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Eye movements of the blind.

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We investigated a group of patients who were blind because of disease affecting the anterior visual pathways. All subjects showed an inability to maintain steady eye position, with a consequent jerk nystagmus. Blindness from birth was associated with an impaired vestibulo-ocular reflex and inability to voluntarily initiate saccades, although quick phases of nystagmus were maintained. Acquired blindness was associated with relatively preserved vestibulo-ocular responses and the ability to initiate voluntary saccades and smoothly track self-moved targets. Certain features of the eye movements of the blind are similar to those due to cerebellar dysfunction.

Certain aspects of ocular motor control have been better characterized by studying the eye movements of normal individuals made during darkness (the "open loop" condition). We wondered how chronic deprivation of visual feedback due to blindness might affect the neural guidance of ocular movements. Bartels[1] noted that the blind invariably show oculomotor abnormalities and that those who had lost vision at an early age lacked the ability to voluntarily direct their eyes. Ohm[2] noted both pendular and jerk nystagmus, which he ascribed to a vestibular imbalance. We have attempted to assess how visual feedback influences the development and maintenance of function of each of the specific oculomotor subsystems (saccadic, pursuit, and vestibular).

Subjects and methods. We observed the eye movements of 18 blind subjects, 18 through 61 years of age, who were either employed or undergoing training at a vocational center for the blind. Seven subjects had no light perception (four since birth), and none had visual acuity better than 20/200. Loss of vision was due to a variety of abnormalities of the anterior visual pathways, including retrolental fibroplasia, glaucoma, trauma, and congenital rubella. We supplemented our clinical observations with motion pictures. We had only limited success with electro-oculography (EOG), since the ocular diseases from which most subjects suffered usually attenuated the corneoretinal potential. When EOG was possible, an approximate calibration was obtained with ±45° used for extremes of lateral gaze.

Results. All subjects showed a continuous nystagmus, with slow and quick phases usually in the horizontal plane. While they attempted to maintain their eyes in the primary position, the nystagmus would typically be of high frequency (up to 5 Hz), and in many subjects it would, over the course of 20 or 30 sec, reverse direction. In one subject with partial visual loss since an early age, the nystagmus was rapid and downbeating; in another with partial visual loss due to retrolental fibroplasia, a primary torsional component was present. In several subjects with total blindness since an early age, slower vertical oscillations appeared to be superimposed upon a predominantly horizontal nystagmus. In all subjects, attempts to hold eccentric horizontal or vertical eye position caused the nystagmus to become more prominent and "gaze paretic" in type, with exponential drifts back to some null position interrupted by corrective saccades (Fig. 1, A).

Rotational stimuli produced convincing vestibulo-ocular responses in eight subjects, five of whom had some residual vision, two of whom had lost all but light perception during their teens, and one who had been completely blind for 20 years (Fig. 1, C). Subjects who had been totally blind since birth appeared to have either an absent or markedly reduced vestibulo-ocular response, although they described normal sensations of self-rotation. Voluntary saccades were preserved in those subjects who either had partial preservation

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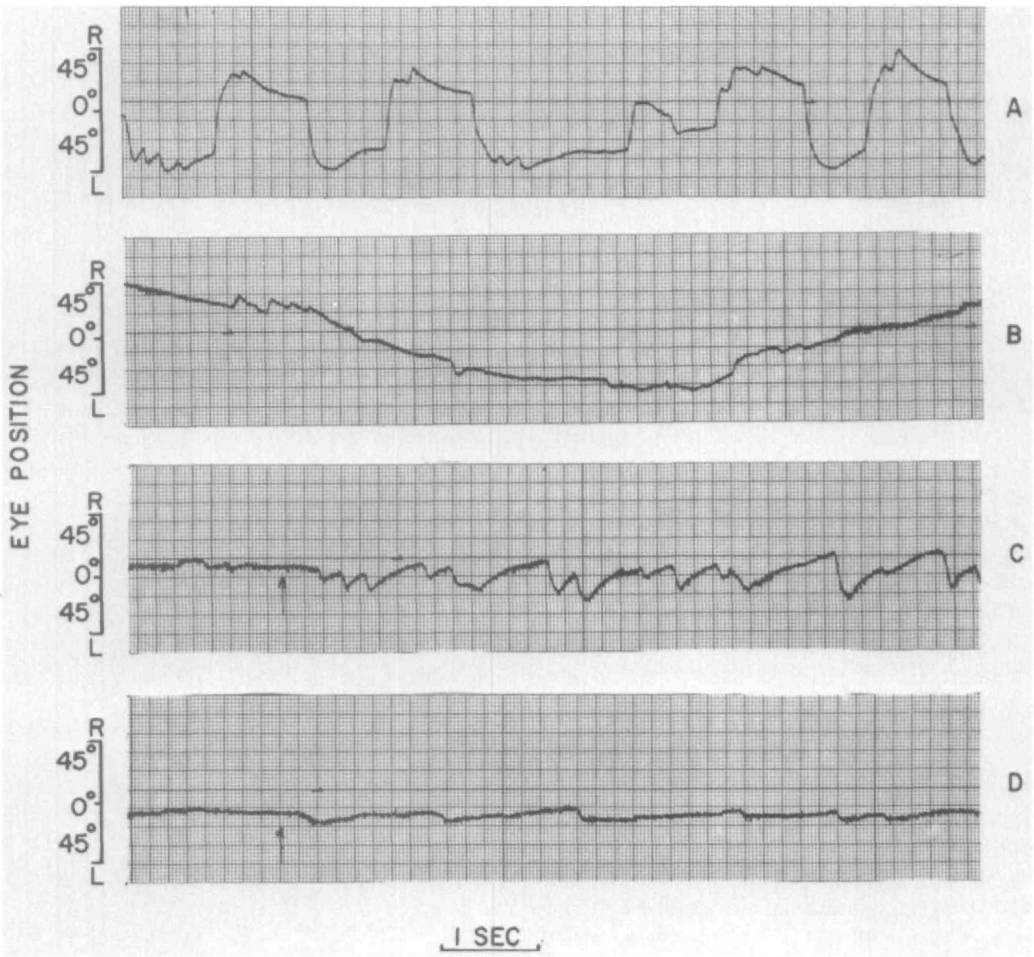

