Volume 32 • Issue 3 • September-December 2020

ISSN: 0976-6677

Kerala Journal of OPHTHALMOLOGY

The Official Journal of Kerala Society of Ophthalmic Surgeons

www.kjophthal.com



HIGHLIGHTS

Emerging retinal diseases and newer terminologies in SD OCT

Astigmatism Correction with Toric Intraocular Lens

NCDs and NCEDs: What is the way forward?

Post graduate surgical training in ophthalmology

Eye Grader - Smartphone App for Clinical Grading Scales in Ophthalmology

Sir Harold Ridley and the Intraocular Foreign Body that made history

Wolters Kluwer

Medknow

ISSN 0976-6677

Kerala Journal of Ophthalmology

Editorial Board

Editor
Sudha V

Managing Editor
Sreeni Edakhlon

Editorial Committee

Biju John C
Lathika Kamaladevi
Prashob Mohan
Sinumol S
Dhanya Cheriyath
Remya Mareen Paulose
Suma Unnikrishnan
John Davis Akkara
Babitha V

Advisory Committee

Anantharaman Giridhar
Ani Sreedhar
Arup Chakrabarti
Elizabeth Joseph K
Ganesh V Raman
Mahesh G
Minu Mathen
C V Anthrayose Kakkanatt
Thomas Cherian
Somasheila Murthy
Santosh G Honavar

Emeritus Editors

Anantharaman Giridhar
Ashok Nataraj
Bastin VA
Gopal S Pillai
KE Eapen
Krishnankutty
Mahadevan K
Mahesh G
Meena Chakrabarti
Narayanankutty K
NSD Raju
PI Mohan
Saikumar SJ
Sasikumar S
Thomas Cherian
TP Ittyerah

Cover Story:

Description:

The cover photo shows an anteriorly dislocated hypermature cataract in a 84 year old lady, with anterior capsular calcification and areas of absorption in the cataract.

Contributed by: Hridya Mohan, Medical Officer, Cataract Services, Aravind Eye Hospital, Coimbatore

Kerala Journal of Ophthalmology

Volume 32 Issue 3 September-December 2020

Contents

Editorial

The silent epidemic – Are we neglecting it?
V. Sudha 219

Guest Editorial

Postgraduate surgical training in ophthalmology
Vivek Oommen Varghese, Thara R. Nair, K. Mahadevan 221

Perspective

Noncommunicable diseases and noncommunicable eye diseases: What is the way forward?
Meena Chakrabarti 224

Ophtha Insta

Pigment sign in vernal keratoconjunctivitis
Amanda Mohanan-Earatt, Sanjeev Srinivas 232

Major Reviews

Emerging retinal diseases and newer terminologies in spectral domain optical coherence tomography
Divya Alex, Anantharaman Giridhar, Mahesh Gopalakrishnan, Shivam Madan, Swati Indurkhya, Swati Haridas, Sailesh Kumar, Dinesh Rungta 234

Astigmatism correction with toric intraocular lens during cataract surgery
Sandhya N, Keerthana Zacharia, Saikumar S. J 244

Original Articles

Ocular biometry in an adult Ghanaian population
Samuel Kyei, Frank Assiamah, Michael Agyemang Kwarteng, Victor Kwesi Ansah 252

Trial for a trial: Is there hope? – Optic Neuritis Treatment Trial protocol in patients with traumatic optic neuropathy
Jainy Joseph Emmatty, Smita Narayan, Rajini K. Cherayath 258

Functional blindness: Blepharospasm with apraxia of eyelid opening
Titap Yazicioglu 263

A study on changes in choroidal thickness in antenatal women with pregnancy-induced hypertension using spectral-domain-optical coherence tomography
Sozhamadevi Subramaniyan, Venkatesh Perumal, Thenmozhi R 268

Clinical presentation and outcome of patients with optic pathway gliomas: A series of twenty patients
Sudha Menon, Girish Menon, Reddy Vyjayanth, Neenu George, Vindhya Jagdish 271

Efficacy of Trash To Treasure Retcam in Diabetic Retinopathy Screening
Prithvi Chandrakanth, K. S. Chandrakanth, A. K. Sheeja Vishwanathan, R. Nirupama, Tresa Mathew 278

Surgical Corner

Basics of fluid air exchange in vitreoretinal surgery
Gaganjeet Singh Gujral 284

Case Reports

Ocular juvenile xanthogranuloma
Shaji Hussain, Anu Mariya Paul 287

Iridocorneal endothelial syndrome – A case of contradictions
Jincy Mariya Paul, Padma B. Prabhu, Charmaine B. Solomon 291

Scleral patch graft as a rescue for infectious scleritis
Hilda K. Nixon, Lilan Bhat, David Pudukadan, Neena Chris William 295

Primary conjunctival non-Hodgkin's marginal zone lymphoma
Nareddy Prasanna 299

Photo Essay

A challenging case of refractory childhood glaucoma associated with bilateral congenital ectropion uveae
V. K. Harshavardhan, Meena Menon 302

Innovations

Eye grader-smartphone app for clinical grading scales in ophthalmology
John Davis Akkara, Anju Kuriakose 305

Clinical Challenge

Unilateral abducent nerve palsy in a diabetic patient with treated cerebellopontine angle tumor
Sanitha Sathyan 311

History Corner

Sir Harold Ridley and the intraocular foreign body that made history
C. Biju John 315

Journal Review

S Reshmi 321

PG Corner

Bharat Gurnani, Kirandeep Kaur, Poulomi Das 323

Letters to the Editor

Comment on "Role of mitomycin C in the management of ocular surface squamous neoplasia"
Bharat Gurnani, Kirandeep Kaur 329

Comment on ' Comparison of the scleral tunnel constructed with crescent versus razor blade using the anterior segment optical coherence tomography"
Kirandeep Kaur, Bharat Gurnani 331

Kerala Journal of Ophthalmology

General Information

The journal
Kerala Journal of Ophthalmology (KJO), the official publication of Kerala Society of Ophthalmic Surgeons (KSOS) is published as print and online journal.

Abstracting and Indexing information
Baidu Scholar, CNKI (China National Knowledge Infrastructure), EBSCO Publishing's Electronic Databases, Ex Libris – Primo Central, Google Scholar, Hinari, Infotrieve, National Science Library, ProQuest, TdNet, Wanfang Data

The journal is indexed with, or included in, the following:

DOAJ

Information for Authors
There are no page charges for submissions to the journals. Please check http://www.kjophthal.com/contributors.asp for details.

All manuscripts must be submitted online at www.journalonweb.com/kjo.

Subscription Information
Copies are provided free of cost to members of KSOS. A subscription comprises of 3 issues every year. Please include postage.

Annual subscription rates for non-members

Institutional - INR 1200.00 for India and USD 100.00 for outside India
Personal - INR 600.00 for India and USD 50.00 for outside India
For mode of payment and other details, please visit www.medknow.com/subscribe.asp.

Claims for missing issues will be serviced at no charge if received within 60 days of the cover date for domestic subscribers, and 3 months for subscribers outside India. Duplicate copies cannot be sent to replace issues not delivered because of failure to notify publisher of change of address.

The journal is published and distributed by Wolters Kluwer India Private Limited. Copies are sent to subscribers directly from the publisher's address. It is illegal to acquire copies from any other source. If a copy is received for personal use as a member of the association/society, one cannot resale or give-away the copy for commercial or library use.

The copies of the journal to the members of the association are sent by ordinary post. The editorial board, association or publisher will not be responsible for non receipt of copies. If any member/subscriber wishes to receive the copies by registered post or courier, kindly contact the publisher's office. If a copy returns due to incomplete, incorrect or changed address of a member/subscriber on two consecutive occasions, the names of such members will be deleted from the mailing list of the journal. Providing complete, correct and up-to-date address is the responsibility of the member/subscriber.

Nonmembers: Please send change of address information to wkhlrpmedknow_subscriptions@wolterskluwer.com.

Advertising policies
The journal accepts display and classified advertising. Frequency discounts and special positions are available. Inquiries about advertising should be sent to Wolters Kluwer India Private Limited, advertise@medknow.com.

The journal reserves the right to reject any advertisement considered unsuitable according to the set policies of the journal.

The appearance of advertising or product information in the various sections in the journal does not constitute an endorsement or approval by the journal and/or its publisher of the quality or value of the said product or of claims made for it by its manufacturer.

Copyright
The entire contents of the Kerala Journal of Ophthalmology are protected under Indian and international copyrights. The Journal, however, grants to all users a free, irrevocable, worldwide, perpetual right of access to, and a license to copy, use, distribute, perform and display the work publicly and to make and distribute derivative works in any digital medium for any reasonable non-commercial purpose, subject to proper attribution of authorship and ownership of the rights. The journal also grants the right to make small numbers of printed copies for their personal non-commercial use.

Permissions
For information on how to request permissions to reproduce articles/information from this journal, please visit www.kjophthal.com

Disclaimer
The information and opinions presented in the Journal reflect the views of the authors and not of the Journal or its Editorial Board or the Publisher. Publication does not constitute endorsement by the journal. Neither the Kerala Journal of Ophthalmology nor its publishers nor anyone else involved in creating, producing or delivering the Kerala Journal of Ophthalmology or the materials contained therein, assumes any liability or responsibility for the accuracy, completeness, or usefulness of any information provided in the Kerala Journal of Ophthalmology, nor shall they be liable for any direct, indirect, incidental, special, consequential or punitive damages arising out of the use of the Kerala Journal of Ophthalmology. The Kerala Journal of Ophthalmology, nor its publishers, nor any other party involved in the preparation of material contained in the Kerala Journal of Ophthalmology represents or warrants that the information contained herein is in every respect accurate or complete, and they are not responsible for any errors or omissions or for the results obtained from the use of such material. Readers are encouraged to confirm the information contained herein with other sources.

Address
Editorial Office
Dr. Sudha V
MS (Ophthalmology), DNB (Ophthalmology)
Additional Professor, Govt.Medical College, Thrissur Kerala, India.
E-mail: kjoeditor2020@gmail.com
Phone - 9446619280

Published by
Wolters Kluwer India Private Limited.
A-202, 2nd Floor, The Qube, C.T.S. No.1498A/2 Village Marol,
Andheri (East), Mumbai - 400 059, India.
Phone: 91-22-66491818
Website: www.medknow.com

Printed at:
Nikeda Art Printers Pvt Ltd.
Bhandup (W), Mumbai, India.

Editorial

The silent epidemic – Are we neglecting it?

The COVID pandemic has brought life literally to a halt. Health-care systems have weakened and health-care delivery has been put on the back burner. Elderly men and women who suffer from multiple comorbidities are the most affected as restrictions have been imposed on their movements and lifestyles, including getting fresh air and exercise in lieu of them being most susceptible to the virus. The aftermath of the corona pandemic will be equally problematic with vulnerable populations facing socio-economic disadvantages too.

The psychology of the pandemic and the inability to continue hospital visits has led to a situation where neither the patient nor the medical professional can judge the prognosis of many chronic illnesses like diabetes and cardiovascular disease. We in Ophthalmology face additional problems of continuing to provide care in visually debilitating diseases like glaucoma and cataract, especially in community models like camps. Ongoing care for conditions like diabetic retinopathy and glaucoma is of utmost importance to prevent an explosion of a population affected with severe visual impairment or blindness.

The burden of noncommunicable diseases and noncommunicable eye diseases has been highlighted in this issue by the President of the KSOS. Implementation of The Kerala Model "Vision Rehabilitation Centres" has also been marked as a priority by our Society.

Babu *et al*. have noted in their study that a significant number of patients could not get adequate treatment during the lockdown period. The number of patients undergoing intravitreal injections, laser treatment for peripheral tears as well as retinal vascular pathologies has dwindled. In the absence of timely intervention, such patients will show progression to advanced stages.[1] Similarly in the same issue, a survey done concluded that the majority of ophthalmologists in India were not seeing patients during the COVID-19 lockdown, with the near-total cessation of elective surgeries[2] and these so-called elective interventions are mostly critical services to preserve or restore vision. Cataract continues to be the major cause of blindness and responsible for 66.2% of blindness in India.[3]

The Directorate General of Health Services, Ministry of Health and Family Welfare, Government of India, mentions the target of reducing the prevalence of blindness to 0.3% by the year 2020.[4] Under the recently issued guidelines on safe ophthalmology practices in the COVID-19 scenario, the union health ministry has said the screening of patients for cataract and other eye diseases in outreach areas needs to be undertaken after duly following social distancing, hand hygiene, and personal protective measures.[5] Encouraging remote consultations by nongovernmental organization in vision centers, the union health ministry has said identified patients may be called to the base hospital by appointment for cataract surgery, so that backlog of such cases does not build up.

The novel coronavirus pandemic is not coming to an end anytime soon as fresh waves of infection are occurring globally. Many of these degenerative eye diseases will silently and relentlessly progress to blindness. Urgent changes need to be incorporated in Ophthalmic care at all levels, beginning with talking about these issues in all forums available, including Ophthalmic conferences, government-level meetings, and public discourses. Brainstorming by all concerned stakeholders to resolve these issues must be done at the earliest.

The risk of delaying eye care is too high.

Time is running out…….

V. Sudha
Department of Ophthalmology, Government Medical College, Thrissur, Kerala, India
E-mail: sudhajithdr@gmail.com

Address for correspondence: Dr. Sudha V, Department of Ophthalmology, Government Medical College, Thrissur, Kerala, India.
E-mail: sudhajithdr@gmail.com

REFERENCES

1. Babu N, Kohli P, Mishra C, Sen S, Arthur D, Chhablani D, *et al*. To evaluate the effect of COVID-19 pandemic and national lockdown on patient care at a tertiary-care ophthalmology institute. Indian J Ophthalmol 2020;68:1540-4.
2. Nair AG, Gandhi RA, Natarajan S. Effect of COVID-19 related lockdown on ophthalmic practice and patient care in India: Results of a survey. Indian J Ophthalmol 2020;68:725-30.
3. National Blindness and Visual Impairment Survey India 2015-2019- A Summary report Available from: https://npcbvi.gov.in/write

ReadData/mainlinkFile/File341.pdf. [Last accessed on 2020 Aug 27].
4. National Programme for Control of Blindness and Visual Impairment (NPCB&VI). Available from: https://dghs.gov.in/content/1354_3_National Programme for Control of Blindness Visual .aspx. [Last accessed on 2020 Aug 27].
5. Government of India Ministry of Health and Family Welfare. Guidelines on Safe Ophthalmology Practices in COVID-19 Scenario. Government of India Ministry of Health and Family Welfare.; August 19, 2020. Available from: https://www.mohfw.gov.in/pdf/GuidelineforEyeCare.pdf. [Last accessed on 2020 Aug 27].

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

Submitted: 28-Aug-2020 **Accepted:** 01-Sep-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_128_20	

How to cite this article: Sudha V. The silent epidemic – Are we neglecting it? Kerala J Ophthalmol 2020;32:219-20.

Postgraduate surgical training in ophthalmology



"Fill the cup of joy with the nectar of pleasure and enjoy life, before you too into the dust descend"

- DR. K. Mahadevan

Although it probably takes more than a decade of excruciating hard work and a highly precise skill-set to get there, the evolving prospects in Ophthalmic surgery are still the most exciting and rewarding area of the subject for most practicing Ophthalmologists. Whether it is the prospect of granting the gift of clear vision to the masses or the enviable high obtained by working on the cutting edge developments such as the latest Lasers or Corneal transplants, surgery is for many, the pinnacle of ophthalmology. It always attracts the best and brightest doctors in training, almost all of whom have a certain level of assertiveness and confidence as hallmarks. A surgeon has to be constantly up to date in this ever-changing field, regularly unlearning outdated methods and absorbing newer techniques that develop alongside every recent innovation.

THE LEARNING CURVE

From the beginning of PG training to the acquisition of the much coveted degree, it makes for around 3 years of theoretical study coupled with core surgical training. These surgically formative years are his years of self-discovery. Opportunity to learn from the very best hands, who rose within their surgical specialties entirely on the basis of their dexterity and finer skills, is often the main highlight of this training period. The interactions with such high caliber surgeons often rub off on a trainee kindling a newly discovered love for the surgery. The niche area of surgery one wants to further specialize in is something that an aspiring ophthalmic surgeon only zeroes in during his PG years.

GLEANING THE BASICS

Many top tier colleges in the country have, since many years now, resorted to teaching their postgraduate students the functional structure and mechanism of human eyes on the samples of goats' eyes. Goat eyes [Figures 1 and 2] that are economically sourced from slaughter houses have evolved as a relatively simple and inexpensive technique to train budding ophthalmology surgeons, preparing them to get over their initial awe in doing surgical procedures on a human eye. It provides the opportunities to hone their surgical dexterity, depth perception, handling of the operating microscope and phacomachine by hands-on training [Figure 3]. Postgraduates are encouraged to use the Head-Loupe for better clarity and more magnified view, even during minor op-based surgical procedures so that their eventual transition to the Operating Microscope is made much easier.

AGE OF SIMULATORS

Simulation is becoming hugely popular in every surgical field, with Ophthalmology being no exception. Technology-enhanced simulation training[1,2] has been shown to have positive knowledge, skills, and behavior-related outcomes in budding surgeons which reflect positively on the patient-related

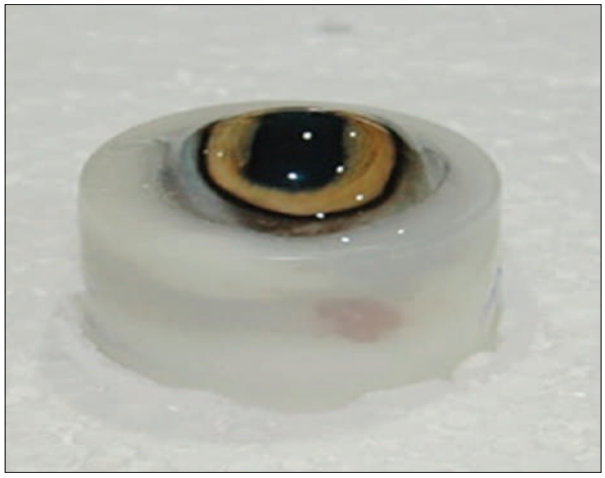


Figure 1: Mounted Goat Eye

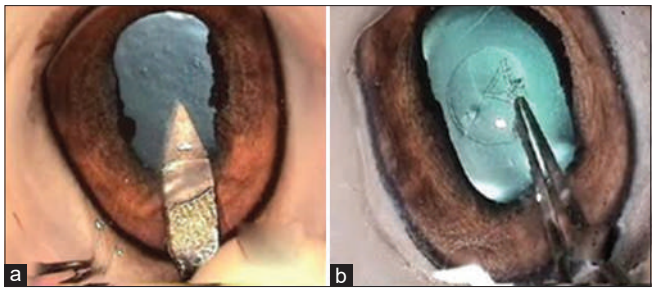


Figure 2: a) Clear Corneal Incision, b) Continuous Curvilinear Capsulorhexis



Figure 4: Traditional Teaching Methods



Figure 3: Hands On surgical training



Figure 5: Surgical Training Kit

prognosis. Although bed-side teaching using real patients is the more traditionally acceptable norm, especially in the medical specialties, this method is being increasingly replaced by simulator training. Without taking away from the true learning experience from real patients in the clinical setting [Figure 4], this change in the teaching paradigm has been largely made possible by the enviable technological innovations happening in this regard, factoring in the patients' comfort or time constraint in a busy OPD. The simulations [Figure 5] are good platforms for the surgical residents to practise and refine their surgical techniques without jeopardizing patient care and inviting unnecessary litigations. It is a dress rehearsal where mistakes can be made and lessons learned without compromising on the quality of care that can be ultimately delivered.

Various other forms of medical simulation such as wet laboratories [Figure 6], mannequins, cadavers, simulated patients, simulated hospital environments, simulated laser or surgical models, and virtual reality[3] are also being designed to enhance the postgraduate resident's learning experience.

THE JAPANESE INFLUENCE

A novel and revolutionary eye surgery training system has been developed by a major Japanese Enterprise for teaching, practicing and mastering phacoemulsification, and other ophthalmic surgeries. They aim to replace animal eyes in surgical learning and comprises of three types of realistic simulation training kits. This includes the "Dry Lab" for learning instrument manipulation, and "Wet Lab" for learning phacomachine operations, before attempting to perform the actual surgeries. The recently developed "Multi Lab" seeks to improve surgical skills for difficult and complicated cases in phacoemulsification in addition to various other ophthalmic surgical techniques.

THE PROACTIVE INITIATIVE

A leading Indian Pharmaceutical company in joint effort with the Vitreo-Retinal Developmental Program hosts and houses many major training initiatives in the Ophthalmic "Centers for Excellence" pan India. This undertaking came into the fore for the first time as early as 2014, in the premier institution of Susrut Eye Hospital and Research Center, West Bengal. The continuing program provides a common platform to trained eye



Figure 6: Wet Lab

surgeons from all over India and even neighboring countries such as Bangladesh and Nepal for finessing their surgical skills and learning newer emerging techniques in the treatment of ocular morbidities. The key agenda of this 2 week long Skill-enhancement initiative is to render formal training to VR surgeons intending to shift from 20-gauge vitrectomy to micro-incision vitrectomy surgery, especially in the surgical management of the Diabetic Retinopathy cases that are currently on the rise. Such companies that serve as the prime resource of the vitrectomy surgical system and consumables for the learning also plays a key role financing these training programs, as does other nongovernmental organizations that support the surgery cost. By 2016, the initial Susrut program expanded their scope to other select Institutions such as Aravind Eye Hospitals at Coimbatore, Madurai, and Pondicherry.

Similar training programs are also being undertaken in Specialist Training Institutes, targeted toward enhancement of cataract surgical capabilities among practicing surgeons from the various parts of the world. The Phaco Development Program is one such initiative aimed at building up expertise in phaco-based procedures at the community level.

THE AURA OF A SURGEON

The strength of all these educational training initiatives stems from the commitment and dedication shown by practicing eye surgeons to continue their learning curve, sharpening and further strengthening their surgical capabilities to ensure enhanced positive eye surgery outcomes for their patients.

If the prospect of such long years of study, unending training and re-training, with grueling hours of work and the onus of making split-second life-saving decisions fills you with excitement rather than the customary response of dread, you are definitely born to be a surgeon. The modern day surgeon is, therefore, without question an extreme machine, with life in it.

Acknowledgments

1. India's Vitreo Retinal Development Project : Providing Advanced Training to Retina Surgeons, Pravin U. Dugel, MD, American Society of Retina Specialists
2. "Kitaro Surgical Training System" – Junsuke Akura, Kiran Pokharel
3. Alcon Laboratories, Karnataka, India
4. Sree Gokulam Medical College, Venjaramoode, Thiruvananthapuram, Kerala.

Vivek Oommen Varghese, Thara R. Nair, K. Mahadevan

Department of Ophthalmology, Sree Gokulam Medical College, Thiruvananthapuram, Kerala, India

Address for correspondence: Dr. K Mahadevan, Department of Ophthalmology, Sree Gokulam Medical College, Thiruvananthapuram, Kerala, India.
E-mail: eyemahadevan@rediffmail.com

REFERENCES

1. Alwadani S. Cataract surgery training using surgical simulators and wet-labs: Course description and literature review. Saudi J Ophthalmol 2018;32:324-9.
2. Saleh GM, Lamparter J, Sullivan PM, O'sullivan F, Hussain B, Athanasiadis I, *et al*. The international forum of ophthalmic simulation: developing a virtual reality training curriculum for ophthalmology. Br J Ophthalmol 2013;97:789-92.
3. Solverson DJ, Mazzoli RA, Raymond WR, Nelson ML, Hansen EA, Torres MF, *et al*. Virtual reality simulation in acquiring and differentiating basic ophthalmic microsurgical skills. Simul Healthc 2009;4:98-103.

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

Submitted: 23-Aug-2020 **Accepted:** 24-Aug-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_124_20	

How to cite this article: Varghese VO, Nair TR, Mahadevan K. Postgraduate surgical training in ophthalmology. Kerala J Ophthalmol 2020;32:221-3.

Perspective

Noncommunicable diseases and noncommunicable eye diseases: What is the way forward?

ABSTRACT

As humans conquered indectious communicable diseases, nutritional deficiency as well as conditions responsible for maternal and perinatal mortality and lifespan increased, new problems such as diabetes, hypertension, cardiovascular, respiratory, and renal diseases, mental health, and drug abuse became significant health issues. Cloistered under a single umbrella term noncommunicable diseases (NCDs), these conditions as well as noncommunicable eye diseases are characterized by a course that is prolonged with multiple morbidities and an unequal distribution that puts the poor and the vulnerable at a disadvantage. Our country is going through the phase of communicable disease to noncommunicable disease (NCD) transition. And hence it is absolutely vital for us to plan and execute effective strategies to control these conditions.

Keywords: Noncommunicable disease, noncommunicable eye disease, vision rehabilitation centers

INTRODUCTION

For the most part of our history, the main threats to a healthy life were natural calamities, communicable diseases, malnutrition, injuries, and complications of childbirth. After a prolonged and successful battle against these entities, our health-care system was able to streamline a very effective strategy for early recognition, prevention as well as effective management of these disease entities. As humans conquered these conditions and lifespan increased, new problems such as diabetes, hypertension, cardiovascular, respiratory, and renal diseases, mental health, and drug abuse became significant health issues. Cloistered under a single umbrella term noncommunicable diseases (NCDs), these conditions are characterized by a course that is prolonged with multiple morbidities and an unequal distribution that puts the poor and the vulnerable at a disadvantage [Figure 1 and Table 1].

Submitted: 01-Aug-2020 **Accepted:** 01-Aug-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_114_20	

The threat to a healthy life from these diseases is greater due to the lack of a proper health-care system response or an effectively planned strategy to contain them.

WHY ARE NONCOMMUNICABLE DISEASES A SERIOUS PUBLIC HEALTH PROBLEM?

NCDs have become the most dominant global public health challenge of the 21st century. Sixty-three percent of the annual global death toll and untold morbidity and disability caused by NCDs makes this entity a real public health problem. In India, 62% of all deaths and 55% of all disability-adjusted life years (DALYs) in 2016 were attributable to NCDs. NCDs not only have a serious impact on human health but also exert a severely detrimental effect on the country's economic growth. NCDs and mental health conditions could cause the world $47

Meena Chakrabarti

Retina and Vitreous Services, Chakrabarti Eye Care Centre, Trivandrum, Kerala, India

Address for correspondence: Dr. Meena Chakrabarti, Retina and Vitreous Services, Chakrabarti Eye Care Centre, Indian Oil Petrol Pump, Kanjirampara P.O., Trivandrum, Kerala, India.
E-mail: drmeenachakrabarti@gmail.com

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Chakrabarti M. Noncommunicable diseases and noncommunicable eye diseases: What is the way forward? Kerala J Ophthalmol 2020;32:224-31.

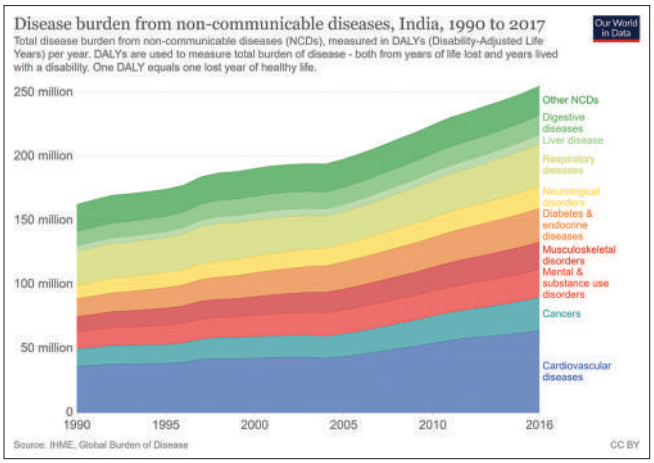


Figure 1: Disease burden from noncommunicable diseases, India 2016 (source: IHME, Global Burden of Disease)

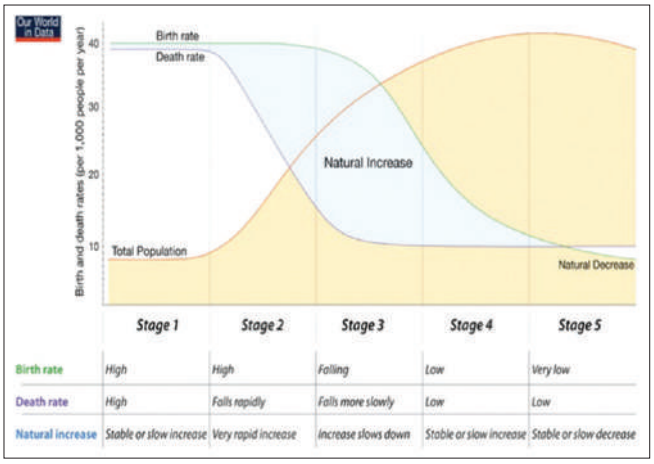


Figure 2: Epidemiological transition (source: https://www.ourworldindata.org)

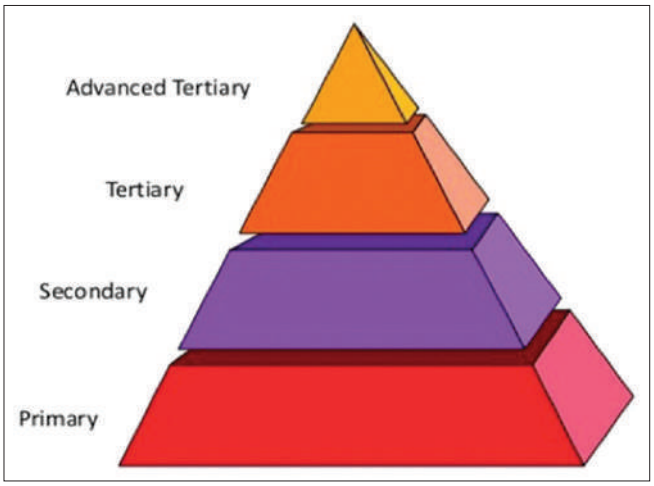


Figure 3: A successful strategy to tackle the enormous burden of NCED in our country Adapted from Das *et al.* IJO 2018;66 (11):1532-8

trillion in lost economic output from 2010 to 2030 if urgent actions are not enforced to prevent or treat these conditions.

