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Refractive Error Characteristics in Patients with Congenital Blepharoptosis Before and after Surgery

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ABSTRACT

Objective: To surgically manage patients with congenital blepharoptosis and compare the refractive errors before and after intervention

Study Design: Quasi-experimental study.

Place and Duration of Study: Department of Orbit and Oculoplastic surgery, Armed Forces Institute of Ophthalmology Rawalpindi from February 2022 to December 2022.

Methodology: Patients with congenital ptosis who underwent surgical management at our setup and those who stick to post-operative follow up for at least 6 weeks were included in the study. An average of 6 readings were taken of an automated refractometer to measure pre- and post-operative refractive error.

Results: Out of total 20 patients, 11(55%) were males while 9(45%) were females. Age of patients who were surgically managed ranged from 3 to 13 years. All patients had pre-op diagnosis of unilateral simple congenital ptosis. There was significant difference (in terms of reduction of refractive error) in pre and post op refractive error measured in diopters.

Conclusion: Congenital blepharoptosis may present as a wide variety of refractive errors and upper lid blepharoptosis surgery repair have shown to improve the refractive error secondary to induced astigmatism. However, no particular surgery technique is found to be superior to other in our study sample and all surgical procedures had proven to be safe and effective approach towards blepharoptosis repair.

Keywords: Blepharoptosis, Congenital ptosis, Ptosis repair surgery, Refractive error.

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INTRODUCTION

Blepharoptosis is defined as the drooping of the upper eyelid resulting in covering of the superior aspect of cornea. The drooped eyelid may or may not cover the visual axis and thus affect vision. The normal adult upper eyelid margin lies 0.5-2mm below the superior corneal limbus and is highest just nasal to the pupil.[1] Ptosis can present at any age group but for the purposes of this study we will focus on congenital ptosis. Congenital ptosis is the narrowing of the palpebral fissure due to a drooping upper lid margin presenting within the first year of life.[2] Presentation can be both unilateral and bilateral and can occur in isolation or in association with some other ocular as well as systemic abnormality/disorder.[3] It also conventialy referred to as ptosis.

While in adults the ocular system is fully developed and thus ptosis in itself is benign (although it may be a sign of a more sinister underlying cause), children are more seriously affected by ptosis as the narrowed palpebral fissure can impair light entry into the eye and thus the affected eye is left unused and may develop sensory deprivation amblyopia. Drooping of upper lid is also known to cause changes in corneal topography and thus lead to astigmatism which may cause refractory or anisometropic amblyopia if left untreated. Congenital ptosis is a non-progressive condition but it can be associated with significant defects of normal visual development and thus needs to be managed adequately to prevent long term complications.[2]

The levator palpabrae superioris (LPS) and the superior rectus (SR) muscles are responsible for the elevation of the upper eyelid and dysfunction in one or both can lead to ptosis. The reasons for their dysfunction are multiple and highly varied and so management depends on identifying the underlying cause. Over the years multiple theories have been proposed regarding its pathogenesis. While initially it was that defective muscle development was the primary reason for congenital ptosis, newer theories suggest alternative causes including levator palpebrae superioris muscle dysgenesis and disorders of muscle denervation.[4,5]

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Early treatment is essential to prevent development of visual defects such as amblyopia, refractive errors and astigmatism. Surgical treatment should be considered only when non-surgical options are exhausted or when the ptosis is interfering with the visual field axis.[6] Multiple surgical approaches are available with the frontalis sling procedure being the most commonly used. Surgical repair of ptosis can often involve multiple surgical procedures and the exact type of procedure used depends on the levator function and the amount of ptosis. These procedures can be done separately or as a combined single procedure.[7] After surgical correction of unilateral ptosis, contralateral eyelid may become ptotic. Due to Herings law which states that ocular yoke muscles receive equal innervation, the brain increases innervation to both LPS muscles to clear the visual axis and counteract the ptotic eyelid. After unilateral ptosis repair this innervation is decreased and thus, correspondingly the contralateral normal eyelid may experience lowering.[8]

The objective of this study is to surgically manage patients with congenital blepharoptosis and compare the refractive errors before and after intervention. We aim to explore how effective is the surgical management of congenital ptosis while also attempting to compare the different surgical procedures used for ptosis repair and how they compare against one another.

METHODOLOGY

The quasi-experimental study was carried out department of Orbit and Oculoplastic surgery, Armed Forces Institute of Ophthalmology Rawalpindi for a duration of 10 months from February 2022 to December 2022 after approval from Institutional review board (274/ERC Dated 01 May 2022). A sample size of 20 was calculated using reference prevalence for congenital ptosis to be 1.26%, and a confidence level of 95%.[1] OpenEpi Online Software was used to calculate the sample size. Written informed consent was sought from the parents of all the patients. Patients with congenital ptosis who underwent surgical management at our setup and those who stuck to post-operative follow up for at least 6 weeks were included in the study. Patients with myogenic, neurogenic or traumatic causes of ptosis, bilateral congenital ptosis or patients who had any other significant ocular pathology and or amblyopia were excluded from the study. A preformed Microsoft Excel sheet was used for collection of data by a single investigator under headings of different variables including age, gender, diagnosis, laterality of ptosis, severity of ptosis, pre-op lid contour, pre-op refractive error and spherical equivalent in diopters, surgical technique used, post-op Marginal reflex distance (MRD) 1 post-op lid contour, post-op refractive error and spherical equivalent in diopters, post-op inter eye symmetry and post-operative complications. Ptosis was classified in to three grades based on MRD 1 i.e., distance from corneal reflection to upper lid margin, between both eyes; mild (2mm), moderate (3 mm) and severe (4mm). Upper lid crease was graded as absent (no lid crease), poor (some contour/crease forms) and fair (Upper lid crease present) by a senior oculoplastic surgeon.

