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Second Edition

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Electronystagmography/ Videonystagmography (ENG/VNG)

Second Edition

Devin L. McCaslin, PhD





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Foreword

I had been doing ENG at Ohio State for a couple of years when my department chairman said to me, "Charlie (he always called me 'Charlie'), why don't you write a book about this?"

I said, "But . . . but . . . Doctor Saunders (I always called him 'Doctor Saunders'), I've only been doing ENG for a couple of years."

He said, "Very good, Charlie. I'll put you in touch with my publisher."

His publisher did agree to publish the book, but first I had to get a physician to be co-author. My first choice was Hugh Barber of Sunnybrook Hospital in Toronto. He conducted the most elegant dizziness clinic I've ever attended. Patients came to him from all over Canada and many were desperate. He entered the exam room trailing a retinue of fellows and residents and other hangers-on like me and spent considerable time with each patient—probing, inquiring, listening, reassuring—and many of them were in tears when we left. Once outside, he went over his findings, diagnosis, and treatment plan with us, and his clinical acumen often blew us away.

So I called him up and said, "Doctor Barber (I always called him 'Doctor Barber'), would you perhaps agree to be co-author with me on a book about ENG?"

To my great surprise, he said, "Sure, Charlie (he always called me 'Charlie'), let's give it a go." So began a collaboration that lasted for three decades.

Our book, *Manual of Electronystagmography*, was published in 1976, reissued as a second edition in 1980, and dropped by the publisher in 1983. Thereafter it seemed to have a life of its own. Del Bloem of ICS Medical published a paperback edition for many years, and I've heard there were bootleg versions as well. In those days, I conducted ENG courses and attendees would sometimes ask me to autograph their copies of the book, often tattered and held together with rubber bands. By then, *Manual of Electronystagmography* had gotten seriously out of date. Eye movements

were recorded on strip charts and nystagmus slow phase eye velocity was measured with a ruler. There was no mention of videonystagmography or the various forms of BPPV or their treatments and nothing about the advances in eye movement neurophysiology in the last 35 years. A new book was long past due.

So along comes this young fella, Devin McCaslin (Master's degree in Audiology from Wayne State University, PhD in Hearing Science from The Ohio State University, currently Director of the Vestibular and Balance Program at the Mayo Clinic in Rochester, Minnesota). Dr. McCaslin published *Electronystagmography/Videonystagmography* (*ENG/VNG*) in 2012 and it was quite a book—well reviewed, widely read by clinicians, and widely adopted by instructors of ENG/VNG courses.

I can think of three reasons why Dr. McCaslin's book was so good:

- 1. Dr. McCaslin is an experienced clinician. He has spent many years doing ENG/VNG and continues to see patients despite many other demands upon his time. At one point in my own career, I decided I needed to spend my precious time writing and teaching, so I stopped seeing patients. That was a mistake. I quickly got out of touch and ended up mostly parroting hearsay. Soon I began seeing patients again and thereafter became a better writer and teacher. Dr. McCaslin hasn't made that mistake. He writes from firsthand experience in the clinic. You can tell.
- 2. Dr. McCaslin is a serious scholar. He reads and winnows basic research on the neurophysiology of eye movements, and as a consequence, is able to tell us the causes (insofar as they are known) of the abnormalities we see in the clinic. This information alone is worth the price of the book.
- 3. Dr. McCaslin is a lucid writer. To be a lucid writer, you must of course be a lucid thinker,

but that's not enough. First drafts are always terrible (at least mine always were). So you have to lay down the hours, painstakingly going over every sentence again and again until the words on the page say exactly what you mean. Dr. McCaslin does that.

Now we have before us the second edition of Dr. McCaslin's book, and it's even better than the first. Same firsthand clinical experience, same serious scholarship, same lucid writing, but now there's updated and expanded information on eye movement neurophysiology and a new set of illustrations. Also new is a whole chapter listing common (and some not so common) dizzinesscausing disorders. Dr. McCaslin has outdone himself here. For each disorder, he provides historical background, pathophysiology, clinical presentation, laboratory findings, and treatment. He also provides several useful appendices—a dizziness questionnaire, suggested alerting tasks to be used during caloric testing, a table listing reliability and localizing value of various ENG/VNG findings, and best of all, a delightful brochure for children describing the ENG/VNG procedure. (With minor modification, I think this would also work for adults.)

