Congenital ptosis—

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New concepts and new treatments

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Abstract

Treatment of congenital ptosis can have significant effects on visual and psychosocial development. Traditionally, treatment has been based on the extent of ptosis, the presence of amblyopia, and the function of the levator muscle. Recently, there have been significant shifts in the approach to congenital ptosis. These include the adoption of the frontalis flap procedure and the timing of surgery. This article will review these recent changes to update the reader on the current approach to congenital ptosis.

Congenital ptosis can be isolated or associated with other conditions. In the past, the condition was thought to be a myopathic problem. It is now believed that most congenital ptosis is caused by a dysinnervation of the levator muscle during development. This places congenital ptosis under the umbrella of congenital cranial dysinnervation disorders (CCDD), therefore suggesting a neurogenic cause.[1] There are a number of associated syndromes with congenital ptosis, including blepharophimosis-ptosis-epicanthus inversus syndrome (BPES), congenital fibrosis of the extraocular muscles (CFEOM), and Marcus Gunn jaw-winking syndrome. In evaluating congenital ptosis, it is important to rule out these other associated syndromes, as their treatment will differ from "simple" congenital ptosis. Other conditions to consider when evaluating a patient for congenital ptosis are causes of mechanical ptosis including infantile hemangiomas and plexiform neurofibromas.

Congenital ptosis should be present at birth and not significantly change. However, recognition of the ptosis may not occur immediately after birth. If there is any question with regards to the onset of ptosis, previous photographs of the patient should be inspected. The condition can be unilateral or bilateral, and this has implications for treatment; however, the condition is more commonly unilateral with a left-sided predominance.[2] The most important issue to address at the first visit is the presence or absence of amblyopia or preference of one eye over the other. Visual preference or eye dominance is usually noted on the side without ptosis, even if there is no amblyopia present, with equal visual acuity on both sides. It is not uncommon for patients to have monofixation syndrome without amblyopia. Treatment of congenital ptosis usually involves a team approach including a pediatric ophthalmologist and oculoplastic surgeon. If amblyopia is detected, this should be treated, usually with occlusion therapy. If there is no amblyopia present, or if the patient has a positive response to occlusion therapy, surgery can usually be delayed. An acceptable delay is typically up to about the age of two years, when the child is thought to be at lower risk from anesthesia.[3] Usually, all congenital ptosis will be treated before the age of 5 years, and therefore before the child enters school. A recent study showed evidence of significant psychosocial issues in patients untreated for their congenital ptosis before starting school compared to controls, supporting the importance of this timing.[4]

Treatment prior to the age of two years is usually performed for significant ptosis that has not responded to occlusion therapy. Another reason for early treatment is if the ptosis is causing a significant abnormal head position, usually with the chin raised. Although the abnormal head position is not preferred, it is usually an indicator that there is no amblyopia present.

The preferred surgery for a patient with congenital ptosis depends on several factors: bilateral or unilateral disease, brow elevation, amount of ptosis, and the quality or strength of the levator muscle. With bilateral congenital ptosis, one may lean toward a frontalis suspension procedure. If there is unilateral ptosis but the patient is able to lift the brow on the side of the ptosis, then one may lean toward a unilateral frontalis suspension. However, most

decisions about the type of surgery revolve around the extent of ptosis and the strength of the levator muscle. With more ptosis and less levator strength, one would lean toward a frontalis suspension procedure. With less ptosis and more levator strength, a levator-based procedure or a posterior approach (Muller muscle-conjunctival resection [MMCR]) would be favored.

An MMCR can be performed for minimal ptosis (2 mm or less), with good levator strength, that responds to a drop of 2.5% phenylephrine. If the eyelid lifts but not to a desired height after phenylephrine application, a limited tarsectomy of up to 2 mm can be added.[5] The amount of tarsectomy to add should be twice the amount of residual elevation desired after phenylephrine instillation. For example, if phenylephrine lifts the lid 1 mm, but 2 mm elevation is desired, then a 2 mm tarsectomy would be added to the 9.0 mm MMCR, to account for the 1 mm of residual elevation desired. More than 2 mm of tarsectomy should not be performed, as this results in an unstable tarsus that can be difficult to reoperate.

