A CLASSIFICATION OF EYE MOVEMENT ABNORMALITIES AND STRABISMUS (CEMAS)

Report of a National Eye Institute Sponsored Workshop

From the Committee for the Classification of Eye Movement Abnormalities and Strabismus (CEMAS) Workshop. A complete list of participants is given at the end of this article.

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INTRODUCTION

Classification systems have been part of the biologic and natural sciences for recorded history[1-3]. Their constructions vary and have been based solely on individual anecdotal observations to structured organized prospective collections of data using sophisticated technology. Most classification systems form the basis for our understanding of natural and pathologic processes. They allow an approach to further investigations and the application of scientific methods for use in the treatment of disease. Individuals, institutions and groups of providers for a variety of purposes use classification systems, which include administrative, research, education, categorization, patient documentation, care improvement, quality assessment and communication[4, 5].

Examples of recent clinical medical classification systems that have provided for new methods of research and treatment include the International Classification of Disease Manual (ICD), The American Psychiatric Association's Diagnostic and Statistical Manual of Mental Disorders (DSM), The International Classification of Primary Care (ICPC), and The Current Procedural Terminology (CPT) coding system[6-9]. The ICD manual was initially a uniform classification of the causes of death. It is based on the official version of the World Health Organization's (WHO) 9^{th} revision, International Classification of Diseases. It is now used to report diseases and health related problems, as well as mortality. The resulting ICD-9 covers the entire medical domain. In use since January 1979, ICD-9-CM provides a diagnostic coding system that is more precise than those needed only for statistical groupings and trend analysis. A specific requirement of the Medicare Catastrophic Coverage Act of 1988 required health care professionals to include ICD-9-CM codes on their Medicare claim forms.

The International Classification of Primary Care (ICPC) is the official classification system of the World Organization of Family Doctors developed to order the medical domain into classes on the basis of their relevance for international primary care. ICPC emphasizes symptom- and complaint-diagnoses, especially at the start of episodes of care, when a more precise diagnosis may not be possible. The Current Procedural Terminology (CPT) system involves organizing procedures using five-digit codes that are used administratively by health care systems to compensate for patient care[6, 7].

There are several examples in medicine in which the development of a workable classification made way for a successful clinical contribution. Most noteworthy, and familiar to all ophthalmologists are the Diabetic Retinopathy Study and the International Classification of Retinopathy of Prematurity, both sponsored by the National Eye Institute in the 1970's and 1980's respectively[10] [11]. These classification systems have played key roles in subsequent natural history studies and collaborative clinical trials that continue to provide the ophthalmic community with important information regarding these two diseases. In addition to serving as standards for classification, these systems have been invaluable in the instruction and training of eye-care professionals.

The field of strabismus and eye movements has seen an explosion in scientific investigation and clinical expertise[12-16]. Clinicians and scientists who represent this

field come from a multitude of disciplines including, but not limited to, ophthalmology, optometry, behavioral psychology, psychiatry, neurology, otolaryngology, neurobiology, visual psychophysics, engineering and mathematics. Although there are numerous published classification systems for strabismus and eye movement disorders, there is no system that crosses scientific disciplines or cultural boundaries. For example, the diagnosis of accommodative esotropia describes one condition in New York and another in Ecuador, London or India. Definitions and defining characteristics may also change meaning across eye disciplines. This lack of uniformity interferes with cooperative effort among various disciplines involved in the care of these patients and makes collaborative multicenter natural history studies and/or clinical trials, at best difficult, and at times impossible.

This report summarizes the effort begun with the submission of an Intramural National Eye Institute Protocol in December of 1998 that has culminated in a National Eye Institute, National Institutes of Health sponsored workshop. Experts from varied disciplines and national geographic locale have established a modern and lively (potential for change and growth) classification system of strabismus and eye movement disorders. This classification system is based on current clinical and scientific information and can be used as a resource for scientists (multicenter trials), clinicians (directing patient care), and educators (student, resident, and fellow teaching).

BACKGROUND

Preliminary Work

Ocular motility classification systems include involuntary or limited ocular movements, interactions between accommodation and convergence, dynamic properties of accommodative and fusional vergence, interactions between vergence eye movements and other visual motor systems, violations of Herring's law associated with specific components of vergence, and sources of fixation disparity.

The classification and categorization of eye movement disorders and strabismus have evolved primarily as a result of clinical and historical observations, responses to treatments and eye movement recordings without uniformity or cross-discipline referencing [17-22]. The ICD-9-CM lists 78 conditions and diagnoses codes that cover the discipline of strabismus and eye movement disorders. However, these "codes" do not come with accompanying definitions. An individual practitioner or researcher bases classification coding on an individual patient at each visit. Thus, the use of this classification system precludes uniformity and promotes variability.

Currently, the clinical approach to diagnosis of strabismus has some standards. Certain tests are to be performed to determine 1) the status of the binocular sensory system ("fusion") in the habitual head position under normal viewing conditions and in the primary position at near and distance; 2) the presence and characteristics of fusional vergence, and, 3) the ocular deviations that exist at distance, different positions of gaze and at near. These are determined by the cover tests and the prism cover test. 4)

Incomitance of the strabismus, A-V phenomena and mechanical weaknesses and restrictions are assessed by observations of eye movements and duction tests. Finally, 5) the complete state of the sensory system is made based on the history, visual acuity and special subjective "sensory" examinations. This approach usually allows an accurate and reproducible examination to be performed in an objective manner, thus making it possible to compare clinical findings over time and to allow the formulation of a rational plan of management. This system of diagnosis is simple to follow and understand and economic on time. Unfortunately, classification systems are not uniform, even with the use of standard clinical examination methods[23-31]. These varied systems cannot be used for prospective multicenter clinical research trials. Recently, a classification of esotropia was developed for a multicenter trial and used during the prospective analysis of prism adaptation prior to surgical correction[32]. However, this study was multicenter but not multidiscipline.

The purpose of the CEMAS workshop and the resulting document is to provide a foundation of systematic classification of primary eye movement abnormalities and strabismus conditions that can be utilized for clinical research. The delineation of inclusion and exclusion criteria for involvement in clinical studies are often the most crucial parts of study design. This document can provide research groups with assistance in designing clinical trials for eye movement abnormalities and strabismus and begin to introduce a more specific non-eponymic nomenclature.

METHODS

After submission of a classification protocol in December of 1998, and, at the request of the director of the NEI, an informal working group consisting of 7 members (participants numbers 3, 6, 9, 13, 14, 18 and 20 listed at the end of the document) from 5 institutions met in the summer of 1999 to discuss the possibility of a strabismus and eye movement classification workshop. There was a consensus by this group that a project of this type would result in a valuable contribution to the eye care field and fields of physiology encompassing eye movements, strabismus and amblyopia. There was also a consensus that this time period was appropriate to begin this project. With the trend towards outcome-based research and multicenter therapeutic trials, uniform classification of diseases would provide valuable foundations to begin these studies. Based on the recommendation of this group the National Eye Institute agreed to sponsor a workshop.

The Plan of the 2-Day Workshop consisted of the following:

a. Preparation of a focused, formal planning document.

b. Identification of the scope of the workshop.

c. Identification of topic areas for classification.

d. Selection and organization of the classification scheme.

e. Planning of a detailed agenda.

f. Suggestions for additional committee members.

g. Three subcommittees were organized, each with a chair representing the three major diagnostic categories (i.e., Horizontal Deviations; Vertical Deviations and Special Forms of Strabismus; and Nystagmus and Involuntary Ocular Movements.)

On February 9th and 10th, 2001, 22 scientists and physicians, representing some of the Nation's most experienced clinical and basic science investigators in the diagnosis, treatment, and etiology of eye movement abnormalities and strabismus from varied disciplines (participants are listed at the end of this article) met for a two day workshop on the **C**lassification of **E**ye **M**ovement **A**bnormalities and **S**trabismus (CEMAS) at the National Eye Institute, National Institutes of Health Campus, Bethesda, MD. The following document is the result of the CEMAS workshop and subsequent collaboration by participants.

RESULTS

The design and conceptual foundation of the CEMAS classification scheme follows traditional ideas and attempts to remain clinically pragmatic. The scheme describes three broad categories of abnormalities: 1) Horizontal Deviations, 2) Vertical Deviations and Special Forms of Strabismus, and finally, 3) Nystagmus and Other Ocular Oscillations and is comprised of eight major areas. These include:

The horizontal heterotropias are classified first by direction, (i.e., eso-deviations and exo-deviations) followed by subdivision into those eso- and exo-deviations that are "comitant" and those that are "non-comitant." Throughout this classification an attempt has been made to provide some consistency with previous schemes while removing eponyms and consolidating those conditions felt to be along a spectrum of a single disease entity. Where relevant, previously used terms are indicated in square brackets (i.e., [old:].) We have used the phrase "syndrome" to specify a disease entity that has common underlying clinical criteria, but may vary with respect to some associated

findings. Each clinical condition listed first in outline form has a "description" box that outlines the major criteria for diagnosis, common associated findings, and some general comments.

GENERAL OUTLINE:

I. OCULAR MOTOR ASPECTS OF VISION

II. SENSORY ASPECTS OF BINOCULAR VISION

III. HORIZONTAL HETEROTROPIAS

A. Concomitant Esodeviations

1. Infantile Esotropia Syndrome
2. Accommodative Esotropia
    a. Pure Refractive
    b. Non-Refractive
    c. Mixed
3. Monofixation Esotropia Syndrome
4. Basic Non-Accommodative Esotropia
5. Esotropia And Visual or Neurologic Abnormality (e.g., sensory esotropia)
6. Intermittent Esotropia
7. Divergence Insufficiency Esotropia (paresis, paralysis)
8. Mixed (Partially Accommodative) Esotropia.

B. Non-Concomitant Esodeviations

1. Cranial Nerve Disease (palsy/paresis, congenital/developmental anomaly, tumor, vascular, inflammatory, trauma, immune, etc.)

a. Nuclear Location

b. Fasicular Location

c. Subarachnoid Location

d. Cavernous Sinus Location

e. Orbital Location

2. Neuromuscular Junction (Myasthenia, etc)
3. Muscular Disease (congenital/developmental anomaly, tumor, vascular, inflammatory, trauma, immune, etc.)
4. Oribital Disease (pulley, congenital/developmental, tumor, vascular, inflammatory, trauma, immune, etc.)

