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CLINICAL STUDY

Refractive errors and strabismus in Asian patients with Down syndrome

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Abstract

Purpose **The purpose of this study was to investigate the prevalence and patterns of refractive errors and strabismus in Asian patients with Down syndrome, as they relate to age.**
Methods **A total of 261 Korean patients with Down syndrome were examined between March 1999 and May 2007. Eighty-nine patients were excluded from the study. The remaining 172 patients were divided into four age groups (<3 years, 3–<6 years, 6–<9 years, and ⩾9 years). Full ophthalmologic examinations and strabismus assessments were performed. Refractive errors were defined as follows: myopia ⩾−1.00 D, hyperopia ⩾+1.00 D, astigmatism ⩾±1.00 D, and anisometropia as a refractive difference between the two eyes ⩾1.00 D.**
Results **Hyperopia (46.5%) was slightly more common than myopia (40.1%). The prevalence of myopia increased with age, whereas that of hyperopia decreased. Astigmatism was found in 66.8% of patients, and astigmatism ⩾2 D was found in 16.8% of patients. Anisometropia was identified in 29.7% of patients, and the incidence of anisometropia correlated significantly with age as well as with astigmatism. Esotropia (22.1%) was twice as common as exotropia (10.5%). The prevalence of esotropia increased with age, but that of exotropia decreased. Fifty patients were found to have nystagmus (29.1%).**
Conclusions **In Asian patients with Down syndrome, esotropia was more common than exotropia and hyperopia was more common than myopia. The prevalence of exotropia and astigmatism was much higher in this study than has been previously reported.**
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Keywords: Down syndrome; nystagmus; refractive errors; strabismus

Introduction

Down syndrome is a common chromosomal disorder. It is associated with numerous ocular manifestations, including eyelid problems, keratoconus, Brushfield spots of the iris, refractive error, strabismus, and nystagmus.[1,2] Many studies have reported an increasing prevalence of refractive errors with age in patients with Down syndrome.[3,4] The prevalence of strabismus in these patients has been reported to be between 19 and 70%, with esotropia representing the vast majority of strabismus cases. Exotropia has been identified in only 6% of cases or less.[5,6]

The prevalence of refractive errors differs among different ethnic groups. Asians have the highest prevalence of myopia. In Asian patients with strabismus, exotropia is the most common type.[4,7,8] It is likely that the ocular clinical manifestations of Down syndrome also differ according to ethnicity. However, there is limited information concerning Down syndrome in Asians.[9] Moreover, there is no detailed description of strabismus or refractive errors in Asian patients with Down syndrome. This study was designed to survey the prevalence of refractive errors and strabismus in Asian patients with Down syndrome, as well as to analyse age-related changes in refractive errors and strabismus in patients with Down syndrome.

Materials and methods

One investigator (JMH) examined 261 Korean patients with Down syndrome between March 1999 and May 2007. None of the children was institutionalized. Eighty-nine patients were excluded from the study because they had undergone prior strabismus surgery or had an angle of strabismus that could not be measured secondary to poor cooperation. In the remaining 172 patients, 85 patients (49.4%) were male, and

87 (50.6%) were female. Patients were divided into four age groups: group I, <3 years; group II, 3–⩾6 years; groups III, 6–<9 years; and group IV, ⩾9 years of age. The mean age was 18.51 ± 11.04 months ($n = 45$) in Group I, 55.20 ± 10.79 months ($n = 59$) in Group II, 89.80 ± 11.87 months ($n = 44$) in group III, and 128.21 ± 20.68 months ($n = 24$) in group IV.

A full ophthalmologic examination was performed and included assessment of best-corrected visual acuity, measurement of intraocular pressure, slit-lamp examination, and fundus examination. Cycloplegic refraction was performed 45 min after administering 3–5 drops of 1% cyclopentolate by one investigator (JMH). Refractive errors were recorded as spherical equivalents. Refractive errors were defined as follows: emmetropia between −1.00 D and +1.00 D; myopia ⩾−1.00 D; hyperopia ⩾ +1.00 D; astigmatism ⩾ ± 1.00 D of cylinder; significant astigmatism ⩾ ± 3.00 D of cylinder; anisometropia, an interocular difference ⩾1.00 D; and significant anisometropia, an interocular difference ⩾2.00 D.

Strabismus was evaluated using the alternating prism cover test or Krimsky test. Sixty-three percent of the patients in group I and one patient in group II were evaluated using the Krimsky test. The alternating prism cover test was applied to all other patients. Quantification of the angle of strabismus was measured at 5 metres for distance and 33 cm for near. Infantile esotropia was defined as a constant angle with an onset before 6 months of age. Patients were assessed for the presence of nystagmus or abnormal head posture.

