

## CONGENITAL ESOTROPIA AND NYSTAGMUS. EVALUATION OF SURGICAL OUTCOMES

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### Abstract

Both congenital and infantile esotropia are frequently associated with nystagmus. This retrospective study evaluated a relationship between the presence of nystagmus and surgical outcome in children treated for congenital esotropia. The clinical notes of 316 consecutive patients who underwent surgical correction for congenital esotropia between 1985 and 1997 were evaluated. Preoperative clinical characteristics and indications for additional strabismus surgery because of a residual or consecutive deviation were noted. The required minimum follow-up after the initial surgical correction was 12 months. A total of 196 patients met the inclusion criteria; of these, 57 (29%) had latent or manifest nystagmus (group A), and 139 (71%) had no nystagmus (group B). In groups A and B, 31 (54%) and 39 (27 %) children, respectively, required repeat surgery ( $P=0.096$ ). It is concluded that nystagmus associated with congenital esotropia may increase the risk of additional strabismus surgery for residual or consecutive deviations.

### Key words

Congenital esotropia, Nystagmus, Surgery

### INTRODUCTION

Congenital esotropia is an esodeviation with onset before 6 months of age, with an incidence of about 1%. The angle of deviation is usually very large, greater than 30 prism dioptres (PDs), and is approximately the same regardless of whether the eye is focused on a distant or a near object. Congenital esotropia may be associated with nystagmus, which is very common dissociated vertical deviations and inferior oblique muscle overaction. Nystagmus is usually latent, becoming manifest when one eye is occluded. In patients with congenital esotropia, the presence of nystagmus, either latent or manifest, as well as other conditions, such as amblyopia or vertical deviations, may adversely affect the outcome of surgery for congenital esotropia (1–5). In this study, a relationship between nystagmus and the surgical outcome in children treated for congenital esotropia was investigated on the basis of clinical data available over a period of 12 years.

## MATERIALS AND METHODS

The clinical notes of 316 consecutive patients who underwent surgical correction for congenital esotropia at the Department of Paediatric Ophthalmology between 1985 and 1997 were reviewed and the patients to be included in the study were selected on the basis of the following criteria: onset of unaccommodative esotropia by 6 months of age, no concurrent ocular or neurological abnormalities, no previous strabismus surgery, and a minimum follow-up of 12 months after the first correction of congenital esotropia. Further information sought for each selected patient included: preoperative and postoperative visual acuity, spherical equivalent determined by cycloplegic refraction, presence or absence of amblyopia, presence or absence of nystagmus (latent or manifest), dissociated vertical deviation, when present, the maximum angle of deviation found on distant or near alternate prism covering testing. Only measurements recorded for primary positions were taken into account. Of the 316 patients whose records were investigated, 196 met the inclusion criteria.

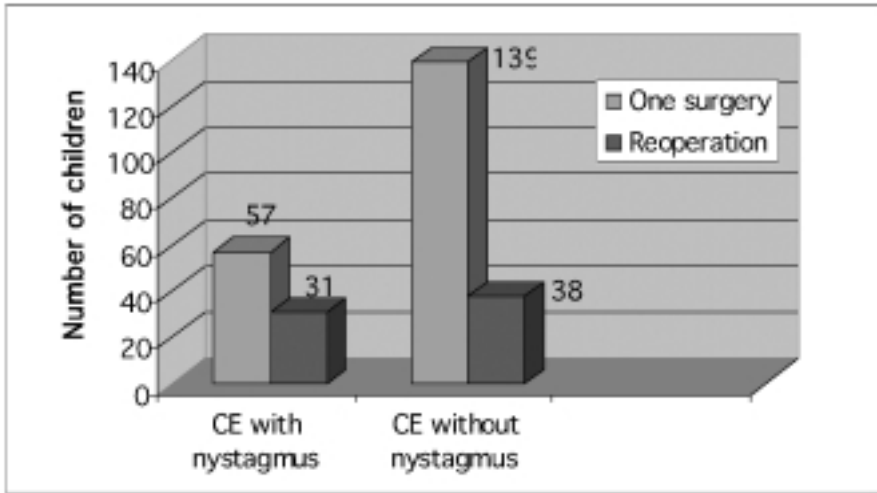
All 196 children underwent medial rectus muscle recession. In 24 patients this was combined with either inferior oblique muscle myectomy or anterior transposition for correction of congenital esotropia. Bilateral medial rectus muscle recessions were performed according to the guidelines for surgical dosage and techniques (6). For the purpose of this study, an esodeviation of 15 PDs or an exodeviation of >10 PDs on the last examination in the patient's record was regarded as an indication for repeat surgery.