C. Concomitant Exodeviations

1. Infantile Exotropia Syndrome
2. Intermittent Exotropia
    a. Low Accommodative Convergence
    b. Normal Accommodative Convergence
    c. High Accommodative Convergence
3. Monofixation Exotropia Syndrome
4. Basic Exotropia
5. Exotropia Associated with Visual or Neurologic Abnormality (.g., sensory exotropia)
6. Convergence Insufficiency Exotropia

D. Non-Concomitant Exodeviations

1. Cranial Nerve (palsy/paresis, congenital/developmental anomaly, tumor, vascular, inflammatory, trauma, immune, etc.)
    a. Nuclear Location
    b. Fasicular Location
    c. Subarachnoid Location
    d. Cavernous Sinus Location
    e. Orbital Location
2. Neuromuscular Junction Disease (Myasthenia, etc)
3. Muscular Disease (congenital/developmental anomaly, tumor, vascular, inflammatory, trauma, immune, etc.)
4. Oribital Disease (pulley, congenital/developmental, tumor, vascular, inflammatory, trauma, immune, etc.)

IV. HORIZONTAL HETEROPHORIAS

1. Esophoria
    a. Divergence Insufficiency
    b. Convergence Excess
    c. Basic
2. Exophoria
    a. Divergence Excess
    b. Convergence Insufficiency

c. Basic

3. Fusional vergence dysfunction

V. CYCLOVERTICAL HETEROTROPIAS AND SPECIAL FORMS OF STRABISMUS

A. Apparent Oblique Muscle Dysfunction

1. Over-Elevation in Adduction (OEA) [Old, Inferior Oblique Overaction]
    a. Primary
    b. Secondary
2. Under-Elevation in Adduction (UEA) [Old, Inferior Oblique Underaction]
    a. Primary
    b. Secondary
3. Over-Depression in Adduction (ODA) [Old, Superior Oblique Overaction]
    a. Primary
    b. Secondary
4. Under-Depression in Adduction (UDA) [Old, Superior Oblique Underaction]
    a. Primary
    b. Secondary

B. Cyclovertical Deviations of Paretic Origin

1. Unilateral Superior Oblique Paresis (Congenital/Decompensated)
2. Superior Oblique Paresis (Non-Congenital [old “acquired’])
3. Bilateral Superior Oblique Paresis
4. Monocular elevation deficiency [old “double elevator palsy”]

5. Monocular depression deficiency [old "double depressor palsy"]

C. Dissociated Strabismus, Cyclovertical Deviation

1. Dissociated Cyclovertical Deviation

D. Restrictive/Mechanical Strabismus

1. Cyclovertical Deviations Secondary to Muscular Disease
2. Cyclovertical Deviations Associated with Orbital Bony Disease
3. Iatrogenic Cyclovertical Deviations, ("Induced Adhesive Syndromes")

E. Neuro-Myogenic Strabismus

1. Myasthenia Gravis
2. Chronic Progressive External Ophthalmoplegia
3. Internuclear Ophthalmoplegia
4. Skew Deviation

F. Special Forms

1. Co-Contractive Retraction Syndrome (CCRS, Types 1-3) [Old Duane]
2. Co-Contractive Retraction with Lower Cranial Neuropathy (CCRS, Type 4) [Old, Moebius]
3. Co-Contractive Retraction with Jaw-Eyelid Synkinesis Syndrome (CCRS, Type 5) [Old, Marcus Gunn]
4. Co-Contractive Retraction with Exotropia [Old Synergistic Divergence and "Y" Exotropia] (CCRS Type 6)
5. Restrictive Hypotropia in Adduction (RHA) [Old, Brown Syndrome]
6. Congenital Fibrosis of the Extraocular Muscles (CFEOM)

VI. CYCLOVERTICAL HETEROPHORIAS

1. Hyperphoria
2. Vertical fixation disparity
3. Latent hyperphoria
4. Pure Cyclophoria

VII. ACCOMMODATIVE DISORDERS

1. Paralysis
2. Infacility
3. Insufficiency
4. Excess

VIII. NYSTAGMUS AND OTHER OCULAR MOTOR OSCILLATIONS

A. Physiological Fixational Movements

1. Microtremor
2. Slow Drifts
3. Microsaccades

B. Physiological Nystagmus

1. Vestibular Nystagmus
2. Optokinetic Nystagmus
3. Eccentric Gaze Nystagmus

C. Pathologic Nystagmus

1. Infantile Nystagmus Syndrome (INS)
2. Fusion Maldevelopment Nystagmus Syndrome (FMNS)
3. Spasmus Nutans Syndrome (SNS)
4. Vestibular Nystagmus
    a. Peripheral Vestibular Imbalance
    b. Central Vestibular Imbalance
    c. Central Vestibular Instability
5. Gaze-Holding Deficiency Nystagmus
    a. Eccentric Gaze Nystagmus
    b. Rebound Nystagmus
    c. Gaze-Instability Nystagmus ("Run-Away")
6. Vision Loss Nystagmus
    a. Pre-chiasmal
    b. Chiasmal
    c. Post-chiasmal
7. Other Pendular Nystagmus and Nystagmus Associated with Disease of Central Myelin
    a. Multiple Sclerosis, Peliazaeus-Merzbacher, Cockayne's Perioxisomal disorders, Toluene abuse.
    b. Pendular Nystagmus Associated with Tremor of the Palate.
    c. Pendular Vergence Nystagmus Associated with Whipple's Disease.
8. Ocular Bobbing (Typical and Atypical)

9. Lid Nystagmus

D. Saccadic Intrusions and Oscillations

1. Square-Wave Jerks and Oscillations
2. Square-Wave Pulses
3. Saccadic Pulses (Single and Double)
4. Induced Convergence-Retraction
5. Dissociated Ocular Oscillations
6. Hypermetric Saccades
7. Macrosaccadic Oscillations
8. Ocular Flutter
9. Flutter Dysmetria
10. Opsoclonus
11. Psychogenic (Voluntary) Flutter
12. Superior Oblique Myokymia

E. Generalized Disturbance of Saccades

F. Generalized Disturbance of Smooth Pursuit

G. Generalized Disturbance of Vestibular Eye Movements

H. Generalized Disturbance of Optokinetic Eye Movements

DISCUSSION:

One of the most famous scientific classifications was by Carl von Linne, Linnaeus in Latin [33]. His original plant coding wed the topography of number of pistils and stamens. But in 1750 he progressed to a binomial nomenclature for animals and plants based on genus and species, for example, Homo sapiens and, like humans, who use surname and forename for identification [33]. These are both double-axis classification systems. Classification systems do not primarily provide names but they provide structure to order objects in classes according to established criteria. Identification of an

object (like a diagnosis) requires a correct name (label). There is only partial harmony in the relationship between the available classification systems and the iterations of formal administrative systems, such as the ICD-9. The ICD-9 was primarily designed to record diagnoses, causes of death and to characterize treatments, but numerous other variables are relevant in the practice of clinical medicine and clinical research. The new ICD-10, however, now provides a nomenclature of diseases recognized by the international as well as national community. Even with these recent modifications, the ICD-10 as such is not the most appropriate tool for most research needs and many administrative needs by our subspecialty[6, 7].

This document contains nomenclature that represents professional jargon regarding strabismus and eye movement disorders. The first question concerning any system is whether it will be etiologic or descriptive. If history shows anything, it is that we are not satisfied with descriptive. But, because of the severe and profound limits to our knowledge of the underlying etiology of most of these disorders, this workshop and its members have produced a hierarchical, categorical and descriptive system. We then both, "split" and "lumped" all disorders based on our current state of knowledge and traditional classification systems with a further goal of eliminating eponyms when possible. We also recognize that it is of great importance whether a diagnostic system includes only "extreme" cases or also deals with those closer to normality. We have not attempted to define the complete "spectrum" of these disorders but have provided criteria from which to group patients for future study.

It is not unusual for a medical discipline to exhibit wide disagreement on the meanings and use of terms and concepts. These differences reflect a rich diversity of experience, background and theoretical and technological perspectives among the individuals involved in the discipline. Although there are numerous reports and texts proposing multiple classifications schemes, there are no unifying paradigms covering diagnosis in the areas of eye movement abnormalities and strabismus (See Texts 1-48). Furthermore, there are no cross-disciplinary agreements on the definitions, contents, and varieties of disorders. Only through cooperative, conceptual classification and the identification of weaknesses and potential utility in classificatory models can we fuel that dialog, which will lead toward consensus and, ultimately, toward more focused research, theory-building and patient care. We have provided this document as a first step in this direction.

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DESCRIPTION BOXES

I. OCULAR MOTOR ASPECTS OF VISION

	OCULAR MOTOR SYSTEM
Criteria	**1. Normal –** Full versions and ductions, normal fusional vergence amplitudes, accurate and normal speed saccades, normal gain pursuit and vestibular movements, no pathologic oscillations or intrusions. **2. Subnormal (abnormal) –** Any disturbance in 1 above.

II. SENSORY ASPECTS OF BINOCULAR VISION

	BINOCULAR SENSORY SYSTEM
Criteria	**1. Normal -** Bifixation with normal visual acuity in each eye, no strabismus, no diplopia, normal retinal correspondence, normal fusional vergence amplitudes, normal stereopsis. **2. Subnormal (abnormal) –** 1 or more of the following; anomalous retinal correspondence, suppression, deficient to no stereopsis, amblyopia, decreased fusional vergence amplitudes. **3. Absence of Binocular Vision.**

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III. HORIZONTAL HETEROTROPIAS

A. CONCOMITANT ESODEVIAITONS

Disease Name	**1. INFANTILE ESOTROPIA SYNDROME [Old-congenital esotropia]**
Criteria	Infantile onset (first 6 months), constant, large angle esotropia, unresponsive to plus lenses, neurologically healthy child.
Common Associated Findings	Dissociated deviations, apparent oblique dysfunction, infantile nystagmus or fusion maldevelopment nystagmus, (adult strabismic with history of infantile strabismus esotropia surgery), cross fixation, alphabet motility patterns, monofixation syndrome may be present, OKN and monocular naso-temporal pursuit asymmetry.
General Comments	Monofixation syndrome is usual best treatment result possible. Spontaneous resolution is rare.

