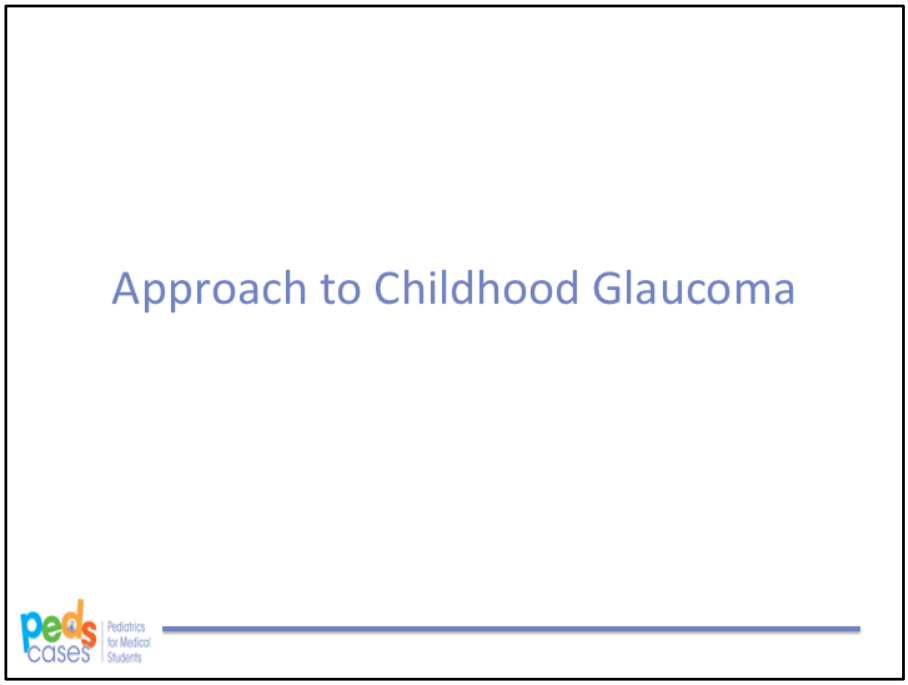
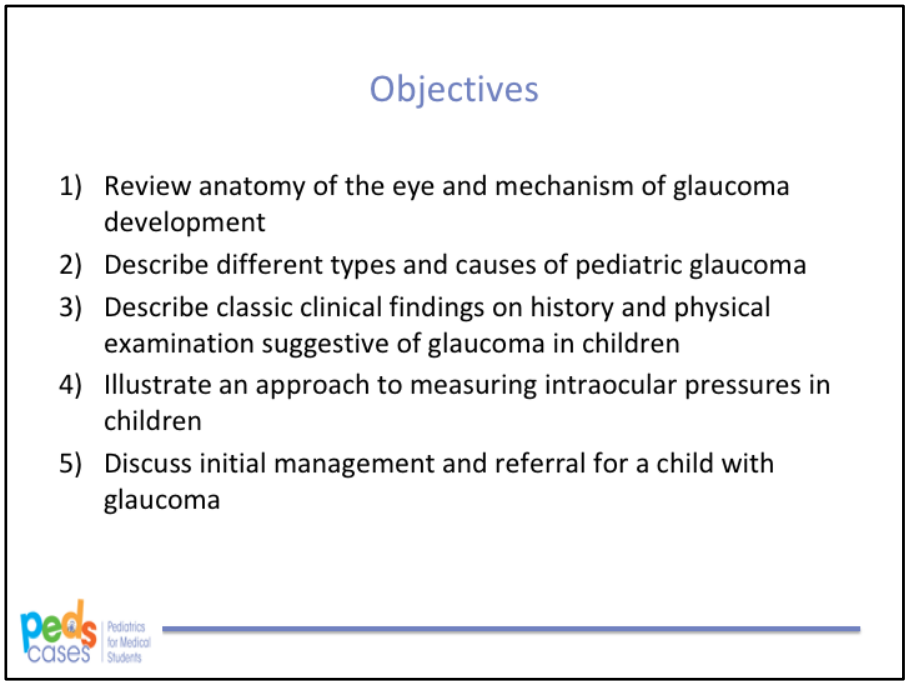
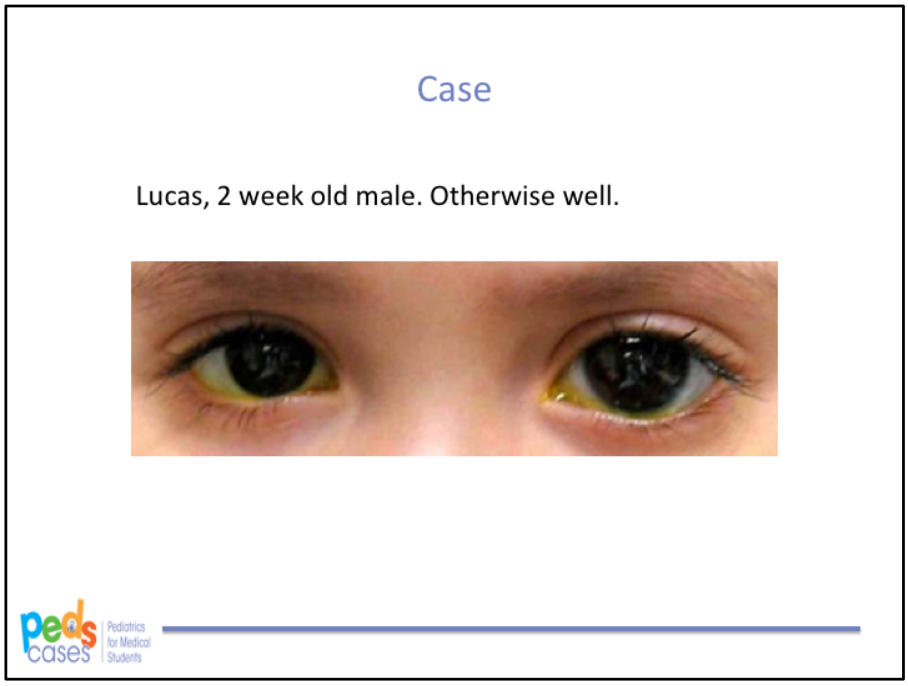



Hi everyone, my name is Jenny Ma. I'm a medical student at the University of Alberta. This podcast will go over an approach to glaucoma in children and was developed in collaboration with Dr. Chris Novak, a pediatric resident at University of Alberta. Glaucoma is a group of diseases characterized by damage to the optic nerve that often occurs when the intraocular pressure is too high. This can result in severe and irreversible vision loss. The vision loss associated with glaucoma usually starts from the periphery so many patients do not notice it until the visual field defect is advanced. Thus it is important for primary care providers and pediatricians to notice the symptoms and signs of glaucoma. Despite similarities to glaucoma in adults, the clinical findings and surgical management of pediatric glaucoma vastly differ.

