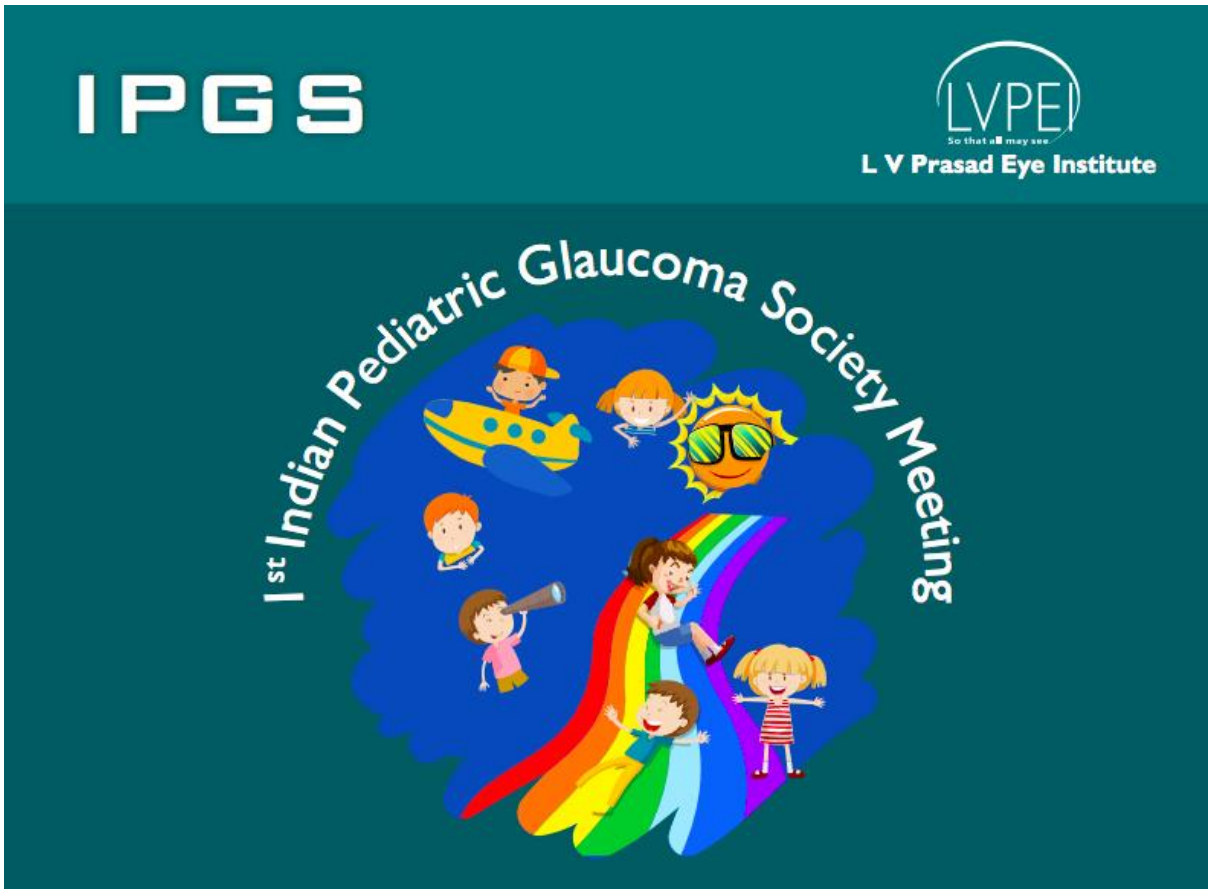

Learnings from IPGS 2019

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Vision Assessment

By 2-6 months of age child fixes and follows

Central, Steady and Maintained fixation – suggests vision in both eyes is good

Crucial Milestones	Age	Significance
Stable eye contact	8 weeks	
Social smile	3-4 months	Indicates accommodation is developing
Awareness of hands	3-4 months	Visual conception
Watch hand movements	5 months	
Eye positioned straight	5 months	Requisite for binocular vision If squint noted, needs cycloplegic refraction
Goal oriented hand movements	6-7 months	
Recognition of familiar faces	7-9 months	
Pictures in books Picking up objects	9-12 months	
Stereopsis	5 years	

Age	Chart used
Pre-Verbal children	Preferential looking test Teller Acuity Cards (TAC) Cardiff
Pre School children	Lea Symbols Sheridan Gardiner Chart

TAC can be done in children who have developed neck holding (>3mo)

Visual acuity in children:

- Matches adults visual acuity on teller acuity cards by 18 months
- Matches adults visual acuity on Snellen's by 3 years

In children >1 year of age, following refractive errors are considered significant and should be treated