Levator resection surgery should be performed for patients with 3 mm or less of ptosis with a viable levator muscle. The amount of levator to resect is based on the amount of ptosis and the strength of the levator muscle. In general, this surgery is reserved for 2 to 3 mm of ptosis. There are several indicators of levator strength. Traditionally, levator function (the distance the eyelid moves from downgaze to upgaze, measured in millimeters) has been used to determine levator health. However, this can be difficult to measure, especially in young children. Furthermore, studies have demonstrated that levator function is not necessarily the best measure of levator strength or health.[6] Other indicators of levator health include the amount of ptosis (significant ptosis in the congenital condition indicates poor levator health) and a distinct eyelid crease. A distinct eyelid crease is thought to be associated with a stronger levator muscle. Finally, the levator muscle can be evaluated intraoperatively to determine its quality. If there are visible muscle fibers, then the muscle can often be used for a levator resection; however, if the levator is extensively infiltrated with fat, a frontalis suspension procedure should be performed. There are many algorithms addressing the amount of levator to resect. This can be simplified to resecting minimal (15 mm), moderate, (20 mm), or maximal (25 mm or more) amounts depending on the amount of ptosis and the health of the muscle.

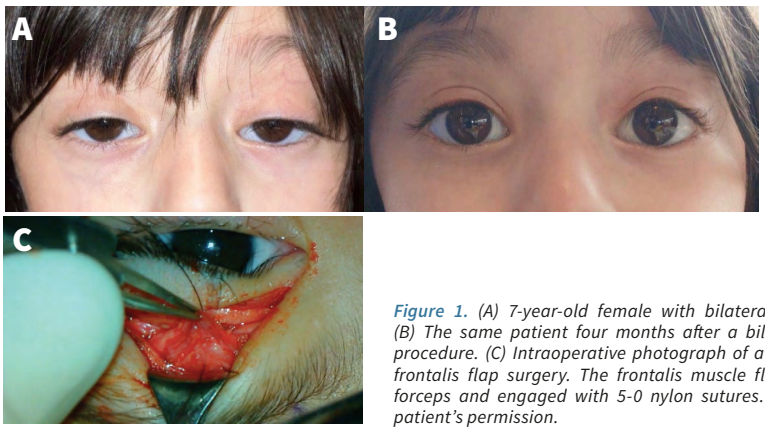

Figure 1. (A) 7-year-old female with bilateral congenital ptosis. (B) The same patient four months after a bilateral frontalis flap procedure. (C) Intraoperative photograph of a patient undergoing frontalis flap surgery. The frontalis muscle flap is held with the forceps and engaged with 5-0 nylon sutures. Published with the patient's permission.

Frontalis suspension surgery is performed to bypass the levator muscle and make a link between the frontalis muscle and the tarsus. The gold-standard material to make this link is autogenous fascia lata.[7] However, it is difficult to obtain an adequate length (usually 12–15 cm) of fascia lata before the age of four years, although some authors have described success in children less than three years of age.[8] Prior to 3 or 4 years of age, an implant is used for a frontalis sling. Appropriate implants include silicone and Gore-Tex. The problem with these materials is that they usually fail with time, and the patient will need to undergo additional surgery involving the fascia lata later in life. In addition, implants can become infected or extrude.

A recent advance in frontalis suspension surgery is the introduction of the frontalis muscle flap procedure.[9-11] In the frontalis flap procedure, a direct link is made between the frontalis muscle and the tarsus (**Figure 1**). The advantage of this procedure is that it does not have a lower age limit, and it appears to be durable. In addition, the entire surgery can be performed through an eyelid crease incision, while frontalis sling procedures require one to three incisions at or above the eyebrow, which can sometimes heal unsatisfactorily. A modification of the frontalis flap procedure involves the additional inclusion of a levator resection in combination with the frontalis flap, which helps avoid eyelid "pop" (lifting of the eyelid from the surface of the eye on downgaze with brow elevation); this addition also helps in the formation of the eyelid crease.[12] Eyelid crease formation is critical in any congenital ptosis surgery, both for cosmetic reasons and to prevent eyelash ptosis.

There are many potential complications after any congenital ptosis procedure. These include bleeding, infection, reoperation, contour abnormalities, over-correction, and under-correction. Lagophthalmos after any of the ptosis procedures is possible and may be long-lasting. In general, the more severe the ptosis, the greater the likelihood of post-operative lagophthalmos. The parents should expect significant lagophthalmos immediately after surgery which improves with time. Fortunately, most children tolerate lagophthalmos well, due to the high quality of their tears. Long-term lubrication may be required, depending on the extent of lagophthalmos.

