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Pediatric Ptosis

Developed by Arshdeep Singh Marwaha and Dr. Jane Gardiner for PedsCases.com.
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Introduction:

[Hi! My name is Arshdeep and I am a 3rd medical student at the University of British Columbia in Vancouver, Canada. The topic of this podcast is ptosis in children. We will start with a clinical vignette, some background information regarding what ptosis is, and a brief review of relevant anatomical structures of the eye. The second half of this talk will be about an approach to pediatric ptosis whenever you encounter this problem. Without further ado, lets jump right in!]

Clinical Vignette

It's the first day of your pediatric ophthalmology elective and your first consult is a 7 month-old female patient referred to the Ophthalmologist by her pediatrician for evaluation of a possible left upper eyelid ptosis. The parents tell you that the eyelid has been "droopy" since birth and has been getting worse progressively. The baby was born at term without any complications during delivery and there was no history of trauma. The referral information indicates that she has meet all her developmental milestones on time. She is not taking any medications, has no known drug allergies, and no family history of ptosis.

Upon general examination, the patient appeared well. She was hemodynamically stable, no obvious skin changes noted, and no facial deformities seen. The child was in the 50th percentile for height, weight, and head circumference. The visual acuity exam, she fixes and follows bilaterally. However, you notice that with the right eye she fixes more with the right eye than left. Pupillary exam was unremarkable. You next inspect the eye and notice the left upper eyelid is lower than you would expect. There was no gaze preference or palpable masses on the eyelid or periorbital area. Your preceptor tells you the anterior segment exam and dilated fundus exam were normal. External measurements showed that the left palpebral fissure was 5 mm and 9mm on the right. Levator function test showed poor excursion on the left.

Your preceptor then asks what you think the cause of her droopy eyelid is and what should be done next.

[Let's have a quick review!]
Background information

What is Ptosis?

A drooping eyelid is also called ptosis or blepharoptosis. This is a condition in which the eye lid falls to a position lower than it normally is; normal position of the eye lid is around 0.5-1.0mm below the superior limbus, which is the superior border between the cornea and sclera. As a result, it can cover all or part of the pupil, limiting the superior visual field first and then affecting central vision.

Ptosis can affect one or both eyes and does not demonstrate a predilection based on ethnicity or biological sex. Ptosis can be congenital or acquired. Congenital ptosis is often isolated and typically not associated with a systemic disease. If an eyelid droop develops after a few weeks of life, there may be an underlying disease to consider which would require further neurological and physical examination.

Fortunately, treatment of this condition can allow for normal visual development as well as cosmetic appearance – therefore early recognition of ptosis is imperative!

Muscle of the eye lid and nerve innervation

[Before continuing our discussion, let's review the muscles and nerves involved in the movement of our eyelids.]

- The Facial Nerve (CNVII) innervates the orbicularis oculi, frontalis, procerus, and corrugator supercilii muscles, and supports eyelid *protraction or closure.*
- Oculomotor nerve (CN III) innervates the main upper eyelid *retractor*, the levator palpebrae superiorus, via its superior branch. The oculomotor nerve also innervates the inferior rectus muscle, causing lower lid retraction during downward gaze.
    - Important structures to know about in the context of ptosis are the levator palpebrae superiorus muscle and the levator aponeurosis. The levator palpebrae superiorus is connected to a thick layer of connective tissue on the upper eye lid called tarsus via the levator aponeurosis fascial tissue. The muscle pulls on the connective tissue to lift the eyelid.
- Sympathetic fibers assist with both upper and lower lid retraction by innervating the superior and inferior tarsal muscles, respectively. This only contributes to about 1 mm of upper eyelid elevation

