

# OPHTHALMIC FEATURES OF CHILDREN WITH ANIRIDIA PRESENTING TO ALEXANDRIA MAIN UNIVERSITY HOSPITAL

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## INTRODUCTION

Aniridia is defined as Partial or complete absence of the iris. It may be associated with systemic disorders and ocular disorders. It presents as congenital aniridia which is rare and may be acquired after trauma or ocular surgery. The most common variant of congenital aniridia is a de novo mutation of the PAX6 gene. Presentation may be by poor vision since childhood. Nystagmus and strabismus are common. Iris may be complete absence. Signs of keratopathy can appear in first decade of life. The anterior chamber is open on gonioscopy, and congenital glaucoma / buphthalmos is rare in aniridia. However, within the 2nd -decade angle closure due to peripheral anterior synechia may lead to an increase in the intraocular pressure. Gonioscopy may show iris fibers which lie at the space between the rudimentary small hypoplastic iris and the trabecular meshwork. The lens in aniridia may show superior subluxation (clear or cataractous), ectopia lentis (noted in up to 56% of cases). Foveal hypoplasia are also present.

## AIM OF THE WORK

The study aim was to report on the ophthalmic eye features with aniridia in children presenting to Alexandria Main University Hospital from 2005 to 2020.

## PATIENTS AND METHODS

This study was a retrospective chart review. From 2005 to 2020 ophthalmic records of children presenting with Aniridia to the Ophthalmic outpatient clinic of Alexandria Main University Hospital were reviewed.

Data extracted: Gender and age (from 1 month to 16 years old), history (antenatal, natal, and postnatal), surgical and medical history, family history, and data on parental consanguinity. Clinical examination data (Anterior segment examination data by portable slit lamp: Cornea, Iris, dilated pupil, and lens. Intraocular pressure by Perkin's applanation tonometer and Fundus examination by indirect ophthalmoscopy. Axial length, Refraction, and systemic examination data if available). Aniridia was defined by partial or complete absence of the iris. Glaucoma was defined by both an IOP above 16mmHg and a C/D ratio above 0.4. Presence of one defined as a glaucoma suspect. For children subjected to glaucoma surgery details of the surgery were extracted from the records as well as follow-up data for up to 36 months.

## RESULTS

Table 1 : Relation between sex and postoperative IOP

IOP (value)	Sex		U	p
	Males	Females		
<b>1 month</b>	(n = 3)	(n = 5)		
Mean ± SD.	12.67 ± 8.33	10.40 ± 9.74	4.50	0.393
Median (Min. – Max.)	10.0 (6.0 – 22.0)	4.0 (2.0 – 22.0)		
<b>3 months</b>	(n = 3)	(n = 3)		
Mean ± SD.	17.0 ± 8.19	17.0 ± 10.15	4.50	1.000
Median (Min. – Max.)	15.0 (10.0 – 26.0)	15.0 (8.0 – 28.0)		
<b>6 months</b>	(n = 3)	(n = 5)		
Mean ± SD.	19.33 ± 5.77	17.40 ± 11.65	6.0	0.786
Median (Min. – Max.)	16.0 (16.0 – 26.0)	11.0 (6.0 – 30.0)		
<b>12 months</b>	(n = 4)	(n = 4)		
Mean ± SD.	14.75 ± 2.22	13.0 ± 6.16	8.0	1.000
Median (Min. – Max.)	15.0 (12.0 – 17.0)	15.0 (4.0 – 18.0)		
<b>24 months</b>	(n = 4)	(n = 2)		
Mean ± SD.	11.50 ± 1.91	14.0 ± 2.83	1.500	0.267
Median (Min. – Max.)	11.0 (10.0 – 14.0)	14.0 (12.0 – 16.0)		
<b>36 months</b>	(n = 3)	(n = 1)		
Mean ± SD.	10.0 ± 5.29	16.0	0.500	0.500
Median (Min. – Max.)	8.0 (6.0 – 16.0)			