NCDs account for:

- 62% of all deaths in India
- 40% of all hospital stays
- 35% of all recorded outpatient department (OPD) visits
- 55% of DALYs
- 26% probability of premature deaths
- Increased out-of-pocket expenditure (OOPE) for hospitalization for NCDs (from 32% [1995–1996] to 47% in 2004).

Thus, it is estimated that NCDs reduce the gross domestic product of India by at least 1%, and hence, the economic consequences of NCDs cannot be overstated. NCDs push a large number of persons with fragile and unstable financial status into poverty with its high potential for incurring OOPE for hospitalization to manage the several chronic disease-related sequelae.[1-5]

NCDs exert a deleterious effect on the country's economic growth in several other diverse ways also. They account,for increased health-care expenditure for the health-care system, the family as well as the individual, and is responsible for the affected persons opting for early retirement, thereby reducing the available labor and reducing productivity. Productivity is further reduced when the afflicted employee (due to his chronic ill health) is unable to perform his duties satisfactorily.

As the impact of NCD increases, fuelled by the lack of a proper health-care system response and an increased aging population, deaths due to NCDs are expected to spiral skywards with the maximum increase in the middle- and low-income groups.

COMMUNICABLE FACTS FOR THE COMMUNITY THE MYTHS SURROUNDING NONCOMMUNICABLE DISEASES

Myth No 1 – Noncommunicable diseases are primarily urban diseases of the rich and the elderly

The myth that NCDs are seen only in urban India was busted as early as 2003, when the NCD mortality in rural India was shown to be comparable to the mortality due to communicable diseases, maternal and neonatal conditions, and nutritional disorders. This showed that our country was going through the phase of communicable disease-to-noncommunicable disease (NCD) transition. The rate of transition is directly related to the level of social development, education, and degree of urbanization and was seen to occur faster in the more developed metropolitan cities with the poorer states also racing to catch up. This transition is evident if we examine the DALY due to NCDs which increased by 65% (1990–2016) in the late transition states compared to the 36% increase in the early transition

Table 1: Disease burden from noncommunicable diseases, India 2016 (Source: IHME, Global Burden of Disease)

Disease (noncommunicable)	Burden
Digestive disorders	15.23 million
Liver diseases	7.25 million
Respiratory diseases	33.13 million
Neurological disorders	17.33 million
Diabetes and endocrine disorders	26.16 million
Musculoskeletal disorders	21.35 million
Mental disorders and substance abuse	22.10 million
cancers	25.77 million
CVS disorders	64.22 million
Others	22.56 million
Total	255.09 million

CVS: Cyclic vomiting syndrome

states during the same period of time. It is also disheartening to note that the NCD burden is increasing at a greater rate among the poor in India in whom the onset of these diseases is at an earlier age. The majority of the deaths due to NCDs in India are premature and preventable, and additionally, the years of productive life lost due to NCDs are greater than those due to communicable diseases.

Myth No 2 – Increase in noncommunicable diseases represents economic growth

Until a few years back, NCDs, especially diabetes and hypertension, were considered as signs of prosperity and wealth. This scenario has changed dramatically with the NCD burden increasing at an alarming rate among the poorer and marginalized sections of the society. The triad of reduced productivity during the most active phase of life, increasing OOPE to treat chronic sequelae of NCDs as well as the added burden on an already stressed health-care system in a highly populated country like India has created an awareness in governmental and nongovernmental agencies on the tremendous detrimental effect of NCDs on economic growth.

Myth No 3 – Noncommunicable diseases are incurable, self-inflicted with difficult-to-change behavioral risk factors and the state has no role in its prevention

Most of the NCDs are incurable, but the multiple morbidities, premature deaths, and disabilities sue to NCDs are preventable using evidence-based interventions. There is good quality evidence which suggests that lifestyle interventions and an emphasis on a daily physical activity are useful in delaying the onset and slowing the progression of diabetes, hypertension, and cardiovascular diseases.

The role of the state administration: Initiating changes in the society to trigger these behavioral modifications and providing incentives to make healthy choices on the responsibilities of the state. Smart urban life planning, affordable and accessible healthy foods, quality health education, and limitations on the advertising of unhealthy food products are strategies to reduce NCDs.

Low-cost solutions ("best buys" – actions) that should be enforced to accelerate results in terms of lives saved, disease prevented, and heavy costs avoided are as follows:

- Banning smoking in public places, thereby protecting people from tobacco smoke
- Creating public awareness about the dangers of tobacco and alcohol use (using both the visual, digital, and print media)
- Enforcing bans on tobacco and alcoholic product advertising, promotion, and sponsorship
- Restricting access to retailed alcohol
- Promoting public awareness on healthy diet and physical activity
- Enforcing drink-driving laws
- Restriction on marketing food and beverages high in salts, fats, and sugar, especially to children
- Promoting school-, workplace-, and community-based physical activity programs
- Enforcing population-wide preventive interventions
    - Vaccination against hepatitis B, a major cause of liver cancer
    - Vaccination against human papillomavirus, the main cause of carcinoma cervix
    - Protection against environmental and occupational hazards such as aflatoxins, asbestos, silica dust, and contaminants in potable water
    - Screening for breast, oral, colon, and cervical cancers.

The health-care system should combine population-based and individual interventions for primary prevention, early detection, treatment, and palliative care.

Currently, the health care for NCDs has its main focus on hospital-centered acute care which is a very expensive way of tackling NCDs and will not contribute to the reduction of NCD burden. To ensure primary prevention and early detection of NCDs, the integration of these diseases into the primary health-care package is mandatory. Cost-effective and sustainable health-care interventions should be offered to high-risk individuals such as:

- Interventions for the prevention and management of diabetes (which have definitely reduced costs while improving health) , include glycemic control, blood pressure control, foot care, diabetic retinopathy (DR) screening, screening for nephropathy
- Inclusion of chronic respiratory diseases also in lung

Table 2: An easily replicable model adopted by one of the major tertiary eye care centers in South India (Adapted from Das *et al.* IJO 2018;66 (11):1532-8)

STRUCTURE	POPULATION SERVICED	TECHNICAL PERSONNEL	LEVEL OF CARE	QUANTUM OF CARE
PRIMARY	50,000	VISION TECHNICIAN	SPECTACLE DISPENSING REFER	40% OF VISUALIMPAIRMENT (URE)
SECONDARY	500,000	OPHTHALMOLOGISTS VISION TECHNICIAN SURGICAL ASSISTANT	COMMON EYE EXAMINATION COMMUNITY CARE SURGERY OF COMMON EYE DISORDERS	70% OF VISUAL IMPAIRMENT (URE + CATARACT)
TERTIARY	5 MILLION	OPHTHALMOLOGIST OPTOMETRISTS NURSES REHABILITATION MICROBIOLOGY PATHOLOGY EYE BANKING	SECONDARY LEVEL CARE + ALL EYE SURGERIES CORNEAL TRANSPLANT LOW VISION AND REHAB TRAINING & CLINICAL RESEARCH	90% OF VISUAL IMPAIRMENT (URE + CATARACT + SURGERY FOR GLAUCOMA & DR)
ADVANCED TERTIARY	50 MILLION	TERTIARY LEVEL+ BASIC SCIENTISTS POLICY MAKERS	TERTIARY LEVEL CARE + TRANSLATIONAL RESEARCH POLICY MAKING & PLANNING	100% OF VISUAL IMPAIRMENT POLICY EXECUTION

heath programs designed for prevention, treatment, and control of tuberculosis. This program should explore the procurement of quality-assured inhaled drugs at affordable cost

- High-risk individuals and those with established cardiovascular disease can be treated with low-cost generic medicines (regimen of aspirin, statins, and blood pressure-lowering agents) that significantly reduce the risk of death or a major cardiac event. When coupled with cessation of smoking the therapeutic benefit can be doubled
- Creating access to pain-relieving agents (morphine) and staff trained in palliative care, by forming mobile palliative units for home visits and necessary care of the terminally ill patients.

Financing and strengthening the health-care system to deliver cost-effective individual interventions through a primary health-care approach is a pragmatic first step to achieving the long-term vision of universal care coverage for NCDs. Strengthening political commitments and according a high priority to NCD programs are also key steps in expanding the health-care system capacity to tackle NCDs. For this to occur, we need to improve our basic technologies, the health workforce, health information, funding, etc., to ensure that the primary care package delivers the abovementioned cost-effective interventions to the public.

NONCOMMUNICABLE EYE DISEASES

Global efforts to address NCDs have largely excluded reference to the massive social and economic impacts of blinding conditions such as cataract, glaucoma, diabetes, and age-related macular degeneration (AMD). These efforts focused mainly on how mortality rates could be reduced, and hence, diseases such as cancer, respiratory illnesses, and those caused by tobacco and poor diet received all the attention in discussions on tackling the NCDs.

The World Health Organization's (WHO) most recent figures show that 285 million people globally are vision impaired, of whom nearly 40 million are blind. Around 50% of all blindness and vision impairment in developing countries is caused by cataract, and a further 10% of blindness is caused by glaucoma. Four percent of blindness and vision impairment is caused by diabetes, one of the primary diseases being addressed within the NCD framework.

There is growing evidence to support a relationship between blindness and mortality. Studies have shown that up to 60% of children who become blind die within 2 years. The DALY measure of time lived with a disability combined with time lost due to premature mortality for vision impairment (mostly caused by cataract, glaucoma, and refractive error) is the 6th largest cause of DALYs globally. Vision impairment has a 3% share of global DALYs – more than unhealthy diet (1%–2%) and physical inactivity (2.1%), and only fractionally less than those resulting from cancer (5.1%), respiratory disease (3.9%), and harmful use of alcohol (4.5%) and tobacco use (3.7%)

Although cataract and uncorrected refractive errors remain by far the leading causes of visual impairment, several other eye conditions have emerged as significant threats to people's vision – those which are noncommunicable (or chronic) and which become more prevalent with aging. They are:

- DR
- glaucoma
- AMD.

In addition, the ocular adverse effects of abuse of tobacco, alcohol, and illicit drugs have also emerged as a social threat and health hazard. Drug and alcohol abuse can produce a variety of ocular and neuro-ophthalmic manifestations. Novel substances of abuse and novel routes of administration may surprise even the most astute physician if substance abuse is not suspected.

Unlike cataract or refractive error, for which surgery or a pair of spectacles can restore vision, these diseases all affect the back of the eye, are incurable, and require ongoing lifetime follow-up and management

Over the last two decades, these noncommunicable eye diseases (NCEDs) have become much more significant due to two main reasons:

1. The demographic transition (the fact that more people are living longer) is taking place due to better nutrition and accessibility to a more comprehensive health-care service. Since DR, glaucoma, and AMD mainly affect the elderly, demographic transition has resulted in an increase in the incidence of these conditions
2. The epidemiological transition means a change in population health due to changes in lifestyle. NCDs, particularly cardiovascular disease and diabetes (with its complications, including DR), are increasing because of changes in lifestyle with urbanization. People are less active, eat more, and eat unhealthy junk food. Smoking is prevalent in many populations and is a known risk factor for AMD Figure 2.

Both demographic and epidemiological transition has already occurred in high-income and many of the middle-income countries and is starting to become noticeable in low-income countries also.

Strategic planning and availability of services for the prevention, early detection, and management of DR, glaucoma, and AMD has been enforced in many high-income countries by providing comprehensive eye care services that are integrated into their national health systems.

In addition, to enhance the quality of life of people with permanent, severe vision loss, the provision of low vision services as well as vision and occupational rehabilitation services have been made available. Most low-income countries, and some middle-income countries, however, are not able to provide adequate services to prevent and manage these diseases as there is a shortage of skilled eye care professionals with the competence to effectively handle these eye conditions. The eye health services in these countries also lack an interdisciplinary, patient-centered approach, which is critical for the successful management of NCEDs.

What are the priorities for action?

Major contributions to eye care development in India have come from the ophthalmologists (mass cataract surgery in the early 1900s; major participation of nongovernment organizations), policymakers (National Program for Control of Blindness and Visual Impairment, 1976; systematic development under the World Bank-assisted India Cataract Project, 1995–2002), and the industry (manufacturing of affordable surgical instruments and medicines). Although the country can boast of higher cataract surgical coverage and near-total elimination of trachoma, there is an increasing prevalence of DR and undetected glaucoma.

India is presently at the crossroads of adherence to the old successful model of service delivery and adoption of new innovative methods of teaching and training (workforce development and skill-based training) relevant medical research and product development. In the absence of these new approaches, the initial gains in eye care cannot be furthered in India. A new approach combining the best of "old" tradition and the "new" technology is required to further the future of eye care in our country.[6-8] Thus, the future of eye care in our country is dependent on:

1. Availability of committed, trained eye care professionals who have the equipment they need and are working within an adequate infrastructure and are trained to provide comprehensive eye examinations. They should be confident in managing NCEDs, including the competency, to perform patient counseling.

There is a huge disparity in the quality of training available to the ophthalmic assistants and optometrists in our country. The duration of training varies from 6 months to 4 years, with a similar variation in the quality of training. These variations in training are reflected in the quality of eye care services available in our country. On the one hand, we have well-equipped and staffed tertiary eye care referral centers while on the other end of the spectrum are centers lacking appropriately trained professionals and basic infrastructure. Rural–urban disparities of eye care services make easy availability of eye care facilities difficult for the rural population. More than 70% of the Indian population reside in rural areas where only 25% of ophthalmologists practice. Eye surgeon–population ratio varies from 1:20,000 in urban areas to 1 in 2, 50,000 in rural areas.

2. Necessary equipment and infrastructure (including good record-keeping processes) need to be put in place so that,

once trained, eye care providers can examine, manage, and follow up patients. Nonavailability of proper equipment is one of the main obstacles hampering appropriate eye care delivery. In addition, there are centers where equipment are available, but lack of maintenance results in suboptimal utilization of the equipment. Inadequate ophthalmologic equipment management training also acts as a barrier in utilization of equipment. High maintenance costs and location of the eye care facility in remote areas again pose challenges for maintenance of equipment.

3. Adequate and realistic patient counseling: As these conditions require often lifelong follow-up, they will have a major long-term impact on patients, both in terms of their time and compliance with the treatment regimen and in terms of the lifelong costs of managing their condition. An awareness drive should be started, and at all contact points (OPD, camps, schools, offices, etc.), trained counselors should explain about the importance of early detection, periodic checkup, compliance to therapy as well as long-term follow-up. In a study of the predictors of and barriers associated with poor follow-up in glaucoma patients, lack of knowledge about the blinding sequelae and perceptional barriers were responsible in 40% of patients with poor compliance to follow-up. If this is the level of awareness regarding the disease and the need for follow-up in patients with established disease, the awareness levels in those without established disease will definitely be worse.
4. Establishing health insurance-based financing of health care, which includes eye care, appears to be the optimal way to prevent vision loss in those individuals who may not be able to afford out-of-pocket payments. There are about 16 government health insurance schemes in India such as the Ayushman Bharat, Rashtriya Swasthya Bima Yojana, or the Kerala government-sponsored Karunya Health Scheme. A Government Health Insurance Scheme is a state or central government-powered health insurance initiative for its citizens, directed toward enhancing the health-care quotient of the region by offering low-priced insurance policies with a sizeable sum insured. Such policies are usually offered on an annual basis at a low price to encourage people below the poverty line to avail insurance.

Patients must be able to comply with the treatment regimens, which means that the medicines they need have to be available and affordable. This means that the procurement and distribution logistics of eye medicines using bulk purchasing to drive prices down and to ensure quality control and standardization have to be strictly enforced.

5. Availability of low vision services which are affordable. Even in high-income countries, there may be uneven coverage of the population, and most low vision services are provided in urban areas, resulting in limited access. Adequate low vision and rehabilitation services should, therefore, be an integral part of comprehensive eye care. Once people have lost their sight due to an NCED, there is usually no way to restore vision, so rehabilitation and low vision are currently the only remaining intervention.

The Kerala Model "VISION REHABILITATION CENTERS:" To ensure provision for adequate accessibility to low visual aid facilities, the Kerala Society of Ophthalmic Surgeons (KSOS) have started a pilot project to train optometrists and facilitate setting up of vision rehabilitation centers within our state.

The participating institutes sign a MoU with KSOS to run low vision centers with commitment for a minimum period of 3 years without looking at the financial viability of the project. The institutions provide (a) space – minimum of 100–150 sq ft, (b) service of trained personal in low vision, and (c) consultant in medical retina (full time/part-time) and should have easy access to optical coherence tomography and Humphrey Field Analyser (HFA). The management personnel of the selected institutions/hospitals are trained regarding the functioning of low vision centers. The trained optometrists serve the designated center for a period of 12 months after completion of training. They will, in turn, train other optometrists deputed by KSOS MoU-signed institutions in low vision service. The service provider should also take the initiative to conduct outreach programs for case detection. This program may be considered as our first step toward ensuring provision for adequate accessibility to low visual aid facilities in our state.

A "bottom-up" pyramid represents a model of holistic eye care for creation of sustainable permanent facilities within communities, staffed and managed by locally trained human resources, and linked effectively with successively higher levels of care. This is the model adopted by one of the major tertiary eye care centers in South Indian and is an easily replicable model Table 2 and Figure 3.

- At the base of the pyramid are VISION GUARDIANS that represent community involvement and comprise trained young people who keep a close vigil on the eye health of about 5,000 persons within communities through door-to-door surveys and other informal means
- VISION CENTERS form the next level and serve the primary eye health needs of the community and are staffed by persons from the local community, catering to a cluster of villages, individually servicing a target of around 50,000 people
- SECONDARY EYE CARE CENTERS are networked to the

vision centers, and each serves a population of 500,000 persons. These centers provide care that can diagnose the complete range of ophthalmological diseases and offer high-quality surgical care for cataract – the most common cause of blindness. These centers draw upon local talent too, with team members recruited from the local community and trained at an advanced tertiary centers

- TERTIARY CARE HOSPITALS/TRAINING CENTERS are linked to secondary centers, and each serves a population of 5 million persons. These centers provide a comprehensive range of services and also serve as training centers to the secondary centers.
- CENTERS OF EXCELLENCE are linked to tertiary centers, and each serves a population of 50 million persons. These centers treat complex diseases, train the trainers in subspecialties and rehabilitation, and engage in advocacy.

This innovative approach ensures eye care delivery that

- Combines excellence with equity: Quality eye care for all
- Sustainability: Each center becomes self-supporting through income-generating activities and cross-subsidization of services
- Team approach: Complementary roles for each team member for an efficient and cost-effective system
- Community participation: Strong community ownership at the primary and secondary levels, with over 50% of staff recruited locally and contributions from the community in cash and kind.

This replicable model is appropriate for developing countries, as it does not depend on external funding or expertise in the long term.

While India is waking up to the need for eye care with the government making policy initiatives at both the state and center levels, a lot still needs to be done. The entry of several service providers and active participation from NGOs through various initiatives has also slightly assisted in raising the awareness of eye care in India. One such initiative is VISION 2020 that was launched by the WHO together with more than 20 international nongovernmental organizations that aim to provide technical support and advocacy to the prevention of blindness activities worldwide by the year 2020.

Another initiative that has seen some success is the Global Action Plan (GAP) which is a commitment endorsed by all WHO Member States to improve eye health for everyone ("Universal Eye Health") by 2018. GAP is now the most important strategic document in eye health. This initiative represents a significant step forward toward achieving "universal access" to eye health. Approximately 25 VISION 2020 workshops take place every year at local and international levels, tackling specific national or regional priorities for the GAP. These workshops help to align GAP's objectives in tandem with the specific region.

CONCLUSION

- NCDs kill 41 million people each year, equivalent to 71% of all deaths globally
- Each year, 15 million people die from a NCD between the ages of 30 and 69 years; over 85% of these "premature" deaths occur in low- and middle-income countries
- Cardiovascular diseases account for most NCD deaths, or 17.9 million people annually, followed by cancers (9.0 million), respiratory diseases (3.9 million), and diabetes (1.6 million)
- These four groups of diseases account for over 80% of all premature NCD deaths
- Tobacco use, physical inactivity, the harmful use of alcohol, and unhealthy diets all increase the risk of dying from a NCD
- Detection, screening, and treatment of NCDs, as well as palliative care, are key components of the response to NCDs.

The "causes of the causes" of NCDs make them difficult to address; proximal causes include raised cholesterol, blood pressure, and glucose; intermediate causes include tobacco, poor diet, physical inactivity, and harmful use of alcohol. These risks are largely human-made and relate to how we live, age, work, and play. Distal causes include urbanization, population aging, and trade. Premature death and disability due to NCDs can, therefore, be viewed as failures of a broader socioeconomic system.

Thus, for providing a permanent solution to prevention and effective management of avoidable morbidity due to NCDs and NCEDs, the following factors should be integrated into the existing health-care facilities.

1. Effective strategies for disease prevention and control
2. Training of required personnel
3. Infrastructure development
4. Strengthening existing health care
5. Development of affordable technology
6. Advocacy and resource mobilization
7. Revamping of medical education for improved knowledge and skills.

A large part of the population whom we are targeting are illiterate and underprivileged, largely in rural and also in urban areas. These subsets of our population are lacking in awareness of health issues and have not yet adopted a health-seeking behavior. There is a strong need for an advocacy to create awareness and bring about behavioral change using multiple mediums ranging from folk theater to community radio, panchayat, and peer group influence. All medium of communication should be utilized to achieve this.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Available from: http://www.globalburden.org/.
2. Available from: http://www.vision2020.org/documents/IAPB%20Documents/Briefing%20papers%20etc/Health%20 Systems%20and%20Eye%20Health-BP.pdf.
3. World Health Organization. The World Health Report 2003 p160-165; WHO, Global Status Report on NCDs. p. 56, 89.
4. Serge Resnikoff, *et al*. Non – Communicable Eye Disease: Facing the Future. Comm Eye Health 2014;27:87. Published online 10 December, 2014.
5. Universal Eye Health: A Global Action Plan 2014–2019. Available from: http://www.who.int/blindness/actionplan/en/.
6. Prevalence and Attributable Health Burden of chronic Respiratory Diseases, 1990–2017: A Systematic Analysis for the Global Burden of Disease Study 2017. *GBD Chronic Respiratory Disease Collaborators* The Lancet Respiratory Medicine *2020;8:6.*
7. Das T, Panda L. Imagining eye care in India (2018 Lalit Prakash Agarwal lecture). 2018;66 (11):1532-8.
8. Vijaya L, George R. Prevalence of angle-closure disease in a rural South Indian population. Arch Ophthalmol 2006;124:403-9.

Pigment sign in vernal keratoconjunctivitis

CLINICAL SIGN-PIGMENT SIGN IN VERNAL KERATOCONJUNCTIVITIS

Multiple fine, granular, discrete, dot-like brown pigments are seen in the perilimbal bulbar conjunctiva of children and young adults diagnosed with vernal keratoconjunctivitis (VKC) [Figures 1 and 2].[1]

SEEN IN

Vernal Keratoconjunctivitis.

DESCRIBED BY

Rao and Padmanabhan in 2002.[1]

CARDINAL FEATURES

Multiple faint to dark brown, discrete, fine pigments are seen scattered in the perilimbal bulbar conjunctiva of at least one eye.[1]

The most common site of pigmentation is the interpalpebral conjunctiva followed by the inferior and superior bulbar conjunctiva.[2]

Pigmentation is noted to occur in all grades of VKC. However, it is of greater clinical importance in the diagnosis of mild or quiescent forms of the disease when palpebral and limbal changes are not very characteristic.[2]

There is no relationship of density of pigmentation with age, severity, or chronicity of the disease. The pigmentation persists even when the disease is in remission.[3]

WHY IT OCCURS?

It is hypothesized to be the result of complex cellular immunity interactions occurring on the ocular surface, with the limbus serving as the seat of antigen–immune system interaction. The pigmentation may be representative of the afferent activity occurring, due to the transportation of foreign antigens to regional lymph nodes, by the Langerhans cells in a milieu of mast cells and melanocytes.[2]

WHAT ELSE TO EXAMINE?

Other signs/associations of vernal keratoconjunctivitis

i. Tarsal papillae, cobblestone papillae, limbal nodules, Horner–Trantas dots, pseudogerontoxon, superficial punctate keratitis, corneal erosion, epithelial defect, plaque formation, shield ulcer
ii. Examine for corneal ectasia, cataract, and glaucoma.[4]

Differential diagnosis and signs to exclude them

i. Vitamin A deficiency may show perilimbal pigmentation, which can be differentiated from VKC by the presence of Bitot's spots and the absence of other limbal and palpebral changes[3]

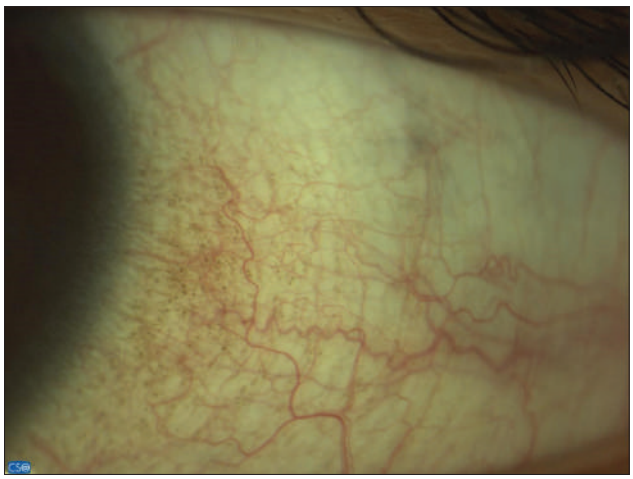


Figure 1: Pigment sign seen on the temporal conjunctiva of the left eye

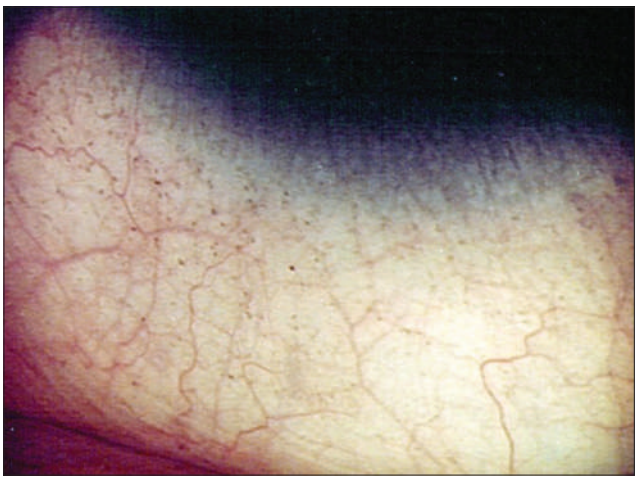


Figure 2: Pigment sign seen on the inferior bulbar conjunctiva of the left eye

© 2020 Kerala Journal of Ophthalmology | Published by Wolters Kluwer - Medknow

ii. In chemical injury, pigmentation of the bulbar conjunctiva may be seen. Apart from the history of trauma and associated signs, this pigmentation tends to have a blotchy appearance and lacks the fine granularity seen in VKC.[3]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Amanda Mohanan-Earatt, Sanjeev Srinivas

Cornea Services, Darshan Eye Care, Chennai, Tamil Nadu, India

Address for correspondence: Dr. Amanda Mohanan-Earatt, Darshan Eye Care, Anna Nagar, Chennai - 600 040, Tamil Nadu, India.
E-mail: dramandamohanan@gmail.com

REFERENCES

1. Rao SK, Padmanabhan P. Perilimbal conjunctival pigmentation in vernal keratoconjunctivitis: A new sign. Cornea 2002;21:432.
2. Rao SK, Meenakshi S, Srinivasan B, Baluswamy S. Perilimbal bulbar conjunctival pigmentation in vernal conjunctivitis: Prospective evaluation of a new clinical sign in an Indian population. Cornea 2004;23:356-9.
3. Luk FO, Wong VW, Rao SK, Lam DS. Perilimbal conjunctival pigmentation in Chinese patients with vernal keratoconjunctivitis. Eye (Lond) 2008;22:1011-4.
4. Kumar S. Vernal keratoconjunctivitis: A major review. Acta Ophthalmologica 2009;87:133-47.

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

Submitted: 03-Jun-2020 **Accepted:** 06-Jun-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_77_20	

How to cite this article: Mohanan-Earatt A, Srinivas S. Pigment sign in vernal keratoconjunctivitis. Kerala J Ophthalmol 2020;32:232-3.

Major Review

Emerging retinal diseases and newer terminologies in spectral domain optical coherence tomography

ABSTRACT

The retina practice has changed in such a way that diagnosis is incomplete without an optical coherence tomography (OCT) image. This article gives a brief update on the emerging spectrum of retinal diseases and newer terminologies in the spectral domain OCT to keep pace with the rapid advancements in the field of imaging.

Keywords: Emerging retinal diseases, newer terminologies, spectral-domain optical coherence tomography

INTRODUCTION

Introduced by researchers at Massachusetts Institute of Technology in the early 1990s,[1] optical coherence tomography (OCT) has allowed ophthalmologists to see the eye from a different perspective. With the latest improvements in imaging, spectral-domain OCT (SD-OCT) allows unrivaled details of the retina and choroid. SD-OCT acts as an optical biopsy,[2] and is used in the diagnosis and follow-up of various retinal and choroidal diseases involving the macular region.

In 2014, at International Nomenclature OCT (INOCT) Panel, Staurenghi *et al*.[3] proposed a nomenclature system for normal anatomic landmarks in SD-OCT and the panel suggested a consensus regarding the most proper terminology [Figure 1].

