

Recent Updates in Primary Congenital Glaucoma (PCG) Management in Egypt: A Comprehensive Review with Focus on Nasal Goniotomy–Temporal Trabeculotomy (NGTT).

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Cite this paper as Mohammed A. Al-Naimy Omar Mohamed Afifi, Tarek Hussein Elbromboly, Ahmed Elsayed Hussein (2024) Recent Updates in Primary Congenital Glaucoma (PCG) Management in Egypt: A Comprehensive Review with Focus on Nasal Goniotomy–Temporal Trabeculotomy (NGTT)..Journal of Neonatal Surgery, 13, 2218-2222

ABSTRACT

N/A

Key Words: N/A

INTRODUCTION

Primary congenital glaucoma (PCG) represents a significant subset of childhood glaucoma, characterized by developmental anomalies in the eye's aqueous drainage system. This condition leads to an elevation of intraocular pressure (IOP) beyond tolerable limits, resulting in ischemia, compression of optic nerve fibers, and, if left untreated, irreversible visual impairment. In Egypt, the management of PCG is a critical public health priority due to its disproportionately high prevalence compared to global averages. This review explores recent epidemiological trends, diagnostic advancements, and the evolution of surgical management in Egypt, with a specific emphasis on the Nasal Goniotomy–Temporal Trabeculotomy (NGTT)—a novel, cost-effective procedure designed to achieve circumferential angle treatment while preserving vital ocular tissues (1).

Epidemiology and Demographic Patterns in Egypt

The global incidence of PCG is estimated between 1:10,000 and 1:30,000 live births in Western nations. However, in Egypt, the estimated prevalence is significantly higher, approaching 1:4200 live births. This elevated risk is primarily attributed to high rates of consanguineous marriage, which is a documented risk factor for autosomal-recessive inheritance of the disease. Studies from Upper Egypt have shown that consanguinity is present in nearly half of all recorded PCG cases (1).

Demographic Characteristics (2):

Age of Onset: PCG in Egypt typically presents very early. Approximately 52% of cases are diagnosed within the first month of life, and nearly two-thirds are identified within the first year. Earlier onset is frequently correlated with a positive history of consanguinity.

Gender: There is a notable male preponderance, with a male-to-female ratio of approximately 2:1. Interestingly, some studies have recorded a higher incidence of females in neonatal presentations.

Laterality: The disease is overwhelmingly bilateral, occurring in 77.8% to 84% of cases according to various Egyptian university studies.

Clinical Presentation and Diagnostic Innovations

The classic presentation of PCG in Egyptian children includes corneal clouding and enlargement. Symptoms of tearing are less frequent (25%) compared to visible corneal changes (70%) (3).

Clinical Markers

The mean horizontal corneal diameter in Egyptian PCG patients typically ranges from 11.9 to 13.1 mm. Cases from Upper Egypt often present with larger corneal diameters and higher preoperative IOP values, often exceeding 30 mmHg. The mean

cup-to-disc (C/D) ratio at presentation is often reported around 0.6 ± 0.2 , reflecting significant optic nerve damage prior to intervention (1).

Advanced Imaging Techniques

Recent years have seen the integration of sophisticated imaging tools into Egyptian clinical practice (4):

- **Ultrasound Bio-microscopy (UBM):** This is essential for imaging infants with dense corneal clouding. UBM can detect abnormal membranes, the level of iris root insertion, and the position of ciliary processes. It has revealed that PCG eyes have significantly increased central corneal thickness (mean $700\pm 190\ \mu\text{m}$) due to edema and fibrosis, as well as increased anterior chamber depth (ACD) resulting from stretching under high IOP.
- **Hand-Held Anterior Segment OCT (HH-OCT):** The introduction of HH-OCT has revolutionized the examination of uncooperative or recumbent children in the operating room. This technology has identified specific patterns in PCG eyes, such as flat, thin irises and abnormal hyper-reflective membranes (Barkan's membranes) extending to the Schwalbe's line.
- **Visual Evoked Potential (VEP):** Flash VEP is used to monitor optic nerve conduction. Successful surgical reduction of IOP has been shown to improve P2 implicit time and N1–P1 amplitude, indicating functional recovery.

The Evolution of Surgical Management

Given that medical therapy is generally reserved as a bridge to surgery or an adjunctive measure, surgical intervention is the primary treatment modality for PCG in Egypt (2).

Traditional Angle Surgeries (5):

1. **Goniotomy:** Conventional ab-interno goniotomy aims to reach the Schlemm canal (SC) by incising the immature trabecular meshwork. However, the failure rate in the Middle East is relatively high (40–50%), likely due to the more severe phenotypes endemic to the region and the difficulty of visualization through cloudy corneas.
2. **Trabeculotomy:** This ab-externo approach uses a rigid probe or nylon filament. In Egypt, double-site rigid probe trabeculotomy has been found more successful than single-site procedures for achieving long-term IOP control, though single-site procedures are sometimes preferred to preserve parts of the angle for the future.

