Improved Visual Acuity after Frontalis Sling Surgery for Simple Congenital Ptosis

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ABSTRACT

INTRODUCTION Congenital ptosis is malpositioning of the eyelids that, when moderate or severe, can negatively affect visual development during its critical period, resulting in amblyopia: diminished visual acuity with no apparent organic cause. Early diagnosis and timely treatment are essential for preventing amblyopia. Congenital ptosis is uncommon but poses a challenge to any ophthalmologist; the only treatment is surgical. Among these patients in Cuba, those with the most complex clinical characteristics are generally referred to the Ramón Pando Ferrer Ophthalmology Institute in Havana.

OBJECTIVE Characterize visual acuity outcomes obtained in patients seen at this Institute who received surgery for simple congenital ptosis using the frontalis sling procedure.

METHOD A descriptive prospective longitudinal study was conducted to describe visual acuity outcomes in 11 patients with a diagnosis of isolated congenital ptosis seen in the Oculoplastic Service of the Ramón Pando Ferrer Ophthalmology Institute between January and July 2009 and operated on using the frontalis sling procedure. The majority exhibited severe visual acuity impairment (0.1–0.5) prior to surgery. Variables employed were age, sex, degree of ptosis, degree of ptosis correction, visual acuity, and complications during surgery and postoperatively.

RESULTS Male patients aged 1–4 years predominated. Visual acuity improved in 100% of patients, to varying degrees. Prior to surgery, 72% had visual acuity of 0.1–0.5. Six months post-surgery, with visual rehabilitation, 90.9% exhibited visual acuity of >0.5. In 81.8% of patients, palpebral ptosis was fully corrected. Complications were minimal: injury to the palpebral tarsus and undercorrection were the most common and did not affect final surgical outcome or interfere with rehabilitation.

CONCLUSIONS Correction of congenital ptosis using the frontalis sling technique yielded satisfactory visual acuity outcomes, contributing to visual rehabilitation of the affected patients.

KEYWORDS Blepharoptosis/congenital, blepharoplasty, frontalis sling, amblyopia, Cuba

INTRODUCTION

Palpebral ptosis or blepharoptosis is descent of the free border of the upper eyelid below its normal position, or involuntary drooping of the upper eyelid when the person is actively focusing on a fixed point, resulting in a narrowing of the palpebral fissure, smoothing of the eyelid and potential disappearance of the palpebral fold. It can be congenital or acquired; unilateral or bilateral; constant or intermittent; associated with a localized condition or systemic disease. It can produce functional limitations, aberrations in neck or body posture and aesthetic and psychological alterations. It is a condition well known in the fields of plastic surgery and ophthalmology. However, its complex etiology and difficult (and sometimes frustrating) treatment cause many professionals in the field to ignore it.[1]

There are many classifications for ptosis, anatomical and etiological. Etiologically, ptosis is classified as: myogenic, aponeurotic, neurogenic, mechanical, traumatic and pseudoptosis.[2,3]

Myogenic ptosis can be associated with weakness of the superior rectus muscle, present in blepharophimosis syndrome, chronic progressive external ophthalmoplegia, oculopharyngeal syndrome, progressive muscular dystrophy, myasthenia gravis, congenital fibrosis of the extraocular muscles, myotonic dystrophy and the simple or isolated congenital form—the latter accounting for some 50–60% of cases.[1]

Simple or isolated congenital ptosis is present from birth, other than resulting from birth trauma. It is an inherited dominant, recessive, or multifactorial autosomal condition involving a defect in the development of the levator muscle, generally unilateral (Figure 1). If bilateral (Figure 2), the patient tends to compensate by elevating the chin and looking downward, which can lead to abnormal head and neck posture at whatever age the condition appears, as well as significant aesthetic impact.[1,3]

It is important to correct congenital ptosis, as it can cause amblyopia, which, depending on its etiology, can be: strabismic due to loss of eye parallelism; isoametropic due to bilateral refractive

Figure 1: Unilateral ptosis.

