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Refractive Status and Amblyopia in Simple Congenital Ptosis

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ABSTRACT

Purpose: The aim of the present study was to evaluate the refractive condition and visual status of simple congenital ptosis. **Materials & Methods:** This analytical cross-sectional study was carried out in a tertiary-based eye hospital for the period of six (6) months from January to June 2016 on 56 ptotic eyelids of 50 patients. **Results:** Age group was 9 to 38 years (Male 56%) and female 44%)) with the mean is 17.7 (±5.939) years. Unilateral was 88% and bilateral 12%. Regarding uncorrected visual acuity 52.3% had visual acuity 6/6, 6.8% had 6/9, 13.6% had 6/12, 11.4% had 6/18, 6.8% eyes had both 6/24 and 6/36 and only 2.3% had less than 6/60. In fellow eyes, visual acuity was 6/6 in 86.4% and 6.8% eyes was found 6/9 and 6/18 visual acuity in each (p=0.0001). In this study myopia was more common in ptotic eye (29.8%) than fellow normal eye (8.1%). Again, myopic astigmatism (16.2%) was more than hypermetropic astigmatism (5.4%) in ptotic eye and both were more common in in ptotic eyes. Astigmatism is more common in severe cases (50.0%) than moderate (15.4%) and mild (5.5%) cases. Myopia is also more common in severe cases (50.0%) than mild (16.7%) and moderate (7.7%) cases (p=0.0284). Among all emmetropic eyes, mild ptosis was found in 66.7% cases and moderate ptosis was found in 33.3% cases. Amblyopia was detected in 14% cases of total unilateral ptosis. **Conclusion:** Refractive error is more commonly detected in simple congenital ptosis depending on its severity. and amblyopia is more prevalent in severe unilateral ptosis.

Keywords: Blepharoptosis, congenital ptosis, Refractive error, myopia, astigmatism.

I. INTRODUCTION:

Ptosis is the drooping of upper eyelid, and it can affect all age groups.[1] BeardCdescribed eyelid drooping from normal position of 2 mm or less as mild, 3 mm as moderate, and 4 mm or more as severe.[2] As discussed later, these measurements may come into play in pre-surgical planning. According to etiological factors ptosis is classified into four categories which are Neurogenic, Myogenic, Aponeurotic and Mechanical[3].

Ptosis could be congenital or acquired.Congenital ptosis caused by an isolated developmental anomaly of the levator palpebrae muscle or poorly developed levator muscle.Congenital Ptosis is considered if present at birth or if it is diagnosed within the first year of life.[4] Congenital ptosis is common than acquired ptosis. However, Ptosis may present significant problems in children like cosmetic, functional and psychosocial.[5-7]The term acquired ptosis should be used to describe ptosis that was not present at birth but developed later in life not necessarily in adulthood.[7] The Management of Pediatric ptosis is challenging than adult ptosis because of extraconsiderations such as amblyopia, difficulty of examination, surgery under general anesthesia, and the age at which surgery should be performed.[8]

Congenital ptosis may be classified into Simple congenital ptosis, and complicated congenital ptosis. Both genders can affect. Unilateral and bilateral ptosis may occur from multiple causes and in varying amounts. Congenital ptosis may be classified into congenital simple ptosis, congenital aponeurotic ptosis, complicated congenital ptosis such as congenital ptosis with monocular elevation defect (MED), Marcus gun jaw winking ptosis (MJW), MJW ptosis with MED, Blepharophimosis syndrome (BPS) and congenital fibrosis syndrome (CFS). Usually, the most

common type of congenital ptosis results from a poorly developed levator muscle (myogenic cause). The most common type of acquired ptosis is caused by stretching or disinsertion of levator aponeurosis (aponeurotic cause)**.** Ptosis since birth with no other ocular or systemic association is diagnosed as simple congenital ptosis. It is the most common form of childhood ptosis occurring in 1 in 842 births.[9]

Congenital ptosis is generally unilateral (70%), but may be bilateral, and can be isolated or associated with disease of one or more of the extraocular muscles and/or other systemic conditions. Children with this condition suffer from obstructed vision in their upper visual quadrants and frequently require surgery to elevate their eyelids.[4] There can be a familial association in congenital ptosis, and sometimes the family member's condition may be so mild that it may not be apparent to the family. Perhaps 75% of cases of pure congenital ptosis are unilateral; however, associated eye conditions include amblyopia (20%) that may be secondary to the occlusion, astigmatism, anisometropia, or convergent strabismus.[7]Literature reveals Simple congenitalptosis accounted for 76% of childhood ptosis, whereas all other forms were relatively rare [10]. Congenital ptosis is often associates with visual impairment. Patients with congenital ptosis have higher rate of amblyopia due to greater prevalence of strabismus and refractive error although stimulus deprivation amblyopia is less common [11]**.**