INTERNATIONAL NOMENCLATURE OPTICAL COHERENCE TOMOGRAPHY DEFINITIONS OF THE NORMAL ANATOMIC LANDMARKS IN SPECTRAL DOMAIN-OPTICAL COHERENCE TOMOGRAPHY

Moving from within the vitreous toward the outer layers of the globe, SD-OCT detects neurosensory retina resting between two layers of extreme hyperreflectivity. The first layer is at the vitreoretinal interface and the retinal nerve fiber layer. The second lay corresponds to the retinal pigment epithelium (RPE) – Bruch's membrane complex. The INOCT agreed on the adequate nomenclature for the outer retinal bands depending on the reflectivity of the layer.

The external limiting membrane band (ELM) - is located at the boundary between the cell bodies (nuclei) and the inner segments (IS) of the photoreceptors and comprises clusters of junctional complexes between the Müller cells and the photoreceptors.

Myoid zone (MZ) - the hyporeflective region between ELM and ellipsoid zone (EZ) corresponds to the innermost segment of photoreceptors. The reduced reflectivity of this zone is due to the lower packing density of mitochondria.

Divya Alex, Anantharaman Giridhar, Mahesh Gopalakrishnan, Shivam Madan, Swati Indurkhya, Swati Haridas, Sailesh Kumar, Dinesh Rungta

Department of Vitreoretinal Services, Giridhar Eye Institute, Kochi, Kerala, India

Address for correspondence: Dr. Divya Alex, Department of Vitreoretinal Services, Giridhar Eye Institute, Ponneth Temple Road, Kadavanthra, Kochi - 682 020, Kerala, India.
E-mail: alex.divya@gmail.com

Submitted: 22-Jul-2020 **Accepted:** 27-Jul-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_105_20	

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Alex D, Giridhar A, Gopalakrishnan M, Madan S, Indurkhya S, Haridas S, *et al*. Emerging retinal diseases and newer terminologies in spectral domain optical coherence tomography. Kerala J Ophthalmol 2020;32:234-43.

© 2020 Kerala Journal of Ophthalmology | Published by Wolters Kluwer - Medknow

EZ - the hyperreflective region adjacent to the MZ is the interface between inner and outer photoreceptors (previously referred as IS-outer segments [OS] junction). They are packed with mitochondria and have the potential for high reflectivity.

The interdigitation zone (IZ) - corresponds to the contact cylinder represented by the apices of the RPE cells that encase the cone OS. This layer was previously referred to as cone outer segment tips/Verhoeff's membrane,[4] or rod outer segment tips, and it is not always distinguishable from the underlying RPE layer.

Photoreceptor disruption can be visualized on OCT as the loss of integrity or absence of the outer retinal layers: The ELM, EZ, and IZ. Disruptions of these layers on OCT have been shown to correlate with worse visual acuity and retinal sensitivity in many retinal diseases.[5]

ENHANCED DEPTH IMAGING

The near-infrared 800 nm light source that is used in conventional OCT systems is scattered by the photoreceptor layer, and as a result, the signal reflected from the choroid is weak. Superior quality choroidal imaging can be obtained with a slight modification in conventional OCT systems with a longer wavelength of light, giving a low signal-to-noise ratio (Swept-source) and an improved high-quality eye-tracking ability, better image averaging capability as used in enhanced depth imaging (EDI). Morphological and vascular analyses of the choroid have been revolutionized with the advent of EDI. Studies have found the mean subfoveal choroidal thickness in healthy eyes to be 287 ± 76 mm using EDI.[6] The variants are better known as leptochoroid and pachychoroid [Figure 2].

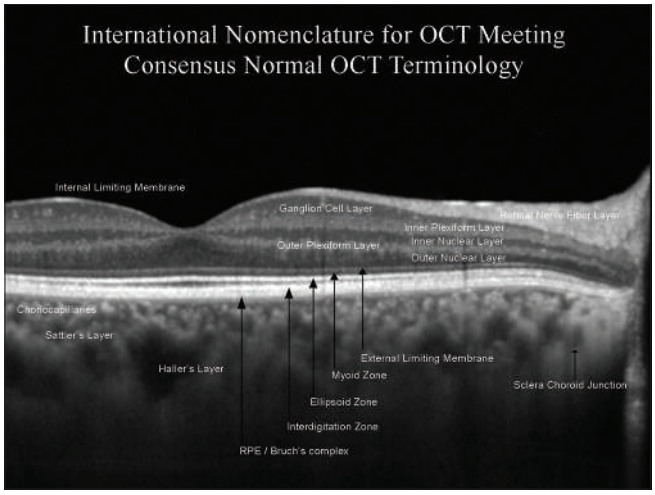


Figure 1: Nomenclature for normal anatomic landmarks seen on spectral domain-optical coherence tomography images proposed and adopted by the international nomenclature for optical coherence tomography panel

Leptochoroid

Leptochoroid is defined as extreme choroidal thinning in SD OCT.[7] In Greek, Lepto means thin or fine. It is a new clinical entity characterized by geographical hyperpigmented fundus centered in the macula, with surrounding relative hypopigmented fundus and associated with decreased choroidal thickness. Leptochoroid is not specific to high myopia.[8] Spaide described an entity of age-related choroidal atrophy (ARCA) affecting older individuals with a myopic error of 6 diopters, mean age being 80.6 years, mean choroidal thickness being 69.8 mm, and mean visual acuity being 20/40.[9] Eyes with ARCA may demonstrate extreme leptochoroid; however, they have a discrete appearance from eyes with high myopia. ARCA is characterized by a tessellated fundus appearance and rarefaction of the choroidal vessels under the macula, whereas highly myopic eyes with extreme leptochoroid have prominent choroidal vasculature instead of rarefaction of the choroidal vessels. In addition, eyes with ARCA may have reticular pseudodrusen. These two entities suggest that choroidal thickness alone is not a reliable indicator of visual function.

Pachychoroid

The term pachy originates from the Greek, which means thick. Pachychoroid is a relatively novel concept describing

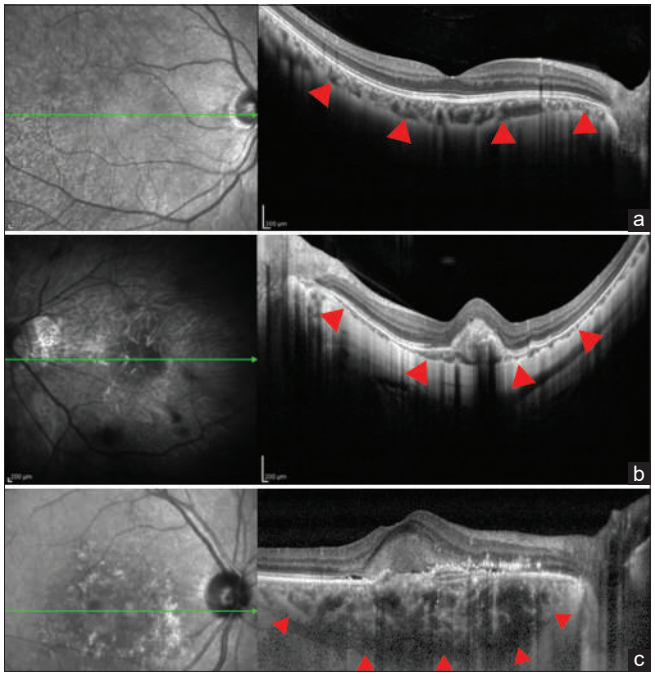


Figure 2: Enhanced depth imaging showing better delineation of the sclero-choroidal boundary (red triangles) (a) Spectral domain-optical coherence tomography of a healthy retina revealing a normal choroidal thickness. (b) Spectral domain-optical coherence tomography of a pathological myopic eye with choroidal neovascular membranes revealing extremely thin choroid (Leptochoroid). Note the reduced choroidal thickness further away from the macula (c) spectral domain-optical coherence tomography of an eye with polypoidal choroidal vasculopathy showing a diffusely thick choroid (Pachychoroid) with compression of choriocapillaris and dilated vessels at Haller's layer

a phenotype characterized by focal or diffuse choroidal thickening, attenuation of the choriocapillaris overlying dilated choroidal vessels in the Hallers layer (pachyvessels) and associated with progressive RPE dysfunction and neovascularization.[10] Several clinical manifestations have been described to reside within the pachychoroid disease spectrum, including central serous chorioretinopathy (CSCR), pachychoroid pigment epitheliopathy (PPE), pachychoroid neovasculopathy, polypoidal choroidal vasculopathy/ aneurysmal type 1 neovascularization, focal choroidal excavation (FCE) and peripapillary pachychoroid syndrome (PPS). Newer entities are discussed.

PACHYCHOROID PIGMENT EPITHELIOPATHY

The term PPE was first introduced by Warrow *et al*.[11] The clinical appearance of the pigment epitheliopathy included mottling of the RPE, irregular areas of RPE elevation termed "drusenoid RPE lesions" with the absence of soft drusen [Figure 3]. Since none of the eyes had manifested neurosensory detachment, PPE was considered a forme fruste of CSCR. Indocyanine green (ICG) shows increased choroidal hyperpermeability as mid-phase hyperfluorescence that co-existent with the areas of RPE disturbances. These patients were often misdiagnosed with pigmentary age-related macular degeneration (AMD), pattern dystrophy, or retinal pigment epithelitis. However, PPE is usually asymptomatic and usually found in the fellow eyes of CSCR. RPE elevation with microbreak appearance can be a prerunner to subretinal fluid (SRF) formation later on. It was subsequently observed that patients with PPE could go on to develop type one neovascularization, with or without aneurysmal (polypoidal) lesions, without necessarily developing CSCR.[12]

PACHYCHOROID NEOVASCULOPATHY (PCN/PNV)

Pang and Freund described the evolution of type one neovascularization in eyes over backdrop changes consistent with PPE, and coined the term PNV.[13] Characteristic features of PNV include the presence of type 1 neovascularization, which appears on OCT as a shallow irregular separation of the RPE from Bruch's membrane, which appears as "double layer sign (DLS)" overlying pachyvessels.[14] The presence of hyporeflectivity in DLS is a crucial marker of chronic CSCR, whereas hyperreflectivity in DLS distinctly correlated with PCN and polypoidal choroidal vasculopathy, indicative of underlying neovascular tissue complex.[15] The presence of neovascularization can be confirmed by detection of leakage on FA, typically in the form of late leakage with undetermined origin, and a corresponding late staining "plaque" on ICGA. With the advent of OCT angiography (OCTA), the diagnosis of PCN is more straightforward. OCTA reveals a tangled filamentous branching vascular network (BVN) with a flow signal corresponding to the DLS. The sensitivity of OCTA in detecting a BVN is more than that of ICG [Figure 3].

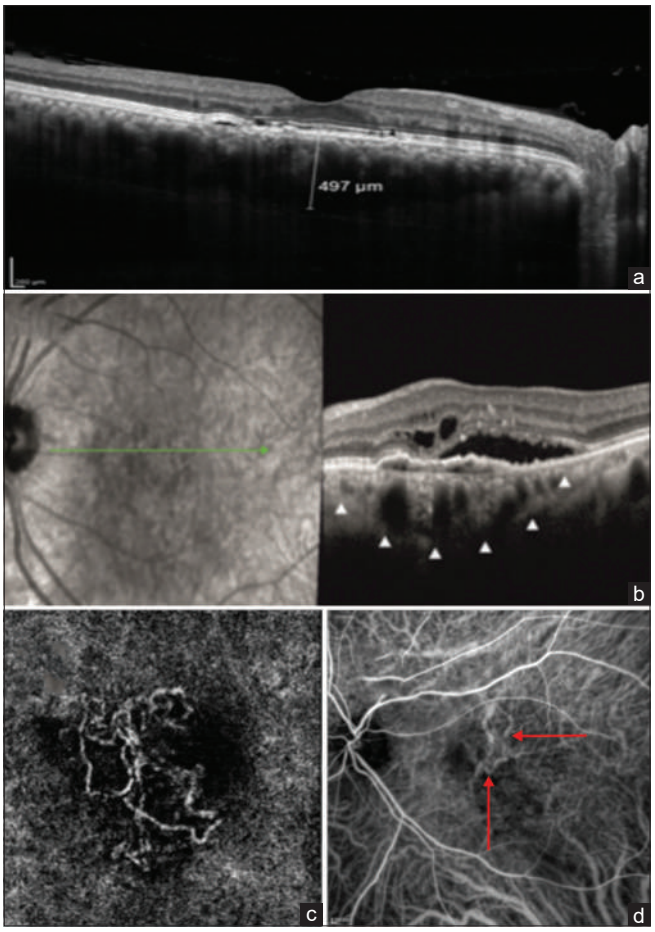


Figure 3: a) Foveal section spectral domain-optical coherence tomography of an eye with pachychoroid pigment epitheliopathy showing irregular retinal pigment epithelium with patchy loss of ellipsoid, pachyvessels, and thick choroid. (b) Spectral domain-optical coherence tomography of an eye with pachychoroid neovasculopathy (PCN) showing a hyperreflective double layer sign, subretinal and intraretinal fluid, pachyvessels, and pachychoroid. (c) Optical coherence tomography-angiography detecting an entangled branching vascular network with peripheral anastomosis (d) Indocyanine green showing the branching vascular network. Both corresponding to the level of PCN

PERIPAPILLARY PACHYCHOROID SYNDROME

PPS was described by Phasukkijwatana *et al*. as a distinct variant of the pachychoroid disease spectrum, in which maximal choroidal thickness occurs close to the optic nerve rather than subfoveally.[16] These patients typically present with nasal macular intraretinal and/or SRF and crowded disc and occasional optic nerve edema [Figure 4]. Peripapillary choroidal congestion with a compartment-like effect on the peripapillary region was proposed as an etiologic mechanism. The association of PPS with choroidal folds, short axial length, and hyperopia indicated similarities with Uveal Effusion

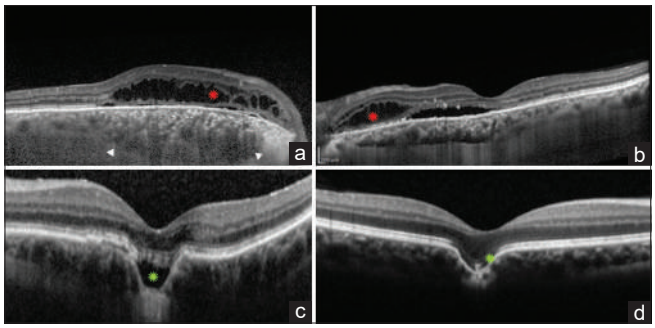


Figure 4: (a and b) Spectral domain-optical coherence tomography enumerating the features of peripapillary pachychoroid syndrome. Note the nasal intraretinal schisis (red asterisk) with maximum choroidal thickness nasally near the optic nerve head (ONH) (white triangle). (c) Spectral domain-optical coherence tomography imaging of a nonconforming focal choroidal excavation where the photoreceptor tips are detached from the underlying retinal pigment epithelium, with a hyporeflective cleft (green asterisk). (d) Spectral domain-optical coherence tomography imaging of a conforming focal choroidal excavation, where the photoreceptor tips are in direct contact with the retinal pigment epithelium

syndrome (UES). Serous macular detachments diffuse granular intense hyperfluorescence in early and late ICGA as usually seen only in UES and makes it distinct. It is important to recognize this clinical entity, which can be confused with posterior uveitis and neuro-ophthalmologic disorders to avoid unnecessary interventions.

PACHYDRUSEN

Coined by Spaide,[17] Pachydrusen is a relatively new entity characterized by yellowish-white Sub-RPE drusenoid deposits in patients with the thick choroid [Figure 5]. The various locations described for the pachydrusen are in the posterior pole sparing the macula, peripapillary area, and near arcades, while the distribution pattern can be solitary, scattered, or clustered having a better-defined outer border. Choroidal morphology under pachydrusen showed increased Haller's layer thickness with attenuation of choriocapillaris. Whether the occurrence of these changes is a causal or resultant phenomenon needs to be elucidated.

RETICULAR PSEUDO DRUSEN OR SUBRETINAL DRUSENOID DEPOSITS

Subretinal drusenoid deposits (SDD) is unique from other subtypes of drusen by being located above the level of the RPE [Figure 5]. Although they can appear in individuals with no other apparent pathology, their highest rates of occurrence are in association with AMD. Reticular pseudodrusen are also documented in diseases such as Sorsby's fundus dystrophy, pseudoxanthoma elasticum, and acquired vitelliform lesions.[18] They are found bilaterally, more in females and with increased age. On histological examination, RPD has been shown to have

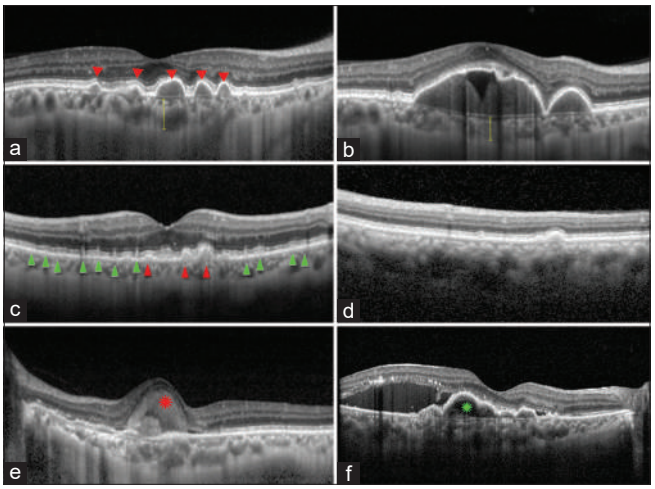


Figure 5: (a-d) Images showing various types of drusen. (a) Spectral domain optical coherence tomography imaging of soft drusen (red triangle) located below the retinal pigment epithelium. Mild disruption of the ellipsoid zone is visible. No signs of subretinal fluid or choroidal neovascular membranes. (b) large soft confluent drusen obscuring the foveal contour (c) Spectral domain optical coherence tomography demonstrating multiple reticular pseudodrusen as hyper-reflectance between the ellipsoid and retinal pigment epithelium (green triangle) in contrast to typical soft drusen (red triangles) which clearly reside below retinal pigment epithelium. (e) Spectral domain optical coherence tomography image showing the presence of subretinal hyperreflective membrane (SHRM, red asterisk) in type 2 classic choroidal neovascular membranes. (f) Spectral domain-optical coherence tomography image demonstrating a type 1 choroidal neovascular membranes with a fibrovascular pigment epithelial detachment (green asterisk) without SHRM

discrete compositions from typical drusen, suggesting different pathways of pathogenesis. SDD is thought to be a precursor lesion for Type 3 choroidal neovascular membranes (CNVM) or retinal angiomatous proliferation.

Zweifel *et al*.,[19] have proposed a three-stage classification for SDD using the amount of hyperreflective material visible on SDOCT and the level of distortion to the ellipsoid layer. Stage 1 is defined by the diffuse deposition of granular hyperreflective material between the RPE and the EZ. Stage 2 represents the progression of material accumulation between the RPE and EZ, forming small mounds and large enough to distort the contour of the EZ. In stage 3, SDD takes on a conical shape, in contrast to the gently rounded appearance of stage 2 lesions, and punch through the ellipsoid boundary.

FOCAL CHOROIDAL EXCAVATION

FCE is a concavity in the choroid occurring without any adjacent scleral abnormality or ectasia and normal-appearing overlying retina.[20] Origin of FCE can be congenital posterior malformation or acquired. FCE is now considered as a distinct entity in pachychoroid spectrum. FCE lesions could represent the scarring of choroidal connective tissue from a previous inflammatory process. Presumably, scar contraction could

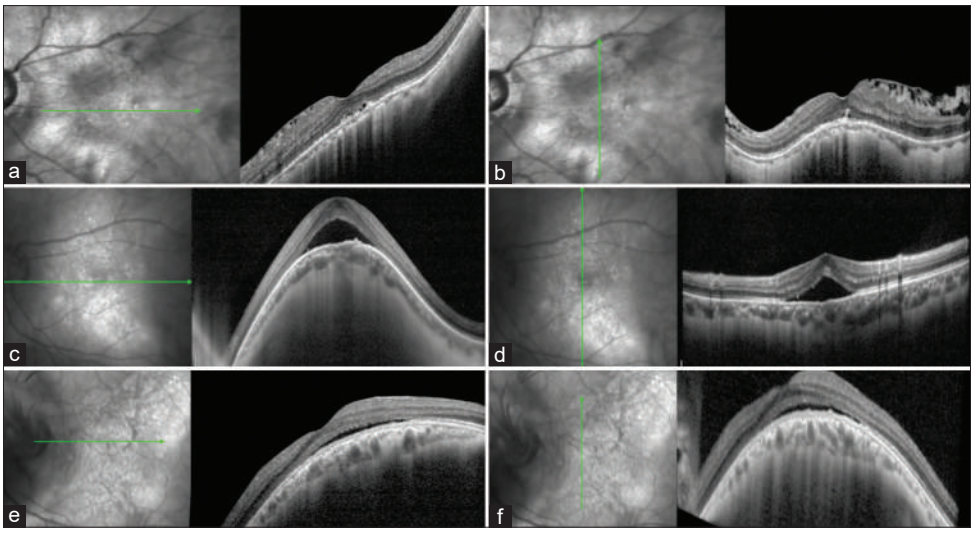


Figure 6: The different configurations of dome-shaped macula. (a and b) Horizontal dome-shaped macula as seen only in vertical optical coherence tomography imaging. Note the ellipsoid zone disruption at the fovea and epiretinal membrane. (a) Horizontal optical coherence tomography fails to reveal the dome-shaped macula. (b) A vertical optical coherence tomography section across the fovea shows an inward protrusion of the macula due to dome-shaped macula. (c and d) Vertical dome-shaped macula revealed only in horizontal optical coherence tomography imaging. Vertical optical coherence tomography fails to show the dome-shaped macula. Both images show subretinal fluid indicative of dome-shaped maculopathy (c) Round dome as seen in both vertical and horizontal optical coherence tomography imaging

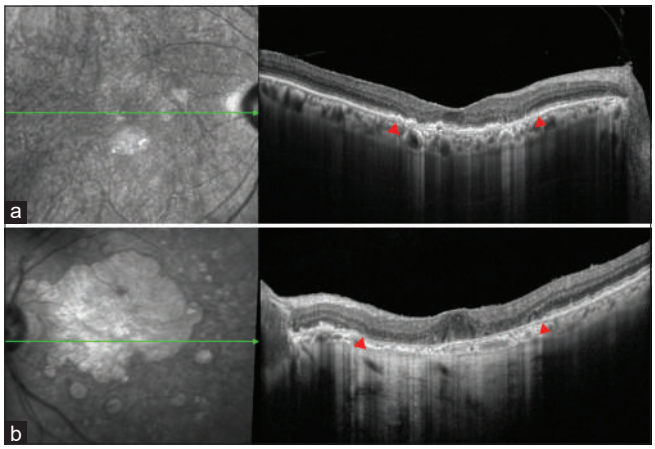


Figure 7: (a) Spectral domain optical coherence tomography image demonstrating features of incomplete retinal pigment epithelium and outer retinal atrophy. Note the irregular loss of retinal pigment epithelium band, and interrupted photoreceptor degeneration and discontinuous reverse shadowing (enhanced visibility of choroid due to loss of retinal pigment epithelium). (b) Spectral domain optical coherence tomography image demonstrating features of complete retinal pigment epithelium and outer retinal atrophy. Note the complete absence of retinal pigment epithelium, ellipsoid zone, external limiting membrane band with thinning of outer nuclear layer, and continuous reverse shadowing

draw the choroid and RPE toward the sclera producing FCE. Abnormal structure or configuration of the overlying RPE or Bruch's membrane in FCE may predispose to CNVM.[21]

Two patterns of excavation have been described. In conforming FCE, the photoreceptor tips are in direct contact with the RPE, whereas in nonconforming FCE the photoreceptor tips appeared to be detached from the underlying RPE, and a hyporeflective cleft can be observed in the intervening space [Figure 4]. Nonconforming FCE can be misdiagnosed as SRF.

DOME SHAPED MACULA

It is the forward convex bulge of the macula that is observed in highly myopic eyes with or without posterior staphylomas, usually detected in vertical and horizontal OCT scans. Three dome-shaped macula (DSM) configurations have been described by Caillaux *et al*.:[22]

- Horizontal oval-shaped dome (63%) – dome can be visible only in vertical OCT
- Vertical oval-shaped dome (16%) – dome can be visible only in vertical OCT
- Round dome (21%) – dome present on both horizontal and vertical OCT scans [Figure 6].

DSM was thought to be secondary to ingrowth of the choroid, but recent research indicates that the main problem is focal scleral thickening in the foveal area.[23,24] One of the major complications in these patients is the loss of vision secondary to the SRF accumulation. DSM mimics CSCR and results in mismanagement and shows the importance of vertical OCT scan. The condition is usually managed conservatively due to conflicting evidence of treatment success using anti-vascular endothelial growth factor (VEGF) injections, oral spironolactone, and PDT.

SUBRETINAL HYPERREFLECTIVE MATERIAL

Subretinal hyperreflective material (SHRM) is a morphological feature identified on the SD-OCT as hyperreflective material

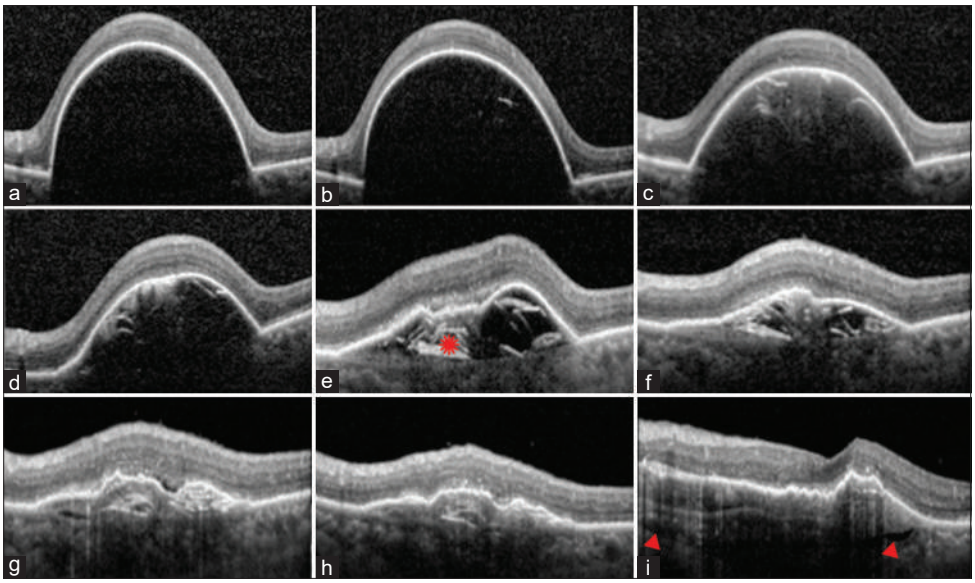


Figure 8: (a-i) Serial spectral domain optical coherence tomography images are showing the evolution of multi-layered pigment epithelial detachment from an extralarge pigment epithelial detachment. Note the formation of neovascular tissue within the pigment epithelial detachment in layers (red asterisk) and pre choroidal cleft (Red Triangle) with triple-layer sign

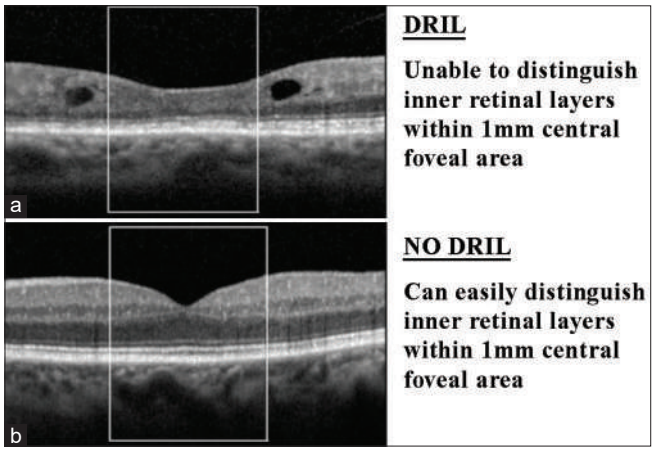


Figure 9: Representative spectral domain optical coherence tomography images showing combinations of the presence or absence of disorganization of retinal inner layers in diabetic macular edema. The central 1 mm foveal area is enclosed in the white box

located between neurosensory retina and RPE [Figure 5]. The range of SHRM seen in macular disease includes neovascular tissue, fibrin, exudates, vitelliform material, and hemorrhage. OCTA can distinguish vascular from avascular SHRM. OCTA reveals a strong intrinsic flow signal with an entangled network in a vascular SHRM.[25]

SHRM has a clear role as a biomarker or prognostic factor in neovascular AMD management.[26] The presence of foveal involving SHRM, well-defined SHRM borders, and thicker SHRM at baseline are associated with poor visual outcome after anti-VEGF therapy even though there is a reduction in the size of SHRM.[27,28] SHRM in wet AMD should be differentiated from fibrin in CSCR. The thickness of the choroid and vacuole sign,[29] will help to differentiate the same.

