Ocular findings in children with nonsyndromic cleft lip and palate

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SUMMARY: Yaman A, Saatçi P, Arıkan G, Soylu A, Saatçi AO, Kavukçu S. Ocular findings in children with nonsyndromic cleft lip and palate. Turk J Pediatr 2009; 51: 350-353.

The aim of this study was to evaluate ocular findings in children with nonsyndromic cleft lip and palate. Fifty-seven consecutive patients with cleft lip and/or palate seeking orthodontic treatment during 2006 were examined prospectively from an ophthalmological standpoint.

Mean age of the patients was 9.2 years (range: 15 days to 18 years). Of the 57 children in total, five cases (8.7%) had cleft lip, six cases (10.5%) had isolated cleft palate and 46 cases (80.7%) had both cleft lip and palate. Thirty-seven of 46 cases with cleft lip and palate were unilateral and 20 were bilateral. Eleven of the 57 patients (19.1%) had ocular findings including congenital nasolacrimal duct obstruction (5 patients), ptosis (1 patient), bilateral iris coloboma (1 patient), dermoid tumor (1 patient), vernal conjunctivitis (1 patient), and esophoria (1 patient). Twenty patients (35%) had one or more systemic abnormalities such as motor mental retardation, hearing loss, syndactylia, growth retardation, double urinary tract, vesicoureteral reflux, penile nevus, hypospadias, non-redundant testis, inguinal hernia, mitral valve prolapsus, ventricular septal defect, complete right bundle branch block, and hirsutism.

Though not very often, cleft lip and palate patients may have several associated ocular changes, and these patients should also be examined by ophthalmologists.

Key words: cleft lip, cleft palate, eye, malformation.

Cleft lip and/or cleft palate are among the common congenital anomalies of the head and neck region. The majority of cleft lips and/or cleft palates are believed to be caused by the multifactorial inheritance whereby several genes act in concert with environmental agents[1,2]. The incidence varies according to race, geographic location, sex, and nationality. In Turkey, incidence of cleft lip ± cleft palate is 0.95‰ and the incidence of isolated cleft palate is 0.77‰[3].

In this prospective clinical study, we looked for the ophthalmic features of patients with nonsyndromic cleft lip and/or cleft under 18 years of age seeking orthodontic help.

Material and Methods

Consecutive patients with cleft lip and/or palate who sought orthodontic help at the Dental Hospital, İzmir, Turkey, throughout the calendar year of 2006 were evaluated from an ophthalmologic standpoint. Ophthalmic examination was carried out at the Ophthalmology Department of Dokuz Eylül University. Ophthalmic evaluation comprised visual acuity assessment, cover-uncover test, cycloplegic refraction, anterior segment examination with slit-lamp or hand-held slit lamp, measurement of corneal diameter and axial eye length, evaluation of distance between medial and lateral canthus, and dilated fundus examination. Visual acuity was tested with Snellen letters and Teller acuity cards. If the child was unable to cooperate, the ability to fix and follow a target was assessed. Amblyopia was defined as[1] a difference between the two eyes ≥2 lines of best corrected visual acuity with Snellen test,[2] a lack of central, steady, maintained fixation in infants, or[3] a difference between two eyes ≥ one octave of visual acuity with Teller

acuity card. All data were carefully documented. All patients were also referred to the Department of Pediatrics to rule out syndromic patients and no syndrome was yet to be diagnosed. To better analyze the data, refractive status, inner and outer canthal distance, mean axial length and corneal diameter were assessed according to age groups (0-2 years, 3-8 years, 8-13 years, 14-17 years).

Results

Fifty-seven patients comprised the study population. Twenty-nine of 57 patients (50.8%) were female and 28 (49.1%) were male. The age at examination ranged between 15 days and 17 years (average: 9.2 years), and age distribution of the study group is seen in Figure 1. Five children (8.7 %) had cleft lip, 6 (10.5%) isolated cleft palate and 46 (80.7%) both cleft lip and palate. Of the 46 cases with cleft lip and palate, 37 (80.4%) had unilateral and 20 bilateral clefts.

Ophthalmic Findings

Refractive status, mean inner and outer canthal distance, and mean axial length are illustrated in Figures 2-5.

Twelve eyes (10.5%) had myopic total spherical power, whereas 38 eyes (33.3%) had hyperopic total spherical power and 64 eyes (56.1%) had emmetropia. One of the 12 myopic eyes had myopia >5 D, while 7 of the 38 hyperopic eyes had hyperopia <3 D. Nine eyes (7.8%) had astigmatism ≥1.0 diopters. One patient had anisometropic amblyopia. The mean corneal horizontal diameter was 11.21±0.72 mm (range: 9 to 13 mm). Five patients (8.7%) had congenital nasolacrimal duct obstruction (4 unilateral, 1 bilateral) and 1 patient had unilateral ptosis. One patient (0.8%) with unilateral cleft lip and palate had bilateral iris coloboma (Figs. 6a and b). Dermoid tumor was noted in 1 patient. Vernal conjunctivitis was present in 1 patient and esophoria was the motility problem in 1 patient.

