

EDITORIAL

Clarity of Words and Thoughts About Strabismus

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“HOW OFTEN MISUSED WORDS GENERATE MISLEADING thoughts.” Herbert Spencer, English philosopher, 1820–1903, from *Principles Of Ethics*.¹

In this issue, Donahue and associates present a series of patients in whom well-defined neurologic disorders produced a pattern of incomitant vertical strabismus corresponding by the three-step test to the pattern traditionally believed to represent inferior oblique palsy.² Donahue and associates instead attribute the strabismus to a skew deviation, which might practically be defined as a supranuclear vertical strabismus arising from an abnormality of the ocular tilt reaction mediated by otolith-ocular reflex pathways.² Skew deviations need not be comitant, and are

See also pp. 751–756.

typically modulated by changing otolithic orientation relative to gravity during head tilt. Based on the finding of excyclotorsion in the hypotropic eye of each patient, the authors conclude that despite a three-step test indicating paralysis of the involved inferior oblique muscle, the inferior oblique is actually overacting. The reader is thus left to conclude that the three-step test, traditionally considered the gold standard for defining a weak cyclovertical extraocular muscle, may show identical findings in the presence of excessive contraction of the same muscle!

This paradox is more than a nuisance. If one considers this situation in a broad perspective, the consequences of the findings of Donahue and collaborators seriously undermine traditional thinking in the field of ocular motility. Do we really believe that both “underaction” and “overaction” of the same extraocular muscle can produce identical findings on a clinical test that we believe represents the defining measure of extraocular muscle function? We

should not be surprised if this concept confuses students and ophthalmology residents. More seriously, this sort of problem with diagnostic terminology is likely to confuse even experienced practitioners, leading to inappropriate diagnoses and treatments.

The cause of this conundrum may be traced to the history of ophthalmology as seen from the perspective of the philosophy of science. Scientific philosopher Thomas Kuhn, in his epic work *The Structure of Scientific Revolutions*, noted that scientific thinking is generally conducted within the framework of a *paradigm*.³ A paradigm is a particular approach to thinking about and working within a scientific field, such as the one of strabismus, and incorporates specific concepts regarding mechanisms. Essential aspects of a paradigm are typically embodied in the terminology used by a group of practitioners of the paradigm to describe phenomena, and the sorts of phenomena that can be accepted as evidence regarding observed behaviors. For example, relativistic physics is a modern paradigm that has supplanted an older paradigm that supposed that light waves are transmitted through an hypothetical substance called ether.

The currently prevailing paradigm in the field of strabismology is a rather venerable concept of the actions of the extraocular muscles that can be found in textbooks dating back to the middle of the 19th century in a form perfectly familiar to the modern reader. At that time, the rotations of the two eyes were observable clinically, and binocular alignment could be measured using devices such as prisms. At first, prosaic Latin terms like “strabismus sursoadductorius” were used to describe in neutral fashion phenomena, such as over-elevation in adduction, that were directly observed. There was initially no attempt to interpret the mechanism. Based on the classical gross anatomy of the extraocular muscles and orbit, theories were developed of the action of each oculorotary muscle. Clinical phenomena were observed, and astute clinicians made initial guesses as to the pathogenesis of strabismus. These guesses about pathology included “overactions” and “underactions” of specific extraocular muscles as causes of strabismus that changed in relationship to the theorized directions of action of these muscles. Since specific mechanistic causes of the observed motility abnormalities were inapparent, and since no tests were available to distinguish

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among mechanisms of muscle imbalance anyway, clinicians yielded to the temptation to abandon cumbersome Latin descriptive terms in favor of seemingly parsimonious English terms. Terms like muscle “overaction” and “underaction” became commonplace jargon for clinical findings, and appeared reasonable since they correlated with observations of eye rotations on duction and version testing. However, these definitions have always been *tautologous*, in that the mere observation of an excessive or insufficient amount of rotation of the eye in a particular direction was the defining characteristic of the muscles’ overaction or underaction, respectively. The descriptive terms such as muscle overaction and underaction have never been correlated with any other measure of the force development of an extraocular muscle. In brief, these traditional terms have never been validated.

The three-step test is another example of tautologous definition of clinical entities. Based on ingenious reasoning, the Bielschowsky head-tilt phenomenon has been extended and generalized to presumably permit the diagnosis of palsy of any of the cyclovertical extraocular muscles. Implicit in this logic is the concept that all patients begin with an array of extraocular muscles that is normal and highly stereotypic, so that each of these muscles normally acts in the same way. Further implicit is the idea that the only primary abnormality of an extraocular muscle is weakness. Neither of these assumptions seems plausible in the modern era, and it is increasingly recognized that the three-step test is unreliable for the diagnosis of many forms of cyclovertical strabismus.⁴

New scientific knowledge is highlighting the inadequacy of traditional terminology in strabismus and ocular motility. Until recently, the activity of the extraocular muscles could only be indirectly inferred from examination of the orientation and movement of the eyes, supplemented in a limited way by electromyography and force generation testing. Within the last decade, improvements in the technology of magnetic resonance imaging (MRI) have enabled the direct examination of the locations, pulling directions, size, and contractility of each of the extraocular muscles in living people.⁵ When multipositional MRI showed the paths of extraocular muscles to differ markedly from the traditionally assumed shortest paths over the globe, the anatomy of the extraocular muscles and orbital connective tissues was reexamined at the microscopic level using modern immunohistochemical techniques.⁶⁻⁸ These studies have produced a very different picture of extraocular muscle anatomy and function. For example, it is now recognized that each of the extraocular muscles passes through a pulley that redirects muscle force and acts as the muscle’s functional origin. For all of the extraocular muscles except the superior oblique, the location of this pulley is under active muscular control, and thus the pulling direction of each of these muscles is subject to change.⁷ It is now recognized that disorders of the extraocular muscle pulleys and their associated connective tissues

