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Research Article

Amblyogenic Factors in Congenital Lid Ptosis

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Abstract

Purpose: Assess the incidence of amblyopia and evaluate its correlation to the refractive errors, strabismus, deprivation, and severity of 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, and strabismus and ptosis assessment. Cases with acquired ptosis were excluded. Person'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 years 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. Amblyopia was found in 15 (22.5%) of the patients, 14 (20.9%) in unilateral and one (1.5%) in bilateral cases. Refractive error (9%) was found to be the main cause of amblyopia followed by deprivation (6%) and strabismic (3%) amblyopia. While mixed causes, 3 (4.4%) cases. A significant refractive error correlation to amblyopia was mainly due to anisometropia and with the rule astigmatism. Severity of lid ptosis is highly associated with amblyopia and deprivation amblyopia mainly. While head posturing acted as a protective mechanism against deprivation amblyopia.

Conclusion: High incidence of amblyopia in association with congenital ptosis, therefore, it is highly recommended to fully assess those patients for the presence of any amblyogenic factor in the ptotic eye by measuring the ptosis, refractive error assessment, ocular motility, and visual rehabilitation after surgical correction of ptosis.

Keywords: Congenital lid ptosis; Strabismus; Refractive error; Deprivation; Amblyopia

Key Message

Congenital lid ptosis is not the only problem to be concerned with. Amblyopia is the main consequences that can be avoided by finding and treating the cause.

Introduction

Congenital ptosis refers to the drop of the upper eyelid that is usually presented since birth or the first year of life [1-5]. It might be developed due to neurogenic causes like congenital third nerve paresis and congenital Horner syndrome or myogenic due to poor levator or superior rectus muscles function [2,5,6]. The prevalence of congenital ptosis is 0.18%-1.41% [7-9]. Congenital ptosis might be associated with refractive errors, strabismus, or occlusion of the visual axis that increases the risk of developing amblyopia [10-18].

Amblyopia is usually unilateral, rarely bilateral [5,19]. The incidence of amblyopia has been reported to be higher in patients with congenital ptosis (13.8% [20]) than in general population [17,21-23] (1.2%-4.4% [24]) due to the association of ptosis with refractive errors, strabismus and occlusion of visual axis [21-23,25,26]. Refractive errors are more frequently responsible for amblyopia than the other causes of amblyopia, which include anisometropia, isoametropic and meridional amblyopia. Strabismic amblyopia results from monocular suppression of the deviating eye [5,19]. Deprivation amblyopia occurs due to lid occlusion of the visual axis, but it is usually rare due to the head decompensation mechanism in human that causes abnormal head postures including head tilt and compensatory chin elevation which is more obvious in bilateral ptosis [4,5,23,26,27]. That is why several authors claim that amblyopia is rarely developing from ptosis alone without other refractive or strabismic causes [12,26,27]; however, other authors support the idea that the depth of amblyopia is directly proportional to the severity of ptosis [13,15,28].

In this study, 67 patients with congenital ptosis were examined to assess the incidence of amblyopia and evaluate its correlation to the refractive errors, strabismus, deprivation and severity of ptosis.

Patients and Methods

Sixty-seven patients with congenital ptosis had been examined in the Oculoplastic Ophthalmology Department 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 participant has 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. The severity of ptosis was classified to mild, moderate and severe depending on Marginal Reflex Distance 1 (MRD1) which was measured from the center of the upper lid to the pupillary light reflex. MRD1 equals 4 mm to 5 mm was considered to be normal, mild (MRD1 2 mm to 3mm), moderate (MRD1 1 mm to 2 mm) and severe (MRD1 0 or less).

Visual acuity was measured by Snellen chart. Amblyopia was considered in patients when the Best Correction of Visual Acuity

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(BCVA) difference is two lines with Snellen chart between the two eyes, or the BCVA is less than 6/60 (1 log MAR). Refractive error was measured by cycloplegia refraction using 1% cyclopentolate in younger patients and manifest refraction in older patients. Difference in refraction >1 diopter between the two eyes was considered as anisometropia. Astigmatism equals to 1D or more, hypermetropia more than 4D and myopia more than 6D were considered as amblyogenic.

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. Deprivation amblyopia is considered in cases that lack significant refractive error or strabismus and ptosis was severe that occlude the visual axis without compensatory head posture. The cause of amblyopia was categorized according to the above criteria to refractive (myopic, hypermetropia, astigmatism), strabismic and deprivative amblyopia. If the patient had more than one component, then the patient would be classified as ''Mixed".