Fig. 1. Monocular EOG records from a 55-year-old subject who lost all vision during his teens due to progressive optic nerve dysfunction. Eye position is an approximate calibration based on extremes of gaze. **A**, Voluntary saccades which have normal peak velocity–amplitude relationships but which are followed by postsaccadic drift of variable time course and direction. **B**, Smooth eye movements generated when the subject attempts to pursue his outstretched hand. **C**, Ocular response to a 60°/sec counterclockwise rotation of the head. **D**, Partial cancellation of the vestibulo-ocular reflex when the subject attempts to direct his eyes towards his outstretched hand which is rotating in phase with his head. Same stimulus as in **C**.

of sight or had lost vision in later life. However, subjects usually were unable to make small saccades between the location of their outstretched thumbs and significantly overshot the targets. Saccade dynamics as assessed by the peak velocity–amplitude relationship were normal. Congenital total blindness resulted in a characteristic inability to sense the position of, and thereby consciously direct, the eyes in the orbit. When instructed to move their eyes in a specific direction, these subjects would usually thrust their heads from side to side with no accompanying modulation of their continuous nystagmus. In addition, these subjects had little voluntary control of lid movement and tended to maintain a constant partial eye closure.

Drift of the eyes after both saccades and quick phases was the rule and occurred both centripetally and centrifugally with a variable time course (time constant ranging from 200 msec to several seconds) (Fig. 1, A).

In most subjects the resting nystagmus made it difficult to detect pursuit eye movements even

when the subjects attempted to track a self-generated hand movement. However, three individuals who had vision reduced to 20/200 (one since early life) could generate some smooth following movements, and one subject produced smooth visual following movements despite the complete absence of vision for more than 20 years (Fig. 1, B). He also had other evidence of preserved visual following: he could cancel his vestibulo-ocular response during rotation by attempting to direct his eyes at his own outstretched hand which was moving with his head (compare Fig. 1, *C* and *D*).

Discussion. These results suggest that visual inputs are needed to maintain the normal performance of all classes of eye movement. Vision may also be necessary for the development of certain oculomotor subsystems. What neural systems might utilize visual information to monitor and sustain appropriate eye movements?

A clue is given by the many similarities between the eye movements of our blind patients and those of patients and experimental animals with cerebellar lesions.[3–7] Common features include inadequate gaze holding, a "wandering" null point, and postsaccadic drift. Both the vestibulocerebellum and dorsal vermis, which appear to be important for normal gaze holding, appropriate vestibulo-ocular responses, and saccadic accuracy, receive visual inputs. Our observations suggest that deprivation of visual inputs to the cerebellum and lesions of the cerebellum itself produce similar ocular motor abnormalities.

Visual inputs also appear to be important for the normal development of the vestibulo-ocular reflex since cats dark-reared for up to 1 year show a significant reduction of the gain of this reflex.[8, 9] Our subjects who had been blind from an early age had either absent or significantly diminished vestibulo-ocular responses, although more sensitive eye recording methods might have determined remnants in all. However, both partial blindness and loss of vision later in life were compatible with clear preservation of some vestibular function. This evidence suggests that visual inputs are more important in the early years of life to fashion the neural machinery responsible for the vestibulo-ocular responses.

Present results suggest that unless there has been visual experience and subsequent development of sense of eye position or change in direction of gaze, voluntary saccades cannot be made. The nystagmus quick-phase mechanism is preserved, however, with normal peak velocity–amplitude relations.

We were able to demonstrate preservation of the ability to generate smooth following eye movements in one subject despite years of blindness. This would support psychophysical evidence that both limb proprioception and possibly efference are important in the generation of smooth visual following.[10, 11] This same subject could also use similar means to modulate the gain of his vestibulo-ocular reflex. He had retained clear visual memories and commented that he was forcefully trying to imagine seeing his outstretched hand during the pursuit tasks.

In conclusion, these results emphasize the importance of visual inputs in molding and maintaining appropriate oculomotor behavior. Most impaired were oculomotor functions dependent in part on the cerebellum: steady holding of eye position between refixations and maintenance of an appropriate vestibulo-ocular reflex.

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