SPONTANEOUS NYSTAGMUS IN CON- GENITAL SYPHILIS

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The phenomenon of spontaneous nystagmus, of otitic origin, in children the subjects of hereditary syphilis, is one that is well known to the continental otologists. It is constantly referred to by them in their writings and lectures, but does not appear in the English literature devoted to pædiatrics, otology or syphilology.

Hennebert¹ drew attention to the syndrome, and it goes by the aponym 'Hennebert's syndrome,' or 'fistel symptom ohne fistel' (fistula symptom, without fistula). Alexander³ and Ruttin have subsequently recorded their observations.

According to these otologists, children afflicted with hereditary syphilis may exhibit one or other of the following :—

1. Attacks of giddiness and nystagmus, coming on spontaneously, and noted by the parent. The attack is of short duration, and may be precipitated by pulling on the tragus or insertion of a finger in the external auditory meatus. It is not a constant phenomenon and is transient.

2. In this group, on a routine examination, it is discovered that by compression or aspiration of the air in the external auditory meatus, using a Siegle's speculum, or even by mere compression with the finger in the external auditory meatus, spontaneous nystagmus can be elicited. On otoscopic examination, an intact tympanic membrane will be found. Hence the description, 'fistula symptom, without fistula.' Again the phenomenon is not constant, but transient, that is to say, it cannot necessarily be elicited at any other given time. Such significance is attached to the demonstration of this sign by the continental observers that it is taken to be pathognomonic of hereditary syphilis. It is not a common phenomenon, and is not present in all cases of this malady.

Anti-luetic treatment is said to cure the condition.

Alexander³ states, *inter alia*, in discussing this symptom that 'no cause is known, that it occurs from the age of 3 months onwards, that it may be continuous or periodic, that giddiness and nystagmus occur, and that pulling on the tragus or compression of the meatus with the finger will produce the phenomenon.' He also gives two aural stigmata that are characteristic of congenital syphilis, namely, a shortened bone conduction with normal hearing, and the 'fistel symptom, ohne fistel.'

Hastings² records a case in a girl of 20, where it was this symptom that led to the diagnosis of hereditary lues. This patient was deaf in the left ear. The membrane was intact, and compression of the meatus gave nystagmus to the right, and caused giddiness. She also had two other stigmata of the disease, interstitial keratitis and Hutchinson's teeth. Three weeks later Hastings was unable to elicit the nystagmus again.

The pathology of the condition is not settled as yet. Alexander considers it due to weakening of the annular ligament which encircles the foot piece of the stapes and anchors it in position. Ruttin is driven to the assumption that it is due to some congenital change affecting the inner ear. The varied nature of the opinions hazarded is due to the fact that congenital lues is not a lethal disease; that the few cases that have come to autopsy with a report as to the condition of the ear, no record of spontaneous nystagmus was elicited. There is yet to be recorded the findings of the middle and inner ear in a case in which Hennebert's syndrome has been previously observed.

The phenomenon is not a common concomitant of the malady, and an explanation may be suggested more consistent with its variability, its transiency, its reaction to anti-syphilitic treatment, and its rarity. It is well known that in a case of chronic suppurative otorrhœa, a fistula symptom develops if the osseous covering of the horizontal semi-circular canal is eroded by the supuration. If this osseous wall is destroyed by a gummatous infiltration, or is thinned by a process comparable to that which produces cranio-tabes in the vault of the skull in congenital syphilitics, the same condition obtains as in a fistula. Thus any variation in the intra-tympanic pressure is readily transmitted to the horizontal semi-circular canal, and so produces the 'fistula symptom without fistula' syndrome. This would account for the infrequency of the phenomenon, as it is only when the thinning process is localized to the layer of bone over the horizontal semi-circular canal that the nystagmus is produced.

Through the courtesy and encouragement of Dr. D. Nabarro, I was able to examine some 40 cases of congenital syphilis, in search of any symptoms similar to those recorded above. The attached figures shew the results obtained.

In no case had the nystagmus been recorded or observed by the physicians under whose care the children were, nor had the parent in any case observed it. Only by testing the children for nystagmus was the symptom detected. All the children examined were under anti-luetic treatment for years, many from birth to 12. No occasions presented for the examination of any of the cases before treatment had been instituted.

The results of these examinations were as follows:—

Number of cases investigated	40.
Ages	3 months to 18 years (29 cases between 1 and 12 years).
No nystagmus elicited	27 cases.
Nystagmus	13 cases.
? other origin than ear	5 cases.
True spontaneous nystagmus	8 cases.

The doubtful cases were as follows :—

- 1 case aged 5 was a mental defect and the subject of fits ;
- 1 case had bilateral chronic suppurative otitis media ;
- 1 case was blind and the eye movements were inco-ordinate ;
- 2 cases had a strabismus and the nystagmus was elicited without them wearing their spectacles.

The positive fistula symptom cases were as follows : the tympanic membrane was intact in all the cases.

CASE 1. Age 12, male, spontaneous nystagmus not increased by compression of the meatus. Nystagmus horizontal to one side only, transient and not consistent. Many stigmata of congenital syphilis, e.g., rhagades, Hutchinson's teeth, hoarseness, etc. W.R. negative.

CASE 2. Age 12, female. Finger in the left meatus produces nystagmus to the left. It is slight and transient. W.R. positive.

CASE 3. Age 10, female. Nystagmus to the right slight, with or without meatal pressure. W.R. positive.

CASE 4. Age 9, male. W.R. negative. Nystagmus to the right, slight, horizontal, and unaffected by increased meatal pressure.

CASE 5. Age 9, male. Horizontal nystagmus, ? slightly increased by meatal pressure. W.R. positive.

CASE 6. Age 5, female. Marked nystagmus to the left, increased by meatal pressure. W.R. positive.

CASE 7. Age 11, female. Nystagmus to the right, slightly increased by insertion of finger in the left meatus. W.R. positive.

CASE 8. Age 10, female. Slight transient nystagmus to the left, horizontal. Not increased or affected by the compression of the meatus. Has interstitial keratitis. W.R. positive.

Summary.

1. All the cases investigated are those who have been subjected to anti-luetic treatment, many for years or from birth, so that the incidence of the spontaneous nystagmus is that amongst treated cases. No untreated cases were examined.

2. In all 8 cases the drums were intact, and as far as could be elicited the hearing was normal.

3. No vestibular or caloric tests were performed for obvious reasons.

4. The phenomenon was not a constant one.

5. Although spontaneous nystagmus was elicited in these cases, no one instance satisfied the criterion of Hennebert, namely, that compression and rarefaction of the column of air in the meatus produced an exaggeration of the nystagmus.

6. Thus one is driven to the conclusion from this small series of cases, that nystagmus, as tested for in the usual manner, is a not uncommon concomitant of hereditary lues, among treated cases in infants and children, although a true Hennebert's phenomenon was not demonstrated in any of these treated cases. What part the anti-luetic treatment has played in preventing the production of this phenomenon must remain a matter for conjecture.

The attention of pædiatric physicians is drawn to this phenomenon in the hope that all cases of hereditary lues will be examined with a view to eliciting the phenomenon. A record of their results will settle the question as to the diagnostic significance of the phenomenon in congenital syphilitics. It is the pædiatrist who sees all these cases. They do not reach the otologist, hence any early investigation must come from the pædiatrist.

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REFERENCES.

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2. Hastings, *J. Am. Med. Ass.*, Chic., LXV.
3. Alexander, *Ohrenkrankheiten im Kindesalter*, 348.