The four different age groups were compared with respect to refractive error and strabismus. The data were analysed using SPSS v. 15.0. The one-way ANOVA test was used to assess the differences among the groups, and Pearson's χ^2-test was used to evaluate the correlation between refractive error and age. Differences were considered significant if the P-value was <0.05.

Results

Refractive errors

The prevalences of hyperopia and myopia were 46.5% and 40.1%, respectively. High myopia (>−6.00 D) was found in 10.5% of patients among the four groups, and in up to 23% in group IV. The prevalence of myopia increased with age, whereas that of hyperopia decreased.

Astigmatism was found in 125 patients (66.8%), with significant astigmatism ⩾ ± 3.00 D found in 29 patients (16.8%). Astigmatism was most commonly found in group III (79.5%), and significant astigmatism was most commonly found in group IV (29.2%; Figure 1a). However, there was no significant relationship between astigmatism and age ($P = 0.082$). Anisometropia ⩾1.00 D was found in 51 patients (29.7%), and anisometropia ⩾2.00 D was found in 17 patients (11.0%; Figure 1b).The highest prevalence of anisometropia was observed in group IV (57.7%), which also had the highest prevalence of significant anisometropia (19.2%). Anisometropia increased with age ($P = 0.039$, correlation coefficient = 0.157). However, there was no significant difference among the four groups with regard to the rate of increase ($\chi^2 = 3$, F = 170, $P = 0.195$). A significant positive correlation was found between astigmatism and anisometropia ($P = 0.000$).

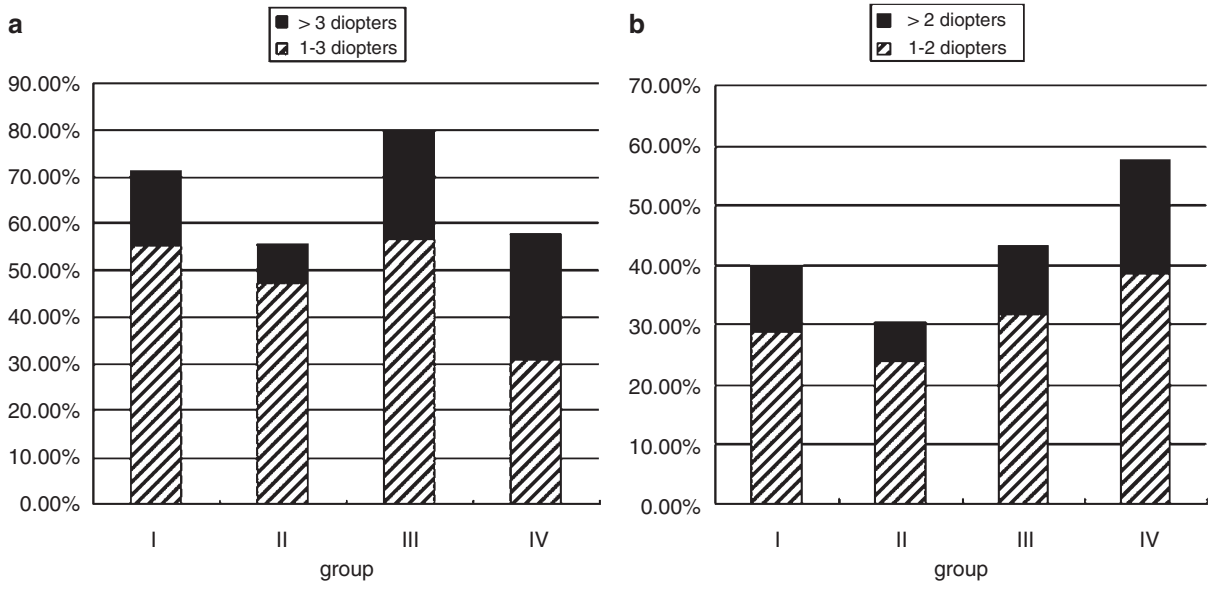

Figure 1 (a) Prevalence of astigmatism in each group. (b) Prevalence of anisometropia in each group.