A study of sixty-three patients with congenital ptosis from Taiwan found high astigmatism (>2.5D) in 25.3% of patients, anisometropia in 30.2%, amblyopia in 39.7%, and strabismus in 11.1%.[12] Sixty four percent of patients with amblyopia and ptosis had high astigmatism (>2.5D). No new amblyopia developed after surgery to correct ptosis. Hornblass [13] found amblyopia associated with congenital ptosis in 19% of patients. Patients with congenital ptosis have higher rate of amblyopia due to greater prevalence of strabismus and refractive error although stimulus deprivation amblyopia is less common**.** Due to lack of proper knowledge and treatment facilities ptosis is ignored and many children are suffering from different refractive errors as well as amblyopia. Therefore, it is very urgent to evaluate visual acuity as well as refractive errors of the patients presented with ptosis. The result of the study indicates the visual status of the children having congenital ptosis. Thus, this study was undertakento assess the visual acuity, to find out the visual condition, refractive status, amount of ptosis, and the frequency of amblyopia in the study subjects.

II. METHODOLOGY:

This analytical cross-sectional study was carried out in the oculoplastic clinic of Sheikh Fazilatunnesa Mujib Eye Hospital and Training Institute, Gopalganj, Bangladesh from the period of January 2016 to December 2019 for a period of 4 years. We included all the patients with unilateral or bilateral simple congenital ptosis at any ages above 8 years irrespective of gender, who attended in the hospital. We excluded all acquired ptosis, aponeurotic ptosis, and complicated congenital ptosis. Simple congenital ptosis less than 8 years of age also excluded from this study.The sampling technique was purposive random sampling method. This purposive sampling was used as per inclusions and exclusion criteria. Overall ophthalmic examination includingvisual acuity, andrefractive status were measured in both eyes of all study subjects.We studied the demographic variables like age, gender, and Outcome variables like visual acuity, refractive status, amblyopia, and amount of ptosis.All data were recorded systematically in preformed data collection form (questionnaire) and quantitative data were expressed as mean and standard deviation and qualitative data were expressed as frequency distribution and percentage. Statistical analysis was performed by using window-based computer software devised with Statistical Packages for Social Sciences (SPSS-20) (SPSS Inc, Chicago, IL, USA). 95% confidence limit was taken. Probability value <0.05 was considered as level of significance. The association between two qualitative variables was measured by Chi-Square test.

III. RESULTS:

We evaluated 56 ptotic eyelids of 50 eyes from 9 to 38 years of age. Among them, male is predominant than female which is 28 (56.0%) cases and 22(44.0%) cases respectively. The male and female ratio was 1.3:1. Unilateral ptosis was 88% and bilateral (**Fig. 1a**) was only 12%. Involvement of left eye is more common than right eye which is 75% and 25% cases respectively. The age group of the patients was categorized into 3 groups like as 8

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to18 years, 18 to 28 years, and 28 to 38 years (**Table-1**). According to age of presentation, the most common age group was the 8 to 18 years which was 33 (66.0%) cases followed by 18 to 28 years and 28 to 38 years age group which were 16 (32.0%) cases and 1 (2.0%) case respectively. The mean age with a standard deviation is 17.7 years (± 5.939) and the minimum age was 9 years and maximum were 38 years.

Table 1: Distribution of the Study Population according to Age of presentation (n=50)

Age group (years)	**Male**	**Female**	**Unpaired t test** **df** **P value**
8 to 18	20	13	t = 0.3030
18 to 28	07	08	df = 4
28 to 38	01	01	P = >0.5^{ns}
Total	**28 (56%)**	**22 (44%)**	

Among the 44 unilateral cases, the rate of mild, moderate (**Fig. 1b**),and severe ptosis (**Fig. 1c**) was 32%, 39% and 29% respectively. Among all the unilateral ptotic eyes, 52.3% eyes had visual acuity 6/6 (**Table-2**), 7% eyes had 6/9, 14% eyes had 6/12, 11% eyes had 6/18, 7% had both 6/24 to 6/36 and only 2.3% eye had6/60 or less visual acuity. In the fellow eyes, visual acuity was 6/6 to 6/9 in 93.2% of cases.The difference of visual acuity between ptotic and fellow eye is statistically highly significant (p=0.0001).According to visual acuity in bilateral cases. 6/6 vision was in 5 (83.3%) cases and only 1 (16.7%) case had visual acuity 6/12.

