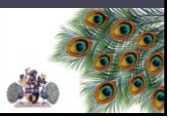
# Childhood Glaucoma in the 21st Century

Egyptian Society of Glaucoma
19th Annual Meeting

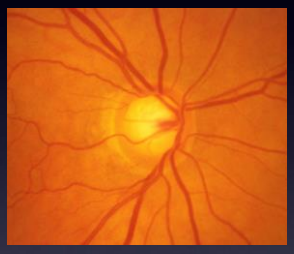
Maria Papadopoulos
Glaucoma Service
MEH, London, UK

Moorfields Eye Hospital NHS Foundation Trust



## Childhood Glaucoma

- Raised IOP damages optic nerve & eye



Glaucoma

Right buphthalmos with raised IOP in infancy

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before surgery

after surgery

Courtesy J Brandt

# Childhood Glaucoma

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  - more likely [1]
  - more likely to fail [2,3]
  - more challenging (↑complications) [4]


1. Taylor RH, Ainsworth JR, Evans AR, Levin AV. The Epidemiology of Pediatric Glaucoma: The Toronto Experience. *J AAPOS* 1999
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- Amblyopia important cause of reduced vision


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# Primary congenital glaucoma (PCG)

- Isolated trabeculodysgenesis

'immature angle'
arrest of angle maturation

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  1 /4,250 Egypt [2]
  1/18,500 live births UK [3]

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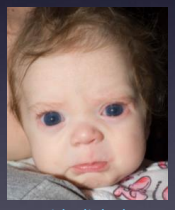
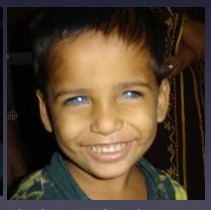
  Middle East & South Asia: more severe phenotype, late



Buphthalmos with slight hazy corneas (West)



Severe buphthalmos & marked corneal oedema (Middle East & India)

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- Genetics: sporadic
  AR: *CYP1B1* mutations
  AD: *TEK* mutations

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# Management

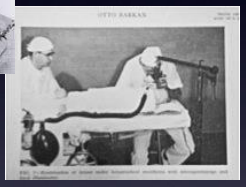
- Control IOP
  - Medication
  - Surgery* - mainstay of treatment
- Visual rehabilitation: treat refractive errors & amblyopia
  - Glasses (tinted/photochromatic), low vision aids, patching
- Manage associated ocular problems
- Advice with schooling and social support
- Management of whole family (genetic counselling, siblings)

# Surgery

- Angle surgery
  - Goniotomy
  - TrabeculOtomy
  - TrabeculOtomy + TrabeculECtomy (CTT) (Middle East & South Asia)
- TrabeculECtomy
- Glaucoma drainage devices
- (Cyclodestruction)

# Angle Surgery

## Goniotomy (Barkan, 1942)

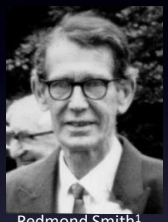


need clear cornea

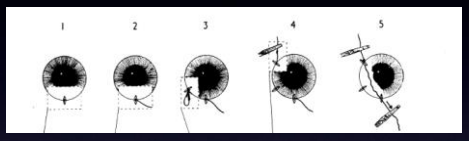
Barkan O. Operation for congenital glaucoma. *Am J Ophthalmol* 1942;25:552-68.

# Angle Surgery

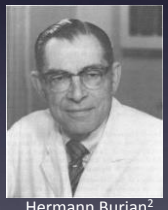
## Trabeculotomy (1960)



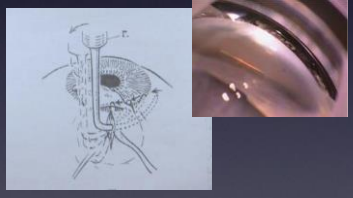
Redmond Smith[1]



*nylon suture*
incision of
Schlemm canal



Hermann Burian[2]