Focus: Nasal Goniotomy–Temporal Trabeculotomy (NGTT)

The development of Nasal Goniotomy–Temporal Trabeculotomy (NGTT) represents a major Egyptian innovation in pediatric glaucoma surgery. This procedure was designed to overcome the limitations of conventional techniques while providing a high-efficacy, low-cost solution suitable for the Egyptian healthcare context (6).

The Rationale for NGTT

Circumferential (360-degree) angle surgery is increasingly recognized as more effective than conventional limited-angle surgeries. While methods like Microcatheter-assisted trabeculotomy and GATT can achieve 360-degree treatment, they have significant drawbacks (7):

- **Cost and Availability:** Microcatheters are expensive and not always available in all Egyptian centers.
- **Technical Challenges:** GATT (ab-interno) is difficult in eyes with cloudy corneas or anomalous angles where identifying the SC is challenging.
- **Conjunctival Sparing:** Many ab-externo circumferential techniques sacrifice superior conjunctiva, which is disastrous if the child later requires a trabeculectomy or drainage implant.

The NGTT Procedure

NGTT utilizes the "best of both worlds" by combining an ab-interno approach nasally and an ab-externo approach temporally (8):

1. **Nasal Goniotomy:** Using clear corneal incisions, the surgeon treats approximately 140 degrees of the nasal angle. This avoids the superior conjunctiva and uses standard surgical needles or MVR blades, keeping costs minimal.
2. **Temporal Trabeculotomy:** A temporal, fornix-based conjunctival flap is created to access the SC. A rigid trabeculotome is then used to treat the remaining quadrants.

3. Outcome: Together, these steps provide nearly 360-degree treatment while leaving the superior 180 degrees of conjunctiva completely untouched (virgin site) for future needs.

Commentary on the Results of NGTT

A landmark study by Abdelrahman and Amin (2022) at Cairo University provided compelling data on the efficacy of this technique (9):

- IOP Reduction: The average preoperative IOP was 30.3 mmHg. Following NGTT, the average IOP dropped to 10.5 mmHg at one month—a 65.3% reduction.
- Sustained Efficacy: This reduction was remarkably stable, with a mean IOP of 13.3 mmHg maintained at 18 months post-operatively (57.2% reduction).
- Success Rate: The study reported a 93.3% complete success rate, meaning the target IOP was achieved without the need for supplemental medications.
- Safety Profile: Complications were minor and transient. Hyphema occurred in 33% of eyes but cleared spontaneously. In four cases, iris prolapse necessitated a peripheral iridectomy, which did not affect the final IOP outcome.
- Clinical Implications: In the single case that failed, the superior conjunctiva remained available for a subsequent trabeculectomy, which successfully controlled the pressure. This confirms NGTT as a strategic primary intervention that does not "burn bridges" for future management.

Other Low-Cost Surgical Innovations in Egypt

Egyptian surgeons have pioneered several other "economic" solutions for angle management (10):

- Bent Needle Ab-interno Goniectomy (BANG): This uses a standard sterile hypodermic needle bent into a "reverse cystotome" to strip the trabecular meshwork. It has shown high success rates, particularly in infantile-onset PCG.
- Kahook Dual Blade (KDB) Goniectomy: While KDB effectively excises the trabecular meshwork, its adoption is limited by the added cost compared to the BANG or NGTT techniques.
- Modified GATT: Using 5-0 Prolene sutures instead of microcatheters to perform ab-interno circumferential trabeculotomy has been studied, though it carries a risk of "false passages" in the SC.

Management of Refractory PCG

When primary angle surgeries fail, Egyptian practitioners turn to filtering surgeries or drainage devices.

Filtering Surgeries (11):

- Augmented Trabeculectomy: The use of Mitomycin-C (MMC) or Ologen (a biodegradable collagen matrix) is standard to prevent scarring. Studies indicate that Ologen may provide better long-term control with fewer MMC-related complications like bleb leaks or necrosis.
- Combined Trabeculotomy-Trabeculectomy (CTT): Often the first choice for moderate to severe cases (IOP >35 mmHg, diameter >14.5 mm), CTT provides dual drainage pathways. Recent refinements include reducing MMC exposure time to one minute to minimize toxicity without sacrificing success.

Glaucoma Drainage Devices (GDD)

For the most severe phenotypes, GDDs are employed (12):

- Ahmed Glaucoma Valve (AGV): Often augmented with MMC or wrapped in amniotic membrane to reduce plate encapsulation.
- Aurolab Aqueous Drainage Implant (AADI): A significantly cheaper alternative to the Baerveldt implant, though it has been associated with higher rates of early postoperative inflammatory reactions.