COMPLETE RETINAL PIGMENT EPITHELIUM AND OUTER RETINAL ATROPHY AND INCOMPLETE RETINAL PIGMENT EPITHELIUM AND OUTER RETINAL ATROPHY

The classification of atrophy meeting recently proposed a consensus definition and nomenclature for SD-OCT defined geographical atrophy in the setting of AMD.[30]

They defined complete retinal pigment epithelium and outer retinal atrophy (cRORA): (1) a region of choroidal hypertransmission of at least 250 ∝m in diameter, (2) a zone of attenuation or loss of the RPE of at least 250 ∝m in diameter, and (3) evidence of overlying photoreceptor degeneration (loss of the IZ, EZ, and ELM and thinning of the outer nuclear layer). Incomplete retinal pigment epithelium and outer retinal atrophy (iRORA) does not fulfill all three criteria for cRORA and typically demonstrate discontinuous hypertransmission, a present but irregular or interrupted RPE band, and interrupted photoreceptor degeneration [Figure 7].Complete Outer Retinal Atrophy (cORA) is defined by continuous absence of the EZ and interdigitation zone and severe thinning of the outer retina with an intact RPE band. Incomplete Outer Retinal Atrophy (iORA) demonstrates thinning of the outer retina, an intact RPE band, and no hypertransmission. Visual acuity will be worst in cRORA.

MULTILAYERED PIGMENT EPITHELIAL DETACHMENT

In 2014, Freund *et al*. coined the term multilayered

pigment epithelial detachment (PED) for organized layers of hyperreflective bands between the RPE monolayer and Bruch's membrane within vascularized PEDs.[31] It is comprised fibrocellular tissue with contractile properties organized in a spindle-shaped configuration. Khan *et al*.,[32] noted the presence of a prechoroidal cleft in multi-layered PED and described as one component of a "*triple-layer" sign*: Sub-RPE neovascular tissue, the hyporeflective space, and the underlying choroid [Figure 8].

Although the appearance of multilayered PED can be rather dramatic, these eyes maintain surprisingly good visual acuity, presumably because the neovascular and subsequent cicatricial process is confined to the sub-RPE space and the neovascular tissue may provide oxygenation or nutritional support to the outer retina and RPE, protecting against involution and geographic atrophy.

DISORGANIZATION OF RETINAL INNER LAYERS

Disorganization of retinal inner layers (DRIL) has been defined by Sun *et al*.,[33] as the lack of distinguishable boundaries between the ganglion cell– inner plexiform layer (IPL) complex, inner nuclear layer, and outer plexiform layer in the central 1000 μ in DME patients [Figure 9]. The development of DRIL is not specific to DME, it is a common response to retinal stress. DRIL is a negative predictive OCT biomarker. Centrally located DRIL and the extent of disruption were both associated with worse visual acuity. For each 100 μm increase in DRIL, there is a negative impact of approximately 6 letters, which is more than 1 line on the ETDRS chart. It has been hypothesized that the disorganization of the inner retina occurs when bipolar axons snap when their elasticity limit has been exceeded because of edema. It has also been suggested that DRIL represents the loss of bipolar, amacrine, or horizontal cells within the inner retinal layers. Recently, it has been found that DEX implant has the potential to ameliorate DRIL and is related to the architectural effect on the Muller cells.[34]

LAMELLAR HOLE ASSOCIATED EPIRETINAL PROLIFERATION

Lamellar Hole associated epiretinal proliferation is the epiretinal proliferation arising from Muller cells seen in the degenerative lamellar macular hole (LMH). LMH can be tractional or degenerative [Figure 10].[35] Degenerative LMH has a "top hat" morphology and is associated with the presence of intraretinal cavitation, potentially affecting all retinal layers. A foveal "bump" of spared retinal tissue is frequently present, as well as outer retinal disruption. There won't be no visual benefit after surgery, despite anatomical improvement. Tractional LMH has a "moustache" appearance and is associated with the presence of Epiretinal or premacular membrane. These lesions present with a "schitic" morphology, with a sharp split located in the Henle's layer, which separates the inner and outer retina. Foveal photoreceptors are frequently spared and usually have better visual acuity.

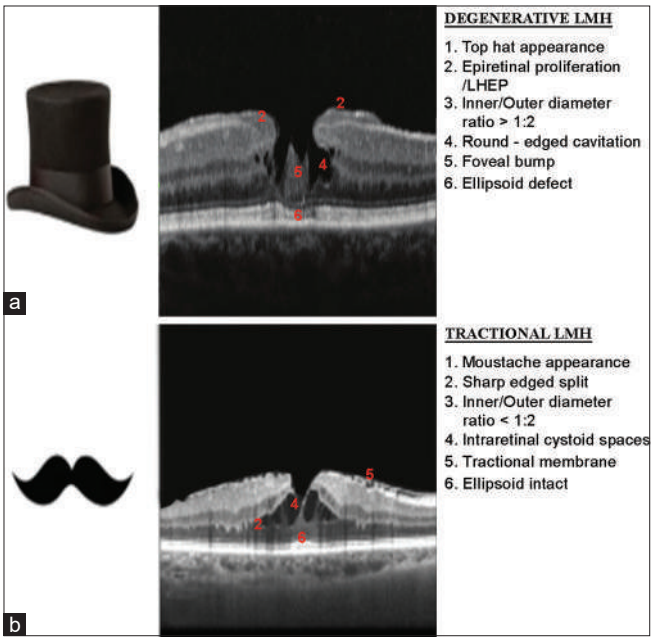


Figure 10: The features of both variants of the lamellar macular hole (a). Degenerative LMH (b)Tractional LMH

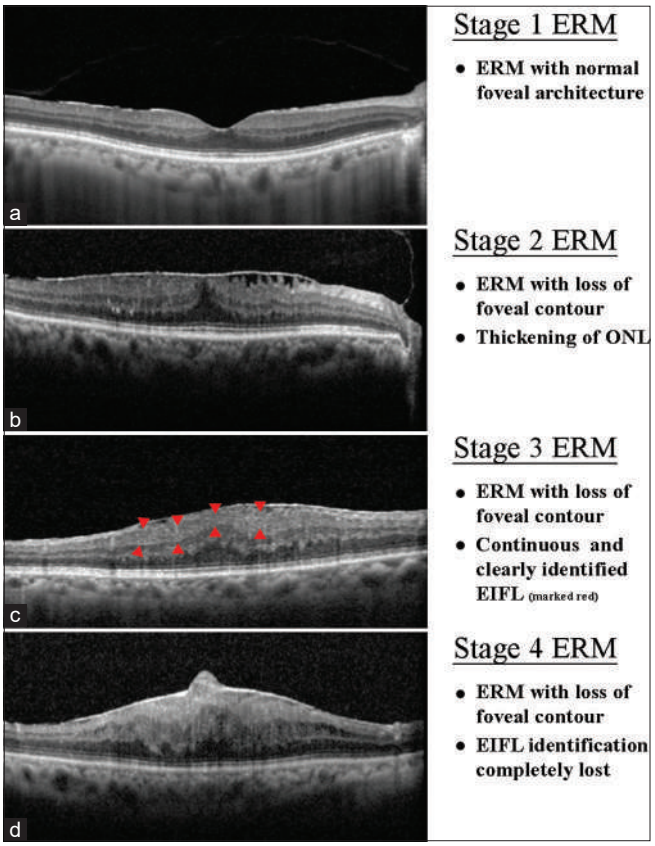


Figure 11: (a-d) The four stages of the epiretinal membrane. Ectopic inner foveal layers marked between the red arrows in (c)

ECTOPIC INNER FOVEAL LAYERS

The chronic anteroposterior and centripetal traction caused by the epiretinal membrane (ERM) may induce the displacement and reorganization of the inner retinal layers, creating a continuous floor of inner retinal tissue extending from the inner nuclear layer (INL) and IPL across the central fovea and referred to as ectopic inner foveal layers [Figure 11]. Ectopic inner foveal layer formation represents an important sign of ERM progression and it is the keystone of a newly proposed SD-OCT staging scheme for idiopathic ERM.[36] OCTA taken in Stage 3 ERM with ectopic inner foveal layers (EIFL) has shown significantly distorted or absent FAZ. EIFL is associated with reduced pre- and post-operative visual acuity. The thickness of EIFL has a negative correlation with visual outcome.

DENGUE-INDUCED INFLAMMATORY, ISCHEMIC FOVEOLITIS AND OUTER MACULOPATHY

Approximately 5%–8% of patients with dengue fever are at a risk of developing permanent visual loss due to posterior segment involvement. The dengue-induced inflammatory, ischemic foveolitis, and outer maculopathy (DIII-FOM) encompass pathological changes induced by ischemia and inflammation of predominantly the outer retina. Inflammatory insult is evidenced in SDOCT by the presence of vitreous cells, cystoid spaces in Henle's layer, "conical" foveal elevations, presence of outer plexiform and outer nuclear hyper-reflectivity and disruption of ELM, EZ and inter-digitation zone. Immune complex deposition in the deep capillary plexus may be the reason for retinal ischemia. Although the inflammatory insult is controlled by early initiation of corticosteroids, the recovery of ischemic insult is unclear, resulting in permanent scotoma.[37]

ACUTE MACULAR NEURORETINOPATHY AND PARACENTRAL ACUTE MIDDLE MACULOPATHY

Both lesions present with acute onset of paracentral scotomas. First described by Bos and Deutman,[38] in 1975, acute macular neuroretinopathy (AMN) is a rare retinal disorder that typically affects a young healthy woman in their teens-30's. The classic funduscopic finding in AMN is the presence of single or multiple dark, well-defined, wedge-shaped intraretinal lesions pointing to the fovea, often in a flower petal arrangement. SD-OCT demonstrates band-like hyper-reflective plaques at the ONL/OPL junction indicating disruption of photoreceptor cell bodies and their axons.[39,40] With the resolution of this ONL/OPL hyper-reflectivity, thinning of the ONL and EZ disruption occurs.

More recently, Sarraf *et al*.[41] identified characteristic AMN lesions at the level of the INL, a novel SD-OCT finding referred to as paracentral acute middle maculopathy (PAMM) [Figure 12]. PAMM may be idiopathic and may even develop in young and healthy individuals with an otherwise normal ocular examination. If so, appropriate systemic workup to exclude systemic or cardiovascular risk factors is advisable. Although the visual prognosis is good, the complete resolution of scotomas never occurs in both lesions.[42]

Financial support and sponsorship
Nil.

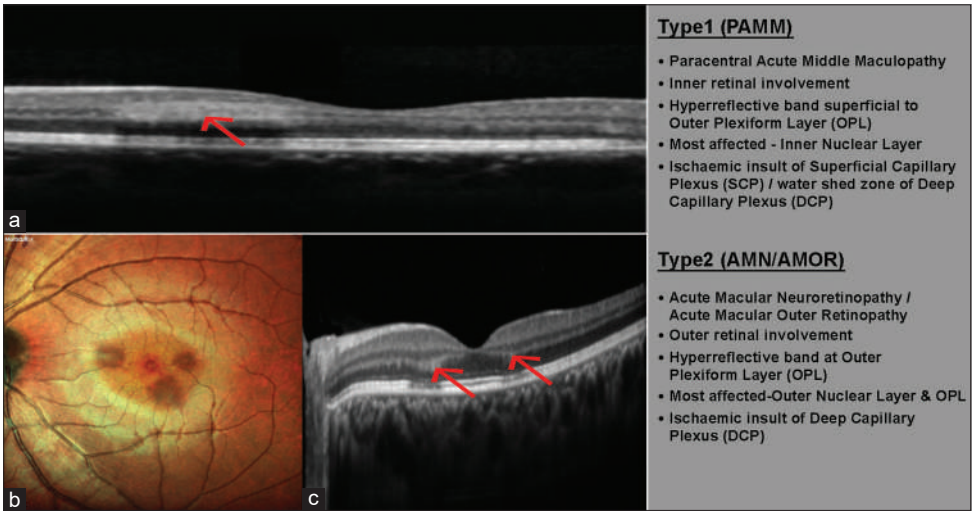


Figure 12: (a) Spectral domain optical coherence tomography of paracentral acute middle maculopathy shows hyperreflective band-like lesion located above the outer plexiform layer in the paracentral macula (b) Multicolour fundus photograph showing wedge-shaped lesions at the macula with apex directing toward the macula. (c) Spectral domain optical coherence tomography of acute macular neuroretinopathy shows hyperreflective band-like lesion located at the junction of the outer plexiform and inner nuclear layer

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Huang D, Swanson EA, Lin CP, Schuman JS, Stinson WG, Chang W, *et al*. Optical coherence tomography. Science 1991;254:1178-81.
2. Gabriele ML, Wollstein G, Ishikawa H, Kagemann L, Xu J, Folio LS, *et al*. Optical coherence tomography: History, current status, and laboratory work. Invest Ophthalmol Vis Sci 2011;52:2425-36.
3. Staurenghi G, Sadda S, Chakravarthy U, Spaide RF, International Nomenclature for Optical Coherence Tomography (IN-OCT) Panel. Proposed lexicon for anatomic landmarks in normal posterior segment spectral-domain optical coherence tomography: The IN•OCT consensus. Ophthalmology 2014;121:1572-8.
4. Verhoeff FH. A hitherto undescribed membrane of the eye and its significance. Boston Med Surg J 1903;149:456–8.
5. Gin TJ, Wu Z, Chew SK, Guymer RH, Luu CD. Quantitative analysis of the ellipsoid zone intensity in phenotypic variations of intermediate age-related macular degeneration. Invest Ophthalmol Vis Sci 2017;58:2079-86.
6. Margolis R, Spaide RF. A pilot study of enhanced depth imaging optical coherence tomography of the choroid in normal eyes. Am J Ophthalmol 2009;147:811-5.
7. Francis JH, Habib LA, Abramson DH. Peripheral leptochoroid: Clinical and anatomical findings. Br J Ophthalmol 2018;102:120-5.
8. Pang CE, Sarraf D, Freund KB. Extreme choroidal thinning in high myopia. Retina 2015;35:407-15.
9. Spaide RF. Age-related choroidal atrophy. Am J Ophthalmol 2009;147:801-10.
10. Ayachit G, Ayachit A, Nadgir H, Joshi S. Validating the pachychoroid disease spectrum using multimodal imaging. Indian J Ophthalmol 2018;66:10224
11. Warrow DJ, Hoang QV, Freund KB. Pachychoroid pigment epitheliopathy. Retina 2013;33:1659-72.
12. Pang CE, Freund KB. Pachychoroid pigment epitheliopathy may masquerade as acute retinal pigment epitheliitis. Invest Ophthalmol Vis Sci 2014;55:5252.
13. Pang CE, Freund KB. Pachychoroid neovasculopathy. Retina 2015;35:1-9.
14. Sato T, Kishi S, Watanabe G, Matsumoto H, Mukai R. Tomographic features of branching vascular networks in polypoidal choroidal vasculopathy. Retina 2007;27:589-94.
15. Sheth J, Anantharaman G, Chandra S, Sivaprasad S. "Double-layer sign" on spectral domain optical coherence tomography in pachychoroid spectrum disease. Indian J Ophthalmol 2018;66:1796-801.
16. Phasukkijwatana N, Freund KB, Dolz-Marco R, Al-Sheikh M, Keane PA, Egan CA, *et al*. Peripapillary pachychoroid syndrome. Retina 2018;38:1652-67.
17. Spaide RF. Disease expression in nonexudative age-related macular degeneration varies with choroidal thickness. Retina 2018;38:708-16.
18. Finger RP, Wu Z, Luu CD, Kearney F, Ayton LN, Lucci LM, *et al*. Reticular pseudodrusen: A risk factor for geographic atrophy in fellow eyes of individuals with unilateral choroidal neovascularization. Ophthalmology 2014;121:1252-6.
19. Zweifel SA, Spaide RF, Curcio CA, Malek G, Imamura Y. Reticular pseudodrusen are subretinal drusenoid deposits. Ophthalmology 2010;117:303-120.
20. Margolis R, Mukkamala SK, Jampol LM, Spaide RF, Ober MD, Sorenson JA, *et al*. The expanded spectrum of focal choroidal excavation. Arch Ophthalmol 2011;129:1320-5.
21. Xu H, Zeng F, Shi D, Sun X, Chen X, Bai Y. Focal choroidal excavation complicated by choroidal neovascularization. Ophthalmology 2014;121:246-50.
22. Caillaux V, Gaucher D, Gualino V, Massin P, Tadayoni R, Gaudric A. Morphologic characterization of dome-shaped macula in myopic eyes with serous macular detachment. Am J Ophthalmol 2013;156:958-670.
23. Gaucher D, Erginay A, Lecleire-Collet A, Haouchine B, Puech M, Cohen SY, *et al*. Dome-shaped macula in eyes with myopic posterior staphyloma. Am J Ophthalmol 2008;145:909-14.
24. Imamura Y, Iida T, Maruko I, Zweifel SA, Spaide RF. Enhanced depth imaging optical coherence tomography of the sclera in dome-shaped macula. Am J Ophthalmol 2011;151:297-302.
25. Dansingani KK, Tan ACS, Gilani F, Phasukkijwatana N, Novais E, Querques L, *et al*. Subretinal Hyperreflective Material Imaged With Optical Coherence Tomography Angiography. Am J Ophthalmol 2016;169:235-48.
26. Willoughby AS, Ying GS, Toth CA, Maguire MG, Burns RE, Grunwald JE, *et al*. Subretinal hyperreflective material in the comparison of age-related macular degeneration treatments trials. Ophthalmology 2015;122:1846-5300000.
27. Pokroy R, Mimouni M, Barayev E, Segev F, Geffen N, Nemet AY, *et al*. Prognostic value of subretinal hyperreflective material in neovascular age-related macular degeneration treated with bevacizumab. Retina 2018;38:1485-91.
28. Kumar JB, Stinnett S, Han JIL, Jaffe GJ. Correlation of subretinal hyperreflective material morphology and visual acuity in neovascular age-related macular degeneratioN. Retina 2020;40:845-56.
29. Rajesh B, Kaur A, Giridhar A, Gopalakrishnan M. "Vacuole" sign adjacent to retinal pigment epithelial defects on spectral domain optical coherence tomography in central serous chorioretinopathy associated with subretinal fibrin. Retina 2017;37:316-24.
30. Sadda SR, Guymer R, Holz FG, Schmitz-Valckenberg S, Curcio CA, Bird AC, *et al*. Consensus definition for atrophy associated with age-related macular degeneration on OCT: Classification of atrophy report 3. Ophthalmology 2018;125:537-48.
31. Rahimy E, Freund KB, Larsen M, Spaide RF, Costa RA, Hoang Q, *et al*. Multilayered pigment epithelial detachment in neovascular age-related macular degeneration. Retina 2014;34:1289-95.
32. Khan S, Engelbert M, Imamura Y, Freund KB. Polypoidal choroidal vasculopathy: Simultaneous indocyanine green angiography and eye-tracked spectral domain optical coherence tomography findings. Retina 2012;32:1057-68.
33. Sun JK, Lin MM, Lammer J, Prager S, Sarangi R, Silva PS, *et al*. Disorganization of the retinal inner layers as a predictor of visual acuity in eyes with center-involved diabetic macular edema. JAMA Ophthalmol 2014;132:1309-16.
34. Zur D, Iglicki M, Sala-Puigdollers A, Chhablani J, Lupidi M, Fraser-Bell S, *et al*. Disorganization of retinal inner layers as a biomarker in patients with diabetic macular oedema treated with dexamethasone implant. Acta Ophthalmol 2020;98:e217-e223.
35. Pang CE, Spaide RF, Freund KB. Epiretinal proliferation seen in association with lamellar macular holes: A distinct clinical entity. Retina 2014;34:1513-23.
36. Govetto A, Lalane III RA, Sarraf D, Figueroa MS, Hubschman JP. Insights into epiretinal membranes: Presence of ectopic inner foveal layers and a new optical coherence tomography staging scheme. American journal of ophthalmology. 2017 Mar 1;175:99-113.
37. Agarwal A, Aggarwal K, Dogra M, Kumar A, Akella M, Katoch D, *et al.* OCTA Study Group. Dengue-induced inflammatory, ischemic foveolitis and outer maculopathy: A swept-source imaging evaluation. Ophthalmology Retina. 2019 Feb 1;3 (2):170-7.
38. Bos PJ, Deutman AF. Acute macular neuroretinopathy. Am J Ophthalmol 1975;80:573-84.
39. Miller MH, Spalton DJ, Fitzke FW, Bird AC. Acute macular neuroretinopathy. Ophthalmology 1989;96:265-9.

40. Fawzi AA, Pappuru RR, Sarraf D. Acute macular neuroretinopathy: Longterm insights revealed by multimodal imaging. Retina. 2012;32:1500-1513.
41. Sarraf D, Rahimy E, Fawzi AA, Sohn E, Barbazetto I, Zacks DN, *et al*. Paracentral acute middle maculopathy: A new variant of acute macular neuroretinopathy associated with retinal capillary ischemia. JAMA Ophthalmol 2013;131:1275-87.
42. Chen X, Rahimy E, Sergott RC, Nunes RP, Souza EC, Choudhry N, *et al*. Spectrum of Retinal Vascular Diseases Associated With Paracentral Acute Middle Maculopathy. Am J Ophthalmol 2015;160:26-340.

Major Review

Astigmatism correction with toric intraocular lens during cataract surgery

ABSTRACT

Currently toric intraocular lens (TIOL) implantation is the most reliable method to correct pre-existing corneal astigmatism of 1D or more at the time of cataract surgery. A good surgical outcome depends on proper patient selection, accurate preoperative measurements, precise intraocular lens (IOL) power calculation and execution of proper surgical plan. Accurate keratometry measurement on a pristine cornea is crucial in TIOL power calculation. The recognition of the importance of posterior corneal astigmatism and its inclusion into various formulas and recent methods to measure it directly have resulted in better postoperative refractive outcomes. Fine-tuning of surgery with digital guidance in TIOL alignment and new lens designs with better rotational stabilities have helped achieve more predictable outcomes. Postoperative IOL rotation is the major cause of patient dissatisfaction after TIOL surgery. However, various correction algorithms and formulas are available which can help optimal corrective re rotation to address this problem satisfactorily.

Keywords: Astigmatism, cataract surgery, toric intraocular lenses

INTRODUCTION

The aim of modern cataract surgery has shifted from just removal of cataract to the provision of spectacle free vision. Currently, around 40% of people undergoing cataract surgery have astigmatism of more than 1D and 8% more than 2D.[1-3] Residual postoperative astigmatism is an important cause of patient dissatisfaction after cataract surgery.

Astigmatism can be corrected by limbal relaxing incisions, astigmatic keratotomies, opposite clear corneal incisions, or by currently toric intraocular lenses (TIOLs). Corneal incision techniques are less predictable and results are more likely to regress over time.[4,5] In comparison, TIOLs are more predictable and stable.

At present, standard TIOLs are available in cylinder powers of 1.5D to 6.0D and are intended to correct preexisting regular corneal astigmatism from 0.75D to 4.75D.[6] Extended series and custom made TIOLs can correct cylinders up to 11-12D.

Submitted: 02-Sep-2020 **Accepted:** 03-Sep-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_133_20	

Postoperative residual refractive corneal astigmatism of up to 0.5D may not need correction, but astigmatism of 0.75D or more should be addressed in patients seeking spectacle independence, especially in those choosing multifocal intraocular lenses (IOLs).[7]

The 1st TIOL was a three piece non-foldable lens introduced by Shimizu *et al*. in 1992.[8] The first foldable TIOL became available in 1994.[9] However, these lenses had a relatively high postoperative rotation rate. Since then, various modifications have come in TIOL models.

TIOL is different from the spherical IOL in its toricity on the anterior, posterior, or both surfaces. There are marks near the haptic optic junction, which represent the plus axis cylinder

Sandhya N, Keerthana Zacharia, Saikumar S. J

Department of Cataract Services, Giridhar Eye Institute, Kochi, Kerala, India

Address for correspondence: Dr. Sandhya N, Senior Consultant, Giridhar Eye Institute, Kochi, Cochin - 682 020, Kerala, India.
E-mail: drsandyan@yahoo.com

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Sandhya N, Zacharia K, Saikumar SJ. Astigmatism correction with toric intraocular lens during cataract surgery. Kerala J Ophthalmol 2020;32:244-51.

of the lens, which needs to be aligned with the steep axis of the cornea. Successful TIOL outcome depends on proper patient selection, acquisition of accurate preoperative data, precise IOL power calculation and execution of the proper surgical plan.

PATIENT SELECTION

Patients with regular bowtie astigmatism are most suitable for TIOL implantation. Patients with irregular astigmatism are not good candidates though spectacle dependence can be reduced with TIOLs.[10]

Patients must have a healthy and pristine ocular surface to obtain reliable and reproducible keratometry readings. Dry eye and other ocular surface diseases should be addressed before taking corneal measurements. Remember to measure the cornea before refraction, tonometry and dilatation.

A stable IOL-bag complex is a prerequisite for the rotational stability of the TIOL. Zonular instability and posterior capsule dehiscence are relative contraindications for TIOL implantation. Patients who have undergone prior intraocular surgeries may not achieve the intended results with TIOLs due to the surgically induced anatomical changes and the increased chance of IOL rotation.[11]

INTRAOCULAR LENS POWER CALCULATION

Manifest refraction should never be taken into account for IOL power calculations because it involves both corneal and lenticular astigmatism. Only corneal astigmatism is considered when planning TIOL.

In with-the-rule (WTR) astigmatism, the vertical corneal meridian is steeper. In against the rule (ATR) astigmatism, the horizontal meridian is steeper. Normally anterior cornea has WTR astigmatism and this gets modified by the posterior cornea, which has ATR astigmatism and hence this needs to be taken into account in TIOL power calculations to avoid overcorrection of WTR (by a factor 0.5D) and under correction of ATR astigmatism (by a factor 0.3D).[12] Because of this, the threshold for putting a TIOL in the case of WTR astigmatism is around 1.25D and for ATR astigmatism around 0.4–0.5D. Studies have also shown that ignoring posterior corneal astigmatism (PCA) may lead to axis errors of $7.4° \pm 10.3°$ due to the lack of accurate prediction of total corneal astigmatism (TCA.)[13]

Precise keratometry (K) measurements are important in identifying the correct power and axis of corneal astigmatism. The keratometers and Placido based topography systems measure only the anterior corneal curvature. They assume a fixed ratio between the anterior and posterior curvature and estimate the PCA.[14] Point-source color LED topographer, OCT and Scheimpflug image-based tomographers measure anterior and posterior corneal curvatures. Devices using these technologies, including Galilei, Pentacam HR, and Cassini, provide TCA and power. Although the use of direct measurements of PCA is theoretically useful in decreasing postoperative astigmatism after TIOL implantation, studies have shown that outcomes in normal eyes using these measurements are not better than those achieved by using mathematical models that estimate PCA such as Barrett's toric calculator.[15]

Be cautious if the average corneal power difference between the two eyes is >0.9D, and the patient cannot adequately fixate during keratometry measurements. It is better to further validate the measurements if astigmatism is >2.5D and if corneal power is <41.0D or >47.0 D or if the corneal diameter is <10.75 mm or >13.0mm.

For better outcomes you can take keratometry measurements from 2 devices that use different principles.[16,17] Cases with similar steep corneal meridian on different devices are good candidates for TIOL implantation. However, if significant variability in both the axis and magnitude of TIOL is observed on different devices, the patient should be evaluated to rule out coexistent ocular comorbidities.[11] The visual outcomes may not be satisfactory in such cases. The highest precision for planning TIOL power and axis is achieved by combining the keratometry and OCT data.[11]

An ideal IOL power calculation formula should take into account the surgically induced astigmatism (SIA), PCA as well as the effective lens position (ELP).[11] An incision into the cornea will alter its astigmatism. However, the magnitude and direction of change depend on the age of the patient, size and shape of the incision, location on the cornea and corneal biomechanical properties. Prediction of SIA is often difficult, and therefore, many surgeons use a value of approximately 0.5D. However, recent reports have shown that the range of SIA is less predictable and a "centroid" value defined as a two dimensional mean of SIA of 0.1D can be used for temporal MICS.[18]

In general, there are two approaches for TIOL calculations. The first is to use mathematical models (e. g. Barrett toric calculator (BTC), Abulafia-Koch) that estimates the predicted postoperative residual refractive astigmatism based on anterior corneal measurements. The second is the use of TCA measurements (Scheimpflug imaging with Panacea Toric Calculator, ray tracing software such as PhacoOptics).

Comparative studies found that in normal eyes, the direct measurement approach was not superior to the estimated mathematical approaches.[19]

Various IOL power calculating formulas are available, and currently the BTC is the most widely used. It takes into account both PCA and ELP and is robust throughout the axial lengths. It is available online through the ASCRS and APACRS websites. BTC also has the option of input of 3 different K readings from three different devices through integrated K and can also use the measured K values.

In addition, most of the IOL manufacturers have their own toric calculators, most of which take into account PCA like AcrySof toric calculator and the Tecnis toric calculator, which are available online.

Studies have been performed comparing BTC and new Alcon AcrySof toric calculator and found that these two methods are highly accurate to calculate toric lens power compared to older methods.[20] A study comparing AcrySof and Tecnis calculators found that the Tecnis calculator tends to predict a higher astigmatic power, however, no postoperative results were compared.[21] All these calculators incorporate PCA and ELP, still tend to overcorrect WTR astigmatism and under correct ATR astigmatism.

SPECIAL SITUATIONS

Eyes with a history of previous corneal surgery or irregular contours due to diseases such as Keratoconus poses a challenge in IOL power calculations. Traditional keratometry measures only the anterior cornea and estimates the diopteric power of cornea based on a fixed ratio of the radii of the two surfaces. Corneal refractive surgery alters the anterior surface without altering the posterior surface of the cornea, affecting the ratio unpredictably. Barrett True-K TK Toric Calculator can be of help in these eyes because it incorporates the direct measurements of the posterior cornea avoiding estimation errors.[22]

In eyes with keratoconus there is an irregular surface, and the apex is offset. TIOL implantation can be deferred in such situations. However, if you are planning a TIOL, elevation-based topography devices (Pentacam or Galilei) have been reported to be advantageous to measure both the anterior and posterior corneal curvatures and to calculate the TCA.[23]

EXECUTION OF PROPER SURGICAL PLAN

The most important aspect in TIOL implantation is marking the steep axis of the cornea and aligning the mark on the TIOL with it. This is because one degree of misalignment of TIOL from the intended axis can cause a loss of approximately 3% of the effective cylinder power, and 30° misalignment can cause complete loss of its toric effect. The axis marking can be done by iris fingerprinting technique, image-guided systems, intraoperative aberrometry based systems or manually.