Table 2: Distribution of the involved eye among the study population according to visual acuity in unilateral Cases (Ptotic eye n=44, Fellow eye n=44)

Visual acuity	**Eye Involved**		**Total**
	Ptotic Eye	**Fellow Eye**	
6/6	23(52.3%)	38(86.4%)	61(69.3%)
6/9	3(6.8%)	3(6.8%)	6(6.8%)
6/12	6(13.6%)	0(0.0%)	6(6.8%)
6/18	5(11.4%)	3(6.8%)	8(9.1%)
6/24	3(6.8%)	0(0.0%)	3(3.4%)
6/36	3(6.8%)	0(0.0%)	3(3.4%)
<6/60	1(2.3%)	0(0.0%)	1(1.1%)
Total	**44(100.0%)**	**44(100.0%)**	**88(100.0%)**

* Pearson Chi-Square test has been performed corrected by Fisher's exact test
*p value=0.0001; Pearson Chi-Square value=17.189.

Myopia is more in ptotic eye than fellow eye which was 29.8% and 8.1% respectively. Again, myopic astigmatism is more than hypermetropic astigmatism in ptotic eye which was

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16.2% and 5.4% respectively and both the refractive error was more in ptotic eye than normal eye. Emmetropia is more in normal eye than ptotic eye which is 83.8% and 48.6 % respectively. Significant refractive error is present in unilateral ptotic eye (P=0.016). Emmetropia was found in 48.6% ptotic eyes and 83.8% in fellow eyes. Astigmatism was found 21.65% in ptotic eyes and 5.4% in fellow eyes.

Table 3: Distribution of refractive error according to the severity of ptosis in unilateral cases (n=37)

Parameter	Mild ptosis	Moderate ptosis	Severe ptosis	Total
Emmetropia	14(77.8%)	10(76.9%)	0(0.0%)	24 (64.9%)
Myopia	3(16.7%)	1(7.7%)	3 (50.0%)	7(18.9%)
Hypermetropia	0(0.0%)	0(0.0%)	0(0.0%)	0(0.0%)
Astigmatism	1(5.5%)	2(15.4%)	3 (50.0%)	6(16.2%)
Total	**18(100.0%)**	**13(100.0%)**	**6(100.0%)**	**37(100.0%)**

* Pearson Chi-Square test has been performed corrected by Fisher's exact test
*p value=0.0284; Chi-square value=6.52

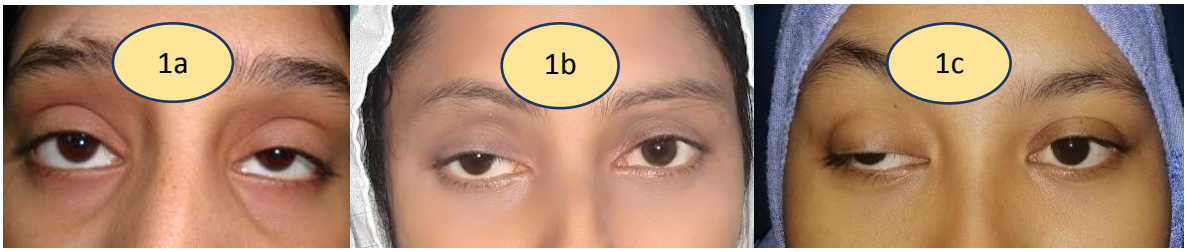

Figure 1a-c: a. Bilateral simple congenital ptosis, b. Right moderate simple congenital ptosis, c. Right severe simple congenital ptosis.

Myopic Astigmatism (16.2%) was more than hypermetropic astigmatism (5.4%) in ptotic eyes.Astigmatism was present in 50% of severe ptosis, 15.4% of moderate ptosis and 5.5% of mild ptosis (**Table-3**).Myopia was commonly found in ptotic eyes. Myopia was in 50% of Severe cases, 16.7% Mild ptosis and 7.7% of Moderate ptosis. In Bilateral cases, Emmetropia was found in 8 mild ptotic eyes and 2 moderate ptotic eyes. Myopia was in only 2 moderateptotic eyes.

Amblyopia was found in 7 ptotic eyes and total Unilateral ptotic eyes was 44. The frequency of amblyopia in total unilateral ptosis is 16%.The severe unilateral ptosis was 13 (23.2%), thefrequency of amblyopia in severe unilateral ptosis was 54% (7 in 13 cases). Amblyopia was found in only severe unilateral ptotic eyes.