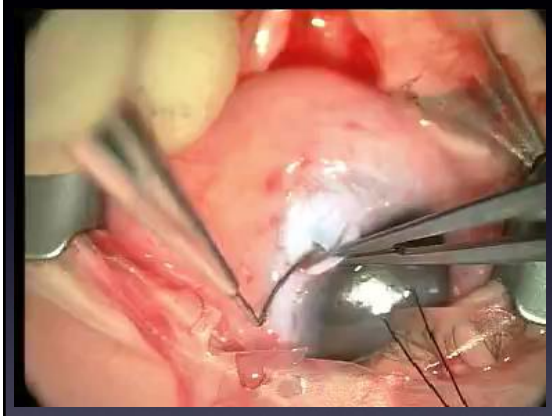


*trabeculotome*
(metal probe)
incision of
Schlemm canal

'trabeculotomy ab-externo'

1. Smith R. A new technique for opening the canal of Schlemm. Preliminary report. *Br J Ophthalmol* 1960
2. Burian HM. A case of Marfan's syn with glaucoma:descr of new type op for developmental glaucoma (trabeculotomy ab externo). *AJO* 1960

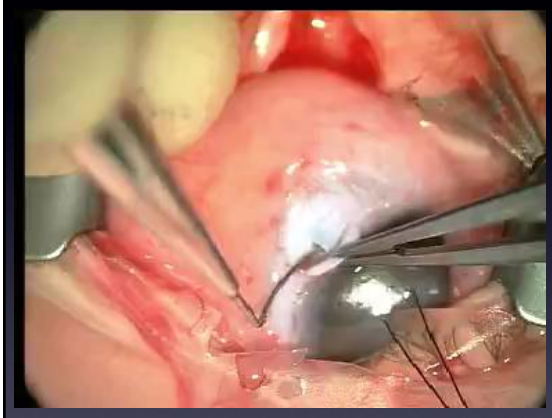
# TrabeculOtomy

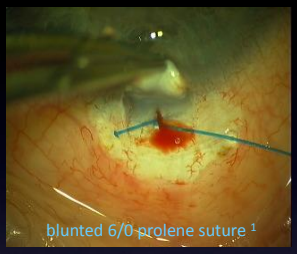


Probe Trabeculotomy

≈ 120° (often needs to be repeated)

# TrabeculOtomy





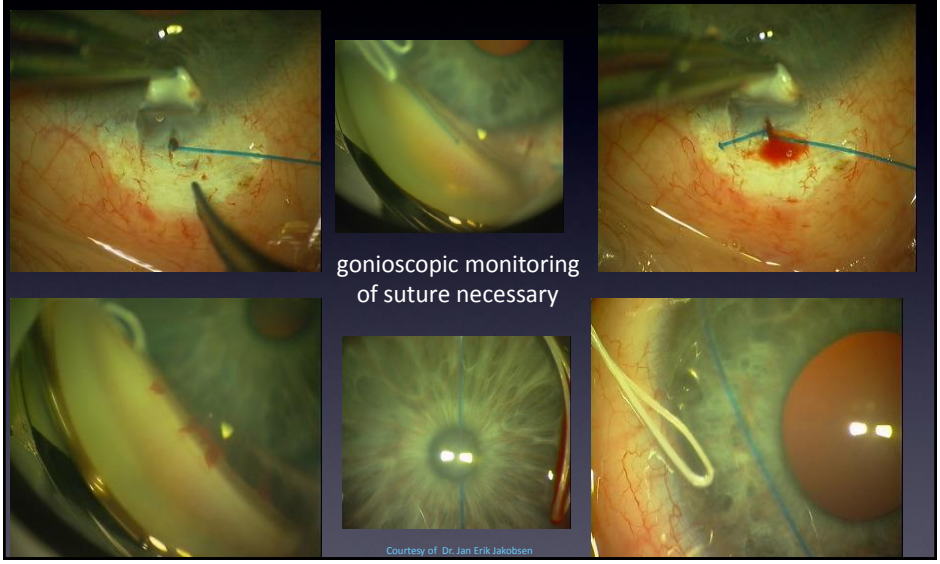

Probe Trabeculotomy

≈ 120° (often needs to be repeated)

