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AMBLYOPIA IN SIMPLE CONGENITAL PTOSIS: PREVALENCE AND CAUSES

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ABSTRACT

Purpose: To report the prevalence and causes of amblyopia among patients of Simple congenital ptosis. **Design:** Prospective observational. **Material and Method:** A prospective observational single institutional study was conducted on 37 eyes of 35 patients having Simple Congenital Ptosis. Visual acuity was measured using Snellen acuity testing. **Results**: The study conducted on 37 eyes of 35 patients revealed that 13.5% (05) of patients with simple congenital ptosis had amblyopia. Among these patients, the majority of patients (80%) had amblyopia as result of occlusion by ptotic lid while only 20% patients had amblyopia as a result of refractive errors. **Conclusion:** Early examination and follow-up of patients with Simple congenital ptosis is important to diagnose and treat any subsequent amblyopia. Surgery may be indicated should stimulus deprivation amblyopia develop.

KEYWORDS: Amblyopia, Ptosis, Visual acuity.

INTRODUCTION

Blepharoptosis is an abnormal low-lying upper eyelid margin with the eye in the primary position of gaze.[1] Congenital ptosis is present at birth or in the first year of life.[2] Congenital ptosis can cause cosmetic, functional and psychosocial problems in children.[1] In addition, it can result in abnormal postures such as backward tilt of the head, chin and brow elevation in some severe cases.[2-4] The functional impairment leads to abnormal visual development, resulting in a long-lasting uncorrected astigmatism or deprivational amblyopia. This form of amblyopia is defined as a disruption in the normal image-forming ability of the eye early in life caused by diminished performance of the visual system and severe reduction of the visual acuity due to an obscured visual field.[5] The incidence of amblyopia in the overall population has been assessed at ~3%. However, a paper published by **Willshaw**[1]**et al** provides information of an overall rate of amblyopia of up to 26.45% in a group of pediatric patients with blepharoptosis of different aetiologies. Among them, 18.7% have visually significant refractive error, while 14.19% are present with strabismus. Patients with congenital ptosis have to be closely monitored in order to preserve their visual function and establish timely and appropriate medical or surgical treatment.[6]

MATERIALS AND METHODS

A Prospective Observational single institutional study was conducted, in the Post Graduate Department of Ophthalmology, Government Medical College, Srinagar. In this study, 35 patients (37 eyes) were included, among which 18 were females and 17 were males. After obtaining the ethical clearance from the institutional ethical committee the study was conducted for the period of 02 years (Jan. 2018-Jan. 2020). Patients of all ages and both sexes having simple congenital ptosis were included, whereas, patients of acquired ptosis, complicated ptosis, ptosis associated with Marcus Gunn Jaw Winking phenomenon, ptosis with third nerve misdirection, impaired corneal sensitivity, dry eyes, ptosis with strabismus, poor bells phenomenon and mechanical ptosis were excluded from the study.

Patients of simple congenital ptosis were diagnosed clinically on the basis of history, old photographs and clinical signs. General examination and detailed ocular examination were done. Visual acuity was measured by Snellen testing. Clinically Amblyopia was defined as unilateral or bilateral reduction in the best-corrected visual acuity caused by vision deprivation and /or abnormal binocular interaction, without a visible organic cause which commensurate with this visual loss. A best corrected visual acuity less than 6/12 (20/40) was labeled as bilateral amblyopia and a difference of two or more lines between normal and amblyopic eye was required to classify it as unilateral amblyopia. After patients were diagnosed with amblyopia, initial and subsequent refractions were determined following the topical administration of 1% cyclopentolate in younger patients and by a manifest refraction for older patients. All refractions were converted into their spherical equivalent. Significant refractive error was defined as the presence of anisometropia of at least one diopter

difference between the spherical equivalents of each eye, hyperopia ≥ 3 diopters or astigmatism ≥ 1 diopter. The evaluation of ptosis was done by taking lid measurements such as Palpebral Fissure Height (PFH), Marginal Reflex Distance 1 (MRD1), Marginal Reflex Distance 2 (MRD2), Marginal Limbal Distance (MLD), Marginal Crease Distance (MCD) and Levator Function (LF).

Figure 1: Right Sided Severe Ptosis.

RESULTS

The results of this study are as following.