MANUAL MARKING

Here first, the reference axis and then the target axis are marked. Reference marking is the marking of 3 and 9 o'clock meridia. This should be done in the sitting position to take care of cyclorotation happening on lying down. This is approximately 2°–4° on average but can be up to 15° in individual patients.[24]

Adequate topical anesthesia should be administered, and the patient should be looking at the distance to avoid convergence. The cornea should be dry and the marking should be done on the cornea and not on the conjunctiva (conjunctiva is mobile). The marking may be performed with a skin marking pen in a freehand manner or with the help of various devices such as a thin slit beam [Figure 1], weighted thread, pendulum marker, or Nuijts Solomon bubble marker[12] [Figure 2]. This is followed by the intraoperative alignment of these reference marks to the degree gauge on a fixation ring, and the target axis is then marked with a corneal meridian marker [Figure 3a-c]. Anterior stromal puncture using an ink-stained 26G needle produce marks lasting longer.[25]

One step axis marking may be done with various devices such as tonometer markers, Neuhann one step toric bubble marker, and Geuder Gerten Pendulum marker. A mean error

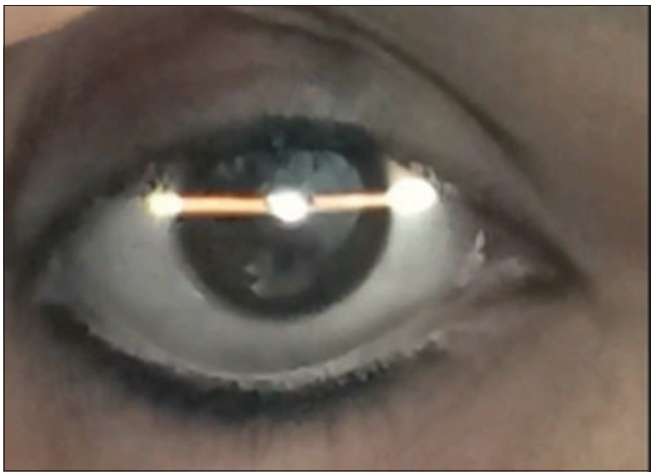


Figure 1: Reference axis marking with slit beam. Mark the point where the bright reflex meets the limbus

of 2.4° ± 0.8° has been observed during axis marking with a bubble marker.[26] The least rotational deviation was observed with a pendular marker (mean 1.8°) in a study comparing four methods of manual marking.[27]

Osher ThermoDot Marker employs a bipolar cautery to create an ink-free, precise reference mark. Other marking systems include the RoboMarker system and electronic TIOL markers.

The manual marking methods have some inherent sources of errors such as smudging and fading of the dye, irregular and broad marks, which can be minimized by a meticulous approach. Digital technologies can be used to further refine this.

DIGITAL TECHNOLOGIES

In 2010, Osher[28] introduced the concept of iris-fingerprinting, which uses the landmarks of the iris to place the axis marks. The Osher Toric Alignment System constituted the basis for the modern image-guided systems.

These technologies can exclude human errors, transcription errors and provide a higher degree of accuracy in intraoperative TIOL alignment. Interconnected devices can measure IOL power, process data and share information to the surgical microscope. These technologies include real-time eye-tracking that compensates for cyclorotation and eye movements to ensure that the superimposed visual overlay always remains in the same position in relation to the eye.

There are three main principles used by these technologies:

1. Limbal registration (image-guided) [Table 1]: An image of the vascularization of the conjunctiva and sclera is captured during biometry and matched with the image obtained by the surgical microscope. An overlay then indicates the steepest axis. Alcon VERION Image-Guided System [Figure 4], Zeiss Callisto Eye with Z align [Figure 5], and TrueVision 3-D Surgical System work on this principle.

Factors altering conjunctival vasculatures like pharmacologic mydriasis, application of topical anesthesia and conjunctival chemosis, ballooning or bleeding may interfere with intraoperative registration in these devices.

2. Intraoperative wavefront aberrometry (IA): IA devices such as ORA system and Holos IntraOp perform a real-time assessment of the aphakic or pseudophakic refraction to provide feedback for TIOL power calculation and alignment.

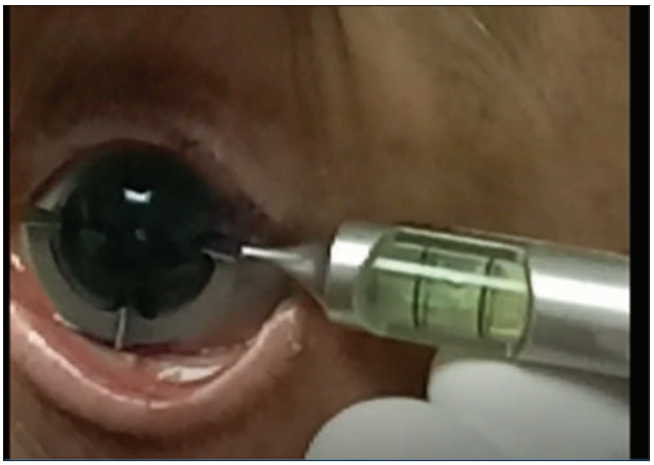


Figure 2: Reference marking with bubble marker. Mark the cornea at the limbus when the bubble is in the center

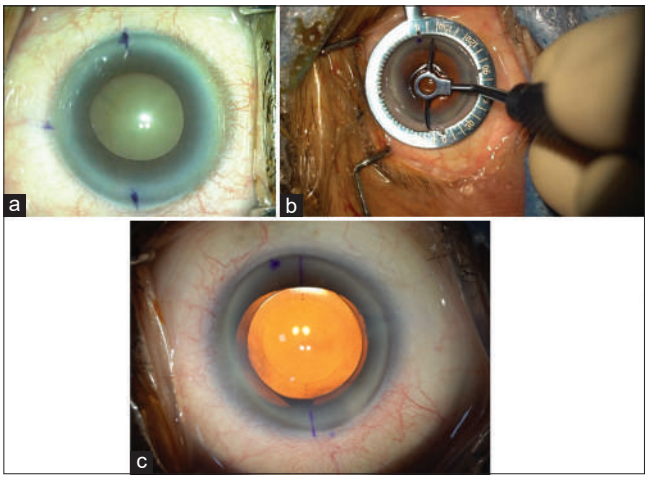


Figure 3: (a) Patient under microscope after reference axis marking. (b) Target axis marking under microscope. (c) Final alignment of the TIOL with the target axis. The 3 dots near the haptic optic junction aligned with the target axis marks

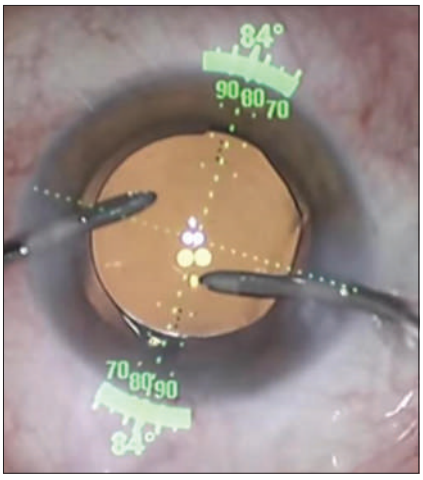


Figure 4: VERION image guided axis alignment

Solomon *et al*. found that the current generation formulas produce clinical outcomes with TIOLs that are as good

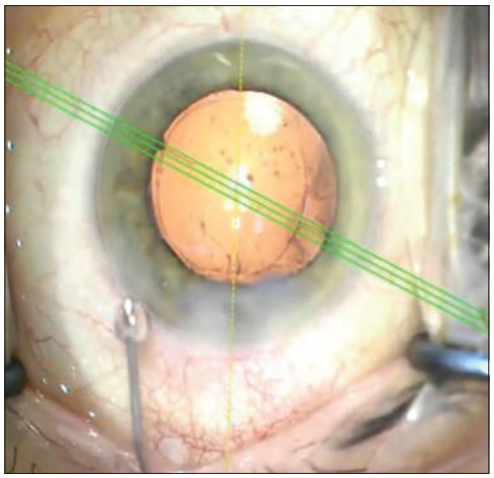


Figure 5: Axis alignment with Zeiss Callisto

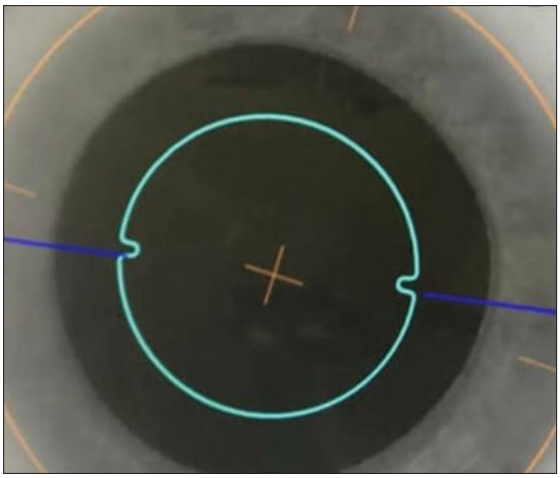


Figure 6: Intelliaxis refractive capsulorhexis

Table 1: Image-guided systems for toric intraocular lens alignment

Image guided system	Preoperative image capture	Intraoperative alignment
Osher Toric Alignment System (OTAS, Haag-Streit, Switzerland)	Slit-lamp photograph	Software with protractor to assign a degree to unique iris landmarks (crypts, nevi, brushfield spots, etc.) and superimpose the degrees on the photograph
iTRACE System (Tracey Technologies, Houston, Tx)	iTrace visual function analyzer	Zaldivar caliper toric tool-provides location of iris/limbal landmarks and their distance in degrees from target axis
VERION (Alcon)	VERION reference unit	VERION digital marker-graphic overlay of target axis based on scleral blood vessels, limbus, and iris details as reference landmarks
CALLISTO Eye and Z Align (Carl Zeiss Meditec)	IOL master 500/700	Graphic overlay of target axis based on limbal vascular landmarks
TrueGuide (TrueVision 3D Surgical system)	i-Optics Cassini, corneal LED topographer	Graphic overlay of target axis based on limbal vascular landmarks

LED: Light-emitting diode, IOL: Intraocular Lens

or better than those achieved with IA.[29] The accuracy of aberrometry readings may be affected by intraoperative corneal edema, pressure by eyelid speculum, presence of air bubbles or clumps of the dispersive ocular viscoelastic device (OVD) in the anterior chamber, centrally located cortex or by dry corneal surface.[30] Before obtaining the readings, ensure a uniform fundal glow and fill the anterior chamber uniformly with OVD to maintain an intraocular pressure of around 20-30.

3. Iris fingerprinting (iris detection): LENSAR uses IntelliAxis Refractive Capsulorhexis to create notches in the anterior capsulorhexis to indicate the steep axis [Figure 6]. Here combining this technology with corneal shape analysis ensures marking of the steep corneal axis by the femtosecond laser in a way that compensates for cyclorotation.

The markings can be virtual that are visible only as projections during surgery (image-guided systems, IA) or real semi-permanent or permanent (intrastromal corneal or capsular markings). Permanent marking is helpful to monitor postoperative misalignment of TIOL.

SURGICAL PROCEDURE

Phacoemulsification is not different from the standard one. The capsulorhexis should overlap the optic of the IOL all around uniformly for better centration of the IOL. The ideal capsulorhexis diameter should be adjusted to the IOL optic diameter and ranges from approximately 4.5-5.5 mm. The mark on the IOL needs to be aligned with the target axis. First, the lens is grossly aligned by rotating it clockwise as it unfolds till it is left about 3°–5° anticlockwise to the final desired axis. Rotate it to the final axis after removing OVD completely and tap the lens gently onto the posterior capsule. While hydrating the corneal incision, care should be taken not to propagate the fluid wave behind the IOL and not to leave the eye hypotonous as both can lead to untoward rotation of the implant. Hydrating the second paracentesis before removing the irrigation handpiece from the eye helps to reduce chamber collapse and rotation of the IOL after the infusion is removed.

In high myopes and in whom the previously operated eye had undergone significant postoperative rotation of IOL, insertion of capsular tension ring (CTR) is an option to increase the rotational stability by increasing the posterior capsule-IOL contact. In a study by Vokrojova *et al*. presence

Table 2: Material, design and power of currently available mono focal toric intraocular lens

Toric IOL	Material	Design	Spherical power (d)	Cylindrical power (d)
AcrySof Toric IOL (Alcon)	Hydrophobic acrylic, aspheric	Flexible loop haptic with stable force technology	+6.0-+34.0	1.50-6.0 (0.75 steps)
AT TORBI (Zeiss, Oberkochen, Germany)	Hydrophilic acrylic with hydrophobic surface, aspheric	Plate haptic	−10.0-+32.0	1.0-12.0 (0.50 steps)
T-flex (Rayner)	Hydrophilic acrylic, aspheric	Loop haptic with AVH technology	−10.0-+35.0	1.0-11.0 (0.25 steps)
Precizon Toric (Ophtec)	Hydrophilic acrylic, aspheric	Biconvex, transitional conic toric design, offset- shaped haptic	+1.0-+34.0	1.0-10.0 (0.50 steps)
LentisT plus (Oculentis)	Hydrophilic acrylic with hydrophobic surface, aspheric	Plate haptics	−10.0-+35.0	0.25-12.0 (0.75 steps/0.01 custom made)
Hoya iSert Toric Preloaded IOL system (Hoya)	Hydrophobic acrylic with PMMA haptic tips, aspheric	PMMA modified C-loop haptic	+6.0-+30.0	1.5-6.0 (0.75 steps)
Staar Toric IOL (Staar Surgical company) Morcher 89A, 92S (Morcher GmbH)	Silicone hydrophilic acrylic	Plate haptic Bag-in-the-lens	+9.50-+28.5 +10.0-+30.0	2.0 or 3.50.50-8.0 (0.25 steps)
TECNIS Toric IOL (Johnson and Johnson, New Brunswick, NJ)	Hydrophobic acrylic, aspheric	"Tri-Fix" modified C-haptic integral with optic	+5.0-+34.0	1.50-6.0 (0.50-1.0 steps)

AVH: Anti- vaulting haptic, PMMA: Polymethyl methacrylate, IOL: Intraocular Lens

Table 3: Material, design and range of power of commercially available multifocal toric intraocular lens

Multifocal Toric IOL	Material	IOL design	Multifocal technology	Near addition (D)	Spherical power (D)	Cylinder power (D)
Acrysof IQ Restor toric (Alcon)	Hydrophobic acrylic	Loop haptic with stable force technology	Diffractive + refractive	+2.5, +3.0	+6.0-+34.0	1.0-3.0 (0.5/0.75 steps)
TECNIS Symfony Toric	Hydrophobic acrylic	Tri-fix haptics offset from optic	Diffractive with echelette feature	Extended range of vision	+5.0-+34.0	1.5-3.75 (0.75 steps)
TECNIS multifocal toric	Hydrophobic acrylic	Haptics offset from optic for 3-point fixation	Diffractive	+4.0	+5.0-+34.0	1.5-4 (0.75/1.00 steps)
PhysIOL (FineVision Toric)	Hydrophilic acrylic	Double C-loop quadripod haptic	Diffractive trifocal	+3.5 for near vision and +1.75 for intermediate vision	+6-+35	1.0-6.0
M-flex T (Rayner)	Hydrophilic acrylic with hydrophobic surface	Loop haptic with AVH technology	Refractive	+3.0 or+4.0	+14.0-+32.0	1.5-6.0 (0.5 steps)
Acri Lisa Toric (Carl Zeiss Meditec)	Hydrophilic acrylic with hydrophobic surface	Four-point plate haptic	Diffractive	+3.75	−10.0-+32.0	1.0-12.0 (0.50 steps)
Lentis Mplus toric (Oculentis)	Hydrophilic acrylic with hydrophobic surface	Both C-loop and plate haptic design	Sector-shaped refractive segment	+3.0	0.0-+36.0	0.25-12.0 0.75/0.01 (custom made) steps
Trulign toric (Bausch and Lomb)	Silicone with silicone and polyimide haptic	Modified plate haptic lens with hinges across the plates adjacent to the optic	Active focus design-apodized diffractive optic	+1.0	+4-+33D	1.25, 2.00, 2.75
PanOptix Toric IOL (Alcon, Fort Worth, TX)	Hydrophobic acrylic, aspheric	Loop haptic with stable force technology	Trifocal, inner diffractive and an outer refractive zone	+2.17 D intermediate add, +3.25 D near add	+6.0-+34.0 D	0.75-2.57
AT LARA Toric 929 MP (Zeiss, Oberkochen, Germany)	Hydrophilic acrylic, aspheric		EDOF diffractive	Depth of focus extension 0.95 and 1.9D	−4.0-+32.0D	1.0-12.0D

AVH: Anti-vaulting haptic, IOL: Intraocular Lens, EDOF: Extended Depth of Focus

or absence of CTR did not make a significant difference in the rotational stability of the TIOL in normal eyes, but in eyes with an axial length of 24 mm or greater, better IOL alignment was observed in the group with CTR.[31] Long axial length is associated with larger capsular bags and lower power IOLs, which are thinner.

CURRENT INTRAOCULAR LENS MODELS

Currently, available IOL designs are shown in Tables 2 and 3. The choice of IOL depends on the surgeon's comfort and patient expectations.

POSTOPERATIVE MANAGEMENT

Patients undergoing TIOL implantation do well with excellent visual outcomes; however, these lenses can rotate after surgery, especially in the early postoperative period, with a majority occurring within 10 days after surgery.[32]

Accuracy of IOL alignment depends on the precision of marking of the target axis, uniformity of IOL overlap with the capsulorhexis, completeness of removal of OVD, water tightness of incisions, and on a stable IOP. Stability also depends on the material, design, and overall diameter of the IOL, axial length of the eye, and capsule contraction. Hydrophobic acrylic IOLs showed the highest capsular adhesive properties, followed by hydrophilic acrylic IOLs and finally silicone IOLs.[11] Limiting physical activities in the first 24 h after surgery has also shown to reduce the chance of IOL rotation.

In a study conducted by Miyake *et al*. on AcrySof toric IOLs, postoperative rotation of >10° was observed in only 1.68% of eyes after 2 years[33] Lee and Chang *et al*.[34] found that nearly all of the TIOL rotations occurred within the first postoperative hour. The mean rotation was 3.79° for tecnis TIOLs and WTR axis IOL orientation was more associated with >10° rotation, which could reflect the effect of gravity on haptic position for vertically oriented IOLs. In their study, the Tecnis TIOLs predominantly rotated counter-clockwise irrespective of the IOL's axis. Postoperative toric IOL misalignment was the major factor for unsatisfactory visual outcomes after surgery, with realignment needed in up to 0.65%–3.3% of cases.[11]

The IOL position should be confirmed postoperatively if a significant difference is found between expected and measured postoperative refraction. The axis of implanted TIOL may be assessed in the postoperative period at the slit lamp with a rotating slit and rotational gauge. The 10° steps on the slit lamp measuring reticle limit the accuracy of this method. Another easy way to measure the axis of TIOL is using various camera-enabled cellular phones and computer software.

In cases of postoperative misalignment, www.astigmatismfix.com can be used to determine the axis of TIOL realignment.[35] This calculator uses the patient's postoperative refraction, IOL power, and axis to predict the new axis of alignment. Barrett Rx Formula can also be used to determine possible piggyback IOL and ideal IOL axis.

Early rerotation tends to lead to improved outcomes. In cases where the lens is felt to be at high risk of repeat rotations, the IOL optic may be captured in the capsular bag to improve rotational stability.[36] In cases with larger amounts of postoperative refractive error which cannot be completely negated by IOL rotation alone, options include IOL exchange, piggyback IOLs, or corneal ablative procedures.

Newer modular IOL systems in clinical trials, such as the ClarVista Harmoni, which have an outer base with haptics and an inner removable optic, would theoretically allow for easier IOL explantations. Another technology to manage postoperative refractive outcomes is the Light Adjustable Lens (LAL, RxSight).[36] When it is selectively exposed to ultraviolet light, it undergoes spherical and cylindrical power changes and allows the surgeon to adjust the final refraction after the lens has stabilized in the eye.

CONCLUSION

The outcome of TIOL implantation has improved dramatically in recent times. Improved corneal measurement techniques, predictive IOL formulas, newer IOL designs and intraoperative alignment techniques have helped surgeons to provide excellent uncorrected visual acuity.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Holland E, Lane S, Horn JD, Ernest P, Arleo R, Miller KM. The AcrySof Toric intraocular lens in subjects with cataracts and corneal astigmatism: a randomized, subject-masked, parallel-group, 1-year study. Ophthalmology 2010;117:2104-11.
2. Hoffmann PC, Hütz WW. Analysis of biometry and prevalence data for corneal astigmatism in 23,239 eyes. J Cataract Refract Surg 2010;36:1479-85.
3. Ferrer-Blasco T, Montés-Micó R, Peixoto-de-Matos SC, González-Méijome JM, Cerviño A. Prevalence of corneal astigmatism before cataract surgery. J Cataract Refract Surg 2009;35:70-5.
4. Núñez MX, Henriquez MA, Escaf LJ, Ventura BV, Srur M, Newball L, *et al*. Consensus on the management of astigmatism in cataract surgery. Clin Ophthalmol 2019;13:311-24.
5. Kaufmann C, Peter J, Ooi K, Phipps S, Cooper P, Goggin M, *et al*. Limbal relaxing incisions versus on-axis incisions to reduce corneal astigmatism at the time of cataract surgery. J Cataract Refract Surg 2005;31:2261-5.
6. Khan MI, Ch'ng SW, Muhtaseb M. The use of toric intraocular lens to correct astigmatism at the time of cataract surgery. Oman J Ophthalmol 2015;8:38-43.
7. Villegas EA, Alcón E, Artal P. Minimum amount of astigmatism that should be corrected. J Cataract Refract Surg 2014;40:13-9.
8. Shimizu K, Misawa A, Suzuki Y. Toric intraocular lenses: correcting astigmatism while controlling axis shift. J Cataract Refract Surg 1994;20:523-6.
9. Grabow HB. Early results with foldable toric IOL implantation. Eur J

Implant Refract Surg 1994; 6:177-8.
10. Luck J. Customized ultra-high-power toric intraocular lens implantation for pellucid marginal degeneration and cataract. J Cataract Refract Surg 2010;36:1235-8.
11. Kaur M, Shaikh F, Falera R, Titiyal JS. Optimizing outcomes with toric intraocular lenses. Indian J Ophthalmol 2017;65:1301-13.
12. Koch DD, Jenkins RB, Weikert MP, Yeu E, Wang L. Correcting astigmatism with toric intraocular lenses: Effect of posterior corneal astigmatism. J Cataract Refract Surg 2013;39:1803-9.
13. Ho JD, Tsai CY, Liou SW. Accuracy of corneal astigmatism estimation by neglecting the posterior corneal surface measurement. Am J Ophthalmol 2009;147:788-95, 795.e1-2.
14. Goggin M, Zamora-Alejo K, Esterman A, van Zyl L. Adjustment of anterior corneal astigmatism values to incorporate the likely effect of posterior corneal curvature for toric intraocular lens calculation. J Refract Surg 2015;31:98-102.
15. Ribeiro FJ, Ferreira TB, Relha C, Esteves C, Gaspar S. Predictability of different calculators in the minimization of postoperative astigmatism after implantation of a toric intraocular lens. Clin Ophthalmol 2019;13:1649-56.
16. Woodcock MG, Lehmann R, Cionni RJ, Breen M, Scott MC. Intraoperative aberrometry versus standard preoperative biometry and a toric IOL calculator for bilateral toric IOL implantation with a femtosecond laser: One-month results. J Cataract Refract Surg 2016;42:817-25.
17. Browne AW, Osher RH. Optimizing precision in toric lens selection by combining keratometry techniques. J Refract Surg 2014;30:67-72.
18. Canovas C, Alarcon A, Rosén R, Kasthurirangan S, Ma JJK, Koch DD, *et al*. New algorithm for toric intraocular lens power calculation considering the posterior corneal astigmatism. J Cataract Refract Surg 2018;44:168-74.
19. Ferreira TB, Ribeiro P, Ribeiro FJ, O'Neill JG. Comparison of methodologies using estimated or measured values of total corneal astigmatism for toric intraocular lens power calculation. J Refract Surg 2017;33:794-800.
20. Ferreira TB, Ribeiro P, Ribeiro FJ, O'Neill JG. Comparison of astigmatic prediction errors associated with new calculation methods for toric intraocular lenses. J Cataract Refract Surg 2017;43:340-7.
21. Park HJ, Lee H, Woo YJ, Kim EK, Seo KY, Kim HY, *et al*. Comparison of the astigmatic power of toric intraocular lenses using three toric calculators. Yonsei Med J 2015;56:1097-105.
22. Abulafia A, Hill WE, Koch DD, Wang L, Barrett GD. Accuracy of the Barrett True-K formula for intraocular lens power prediction after laser *in situ* keratomileusis or photorefractive keratectomy for myopia. J Cataract Refract Surg 2016;42:363-9.
23. Ghiasian L, Abolfathzadeh N, Manafi N, Hadavandkhani A. Intraocular lens power calculation in keratoconus; A review of literature. J Curr Ophthalmol 2019;31:127-34.
24. Chang J. Cyclotorsion during laser *in situ* keratomileusis. J Cataract Refract Surg 2008;34:1720-6.
25. Bhandari S, Nath M. Anterior stromal puncture with staining: A modified technique for preoperative reference corneal marking for toric lenses and its retrospective analyses. Indian J Ophthalmol 2016;64:559-62.
26. Visser N, Berendschot TT, Bauer NJ, Jurich J, Kersting O, Nuijts RM. Accuracy of toric intraocular lens implantation in cataract and refractive surgery. J Cataract Refract Surg 2011;37:1394-402.
27. Popp N, Hirnschall N, Maedel S, Findl O. Evaluation of 4 corneal astigmatic marking methods. J Cataract Refract Surg 2012;38:2094-9.
28. Osher RH. Iris fingerprinting: new method for improving accuracy in toric lens orientation. J Cataract Refract Surg 2010;36:351-2.
29. Solomon KD, Sandoval HP, Potvin R. Evaluating the relative value of intraoperative aberrometry versus current formulas for toric IOL sphere, cylinder, and orientation planning. J Cataract Refract Surg 2019;45:1430-5.
30. Stringham J, Pettey J, Olson RJ. Evaluation of variables affecting intraoperative aberrometry. J Cataract Refract Surg 2012;38:470-4.
31. Vokrojová M, Havlíčková L, Brožková M, Hlinomazová Z. Effect of capsular tension ring implantation on postoperative rotational stability of a toric intraocular lens. J Refract Surg 2020;36:186-92.
32. Kramer BA, Berdahl JP, Hardten DR, Potvin R. Residual astigmatism after toric intraocular lens implantation: Analysis of data from an online toric intraocular lens back-calculator. J Cataract Refract Surg 2016;42:1595-601.
33. Miyake T, Kamiya K, Amano R, Iida Y, Tsunehiro S, Shimizu K. Long-term clinical outcomes of toric intraocular lens implantation in cataract cases with preexisting astigmatism. J Cataract Refract Surg 2014;40:1654-60.
34. Lee BS, Chang DF. Comparison of the rotational stability of two toric intraocular lenses in 1273 consecutive eyes. Ophthalmology 2018;125:1325-31.
35. Berdahl JP, Hardten DR. Residual astigmatism after toric intraocular lens implantation. J Cataract Refract Surg 2012;38:730-1.
36. Tarentino AL, Maley F. A comparison of the substrate specificities of endo-beta-N-acetylglucosaminidases from *Streptomyces griseus* and *Diplococcus pneumoniae*. Biochem Biophys Res Commun 1975;67:455-62.

Original Article

Ocular biometry in an adult Ghanaian population

ABSTRACT

Purpose: To measure ocular biometry and its correlates in an adult population. **Materials and Methods:** The measured biometric data included central corneal thickness (CCT), corneal power (K), anterior chamber depth (ACD), lens thickness (LT), vitreous chamber depth (VCD), axial length (AL), and spherical refractive error (SER) in 162 adult eyes. **Results:** The mean CCT was 506.7 ± 39.2 µm, mean K was: K1 = 42.50 ± 2.00 D, K2 = 43.50 ± 2.44 D, the mean AL was 24.2 ± 1.3 mm, ACD had a mean of 3.5 ± 0.6 mm, LT (mean of 3.9 ± 0.8 mm), VCD (mean of 16.4 ± 1.39 mm), and SER ranged from -10.00 D – +8.00 D. There were no significant correlations between the CCT and SER ($r = 0.06$, $P = 0.44$), LT ($r = 0.06$, $P = 0.59$), ACD ($r = 0.12$, $P = 0.12$), AL ($r = 0.02$, $P = 0.98$), and VCD ($r = 0.05$, $P = 0.68$). Pearson's correlation coefficient tests showed that AL was significantly positively correlated with ACD ($r = 0.4$, $P < 0.001$) and VCD ($r = 0.73$, $P < 0.001$) and negatively correlated with LT ($r = 0.22$, $P < 0.05$). A significant negative weak correlation was found between LT and ACD values ($r = 0.22$, $P < 0.05$). **Conclusion:** A thin CCT was observed among an adult Ghanaian population. CCT was found to be independent of AL. Correlation among ocular biometry showed that AL positively correlated with ACD and negatively correlated with LT.