may cause a strabismus. In particular, static malpositioning of rectus pulleys may produce A- and V-pattern strabismus that simulates traditional patterns of oblique muscle dysfunction,⁹ and dynamic instability of pulleys can produce complex patterns of incomitance that can also simulate restrictive strabismus such as Brown’s syndrome. Pertinent to the current paper by Donahue and collaborators, MRI has in some strabismic patients demonstrated superior displacement of the lateral rectus pulley that gives the lateral rectus elevating as well as abducting action.¹⁰ Since the elevated, abducting eye is the only one that can fixate during version testing in tertiary gaze, Herring’s law will in the presence of this abnormality result in the supply of subnormal elevating innervation to the nonfixating eye in adduction. This adducting eye will underelevate and thus be mistaken to have inferior oblique weakness based on traditional thinking. Similar abnormalities of extraocular muscle pulleys are not uncommon, and have different neurologic implications and may require treatment than that appropriate for oblique muscle weakness caused by cranial neuropathy.

It is useful to reminisce upon our early experiences as medical students learning physical diagnosis. Upon questioning by the clinical professor, an eager young medical student hearing crackles in a patient’s lung base for the first time may blurt out, “Oh, the patient has pneumococcal pneumonia!” The wise old professor will typically acknowledge that pneumococcal pneumonia is indeed one possibility, but will counsel the student to consider a differential diagnosis including other causes such as pulmonary edema. We as ophthalmologists have long been guilty of similar diagnostic behavior when we have naively attributed highly specific extraocular muscle pathology, and usually cranial neuropathy, to nonspecific patterns of strabismic incomitance such as those demonstrated by the three-step test.

Some might respond that changes in our terminology are unnecessary, since sophisticated strabismologists understand that terms like “inferior oblique overaction” are really only euphonious shorthand describing a clinical observation of an ocular version abnormality (perhaps also associated with a V pattern and excyclotropia), and that sophisticated minds can now consider other pathophysiologies such as rectus pulley disease. Perhaps so, but not without a significant detriment to the clarity of our thinking. Consider the situation if general physicians elected to term all crackles auscultated in the lung base as “pneumococcal pneumonia.” Sophisticated internists could constantly remind themselves that “pneumococcal pneumonia” is sometimes due to a gram-positive bacterial pneumonitis, but that it can also be caused by other conditions such as pulmonary edema. While this peculiar use of the term “pneumococcal pneumonia” might not always be harmful when used by highly sophisticated internists, it seems highly likely to motivate residents to the excessive use of antibiotics, and insufficient use of

TABLE 1. Substitution of Descriptive for Conclusionary Terms in Ocular Motility

| Prevalent Conclusionary Term | Suggested Descriptive Term |
|------------------------------|-------------------------------|
| Inferior oblique overaction | Over-elevation in adduction |
| Inferior oblique underaction | Under-elevation in adduction |
| Superior oblique overaction | Over-depression in adduction |
| Superior oblique underaction | Under-depression in adduction |

diuretics. Internists thus appropriately use the term “crackles” for the sounds heard in the chest, and then consider a differential diagnosis for this objective finding.

We would make a good start in remedying our terminological morass if we would employ clinically descriptive terms for pathologic findings rather than terms that misleadingly imply specific pathogenic mechanisms (see Table 1). Since, for example, there are multiple reasons for an eye to exhibit “over-elevation in adduction,” we should employ this term rather than the more common, yet unjustifiably conclusionary term “inferior oblique overaction.” The observation of “under-depression in adduction” is an objective and reproducible finding not embodying any assumptions about its pathogenesis; on the contrary, the more commonly employed term “superior oblique underaction” gives the very strong implication that the finding is known to result from impaired contractility of the superior oblique muscle. Our traditional terminology, which immediately imputes causality to specific oblique muscles, invites us to disregard alternative pathologies and immediately jump to management that might only be appropriate to the condition we have by our terminology just presumed. We are really behaving like the naive medical student who diagnoses pneumococcal pneumonia based exclusively on the auscultation of rales. Our thinking would be clearer, and our patients better served, if we considered a differential diagnosis for clinical findings.

Another good start could be made in resolving our terminological problems in strabismology if we will recognize and avoid tautologous diagnoses. For example, we should recognize that the three-step test only provides diagnostic information that may identify abnormalities of cyclovertical muscles. If in the past we permitted this test to define entities such as “inferior oblique palsy” and “superior oblique palsy,” then we should reconsider the validity and definition of these diagnoses in light of modern methods of validation. We would be more consistent with the practices in general neurology and general medicine if we considered “palsy” and “paresis” to repre-

sent the absence and deficiency, respectively, of contractile force generation by a muscle. Neurologists do not diagnose weakness of specific muscles by patterns of gait, but by direct tests and observations of individual muscle contractility. Since it is now possible for us directly to observe muscle function in patients, we ought to regard this measure as the gold standard applicable to the definitions of paralysis and paresis of extraocular muscles.

Great opportunities are now developing for the application of modern techniques for the clinical study of extraocular muscle structure and function in strabismus. Application of these technologies for the benefit of our patients will require that strabismologists clear away old terminology that confounds our thinking, and adopt nomenclature that is more logical. An organized effort in this direction, sponsored by the United States National Eye Institute, is the interdisciplinary working group on the Classification of Eye Movement Abnormalities and Strabismus (CEMAS). Ophthalmologists should be open-minded about imminent improvements in the nomenclature of strabismus and related disorders of ocular motility.

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