

Upper Eyelid Ptosis Revisited

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Abstract

Blepharoptosis, commonly referred to as ptosis is an abnormal drooping of the upper eyelid. This condition has multiple etiologies and is seen in all age groups. Ptosis results from a congenital or acquired weakness of the levator palpebrae superioris and the Muller's muscle responsible for raising the eyelid, damage to the nerves which control those muscles, or laxity of the skin of the upper eyelids. Ptosis may be found isolated, or may signal the presence of a more serious underlying neurological disorder. Treatment depends on the underlying etiology. This review attempts to give an overview of ptosis for the primary healthcare provider with particular emphasis on the classification, salient features of each of the subtypes, clinical evaluation, and treatment.

Introduction

Blepharoptosis or ptosis (pronounced "toe-sis") is one of the most common eyelid disorders encountered in ophthalmology. It refers to the unilateral or bilateral abnormal drooping of the upper eyelid. It usually occurs from a partial or complete dysfunction of the muscles that elevate the upper eyelid: the levator palpebrae superioris and the Muller's muscle.¹ Ptosis can be classified as congenital or acquired. The most common cause of congenital ptosis is myogenic due to the improper development of the levator muscle. A more comprehensive classification of ptosis is based on etiology and includes myogenic, aponeurotic, neurogenic, neuromuscular, mechanical, neurotoxic, traumatic, and pseudoptotic. Ptosis that obstructs the pupil may interfere with the normal development of vision, resulting in amblyopia in children.² In adults it may impair the field of vision and interfere with activities of daily living.^{2,3} Thus, the early diagnosis and treatment of ptosis is an important prognostic factor in its management.

Epidemiology of Ptosis

Although ptosis is commonly encountered in patients of all ages, there are insufficient statistics regarding the prevalence and incidence of ptosis in the United States and globally.² There is no known ethnic or sexual predilection.² However, there have been few isolated studies on the epidemiology of ptosis. A study conducted by Baiyeroju et al, in a school and a private clinic in Nigeria, examined 25 cases of blepharoptosis and found during a five-year period that 52% of patients were less than 16 years of age, while only 8% were over 50 years of age. There was a 1:1 male to female ratio in the study with the majority (68%) having only one eye affected. The most common cause of blepharoptosis in the study was congenital (56% of patients). The prevalence rate in the school survey was found to be 1.2%.⁴ Ptosis has been increasingly recognized in the elderly population, particularly after cataract extraction or lens replacement.⁵ This is probably due to the stretching or disruption of the levator muscle or its aponeurosis when the eyelid is retracted with a speculum during surgery.⁶

Anatomy

The closure of the eyelids is facilitated by the protractors of the eyelids: circumferential orbicularis oculi muscle, which is innervated by the facial (seventh cranial) nerve. The elevators of the upper eyelid are the levator palpebrae superioris and the Muller's muscle. The levator palpebrae superioris is the main upper eyelid elevator and is innervated by the oculomotor (third cranial) nerve. The Muller's muscle is a smooth muscle that arises from the undersurface of the levator and inserts into the superior tarsus. The Muller's muscle is innervated by the sympathetic nervous system. The muscle is responsible for the over-elevation of the eyelid when a patient becomes excited or fearful and leads to mild ptosis with fatigue or inattention.

Clinical Presentation

The presence of ptosis is usually an isolated finding; however, it can be part of a group of presenting symptoms of other medical conditions.

History

A detailed history of present illness includes asking about the onset, duration, variability, progression, and severity of ptosis. Also investigate whether there is involvement of one eye or both eyes simultaneously. In the past ocular history it is vital to elicit a history of eye trauma, previous eye surgeries, contact lens use, and previous eye diseases, such as dry eyes and thyroid eye disease. One should also ask about associated symptoms, such as diplopia,odynophagia, peripheral muscle weakness, and symptoms of cardiac conduction abnormalities, which may give a clue to the diagnosis of associated systemic diseases, such as myasthenia gravis, myotonic dystrophy, and Kearns Sayre's. A complete medical history is important for the diagnosis of associated systemic diseases and surgical planning. Inspecting old photographs and eliciting information regarding familial predisposition for ptosis is helpful in the assessment of the severity of ptosis.

Symptoms

Most patients present with drooping eyelids, giving a sleepy or tired appearance. The patients may complain of blurred vision, increased tearing,² and diminished upper visual fields. There is increased difficulty with activities of daily living, such as driving, reading, and climbing stairs, affecting the patient's quality of life.⁷ They may complain about frontal tension headache from overuse of the forehead muscles in attempts to elevate the eyelids indirectly by elevating the eyebrows.²

Undiagnosed congenital ptosis may result in amblyopia.² On inspection, the patients, especially children may assume a head-tilt backwards and chin-up position.² The individual may be seen lifting the eyelid with a finger or using multiple fingers to raise the entire eyebrow in an effort to view from under the drooping eyelids.² Symptoms of diplopia may be seen in ptosis associated with myasthenia gravis or third nerve palsy.

Signs

A comprehensive ophthalmic examination should be done in all cases.

