Pediatric Blepharoptosis

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Abstract

Congenital blepharoptosis, caused by levator muscle dysgenesis, presents at birth and may lead to disturbed visual development and function. Other causes of ptosis in pediatric patients can be myogenic, neurogenic, mechanical, or traumatic. Timely correction is, therefore, critical, and careful preoperative planning and intraoperative considerations are crucial to achieve optimal outcomes and minimize potential complications. The various surgical techniques, including the frontalis suspension or sling, levator resection and advancement, Müller's muscle conjunctival resection (the Putterman procedure), and modified Fasanella-Servat procedure are each associated with distinct indications, benefits, and drawbacks, necessitating a unique tailored approach to each surgical candidate.

Keywords

- pediatric ptosis
- blepharoptosis
- droopy eyelid

Blepharoptosis, commonly abbreviated as ptosis, is defined as an abnormally low-lying upper eyelid margin on primary gaze. Congenital blepharoptosis occurs in isolation or concurrently as an ocular or systemic syndrome. The surgical repair of ptosis in pediatric patients is particularly challenging because of aesthetic and functional implications. Although not all children require surgical repair, the timely correction of pediatric ptosis is crucial in maintaining normal visual development and function.

Epidemiological data are dependent on the ethnic group studied, thereby limiting the widespread applicability of published results. A retrospective study of children in Minnesota revealed simple congenital ptosis occurring in 1 out of 842 births and more commonly affecting the left side (55%).¹ A Chinese study found a prevalence of 0.18% in the general population with most cases sporadic in nature, 18.4% demonstrating autosomal dominant inheritance, and 14.5% demonstrating autosomal recessive inheritance.² Results from Egypt demonstrated unilateral congenital ptosis in 65% of instances, with left-sided occurring more commonly than right-sided (74%) ptosis.³

Anatomy

The repair of congenital ptosis requires familiarity with the functional mechanisms responsible for eyelid elevation and surface anatomy. In normal anatomy, the upper eyelid should generally fall between 1 to 2 mm below the upper limbus. Eyelid elevation is primarily provided by the levator palpebrae superioris (LPS) muscle, which is innervated by the superior branch of cranial nerve III. This nerve also innervates the superior rectus muscle. Whitnall's ligament converts the vector of force from the LPS muscle from horizontal to vertical, providing support for the upper eyelid. The LPS muscle originates on the lesser wing of the sphenoid and inserts on Whitnall's ligament, continuing inferiorly to the tarsus as an aponeurosis. Müller's muscle, innervated by the involuntary sympathetic nervous system, provides secondary function in eyelid elevation. Originating from the inferior surface of the levator and inserting on the superior edge of the tarsus, it is responsible for ~2 to 3 mm of eyelid elevation in most people.

The longitudinally oriented frontalis muscle interdigitates with the horizontally oriented orbicularis oculi muscle, which allows for vertical movement of the superior orbit. In a ptotic

patient, the frontalis is often used as an accessory eyelid elevator. Its distinct anatomical characteristics allow it to be used as a sling or muscle flap to treat blepharoptosis in patients with poor levator function.

Indications and Considerations

Congenital ptosis most commonly results in unilateral poor levator function,^{1,3} more specifically a myogenic development abnormality, found in 76% of childhood ptosis cases.⁴ Other etiologies include a defect in the LPS muscle with fibrosis and decreased skeletal muscle fibers^{5,6} and levator disinsertion secondary to trauma at birth.⁷

Ptosis can be unilateral or bilateral and may be seen either in isolation or in conjunction with other syndromes. Instances of unilateral congenital ptosis are particularly challenging with regards to achieving good symmetry and desired aesthetic outcomes. With simple, unilateral congenital ptosis, most surgeons choose to operate only on the affected side.⁸ However, proponents of bilateral surgery in cases of unilateral ptosis argue that this approach provides better synkinetic eye movements. Hering's law of equal innervation states that when one eyelid is ptotic, the brain increases innervation to both LPS muscles. As a result, a normal appearing eyelid may appear so due to pseudoretraction. After the repair of unilateral ptosis, the innervation to bilateral LPS muscles is decreased, resulting in ptosis of the originally normal lid.⁹ The surgeon should anticipate the possibility of contralateral ptosis and consider a bilateral balanced ptosis repair if necessary.

