

# Assessing amblyogenic factors in 100 patients with congenital ptosis

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## Abstract

- **AIM:** To study the frequency of amblyogenic factors in patients with congenital ptosis.
- **METHODS:** In this cross-sectional study, 114 eyes of 100 patients with congenital ptosis more than 1 year old were included. Amblyopia was defined as best-corrected visual acuity (BCVA) less than 10/10 or a difference between the two eyes of at least 2/10. In patients too young to be measured by the linear Snellen E test, fixation behavior was observed. Different types of amblyopia were assessed for each patient as: 1) anisometropic amblyopia: astigmatic anisometropia  $\geq 1$  dpt, hyperopic spherical anisometropia  $\geq 1$  dpt, myopic spherical anisometropia  $\geq -3$  dpt (with cycloplegia); 2) strabismic amblyopia, and 3) stimulus deprivation amblyopia (SDA). Then the total incidence of amblyopia and each type of it were obtained. Patients with uni- and bi-lateral ptosis were also compared.
- **RESULTS:** The incidence of amblyopia in ptotic eyes was 39/114 (34.2 %), and for each specific cause was: refractive amblyopia in 29.8%, SDA in 10.5%, strabismic amblyopia in 4.3%. Amblyopia was more frequent in severe ptosis, 76% in patients with covered optical axes (OA), compared to non-covered OA (22.5%). In unilateral ptosis with covered OA, astigmatic anisometropic amblyopia was more frequent, and in bilateral ptosis with at least one eye covered OA, spherical anisometropic amblyopia was more frequent. In both unilateral and bilateral ptosis, SDA was more common if the OA was covered.
- **CONCLUSION:** As refractive anisometropic amblyopia is more prevalent than SDA, paying attention to all causes of amblyopia may be important in preventing amblyopia in a child with a ptotic eye.

## INTRODUCTION

Congenital ptosis may lead to amblyopia and more severe ptosis may lead to more severe amblyopia. Also other causes of amblyopia other than stimulus deprivation amblyopia (SDA) may play a role in the development of amblyopia in congenital ptosis<sup>[1]</sup>. In this regard, some authors found a small percentage of SDA in patients with congenital ptosis<sup>[2-6]</sup>. The purpose of this study was to investigate the frequency of anisometropic, strabismic, and SDA in congenital ptosis. Moreover, the effect of unilateral or bilateral ptosis and severity of ptosis on amblyopia was studied.

## MATERIALS AND METHODS

**Materials** In this cross-sectional study, 100 patients with congenital ptosis presented to oculoplastic clinic in Farabi Eye Hospital between Jul. 2007 and Dec. 2009 were examined. The inclusion criterion was any patient with congenital ptosis more than 1 year old. Exclusion criteria were: only one seeing eye; age less than one year (because of the possibility of physiological hyperopia and astigmatism), secondary ptosis related to tumors especially neurofibromas and hemangiomas, myopathies, myasthenia gravis trauma; retinal disease; optical media opacities; third nerve palsy; double elevator palsy; blepharophimosis syndrome; neurologic and systemic diseases; Marcus Gunn jaw-winking syndrome; previous eye or ptotic surgery.

**Methods** Initially, a complete ocular examination was performed for each patient with congenital ptosis and after passing inclusion and exclusion criteria, the following data were obtained: visual acuity was measured by linear Snellen

E test. Amblyopia was defined as visual acuity <10/10 or a difference between two eyes of at least 2/10. In patients too young to be measured by the linear Snellen E test, fixation behavior was observed and if both eyes were straight, induced tropia test was done with a 10-15 diopter base-down prism over one eye to induce vertical prism and then fixation behavior was assessed. Amblyopia was considered present if there was evidence of preferential fixation. Ptosis was classified as mild (lid droop less than or equal to 2mm, or margin reflex distance +1.5mm or more), moderate (lid droop between 2mm and 4mm, or margin reflex distance +0.5), or severe (lid droop greater than or equal to 4mm, or margin reflex distance -0.5). Consequently, in mild and moderate ptosis the optical axis was not covered, and in severe ptosis was covered. The presence of strabismus was determined by orthoptic examination. The full cycloplegic refraction was then performed using cyclopentolate, 1% in children aged over three years old and 0.5% in those under three, 1 drop, two times with an interval of five minutes and after 30 minutes past the first instillation refraction was performed. Amblyogenic refractive errors were defined if: differences between eyes of 1.0 diopter (dpt) or more of hyperopic or astigmatic anisometropia and, -3 dpt or more of myopic anisometropia were present. Astigmatism was classified with-the-rule ( $0^\circ \leq \text{axis} \leq 30^\circ$ , or  $150^\circ \leq \text{axis} \leq 180^\circ$ ), against-the-rule ( $60^\circ \leq \text{axis} \leq 120^\circ$ ), or oblique ( $30^\circ < \text{axis} < 60^\circ$ , or  $120^\circ < \text{axis} < 150^\circ$ ).