- ✓ Myopia > -2 D
- ✓ Hypermetropia > +3D
- ✓ Astigmatism > +/- 1.5 D

IOP and Tonometry

1. Normal IOP values in children (under GA, with Applanation tonometer)
    - <3 months: <8 mm Hg
    - 6-9 months: <12 mm Hg
    - Increases 1 mm Hg per year up to 12 years, reaching adult levels

IOP is underestimated by at least 5 mm Hg under EUA

2. Preferred Tonometer
    - Perkins tonometer – Gold standard
    - Tonopen can be used - overestimates IOP
    - ICare ic200 – rebound tonometer with longer probe- can be used in supine position

3. Effect of anaesthetic agents on IOP

IOP	Anaesthetic Agents
Increase	Ketamine, Succinyl choline
Mild reduction	Midazolam, Sevoflurane, Methohexital
Significant reduction	Halothane

4. Factors affecting IOP during EUA (factors other than anaesthesia)
    - Speculum – raises IOP an average of 4mmHg
    - Type of speculum – Wire speculum> screw speculum
    - Degree of separation of lids
    - Globe manipulation (sclera depression)

5. Airway management during EUA

Type of Airway	Effect on IOP	Mechanism
Facemask	Increase in IOP	Inadvertent pressure on the eye
LMA	Closer to actual IOP	Hemodynamic response less
Laryngoscope used during endotracheal tube insertion	Increase in IOP	Stimulates adrenergic response and increases the outflow resistance

6. Recommendations during EUA
    - Wait for at least 2-3 minutes after anesthesia induction to check for IOP
    - Avoid use of speculum and gently separate the lids by hands/bud
    - Measure IOP before any contact procedure
    - Similar tonometer to be used in repeated examinations

Goniotomy

1. Indications:
    - PCG – *Best indication*
    - Sturge-Weber Syndrome
    - Uveitic glaucoma

    } *PCG-like angles*

    - Secondary glaucoma associated with Congenital Rubella

2. Poor prognostic factors for goniotomy:
    - Early presentation (at birth or <1-month age)
    - Sematic and Indian races
    - Higher IOP at presentation
    - HCD >14mm
    - Axial length >24mm

3. Goniolens - Khaw, Barkan, Swan-Jacob

4. Mechanism of Action:
    - Incise the impermeable membrane
    - Reduces the iris insertion causing increase in outflow

5. Procedure:
    - Inject intracamerally Pilocarpine, avoid apraclonidine as it causes pupillary dilatation
    - Locking forceps (at 6 and 12’0 clock)
    - Place the Goniolens with OVD on cornea

- Initially lower magnification and slowly increase the magnification as we enter the eye
- Lower the iris insertion from TM

6. Intraoperative tips to get clear cornea during goniotomy
    - Debride the corneal epithelium with alcohol soiled cotton buds
    - Debride the epithelium with Bard parker handle
    - Control IOP preoperatively with AGMs – especially pilocarpine

Advantages	Disadvantages
• Spares conjunctiva • Short surgical time • Allows direct view	• Needs special equipment • Visibility of angles is a prerequisite

Primary Congenital Glaucoma

1. Target IOP in children in PCG is usually below 15 mm Hg
    - Children have thinner lamina crib Rosa and thinner sclera, so a target IOP cut off of 21 mm Hg is not adequate
    - Usually taken as IOP < 15mmHg, preferable IOP <12mmHg
    - 70% cases show reversal of cupping if IOP of <12mmHg

2. Preferred surgery for PCG:
    - Goniotomy: Western population
    - Deep sclerectomy/combined Trabeculotomy and trabeculectomy in Saudi Arabia
    - Combined trabeculotomy and trabeculectomy in Indian population

3. Trabeculotomy
    - Small bleed in anterior chamber indicates correct plane
    - Massive AC bleed indicates injury to ciliary body
    - Post-operative pilocarpine should be given when trabeculotomy alone is performed as it helps keep the trabeculotomy open

4. Schedule for Post-operative EUA (The below is only if everything is going well after surgery)
    - A month after surgery
    - Every 3-4 months for the first year, then every 6 months

During EUA, after ocular examination, utilize the opportunity to examine the child for systemic features also, especially in suspected syndromes

5. Surrogates that indicate stable disease/successful surgical outcome in a child:
    - Reversal or stable CDR
    - Clear Cornea
    - Stable Axial length
    - Visual acuity and visual fields

6. Surrogates that indicate glaucoma progression in children
    - Increased IOP
    - Increase in Horizontal corneal diameter
    - Increase in Axial length
    - Myopic shift