Disease Name	2. ACCOMMODATIVE ESOTROPIA
Criteria	**"REFRACITVE" Normal Accommodation Induced Convergence (Basic) (Old "normal" AC/A)** = esotropia eliminated by hyperopic spectacles **"NON-REFRACTIVE" Increased Accommodation Induced Convergence (Divergence Excess) (Old "high" AC/A)** = esotropia at near only and eliminated with plus lenses at near, e.g., bifocal. **"MIXED" = (Combination of above)** esotropia at distance and greater at near associated with hyperopia and responds to hyperopic correction at distance with bifocal for near.
Common Associated Findings	Onset usually later than one year of life, usually begins at near and is initially intermittent, apparent oblique dysfunction and alphabet motility patterns.
General Comments	Untreated condition leads to amblyopia, asthenopia, loss of any binocular function, and constriction of visual field.

Disease Name	3. MONOFIXATION ESOTROPIA SYNDROME [Old – Microtropia]
Criteria	Small angle esotropia to no tropia, macular scotoma in non-fixing eye with anomalous retinal correspondence.
Common Associated Findings	Can be primary, genetic or acquired after surgical treatment of infantile strabismus, can be associated with anisometropia, amblyopia often present, stereopsis present but poor, alternate cover test may reveal larger deviation than simultaneous cover test. Good fusional vergence amplitudes.
General Comments	Promotes stable ocular alignment and sensory status. Can deteriorate into constant, larger angle esotropia, requiring surgical treatment.

Disease Name	4. BASIC NON-ACCOMMODATIVE ESOTROPIA
Criteria	Comitant esotropia, onset after infancy, may be acute, in a healthy child with no eye or brain disease.
Common Associated Findings	Intermittent, variable deviation, may report diplopia or blurred vision, may have amblyopia, occasional apparent oblique dysfunction, alphabet motility patterns often present, no or some response to correction of hyperopia, no ocular motor signs of infantile strabismus syndrome.
General Comments	Untreated may result in amblyopia.

Disease Name	5. ESOTROPIA AND VISUAL OR NEUROLOGIC DISEASE [Old –"Sensory Esotropia"]
Criteria	Comitant esotropia in an infant or child with eye and/or brain disease.
Common Associated Findings	Onset variable, constant, variable deviation. May have diplopia, occasional apparent oblique dysfunction, no response to correction of hyperopia, may have some ocular motor signs of infantile strabismus syndrome.
General Comments	Can occur at any age.

Disease Name	6. INTERMITTENT ESOTROPIA
Criteria	Onset greater than 1 year, intermittent, moderate angle, comitant esotropia, normal vision, bifixation, healthy eye and child. Includes "cyclical" forms.
Common Associated Findings	May have diplopia, blurred vision, usually no amblyopia, no response to correction of hyperopia, no ocular motor signs of infantile strabismus syndrome.
General Comments	Some overlap with number basic non-accommodative esotropia.

Disease Name	7. DIVERGENCE INSUFFICIENCY ESOTROPIA
Criteria	Constant or intermittent esotropia, greater at distance (by at least 10 prism diopters) than at near, normal versions and ductions, low AC/A ratio, no amblyopia, bifixation at near, healthy brain and eye.
Common Associated Findings	May have diplopia, blurred vision, little or no response to correction of hyperopia, and no ocular motor signs of infantile strabismus syndrome. Reduced fusional divergence amplitudes at far, no refractive pattern.
General Comments	Occasionally a CNS tumor is underlying etiology.

Disease Name	8. MIXED (PARTIALLY ACCOMMODATIVE) ESOTROPIA
Criteria	Onset usually greater than 1 year, hyperopia with incomplete response to spectacles and bifocals. Normal versions and ductions, no bifixation, healthy brain and eye.
Common Associated Findings	Typically, constant esotropia with strong monocular fixation preference, sensory adaptations, amblyopia and poor stereoacuity are often present.

B. NON-COMITANT ESODEVIATIONS

Disease Name	1. CRANIAL NEUROPATHY, 2. NEUROMUSCULAR, 3. MUSCULAR AND 4. ORBITAL DISEASES
Criteria	Non-comitant esotropia with clinical, electrophysiologic, laboratory, or neuroradiologic evidence of cranial nerve dysfunction, neuromuscular, muscular or orbital disease.
Common Associated Findings	Saccadic aelocity/amplitude relationship subnormal, forced generation and forced duction abnormalities. Amblyopia present with young age. Common etiologies include congenital tumor, vascular, inflammatory, trauma, immune associations/syndromes, onset underlying disease/etiology dependent, diplopia, blurred vision, vision loss, suppression and ARC possible, neurologic symptoms, no response to correction of hyperopia.
General Comments	Topically diagnosed, e.g., nuclear, fasicular, sub-arachnoid, cavernous sinus, orbital structural or possibly orbital "pulley" disease. Associated syndromes may include the co-contractive retraction syndromes (CCRS), and fibrosis syndromes.

C. CONCOMITANT EXODEVIATONS

Disease Name	1. INFANTILE EXOTROPIA SYNDROME [Old-congenital exotropia]
Criteria	Infantile onset (first 6 months), constant, large angle exotropia, neurologically healthy child.
Common Associated Findings	Dissociated deviations, apparent oblique dysfunction, infantile nystagmus of fusion maldevelopment nystagmus, reduced stereopsis, alphabet motility patterns, monofixation syndrome may be present, OKN and monocular naso-temporal pursuit asymmetry.
General Comments	Monofixation syndrome is usual best treatment result possible. Spontaneous resolution is rare.

Disease Name	2. INTERMITTENT EXOTROPIA
Criteria	Onset usually greater than 1 year of life, intermittent, moderate angle deviation at both distance and near, usually normal vision both eyes and good stereopsis with bifixation, healthy eye and child. **Normal Accommodation Induced Convergence (Basic) (Old "normal" AC/A) –** near deviation within 10 prism diopters of distance deviation **Increased Accommodation Induced Convergence (Divergence Excess) (Old "high" AC/A) –** near deviation 10 prism diopters less exotropic than distance **True DE –** patch test, no or slight increase in near deviation **Simulated DE –** patch test, near deviation increases to within 10 diopters of distance deviation **Decreased Accommodation Induced Convergence (Convergence Insufficiency) (Old "low "AC/A) -** near deviation 10 prism diopters more exotropic than distance, reduced near point of convergence, reduced convergence amplitudes at near.
Common Associated Findings	Squinting in bright light, may have diplopia, may have ARC and suppression with eye deviation. Amblyopia not present, reduced positive fusional vergence.
General Comments	No consistent refractive pattern.

Disease Name	3. MONOFIXATION EXOTROPIA SYNDROME
Criteria	Small angle exotropia to no tropia, macular scotoma in non-fixing eye with anomalous retinal correspondence.
Common Associated Findings	Can be primary, genetic or acquired after surgical treatment of infantile strabismus, can be associated with anisometropia, amblyopia often present, stereopsis present but poor, alternate cover test may reveal larger deviation than simultaneous cover test.
General Comments	Can deteriorate into constant exotropia, requiring surgical treatment.

Disease Name	4. BASIC EXOTROPIA
Criteria	Comitant exotropia, onset after infancy in a healthy child with no eye or brain disease.
Common Associated Findings	Constant, variable deviation, amblyopia, occasional apparent oblique dysfunction and alphabet motility patterns often present, normal to variable accommodation induced convergence, no ocular motor signs of infantile strabismus syndrome.
General Comments	No consistent refractive error.

Disease Name	5. EXOTROPIA AND VISUAL OR NEUROLOGIC DISEASE [Old – "Sensory Exotropia"]
Criteria	Comitant exotropia in an infant or child with eye and/or brain disease.
Common Associated Findings	Onset variable, constant, variable deviation. May have diplopia, occasional apparent oblique dysfunction, no response to correction of hyperopia, may have some ocular motor signs of infantile strabismus syndrome.
General Comments	Can occur at any age.

Disease Name	6. CONVERGENCE INSUFFICIENCY EXOTROPIA
Criteria	Onset greater than 1 year, no distance exotropia with near exotropia, normal versions and ductions, vision, bifixation, healthy brain and eye.
Common Associated Findings	No family history. May have diplopia, blurred vision, and no ocular motor signs of infantile strabismus syndrome.
General Comments	No consistent refractive error.

D. NON-COMITANT EXODEVIATIONS

Disease Name	1. CRANIAL NEUROPATHY, 2. NEUROMUSCULAR, 3. MUSCULAR AND 4. ORBITAL DISEASES
Criteria	Non-comitant exotropia with clinical, electrophysiologic, laboratory, or neuroradiologic evidence of cranial nerve dysfunction, neuromuscular, muscular or orbital disease.
Common Associated Findings	Saccadic velocity/amplitude relationship subnormal, forced generation and forced duction abnormalities. Amblyopia present with young age. Common etiologies include congenital tumor, vascular, inflammatory, trauma, immune associations/syndromes, onset underlying disease/etiology dependent, diplopia, blurred vision, vision loss, neurologic symptoms, no response to correction of hyperopia.
General Comments	Topically diagnosed, e.g., nuclear, fasicular, sub-arachnoid, cavernous sinus, orbital structural or possibly orbital "pulley" disease. Associated syndromes may include the co-contractive retraction syndromes (CCRS) and fibrosis syndromes.

IV. HORIZONTAL HETEROPHORIAS

Disease Name	1. ESOPHORIA
Criteria	Esodeviation with alternate cover testing, generally reduced negative (base-in prisms O.U.) horizontal fusional vergence; can be comitant or non-comitant, no refractive pattern, normal vision, bifixation, healthy eye and child. **a. Divergence Insufficiency = greater deviation at distance** **b. Convergence Excess = greater deviation at near** **c. Basic = distance and near deviation equal** **d. Eso Fixation Disparity = deviation < 1 prism diopter (10 min/arc), phoria only, decreased fusional amplitudes +/- central suppression.**
Common Associated Findings	History of discomfort associated with visual tasks, intermittent diplopia, avoidance of reading or other intense visual tasks, car sickness, asthenopia, intermittent diplopia, may cover one eye, rubbing eyes, tearing, avoidance of near work.
General Comments	No consistent refractive error.