An average of 6 readings were taken of an automated refractometer to measure pre-and post-operative refractive error. Spherical equivalent (by adding half of the cylindrical power to spherical power) was taken for cylindrical errors for ease of standardization and data analysis. Data was analyzed using IBM SPSS version 22 software (Chicago, Illinois). Patient confidentiality was maintained at each and every step of study.

RESULTS

Out of total 20 patients, 11(55%) were males while 9(45%) were females. Age of patients who were surgically managed ranged from 3 to 13 years. All patients had pre-op diagnosis of unilateral simple congenital ptosis. Twelve patients (60%) had involvement of right side while rest of them had left sided ptosis. Classification of ptosis on the basis of pre-operative MRD1 revealed severe ptosis in 4(20%), Moderate ptosis in 9(45%) and mild ptosis in 7(35%) patients.

Pre-Operative Upper lid contour was absent in 8(40%), poor in 5(25%) and fair in 7(35%) patients. Four (20%) patients underwent levator aponeurosis advancement, 8 underwent levator resection and 5(25%) underwent Unilateral frontalis sling, 2(10%) underwent Muller muscle resection and only 1(5%) patient had bilateral frontalis sling procedure.

There was significant difference in pre and post op refractive error measured in diopters where a difference of 0.25 Diopter was documented in 6(30%) cases, 0.50 Diopters in 4(20%) cases, 1 Diopter in 2(10%) cases and 9 (45%) cases showed no difference in pre and post op refractive error correction. Post-operative eyelid symmetry achieved using different surgical approaches is shown in Table-I. Post-operative complications are shown in Table-II.

Table-I: Post-operative outcomes using Different Surgical Procedure for Blepharoptosis Repair (n=20)

		Surgical Procedure for Blepharoptosis Repair				
		Levator Aponeurosis Advancement	Levator resection	Unilateral Frontalis Sling	Muller Muscle Resection	Bilateral Frontalis Sling
Post-operative symmetry between two eyes	<1.5mm	2(10%)	6(30%)	1(5%)	2(10%)	1(5%)
	1.5 to 2.5mm	1(5%)	2(10%)	3(15%)	0	0
	>2.5mm	1(5%)	0	1(5%)	0	0

Table-II: Post-Blepharoptosis Repair Surgery Complications(n=20)

Post-operative Complications	Frequency
Lagophthalmos	1(5%)
Overcorrection	4(20%)
Hematoma	1(5%)
Dyplopia	1(5%)
Under correction	3(15%)
Corneal Abrasion	1(5%)
Lid lag	1(5%)
No Complication	8(40%)

DISCUSSION

Ptosis is a fairly common eyelid disorder present within all age groups with a wide range of causes. Congenital ptosis however, is rarer and large-scale surveys for its prevalence have not yet been widely conducted so data regarding that is scarce. One of the largest studies having been conducted in this regard was in China. The study showed the prevalence of congenital ptosis in a sample size of seven million people to be 0.18%. Among these, majority of the cases were sporadic; although a genetic tendency was also seen with 18.4% presenting with autosomal dominant inheritance and 14.5% presenting with autosomal recessive inheritance.[9] Other studies having been conducted show a more common incidence of congenital ptosis, with a study being conducted in Egypt identifying 336 children with ptosis over a nine-year period of which 69% had congenital ptosis.[10] A review of cases of childhood ptosis over a 40-year period in Olmsted County, Minnesota was conducted. 107 cases were identified, with an incidence of 7.9 per 100,000. 89.7% had congenital ptosis, with 12% having a positive family history. The rate of congenital ptosis was 1 in 842 births. Only 3% were bilateral, and there was a slight predominance of left ptosis (55%).[11]

There are a wide range of underlying causes for the ptosis. While many cases are sporadic, genetic association has been observed in previous studies. Conditions causing congenital ptosis include CFEOM (congenital fibrosis of extraocular muscles), CN (cranial nerve) III dysfunction, Duane retraction syndrome and BPES (Blepharophimosis, blepharoptosis, epicanthus inversus and telecanthus) disorder. The more common genetic cause of congenital ptosis is Marcus Gunn syndrome accounting for almost 5% of all congenital ptosis cases 6. PTOS1 gene was the first to be identified as a potential cause of isolated congenital ptosis,[12] and in 2002 McMullan *et al.*, described another form of congenital ptosis which was inherited in an X-linked dominant form.[13]

