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# Outcomes of Early Correction of Congenital Myogenic Ptosis Using Transconjunctival Levator Plication

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**ABSTRACT**

**Introduction:** Early correction of congenital ptosis may be indicated due to a risk of amblyopia or because of an abnormal head tilt. One of the main problems, of planning ptosis surgery in very young children, is the inability to measure the levator function. **Aim:** The aim of the article was to analyze the early correction of congenital myogenic ptosis. **Methods:** This was a retrospective, interventional, case series study, conducted on 12 eyes of 12 patients with unilateral, mild to moderate, congenital myogenic ptosis. Surgical correction of ptosis was performed by transconjunctival levator muscle plication. Pre- and postoperative measurements of the upper lid margin to central corneal reflex (MRD1) and upper lid skin crease height (UEC) were obtained, as well as the presence or absence of a reaction to topically applied phenylephrine 2.5% solution. **Results:** The mean age of the patients was 29.83 months (range 14-45 months). A negative phenylephrine test was noted in only 3 (25%) of cases. Equalization of upper lid height was achieved in 6 (50%), and a hypocorrection of up to 1 mm was noted in 4 (33%) of patients. There was only 1 hypercorrection of 1 mm, noted in the first postoperative month. In one case of hypocorrection of 2 mm, the height of the lid dropped between the 1 and 3 months follow up. Subsequent revision surgery was performed, with a good outcome. With regard to the upper lid skin crease height (UEC), the mean preoperative difference in relation to the contralateral (non-operated) lid, was 2.16 mm, whereas the average postoperative or final difference was 0.41 mm. **Conclusion:** Correction of myogenic ptosis in small children, using transconjunctival levator plication, in whom levator function cannot be measured, may have a satisfactory postoperative outcome.

**Keywords:** Blepharoptosis congenital, blepharoptosis/surgery, eyelids surgery

**1. INTRODUCTION**

Congenital ptosis is a condition in which the upper eyelid is in a position that is lower than normal and becomes apparent at birth or during the first year of life (1). It may be present on one or both eyes. Congenital myogenic ptosis is a consequence of underdevelopment or dysgenesis of the levator muscle fibers, which are substituted by adipose or fibrous tissue, thus decreasing the ability of the levator muscle to contract (2). There are also proposals that the fatty infiltration of the levator muscle could be a degenerative process (3).

Congenital ptosis can induce visual problems such as myopia, astigmatism, anisometropia, amblyopia, torticollis and strabismus. Although there is no doubt that complete obstruction of the pupillary axis, in severe congenital ptosis, is an indication for early correction (4), there is lack of consensus relating to the timing of surgery in mild to moderate ptosis, though there is evidence to suggest that even children with mild congenital ptosis may be at risk of developing anisometropic and strabismic amblyopia (5).

One of the main problems of planning an operation of congenital myogenic ptosis, in very young children, is the inability to reliably measure the levator function (LF), which is one of the parameters necessary in the preoperative calculation of the amount of levator resection (6). Other parameters such as the margin to reflex distance (MRD1) and upper lid skin crease height (UEC) can be indirectly measured from a photograph, taken when the patient is looking straight ahead. This is usually possible, even with the most uncooperative of small children.

Intraoperative adjustment of the upper lid height, based on the preoperative measurements of levator function, has been described by Berke (7), to aid in a more precise achievement of the desired postoperative eyelid position.

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Many methods of congenital ptosis correction have been in use, but one could classify all of them into anterior or posterior, the former being done transcutaneously and the latter transconjunctivally. Depending on the tissues which are manipulated, in order to correct the congenital ptosis, they can be classified into Mullers muscle resection, levator resection/plication and frontalis suspension techniques.

Recently, a minimally invasive, posterior or transconjunctival approach levatorpexy for congenital ptosis, has been described in mild and moderate cases (8) and subsequently, also, for ptosis with a poor levator function (16).

## 2. AIM

The aim of the article was to analyze the outcome of transconjunctival levator plication in the early correction of congenital myogenic ptosis.

## 3. METHODS

### Preoperative considerations and examinations

This was a retrospective, interventional, case series study, conducted on 12 eyes of 12 patients with unilateral, mild to moderate, congenital myogenic ptosis. All the patients were operated in two centers, using transconjunctival levator plication, by a single surgeon, in the period June 2018-December 2019. Only patients between the age of 1-4 years (12-48 months), in whom the levator function (LF) could not be reliably measured, were included in the study.

