

# Voluntary nystagmus in five generations

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**SYNOPSIS** Voluntary nystagmus is not a pathological condition. By voluntary effort, rapid synchronous pendular eye movements in the horizontal plane can be performed. Voluntary nystagmus has a frequency of 10 to 25 Hz, amplitude up to 6°, and can be maintained for up to 35 s. Oscillopsia is a concomitant feature. It has been reported in more than 100 individuals since 1855 but only once in three siblings, or in two generations. This paper reports five cases in five generations of one family and autosomal dominant inheritance with high penetrance is suggested.

Voluntary nystagmus does not appear to be pathological, though very few individuals are capable of producing this form of pendular nystagmus. Voluntary control is, however, limited to initiation and duration; frequency and amplitude of nystagmus cannot be influenced. It has been reported only once in siblings (Goldberg and Jampel, 1962) and Keyes (1973) described voluntary nystagmus in two generations of a family. It seemed worthwhile, therefore, to report a family in which voluntary nystagmus had been found in five generations.

One of the authors (R.R.) is a member of the

above-mentioned family. He himself can produce voluntary nystagmus and, as a neurologist, noticed its frequent incidence in his family. In five generations there is a total of 25 members, of whom 18 were either available for testing or sufficient information could be obtained. Five members, three male and two female, one in each generation, could readily produce voluntary nystagmus, whereas the remaining 13 members could not, despite many attempts. The family's pedigree is shown in Fig. 1.

### CASE 1

R.R. is a male, aged 39 years, healthy, with normal EEG and vision except a slight red-green colour-blindness. He has no spontaneous nystagmus nor

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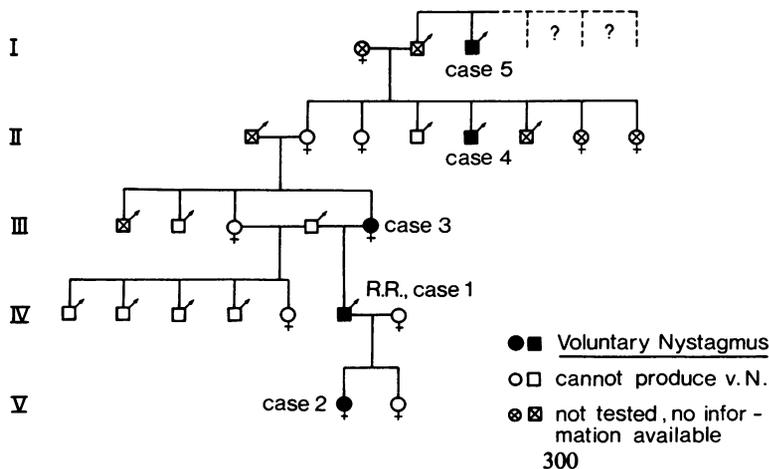


FIG. 1 Pedigree of the family with voluntary nystagmus throughout five generations. One of the authors (R.R.) is listed as case 1.

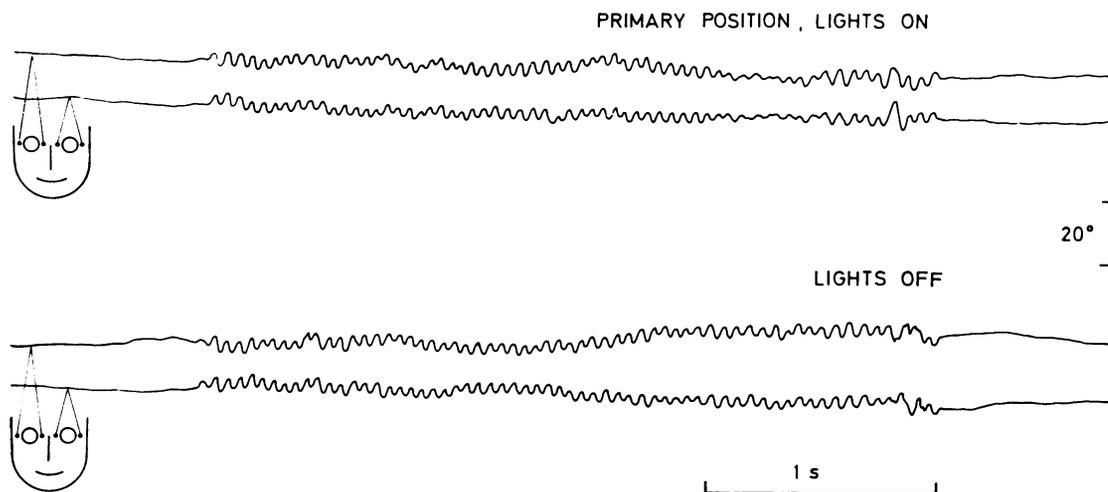


FIG. 2 Electronystagmographic d.c. recordings of the voluntary nystagmus with the eyes open and closed.

ocular dysmetria and normal optokinetic nystagmus in all directions. At about the age of 8 years he had for the first time observed that he could voluntarily without imitating anyone else make his surroundings 'jiggle' or 'dance'. Later, he recognized these high frequency ocular oscillations as voluntary nystagmus. Neither his half-sister nor his four half-brothers could produce voluntary nystagmus despite repeated effort. In later years he neglected his ability and did not show it to his daughters.