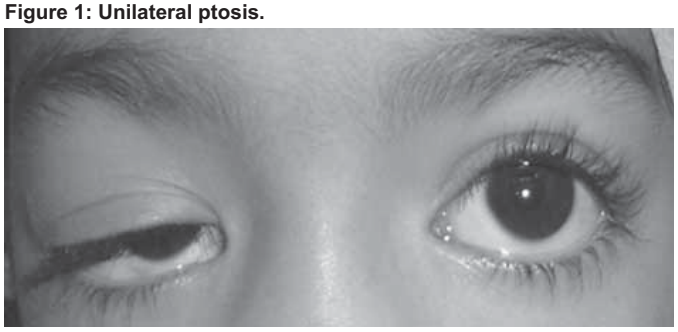
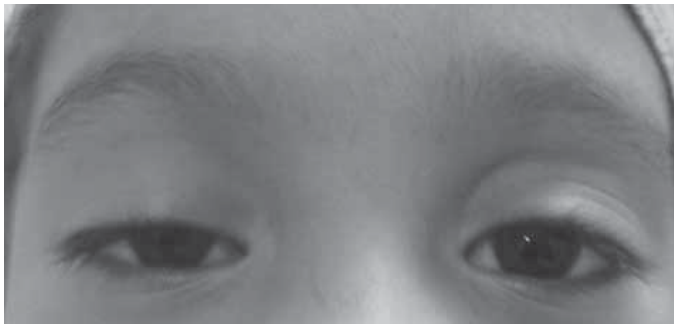


Figure 2: Bilateral ptosis, greater in the right eye.



defects; anisometropic due to unequal refraction defect of ≥1.50 D (diopters) and stimulus deprivation, secondary to an obstruction in the anterior visual pathway, such as the object of this study, palpebral ptosis.[4,5] Amblyopia from stimulus deprivation is rare and it is hard to find accurate estimates of its prevalence; it probably accounts for less than 3% of all amblyopia cases.[6]

Amblyopia is unilateral, or less commonly, bilateral reduction of the best corrected visual acuity (VA) not directly attributable to structural damage of the eye or posterior visual pathway. It occurs during the critical postnatal period in which the visual cortex remains labile enough to adapt to experiential or environmental influences. This sensory plasticity is at its height during the first 2 years of life, but there is potential for change, although to a lesser extent, up to 7 or 8 years, making visual rehabilitation possible.[7] Amblyopia is the primary cause of monocular vision loss in people aged 20–70 years, surpassing diabetic retinopathy, glaucoma, age-related macular degeneration and cataracts.[8]

Since 80% of learning in school before age 12 depends on vision and two out of every three school failures are attributable to visual impairment, early rehabilitation is vital.[8,9] Amblyopia is a preventable cause of visual deficit in a population that could be economically active in society, with a prevalence of 2–4% in the general population, 3–4% in preschool children and 2–7% in school-age children.[9,10]

Moderate or severe ptosis requires surgery.[1] Severe blepharoptosis, associated with poor levator function, is correctable with the frontalis sling procedure, described in 1909 by Payer, who used fascia lata;[1] in 1992, Wright made significant improvements to the procedure. Other materials have been used in frontalis sling surgery for ptosis—for example, sclera, silicone, polyfilament sutures, polyester, carbon and umbilical vein. In 1965, Gutman began using fascia lata irradiated with cobalt gamma rays.[1,11] Downes and Collin described the use of nonabsorbent synthetic materials for the first time in 1989.[1]

There are various techniques today for correcting moderate and severe ptosis in patients with good levator function, among them: the Fasanella-Servat procedure, levator resection, reinforcement of the aponeurosis and other techniques involving the upper eyelid levator aponeurosis. When levator muscle function is poor or nil, as was the case in all our patients with simple congenital ptosis, the frontalis sling procedure is indicated.[1]

The Ramón Pando Ferrer Ophthalmology Institute (ICO, its Spanish acronym) is a tertiary care institution within Cuba's public health system. It is a national referral center for treatment of this and other eye pathologies, receiving patients from all over the country.[12] While simple congenital ptosis is uncommon, patients seen at ICO are generally those whose condition could not be successfully treated at the secondary care level.