Statistical analysis was performed by using SPSS for Windows, ver. 15 (SPSS Inc, Chicago, IL, USA). Person'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 years 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 incidence of amblyopia and its significance related to each cause are listed in Table 1.

Amblyopia was found in 15 (22.5%) of the patients, 14 (20.9%) in unilateral and one (1.5%) in bilateral cases. Refractive error (9%) was found to be the main cause of amblyopia followed by deprivation (6%) and strabismic (3%) amblyopia. While mixed causes, 3 (4.4%) cases, were presented as strabismus and deprivation amblyopia in one case, refractive error, and strabismus in the second case and combination of refractive, deprivation and strabismic in the third case. Types of amblyopia and its significance are listed in Table 2.

Refractive amblyopia was considered if the Best Corrected Visual Acuity (BCVA) of the patient was less than 6/6 (Log MAR 1), therefore, the refractive amblyopia was reported in two forms as anisometropia and astigmatism only. A significant refractive error correlation to amblyopia was noticed, mainly due to anisometropia and with the rule astigmatism, while it was insignificant to against the rule astigmatism, as it is shown in Table 3.

Severity of lid ptosis and abnormal head posture and their correlation to amblyopia, in all types, were assessed. A total of 85 ptotic eyes of 67 patients were included and it was found that severe ptosis was significantly associated with amblyopia, especially due to deprivation amblyopia (6 eyes with sever ptosis were associated with deprivation amblyopia (p-value 0.004)). While mild and moderate ptosis were insignificantly associated with amblyopia (Table 4).

Regarding the abnormal head posture, it is found that it was insignificantly related to amblyopia; it was noticed in 7 (10.4%) of the patients all had sever bilateral ptosis but only 2 patients has had amblyopia (p-value 0.678).

Table 1: Congenital Ptosis Etiology.

Causes		Amblyopia	p-value
Simple congenital ptosis	53 (79.1%)	8 (11.9%)	0.005
Marcus Gunn	5 (7.5%)	3 (4.5%)	0.036
Mono-elevation deficit	5 (7.5%)	1 (1.5%)	0.894
Congenital third nerve palsy	2 (3%)	2 (3%)	0.008
Congenital fibrosis syndrome	1 (1.5%)	1 (1.5%)	0.061
Blepharophimosis syndrome	1 (1.5%)	0 (0%)	0.588
Total	67 (100%)	15 (22.5%)	

Table 2: Types of Amblyopia.

Causes	Amblyopia	(% of total patients)	(% of amblyopic patients)	p-value
Refractive	6	(9%)	(40%)	<0.001
Deprivation	4	(6%)	(26.6%)	<0.001
Strabismus	2	(3%)	(13.3%)	0.008
Mixed[1]	3	(4.5%)	(20%)	0.001
Total	15	(22.5%)	(100%)	

[1]Mixed includes amblyopia due to multiple causes

Table 3: Refractive Amblyopia.

Refractive error	Amblyopia	(% of total patients)	(% of amblyopic patients)	p-value
Anisometropia	3	(4.5%)	(20%)	0.001
Astigmatism	3	(4.5%)	(20%)	0.001
WTR[1]	2	(3%)	(13.3%)	<0.001
ATR[2]	1	(1.5%)	(6.70%)	0.061

[1]WTR: With the Rule Astigmatism; [2]ATR: Against the Role Astigmatism

Table 4: Severity of Ptosis and Amblyopia.

Severity	Number of eyes (%)	Amblyopia associated	p-value
Mild	30 (35.3)	1 (6.3%)[1]	0.685
Moderate	25 (29.4%)	5 (31.2%)	0.858
Severe	30 (35.3%)	10 (62.5%)	0.011
Total	85 (100%)	16	

[1]Percentage was calculated from the total number of eyes not the patients

Discussion

The risk of amblyopia in congenital ptosis is considered the cornerstone in assessing, treating and postoperative follow up of the patients. Its incidence had been estimated to be 3.0% to 3.2% in general population and higher among patients with congenital ptosis. Previous studies had estimated the range of amblyopia to be between 14% to 48% in all forms of congenital ptosis [10-17,21,23,27,29-32]. In this study, the incidence of amblyopia was estimated at 22.5% of the 67 patients involved in this study, in most of the patients (20.9%), amblyopia was due to unilateral ptosis and only one patient due to bilateral ptosis. The commonest form of congenital ptosis is simple congenital ptosis [8,17]. Among the 67 patients, 53 patients were diagnosed with simple congenital ptosis; eight of them (15%) have had amblyopia. This is relatively close to Griepentrog et al. [17] (14.8%), Srinagesh et al. [14] (25.3%) and Lin et al. (21.5%) [16].