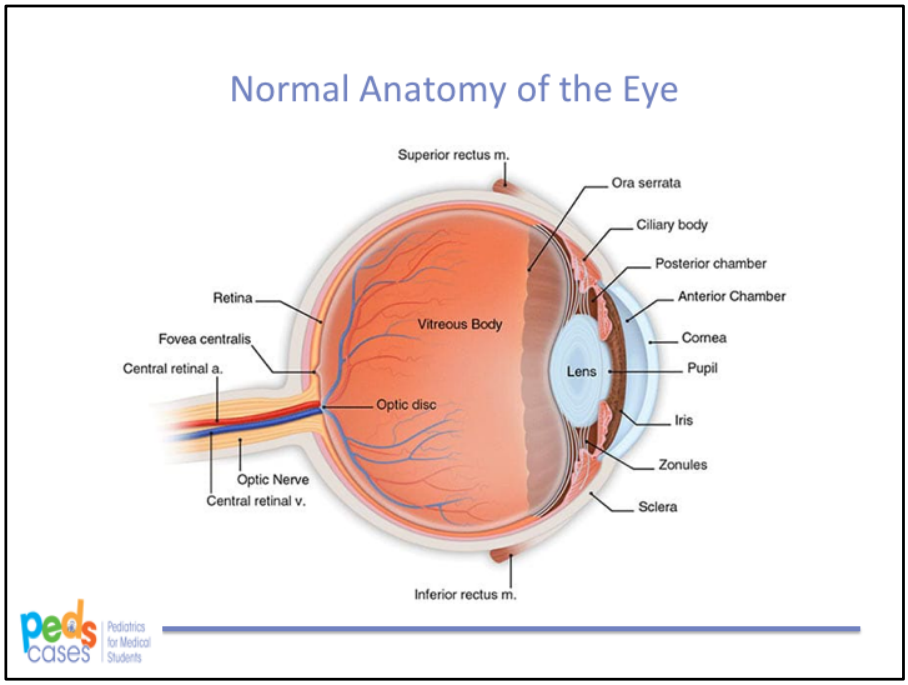



After listening to this podcast, the learner should be able to:

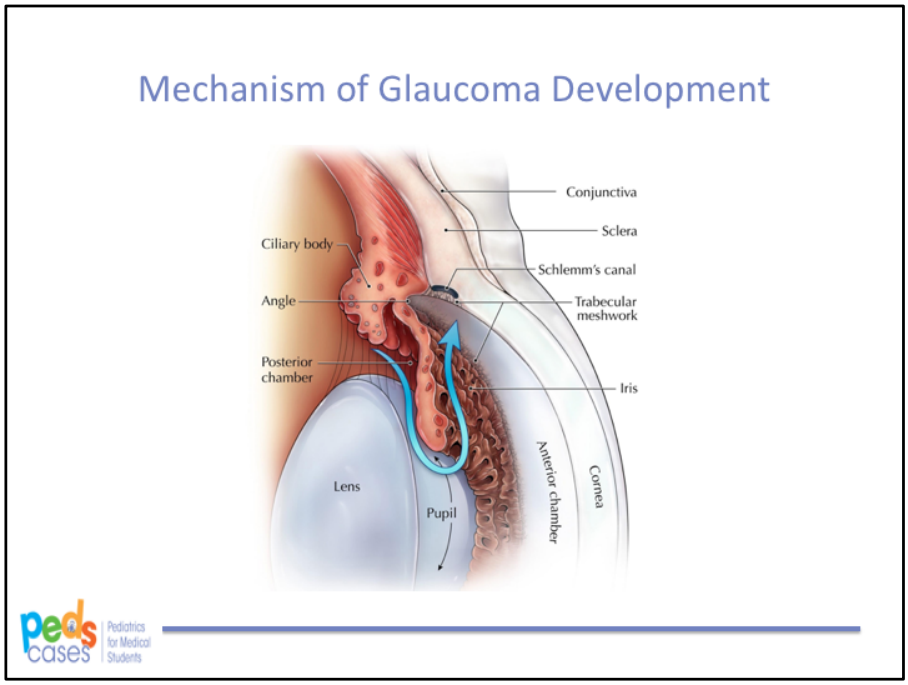
1) Understand the anatomy of the eye and mechanism of glaucoma development
2) Describe different types of pediatric glaucoma
3) Recognize classic clinical findings on history and physical examination suggestive of glaucoma in children
4) Illustrate an approach to measuring intraocular pressures in children
5) Discuss initial management and referral for a child with glaucoma



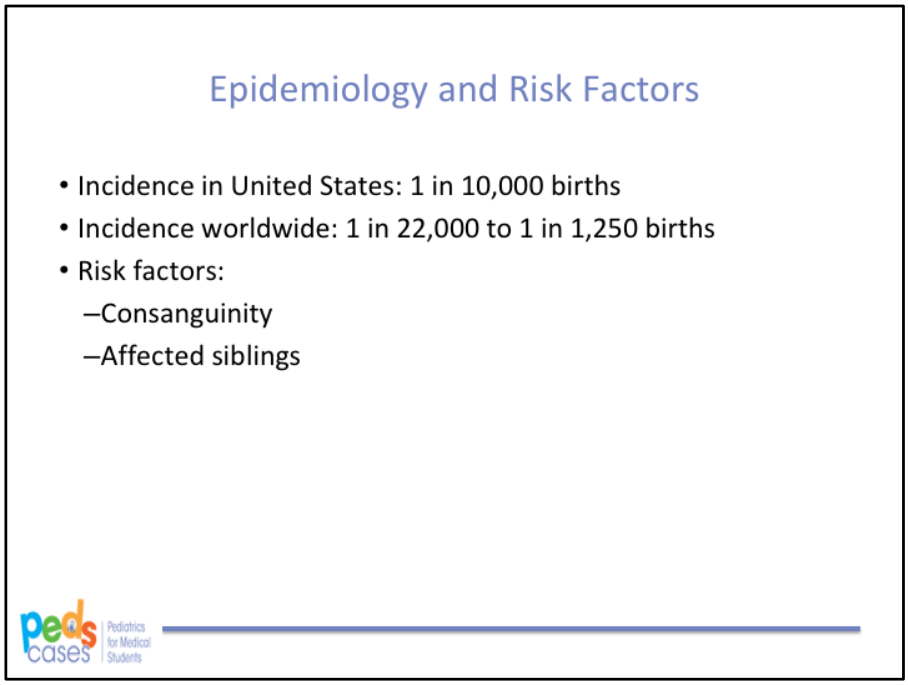

Let's start with a case:
You are working in a community clinic and are scheduled to see Lucas, a 2-week-old male, presenting for his well-baby check. Lucas has been well but his parents are concerned that one of his eyes seems bigger than the other. They have also noticed he is tearing excessively, seems sensitive to light, and does not want to open his eyes much. What is causing these findings and should you be concerned?



