Outcome of Silicone Sling Frontalis Suspension in Isolated Uncomplicated Congenital Ptosis vs. Complicated Ptosis

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Article

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Abstract

Objectives

To compare the outcome of silicone sling frontalis suspension (FS) surgery in children with isolated uncomplicated congenital ptosis (IUCP) vs. children with complicated ptosis.

Methods

A retrospective medical chart review of all pediatric patients who underwent silicone sling FS surgery between 2009–2020 at a single center were included. Patients were divided according to ptosis type: IUCP vs. complicated ptosis. Pre-and post-operative margin-to-reflex distance (MRD<sub>1</sub>) measurements were determined from clinical photographs. Main outcome measures were differences in improvement in eyelid height, reoperation rate, and timing between the groups.

Results

Two-hundred and eight children were included: 139 IUCP and 69 complicated cases, 83 females (40%), mean (± SD) age at intervention was 1.9 ± 2.9 years. Complicated cases included: blepharophimosis epicanthus inversus syndrome (n = 35), Marcus Gunn jaw winking (n = 12), oculomotor palsy (n = 8), congenital fibrosis of extraocular muscles (n = 3), chronic progressive external ophthalmoplegia (n = 3), and others. Mean MRD1 improved by an average of 1.6 mm in both groups. Repeat ptosis repair was performed in 50/171 (29%) patients without a history of failed ptosis procedures, and this rate was similar between IUCP and complicated cases. Children under 3 years of age had higher rates of repeat ptosis repair than older children (n = 59/175, 34% vs n = 5/33, 15%, p = 0.03 chi²).

Conclusions

Silicone sling frontalis suspension has a favorable outcome in 70% of pediatric patients. Preoperative and final MRD<sub>1</sub> and reoperation rates were similar between both groups, suggesting that despite the higher complexity in atypical cases, the outcome is similar.

Introduction

Congenital ptosis is usually isolated to the eyelid, and not associated with other ocular, adnexal, and/or systemic disorders. It is usually sporadic, without a known genetic variant or family history. It is caused by abnormal embryonic development of the levator palpebrae muscle, which is commonly dystrophic, with fatty and fibrotic changes replacing healthy muscle fibers. Although the congenital isolated myogenic type is most common, other more complex ptosis types can also be observed in children, more
commonly associated with other systemic and genetic disorders. These include myogenic, aponeurotic, neurogenic, and mechanical, types.

As the surgical management of isolated uncomplicated congenital ptosis (IUCP) is well established, the repair of complex ptosis cases it is often perceived as more challenging. This may be due to abnormal anatomical/neurological structures or otherwise related to factors associated with the general medical situation. Successful outcomes from both the cosmetic and functional perspective are also intriguing, as for some cases, an under-correction is the intended goal, such as for corneal exposure issues from a reduced Bell's phenomenon as seen with CN3 palsy or CFEOM. Despite the various types of complex ptosis and the importance of successful management in preventing amblyopia, peer-reviewed literature on this subject is sparse.

The purpose of this study was to compare the surgical outcomes of frontalis suspension in IUCP vs. pediatric complicated ptosis using a silicone sling for frontalis suspension. We evaluated the re-operation rate and timing between the initial and repeat procedures.

**Methods**

This was a retrospective chart review of all pediatric ptosis repairs treated at The Children’s Hospital of Philadelphia (CHOP) with silicone sling frontalis suspensions (FS) over 12 years (2009–2020). Isolated uncomplicated congenital ptosis (IUCP) as well as other complicated cases were included. Complicated ptosis types included: Blepharophimosis, ptosis, and epicanthus inversus syndrome (BPES), paralytic ptosis, congenital fibrosis of the extraocular muscles (CFEOM), Marcus Gunn jaw wink, chronic progressive external ophthalmoplegia (CPEO) including Kearns-Sayre syndrome, and mechanical ptosis. In repeat FS procedures, only the first procedure was included for each patient.

**MRD1 measurements**

Pre- and post-operative MRD\textsubscript{1} measurements were determined from clinical photographs using ImageJ software (nih.gov).

The study adhered to the tenets of the Declaration of Helsinki and was HIPAA-compliant, with CHOP Institutional Review Board (IRB) approval. Cases without a second ptosis repair were defined as successful or having a favorable outcome.

**Statistical Analysis**

The Chi-square and binary logistic regression tests were used to calculate proportional differences between categorical groups. Paired samples t-test was used to compare pre- and postoperative MRD\textsubscript{1} measurements. Independent samples t-test and ANOVA were used to compare other continuous
variables. Kaplan-Meier survival analysis was used to estimate the cumulative of re-operation over time. In bilateral cases, the right eye was arbitrarily selected. Statistical analysis was carried out using SPSS (version 26, SPSS Inc., Chicago, IL). All results are presented as mean ± standard deviation.