The results were statistically analysed using Student's *t* test and presented as mean  $\pm$  SD values.

## RESULTS

The patient population of 196 children was further classified according to the presence (group A, *n* = 57) or absence (group B, *n* = 139) of nystagmus. In group A, nystagmus was preoperatively diagnosed as manifest in 31 and latent in 26 patients. The mean age at the initial operation for all 196 patients was 15 months (range, 4 to 51 months), for groups A and B it was  $17 \pm 1.9$  months and  $11 \pm 7.4$  months, respectively. In the whole patient population, the mean follow-up was 34 months (range, 13 to 95 months) and, in groups A and B, the mean periods were 41 months (range, 14 to 95 months) and 35 months (range, 12 to 76 months), respectively. The mean spherical equivalent for each eye was +1.73 D, regardless of whether the patient had nystagmus or not. The mean difference in spherical equivalent between both eyes (anisometropia) was the same in both groups.

In group A, 31 patients (54 %) had already had a repeat surgery or were indicated for one according to the criteria defined above. Indications for repair included undercorrection (residual esotropia) in 22 (72 %) and overcorrection (consecutive exotropia) in 9 (28 %) patients. In group B, 38 (27 %) patients either had had or were indicated for repeat surgery for either undercorrection or overcorrection (32 and 6 children, i.e., 83% and 17 %, respectively). Overall, 69 from 196 patients (35 %) had had or would require repeat surgery. In most of these patients, the reason for repeat operation was undercorrection whether or not the patient had nystagmus. The average undercorrection was 17 PDs (range, 5–35 PD, esotropia), and the average overcorrection was 14 PDs (range, 6–25 PD, exotropia). However, the difference between groups A and B in their requirements for repeat surgery was not significant (*P* = 0.096; *Fig. 1*).



*Fig 1.*

Outcomes of surgical treatment in children with congenital esotropia in relation to the presence or absence of nystagmus. Group A (n = 57), children with nystagmus; group B (n = 139), children without nystagmus.

The average preoperative deviation was 47 PD (range, 30–60 PD) in group A and 41 PD (range, 20–70 PD) in group B. This difference was not statistically significant.

When the presence of amblyopia was assessed, the following information was obtained: amblyopia was diagnosed in 21 % (42) of all the patients, in 28 % of the patients who had had or would require repeat surgery (19/69) and in 25 % of the patients without indications for repair of either under- or overcorrection (35/139). There were no differences among these relative numbers. In groups A and B, amblyopia was present in 15 (26 %) and 27 (19 %) patients, respectively. This difference was not significant.

#### DISCUSSION

Congenital esotropia is usually associated with inferior oblique muscle overaction, dissociated vertical deviation, and congenital nystagmus (1–8). Congenital nystagmus may be manifest or latent. Manifest congenital nystagmus is characteristically pendular with no change on unilateral occlusion and is rarely associated with congenital esotropia, whereas latent nystagmus is a jerk-type nystagmus that increases on unilateral occlusion and is often associated with congenital esotropia. Von Noorden (2) has reported an overall incidence of in

25% of the patients in patients with congenital esotropia and has suggested that the pathophysiology of congenital esotropia may be different in eyes affected also with congenital nystagmus, as compared to eyes without nystagmus, and this may also results in a poorer outcome of surgical treatment. Some of the previous studies have related preoperative clinical characteristics to the outcome of surgery (2, 4, 5, 9, 10).

