Egocenter Location in Children Enucleated at an Early Age

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In normal binocular vision the origin of judgments of visual direction is the egocenter located on the median plane of the head, midway between the two eyes. For children enucleated at an early age there is abrupt cessation of binocular input to the visual cortex. We evaluated the effect of early removal of one eye on the location of the egocenter, comparing the results in enucleated children to normal binocular children tested monocularly. We found a significant shift of the egocenter towards the remaining eye of the enucleates that was unrelated to the age at enucleation. At least up to 4 years of age, egocenter location is plastic and can be modified by the complete monocularity imposed by enucleation. Invest Ophthalmol Vis Sci 29:1348–1351, 1988

One of the important functions of the human visual system is to specify the direction of objects in visual space. Underlying the performance of this function is the hypothesis that binocular judgments of visual direction originate from a single point or center that is approximately midway between the two eyes.1 This point has been variously referred to as the cyclopean eye, the visual egocenter, the binocular and the projection center; we will use the term egocenter. In humans with normal binocular vision it has been shown that the egocenter hypothesis is valid.2 Anecdotal reports of Church (cited by Barbeito3) described that 1.5-year-old infants would place a tube between their eyes when asked to sight through the tube. Barbeito subsequently confirmed this "cyclops" effect. Adults also use the egocenter when not constrained by a monocular alignment task.4

In retinoblastoma, since vision is restricted to only one eye from a very early age, the normal development of egocentric projection may be impeded. A malignant tumor arises in the retina in infancy to eventually fill the globe (and, if left untreated, it can invade the optic nerve and brain to cause death). Enucleation of the eye (usually before the age of 4) is therefore frequently obligatory as a life-saving treatment. Consequently there is an abrupt cessation of binocular input to the visual cortex during the critical period for vision development. At the Hospital for Sick Children in Toronto, Canada, a large number of children with retinoblastoma who have had one eye removed are ophthalmology clinic patients and able to participate in vision experiments. We were interested in determining if the location of the egocenter was altered in these children after enucleation. Hering5 to whom the original formulation in 1861 of the laws of visual direction has been attributed, (although see Ono6 for evidence that these concepts were first proposed by Wells as early as 1792) showed prescience when he also suggested that for enucleates the principle that visual direction judgments originated from the egocenter would not apply. Rather, Hering proposed that for one-eyed observers the cyclopean eye shifts to the remaining eye. We wished to determine how accurate Hering was about the location of the egocenter in enucleates. In addition, because our population included children enucleated between the ages of 1 month to 4 years, we expected to be able to find evidence for a critical period in the development of the egocenter.

Materials and Methods

At least four reliable methods of determining the egocenter in adults have been described.7 We adopted and modified Roelof's method so that it would be suitable for use with young children. It is a principle of visual direction that a line along the visual axis will appear to be on the cyclopean axis; that is, the line that passes through the egocenter. In Roelof's original experiment the subject was required to look monocularly at the front of a tube that had been objectively
aligned along the visual axis. The apparent direction of the tube was specified as the projection of the front of the tube to the point on the subject's face toward which the tube appears to point. Presumably this point represented the egocenter so that the projection passed through it. In our experiment the equivalent of the tube was a displaceable, pivoting rod 1.5 mm in diameter. Referred to as the "fireman's hose," it emerged from the base of a 4.5 cm high toy fireman that the child was asked to fixate and travelled the length of the board (Fig. 1). This toy fireman sat in the middle of one end of a wooden board whose dimensions were 33 cm × 35.5 cm. Directly opposite the fireman an indentation was cut into the board in which the bridge of the child's nose was inserted. In separate sets of trials the rod was set by the experimenter to point either along the child's midline or along the eye's visual axis (considered the center of the child's pupil, as observed by the experimenter). Underneath the board, but attached to it, was a metal lever that pivoted from the same point as the rod on the upper surface. The child was instructed to grasp this lever with both hands and to move it to that position which placed it directly beneath the fireman's hose, thus along the same visual direction. At the completion of this task the experimenter read the position of the lever (to the nearest millimeter) on a metric ruler taped beneath the board. Neither the lever nor ruler were visible to the child.

The experiment began with the fireman's hose aligned along the child's midline. Between three to six measures were obtained. The experimenter then aligned the fireman's hose along the visual axis and three to six measures on this condition were obtained. It was necessary to reduce the number of trials when the child demonstrated fatigue or a decrease of concentration for the task. These factors tended to increase the variability of the measures. Fixation was monitored by the experimenter, and, when necessary, trials were repeated when fixation was not accurately maintained on the fireman. The child's head position, while not actively restrained, was monitored and kept aligned with the apparatus.

