Published in final edited form as:

Ophthalmic Physiol Opt. 2008 May ; 28(3): 283–290.

Monocular Fixation with the Optic Nerve Head: a case report

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Abstract

Purpose—To document and discuss the case of a patient with left esotropia (ET) who uses the left eye optic nerve head (ONH) for monocular “fixation”.

Case Report—The patient was an 80 year-old male with left ET from early childhood. Retinal tracking monocular fixation measurements with a Nidek MP-1 revealed stable fixation within the left ONH area. In an attempt to challenge the initial observation, further assessments of fixation were performed with a smaller target size and requiring various gaze positions. MP-1 fixation data showed remarkably stable monocular fixation ($\pm 1^\circ$ over 30 sec) mostly within the left ONH for all the target sizes and positions of gaze tested. Additional clinical binocular evaluations showed concomitant left ET $\sim 28\Delta$, no movement with cover test regardless of fixation target, and no significant monocular motility restrictions. Visuoscopy also revealed fixation at the left ONH. There was a strong family history of ET, but none of the other affected descendants tested ($n=3$) demonstrated the same behaviour.

Conclusions—This is the first report documenting abnormally developed monocular ocular motor system with principal visual direction and zero retinomotor value shifted from the fovea to the ONH. We do not believe that there is any direct visual input from the ONH. The patient may use visual information obtained by glancing with peripapillary areas to determine the target position (though this was largely ruled out), or obtain position information from the average luminance produced by scattered light around the ONH margin. The abnormal oculocentric direction might then be combined with extra retinal information (efferent copy or extraocular muscle proprioception) of the eye location in the orbit to stabilize the fixation. This patient does not have the Blind Spot Syndrome as previously described (Swan, 1948). Possible aetiologies are discussed. We propose the use of a retinal perimeter for documentation of eccentric fixation in strabismus.

Keywords

Blind Spot; Optic Nerve Head; Eccentric Fixation; Principal Visual Direction; Retinomotor Value; Esotropia; Retinal Perimeter

In early acquired strabismus the visual system typically develops sensory adaptations to avoid the symptoms of diplopia and visual confusion (Pickwell, 1984), often reported as “double vision”. If these adaptations did not develop, the patient would experience diplopia, as the image of an object seen with the fovea of the nondeviated eye would also be seen with a peripheral retinal area of the deviated eye (retinal images would not fall on corresponding points); and visual confusion, as the fovea of the deviated eye would receive an image different to that falling on the fovea of the non-deviated eye (two corresponding points would receive different images which would appear superimposed, in the same direction). These phenomena are usually reported, when they occur in central retina, by patients who develop strabismus as adults (at an age when they typically do not develop adaptations). While diplopia and visual confusion occur only under binocular viewing conditions (as a consequence of strabismus), the resultant adapting mechanisms may function under either binocular viewing conditions or may persist under monocular conditions.

The sensory adaptive changes that may occur during binocular vision are suppression and abnormal retinal correspondence (ARC) (Von Noorden, 1969; Pickwell, 1984). A suppression scotoma may appear in the central retina of the deviated eye to avoid visual confusion and may extend (nasally in ET) to avoid diplopia (Pickwell, 1984). ARC is a binocular adaptation of the visual direction system, measurable only under binocular conditions, where objects are perceived as localized in the same direction in spite of their images falling on normally non-corresponding retinal locations (Pickwell, 1984).

These adaptation mechanisms may persist monocularly in the form of amblyopia or eccentric fixation. Amblyopia is the developmental impaired vision (of no organic cause) typified by reduced visual acuity, distorted space perception and incorrect localization of objects in space (Bedell *et al.*, 1985; Skottun *et al.*, 1986; Fronius *et al.*, 2004). It may be a consequence of long term binocular suppression that remains under monocular conditions - although some advocate that it may precede the strabismus (Schor, 1978). Strabismic amblyopia is most commonly associated with constant ET (Quah *et al.*, 1991). Eccentric fixation refers to abnormal monocular visual fixation with an area other than the fovea (Von Noorden, 1970; Kirschen and Flom, 1978; Pickwell, 1984). The eccentric fixation locus typically occurs within 5deg ($\sim 10\Delta$) from the fovea (Flom *et al.*, 1980; Pickwell, 1981; Bedell *et al.*, 1990; Matilla *et al.*, 1995; Cleary and Thompson, 2001) and it may be stable or unstable.