Our results indicate that nystagmus, when associated with congenital esotropia, may increase the risk of achieving a poor surgical outcome. Fifty-four percent of our patients with nystagmus and congenital esotropia had or required repeat surgery, according to our criteria whereas only 27% of the patients without nystagmus had or required repeat surgery for congenital esotropia. This difference was not statistically significant and our results are in accord with those of several authors (1,4) who found the presence of latent nystagmus to be predictive of a less satisfactory surgical outcome in patients with congenital esotropia. *Keenan and Willshaw* (5) found no effect of nystagmus on surgical outcome; however, the number of patients reported in that study is small. Most patients requiring additional surgery in our study had a residual esotropia (undercorrection), whether or not nystagmus was present. Patients with nystagmus had a somewhat greater likelihood of being overcorrected after the initial surgery than had those without nystagmus; this difference, however, was not statistically significant. There was no significant difference in the angle of preoperative deviation, mean spherical equivalent, or duration of follow-up between our two groups. The occurrence of nystagmus in our study is in agreement with the findings of *von Noorden* and others (1,2). Interestingly, 9 of 37 patients with preoperative manifest nystagmus converted to latent nystagmus after strabismus surgery. Others have reported similar experience with the conversion of manifest to latent nystagmus after strabismus surgery (11).

Several studies have reported an association between amblyopia and a less favourable surgical outcome in the treatment of congenital esotropia (4,5). Amblyopia did not appear to increase the risk of significant residual or secondary deviation in our study.

Comparisons among studies are difficult because, in each, the evaluation is based on different criteria of a successful outcome. Small (10 PDs, esotropia or exotropia) or large (20 PD) deviations after surgery have been used by some authors as their criteria for successful surgical outcome (2,5,10). Others have used subnormal binocular vision and fusion as markers of a successful outcome (2,4,10). Some authors even considered the outcome to be successful when their patients required multiple surgeries to achieve an acceptable alignment (4,10). In our study, patients requiring repeat surgeries were evaluated as having a poor outcome of treatment.

This retrospective study shows that, in patients treated surgically for congenital esotropia, the presence of nystagmus may increase the risk of development of

residual or secondary strabismus requiring repeat surgery, which will make the long-term postoperative results of congenital esotropia less successful.

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## KONGENITÁLNÍ EZOTROPIE A NYSTAGMUS. VÝSLEDKY CHIRURGICKÉ LÉČBY

### S o u h r n

Kongenitální a infantilní esotropie (konvergentní šilhání) je často spojena s nystagmem. Cílem retrospektivní studie je zhodnocení vztahu přítomnosti nystagmu a jeho vlivu na výsledky chirurgické léčby vrozené a infantilní ezotropie. Studie hodnotí 316 dětí u kterých byla provedena chirurgická korekce vrozené ezotropie v letech 1985 až 1997. Je srovnáván předoperační klinický náález a následná potřeba dodatečného operačního zákroku pro reziduální nebo následné šilhání. Minimální sledovací doba byla 1 rok po prvním operačním zákroku. Z celkového počtu 316 dětí splňovalo kriteria pro zařazení do studie 196 dětí, 57 dětí (29%) mělo latentní nebo manifestní nystagmus spojený s vrozenou ezotropií, a 139 dětí bylo bez nystagmu. Třicet jedna dětí z 57 (54%) s nystagmem vyžadovalo reoperaci. U třiceti osmi dětí ze 139 (27%) bez nystagmu bylo potřeba provést reoperaci ( $P=0.096$ ). Nystagmus spojený s kongenitální ezotropií zvyšuje riziko dodatečných operací pro reziduální či následné šilhání. Jsou potřebné další studie k objasnění vlivu ostatních faktorů na výsledky chirurgické léčby dětí s vrozenou ezotropií.

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