- The examination begins with careful external examination along with palpation of the eyelids and the orbital rim. Evaluate any clinical evidence of relative proptosis or enophthalmos in each eye.
- Inspection of eyebrows at baseline should be done, since patient may compensate by lifting eyebrows with the frontalis muscle.
- The patient's head posture should be noted; document chin-up position.

- Best corrected visual acuity and cycloplegic refraction should be done. This helps to identify refractive errors and amblyopia. In infants, note if baby can fixate, maintain, and follow light.
- Pupillary examination is important. Pupillary size and iris color differences between the two eyes should be examined to rule out Horner's syndrome.
- Extraocular muscle movement should be evaluated with a note of diplopia - in chronic progressive external ophthalmoplegia, there may be extraocular muscle weakness together with ptosis.
- Evaluation of strabismus if present.
- Slit lamp examination should note the presence of any anterior segment pathology, such as dry eyes, corneal exposure, or exposure keratopathy.
- Dilated fundus examination should rule out any posterior segment pathology, such as abnormal retinal pigmentation seen in Kearns Sayre's syndrome.

Eyelid measurements

To quantify the severity of ptosis, various eyelid measurements should be taken with the face held in the frontal plane and with frontalis muscle relaxed.² Frontalis contracture should be neutralized by holding the brows in their relaxed position. If the droopy lid obscures the lid margin, the examiner should lift the brows to observe the lid level for measurement. The examiner should note the following:

- The **palpebrae fissure height (PF)** is the distance between the upper and lower eyelid margins at the axis of the pupil. Normal measurement is 9 to 12 mm.
- **Marginal reflex distance (MRD)** is the distance between the central corneal light reflex and upper eyelid margin with eyes in primary position. The severity of ptosis is better determined with MRD than PF measurements as lower lid malpositions are eliminated. Normal MRD is 4-5 mm.
- **Levator function** should be evaluated in all cases. The patient looks downward as a measuring device is positioned with a mark adjacent to the upper lid margin. With the examiner's hand eliminating any brow action by the patient, the patient looks upward as far as possible without changing his/her head position. The amount of lid elevation is recorded in millimeters (mm) of levator function.

Classification of levator function:

Poor: 0 – 5 mm lid elevation

Fair: 6 – 11 mm lid elevation

Good: > 12 mm lid elevation

- **Upper eyelid crease** position is the distance from the upper eyelid crease to the eyelid margin. It is normally 7-8 mm in males and 9-10 mm in females.

The position of the ptotic eyelid in down-gaze should be assessed, as it helps in differentiating congenital from acquired ptosis. Congenital ptotic eyelid remains higher in down-gaze due to lid lag.

Ancillary testing

- Corneal sensitivity should be tested in all cases. This is important because of the potential for incomplete lid closure and exposure keratopathy after surgical correction.
- Bell's phenomenon should be tested to evaluate the risk of exposure keratopathy if surgery is being planned.
- Jaw winking phenomenon - a brisk upper eyelid retraction will be elicited when the patient is asked to open and close his/her mouth or with protrusion or lateral movement of the jaw.
- Schirmer's test to evaluate tear function and fluorescein test for tear break-up time and tear meniscus may be done in individuals suspected of having dry eyes due to the potential risk of incomplete eyelid closure and exposure keratopathy following surgical correction.
- Serial external photographs of the eyes and the face may be included in the patient's record for documentation, especially in children, when direct measurements may be difficult.
- Visual field testing with the eyelids in ptotic state and then artificially elevated may be done to determine the level of functional improvement expected if surgery is being contemplated.^{3,7}
- Tensilon test is traditionally done to rule out myasthenia gravis.⁸ The ice pack test may be alternatively done in suspected myasthenia.⁹ An ice pack is applied to the eyelids for two minutes, and then the examiner looks for improvement of ptosis.

- Sympathomimetic agents, such as phenylephrine or apraclonidine,¹⁰ can be instilled under the eyelid to test the function of Muller's muscle.

Discussion

Types of Ptosis

Ptosis is broadly classified into congenital and acquired, based on age of onset of the ptosis. Ptosis that is present at birth or within the first year of life is called congenital ptosis. This is usually an isolated presentation and rarely may be associated with other findings when it is considered to be non-isolated. Ptosis that presents after the age of one year is termed acquired ptosis. This may again be an isolated or a non-isolated presentation. (See appendix 1 for diagnosis of ptosis). Ptosis may also be classified by etiology: aponeurotic, neurogenic, myopathic, neuromuscular, neurotoxic, mechanical, traumatic, and pseudoptosis.

