EVIDENCE FOR A TEST OF DYNAMIC OTOLITH FUNCTION CONSIDERED IN RELATION TO RESPONSES FROM A PATIENT WITH IDIOPATHIC PROGRESSIVE VESTIBULAR DEGENERATION

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SUMMARY PAGE

THE PROBLEM

Several explanations have been advanced concerning the persistent nystagmus that is provoked by constant velocity rotation about an Earth-horizontal axis. In one view, this response is attributed to stimulation of the otolith system, and in another, to stimulation of the semicircular canals. To obtain evidence on this issue, an examination was made of nystagmus responses elicited by Earth-horizontal axis rotation in a patient who possessed residual otolith function, but whose loss of semicircular canal function was nearly complete.

FINDINGS

The patient displayed a clear (but abnormal) nystagmus response during rotation about an Earth-horizontal axis, confirming the hypothesis that this response depends upon the otolith system. This test appears to measure dynamic otolith function and therefore provides a useful supplement to other vestibular tests.
INTRODUCTION

When man is rotated at a constant speed of 60 deg/sec about an Earth-vertical axis with the head positioned so that the horizontal canals are in the plane of rotation, primary vestibular nystagmus subsides about 40 sec after the initial angular acceleration ends. When the rotation axis is horizontal but all other aspects of the rotation are the same, nystagmus persists for as long as the rotation continues (1, 5). The important difference in these two stimulus situations is as follows: When the rotation axis is vertical, the vestibular structures maintain a constant orientation relative to gravity, and it is well known that the horizontal vestibular nystagmus in this situation depends upon stimulation of the horizontal semicircular canals by angular acceleration. In contrast, when the rotation axis is horizontal, the vestibular structures undergo continual reorientation relative to gravity, and although the initial horizontal nystagmus is partially attributable to angular acceleration, the long-persisting response derives from some additional sensory input which is not dependent upon angular acceleration but is dependent upon change in orientation relative to gravity.

This additional input comes from the vestibular structures; men with bilateral loss of labyrinthine function do not yield an unequivocal nystagmus response during rotation about an Earth-horizontal axis, and their subjective experiences are quite different from those of men with normal function (5). Conflicting views have been expressed concerning the specific vestibular mechanisms responsible for this kind of persistent nystagmus. Benson and Bodian (1) postulated a novel mode of semicircular canal stimulation, whereas Guedry (5) suggested that the response is caused by stimulation of the otolith organs. Two animal experiments support the latter hypothesis. Janeke et al. (6) found that sectioning the utricular nerve in rabbits abolished sustained nystagmus during rotation about an Earth-horizontal axis. Correia and Money (2) reported that a clear nystagmus response during rotation about an Earth-horizontal axis was still present in cats after all six semicircular canals had been blocked, even though no response could be elicited in these operated animals by strong angular acceleration about an Earth-vertical axis. Taken together, these two experiments on animals indicate that the sustained nystagmic response during rotation about an Earth-horizontal axis is provoked by the otolith system. Recent study of a clinical case appears to extend this conclusion to man. A complete description of the patient appears elsewhere (4); only the findings relevant to the present discussion are summarized herein.

CASE REPORT

The subject, a healthy appearing man 20 years of age, had always been well and active except for persistent dizzy spells since childhood, which had been ascribed to "nerves." After a careful evaluation of the vestibular organs at the Naval Aerospace Medical Research Laboratory in Pensacola, a diagnosis was made of "idiopathic progressive vestibular degeneration." Hearing was normal. Variability in test results and spontaneous attacks of dizziness and vertigo indicated that the pathological process was active at the time he was being tested. Three years later, loss of vestibular function was found to be virtually complete.
The significance of this case to the present discussion lies in the pattern of functional loss at the time the tests were conducted. Thermal stimulation and strong angular acceleration about an Earth-vertical axis were used to assess semicircular canal function. These tests indicated that horizontal canal function was either severely reduced or absent. Otolith function was assessed by the ocular counterrolling (7) and the oculogravic illusion (3) tests. Scores on the former were low, but normal, and on the latter were well within the normal range. Thus both tests indicated the presence of otolith function.

The patient was rotated at 10 rpm about an Earth-horizontal axis for 90 sec on two separate days. On each occasion he displayed a clear nystagmus response and thus differed from men without labyrinthine function who do not yield any clear nystagmus in this situation. However, the patient's nystagmus response also differed from the response elicited from normal individuals, as illustrated in Figure 1. During rotation in a clockwise direction, a normal subject displays a right-beating nystagmus which is diminished as he rotates through the right-ear-down position and is augmented as he rotates through the left-ear-down position. During counterclockwise rotation, he displays a left-beating nystagmus which is diminished as he rotates through the left-ear-down position and enhanced as he rotates through the right-ear-down position (Figure 1, upper panel). On the first day of testing, the patient displayed a nystagmus which reversed direction from right-beating to left-beating just after the nose-down portion was passed, irrespective of the direction of rotation (Figure 1, middle panel). Ten days later the patient was tested again. On this day he had experienced dizziness prior to testing, and he possessed a strong left-beating spontaneous nystagmus. During clockwise rotation about an Earth-horizontal axis, he exhibited the same type of direction-reversing nystagmus he had displayed on the first day of testing. During counterclockwise rotation, his nystagmus response was comparable to that of a normal individual (Figure 1, lower panel). Thus the positional modulation of his response to clockwise rotation was abnormal on both days, while his nystagmus during counterclockwise rotation changed from one test to the next, indicating that the vestibular response system was in a state of change, probably due to an active pathological process.

DISCUSSION

The nature of the pathological process was such that, at the time he was tested, the patient still possessed otolith function, while horizontal canal function had been lost and vertical canal function was severely reduced. Thus his vestibular system functionally resembled those of the cats whose otolith function remained intact after their semicircular canals had been surgically blocked by Correia and Money (2). However, in at least one important respect, his vestibular system probably differed from those of Correia and Money's cats. Correia and Money were able to block the semicircular canals without damaging the ampullae. Therefore, it is likely that spontaneous inflow from the horizontal canal cristae was normal. On the other hand, the pathological process seen in this patient can reasonably be presumed to have eliminated spontaneous activity. The fact that the patient displayed a clear horizontal nystagmus response during rotation about an Earth-horizontal axis supports the conclusion that this response
Comparison of patient's response with a typical normal response. Nystagmus illustrations are drawings (not actual records) to clarify the nature of the differences between the patient and a normal person. Second test was performed 10 days after first.
depends upon the otolith system. In addition, the indication is that the response may be independent of spontaneous inflow from the semicircular canals.

On the basis of present evidence, it appears that Earth-horizontal axis rotation, which yields an easily quantified nystagmus response, might provide a useful supplement to other tests of otolith function.
REFERENCES


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A patient is described who possessed residual otolith function, but whose loss of canal function was complete for the horizontal and nearly complete for the vertical canals. A clear (but abnormal) nystagmus response was elicited during rotation about an Earth-horizontal axis, confirming the conclusion, based on animal experiments, that this response depends upon the otolith system. This test appears to measure dynamic otolith function and therefore provides a useful supplement to other vestibular tests.
Vestibular apparatus

Linear acceleration

Otolith function

Nystagmus

Otolith test