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Strabismus Associated with Neurological Impairment: Retrospective Case Series

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Abstract

Purpose: To describe the clinical features and to discuss the results of strabismus surgery or the natural course of ocular deviations in a group of patients with neurological problems.

Material and Methods: The medical records of patients with manifest ocular deviations and neurological disorders were reviewed. The main outcome measures were the possible etiology of neurological impairment, clinical features and the results of strabismus surgery.

Results: A total of 117 (60 males, 57 females) patients were enrolled in the study. The mean age of patients was 5.6 ± 4.4 years. 58 patients (49.6%) were lost from follow-up. The most common type of strabismus was esotropia (56.4%). 36 patients (30.8%) underwent operations for strabismus. 13 patients (11.1%) had consecutive deviations during follow-up period.

Conclusions: Ocular deviations may be associated with neurological impairment. Strabismus surgery may provide good cosmesis in this group of patients. However, spontaneous and postoperative consecutive deviations should be kept in mind.

Keywords: Cerebral palsy; Esotropia; Exotropia; Neurological impairment; Strabismus

Introduction

The normal development of eye, vision and ocular movements and so the gaze systems depend on normal vissual function [1]. Some ophthalmological problems are more commonly seen in patients with neurological disorders and ocular misalignment is among the ocular problems encountered in these patients. The ocular deviations in this group requires careful assessment and follow-up because it is often difficult to have accurate measurements thus the management of strabismus is challenging [2].

The aim of the study was to evaluate the type of strabismus in a sample of patients with neurological impairment mainly with mental retardation due to variable causes, and to describe the clinical features and surgical results if performed.

Material and Methods

The study was conducted in Department of Ophthalmology, Pediatric Ophthalmology and Strabismus Section. A retrospective analysis of medical records of all patients who had strabismus and associated neurological impairment mainly mental retardation was performed.

A total of 2237 medical records were reviewed and 117 of them were found to be eligible according to present study's aforementioned inclusion criteria. The etiology and neurological assessments were based on the historical findings. Complete ophthalmological assessment was performed. Depending on the age and developmental status of the patient, Snellen letters or LEA symbols were used to assess visual acuity when possible. The cover-uncover test and Prism Cover Test (PCT) were used to assess ocular alignment at near and distance. Krimsky test was used to evaluate objective angle of deviation for uncooperative children. Titmus fly stereotest was used to examine stereoscopic vision. Worth test was used to assess fusion for cooperative child. The age at surgery, number of operations, the final status of ocular deviation, stereoacuity and fusion were also recorded. Surgical procedures were carried out under general anesthesia for all patients in a dedicated operating room.

All retrospective review was performed in strict compliance upon approval of Institutional Review Board, and all aspects of the study were carried out according to the tenets of the Declaration of Helsinki.

Statistical analyses were performed using SPSS software for Windows version 15.0 (Statistical Package for the Social Sciences, SPSS, Inc., Chicago, IL). Mann Whitney U test was used for non-parametric data. Wilcoxon signed rank test was used to compare quantitative variables in the matched sample. Arithmetic mean, standard deviation, median, range, frequency and percentage were used as descriptive statistics. The inital and final findings were given and compared only for patients who had initial and final visits. Results were accepted as statistically significant when p was <0.05.

Results

A total of 117 consecutive patients including 60 (51.3%) males and 57 females (48.7%) were enrolled in the study. The mean age of males was 5.90 ± 4.71 years, and the mean age for females was 5.39 ± 4.11 years (p=0.939). Of the 117 patients, 58 (49,6%) were examined only once, and did not visit the clinic again. Therefore, the final visit could be performed for only 59 (50.4%) patients. The median follow-up duration for these patients was 3 years (1-13). The underlying neurological problems were given in detail in Table 1.