This study's objective was to characterize visual acuity outcomes in patients after frontalis sling surgery for simple congenital ptosis.

METHODS

A descriptive prospective longitudinal study was conducted to characterize VA outcomes in ICO patients undergoing frontalis sling surgery for simple congenital ptosis. These patients were seen in the ICO's Oculoplastic Service between January and July 2009; they came from various provinces in Cuba, referred to tertiary care because their condition could not be treated at the secondary level.

The study population consisted of 11 patients who underwent frontalis sling surgery for congenital ptosis, having met the following criteria:

Inclusion criteria Patients aged >1 year and <10 years, with moderate or severe simple congenital ptosis.

Exclusion criteria Patients with prior palpebral diseases: blepharitis, trichiasis, entropion, symblepharon; with ophthalmologic problems: corneal scarring, dry eyes, glaucoma and retinal detachment; with uncontrolled chronic diseases. Patients whose parents refused consent.

Ethical considerations Parents of patients meeting inclusion criteria received an explanation of the surgical procedure, its objectives and potential complications. Their written consent was obtained for their children's participation in this research and to permit publication of data and photographs, maintaining subject anonymity. The study was approved by the ICO Ethics Committee.

Procedures Assessment of surgical patients was based on the following parameters:

Visual Acuity, using visual preference and optokinetic nystagmus in preverbal children. In verbal children, depending on their age, Kay pictures (≥3 years) and the Snellen E chart (≥5 years) were used.

Complete ophthalmologic exam of all patients, consisting of clinical assessment of the adnexa (eyebrows, eyelids, eyelashes and conjunctiva), anterior segment (sclera, cornea, anterior chamber), media (aqueous humor, crystalline lens and vitreous humor), and fundus. Special attention was paid to palpebral fissure height (normal: 10 mm on average, although varying with age) and levator function (nil: 0–3 mm; fair: 4–7 mm; good: 8–12 mm; excellent: >13 mm). The latter diagnostic criterion was important because it determined the surgical technique to be used. Margin-reflex distance was assessed (normal: 4 mm approximately), as were presence of superior palpebral sulcus, Bell's phenomenon, tear production, dominant eye and corneal sensitivity.

Diagnostic criteria Ptosis was considered present when: the upper palpebral border occluded 1.5–2 mm of cornea in primary gaze position; the palpebral fissure height was <9 mm high in its central or paracentral zone in primary gaze position and without frontalis activity; the distance from the center of the pupil to the upper palpebral border was <4 mm approximately.[3]

Study variables The variables employed and their definitions can be found in Table 1.

Surgical technique The frontalis sling procedure was chosen because all patients had nil or virtually nil function of the frontalis muscle—a characteristic nearly always present in congenital ptosis, for which this is the indicated technique.

Table 1: Study variables

Variable	Parameters
Age at time of surgery (years)	1–4 5–7 ≥8
Sex	Male Female
Laterality of ptosis	Unilateral Bilateral
Degree of ptosis	Mild: ≤2 mm Moderate: 3–4 mm Severe: >4 mm
Degree of ptosis correction	Corrected: Palpebral margin at 1.5 mm below the corneal limbus. Undercorrected: Improvement in palpebral margin position, but ptosis still present to some degree. Not corrected: Same as prior to surgery
Amblyopia classification (visual acuity, VA)	Mild amblyopia VA >0.5 Moderate amblyopia VA 0.1–0.5 Profound amblyopia VA <0.1
Intraoperative complications	Anesthesia reaction Hemorrhage Damage to palpebral tarsus Damage to surrounding tissue Eyeball injury
Postoperative complications	Suture dehiscence Suture exposure on frontalis Granuloma Undercorrection Palpebral border irregularity