Systemic Findings

Associated systemic findings are summarized in Table I.

Discussion

We have investigated the type and incidence of associated eye features and anomalies in children with cleft lip and/or palate seeking orthodontic help.

As children with cleft lip and/or palate may have associated syndromes and various congenital defects, various authors have assessed the most common systemic associations. To the best of our knowledge, detailed prospective eye examination in children with cleft lip and/or palate was not performed previously. Data on the prevalence of associated malformations in infants with clefts were collected prospectively between 1975 and 1992 on all infants born in greater Stockholm, Sweden by Milerad et al.[4]. Of the 616 cleft infants born during this period, 21% had associated malformations and 12 cases had eye malformation (mostly colobomas). No other additional information was mentioned

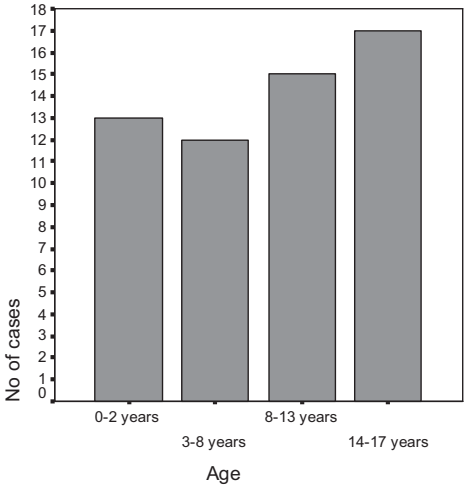

Fig. 1. Age distribution of the study group.

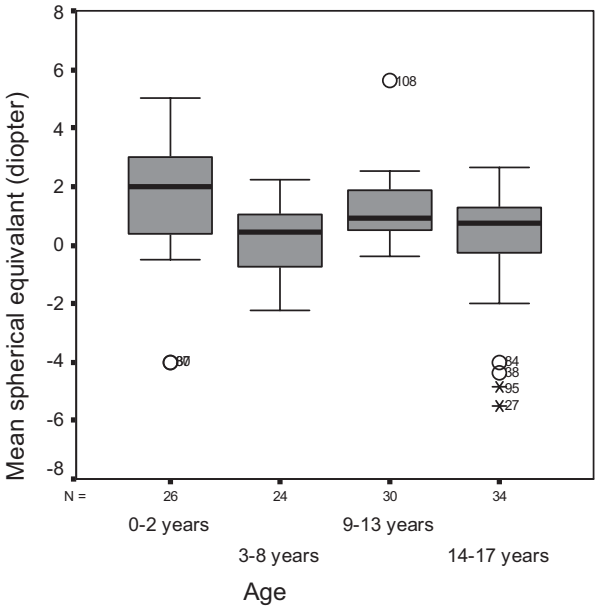

Fig. 2. Mean spherical equivalent of the study group according to age.

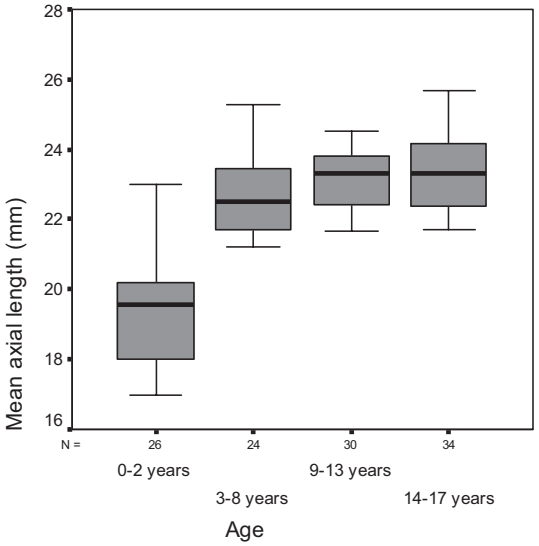

Fig. 3. Mean axial length of the study group according to age.

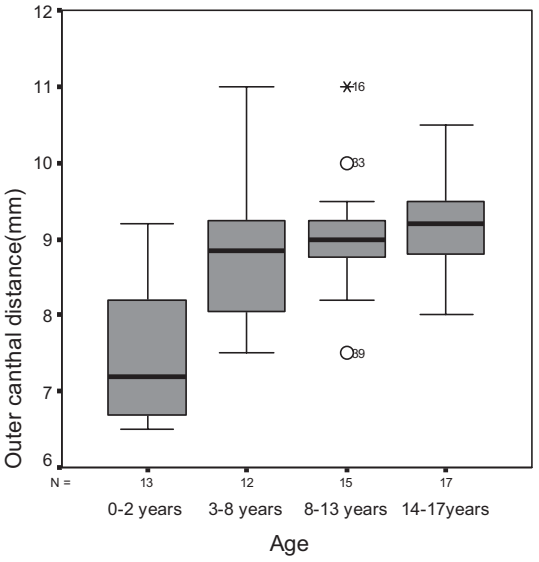

Fig. 5. Distribution of outer canthal distance of the patients according to age.

