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THE VESTIBULO-OCULAR REFLEX AND VELOCITY STORAGE IN SPINOCEREBELLAR ATAXIA 8

J.H. ANDERSON[1*], M.C. YAVUZ[2], B.M. KAZAR[3], P. CHRISTOVA[1], AND C.M. GOMEZ[4]

[1] Department of Otolaryngology, University of Minnesota, Minneapolis, MN, USA; [2] Department of Computer Science and Engineering, University of Minnesota, Minneapolis, MN, USA; [3] Department of Electrical and Computer Engineering, University of Minnesota, Minneapolis, MN, USA and [4] Department of Neurology, University of Minnesota, Minneapolis, MN, USA

INTRODUCTION

The vestibulo-ocular reflex (VOR) helps to maintain gaze, the position of the eyes in space, during head movement. During brief, transient rotations the angular VOR causes the eyes to rotate opposite to the head rotation, thereby helping to keep the visual world stable on the retina. However, when the movement of the head is sustained, the VOR will no longer be present. After reaching a constant head velocity, the eye velocity will decay to zero in darkness. The characteristics of this decay are determined in part by a neural network, referred to as velocity storage (15, 17), which helps to prolong the time course for decay so that the VOR continues even after the neural activity of the vestibular afferents has returned to baseline. In addition, there can be a reversal in the direction of the eye velocity (i.e., the eyes move in the same direction as the head) that is dependent on both adaptation in the transduction process and the brainstem network (7, 8).

The aim of the present work was to develop a model for the decay and reversal of the horizontal VOR in patients who have spinocerebellar ataxia (SCA) subtype 8 (13). Clinical evaluations and MRI (5) indicate that the pathology in SCA8 includes a degeneration of the cerebellum, a structure that is important for controlling the gain and direction of the VOR (19) and the spatial relationship of the VOR to a gravitoinertial reference frame (11, 16). The SCAs are a group of neurodegenerative diseases characterized by progressive instability of posture and gait, incoordination, ocular motor dysfunction, and dysarthria due to degeneration of cerebellar and brainstem neurons. Recent studies have established that there are more than 20 genetically distinct subtypes, and clinical observations suggest that eye movements and postural stability are universally but differentially impaired across the subtypes. In SCA8 the pathology seems primarily to involve the cerebellum and not other CNS structures.

Previous studies with the SCAs and episodic ataxia II have shown that otolith-ocular reflexes (1, 4, 6) and visual-vestibular interactions (18) are impaired, and there can be either an increase (9) or decrease (3) in the gain of the angular VOR,

Address for correspondence: Dr. J.H. Anderson, Department of Otolaryngology, University of Minnesota, Mayo Mail Code 396, 420 Delaware St. S.E., Minneapolis, MN 55455,USA