Central Corneal Thickness in Congenital Aniridia and its Role in Glaucoma in an Indian Population

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Aim: *To determine the range and distribution of central corneal thickness in patients with aniridia and compare with age- and sex-matched controls.*
Methods: *This was a prospective cross-sectional observational case-control study performed at a tertiary eyecare hospital. All patients diagnosed with aniridia from June 2006 to April 2008 were enrolled. Patients were excluded if they had corneal oedema, central corneal scars, central aniridic keratopathy, or any history or signs suggestive of previous surgery or injury. Central corneal thickness was measured by ultrasound pachymetry and the mean of 5 measurements per eye was used as the study measurement. The results were compared with those of age- and sex-matched controls.*
Results: *The mean central corneal thickness in the aniridia group was 0.603 mm compared with 0.566 mm in the control group (p = 0.0083).*
Conclusions: *This study has determined the range and distribution of CCT in Indian patients with aniridia and substantiates the fact that patients with aniridia in an Indian cohort also have significantly thicker corneas. This needs to be considered for making accurate IOP measurements in these patients to ascertain the appropriate management.*

Keywords: *Aniridia, Cornea, Glaucoma*

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Introduction

Congenital aniridia is a hereditary condition associated with partial or complete absence of iris. Congenital aniridia has a low incidence rate of 18 per 100,000 live births (0.018%). Other associated findings include abnormalities of the conjunctiva and corneal surface, hypoplasia of the fovea and\or the optic nerve, nystagmus, and glaucoma.[1]

Autosomal dominant aniridia is the most common form, and is not associated with any systemic disorder. Congenital sporadic aniridia is found in association with Wilms tumour nephroblastoma, genitourinary abnormalities, and mental retardation (Miller syndrome) and is known as WAGR syndrome (Wilms tumour, Aniridia, Genitourinary abnormalities, Retardation). WAGR syndrome is linked with partial deletions of the short arm of chromosome 11 (11p13). Autosomal recessive aniridia is the least common type and is associated with cerebellar ataxia and mental retardation (Gillespie syndrome).

Glaucoma develops in approximately 50% of patients with aniridia, usually after the second decade of life when anatomical changes occur in the angle secondary to adhesion of peripheral iris strands to the outflow structures. The iris strands bridge the space between the iris stump and trabecular meshwork, and progressive contracture of the iris strands leads to angle closure glaucoma. Goniodysgenesis is also noted in some patients.

In the presence of corneal scars precluding the fundus view and the impracticality of visual field examination due to associated nystagmus, the diagnosis of glaucoma on the basis of elevated intraocular pressure (IOP) alone is not possible and relative afferent papillary defect cannot be detected. Therefore, IOP measurement corrected for central corneal thickness (CCT) is essential for diagnosis,[2,3] as aniridia is associated with thicker corneas.[4,5]

This study was performed to determine the range and distribution of CCT in patients with aniridia and compare with age- and sex-matched controls.

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Table 1. Central corneal thickness measurements among patients with aniridia and age- and sex-matched control participants.

Patient number	Aniridia				Controls			
	Age (years)	Sex	Central corneal thickness (mm)		Age (years)	Sex	Central corneal thickness (mm)	
			Right	Left			Right	Left
1	0.5	Male	0.496	0.548	0.5	Male	0.454	0.453
2	4	Female	0.584	0.562	4	Female	0.738	0.761
3	5	Male	0.536	0.535	5	Male	*	0.567
4	5	Male	0.630	0.633	5	Male	0.584	0.576
5	6	Male	0.568	0.588	6	Male	0.584	0.591
6	8	Female	0.617	0.602	8	Female	0.566	0.568
7	8	Female	0.661	0.644				
8	9	Male	0.584	0.584	9	Male	0.541	0.545
9	9	Female	0.619	0.622	9	Female	0.584	0.601
10	9	Female	0.639	0.611				
11	10	Female	*	0.632	11	Female	0.563	0.483
12	12	Male	0.656	0.649	12	Male	0.568	0.552
13	12	Male	0.627	0.630				
14	13	Female	0.638	0.624	13	Female	0.556	0.583
15	13	Male	0.641	0.651				
16	22	Male	0.582	0.584	22	Male	0.519	0.519
17	24	Female	0.546	0.566	24	Female	0.535	0.552
18	30	Male	*	0.617	30	Male	0.568	0.568

* Central corneal thickness not measured due to corneal pathology.