Congenital ptosis can be mild, moderate or severe depending the degree of the eyelid being covered and classified on the basis of difference of MRD1 between 2 eyes. Mild is 2mm, moderate is 3mm, and severe is 4mm. in even more severe cases children may present in a "chin-up" position due to inability to see while looking straight.[10] While surgical intervention should be deferred to only when all non-surgical options are exhausted, in most cases of severe congenital ptosis, surgery of some kind is needed to prevent development of refractory errors and long-term vision defects. In addition, due to difficulty seeing, children tend to tilt their head backs in a "chin-up" position which if not corrected can also lead to orthopedic problems in the future.[14]

The particular need for surgical intervention arises due to the high chance of development visual defects with congenital ptosis of which the most commonly encountered are myopia, amblyopia, strabismus, anisometropia and refractory errors. A systematic review conducted to assess prevalence of these complications shows that among 24 studies conducted, 2589 individuals suffered from congenital ptosis, 30.2% developed myopia, 22.7% developed amblyopia and 17% developed anisometropia. The most alarming finding is that although treatments exist for congenital ptosis, children often present too late and complications have already developed by that time. Strabismus has a 1-5% prevalence in the general population while the meta-analysis revealed that prevalence of strabismus in congenital ptosis patients is a much higher 19.6%.[15] There is no mention of pre- and post-op comparison in the study.

Multiple surgical approaches are available to correct ptosis. The most widely used is the frontalis sling procedure which aims to form a connection

between the frontalis muscle and tarsus of upper eyelid.[10] In patients with good levator function, the levator resection procedure can be used which attempts resection and advancement of levator aponeurosis. This procedure has the additional advantage of allowing intraoperative adjustment of the amount of eyelid elevation.[16] Other procedure that can be used include the Whitnall sling and the Fasanella-Servat procedure. The Whitnall sling converts the action of the LPS from a horizontal to a vertical plane and provides greater upper eyelid support.[17] The Fasanella-Servat procedure involves the removal of up to 3mm of superior tarsus, conjunctiva, Muller muscle and the levator muscle. It is generally used in cases of mild ptosis with good levator function.[15] Our study compared the different procedures for ptosis correction and revealed that while all procedures improved post-op refractive errors; no particular procedure was superior to the other.

Our study showed a significant difference in pre and post op refractive errors. 0.25 Diopter correction was documented in 6(30%) cases, 0.50 Diopters in 4(20%) cases, 1 Diopter in 2(10%) cases and 9(45%) cases showed no difference in pre and post op refractive error correction. In a study comparing pre- and post-op refractive errors, a mean change in refractive error of 0.40 Diopters was observed.[18] Refractive error correction was maintained during a follow-up period of 12 months.[18] In another study assessing change in refractive error after unilateral levator resection for congenital ptosis, the mean change in refractive error from pre-operative measurements was a 1.23 Diopters decrease in sphere with a range of 0–3.5 Diopters.[19] In both of these studies, the change in refractive error was measured in all patients regardless of whether they had amblyopia or not and thus a difference in the refractive error correction compared to our study may be observed. A literature review of refractive error changes following ptosis surgery shows a mean change in refractive error of 0.40 Diopters, however the results were not statistically significant.[20]

Majority of patients had a post-op symmetry of <1.5mm regardless of the type of procedure conducted. While Unilateral Frontalis Sling procedure also produced >1.5mm post-op symmetry in 75% in our study, it is an extremely small sample size and thus significant conclusions are difficult to draw as to whether it is superior to other procedures or not. However, in a study comparing frontalis sling to levator resection, frontalis sling showed a significantly greater improvement in MRD-1 (4.46±2.19) compared to levator resection (1.85±2.5).[21]

LIMITATIONS OF STUDY

The sample size consisted of only 20 patients which may lead to a less than ideal power for the study. The condition is rare and so while a larger sam-ple size may be more ideal, patients presenting with simple unilateral congenital ptosis, without any ambly-opia or strabismus are infrequent. In addition, patients may have some undiagnosed underlying condition causing the ptosis and thus affecting long term results of the surgery. Prolonged assessment of patients with serial checkups might be helpful to assess for any such conditions.

CONCLUSION

Congenital blepharoptosis may present as a wide variety of refractive errors and upper lid blepharo-ptosis surgery repair have shown to improve the re-fractive error secondary to induced astigmatism. However, no particular surgery technique is found to be superior to other in our study sample and all surgical procedures had proven to be safe and effective approach towards blepharoptosis repair.

Conflict of Interest: None

Authors Contribution

Following authors have made substantial contributions to the manuscript as under:

TAK: & MS: Supervision, Conception, Study design, analysis and Interperitation of data, Critically reviewed manuscript & approval for the final version to be published.

SN: & AR: Co-supervision, Data entry, analysis and interpretation, manuscript writing & approval for the final version to be published.

TL: & MA: Critically reviewed, Drafted manuscript & approval for the final version to be published.

MAR: Data colllection, Entry and analysis of data, preparation of rough draft & approval for the final version to be published.

WN:, RJ:, ZBI:, UF: Data collection and entry & approval for the final version to be published.

Authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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