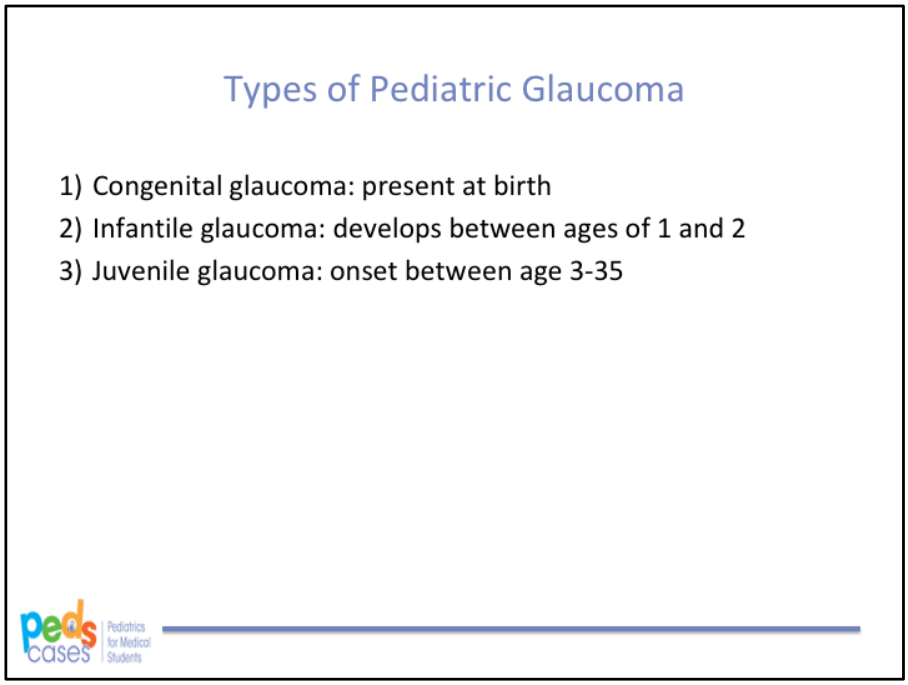

Before talking more about glaucoma, let's first review the anatomy of the eye. There are 3 chambers to the eye: anterior chamber (between the iris and cornea), posterior chamber (between the iris and lens), and vitreous chamber (behind the lens). The "angle" that is often referenced in glaucoma is formed between the iris and cornea. The ciliary body is a sphincter body that surrounds the lens. One of the functions of the ciliary body is to produce aqueous humour that fills the anterior and poster chambers. The aqueous humour travels out through the pupil to the trabecular meshwork and flows out of the eye through Schlemm's canal.



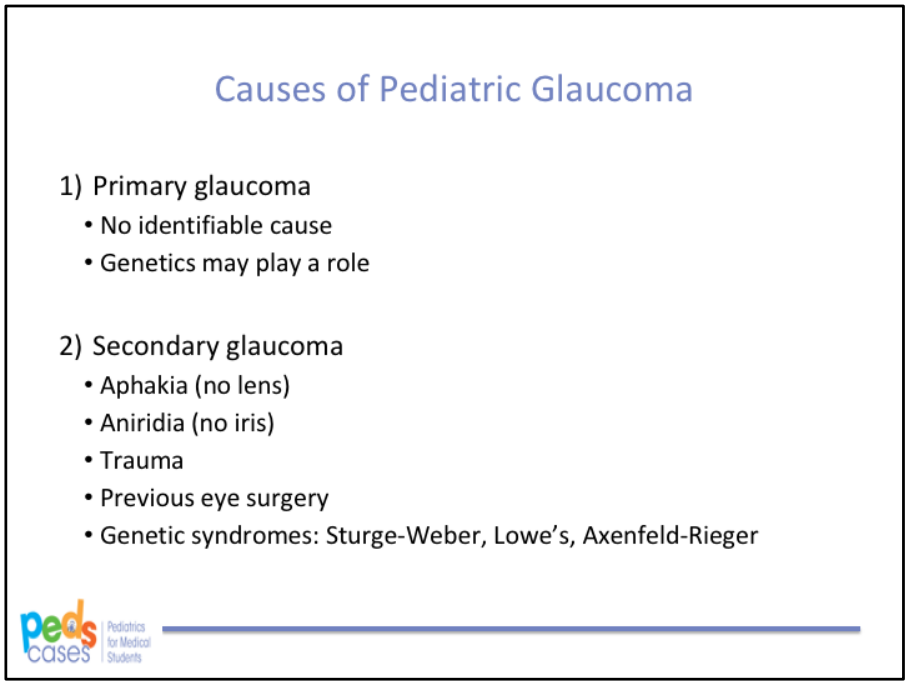

Glaucoma is characterized by damage to the optic nerve and corresponding visual field defects, often associated with elevated intraocular pressures (IOP). Most often, the elevated IOP is due to the eye continually making the fluid it needs, without the ability to sufficiently drain the fluid out. This can be seen with dysfunction of either the trabecular meshwork or Schlemm's canal. When drainage of this fluid is impaired, fluid accumulates within the anterior chamber. Intraocular pressures begin to rise and can lead to the two hallmark features of glaucoma: 1) optic nerve damage and 2) visual field defects.