360° Trabeculotomy

1 Beck AD, Lynch MG. *Arch Ophthalmol* 1995

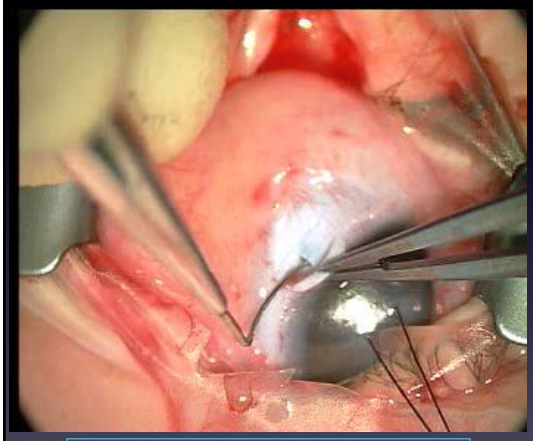
# 360° TrabeculOtomy
## blunted 6/0 prolene suture




# 360° TrabeculOtomy
## blunted 6/0 prolene suture

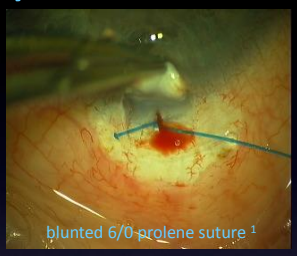

1. Neely DE. *J AAPOS* 2005
2. Verner-Cole EA. *Am J Ophthalmol* 20
3. Chang TC, Cavuoto KM. *J Ophthalmol* 2015

# TrabeculOtomy



Conventional probe Trabeculotomy

≈ 120° (often needs to be repeated)



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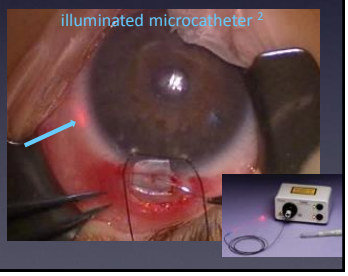
360° Trabeculotomy

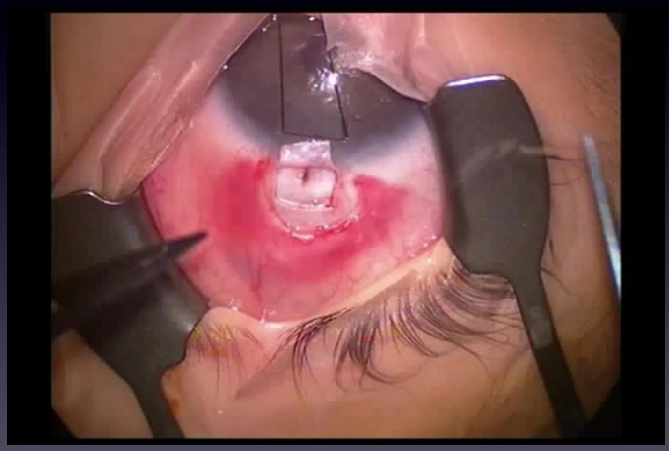


illuminated microcatheter [2]

1 Beck AD, Lynch MG. *Arch Ophthalmol* 1995
2. Sarkisian SR. *J AAPOS* 2010

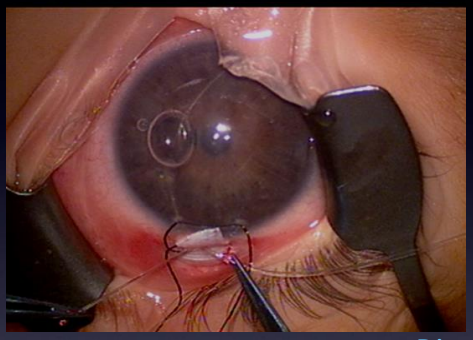
# 360° TrabeculOtomy

## Illuminated microcatheter (iTrack)



# 360° TrabeculOtomy

## Illuminated microcatheter



### Advantages

- safe, regardless of corneal clarity
- once, minimises delay in controlling IOP
- 360° TrabO more successful than probe TrabO [1]
- 360° TrabO = Combined TrabO + TrabEC with MMC [2]
- can be performed ab interno through corneal incision (GATT) [3]

### Disadvantages

- time consuming (ab externo)