IV. DISCUSSION:

56 ptotic eyes of 50 patients of both genders above 8 years of age having congenital simple ptosis unilateral or bilateral are studied for this study. Male (56.0%) is predominant than female (44.0%). The male and female ratio is 1.3: 1. Similar result has been published by Ducasse et al and has mentioned that male sex predominates in congenital ptosis.[14] Berry-Brincat and Willshaw have reported that male preponderance in the occurrence of congenital ptosis which is consistent with the present result.[15]

The distribution of the study population according to age of presentation is recorded. Out of 50 patients, the most common age group is the 8 to 18 years which is 33(66.0%) cases followed by 18 to 28 years and 28 to 38 years age group which are 16(32.0%) cases and 1(2.0%) case respectively. The mean age with a standard deviation is 17.7 years (±5.939) and the minimum is 9 years and maximum are 38 years. In majority of instance the congenital ptosis occurs at the early age. Our result is comparable to the study by Ahmadi and Sire[7] where they have mentioned that congenital ptosis is occurred after birth. Interestingly patients usually ignored this due to lack of knowledge.

Unilateral ptosis is found more common than bilateral which is 44(88.0%) cases and 6(12.0%) cases respectively. Involvement of left eye (75%) is more common than right eye (25%). Similar result presented by Allard and Durairaj[4] who have mentioned that congenital ptosis is

generally unilateral (70%); however, it may be bilateral which is less common, and this agrees with present study.

Among all unilateral ptotic eye 52.3% eyes had visual acuity 6/6, 6.8% eyes had 6/9, 13.6% had 6/12 visual, 11.4% had 6/18, 6.8% eyes had both 6/24 and 6/36 and only 2.3% eyes had visual acuity 6/60 or less. Again, among all fellow eyes visual acuity is 6/6 in 86.4% eyes and in 6.8% eyes is 6/9 and 6/18 in each. The difference of visual acuity between ptotic and fellow eye is statistically highly significant (p=0.0001). In bilateral ptotic eye 83.3% eyes had visual acuity 6/6 and only 16.7% case had 6/12.Similar result has been reported by Ducasse et al[14] who has mentioned that majority ptotic eyes have problem in visual acuity.

In unilateral cases excluding amblyopia, myopia is more common in ptotic eye (29.8%) than normal eye (8.1%). Similar finding has been reported by Griepentrog et al[10] who has mentioned that incidence of myopia is increased in ptosis patient**.** Again, in this study myopic astigmatism (16.2%) is more common than hypermetropic astigmatism (5.4%) in ptotic eye and both the refractive error is more common than fellow normal eye. Corneal topographic changes and induced astigmatism have been reported with ptosis [16] which is consistent with present study. Emmetropia is more prevalent in normal eye (83.8%) than ptotic eye (48.6%). A difference of refractive status between ptotic and fellow eye is found which is statistically highly significant. From this result it has been established that vision problems may accompany congenital ptosis like astigmatism, strabismus and amblyopia which agrees with Srinagesh et al [17]. In another study Kao et al [18] has reported that patients with congenital ptosis found high astigmatism in 25.3% of patients, anisometropia in 30.2%, amblyopia in 39.7%, and strabismus in 11.1%. Astigmatism is more common in severe cases (50.0%) than moderate (15.4%) and mild (5.5%) cases. Myopia is also more common in severe cases (50.0%) than mild (16.7%) and moderate (7.7%) cases. In bilateral cases among all emmetropic eyes mild ptosis is found in 66.7% cases and moderate ptosis is detected in 33.3% cases The difference between ptotic and fellow eye is statistically

All 7(100.0%) amblyopia cases of severe ptosis are found in unilateral cases. Amblyopia is detected in 16.0% cases of all unilateral ptosis. Amblyopia was detected in 54.0% cases of severe unilateral ptosis. Similarfindings havebeen reported in other study[18]that patients with congenital ptosis found high astigmatism in 25.3% of patients, anisometropia in 30.2%, amblyopia in 39.7%, and strabismus in 11.1%. Hornblass et al [13] found amblyopia associated with congenital ptosis in 19% of patients. significant (p=0.0284). This similar rate is comparable with previous study [19].

A study from Japan[20] showing the incidence of amblyopia is 48% in congenital ptosis. Among them 26% amblyopia due to ptosis and only 12% was stimulus deprivation amblyopia due to severe ptosis. Other study showing 6.9% amblyopia due to congenital ptosis and another study presented 19% amblyopia due to congenital Ptosis.[21] In our study, Myopia is more common in ptotic eyes than fellow eyes. Amblyopia is 14% of total congenital ptosis, 16.0% cases of total unilateral ptosis, and 54.0% cases of severe unilateral ptosis.Refractive errors such as myopia, astigmatism is more common in simple congenital ptosis. Amblyopia is more prevalent in severe unilateral ptosis. These studies confirm many classic studies on the association of ptosis and myopia and astigmatism that may cause amblyopia.

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