Who is this book for? It should be required reading for all clinicians who perform ENG/VNG and for all students who aspire to do so. It should also be read by referring physicians. ENG/VNG test results alone rarely yield a diagnosis, but they often provide useful information to physicians who understand their implications and relate them to other medical data available for diagnosis. One need not look elsewhere to find cogent descriptions of ENG/VNG results and their implications. They're in this book. I learned a lot. You will too.

> Charles W. Stockwell, PhD July, 2019

The ENG/VNG examination is comprised of a number of tests that each evaluates a different aspect of the balance system. Findings are then used to assist the physician or therapist in the diagnosis and treatment of the dizzy patient. The first edition of this text, and now the second edition, is written with the purpose of providing a resource "handbook" that provides practical descriptions of the tests for the practicing clinician, as well as for the graduate student first learning about the ENG/VNG examination. In order to achieve this goal, the book is written from the perspective of a clinician with the intent of providing a text that can be used in the clinical environment.

What's new in the second edition? Based on feedback from readers of the first edition, a significant amount of information has been added to select chapters. Chapter 1 has been extensively expanded by providing a deeper level of detail to the practical anatomy and physiology of the ocular motor system. This chapter also incorporates new illustrations to supplement the additional text. In Chapter 3, new material has been added regarding recently developed questionnaires that can be utilized for assisting with the case history, as well as the addition of tests that can be used to assist the clinician at the bedside. Chapter 4 has added new information with the addition of more examples and descriptions of commonly encountered ocular motor disorders and their underlying pathophysiological mechanisms. In Chapter 5, an entire set of new illustrations are provided to better guide the clinician in the testing and treatment of benign paroxysmal positional vertigo. The ter-

minology associated with positioning vertigo has changed since the initial edition of this text and this chapter reflects those changes. A new chapter has been added entitled Common Vestibular Disorders: Clinical Presentation. The purpose of this chapter is to provide the clinician with a basic understanding of the pathophysiology of the most commonly encountered disorders, the most frequent laboratory test findings, and currently accepted treatments for these disorders. When appropriate, Bárány Society Diagnostic criteria for the disorders are presented. Finally, 10 illustrative cases with accompanying eye movement videos have been added and can be accessed using the PluralPlus companion website. These cases are intended to be used in conjunction with the descriptions of a number of the disorders described in the text.

The ENG/VNG examination is a technically challenging set of tests where the correct interpretation by the clinician is critical to the patient's diagnosis and management. When performed and interpreted correctly, the ENG/VNG provides unique information for those managing dizzy patients and can expedite treatment. Alternatively, when test findings are misinterpreted or over-interpreted, the result can be delays in treatment or worse, inappropriate procedures being recommended. This book is written with the purpose of providing the clinician doing ENG/VNG with a readily accessible source for test protocols, interpretation of the various subtests, and the background information to make recommendations regarding the source of patients' dizziness.

Acknowledgments

This book is a product of my professional experiences with those that I have had the opportunity to work with over the years. My great friend and collaborator Gary P. Jacobson, PhD, first provided the spark that drove my interest in this area, and then he continued to guide my learning throughout the years. He is one of the early pioneers in the area of balance and continues to be a tireless advocate for our profession in this area of study.

Although this book has my name on the cover, it is ultimately my collaborations with others in the profession and what I have learned from them throughout the years that is embedded in the content. I must thank Drs. Bob Burkard,

Neil Shepard, and Paul Kileny for their friendship, kindness, support, and guidance throughout the years.

It was my mother, Laurie LaFleur, who first exposed me to this field and showed me how rewarding a profession could be. Her sound guidance throughout the years and "lead by example" approach to how to persevere, no matter how dire the situation, has been invaluable in every aspect of my life. These skills are particularly useful when writing a book.

Finally, I must thank my wife, Heather, and children, Molly and Declan, for their support and love during this project and throughout my career.

This text comes with supplementary case videos on a PluralPlus companion website. See the inside front cover for instructions on how to access the website.

