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Essential infantile esotropia

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Abstract

Aim : To determinate the **prevalence of congenital infantile esotropia (ETI)** in strabismic **population with Down syndrome** versus strabismic control group from the general **Macedonian population**. To present subject age, sex distribution, maternal age and spherical equivalent in each of the two analyzed groups.

Patients and methods: Population- based retrospective **case- control study** was conducted in which a study group (52) (strabismic group with Down syndrome) and a control group (257) (strabismic group of the general healthy population of children and young adults) were analyzed. Each subject underwent strabismic examination and calculated spherical equivalence based on refractive status in short-term cycloplegia. Statistical processing included the preparation of percentages of structure and average value.

Results: Out of a total of 52 children and young adults with Down syndrome and strabismus group, 4 subjects showed the presence of ETI (4/52) 7.7%. Out of a total of 257 children and young adults without Down syndrome from healthy general population, 13 subjects showed the presence of ETI (13/257) 5.0%. The average value of the spherical equivalent in the ETI Down syndrome strabismus group was +1.69 D, while the average value of the spherical equivalent in the ETI general population control group was +2.20 D

***Conclusion*: The prevalence of ETI in the Down syndrome strabismic population was 7.7% versus 5.0 % in general strabismic population** . The spherical equivalent results in our study showed a small hypermetropia, as already published studies on refractive status in ETI.

***Keywords**:* essential infantile esotropia, prevalence, spherical equivalent, treatment.

Introduction

Congenital infantile esotropia (or essential infantile esotropia) (ETI) is classically defined as a convergent strabismus (esotropia) with a very large angle of curvature, occurring in the first 6 months of life [(1)]. The prevalence of strabismus in general is 4-6%, while the prevalence of congenital esotropia in the general population is less than 1%.

Mohoney et al. [(2)] reported a prevalence of 27 cases per 10,000 neonates, while Archer et al. [(3)] reported an incidence of 0.5% in a study group of 582 children from the general population. The strabismus angle in congenital esotropia is very large, from 30-50 prism diopters (pdp), and even more. The strabismus is alternating with typical cross-fixation. T he refractive errors are small, the possibility of developing binocular vision is limited and the neurological findings are normal[(4)]. Other additional features of ETI are dissociated vertical deviation (DVD) and latent or manifest strabismus.

The etiology of ETI is controversial and still remains unknown. Historically, there are two basic theories about the cause of ETI: sensory and motor. The sensory or Worth's theory proposes that ETI is caused by the absence of cortical fusion potential. The motor or Chavasse's theory proposes that motor impairment is primary and that poor binocular sensory status occurs secondarily [(1)]. Modern understandings of the pathogenesis of ETI according to Campos [(5)] are that there is no normal motor fusion,i.e. there is a direct impairment in the center of the motor fusion. That center is located in the mesencephalon.

A NIH (National Institute of Health) sponsored a multicenter US study called the Congenital Esotropia Observational Study (CEOS), led by Kenneth Wright [(1)], reported that 43% of examined large-angle ETIs were noticed by a parent or a guardian at birth, while 23% were first observed in the first month of life [(6)]. The exact age of onset of ETI is important for the accurate defining of the condition as congenital or acquired. In summary, the occurrence of this form of esotropia is variable, with some cases being really congenital, while some acquired even a few months after birth.

Costenbader[(7)] reports that 50% of the 500 examined children with ETI had significant hypermetropia ranging from +2.25 D (diopters) to over +5.00 D. Wright and CEOS [(6)] showed that mild to moderate hypermetropia was present in most respondents, with 20% above +3.00 D, 12% above +4.00 D, and less than 10% were myopic. Birch et al.[(8)], Hiles et al.[(9)], and Mutti et al.[(10)] all report similar results on CEOS. From published studies, subjects with ETI have on average similar refractive errors as the general population of children at that age.

Our population- based study investigate the prevalence of ETI in strabismic population with Down syndrome and the strabismic control group of the general population, as well as age, sex distribution, maternal age and spherical equivalent (as a numerical expression of the refractive error) in each of the two groups.

Patients and methods

A population- based retrospective case - control was conducted in which a study group (Down syndrome group with strabismus) and a control group (general population healthy group of children and young adults with strabismus) were analyzed. The study group consisted of 52 children and young adults who met both inclusion criteria: to have clinically determined Down syndrome and to have strabismus. The control group consisted of 257 children and young adults who had strabismus but no Down syndrome and belonged to the healthy general population. Down syndrome children and young adults were examined between March 2007 and July 2009, and an appropriate Van Cleeve protocol was opened for each subject [11, 12]. Patients from the control group belonging to the healthy general population group were examined between July 2003 and December 2012 and each patient had a history of strabismus according to Von Noorden[13].

Children and young adults from the Down syndrome group were examined in the Private Polyclinic Medika Plus in Skopje, Macedonia and the Private Polyclinic Svjetlost in Zagreb, Croatia , as well as in local private ophthalmological offices in 8 other cities in North Macedonia and 3 other cities in Croatia. Children and young adults from the general population control group were examined in Private Polyclinic Medika Plus , Skopje and the Eye Diseases Center "Professor Kosta Janev", Skopje.

Comparatively, in the examined Down syndrome group and general population control group, the prevalence of congenital infantile esotropia was analyzed, as well as the average age of the subjects , gender distribution, maternal age at the time of childbirth and the spherical equivalent .