U: Mann Whitney test

p: p value for comparing males and females and postoperative IOP

Table 2 : Correlation between age at presentation and postoperative IOP

IOP (value)	N	Age (at presentation in months)	
		r <sub>s</sub>	p
<b>1 month</b>	8	-0.847*	0.008*
<b>3 months</b>	6	-0.672	0.144
<b>6 months</b>	8	-0.939*	0.001*
<b>12 months</b>	8	-0.555	0.153
<b>24 months</b>	6	-0.123	0.816
<b>36 months</b>	4	0.000	1.0000

r<sub>s</sub>: Spearman coefficient

\*: Statistically significant at p ≤ 0.05

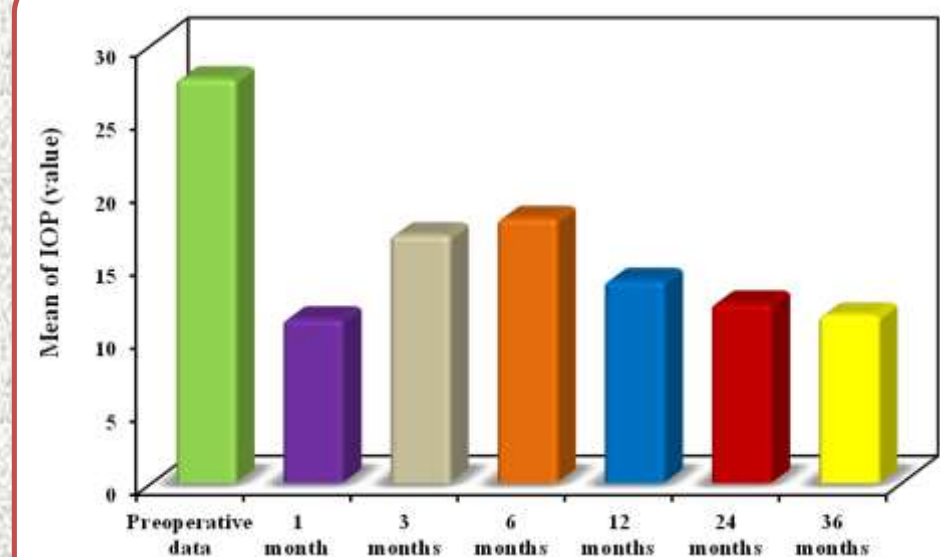


Figure 1: Distribution of the studied cases according to IOP (value)

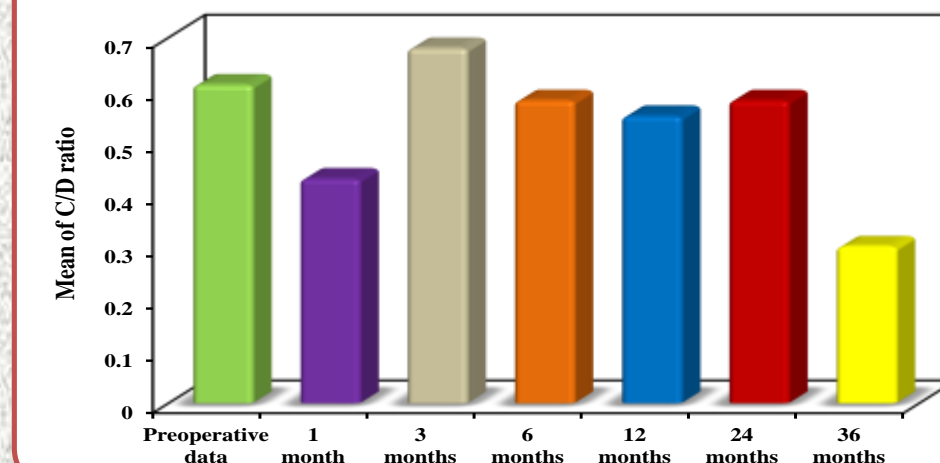


Figure 2: Distribution of the studied cases according to C/D ratio.

## CONCLUSION

Glaucoma is still one of the most difficult aspects of aniridia. Eyes with aniridia typically develop glaucoma in late childhood or early adulthood as the drainage angle undergoes progressive anatomical changes. The procedure of combined trabeculotomy - trabeculectomy with mitomycin C for glaucoma with aniridia has a success rate of 50% and results in a reduction of IOP that is sustained for at least 3 years.