Table 1: Visual acuity of study eyes.		
Visual Acuity	**Frequency**	**Percentage**
6/6-6/9	32	86.5
6/12-6/18	3	8.1
6/24-6/36	2	5.4
< 6/60	0	0.0
Total	37	100

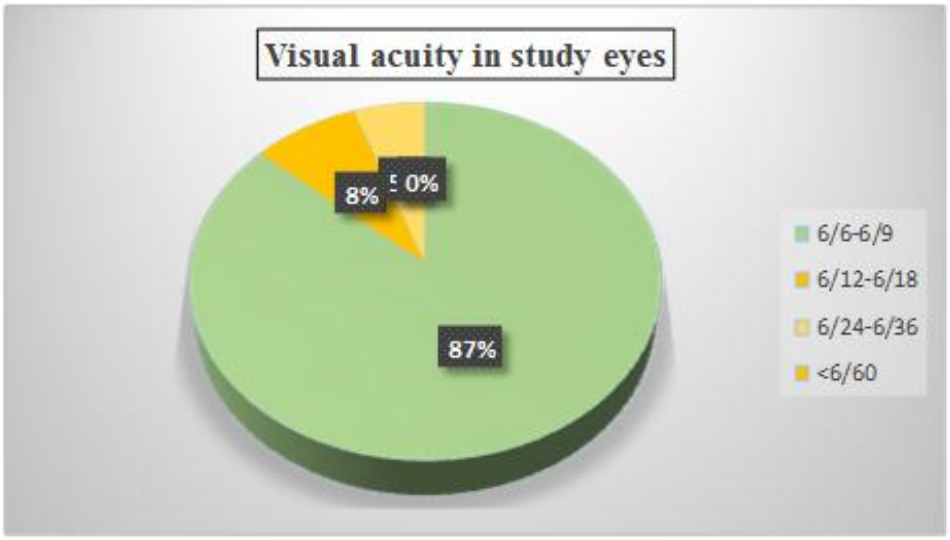

FIGURE2

The visual acuity between 6/12 and 6/18 was seen in only 03 eyes of ptosis patients while only 02 study eyes had a visual acuity between 6/24 and 6/36. This implies that only 13.5% (05) of study eyes with simple congenital ptosis had amblyopia. The majority of study eyes (86.5%) had no significant visual error as result of the simple congenital ptosis. Moreover, there was no study eye with visual acuity <6/60. (TABLE 1) (FIGURE 2).

Table 2: Causes of Amblyopia in study eyes of Simple Congenital Ptosis patients		
Cause	**Frequency**	**Percentage**
Stimulus deprivation amblyopia	4	80
Amblyopia due to significant refractive error	1	20
Total	5	100

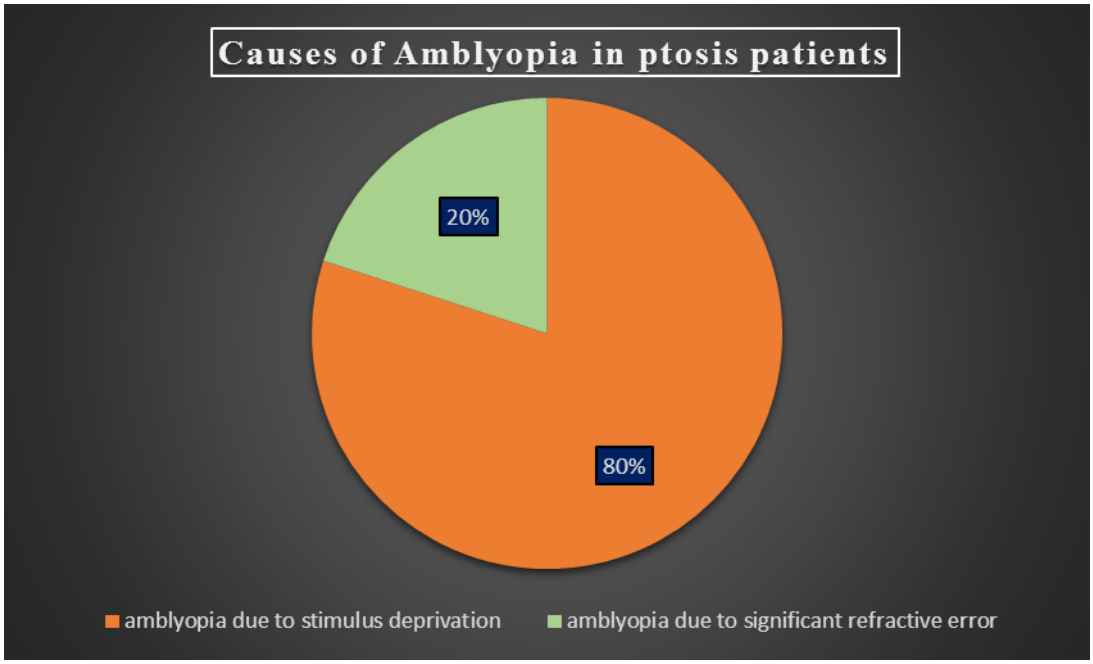

FIGURE 3

The study also revealed that in the majority (80%) of the patients, amblyopia occurred as a result of stimulus deprivation due to the drooping eyelid covering the pupillary axis especially in unilateral severe ptosis patients. The amblyopia in the rest of 20% (01) of patients was a result of significant refractive error. As this study was conducted on patients of simple congenital ptosis, the amblyopia due to strabismus was not included in the study. (TABLE 2) (FIGURE 3).