Electronystagmographic d.c. recordings (ENG) in light and dark are shown in Fig. 2. Voluntary nystagmus was also seen with the eyes closed. A constant frequency of 17 to 18 Hz with amplitudes of about 4° was measured. Voluntary nystagmus could even be elicited at 20° gaze angles, but only for short durations and with smaller amplitudes. It could be maintained for up to 15 s, but showed rapid deterioration when repeated at short intervals.

#### CASE 2

The daughter of R.R., age 8 years, is physically and mentally normal with normal EEG and vision. She discovered her ability for voluntary nystagmus at the age of 6 years and was astonished to learn that her father could jiggle his eyes as well. Her older sister cannot imitate this well-established voluntary nystagmus despite repeated effort.

On electronystagmographic d.c. recordings a frequency of 17 Hz and amplitudes up to 4° were measured. All ENG recordings were identical with that of case 1, except that voluntary nystagmus could only be maintained up to 8 s.

#### CASE 3

The mother of R.R. died when he was 1 year old. According to her brothers and sisters, she often jiggled her eyes at will as a child to the amusement of her family. Her siblings could not imitate this.

#### CASE 4

For four of the seven siblings in this generation, exact information has been offered by one of R.R.'s great-aunts. She reported that one of her brothers could voluntarily oscillate his eyes at a high frequency, but it is not known when he acquired this ability. This great-uncle to R.R. was a pilot in the first world war. He can, therefore, be considered physically and mentally normal, including vision and eye movements.

#### CASE 5

From the same great-aunt it is known that her uncle could jiggle his eyes at will and, by so doing, amused his nephews and nieces. She recalls several occasions when she, her brothers and sisters had tried to imitate voluntary nystagmus but, except for her brother (case 4), without success.

The electronystagmographic recordings of cases 1 and 2 demonstrated that voluntary nystagmus was pendular, horizontal and synchronous in both eyes. Differences in frequency, amplitudes, or wave form between the two eyes could not be detected.

#### DISCUSSION

More than 100 cases of voluntary nystagmus

have been reported in the literature. They agree in wave form, frequency range, amplitudes, initiation procedures, and duration (Bruckner, 1917; Westheimer, 1954; Goldberg and Jampel, 1962; List and Collins, 1964; Coren and Komada, 1972; Blumenthal, 1973; Keyes, 1973). These criteria, identical with our ENG recordings in cases 1 and 2, are shown in Fig. 3. When the age of initiation of voluntary nystagmus is mentioned, it most commonly occurred spontaneously during childhood. The organic basis of selection of individuals and how they detect their ability to produce the phenomenon is not as yet known.

Voluntary nystagmus can be maintained normally for 5–10 s (35 s maximum, Coren and Komada, 1972), and is associated with oscillopsia. Frequencies have been measured usually between 15 and 20 Hz (Westheimer, 1954), the fastest being 23 Hz (Goldberg and Jampel, 1962). Visual acuity and other ophthalmological criteria are normal. Except for voluntary nystagmus, no conspicuous abnormalities have been detected in the cranial nerves nor in the general neurological status. Further well-established characteristic properties are that this high frequency pendular nystagmus can be initiated voluntarily independently of fixation, eye position, or whether the eyes are open. A convergence or wide opening movement of the eyes is often necessary for its initiation. The capability of producing it is limited by fatigue.

Different opinions have been presented concerning the aetiology of voluntary nystagmus, particularly whether it can be learned (Bruckner, 1917; Lindner, 1941). The possibility of a

Voluntary nystagmus	
(summary of literature and own cases)	
Frequency	15–20 Hz: min: 8 Hz– max: 23 Hz
Amplitude	3°–8°
Type of nystagmus	Conjugate, pendular, horizontal, seldom monocular
Duration	Max. 20–30 s
Initiation	With and without fixation at all eye positions, in light and darkness, even with closed eyes
Accompanying symptoms	Oscillopsia with visual blurring
Pathological significance	None, otherwise healthy persons
1. Appearance	Mostly between age 8 to 15 yr
Aetiology	(a) Imitation (b) Hereditary

Characteristics not under voluntary control

FIG. 3 *Characteristic properties of voluntary nystagmus as summarized from the literature and from present cases.*

hereditary form was discussed by Goldberg and Jampel (1962). They described voluntary nystagmus in three of six siblings whose parents could not elicit voluntary nystagmus. The pedigree of this family, as well as that of a family reported by Keyes (1973) with voluntary nystagmus in two generations, is shown in Fig. 4.