All the patients underwent complete ophthalmological examination by a pediatric ophthalmologist (LM), who referred them for ptosis correction. The indication was based on the orthoptic findings, which were suggestive of impending amblyopia, such as loss of fixation by the eye on the ptotic side, in comparison to the contralateral eye, during an induced tropia test. Another indication was an abnormal upward head tilt, or torticollis, which could cause functional and structural damage to the cervical spine.

Taking into consideration the lack of cooperation on the part of the very young child, the preoperative evaluation included making a digital photograph (Figure 1a), which was the basis for the measurement of the preoperative upper lid margin to central corneal reflex (POMRD1) parameter. The second parameter was the phenylephrine test (PE test). This test was performed by instilling one drop of 2.5% solution into the eye on the affected side. After 10 minutes another digital photograph was taken to evaluate the result (Figure 1b).

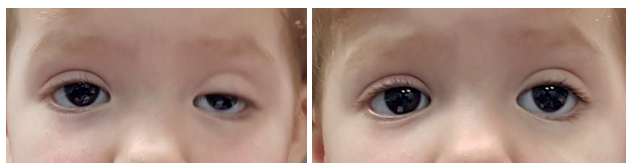


Figure 1. Phenylephrine test: a) Ptosis on the left side prior to instillation of 2.5% phenylephrine; b) Elevation of ptotic lid 10 minutes into the test

The result of the PE test was graded as positive or negative, the former meaning any visible elevation in the position of the ptotic lid, and the latter meaning complete absence of response.

### Surgical technique

All operations were performed in general anesthesia.

The surgical technique was based on the one originally described by All-Abadi and co-workers (8), with some modifications, which are described in the following text. The first author has had the opportunity to learn this technique, during his visit to Dr. Raman Malhotra in 2018.

The existing or intended upper eyelid crease was marked by a skin marker dot on the skin, in vertical alignment with the lowest point on the eyelid margin, with the eye closed. The upper lid was everted using a traction suture and a Desmarres retractor. The upper fornical conjunctiva was infiltrated with an injection of 0.5 ml solution lidocaine 2% and adrenaline 1/100 000, in order to reduce bleeding and the amount of general anesthetic use. Bipolar diathermy and epithelial debridement was performed at the upper tarsal border/fornical conjunctiva junction, where an incision was made. Conjunctiva and Mullers muscle were dissected from the posterior surface of the levator aponeurosis, up to and beyond the junction of the aponeurosis with the levator muscle (Figure 2a). Bipolar diathermy was used in hemostasis. A 5-0 polyglactic acid (PGA) suture (Vicryl<sup>®</sup>, Ethicon<sup>®</sup>), on a ¼ circle spatulated needle, was passed, tangentially, through the upper border of the tarsus, at the highest point of curvature.



Figure 2. Surgical technique: a) Dissection of the conjunctiva and Mullers muscle from the posterior surface of the levator aponeurosis; b) Placement of the suture through the junction of the aponeurosis and levator muscle

Then, one arm of the suture was passed, with a fore-hand motion, partial thickness, through the junction of the levator aponeurosis and levator muscle and tied with a temporary suture (Figure 2b). The intraoperative upper lid height was measured in the following manner: with the main operating light turned away from the operating field and only the surgeon headlight illuminating the eye, the surgeon uses a caliper to measure the distance from the central corneal reflex to the upper lid margin. The measured distance was the intraoperative MRD1 (IOMRD1). The aim was to set the upper lid margin at a certain height, based on a modified nomogram (Table 1). This was done by adjustment of the height of the suture, which, in most cases, meant that the height of the suture needed to be increased. In case the preoperative phenylephrine test was negative, the aimed

POMRD1	IOMRD1
0	3
1	2
2	1
3	0

Table 1. Intraoperative adjustment of eyelid height. Preoperative eyelid height (POMRD1), intraoperative upper lid position (IOMRD1), in millimeters.

IOMRD1 was increased by 1 mm. After achieving the desired IOMRD1, the second arm of the suture was passed 1 mm above the first one, in a backhand motion and then both arms of the sutures were tied internally with a throw sequence of 2-2-1. Next, both arms of the suture were externalized through the marked dot on the skin and tied with a throw sequence of 1-1-1.

Antibiotic ointment was applied to the eye surface and the eye was padded with the cotton gauze. A temporary non-resorbable Frost suture, passed through the lower lid and taped to the upper lid, was used if necessary.

The minimum follow up regimen was on the first day after surgery, after that, postoperatively one week, one month and three months.