The perceived direction of an object in space is determined by the retinal location it stimulates in conjunction with extraretinal information (e.g. eye position in orbit). Each retinal locus has a retinal visual direction associated with it. In a normal eye, the fovea has the principal visual direction (PVD) that is associated with the straight-ahead egocentric visual direction when the eye is in the primary position of gaze. Likewise, each retinal locus has a motor value, which means that it induces a fixed magnitude eye movement (saccadic) during the fixation reflex that brings the fovea to the direction of the object that stimulated that retinal location. The zero retinomotor value (ZRMV) (normally at the fovea) is the retinal point to which the eye makes a reflex saccadic re-fixation movement when presented with a peripheral stimulus (Ciuffreda, 1991; Steinman *et al.*, 2000). Usually the new monocular fixation locus in eccentric fixation becomes associated with the PVD and the ZRMV (Von Noorden, 1970; Ciuffreda *et al.*, 1979; Bedell and Flom, 1981), although some authors argue that the PVD may remain unchanged (Pickwell, 1984).

ET with an angle of deviation of about 15deg ($\sim 28\Delta$) is not rare (Swan, 1948; De Muelenaere and Hambresin, 1956; Guzzinati, 1956; Prizner, 1975; Olivier and Von Noorden, 1981; Birch *et al.*, 2004). In these cases, the fovea of the deviated eye is directed at an object whose image falls within the physiological blind spot of the non-deviated eye and vice versa. It has been suggested that this phenomenon may be a mechanism to avoid diplopia (Swan, 1947) and visual confusion. Swan (1948) defined the *Blind Spot Syndrome* as a type of adaptation to ET with angle of deviation of about 15deg, with occasional diplopia, normal retinal correspondence, and normal visual acuity. Others have reported cases of *Blind Spot Syndromes* (De Muelenaere and Hambresin, 1956; Guzzinati, 1956; Uemura, 1964; McKenzie *et al.*, 1970; Prizner, 1975; Harrer, 1984). However, controversy surrounds the nature of these cases. The case we report here is not a case of *Blind Spot Syndrome* (see Discussion section).

We have documented, using an objective method of assessing visual direction, a case of left ET wherein (under left monocular viewing) the left PVD and ZRMV are located within the left physiological blind spot.

CASE REPORT

Patient

The patient was an 80 year-old male with left ET and amblyopia documented to exist at least since age 3 (figure 1) and with no reported change in visual acuity or angle of deviation over time. He did not receive surgical, refractive, or visual training treatment. No other health problems or illnesses occurring during birth or childhood were reported.

His right eye has reduced visual acuity due to a non-arteritic anterior ischemic optic neuropathy (NAION) diagnosed 4 years before our evaluation. His visual acuity was normal, recorded Snellen 20/20 (6/6), in this eye prior to the NAION acute event.

Procedures

The patient came to our research laboratory to participate in a low vision research project. During the routine visual exam conducted prior to the experimental procedures, we conducted retinal fixation measurements with a Nidek MP-1 retinal perimeter (Nidek Technologies, Vigonza, Italy, software version available in 2004: MP1 SW1.4.1. SP1). Measurements were made monocularly with the other eye occluded with an eye-patch. During the retinal fixation exam, the patient was asked to look at a fixation target (a white cross) for 30 sec. An auto-tracking system recorded the patient's retinal movements, frame by frame, using a reference retinal feature chosen by the examiner on the IR retinal image. Thus the fixation locus and its variability over time were evaluated. During 30 seconds of net retinal fixation recordings, 750 retinal fixation points were recorded (rate 25Hz), skipping images lost due to blinking or other causes. The system allowed placement of the fixation cross into any desired position within the (45deg) screen seen by the patient, enabling evaluation of fixation at different gaze positions. A colour retinal photography was obtained and registered with the IR image so that the results were displayed on the colour image.

The initial fixation target used was a white cross of 2x2deg positioned centrally hence requiring straight ahead gaze position. Fixation data showed that the right eye fixated with an area slightly superior to the fovea (consistent with central field loss due to NAION, figure 2) and the left eye fixated within the optic nerve head (ONH) (figure 2).