It is important that the surgeon consider syndromes that may be associated with congenital ptosis. These include Horner's syndrome, Marcus Gunn's jaw-winking syndrome, congenital fibrosis of extraocular muscles, Duane's retraction syndrome, and blepharophimosis eyelid syndrome. Additional considerations are warranted, as Larned et al observed significantly higher rates of congenital heart disease in otherwise normal congenital ptosis patients.¹⁰

Although generally nonprogressive, ptosis may be associated with abnormal visual development and function.¹¹ Surgical repair is indicated in instances where the upper eyelid interferes with the visual axis, resulting in stimulus deprivation or astigmatism that is amblyogenic.¹² The best-suited surgical approach is primarily based on levator function, the degree of ptosis, and the patient's response to 2.5% phenylephrine testing. Other considerations include the eyelid crease location, symmetry, and eyelid contour.¹²

The timing of surgical treatment must be tailored to the severity of ptosis and the wishes of the patient and family. Severe ptosis that threatens visual development should be considered after 6 weeks of age, whereas only mild ptosis with only a minimal asymmetry to the eyelids can simply be observed. For more moderate ptosis, corrective surgery can be performed at any age, but ideally is delayed until after the age of 6 months.

The most commonly occurring complications include overcorrection and undercorrection, which may require surgical revision. The surgeon must carefully balance sufficient eyelid elevation with the prevention of lagophthalmos, which is more likely to occur with overcorrection. Regardless of the

surgical approach, lagophthalmos may occur postoperatively. Other complications include wound dehiscence, hematoma, infection, entropion, ectropion, and corneal abrasion or irritation. Overcorrection resulting in lagophthalmos may lead to dry eye and corneal irritation.^{13,14}

Evaluation and Examination

Preoperative assessment is necessary to accurately determine the diagnosis and select the appropriate treatment modality. Upon noticing signs of congenital ptosis, the patient should undergo a thorough ophthalmologic examination. A relevant history should be taken, including birth history, eye trauma, and family history of ocular pathologies.

Accurate preoperative assessment is critical to predict optimal postoperative eyelid height and stable outcomes. The examination is initially focused on the function of the LPS muscle. An assessment of function is performed by immobilizing the brow and measuring the upper eyelid movement from downward to upward gaze. Normal excursion is defined as 10 to 15 mm, fair function is 6 to 9 mm, and poor function is 5 mm or less.¹⁵

The marginal reflex distance-1 (MRD-1) is defined as the distance between the center of the pupillary light reflex and the upper eyelid margin with the eye in primary gaze. Normal MRD-1 is 4.0 to 4.5 mm, and a normal palpebral fissure is ~10 mm in an adult, but is 5 to 7 mm in infants.¹⁶ In addition, in-office 2.5% phenylephrine testing is critical to assess for the function of Müller's muscle surgery. In general, if the desired eyelid height and symmetry is achieved with this test a modified Fasanella or conjunctival-Müllerectomy ptosis repair can be performed. Finally, testing for the presence of a Bell's phenomenon is critical. This is the involuntary upward deviation of the eye with eyelid closure. If this reflex is missing, there is an increased risk for corneal exposure and surgical undercorrection should be considered.

Surgical Techniques

The primary surgical approaches for congenital ptosis include the frontalis sling, levator resection, and Müller's muscle-conjunctival resection with or without a tarsectomy. The decision of which surgical approach to use is primarily guided by levator function, the degree of ptosis, and the response to 2.5% phenylephrine. The remainder of this discussion will focus on the primary methods of repair, including the frontalis sling and Müller's muscle resection.

Frontalis Suspension/Sling

The frontalis sling is the most commonly used method to treat congenital ptosis and is indicated in severe cases with levator function of 4 mm or less.¹⁷ The principle behind this approach is to create a link between the frontalis muscle and the upper lid tarsus, utilizing the frontalis to assist in eyelid elevation.^{15,18} For satisfactory outcomes, the eyelid must have good excursion on passive movement.¹⁵ A variety of autologous and synthetic materials can be used in the frontalis sling. Autologous materials include fascia lata autografts, banked/preserved fascia lata,

the palmaris longus tendon, and the temporalis fascia. Synthetic materials consist of monofilament or polyfilament nylon, Prolene, Mersilene mesh, silicone, polytetrafluoroethylene, chromic gut, collagen, stainless steel, silk, and polyester. The most commonly used autologous and synthetic options are tensor fascia lata and silicone, respectively.

When performing frontalis suspension surgery, it is crucial to use the most reliable and effective sling material to optimize outcomes. Synthetic materials are readily available, but are more prone to recurrence, exposure, wound granulo-ma, or infection postoperatively.^{8,19,20} Cable suture slings, such as Prolene or nylon, are particularly susceptible to recurrence; therefore, they are primarily indicated as tempo-rary treatment to prevent amblyopia in severe ptosis.^{20–22} Expanded polytetrafluoroethylene (ePTFE) is a synthetic porous fluoropolymer with a low recurrence rate^{23,24} and an infection rate between 7% and 45.5%.^{20,23} Silicone provides the benefits of easy reversibility and increased elasticity, which decreases the risk of lagophthalmos (►Fig. 1).^{8,25} Mersilene is a nonabsorbable knitted polyester mesh that is both flexible and porous, allowing tissue ingrowth.¹²

In terms of autologous materials, preserved fascia lata is less commonly used due to high recurrence rates and the possible risk of transmitting infectious diseases.^{20,26} Autolo-gous fascia lata, on the other hand, is considered the gold standard for treatment. Due to its low complication and recurrence rate, it is deemed the best for long-term correction (►Figs. 2 and 3).⁸ Disadvantages include the necessity of a

Fig. 2 Autogenous fascia lata sling ptosis repair. (A) Harvesting fascia lata from the lateral leg. A fascia lata stripper is used. (B) The desired eyelid height is set by tightening the fascia lata strips.

