PATIENT CHARACTERISTICS OF CHILDREN PRESENTING WITH PRIMARY CONGENITAL GLAUCOMA TO A TERTIARY REFERRAL CENTER

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Introduction

Primary congenital glaucoma (PCG) is a rare, genetically derived, developmental defect in the trabecular meshwork and anterior chamber angle that occurs without other significant ocular or systemic anomalies. Other features, e.g., elevated intraocular pressure (IOP); corneal enlargement, or presence of Haabstriae or corneal opacity, or both; and progressive optic nerve cupping, may also be present. The incidence of PCG ranges from ~1 in 10,000–30,000 births in Western countries to the highest of 1:1250 in Slovakian Gypsies. Defined ethnic groups, e.g., Pakistani, Bangladeshis, and Indians, with an elevated level of consanguinity within them, tend to have increased incidences. The child's glaucoma manifests with one or more of the "classic triad" of findings: epiphora, photophobia, and blepharospasm. Surgical intervention constitutes the definitive treatment modality for PCG. Medications play an adjunctive role to surgery in the therapy of PCG, Preoperatively, medications may help clear the cornea to facilitate goniotomy, and postoperatively, they may help control IOP until the adequacy of the surgical procedure has been verified.

Aim of the work

The aim of this study was to report the presenting characteristics of the children presenting with Primary Congenital Glaucoma to Alexandria Main University Hospital.

Subjects and Methods

Patients: The study was a retrospective chart review of the medical records of children presenting with Primary Congenital Glaucoma to the Ophthalmology department of Alexandria Main University Hospital from the year 2005 till the year 2018.

Inclusion criteria: Patients < 18 years with primary congenital glaucoma.

Exclusion criteria:

- Patients> 18 years on presentation. -Patients with Secondary glaucoma.

Methods: The following data was extracted from the patients' preoperative medical records: Patients Demographics, Antenatal history, Natal and Postnatal history and Clinical ophthalmic examination (and investigations) preoperatively: Initial office examination was followed by an examination under general anesthesia to obtain information including: Intraocular pressure (measured using Perkins hand-held applanation tonometer) (measured using catroveijo surgical caliper as horizontal white-to-white) (using indirect ophthalmoscope if the media clarity allowed visibility of posterior segment structures).

Axial length measurement (using A-scan ultrasonography).

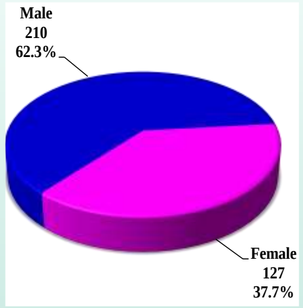
B-scan ultrasound if media clarity precluded clinical fundoscopy.

Central Corneal Thickness measurement (using the Pachmate ®, DGH technology, when this was available).

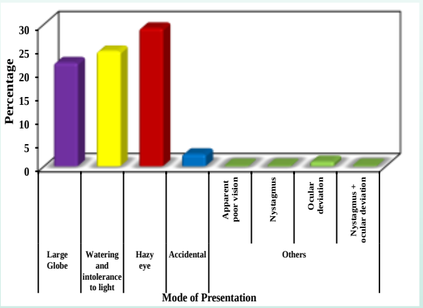
Results

Table 1: This study was conducted on 503 eyes of 337 children with the following results

Demographic characteristics of study children	No.	%
Study children	337	100.0
Male	210	62.3
Female	127	37.7
Age at presentation (months) **(Age at which the child came asking for medical advice)**	(n=337)	
Min. – Max.	0.53 – 109.93	
Mean ± SD.	8.34 ± 11.58	
Median (IQR)	5.0 (3.27–7.90)	
Age at onset (months) **(Age at which the child developed first symptoms of the disease)**	(n=268)	
Min. – Max.	0.0 – 75.0	
Mean ± SD.	3.33 ± 6.97	
Median (IQR)	2.0 (0.0–4.0)	
Mode of Presentation		
Large Globe	110	21.9
Watering and intolerance to light	122	24.3
Hazy eye	147	29.2
Accidental	13	2.6
Others	25	5.0
Apparent poor vision	1	0.2
Nystagmus	1	0.2
Ocular deviation	5	1.0
Nystagmus + ocular deviation	1	0.2
Parental consanguinity	(n=332)	
Negative	196	58.2
Positive	136	40.4
Mode of Delivery	(n=330)	
NVD	167	49.6
CS	163	48.4
Term	(n=325)	
Full term	316	93.8
Preterm	9	2.7



Study childrenof study children % from total (n=337).



Mode of presentation of study children % from total (n=337).

Conclusion

In children presenting with PCG to Alexandria Main University Hospital:

1. There is a slight male predominance.
2. Children present at an age that is 5 months after onset of symptoms and 3 months later than the standard for other populations.
3. The most common presentations are a cloudy cornea, a big cornea and light intolerance.
4. Factors significantly related to patients clinical characteristics include age at presentation, sex of patient, parental consanguinity, antenatal history, family history and preterm delivery.
5. Hazy cornea was the most presenting symptom, but clear cornea does not exclude the diagnosis and increase intraocular pressure is not the presenting symptom.
6. PCG running in families tends to be more severe than in sporadic cases.
7. Consanguineous parents had children with more severe disease.