# Kongenitaler Nystagmus

## Congenital Nystagmus Waveforms and Foveation Strategy

### Kurvenformen des kongenitalen Nystagmus und Strategie der „Foveation“

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**Schlüsselwörter:** Congenitaler Nystagmus, Kurvenformen, Foveation, Nystagmus-Behandlung.

**Zusammenfassung:** Genaue Messungen der Augenbewegungen an 100 Personen mit kongenitalem Nystagmus brachten eine sichere Grundlage für die Klassifizierung der zahlreichen Bewegungsformen. Es ergaben sich objektive Definitionen, die auf meßbaren Größen beruhen und nicht auf dem „klinischen Eindruck“. Die Genauigkeit der Ableitungen gewährte, zusammen mit Filmaufnahmen des Fundus, die während der Fixation eines Laser-Zielpunktes ausgeführt wurden, Einblicke in den Mechanismus des kongenitalen Nystagmus. Viele der Kurvenformen erklären sich aus dem Versuch, das Zielobjekt möglichst lange auf der Fovea zu belassen. Diese Strategie erfolgt im Interesse einer Verbesserung der Sehschärfe.

The definition and waveform categorizations presented in this paper are the result of careful study of the eye movement recordings made of 100 patients with congenital nystagmus (CN). These were dc-coupled, high-bandwidth (100 Hz) recordings made using a noise-free infra-red reflection technique. Eye position and velocity records were made for both eyes simultaneously. Foveation strategies were verified using a laser-target retinal cinematographic technique (Dell'Osso, 1973a). The results of these studies have been a greater understanding of this ocular motor instability (CN) and the therapeutic use of composite prisms and surgery to improve visual acuity. Secondarily, many false or misleading clinical impressions have been exposed and reliance upon them discredited.

The clinical catch phrases which have led the misunderstanding of the nature of CN are: "movements of equal speed in each direction", "fast phase", "slow phase" and finally the concept of "sensory-detect" and "motor-detect" CN. Clinical observation can yield only a gross, oversimplified description of the complex waveforms of CN. Only estimates of mean velocities and time intervals can be made rather than peak velocities and partial time intervals for the different types of movements in the same direction. There exist pendular CN waveforms whose directional components are of unequal speed and jerk waveforms whose directional components are of equal speed. Thus, while looking at a CN patient's eyes, the criterion of "equal speed" cannot be used to determine waveform. Similarly, there exists a pendular CN waveform with an apparent "fast phase" and "slow phase"; there is also a jerk waveform where the movement in the jerk direction ("fast phase") takes longer than that in the other direction ("slow phase"). Obviously, such waveforms have been misdiagnosed in the past (especially if recordings of eye velocity were not used) and therefore, claims in the literature regarding etiological inferences of waveforms or directional effects of superimposed latent components cannot be supported. Indeed, the whole concept of "sensory" and "motor" defect waveforms is flatly contradicted by accurate waveform recordings and all CN is revealed to be an ocular motor instability, whatever the resulting waveform.

Retinal cinematography and dc-coupled recordings have established that descriptions of pendular CN as to-and-fro movements across the line of regard are false. Actually, the eyes rest on the target (foveation) at one or the other peak of the oscillation (Dell'Osso, 1973a). Both the side to which the eyes are biased and the frequency of bias reversals are idiosyncratic and affected by gaze angle and psychophysiologic factors. Jerk nystagmus has been shown to consist of a slow, accelerating drift of the eyes off target followed by a saccade which stops the drift and either fully or partially corrects eye position (Dell'Osso et al., 1974). Both types are slow eye movement defects reflecting instability in the slow eye movement subsystem.

One final clinical misconception which concerns the genesis of CN is its relation to ambient or retinal illumination (eyelid position). Previous observations attempting to relate either of these factors to CN genesis were misleading because the key variable of fixation attempt was never considered. It is the very attempt to fixate or to direct the eyes which brings on the CN oscillation. Statements such as, "nystagmus disappeared behind closed lids but increased in the dark" are irrelevant since the patient's fixation attempts were not monitored while making these observations. The discovery that fixation attempt was related to CN genesis is consistent with observations of decreased or no nystagmus in patients who are not attending to a visual input (e.g. when daydreaming) and with the opposite condition whereby attempts to read lower on the acuity chart produce intensified nystagmus and even head nodding. This vicious cycle of increased effort causing increased nystagmus which decreases acuity is fully discussed elsewhere (Dell'Osso, 1973b).

Our studies have resulted in the following two definitions for pendular and jerk CN waveforms:

**Pendular.** An ocular motor instability of the slow eye movement subsystem resulting in periodic motion of the eyes away from and back to the intended gaze angle (or target) such that the waveform is approximately sinusoidal. Occasionally small foveating saccades will be present on the peaks corresponding to target foveation.

**Jerk.** An ocular motor instability of the slow eye movement subsystem resulting in a periodic drift of the eyes away from the intended gaze angle (or target) which requires a saccade in the opposite direction to stop the slow eye movement. The saccade may either fully refoveate the target or begin a slow eye movement in the proper direction for refoveation. The direction of the jerk nystagmus is defined as the direction of this corrective saccade.

Key in these definitions are that both types: result from slow eye movement instabilities; cause the eyes to move away from and back to the target; and may contain small breaking saccades (Dell'Osso, Daroff, 1976) which sometimes achieve target foveation. The direction of jerk nystagmus is always that of the corrective saccade regardless of its foveating ability or the time required for foveation. These definitions, consistent with recorded data, correct previous errors and assumptions and allow a meaningful, systematic classification of CN waveforms.