Disease Name	2. EXOPHORIA
Criteria	Exodeviation with alternate cover testing, generally reduced positive (base-out prisms O.U.) horizontal fusional vergence; can be comitant or non-comitant, no refractive pattern, normal vision, bifixation, healthy eye and child. **a. Divergence Excess = greater deviation at distance** **b. Convergence Insufficiency = greater deviation at near** **c. Basic = distance and near deviation equal** **d. Exo Fixation Disparity = deviation < 1 prism diopter, phoria only, decreased fusional amplitudes +/- central suppression.**
Common Associated Findings	History of discomfort associated with visual tasks, intermittent diplopia, avoidance of reading or other intense visual tasks, car sickness, asthenopia, intermittent diplopia, head tilt, may cover one eye, rubbing eyes, tearing, avoidance of near work.
General Comments	No consistent refractive error.

Disease Name	3. FUSIONAL VERGENCE DYSFUNCTION
Criteria	Low phoria, distance and near reduced positive and negative fusional vergence amplitudes in all horizontal directions.
Common Associated Findings	May be associated with head trauma and other cerebral disease, poor vergence facility testing, good stereoacuity.
General Comments	May be associated with other ocular motor (pursuit and vestibulo-ocular) dysfunction.

V. CYCLOVERTICAL HETEROTROPIAS AND SPECIAL FORMS OF STRABISMUS

A. APPARENT OBLIQUE MUSCLE DYSFUNCTION

Disease Name	**1. OVER-ELEVATION IN ADDUCTION (OED) [Old inferior oblique overaction]**
Criteria	Upshoot or over-elevation of adducted eye **Primary -** minimal or no vertical deviation. in primary gaze, normal motility of antagonist superior oblique, no head tilt, negative 3 step test absence of subjective torsion, vertical deviation is greatest in upgaze rather than downgaze, can be unilateral, bilateral more frequent, possible "V" pattern. Usually associated with horizontal deviations, onset 1-2 years of age. **Secondary** - significant vertical deviation in primary gaze, underaction superior oblique, compensatory head tilt to contralateral side, positive 3-step, excyclotorsion possible "V" pattern, may be history of head trauma or cranial nerve IV palsy, diplopia, asthenopia.
Common Associated Findings	None or may be chin up posture with "V" pattern exotropia or chin down posture with "V" pattern esotropia.

Disease Name	**2. UNDER-ELEVATION IN ADDUCTION (UED) [Old inferior oblique underaction]**
Criteria	Downshoot or under-elevation of adducted eye. **Primary -** minimal or no vertical deviation in primary gaze, normal motility of antagonist superior oblique, no head tilt, negative 3 step test, absence of torsion, vertical deviation is greatest in downgaze rather than upgaze, can be unilateral, bilateral more frequent, possible "A" pattern, usually associated with infantile strabismus, onset 1-2 years of age. **Secondary** - significant vertical deviation in primary gaze, compensatory head tilt to same side, positive 3-step, incyclotorsion possible "A" pattern, may be history of orbital trauma or cranial nerve III palsy, diplopia, asthenopia.
Common Associated Findings	None or may be chin up posture with "V" pattern exotropia or chin down posture with "V" pattern esotropia.

Disease Name	3. OVER-DEPRESSION IN ADDUCTION (ODA) [Old superior oblique overaction]
Criteria	Downshoot or over-depression of adducted eye. **Primary -** minimal or no vertical deviation in primary gaze, normal motility of antagonistic inferior oblique, no head tilt, negative 3 step. Absence of torsion, vertical deviation is greatest in downgaze, can be unilateral, bilateral more frequent, possible "A" pattern. **Secondary** - significant vertical deviation in primary gaze, compensatory head tilt to opposite side, positive 3-step, incyclotorsion possible "A" pattern, may be history of orbital trauma or, diplopia, asthenopia.
Common Associated Findings	Onset before age of 4, usually associated with esotropia or exotropia, may be present in isolation, may be associated with neurologic abnormalities (e.g., hydrocephalus, meningomyelocele, cerebral palsy). May be chin down posture with "A" pattern exotropia or chin up posture with "V" pattern esotropia.

Disease Name	4. UNDER-DEPRESSION IN ADDUCTION (UDA) [Old superior oblique underaction]
Criteria	Upshoot or under-depression of adducted eye. **Primary -** minimal or no vertical deviation in primary gaze, normal motility of antagonistic inferior oblique, no head tilt, negative 3 step. Absence of torsion, vertical deviation is greatest in downgaze, can be unilateral, bilateral more frequent, possible "V" pattern. **Secondary ("superior oblique paresis")** - significant vertical deviation in primary gaze, overaction of inferior oblique, compensatory head tilt to opposite side, positive 3-step, excyclotorsion possible "V" pattern, may be history of orbital trauma or, diplopia, asthenopia.
Common Associated Findings	May decompensate after years of stability.

B. CYCLOVERTICAL DEVIATIONS OF PARETIC ORIGIN

Disease Name	1. UNILATERAL SUPERIOR OBLIQUE PARESIS (CONGENITAL/DECOMPENSATED)
Criteria	Significant vertical deviation in primary gaze, overaction of inferior oblique, compensatory head tilt to opposite side, positive 3-step, excyclotorsion possible "V" pattern, generally good stereopsis, absence of torsional diplopia, expanded vertical fusional amplitudes, early onset, photographs with abnormal head posture evident, asthenopia, intermittent diplopia (cyclo-vertical or diagonal), loose superior oblique tendon with forced traction testing or direct observation.
Common Associated Findings	May have facial asymmetry (shallow side of face on side of head tilt), head tilt, face turn, may cover one eye, "spread of comitance."

Disease Name	2. SUPERIOR OBLIQUE PARESIS (ACQUIRED)
Criteria	Cyclo-vertical deviation in primary position, positive 3 step, underaction of paretic SO and/or overaction ipsilateral IO, compensatory head posture, excyclotorsion, "V" pattern, generally good stereopsis, torsional diplopia, no expanded vertical fusional amplitudes, history of head trauma or other acquired brain injury, asthenopia, cervical discomfort, head tilt, face turn, may cover one eye.
Common Associated Findings	May develop a "spread of comitance," may decompensate and an atrophic superior oblique muscle belly can be seen on MRI Imaging.

Disease Name	3. BIILATERAL SUPERIOR OBLIQUE PARESES
Criteria	Small vertical deviation in primary position, "V" pattern deviation with esotropia in downgaze and exotropia in upgaze, hyper deviation switches from right hypertropia in left gaze to left hyper in right gaze, excyclodeviation often exceeds 10 degrees, right hyper/incyclotropia with right head tilt and left hyper/incyclotropia with left head tilt.
Common Associated Findings	Usually associated head trauma, intermittent torsional diplopia, asthenopia, no facial asymmetry, head tilt (in asymmetric cases), or face turn (but may have chin down), may cover one eye.

Disease Name	4. MONOCULAR ELEVATION DEFICIENCY (MED) [Old Double Elevator Palsy]
Criteria	Unilateral limitation of upgaze above the midline, hypotropia on affected side, limited upgaze same in adduction or abduction, forded duction may show restriction to elevation, possible abnormal head posture, may have associated ptosis. Bells phenomenon reduced.
Common Associated Conditions	Associated amblyopia, possible history of other neurologic problems, general systemic problems, diplopia if recent adult onset, otherwise no symptoms, MRI imaging may show hypoplastic superior rectus, levator complex.

Disease Name	5. MONOCULAR DEPRESSOR DEFICIENCY (MDD) [Old Monocular Depressor Palsy]
Criteria	Unilateral limitation of downgaze below the midline, hypertropia on affected side, limited downgaze same in adduction or abduction, upper lid retraction when the uninvolved eye fixates, forced ductions may be positive.
Common Associated Findings	Possible history of other neurologic problems or general systemic problems, diplopia if recent adult onset, otherwise no symptoms, possible anomalous head posture.

C. DISSOCATED STRABISMUS COMPLEX

Disease Name	1. DISSOCIATED CYCLOVERTICAL DEVIATION
Criteria	Intermittent unilateral or bilateral upward and excyclo-excursion of nonfixating eye, increase in non-fixing eye with reading.
Common Associated Findings	Fusion maldevelopment nystagmus syndrome present, excyclotorsion, anomalous head posture, history of infantile strabismus syndromes, cosmetic concern, worse during periods of inattention, illness, ARC and suppression may be present, red glass test: red light always below the white light. Decreased stereopsis or strabismus often present in 1st degree relatives.
General Comments	Stable to more manifest deviation over time.

D. RESTRICTIVE/ MECHANICAL CYCLOVERTICAL DEVIATIONS

Disease Name	1. CYCLOVERTICAL DEVIATIONS SECONDARY TO MUSCULAR DISEASE
Criteria	Cyclovertical deviation with history of thyroid disease, infection, inflammation, tumor, trauma and positive forced duction testing.
Common Associated Findings	Chemosis, periorbital congestion, lid retraction and lid lag, exophthalmos, optic neuropathy, esotropia or exotropia may coexist, diplopia, imaging shows thickened muscle bellies, tendons spared (thyroid) tendons not spared (tumor, other immune or infection), hypotropia in primary gaze, limited supraduction.
General Comments	Treatments may help restore some binocular vision.

Disease Name	2. CYCLOVERTICAL DEVIATIONS SECONDARY TO ORBITAL FRACTURE
Criteria	Cyclo-vertical deviation after history of trauma with positive imaging and forced duction testing.
Common Associated Findings	Variable pattern deviation (with or without horizontal component) depending on muscle involvement and restrictive factors, prior facial trauma, diplopia, facial asymmetry, anomalous head position limited supraduction, facial asymmetry, relative exophthalmos or exophthalmos, saccadic velocity analysis can show restrictive or combination restrictive/paralytic pattern.

Disease Name	3. IATROGENIC VERTICAL DEVIATIONS (INDUCED "ADHESIVE SYNDROMES")
Criteria	Non-comitant cyclo-vertical deviation after history of medical/surgical treatments of the orbit (e.g., radiation, chemotherapy, muscle procedures or implants) and positive forced duction testing.
Common Associated Findings	May have a horizontal component, associated diplopia and occasional orbital pain or inflammation. May have torsional diplopia, asthenopia, a head tilt, or cover one eye.
General Comments	Many resolve spontaneously and if not may require multiple procedures to gain some binocular function.