1. El Sayed Y, Gawdat G. *Acta Ophthalmol* 2017
2. Temkar S et al. *Am J Opthalmol* 2014
3. Grover DS et al. *BJO* 2015

# Surgery

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  - Goniotomy
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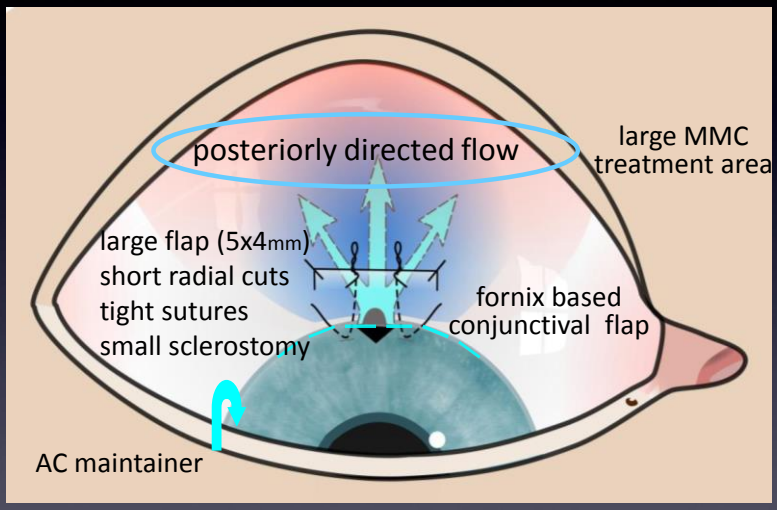
  - TrabeculOtomy + TrabeculECtomy (CTT) (Middle East & South Asia)
- TrabeculECtomy
- Glaucoma drainage devices
- (Cyclodestruction)

# Paediatric TrabeculECtomy: contemporary




Moorfields Safer Surgery System (MSSS)

# Paediatric MMC TrabeculECtomy in infants

American Academy of Ophthalmology
The Eye M.D. Association

**Long-Term Outcomes of Trabeculectomy Augmented with Mitomycin C Undertaken within the First 2 Years of Life**

Hari Jayaram, PhD, FRCSEd,[1,2,3] Richard Scawn, FRCOphth,[1] Francisco Pooley, MD,[1,4] Mark Chiang, MBBS, FRANZCO,[1,2,5] Catey Bunce, MSc, DSc,[5,6] Nicholas G. Strouthidis, PhD, FRCOphth,[1,2,7,8] Peng Tee Khaw, PhD, FRCOphth,[1,2] Maria Papadopoulos, MBBS, FRCOphth[1,2]

- 40 phakic eyes
- 80% PCG failed goniotomy
- Moorfields SSS Trabeculectomy
- MMC subconj & under scleral flap

week 1

week 6

3 months

diffuse, elevated bleb

1. Jayaram H et al. Long term outcomes of trabeculectomy augmented with MMC undertaken within the first 2 yrs of life. *Ophthalmology* 2015

# Paediatric MMC TrabeculECtomy in infants

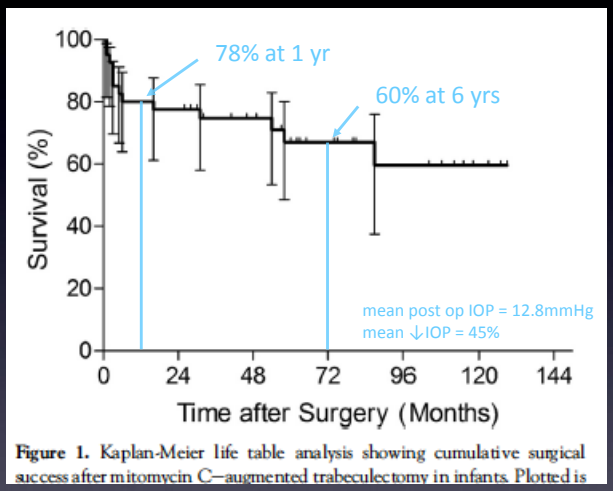



**Figure 1.** Kaplan-Meier life table analysis showing cumulative surgical success after mitomycin C–augmented trabeculectomy in infants. Plotted is