Fig. 1 (A) Silicone sling surgery. The sling is staged to a needle that is passed in the pretarsal/suborbicularis plane. (B) Rhomboid configuration. The sling is passed to the brow via two brow incisions. The silicone rod is passed through a silicone sleeve and the eyelid is elevated to the desired intraoperative height. (C) Pre- and postoperative photo of a patient with left congenital ptosis treated with a frontalis silicone sling.

Fig. 3 Pre- and postoperative autogenous fascia lata sling ptosis repair. (A) Patient with isolated right congenital ptosis. (B) Postoperative photo taken 6 months after surgery. Note the left upper eyelid is now lower as compared in (A). This is due to Hering's law.

second operative site on the leg with associated scarring, the possibility of contracture and fibrosis, and a minimum patient height of 38 inches.^{18,19} Additionally, rapid integration with surrounding tissue may hinder attempts at revision, and resorption may limit long-term success rates.^{8,20} As a result, achieving appropriate eyelid height fixation intraoperatively is critical when using autogenous fascia lata. Autogenous temporalis fascia has been used as an alternative and has demonstrated similar rates of success.²⁷

With children, some authors describe a strong prioritization of aesthetic considerations for parents, with the desire to

limit revisions.⁸ In these instances, nonextensible materials such as autologous fascia lata ePTFE may be better suited.⁸ The eyelid crease formation is dependent not only on the location of the eyelid skin incision, but also the fixation of the fascia to the superior portion of the tarsus.²⁸

The surgeon can utilize either a single-loop or double-pentagon design to perform the frontalis sling. Between the two approaches, recurrence rates, functional outcomes, and cosmetic results are equivalent.²² Multiple modifications to the traditional frontalis sling procedure have arisen. Increasing the surface area connection through broad fixation of the fascia to the frontalis muscle improves strength and durability.²⁸ Recent studies have demonstrated that tarsal fixation of silicone, fascia lata, or ePTFE to the tarsal plate through a direct eyelid crease incision improves success rates.^{23,29,30}

As with any foreign material, inflammation, infection, and extrusion are possible complications that may require antibiotics or sling explantation.¹² It has been demonstrated that in patients with severe congenital ptosis undergoing correction using fascia lata, postoperative eyelid height can be predicted by overcorrecting intraoperatively.³¹ Overcorrection occurs more frequently than undercorrection or recurrence after frontalis suspension utilizing autogenous fascia lata.¹⁹ Lagophthalmos is a relatively common complication following this procedure, particularly in the immediate postoperative period.¹⁵

Levator Resection and Advancement

Levator resection and advancement is generally used in patients with good levator function, defined as 5 mm or greater.^{12,18} In this technique, exposure is attained through a superior eyelid crease incision with dissection down to the levator aponeurosis, which is then folded or excised and

Fig. 4 Anterior levator resection. (A) Intraoperative photo showing the levator aponeurosis that is being sutured to the tarsus with three 5-0 Vicryl sutures. (B) Preoperative photo of a patient with a right Marcus Gunn jaw wink and ptosis. (C) Postoperative photo taken 1 year after an anterior levator resection. The levator muscle was not extirpated and the wink remains, but is much less noticeable with the eyelid in a better position.

subsequently reattached to the superior tarsus.¹² This superior crease incision allows for concurrent removal of redundant skin and orbicularis oculi muscle, as well as skin crease reformation in the non-Asian eyelid¹⁵

There are two schools of thought when approaching the amount of levator to resect. The first describes approximating the eyelid at the appropriate position and excising excess tissue (—Fig. 4). However, this technique is subject to variable outcomes based on decreased ocular muscle tone from anesthesia and gaze position.¹³ Alternatively, algorithms based on levator function and the degree of ptosis have been devised. A straightforward formula of 4 mm of resection per 1 mm of ptosis has been utilized,³² in addition to more-comprehensive algorithms.⁷ These algorithms are reliant on preoperative measurements and do not account for the quantity or the quality of the levator muscle.¹³ Therefore, a familiarity with multiple methods is recommended for optimal results.