- Case 1: Downbeat Gaze Nystagmus (1 video)
- Case 2: Bidirectional Gaze-Evoked Nystagmus (2 videos)
- Case 3: Hypermetric Saccades (1 video)
- Case 4: Infantile Nystagmus (2 videos)
- Case 5: Ocular Flutter (1 video)
- Case 6: Slow Saccades (2 videos)
- Case 7: Spontaneous Vestibular Nystagmus (4 videos)
- Case 8: Posterior Canal Benign Paroxysmal Positional Vertigo (1 video)
- Case 9: Square Wave Jerks (1 video)
- Case 10: Geotropic Horizontal Canal Benign Paroxysmal Positional Vertigo (2 videos)

Neural Control of Eye Movements

INTRODUCTION

One of the ways that an examiner can obtain information about a patient who complains of dizziness is to observe the patient's eye movements in response to different stimuli. In some instances, the observation of the eyes can be more sensitive than magnetic resonance imaging in localizing and identifying impairments that can result in balance disturbances. An examiner who is knowledgeable about the neurology of eye movements is afforded the ability to distinguish between impairments involving the central nervous system and the peripheral vestibular system.

HIERARCHY OF THE OCULOMOTOR SYSTEM

The neural control of human eye movements is organized in such a way as to allow an individual to explore their world in an effective way. When an object of visual interest is identified, three factors must be in play in order to observe it in detail (Schor, 2003): first, where the target is located; second, whether the target is moving or stationary; and third, whether the observer is moving or stationary. Each of these considerations must be taken into account because of the physiological limitations of the retina. The retina is composed of two types of photoreceptor cells known as rods and cones. Cones are concentrated primarily in and around the fovea, making it the part of the eye that has the highest spatial sensitivity and the part responsible for visual acuity (Figure 1–1). In this regard, when an observer wants to see a visual target with any detail, the oculomotor system (OMS) must align the two foveae so that the target falls on them. A single type of eye movement is inadequate



FIGURE 1–1. A diagram of the eye illustrates the location of the fovea.

to keep targets of interest on the foveae in all situations and is the reason why multiple eye movement systems exist. Depending on the task required to observe a target, different eye movement systems, with separate and independent neural pathways are recruited. Each of these systems employ different brain structures to process the information about the target, which ultimately converges in the "final common pathway."

Physiologists have organized the OMS into a hierarchy where each component has a different level of processing. Many authors organize the OMS into three components: (1) motor system, (2) premotor system, and (3) type of eye movement system. The motor system (i.e., the part of the system directly involved with movement of the eyes) moves the eye in the orbit and consists of the oculomotor nerves and the extraocular eye muscles. The premotor system organizes the neural input coming from higher centers (e.g., cerebral cortex and midbrain) and relays these commands to the motor system. The premotor system is located in the brainstem. Together, the motor and premotor systems comprise what has been termed the "final common pathway." Four primary control systems provide input to the final common pathway; these include the saccade, pursuit, optokinetic, and vestibular systems.

These systems all work together to enable an observer to clearly perceive objects of interest and explore the surrounding environment. First, the saccade system enables an observer to quickly bring a visual target identified in the peripheral field of vision system onto the fovea. The pursuit system is recruited when a target is moving slowly and the observer wishes to track it. The optokinetic and vestibular systems work together to keep the fovea centered on a target when the head is moving. The following section discusses the actions and neural generators of each of these functional classes of eye movements.

THE FINAL COMMON PATHWAY OF THE OCULOMOTOR SYSTEM

In order for the clinician to be able to make judgments about whether an eye movement is normal or abnormal, he or she needs to have a basic knowledge about the structure and function of the six eye muscles. Through a complex series of contractions and relaxations of six extraocular muscles (EOM), the globe can move in three axes: horizontal (i.e., z-axis), vertical (i.e., y-axis) and torsional (i.e., x-axis) (Figure 1–2). The various rotations of the eye in these three directional planes are, by convention, described as ductions, versions, and vergences.

Sherrington (1947) described the final common pathway component of the OMS as being composed of the ocular motor nerves and the extraocular muscles (EOMs). When the eye is directed straight ahead, it is referred to as being in the primary position. There are six EOMs that control the movement of each eye; these include the medial rectus, lateral rectus, superior rectus, inferior rectus, superior oblique, and inferior oblique (Figure 1–3). Each EOM has a primary action that refers to its rotational effect on the eye in the primary position. There are also secondary and tertiary actions that the muscle has on the eye (Figure 1–4). The EOMs are housed within the bone of the orbit. For each eye, these six muscles each have an opposing counterpart comprising three pairs. Each muscle in a pair moves the eye in the same plane, but in the opposite direction. The pairs are the medial rectus and lateral rectus, superior rectus and inferior rectus, and the superior and inferior oblique. If the eye is to be moved, the opposing counterpart (i.e., antagonist muscle) must be relaxed, and the muscle pulling the eye in the direction of interest (i.e., agonist muscle) must be contracted. That is, the agonist muscle will pull the eye in the direction of the target, and the antagonist muscle moves the eye away from the object of interest.