Failed cases

all uncontrolled IOP

Successful cases

96% off meds

44% 6/12 or better

1/40 cystic bleb

1. Jayaram H et al. Long term outcomes of trabeculectomy augmented with MMC undertaken within the first 2 yrs of life. *Ophthalmology* 2015

# Paediatric MMC TrabeculECtomy in infants

## Complications

| | Jayaram et al (MEH)[1] | Beck et al [2] |
|---|---|---|
| Choroidal effusions | 4 / 40 (10%)* | 4 / 24 (16.7%) |
| Chronic hypotony | 0/40 | 1 / 24 (4%) |
| Late bleb leak | 0 / 40 | 3 / 24 (12.5%) |
| Bleb related infection | 1 / 40 (2.5%) blebitis | 2 / 24 (8%) endophthalmitis |
| Corneal decompensation | 0 / 40 | 2 / 24 (8%) |
| Cataract | 3/26 (11.5%) | 2 / 24 (8%) |

* All 4 cases of effusions had successful outcome (3 AC viscoelastic inj)

1. Jayaram H et al. Long term outcomes of trabeculectomy augmented with MMC undertaken within the first 2 yrs of life. *Ophthalmology* 2015
2. Beck AD, Freedman S, Kammer J, Jin J. Aqueous shunt devices compared to Trabeculectomy with MMC for children in 1st two years of life. *Am J Ophthalmol* 2003

# Redo Paediatric MMC TrabeculECtomy

- Role of Avastin (Bevacizumab)

Evaluation the adjunctive use of combined bevacizumab and mitomycinc to trabeculectomy in management of recurrent pediatric glaucoma

RA Mahdy, SM Al-Mosallamy, MA Al-Aswad, A Bor'i and WM El-Haig

CLINICAL STUDY

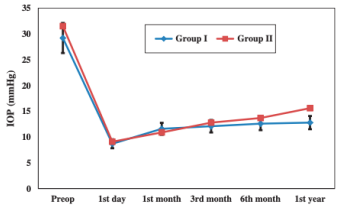



**Figure 1** Linear regression curve showing preoperative and postoperative follow-up of the IOP with error bar.

24 eyes (previous traby ± MMC)

prospective

- MMC + Avastin (2.25 mgs) subconj Gp I
- MMC alone (0.4mgs/ml, 3 mins) Gp II

Mahdy RA, Al-Mosallamy SM, Al-Sawad MA,Bor'i A, Wm E-H. Evaluation the adjunctive use of combined bevacizumab and mitomycin C to trabeculectomy in the management of recurrent pediatric glaucoma. Eye (Lond). 2016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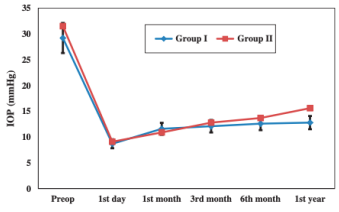



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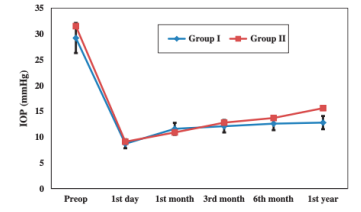


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1 chronic hypotony case MMC + Avastin
1 endophthalmitis case MMC alone

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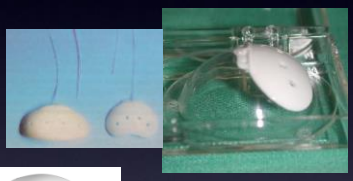
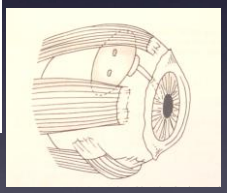
- (Cyclodestruction)

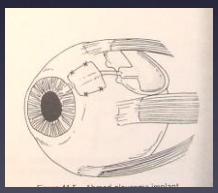
# Glaucoma Drainage Devices

- Operation of choice for refractory childhood glaucomas
  - failed trabeculectomy and primary intervention (e.g. aphakia / uveitis)



Baerveldt & AADI implant
(non flow-restricted)



Ahmed implant
(flow-restricted)

# Glaucoma Drainage Devices

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- Similar success rates for all implants[1,2] ≈ 50% 5 years + meds

1. Tanimoto SA, Brandt JD. Options in pediatric glaucoma after angle surgery has failed. *Curr Opin Ophthalmol*. 2006
2. Chen A et al. Valved Glaucoma Drainage Devices in Pediatric Glaucoma: Retrospective Long-term Outcomes. *JAMA Ophthalmol* 2015.