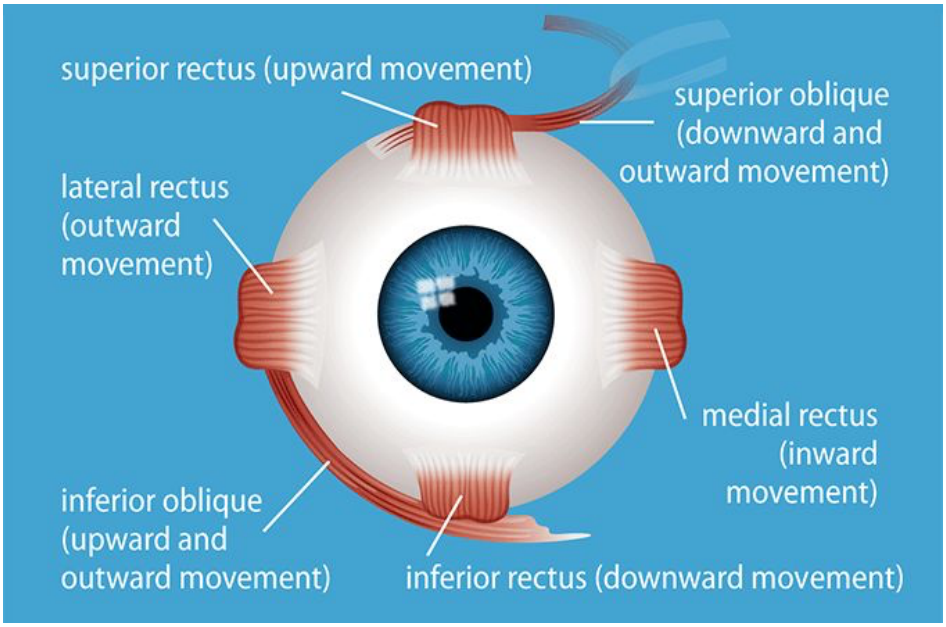

Image from: https://www.allaboutvision.com/eye-care/eye-anatomy/eye-muscles/

Types of Ptosis

The underlying cause of ptosis can be due to involutional (aponeurotic), myogenic, neurogenic, or mechanical issues. We'll discuss each of these in turn to explain what they mean. For each cause, it is important to differentiate whether ptosis is unilateral or bilateral.

1. Involutional (aponeurotic)

   Aponeurotic ptosis is the most common cause of acquired ptosis, and the most common cause of ptosis overall. It occurs when there is stretching or disinsertion of the levator aponeurosis, thereby inhibiting the function of the levator muscle.

   Though this is a common problem in adults, congenital aponeurotic ptosis is relatively uncommon. Most pediatric cases of involutional ptosis occurring at birth are due to trauma during delivery. Therefore, it is important during history to ask about important risk factors, including forceps delivery, vacuum extraction, traumatic fetal rotation, and shoulder dystocia.

2. Myogenic

   Myogenic ptosis can either be congenital or acquired. The most common cause of myogenic ptosis is congenital ptosis. It is characterized by limited eyelid elevation due to weakness of the levator muscle. Often, congenital ptosis has an idiopathic etiology. However, it can also be caused by systemic disorders

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including myotonic dystrophy, oculopharyngeal muscular dystrophy (OPMD), and chronic external ophthalmoplegia (CPEO). These disorders are a result of inherited genetic mutations. In congenital myogenic ptosis, the levator muscle in patients with ptosis are dystrophic. The muscle and the aponeurosis appear to be infiltrated by fat and fibrotic tissue.

3. Neurogenic

   Neurogenic ptosis results from damage to the oculomotor nerve or sympathetic nerve fibers. Causes of neurogenic ptosis in children include third nerve palsy (pupil may be small and non-reactive), Horner syndrome (ipsilateral ptosis, miosis, & anhidrosis), and myasthenia gravis.

4. Mechanical Issues

   This occurs when the eyelid is too heavy for the levator muscle to elevate it. This can happen due to conditions such as blepharochalasis, orbital fat prolapse and eyelid tumors including disorders such as neurofibromas in neurofibromatosis type 1. The increased weight on the eyelid also causes the eyelid skin to stretch.

Why is ptosis a significant problem for children?

Babies need good vision to learn and grow! Having ptosis puts a child at risk for delayed vision development, which can have downstream consequences such as delays in reaching age-specific developmental milestones.