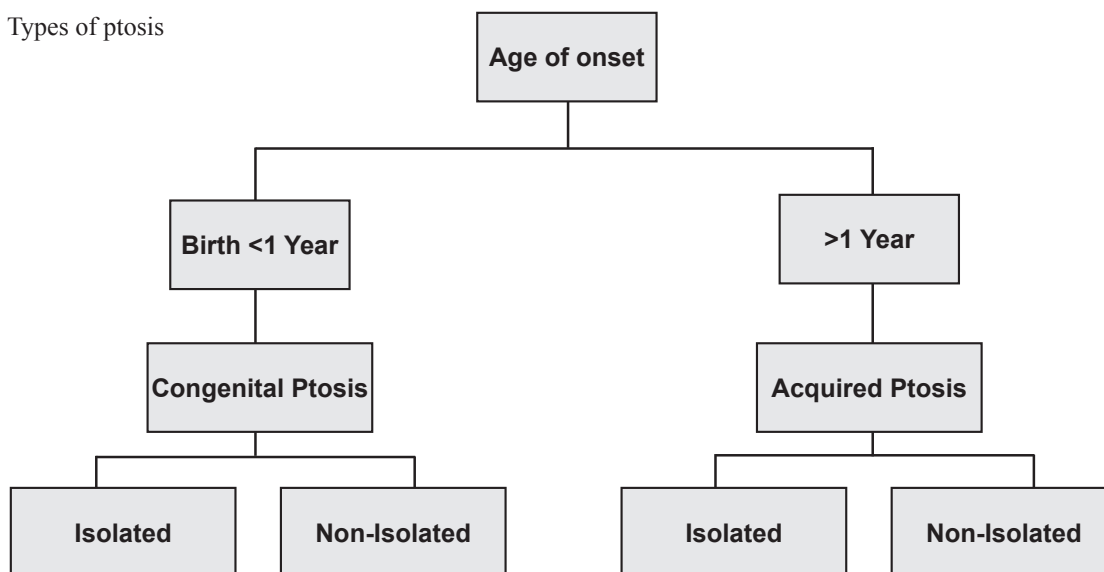
Congenital Ptosis

Abnormal drooping of the upper eyelid, present at birth or within the first year of life is termed congenital ptosis. The frequency of congenital ptosis in the United States has not been reported. In almost 75 percent of the cases only one eye is affected.¹¹

The majority of congenital ptosis is due to myogenic dysgenesis of the levator muscle. In these cases, rather than having normal muscle fibers, fibrous and adipose tissue are present in the muscle belly instead.¹² Thus there is a reduction or absence of functional muscle, impairing the ability of the levator to contract and elevate the eyelids.¹³

Most congenital ptosis is isolated and does not affect vision. In severe cases, the drooping eyelid may occlude all or part of the pupil and may interfere with vision, resulting in amblyopia.^{2,14} The droopy eyelids may "compress" the eyeball leading to astigmatism.¹⁵ A compensatory head posture, such as chin-

Appendix 1: Types of ptosis



up position, may be adopted by children with severe bilateral congenital ptosis to obtain good vision.² Some children may mechanically lift the ptotic eyelid with their finger in order to see clearly.²

Congenital ptosis may occur through autosomal dominant inheritance.¹⁶ Common familial occurrences suggest that genetic or chromosomal defects are likely.

Isolated congenital ptosis

- Simple congenital ptosis** due to a dystrophy of the levator muscle is the most common type of congenital ptosis. The patients usually manifest a lid lag on down-gaze and higher than normal or poorly formed lid crease. Rarely, lagophthalmos may be seen.
- Synkinetic ptosis** — rarely, congenital ptosis can occur due to an aberrant innervation of the levator muscle by the mandibular branch of the trigeminal nerve, resulting in Marcus Gunn Jaw-Winking Syndrome. In this syndrome, there is a brisk upper eyelid retraction when the ipsilateral pterygoid muscle contracts during mastication, jaw thrusting to the contralateral side, jaw protrusion, chewing, smiling, or sucking.^{17,18,19, 20} This phenomenon is discovered early as the infant is bottle- or breastfed.
- Aponeurotic ptosis** — congenital aponeurotic defects result from a failure of the aponeurosis to insert on the anterior surface of the tarsus or from birth trauma following forceps delivery. The skin crease may remain normal or high depending on where the aponeurosis is affected. The levator function is usually good, and there is no lid lag on down-gaze.