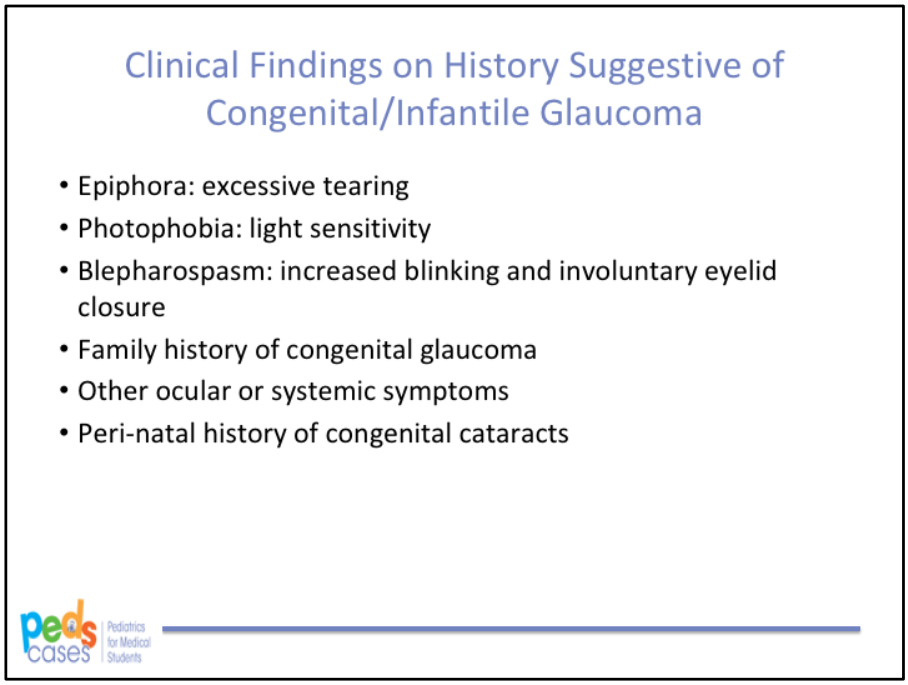

Although glaucoma in adults is a common finding, childhood glaucoma is relatively rare. While worldwide estimates vary, the incidence of primary congenital glaucoma and infantile glaucoma in the United States is around 1 in 10,000 births. Risk factors for glaucoma include consanguinity and previously affected siblings. The risk of congenital glaucoma is around 5% in the second child and increases to 25% with two affected siblings. Also, there is an increased risk of glaucoma in infants and children who have had surgery for congenital cataracts.




One way to classify glaucoma is based on the age of onset. Primary congenital glaucoma presents at birth and is the most common form of childhood glaucoma. This condition is more common in males, typically bilateral, and does not have racial or geographic preference. There is no way to prevent congenital glaucoma so early detection and treatment is key to maintaining vision. Infantile glaucoma develops in a child between the ages of 1 and 2 years. Juvenile glaucoma has an onset between 3-35 years old and tends to develop without any obvious symptoms, similar to adult glaucoma. Patients with juvenile glaucoma often have a family history. The classic signs and symptoms mentioned here are more pertinent to congenital and infantile glaucoma.

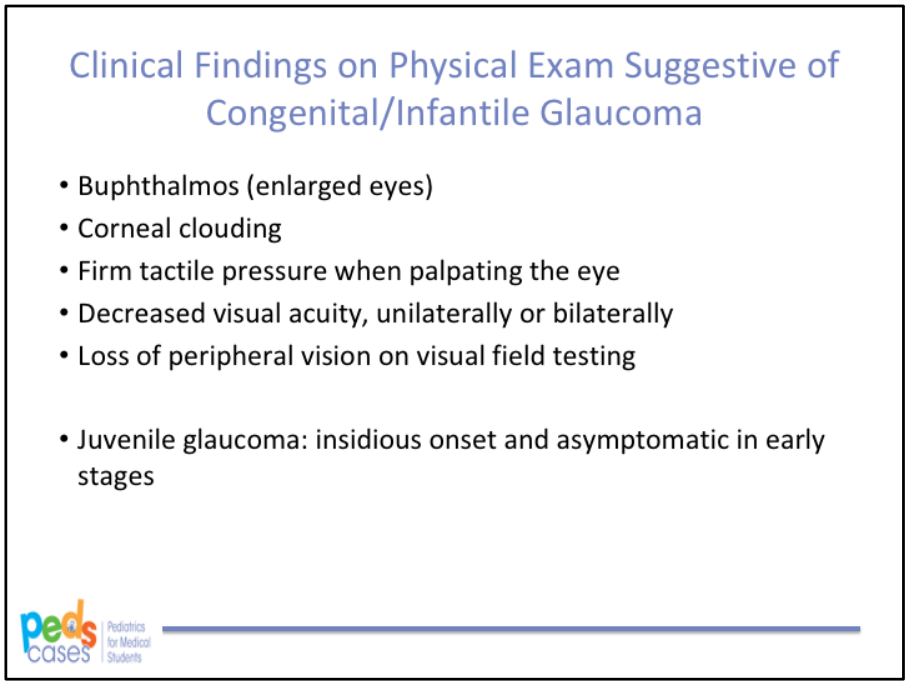



Another way of classifying glaucoma is based on the etiology as primary or secondary. Primary glaucoma is defined as being caused by a primary ophthalmologic cause, with no other identifiable cause or systemic symptoms. Several theories exist for the mechanism of primary congenital glaucoma. These include the formation of a membrane covering the anterior chamber angle, trabecular meshwork obstruction, or developmental arrest of the anterior chamber tissue in utero. Most cases are sporadic but about 10% of primary congenital/infantile glaucoma cases are inherited and there is a higher risk in consanguineous families. Pediatric glaucoma can also be secondary to another condition. Aphakic glaucoma, occurring after pediatric cataract extraction, is the most common form of secondary pediatric glaucoma. Aniridia (the absence of an iris), eye trauma, and previous eye surgery are also associated with a higher incidence of glaucoma. In addition, several rare genetic syndromes are associated with glaucoma such as Sturge-Weber, Lowe's, and Axenfeld-Reiger. Not all patients with these conditions will develop glaucoma but they are at a higher risk so they should be monitored regularly. You should consider Sturge-Weber syndrome in a child born with a large port wine stain on their face, also known as a capillary malformation.




Findings on history that are suggestive of glaucoma include:

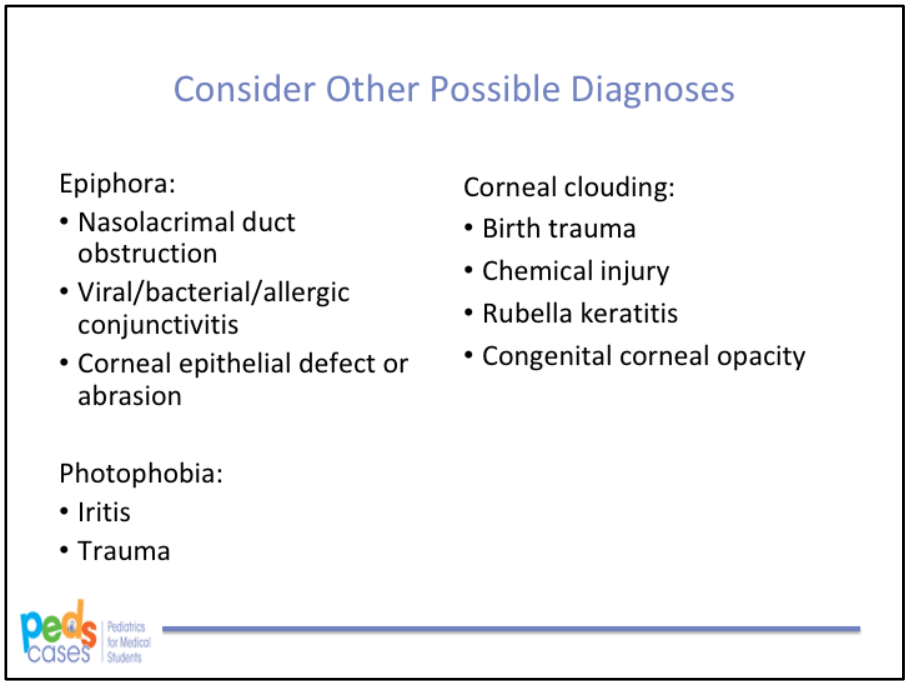
- Epiphora: excessive tearing
- Photophobia: light sensitivity
- Blepharospasm: increased blinking and involuntary eyelid closure
- Family history of congenital glaucoma
- Peri-natal history such as cataract extraction for congenital cataracts




