# Nystagmus in periventricular leucomalacia

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**Abstract**

***Background/aims*—Periventricular leucomalacia (PVL) is a lesion in the immature brain involving the optic radiation. Children with PVL have visual problems including crowding, visual field defects, strabismus, and visual perceptual/cognitive deficits, together with nystagmus. They often have optic nerve hypoplasia seen either as small discs or as large cupping of normal sized optic discs. This study aimed to perform eye movement recordings in a group of children with PVL in order to characterise and classify the nystagmus.**

***Methods*—19 children with PVL on cerebral imaging underwent eye movement recordings with the Ober-2 infrared reflection technique.**

***Results*—16 of the 19 subjects had horizontal nystagmus.**

***Conclusion*—The present study shows that nystagmus is commonly seen in children with PVL.**

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Recent studies in children with periventricular leucomalacia (PVL) have revealed that many of these children exhibit a visual function disability including crowding, visual field defects, and visual/cognitive deficits.[1] Optic nerve anomalies associated with PVL has been described as optic nerve hypoplasia (ONH),[2] but also large cupping of normal sized optic discs, considered to be a variant of ONH.[3] In addition, the clinical impression has been that these children have nystagmus, strabismus, and other ocular motor disturbances.

Periventricular leucomalacia is a brain lesion caused by episodes of hypoxia/ischaemia at a gestational age of 24–34 weeks.[4] The lesion affects the corticospinal and/or the geniculostriate tracts, giving rise to spastic diplegia[5] and/or visual impairment.[1 6–9] Visual impairment caused by PVL is classified as a type of cerebral visual impairment (CVI).

Nystagmus has been reported absent in CVI[2 10] However, van Nieuwenhuizen[6] described latent or manifest nystagmus in 17 of 26 children having CVI but no ocular or optic nerve disorder. Jacobson and coworkers[1] also found nystagmus in many children in their study of the visual functions in PVL.

Early onset nystagmus is usually classified as congenital nystagmus (CN).[2] Although CN can occur without any known ocular anomaly (that is, congenital motor nystagmus),[11–13] several reports indicate that when performing a more extensive investigation, including electroretinography, in children with CN, this diagnosis seems to be linked to an ocular anomaly or an anterior visual pathway disorder more often than previously thought.[11–13] However, the presence or absence of an underlying sensory visual disorder cannot be predicted from the clinical features of the patient or the waveform of the nystagmus,[14] although certain waveform characteristics are commoner in children with ocular or anterior visual pathways disorders.[15] Congenital nystagmus includes up to 12 different waveforms,[15] with the majority of cases exhibiting either pendular or slow phase exponentially increasing velocity profiles. The vector is usually limited to the horizontal plane and the eyes move conjugately in direction, frequency, and amplitude. It can also have a latent component—that is, increased intensity (frequency and amplitude), when occluding one eye.

Latent/manifest latent nystagmus (LMLN) is mainly seen in children with infantile esotropia[2 16] and amblyopia. Strabismus seems to be essential in LMLN[14] and these children may also have a visual pathway disorder. LMLN can be distinguished from CN by the nystagmus waveform which usually has a slow phase decreasing velocity profile in LMLN. The nystagmus in LMLN is usually conjugate. It is present with both eyes open and increases in amplitude and frequency by occluding one eye. The slow phase is directed away from the fixating eye and the non-fixating eye may be either occluded (as in latent nystagmus, LN) or suppressed, although both eyes are open (as in manifest latent nystagmus, MLN).[14] Thus, the main difference between CN and LMLN is that in waveforms, and eye movement recordings are necessary to confirm the diagnosis of LMLN.

Because of our clinical observations of ocular motor disturbances such as nystagmus and strabismus in children with PVL, we performed eye movement recordings in a group of children with PVL to characterise and classify the nystagmus.

## Material and methods

### SUBJECTS

Nineteen children (nine boys and 10 girls; see Table 1 for details) participated in the present study. The children had either been referred to Tomteboda Resource Centre (TRC) in Stockholm, a Swedish national educational centre for visually impaired children (12 children) or recruited from patients with spastic diplegia, referred to the department of paediatric ophthalmology, Huddinge University Hospital because of strabismus or subnormal visual acuity when they were of preschool age (seven children). All of them had had their visual function assessed repeatedly since they were