The postoperative regimen consisted of tobramycin/dexamethasone eye drops, applied 4x daily, for two weeks. The absorbable 5-0 PGA suture was left to fall out spontaneously.

**Postoperative evaluation and follow up**

The preoperative photographs, as well as from the three months follow up, were examined, in order to determine the pre- and postoperative parameters. The measurements were based on the average horizontal corneal diameter in this age group, which is 11.7 mm (9), and, in this study, was approximated at 12 mm. The digital photograph of the eye was enlarged, on the computer screen, so that the horizontal diameter could be used as a ruler scale, for the pre- and postoperative measurement of MRD1 and UEC.

**4. RESULTS**

Data relevant to the study group is presented in Table 2. Representative pre- and postoperative photos are presented in Figures 3 and 4.

The mean age of the patients was was 29.83 months (range 14-45 months). The gender structure was 7 males and 5 females. The mean follow up period was 7 months (range 3-12 months).

A negative phenylephrine test was noted in only 3 (25%) of all cases.

The preoperative MRD1 (POMRD1) range was 0-3 mm, the average value being 1.58 mm. The mean postoperative or final MRD1 (FMRD1) was 3.41 mm (range 2-5 mm). Equalization of eyelid height, with the contralateral (non operated) upper lid, was achieved in 6 (50%), and a hypocorrection of up to 1 mm was noted in 4 (33%)

of patients. There was only 1 hypercorrection of 1 mm, noted in the first postoperative month (patient No.7). In this case the parents were instructed to massage the eyelid, in the early postoperative period, which resulted in equalization of upper lid height at the final follow up. In one case of hypocorrection of 2 mm (patient No.5), the height of the lid dropped between the 1 and 3 months

Pt. No.	Gender (M/F)	Age (months)	POMRD1 (mm)	FMRD1 (mm)	IEMRD1 diff. (mm)	POUEC diff. (mm)	FUEC diff. (mm)	PE test (+/-)
1	M	22	0	2	-1	2	1	-
2	M	34	2	4	0	3	0	+
3	F	14	1	3	0	2	0	+
4	M	19	1	3	-1	3	0	+
5	F	25	0	2	-2	4	2	+
6	F	41	3	4	0	1	0	-
7	M	37	2	5 (4)	+1 (0)	2	1	+
8	F	26	3	4	0	1	0	-
9	M	25	0	3	-1	3	0	+
10	M	37	3	4	0	1	0	+
11	F	33	1	3	-1	3	1	+
12	M	45	3	4	0	1	0	+

Table 2. Data relevant to the study group. Preoperative MRD1 (POMRD1), final MRD1 (FMRD1), intereye MRD1 difference (IEMRD1 diff.), preoperative UEC (POUEC), final UEC (FUEC), phenylephrine test (PE test)



Figure 3. Child with a left side ptosis: a) Preoperative photo. Note: the child is agitated and crying, even with photography alone; b) Postoperative result.



Figure 4. Child with a right side ptosis: a) Preoperative; b) Postoperative result.

follow up. Subsequent revision surgery was performed, with a good outcome. With regard to the upper lid skin crease height (UEC), the mean preoperative difference in relation to the contralateral (non-operated) lid, was 2.16 mm, whereas the average postoperative or final difference was 0.41 mm.

**5. DISCUSSION**

In the study concerning the effect of surgical correction of congenital ptosis on amblyopia, Lin et al. found that all of the amblyopia cases were on the ptotic side and in case of bilateral ptosis, on the more ptotic side (10). Their study included patients aged 2 months to 17 years, whereas 32% of the study group were aged 1-4 years.

Quaranta-Leoni and coworkers have found that the functional and cosmetic results of levator resection, in

congenital ptosis, is more favorable if the surgery is done before the age of 4 years (11). They have also found an increase in postoperative levator function, which was more pronounced in the age group of 2-4 years. However, children below the age of 3 years, in whom the levator function could not be measured, were excluded from the study.

Kang et al. established a statistically significant negative correlation between the preoperative MRD and LF, and the amount of intraoperative lagophthalmos, in levator resection for congenital ptosis (12). They have concluded that taking into account both preoperative variables, rather than any one of them alone, results in a more favorable surgical outcome.