Non-isolated congenital ptosis

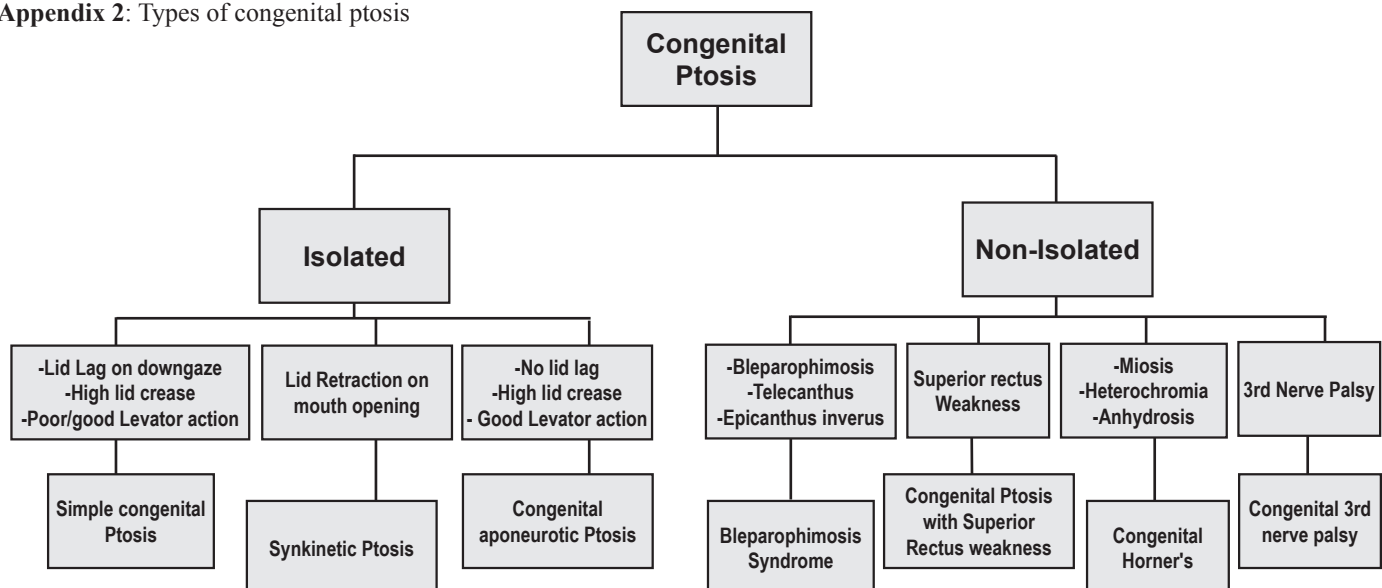
- Due to close embryological development of the levator and superior rectus muscles, congenital ptosis may be associated with **superior rectus weakness**.

- Blepharophimosis Syndrome** is another rare autosomal dominant inherited condition. It presents with the characteristic features of blepharophimosis, epicanthus inversus, telecanthus, and ptosis.²¹ This disorder is associated with a high incidence of amblyopia.²² Other non-ocular associations include ovarian failure, arched palate, and cardiac defects.
- Neurogenic Ptosis-Horner's Syndrome** occurring in infancy can present with ptosis, miosis, anhidrosis, and progressive heterochromia. The lighter colored iris is ipsilateral to the affected side. The lesion may occur anywhere along the oculosympathetic pathway. It is important to evaluate the patient for a possible etiology,²³ such as congenital varicella, tumors of the neck and mediastinum, and vascular lesions of the internal carotids or subclavian artery.
- Congenital Third Cranial Nerve Palsy** may be partial or complete. It may present with ptosis together with inability to depress, elevate, or adduct the eye. The pupil may be dilated. Signs of aberrant regeneration are rare, but, if present, the pupil may be paradoxically small and non-reactive. (See Appendix 2 - Types of congenital ptosis.)

Other causes of ptosis in children

- Birth trauma**
- Duane syndrome** — in this condition, the sixth cranial nerve fails to innervate the lateral rectus muscle. Instead, the muscle acquires an innervation from the third cranial nerve. Although the synkinesis produced does not involve lid innervations, enophthalmos with apparent ptosis may result.
- Periorbital tumor** or other deep orbital tumors may produce proptosis with ptosis.
- Congenital Fibrosis of the Extraocular Muscles (CFEOM)** is a non-progressive, autosomal dominant ocular disorder, resulting in fibrosis of the extraocular muscles.²⁴ This disorder is characterized by bilateral ptosis and external

Appendix 2: Types of congenital ptosis



ophthalmoplegia, with a compensatory backward tilt of the head. The pathophysiology of this disorder could be due to a primary neurogenic or myopathic etiology.

- **Kearns-Sayre syndrome** is a mitochondrial deletion disorder that is characterized by progressive external ophthalmoplegia, heart block, retinitis pigmentosa, and central nervous system manifestations. This condition begins in childhood but is rarely present at birth. It generally becomes symptomatic in the first or second decade of life. Bilateral ptosis is a prominent feature of this syndrome.²⁵
- **Myotonic dystrophy** is an autosomal dominant disorder characterized clinically by myotonia and progressive muscle weakness.²⁶ Patients may present with polychromatic cataracts, gonadal atrophy, or premature thinning and/or loss of hair. Myotonic dystrophy causes generalized weakness, usually beginning in the muscles of the hands, feet, neck, or face and then progressing to other muscle groups, such as the heart. Bilateral ptosis may be seen. Symptoms may appear at any time from infancy to adulthood. A severe form of myotonic dystrophy, congenital myotonic dystrophy, or Thomsen's disease may appear in newborns of mothers with myotonic dystrophy.
- **Myasthenia gravis** causes unilateral or bilateral fluctuating ptosis. This may be seen in children.
- **Pseudoptosis** — Decreased orbital mass (e.g., unilateral smaller eye, fat atrophy, blowout fracture) may produce the appearance of ptosis secondary to the decreased volume of orbital contents.

Acquired Ptosis: This is classified into Isolated or Non-Isolated Ptosis.