In order to challenge this surprising observation, additional measurements were taken on the left eye. It was possible that the patient was not looking at the cross as instructed, but rather "holding" (Steinman *et al.*, 1969) his eye in that straight ahead gaze position. The patient might have been using the peripherally visible edge of the screen and the observation tubes as visual cues to help him direct his ONH to the centre of the screen (Sansbury *et al.*, 1973). It was also possible that he was obtaining visual information from an area near the ONH for fixation. Since the initial cross was large (2x2deg), the ends of two of the arms of the cross might have been visible in peripapillary retina. Therefore, additional measurements of fixation with the left eye were taken using a smaller fixation cross (1x1 deg) which was positioned at various locations within the screen seen by the patient, requiring different gaze positions.

Findings

1. Fixation measurements with Nidek MP-1—The patient followed the target (white cross on black background) with his left eye easily into all positions of gaze and at each position demonstrated a remarkably stable monocular fixation during the exam (the duration of the total fixation time required to record 30 sec of valid data varied between the 7 positions [55 ± 12 sec]). For most gaze positions, the fixation was stable (based on the instrument's manual definition - more than 75% of the fixation points are within a 2deg circle) (Nidek MP-1 User

Manual v1.5.0). On average the gaze was within a 2deg diameter circle for $73 \pm 11\%$ of the testing time (30sec) and within 4deg for $97 \pm 3\%$ of the time.

Custom software was used to calculate fixation eccentricity (figure 3). The rim of the ONH (blue crosses) is manually marked (variability $\leq 0.13\text{deg}$) (Woods *et al.*, 2007), and the program fits an ellipse (black continuous line). The centre of the ellipse is considered the centre of the ONH (black cross). The location of the fovea is calculated from the centre of the ONH (green dot). The red circle represents the 95% confident limits. The white cross and the cluster of cyan dots represent the area used for fixation. The estimated fovea position is based on an average from studies with normally sighted subjects (Hu *et al.*, 1994; Rohrschneider, 2004; Rohrschneider *et al.*, 2005; Timberlake *et al.*, 2005): 15.3deg temporal and 1.5deg inferior to the centre of the ONH (figure 3).

With the 2x2deg target size, the right eye showed peri-central fixation, with the fixation locus situated 13.6deg temporal and 0.4deg superior to the centre of the ONH (figure 3), near but not at the fovea for that eye. Under the same conditions, the left eye used the superior-inner part of the ONH (0.5deg temporal and 1.4deg superior to the centre of the ONH) as its fixation locus (figure 3).

With the smaller target size (white cross, 1x1deg), the fixation cross was located within the optic ONH margins in the left eye for most gaze positions (figure 4), except for the extreme left gaze positions when it lay outside the ONH margins. There was a small variation in the exact location of the left monocular fixation at various gaze positions, with a fixation location of $0.2\text{deg} \pm 1.5\text{deg}$ (mean $\pm 1\text{SD}$) temporally and $1.4\text{deg} \pm 0.5\text{deg}$ (mean $\pm 1\text{SD}$) superiorly from the centre of the ONH. In most gaze positions the whole fixation cross was within the ONH during all the recorded time.

Potential spatial misalignment and head rotation have to be considered in the calculation of the location of the fixation location. Since, unlike the original SLO, the Nidek MP-1 uses a different light source (IR) to track the fundus image than to present the stimuli, there is potential for misalignment between these two systems (Woods *et al.*, 2007). When applying the correction factor proposed by Woods *et al.* (2007) for the misalignment found in the MP-1 used in this study, the fixation location for the left eye would be shifted further into the centre of the ONH (approximately 0.2deg vertically and 0.7deg horizontally). Therefore, with this systematic error correction, the whole fixation cross would be located within the ONH in all gaze positions.

2. Additional optometric examinations—External observation revealed no facial or postural anomalies. Visual acuity (single letter, Test Chart PRO2000, Thomson Software Solutions, Herts, UK; www.thomson-software-solutions.com) for the right eye was 6/30 (20/100) and for the left eye 6/480 (20/1600). Visual acuity for his left eye's fovea (measured by asking him to fixate at a target located at 15deg to the left of the visual acuity target position) was 3/387 (20/2580).