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*Table 1 Clinical data*

| *Patient no* | *Sex* | *Year of birth* | *GA (weeks)* | *BW (g)* | *Duration of initial hospitalisation (days)* | *Duration of mechanical ventilation (days)* | *HC* |
|---|---|---|---|---|---|---|---|
| 1 | M | 1980 | 28 | 930 | 74 | 0 | − |
| 2 | M | 1982 | 30 | 1270 | 122 | 11 | + |
| 3 | F | 1986 | 26 | 1070 | 205 | 14 | + |
| 4 | F | 1986 | 26 | 710 | 350 | 183 | − |
| 5 | F | 1986 | 29 | 1000 | 68 | 2 | − |
| 6 | M | 1986 | 33 | 1900 | 40 | 0 | − |
| 7 | F | 1988 | 30 | 1350 | 59 | 3 | − |
| 8 | F | 1988 | 31 | 2075 | 23 | 2 | − |
| 9 | F | 1989 | 33 | 2640 | 23 | 0 | − |
| 10 | M | 1989 | 28 | 1260 | 90 | 19 | − |
| 11 | F | 1989 | U | 1350 | U | 0 | + |
| 12 | F | 1990 | 27 | 870 | 77 | 3 | − |
| 13 | M | 1977 | 33 | 1900 | 28 | 2 | − |
| 14 | M | 1986 | 30 | 1490 | 44 | 0 | − |
| 15 | F | 1986 | 35 | 1529 | 29 | 0 | − |
| 16 | F | 1986 | 28 | 1280 | 106 | 11 | + |
| 17 | M | 1989 | 29 | 1550 | 56 | 0 | − |
| 18 | M | 1990 | 29 | 1550 | 38 | 0 | − |
| 19 | M | 1990 | 34 | 1463 | 47 | 0 | − |

GA = gestational age at birth; BW = body weight in grams at birth; U = unknown (adopted child); HC = hydrocephalus. Patients 1–14 have low visual acuity; patients 15–20 have normal visual acuity.

4 years of age. The children were between 5 and 18 years old at the time of the ophthalmological investigations and eye movement registration session. They had a gestational age between 26 and 35 weeks (median 29.5 weeks) and their birth weight ranged between 710 and 2640 g (median 1350 g; Table 1). All children showed signs of PVL, visualised on computed tomography (CT) or magnetic resonance imaging (MRI; Table 2). Children with known retinopathy of prematurity (ROP) of greater severity than stage 2[17] were excluded. One child (patient 3) was included, although she had a cerebral lesion besides PVL (a leptomeningeal cyst of iatrogenic origin caused by a complication of a ventricular shunt earlier; this frontal lobe lesion was considered have no significance for the functions assessed in the present study). A neurodevelopmental assessment was performed in all children. Fifteen children had a normal intellectual level, three had mild mental retardation, and one had severe mental retardation. All children in the present study group showed an uneven cognitive profile, performing better in verbal than in visuospatial tasks. Fifteen children had a cerebral paresis syndrome (Table 2), as defined by Hagberg and coworkers,[18] while the remaining four did not. Thirteen of the 19 children had formerly been included in a previous study on optic nerve characteristics in PVL.[3]

### OPHTHALMOLOGICAL EXAMINATION

A neuro-ophthalmological examination was performed in all children. This included an assessment of linear optotype acuity at a distance, cover test, refraction in cycloplegia, ophthalmoscopy, fixation (visuscope), and evaluation of visual fields with the Stycar confrontation technique or Goldmann perimetry (Tables 3 and 4).

### CEREBRAL IMAGING

Cerebral imaging was performed with computed tomograph (CT) (12 subjects) or magnetic resonance imaging (MRI) (seven subjects). The technical quality of the images was judged very good in all patients. Although several examinations had been performed in most patients in the present study, the most recent examination was used for evaluation (Table 2). We consider the images to document a permanent end stage of the brain lesion since it was performed after completion of the myelinisation.[19]

### EYE MOVEMENT RECORDINGS

All children underwent binocular eye movement recordings (sampling frequency 500 Hz) with the Ober-2 infrared reflection technique. The child's head was fixated by a bite board and all recordings were made in dim light or a completely dark room. Targets were presented on a computer screen at a distance of about 45 cm. The target was in the form of a flashing dot measuring $3 \times 3$ mm, with a bright contrast to the otherwise dark screen. All children were first tested so they could see the target on the screen in all testing directions and the test was modified because of visual field defects in some