The arrangement of the EOMs enables the eye to be moved in the horizontal plane (back and forth) and the vertical plane (up and down). A third type of movement is torsional. This is a rotation of the eye around the line of sight (an imaginary line that connects the eye with the target). Sherrington's law of reciprocal innervation refers to the process that when an agonist muscle is contracted, a simultaneous equivalent relaxation must take place in the corresponding antagonist muscle. This process is mediated primarily through neural structures in the brainstem.



sor eye movements that rotate the eye in the y-axis. D. Elevator eye movements that rotate the eye in the y-axis. E. Lateral rotational eye movements that rotate the eye in the x-axis. F. Medial rotational eye movements that rotate the eye in the x-axis. (By Eye_movements_abductors.jpg: Patrick J. Lynch, medical illustrator derivative work: Anka Friedrich [talk]—Eye_movements_abductors.jpg [A], Eye_movements_adductors.jpg [B], Eye_movements_Depressor.jpg [C], Eye_movements_elevators.jpg [D], Eye_movements_lateral_rotators.jpg [E], Eye_movements_media_rotators.jpg [F], CC BY FIGURE 1-2. A. Abducting eye movements that rotate the eye in the z-axis. B. Adducting eye movements that rotate the eye in the z-axis. C. Depres-2.5, https://commons.wikimedia.org/w/index.php?curid=7493751)



FIGURE 1–3. The six extraocular muscles of the eye. (Courtesy of Patrick Lynch, Yale University School of Medicine)

Herings law of equal innervation refers to the two eyes being yoked during an eye movement. Specifically, the law states that during a conjugate movement of the eyes, the paired agonist muscles and antagonist muscles must receive equivalent neural input so that both eyes move together.

When patient's OMS is being evaluated clinically, the eye movements are all referenced to the primary position, which is when the eyes are in their natural resting state looking forward. When patients deviate their gaze eccentrically (i.e., right, left, up, or down) the eye movement is referred to as a secondary position. When an observer gazes up and to the right or down and to the left, these are considered tertiary positions (see Figure 1–4). These types of actions of the EOMs allow for movement of the eyes in three directional planes: horizontal, vertical, and torsional (Table 1–1). In



FIGURE 1–4. The different gaze positions and the primary agonist muscles that move the eye into position. MR = medial rectus; LR = lateral rectus; SR = superior rectus; IR = inferior rectus; SO = superior oblique; IO = inferior oblique. (From Barber & Stockwell, 1976)

the real world, the majority of eye movements are complex requiring various levels of activation and inhibition of all the EOMs. A comprehensive overview of this topic is given by Leigh and Zee (2006).

Cranial Nerves and Nuclei of the Oculomotor System

The EOMs are innervated by oculomotor neurons (OMNs) located on each side of the midline of the brain (Figure 1–5). These cranial nuclei receive eye movement information from the premotor center and relay it through projections to innervate the EOMs (Figure 1–6). The cell bodies of these nerves form the three oculomotor nuclei: the third nucleus (oculomotor), the fourth nucleus (trochlear), and the sixth nucleus (abducens nucleus).

Oculomotor Nerve (Cranial Nerve III)

The oculomotor nuclei are located in the dorsal midbrain near the floor of the third ventricle. From the periaqueductal gray matter of the midbrain, the nerve passes through the medial longitudinal fasciculus (MLF) and emerges from the cerebral peduncle and forms the oculomotor nerve trunk. The nerve then travels through the subarachnoid space, over the petroclinoid

Extraocular Muscle	Primary Action	Secondary Action	Tertiary Action
Medial rectus	Moves eye inward		
Lateral rectus	Moves eye outward		
Superior rectus	Moves eye upward	Rotates top of eye toward nose	Adduction
Inferior rectus	Moves eye downward	Rotates top of eye away from nose	Adduction
Superior oblique	Rotates top of eye toward nose	Moves eye downward	Abduction
Inferior oblique	Rotates top of eye away from nose	Moves eye upward	Abduction

Table 1–1. Primary, Secondary, and Tertiary Eye Movements Controlled by Extraocular Muscle