Strabismus

Approximately one-third (34.3%) of the patients had strabismus (Tables 1 and 2). Esodeviation was identified in 39 patients (22.1%), exodeviation in 18 patients (10.5%), and hypertropia in three patients in group III. The most common type of esotropia was basic esotropia (68.2%), and the most common type of exotropia was also the basic type (75%). Only one patient was diagnosed with esotropia before 6 months of age. Accommodative esotropia was found in 12 patients (four refractive accommodative esotropia, two non-refractive accommodative esotropia with a high AC/A ratio, and six partially accommodative esotropia). The prevalence of esotropia increased with age, but that of exotropia decreased (Figure 2a). The mean esodeviation was 30 (range: 16–50) prism dioptres (PDs) in group I, 21 (range: 14–35) PD in group II, 22 (range: 6–40) PD in group III, and 36 (range: 12–70) PD in group IV, with no clinically significant differences observed among the four groups. The magnitude of exotropia could not be compared because of the small number of subjects. The esotropia patients had myopia and hyperopia in an equal ratio (41.5%). Hyperopia was slightly more common than myopia in the exotropia patients (Figures 2b).

Other findings

Nystagmus was noted in 50 patients (29.1%), including 17 patients (37.8%) in group I, 17 patients (28.8%) in group II, nine patients (20.5%) in group III, and seven patients (29.2%) in group IV. Orthotropia was observed in 31 nystagmus patients, esotropia in 16 nystagmus patients, and exotropia in three nystagmus patients. Head turn was found in one patient (0.6%), and head tilt in three patients (1.7%).

Discussion

The purpose of this study was to report our experience with 172 patients with Down syndrome, the largest series of Asian patients reported thus far. Although it is difficult to compare our results with previous studies because of the variegated definitions of refractive error, the prevalences of myopia and hyperopia in our study appear to be similar to those reported in earlier studies.[10] However, the prevalence of astigmatism was different from those studies, with an almost twofold increase noted in our study.[3] In addition, the prevalence of astigmatism increased with age in earlier studies.[4] However, in our study, astigmatism did not correlate with age. Group III had the highest prevalence of astigmatism among the four groups, although significant astigmatism was most commonly seen in group IV.

Anisometropia was diagnosed in 51 patients (29.7%) in our study. Almeder *et al*[11] reported that the prevalence of persistent anisometropia might be only 1% in developmentally normal children. Furthermore, anisometropia increased with age in our study ($P = 0.039$, correlation coefficient = 0.157), and significant anisometropia was noted in 19 patients. Therefore, optical correction is essential to prevent anisometropic amblyopia in these children. This increase in the prevalence of refractive errors in children with Down syndrome might reflect a failure of emmetropization.[12]

Earlier reports have documented that esotropia is common and exotropia is uncommon in individuals with

Table 1 Classification of strabismus in 57 patients with Down syndrome

Type of deviation	*0–2 years*	*3–5 years*	*6–8 years*	*>9 years*
Esophoria	—	—	1	1
Accommodative ET				
Refractive ET	—	4	—	—
Partially accommodative ET	1	3	2	—
Non-refractive accommodative ET (high AC/A ratio)	—	—	2	—
Non-accommodative ET				
Infantile ET	1	—	—	—
Acquired (basic) ET	2	5	9	8
Exophoria	—	3	—	—
Basic XT	7	3	2	—
Simulated divergence excess type XT	—	1	—	—
Divergence excess type XT	—	—	1	—
Convergence insufficiency XT	—	—	—	1

ET = esotropia; XT = exotropia.

Table 2 Comparison of clinical characteristics between patients in the current study and patients in earleir Down syndrome studies

	Current study (N = *172; %*)	*Wong and Ho*[9] (N = *140; %*)	*Roizen et al*[13] (N = *77; %*)	*Caputo et al* [3] (N = *187; %*)	*Shapiro and France* [14] (N = *53; %*)	*Jaeger*[1] (N = *7; %*)
Refractive errors	88.2	58	35	30	63	53.5
Hyperopia	46.5 (⩾ +1 D)	10.2	6.5 (⩾ +2 D)	20.9 (⩾ +3 D)	35 (> +1 D)	18.3 (⩾ +4 D)
Myopia	40.1 (⩾ −1 D)	8.6	14.3 (⩾ −2 D)	22.5 (⩾ −1 D)	37 (> −1 D)	19.7 (⩾ −4 D)
Astigmatism	66.8 (⩾1 D)	2.9 (>1 D)	6.5 (⩾1.5 D)	22.5 (⩾1 D)	25 (>1 D)	15.5 (⩾ −2.5 D)
Anisometropia	29.7	NA	NA	NA	NA	NA
Strabismus	34.3	20	27	57	43	41.3
Esotropia	22.1	18.6	21	51.8	41.2	37.3
Exotropia	10.5 (29.8)[a]	1.4 (7)[a]	1 (5.7)[a]	2.1 (3.7)[a]	1.8 (4)[a]	2.7 (6.5)[a]
Nystagmus	29.1	11	19.5	29	9	20

NA = not available.
Parenthesis-bound figures in the specific refractive error rows represent the definitions of refractive error used in each report.
[a]Percentage of strabismus represented by exotropia in patients with Down syndrome.