Table I. Systemic Findings of Patients

Systemic findings	No of cases
Mental retardation	5
Hearing loss	9
Syndactylia	1
Growth retardation	2
Double urinary tract	1
Vesicoureteral reflux	1
Penile nevus	1
Hypospadias	1
Non-redundant testis	1
Bilateral inguinal hernia	1
Mitral valve prolapse	1
Ventricular septal defect	1
Complete right bundle branch block	1
Hirsutism	1
Preauricular skin tag	1

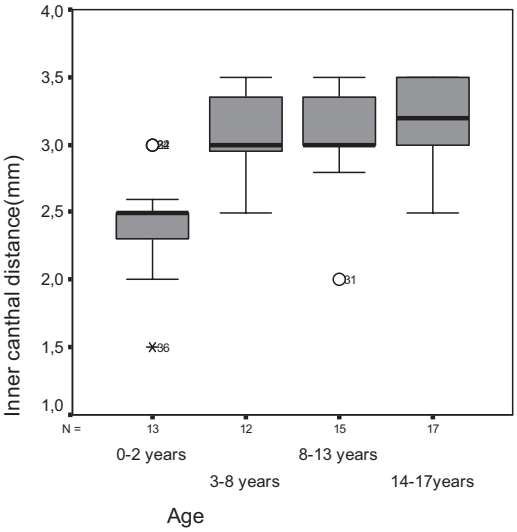

Fig. 4. Distribution of inner canthal distance of the patients according to age.

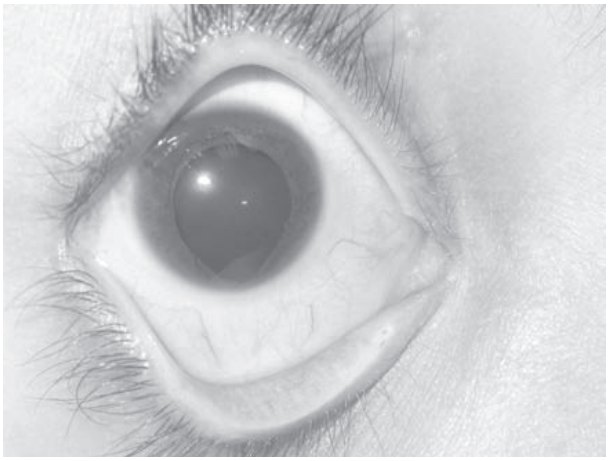

Fig. 6a. Right eye, iris coloboma.

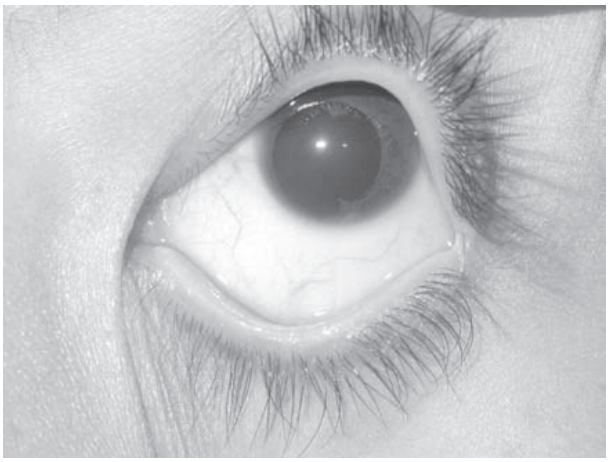

Fig. 6b. Left eye, iris coloboma.

about eye features. Elahi et al.[5] surveyed 61,156 live births during a 4.5-year study period in a northwest frontier province of Pakistan and found 117 children with cleft lip and/or cleft palate; 11 patients with nonsyndromic type had various anomalies, including one patient with telecanthus.

Tunçbilek et al.[6] classified and investigated the incidence of additional malformations and syndromes associated with 1,220 cleft lip and palate patients and showed that only 9 patients had ophthalmic findings (2 cases, anophthalmos; 2 cases, strabismus; 1 case, iris coloboma; 1 case, glaucoma; 1 case, limbal dermoid; 1 case, eyelid deformity, and 1 case, microphthalmia).

Refraction, axial length, and inner and outer canthal distance tend to change gradually during the growth process as also observed in the different age groups of our study. For instance, refractive errors change with time. Most infants are born hyperopic and become near emmetropic by 6 to 8 years of age. In our study, five patients had congenital nasolacrimal duct obstruction. However, congenital nasolacrimal duct obstruction is clinically evident in 20% of all infants and approximately 95% of infants became asymptomatic by the age of one year[7]. Therefore, nasolacrimal duct obstruction was most likely a coincidental finding in our group of patients. One patient with unilateral ptosis, one patient with bilateral iris coloboma, and one patient with dermoid tumor represent the significant eye features present in our group.

Though eye findings are not very common in cleft lip and/or palate patients, as seen in our study, there can still be important eye changes such as iris colobomas. Therefore, patients with cleft lip and/or cleft palate should also be examined ophthalmologically at least once.

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