Isolated Acquired ptosis

- a. **Aponeurotic ptosis** — in adults the most common cause of acquired ptosis is an abnormality in the levator aponeurosis, i.e. a dehiscence, disinsertion, or stretching.²⁷ This type of ptosis most often affects the elderly but can occur in younger patients as well. This has previously been discussed in congenital ptosis. In the elderly the degeneration of the levator muscle or thinning of aponeurosis with advancing age may result in **involutional ptosis**. In younger patients, repeated manipulation of upper eyelid during contact lens wear (both hard and soft lenses²⁸) may cause disinsertion of levator aponeurosis. Aponeurotic ptosis occurs frequently after ocular surgery (cataract surgery⁶ and glaucoma filtration surgery²⁹) and also following trauma. A variety of mechanisms may be responsible following surgery, such as direct injury to the aponeurosis, stretching or damage from postoperative swelling, use of rigid eyelid speculum, myotoxic effects of local anesthetics used, or damage to the levator muscle from bridle sutures.³⁰

The ptosis could be unilateral or bilateral and vary in severity. Either raised or absent upper lid crease due to slackened attachments of the aponeurosis to the tarsal plate and orbicularis oculi may be seen.³¹ There is also thinning of the upper eyelid and a deep superior eyelid sulcus. The levator function is essentially normal. There may be ptosis in primary gaze, but it typically worsens in down-gaze and obstructs vision, especially when reading. The clinical presentation includes a history of increasing ptosis towards the end of the day because of fatigue of the Muller's muscle and may lead to the erroneous diagnosis of myasthenia.

Other causes of aponeurotic ptosis include eyelid edema from infection or allergy, blepharochalasis, pregnancy, chronic use of topical steroids, and frequent lid rubbing.

Non-isolated acquired ptosis

- a. **Neurogenic ptosis** is rare but requires further evaluation when present to rule out the possibility of a severe underlying neurological disorder. Neurogenic ptosis usually results from either oculomotor nerve palsy or Horner's syndrome. Rarely, central nervous system abnormalities,³² such as unilateral or bilateral cortical ptosis and paradoxical supranuclear inhibition of the levator muscle, may lead to neurogenic ptosis. Complete oculomotor nerve palsy is recognizable when the patient has a combination of profound ptosis, ophthalmoplegia, diplopia, and a mid-dilated and poorly reactive pupil.

Disorders of the oculomotor nucleus usually present with bilateral ptosis. Signs of mesencephalic dysfunction may also be present. The disorder may be congenital (aplasia or dysplasia of the nucleus) or acquired (ischemia, inflammation, infiltration, compression, and metabolic and toxic processes).

Disorders of the oculomotor nerve may occur anywhere from its origin in the oculomotor nucleus in the midbrain to its peripheral innervations of the extraocular muscles within the orbits.³³ The pathology may result from trauma, tumors, aneurysms, vasculopathy as in diabetes, multiple sclerosis, and infection. Fascicular lesions present with signs of unilateral, partial, or complete third nerve palsy often associated with contralateral hemiplegia, cerebellar ataxia, rubral tremor, or signs of midbrain dysfunction. Ptosis in such fascicular lesions will be unilateral, of variable severity, associated with weakness of other extraocular muscles, and pupil involvement. The ocular findings may be similar in a peripheral lesion of the oculomotor nerve. Often in peripheral lesions, ptosis may be isolated or may precede other signs of third nerve dysfunction. In peripheral oculomotor nerve dysfunction caused by aneurysms, accompanying symptoms of headache may be present. Lid synkinesis may occur from aberrant regeneration following oculomotor nerve palsy. Vasculopathic third nerve palsies usually spare the pupil and do not result in aberrant regeneration.

- b. The **acquired form of Horner's syndrome** occurs secondary to trauma, neoplastic insult, stroke, or vascular disease of the sympathetic pathway. The pathology may involve

the hypothalamus, the preganglionic, or the post-ganglionic sympathetic pathway. Patients present with ipsilateral mild ptosis, ipsilateral miosis, and anhidrosis. The acquired form can be distinguished from the congenital form by the absence of iris and areola hypopigmentation.