# Glaucoma Drainage Devices

- Role of Avastin (Bevacizumab) or MMC

ORIGINAL STUDY

Adjunctive Use of Bevacizumab Versus Mitomycin C With Ahmed Valve Implantation in Treatment of Pediatric Glaucoma

*Reda Abdel Rahman Mahdy, MD*

- prospective, randomised, masked
- 60 eyes, S3 paediatric model (85mm²)

Madhy RAR. Adjunctive use of bevacizumab versus mitomycin C with Ahmed valve implantation in treatment of pediatric glaucoma. *J Glaucoma* 2011

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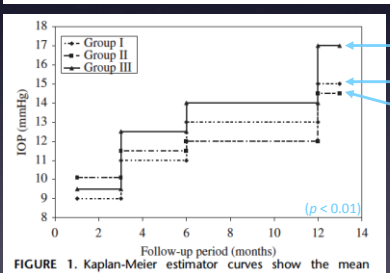


**FIGURE 1.** Kaplan-Meier estimator curves show the mean estimated intraocular pressure (IOP) during the follow-up period of the 3 studied groups.

Control

Bevacizumab (Avastin) 1.25mg subconj

MMC 0.4mgs/ml 3 mins

IOP control: MMC & Avastin better than Control Gp

Madhy RAR. Adjunctive use of bevacizumab versus mitomycin C with Ahmed valve implantation in treatment of pediatric glaucoma. *J Glaucoma* 2011

# Glaucoma Drainage Devices

- Role of Avastin (Bevacizumab) or MMC

ORIGINAL STUDY

Adjunctive Use of Bevacizumab Versus Mitomycin C With Ahmed Valve Implantation in Treatment of Pediatric Glaucoma

*Reda Abdel Rahman Mahdy, MD*

- prospective, randomised, masked
- 60 eyes, S3 paediatric model (85mm²)

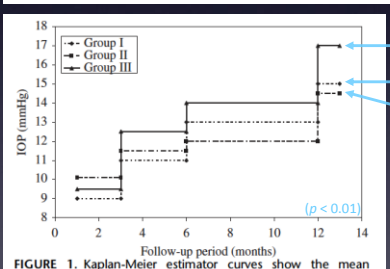


FIGURE 1. Kaplan-Meier estimator curves show the mean estimated intraocular pressure (IOP) during the follow-up period of the 3 studied groups.

Control

Bevacizumab (Avastin) 1.25mg subconj

MMC 0.4mgs/ml 3 mins

IOP control: MMC & Avastin better than Control Gp

Success: MMC (90%) / Avastin (80%) / Control (60%)

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COMPLICATIONS Avastin MMC Control

TABLE 5. The Complications Encountered in the 3 Studied Groups

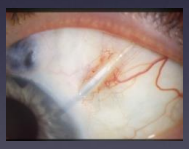
| | Group I=20 Eyes | | Group II=20 Eyes | | Group III=20 Eyes | | Total 60 Eyes | |
|---|---|---|---|---|---|---|---|---|
| | No. | % | No. | % | No. | % | No. | % |
| Operative | | | | | | | | |
| Hyphema | – | – | 2 | 10 | 2 | 10 | 4 | 6 |
| Lens touch | 2 | 10 | – | – | – | – | 2 | 3 |
| Early postoperative | | | | | | | | |
| Shallow AC | 8 | 40 | 6 | 30 | 8 | 40 | 22 | 37 |
| Lens opacity | 2 | 10 | – | – | – | – | 2 | 3 |
| Choroidal effusion | 4 | 20 | 6 | 30 | 4 | 20 | 14 | 23 |
| Corneal tube touch | – | – | – | – | 2 | 10 | 2 | 3 |
| Late postoperative | | | | | | | | |
| Tube exposure | – | – | 4 | 20 | 2 | 10 | 6 | 10 |
| [illegible] | [illegible] | [illegible] | [illegible] | [illegible] | [illegible] | [illegible] | [illegible] | [illegible] |
| Infection and loss of PL | – | – | 2 | 10 | – | – | 2 | 3 |