There are a number of findings on physical exam that are suggestive of glaucoma. The following findings can be readily observed by a general physician:

- Buphthalmos (enlarged eyes): the affected eyes can become enlarged because collagen in the sclera and cornea can stretch from increased IOP. This enlargement does not occur in adult glaucoma.
- Firm tactile pressure when palpating the eye
- Corneal clouding: this depends on how acute the pressure elevation is. When there is an acute IOP rise, children will present with corneal clouding, which may also be present at birth. Firm tactile pressure in these cases can be apparent and helpful in differentiating other causes of corneal opacification. However gradually increasing pressure may result in little to no corneal clouding.
- Decreased visual acuity, unilaterally or bilaterally
- Loss of peripheral vision on visual field testing

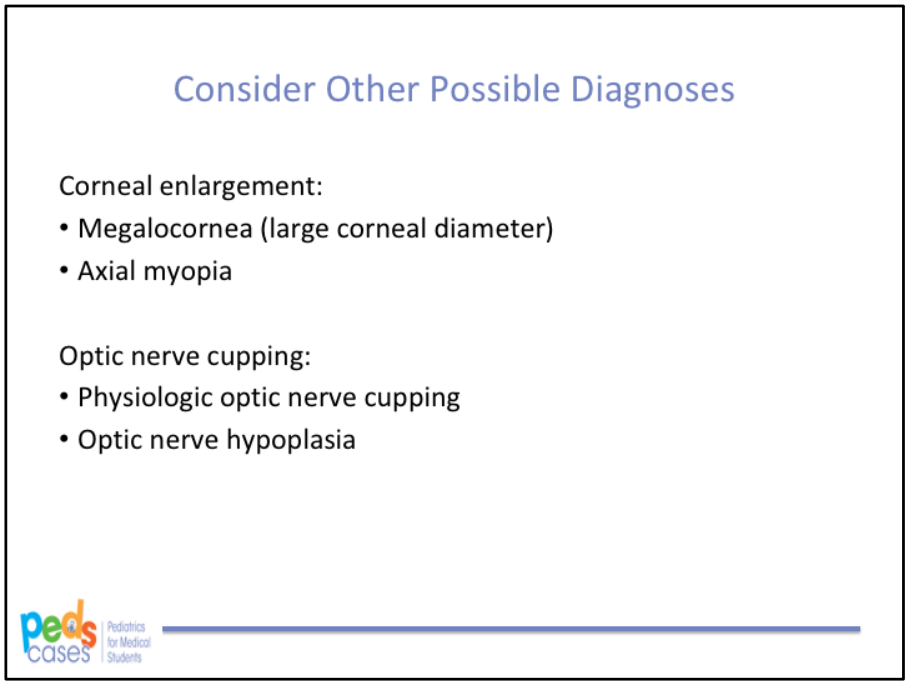
Juvenile glaucoma  has more of an insidious onset similar to glaucoma in adults, and is asymptomatic in the early stages without the clinical findings on history and physical exam mentioned here. Symptoms are rare but may include blurred vision and headaches from elevated IOP. In later stages of the disease, there may be significant vision loss.




It is important to consider other diagnoses in your differential when a child presents with signs and symptoms of congenital glaucoma since it is a relatively rare entity. For epiphora, nasolacrimal duct obstruction is an important differential. Children may present with excessive tearing since blockage of the nasolacrimal duct prevents tear drainage. This is usually accompanied by yellow/green discharge and usually resolves by age 1. To learn more about this diagnosis, please see our PedsCases podcast on Nasolacrimal Duct Obstruction. Viral/bacterial/allergic conjunctivitis may cause also epiphora, with a history of recent illness or allergies. A corneal epithelial defect or abrasion, observed with fluorescein staining on slit lamp, should also be considered.

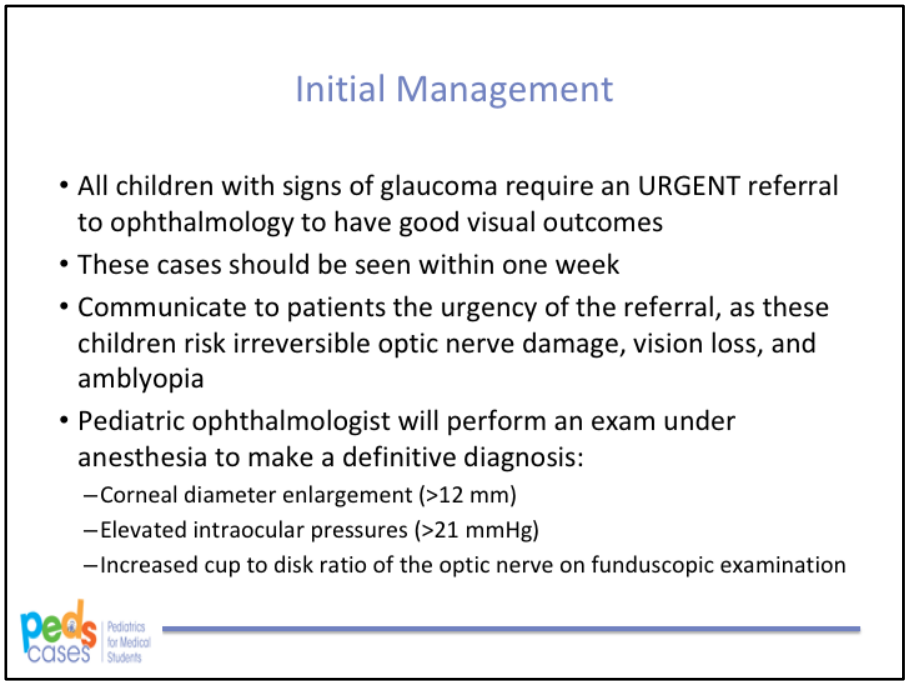
For photophobia, it is important to consider iritis, which will have signs of inflammation such as cells and flare in the anterior chamber observed on slit lamp. Also consider a history of trauma to the eye.

Some differential diagnoses for corneal clouding include birth trauma or chemical injury from history. On pre-natal history, it is important to note any TORCH infections in the mother in the pre-natal period since rubella keratitis may cause corneal clouding. In addition, this is may be a congenital corneal opacity, with absence of other signs of congenital glaucoma.