- c. **Myopathic ptosis** (acquired) results from any pathology that affects the levator muscle. This is usually seen in mitochondrial myopathies, such as **Chronic Progressive External Ophthalmoplegia (CPEO)**, and other muscular disorders, such as **Oculopharyngeal Muscular Dystrophy (OPMD)** and **Myotonic Dystrophy (MD)**. In a retrospective study by Wong *et al.*, CPEO was found to comprise 43 percent of the myogenic ptosis, along with 18 percent for OPMD, and 18 percent for MD.³⁴ Disorders of mitochondrial function may involve the extraocular and eyelid muscles producing CPEO. This condition presents with bilateral slowly progressive symmetric ptosis, orbicularis oculi weakness that prevents complete closure of the eyes, and impaired eye movements.³⁵ Since all eye movements are reduced equally, diplopia may not occur or is typically a late manifestation. Patients with ptosis caused by mitochondrial myopathies may present at any age. These patients frequently present late in life. Excessive forehead wrinkling and chin lift to facilitate vision may be observed in these patients. In the **Kearns-Sayre variant of mitochondrial myopathy**, patients may present with CPEO, along with retinitis pigmentosa and abnormal cardiac conduction, beginning in their first or second decade. **Oculopharyngeal Muscular Dystrophy** is a rare, autosomal dominant myopathy that is characterized by late-onset (usually after 45 years old) ptosis, progressive tongue atrophy, dysphasia, dysarthria, and proximal lower extremity weakness.³⁵ **Myotonic Dystrophy** is another autosomal dominant disorder with clinical findings of ptosis and muscle dystrophy of the face, jaw, and neck.²⁶ Other associated abnormalities include ophthalmoparesis, cataract, frontal balding, cardiac conduction defects, and variable intellectual impairment. The levator muscle may also be damaged by a variety of inflammatory (sarcoidosis), infiltrative (lymphoid), or ischemic processes affecting the orbit.
- d. **Neuromuscular ptosis** — in myasthenia gravis, ptosis and diplopia are frequently the initial presenting symptoms of both the ocular and the generalized forms of the disease.³⁶ This is an autoimmune disorder whereby antibodies block, alter, or destroy the postganglionic acetylcholine receptors in the neuromuscular junction of skeletal muscles, thereby preventing muscle contraction. The ptosis may be unilateral or bilateral and frequently asymmetric. It may be isolated or associated with varying degrees of ophthalmoplegia, resulting in coexisting diplopia. The hallmark feature of myasthenia gravis is the fluctuating weakness and fatigability of the muscles, typically worsening with activity. Exaggerated ptosis with sustained upward gaze for at least 30 seconds, positive Cogan's upper eyelid-twitch sign, orbicularis oculi fatigue causing weakness of eye closure, and reversal of ptosis with rest and local application of ice pack may be appreciated. However, a similar tendency to fatigue may also be seen in aponeurotic ptosis and oculomotor nerve palsy.
- e. **Mechanical ptosis** occurs as a result of excessive weight, usually from a neoplasm on the upper eyelid, making it too heavy for the levator muscle to perform its function. The most common causes are benign or malignant neoplasm of the eyelid, such as a hemangioma, chalazion, neurofibroma, and dermoid cyst with greater ptosis occurring in the area of the mass. Cicatricial changes in the tarsal conjunctiva and superior fornix following trachoma may result in a restrictive type of ptosis. Blepharochalasis is a rare condition of unknown etiology that affects young people and is usually hereditary. It manifests with repeated transient attacks of eyelid edema and erythema that starts around puberty and leads to ptosis during the attacks.³⁷ After the attacks, permanent changes may develop in the lid, such as thinning, wrinkling, and discoloration of skin or dehiscence of aponeurosis also leading to ptosis. Ptosis can also be seen following enucleation because the absence of support to the levator by the globe permits the lid to droop.³⁵⁻³⁸ Entrapment of the levator in orbital fracture or encroachment by an orbital foreign body may also mechanically interfere with the function of the levator leading to ptosis.
- f. **Neurotoxic ptosis** is a classic symptom of envenomation³⁹ by elapids such as cobras, kraits, etc. Neurotoxic ptosis is a precursor to respiratory failure and eventual suffocation caused by complete paralysis of the thoracic diaphragm. Urgent medical intervention is, therefore, required.
- g. **Pseudoptosis** is a form of ptosis that occurs due to abnormalities other than those found in the eyelid elevators. Pseudoptosis may be found on the side of the eye that is abnormal in size, shape, or position; for example, anophthalmos, microphthalmos, and phthisis bulbi. Pseudoptosis has also been described in dermatochalasis, where excessively loose upper eyelid skin appears to overhang the lid margin and simulate ptosis.⁴⁰ Pseudoptosis may be seen in the contralateral normal eye of persons with unilateral lid retraction and proptosis from thyroid ophthalmopathy, as well as those with fixating hypertropic eye. Persons with down-gaze palsy and those with narrowed lid fissure following previous facial nerve palsy may also present with pseudoptosis.
- h. **Traumatic ptosis** — Traumatic blepharoptosis may develop following an eyelid injury with ensuing damage to the lid elevator muscles, the levator aponeurosis or disruption of the neural input. Hence, eyelid trauma may lead to a myogenic, aponeurotic or a neurogenic ptosis.
- i. **Brow ptosis** is fairly common in the general population, especially in those above 50 years and those with dermatochalasis⁴¹ (a condition with redundant skin and muscle of the eyelid). Brow ptosis is a condition in which the eyebrow droops or sags. Laxity in the forehead muscles allows the eyebrows to fall. With this, the skin below the eyebrow also falls into the upper eyelid space, making the

upper eyelid fold heavy. With this encroachment of tissue onto the upper eyelid, frontalis muscle acts to lift the eyebrows, accounting for the forehead lines associated with eyebrow ptosis. Brow ptosis may be asymmetric. Gravity and age as well as genetic inclination and physiognomy all play a part in the etiology of brow ptosis. Aging changes in the eyelids and face result from a loss of tone in the various layers underlying the skin. Patients report symptoms of ocular fatigue secondary to the continuous action of the frontalis muscle. They may also complain of limitation in superior visual field due to overhanging skin.⁴⁰ (See Appendix 3 - Types of acquired ptosis.)

medical therapy, such as with cholinesterase inhibitors, corticosteroids, azathioprine, and diaminopyridine.