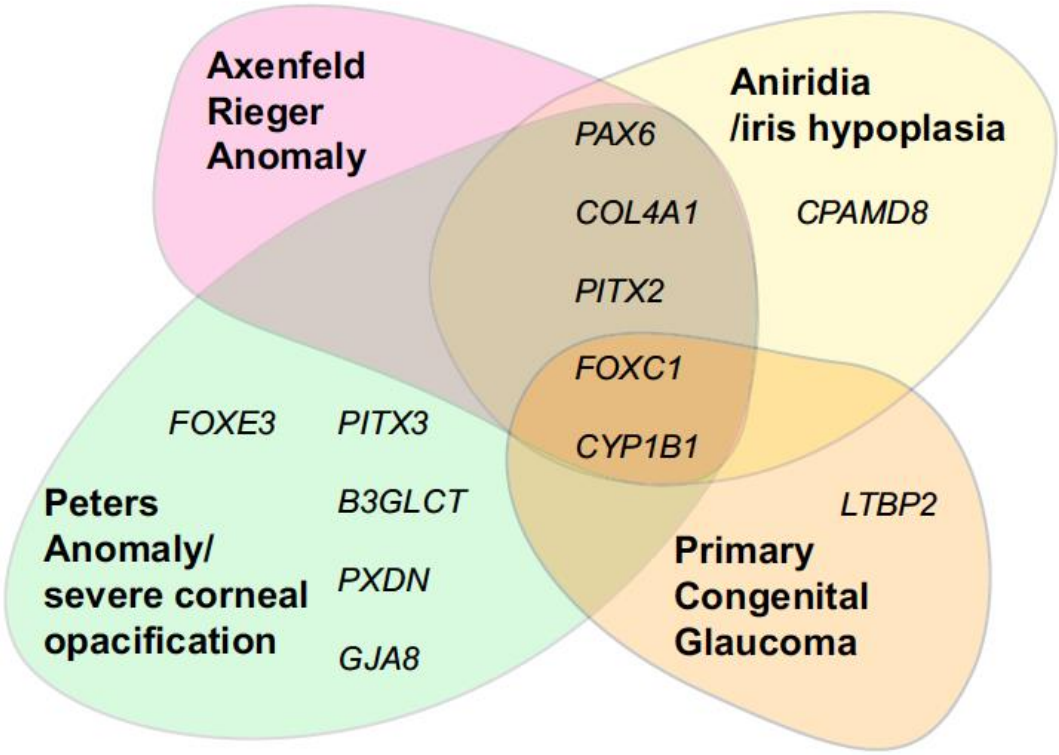
Follow-up with refraction and amblyopia very important

Axial length >26 mm is a risk for retinal detachment

Congenital Megalocornea	Buphthalmos
More common in boys	
X- Linked Recessive	
AC Depth/Total Axial Length >0.2	Both anterior and posterior segment depth is more
Regular astigmatism	Irregular astigmatism

Genetics

Mutation	Disease	Typical features
Myocilin	POAG/JOAG	
CYP1B1 gene	PCG	
PAX6	Aniridia	
FOXC1	Axenfeld-Reiger spectrum	
PITX2	A-R spectrum	
COL4A1	A-R spectrum	Microcornea, Cataract, iris abnormalities
CREBBP	Rubenstein-Taybi syndrome	
SLC4A4		Renal tubular acidosis, developmental delay, band keratopathy, glaucoma
CYP1B1+FBN1 (fibrillin)	JOAG	JOAG+Subluxated lens
SLC4A11	CHED	



Ma AS, Grigg JR, Jamieson RV. Phenotype-genotype correlations and emerging pathways in ocular anterior segment dysgenesis. Hum Genet. 2019 Sep;138(8-9):899-915

Diagnosis of pediatric glaucoma is clinical and genetic testing is usually not needed

Indications of genetic testing:

1. Family counselling is desired and is reasonable
2. The phenotype suggests a risk for extraocular disease
3. The glaucoma is in the setting of systemic disease
4. The diagnosis is questionable (certain populations)

Refractory Glaucomas

Pearls:

- Prevention is the key

- First chance is the best chance
  - ✓ Choose the first surgery wisely
  - ✓ Eyes with choroidal haemangiomas/ post cataract surgery glaucomas/ cornel grafts – Prefer tubes

- If one operation fails- do not repeat the same in the other eye

- No surgical short cuts

- Do not underestimate complications of GDD
  - ✓ Infantile Sturge Weber syndrome- 250 mm^2 baerveldt is used(implant with a lower surface area is preferable), No mitomycin
  - ✓ Microphthalmos need smaller implants
  - ✓ The two factors which determine the success of GDD are surface area of the implant and thickness of the capsule
  - ✓ To maximize the efficacy of second implant- Take second implant with larger surface area, do it in a place where conjunctiva is still naïve, use MMC

- Know when to stop

Anaesthesia in children

1. Use of dexmedetomidine for EUA in pediatric glaucoma cases
    - Given intranasally by using atomizer
    - It prevents need of inhalational anaesthesia that carries risk of effecting the neurodevelopment
    - Prevents need of intravenous cannulation for medication
    - Child should be monitored by anaesthesiologist when dexmedetomidine is being used as it can lower the heart rate
    - Dosage: 3-3.5 micrograms/kg intranasally divided between two nostrils