The congenital infantile esotropia was defined as a constant deviation occurring in the first 6 months of life, heteroanamnestic information obtained from a parent or a guardian.

Strabismological assessment included: 1) deviation in the primary position at distance and near using the Hirshberg test with the presence of the corneal light reflex; 2) Cover- and Cover/Uncover test ; 3) ocular motility in 9 diagnostic directions; 4) punctum proximumconvergenciae (PPC).

The classification of the type of strabismus was made according to RCOPHT (Royal College of Opthalmologists) [14] on: 1) Esotropia infantilis essentialis (congenital infantile esotropia - ETI); 2) acquired esotropia; 3) exotropias and 4) vertical strabismus.

The refractive status was determined in the short-term cycloplegia of the Potec Auto-Ref-Keratometar PRK-5000, Daejon, Korea, using Cyclopentolate 1% drops, three to five times, one drop each time, at aninterval of 15 minutes. A spherical equivalent was calculated for each

subject and emmetropia was defined as a refractive error between -0.75 D and+0.75 D spherical equivalent. Myopia was defined as a spherical equivalent less than -0.75 D and hypermetropia as a spherical equivalent larger than +0.75 D.

The statistical processing included the derivation of percentages of structure and average value.

Results

Out of a total of 52 children and young adults with Down syndrome and strabismus group, 4 subjects showed the presence of ETI (4/52) 7.7%. Out of a total of 257 children and young adults without Down syndrome from healthy general population, 13 subjects showed the presence of ETI (13/257) 5.0%.

The average age of the subjects with ETI in the Down syndrome strabismus group was 8.5 (age range 3 to 16 years), while the average age of the subjects with ETI in the general population control group was 3.7 (age range 0.8 to 14 years).

Regarding the gender distribution in the ETI Down syndrome strabismus group, all 4 respondents were female, while in the ETI general population control group 61.5% (7/13) were female, and the remaining 38.5% (5/13) were male.

The average age of the mother at the time of childbirth in the ETI Down syndrome strabismus group was 22.7 (age range 21 to 24 years), while in the ETI general population control group was 30.8 (age range 22 to 38 years) .

The average value of the spherical equivalent in the ETI Down syndrome strabismus group was +1.69 D, while the average value of the spherical equivalent in the ETI general population control group was +2.20 D.

Discussion

The prevalence of the congenital infantile esotropia in the Down syndrome population with strabismus in Macedonia showed the value of 7.7 %, while in the healthy general population with strabismus of 5.0 %. In the electronic search of Pub Med, there was no study comparing the prevalence of ETI in Down syndrome and the healthy general population with strabismus. A limitation of the study is that the Down syndrome study group was small and there was difference in the average age of the subjects. Contrary to expectations, the ETI eximinees from the Down syndrome study group with strabismus showed lower average age of the mother at the time of childbirth.

The average value of the spherical equivalent, as a numerical value of the refractive error, in the ETI Down syndrome group showed a value of +1.69 D, while in the ETI general control group +2.20 D. The results of our study fall within the scope of low hypermetropia as already published studies on refractive status in ETIs [6, 8, 9, 10].

The treatment of ETI is usually surgical. In children with ETI, hypermetropia higher than +2.00 D should be corrected with glasses [(1)]. In the case of amblyopia, occlusion of the dominant eye is required[(4)]. Complete cure is not possible even with early surgery, and the best outcome that can be achieved is subnormal binocular vision, with a small angle of deviation. Normal binocular vision (and stereo vision) does not exist even in those patients having orthotropia achieved by surgery and who have undergone surgery at an early age. Alternating strabismus with central fixation and a small angle strabismus is considered a satisfactory outcome of the treatment[(4)].

The standard surgical approach is a bilateral recession of the medial rectus muscle, using standard surgical schemes [(1)]. In adult patients with irreversible significant amblyopia, surgery is limited to the amblyopic eye by performing medial rectus muscle recession and resection of the lateral rectus muscle [(1)].

Early surgical intervention covers the period from the 6th month to the second year. Historically, it has been a standard approach with limited motor and sensory outcome. In human subjects, the critical period for the development of binocular vision is the first 3-4 months of life, a finding which brought the Nobel Prize to Hubel and Weisel[(15)].

At the 35th Congress of the European Strabismology Association (ESA, 2013), Israeli authors in a tertiary center, in a group of 83 children with ETI retrospectively analyzed the risk factors for reoperation of ETI[(16)]. Emphasizing the limitations of their study, due to the small sample of patients, the authors concluded that increase of the medial muscle recession size and the more aggressive anti-amblyopic treatment, as well as the early surgical intervention may prevent reoperation in patients with ETI. The primary versus the residual angle deviation at near, the amblyopia, and the accommodation convergence/accommodation index are significant predictors of reoperation in children with ETI.

At the same ESA congress, 2013, Graef and German authors, in a university tertiary center, in a retrospective study of 61 children with ETI with a large angle of deviation, operated with an additional method of posterior fixation suture (Faden-surgery), show that the success rate is similar to the standard method alone without the additional method. Performing the standard method with the additional Faden-surgery, which they performed exclusively on ETI with ultra-large angle of deviation, is technically much more difficult to perform and requires extensive surgical experience[(17)].

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