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Research Article

Prevalence of Strabismus in Patients with Congenital Ptosis

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Abstract

Purpose: To assess the prevalence of strabismus, and its presentation in different causes of congenital ptosis.

Patients and methods: Sixty-seven patients with congenital ptosis had been examined in the Oculoplastic Ophthalmology Department. All patients underwent full ophthalmological assessment including Best Corrected Visual Acuity (BCVA), refractive error assessment, neurological, ocular motility, strabismus examinations and ptosis assessment. Cases with acquired ptosis were excluded. Strabismus was defined by the presence of constant or intermittent horizontal squint (10 or more prism diopters), vertical squint (2 or more prism diopters) or other movement disorders. Pearson's Chi-square test was used to test significance and p-value <0.05 was considered significant.

Results: Sixty-seven patients who have had congenital ptosis were evaluated; mean age (± SD) was 14.95 (± 8.615) years ranging from 4 to 35 years. They were 34 (50.7%) male and 33 (49.3%) female patients; 49 (73.1%) unilateral and 18 (26.9%) bilateral ptosis. Strabismus was found in 14 (20.8%) of the patients, 11 patients have had unilateral lid ptosis and 3 patients bilateral ptosis. 4 patients had horizontal strabismus, 7 vertical strabismus mainly due to Mono-elevation deficit and 3 combination of both (vertical and horizontal) including 3rd nerve palsy and congenital fibrosis syndrome.

Conclusion: With simple myogenic congenital ptosis patients presented with horizontal strabismus while in other etiologies of congenital ptosis (Marcus Gunn Jaw Winking, congenital fibrosis syndrome, mono-elevation deficit syndrome and congenital 3rd n palsy) the strabismus was mainly vertical one. In the current study, amblyopia was not common among patients with simple myogenic congenital ptosis probably because of counter effect of compensatory head posture or frontalis muscle recruitment. Management of functional element and aesthetic concerns must be aimed in a way that reduces the economical and psychosocial burdens on the patients and their families.

Keywords: Strabismus; Congenital lid ptosis; Binocularity; Amblyopia

Introduction

Congenital ptosis refers to the drop of the upper eyelid that is usually presented since birth or the first year of life [1-4]. It can be developed due to neurogenic causes (congenital third nerve paresis and congenital Horner syndrome) or myogenic due to poor levator or superior rectus muscle's function [2,5]. The prevalence of congenital ptosis is 0.18%-1.41% [6,7]. Congenital ptosis might be associated with strabismus [8-11] whether horizontal or vertical strabismus that might be depending on the cause of congenital ptosis. The incidence of strabismus in congenital ptosis is ranging from 10.3%-32% [9,10,12-14].

In this study, 67 patients with congenital ptosis were examined to assess the prevalence of strabismus, and its presentation in different causes of congenital ptosis.

Patients and Methods

Sixty-seven patients with congenital ptosis had been examined in the Oculoplastic, Ophthalmology Department at Ibn Al-Haithem teaching eye hospital, and Iraq during the period from February 2017 to March 2019. All patients underwent full ophthalmological assessment including Best Corrected Visual Acuity (BCVA), refractive error assessment, neurological, ocular motility, strabismus examinations and ptosis assessment. Cases with acquired ptosis were excluded. Ethics approval for this study was obtained from the Ethics Supervisory Committee of Ibn Al Haitham Teaching Eye Hospital and the study participants have given consent to participate as well as consent to publish the data.

Ptosis was assessed by measuring margin-reflex distance, palpebral fissure height, levator function, upper lid crease and pretarsal show. Marginal Reflex Distance 1 (MRD1) was measured from the center of the upper lid to the pupillary light reflex.

Strabismus was defined by the presence of constant or intermittent horizontal squint (10 or more prism diopters), vertical squint (2 or more prism diopters) or other movement disorders.

Due to the Iraq conditions in certain areas involved in war zones during the period of data collection, delayed diagnosis in certain cases was the main issue for the late presentation, in addition to the unawareness of some patients in rural areas about the seriousness of the condition. Data was collected for older patients according to history, examination, and previous hospital records. Cases were excluded from the study if there were any suspicions of acquired causes.

Statistical analysis was performed by using SPSS for Windows, ver. 15 (SPSS Inc, Chicago, IL, USA). Pearson's chi-square test was used to test significance and p-value <0.05 was considered significant.

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Results

Sixty-seven patients who have had congenital ptosis were evaluated; mean age (± SD) was 14.95 (± 8.615) years ranging from 4 to 35 years. They were 34 (50.7%) male and 33 (49.3%) female patients, 49 (73.1%) unilateral and 18 (26.9%) bilateral ptosis. The causes of congenital ptosis of the involved patients and the prevalence of strabismus are listed in Table 1.