Results

Two-hundreds and eight children were included: 139 IUCP and 69 complicated cases, 83 females (40%), mean (± SD) age at intervention was 1.9 ± 2.9 years. Complicated cases included: blepharophimosis epicanthus inversus syndrome (BPES, n = 35) (Fig. 1), Marcus Gunn jaw winking syndrome (MGJW, n = 12), oculomotor palsy (n = 8), congenital fibrosis of extraocular muscles (CFEOM, n = 3) (Fig. 2), chronic progressive external ophthalmoplegia (CPEO, n = 3 including one case of Kearns Sayre), and single cases of eyelid hemangioma (PHACES), s/p rhabdomyosarcoma, Treacher Collins with left eyelid coloboma, traumatic, monocular elevation paresis, severe developmental hypoplasia of the extraocular muscles, s/p enucleation.

A history of previous ptosis repair surgery at presentation was more common in the complicated group (n = 21, 30% vs. n = 16, 11% respectively, p = 0.002, χ²), and children with complicated ptosis were operated on at an older age: 3.1 vs. 1.4 years (P = 0.002, independent samples t-test). 74 (36%) children underwent bilateral surgery, and this was more common in complicated cases (25% vs. 56%, p < 0.001, χ²). Mean MRD1 improved by an average of 1.6 mm in both groups. Most parents in both groups (70%) were satisfied with the surgical outcome. Repeat ptosis repair was performed in 50/171 (29%) patients without a history of failed ptosis procedure, and this rate was similar between IUCP and complicated cases (p = 0.8, independent samples t-test).

Overall, repeat ptosis repair was performed in 64 (31%) patients, 31.2 ± 20.9 months after the initial procedure. Children under 3 years of age had higher rates of repeat ptosis repair than older children (n = 59/175, 34% vs n = 5/33, 15%, p = 0.03 chi²).

Outcome by ptosis type

Surgical intervention outcomes – stratified by complicated ptosis groups vs. isolated uncomplicated congenital ptosis are detailed in Table 1. Overall, Cases of BPES had similar pre & postoperative MRD, similar intervention rates & timing. Cases of MGJW had similar pre & postoperative MRD, however a higher intervention rate (50%) - although not statistically significant - and a shorter period until the repeated procedure (13.3 vs. 32.8 months, p = 0.002, independent samples t-test) (Fig. 3). Cases of CN3 palsy had lower preoperative MRD1 (-0.7 vs. 0.2 mm, p = 0.03, independent samples t-test), and cases of CFEOM had higher postoperative MRD1 (2.8 vs. 1.8 mm, p = 0.007, independent samples t-test).

Discussion
This study compared surgical outcomes of frontalis suspension silicone slings for pediatric ptosis between simple and complex cases. Children with complicated ptosis were operated on at an older age compared with IUCP, and had higher rates of bilateral intervention. The surgery resulted in a favorable outcome, with a similar improvement of MRD1, and a similar re-operation rate of 29%. Children over 3 years of age had lower rates of repeat ptosis repair (15%).

While IUCP is the most common pediatric ptosis type, other more complex ptosis types can also be observed in children. The latter are more commonly associated with other systemic and genetic disorders. These include myogenic, aponeurotic, neurogenic, and mechanical types. Myogenic ptosis types include the blepharophimosis syndrome (BPES), congenital fibrosis of the extraocular muscles (CFEOM), muscular dystrophies, chronic progressive external ophthalmoplegia (CPEO), and myotonic dystrophy. Thyroid eye disease, while usually associated with eyelid retraction, can rarely present as a myogenic ptosis due to inflammatory infiltration of the eyelid retractors. Eyelid trauma such as forceps injury during birth can cause aponeurotic ptosis, usually accompanied by a higher-than-normal lid crease.

Neurogenic ptosis can develop secondary to defects in innervation occurring during embryonic development or acquired later in life. Examples include third nerve palsy or paresis, and congenital Horner syndrome due to abnormalities involving the sympathetic innervation. Abnormal or synkinetic connections can occur between the third and fifth cranial nerves, producing a Marcus Gunn jaw-winking type of ptosis. Although not truly a neurogenic cause, myasthenia gravis can also lead to ptosis in the pediatric age group, which may manifest with fatigue and with extraocular muscle function disorders.

