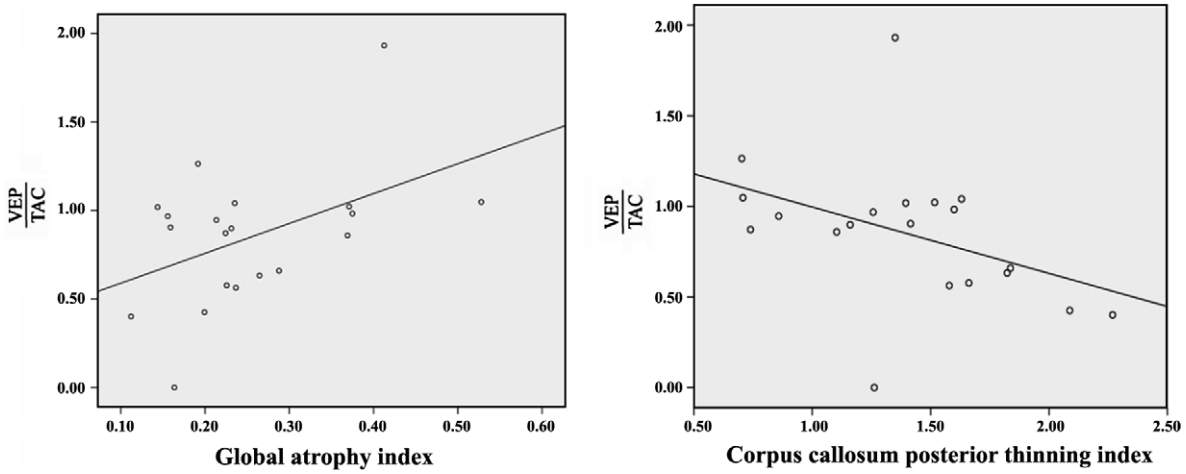

Fig. 3. Correlations between VEP/TAC ratio and brain damage.

In term-born children with hypoxic-ischemic encephalopathy or in those with multiple neurological handicap and CVI, a diffuse bilateral involvement of cortical brain structures is generally present. This is often associated with a global developmental delay and important attentional deficits, which are likely to affect to a greater extent the performance on behavioural than electrophysiological testing. This would explain the relative deficit of PL acuity, particularly in those children with greater developmental delay (Orel-Bixler et al., 1989). Our finding of an inverse pattern of distribution of acuity, i.e. PL acuity significantly better than VEP acuity, in children with PVL may be interpreted in the light of these reports. In fact, the typical site of brain damage in PVL children is the white matter around the posterior horns of the lateral ventricles, i.e. the region was the optic radiations are located. This may result in a direct damage of the geniculo-striate pathway, but with a relative sparing of cortical visual structures. As a consequence, it may be hypothesised that the plastic mechanisms of adaptive reorganization based on the reinforcement of the extra-striate visual pathways are virtually unconstrained and thus potentially able to produce a good recovery of function. In these conditions, the effects of brain reorganization might be better appreciated with behavioural procedures, that are likely to benefit more from the process of plasticity, as opposed to electrophysiological techniques, that are highly dependent on direct anatomical integrity of primary visual pathways. This interpretation is also consistent with animal studies showing a substantial sparing of visual orientation and pattern recognition in adult cats who underwent complete resection of the primary visual cortex at birth, as opposed to those operated later in life. Following early lesions, spared visual functions are remapped across the cortical surface and redistributed to a more spread network of functionally distinct areas of the visual system, which are able to compensate to a great extent for the primary visual brain damage (see, Payne & Lomber, 2002 for a review).

The high correlation we found between the VEP/Teller ratio, which reflects the concordance of the two measures, and the anatomical indexes of brain damage, may support the hypothesis of a differential sensitivity of the two techniques to periventricular brain damage. In fact, the relative prevalence of Teller acuity was found to be significantly correlated in particular to the degree of posterior thinning of the corpus callosum. We propose that in our cohort the relative hypotrophy of the splenium of the corpus callosum may represent an indirect measure of the integrity of the occipital cortex itself, as in this area the fibers connecting the two homologous occipital cortices are located (occipital-callosal fiber tracts) (Berardi, Bodis-Wollner, Fiorentini, Giuffre, & Morelli, 1989; Dougherty, Ben-Shachar, Bammer, Brewer, & Wandell, 2005; Dougherty et al., 2005; Saint-Amour, Lepore, Lassonde, & Guillemot, 2004). This is also supported, in the animal model, by the plastic changes in the occipital-callosal fiber tracts following visual impairment (Watroba, Buser, & Milleret, 2001). On the other hand, the degree of global atrophy might be related to the level of extra-striate visual pathway expansion, as suggested by numerous animal studies exploring plastic modifications and fiber restructuring in the visual system following a perinatal damage (see Payne & Lomber, 2002 for a review). In this view, the discrepancy between visual acuity assessed with electrophysiological or behavioural techniques might be interpreted as the effect of an involvement of the primary visual structures together with a sparing of extra-striate plastic potentials.

In conclusion, we showed a high correlation between electrophysiological and behavioural techniques in children with PVL, similar to what has been reported in other groups of children at high risk for CVI. However our results also suggest that behavioural measures can be, at least in PVL children, a better expression of visual functionality, as they reflect to a certain extent the efficacy of the compensatory mechanisms following brain damage (neural plasticity). This is consistent with animal studies showing how neural networks alternative to the primary visual pathways may be significantly enlarged and gives rise to a high degree of functional compensation. This proposal needs to be confirmed by the use in prospective studies of different imaging methodologies able to provide better and more direct measures of structural injury or pathway reorganisation, such as diffusion tensor imaging or voxel based morphometry.

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