SPECIAL ARTICLE

# "Sensory" and "Motor" Nystagmus

## *Erroneous and Misleading Terminology Based on Misinterpretation of David Cogan's Observations*

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Most patients with infantile nystagmus syndrome (INS)[1] (formerly known as *congenital nystagmus*) exhibit nystagmus with several of the waveforms first identified in 1975.[2] The past 45 years of recording and analyzing nystagmus waveforms have repeatedly demonstrated that most INS waveforms result from the same deficit in one of the several ocular motor subsystems, specifically smooth pursuit.[3-6] This applies to all patients with INS whether they have associated visual sensory deficits, are familially predisposed to have INS (ie, there is a true genetic anomaly), or exhibit INS without an associated ocular or central nervous system deficit (so-called idiopathic). Despite this eye-movement data, textbooks in ophthalmology, neuro-ophthalmology, and neurology as well as current peer-reviewed literature continue to use the terms *sensory* and *motor* nystagmus to describe the ocular oscillations of INS. Those descriptions imply not only that there are at least 2 different types of nystagmus but also that they have independent pathophysiology caused by different primary developmental deficits; both implications are unsupported by decades of ocular motor electrophysiological data. The adoption of this terminology and its presumed ability to clinically differentiate pendular from jerk waveforms are responsible for the misdiagnosis of many cases of INS. Ocular motor recordings demonstrate that owing to the complexity of INS waveforms, clinical differentiation is impossible in many patients. These recordings also show that another type of nystagmus of infancy, fusion maldevelopment nystagmus syndrome (formerly known as *latent/manifest latent nystagmus*), cannot be differentiated from INS with a latent component; Alfred Kestenbaum, MD, first noted the existence of these 2 clinically similar types of nystagmus in his 1946 textbook and its expanded 1961 second edition.[7]

In the population of infants and children with INS, most studies[8-10] report a prevalence of associated afferent visual system diseases of greater than 50%, ie, optic nerve or foveal hypoplasia, retinal disease, ametropia, and congenital cataracts. Owing to this association, it is important that an ophthalmologist evaluate the eye and visual system in these patients. A thorough examination includes testing monocular and binocular vision, testing refraction, and ophthalmoscopy. Many times additional testing is indicated, ie, formal color vision and visual fields, dark adaptation, electroretinography, visual evoked potentials, or orbital and central nervous system imaging such as magnetic resonance imaging. This evaluation helps with understanding systemic diagnosis, visual system prognosis, and potential treatment options.

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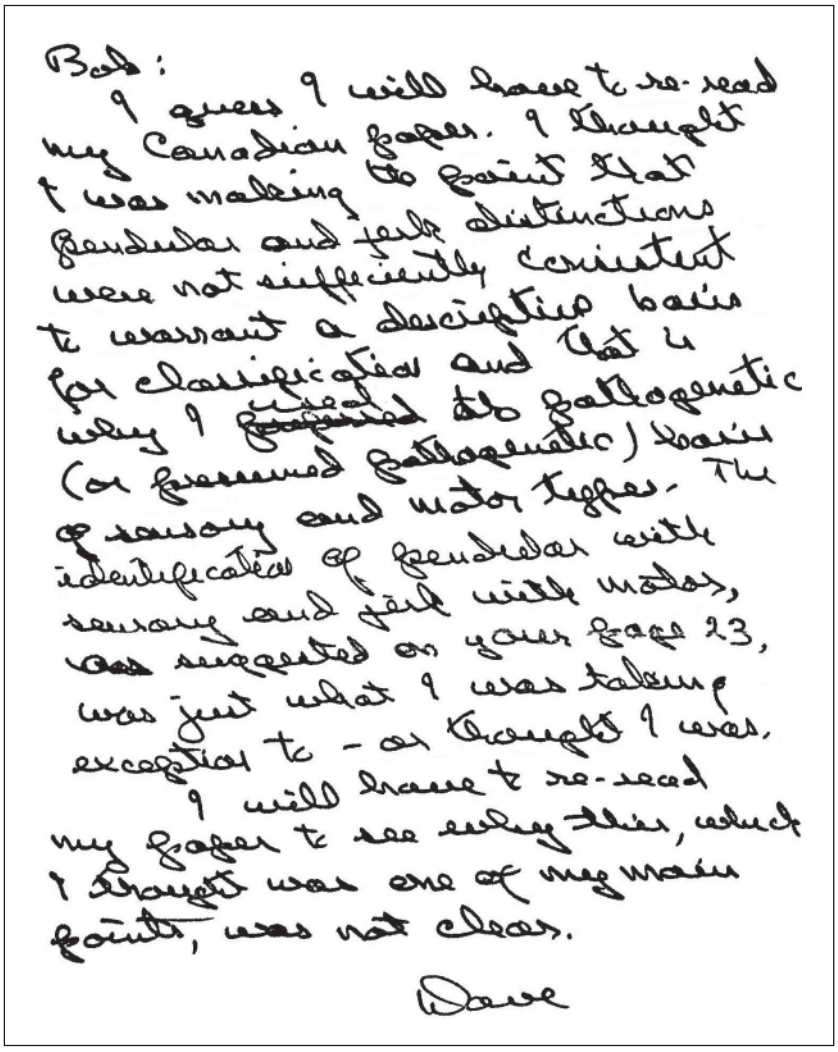
Bob:
I guess I will have to re-read my Canadian paper. I thought I was making the point that pendular and jerk distinctions were not sufficiently consistent to warrant a descriptive basis for classification and that is why I ~~preferred~~ used the pathogenetic (or presumed pathogenetic) basis of sensory and motor types. The identification of pendular with sensory and jerk with motor, as suggested on your page 23, was just what I was taking exception to—or thought I was. I will have to re-read my paper to see why this, which I thought was one of my main points, was not clear.
Dave

**Figure.** Hand-written note from David Cogan, MD, to one of us (R.B.D.) dated June 5, 1974, after R.B.D.'s presentation at the Wenner-Gren Conference in Stockholm, Sweden. The text of the note is as follows: "Bob: I guess I will have to re-read my Canadian paper. I thought it was making the point that pendular and jerk distinctions were not sufficiently consistent to warrant a descriptive basis for classification and that is why I ~~preferred~~ used the pathogenetic (or presumed pathogenetic) basis of sensory and motor types. The identification of pendular with sensory and jerk with motor, as suggested on your page 23, was just what I was taking exception to—or thought I was. I will have to re-read my paper to see why this, which I thought was one of my main points, was not clear. Dave."