The levator plication is a similar technique whereby the levator aponeurosis is exposed, but not divided or elevated. Instead, the levator aponeurosis is tightened by vertical plication sutures. The benefit to this option is that it requires less dissection than the levator advancement, which also contributes to the drawback of a potential bulge from redundant tissue.¹⁵

The advantages of this technique include low rates of reoperation,³³ the preservation of Müller's muscle and Whitnall's ligament,³⁴ and the lack of foreign material, which may reduce the risk of infection.¹² In addition, the small eyelid incision (8–13 mm) allows for less local anesthetic and tissue distortion, less edema, shorter operative times, and a more rapid recovery from surgery.⁹ As with other techniques, undercorrection is a common complication. Other complications include overcorrection, infection, hematoma, poor lid contour, and amblyopia.¹³

Müller's Muscle Surgery

The two effective procedures to resect Müller's muscle is the Putterman procedure, which is a resection of Müller's muscle and conjunctiva. When this tissue complex is clamped with a portion of the tarsus, the procedure is called a modified Fasanella-Servat.

The Müller's muscle conjunctival resection technique (Putterman) is indicated in etiologies where topical 2.5% phenylephrine, stimulating Müller's muscle through its sympathomimetic activity, improves ptosis of the eyelid.^{12,18} However, favorable outcomes have also been described in patients who do not respond to phenylephrine.³⁵ As this technique is dependent on the function of both the LPS and Müller's muscles, it is indicated for cases of mild ptosis.

Eyelid height cannot be adjusted intraoperatively. Instead, the amount of tissue resection is determined preoperatively. When phenylephrine testing yields a normal eyelid height, a resection of 8.25 mm is indicated.³⁶ Mercandetti et al described a ratio of 1 mm of resection for each 0.32 mm of eyelid elevation.³⁷ A separate modification by Dresner entailed a resection amount dependent on the response to phenylephrine testing. If at least 2 mm of eyelid elevation

Fig. 5 Modified Fasanella-Servat procedure. (A) The eyelid is everted; the tarsus, the conjunctiva, and the Müller's muscle are clamped. (B) After resecting the clamped tissue (not pictured), the Müller's muscle is sutured in a vertical mattress fashion to the tarsus with a 5–0 Nylon suture. (C) The suture is anteriorized to the eyelid skin and tied, which is removed 7 days later in the office. (D) Pre- and postoperative 2.5% phenylephrine testing in the office, and a 1-year postoperative right modified Fasanella-Servat procedure. Note the final eyelid height is well approximated by the phenylephrine test in (B).

result, a resection of 4 mm was performed for 1 mm of ptosis, 6 mm of resection for 1.5 mm of ptosis, 10 mm of resection for 2 mm of ptosis, and 11 or 12 mm of resection for greater than 3 mm of ptosis.³⁸ Perry et al hypothesized that the complete or near-complete excision of the Müller's muscle accomplishes the same degree of eyelid elevation as phenylephrine testing and that resection of the tarsus yields a 1:1 ratio of eyelid elevation.³⁶

The posterior eyelid approach originally described in this technique conceals any visible scarring.³⁹ The primary advantage of this technique over the Fasanella-Servat procedure described below is that the tarsus is spared.¹² Unique to this procedure is the risk of corneal abrasion or ulceration from an exposed suture on the palpebral conjunctiva.¹² Others have argued that the removal of goblet cells and accessory lacrimal glands may increase dryness of the eye.⁴⁰

The Fasanella-Servat procedure, also known as tarsoconjunctival mullerectomy, has the same indications as the Putterman technique. This is a posterior approach where the upper eyelid is everted, allowing for the use of hemostatic clamps to grasp the conjunctiva, the tarsus, and Müller's muscle (► Fig. 5).¹⁵ In one study by Pang et al, a 76.5% success rate was noted in the treatment of congenital ptosis using this technique.⁴¹ Berry-Brincat et al demonstrated similar results, with a success rate of 71% and a reoperation rate of 20%.⁴ Complications from the Fasanella-Servat procedure include dermatochalasis, duplicate eyelid creases, eyelid contour abnormalities, hematoma, wound dehiscence, pyogenic granulomas, and bleeding.⁴¹ Other disadvantages include limited flexibility, the inability to adjust the amount of ptosis correction, and an increased risk of corneal abrasion with the posterior approach and resection.⁴²

Conclusion

Clinically significant congenital ptosis in pediatric patients is best treated by surgical means. Determination of the underlying pathology in each case is important to navigate optimal treatment strategies. The surgeon must understand the complex eyelid anatomy in addition to being well versed in the various benefits, drawbacks, and indications of each technique. The primary techniques most commonly used include the frontalis sling with silicone or autogenous fascia lata, levator resection and advancement, and Müller's muscle resection.

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