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Etiology	No. of patients (n=117)	% of study population
Kernicterus	4	3.4
Cerebral palsy	23	19.7
Corpus callosum agenesis	5	4.4
Meningitis	4	3.4
Prematurity	8	6.8
Microcephalus	9	7.7
Silver Russel syndrome	1	0.8
Epilepsy	13	11.1
Prader-Willi syndrome	1	0.8
Phenylketonuria	3	2.6
Cornelia de Lange syndrome	2	1.7
Down syndrome	3	2.6
Rubinstein Taybi syndrome	1	0.8
Hallerman-Streiff syndrome	1	0.8
Joubert syndrome	1	0.8
Cerebrovascular event	9	7.7
Unidentified	29	24.9

Table 1: Neurological disorders associated with strabismus in the study population.

		Visits No. of patients (%)	
		Initial	Final
Operation (+)	**No deviation**	0 (0)	10 (27.8)
	Esotropia	20 (55.5)	12 (33.3)
	Exotropia	16 (44.5)	14 (38.9)
Total		36 (100)	36 (100)
Operation (-)	**No deviation**	0 (0)	4 (17.4)
	Esotropia	13 (56.5)	10 (43.5)
	Exotropia	10 (43.5)	9 (39.1)
Total		23 (100)	23 (100)

Table 2: Types of deviations at the initial and final visits.

Binocular function	With operation No. of patients		Without operation No. of patients	
	Initial	Final	Initial	Final
Fusion[a]	1	3	4	4
Stereopsis[b]	3	5	4	7
Total	4	8	8	11

[a]Fusion: Four dots on Worth test
[b]Stereopsis : by Titmus fly test

Table 3: The number of patients who had fusion and stereopsis during follow-up. Only patients who had initial and final visits were enrolled in the analysis.

The median best corrected visual acuity at initial visit was 0.6 (0.05-1.0) for 44 of 117 patients whose visual acuity could have been evaluated. Of these 44 patients, 17 were amblyopic among whom 7 had anisometropic, 8 were thought to have strabismic amblyopia and 2 had mixed. The median best corrected visual acuity at the final visit was 0.6 (0.05-1.0) (36/59 patients' visual acuity could be tested) and 11 patients were amblyopic (5/11 anisometropic, 6/11 strabismic). There was only 22 patients, whose visual acuity could have been tested at both of initial and final visits. The mean initial visual acuity of these patients was 0.6 (0.05-1.0) and it was 0.5 (0.05-1.0) finally. The difference was statistically insignificant (p=0.607).

At the initial visit, 66 patients (56.4%) had esotropia, 51 patients (43.6%) had exotropia whereas in 59 patients who had a final visit 14 (23.7%) had no deviations, 22 (37.3%) had esotropia and 23 (39.0%) had exotropia.

Twelve of 117 patients (10.3%) had nystagmus in varying degrees, and in various types. Twenty of 117 (17.1%) patients were using spectacles. Cycloplegic retinoscopy revealed hyperopia (higher than +0.50D) in 65 (55.5%) patients, myopia (higher than -0.50D) in 33 (28.2%) patients and astigmatism in 22 (18.9%) patients (higher than 0.50D) .

The strabismus surgery was performed in 36 patients (30.8%). The median number of operation was 1 (1-3). Of the 36 patients, 28 (77.8%) had 1 operation, 7 (19.4%) had 2, and one (2.8%) had 3 operations for strabismus. The mean age of operation was 6.59 ± 5.35 years. All the patients who had a strabismus surgery, had a final visit. The most commonly performed operation was recession-resection procedure in 19 patients (52.7%), followed by recession of bilateral medial recti in 12 patients (33.3%), and recession of inferior oblique in 7 patients (19.4%). The initial median near deviation was 50 Δ (20-80) and the final median near deviation was 20 Δ (0-50) in patients who had strabismus surgery (p=0.001). The initial and final distance deviations were 45 Δ (20-60) and 10 Δ (0-30) respectively (p=0.109).

However, the initial and final median deviations of 23 patients who had no operation but who had a final visit were 35 Δ (10-80) and 25 Δ (0-80) for near (p=0.03) and 14 (0-50) and 8 (0-40) for distance (p=0.465) respectively. The difference of final deviations of patients with and without operations was not significant (p=0.673).