As with other authors, we defined SDA as a result of congenital ptosis alone when no other reasons for amblyopia, such as amblyogenic refractive errors or strabismus were found. We investigated the following factors in congenital ptosis: 1) The frequency of amblyopia in congenital ptosis and the frequency of each causes of amblyopia; 2) These data were gathered separately for uni- and bilateral ptosis and amblyogenic factors were evaluated in uni- and bilateral ptosis groups to identify which cause or causes of amblyopia were more significant with respect to uni- and bilateral ptosis. In addition, we also considered the impact of covered optical axis to find out which causes of amblyopia were more significant in more severe ptosis.

**Statistical Analysis** Categorical data were compared using the Chi-square test. The level of significance was 0.05 with all statistical tests, using SPSS for Windows.

## RESULTS

From 100 patients with congenital ptosis, 61% were male and 39% were female. Of those patients, 86% had unilateral and 14% had bilateral ptosis. Positive family history was present in 23% of patients. Overall, the incidence of amblyopia in congenital ptosis was 34.2% (39 out of 114 eyes); compared to the prevalence of amblyopia in general population of 3.2-4.6%<sup>[7-8]</sup>, was statistically highly significant ( $P < 0.001$ ). The incidence for each specific cause of

**Table 1 Percentage of different types of Amblyopia in uni-and bilateral ptotic eyes with open or covered optical axis**

Amblyopia	Ptosis	Optical axis	
		Not covered (%)	Covered (%)
Astigmatic anisometropic	Unilateral	11.6	35.3 <sup>b</sup>
	Bilateral	10.0	25.0
Spherical anisometropic	Unilateral	10.1	17.6
	Bilateral	10.0	50.0 <sup>a</sup>
SDA	Unilateral	7.2	29.4 <sup>b</sup>
	Bilateral	0	25.0 <sup>a</sup>
Strabismic	Unilateral	4.3	5.9
	Bilateral	5.0	0

<sup>a</sup> $P < 0.05$ , <sup>b</sup> $P < 0.01$  covered vs not covered

amblyopia in 114 ptotic eyes was: refractive amblyopia in 29.8% (34 eyes), SDA in 10.5% (12 eyes), strabismic amblyopia in 4.3% (5 eyes); concerning the fact that each eye may have more than one cause of amblyopia. The incidence of amblyopia in unilateral congenital ptosis was 34.9% (30 out of 86 ptotic eyes had amblyopia). The incidence of amblyopia of the fellow eye was 6/86 (6.9%). In bilateral ptosis, the incidence was 32.1% (5 patients with amblyopia affecting one eye and 2 patients with amblyopia of both eyes were observed, making a total of 9 out of 28 eyes). Statistically, the incidence of amblyopia was not different between unilateral and bilateral ptosis in ptotic eye ( $P = 0.79$ ); but for specific causes of amblyopia when severity of ptosis was more, there were some differences.

In unilateral ptosis, eyes with severe ptosis (with covered optical axes) were amblyopic in 76.5% of cases (13 out of 17 eyes) and showed a higher frequency of amblyopia than those with non-covered optical axes: 24.6% (17 out of 69) which is statistically significant ( $P < 0.05$ ). In bilateral ptosis, the frequency of amblyopia was roughly the same; amblyopia was present in 6 eyes out of 8 (75.0%) when ptosis was severe (with covered optical axis) versus 3 out of 20 (15%) in non-covered optical axis, which is also statistically significant ( $P < 0.05$ ).