Keywords: Anterior chamber depth, axial length, central corneal thickness, Ghana, spherical refractive error

INTRODUCTION

Biometric data of the eye are important clinical parameters due to the influence of these measures on the calculation of intraocular lens implant, refractive errors and intraocular pressure measurements among patients with glaucoma.[1-7] Parameters such as anterior chamber depth (ACD), vitreous chamber depth (VCD), axial length (AL), corneal curvature (CC) or astigmatism, and lenticular thickness (LT) may help explain the pathophysiological dynamics of common eye disorders.[8,9] Biometric studies have shown that ALs account for differences in refractive status among age groups.[10-19]

The AL refers to the distance between the anterior pole and the posterior pole of the eye which ranges from 23 to 25 mm in an adult eye (mean of 23.30 mm).[20,21] Age, gender, refractive error, corneal curvature, and racial and ethnic variations have been observed in reported studies in Africa and African Americans.[7,19,22-30]

A study by Ntim-Amponsah *et al.*[31] compared the central corneal thickness (CCT) among glaucomatous (cases) and nonglaucomatous (controls) population in Ghana and found no significant difference between mean CCT of cases and controls. Another study by Ntim-Amponsah *et al.*,[32] normative CCT among 169 Ghanaians were found to be different from neighboring countries and as such cannot be generalized as representative of Ghanaians. This highlights the need for

Samuel Kyei, Frank Assiamah[1], Michael Agyemang Kwarteng[2], Victor Kwesi Ansah[3]

Department of Optometry and Vision Science, School of Allied Health Sciences, College of Health and Allied Sciences, University of Cape Coast, [3]Bishop Ackon Memorial Christian Eye Centre, Cape Coast, [1]Inter-Star Eye Clinic and Laser Center, Accra, Ghana, [2]Discipline of Optometry, College of Health Sciences, University of KwaZulu-Natal, Durban, South Africa

Address for correspondence: Dr. Michael Agyemang Kwarteng, Discipline of Optometry, College of Health Sciences, University of KwaZulu-Natal, Durban, South Africa.
E-mail: kwartengmichaelagyemang@gmail.com

Submitted: 07-Apr-2020 **Revised:** 13-Apr-2020
Accepted: 16-Apr-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_38_20	

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Kyei S, Assiamah F, Kwarteng MA, Ansah VK. Ocular biometry in an adult Ghanaian population. Kerala J Ophthalmol 2020;32:252-7.

geographic-specific studies on ocular biometry and how they correlate with common ocular disorders.

To fill this gap, there is a need for current studies on ocular biometry among the Ghanaian population. Hence, this prospective study aimed to determine ocular biometry and its correlates in a sample population of native adult Ghanaians attending a referral eye clinic in Ghana.

MATERIALS AND METHODS

Study setting

This study was carried out at the premises of the Christian Eye Center, Cape Coast. The center is the most utilized eye care institution in the Central region of Ghana serving other regions due to the use of advanced technology for cataract and glaucoma surgery among others.

Study design

This was a hospital-based prospective study of patients visiting the center. The study involved measuring and collating biometric data from patients. It sought to explore the relationship between biometric parameters and refractive error.

Sampling technique

The sampling method was nonprobability convenience sampling. The sampling method was based on the fact that the study involved all clients at the center during the study period.

Inclusion and exclusion criteria

The study included all clients who had no ocular disease other than refractive error and those aged 18 and older. Patients below the age of 18 years and patients with steep corneas (keratometric [K] readings of more than 48 D [diopter]) or abnormally thin or thick corneas (CCT <350 μm or CCT > 650 μm), previous contact lens use, along with known glaucomatous individuals were excluded from the study.

Ethical review

The study adhered to the tenets of the Declaration of Helsinki and approval was sought from the "Institutional Review Board of the University of Cape Coast (UCCIRB/CHAS/2018/65). The informed consent of the participants was obtained. There were no risks and/or discomfort associated with participating in the study, and no financial remunerations were offered to the participants. Participation in this study was voluntary, and participants were informed that they could withdraw their participation at any point and that in the event of refusal/withdrawal of participation, they will not incur penalty or loss of treatment or other benefits to which they would normally be entitled.

Data collection procedure

Data collection involved the use of a data extraction sheet to collect data on demographics, and ocular biometry data.

The data extracted included:

1. The examination of the anterior segment was performed on each participant using a slit-lamp biomicroscope
2. The examination of the posterior segment was conducted with an ophthalmoscope and slit-lamp biomicroscope
3. Ocular biometry was measured among participants: Wavelight oculyzer II (Alcon surgical, Fort Worth, Texas, USA) for central cornea readings and keratometry, ultrasound device: US4000 EchoScan (Nidek Co., Ltd., Japan) for biometry, and KR 9000 Auto REF (Perlong Medical Equipment Co., Ltd., Jiangsu, China) for autorefraction
4. Refractive error was calculated in diopters as the spherical equivalent of spherical refractive error plus half of the cylindrical refractive error
5. The two major corneal radii separated by 90° were averaged to give corneal curvature and power.

Data analysis

Data were analyzed using the IBM SPSS version 21 (SPSS Inc, Chicago, USA). Categorical data were presented as frequencies. Pearson's correlation coefficient was used to determine the association between ocular biometric variables. $P < 0.05$ was considered statistically significant.

RESULTS

Demographics

One hundred and sixty-two eyes of 100 participants were involved in the study. Their ages ranged from 21 to 91 years (mean age = 63.75; standard deviation ± 12.84 years) [Table 1].

Ocular biometry

In this study, ocular biometry was evaluated among participants. The median of AL was 24.1 mm, the median of ACD was 3.5 mm, LT with a median of 4.0 mm, VCD had a median of 16.3 mm, and a median CCT of 505.5 μm (409 μm–593 μm) [Table 2].

Table 1: Age distribution

The age group of participants		Frequency (%)
Youth	21-35	8 (4.9)
Adults	36-59	47 (29)
Elderly	60 and older	107 (66)
Total		162 (100.0)

Corneal astigmatism

Corneal astigmatism was determined among the participants, 83 (51.2%) had astigmatism >1.00 D, while the remaining 79 (48.8%) had astigmatism of 1.00 D and below [Table 3].

Spherical refractive error

The dominant error of refraction in this study was myopia (57.4%) followed by hypermetropia (31.5%) [Table 4].

Correlations between biometric parameters, refractive error, and central corneal thickness

A Pearson product-moment correlation coefficient was computed to assess the relationship between ocular biometry variables and refractive error. There was a strong positive correlation coefficient between AL and VCD ($r = 0.73$, $n = 162$, $P < 0.001$). The correlations between CCT, SRE, and other biometric parameters are shown in Table 5 as well as Figure 1.

DISCUSSION

This study provides the first ocular biometry data among healthy Ghanaian adults. The study participants were mainly the elderly. A thin mean central corneal thickness (CCT) was recorded in this study due to age factor. This is consistent with other studies, which have reported that age is an important factor in corneal thickness.[33,34] They reported that the thinner cornea is associated with old age. Hence, as one age, CCT decreases. This cornea thinning is a result of a reduction in the density of keratocytes, collagen fiber degeneration, and a decrease in the distance between fibers in the cornea.[33,34] A similar study by Mashige and Oduntan[7] reported an average CCT of 493. 05 ± 33.2 μm among the Black South African population. Also, a study by Bagus *et al*.[3] reported a similar finding of 494 μm in the same population with a median age of 67 years. Furthermore, in a study by Ntim-Amponsah *et al*.[32] they reported that a sample of participants aged >50 years had a thin CCT of 516 μm which continues to decrease with age. In contrast, a study in Ghana by Ntim-Amponsah *et al*.,[32] reported a higher mean CCT of 533.3 μm among a healthy population with a mean age of

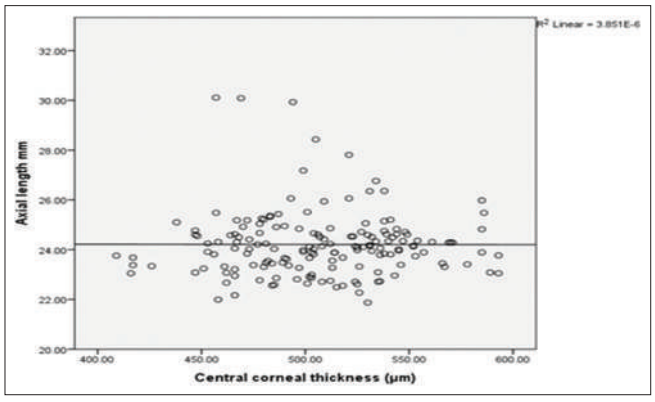


Figure 1: A scatter plot showing the correlation between axial length and central corneal thickness

Table 2: Mean of ocular biometry variables

Variables	Mean±SD	95% CI	Median	Range
AL (mm)	24.2±1.3	24.0-24.4	24.1	21.9-30.1
ACD (mm)	3.5±0.6	3.5-3.6	3.5	1.9-6.9
LT (mm)	3.9±0.8	3.8-4.1	4.0	2.0-6.0
VCD (mm)	16.4±1.39	16.1-16.7	16.3	14.0-21.0
CCT (μm)	506.7±39.2	500.6-512.8	505.5	409.0-593.0
K1 reading (D)	42.50±2.00	42.20-42.80	42.50	38.50-46.50
K2 reading (D)	43.50±2.44	43.10-43.90	43.50	39.75-49.50
SRE (D)	−0.71±2.30	−1.1-−0.4	−0.75	−10.00-+8.00
MMyopia (D)	−2.16±1.68	−2.52-−1.81	−1.50	−10.00-−0.50
MHyper (D)	+1.66±1.49	+1.23-+2.08	+ 1.25	+0.50-+8.00

CCT – Central corneal thickness, AL – Axial length, ACD – Anterior chamber depth, LT – Lens thickness, VCD – Vitreous chamber depth, K1 – Flat keratometry, K2 – Steep keratometry, SRE – Spherical refractive error, MMyopia – Magnitude of myopia, MHyper – Magnitude of hypermetropia, SD – Standard deviation, CI – Confidence interval

Table 3: Classification of corneal astigmatism

Power	Frequency (%)
0.00-0.50 D	35 (21.6)
0.51-1.00 D	44 (27.2)
1.01-1.50 D	43 (26.5)
1.51-2.00 D	17 (10.5)
2.01-2.50 D	13 (8.0)
≥2.51	10 (6.2)
Total	162 (100.0)

Table 4: Type of Spherical refractive error

Refractive error	Range	Frequency (%)
Myopia	At least −0.50 D	93 (57.4)
Hypermetropia	At least +0.50 D	51 (31.5)
Emmetropia (x)	−0.50 D < X < +0.50 D	18 (11.1)
Total		162 (100.0)

Table 5: Correlation between ocular biometric variables

Variables	*R*	R^2	*P*
CCT versus AL	−0.02		0.98
CCT versus ACD	−0.12	0.015	0.12
CCT versus LT	0.06	0.003	0.59
CCT versus VCD	−0.05	0.002	0.68
CCT versus SRE	0.06	0.004	0.44
SRE versus ACD	−0.50	0.003	0.53
SRE versus AL	−0.47	0.22	<0.001
AL versus ACD	0.40	0.16	<0.001
AL versus LT	−0.22	0.04	<0.05
AL versus VCD	0.73	0.53	<0.001
LT versus ACD	−0.22	0.05	<0.05

CCT – Central corneal thickness, AL – Axial length, ACD – Anterior chamber depth, LT – Lens thickness, VCD – Vitreous chamber depth, SRE – Spherical refractive error

34.09 ± 12.14. The difference in the means of CCT in these studies is the aging factor associated with corneal thinning.

In this study, the mean AL was greater than that reported among African descends. An average AL of 21.02 mm[35] and 23.50 mm[36] in Nigeria, 23.05 mm in South Africa,[9] 23.09 mm in Sudan,[26] and 23.7 mm in Egypt[37] has been reported. The reason for a longer AL in this study might be due to scleral thinning and eyeball elongation in myopic eyes which happen to be the dominant error of refraction in this study. Our study reported a higher mean age along with a longer AL which is consistent with studies by Yin *et al*.[38] and Nangia *et al*.[39] However, it contrasts studies that have reported that AL decreases with age.[9,35,40]

The mean ACD in this study was higher than that reported by Mashige and Oduntan,[9] among South Africans but consistent with that of Nagra *et al.,*[41] who reported ACD of 3.55 mm among Britons. It has been established that accurate ACD measurements potentially prevent refractive errors among patients after intraocular lens implantation more than corneal power or AL measures among patients undergoing cataract surgery.[42] A deep ACD has been observed among myopes, which correspond with a high number of myopic eyes in this study [Table 4].

The mean lens thickness (LT) in this study was consistent with that of Mallen *et al*.[43] who reported an average LT of 3.85 mm. The LT is this study is higher than that reported by Mashige and Oduntan:[9] 3.69 mm, Osuobeni:[44] 3.72 mm but lower than reports by Hashemi *et al*.:[45] 4.28 mm, Shufelt *et al*.:[46] 4.28 mm, He *et al*.:[47] 4.44 mm and Olsen *et al*.:[42] 4.68 mm and 4.65 mm in the right and left respectively. A thicker lens observed in this study is due to the higher mean age, since LT increases as one age. Similar studies have reported the effect of age on LT.[42,45-47] The anterior lens capsule thickens gradually from the neonatal stage to the seventh decade of life before stabilizing due to the continual addition of lens fibers.[48]

VCD is deeper among myopes than hyperopes and emmetropes as suggested by other researchers[40] which is consistent with VCD findings in this study due to the high number of myopes. A study by Hashemi *et al*.[44] reported a mean VCD of 15.72 mm as well as a recent study by Takkar *et al*.,[49] also reported an average VCD of 15.38 mm. The VCD reported in these studies were lower than what is reported in this current study. However, a study by Saka *et al*.[50] reported an average VCD of 21–22.3 mm among participants with high myopia. It can be concluded that a mean refractive error of approximately-0.75 D might have resulted in an appreciable increase in VCD among the eyes.

Corneal astigmatism had a significant visual effect on the sample population with more than half of the participants having astigmatism of more than 1.00 D [Table 3]. This is similar to a study in Africa by Bagus *et al*.[3] which reported 45% of Black South Africans with astigmatism of 1.00 D or greater. Information on corneal astigmatism is helpful to surgeons in ensuring a reduction in postoperative astigmatism among patients due to cataract surgery. Hence, the use of toric intraocular lenses as recommended, can be used among the Ghanaian population.

There was a poor correlation between CCT and AL in this study [Figure 1 and Table 5]. Studies have reported that CCT is not correlated with AL, making them two independent variables in ocular biometry.[2,33,34,51,52] AL was positively correlated with ACD and negatively correlated with LT. These findings correlate with studies by Chen *et al*.,[2] Mashige and Oduntan[7,9] Olsen *et al*.[42] and Osuobeni.[44] ACD has great effects on the pathogenesis of ocular conditions such as refractive errors and glaucoma.[9] There was a negative association between ACD and LT in this study which correlates with previous studies.[9,44,53,54] The gradual thickening of the anterior capsule of the lens will decrease the ACD since it happens to be in the anterior chamber.

CONCLUSION

A biometry profile was established in native Ghanaians for the first time. CCT was found to be independent of AL because there was no significant relationship between the two variables. There was no association between CCT and ACD, LT, VCD, and SRE. Intercorrelation among ocular biometry showed that AL positively correlated with ACD, VCD and negatively correlated with LT.

Acknowledgment

The authors are grateful to the Management of Bishop Ackon Memorial Christian Eye Centre, Cape Coast, Ghana.

Financial support and sponsorship

This study was solely funded by the authors.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Damji KF, Muni RH, Munger RM. Influence of corneal variables on accuracy of intraocular pressure measurement. J Glaucoma 2003;12:69-80.
2. Chen MJ, Liu YT, Tsai CC, Chen YC, Chou CK, Lee SM. Relationship between central corneal thickness, refractive error, corneal curvature, anterior chamber depth and axial length. J Chin Med Assoc 2009;72:133-7.

3. Bagus T, Alberto K, Muteba M, Makgotloe A. Analysis of corneal biometry in a black South African population. Afr Vision Eye Health 2019;78:a495.
4. Gharaee H, Abrishami M, Shafiee M, Ehsaei A. White-to-white corneal diameter: Normal values in healthy Iranian population obtained with the Orbscan II. Int J Ophthalmol 2014;7:309-12.
5. Zha Y, Feng W, Han X, Cai J. Evaluation of myopic corneal diameter with the Orbscan II Topography System. Graefes Arch Clin Exp Ophthalmol 2013;251:537-41.
6. Hashemi H, Khabazkhoob M, Emamian MH, Shariati M, Yekta A, Fotouhi A. White-to-white corneal diameter distribution in an adult population. J Curr Ophthalmol 2015;27:21-4.
7. Mashige KP, Oduntan OA. Corneal parameters and their correlations with refractive error, axial length, anterior chamber depth and lens thickness in black South Africans. Guoji Yanke Zazhi (Int Eye Sci) 2017;17:597-603.
8. Lim KJ, Hyung SM, Youn DH. Ocular dimensions with aging in normal eyes. Korean J Ophthalmol 1992;6:19-31.
9. Mashige KP, Oduntan OA. Axial length, anterior chamber depth and lens thickness: Their intercorrelations in black South Africans. Afr Vision Eye Health 2017;76:a362.
10. Stenstrom S. Investigation of the variation and the correlation of the optical elements of human eyes. Am J Optom Arch Am Acad Optom 1948;58:1-71.
11. Sorsby A, Benjamin B, Sheridan M, Stone J, Leary GA. Refraction and its components during the growth of the eye from the age of three. Memo Med Res Counc 1961;301:1-67.
12. Larsen JS. The sagittal growth of the eye. IV. Ultrasonic measurement of the axial length of the eye from birth to puberty. Acta Ophthalmol (Copenh) 1971;49:873-86.
13. Larsen JS. The sagittal growth of the eye. III. Ultrasonic measurement of the posterior segment (axial length of the vitreous) from birth to puberty. Acta Ophthalmol 1971;49:441-53.
14. Grosvenor T, Scott R. Three-year changes in refraction and its components in youth-onset and early adult-onset myopia. Optom Vis Sci 1993;70:677-83.
15. Lin LL, Shih YF, Lee YC, Hung PT, Hou PK. Changes in ocular refraction and its components among medical students—a 5-year longitudinal study. Optom Vis Sci 1996;73:495-8.
16. Adams AJ. Axial length elongation, not corneal curvature, as a basis of adult onset myopia. Am J Optom Physiol Opt 1987;64:150-2.
17. McBrien NA, Millodot M. A biometric investigation of late onset myopic eyes. Acta Ophthalmol (Copenh) 1987;65:461-8.
18. Jiang BC, Woessner WM. Vitreous chamber elongation is responsible for myopia development in a young adult. Optom Vis Sci 1996;73:231-4.
19. Aghaian E, Choe JE, Lin S, Stamper RL. Central corneal thickness of Caucasians, Chinese, Hispanics, Filipinos, African Americans, and Japanese in a glaucoma clinic. Ophthalmology 2004;111:2211-9.
20. Prager TC, Hardten DR. Immersion biometry and optical coherence biometry offer clinical advantages. Ophthalmol Manage 2012;16:28-31.
21. Goldschmidt E. Refraction in the newborn. Acta Ophthalmol (Copenh) 1969;47:570-8.
22. La Rosa FA, Gross RL, Orengo-Nania S. Central corneal thickness of Caucasians and African Americans in glaucomatous and nonglaucomatous populations. Arch Ophthalmol 2001;119:23-7.
23. Brandt JD, Beiser JA, Gordon MO, Kass MA. Ocular Hypertension Treatment Study (OHTS) Group. Central corneal thickness in the ocular hypertensive study (OHTS). Ophthalmology 2001;108:1779-88.
24. Shimmyo M, Ross AJ, Moy A, Mostafavi R. Intraocular pressure, Goldmann applanation tension, corneal thickness, and corneal curvature in Caucasians, Asians, Hispanics, and African Americans. Am J Ophthalmol 2003;136:603-13.
25. McBrien NA, Adams DW. A longitudinal investigation of adult-onset and adult-progression of myopia in an occupational group. Refractive and biometric findings. Invest Ophthalmol Vis Sci 1997;38:321-33.
26. Mohamed NY, Hassan MN, Ali NAM, Binnawi KH. Central corneal thickness in Sudanese population. Sud J Ophthalmol 2009;1:29-32.
27. Iyamu E, Memeh M. The association of central corneal thickness with intraocular pressure and refractive error in a Nigerian population. Online J Health Allied Sci 2007;6:1-7.
28. Iyamu E, Ituah I. The relation between central corneal thickness and intraocular pressure: A comparative study of normal and glaucoma subjects. Afr J Med Med Sci 2008;37:345-53.
29. Babalola OE, Kehinde AV, Iloegbunam AC, Akinbinu T, Moghalu C, Onuoha I. A comparison of the Goldmann applanation and non-contact (Keeler Pulsair EasyEye) tonometers and the effect of central corneal thickness in indigenous African eyes. Ophthalmic Physiol Opt 2009;29:182-8.
30. Eballe AO, Koki G, Ellong A, Owono D, Epée E, Bella LA, *et al*. Central corneal thickness and intraocular pressure in the Cameroonian nonglaucomatous population. Clin Ophthalmol 2010;4:717-24.
31. Ntim-Amponsah CT, Seidu A, Essuman V, Fordjour G, Tagoe N, Coker A, *et al*. A study of central corneal thickness in glaucoma and non-glaucoma patients in West African population. Cornea 2012;31:1093-6.
32. Ntim-Amponsah CT, Essuman VA, Edirisuriya-Khair RD. A study of central corneal thickness in normal Ghanaians. Niger J Ophthalmol 2007;15:1-4.
33. Nemesure B, Wu SY, Hennis A, Leske MC, Barbados Eye Study Group. Corneal thickness and intraocular pressure in the Barbados eye studies. Arch Ophthalmol 2003;121:240-4.
34. Muhammed AH, Joma AK, Abdulgani AI. The relation between central corneal thickness and axial length in a sample of Erbil population. Zanco J Med Sci 2015;19:1096-103.
35. Ogbeide OU, Omoti AE. Ultrasonographic ocular diameters in Nigerians. West Afr J Med 2009;28:97-101.
36. Iyamu E, Iyamu JE, Amadasun G. Central corneal thickness and axial length in an adult Nigerian population. J Optom 2013;6:154-60.
37. Abdelaziz A, Mousa A. Ocular axial length measurement using regular ultrasound and IOL master for different refractive errors in Egyptian population. Med J Cairo Univ 2014;82:159-65.
38. Yin G, Wang YX, Zheng ZY, Yang H, Xu L, Jonas JB, *et al*. Ocular axial length and its associations in Chinese: The Beijing Eye Study. PLoS One 2012;7:e43172.
39. Nangia V, Jonas JB, Sinha A, Matin A, Kulkarni M, Panda-Jonas S. Ocular axial length and its associations in an adult population of central rural India: The Central India Eye and Medical Study. Ophthalmology 2010;117:1360-6.
40. Bhardwaj V, Rajeshbhai GP. Axial length, anterior chamber depth-a study in different age groups and refractive errors. J Clin Diagn Res 2013;7:2211-2.
41. Nagra M, Gilmartin B, Logan NS. Estimation of ocular volume from axial length. Br J Ophthalmol 2014;98:1697-701.
42. Olsen T, Arnarsson A, Sasaki H, Sasaki K, Jonasson F. On the ocular refractive components: The Reykjavik Eye Study. Acta Ophthalmol Scand 2007;85:361-6.
43. Mallen EA, Gammoh Y, Al-Bdour M, Sayegh FN. Refractive error and ocular biometry in Jordanian adults. Ophthalmic Physiol Opt 2005;25:302-9.
44. Osuobeni EP. Ocular components values and their inter-correlations in Saudi Arabians. Ophthalmic Physiol Opt 1999;19:489-97.
45. Hashemi H, Khabazkhoob M, Miraftab M, Emamian MH, Shariati M, Abdolahinia T, *et al*. The distribution of axial length, anterior chamber depth, lens thickness, and vitreous chamber depth in an adult population of Shahroud, Iran. BMC Ophthalmol 2012;12:50.
46. Shufelt C, Fraser-Bell S, Ying-Lai M, Torres M, Varma R. Los Angeles Latino Eye Study Group. Refractive error, ocular biometry, and lens opalescence in an adult population: The Los Angeles Latino Eye Study.

Invest Ophthalmol Vis Sci 2005;46:4450-60.
47. He M, Huang W, Li Y, Zheng Y, Yin Q, Foster PJ. Refractive error and biometry in older Chinese adults: The Liwan eye study. Invest Ophthalmol Vis Sci 2009;50:5130-6.
48. Krag S, Andreassen TT. Mechanical properties of the human posterior lens capsule. Invest Ophthalmol Vis Sci 2003;44:691-6.
49. Takkar B, Gaur N, Saluja G, Rathi A, Sharma B, Venkatesh P, *et al*. Evaluation of the vitreous chamber depth: An assessment of correlation with ocular biometrics. Indian J Ophthalmol 2019;67:1645-9.
50. Saka N, Ohno-Matsui K, Shimada N, Sueyoshi S, Nagaoka N, Hayashi W, *et al*. Long-term changes in axial length in adult eyes with pathologic myopia. Am J Ophthalmol 2010;150:562-80.
51. Oliveira C, Tello C, Liebmann J, Ritch R. Central corneal thickness is not related to anterior scleral thickness or axial length. J Glaucoma 2006;15:190-4.
52. Shimmyo M, Orloff PN. Cornea thickness and axial length. Am J Ophthalmol 2005;139:553-4.
53. Mei L, Zhonghao W, Zhen M, Yimin Z, Xing L. Lens thickness and position of primary angle closure measured by anterior segment optical coherence tomography. J Clin Exp Ophthalmol 2013;4:281.
54. Hashemi H, Jafarzadehpur E, Ghaderi S, Yekta A, Ostadimoghaddam H, Norouzirad R, *et al*. Ocular components during the ages of ocular development. Acta Ophthalmol 2015;93:e74-81.

Original Article

Trial for a trial: Is there hope? – Optic Neuritis Treatment Trial protocol in patients with traumatic optic neuropathy

ABSTRACT

Aims: The aim of this study was to analyze the clinical profile, visual outcome, factors predicting the visual outcome, and response to systemic methylprednisolone as per the Optic Neuritis Treatment Trial (ONTT) protocol in patients with traumatic optic neuropathy (TON). **Study Design:** This was a prospective observational study. **Subjects and Methods:** All cases who had decreased visual acuity (VA) following impact head/orbital injury were evaluated with detailed history, and ophthalmic evaluation was done. Those who were diagnosed to have indirect TON were treated with intravenous methylprednisolone 500 mg twice daily for 3 days, followed by oral prednisolone 1 mg/kg for 11 days. Visual outcome was assessed at 1 week, 1 month, 3 months, and 6 months. The results were statistically analyzed. **Results:** 34.65% had severe loss of vision (<6/60 vision) at presentation. Twenty patients received early treatment (within 3 days) and six patients received treatment after 3 days. Fourteen eyes (53.84%) showed an improvement in VA at 1 month. Among those who had early treatment, 60% improved, but it was not statistically significant. An initial VA of <6/60 and the presence of brain contusions were significantly associated with poor visual outcomes. **Conclusions:** There is no statistically significant improvement in visual outcome with early initiation of therapy. Severe initial visual loss and presence of brain contusions predict poor visual outcomes. Selected patients will benefit with corticosteroids, and the ONTT protocol with minimal adverse reactions would be a safer choice in them.

Keywords: Methylprednisolone, Optic Neuritis Treatment Trial, traumatic optic neuropathy

INTRODUCTION

The optic nerve is prone to indirect and direct trauma, causing functional impairment of vision. Optic nerve injuries can occur with head injury due to road traffic accidents (RTAs) or falls. However, the diagnosis of optic nerve injury may be delayed because of focus on other life-threatening injuries. The signs of optic nerve trauma are clinical, and the examination of acutely injured is often difficult. Indirect damage to the optic nerve is the most common form of traumatic optic neuropathy (TON) and is reported following 0.5%–5% of all closed-head trauma.[1] It is defined as traumatic visual loss that occurs without external or initial ophthalmoscopic evidence of injury to the eye or the optic nerve. Indirect injuries occur due to concussive forces that are transmitted to the optic nerve as a result of orbit or cranial trauma. This impact produces a shock wave which leads to optic nerve avulsion or posterior indirect TON.[1] After the immediate injury, swelling of the optic nerve within the canal causes secondary retinal ganglion cell loss.[2] Direct TON, on the other hand, results from direct trauma to the optic nerve from sharp objects, missiles, and bony fragments.

Jainy Joseph Emmatty, Smita Narayan[1], Rajini K. Cherayath[2]

Department of Ophthalmology, Amala Institute of Medical Sciences, Thrissur, [1]Department of Ophthalmology, Regional Institute of Ophthalmology, Thiruvananthapuram, [2]Department of Ophthalmology, Government Medical College, Manjeri, Kerala, India

Address for correspondence: Dr. Smita Narayan, 3B, Condor Marigold, Near DPI, Thycaud P.O, Trivandrum - 695 014, Kerala, India.
E-mail: drsmita.narendranath@gmail.com

Submitted: 14-Feb-2020 **Revised:** 02-Mar-2020
Accepted: 20-Mar-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_15_20	

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Emmatty JJ, Narayan S, Cherayath RK. Trial for a trial: Is there hope? – Optic neuritis treatment trial protocol in patients with traumatic optic neuropathy. Kerala J Ophthalmol 2020;32:258-62.