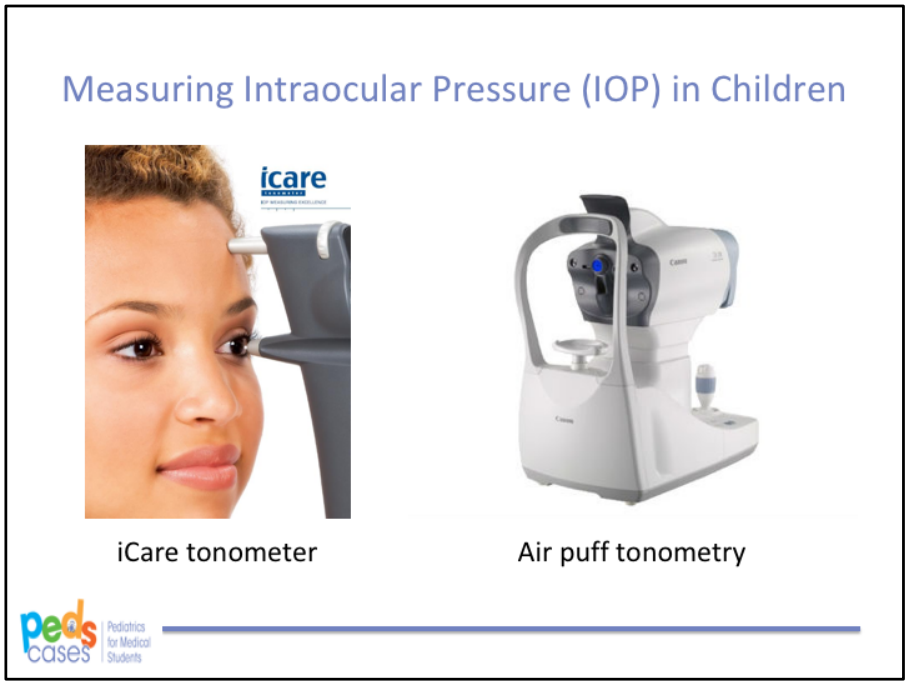

For corneal enlargement, it may be a developmental defect known as megalocornea (large corneal diameter). There will be absence of other signs of congenital glaucoma. Axial myopia may also cause corneal enlargement because there is elongation of the axis of the eye.

A pediatric ophthalmologist will most likely examine the optic nerve but if optic nerve cupping is observed on fundoscopy, some differentials to consider include physiologic optic nerve cupping and optic nerve hypoplasia. This results from underdevelopment of the optic nerve resulting in a small looking optic disc .



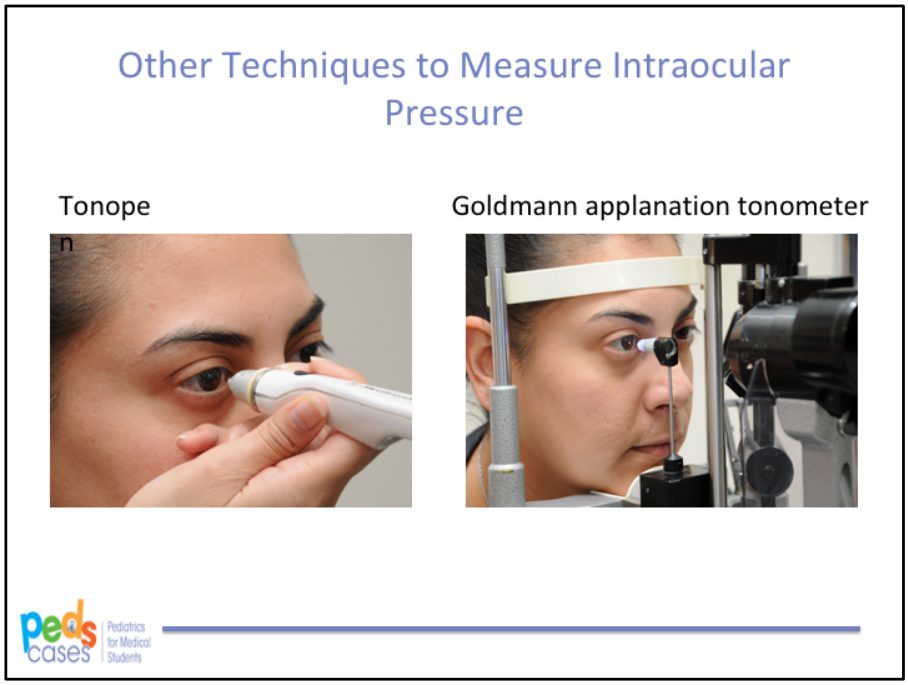

After discovering glaucoma on history and physical exam, all children require an urgent referral to an ophthalmologist experienced at examining children. They should ideally be seen within one week. Your referral should include a detailed medical history (including any associated systemic symptoms), family history of ophthalmic disease including congenital glaucoma, and any findings on physical exam. Communicate to the family the urgency of this referral, as delays can cause irreversible optic nerve damage, vision loss, permanent corneal enlargement, irregular astigmatism and amblyopia. A pediatric ophthalmologist can perform an exam under anesthesia to make a definitive diagnosis and take the necessary steps towards treating this condition. Some specialized assessments they can do include:

- Corneal diameter enlargement (>12 mm) measured with a caliper under topical or general anesthesia
- Elevated intraocular pressures (>21 mmHg) measured under general anesthesia
- Increased cup to disk ratio of the optic nerve on fundoscopy, indicating damage to the optic nerve and death of nerve fibers

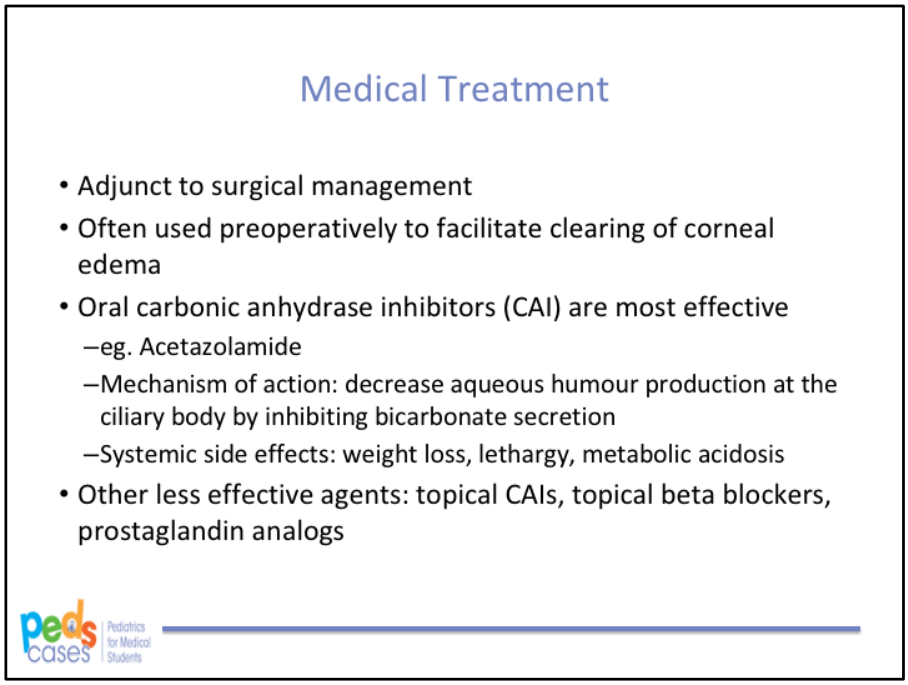



Since elevated intraocular pressure (IOP) is associated with glaucoma, accurate measurement of IOP is critical for diagnosis of glaucoma in both children and adults. Normal pressures will be in the range of 10-21 mmHg.