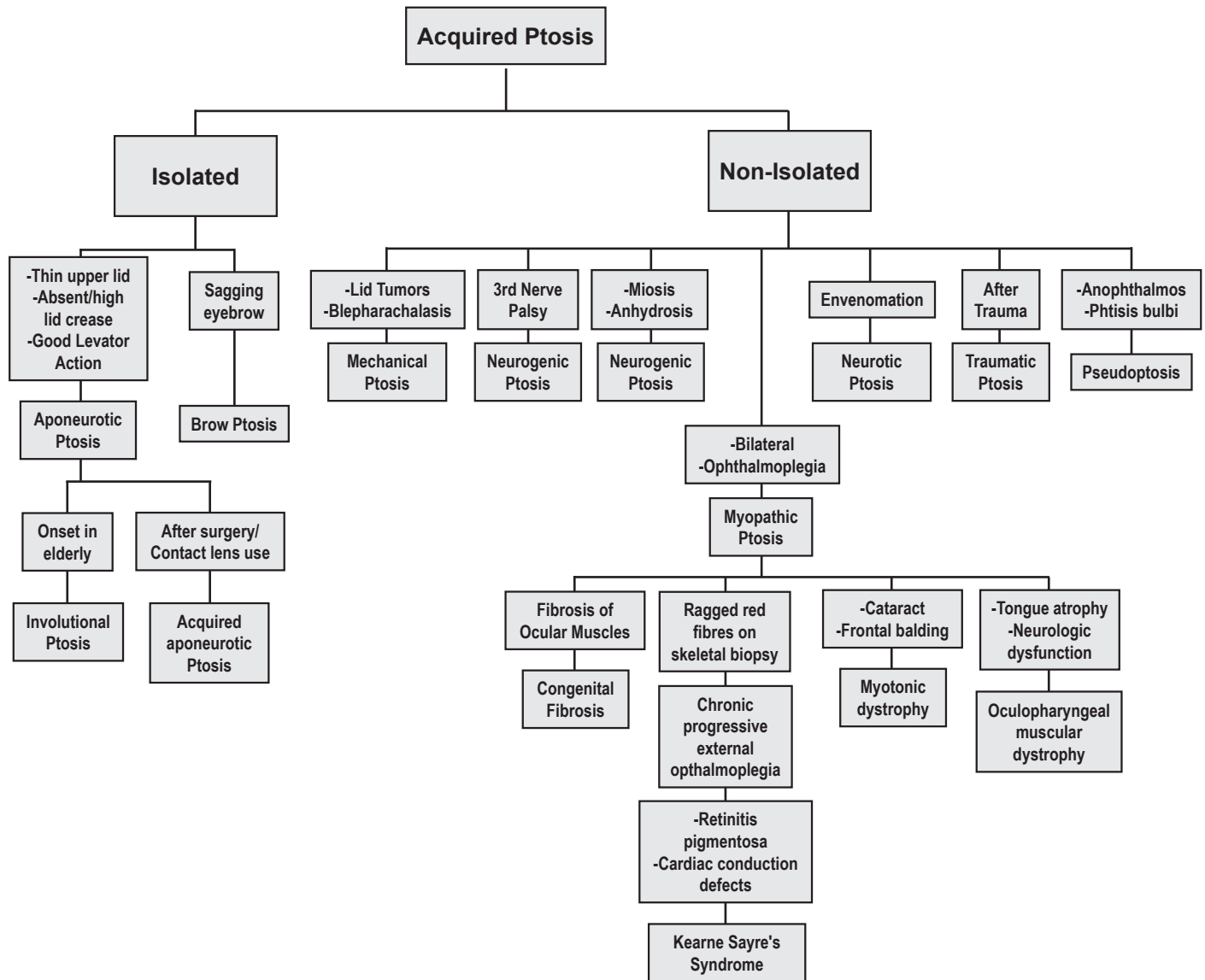
Ptosis usually does not improve over time and nearly always requires corrective surgery. In mild cases of congenital ptosis observation is sufficient, if no signs of amblyopia, strabismus, and abnormal head posture are present. However, patients should be monitored every 3-12 months for signs of amblyopia and for their head posture. External photographs can be helpful in monitoring patients. If the patient acquires a chin-up posture due to the worsening of ptosis, surgery may be indicated. If the child has strabismus together with ptosis, surgery to correct strabismus is usually done prior to ptosis surgery. Following surgery for ptosis, amblyopia treatment should continue. Usually surgery for ptosis is performed around 3-4 years of age as measurements may be more accurately performed around this time.

Conservative treatment of ptosis include the use of an adhesive *eye-putti*, as in myotonic dystrophy, to affix the upper eyelid to

Treatment

The treatment of ptosis depends on the underlying etiology. Successful treatment of underlying medical problems may sometimes correct the ptosis, as in myasthenia gravis. Cases of ocular or generalized myasthenia gravis are responsive to

Appendix 3: Types of acquired ptosis



the supraorbital structures, or inserting eyelid crutches into eye-glasses as a temporary measure to hold up the eyelid.