## THE PROBLEM

For the past 40 years, eye-care professionals, residents in training, and other physicians have been misinformed about the true nature of INS and instead misled by this persistent, simplistic, and erroneous dual-waveform, dual-cause concept. As a result, some scientists and many eye-care professionals and neurologists have been trapped in a clinical paradigm that is fraught with diagnostic and therapeutic pitfalls. How did this erroneous notion evolve? We traced it to a seminal article by the late David Cogan, MD,[11] one of the keenest clinical observers of nystagmus phenomenology. In his article, Cogan linked the waveform terms *pendular* and *jerk* with the presumed pathogenetic deficits *sensory* and *motor*. Indeed, a cursory reading of his article may create the mistaken impression that Cogan thought that what looked clinically like pendular nystagmus was due to a primary sensory disorder, whereas what looked like jerk nystagmus was due to a primary motor disturbance.

Did Cogan's article really make that assertion and did he truly believe that waveforms that appeared to be pendular identified the primary cause of that infantile nystagmus as a sensory deficit and that jerk waveforms identified a motor cause? We will show that the answer to both questions is no.

## THE DATA

### Cogan's Article

In his 1967 article, Cogan made the association of waveform types with *presumed* pathogenetic cause in several places. The following are examples: "First is the type which is frequently called pendular nystagmus but which I would like to call sensory-defect nystagmus,"[11] or, "The type of congenital nystagmus which, as I shall attempt to show, depends on a defect in the efferent mechanism is customarily described as being the jerk type."[11] However, as regards pendular nystagmus, Cogan was careful to state, "This, perhaps this is not a good name because the nystagmus becomes jerk type on gaze to either side."[11] As to the jerk type, he stated, "This is not a wholly adequate basis for characterization, however, since the nystagmus may be pendular in the in-between positions of gaze."[11] Thus, patients could exhibit both types of waveform, rendering as artificial the division of what we now know as INS into 2 types. Cogan also theorized that his sensory-defect-type nystagmus resulted from failure of the fixation reflexes early in life, but he failed to justify either the resulting waveform or the many cases of pendular nystagmus documented at birth. Similarly, for motor-defect-type nystagmus, he theorized about both the optokinetic and vestibulo-ocular pathways that he presumed to be defective but which subsequent eye-movement recordings demonstrated to be intact along with smooth pursuit.[12,13] The confusion created by this article persists to this day.

### Cogan's Note

In the concluding and summarizing address of the Clinical Sciences Section of the Symposium on Basic Mechanisms of Ocular Motility and Their Clinical Implications at the Wenner-Gren Center, Stockholm, Sweden, June 3 through 5, 1974, our eye-movement data–driven conclusion contradicting the oft-cited clinical connections made between pen-

dular waveforms and sensory deficits as well as between jerk waveforms and motor deficits was described.[14] Although the published chapter did not refer to Cogan, the presentation did. Cogan was in the audience and wrote a note to one of us (R.B.D.) who delivered the paper. A copy of that hand-written note is shown in the **Figure**. In it, Cogan stated that in his 1967 article, he thought he was making the point that "pendular and jerk distinctions were *not* [emphasis added] sufficiently consistent to warrant a descriptive basis for classification." Thus, despite not having accurate eye-movement data, he recognized that patients with or without sensory deficits had what appeared clinically as both pendular and jerk waveforms. He also stated that he used a "pathogenetic (or presumed pathogenetic) basis of sensory and motor types"; again, that presumption of 2 types of nystagmus was based solely on clinical impression, not on objective eye-movement data. Furthermore, he stated, "The identification of pendular with sensory and jerk with motor, as suggested on your page 23, was just what I was taking exception to—or thought I was."

## COMMENT

A critical reading of Cogan's article revealed that he not only recognized the intermixing of INS waveforms in most patients (at least based on clinical impression) but also did not think one could use waveforms to differentiate his 2 presumed pathogenetic causes for the nystagmus. However, the article did contain repeated instances coupling pendular and jerk waveforms with sensory and motor deficits, respectively. Subsequent research over 4 decades has established that the actual waveforms exhibited by patients with INS are not causally related to the presence or absence of visual sensory deficits and that the latter are neither necessary nor sufficient to cause INS (ie, they are not the direct cause of INS but may be an additive factor). Rather, INS is caused by a developmental instability in the ocular motor system that can either coexist with any of a number of associated visual conditions or occur in patients with no known visual deficits.

In summary, a careful reading of both Cogan's article and his note indicates that the coupling of waveforms with presumed types or causes of INS was not his intent; it is equally clear from subsequent eye-movement research that such a coupling was incorrect. We hope that all mention of "sensory" vs "motor" nystagmus be expunged from future textbooks, peer-reviewed literature, and teaching material used to train health-care professionals. The necessity for quantitative eye-movement analysis to understand and model the neuropathophysiology, make accurate, repeatable diagnoses, determine the best therapeutic approach for each patient, and assess the efficacy of each therapy has been repeatedly demonstrated.

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