- shallow AC similar rates all Gps
- tube exposure MMC & Control Gps
  - no donor graft used
  - tube under partial thickness scleral flap

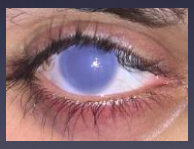

Madhy RAR. Adjunctive use of bevacizumab versus mitomycin C with Ahmed valve implantation in treatment of pediatric glaucoma. *J Glaucoma* 2011

# Glaucoma Drainage Devices

- Operation of choice for refractory childhood glaucomas
  - failed trabeculectomy and primary intervention (e.g. aphakia / uveitis)
- Similar success rates for all implants[1,2] ≈ 50% 5 years + meds
- High complication rates [3]
  - Consistently reported > commonly in children vs adults:
    - tube malposition, migration, retraction (surgery 1/3 cases)
    - tube erosion
    - endophthalmitis



GDD tube & plate erosion



Corneal decompensation following GDD

1. Tanimoto SA, Brandt JD. Options in pediatric glaucoma after angle surgery has failed. *Curr Opin Ophthalmol*. 2006
2. Chen A et al. Valved Glaucoma Drainage Devices in Pediatric Glaucoma: Retrospective Long-term Outcomes. *JAMA Ophthalmol* 2015.
3. Chen TC et al. Pediatric Glaucoma Surgery. A report for the AAO. *Ophthalmology* 2014

# Quality of Life (QoL)

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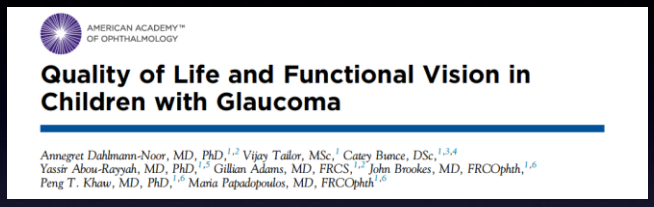
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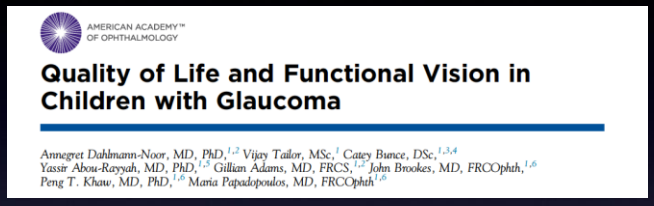
**Quality of Life and Functional Vision in Children with Glaucoma**

*Annegret Dahlmann-Noor, MD, PhD,[1,2] Vijay Tailor, MSc,[1] Catey Bunce, DSc,[1,3,4] Yassir Abou-Rayyah, MD, PhD,[1,5] Gillian Adams, MD, FRCS,[1,2] John Brookes, MD, FRCOphth,[1,6] Peng T. Khaw, MD, PhD,[1,6] Maria Papadopoulos, MD, FRCOphth[1,6]*

- 119 children with glaucoma (5-16yrs)
- self reporting questionnaires

Dahlmann-Noor A et al. Quality of Life and Functional Vision in children with glaucoma. *Ophthalmology* 2017

# Quality of Life (QoL)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Children with glaucoma:

- report markedly ↓ functional visual ability, VR- & HR-QoL scores compared to normal
- older children report less impairment than younger children
- parents state greater impact on QoL than children themselves
- HR-QoL scores similar to other chronic conditions (e.g. liver transplants) and cancer

Dahlmann-Noor A et al. Quality of Life and Functional Vision in children with glaucoma. *Ophthalmology* 2017

# Childhood Glaucoma in the 21st Century

## Conclusion

- Rare but significant cause of visual impairment
- Many unique features
- Management is challenging, especially surgery
- Advances mainly in surgery
  - 360° trabeculotomy, modified trabeculectomy technique, prospective evidence for anti-scarring agents (MMC, Avastin)
- Significant impact on child's QoL