Although TON can be diagnosed by a careful history taking and examination, its appropriate management has not been well defined. Different approaches, including different dosages of steroids (60 mg to 7 g/day), surgical decompression of optic canal (via intracranial, transethmoidal, endonasal, sublabial, or other techniques), and observation alone, have been suggested, but conclusive evidence to establish a standard approach to this devastating cause of visual loss is lacking.[3] Although visual outcome after indirect TON may be quite variable, approximately 50% of the patients are left with "light perception" or "no light perception" vision, making it a significant cause of permanent visual loss.[1]

The role of steroids in TON is controversial. In most cases, the treatment is based on individual decision. Despite being an important cause of permanent visual loss, there are few studies in this regard from Kerala. We undertook this study to identify the clinical profile, associated injuries, visual outcome, prognostic factors, the role of steroids, and the impact of early initiation of therapy in patients with TON. Knowledge of these factors would help in formulating treatment protocol and fuel further research in the field.

SUBJECTS AND METHODS

After getting the institutional ethics committee approval, all patients who were diagnosed to have indirect TON were included in the study. However, those with major head trauma, unconscious patients, penetrating eye injury patients, and patients needing decompression surgery were excluded. Those with posttraumatic visual loss not related to optic nerve dysfunction (open-globe injury, traumatic cataract, retinal detachment, choroidal rupture, and vitreous hemorrhage) and those with preexisting visual loss were also excluded.

After getting a detailed informed consent, data were collected regarding age, sex, the cause of blindness (timing and nature of trauma), level of consciousness after trauma, site of injury, onset and duration of symptoms, the time of presentation, and other systemic illness. A baseline neuro-ophthalmic examination for visual acuity (VA), relative afferent pupillary defect (RAPD), field of vision, and color vision was done. Slit-lamp examination was done to rule out penetrating injury, traumatic cataract, hyphema, and uveitis. Ophthalmoscopy was performed to see for optic nerve avulsion, choroidal rupture, and pallor. Extraocular movements were also assessed. X-rays of the skull and the orbit were taken. High-resolution computed tomography (CT) scan of the head and orbit was taken in all cases to assess orbital wall fractures, hemolysins, and to rule out the presence of optic canal fracture or bony fragments impinging the optic nerve. Magnetic resonance imaging was done in those with worsening of vision to look for optic nerve edema in the optic canal, optic nerve sheath hematoma, and intracranial injuries that were inadequately detailed with CT imaging.

Included patients were treated with intravenous methylprednisolone 500 mg twice daily for 3 days, followed by oral prednisolone (1 mg/kg) for 11 days. Visual outcome was assessed at 1 week, 1 month, 3 months, and at 6 months. At each visit, patients' VA, pupillary reaction, color vision, visual fields, and fundus were assessed. Two- or more-line improvement in VA was considered as a significant improvement.

The collected data were analyzed statistically using SPSS version 16.0 (SPSS Inc., 233 South Wacker Drive, Chicago, IL). Quantitative variables are expressed as means and qualitative parameters as proportions. Correlation between the quantitative parameters was analyzed using independent sample Student's *t*-test and between qualitative parameters using the Chi-square test. $P < 0.05$ was considered statistically significant.

RESULTS

Twenty-six eyes of 26 patients were included in the study. The mean age of the patients was 37 years. Patients in the age group of 31–40 years were the most affected (38.45%), followed by the patients in the 21–30 years' age group (34.15%). Out of 26 patients, 25 (96.15%) were male. Mode of injury was RTA in 25 (99.15%) patients, and one patient developed optic neuropathy following an assault. The left eye was involved in 16 patients (61.54%). The side of the face was the site of injury in 15 patients (67.7%) and the forehead in 11 patients (42.3%). There was a history of transient loss of consciousness in nine patients (34.61%).

In our study, nine patients (34.65%) had severe loss of vision (<6/60) at presentation. Grade 1, Grade 2, and Grade 3 RAPDs were seen in 1 patient (3.84%), 14 patients (53.84%), and 11 patients (42.32%), respectively. Extraocular movements were full in 22 patients (84.61%), while mild restriction was noted in 4 patients (15.39%). Fundus was normal in all patients (100%). Associated injuries noted were lacerated wound over ipsilateral forehead/eyebrow in 17 patients (65.38%), periorbital ecchymosis in 24 patients (92.30%), subconjunctival hemorrhage in 22 patients (84.61%), and periorbital emphysema in 7 patients (26.92%). Other injuries seen were degloving injury of the cheek in one patient and avulsion of the pinna in one patient. Radiological findings

included minor brain contusions in six patients and multiple bone fractures including the frontal bone and orbital wall fractures (mainly lateral wall and roof). Majority of the patients presented with more than one bony fracture of orbit, and three patients (11.53%) had no fractures.

Twenty patients received early treatment (within 3 days), and six patients received treatment after 3 days [Figure 1]. Treatment was initiated late in six patients (23.08%) due to their delayed presentation. Of the 26 eyes included in the study, 14 eyes (53.84%) showed an improvement in VA at a 1-month follow-up, and thereafter, none improved. Of these, eight patients had shown an improvement in visual outcome within 1 week of initiation of therapy. Two patients deteriorated at the end of 3 months. Primary optic atrophy was seen in ten patients (38.46%) and temporal pallor in nine patients (34.61%) at the end of 1 month. All the patients showed optic disc pallor at the end of 3 months.

Among those who had early treatment, 12 out of 20 eyes (60%) improved while only two of the six eyes (33%) in the late treatment group had a significant improvement in the VA. On the Chi-square test, however, the result did not attain statistical significance ($P = 0.365$). The mean age of the patients who had good visual outcome following therapy was 35.2 years compared to 37.4 years in the group that had a poor visual outcome. However, this result was not statistically significant ($P = 0.562$). Of the nine patients who had an initial VA (IVA) <6/60, seven patients (78.78%) had no significant improvement in the final visual outcome. Thus, an IVA of <6/60 was significantly associated with a poor visual outcome ($P = 0.038$). Among the six patients with brain contusions, only 16.67% had a significant improvement in VA, while 65% of the patients without contusions improved [Figure 2]. The presence of associated brain contusions (83.33% vs. 35%) was associated with poor visual outcomes. The result was statistically significant ($P = 0.049$).

DISCUSSION

Twenty eyes of 26 participants were enrolled in our study. Majority of the participants were in the 20–40 years group. This is similar to studies conducted by Sadeghi *et al*.[3] (mean age of 24.1 years) and Lee *et al*.[4] (mean age of 33 years). In our study, 96.15% of the patients were male. The male-to-female ratio 3:1 in the study by Sadeghi *et al*.[3] Motor vehicle accidents accounted for 83.3% of injuries in our study. Similar findings were showed in Sadeghi *et al*. study (71.4%)[3] and Lee *et al*. study[4] (83.3%).

The most common site of indirect optic nerve injury is in the optic canal.[5] Chou *et al*.[6] had proposed that the damage of optic nerve at microscopic level, including contusion necrosis, nerve fiber tears and nerve infarction secondary to closed-space edema, hemorrhage, thrombosis, vasospasm, impingement by bone spicules, and shearing of dural vessels in the optic canal could result in TON. The ocular manifestations most commonly associated with optic nerve injury were periorbital hematoma and subconjunctival hemorrhage, with no changes in the optic nerve. Periorbital hematoma was present in all cases in the study by Lee *et al*.[4] In our study, periorbital ecchymosis was seen in 24 patients (92.30%) and subconjunctival hemorrhage in 22 patients (84.61%).

Our study showed that majority of the patients (82.46%) presented with more than one bony fracture of the orbit and 3 patients (11.53%) had no fractures. Lee *et al*.[4] observed that 79.2% of the patients presented with more than one bony fracture of the skull and/or orbit and 5 patients (20.8%) had no fractures. The presence of these signs may suggest the presence of TON in cases without evidence of optic nerve impingement or compression.

Methylprednisolone therapy is recommended as the initial treatment of choice because of its neuroprotective mechanism. Of the 26 eyes included in the study, all patients had received treatment with steroids. In our study, 14 eyes (53.84%) showed an improvement in VA at 1-month

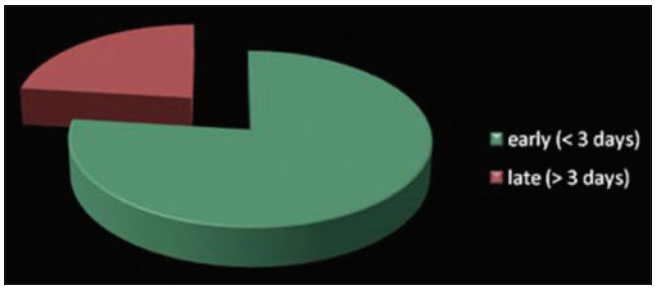


Figure 1: Initiation of treatment in patients (early vs. late)

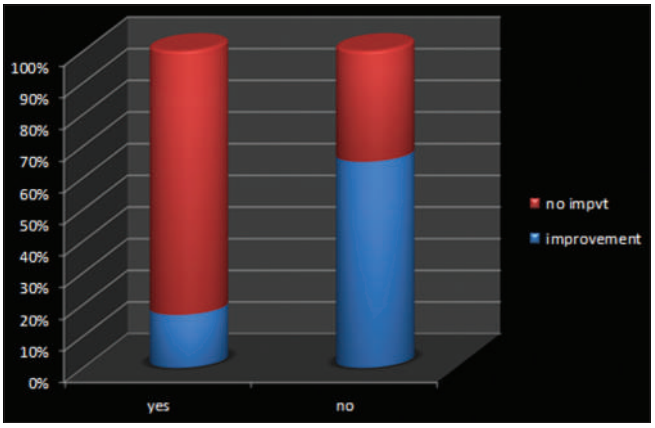


Figure 2: Visual outcome in patients with brain contusions

follow-up which was maintained during the follow-up period of 6 months. Response to steroids varies in some of the previous studies. The largest series on steroid treatment was published by the International Optic Nerve Trauma Study,[7] which reported an improvement in 54% of the patients after 3 months of follow-up. Thirty-seven percent of the patients treated with megadose steroids in Sadeghi *et al*. study[3] showed improvement after 3 months' follow-up. In the study conducted by Joseph *et al*.,[8] 9 of 16 patients showed a significant improvement on steroids. According to Lee *et al*.,[4] 91.7% of the patients treated with intravenous methylprednisolone 250 mg for 3 days, followed by oral prednisolone 1 mg/kg for 11 days, had shown at least one-line improvement of VA, and it was statistically significant ($P > 0.05$). Among those eyes which were treated conservatively, 77.8% had shown at least one-line improvement of VA.

Very high-dose corticosteroids limit free-radical amplification response to the trauma. The Corticosteroid Randomization after Significant Head Injury trial,[9] a large randomized, placebo-controlled study, evaluated the effect of early administration of 48 h infusion of high-dose methylprednisolone on the risk of death and disability after head injury and found a small but statistically significant increase in the risk of death within 2 weeks after head injury in the group of corticosteroids compared with placebo (21 vs. 18) ($P < 0.0001$). Regarding the systemic side effects of steroids in our study, two patients developed hyperglycemia and four had mild gastrointestinal symptoms which were managed medically. Thus, the Optic Neuritis Treatment Trial (ONTT) protocol could be considered safer than high-dose steroids.

Cook *et al*.,[10] in a meta-analysis, reported that recovery of vision in patients treated with steroids or surgical decompression of the optic canal was significantly better than recovery in patients receiving no treatment. The same conclusion, however, has not been supported by the IONTS study which showed neither corticosteroids nor optic canal surgery is effective as the standard of care for patients with TON. Hence it was suggested that every ophthalmologist should decide to treat or not to treat on a case to case basis.[7]

Rajiniganth *et al*.[11] reported a visual improvement in 16 (70%) of 23 patients in whom treatment was started before 7 days after injury. The difference between the two groups was significant ($P < 0.01$). Panja *et al*.[12] also showed that late initiation of therapy >7 days was associated with poor visual outcome. Yang *et al*.[13] also observed a better improvement in those treated within 7 days ($P = 0.056$). On the other hand, Levin *et al*.[7] stated that there was no indication that the dosage or timing of corticosteroid treatment or the timing of surgery was associated with an increased probability of visual improvement. In our study, among those who had early treatment, 12 out of 20 eyes (60%) improved while only 2 of the 6 eyes (33%) in the late treatment group had a significant improvement in the VA. Although this suggested a trend toward a better outcome with early initiation of therapy, the result did not attain statistical significance ($P = 0.365$) [Table 1]. This could probably be due to the limited number of patients included in the study. In our study, eight patients had shown an improvement in visual outcome within 1 week of initiation of therapy and six eyes more showed an improvement in VA at 1-month follow-up, and thereafter, none of the eyes improved. Two patients deteriorated at the end of 3 months. In the study conducted by Sadeghi *et al*.,[3] VA improved by one line in eight eyes (28.6%) immediately after treatment and in ten eyes (37%) after 3 months.

The significant prognostic factors observed in this study were degree of initial visual loss and the presence of brain contusions. In our study, patients who had an IVA >6/60 had a better visual outcome (70.58%) compared to those with an IVA < 6/60 (22.22%), and the result was statistically significant ($P = 0.038$) [Table 2]. Yang *et al*.[13] revealed that initial visual acuity (IVA) was a statistically significant factor affecting the outcome of TON ($P = 0.006$). According to Chen *et al*.,[14] patients with initial vision better than light perception benefitted more from treatment than the patients with no light perception. Wang *et al*.[15] also showed that initial VA better than light perception was associated with better outcomes (100% vs. 27%, $P < 0.05$).

The second statistically significant factor affecting the outcome identified in our study is the presence of brain contusions, and it was associated with poor visual outcomes (83.33% vs. 35%).

A study done by Carta *et al*.[16] showed that age >40 years is a significant risk factor (RR = 1.79) for poor outcome

Table 1: Visual outcome of patients based on the initiation of treatment

Vision	Improved (%)	No improvement (%)	χ^2, *P*
Early treatment	12 (60)	8 (40)	1.321, 0.365
Late treatment	2 (33)	4 (66)	

Table 2: Initial visual acuity and visual outcome

	Improved (%)	No improvement (%)	χ^2, *P*
VA <6/60	2 (22.22)	7 (78.78)	5.539, 0.038
VA >6/60	12 (70.58)	5 (29.42)	

VA – Visual acuity

Table 3: Final visual outcome and history of loss of consciousness

	Improved (%)	Static (%)	χ^2, *P*
History of LOC	3 (33.4)	6 (66.6)	0.484, 0.41
No history of LOC	10 (58)	7 (42)	

LOC – Loss of consciousness

in patients with TON. They speculate that recovery of vision in such patients may be impaired by age-related axonal lipoperoxidation and membrane hydrolysis occurring after the trauma. However, studies conducted by Wang *et al*.[15] and Rajiniganth *et al*.[11] did not show any significant difference in prognosis based on age. In our study, the mean age of the patients who had good visual outcome was 35.2 years compared to 37.4 years in the group that had a poor visual outcome which was not statistically significant ($P = 0.562$). Carta *et al*.[16] reported that loss of consciousness associated with TON and the absence of recovery of VA after 48 h of steroid therapy were poor prognostic factors. Our study also showed a similar trend that a history of unconsciousness is a predictor of poor visual outcomes (66.6% vs. 33.4%). However, the result was not statistically significant ($P = 0.41$) [Table 3].

CONCLUSIONS

The pathogenesis of TON is inconclusive, with various possible mechanisms causing the visual loss. The management of TON is decided mainly on the basis of personal preference. Based on this study, we suggest that selected patients will benefit from corticosteroids and the ONTT protocol is safe with minimal adverse reactions.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Miller NR, Walsh FB, Hoyt WF, editors. Walsh and Hoyt's Clinical Neuro-Ophthalmology. Philaldelphia: Lippincott Williams & Wilkins; 2005;2:2380.
2. Yu-Wai-Man, P, Griffiths PG. Steroids for traumatic optic neuropathy. Cochrane Database of Systematic Reviews 2013.
3. Sadeghi TA, Lasheyei A, Tabassi A. Visual outcome of traumatic optic neuropathy in patients treated with intravenous megadose of steroids." ACTA MEDICA IRANICA 2005;43:110-4.
4. Lee KF, Muhd Nor NI, Yaakub A, Wan Hitam WH. Traumatic optic neuropathy: A review of 24 patients. Int J Ophthalmol 2010;3:175-8.
5. Yu-Wai-Man P. Traumatic optic neuropathy-clinical features and management issues. Taiwan J Ophthalmol 2015;5:3-8.
6. Chou PI, Sadun AA, Chen YC, Su WY, Lin SZ, Lee CC. Clinical experiences in the management of traumatic optic neuropathy. Neuro Ophthalmol 1996;16:325-36.
7. Levin LA, Beck RW, Joseph MP, Seiff S, Kraker R. The treatment of traumatic optic neuropathy: The International Optic Nerve Trauma Study. Ophthalmology 1999;106:1268-77.
8. Joseph MP, Lessell S, Rizzo J, Momose KJ. Extracranial optic nerve decompression for traumatic optic neuropathy. Arch Ophthalmol 1990;108:1091-3.
9. Bracken MB. CRASH (Corticosteroid Randomization after Significant Head Injury Trial): Landmark and storm warning. Neurosurgery 2005;57:1300-2.
10. Cook MW, Levin LA, Joseph MP, Pinczower EF. Traumatic optic neuropathy. A meta-analysis. Arch Otolaryngol Head Neck Surg 1996;122:389-92.
11. Rajiniganth MG, Gupta AK, Gupta A, Bapuraj JR. Traumatic optic neuropathy: Visual outcome following combined therapy protocol. Arch Otolaryngol Head Neck Surg 2003;129:1203-6.
12. Panja S, Gupta A, Gupta A, Prabhakar S. Determinants of outcome in traumatic optic neuropathy. Otolaryngol Head Neck Surg 2010;143 2 Suppl: P128.
13. Yang WG, Chen CT, Tsay PK, de Villa GH, Tsai YJ, Chen YR. Outcome for traumatic optic neuropathy-surgical versus nonsurgical treatment. Ann Plast Surg 2004;52:36-42.
14. Chen HY, Tsai RK, Wang HZ. Intravenous methylprednisolone in treatment of traumatic optic neuropathy. Kaohsiung J Med Sci 1998;14:577-83.
15. Wang BH, Robertson BC, Girotto JA, Liem A, Miller NR, Iliff N, *et al*. Traumatic optic neuropathy: A review of 61 patients. Plast Reconstr Surg 2001;107:1655-64.
16. Carta A, Ferrigno L, Salvo M, Bianchi-Marzoli S, Boschi A, Carta F. Visual prognosis after indirect traumatic optic neuropathy. J Neurol Neurosurg Psychiatry 2003;74:246-8.

Original Article

Functional blindness: Blepharospasm with apraxia of eyelid opening

ABSTRACT

Purpose: The purpose was to evaluate the effectiveness of protractor myectomy surgery in patients with severe blepharospasm whose daily life is severely impaired. **Materials and Methods:** Records of fifty patients with benign, essential blepharospasm were examined retrospectively and seven were included in the study. All patients underwent a complete ophthalmological and neurological examination. The severity and frequency of spasm was determined by Jankovic Rating Scale, ranging from 0 to 4, and six daily activities were determined by Blepharospasm Disability Index. The primary indication for protractor myectomy was poor response to serial botulinum toxin injections. The main outcome measure was based on whether the spasms were fully healed or the need for additional botox application. **Results:** Fourteen eyes of seven patients with significant functional disability were included in the study. Excessive blinking and visual disability were the most common symptoms seen in all patients, and all had previously used one or more medications. Myectomy surgery was performed in all patients who were resistant to serial botulinum toxin injections. Of the 14 eyelids, 85.7% showed resolution of spasms and no longer required botulinum toxin treatment following surgery. One patient with keratitis sequelae and dry eye was given botulinum 1 month after the surgery. One patient with residual blepharospasm in the lower eyelid underwent lower eyelid myectomy and blepharoplasty. Transient periorbital edema, hematoma, and echymosis developed following surgery. The patients were followed up for 2 years, and excellent cosmetic result with no contracture was observed in all patients. **Conclusion:** Protractor myectomy is an effective treatment modality in cases of blepharospasm with severe functional disability.

Keywords: Apraxia, blepharospasm, botulinum toxin, myectomy

INTRODUCTION

Benign essential blepharospasm (BEB) is a type of focal dystonia characterized by excessive, involuntary, repetitive contractions of the protractor muscles (orbicularis oculi, procerus, and corrugator muscles).[1,2] The spasms have a significant impact on the daily activities of patients by causing visual disturbance or functional blindness, anxiety, and depression.[3,4]

The exact cause of BEB is unknown. It is thought to be multifactorial, such as interaction of certain genetic and environmental factors.[4,5] As it is known, the blink reflex is controlled by the basal ganglion (extrapyramidal dopaminergic circuit), and any abnormalities in the basal ganglion and dopaminergic system create BEB. It is also known that progressive loss of dopamine in the substantia nigra with aging increases the trigeminal blink reflex. This relationship between them explains why BEB is common in older age.[4]

The involuntary eyelid closure gradually worsens and results in apraxia of eyelid opening (AEO).[6] It is a nonparalytic motor abnormality and characterized by difficulty in opening the eyelids in the absence of visible contractions of the orbicularis oculi muscle.[6,7] It is more common in women, and the peak age of onset is in the sixth and seventh decades of life.[8]

Submitted: 10-Apr-2020 **Revised:** 25-Apr-2020
Accepted: 15-Jun-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_42_20	

Titap Yazicioglu
Eye Clinic, Kartal Research and Training Hospital, Istanbul, Turkey

Address for correspondence: Dr. Titap Yazicioglu, Konaklar mh. Sebboy Sok. Petekler Sit. G/B D: 18, Yenilevent, Istanbul, Turkey.
E-mail: t_yazicioglu@hotmail.com

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Yazicioglu T. Functional blindness: Blepharospasm with apraxia of eyelid opening. Kerala J Ophthalmol 2020;32:263-7.

The most effective treatment option for BEB is botulinum neurotoxin Type A. It is well tolerated, but sometimes, the disease can be severe and resistant to botulinum toxin treatment.[9,10] Such patients are candidates for surgical treatment.[5,9,11] Many surgical techniques, such as peripheral facial neurectomy by alcohol injections, surgical sectioning or percutaneous thermolysis, and selective peripheral facial nerve avulsion, have been used to interrupt the pathway from the motor cortex to the orbicularis oculi muscles.[1,12] Among them, upper eyelid myectomy procedure, that involves removal of the orbital, preseptal, and pretarsal orbicularis muscles, as well as correction of the associated eyelid deformities, has been shown to be a very effective treatment in refractory cases.[10,13]

Restricted daily activities of patients with eyelid apraxia may cause anxiety and depression. In this study, we intended to show the positive effects of surgery on their severely restricted life.

MATERIALS AND METHODS

In this retrospective study, fifty consecutive patients with blepharospasm treated with botulinum toxin were evaluated, and only seven of them who were refractory to botulinum toxin were included in the study. All procedures adhered to the tenets of the Declaration of Helsinki, and local approval form was received from the ethical committee. Written informed consent was obtained from every patient who participates in the study.

All patients had a full neurological examination to rule out other underlying neurological conditions, and a complete ophthalmological examination was conducted.

Patients with previous eyelid surgery or eyelid retraction were excluded from the study. Demographic features of the disease, duration and severity of spasm, presence of depression and anxiety, ocular irritation, burning sensation, watering, photophobia, sensory tricks, treatment received prior to surgery, number and type of surgery, duration of relief from spasms after surgery, complications or secondary effects of surgery, and further treatment data to obtain relief from blepharospasm after surgery were all observed and are summarized in Table 1.

While evaluating the objective signs of spasms, we used Jankovic Rating Scale – a 5-point scale ranging from 0 to 4, where 0 indicates no symptoms and 4 indicates the most severe and frequent symptoms. We also used the Blepharospasm Disability Index in evaluating each patient's ability to drive, read, watch television, perform household tasks, and walk and were categorized as mild, moderate, or marked.

The primary indication for protractor myectomy was poor response to serial botulinum A toxin injections. The surgery was done under general anesthesia. Orbicularis fibers (preseptal, pretarsal, and orbital) and brow protractor muscles (corrugator, supraciliaris, and procerus) was carried out through a brow and lidcrease skin incision. Care was taken to the supraorbital nerve. In case of lower eyelid blepharospasm, we performed lower eyelid myectomy. The lower eyelid skin was dissected from the orbicularis muscle, and all visualized portions of the orbicularis muscle were removed. Additional eyelid deformities, such as dermatochalasis, were also taken care simultaneously in the necessary cases. The skin was closed with 6/0 Vicryl suture, and pressure bandage was applied to the lid.

RESULTS

A total of 14 eyes of 7 patients, 5 women and 2 men, with a mean age of 62.1 ± 6.4 years (range, 55–72 years), were enrolled in the study. The mean duration of illness was 21.4 ± 12.7 months (range, 12–36 months), and the mean follow-up period was 24.2 ± 5.7 months (range, 18–36 months). All the patients had tried one or more medications before. Excessive blinking and visual disability were the most common symptoms in all patients (100%), and the severity and frequency of blepharospasm was evaluated as Grade 4 [Figure 1]. All patients had anxiety-related disease and severe functional disability. There was only one patient who had keratitis sequelae and dry eye as the triggering factor and received botulinum toxin treatment due to blepharospasm.

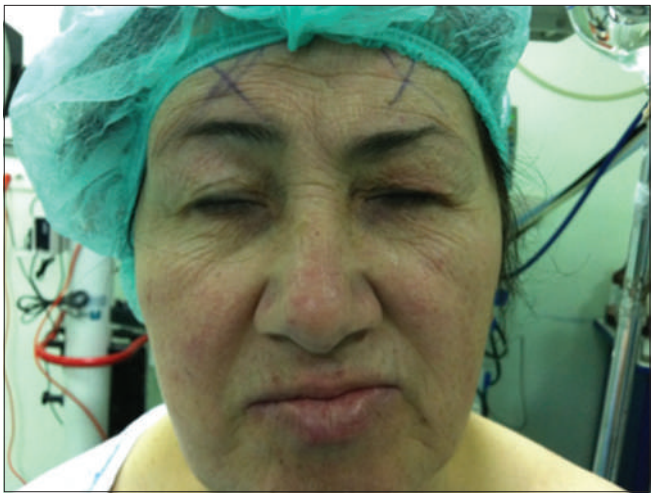


Figure 1: Preoperative with blepharospasm and apraxia of lids demonstrating eyelid spasms

Table 1: General characteristics of the patients

Features	*n*, %					
Number of patients	7					
Female/male	5/2					
Mean age	62.1±6.4 years (range 55-72 years)					
Mean duration of illness	21.4±12.7 months (range 12-36 months)					
Mean duration of follow-up period	24.2±5.7 months (range 18-36 months)					
Complaints at onset (%)						
Increased blinking	7 (100)					
Irritation	1 (14.3)					
Corneal disease	1 (14.3)					
Photofobia	1 (14.3)					
Visual disturbance	1 (14.3)					
Sensory tricks	7 (100)					
Anxiety regarding disease	7 (100)					
Extent of dystonia (%)						
Focal	7 (100)					
Segmental	-					
Generalized	-					
Functional disability grading	**Preoperatively (*n*=7)**			**Postoperatively (*n*=7)**		
	Mild	**Moderate**	**Severe**	**Mild**	**Moderate**	**Severe**
Reading	-	-	7	1	-	-
Watching TV	-	-	7	1	-	-
Walking	-	-	7	1	-	-
Driving	-	-	7	1	-	-
Shopping	-	-	7	1	-	-
Household task	-	-	7	1	-	-
Previous treatment (%)						
Medication	7 (100)					
Serial botulinum toxin injection	7 (100)					
Surgery	-					
Residual blepharospasm (%)						
Patients	2 (28.5)					
Procedure						
Botulinum toxin injection	1 (14.3)					
Lower eyelid myectomy	1 (14.3)					
Complete relief of spasms (%)	6 (85.7)					
Partial relief	1 (14.3)					
Mild-to-moderate irritation in the eye	1 (14.3)					

All patients underwent protractor myectomy surgery [Figure 2]. Of the 14 eyelids, 85.7% showed resolution of spasms and no longer required botulinum toxin treatment following surgery. Only one patient (14.3%) with corneal disease and dry eye required further botulinum toxin injection. The average time for botulinum injection was 30 days. One patient (14.3%) had residual lower eyelid blepharospasm and treated with lower eyelid myectomy combined with lower eyelid blepharoplasty. There was no major morbidity or visual loss and no postoperative superfacial punctate keratitis, and none of the patients experienced marked forehead numbness. All patients had transient periorbital edema, hematoma, and echymosis following surgery. All eyelids had excellent cosmetic results without signs of contracture. The functional disability score was increased to Grade 0 in all patients, and adequate lid opening was achieved 3 months after the surgery [Figure 3]. In the patient with keratitis sequelae and dry eye, the spasms and corneal irritation signs were completely resolved, but due to low vision, he had some difficulty in performing the daily activities.