The proper and reliable measurement of the levator function requires good cooperation on the part of the patient. This parameter is measured by fixating the eyebrow, to eliminate frontalis muscle action, by the thumb of the examiners one hand, and asking the patient to look up and down (or vice versa), at the same time measuring the excursion of the eyelid margin against a ruler, which is held immobile by the other hand, resting on the forehead of the patient (13). During this procedure, the movement of the head is discouraged. However, in small children, the measurement of the levator function is difficult and unreliable. Based on the authors personal experience, as well as on anecdotal information from colleagues in the field, at best, it is an estimation of the eyelid excursion, when the child is motivated to follow an object or light, from the lowest to the highest point in its visual field, or vice versa. Fixation of the eyebrow and the head is very difficult and, together with the placement of a ruler near the eye, elicits an instinctive defensive reaction from the child, who may be agitated just by seeing a medical practitioner (Figure 5).

An indirect sign, which might indicate the amount of levator muscle strength, is the presence of a well defined upper lid skin crease, but this can not be quantified. The Illif test has been described, for the estimation of levator function in small children (14). The test requires that the examiner evert the upper lid, as the child looks down. If the levator function is good, the eyelid will revert to its normal position spontaneously. Apart from being hard to perform, this test, also, has no quantitative value.

Khandwalla et al. have published a method of measuring LF in children with congenital ptosis, using spectacles that have a millimeter scale on each of the lenses (15). The age range was 1-12 years. The measurement method involved holding steady the child's head and fixating the eyebrow by the clinicians thumb, the problems of which have already been elaborated.

Based on the facts outlined above, it is clear that in a certain percentage of small children, in whom a ptosis correction is performed, this is done without a dependable knowledge of the levator function. Based on our experience, this may be the case in children up to the age of 4 years, although, admittedly, there are children younger than this who may be cooperative and older children who are not. In such cases, in which the levator function is not quantified, but is rather a subjective estimate on

the part of the surgeon, the only measurable parameters are the MRD and UEC. The aim of this study was to, exclusively, target this group of patients, knowing that, in practice, they present the biggest challenge to ptosis surgeons.

The phenylephrine test has a certain value in the evaluation of congenital ptosis in a small child. This substance, applied in the form of an eyedrop, as a 2.5% solution, stimulates the adrenergic receptors of the Mullers muscle. A fully positive response can elevate the upper lid from 0.5 mm to more than 2 mm (15). A negative response can be attributed to the gross fatty infiltration of Mullers muscle and its receptors (16). One aspect of the practical use of this test is the demonstration of a potential effect of ptosis correction to the parents of the child. The other aspect is based on the Herings law test, which could help to reveal contralateral latent ptosis. Both aspects rely on a positive response to phenylephrine. Another problem is that most small children start crying when an eyedrop is instilled, which could result in a significant dilution of the substance and consequent spillage over the lower lid skin. One way of verifying that phenylephrine has been sufficiently absorbed is pupil dilation, however, the meantime necessary for this to occur is about 29 minutes in dark irides (17), which is impractical, because the recommended time for the evaluation of upper lid elevation is 10 minutes. Even though it is unclear how a response to phenylephrine correlates to the effectiveness of transconjunctival levator plication, it probably makes sense to take it into account, because the Mullers muscle is advanced at the same time, not excised as in the procedures of Mullers muscle conjunctival resection.

The nomogram we used in our study, to adjust the intraoperative eyelid height, was based on the empirical knowledge that, in congenital myogenic ptosis, the MRD1 is in correlation with LF and that it is highly unlikely that a more ptotic upper lid will have a good levator function, and vice versa, an eyelid with mild ptosis will have a poor levator function. This has been confirmed by Ural et al. who suggest that, in the case of an unknown LF, preoperative MRD1 can be used to select the appropriate type of surgery for congenital ptosis (18).

Our postoperative results are comparable to other studies that have utilized the same surgical technique of posterior levator plication, tucking or levatorpexy (8, 16, 19). The advantages of this type of ptosis operation are the absence of anterior and middle lamella scarring and the fact that no tissue is excised during the procedure. However, the use of only one resorbable suture, for the plication of the levator muscle, may be a cause of a postoperative eyelid height drop, as was the case in one of our patients, who had to be reoperated.

## 6. CONCLUSION

Correction of myogenic ptosis in very young children, using transconjunctival levator plication, in whom levator function cannot be measured, may have a satisfactory postoperative outcome.

- **Author's contribution:** Z.Z. and M.L. gave a substantial contribution to the conception and design of the work and to the acquisition of data. Z.Z. and R.K. gave a substantial contribution to the analysis, interpretation of data and drafting the article. Z.Z, R.K. and M.S. gave substantial contribution to revising it critically for important intellectual content. All authors gave final approval of the version to be published and agreed 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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