Ptosis surgery or blepharoplasty is often indicated for individuals whose ptosis severely obstructs the visual field or interferes

with activities of daily living. However, in many cases, surgery is performed for cosmetic reasons.^{42,43} In adults, the surgery is mostly performed as an outpatient procedure, while the patient is under local anesthesia or has been given a mild sedative; however, general anesthesia may be necessary for pediatric patients.

Appendix 4: Synopsis of upper eyelid ptosis

History	<p>History of Present Illness: Onset, duration, involvement of one or both eyes (simultaneously or one followed by another), variability, progression, severity, inspect old photographs</p> <p>Associated Symptoms: Diplopia, odyndophagia, peripheral muscle weakness, cardiac symptoms, night blindness</p> <p>Past Ocular History: Trauma, surgeries, contact lens use, ocular pathologies, allergies, eyelid edema, dry eyes</p> <p>Family History: Ptosis, musculoskeletal diseases</p>
Comprehensive Ophthalmic Examination	<p>Inspection: Eyebrow position, head posture, eyelid masses, inflammation, heterochromia, proptosis</p> <p>Palpation: Eyelids, orbital rim</p> <p>Best corrected Visual Acuity: In infants, make sure infant can fix and follow light with each eye</p> <p>Pupillary Examination: Pupillary size, reaction</p> <p>Cycloplegic Refraction</p> <p>Strabismus Evaluation</p> <p>Extraocular Muscles Motility: Note paresis, paralysis of muscles</p> <p>Funduscopy Examination: Abnormal retinal pigmentation</p> <p>Exophthalmometer (if proptosis is present)</p> <p>Bell and Jaw-Winking Phenomena Evaluation</p> <p>Corneal Sensitivity</p> <p>Slit-Lamp Examination: With fluorescein stain to examine cornea, tear meniscus, tear break-up time. Note keratopathy, heterochromia iridis</p> <p>Schirmer's Test</p>
Eyelid Measurements	<p>Palpebral Fissure Height (PF): May be < 9 – 12 mm in patients with ptosis, note lid position with down-gaze</p> <p>Marginal Reflex Distance (MRD): Ptosis defined as MRD < 2 mm or an asymmetry of > 2 mm between eyes</p> <p>Levator Function: 0 – 5 mm (poor), 6 – 11 mm (fair), > 12 mm (good)</p> <p>Upper Eyelid Crease: Absent crease or > 7 – 8 mm in males and > 9 – 10 mm in females</p> <p>Lid position in down-gaze</p>
Additional Tests	<p>Suspected Myasthenia Gravis: Serum assay for acetylcholine receptor antibodies, edrophonium chloride test, ice test, electromyography</p> <p>Suspected Mitochondrial Myopathies: Skeletal muscle biopsy with trichrome staining, electrocardiography</p> <p>Suspected Müller's Muscle Dysfunction: Phenylephrine or apraclonidine instilled under eyelid to test function</p> <p>Suspected Neurological Etiologies: Imaging of brain, orbits, cerebrovascular system</p>
Conservative Management	<p>Mild Congenital Ptosis: Routine monitoring every 3-12 months for signs of amblyopia, strabismus, and abnormal head postures; surgery may be indicated if these signs are present</p> <p>Ptosis in Ocular or Generalized Myasthenia Gravis: Responsive to medical therapy (cholinesterase inhibitors, corticosteroids, azathioprin, and daminopyridine)</p> <p>Other Temporizing Measures: Use of adhesive <i>eye-putti</i> to affix upper eyelid to supraorbital structures, inserting eyelid crutches into eyeglasses</p>
Surgical Management	<p>Congenital Ptosis with Strabismus: Surgical correction of strabismus prior to ptosis surgery; Ptosis correction usually performed at 3 – 4 years of age</p> <p>Mild Ptosis, Levator Function > 10 mm: Müller's muscle-conjunctival resection or Fasanella-Servat procedure</p> <p>Moderate Ptosis, Levator Function 5 – 10 mm: Levator palpebrae superioris resection</p> <p>Severe Ptosis, Levator Function < 5 mm: Brow-frontalis suspension</p>

Conclusion

Ptosis is a frequently encountered eyelid malposition in patients of all ages. Blepharoptosis results from underaction of the eyelid protractors relative to the eyelid retractors, causing the eyelid to be lower than its normal anatomic position. It can occur in isolation or is one of the early presenting symptoms of a serious underlying disease. In cases of uncorrected congenital ptosis, the pediatric patient may develop amblyopia and suffer lifelong visual impairments. Also, in young children and teenagers, an abnormal eyelid position may have negative psychosocial effects. In adults with uncorrected ptosis, there may be a reduction of the visual field, which impedes the performance of activities of daily living. Thus, early diagnosis and appropriate management of ptosis is essential and critical to avoid the consequences as discussed. The synopsis of upper eyelid ptosis in Appendix 4 may assist medical practitioners in the clinical evaluation of patients with ptosis presenting to their clinic and in making appropriate management decisions.

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