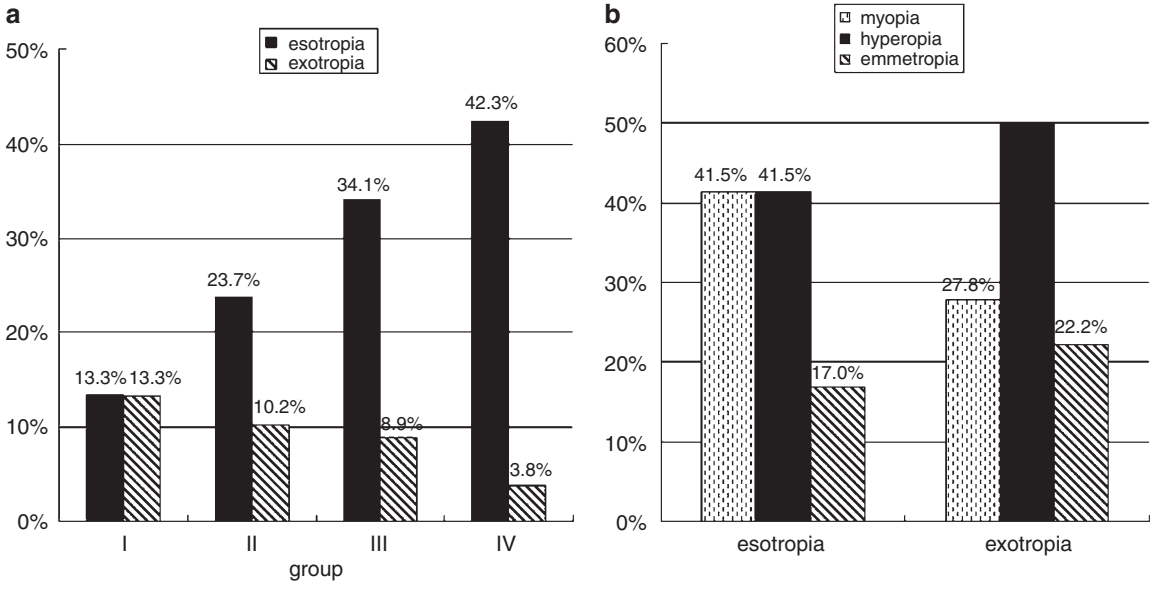

Figure 2 (a) Characteristics of strabismus in each group. (b) Rate of refractive error for each type of strabismus.

Down syndrome (incidence of exotropia 0–10%).[1,3,13,14] In this study, esotropia was the most common type of strabismus.[13,15] However, the prevalence of exotropia was 10.5% and accounted for up to 29.8% of strabismus cases. These findings reflect a much higher prevalence of exotropia than previously reported,[6,16] likely reflecting ethnic differences. The prevalence of strabismus in Korean school age children has been reported to be 3.56%, with 81.4% of those cases reflecting exotropia patients.[17] In other words, the presence of Down syndrome increases the prevalence of strabismus and modulates the racial effects on the prevalence of strabismus type.

Basic esotropia was the most common type of strabismus in our study, which is consistent with earlier reports.[5] Infantile esotropia was found in only one patient, divergent from an earlier study that found infantile esotropia in 49.4% of esotropic Down syndrome patients in Mexico.[18] This disparity might be explained by the low prevalence of infantile esotropia in Koreans,[17] as well as by a possible failure to detect esotropia because of epicanthal folds. In this study, esotropia increased with age, but exotropia decreased. Although hyperopia is usually correlated with esotropia,[19] our esotropia patients showed no difference with regard to the incidence of hyperopia and myopia. Moreover,

hyperopia was found to be more prevalent than myopia in exotropia patients. Woodhouse *et al*[20] reported that strabismus is not significantly associated with a particular type of refractive error. In addition, da Cunha and Moreira[20] reported that accommodative esotropia is of equal prevalence as non-accommodative esotropia. However, our study showed that non-accommodative esotropia was twice as common as accommodative esotropia.

In conclusion, our study showed that esotropia is more common than exotropia and hyperopia is more common than myopia in Asian patients with Down syndrome. These findings differ from those in non-Asian patients with Down syndrome. The prevalences of exotropia and astigmatism were much higher in this study than previously reported studies. These differences should be recognized when evaluating Asian patients with Down syndrome.

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