E. NEURO-MYOGENIC STRABISMUS

Disease Name	1. MYASTHENIA GRAVIS
Criteria	Variability and fatigability in lid position and ocular motility, ptosis (variable), may be unilateral or bilateral, Cogan "lid twitch sign", upper lid retraction, apparent underaction of extraocular muscles (may mimic any ocular motor disorder).
Common Associated Conditions	Age of onset 20-40, ptosis worse after sustained upgaze, abnormalities of saccades, Tensilon test positive, ice pack test, anti-acetylcholine receptor antibodies present and diagnostic ocular motility recording response of saccades to Tensilon.
General Comments	May be associated with systemic myasthenia, other autoimmune diseases, thymoma and variable response to medical treatments.

Disease Name	2. CHRONIC PROGRESSIVE EXTERNAL OPHTHALMOPLEGIA
Criteria	Progressive loss of all extraocular movements and bilateral ptosis. Mitochondrial deletions present in muscle mitochondrial DNA.
Common Associated Findings	Other neurologic abnormalities (pharyngeal weakness, cerebella ataxia, deafness, optic atrophy, dementia), compensatory head posture diplopia, chin up posture, Bell's phenomenon is absent, ragged-red fibers on muscle biopsy. Associated cardiac conduction system/muscle abnormalities.
General Comments	Fair for treatment of ptosis, strabismus but progressively increasing deviation.

Disease Name	3. INTERNUCLEAR OPHTHALMOPLEGIA
Criteria	Brainstem lesion causing limitation of adduction of ipsilateral eye on attempted conjugate gaze away from the affected side, convergence frequently spared, other eye exhibits horizontal abducting oscillation.
Common Associated Findings	May be bilateral and patient may have diplopia, exotropia, normal stereopsis, often other clinical findings of brainstem disease (e.g., multiple sclerosis in young adults and central nervous system vascular disease in elderly). Ocular motility recordings show characteristic adduction/abduction findings.
General Comments	Modest response to prisms or surgery.

Disease Name	4. SKEW DEVIATION
Criteria	Comitant or non-comitant cyclovertical phoria or strabismus due to disease of the brainstem and/or cerebellum.
Common Associated Findings	Generally other neurologic signs are present (brain stem or cerebellar disease), usually no image tilt, diplopia, one eye may appear down and in the other up and out. Hypotropic (low) eye usually on the side of the brain lesion.
General Comments	May be transient.

F. SPECIAL FORMS OF STRABISMUS

Disease Name	1. CO-CONTRACTIVE RETRACTION SYNDROMES 1-3 (CCRS TYPES 1-3) [Old Duane Syndrome]
Criteria	Limitation of abduction and/or limitation of adduction, globe retraction (co-contraction), enophthalmos, palpebral fissure narrowing on adduction. **Type 1** - abduction markedly restricted, adduction normal or mildly restricted, orthotropia or esodeviation in primary gaze. **Type 2 -** adduction markedly restricted, abduction normal or mildly restricted, ortho or exodeviation in primary gaze. **Type 3 -** both abduction and adduction markedly restricted, esodeviation in abduction, exodeviation in adduction.
Common Associated Findings	Non-comitant esotropia or exotropia that varies markedly in gaze and at distance and near, more common in females and left eye, hyperopia is common, compensatory head posture, may be unilateral or bilateral, upshoots and downshoots of affected globe common, may have diplopia in certain positions of gaze often good stereopsis and bifixation with good vision. Can be associated with craniofacial or neck anomalies.
General Comments	Many remain stable.

Disease Name	2. CO-CONTRACTIVE RETRACTION SYNDROME 4 WITH LOWER CRANIAL NEUROPATHY (CCRS TYPE 4) [Old Moebius Syndrome]
Criteria	Non-comitant strabismus due to bilateral or unilateral 6^{th} nerve dysfunction, 7^{th} cranial nerve palsies, usually esotropia, facial weakness, "masklike" facial appearance, +/- "Poland" and tongue anomalies.
Common Associated Findings	Lack of smiling, head turn, difficulty in infancy with sucking, drooling of saliva, incomplete closure of eyelids during sleep, amblyopia is common, lagophthalmos, forced duction test positive possible deletion on long arm of chromosome 13 abnormal speech, autism, peroneal atrophy, pes cavus.
General Comments	Difficult to treat deviation.

Disease Name	3. CO-CONTRACTIVE RETRACTION SYNDROME 5 WITH JAW-EYELID SYNKINESIS (CCRS TYPE 5) [Old Marcus-Gunn jaw-wink]
Criteria	Variable Blepharoptosis and changing lid elevation/retraction when using masticatory muscles.
Common Associated Findings	Amblyopia, may have associated superior rectus "weakness", unusual head posture, lid elevation with opening of mouth. Versions: may be restriction of elevation of affected eye.
General Comments	Stable over time.

Disease Name	4. CO-CONTRACTIVE RETRACTION WITH EXOTROPIA (CCRS TYPE 6) [OLD SYNERGISTIC DIVERGENCE AND "Y" EXOTROPIA]
Criteria	Orthophoria in primary position with incomitant exotropia on attempted unidirectional horizontal gaze or vertical gaze.
Common Associated Findings	May have associated ptosis or abnormal lid movements such as lid retraction with induced exotropia. Usually good binocular function, may have compensatory head posture. Synergistic divergence is occasionally the result of CNS disease.
General Comments	Stable over time.

Disease Name	5. RESTRICTIVE HYPOTROPIA IN ADDUCTION (RHA) [Old Brown Syndrome]
Criteria	Limited elevation in eye adduction (same with versions and ductions), positive forced duction test.
Common Associated Findings	Normal elevation in abduction, minimal or no vertical deviation in primary gaze, "V" pattern exodeviation, bifixation without amblyopia, history of something wrong with eye muscles, may have diplopia, pain with adduction, possible chin elevation. Congenital or due to trauma or inflammation around the trochlear area ("click" syndrome).
General Comments	Stable over time.

Disease Name	6. CONGENTIAL FIBROSIS OF EXRAOCULAR MUSCLES
Criteria	Progressive familial disorder of limited ocular motility and blepharoptosis, histopathologic confirmation of fibrosis of the extraocular muscles.
Common Associated Findings	Significant hyperopia and astigmatism is common, absence of Bell's phenomenon, may be symmetric or asymmetric, optic nerve hypoplasia, chorioretinal coloboma, microphthalmia, head posture and chin elevation amblyopia is common, eyes fixed downward, versions restricted in all positions, usually autosomal dominant. Many forms, e.g., bilateral, unilateral, or only one muscle affected on each eye. Many gene defects now found.
General Comments	Difficult to treat strabismus.

VI. CYCLOVERTICAL HETEROPHORIAS

Disease Name	1. HYPERPHORIA
Criteria	Hyperphoria with alternate cover testing, generally reduced vertical fusional vergence, can be comitant or non-comitant, vertical vergence ranges reduced, horizontal vergence ranges reduced, no refractive pattern, possible excyclotorsion depending on etiology.
Common Associated Findings	History of discomfort associated with visual tasks, intermittent diplopia, avoidance of reading or other intense visual tasks, car sickness, asthenopia, intermittent diplopia, head tilt, may cover one eye, rubbing eyes, tearing, avoidance of near work.

Disease Name	2. VERTICAL FIXATION DISPARITY
Criteria	Reduced vertical fusional vergence, vertical deviation only of less than 1 prism diopter (10 min/arc) at distance and/or near with history of discomfort associated with visual tasks.
Common Associated Findings	Rubbing eyes, tearing, avoidance of near work, vertical associated phoria, reduced horizontal fusional vergence, intermittent diplopia, avoidance of reading or other intense visual tasks and/or asthenopia.

Disease Name	3. LATENT HYPERPHORIA
Criteria	Hyperphoria present after prolonged occlusion and reduced vertical fusional vergence ranges.
Common Associated Findings	History of asthenopia, multiple pair of eyeglasses with no relief, may cover one eye, rubbing eyes, tearing, and avoidance of near work.

Disease Name	4. CYCLOPHORIA
Criteria	Excylclophoria or incyclophoria, associated vertical or horizontal deviation, astigmatic shift from distance to near alternate cover test is positive for cyclophoria, torsion with double Maddox rod, typically horizontal and vertical fusional vergence reduced, reduced stereopsis.
Common Associated Findings	May be history of strabismus, strabismus surgery, acquired brain injury, patient presents with multiple pairs of eyewear, asthenopia, diplopia, torsional diplopia, nausea, car sickness, head tilt, covering one eye, avoiding reading or other visual activities, may be associated vertical or horizontal deviation.

VII. ACCOMMODATIVE DISORDERS

Disease Name	1. ACCOMMODATIVE PARALYSIS (PARESIS OF ACCOMMODATION)
Criteria	Markedly reduced accommodative amplitude (unilateral or bilateral) for age.
Common Associated Findings	No accommodation amplitudes may be unilateral or bilateral; high lag on dynamic retinoscopy, ipsilateral mydriatic pupil; may be ipsilateral III nerve involvement with a noncomitant strabismus.

Disease Name	2. ACCOMMODATIVE INSUFFICIENCY
Criteria	Reduced accommodative amplitude for age.
Common Associated Findings	Reduced monocular accommodative amplitudes. Probable high lag on dynamic retinoscopy.

Disease Name	3. ACCOMMODATIVE INFACILITY
Criteria	Fails ±2.00 flippers monocularly (<11cpm) and binocularly (<6cpm) (using 20/30 print at 40cm).
Common Associated Findings	Typically, normal accommodative amplitudes for age but decreased facility.

Disease Name	4. ACCOMMODATIVE EXCESS
Criteria	Difficulty clearing plus lenses at near. Lead or very low lag on dynamic retinoscopy.
Common Associated Findings	Difficulty relaxing accommodation on retinoscopy, subjective, or when viewing through plus lenses. May have EP at near and may have variable noncycloplegic retinoscopy and subjective refraction.

VIII. NYSTAGMUS & OCULAR MOTOR OSCILLATIONS

A. PHYSIOLOGIC FIXATIONAL MOVEMENTS

Disease Name	1. MICROTREMOR, 2. SLOW DRIFTS AND 3. MICROSACCADES
Criteria	Normally present.
Common Associated Findings	Normal, occasional viewable with microscopy, ocular motility recordings show small amplitude tremor, slow drifts and saccades.
General Comments	Believed to assist retinal/visual function.