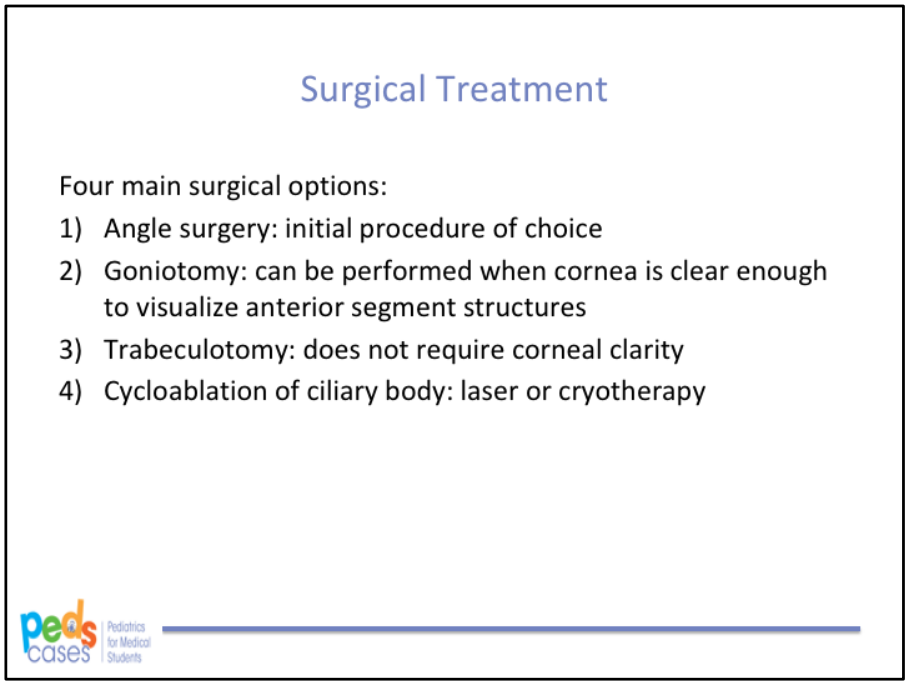
Measuring IOP in children is often challenging. Current methods include rebound tonometry (known commonly by the brand name iCare) and non-contact air puff tonometry . Rebound tonometry has the advantage of not requiring anesthetic drops and is the most common method of measuring pressures in children. For this technique, the child must be upright. To use rebound tonometry, put in a clean probe, ask the child to look straight ahead, and place the forehead support on the patient's forehead as shown in the picture. The probe should be 4-8 mm from the eye and perpendicular to it. Press the measurement button and make sure not to move the tonometer while measuring. There will be a beep after each successful measurement. Six measurements are required for an accurate reading. This is the easiest method to measure pressures in children since it does not require anesthetic drops and is the least invasive.  Air puff tonometry is also a good option if that machine is available in the office, since it does not directly touch the eye.




Other methods to measure IOP include handheld applanation tonometry (known commonly by the brand name Tono-Pen) and the Goldmann applanation tonometer. These two methods are more commonly used in adults. The Goldmann applanation tonometer is most accurate and is used as a gold standard for IOP measurements. However it is a bit more technically challenging and since most offices do not have a slit lamp, the technique for this will not be covered in this video. If you need to hold the eyes open for any of these techniques, be careful to push only on the orbital bones rather than the eye itself as this can artificially elevate pressures.



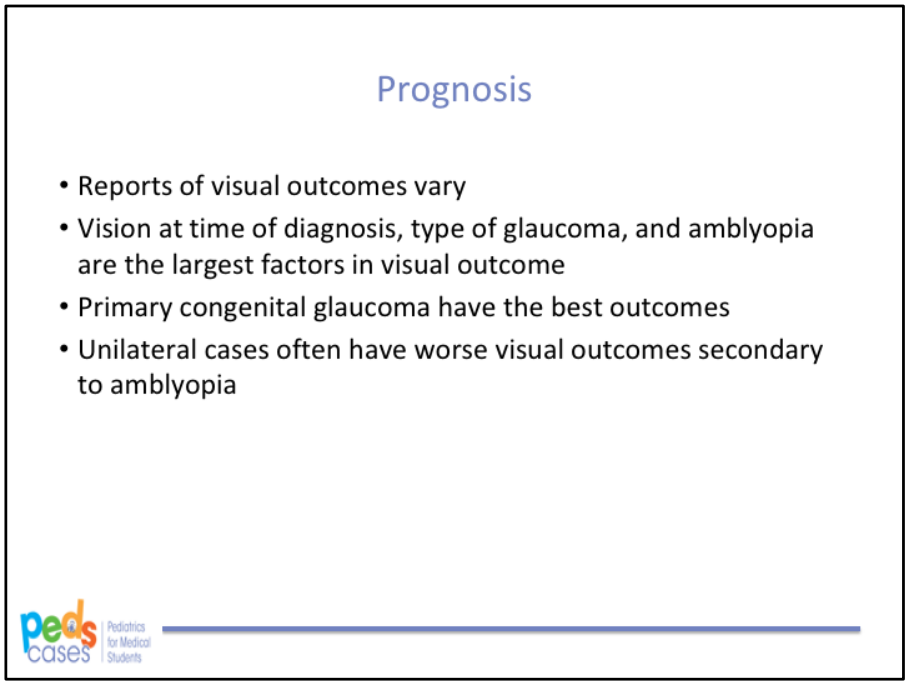

These next two slides on medical and surgical management touch briefly on how a pediatric ophthalmologist might manage the glaucoma after the referral. Medical therapy in pediatric glaucoma is often supplementary to surgical management. It is often used in preoperative treatment to help clear corneal edema. In addition, it can play a role in treating patients who are too unstable to undergo anesthesia. Oral carbonic anhydrase inhibitors such as acetazolamide are the most effective medications. These lower IOP by decreasing aqueous humour production from the ciliary body by inhibiting bicarbonate secretion from the ciliary body. However, it also works at the kidneys to promote diuresis by inhibiting bicarbonate re-uptake in the proximal convoluted tubules, which results in loss of bicarbonate, alkaline urine, and water. Thus patients should be counselled on systemic side effects such as weight loss, lethargy, and metabolic acidosis. Other less effective agents which can be considered include topical carbonic anhydrase inhibitors, topical beta blockers and prostaglandin analogs.