*Table 2 Neurodevelopmental data and neuroradiology findings*

| *Patient no* | *Motor function* | *Intellectual level* | *Cerebral image* | |
|---|---|---|---|---|
| | | | *Method* | *Severity and location of PVL* |
| 1 | mild cp, hpl R | norm | MRI | moderate, R>L, mid, post |
| 2 | moderate cp, dpl | norm | MRI | moderate, R>L, parietal and occipital atrophy, thin CC, shunt R |
| 3 | mild cp, hpl R | norm | CT | mild, R=L, shunt and frontal leptomeningeal cyst R side |
| 4 | no cp | MMR | CT | moderate, R<L, ant, mid, post |
| 5 | no cp | norm | MRI | moderate, R=L, mid, post |
| 6 | mild cp, dpl | norm | CT | moderate, R=L, mid, post |
| 7 | severe cp, dyst, R>L | SMR | CT | severe, R<L |
| 8 | severe cp, R>L | MMR | CT | severe, R=L, ant, mid, post |
| 9 | no cp | norm | MRI | moderate, R=L, mid, post |
| 10 | severe cp,dpl | MMR | CT | severe, R>L |
| 11 | severe cp, dpl | norm | CT | extremely severe, R>L |
| 12 | no cp | norm | MRI | moderate, R=L, post |
| 13 | moderate cp, dpl | norm | MRI | severe, R=L, mid, post |
| 14 | severe cp | norm | CT | severe, R=L |
| 15 | severe cp, dpl | norm | CT | severe, R=L, mid, post |
| 16 | moderate cp, dpl | norm | CT | extremely severe, R=L, ant, mid, post |
| 17 | mild cp, dpl | norm | CT | mild, R<L, mid, post |
| 18 | mild cp, dpl | norm | CT | moderate, R=L, mid, post |
| 19 | mild cp, dpl | norm | MRI | mild R, moderate L, mid, post |

cp = cerebral paresis; hpl = hemiplegia; dpl = diplegia; CT = computed tomography; MRI = magnetic resonance imaging; dyst = dystonia; PVL = periventricular leucomalacia; CC = corpus callosum; NT = not tested; norm = normal; ant, mid, and post refer to PVL location within the white matter; R = right side; L = left side; MMR = mild mental retardation; SMR = severe mental retardation.

*Table 3 Visual functions*

| Patient no | Age at assessment (years) | Refraction | Distant visual acuity | Visual fields |
|---|---|---|---|---|
| 1 | 14 | R −1.0<br>L −1.0 | R 6/30<br>L 3/60 | G: both hemifields R=L ++ |
| 2 | 10 | R +8.0 = −3.5/110°<br>L +6.5 = −2.5/80° | R 6/20<br>L 6/36 | G: hom quad L lower + |
| 3 | 8 | R +7.0 = −1.0/155°<br>L +8.0 = −3.0/30° | R 6/20<br>L 6/30 | both hemifields ++ R<L |
| 4 | 8 | R +2.25 = −0.75/110°<br>L +3.0 = −1.5/65° | Bin 4/60 | both hemifields ++ R<L |
| 5 | 9 | R ± 0<br>L +0.5 | R 6/60<br>L 6/36 | G: both hemifields ++, R=L |
| 6 | 8 | R +2.5 = −0.5/100°<br>L +3.5 | R 6/20<br>L 6/30 | both hemifields down ++ |
| 7 | 6 | R +1.5 = −0.5/0°<br>L +2.0 = −0.5/10° | Bin 6/60 | both hemifields ++ R<L ++ |
| 8 | 7 | R +2.0 = −1.0/90°<br>L +2.0 = −1.0/70° | R 6/36<br>L 6/60 | G: both hemifields ++ R>L |
| 9 | 5 | R +3.0 = −1.5/130°<br>L +3.5 = −1.5/50° | R 6/30<br>L 6/30 | both hemifields down R=L ++ |
| 10 | 5 | R +4.5 = −2.0/130°<br>L +5.0 = −2.0/60° | R 6/30<br>L 6/30 | both hemifields R<L |
| 11 | 6 | R +1.0<br>L +0.5 = −1.0/0° | R 3/60<br>L 3/60 | both hemifields +++ |
| 12 | 5 | R +5.0 = −1.0/160°<br>L +5.0 = −1.0/25° | R 6/36<br>L 6/36 | norm outer limits |
| 13 | 18 | R +0.5<br>L −0.5 | R 6/6<br>L 6/6 | G: both hemifields down, R=L ++ |
| 14 | 9 | R −0.25<br>L +1.0 = −0.5/90° | R 6/9<br>L 6/12 | both hemifields down R=L ++ |
| 15 | 9 | R +2.0<br>L +2.5 | R 6/7.5<br>L 6/9 | norm outer limits |
| 16 | 9 | R +2.5 = −0.75/90°<br>L +2.0 = −0.75/90° | R 6/12<br>L 6/9 | both hemifields down, R=L ++ |
| 17 | 5 | R +4.0 = −1.0/90°<br>L +3.5 = −0.75/80° | R 6/7.5<br>L 6/9 | norm outer limits |
| 18 | 5 | R −0.5<br>L ±0 | R 6/9<br>L 6/6 | norm outer limits |
| 19 | 6 | R +0.5<br>L +0.5 | R 6/5<br>L 6/9 | norm outer limits |