No movement was found in the cover/uncover or alternating cover tests regardless of the fixation target used (e.g. large letter, light source). Hirschberg & Krimsky tests were performed at various gaze positions and results showed left concomitant ET of $\sim 28\Delta$ (figure 5). Ocular motility tests showed unrestricted monocular motility in each eye and reduced abduction in the left eye during binocular testing. Visuoscopy was carried out with a special large fixation target inside the ophthalmoscope and showed fixation of the right eye with an area near fovea and the left eye at the ONH. Finger-pointing showed the same (accurate) response when finger-pointing with the left eye (performed first) or the right eye. He also accurately pointed under a table at the directions of objects placed on the table while he could not see his hand, suggesting

that egocentric mislocalizations are not currently present (Von Noorden *et al.*, 1970). His ability to navigate in a cluttered room with his right eye covered was unimpaired.

3. Other family members—An unusually strong family history of ET was observed in the patient's family (figure 6), with both his sons and one grandson having partially accommodative ET. Therefore, other family members were tested looking for a repetition of the phenomenon, however, none had an ET of about 15deg (table 1).

DISCUSSION

We have documented, for the first time, a case of an abnormal monocular visual system adaptation to ET wherein its fixation locus, PVD and ZRMV have shifted and are now located within the ONH, a visually blind area. Indirect evidence that the patient is using the ONH for left eye fixation is the poor visual acuity (20/1600) found. The size of the smallest letter seen by the patient corresponds to a letter that extends just beyond the limits of the physiological blind spot area (figure 7). This supports the hypothesis that the physiological blind spot has been placed in the centre of the letter when asked to identify it. The abnormal oculocentric direction is subsequently combined with extra retinal information of the eye location in the orbit to give visual direction and egocentric position (Weir *et al.*, 2000).

We are not proposing that there is direct visual input from the ONH. We believe that the patient obtains visual input from areas surrounding the ONH to acquire information on the position of the stimulus, by using scattered light information (Faubert *et al.*, 1999), and then placing the physiological blind spot in the direction of the stimulus. Alternatively, it is possible to "hold" the eye in a particular gaze direction for short periods of time, as shown in normally sighted subjects following specific instructions (Steinman *et al.*, 1969). When doing so, the visual system does not concentrate on details and saccade movements are not elicited. We disregarded this possibility by evaluating his monocular fixation while we located the target at various gaze positions; results showed that the patient was moving his eye following the target and maintaining the ONH as his monocular fixation point. It seems that a complete reorientation of his left eye directionality mapping was developed resulting in the ONH now carrying the straight ahead value. It is, nevertheless, not obvious how the visual system could consistently, accurately and steadily direct the eye to a non-visual area.

We consider two possible aetiologies of the eccentric fixation: (1) the patient may have simply developed "by chance" 15deg of eccentric fixation which corresponds to his 15deg of ET and hence fixation is located at the ONH.

(2) The 15deg angle of ET with the accompanying fixation and primary directionality within the ONH may have developed as an adapting mechanism to avoid visual confusion and diplopia. If a deviation had occurred (primary or as a consequence of reduced VA), and the adaptation mechanism of suppression (or ARC, but not likely in our case given the large deviation) had not been sufficiently deep to avoid diplopia and confusion, a different adaptation mechanism would have been needed. The initial angle of the deviation may have been adjusted, perhaps it was an accommodative ET with variable angle (at least two of the descendants are known to have had accommodative ET); so that the physiological blind spot was used as a central scotoma to avoid visual diplopia and visual confusion—at least centrally.

We believe that the changes in fixation and PVD occurring in this case did result from the need for a complete suppression scotoma in central retina to avoid diplopia and confusion at some point during development. This may be similar to cases with microtropias which may demonstrate eccentric fixation of equal magnitude to the angle of strabismus (Von Noorden, 1969).