Strabismus was found in 14 (20.8%) of the patients, 11 patients have had unilateral lid ptosis and 3 patients bilateral ptosis. Four patients had horizontal strabismus presented with simple congenital ptosis (p-value <0.001), 7 vertical strabismus mainly due to Mono-elevation deficit, including the 2 cases of Marcus Gunn Jaw Winking, and 3 combination of both (vertical and horizontal strabismus) including 3rd nerve palsy and congenital fibrosis syndrome. Types of strabismus presented are listed in Table 2.

Regarding severity of ptosis, 25 (37.3%) of the patients have mild ptosis, 21 (31.3%) have moderate ptosis and 21 (31.3%) have severe ptosis (Table 3).

The relationship between severity of ptosis and the type of strabismus is shown in Table 4. Severe ptosis was more associated with strabismus (64.3%) than mild (14.2%) and moderate (21.4%) ptosis.

Amblyopia was found in 15 (22.5%) of all patients. Amblyopia which has been caused due to strabismus alone was found in 2 (13.3%). However, amblyopia due to multiple causes including strabismus, refractive error and/or deprivation was found in 3 (20%) patients. Types of amblyopia presented in the patients involved in this study are listed in Table 5.

Abnormal head posture was noticed in 7 (10.4%) of all patients with severe bilateral lid ptosis only.

Table 1: Congenital ptosis etiology.

Causes		Strabismus
1. Simple congenital ptosis	53 (79.10%)	4 (7.50%)
2. Marcus Gunn[1]	5 (7.50%)	2 (40%)
3. Mono-elevation deficit	5 (7.50%)	5 (100%)
4. Congenital third nerve palsy	2 (3%)	2 (100%)
5. Congenital fibrosis syndrome	1 (1.50%)	1 (100%)
6. Blepharophimosis syndrome	1 (1.50%)	0 (0%)
Total	67 (100%)	14 (20.80%)

[1]Strabismus associated with Marcus Gunn were due to Mono-elevation deficit.

Table 2: Types of strabismus.

Causes	Number	Esotropia	Exotropia	Hypotropia	Others[1]
1. Simple congenital ptosis	4	1	3	0	0
2. Marcus Gunn[2]	2	0	0	2	0
3. Mono-elevation deficit	5	0	0	5	0
4. Congenital third nerve palsy	2	0	0	0	2
5. Congenital fibrosis syndrome	1	0	0	0	1
Total	14	1	3	7	3

[1]Others: refers to combined strabismus; vertical (hypotropia) and horizontal (exotropia) in the same patient.

[2]Marcus Gunn patients had hypotropia due to Mono-elevation deficit.

Table 3: Severity of ptosis and number of strabismus cases.

Ptosis	Patients (%)	Strabismus [1]
Mild	25(37.30%)	2 (3%)
Moderate	21(31.30%)	3 (4.5%)
Severe	21(31.30%)	9 (13.4%)
Total	67(100%)	14 (20.9%)

Table 4: Severity of ptosis in relation to the types of strabismus.

Ptosis	Horizontal	Vertical	Combined	Strabismus[1]
Mild	1 (7.1%)	1 (7.1%)	0	2 (14.2%)
Moderate	0	3 (21.4%)	0	3 (21.4%)
Severe	3 (21.4%)	3 (21.4%)	3 (21.4%)	9 (64.3%)
Total	4 (28.6%)	7 (50%)	3 (21.4%)	14 (100%)

[1]percentage from total number of the patients with strabismus

Table 5: Types of amblyopia in all patients with congenital ptosis.

Causes	Amblyopia	(% of amblyopic patients)	p-value
Refractive	6	40%	<0.001
Deprivation	4	26.60%	<0.001
Strabismus	2	13.30%	0.008
Mixed[1]	3	20%	0.001
Total	15	100%	

[1]Mixed includes amblyopia due to multiple causes.

Discussion

Congenital lid ptosis may associate with different forms of strabismus. Different etiologies might be involved including refractive, paralytic or restrictive.

In general, the prevalence of strabismus with congenital ptosis was ranging 10.3%-32% [6,9,10,12-19]. In this study, strabismus was found in 14 (20.9%) out of 67 patients involved in this study.

Strabismus associated with congenital ptosis could be due to the association of congenital ptosis with refractive error changes [9,10,15,20] that might cause strabismus. Also, restrictive effect of the muscles and pseudoptosis are another explanation that can be taken in consideration. In addition, strabismus might occur due to visual occlusion and binocularity disruption of the ptotic eyes [12-14,21]. Others explained it due to central nervous system insult in the area of third cranial nerve nuclear complex [10,13].

In this study, both horizontal and vertical strabismus was found. Horizontal strabismus is usually related to simple congenital ptosis even though it is associated with superior rectus weakness. Totally 4 patients presented with horizontal squint (3 exotropia and one esotropia). This finding was similar to other studies [13-15,20]. Almost all cases of congenital ptosis had coexisted refractive error [9]. This plays a major role in development of refractive strabismus which is usually presented as eso or exotropia rather than vertical strabismus.