Visual acuity = visual acuities tested with the LH test linear optotypes; Visual fields = mostly performed with the Stycar confrontation technique; Goldmann perimetry, when performed indicated by G; fd = field defect, ++ = moderate fd, +++ = severe fd; hom = homonymous.

*Table 4 Occurrence of strabismus and nystagmus characteristics*

| Patient no | Clinical | | Ober recordings | | | |
|---|---|---|---|---|---|---|
| | Strabismus | Nystagmus | Nystagmus type | Ampl (degrees) | Freq (Hz) | Latent nystagmus |
| 1 | exo | ## | pend | 0.5–2 | 1 | |
| 2 | eso | ## | SPI | 2–3 | 3 | + |
| 3 | exo | ## | SPD | 3–5 | 3 | |
| 4 | eso | ## | SPI | 1–2 | 5–6 | + |
| 5 | eso | ## | SPD | 3 | 1 | |
| 6 | exo | # | SPD | 3 | 1–2 | + |
| 7 | exo | 0 | SPD | 1–2 | 2–3 | + |
| 8 | exo | # | pend | 3–5 | | |
| 9 | eso | # | SPD | 3 | 2–3 | + |
| 10 | eso | ## | SPD | 3–5 | 2–3 | + |
| 11 | eso | # | SPD | 3–4 | 1–2 | |
| 12 | exo | # | pend | 2 | 1 | |
| 13 | exo | 0 | SPD | 2–3 | 3–4 | + |
| 14 | exo | # | SPD beats | 3 | 1 | + |
| 15 | exo | # | SPD beats | 2 | 1–2 | |
| 16 | exo | # | SPD | 1–2 | 2 | |
| 17 | orto | 0 | SPD | 2–3 | 3 | |
| 18 | eso | # | SPD | 0.5 | 3 | + |
| 19 | eso | 0 | SPI beats | 0.5 | 1–2 | |

exo = divergent strabismus; eso = convergent strabismus; ## = clinically easily detectable nystagmus; # = indicates clinically detectable latent nystagmus or intermittent nystagmus; 0 = no clinically detectable nystagmus; SPI = slow phase increasing velocity waveform; SPD = slow phase decreasing velocity waveform; pend = pendular waveform; beats = less than 5 nystagmus beats per 5 seconds; + = presence of latent form of nystagmus.

subjects. Calibration was done at the beginning of each recording by monocular viewing of a target moving in the four cardinal directions. A two point calibration procedure was then used. The paradigms used were fixation, smooth pursuit, and a saccade test. In the fixation paradigm, the subjects viewed the targets both binocularly and monocularly with both eyes separately. During the fixation paradigm, the subject was asked to keep his/her eyes on the flashing spot in the centre of the screen for about 10 seconds, and then fixation was tested in the four cardinal directions. The children were encouraged during the run of the paradigm to keep their eyes on the spot. In the smooth pursuit paradigm, the subjects mostly viewed the target binocularly. The stimulus moved at a constant velocity (triangular wave) of 5 deg/s for an amplitude of 25 degrees in the horizontal direction and 17 degrees in the vertical direction. In both the horizontal and the vertical planes, smooth pursuit movements were thus elicited in the centrifugal and centripetal directions. The saccade test was performed as refixations of the spot target jumping from the central fixation point to peripheral points in the horizontal and vertical directions. The maximum amplitude was 20 degrees in the horizontal plane and 10 degrees in the vertical plane. All analyses were performed off line on a computer. Nystagmus was defined as a rhythmic pendular movement of the eyes or a rhythmic jerk movement occurring more than 5 times during a 5 second period. If the eyes made rhythmic movements with a distinct nystagmoid waveform but with a low frequency (less than 5 beats/5 seconds), the movements were classified as nystagmus beats (Table 4).