If the child's eyelid droops so much that it blocks vision, amblyopia (also called "lazy eye") can develop. One eye will have better vision than the other. The open eye will develop vision due to the normal visual input, while the ptotic eye will not develop the proper visual pathways and it will become lazy. This is called deprivational amblyopia, as the eye is being deprived or normal visually input. A child with ptosis can also have astigmatism, causing blurry vision and is also a risk of poor visual input and amblyopia. This is called anisometropic amblyopia. Lastly, the child may also develop strabismus (misaligned/crossed eyes) because the vision is not developing normally which predisposes to the eye drifting.

An approach to pediatric ptosis

[Given the substantial impact that ptosis and other eye disorders can have on a child, examination of the eye is integral during routine well-child visits. The Rourke Baby Record, which outlines routine developmental surveillance in children, includes a section on vision. We will move our discussion now to the development of an approach

to ptosis which will be useful for you during your pediatric, family practice or emergency medicine rotations.]

Signs and Symptoms

First let's review some of the signs and symptoms. A child with ptosis often has no symptoms, particularly if it is mild. If they do have symptoms, parents may report seeing their child tilt their head back to see better, complain of difficulties seeing things, or bumping into objects which are over hanging. They may also have gross motor delays (i.e., crawling/walking) due to the importance of vision in movement. Lastly, depending on their age, they may complain of blurred vision, double vision, or reduced vision.

History

All patients presenting with droopy eyelids need a comprehensive medical history, with a special focus on birth history and family history. It is also important to complete a detailed review of systems. In the context of pediatrics, also ask for developmental milestones as well as labor & delivery history.

Some important aspects to probe with the associated reasons include:

- History of red eye, watering, discharge, or pain while blinking may suggest a corneal pathology.
- Any recent medications taken or known allergies?
    - Allergic reactions can cause the eyelid to become edematous and droopy.
- Have either of the parents or child's siblings had the same problem? This may suggest a congenital or inherited cause.
- Has there been a relevant history of trauma to the head or face?
    - Orbital wall fractures can result in pseudoptosis. Trauma can also cause third nerve palsy or a laceration of the levator muscle
- Has the child been diagnosed with cancer?
- Have the parents noted a difference in pupil size, periodic ptosis, or cross eyes?
    - Ipsilateral or same side constriction of the pupil (miosis) and ptosis are suggestive of Horner's syndrome. This is a condition caused by a disruption in the nerve pathway between the brain and the face as well as eye on one side. Possible causes include: stroke, tumor, or spinal cord injury.
    - Periodic ptosis and strabismus are suggestive of myasthenia gravis. This is an auto-immune disease attacking postsynaptic acetylcholine receptors, which disrupts neuromuscular transmission.
- Has the child been diagnosed with a cancer?
    - Metastatic or primary orbital tumors may cause a mass effect, leading to malposition of the eyelid.

Differential Diagnosis

The differential diagnosis for ptosis can be quite broad. Broadly, Ptosis may be due to involutional (aponeurotic), myogenic, neurogenic, or mechanical issues. It is important to determine whether ptosis is unilateral or bilateral. Some of the specific causes include:

- Corneal abrasion
- Corneal Foreign body
- Conjunctivitis (allergic/infectious)
- Horner Syndrome
- OPMD
- CPEO
- Blepharochalasis
- Myasthenia Gravis

Physical Examination

Equipment needed includes: Tumbling E-chart, Snellen chart, pen light, slit-lamp, and measuring tape.

As with any clinical encounter, it is important to start off with a general exam. In this context, a standard head-to-toe pediatric examination should be conducted. Head circumference, weight, and height should also be plotted on the WHO growth charts.

- A general observation should be made regarding the position of the eyelids as well as the direction in which the eye is facing. The upper eyelid crease is often absent in patients with congenital ptosis. Also note any signs of trauma or infection. In a general physician or pediatricians office, visual acuity, examination of the pupils, and observing extraocular muscle function can be performed.
- Visual Acuity
    - Fixation preference for preverbal children
    - Tumbling E-chart (3-5 years old)
    - Snellen Chart (6 years and older)
    - Central and peripheral vision test
- Examination of pupils
    - Note the pupil size and color. Leukocoria upon illumination of the fundus may indicate a problem with the lens (ex. cataracts), retina (ex. retinoblastoma), or vitreous (ex. Hemorrhage). It is also important to observe if the pupils are the same size bilaterally?
    - Illicit and observe the presence of a direct pupillary reflex and consensual pupillary reflex
    - Perform a swinging light test to examine eyes for relative afferent pupillary defect.
- Extraocular muscles