The use of the physiological blind spot as a mechanism to avoid diplopia and visual confusion was suggested by Swan (1947). Swan (1947) studied 296 patients with concomitant ET, 80 of which had a deviation that would overlap the fovea of one eye with the physiological blind spot of the other eye in binocular conditions (he defined ET = 12–17deg). Out of these 80 cases, 7 cases seemed to use the physiological blind spot as the only mechanism to relieve diplopia since there was neither suppression scotoma nor ARC. He identified some common characteristics of these cases including concomitant ET = 12–17deg (~25–35 Δ) at distance and near, occasional diplopia, normal VA in each eye (20/20), normal retinal correspondance and good fusion, and named them as the *Blind Spot Syndrome*. None of these cases continued to fixate with the ONH under monocular conditions.

In 1948, Swan reported a larger series of these cases (102) that fall into the definition of *Blind Spot Syndrome*, 75 of those reported occasional diplopia when asked (seldom volunteered). All showed normal retinal correspondance measured with image after-effects and synoptophore and had potentially good fusional capabilities.

Others have reported cases of *Blind Spot Syndrome* as it was defined by Swan (Guzzinati, 1956; McKenzie *et al.*, 1970; Harrer, 1984). However, its existence has been disputed, particularly the absence of sensory adaptations as described by Swan (Olivier and Von Noorden, 1981). Oliver and von Noorden argued that Swan found normal retinal correspondance because the techniques he utilized were not sensitive enough, and that if Bagolini's lenses are used, ARC would have been found in all cases.

We do not claim to have presented here a case of *Blind Spot Syndrome* and this case does not fulfil the definition of *Blind Spot Syndrome*, primarily because the shift in fixation holds under monocular conditions, the patient has reduced visual acuity and has never reported diplopia.

In this case, we have documented the retinal area used as fixation locus and PVD in a patient with eccentric fixation. Retinal perimeters such as the Nidek MP-1 or SLOs are currently used to determine fixation (preferred retinal locus - PRL) in patients with macula disease. We propose as an additional use of retinal perimeters with real-time eye tracking the assessment of eccentric fixation in strabismus.

Acknowledgements

This work has been supported in part by NIH grants EY05977 and EY12890.

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Figure 1.
Patient's childhood pictures. [a] at 3 years of age and [b] at 5 years of age both showing left ET.

Figure 2.

Fundus photographs and fixation data taken with the Nidek MP-1 during the routine exam ([a] right eye; [b] left eye) using a large fixation cross target (2x2deg). Measurements were performed monocularly. The patient was instructed to “look at the centre of the cross”. The patient reported “I do not see that it is a cross but I know where it is”. The left eye image quality is reduced due to the cataract.

Figure 3. Fundus photographs and fixation data taken with the Nidek MP-1 and processed to calculate the distance from the centre of the disc to the position of the estimated fovea (green dot) and 95% confident limits (red ellipse).

Figure 4. Fundus photographs and fixation data taken with the Nidek MP-1 on the left eye using a smaller fixation target (1x1deg) placed at various locations. Measurements were performed monocularly. The patient was instructed to “look at the centre of the cross”. The patient reported “I do not see that it is a cross but I know where it is”.

Figure 5. Patient's eyes at time of evaluation (age 80) showing the left eye deviation when looking straight ahead at distance.

Figure 6.

Familial genetic tree of the patient's descendants showing the patient (black filled square) and other members affected with left ET (shaded symbols). Males are represented by squares and females by circles, diagonal lines indicate a deceased family member. Some individuals not affected with ET were removed for simplification; the number of such individuals (descendants of the patients' siblings) is given in brackets. Descendants named as Son 1, Son 2 and Grandson are those family members tested in our lab (table 1).

Figure 7.

[a] Illustration of the retinal size for a 20/200 letter – normal VA expected at the edges of the ONH. [b] Illustration of the retinal letter size corresponding to the patient's left eye VA (20/1600) which is just slightly larger than the ONH.

Table 1

Information on the 3 family members tested.

Family Member	Age (yrs)	Dist Cover Test (Δ)	Type of Deviation	VA (single letter) of affected eye	ET Treatment
Patient	80	no movement	left ET	6/480 (20/1600)	none
Son 1	55	6 Δ (~3 rd)	left ET	6/7 (20/25)	refractive error correction single lenses, patching and visual training
Son 2	53	6 Δ (~3 rd)	left ET	6/12 (20/40)	none
Grandson	24	15 Δ (~8 th)	left ET	6/6 (20/20)	refractive error correction, including bifocal lenses