In unilateral ptosis when ptosis was severe (optical axis was covered) relative to mild and moderate ptosis (optical axis was not covered) astigmatic anisometropic amblyopia was statistically more significant ( $P = 0.01$ ), but that was not true in bilateral ptosis ( $P = 0.30$ , Table 1). In bilateral ptosis when ptosis was severe (optical axis was covered) relative to mild and moderate ptosis (optical axis was not covered) spherical anisometropic amblyopia was statistically more significant ( $P = 0.02$ ), and that was not right in unilateral ptosis ( $P = 0.38$ , Table 1). In unilateral ptosis, when optical

axis was covered, SDA (SDA) was statistically more significant than when optical axis was not covered ( $P=0.01$ ); and that was also true for bilateral ptosis ( $P=0.02$ , Table 1). Strabismic amblyopia was not statistically significant between covered optical axis and non-covered optical axis in unilateral ( $P=0.78$ ) and bilateral ( $P=0.52$ ) ptosis (Table 1). In 30 ptotic eyes that had astigmatism, 90% (27 eyes) were with-the-rule, 6.7% (2 eyes) were against-the-rule, and 3.3% (1 eye) was oblique astigmatism.

### DISCUSSION

The incidence of amblyopia in congenital ptosis that was reported by different authors ranged from 6% to 50% of the patients<sup>[1-6,9]</sup>. In our series of patients, we found amblyopia in 34.2% of ptotic eyes. The difference in the incidence of amblyopia may be because of different definition of amblyopia by different authors and different inclusion and exclusion criteria that each author may have. Furthermore, some authors do not consider a second cause of amblyopia if one is predominant.

In our study, we found that if ptosis was more severe the possibility of amblyopia becomes more. In unilateral ptosis when ptosis was severe (with covered optical axis) relative to mild and moderate ptosis the incidence of amblyopia was 76.5% ( $P<0.05$ ); and in bilateral ptosis it was 75% ( $P<0.05$ ). Beneish *et al*<sup>[9]</sup> found no correlation between severity of ptosis and the incidence of amblyopia; but Hornblass *et al*<sup>[4]</sup> stated significant correlation, as our study. The question is when severity of ptosis increases that causes of amblyopia becomes more prominent. We found different results between uni- and bilateral ptosis. In unilateral ptosis when ptosis was severe (optical axis was covered) relative to mild and moderate ptosis (optical axis was not covered) astigmatic amblyopia was statistically more significant, but that was not the case in bilateral ptosis. The reason of that not found in bilateral ptosis may be, bilateral ptosis may induce the same amount of astigmatism in both eyes to cause astigmatic anisometropic amblyopia.

In bilateral ptosis when ptosis was severe (optical axis was covered) relative to mild and moderate ptosis (optical axis was not covered) anisometropic amblyopia was statistically more significant, however that was not right in unilateral ptosis.

The incidence of each type of astigmatism found in congenital ptosis was different between authors. Stärk *et al*<sup>[6]</sup> found equal incidence of against-the-rule astigmatism relative to with-the-rule astigmatism. Charlotte *et al*<sup>[10]</sup> found only the two forms of astigmatism, oblique and

with-the-rule, more often and slightly more pronounced in cases with more severe ptosis. In our series, we found with-the-rule astigmatism was more frequent compared to against-the-rule and oblique astigmatism in ptotic eyes (90% were with-the-rule astigmatism). This may propose that astigmatism may be caused by ptosis relative to the fact that they may be coincidentally associated.

The low incidence of strabismus in our series of ptotic eyes may be the result of our strict exclusion criteria, not including a third nerve or double elevator palsy (in which strabismus is almost always present). We found 4.3% of ptotic eyes had strabismic amblyopia. The incidence of strabismus found by different authors ranged from 6% to 76%<sup>[2-6,9,10]</sup>. The incidence of SDA as the only cause of amblyopia in congenital ptosis found in 10.5% of 114 ptotic eyes in our patients. SDA occurred more frequently in ptosis with covered optical axes in both uni and bilateral ptosis. The frequency of stimulus deprivation amblyopia in congenital ptosis reported in different studies ranged between 0% to 5.6%<sup>[2-6,9,10]</sup>.

In conclusion, since other causes of amblyopia other than SDA (especially refractive anisometropic amblyopia) may play a role in more severe ptosis, paying attention to all causes of amblyopia may be important in preventing amblyopia in a child with a ptotic eye; and simply opening the visual axis with surgery without a pre- and postoperative refraction may not suffice in preventing amblyopia.

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