Methods

The study was conducted at the Department of Glaucoma, Aravind Eye Hospital and Post Graduate Institute of Ophthalmology, Madurai, India. This was a prospective cross-sectional observational case-control study of all patients diagnosed with aniridia from June 2006 to April 2008. Inclusion criteria were children and adults with congenital absence of iris (total or partial) diagnosed clinically as aniridia. Patients were excluded if they had corneal oedema, central corneal scars, central aniridic keratopathy, or any history or signs suggestive of previous surgery or injury.

The study was approved by the institute's ethics committee and adhered to the principles of the Declaration of Helsinki. Informed consent was given by all the participants in the study.

CCT was measured by ultrasound pachymetry using the PACSCAN 300p model (Sonomed Escalon, Lake Success, USA). The average of 5 measurements for each eye was taken to reduce the likelihood of error induced by off-centre measurements in patients with nystagmus. The results were compared with corresponding age- and sex-matched controls.

Two-sample *t* tests were used to test for differences in age and sex between the patients with aniridia and the controls and for difference in mean CCT between the 2 groups.

Results

Twenty seven patients were enrolled during the study period. Nine patients had corneal pathology (corneal scar [n = 4], central aniridic keratopathy [n = 3], corneal oedema [n = 2]) and were excluded. The study population included 10 male and 8 female patients; 2 eyes could not be measured due to corneal pathology. Fourteen age- and sex-matched healthy participants were enrolled as a control group. There were 8 male and 6 female patients; 1 eye could not be measured due to corneal pathology (Table 1).

The mean CCT in the group with aniridia was 0.601 mm in the right eye and 0.605 mm in the left eye for a mean CCT of 0.603 mm. The mean CCT in the control group was 0.566 mm in the right eye and 0.566 mm in the left eye for a mean CCT of 0.566 mm (Table 2). There was a significant difference between the mean CCT of patients with aniridia and the controls by two-sample *t* test ($p = 0.0083$).

The null hypothesis for any significant differences between the average age and sex of patients with aniridia and the controls by two-sample *t* test with equal variances was 0 ($p = 0.6093$ and $p = 0.9801$, respectively).

Table 2. Average values of central corneal thickness in patients with aniridia and age- and sex-matched control participants.

Participants	Central corneal thickness (mm)	
	Right	Left
Aniridia	0.601	0.605
Controls	0.566	0.566
Overall average	0.587	
Aniridia average	0.603	
Control average	0.566	

Discussion

Patients with aniridia have thicker corneas than healthy individuals. Brandt et al have shown a mean CCT of 0.632 mm in patients with aniridia.[4] Whitson et al compared the CCT of a healthy population with that of patients with congenital aniridia and found that patients with aniridia have thicker corneas, with a CCT of 691.0 μm (SD, 75.4 μm) compared with a CCT of 548.2 μm (SD, 21.2 μm) for control participants.[5]

The diagnosis of glaucoma in patients with aniridia is complicated by multiple challenges. Corneal scars, present in most patients with aniridia, preclude an adequate view of the fundus so appreciation of early glaucomatous damage becomes difficult. The presence of nystagmus secondary to optic nerve/foveal hypoplasia compounds the problem, and obtaining a reliable visual field is almost impossible. Clinicians therefore have to depend primarily on the IOP measurements to exclude glaucoma in these patients.[6]

Variations in CCT change the resistance of the cornea to indentation so that it is no longer balanced entirely by the tear film surface tension, which affects the accuracy of the IOP measurement. A thinner cornea requires less force to applanate it than a thick cornea, leading to an underestimation of the true IOP measurement, while a thicker cornea will require more force for applanation, giving an artificially high IOP measurement. The Goldmann applanation tonometer is designed to give accurate readings when the CCT is 520 μm.

An extensive search of the literature has shown that various correction factors have ranged from approximately 1.00 mm Hg to 3.57 mm Hg for every 50 μm deviation.[7] Therefore, corrected IOP measurements for the corresponding CCT is essential before embarking on a diagnosis of glaucoma and subjecting these patients to lifelong medications or surgical intervention. Management of glaucoma is usually surgical for these patients, as pharmacological therapy usually fails, and consists of prophylactic goniotomy, trabeculectomy, and fistulisation procedures.

Tonometry may not be reliable for patients with aniridia, even after correction for the corresponding CCT, emphasizing the importance of regular gonioscopy and optic nerve examination to identify the presence or progression of glaucomatous pathology.

This study has determined the range and distribution of CCT in Indian patients with aniridia and substantiates the fact that patients with aniridia in an Indian cohort also have significantly thicker corneas. This needs to be considered for making accurate IOP measurements in these patients to ascertain the appropriate management.

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