B. PHYSIOLOGICAL NYSTAGMUS

Disease Name	1. VESTIBULAR, 2. OPTOKINETIC, AND 3. ECCENTRIC GAZE NYSTAGMUS
Criteria	Can be present "normally".
Common Associated Findings	No central nervous system pathology, ocular oscillation with: Vestibular (spinning, gravity testing, "cold/warm induced"); Optokinetic (foveal versus full field stimulation); Eccentric gaze (low intensity, horizontal, unsustained, ± rebound); Ocular motility recordings – slow phases are linear.
General Comments	Believed to assist retinal/visual function.

C. PATHOLOGIC NYSTAGMUS

Disease Name	1. INFANTILE NYSTAGMUS SYNDROME (INS) [Old Congenital Nystagmus and "Motor and Sensory" Nystagmus]
Criteria	Infantile onset, ocular motor recordings show diagnostic (accelerating) slow phases.
Common Associated Findings	Conjugate, horizontal-torsional, increases with fixation attempt, progression from pendular to jerk, family history often positive, constant, conjugate, with or without associated sensory system deficits (e.g., albinism, achromatopsia), associated strabismus or refractive error, decreases with convergence, null and neutral zones present, associated head posture or head shaking, may exhibit a "latent" component, "reversal" with OKN stimulus or (a)periodicity to the oscillation. Candidates on Chromosome X and 6 may decrease with induced convergence, increased fusion, extraocular muscle surgery, contact lenses and sedation.
General Comments	Waveforms may change in early infancy, head posture usually evident by 4 years of age. Vision prognosis dependent on integrity of sensory system.

Disease Name	2. FUSION MALDEVELOPMENT NYSTAGMUS SYNDROME (FMNS) [Old Latent/Manifest Latent Nystagmus]
Criteria	Infantile onset, associated strabismus, ocular motor recordings show two types of slow phases (linear and decelerating) plus high-frequency, low amplitude pendular nystagmus (dual-jerk waveform), jerk in direction of fixing eye.
Common Associated Findings	Conjugate, horizontal, uniplanar, usually no associated sensory system deficits (e.g., albinism, achromatopsia), may change with exaggerated convergence ("blockage"), head posture associated with fixing eye in adduction, no head shaking, may exhibit "reversal" with OKN stimulus, no (a)periodicity to the oscillation. Dissociated strabismus may be present. Decreases with increased fusion (binocular function).
General Comments	Intensity decreases with age.

Disease Name	3. SPASMUS NUTANS SYNDROME (SNS)
Criteria	Infantile onset, variable conjugacy, small frequency, low amplitude oscillation, abnormal head posture and head oscillation, improves ("disappears") during childhood, normal MRI/CT scan of visual pathways. Ocular motility recordings – high frequency (>10 Hz), asymmetric, variable conjugacy, pendular oscillations.
Common Associated Findings	Dysconjugate, asymmetric, multiplanar, family history of strabismus, may be greater in one (abducting) eye, constant, head posture/oscillation (horizontal or vertical), usually no associated sensory system deficits may have associated strabismus and amblyopia, may increase with convergence, head bobbing, head posture may be compensatory. Normal fundus exam decreases with increased fusion (binocular function).
General Comments	Usually spontaneously remits in 2-8 years.

4. VESTIBULAR NYSTAGMUS

Disease Name	a. PERIPHERAL VESTIBULAR IMBALANCE
Criteria	Vertigo, nausea, dizzy, oscillopsia, mixed horizontal-torsional trajectory; usually beats away from the side of a vestibular lesion, associated neurologic signs and symptoms, usually acute onset, unsteady gait.
Common Associated Findings	Saccades and smooth pursuit are relatively preserved, skew deviation and ocular tilt reaction may be associated, nystagmus increases when eyes are turned in the direction of the quick phases (Alexander's law. Suppressed by visual fixation; increased when fixation is removed. Horizontal component diminished when patient lies with intact ear down; exacerbated with affected ear down, increased or precipitated by changes in head position, vigorous head-shaking, hyperventilation, mastoid vibration or Valsalva maneuver. Bedside caloric stimulation: unilaterally impaired ability to modulate spontaneous nystagmus, MRI/CT scan of brain may show disease, ocular motility recordings show linear ("constant velocity") slow phases.
General Comments	Prognosis depends on underlying disease.

Disease Name	b. CENTRAL VESTIBULAR IMBALANCE NYSTAGMUS
Criteria	Mixed horizontal-torsional trajectory; usually beats away from the side of a vestibular lesion, associated neurologic signs and symptoms. Usually acute onset of vertigo, nausea, dizzy, oscillopsia, associated with other signs of vestibulocerebellar involvement.
Common Associated Findings	Downbeat, upbeat, torsional, horizontal, jerk seesaw, slow phases may have linear-, increasing-, or decreasing-velocity waveforms, poorly suppressed by fixation of a visual target, may be precipitated or exacerbated or changed in direction by altering head position, vigorous head-shaking (horizontal or vertical), or hyperventilation. Convergence may increase, suppress or convert to upbeat to downbeat nystagmus and vice versa. Commonly associated with impaired smooth pursuit, gaze-evoked nystagmus, gait instability and ataxia. MRI/CT scan of brain reflects underlying disease, ocular motility recordings show linear ("constant velocity") slow phases.
General Comments	Prognosis depends on underlying disease.

Disease Name	c. CENTRAL VESTIBULAR INSTABILITY NYSTAGMUS (PERIODIC ALTERNATING NYSTAGMUS)
Criteria	Vertigo, Mixed horizontal-torsional trajectory; spontaneous change direction of fast phase usually beats away from the side of a vestibular lesion, usually acute onset, can be associated with infantile nystagmus syndrome. If onset after infancy; vertigo, nausea, dizzy and oscillopsia. Associated with other signs of vestibulocerebellar involvement (e.g., platybasia).
Common Associated Findings	May be associated with periodic alternating head turns – the head turns in the direction of the quick phase, and the eyes are moved into a position in the orbit that is the same as the direction of the slow phase – so minimizing the nystagmus by Alexander's law, nystagmus cycle is usually little affected by visual fixation. Vestibular stimuli, such as head rotations, can change or transiently stop nystagmus, downbeat nystagmus and square wave jerks may become more obvious in the brief null period when the horizontal nystagmus wanes and then reverses, MRI/CT scan of brain reflects underlying disease, ocular motility recordings show linear ("constant velocity"), slow phases, or accelerating slow phases if associated with INS.
General Comments	Prognosis depends on underlying disease or association with INS.

5. GAZE-HOLDING DEFICIENCY NYSTAGMUS (Neural Integrator)

Disease Name	a. ECCENTRIC GAZE NYSTAGMUS (AND ASSOCIATED REBOUND NYSTAGMUS)
Criteria	Oscillation quick phases are directed away from central position. Associated neurologic signs and symptoms, usually acute onset vertigo, nausea, dizzy, oscillopsia, involuntary oscillation. Associated with other signs of vestibulocerebellar involvement, gaze-evoked nystagmus is induced by moving the eye into lateral or upgaze; with sustained attempts to look eccentrically, gaze-evoked nystagmus declines and may reverse direction—centripetal nystagmus. After the eyes are then returned to the central position, a short-lived nystagmus with quick phases opposite to the direction of the prior eccentric gaze occurs—**rebound nystagmus.** Frequently associated with cerebellar and brainstem disease, or drug intoxications
Common Associated Findings	MRI/CT scan of brain reflects underlying disease, ocular motility recordings show slow phases that are decelerating.

Disease Name	b. GAZE INSTABILITY ("RUN-AWAY") NYSTAGMUS
Criteria	Oscillation slow phases are directed away from central position. Associated neurologic signs and symptoms, usually acute onset involuntary oscillation, associated with other signs of vestibulocerebellar involvement, nystagmus due to slow phases that carry the eye away from central position, slow phases are accelerating-velocity, may be vertical or horizontal. MRI/CT scan of brain reflects underlying disease ocular motility recordings show slow phases that are accelerating.
Common Associated Findings	Central nervous system pathology common
General Comments	Prognosis depends on underlying disease

6. VISION LOSS NYSTAGMUS

Disease Name	VISION LOSS NYSTAGMUS
Criteria	Ocular oscillation occurring with acquired loss of vision, associated signs and symptoms of monocular or binocular vision loss, underlying disease related, symptoms of vision loss, unilateral or bilateral involuntary oscillations: ***a. Pre-chiasmal Vision Loss*** • Bilateral visual loss in children causes continuous jerk nystagmus, with horizontal, vertical, and torsional components, and a drifting "null" position. • Monocular visual loss causes slow vertical oscillations and low amplitude horizontal nystagmus mainly in the blind eye; in children, especially, pendular nystagmus of the blind eye. ***b. Lesions At The Optic Chiasm*** • Seesaw nystagmus with bitemporal visual field loss. ***c. Post-chiasmal Vision Loss– Lesions Affecting Posterior Cortical Areas*** • Low-amplitude horizontal nystagmus beating towards the side of the lesion.
Common Associated Findings	MRI/CT scan of brain reflects underlying disease, ocular motility recordings show slow phases that are linear or pendular.
General Comments	Prognosis depends on underlying disease

7. OTHER PENDULAR NYSTAGMUS ASSOCIATED WITH DISEASES OF CENTRAL MYELIN

Disease Name	a. PENDULAR NYSTAGMUS AND DISEASES OF CENTRAL MYELIN
Criteria	Ocular oscillation occurring with multiple sclerosis, Pelizaeus-Merzbacher disease, Cockayne's syndrome, peroxisomal disorders, toluene abuse.
Common Associated Findings	Associated signs and symptoms of demyelinating disease, unilateral or bilateral involuntary oscillations, may have horizontal, vertical, and torsional components; their amplitude and phase relationship determines the trajectory of the nystagmus in each eye. Phase shift between the eyes is common (horizontally and torsionally; seldom vertically)— may reach 180 degrees, so that the nystagmus becomes convergent-divergent or cyclovergent. Amplitudes often differ, and nystagmus may appear monocular. Trajectories may be conjugate, but more often are dissimilar. Oscillations sometimes suppress momentarily in the wake of a saccade. Generally greater amplitude in the eye with poorer vision. Internuclear ophthalmoplegia commonly associated, may have an associated upbeat component, may transiently stop or "reset" after large saccades. Ocular motility recordings show quasi-sinusoidal with frequency 2–8 Hz (typically 3–4 Hz).
General Comments	Prognosis depends on underlying disease.