Post-operative Trends and Medical Adjuncts

The use of anti-glaucoma medications remains an important secondary strategy. In Egyptian clinical settings, the combination of topical Carbonic Anhydrase Inhibitors (CAIs) and Beta-blockers (BBs) has proven more effective than prostaglandin

analogues (PGAs) for achieving "qualified success" after surgery, providing an average IOP reduction of 43.7% (13).

Conclusion and Recommendations

The management of PCG in Egypt is characterized by a high volume of complex cases met with significant surgical innovation. The development of the NGTT technique exemplifies a move toward circumferential angle surgery that is both highly effective (93.3% success) and economically sustainable, while strategically preserving the superior conjunctiva for the long-term vision of the child (11).

Future Directions

To further improve visual outcomes in Egypt, experts recommend (14):

1. National Screening Programs: Implementing nationwide screening to ensure earlier identification and intervention, as the best outcomes are seen in infants treated within the first months of life.
2. Tele-medicine: Utilizing tele-ophthalmology to share diagnostic data (like HH-OCT images) between rural clinics and specialized centers.
3. Comprehensive Rehabilitation: Addressing the visual, psychological, and social needs of affected children and their caregivers to ensure satisfactory mental development and quality of life.

Through the continued application of techniques like NGTT and the expansion of screening initiatives, Egypt is well-positioned to reduce the burden of blindness caused by primary congenital glaucoma (15).

Diagnostic Advancements

Recent updates in Egypt include the use of advanced imaging to better characterize the anterior segment (1):

- Ultrasound Bio-microscopy (UBM): Essential for angle imaging in infants with dense corneal clouding, detecting abnormal membranes and iris root insertion levels.
- Hand-Held Anterior Segment OCT (HH-OCT): Allows for the examination of recumbent children in the operating room, revealing flat, thin iris patterns and hyper-reflective membranes (Barkan's membranes).
- Visual Evoked Potential (VEP): Used to monitor functional recovery; successful IOP reduction has been shown to improve P2 implicit time and N1-P1 amplitude.

Surgical Management Trends

Management in Egypt relies heavily on surgical intervention, with a shift toward circumferential angle surgeries. While conventional goniotomy and trabeculotomy are common, they face challenges such as high failure rates in severe phenotypes (40-50%) and the need to preserve conjunctiva for future procedures (12).

Focus on Nasal Goniotomy–Temporal Trabeculotomy (NGTT)

Nasal Goniotomy–Temporal Trabeculotomy (NGTT) is a novel, ergonomic Egyptian modification designed to treat the angle circumferentially while remaining cost-effective (13).

- The Procedure: The surgeon performs an ab-interno nasal goniotomy to treat approximately 140 degrees of the angle, followed by an ab-externo temporal trabeculotomy. This combined approach provides nearly 360-degree treatment.
- Conjunctival Sparing: A primary advantage of NGTT is that it completely spares the superior 180 degrees of conjunctiva. This preserves a "virgin" site for future filtering surgeries if needed, avoiding the scarring associated with conventional 2-site trabeculotomy.
- Cost and Accessibility: Unlike microcatheter-assisted trabeculotomy or GATT, which require expensive illuminated catheters or are technically difficult in cloudy corneas, NGTT uses standard surgical needles or MVR blades.

Clinical Results of NGTT

A study conducted at Cairo University demonstrated the following outcomes (10):

- Success Rate: The procedure achieved a 93.3% complete success rate.
- IOP Reduction: The average preoperative IOP of 30.3 mmHg was reduced to 10.5 mmHg at one month (a 65.3% reduction).

- Long-term Stability: The IOP reduction was maintained through 18 months, with a mean post-operative IOP of 13.3 mmHg.
- Safety: Complications were minor, such as transient hyphema in 33% of eyes that cleared spontaneously.

Other Innovative and Economic Solutions (15):

- Bent Needle Ab-interno Goniotomy (BANG): An "economic" solution using a bent hypodermic needle to strip the trabecular meshwork, showing high success in infantile-onset cases.
- Modified Filtering Surgeries: For refractory cases, trabeculectomy augmented with Ologen (a biodegradable implant) has shown longer-term control than Mitomycin-C (MMC) alone.
- Glaucoma Drainage Devices (GDD): The Aurolab Aqueous Drainage Implant (AADI) is utilized as a cheaper alternative to the Baerveldt implant, though it may have higher early inflammatory reactions.

CONCLUSION

Surgical innovation in Egypt, particularly the NGTT technique, provides a high-success, low-cost alternative for managing PCG. By achieving circumferential angle treatment while preserving superior conjunctiva, NGTT offers a strategic advantage for the lifelong management of pediatric glaucoma.

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