## Results

### NYSTAGMUS

Six children had clinically easily recognisable nystagmus, while nine children had clinically detectable latent or intermittent nystagmus seen with the help of a visuscope with a fixation target (Table 4). Four children had nystagmus recognisable only with eye movement recording. With the Ober-2 recordings 16 of the 19 subjects exhibited horizontal nystagmus. In the remaining three subjects, the ocular movement was classified as nystagmus beats. In most cases, the nystagmus had a variable frequency and amplitude during the recording session and therefore the commonest nystagmus characteristics in each subject are described (Table 4). The nystagmus usually had a horizontal vector, but vertical oscillations of small amplitude and low frequency were seen in many cases. Both conjugate and asymmetrical nystagmus waveforms were seen and in some cases the nystagmus was out of phase.

With the aid of the recordings, we characterised the nystagmoid waveforms and could therefore classify the nystagmus. Of the 16 subjects showing nystagmus, as defined above, 11 had a slow phase exponentially declining waveform (SPD in Table 4; Fig 1), but two had a slow phase exponentially increasing waveform (SPI in Table 4; Fig 2) and three subjects had a pendular nystagmus (Fig 3). Among the subjects with nystagmus beats, two exhibited a SPD waveform, one had a SPI waveform. The SPD nystagmus type had an amplitude of 1–5 degrees and a frequency of 1–6 Hz. The nystagmus could always be recognised in the recordings when the subject used both eyes, but in nine cases it increased in intensity

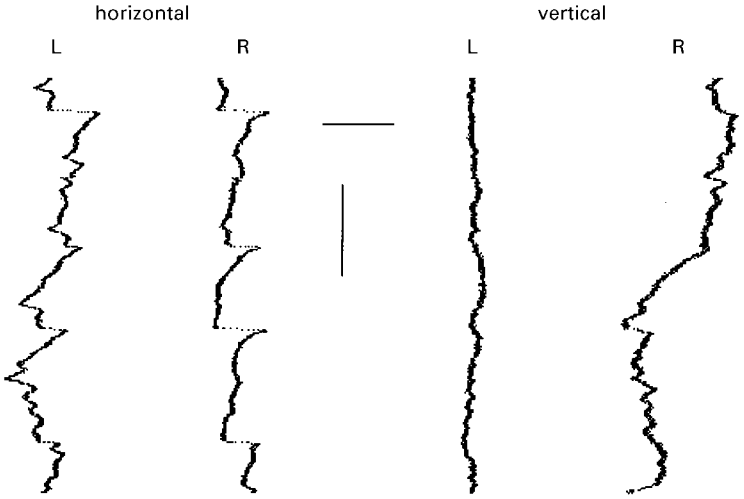


*Figure 1 Eye movement record showing horizontal and vertical traces of the right (R) and left (L) eyes with the Ober-2 system. Patient no 9 when fixating in primary position with both eyes open. Note the variable amplitude and the asymmetrical waveforms with a SPD (slow phase decreasing velocity) waveform most visible in the right eye trace. Note also the difference in vertical stability between the right and left eye. Horizontal bar, 3 degrees; vertical bar, 500 ms.*

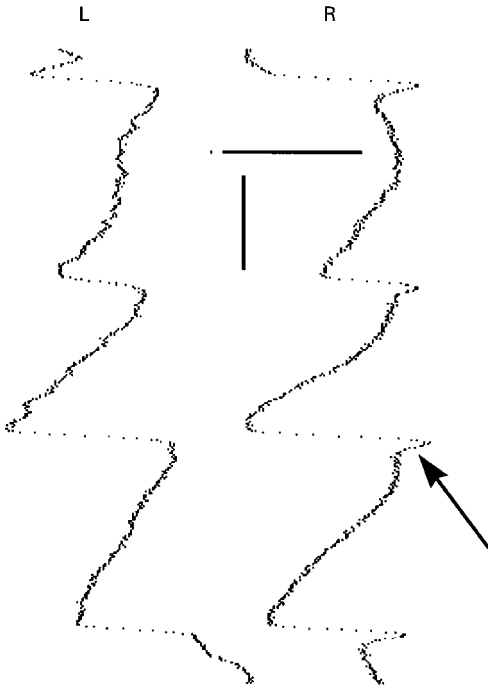


*Figure 2 Eye movement record showing horizontal and vertical traces of the right (R) and left (L) eyes with the Ober-2 system. Patient no 2 with only horizontal traces shown. The record showing a jerk nystagmus with a SPI (slow phase exponentially increasing velocity) waveform with a superimposed foveating saccade (arrow). Horizontal bar, 2 degrees; vertical bar, 200 ms.*

(amplitude and frequency) when the subject used only one eye (that is, latent nystagmus; Table 4).