A last consideration in frontalis suspension surgery is whether to perform bilateral surgery on a child with significant unilateral ptosis. In performing any congenital ptosis surgery, the patient will have lid lag during downgaze on the side of the surgery. This is more so in patients after frontalis suspension surgery. The unilateral lid lag can be significant, and the asymmetry can be noticeable. The argument for performing bilateral surgery for unilateral cases is to give the patient symmetry in terms of not only height and contour of the eyelids but also movement.

The necessity of revision surgery is always a possibility with congenital ptosis. In general, it is thought that levator resection surgery and frontalis sling surgery with autogenous fascia are the most durable. Frontalis suspension with synthetic materials is less durable, and the durability of the frontalis flap procedure is still under study. Early revision is usually reserved for patients who have significant over-correction (causing ocular surface disease not responding to conservative treatment) or under-correction (resulting in amblyopia that does not respond to occlusion therapy). If the over- or under-correction is minor, then revision surgery can be performed on an elective basis.

In summary, treatment of congenital ptosis continues to make strides. Many surgeons have now adopted the frontalis flap procedure with promising results. The goal to give patients symmetrical results before starting school is a priority to aid in adequate psychosocial development.

Key points:

- The timing of treatment of congenital ptosis depends on the presence of amblyopia, abnormal head position, and the prevention of subsequent psychosocial issues.
- Surgical options include Mullerectomy, levator resection, and frontalis suspension.
- Surgical treatment depends on the extent of ptosis, the health of the levator muscle, the presence of amblyopia, and the use of the eyebrow.
- The frontalis muscle flap procedure is becoming the preferred technique for frontalis suspension.

Video 1. Combined frontalis flap with levator resection procedure.

References

1. Gutowski NJ, Chilton JK. The congenital cranial dysinnervation disorders. *Arch Dis Child.* 2015 Jul;100(7):678-81.
2. Griepentrog GJ, Diehl NN, Mohney BG. Incidence and demographics of childhood ptosis. *Ophthalmology.* 2011 Jun;118(6):1180-3.
3. Vutskits L, Davidson A. Update on developmental anesthesia neurotoxicity. *Curr Opin Anaesthesiol.* 2017 Jun;30(3):337-342.
4. Hendricks TM, Griepentrog GJ, Hodge DO, Mohney BG. Psychosocial and mental health disorders among a population-based, case-control cohort of patients with congenital upper eyelid ptosis. *Br J Ophthalmol.* 2023 Jan;107(1):12-16. doi: 10.1136/bjophthalmol-2021-319276.
5. Gazit I, Gildener-Leapman J, Or L, Burkat CN, Pras E, Hartstein ME. Müller's Muscle-conjunctival Resection Combined With Tarsectomy for Treatment of Congenital Ptosis. *Ophthalmic Plast Reconstr Surg.* 2019 Nov/Dec;35(6):619-622.
6. Frueh BR, Musch DC. Evaluation of levator muscle integrity in ptosis with levator force measurement. *Ophthalmology.* 1996 Feb;103(2):244-50.
7. Weaver DT. Current management of childhood ptosis. *Curr Opin Ophthalmol.* 2018 Sep;29(5):395-400.
8. García-Cruz I, Barrancos C, Alonso-Formento N, Albandea AR, Losada-Bayo D, García-Ruiz OA, Sales-Sanz M. Frontalis Suspension Using Autologous Fascia Lata in Children Under 3 Years Old. *Ophthalmic Plast Reconstr Surg.* 2021 Jul-Aug 01;37(4):377-380.
9. Medel R, Vasquez L, Wolley Dod C. Early frontalis flap surgery as first option to correct congenital ptosis with poor levator function. *Orbit.* 2014 Jun;33(3):164-8.
10. Medel R, Molina S, Vasquez LM, Visa J, Wert A, Wolley-Dod C. Frontalis Muscle Flap Versus Maximal Anterior Levator Resection as First Option for Patients With Severe Congenital Ptosis. *Ophthalmic Plast Reconstr Surg.* 2018 Nov/Dec;34(6):565-569.
11. Eton EA, Carniciu AL, Prabhu SS, Wang GM, Kahana A. Treatment of Congenital Ptosis in Infants With Associated Amblyopia Using a Frontalis Muscle Flap Eyelid Reanimation Technique. *Ophthalmic Plast Reconstr Surg.* 2021 Jan-Feb 01;37(1):67-71.
12. Diab MMM, Abd-Elaziz K, Allen RC. Combined levator and frontalis muscle advancement flaps for recurrent severe congenital ptosis. *Eye (Lond).* 2023 Apr;37(6):1100-1106.

Conflict of interest
none