The change of deviations was decribed in detail in Table 2. Five of patients who had esodeviation at initial visit, had exotropia at the final visit whereas 2 of patients who had exodeviation at initial visit, had esotropia at the final visit among the patients who did not have an operation. In the operated group, eso- to exo shift occurred in 4 patients, exo-to eso-shift occurred in 2 patients.

The status of fusion and stereopsis was shown in Table 3. The effect of the type of operation and the type of strabismus on binocularity could not be analyzed because of small number of patients in each group.

Discussion

In the present study, we were able to demonstrate that natural and surgical course of strabismus in patients with neurological impairment may be highly variable.

Fazzi [3] evaluated the presence of visual disorders in a sample of 121 children with central nervous system abnormalities, and observed that 72.7% of patients had strabismus (57.9% esotropia, 42.1% exotropia) [3]. They stated that the clinical manifestations of central visual impairment are heterogeneous, and include different types of refractive errors, abnormalities of visual acuity, visual field, contrast sensitivity, stereopsis, oculomotor abilities, visual perceptual abilities and strabismus [3]. They also stated that visual impairment do not depend only on visual acuity but also on many complex steps of visual perception [3]. Therefore, the visual acuity is not the only factor affecting visual impairment. In the present study, the visual acuity was not the primary outcome measure because the accurate visual evaluation was difficult to perform and unreliable on children with mental disability.

Torb-Pedersen [4] found that congenital abnormalities, low birth weight, prematurity, large head circumference were independent risk factors of strabismus whereas a large angle constant exotropia could be an indicator of underlying neurological disease in their cohort of 1320 cases [4]. They showed that children with congenital abnormalities had 57-65.5% increased risk of strabismus, and all types of congenital abnormalities were significantly associated with exotropia [4].

In the present study, the most common diagnosis was cerebral palsy in 19.7% of patients. Cerebral Palsy (CP) is the motor manifestation of non-progressive brain damage sustained during the period of brain development, and is often found with ocular abnormalities including strabismus [5,6]. The increased survival of premature neonates has resulted in growing number of children with CP [7]. Pennefather and Tin documented the ocular abnormalities in children with CP after premature birth, and observed that the prevalence of strabismus was higher compared to age matched controls without CP, whereas the prevalence of refractive errors was similar [8]. Arnoldi et al [9] reviewed 131 patients with CP and reported that 46% of the patients had decreased vision mainly related to amblyopia (24%) followed by optic nerve abnormality (16%) and cortical impairment (14%) or both [9]. They also found high frequency of binocularity among patients without strabismus [9]. However, in the present study, the visual acuity and binocular function could not be evaluated in a small number of patients.

Pennefather et al investigated the risk factors of strabismus in children born prematurely, and demonstrated an increased risk of strabismus with poor neurodevelopmental outcome in their cohort containing 558 children [10]. They postulated that neurological damage may contribute to a motor predisposition to strabismus and stated that there is no identified specific anatomical site of damage which causes specific type of strabismus [10].

Approximately half of our patients could not be reexamined (58 patients, 49.5%). All the 36 patients who had undergone an operation were seen again in control visits. Of the 81 patients who had not undergone an operation, 55 (67.9%) were lost from follow-up. This finding may show that the main reason of compliance to follow-up was the presence of an operation. The majority of the families may prefer not to bring their child again because of low expectancy about improvement of their children's medical status.

Phillips [11], reviewed the medical records of 29 patients who had intermittent exotropia with near deviation greater than distance deviation, and found a high prevalence of neurological disease, and postulated that this form of strabismus may be a 'soft' sign of underlying neurological impairment [11]. In the present study, exotropia had lower prevalence when compared to esotropia.

The great variability of the data concerning strabismus in neurologically impaired patients may complicate the decision of surgery. The risk of consecutive strabismus is in respect with fusion problems [12]. Bearing in mind that the exact relation between type of ocular deviation and group of underlying neurological disease is not currently determined, the question of the selection of patient for surgery arises.