2. Factors which can make intubation difficult in children
    - Smaller pharynx
    - Bigger and floppier epiglottis
    - Bigger tongue

3. Factors in Sturge Weber Syndrome which affect anaesthesia management
    - Angiomas of the airway
    - Laryngospasm and high output failure
    - Seizure disorder
    - Facial dysmorphism

4. Pearls in pediatric anaesthesia
    - Dysmorphic facies + Airway + cardiac status - 3 important considerations
    - Bronchospasm can cause low SpO2 leading to cyanosis

- Bradycardia can lead to cardiac arrest in children
- 1 mm laryngeal oedema can narrow the airway by 60 %
- Neutral head position is very important - extended position can cause accidental extubation
- Keep a high index of suspicion for pneumothorax in Marfan’s Syndrome

Sturge Weber Syndrome

1. Steps to prevent choroidal effusion in Sturge weber syndrome:
    - Prophylactic sclerostomy 5 mm from limbus in inferotemporal quadrant
    - Use ACM to prevent sudden hypotony
    - Preplaced scleral flap suture
    - Releasable sutures
    - Prophylactic propranolol 2 mg/kg/day in two divided dosages, start 1 week prior to surgery and continue till 6 weeks postop

2. Difluprednate is preferred over prednisolone in cases of postoperative choroidal detachment
    - It has increased affinity to glucocorticoid receptors, thus it is more potent compared with all other steroid molecules
    - It has enhanced penetration to the uvea, due to the acetate ester group at C21
    - As an emulsion, it provides consistent and uniform doses without requiring shaking

3. Choroidal haemangioma in Sturge weber syndrome:
    - Always perform a Distant direct ophthalmoscopy and indirect ophthalmoscopy
    - May not be seen as a discrete lesion
    - Always compare the two eyes - presence of orange-reddish hue (Tomato ketchup appearance) on affected side indicates the presence of choroidal haemangioma
    - Intra operative chemosis may hint towards choroidal effusion and choroidal detachment

Other Secondary Glaucomas

1. Ectropion Uvea
    - Unilateral ectropion: rule out neurofibromatosis
    - Bilateral ectropion: anterior segment dysgenesis

2. Peters anomaly:
    - UBM may help to see the density of corneal scarring and the opacity in these cases is usually denser towards Descemet's membrane. So, doing a DSEK instead of PK can suffice, which is better in an eye with trabeculectomy.
    - Management of cornea haze in peter's anomaly :Optical iridectomy (treatment of choice)

3. In Axenfeld-Rieger Spectrum, glaucoma is due to the deformities in the trabecular meshwork and not due to the iris tissue adhered to Schwalbe's line

4. Glaucoma in aniridia: The mechanism of glaucoma is
    - Impaired Trabecular meshwork function
    - Absent Schlemm's canal
    - Progressive goniosynechiae causing angle closure

5. Glaucoma in ROP
    - Stage V ROP is most commonly associated with glaucoma
    - Glaucoma could be
        - secondary to ROP itself
        - secondary to VR surgery which is more common in Stage V ROP
    - Incidence of glaucoma lesser in cases of eyes where lens sparing vitrectomy has been performed compared to the eyes where lens has been removed

6. Microspherophakia

    - Young age + myopia but shallow anterior chamber suspect microspherophakia
    - Micropherophakia does not mean the lens is small- zonules which normally cause flattening of the lens periphery are abnormal thereby causing a spherical lens.
    - Lensectomy effective in controlling IOP in most of the cases. Can also be planned for high refractive error even if IOP is normal
    - Indications for trabeculectomy or combined surgery
        - ✓ Age<6 years
        - ✓ IOP>32 mm Hg
        - ✓ Advanced disc damage
    - Myopia should be termed as lenticular myopia rather than high myopia in microspherophakia
    - Yag PI may not be very helpful (controversial topic)

7. Post cataract surgery glaucoma
    - Acute glaucoma is usually pupillary block glaucoma — Uveitis (PAS/secclusio/occlusio pupil), Inflammatory membrane, remnant lens matter
    - Chronic open angle glaucoma mechanisms:
        - ✓ Mechanical Theory - Lens removal leads to release of tension on zonules which decreases the pull of TM and decreased aqueous outflow
        - ✓ Chemical Theory – due to inflammatory mediators obstructing TM
        - ✓ Steroid induced
    - Age at cataract surgery if <1 month— high risk of developing glaucoma (risk - 11.8%)
    - >25% of pediatric cataract surgery develop glaucoma

8. If CHED diagnosis is questionable then:
    - Hyperopic refraction can give a clue
    - Genetic testing: SLC4A11 mutations can be present