Disease Name	b. OCULOPALATAL TREMOR OR "MYOCLONUS"
Criteria	Ocular oscillation occurring with oscillation of the palate, associated signs and symptoms of brainstem disease, usually developing slowly, following brainstem stroke, associated with hypertrophic degeneration of the inferior olive, unilateral or bilateral involuntary oscillations, may be vertical (with bilateral lesions) or dysconjugate vertical-torsional, accentuated by eyelid closure, movements of palate and other branchial muscles may be synchronized.
Common Associated Findings	MRI/CT scan of brain reflects underlying disease, ocular motility recordings show waveform as quasi-sinusoidal or cycloid at frequency 1–3 Hz (typically 2 Hz).
General Comments	Prognosis depends on underlying disease.

Disease Name	c. PENDULAR VERGENCE NYSTAGMUS ASSOCIATED WITH WHIPPLE'S DISEASE
Criteria	Ocular oscillation occurring with GI disease "whipples", associated signs and symptoms of gastrointestinal illness, occurs in patients with neurological involvement, who may have minimal systemic symptomsusually convergence-divergence, occasionally vertical; sometimes with associated oscillatory movements of the jaw, face or limbs oculomasticatory (myorhythmia), ocular motility recordings show pendular waveform frequency typically about 1Hz.
Common Associated Findings	Vertical gaze palsy similar to the clinical picture of progressive supranuclear palsy is usually also present.
General Comments	Prognosis depends on underlying disease.

8 OCULAR BOBBING –TYPICAL AND ATYPICAL

Disease Name	OCULAR BOBBING
Criteria **Minor Criteria** **History** **Symptoms** **Signs** **Examination Findings**	Classic ocular bobbing consists of intermittent, usually conjugate, rapid downward movement of the eyes, followed by a slower return to the central position. Reflex horizontal eye movements are usually absent. Associated signs and symptoms of pontine damage, inverse bobbing has an initial downward movement that is slow and the return to midposition is rapid; this has also been called ocular dipping. Reversed ocular bobbing consists of rapid deviation of the eyes upward and a slow return to the horizontal.
Common Associated Findings	Reverse ocular dipping or converse bobbing has been used to describe a slow upward drift of the eyes followed by a rapid return to central position, variants of ocular bobbing are less diagnostically specific.
General Comments	Prognosis depends on underlying disease.

9 LID NYSTAGMUS

Disease Name	LID NYSTAGMUS
Criteria	Eyelids frequently accompany upward movements of vertical nystagmus. Symptoms of central neurologic disease, bilateral involuntary lid and eye oscillations, twitches of the eyelid may accompany horizontal nystagmus, especially in Wallenberg's syndrome (lateral medullary infarction).
Common Associated Findings	Eyelid nystagmus that is evoked by convergence (Pick's sign) with medullary and cerebellar lesions, convergence effort increases innervation to the lids and so may amplify any lid nystagmus.
General Comments	Prognosis depends on underlying disease.

D. SACCADIC INTRUSIONS AND OSCILLATIONS

Disease Name	1. SQUARE WAVE JERKS AND OSCILLATIONS
Criteria	Occurs in patients with cerebral/cerebellar cortical disease, bilateral involuntary, horizontal, small, fast eye movement oscillation, biomicroscopic appearance of fast, back to back, oscillation (no slow phase), pairs of small horizontal saccades (typically < 2 deg) that take the eye away from the target and then return it within 200 msec; often occur in a series (square-wave oscillations).
General Comments	Prognosis depends on underlying disease.

Disease Name	2. SQUARE-WAVE PULSES
Criteria	Occurs in patients with cerebral/cerebellar cortical disease, bilateral involuntary, horizontal, small, fast eye movement oscillation, biomicroscopic appearance of fast, back to back, oscillation (no slow phase), saccadic intrusions that take the eye away from the target and return it within 70–150 msec.
General Comments	Prognosis depends on underlying disease.

Disease Name	3. SACCADIC PULSES (SINGLE AND DOUBLE)
Criteria	Occurs in patients with cerebral/cerebellar cortical disease, bilateral involuntary, horizontal, small, fast eye movement oscillation biomicroscopic appearance of fast, back to back, oscillation (no slow phase), brief, usually small movements away from the fixation point followed by either rapid drift back (saccadic pulse) or an immediate return saccade (double saccadic pulse).
General Comments	Prognosis depends on underlying disease.

Disease Name	4. INDUCED CONVERGENCE RETRACTION
Criteria	Clinical syndrome, vertical gaze palsy, light-near dissociation, midbrain neurologic disease, associated with dorsal midbrain lesions and vertical gaze palsy, bilateral involuntary, horizontal, dysconjugate (convergent), fast eye movement oscillation, elicited either by asking the patient to make an upward saccade, or by using a hand-held optokinetic drum or tape.
Common Finding	Each cycle of the oscillation is initiated by a disjunctive saccade that converges and retracts the eyes.
General Comments	Depends on underlying disease.

Disease Name	5. DISSOCIATED OCULAR OSCILLATIONS
Criteria	Clinical syndrome, unilateral oscillation, associated with abducting eye in INO, muscle paresis, associated signs and symptoms of midbrain lesions, associated with gaze palsy, asymmetric oscillation associated with impaired adduction, usually due to internuclear ophthalmoplegia, initiated by horizontal saccades, or quick phases in response to hand-held optokinetic drum or tape.
Common Associated Findings	Abducting eye appears to show variable nystagmus, but is actually a series of saccades with post-saccadic drifts, that are not apparent in the adducting eye. Each cycle of the oscillation is usually a fast phase overshoot followed by a slow, decelerating drift back (a saccadic pulse).
General Comments	Patching adducting (weak) eye causes the phenomenon to resolve. May be due to adaptive attempts to get the weak, adducting eye on target.

Disease Name	6. HYPERMETRIC SACCADES
Criteria	Ocular motor recordings make diagnosis, bilateral involuntary, horizontal, small, fast eye movement oscillation, biomicroscopic appearance of inaccurate "overshooting" saccades, saccades that overshoot the target with hypermetric corrective saccades.
Common Associated Findings	Associated signs and symptoms of cerebral/cerebellar cortical damage occurs in patients with cerebral/cerebellar cortical disease.
General Comments	Prognosis depends on underlying disease.

Disease Name	7. MACROSACCADIC OSCILLATIONS
Criteria	Occurs in patients with cerebral/cerebellar cortical disease, bilateral involuntary, horizontal, small, fast eye movement oscillation, biomicroscopic appearance of involuntary back-to-back saccades that interrupt fixation, oscillations (hypermetric saccades) around the fixation point that wax and wane, with an intersaccadic interval of about 200 msec.
General Comments	Prognosis depends on underlying disease.

Disease Name	8. OCULAR FLUTTER
Criteria	Occurs in patients with cerebral/cerebellar cortical disease, bilateral involuntary, horizontal, small, fast eye movement oscillation, biomicroscopic or ophthalmoscopic appearance of involuntary back-to-back saccades that interrupt fixation, intermittent bursts of conjugate horizontal saccades without an intersaccadic interval at 10-20 Hz; may be small amplitude.
General Comments	Prognosis depends on underlying disease.

Disease Name	9. FLUTTER DYSMETRIA
Criteria	Occurs in patients with cerebral/cerebellar cortical disease, bilateral involuntary, horizontal, small, fast eye movement oscillation biomicroscopic or ophthalmoscopic appearance of involuntary back-to-back saccades that occur on refixation, Intermittent bursts of conjugate horizontal saccades without an intersaccadic interval at 10-20 Hz induced by a voluntary saccade to a new fixation point, may be small amplitude.
General Comments	Prognosis depends on underlying disease.

Disease Name	10. OPSOCLONUS
Criteria	Random horizontal and/or vertical back-to-back saccades ("saccadomania"), associated signs and symptoms of cerebellar disease (e.g., dancing hands-dancing feet), combined multidirectional, horizontal, vertical, and torsional saccadic oscillations, without an intersaccadic interval, biomicroscopic or ophthalmoscopic appearance of involuntary back-to-back saccades that occur on refixation.
Common Associated Findings	Occurs in patients with neuroblastoma, paraneoplastic phenomenon.
General Comments	Depends on underlying disease.

Disease Name	11. PSYCHOGENIC (VOLUNTARY) FLUTTER
Criteria	Voluntary oscillation, neurologically normal, family history positive symptoms of induced convergence (blurring, diplopia, oscillopsia) bilateral involuntary, horizontal, small, fast eye movement oscillation associated with convergence, unsustained for more than about 30 seconds; often precipitated by convergence. Intermittent bursts of high-frequency (15–25 Hz), conjugate horizontal oscillations.
General Comments	Present in up to 15% of the population.

Disease Name	12. SUPERIOR OBLIQUE MYOKYMIA
Criteria	Recurrent episodes of monocular blurring of vision, or tremulous sensations in one eye, attacks may be brought on by looking downward, by tilting the head towards the side of the affected eye, and by blinking. Most patients with superior oblique myokymia have no underlying disease, though cases have been reported following trochlear nerve palsy, head injury, possible demyelination or brainstem stroke, and with cerebellar tumor.
General Comments	Movements of superior oblique myokymia are often difficult to appreciate on gross examination, but the spasms of torsional-vertical rotations can sometimes be detected by looking for the movement of a conjunctival vessel, as the patient announces the onset of symptoms; they are more easily detected during examination with an ophthalmoscope or slit lamp.

E. GENERALIZED DISTURBANCE OF SACCADES

Disease Name	E. GENERALIZED DISTURBANCE OF SACCADES
Criteria	Delayed initiation (increased latency or reaction time). Abnormal velocity (mainly decreased; apparent increase in some patients with myasthenia gravis) dysmetria hypometria, hypermetria, wrong direction – including lateropulsion). Mismatch of the saccadic pulse and step (leading to glissades).