In subjects with nystagmus classified as SPD, we found a directional preponderance, with the fast phase always directed towards the eye with the better visual acuity. In several cases, a null point was seen and several of the children turned their heads.

The three patients with a SPI nystagmus waveform had an amplitude of 0.5–3 degrees with a frequency of 1–6 Hz (Table 4). The

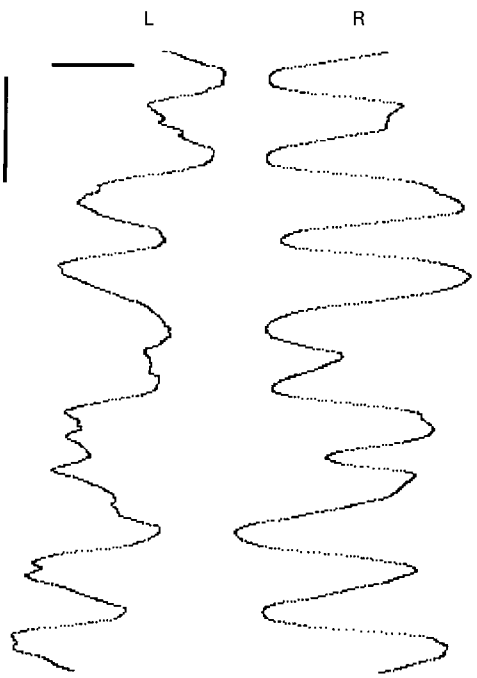


*Figure 3 Eye movement record showing horizontal and vertical traces of the right (R) and left (L) eyes with the Ober-2 system. Patient no 12 with a asymmetrical pendular nystagmus out of phase. Horizontal bar, 4 degrees; vertical bar, 500 ms.*

three patients having pendular nystagmus had an amplitude of 1–3 degrees with a frequency of about 3 Hz (Table 4). The patients with the pendular nystagmus often had different amplitudes in the right and left eyes. No correlation was found between the grade or location of the PVL and the type of nystagmus.

### OTHER OCULAR MOTOR ABNORMALITIES

Although all children seemed able look at the targets during testing, many of the children exhibited very inappropriate smooth pursuit movements. Several children in the study group had great difficulties in performing smooth pursuit when clinically tested and therefore could not trace the target on the computer screen. Among children who could perform smooth pursuit several did this better in one direction than the other. In addition, several of the children seemed to be unable to elicit voluntary saccades to the visually presented targets. This was also a clinical impression in these children. During the clinical investigation it was found that they seemed to be using their heads for redirecting the line of sight and consequently they did not perform appropriate saccades to the visually presented targets since their head was fixated by a bite board.

### CASE PRESENTATION

Patient no 9 (in Tables 1–4) is a girl born at a gestational age of 33 weeks, with birth weight 2640 g. One week before her birth, her mother developed contractions. The cardiac registration then showed a few events of fetal heart deceleration. Delivery was uncomplicated (Apgar score 9-10-10) and the neonatal period was uneventful.