A brief description: The incision line is placed at the cephalic border of the tarsus, 8–10 mm from the upper eyelid's free border in the case of bilateral ptosis, or, in the case of unilateral ptosis, at the supratarsal fold, using the healthy side as a reference. Reference lines for the supraciliary incisions are marked at medial sclerocorneal level, lateral sclerocorneal level and pupil. The full extension of the preseptal portion of the orbicularis muscle is exposed by dissection and the three flaps to be used are marked. The flaps are incised to the full extension of the preseptal portion of the orbicularis muscle: two lateral and one medial. The medial flap is released up to the medial sclerocorneal border; of the two lateral flaps, the upper is medially released up to pupil level and the lower up to the lateral sclerocorneal border.

Next, myorraphy of the remaining orbicularis muscle is performed. Three superciliary cutaneous incisions are made perpendicular to the medial and lateral sclerocorneal and pupil reference lines, and by tunnelization of the subcutaneous palpebral space, the flaps are rotated. The necessary traction is provided for each flap, depending on the degree of ptosis, leaving the sclerocorneal limbus free with an overcorrection of 1 mm, and the flap is attached to the frontalis muscle with a non-absorbable polypropylene suture. The eyelid is sutured, attaching the dermis to the orbicularis muscle in order to simulate the tarsal fold.[13]

At the ICO, silicone and autologous fascia lata have been used for congenital ptosis surgery, but 4/0 polypropylene, a non-absorbable suture, is generally used because it is more readily available. Although no life-long outcomes have been reported for this material, its durability provides a margin of safety to ensure correction in childhood for optimal visual rehabilitation.

Post-surgery, patients were assessed at 24 hours, 7 days, 15 days and 1 month, and monthly thereafter to 6 months. On each of these occasions, palpebral fissure height, surgical wounds on eyelids and brow, and symmetry between the corrected and contralateral eyelid were assessed. A VA exam was performed every three months; however, only VA outcome at 6 months was recorded, as it was the point at which visual rehabilitation had produced the most clinically relevant results.

Postoperative rehabilitation Fifteen days post-surgery at cessation of inflammatory response, patients were examined using cycloplegic drugs to identify any existing refractive defects. These were corrected with lenses for the first month and a half; afterwards, patching commenced for all patients, depending on severity of amblyopia and bearing in mind that the younger a child is, the less patching time is needed to produce the same benefit.[10,14]

Patients with profound amblyopia wore the patch 6 hours per day; those with moderate amblyopia, 4 hours per day, in both cases with at least one hour of near-gaze activities; and patients with mild amblyopia, 2 hours per day, with 20 minutes to 1 hour of near-gaze activities. The dominant eye was always patched to oblige the patient to use the amblyopic eye and thus stimulate visual development, combining this with optical correction.[10,14]

Data sources and processing Data were taken from the patients' clinical histories and processed with SPSS, version 11.5 for Windows, after database creation. Absolute frequencies (number of cases) and relative frequencies (percentages) were determined and summarized in tables.

RESULTS

Of the 11 patients included in the study, the majority (54.5%) were aged 1–4 years. Males predominated (63.6%) (Table 2).

Unilateral ptosis was the most common presentation, constituting 81.8% of cases operated.

Prior to surgery, 63.6% of patients presented moderate ptosis and 36.4% severe. After surgery, there was full correction in 81.8%. Two patients (18.2%) were left with mild palpebral ptosis (Table 3).