The risk of occurence of consecutive deviation seemed to be higher in esotropic patients when compared to exotropic ones. The risk of consecutive deviations is always present after strabismus operations. It is difficult to interpret a conclusion in terms of risk of consecutive deviations because the results may only draw attention to unreliable and unrepeatable measurements. The presence of binocular function also seems to be unexpectable because of early onset of strabismus [13]. Furthermore, the amount of near deviations seemed to be decreased whether an operation had been performed or not.

There are also several limitations of the study as follows: First, this study population did not reflect the general population of children with neurological impairment, because our hospital is a referral hospital mainly not only for pediatric ophthalmology but also for pediatric neurology departments, which brings the risk of selection bias. These children were referred mainly for ophthalmological problems, and the study is based on a referral practice. Furthermore, patients who had manifest deviations were enrolled in the study. Latent deviations require specific attention and may be the essential aim of another study. However, the purpose of the study was not to determine the prevalence of strabismus in a neurologically impaired population but to investigate the natural course of strabismus in this patients. Secondly, the neurological diseases of the patients were recorded according to informations obtained from medical files, and the children were not re-examined by a pediatric neurologist, and the level of mental ability was not graded. There is no control group which consist of strabismic patients without neurological disorders. The latter may highlight a starting point for further studies. The neurological status of the patients and its progression would improve the clinical significance of the results as well because ocular alignment is mainly determined by central control mechanisms and might be affected by the level of control.

Considering the prevalence of ocular deviations accompanied with neurological diseases, this study adds to the literature, the complexity of strabismus in this group of patient, and recommends to individualize the decision of surgery. Therefore our results support the fact that the disruption of specific neural mechanisms play a significant role during the development of strabismus, and instability of measurements may sometimes reveal unpredictable surgical results despite generally accepted surgical amounts.

References

1. Boothe RG, Dobson V, Teller DY (1985) Postnatal development of vision in human and nonhuman primates. Annu Rev Neurosci 8: 495-545.
2. Ma DJ, Yang HK, Hwang JM (2013) Surgical outcomes of medial rectus recession in esotropia with cerebral palsy. Ophthalmology 120: 663-667.
3. Fazzi EI, Signorini SG, Bova SM, La Piana R, Ondei P, et al. (2007) Spectrum of visual disorders in children with cerebral visual impairment. J Child Neurol 22: 294-301.
4. Torp-Pedersen T, Boyd HA, Poulsen G, Haargaard B, Wohlfahrt J, et al. (2010) Perinatal risk factors for strabismus. Int J Epidemiol 39: 1229-1239.
5. Stanley FJ (1994) The aetiology of cerebral palsy. Early Hum Dev 36: 81-88.
6. Ghasia F, Brunstrom-Hernandez J, Tychsen L (2011) Repair of strabismus and binocular fusion in children with cerebral palsy: gross motor function classification scale. Invest Ophthalmol Vis Sci 52: 7664-7671.
7. Bhushan V, Paneth N, Kiely JL (1993) Impact of improved survival of very low birth weight infants on recent secular trends in the prevalence of cerebral palsy. Pediatrics 91: 1094-1100.
8. Pennefather PM, Tin W (2000) Ocular abnormalities associated with cerebral palsy after preterm birth. Eye (Lond) 14 : 78-81.
9. Arnoldi KA, Pendarvis L, Jackson J, Batra NN (2006) Cerebral Palsy for the Pediatric Eye Care Team Part III: Diagnosis and Management of Associated Visual and Sensory Disorders. Am Orthopt J 56: 97-107.
10. Pennefather PM, Clarke MP, Strong NP, Cottrell DG, Dutton J, et al. (1999) Risk factors for strabismus in children born before 32 weeks' gestation. Br J Ophthalmol 83: 514-518.
11. Phillips PH, Fray KJ, Brodsky MC (2005) Intermittent exotropia increasing with near fixation: a "soft" sign of neurological disease. Br J Ophthalmol 89: 1120-1122.
12. Black P (1982) Visual disorders associated with cerebral palsy. Br J Ophthalmol 66: 46-52.
13. Simonsz HJ, Kolling GH (2011) Best age for surgery for infantile esotropia. Eur J Paediatr Neurol 15: 205-208.