



CHILDHOOD GLAUCOMAS

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INTRODUCTION

- The genetics, clinical findings and management of childhood glaucomas differ from that of adult glaucoma.
- Pathogenesis of primary congenital glaucoma- trabeculogysgenesis
- Secondary glaucomas-angle anomalies in most types of childhood glaucomas

INCIDENCE

- Incidence of childhood glaucoma is estimated to be 2.29 per 100,000 patients younger than 20 years old based on a defined U.S. Population study in Olmstead County.
- Primary congenital glaucoma is the most common form of childhood glaucoma, with a reported prevalence of 2.85 cases per 100,000

CLASSIFICATION

- Associated with a wide variety of pathology.
- Several classification systems :
- Primary and secondary
- Age of onset :congenital, infantile, juvenile
- Congenital and developmental glaucomas associated with ocular syndromes and systemic abnormalities fall under the umbrella of primary glaucomas.

CLASSIFICATION

- Causative pathologies ranging from uveitis to congenital cataract surgery, hyphaemas, fall under secondary glaucomas.

CLINICAL FINDINGS

- Manifestations of elevated IOP in children can vary depending on age of onset and rate of pressure elevation.
- Gradually increasing pressure can result in little to no corneal clouding.
- Presentation with buphthalmos and/or symptoms of tearing, blepharospasm and photophobia are more common.
- In contrast, those children with acute pressure elevations present with corneal clouding.

HAAB'S STRIAE



CLINICAL FEATURES

- Haab's striae, which represent breaks in Descemet's membrane, can be present in the absence of elevated pressure .
- This finding signifies a history of elevated IOP associated with rapid eye growth.
- Cupping also present. Can reverse following surgery sometimes.. Not in adults.

CLINICAL FEATURES/ EXAMINATION UNDER ANAESTHESIA

- Rebound tonometry (iCare) and handheld applanation tonometry (Tono-Pen, AccuPen, Kowa and Perkins Tonometer).
- Rebound tonometry has the advantage of not requiring anesthesia
- An exam under anesthesia is essential in diagnosing childhood glaucoma.
- Pre-intubation IOP, refraction, axial length, corneal diameter, gonioscopy, and ultrasound biomicroscopy when visibility is poor, are key components of the exam.

CLINICAL FEATURES/EXAMINATION UNDER ANAESTHESIA

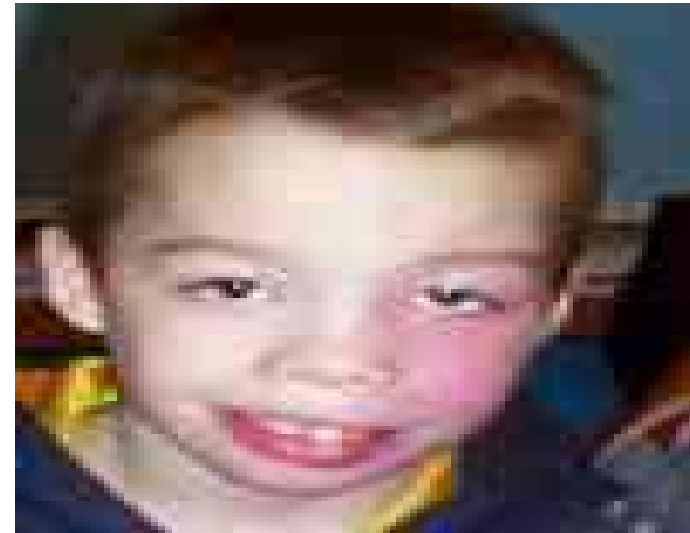
- Progressive myopia, increasing axial length and changing corneal diameter in the face of borderline IOP and cupping are suggestive of fluctuating high pressures.
- Tracking these factors also aids in determining treatment response
- Scleral rigidity reduces after the age of 3 years. Stretched cornea and sclera.

CLINICAL FEATURES

- A large number of syndromes have associated glaucoma.
- The more common syndromes are: Sturge-Weber; Oculocerebrorenal (Lowe); Axenfeld-Rieger; aniridia; and Neurofibromatosis Type 1.

CLINICAL FEATURES

- Sturge-Weber has a sporadic inheritance pattern and is characterized by nevus flammeus (port wine stain) of the face, angioma of the meninges and, rarely, involvement of the airway.



CLINICAL FEATURES

- Axenfeld-Rieger syndrome (ARS) is autosomal dominant
- It is associated with anterior segment abnormalities and is often categorized under anterior segment dysgenesis or goniodysgenesis syndromes, corneal opacity, adhesions between cornea and lens, iris and lens, iris and cornea.

Physical manifestations include: redundant umbilical skin; telecanthus; broad nasal bridge; dental abnormalities (microdontia, oligodontia or hypodontia); and, in some cases, pituitary abnormalities with growth retardation.

AXENFELD REIGER SYNDROME



CLINICAL FEATURES

- Aniridia characterized by hypoplastic iris tissue and is associated with foveal hypoplasia, cataracts, keratopathy secondary to limbal stem cell deficiency and, occasionally, optic nerve hypoplasia. The inheritance pattern is autosomal dominant



CLINICAL FEATURES

- Neurofibromatosis Type 1 has an autosomal-dominant inheritance.
- Café-au-lait spots, freckling of the axial/inguinal area, sphenoid dysplasia, S-shaped plexiform neurofibromas of the lids, optic nerve gliomas, Lisch nodules and choroidal hamartomas. plexiform neurofibromas of the eyelids



TREATMENT

- Medical therapy in pediatric glaucoma is often supplementary to surgical management.
- It is often used for preoperative treatment to facilitate clearing of corneal edema.
- In addition, it can play a role in treating patients who are too unstable to undergo anesthesia.
- Timolol, Prostaglandin analogues

TREATMENT

- Surgery
- Angle surgery is considered the mainstay of treatment for primary congenital glaucoma, with a reported 70 to 90 percent success rate after one to two procedures in patients treated after 3 months of age and before 1 to 2 years of age.
- This success rate significantly diminishes in patients presenting outside of this age range and those who fall in the spectrum of developmental glaucoma.

TREATMENT

- Goniotomy is often the first procedure of choice, but it requires a clear cornea to be performed safely with success rates of 70 to 90 percent.
- Trabeculotomy *ab externo* does not require corneal clarity; however, it requires the technical challenge of finding Schlemm's canal. Traditional trabeculotomy treats 90 to 180 degrees.
- Success rates are comparable to goniotomy
- Combined trabeculotomy/ trabeculectomy
- Trabeculectomy with antifibrotic agents success rates of 60 to 65 percent.

TREATMENT

- Aqueous shunt implantation has shown significantly greater success when compared to trabeculectomy.
- Low endophthalmitis rates have been reported
- Transscleral laser cyclophotocoagulation in recalcitrant cases, using transillumination.

PROGNOSIS

- Vision at the time of diagnosis, type of glaucoma and amblyopia appear to be the largest factors in visual outcomes.
- Children with primary congenital glaucoma have the best prognosis.
- In the setting of well-controlled intraocular pressure, amblyopia is a key factor in vision loss.
- Unilateral cases often have poorer visual outcomes secondary to amblyopia.
- Refraction, monitoring axial lengths , amblyopia management key factors after IOP control.
- Follow up and counselling are essential for both paediatric and adult glaucomas.

THE END