Table 2: Congenital ptosis surgical patients by age and sex

Age in years	Female		Male		Total	
	No.	%	No.	%	No.	%
1–4	2	18.2	4	36.3	6	54.5
5–7	1	9.1	1	9.1	2	18.2
≥8	1	9.1	2	18.2	3	27.3
Total	4	36.4	7	63.6	11	100.0

Table 3: Patients by degree of ptosis pre- and post-surgery

Degree of ptosis	Preoperative		Postoperative (3 months)	
	n	%	n	%
Nil	0	0.0	9	81.8
Mild	0	0.0	2	18.2
Moderate	7	63.6	0	0.0
Severe	4	36.4	0	0.0
Total	11	100.0	11	18.2

Table 4: Patients by visual acuity pre- and post-surgery

Amblyopia classification (visual acuity, VA)	Preoperative		Postoperative (6 months)	
	n	%	n	%
< 0.1 (Profound amblyopia)	2	18.2	0	0.0
0.1–0.5 (Moderate amblyopia)	8	72.7	1	9.1
> 0.5 (Mild amblyopia)	1	9.1	10	90.9
Total	11	100.0	11	100.0

Table 5: Patients with intraoperative complications

Complication	n	%
None	8	72.7
Damage to palpebral tarsus	2	18.2
Hemorrhage	1	9.1
Total	11	100.0

Table 6: Patients with postoperative complications (>3 months)

Complication	n	%
None	7	63.6
Undercorrection	2	18.2
Suture exposure on frontalis	1	9.1
Palpebral border irregularity	1	9.1
Total	4	100.0

In the preoperative period, 72.7% of patients had VA 0.1–0.5 D; however, 6 months after surgery, 90.9% had VA >0.5 (Table 4).

There were few complications. Only 3 patients (27%) experienced intraoperative complications and these did not affect final outcome (Table 5). Undercorrection was the most common complication, present in 2 patients (18.2%) (Table 6).

Figures 3–6 show the surgical procedure's stages and the final outcome in a patient with severe ptosis.

DISCUSSION

Patients with congenital conditions should be seen, diagnosed and treated at an early age, to ensure a better quality of life in childhood and adulthood. The Spanish Society for Reconstructive and Aesthetic Plastic Surgery takes the view that congenital ptosis should generally be corrected before the patient starts school, which in most cases is after age 4. In any case, severe ptosis that can lead to amblyopia should be corrected as soon as possible to preserve normal vision, but never before the age of 1 year.[13,15]

The fact that the majority of our patients had surgery before the age of 4 is the result of early diagnosis, even though they were among the most difficult unresolved cases seen in secondary institutions. Influential in this is the fact that patients in Cuba have free access to health services and are usually assessed first in primary care facilities. From there, they are referred to secondary