Three main groups of CN waveforms emerge from recordings: pendular, jerk and dual. The jerk group is further divided into two subcategories: unidirectional and bidirectional. Comprehensive discussion and examples of all waveforms appear elsewhere

(Dell'Osso, Daroff, 1975; Dell'Osso, 1976). Briefly, there are three pendular waveforms, eight jerk waveforms (four unidirectional and four bidirectional) and one dual waveform. One of the pendular waveforms is easily mistaken clinically for jerk, five of the jerk waveforms usually are mistaken clinically for pendular and the direction of one jerk waveform is clinically reversed.

Good recordings allow for easy identification of that portion of the waveform which corresponds to target foveation. A small flattened part of the waveform which does not vary from beat-to-beat identifies when the eyes are motionless and on-target. It is careful observation of this time-on-target and reduction of nystagmus intensity with gaze angle or convergence that has resulted in the therapeutic use of composite prisms and surgery to increase visual acuity (Dell'Osso et al., 1972).

One final observation of bias reversals has enabled us to assess foveal function. If the flattened portions of oppositely biased nystagmus beats line up (i.e. they are at the same gaze angle) good foveal function is indicated; if there is a difference then foveal aplasia or displasia should be suspected (Dell'Osso, Daroff, 1975).

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## Aussprache

Herr Lang (Zürich):
Gilt ihre Beobachtung, daß der Pendelnystagmus nicht symmetrisch über die Fovea hinweg gleitet, nur für Erwachsene? – Bei Säuglingen sieht man eindeutig einen symmetrischen Pendelnystagmus.

Herr Dell'Osso (Miami):
Although we have not recorded babies' eye movements we have done young children (3–6 years) as well as adults and can say with assurance that as soon as the patient has something he wishes to fixate his CN will be biased to one side or the other. He will not oscillate across the target because that would preclude good vision. Any patient can, of course, shift his bias from one side to the other.

Herr Metz (San Francisco):
Why is the null point broadened by ocular muscle surgery?

Herr Dell'Osso (Miami):
The reason for this increase in the null region is unclear, but we have drastically altered the ocular motor plant by operating and this seems to result in a load-change on an oscillating system such that the oscillations diminish.

Herr Mackensen (Freiburg):
I agree with what you told us concerning nystagmus surgery, but I feel that to drive a motor car should not be an indication for this. Nearly all kinds of congenital nystagmus are activated under psychic excitation, and this will reduce the visual acuity. Therefore it would be better to keep this fellow away from motor traffic.

Herr Dell'Osso (Miami):
I'm happy to hear someone else state that CN is very dependant on psychological factors for that is precisely why the prisms and the operation work; they decrease fixation attempt. As to changing one's life-style by allowing one to drive, not being enough justification for a CN operation, I must disagree. Little anxiety is produced by driving once you gain experience, and the operation has already reduced this anxiety effect on CN as evidenced by the reduced CN intensity post-operatively. Also, I would find it a curious set of values if ophthalmologists could justify operating purely for the patients vanity (which they do) and not to enrich the patient's life by making his schoolwork and participation in sports easier and freeing him with a car.

Herr Strachan (Sheffield):
Has the Faden-Operation been used to help these cases of congenital nystagmus?

Herr Dell'Osso (Miami):
We have not done the Cüppers operation. We have performed recession-resection procedures in these cases.

Herr Jaeger (Heidelberg):
Es ist eine klinische Erfahrung, daß beim congenitalen Nystagmus der binokulare Visus besser ist als der monokulare. Deshalb darf man bei der Visusprüfung bekanntlich bei diesen Patienten nie vergessen, den binokularen Visus mit anzugeben. Kann etwas darüber gesagt werden, ob diese Verbesserung bei binokularem Visus auf alle von Herrn Dell'Osso analysierten Bewegungsabläufe zutrifft oder ob diese verschiedenen Bewegungsabläufe sich in ihrem Verhältnis monokularer Visus/binokularer Visus verschieden verhalten?

Herr Crone (Amsterdam):
Ist periodisch alternierender Nystagmus bei kongenitalem Nystagmus weniger selten als gewöhnlich angenommen wird?

Herr Dell'Osso (Miami):
Periodic Alternating nystagmus (PAN) does occur both as a form of congenital nystagmus (CN) and acquired nystagmus (AN). Many times it is missed because the directional changes are really aperiodic (APAN) and require lengthy viewing and recording to document. The periodicity is sometimes affected by gaze changes so that the motility exam, as normally conducted in the clinic, masks the PAN or APAN. The waveforms distinguish CN from AN.

Herr Körner (Bern):
I would like to ask the speaker whether he has also investigated subjects with acquired pendular and other nystagmus forms. Are there systematic differences in the characteristic of waveforms of congenital versus acquired pendular nystagmus? I have observed a post-saccadic pause of spontaneous nystagmus only in acquired, but not in congenital cases, otherwise there were pronounced similarities.

Herr Dell'Osso (Miami):
One can usually distinguish acquired pendular (P) nystagmus from congenital P nystagmus by looking at eye movement records for fixation bias shifts and/or flattening of the peaks on one side of the oscillation. Either of these signs identifies congenital nystagmus (CN). Also, patients with CN usually have small foveating saccades on the peaks to one side of the oscillation which results in a characteristic CN waveform, pendular with foveating saccades ($P_{FS}$).

If none of the above are present and the waveform is pure P one must rely on the history and look for afferent ocular defects which usually accompany (but do not cause) this type of CN. A third possibility is CN/A which is nystagmus acquired in early infancy and which mimics the CN waveforms described above but is secondary to an afferent defect.

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