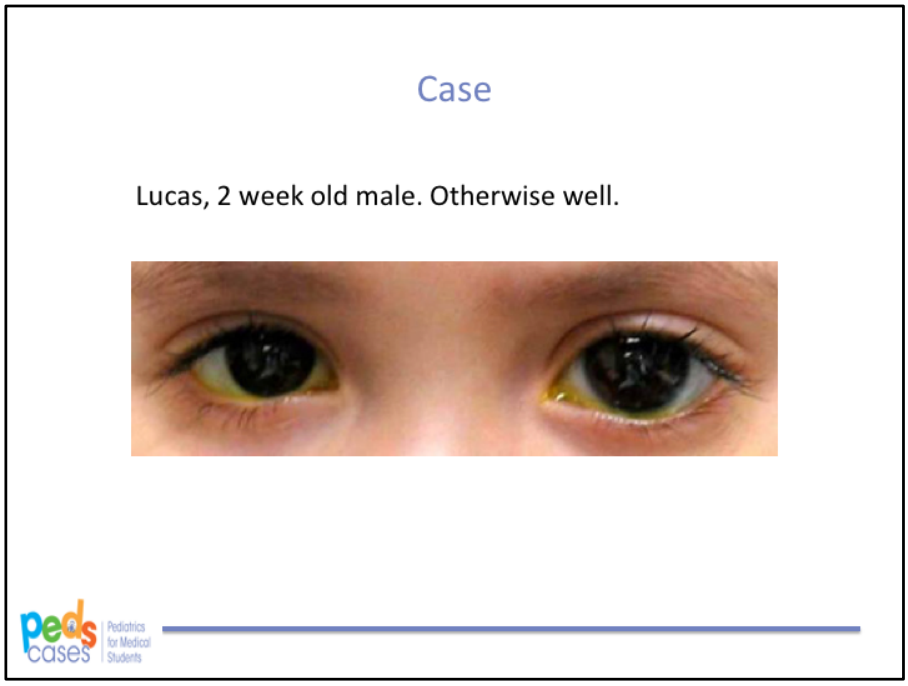

There are 4 major surgical options for primary congenital glaucoma:

1) Early angle surgery is considered the mainstay of treatment, with high success rates in patients treated before 1-2 years old.
2) Goniotomy is preferred when the cornea is clear enough to visualize anterior segment structures.
3) Trabeculotomy is the procedure of choice when the cornea is not clear enough to visualize the angle. But this has the technical challenge of locating Schlemm's canal.
4) Cycloablation of the ciliary body with laser or cryotherapy may be used in difficult to treat cases.

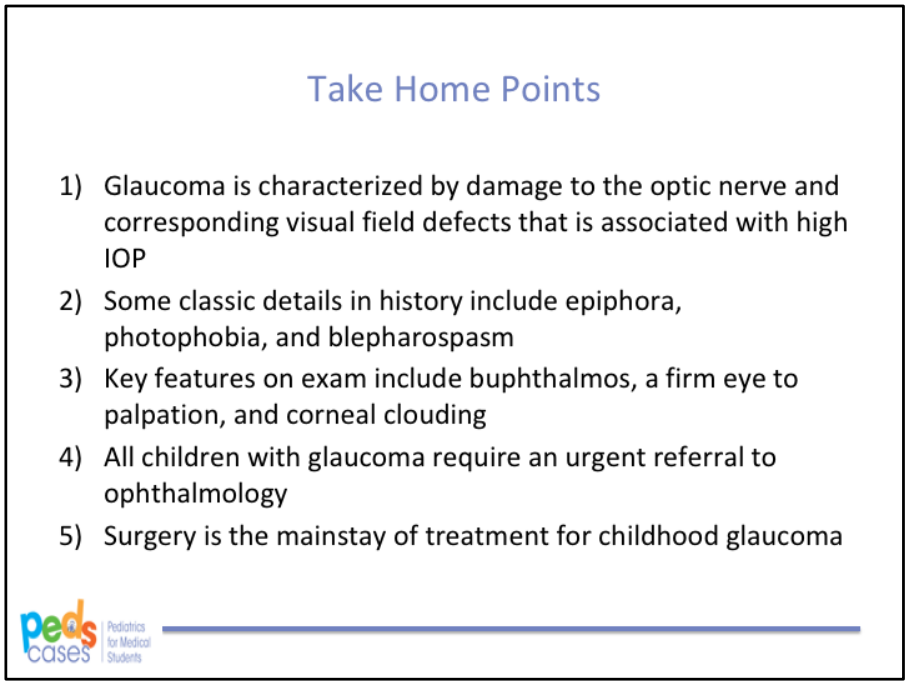
In terms of follow up, patients require frequent follow up in the short term to monitor for infection or excessive inflammation. In the long term, patients require lifelong serial measurements of intraocular pressure, corneal diameter, refractive error, and optic nerve cupping.




Reports of visual outcomes vary. Vision at the time of diagnosis, type of glaucoma and presence of amblyopia are the biggest factors in determining visual outcomes. Children with primary congenital glaucoma have the best prognosis whereas patients with unilateral glaucoma often have worse visual outcomes secondary to amblyopia.




Going back to the case, you find out that Lucas has no other associated symptoms, no previous history of eye trauma or eye surgeries and no family history of congenital glaucoma. On physical exam, you notice that both of Lucas' eyes are enlarged with the left eye being more obvious. You also notice some mild corneal clouding and that his eye is very firm to palpation. You suspect primary congenital glaucoma and explain this to the family. An urgent referral to a pediatric ophthalmologist is made, who measures his intraocular pressure under general anesthesia. Lucas' IOP is 30 mmHg on the right and 35 mmHg on the left. On ophthalmoscope examination of both eyes, his cup to disk ratio of the optic nerve looks higher than normal at 0.5. The diagnosis of primary congenital glaucoma is made and Lucas undergoes an angle surgery several days later. He makes a complete recovery.




The take home points of this podcast are:

1) Glaucoma is characterized by damage to the optic nerve and corresponding visual field defects that is associated with high IOP
2) Some classic details in history include epiphora, photophobia, and blepharospasm
3) Key features on exam include buphthalmos, a firm eye to palpation, and corneal clouding
4) All children with glaucoma require an urgent referral to ophthalmology
5) Surgery is the mainstay of treatment for childhood glaucoma

That concludes the presentation. Thanks for listening to PedsCases podcasts!

# Thanks for Listening!

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1. http://webeye.ophth.uiowa.edu/eyeforum/cases/42-primary-congenital-glaucoma-infantile-glaucoma.htm
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