DISCUSSION

The majority of pediatric blepharoptosis cases are attributable to simple congenital ptosis and 70-80% of those are unilateral.[1,2,6,7] Eyes with congenital unilateral blepharoptosis are more predisposed to develop amblyopia. Amblyopia has an estimated prevalence of 3.0% to 3.2% in the general population.[7] The rate among patients with congenital ptosis has been reported to be higher than that of the general population and preoperatively ranges from 27% to 75%.[3,7-9] Surgical correction may be indicated whenever stimulus deprivation amblyopia due to covered optical axis develops. Previous clinical studies have examined the causes of amblyopia in the general population and have shown that approximately one third are due to anisometropia, one third to strabismus, and the remaining third to a combination of both disorders or a form of occlusive stimulus deprivation.[10] Occlusive stimulus deprivation amblyopia appears to be the least frequent subtype based on the relative rarity of the primary causative factors such as infantile cataract (2 to 4.5 of every 10,000 births) and childhood ptosis (7.9 per 100,000 less than 19 years of age).[10,11] The precise cause of amblyopia among patients with congenital ptosis is controversial. Although some authors have argued that the occlusive effect of the ptotic eyelid(s) does not interfere with visual development, subsequent reports have demonstrated that between 1.6% and 12.3% of patients with a diagnosis of congenital ptosis will have amblyopia solely due to occlusive stimulus deprivation.[7,11]

Visual acuity among the study eyes was measured with Snellen chart, which revealed that majority of patients i.e., 86.5% (32 eyes) had visual acuity ranged between 6/6 to 6/9 at the time of presentation, only 03 eyes (8.1%) had visual acuity between 6/12 to 6/18 and 02 study eyes revealed visual acuity between 6/24 to 6/36. No patient had visual acuity of less than 6/60. Only 13.5% of patients in our study population had Amblyopia. In a study conducted by **Wu SH et al**[12]**,** 84 children underwent levator resection for unilateral congenital blepharoptosis, 16.7% of these patients had amblyopia. Another study conducted by **Mokhtarzadeh A et al**[13]**,** also observed that 07 out of 47 (14.9%) patients had amblyopia. The results in both the studies are comparable to our study. This suggests that amblyopia is not very often seen among the patients of blepharoptosis even if, it is of severe degree. The absence of amblyopia among ptosis patients may be due

to presence of the compensatory head posture including backward tilt of the head and chin elevation. The frontalis overaction causing brow elevation also keeps the visual axis clear, hence preventing occurrence of amblyopia. (FIGURE 4).

Figure 4: Frontalis Overaction to Overcome Puppilary Axis Occlusion by Ptotic Lid.

All cases of amblyopia occurred in patients with unilateral ptosis. In this study, 04 out of 05 study eyes having amblyopia was due to stimulus deprivation as a result of ptosis. Only one patient had Amblyopia due to significant refractive error. The study conducted by **Gripentrog GJ et al**[14] had similar results as of our study. Twelve (14.8%) of the 81 patients diagnosed with simple congenital ptosis had amblyopia, 7 (8.6%) of which were solely due to eyelid occlusion of the visual axis. The causes of amblyopia in the remaining 05 patients: significant refractive error in 3 patients and strabismus in 2 patients. Occlusion of the visual axis was the leading cause of amblyopia in patients with congenital ptosis in this report. This finding is in contrast to large referral-based retrospective studies of congenital ptosis in which the leading causes of amblyopia were strabismus or significant refractive error.[3,7] In a study by **Harrad et al**, 216 cases of simple congenital ptosis referred for oculoplastic evaluation, 37 (17%) patients developed amblyopia, of which 20 (9.3%) cases were due to strabismus and 5 (2.3%) cases were due to stimulus deprivation of the visual axis from the ptotic eyelid. Likewise, in a study by **Dray and Leibovitch**[16] of 130 patients who had surgical correction of their ptosis, 30 (23%) patients were diagnosed with amblyopia, of which 16 (12.3%) cases were due to strabismus and 9 (6.9%) cases were due to occlusion. More recently, in a study of 92 patients with congenital ptosis, 22 (23.9%) of patients were diagnosed with amblyopia, with almost every case occurring in the context of coexisting anisometropia or strabismus. The same authors note that congenital ptosis patients are at risk of developing anisometropic and strabismic amblyopia even if not originally detected, and routine monitoring that includes regular cycloplegic refractions is recommended.[16]

CONCLUSION

Within the limitations of this study it was concluded that the prevalence of amblyopia among patients of simple congenital ptosis was 13.5% and the main cause being stimulus deprivation due to ptotic lid.

We recommend that early examination and follow-up of patients with simple congenital ptosis is important to diagnose and treat any subsequent amblyopia.The long-term results of the study can be evaluated by having a larger sample size and longer duration of study keeping in view the low prevalence of blepharoptosis.

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