The subjects who had undergone enucleation of an eye because of retinoblastoma were all patients at the Ophthalmology Clinic of the Hospital for Sick Children. At the time of testing they ranged in age from as young as 5 years to as old as 29 years of age. The age at enucleation also varied from as young as 3 months to as old as 47 months. To date 35 subjects have been tested and, for all of them, the visual acuity was adequate (20/40 or better) to perform the task. Using both hands they would rotate the lever underneath (bottom half of figure) the board so that it appeared to be directly underneath the hose. Normal children would align the lever with the midline of the head, thus having it point directly to the nose. Enucleated children aligned the lever so that it was almost pointing to the remaining eye.

Subjects the egocenter's location was measured first with the left eye occluded and then with the right eye occluded. Informed consent was obtained in all cases.

Results

The average egocenter deviation of subjects whose remaining eye was the right was compared with the results of normals when they fixated with the right eye. Similarly, for those subjects with only a left eye, their egocenter deviation results were compared to those of normals who fixated with their left. As can be seen in Table 1, for the 17 subjects with a remaining right eye a mean deviation of the egocenter of 1.7 cm (SD 0.9) to the right was obtained. Measurements from the 33 normal right eyes gave a mean deviation
Table 1. Mean (SD) egocenter location in normal and enucleated children (in cm; 0 is midline, + is to the left)

<table>
<thead>
<tr>
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<th>Left eye</th>
<th>Right eye</th>
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<tbody>
<tr>
<td>Normals (N = 33)</td>
<td>0.7 (1.3)</td>
<td>0.1 (1.3)</td>
</tr>
<tr>
<td>Enucleates (N = 35)</td>
<td>1.5 (0.8)</td>
<td>-1.7 (0.9)</td>
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of 0.1 cm (SD 1.3). These locations of the egocenter were, when tested statistically, significantly different (df = 48, t = 4.59, P < 0.001). The 18 subjects whose remaining eye was the left showed a mean deviation of the egocenter of 1.5 cm (SD 0.8) towards the left. From the measures of the left eyes of normals a mean deviation of the egocenter of 0.7 cm (SD 1.3) was obtained. These differences were also significantly different (df = 49, t = 2.56, P < 0.001). The enucleates' egocenter shift did not quite reach to the remaining eye, but rather indicated a shift that was on average about 75% of the distance from the midline to the remaining eye.

When the tests were taken with the hose aligned to the child's actual midline, the measures for both groups were in the expected direction: for normals, when fixing with the right eye the hose appeared to point towards the left eye and vice versa when fixation was with the left eye. For the enucleates the midline measures were similar but displaced in a direction consistent with the shift observed in the location of the egocenter.

To see if there was a correlation of egocenter shift with amount of binocular visual experience we compared age at enucleation against the shifts obtained. Figure 2 shows only a weak, nonsignificant trend in the appropriate direction. There is no evidence for a critical period in the development of the egocenter. In addition, there is no relationship between the amount of time the child has been monocular and the amount of shift in the egocenter (Fig. 3).

Children with bilateral retinoblastoma usually have the disease diagnosed at an earlier age (its presence in family members usually makes parents and doctors more vigilant). In our population, the 12 bilateral cases were enucleated at an average age of 19 months while the unilateral cases were enucleated at 24 months. These differences were not significant, nor were there any differences in the amount of egocenter shift between the two groups. The presence of bilateral disease did not influence our results.

Discussion

This study has shown that when an eye is enucleated a significant deviation in the location of the egocenter in the direction of the remaining eye does occur. Indeed, the shift is, on average, 75% of the distance from the true midline to the remaining eye. The data also suggest that this adaptability is not restricted to a critical developmental period of a few months but rather may extend even beyond 4 years of age. One caution is that we cannot be certain at what age these children actually became visually monocular. By the time of enucleation, the tumor has grown to a size which usually precludes any useful vision. The children who were enucleated in their third or fourth year, however, probably had significant visual experience for the first 2 years. And the children who had tumors located in the peripheral retina also would have had considerable central visual experience before the tumor grew to the size at which it would have been detected.

There is other evidence of extended sensory plasticity in the visual system. For example, Moidell and...
Bedell have shown that in some normal, adult observers small changes in peripheral oculocentric direction can occur. Therapeutic intervention to reduce the amblyopia of strabismics and anisometropes has had some success even in adults. And an anatomic study of the brains of three adults enucleated while they were adults (but many years prior to their death from nonneurological causes) showed differences in ocular dominance columns compared to normals who were binocular at the time of their death. There may be some functions whose plasticity extends well beyond those traditionally defined critical periods.

We are left with the question of when the egocenter shifted in these children. Did it occur in the first week following enucleation? The first hours? We have some evidence for plasticity in the first few days following enucleation in adults. Steinbach et al measured spatial localization changes (using an open-loop pointing task) and found some shifts as early as 24 hr after the enucleation. Unfortunately, their task was completely different from the one used here to test children so we cannot make any comparisons. It remains to be tested, using just-enucleated patients or monocularly patched normal controls and the same egocenter localizing task, whether this center for visual direction is as plastic as it appears to be from the studies of enucleated children.

Key words: egocenter, visual direction, enucleation, sensitive periods, critical periods, visual deprivation, retinoblastoma

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References

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