Australian Orthoptic Journal,
1979-80, Vol. 17

NYSTAGMUS BLOCKING SYNDROME

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Abstract
Nystagmus blocking syndrome is a type of congenital esotropia with features of early onset, large angle and nystagmus on attempted abduction. The characteristics of this syndrome are discussed, together with two case histories. Management usually entails occlusion and surgery. Surgery is generally less predictable than in the uncomplicated congenital esotrope.

Key words
Nystagmus blocking syndrome, congenital esotropia, posterior fixation suture operation.

Nystagmus, which is reduced or absent in adduction is a fairly well recognised entity, and patients can often gain increased visual acuity because of this reduction.

Von Noorden[1] quotes Lafen (1914) and Ohm (1916) as mentioning an entity where infants with nystagmus develop an esotropia to remain in the null point.

Aldestein and Cuppers[1] advanced this concept which they named nystagmus blocking syndrome. In 1966 they documented its features to be:
- esotropia with sudden onset in early infancy, often preceded by nystagmus
- pseudo paralysis of both lateral recti
- appearance of manifest nystagmus as the fixing eye moves from adduction to abduction

Recent authors, namely Gunter Von Noorden[1,2,5] and Craig Hoyt[3] have done much to popularise the syndrome. They both mention the development of abnormal head postures in these children as both eyes are convergent, and the child usually turns his head towards the fixing eye to maintain it in an adducted position. This becomes more obvious when the child's eye is occluded, and in cases where there was no abnormal head posture previously, the occlusion tends to precipitate one.

The incidence of neurological disorders seems to be quite high in patients with nystagmus blocking syndrome. A significant number of reported cases have undergone shunt operations to reduce intracranial pressure (Hoyt, four out of eight cases, Von Noorden, two out of twelve cases).

There seems to be a very fine dividing line between nystagmus blocking syndrome and other types of estropia associated with nystagmus. Some points regarding differential diagnosis should be made clear at this stage.

Nystagmus is not present in adduction but becomes manifest on any attempt to abduct, so that manifest nystagmus is usually seen before the abducting eye has reached the midline. With end point nystagmus, the eye usually has to be in relatively extreme abduction before the nystagmus is manifest.

Nystagmus blocking syndrome can be associated with latent nystagmus but in such cases, the amplitude and often the frequency of nystagmus will increase, and manifest nystagmus will be noted on attempted lateral versions by the time one eye reaches the midline.

Von Noorden[1,2] feels that cross fixation may be misdiagnosed as nystagmus blocking syndrome, especially where there is apparent dysfunction of the lateral recti. However, the cross fixation pattern seen in congenital esotropes does present a different clinical picture as I hope the case histories will show.

The differential diagnosis between bilateral lateral rectus paralysis and nystagmus blocking

syndrome is relatively easy as in the former, neither eye will abduct. However, there is debate about the role of mild lateral recti paresis in the genesis of nystagmus blocking syndrome.

Von Noorden feels that lateral rectus paresis does not play a part in its aetiology. He does state that four of his twelve cases had restriction of abduction but these had "complicating factors".

Hoyt[3] on the other hand, feels that lateral rectus paresis does play a part and mentions that four of his cases could have developed lateral recti paresis during periods of high intracranial pressure before undergoing shunt operations.

It is interesting to note that both Von Noorden[1] and Hoyt[3] found that nystagmus was so fine in some cases that nystagmus blocking syndrome was not diagnosed until the patient underwent visuscope examination.

The mechanism of nystagmus blocking syndrome needs more research. One electromyographical study[4] shows that as the fixing eye moves from abduction to adduction there is incomplete inhibition of the medial rectus in the non-fixing eye. Von Noorden concludes from observation that the convergence mechanism is used, and that it is, at least initially, a purposive squint.

Treatment

Early detection of nystagmus blocking syndrome is essential as it effects a poorer prognosis on cosmetic grounds and I feel that the patient's parents should be aware of this.

Naturally, any significant refractive error should be corrected and occlusion is undertaken to strengthen lateral recti if weak through non use, reverse any amblyopia and promote alternation.

However, with occlusion an abnormal head posture may be exaggerated or one may develop where there was none before.

Cosmetically, something has to be done about the size of the esotropia, but surgery is unpredictable. Theoretically with surgical intervention the neutral zone of nystagmus is shifted to the primary position, enabling good visual acuity post operatively. The most common procedure is the recession/resection and often two operations are needed.

Von Noorden[6] suggests the posterior fixation suture operation, or Fraden operation, as a possible procedure. The medial rectus is sutured to the globe behind the equator, therefore reducing its action without effecting innervation to other muscles. But as von Noorden points out this may be insufficient, even if performed bilaterally, and may need to be combined with a lateral rectus resection.

Both over and under corrections are frequent with surgery. The undercorrections usually are acceptable immediately post operatively (as with case no. 2) and often gradually increase in size. One explanation is that the child is reconverging to block nystagmus again, and this theory could explain some surgical failures that remain at their original angle despite 2 or more operations.

Von Noorden puts no explanation forward to explain his overcorrections, but it seems that generally surgery is far less predictable and that the high incidence of neurological disorders could play a part.

In summary, nystagmus blocking syndrome appears to be caused by hypertonicity of the medial recti resulting from the patient's sustained effort to block nystagmus by converging his eyes. The convergence could well be voluntary initially but becomes involuntary with time. The patient presents with a large angle esotropia with sudden onset in early infancy, possibly with a period of nystagmus preceding it, possible paresis of the lateral recti, and often a head turn in the direction of the fixing eye.

The two following cases illustrate some of the features noted above:

Case 1

Wilson – aged 6 years, has Down's Syndrome and is at present in the Macquarie University Down's Syndrome programme. He has a high I.Q.

His large angle right esotropia was noticed almost immediately after birth. Pregnancy and birth were uncomplicated, and there is no family history of squint.

An abnormal head posture of a head turn to the left eliminated his nystagmus and was first noticed at 2 years 10 months, when he was learning to read. He has a small degree of myopic astigmatism which has been corrected and has worn part time left occlusion to encourage alternation.

Ocular movements show full abduction of each eye but with nystagmus and a reluctance to abduct fully. The nystagmus becomes apparent just before the abducting eye reaches the midline. There is very slight bilateral inferior oblique overaction. With his abnormal head posture Wilson has no nystagmus. Cosmetically his squint is unacceptable.

Case 2

Joanna – aged 3½, first presented at 4 months old with a history of a large left esotropia,

Australian Orthoptic Journal,
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nystagmus and head turn to the right, from birth. Pregnancy and birth were uncomplicated and family history was unremarkable.

She wore part time (R) occlusion until she alternated freely, with a head turn towards the fixing eye. Nystagmus became apparent in the abducting eye as soon as it moved away from the adducted position. Cosmetically her squint was unacceptable so at ten months surgery (left medial rectus recession 6mm, left lateral rectus resection 7mm) was performed.

Immediately post operatively her squint was cosmetically excellent but over the next two years her angle has increased and at present is 30△BO without glasses, 25△BO with glasses.

Cosmetically her squint is fair to good. She has mild hypermetropic astigmatism in her left eye which has been corrected. Ocular movements show manifest nystagmus in abduction more marked in the right than the left eye. Both inferior obliques overact, with minimal underactions of both superior obliques. Her nystagmus is much reduced post operatively and could now be mistaken for gross end point nystagmus.

Acknowledgement

I would like to thank Dr Geoffrey C. Hipwell for his help and encouragement, and for allowing me to present the two case histories.

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