Figure 3: Patient with severe unilateral ptosis at start of surgery

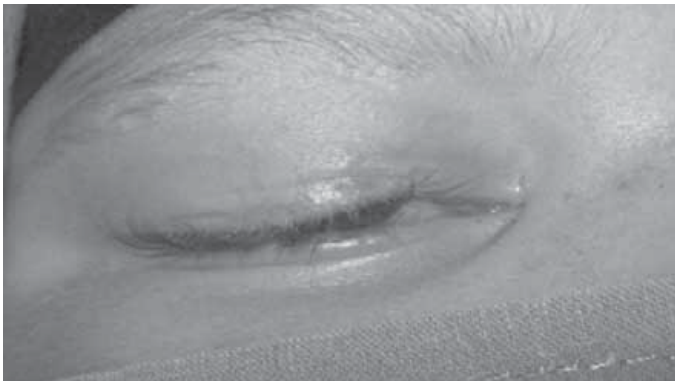


Figure 4: Polypropylene suture used to join tarsus and frontalis muscles

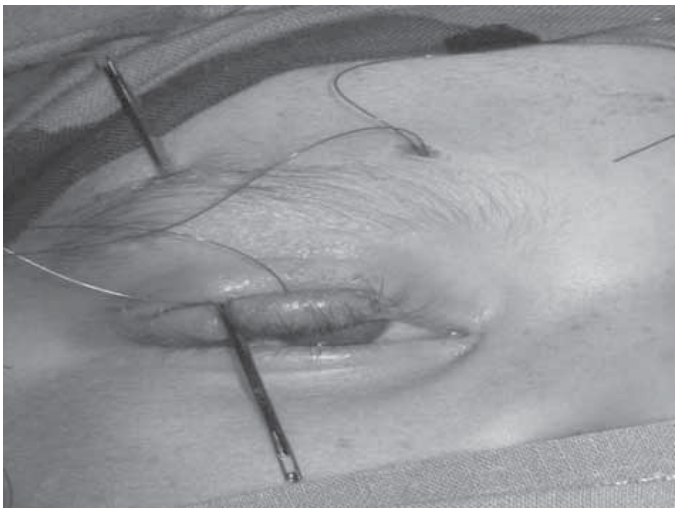


Figure 4: Patient in immediate postoperative period, corrected

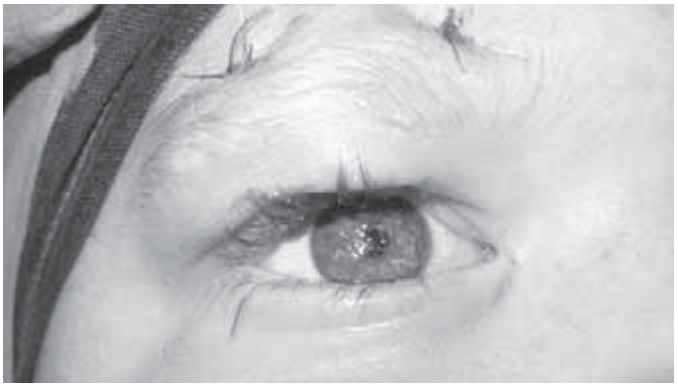
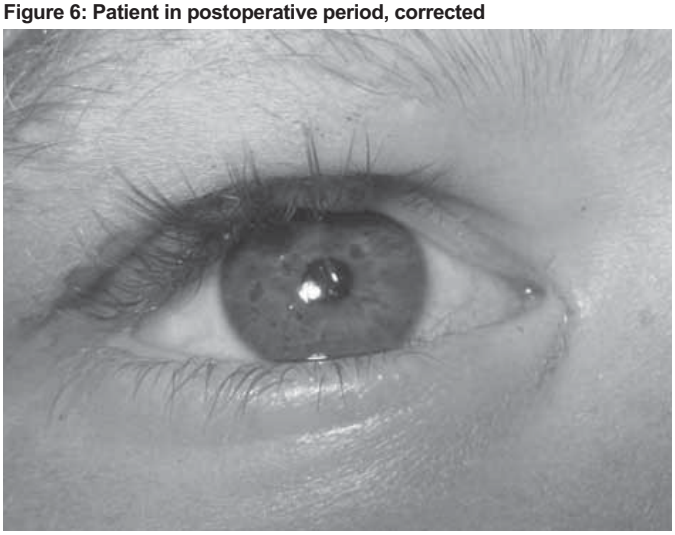


Figure 6: Patient in postoperative period, corrected



care hospitals, where the majority of cases are treated; some are then referred to institutions like ours at the tertiary level.[16]

In this study, unilateral ptosis clearly predominated, similar to Salcedo and Junceda's findings of only 25% bilateral cases.[1,3] In the unilateral form, if the droop completely occludes the pupillary area, it can cause amblyopia in the affected eye and requires immediate surgery. In contrast, when ptosis is bilateral, the patient accommodates by activation of the frontalis muscle and a compensatory head posture (backward neck flexion), so that treatment can be delayed for a reasonable period of time, bearing in mind risk of spinal disorders caused by this position.[1,2]

All patients in the study had a degree of ptosis that impaired their VA, and hence their visual prognosis, requiring surgical intervention. All underwent frontalis sling surgery, indicated for patients with congenital ptosis characterized by poor or nil upper eyelid levator function.[3,17]

Each of the numerous materials that have been used for the frontalis sling has particular advantages. Most surgeons hold that the best surgical material for permanently attaching the eyelid to the frontalis muscle is autologous fascia lata,[17] which is longlasting, easy to harvest and well developed in children aged >5 years. However, its harvest and placement require large incisions, resulting in additional surgery and tissue damage. Silastic tubing may be preferable in cases where the orbicularis muscle is fully compromised (for example, in chronic external ophthalmoplegia).