The leading causes of amblyopia in patients with congenital ptosis are refractive error and strabismus [11-15,21]. Almost all cases congenital ptosis with amblyopia had coexisted refractive error or strabismus [14]. The refractive error was considered to have the major role in amblyopia in congenital ptosis as reported by Oral et al. [12] (71%). and Paik et al. [27] (78%). However, other studies found that the major cause of amblyopia was the strabismus [11,33] and others deprivation amblyopia [17,34]. In the present study we found that the refractive errors in any form is the major factor that caused amblyopia

and it is contributing in about 40% of the amblyopic patients. While occlusion of the visual axis, causing deprivation amblyopia, is about 26.6% of the amblyopic patients and 13.3% due to strabismus and 20% due to combination of multiple factors of refractive error, squint or deprivation.

Refractive error almost always coexisted with congenital ptosis and presented in different forms. The highest prevalence was myopia (30.2%), followed by astigmatism (22.2%), anisometropia (17.3%) and hypermetropia (4.0%) [23]. Despite the presence of refractive error, not all the patients had amblyopia, as it can be corrected by glasses. Therefore, when considering the best correction visual acuity, the major amblyogenic refractive error is astigmatism in the ptotic eye [12,15,27,34], especially with the rule astigmatism [27]. In this study, there were an equal incidence of amblyopia due to astigmatism (20%) and anisometropia (20%), with the rule astigmatism (66.6%) is the main form of astigmatism.

The incidence of strabismus in patient with congenital ptosis ranged between 10.3% to 31.9% in various studies [7,11,14-16,23,29,30,32,33,35-37], this might be secondary to the occlusion of the visual axis by the ptotic eye causing disruption of the binocularity [25,35]. The incidence of amblyopia related to strabismus alone had been reported by Schneider et al. [26] is 6%, and Oral et al. [12] is 3%. Amblyopia due to combination of strabismus, refractive error had been reported by Schneider et al. [26] 11% due to astigmatism with strabismus, Oral et al. [12] 17% due to strabismus and refractive error. In this study the incidence of strabismus is 17.9% and amblyopia related to strabismus is 16.6% of the strabismic patients. Amblyopia due to combination of strabismus, refractive error and deprivation amblyopia has been reported in 3 (20%) of amblyopic patients. Combination of astigmatism and strabismus in the one patient, strabismus and deprivation amblyopia in second patient and combination of astigmatism, deprivation and strabismus in the third case, the last two cases strabismus were due to third nerve palsy.

Regarding deprivation amblyopia, it has been estimated that between 1.6% and 12.3% of the patients with congenital ptosis will have deprivation amblyopia due to occlusion of the visual axis [11-13,15,17,30]. While in Griepentrog et al. [8] study, they found that nearly half of the amblyopic patient with congenital ptosis were due solely to eyelid occlusion of the visual axis, approximately 1 in 7 patients diagnosed with congenital ptosis [7]. This wide variation in incidence may be related to the decompensatory mechanisms that are adopted by the patient to decompensate the occlusion of the visual axis, like chin elevation, frontalis muscle recruitment, that decreased the risk of develop of deprivation amblyopia in even severely ptotic eyes [22,34] especially in bilateral ptotic patients and acting as a protective mechanism against deprivation amblyopia. This was noticed 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. Unilateral ptosis with abnormal head posture has not been recorded in this study.

Severity of ptosis may be correlated with the development of deprivation amblyopia mainly, in addition to the other forms of amblyopia, refractive and strabismic amblyopia [12,27]. In Hornblass et al. [31] study they found a significant relationship between the severity of ptosis and the development of amblyopia. Also Srinagesh et al. [14] found the same correlation with the severity of ptosis and the same with Merriam et al. [30] and Oral et al. [12] studies. In the contrary, Stein et al. [34] and Paik et al. [27] studies found no significant relationship between the severity of ptosis and the development of amblyopia, the same results reported by Beneish et al. [29] and ugurbas et al. [38] In the current study we found a significant relationship between the severe eyelid ptosis and amblyopia as 9 cases (60%) out of 15 cases has sever ptosis while a non-significant relationship for mild and moderate severity, 3 (20%) cases for each.

This study demonstrated the high incidence of amblyopia in association with congenital ptosis, therefore, it is highly recommended to fully assess those patients for the presence of any amblyogenic factor in the ptotic eye by measuring the ptosis, refractive error assessment using cycloplegia refraction, and ocular motility, and, more importantly, visual rehabilitation after surgical correction of ptosis.

Declaration

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