F. GENERALIZED DISTURBANCE OF SMOOTH PURSUIT

Disease Name	F. GENERALIZED DISTURBANCE OF SMOOTH PURSUIT
Criteria	Increased reaction time (latency) of response to sudden onset of target motion abnormal gain (decrease or increase) of: (1) the initiation of pursuit (eye acceleration); (2) sustained pursuit (gain = eye velocity/target velocity, and phase). Abnormal pursuit is often detected clinically by the presence of corrective saccades, which may be "catch-up" ("cog-wheel pursuit") or "back-up" (when pursuit gain is > 1.0).

G. GENERALIZED DISTURBANCE OF VESTIBULAR EYE MOVEMENTS

Disease Name	G. GENERALIZED DISTURBANCE OF VESTIBULAR EYE MOVEMENTS
Criteria	Imbalance of vestibular inputs: (1) semicircular canals (nystagmus, vertigo); (2) otoliths (skew deviation and the ocular tilt reaction). Disturbance of gain (decreased or increased) of the vestibulo-ocular reflexes. Disturbance of phase of the vestibulo-ocular reflexes (including velocity storage). Abnormality of direction of the vestibulo-ocular responses (*e.g.,* perverted nystagmus).

H. GENERALIZED DISTURBANCE OF OPTOKINETIC EYE MOVEMENTS

Disease Name	H. GENERALIZED DISTURBANCE OF OPTOKINETIC EYE MOVEMENTS
Criteria	Abnormalities of the optokinetic system are evaluated by measuring optokinetic after-nystagmus (OKAN), since the optokinetic responses are mainly due to smooth pursuit. Disorders of OKAN are caused by vestibular system disorders (*e.g.,* absent OKAN following loss of vestibular function).

APPENDIX A – SPECIFIC TERMINOLOGY CHANGES

I. HORIZONTAL DEVIATIONS

1. New Terminology:—Infantile Esotropia (Exotropia) Syndrome
Old Terminology: Congenital Esotropia (Exotropia), Infantile Esotropia (Exotropia)

2. New Terminology:—Monofixation Esotropia (Exotropia) Syndrome
Old Terminology: Microtropia, Fixation Disparity, Microstrabismus

3. New Terminology:—Esotropia (Exotropia) and Visual or Neurologic Disease
Old Terminology: "Sensory" esotropia (exotropia), "acquired" esotropia (exotropia), "secondary" esotropia (exotropia).

II. CYCLOVERTICAL AND SPECIAL FORMS OF STRABISMUS

1. New Terminology:—Over-Elevation in Adduction
Old Terminology: Inferior Oblique Overaction.

2. New Terminology:—Under-Elevation in Adduction
Old Terminology: Inferior Oblique Underaction.

3. New Terminology:—Over-Depression in Adduction
Old Terminology: Superior Oblique Overaction.

4. New Terminology:—Under-Depression in Adduction
Old Terminology: Superior Oblique Underaction.

5. New Terminology:—Monocular Elevation Deficiency
Old Terminology: "Double Elevator" Palsy.

6. New Terminology:—Monocular Depression Deficiency
Old Terminology: "Double Depressor" Palsy.

7. New Terminology:—Under-Depression in Adduction
Old Terminology: Superior Oblique Underaction.

8. New Terminology:—Dissociated (horizontal, Cyclovertical) Strabismus Complex
Old Terminology: Dissociated Hyperphoria

9. New Terminology:—Co-Contractive Retraction Syndrome Types 1-3
Old Terminology: Duane Syndrome Types 1-3.

10. New Terminology:—Co-Contractive Retraction Syndrome Type 4
Old Terminology: Moebius Syndrome.

11. New Terminology:—Co-Contractive Retraction Syndrome Type 5
Old Terminology: Marcus-Gunn Jaw-Wink.

12. New Terminology:—Co-Contractive Retraction Syndrome c Exotropia Type 6
Old Terminology: .Synergistic Divergence and "Y" Exotropia

13. New Terminology:—Restrictive Hypotropia in Adduction
Old Terminology: Brown Syndrome.

III. NYSTAGMUS & OCULAR MOTOR OSCILLATIONS

A. NYSTAGMUS

1. New Terminology: Physiological Fixational Movements
Subcategories: Microtremor, Slow Drifts, Microsaccades

2. New Terminology: Physiological Nystagmus
Subcategories: Vestibular, Optokinetic, Eccentric-Gaze (End-Point and Rebound), Caloric, Caloric-After, Rotational, Post-Rotational, Head-Shaking, L-, Pneumatic, Compression, Sigma, Optokinetic After-, Optokinetic After-After, Flash-Induced, Flicker-Induced
Old Terminology: Compensatory, Perrotary, Secondary Phase, Electric, Galvanic, Faradic, Alternating Current, Optic, Optomotor, Panoramic, Railway, Train, Post-Optokinetic, Reverse Post-Optokinetic, End-Gaze, Fatigue

3. New Terminology: Infantile Nystagmus Syndrome
Subcategories: Pursuit-System (P, AP, P_{fs}, PP, PP_{fs}), Vestibular-System (J ± APAN), Unknown (J_{ef}, PC, PJ, BDJ, T, DJ), Hereditary
Old Terminology: **Congenital, Motor, Fixation, Sensory, Nystagmoid**

4. New Terminology:—Fusion Maldevelopment Nystagmus Syndrome
Old Terminology: Latent, Manifest Latent, Latent/Manifest Latent, Occlusion, Monocular Fixation, Unimacular

5. New Terminology:—Spasmus Nutans Syndrome

6. New Terminology:—**Peripheral Vestibular Imbalance**
Old Terminology: Labyrinthine,

7. New Terminology:—Central Vestibular Imbalance
Specific Terminology: Jerk, Geotropic, (A)(po)geotropic, Positional, Positioning, Alcohol-Induced
Old Terminology: Bechterew's

8. New Terminology:—Central Vestibular Instability

9. New Terminology: -Eccentric Gaze Nystagmus

10. New Terminology:—**Gaze-Instability Nystagmus ("Run-Away")**

11. New Terminology: Vision Loss Nystagmus—Pre-chiasmal, Chiasmal, Post-chiasmal
Old Terminology: Hemi-See-Saw

12. New Terminology: Pendular Nystagmus Associated with Disease of Central Myelin—MS, Peliazaeus-Merzbacher, Cockayne's Perioxisomal disorders, Toluene abuse

13. New Terminology: Pendular Nystagmus Associated with Tremor of the Palate

14. New Terminology: Pendular Vergence Nystagmus Associated with Whipple's Disease

B. SACCADIC INTRUSIONS AND OSCILLATIONS
1. New Terminology: Square-Wave Jerks and Oscillations
Old Terminology: Hopping nystagmus, Lightening Eye Movements, Myoclonus, Gegenrucke, Zickzakbewegungen

2. New Terminology: Square-Wave Pulses
Old Terminology: Macro Square-Wave Jerks, Pendular Macro Oscillations, Saccadic Nystagmus, Kippdeviationen, Kippnystagmus

3. New Terminology: Saccadic Pulses (Single and Double)
Specific Nystagmus Terminology: Saccadic Pulse Trains (runs of Saccadic Pulses), Flutter (multiple Double Saccadic Pulses)
Old Terminology: Stepless Saccades (Saccadic Pulses), Abduction Nystagmus or Ataxic Nystagmus (runs of Saccadic Pulses)

4. New Terminology: Induced Convergence-Retraction
Old Terminology: Convergence-Retraction Nystagmus, Nystagmus Retractoris

5. New Terminology: Dissociated Ocular Oscillations
Old Terminology: Abduction Nystagmus

6. New Terminology: Opsoclonus
Old Terminology: Saccadomania, Dancing Eyes, Lightening Eye Movements

7. New Terminology: Psychogenic (Voluntary) Flutter
Old Terminology: Voluntary Nystagmus, Hysterical Flutter, Hysterical Nystagmus, Ocular Fibrillation, Ocular Shuddering, Psychological Nystagmus

C. SYNDROMES
1. Bruns' Syndrome - Gaze-evoked nystagmus in the direction of a cerebellar angle tumor and jerk, vestibular nystagmus in the opposite direction (eyes open) and jerk nystagmus away from the lesion (eyes closed).
2. Lateral Medullary Syndrome - Horizontal-torsional jerk nystagmus beating away from the side of a medullary lesion (eyes open) and towards the lesion (eyes closed). Occasional gaze-evoked and uniocular downbeat nystagmus.

D. SPECIAL TYPES
1. Baers' – Nystagmus after superficial corneal lesion.
2. Bartels' – Induced jerk nystagmus after ±20D lenses and head turn away from primary position; the nystagmus direction is the same as the head turn.
3. Bechterew's – Reappearance of vestibular nystagmus with second unilateral, labyrinthine lesion, after recovery from the first such lesion.
4. Stransky's – Associated, induced nystagmus with attempted, forced opening of one eyelid.

E. GENERAL, DESCRIPTIVE TERMINOLOGY (MANY TYPES OF NYSTAGMUS AND OSCILLATIONS):

Pendular, Jerk, Horizontal, Vertical, Torsional, Cyclohorizontal, Cyclovertical, Diagonal, Elliptical, Upbeat, Downbeat, Rightbeat, Leftbeat, Clockwise, Counterclockwise, See-Saw, Periodic Alternating, Asymmetric (a)Periodic Alternating, Alternating Windmill, Centripetal, Centrifugal, Induced, Muscle-Paretic, Pursuit-After, Spontaneous, Uniocular, Gaze-Evoked, Convergence-Evoked, Intermittent, Drug-Induced, Perverted, Stepping-Around, Epileptic, Ictal, Cervical, Neck-Torsion, Vertibral-Basilar Artery Insufficiency, Bow-Tie

F. GENERAL OLD TERMINOLOGY:

Oblique, Talantropic, Circumduction, Gyratory, Radiary, Rotary, Rotatory, Cyclorotary, Cyclotorsion, Sidebeat, Alternans, Kinetic, Myasthenic, Pseudo-Spontaneous, Occupational, Minor's, Pursuit-Defect, Reflex, Deviational, Gaze-Paretic, Neurasthenic, Seducible, Setting-In, Provoked, Disjunctive, Barbiturate, Lightening Eye Movements, Pseudo-Caloric, Pseudo-Spontaneous, Somatosensory, Reflex, Incyclo-, Excyclo-, Levo-, Dextro-, and variants of all of the above.

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