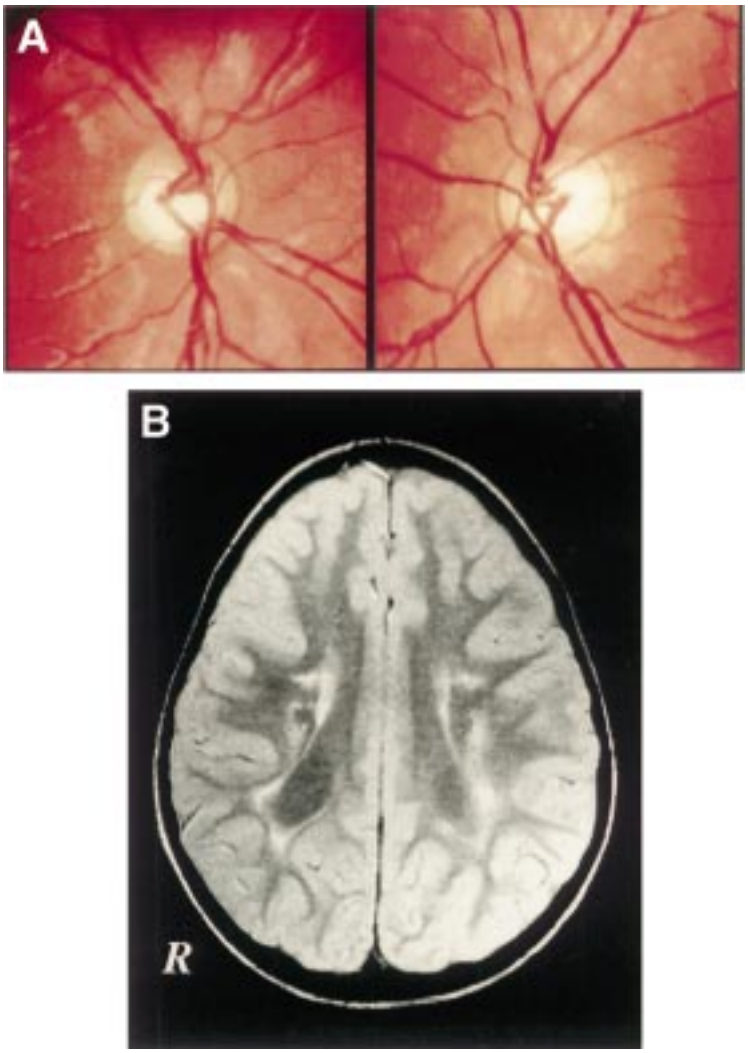


*Figure 4 (A) Fundus photograph of patient no 9 demonstrating large cups in normal sized discs. (B) This single MRI shows typical changes of periventricular leucomalacia with atrophic dilatation of the posterior parts of the lateral ventricles. The amount of periventricular white matter is reduced around the occipital horns and adjacent to the trigone bilaterally. Remaining white matter has an abnormally bright signal on this T2 weighted image, first echo, indicating permanent damage, gliosis. Variations of signal involving left posterior cortical structures represent artefacts in the image.*

The girl was referred to an ophthalmologist at the age of 5 months because of an infantile esotropia with cross fixation. She underwent surgery for strabismus at the age of 1½ years. Latent nystagmus was noted at 2½ years of age. At 4 years of age, she had a visual acuity of R=L 6/30. The optic discs had large cups (Fig 4A) but the intraocular pressure was normal. An MRI of the brain revealed moderate PVL with a side-like distribution and mostly located in the middle and posterior parts of the periventricular white matter (Fig 4B).

Assessment of visual function at the age of 5 years by a multidisciplinary team at TRC demonstrated low vision, crowding, and visual field defects in the lower parts of both hemifields, but preserved colour vision. Refraction was R +3.0=−1.5c 130° L +3.5=−1.5c 50°.

Her cognitive profile was uneven, with considerably higher scores on verbal than on visuospatial tasks. She did not have cerebral palsy, although her movements were not well coordinated.

Eye movement recordings showed a right beating horizontal nystagmus in phase. The nystagmus was mostly asymmetric but had an amplitude of about 1° and a variable frequency between 1–2.5 Hz. The nystagmus waveform had a slow phase exponentially decreasing profile (SPD; Fig 1). The intensity increased markedly when the right eye was covered, but not the left eye (it also changed vector to left beating when the right eye was covered—that is, latent nystagmus). The eyes were mostly relatively stable in the vertical direction (Fig 1). She made better smooth pursuit movements when tracing the target to the right than to the left and saccades could not be elicited with the paradigm used.

## Discussion

The present study was initiated with the clinical impression that children with visual impairment and/or strabismus associated with PVL often had nystagmus and other ocular motor abnormalities. In contradiction to earlier reports,[2 10] it is our opinion that nystagmus is common in children with PVL. Moreover, these children seem to have problems in performing normal smooth pursuit and saccadic eye movements, but one cannot judge from the present study whether these findings are a pure ocular motor problem or indicate a general difficulty with movement perception.[20]

### METHODOLOGICAL CONSIDERATIONS

It must be emphasised that many children in the present study were difficult to investigate both clinically and with the eye movement recording equipment, owing to attention and/or concentration difficulties. Hence, complete ocular motor examination could not be performed in some subjects. However, we believe that the eye movement recordings with the Ober-2 technique used in the present study have been very useful for evaluating eye movement characteristics in PVL children.

The children in this study were selected because of visual impairment or strabismus, in combination with CP. It is therefore possible that the material was biased, excluding those with mild lesions (overlooked, or often regarded as "only strabismics" or CN) or those with more severe neurological sequelae (severe mental retardation combined with CP). The material therefore cannot be regarded as representative of all children with PVL.