Vertical strabismus, on the other hand, was revealed in this study in relation to restrictive and paralytic causes mainly due to mono elevation deficiency. This is caused by either restriction of the inferior rectus or weakness in superior rectus or both [23,24], as well as due to third nerve palsy and congenital fibrosis syndrome. 7 (50%) patients had vertical strabismus which was caused by mono elevation deficiency and 3 (21.4%) of the patients had combination of both horizontal and vertical squint (2 cases of third nerve palsy and one case of congenital fibrosis syndrome). Two of the patients of mono elevation deficiency have had an underlying Marcus Gunn Jaw Winking Phenomenon. It has been reported that up to 25% of monocular elevation deficiency cases were associated with Marcus Gunn Jaw Winking phenomenon [25].

Amblyopia and strabismus are two related factors; one leads to the other. Therefore, amblyopia, in all its forms, is another risk factor for developing strabismus. Refractive error was considered to have the major role in amblyopia in congenital ptosis as reported by Oral et al. [20] (71%) and Paik et al. [26] (78%), and as mentioned above, it can lead to refractive strabismus. On the other hand, deprivation amblyopia can cause sensory loss and deviation of the eye. It has been

estimated between 1.6% and 12.3% of the patients with congenital ptosis will have deprivation amblyopia due to occlusion of the visual axis [10,15,20,27-29]. While in Griepentrog et al. [28] study, they found that nearly half of the amblyopic patients with congenital ptosis were due solely to eyelid occlusion of the visual axis. This wide variation in prevalence may be related to the compensatory mechanisms that are adopted by the patient to compensate the occlusion of the visual axis like chin elevation, frontalis muscle recruitment that decreased the risk of development of deprivation amblyopia in even severely ptotic eyes [30,31], especially in bilateral ptotic patients . This was observed in this study as the abnormal head posture and chin elevation were noticed in 7 (10.4%) out of 67 patients, all with bilateral ptosis and only two patients (3%) had amblyopia.

The prevalence of amblyopia related to strabismus alone had been reported by Schneider and Martus [21] is 6% and Oral et al. [20] is 3%. In this study, amblyopia related to strabismus was seen in 2 (13.3%) of the amblyopic patients. Amblyopia due to combination of strabismus, refractive error and deprivation has been reported in 3 (20%) of amblyopic patients.

The prevalence of strabismus increased with the severity of ptosis, 9 (64.3%) patients had strabismus with severe lid ptosis (3 vertical, 3 horizontal and 3 combinations of both). However, this might be secondary to the cause of strabismus in first place, as restrictive and paralytic causes have secondary association with lid ptosis as in congenital 3rd nerve palsy, congenital fibrosis syndrome and mono-elevation deficit syndrome. Therefore, it is doubtful to consider this relationship as significant. Yet this severe ptosis has another implication on the eye, as it may be correlated with the development of deprivation amblyopia which may lead to secondary ocular deviation.

In Hornblass et al. [32] study, they found a significant relationship between the severity of ptosis and the development of amblyopia. Also Srinagesh et al. [9] found the same correlation with the severity of ptosis and the same with Merriam et al. [29] and Oral et al. [20] studies. On the contrary, Stein et al. [31] and Paik et al. [26] studies found no significant relationship between the severity of ptosis and the development of amblyopia. The same results were reported by Beneish et al. [33] and Ugurbas and Zilelioğlu [34]. In the current study we found a significant relationship between the severe eyelid ptosis and amblyopia as 9 (60.3%) of cases has severe ptosis while a non-significant relationship for mild 2 (14.2%) and moderate severity3 (21.4%).

In conclusion, with simple myogenic congenital ptosis, patients presented with horizontal strabismus while in other etiologies of congenital ptosis (Marcus Gunn Jaw Winking, congenital fibrosis syndrome, mono-elevation deficit syndrome and congenital 3rd n palsy) the strabismus was mainly a vertical one. In the current study amblyopia was not common among patients with simple myogenic congenital ptosis probably because of counter effect of compensatory head posture or frontalis muscle recruitment. However, disruption of fusion resulting in ocular deviation would be an additional indirect cause of amblyopia in congenital ptosis.

Therefore, severity of ptosis induced refractive error and presence of strabismus were related directly to the increased risk of amblyopia among those patients and caused further psychosocial impact on the patient. It is essential to approach the patient and his family in a proper way that needs a conjoint work among oculoplastic surgeons, pediatric ophthalmologists, and pediatricians. Unstable political and security situation in Iraq led to delayed presentation of patients and their management. Both might have caused increased prevalence of strabismus and amblyopia prevalence among ptotic patients in this study.

Severity of ptosis, presence or absence of associated refractive errors, strabismus and amblyopia should be assessed carefully targeting the preservation of vision and binocularity.

Management of functional elements and aesthetic concerns must be aimed in a way that reduces the economical and psychosocial burdens on the patients and their families.

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