Banked irradiated facia lata is especially useful in young children, where taking fascia lata from the thigh is impossible due to poor tendon development. Autologous aponeurosis has long been replaced by non-absorbable sutures, resistant alloplastic materials well-tolerated by the majority of patients that correct ptosis without large eyelid or brow incisions. Synthetic materials have a less long-lasting effect compared with autologous materials, and their use is associated with a higher incidence of secondary infection,[17] but polypropylene sutures are easy to place and can be removed without difficulty if there is overcorrection or a problem on the ocular surface. For these reasons and because of its greater availability, polypropylene was the material used in the surgical management of patients in this study.

Of patients who received surgery, full correction was achieved in all but two. These two patients suffered from severe ptosis (one unilateral and the other bilateral) and were left undercorrected with mild ptosis, at least permitting visual rehabilitation and sparing them the social and psychological isolation that this condition can engender. These patients may also have additional surgery in the future, raising the eyelid to the normal position; this would have cosmetic benefits at least, if not functional ones.

Depending on its severity, drooping of the eyelid can cause refractive defects such as astigmatism, strabismus and more seriously, amblyopia. In this study, VA improved by at least one or two lines on the Snellen Chart, demonstrating once again that deprivation was the cause of diminished vision, as revealed in the literature describing absent or inadequate stimulus as a direct cause of amblyopia.[18–20]

The low incidence of intraoperative and immediate postoperative complications in our patients who received frontalis sling surgery for congenital palpebral ptosis coincides with international reports on this surgical technique.[1,21,22]

Concerning later postoperative complications: undercorrection resulted in two patients, even though the surgical technique selected was correct and all established steps were followed, taking the necessary measurements for each patient into account. This coincides with the literature, which indicates that undercorrection is one of the most common complications of this surgical technique.[3]

Some studies also state that factors influencing rest and postoperative care may have a bearing on appearance of undercorrection, children being a group vulnerable to trauma and other agents that predispose them to this complication.[3,22]

In our study, appearance of complications did not strongly correlate with ptosis severity. Intraoperative complications were highly dependent on the tissue characteristics of each child. In one patient who required a significant correction due to severe ptosis, the tension necessary to raise the eyelid resulted in injury to the tarsus. This complication is generally linked with the fragility of the infantile tarsus, which moreover, we found varies from child to child.

Exposure of the suture on the frontalis appeared in one case, due to the superficiality of sutures, a cause mentioned in the literature.[3] A second case presented irregularity of the palpebral border. For severe irregularities, whether temporal side ("surprised" look) or nasal side ("sad" look), the literature recommends the best solution is to lightly retroinsert 1/3–1/2 of the width of the levator muscle as if dealing with a palpebral retraction.[2,3] Our case was not severe enough to require intervention and the defect has been evolving slowly but favorably.

There are few studies of ptosis in Cuba, and none have followed patients beyond surgery; that is, through visual rehabilitation. Thus, the primary importance of this study is that researchers who evaluated patients and performed surgery also provided follow-up, observing the impact of treatment on improved VA and appearance—important for the children's learning and social relations over time.

A limitation of this study was the small number of subjects, since patients seen in our institution are generally those who have not been successfully treated at the secondary level. Another was its short duration.

CONCLUSIONS

We obtained clinically significant VA improvement using the frontalis sling procedure to correct simple congenital ptosis. This enabled visual rehabilitation, which is important for preventing amblyopia and facilitating children's improved learning and social relations.

We consider it important to accumulate a larger number of patients in a second study, to provide more comprehensive results to guide us in treatment and follow-up of congenital ptosis. An institutional project is being developed based on this premise.

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