Mechanical ptosis is most commonly caused by the inability of the levator muscle to function properly due to the weight of a tumor mass, soft tissue swelling, or foreign body. Examples include plexiform neurofibromas and infantile hemangiomas, both notorious for infiltrating the levator muscle complex and making simple excision difficult. Cicatricial processes secondary to trauma can also mechanically restrict the eyelid in a ptotic position even with an otherwise normally functioning levator.

Each of these rare ptosis types may present a challenge to the oculofacial surgeon. In addition, the appreciation of surgical outcomes is not always straightforward, as perfect symmetry and contour are not always achievable. Patients, as well as physicians’ expectations, may be altered in complex cases. Previous results on ptosis repair in complicated slings mainly include short case series, including BPES, CFEOM, CPEO, and others, with each entity enclosing its unique challenges (Fig. 4). For instance, in BPES simultaneous or successive correction of the epicanthus inversus may affect the eyelid position and contour. In various mechanical ptosis cases, abnormalities of the eyelid and periorbital soft tissue are a caveat for surgical success. Upgaze limitation with deficient Bell’s phenomenon, as observed in neurogenic, CPEO, or CFEOM cases, mandates under-correction to prevent inadvertent exposure keratopathy. The use of an adjustable silicone FS sleeve can also be considered but is somewhat less appropriate for children, in whom the adjustment itself requires general anesthesia. This planned under-correction should be discussed with the parents in advance to prevent unmet expectations. In upgaze limitation, we usually desire an MRD1 of 0-1mm (Fig. 5).
An interesting finding demonstrated in this series is the higher reoperation rate under the age of 3 years; this is in concordance with our clinical experience. We believe that the rapid growth of the cranium in the first years of life is a major cause of the surgical failures observed in the young population, as the slings can more easily break or cheese-wire through the lid tissues.\textsuperscript{14} It is advised to discuss this with the parent of infants, as there is a probable need for a repeated repair by age 4–5 years. At this age, the surgeon may consider a different, potentially more permanent repair such as autogenous fascia lata suspension.

In conclusion, silicone sling frontalis suspension has a favorable outcome in 70\% of pediatric patients and this includes both IUCP and complicated cases. This may encourage surgeons to address complex cases with higher confidence. Preoperative and final MRD, reoperation rate, and parental satisfaction were similar between both groups, suggesting that despite the higher complexity in atypical cases, the outcome is similar. These encouraging findings support the use of FS in complicated cases.

\textbf{Declarations}

\textbf{Financial Support}: None.

\textbf{Conflict of Interest}: All authors report no conflicts of interest.

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\textbf{References}


Table 1

Table 1 is available in the Supplementary Files section.

Figures
Figure 1

A 4-year-old with blepharophimosis epicanthus inversus syndrome (BPES) (A) underwent bilateral double triangle silicone slings frontalis suspension and a bilateral medial canthopexy/plasty (B, 6 days postoperatively) with good postoperative result (C, 5 months postoperatively).
A 1-year-old with congenital fibrosis of extraocular muscles (CFEOM) and severe bilateral ptosis with no Bell’s phenomenon (A). The patient underwent bilateral double-triangle frontalis suspension silicone slings with simultaneous inferior rectus recession with good postoperative result at 4 months postoperatively (B).

Figure 2
Kaplan-Meier curve demonstrating cumulative survival of 13 pediatric subjects with MGJW vs. 139 cases of myogenic ptosis who underwent ptosis repair using frontalis suspension silicone slings. Event was defined as a repeated procedure. The results demonstrated improved survival in the non-MGJW group (p=0.001, Log-Rank Kaplan-Meier). The difference between total rates of repeated procedures between both groups did not reach significance (34% non-MGJW vs. 50% MGJW group, p>0.05, chi-square). The average time to intervention was shorter in the MGJW group (13 vs. 33 months, p=0.002, independent samples t-test).
A 1-month-old with blepharophimosis epicanthus inversus syndrome (BPES) (A) underwent bilateral double triangle silicone slings frontalis suspension with good postoperative result (B, 3 months postoperatively). The ptosis gradually recurred (C, age 3.5 years), and the patient underwent repeated bilateral frontalis suspension using autogenous fascia lata with a bilateral medial canthopexy/plasty at age 4 years (D, 2 months postoperatively). Note the evolving bilateral euryblepharon and lateral canthal
dystopia with the lower lid longer than the upper lid. Correction of this deformity can be planned as a future step.

Figure 5

Pre- and postoperative images of a child with left cranial nerve III palsy and deficient Bells’ phenomenon due to upgaze limitation. In these cases, under-correction is recommended to prevent exposure.

Supplementary Files

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- Table1complexslings7102022.docx