### NYSTAGMUS AND OPTIC NERVE ABNORMALITIES IN PVL

Nystagmus has not been found in cerebral visual impairment,[2 10] but is a common finding in optic nerve hypoplasia.[11–13 21] For example, Gelbart and Hoyt[11] found 119 patients with a sensory disorder in a series of 152 patients with CN. Only 13 patients in their study had pure CN. The commonest diagnosis in the sensory group was ONH. However, the association between ONH and the frequent occurrence of nystagmus in these patients is at present not understood. In PVL, the optic disc often appears to have a large cup, considered a variant of ONH.[3] These discs and the small optic

discs, associated with early, often prenatal, PVL[2 3] are presumably caused by a retrograde trans-synaptic degeneration via the lateral geniculate body at different times during a sensitive period in the immature brain secondary to the lesion in the optic radiation.[3] Children with PVL thus acquire a secondary lesion of the anterior visual pathway. However, the causal connection between the frequently occurring nystagmus in these patients still remains an enigma.

### NEURORADIOLOGY IN PVL

The neuroradiological findings in PVL are well described[19 22] with reduction of the periventricular white matter, enlargement of the lateral ventricles and gliosis in remaining white matter. Today MRI is the method of choice for confirming the clinical impression of PVL. However, CT may demonstrate only the more conspicuous lesions.

In the case report we have presented a girl who was classified as infantile esotropia with nystagmus. She had a normal intellectual level and no cerebral palsy and the MRI investigation had demonstrated a lesion in the optic radiation. In most strabismic children without major neurological deficits, neuroimaging is probably never done. We presume that an optimal neuroimaging procedure, performed in preterm children with infantile esotropia and/or congenital nystagmus with no other neurological deficits, would probably show PVL in some cases.

### NYSTAGMUS WAVEFORMS IN PVL

Twelve children in the study group exhibited a jerk nystagmus, with a slow phase exponentially decaying wave form. This type of nystagmus has been described mostly in subjects with latent nystagmus or latent/manifest latent nystagmus[14 23] The finding that the quick phases were always beating towards the eye with the better visual acuity further indicates that the nystagmus seen is a form of LMLN and not the commonly classified CN, which usually has been associated with an increasing velocity slow phase waveform.[14 16] We therefore regard the nystagmus seen in the subjects with a SPD waveform to be a LMLN which has become manifest as a result of suppression of one eye because of strabismus and amblyopia. The subjects who exhibited the SPI waveform as well as the subjects with pendular nystagmus could well be corresponding to the motor form of CN.

### OTHER OCULAR MOTOR PHENOMENA IN CHILDREN WITH PVL

One of the most striking ocular motor phenomena in these children with PVL was their inability to make smooth pursuit movements as well as saccades to visual stimuli. The reason for this is not clear but there may be various causes. One is that the children with PVL and visual impairment have a deficit in movement perception, as suggested by Dutton and coworkers.[20] Another explanation could be that the prenatal white matter lesions in PVL adjacent to the trigonal area injure the arcuate fibre bundles connecting the striate cortex with area MT (middle temporal visual area).[24] Area MST (medial superior temporal visual area) may also be affected by such a lesion, and neurons in these areas are known to encode not only visual signals but also non-visual signals, presumably an efference copy of the ocular motor command.[25] Therefore, the lesion in PVL might affect the premotor commands for eye movements in areas MT and MST.

## Conclusions

The inappropriate ocular motor function in the PVL children with regard to the smooth pursuit and saccadic performance cannot be explained. The present study has shown that nystagmus is a frequent finding in children with PVL. Furthermore, eye movement recordings in these children can be a useful tool for improving the pathophysiological understanding of this type of nystagmus. This study has shown that one third of the children had clinically easily detectable nystagmus. However, two thirds of the children had clinically detectable latent or intermittent nystagmus or nystagmus beats seen only with the help of a visuscope with a fixation target or the eye movement recording. The nystagmus in these children would easily have been unnoticed during a routine ophthalmological investigation.

The finding of nystagmus in children with PVL is new. The observation can have several explanations. One possibility is that the injury to the immature optic radiation causes a trans-synaptic retrograde degeneration in the optic pathway so that the result in fact is an anterior visual pathway injury. Such an injury is known to be associated with nystagmus although the causal connections have not been established. An alternative explanation of the nystagmus could be an injury to the optic radiation affecting input into the visual integrating circuits which could also affect the premotor commands for eye movements and result in nystagmus.

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