The present investigation describes a family

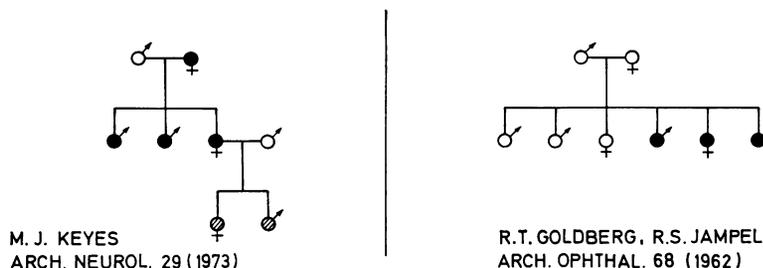


FIG. 4 *Hereditary forms of voluntary nystagmus reported in the literature.*

M. J. KEYES  
ARCH. NEUROL. 29 (1973)

R.T. GOLDBERG, R.S. JAMPEL  
ARCH. OPHTHAL. 68 (1962)

- VOLUNTARY NYSTAGMUS
- CANNOT PRODUCE V.N.
- ⊙ TOO YOUNG FOR TESTING

with evidence of voluntary nystagmus in five generations in which 18 of the 25 members have either been examined or sufficient information was available (Fig. 1). Voluntary nystagmus occurred once in every generation (two males and three females). In case 1 a red-green colour-blindness was present, as is known for other members of the family. Cases 2, 3, and 4 are known to have normal colour vision. Otherwise all members with voluntary nystagmus have a normal case history without auditory or vestibular defects, or congenital nystagmus.

Cases 1 and 2 discovered at the age of 6 and 8 years respectively, their ability voluntarily to control oscillopsia which always accompanies voluntary nystagmus. Case 1 came to know of his deceased mother's voluntary nystagmus only in later years, and he himself never showed his daughter (case 2) this ability. Other members of the family tried repeatedly to produce voluntary nystagmus without success.

It may be of interest, that, on the other hand, we know several children, exclusively deaf mutes, who discovered their ability to produce voluntary nystagmus only after it was suggested to them (Lindner, 1941). These cases will be described by Aschoff *et al.* (in preparation). ENG recordings of voluntary nystagmus show a pendular, sinusoidal form, resembling acquired pendular nystagmus in miners (Ohm, 1954) and in patients with multiple sclerosis (Aschoff *et al.*, 1974) but not congenital nystagmus. Miners' nystagmus and acquired pendular nystagmus in multiple sclerosis oscillate less rapidly: 4–5 Hz. Miner's nystagmus is associated with head tremor, but acquired pendular nystagmus in multiple sclerosis is associated with cerebellar and other neurological deficits. Individuals with voluntary nystagmus, however, must be regarded as normal healthy persons.

Aschoff *et al.* (1974) have shown that pendular nystagmus in multiple sclerosis results from lesions of cerebellar nuclei, or of their efferent connections, which at least partly function as a hold mechanism for limb, body, and eye positions (Kornhuber, 1973). As voluntary nystagmus resembles the pendular nystagmus of multiple sclerosis in wave form, one might consider that voluntary nystagmus results from an ability to turn on and off at will this cerebellar nuclei holding mechanism. The frequency range

of pendular eye movements, 15–20/s must correspond with a natural internal frequency, since rapid alternating, voluntary movements such as tapping reach a maximum of 8–10 Hz. Voluntary nystagmus probably resembles a natural frequency from proprioceptive feedback mechanisms 'cerebellar nuclei—eye muscles—cerebellar nuclei'. Acquired pendular nystagmus in miners and in patients suffering from multiple sclerosis, on the other hand, resembles the natural frequency of the visual hold mechanisms (Kornhuber, 1966). In any case, the ability to produce voluntary nystagmus must be linked with supranuclear anatomical structures and not to the visual system, the vestibular system, eye muscles, or their nuclei.

Our family study demonstrates that voluntary nystagmus, probably one of several forms, is transmitted by a dominant autosomal genetic factor. In the family reported by Goldberg and Jampel (1962) (Fig. 4, right) the transmission could also be recessive but our history clearly shows a dominant transmission with incomplete penetrance. This is illustrated by the fact that R.R.'s grandmother (second generation), who must have transmitted the gene for voluntary nystagmus, could not elicit voluntary nystagmus herself. On the basis of putative father-son transmission from generation I to II (Fig. 1) an X-linked dominant mode of inheritance seems unlikely.

Sporadic cases of voluntary nystagmus, as described in the literature, are most probably determined by genetic factors as well: other members of these families may either have never tried to produce nystagmus or have not been examined for voluntary nystagmus, or the genetic factor involved has only an incomplete penetrance.

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