    - There may be deficiencies in adduction, elevation, or depression of the eyes. What is the alignment? Are the eyes straight or is there a strabismus?
    - This may be difficult to do with an infant or child. You can try using toys to illicit eye movements in the baby

An ophthalmologist may additionally perform the following tests:

- Slit lamp examination
    - Examine the conjunctiva, sclera, eyelid margins for signs of infections (erythema, edema, pus).
    - Observe the cornea for signs of abrasion or erosions. An ophthalmologist can use fluorescein dye to make surface abrasions easier to spot.
- Marginal reflex distance
    - Measure the distance between the upper eyelid margin and the corneal light reflex. This distance is typically 4-5 mm.
- Interpalpebral fissure width
    - Measure the distance between the inner and outer canthi of the eye. This distance is approximately 9 mm. This distance changes based on age. It is therefore important to measure bilaterally.
- Eyelid excursion or levator function
    - Place the thumb on the superior orbital rim to eliminate any contribution of the frontalis muscle on eyelid elevation.
    - Measure the distance between the upper and lower eyelid during down and upward gaze.
- Examination of the fundus
    - Is the red reflex present?
    - In older children, it may be possible to look at the fundus to observe the optic cup to disc ratio,

Who else should be involved in the care?

Referral to an ophthalmologist should be made for a detailed ophthalmic evaluation and determining the etiology of ptosis. The referral to the ophthalmologist should not be delayed, especially when you think the ptosis is interfering with visual development. You may also want to consider a consultation with pediatric neurology if a neurologic etiology is suspected (ex. Horner's Syndrome, Myasthenia gravis).

How is ptosis treated?

If there is a suspected contributing cause such as a systemic condition (i.e. neurologic), or trauma, these need to be addressed.

It is important to note that not all ptosis needs to be treated. Reasons to treat include for vision and for cosmetic reasons. If the ptosis is not impacting vision, treatment can be deferred.

Non-transient forms of Ptosis are treated by surgery of the eye lid. Surgery should be performed as soon as there are signs that there is interference with the child's visual development such as amblyopia or ocular torticollis. These decisions will be made in conjunction with an ophthalmologist. If intervention is not needed urgently, surgery is typically performed between 3-4 years of age. Delaying surgery is beneficial as it allows for more precise preoperative measurements and reduces the anesthetic risk. The two most commonly used surgeries for congenital ptosis are:

1. Levator resection

   This surgery involves shortening the levator-aponeurosis complex. This can be done through the eyelid skin or via an internal approach through the tarsal conjunctiva. A patient needs to have at least moderate levator function (> 4 mm) for this procedure to work. If the levator is not working, then shortening it cannot make it stronger.

2. Ptosis repair by frontalis sling

   The surgery connects the eyelid to the brown with a sling material and utilizes the power of the frontalis muscle to elevate the eyelid. This procedure is indicated when the levator function is less than 4 mm. It creates an asymmetrical appearance of the lids in downgaze. The eyelid with the sling is help up as the eye looks down.

Post-operative considerations

Close post op surgical monitoring is required to ensure the child does not develop exposure of the cornea and conjunctiva (exposure keratoconjunctivits). This is more common in frontalis slings, where the lid may not be able to close fully. In some cases, it may be necessary to undo part of the surgery. Most cases can be managed with extra lubrication before sleep.

The surgeries typically produce great functional and cosmetic results. Associated conditions that developed secondary to the ptosis such as amblyopia or strabismus, may also need treatment. These will be managed by the ophthalmologist. Approximately 50% of patients will require a repeat surgery.

[Going back to our case…]

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The patient's findings were consistent with unilateral isolated congenital ptosis. Her physical exam and family history were unremarkable, making secondary ptosis unlikely. Given that the patient was symptomatic for developing fixation preference for the right eye, surgical intervention is indicated.

[We hope you enjoyed the case. Feel free to leave any comments or suggestions for next time!]

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