DISCUSSION

Intermittent involuntary closure of the eyelids may occur in the presence of spasmodic contractions of the orbicularis oculi or in the failure of levator palpebrae contraction.[3,4] This failure is called AEO.[4]

Treatment of AEO is difficult.[3] Although botulinum toxin provides the most effective treatment, it is difficult to

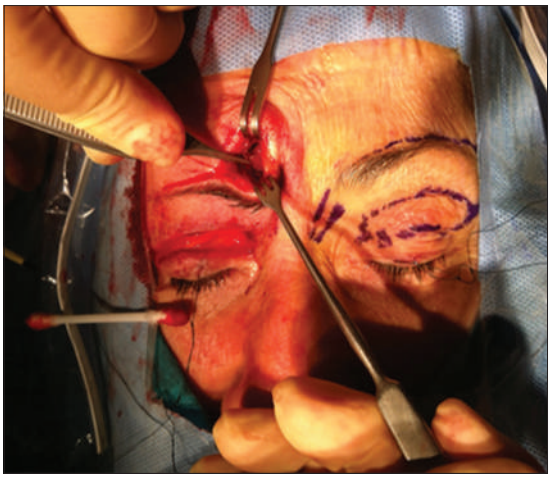


Figure 2: The corrugator superciliaris and procerus muscles between the brows and the base of the nose and also orbicularis muscles are removed



Figure 3: Adequate lid opening was achieved 3 months after the surgery

determine the proper dose, and also the effects of botulinum toxin are reversible and temporary, lasting approximately 3 months.[9,11,14]

It is very important for physicians and patients to be aware of AEO, when botulinum toxin fails to improve spasms despite the production of eyelid weakness.[15] While deciding the myectomy surgery, some conditions must be taken into consideration, such as AEO associated with BEB, blepharospasm-associated deformities, patients who are truly unresponsive to botulinum toxin, and patients who cannot afford or who refuse botulinum toxin injections.[7,15,16]

In a long-term study of patients with botulinum toxin-resistant BEB, it has been stated that subtotal myectomy provides subjective improvement and decreases the long-term need for injections in over 50% of patients.[11,17] In another study, it has been said that among patients who received upper eyelid myectomy or upper and lower eyelid myectomy, there was an improvement in 88% of patients, with only 38% requiring injections after full myectomy.[11,18] Ortisi *et al*. reported that protractor myectomy provides an improvement of the symptoms in 64% of patients and 89% of patients require continued injections of toxin.[10] Rana and Shah described two patients with apraxia of lid opening and blepharospasm, who responded to an initial treatment with botulinum toxin but later stopped responding satisfactorily despite increased doses.[15] Studies have also shown that patients who need botulinum toxin after myectomy surgery often require fewer injections which provide long-term relief.[10]

In our study, excessive blinking and visual disability were the most common symptoms seen in all patients (100%). While evaluating the spasms of patients, we used objective and subjective rating scales, as it is mentioned in the literature that the objective measures may not be sensitive and subjective rating scales may depend too much on the patient's memory expectations and psychological state at the time of rating.[14] According to this evaluation, the frequency and severity of blepharospasm of all patients in our study was Grade 4, and all had severe anxiety and functional disability.

It has been stated that there is a significant association between blepharospasm and anterior ocular segment diseases such as blepharitis and keratoconjunctivitis.[19] Dry eye has been reported frequently in patients with blepharospasm. It is difficult to determine whether dry eye causes blepharospasm, because the blink film increases to compensate for deficiency, or blepharospasm can cause dry eyes, because the periodic blinking effect of the eyelids is very important for the maintenance and renewal of the precorneal tear film.[20] We had one patient who was followed up due to keratitis sequelae and dry eye and received botulinum toxin for blepharospasm which resolved completely. Because of this result, we agree with the idea that these two mechanisms act together.

In our study, the preferred treatment in botulinum toxin-resistant cases was protractor myectomy which was successfully performed in 14 eyelids of seven patients. Nearly 85.7% of the patients showed resolution of spasm and no longer required botulinum toxin treatment following myectomy. One patient (14.3%) had residual lower eyelid blepharospasm and was treated with lower eyelid myectomy combined with lower eyelid blepharoplasty.

Although myectomy surgery is the first option in the treatment of AEO, there may be some surgery-related complications such as considerable swelling and ecchymosis, numbness of the forehead due to supraorbital nerve damage, chronic periorbital lymphedema, exposure keratitis, and

ectropion.[1,7] In our study, except transient periorbital edema, hematoma, and echymosis on the eyelids, we observed no major morbidity or visual loss related to surgery. All eyelids had excellent cosmetic results without signs of contracture, and adequate lid opening was achieved 3 months after the surgery.

CONCLUSION

Blepharospasm with AEO is a troublesome condition that severely restricts daily activities. As the resolution of symptoms from botulinum toxin is transient and requires repeated injections, protractor myectomy surgery is a good option in the treatment of severe cases. Although the small number of patients and retrospective nature of the present study did not allow us to explain the longterm effectiveness of the surgery, we conclude that myectomy surgery has a positive effect on the recovery of the patient's social life.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Bates AK, Halliday BL, Bailey CS. Surgical management of essential blepharospasm. Br J Ophthalmol 1991;75:487-90.
2. Yoon DK. Surgical treatment of essential blepharospasm. Kor J Ophthalmol 1988;2:90-4.
3. Hallett M. Blepharospasm: Recent advances. Neurology 2002;59:1306-12.
4. Sameera I. Minimal orbicularis myectomy: Does it relieve spasms in benign essential blepharospasm? Am J Cosmetic Surg 2015;32:1-10.
5. Hallett M, Evinger C, Jankovic J, Stacy M; BEBRF International Workshop. Update on blepharospasm: Report from the BEBRF International Workshop. Neurology 2008;71:1275-82.
6. Yoon WT, Chung EJ, Lee SH, Kim BJ, Lee WY. Clinical analysis of blepharospasm and apraxia of eyelid opening in patients with Parkinsonism. J Clin Neurol 2005;1:159-65.
7. Jivraj I, Meredith B, Shriver E. "I can't open my eyes": A case of blepharospasm and apraxia of eyelid opening. Ophthalmic Visual Sci 2015;23:1-7.
8. Defazio G, Livrea P, Lamberti P, De Salvia R, Laddomada G, Giorelli M, *et al*. Isolated so-called apraxia of eyelid opening: Report of 10 cases and a review of the literature. Eur Neurol 1998;39:204-10.
9. Jordan DR, Patrrinely JR, Anderson RL, Thiese SM. Essential blepharospasm and related dystonia. Survey Ophthalmol 1989;34:123-31.
10. Ortisi E, Henderson HW, Bunce C, Xing W, Collin JR. Blepharospasm and hemifacial spasm: A protocol for titration of botulinum toxin dose to the individual patient and for the management of refractory cases. Eye (Lond) 2006;20:916-22.
11. Clark J, Randolph J, Sokol JA, Moore NA, Lee HB, Nunery WR. Surgical approach to limiting skin contracture following protractor myectomy for essential blepharospasm. Digit J Ophthalmol 2017;23:8-12.
12. Waller RR, Kennedy RH, Henderson JW, Kenneth RK. Management of blepharospasm. Trans Am Ophthalmol Soc 1985;83:367-85.
13. Georgescu D, Vagefi MR, McMullan TF. Upper eyelid myectomy in blepharospasm with associated of lid opening. Am J Ophthalmol 2008;145:541-7.
14. Wabbels B, Jost WH, Roggenkämper P. Difficulties with differentiating botulinum toxin treatment effects in essential blepharospasm. J Neural Transm (Vienna) 2011;118:925-43.
15. Rana AQ, Shah R. Combination of blepharospasm and apraxia of eyelid opening: A condition resistant to treatment. Acta Neurol Belg 2012;112:95-6.
16. Pariseau B, Worley MW, Anderson RL. Myectomy for blepharospasm. Curr Opin Ophthalmol 2013;24:488-93.
17. Chapman KL, Bartley GB, Waller RR, Hodge DO. Follow-up of patients with essential blepharospasm who underwent eyelid protractor myectomy at the Mayo Clinic from 1980 through 1995. Ophthalmic Plast Reconstr Surg 1999;15:106-10.
18. Anderson RL, Patel BC, Holds JB, Jordan DR. Blepharospasm: Past, present, and future. Ophthalmic Plast Reconstr Surg 1998;14:305-17.
19. Sun Y, Tsai PJ, Chu CL, Huang WC, Bee YS. Epidemiology. PLoS One 2018;26:1-14.
20. Horwath-Winter J, Bergloeff J, Floegel I, Haller-Schober EM, Schmut O. Botulinum toxin A treatment in patients suffering from blepharospasm and dry eye. Br J Ophthalmol 2003;87:54-6.

Original Article

A study on changes in choroidal thickness in antenatal women with pregnancy-induced hypertension using spectral-domain-optical coherence tomography

ABSTRACT

Aim: To evaluate the choroidal thickness using spectral-domain optical coherence tomography (SD-OCT) in antenatal women with pregnancy induced hypertension. **Methods and Material:** In this observational study 100 eyes of 50 pregnant women with pregnancy induced hypertension were included and compared with age matched non- pregnant women. Retinal SD OCT scan was done. The choroidal thickness at the fovea, 500 µm nasal and temporal to the fovea were measured manually. **Statistical analysis used:** Unpaired T test was used. **Results:** The mean choroidal thickness in pregnant women with PIH was 346.23 µm and in normal non-pregnant women it was 296.16 µm. In Multigravida with PIH (317.91 µm) the choroidal thickness was found to be less when compared with primigravida with PIH(359.53). All these results were statically significant. **Conclusions:** This study revealed that there was increase in choroidal thickness in pregnant women with PIH when compared to non-pregnant women. However there was a significant decrease in choroidal thickness in multigravida women with history of PIH and also the thickness of choroid decreased with increase in age.Further studies are needed to correlate choroidal blood flow and choroidal thickness in PIH mother.

Keywords: Choroidal thickness, optical coherence tomography, pregnancyinduced hypertension

INTRODUCTION

Pregnancy-induced hypertension (PIH) and its complications are one of the leading causes of maternal mortality and morbidity worldwide. It complicates approximately 5%–7% of all pregnancies. The pathological changes in PIH are related to vascular endothelial dysfunction, increased systemic vascular resistance, and capillary leak. 40% of PIH patients have some visual disturbances. The most common Changes in the eye include narrowing of retinal arterioles, exudates, retinal edema, anterior ischemic optic neuropathy, transient blindness due to arteriolar spasm, cortical blindness, central serous retinopathy (CSR), Purtscher's-like retinopathy, and retinal and vitreous hemorrhages.[1,2,3]

The photoreceptors are supplied by choroidal vasculature which is responsive to vascular endothelial growth factor (VEGF), which is upregulated in PIH. Acute vasospasm results in a thickened, edematous choroid as compared to normotensive pregnant women.[4] Optical coherence tomography (OCT) is a new noninvasive, rapid, objective, retinal imaging modality that provides high-resolution, cross-sectional images of the retina and choroid with a longitudinal resolution of 3-10 μm. This study aims to assess the changes in choroidal thickness in PIH using OCT.[5-7]

Sozhamadevi Subramaniyan, Venkatesh Perumal, Thenmozhi R

Department of Ophthalmology, Government Mohan Kumaramangalam Medical College Hospital, Salem, Tamil Nadu, India

Address for correspondence: Dr. Venkatesh Perumal, Department of Ophthalmology, Government Mohan Kumaramangalam Medical College Hospital, Salem, Tamil Nadu, India.
E-mail: venkateshophthal@gmail.com

Submitted: 21-Mar-2020 **Accepted:** 28-Mar-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_34_20	

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Subramaniyan S, Perumal V, Thenmozhi R. A study on changes in choroidal thickness in antenatal women with pregnancy-induced hypertension using spectral-domain-optical coherence tomography. Kerala J Ophthalmol 2020;32:268-70.

Table 1: Mean choroidal thickness of women with pregnancy-induced hypertension and nonpregnant women

PIH women (μm)	Nonpregnant women (μm)
346.23	296.16

Unpaired *t*-test - *P*=0.0252 (significant). PIH – Pregnancy-induced hypertension

Table 2: Mean choroidal thickness in pregnancy-induced hypertension women

Nasal (μm)	Fovea (μm)	Temporal (μm)
342.33	346.58	349.79

Table 3: Mean choroidal thickness in multigravida and primigravida - pregnancy-induced hypertension

Multigravida with PIH (μm)	Primigravida with PIH (μm)
317.91	359.53

Unpaired *t*-test - *P*=0.0212 (significant). PIH – Pregnancy-induced hypertension

Table 4: Mean choroidal thickness of newly diagnosed pregnancy-induced hypertension and pregnancy-induced hypertension women on treatment

Newly diagnosed (μm)	Known PIH (μm)
348.14	342.28

Not significant. PIH – Pregnancy-induced hypertension

Table 5: Mean choroidal thickness with increasing age

<20 years (μm)	20-30 years (μm)	>30 years (μm)
351.59	345.93	327.5

The decrease in choroidal thickness with increase in age

SUBJECTS AND METHODS

One hundred eyes of 50 pregnant women with PIH, preeclampsia, and eclampsia were included in this study and compared with age-matched nonpregnant women. Patients with high refractive errors, hypertension, diabetes, Central Serous Chorioretinopathy, or any other retinal pathology involving the macula were excluded from this study. OCT was taken using spectral-domain-OCT. Radial and line protocol scans were taken. Choroidal thickness was measured from the outer border of the retinal pigment epithelium to the inner scleral border at the fovea, and 500 μm nasal and temporal to the fovea. The choroidal thickness measurement was done Manually [Figure 1].

RESULTS

The mean choroidal thickness in pregnant women with PIH was 346.23 μm [Table 1]. It was 346.58 μm, at the fovea, 342.33 μm nasal to the fovea, and 349.79 μm temporal to the fovea [Table 2]. The choroidal thickness in pregnant women with PIH was significantly increased when compared to the choroidal thickness of nonpregnant women (296.16 μm).[8]

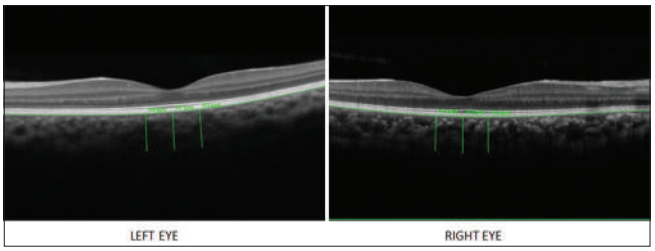


Figure 1: Choroidal thickness was measured from the outer border of the retinal pigment epithelium to the inner scleral border at the fovea, and 500 μm nasal and temporal to the fovea

There was a significant decrease in choroidal thickness in multigravida women with PIH (317.91 μm) when compared to Primigravida with PIH (359.53 μm) [Table 3]. Choroidal thickness in patients with recently diagnosed PIH was 348.14 μm, and it was 342.28 μm in PIH women on treatment [Table 4].

Choroidal thickness decreased with increasing age in PIH women. The choroidal thickness being 351.59 μm in age <20 years, 345.93 μm at 20–30 years, and 327.50 μm >30 years [Table 5].

DISCUSSION

PIH is the development of increase in blood pressure after 20 weeks of gestation in a woman with previously normal blood pressure. In a normal pregnancy, blood volume and erythrocyte count increases, physiological anemia occur, blood pressure drops, and there is an increase in cardiac blood flow and volume, causing an increase in ocular blood flow. Pregnancy is associated with ocular changes that may be physiological or pathological, and these are mostly transient and revert to normal but sometimes can become permanent. PIH is due to abnormal placentation resulting in increased systemic vascular resistance and generalized vasospasm in multiple organs including ocular vasculature. Choroid, which receives 65%–85% of the ocular blood flow, is important for normal functioning of the outer neuroretinal layer.[9]

It has been reported in studies that choroidal thickness in PIH was significantly thicker than healthy nonpregnant women.[10] It is mostly due to an increase in fluid retention in the choroidal layer during pregnancy. However, the choroidal thickness in PIH is low when compared with healthy pregnant women.[11] This decrease in choroidal thickness in pregnant women with PIH is due to increased vasospasm secondary to PIH. Maynard *et al*.[12] described that this is due to high concentration of placental soluble FMS-like tyrosine kinase 1 (VEGF inhibitor) that causes vasospasm and hypertension in PIH.

In this study, choroidal thickness was evaluated to assess the changes in this vascular layer. Choroidal thickness was

measured at the fovea, nasal to the fovea, and temporal to fovea and was compared with age-matched nonpregnant women.

The mean choroidal thickness was 346.58 μm, at the fovea, 342.33 μm nasal to the fovea, and 349.79 μm temporal to the fovea. This was similar to the results of studies by Bhayana *et al*.[13] and Narendran *et al*.[14] where the choroidal thickness was higher temporally.

The mean choroidal thickness in pregnant women with PIH was 346.23 μm, and the mean choroidal thickness in healthy nonpregnant women was 296.16 μm, which is statistically significant ($P = 0.02$). Ataş *et al*.[15] and Garg *et al*.,[7] also reported an increase in choroidal thickness in their studies.

The choroidal thickness in multigravida women with PIH was 317.91.μm, which is less when compared to primigravida women with PIH (359.53 μm). This has not been reported in earlier studies.

There was a decrease in the choroidal thickness with age from 351.59 μm at <20 years to 327.50 μm in mothers >30 years, which was similar to the studies by Margolis and Spaide[16] who reported a 15.6 μm decrease in CT every 10 years, and Ikuno *et al*.[17] showed a decrease of 14 μm for every 10 years.

CONCLUSION

This study revealed that there was an increase in choroidal thickness in pregnant women with PIH when compared to nonpregnant women. However, there was a significant decrease in choroidal thickness in multigravida women with PIH, and also the thickness of choroid decreased with increase in age. Further studies are needed to correlate choroidal blood flow and choroidal thickness in PIH women.

Limitation

In this study, the choroidal thickness in 2nd trimester was not evaluated. This being a tertiary care hospital, most of the patients were admitted for safe confinement in 3rd trimester and admissions were few in 2nd trimister. Evaluation of choroidal thickness in 2nd trimester would help to further understand the pathophysiological changes in the choroid in PIH.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Akhtar Z, Rishi P, Srikanth R, Rishi E, Bhende M, Raman R. Choroidal thickness in normal Indian subjects using Swept source optical coherence tomography. PLoS One 2018;13:e0197457.
2. Rahman W, Chen FK, Yeoh J, Patel P, Tufail A, Da Cruz L. Repeatability of manual subfoveal choroidal thickness measurements in healthy subjects using the technique of enhanced depth imaging optical coherence tomography. Invest Ophthalmol Vis Sci 2011;52:2267-71.
3. Maynard SE, Sukhatme VP, Karumanachi SA. Excess placental soluble FMS-like tyrosine kinase 1 (sFlt1) may contribute to endothelial dysfunction, hypertension and proteiunria in preeclampsia. J Clin Invest 2003;111:649-58.
4. Duru N, Ulusoy DM, Özköse A, Ataş M, Karatepe AS, Ataş F, *et al*. Choroidal changes in pre-eclampsia during pregnancy and the postpartum period: Comparison with healthy pregnancy. Arq Bras Oftalmol 2016;79:143-6.
5. Bakhda RN. Ocular manifestations of pregnancy induced hypertension. DJO 2015:26:88-92.
6. Reddy SC, Nalliah S, George SR, Who TS. Fundus changes in pregnancy induced hypertension. Int J Ophthalmol 2012;5:694-7.
7. Garg A, Wapner RJ, Ananth CV, Dale E, Tsang SH, Lee W, *et al*. Choroidal and retinal thickening in severe preeclampsia. Invest Ophthalmol Vis Sci 2014;55:5723-9.
8. Sheth BP, Mieler WF. Ocular complications of pregnancy. Curr Opin Ophthalmol 2001;12:455-63.
9. Liu R, Kuang GP, Luo DX, Lu XH. Choroidal thickness in pregnantwomen: A cross-sectional study. Int J Ophthalmol 2016;9:1200-6.
10. Peiretti E, Zhang J, Wang H, Yu Q, Tong Q, Lu Q. Enhanced depth imaging optical coherence tomography: A new way measuring choroidal thickness in pregnant women. Journal of Ophthalmology 2017:1–9.
11. Dadaci Z, Alptekin H, Oncel Acir N, Borazan M. Changes in choroidal thickness during pregnancydetected by enhanced depth imaging opticalcoherence tomography. Br J Ophthalmol 2015;99:1255-9.
12. Benfica CZ, Zanella T, Farias LB, Oppermann ML, Canani LH, Lavinsky D. Choroidal thickness in preeclampsia measured by spectral-domain optical coherence tomography. Int Ophthalmol 2019;39:2069-76.
13. Ataş M, Açmaz G, Aksoy H, Demircan S, Ataş F, Gülhan A, *et al*. Evaluation of the macula, retinal nerve fiber layer and choroid in preeclampsia, healthy pregnant and healthy non-pregnant women using spectral-domain optical coherence tomography. Hypertens Pregnancy 2014;33:299-310.
14. Margolis R, Spaide RF. A pilot study of enhanced depth imaging optical coherence tomography of the choroid in normal eyes. Am J Ophthalmol 2009;147:811-5.
15. Ikuno Y, Kawaguchi K, Nouchi T, Yasuno Y. Choroidal thickness in healthy Japanese subjects. Invest Ophthalmol Vis Sci 2010;51:2173-6.
16. Bhayana AA, Kumar V, Tayade A, Chandra M, Chandra P, Kumar A. Choroidal thickness in normal Indian eyes using swept-source optical coherence tomography. Indian J Ophthalmol 2019;67:252-5.
17. Narendran S, Manayath G, Venkatapathy N. Comparison of choroidal thickness using swept-source and spectral-domain optical coherence tomography in normal Indian eyes. Oman J Ophthalmol 2018;11:38-41.

Original Article

Clinical presentation and outcome of patients with optic pathway gliomas: A series of twenty patients

ABSTRACT

Background: Optic pathway gliomas (OPGs) are rare neoplasms where the management strategy is still evolving. The purpose of this study is to present our experience of twenty cases of OPGs with an intent to add to the general fund of knowledge. **Materials and Methods:** Retrospective chart review of patients with OPGs surgically managed in a single tertiary centre over a period of 5 years. Variables analysed include Age at diagnosis, sex, clinical manifestations, neurofibromatosis (NF1) status, tumour location, treatment modality, symptomatic progression, and complications. **Results:** Of the 20 patients with OPG in the study with a mean age of 12.8 years, four patients had NF1 and 16 had sporadic OPG. Dimness of vision (80%) and headache (55%) were the most common presenting symptoms. Majority of the tumours were chiasmatic (40%) in location and vision was affected bilaterally. Subtotal resection could be achieved in 12 (60%) partial decompression in 4 (30%) and gross total resection in 2 (10%). Four patients required additional shunt surgery for hydrocephalus. Postoperative radiotherapy was administered to thirteen patients and only one patient received chemotherapy. At the end of a mean follow up period of 44 months tumour recurrence was noticed in 11/20 (55%) patients. Vision improved in none, remained stable in fifteen and deteriorated in five. Three patients (15%) succumbed to tumour regrowth in the follow up period. **Conclusions:** OPGs are uncommon tumours with unpredictable outcome. Visual deficits are often irreversible and progressive. Curative resection is difficult and adjuvant therapy has variable response. Overall survival rates are good but progression free and survival rates are suboptimal.

Keywords: Chiasma, glioma, neurofibromatosis, optic nerve, orbit

INTRODUCTION

Optic pathway gliomas (OPGs) constitute a rare yet challenging group of intracranial and orbital neoplasms.[1-4] OPGs include a wide spectra of neoplasms with variable extensions, clinical manifestations and management challenges.[1,2] Management strategies have been evolving recently with increasing focus on the role of chemotherapy. Outcome is variable but generally poor, both in terms of vision and overall quality of survival.[3,4] Indian literature on this topic is limited and in the absence of large series a uniform standard of care treatment guideline is yet to be established.[5-7] We present our experience with twenty cases of OPGs with an intent to add to the general fund of knowledge.

Submitted: 12-Apr-2020 **Revised:** 16-Apr-2020
Accepted: 22-Apr-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_43_20	

MATERIALS AND METHODS

We retrospectively reviewed all patients who underwent surgery for an OPG at our institute from January 2015 to December 2019. Variables analysed include age at diagnosis, sex, clinical manifestations, neurofibromatosis (NF1) status, imaging findings, tumour location, treatment modality and recurrence. Visual acuity (VA) was analysed using Snellen Chart and field using Goldman's perimetry. Preoperative computerised tomography (CT) and magnetic resonance

Sudha Menon, Girish Menon[1], Reddy Vyjayanth[1], Neenu George, Vindhya Jagdish

Departments of Ophthalmology and [1]Neurosurgery, Kasturba Medical College, Manipal, Karnataka, India

Address for correspondence: Dr. Sudha Menon, Department of Ophthalmology, Kasturba Medical College, Manipal - 576 104, Karnataka, India.
E-mail: sudha.menon@manipal.edu

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Menon S, Menon G, Vyjayanth R, George N, Jagdish V. Clinical presentation and outcome of patients with optic pathway gliomas: A series of twenty patients. Kerala J Ophthalmol 2020;32:271-7.

imaging (MRI) data were used to determine the location of the tumour. All tumours were classified according to the Dodge classification: Stage I-involving only the optic nerve; Stage II-involving the optic chiasm, with or without optic nerve involvement; and stage III-involving the hypothalamus and/or adjacent structures.[8] The postoperative MRI, was used to determine the extent of surgical resection which was grouped as follows– gross-total resection (GTR removal of >90% of the tumour), subtotal resection (STR removal of 50%–90% of the tumor), and Partial decompression (removal of <50% of the tumour). All patients except those undergoing GTR were advised three-dimensional conformal radiotherapy and/or adjuvant chemotherapy. Statistical analyses were performed using the Fisher exact test, Chi-square analysis, and unpaired *t*-test. $P < 0.05$ were considered significant.

RESULTS

Patients' characteristics

A total of 20 patients with OPG were included in the study. Four patients (20%) had NF1 and 16 had sporadic (non-NF related) OPG. The study group included nine females and eleven males. All the four NF1 patients were males. Their age at surgery ranged from 2 to 40 years, with a mean age of 12.8 years. The demographics and clinical characteristics of the study groups are shown in Table 1.

Table 1: Patient demographics and clinical characteristics

Radiology and Management	Number
Age (years)	
<10	9
11-20	8
>20	3
Gender	
Male	11
Female	9
Visual deficit	
Visual decline	14
Nystagmus	3
Proptosis	2
Diplopia	1
Optic atrophy	13
Intracranial hypertension	
Headache	11
Disturbance in consciousness	3
Seizures	2
Weakness and other deficits	2
Endocrinology	
Obesity	2
Developmental delay	1
Polydypsia with polyuria	3
Neurofibromatosis	
Yes	4
No	16

Signs and symptoms at diagnosis

All the patients were symptomatic before diagnosis. The most common symptoms were diminution of vision (16/20; 80%) and headache (11/20; 55%). One patient presented with an intratumoral bleed and altered sensorium. Two patients (10%) with postchiasmatic lesions with frontal and temporal lobe extensions presented with seizures and contralateral weakness. The most common signs were decreased VA (77.5%), visual field deficits in the form of temporal filed cuts (10/20; 50%), optic atrophy (13/20; 65%) and nystagmus (32.5%). One patient with diplopia and a lateral rectus palsy secondary to raised intracranial pressure. Both the patients with prechiasmatic optic nerve gliomas (Dodge I) tumours had evidence of mild proptosis. Hypothalamic disturbance was noted in the form of obesity (2 patients), polydipsia and polyuria (1) and developmental delay (1).

Surgical management

The location of the tumour was predominantly chiasmatic (Dodge II 14/20; 70%), followed by post chiasmatic (Dodge II 4/20; 20%) and two Dodge I tumours [Figures 1-3]. Of the four patients with NF1, two had Dodge II, one each had Dodge I and Dodge III tumours. All the patients underwent surgery. Pterional craniotomy and a transylvian approach was the preferred approach for most (16/20; 80%) chiasmal tumours. Two patients with Dodge III tumours underwent a sub temporal approach. The two patients with optic nerve gliomas underwent transcranial frontal approach to the orbit. Gross total resection could be achieved in only two tumours (both pre chiasmatic). The rest of the patients had either sub total (12/20) or partial decompression (06/20). Hydrocephalus on diagnosis was detected in eight patients (40%) four (20%) of whom required a shunt in the postoperative period. On histopathology, 17 patients (85%) had a pilocytic astrocytoma (PA) (World Health Organization [WHO] Grade I), 2/20 patients had a diffuse astrocytoma (WHO Grade II) and one patient had anaplastic astrocytoma. Thirteen patients underwent postoperative radiotherapy and one received adjuvant chemotherapy also [Table 2].

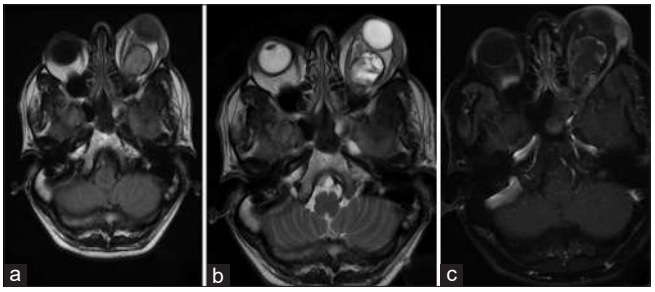


Figure 1: Axial T1 (a), axial T2 (b) and axial contrast T1 (c) images of a Dodge I optic nerve glioma with evidence of axial proptosis

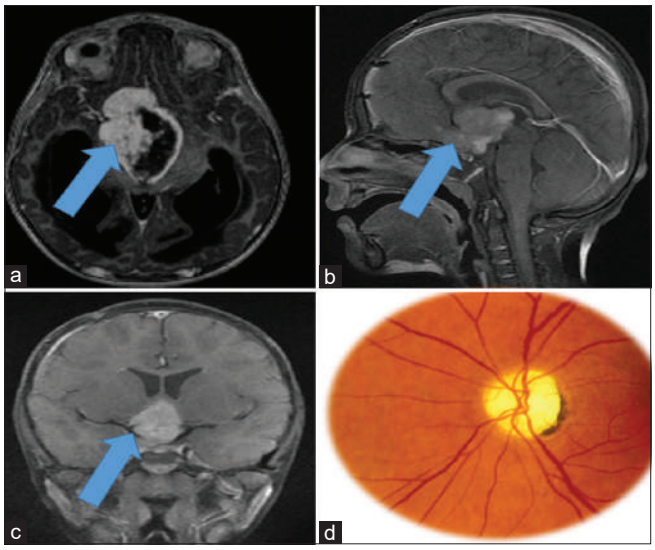


Figure 2: Axial (a), sagittal (b) and coronal (c) magnetic resonance imaging of a Dodge II chasmal tumour with extension up to the floor of the third ventricle (d) Fundoscopy showing primary optic atrophy

Table 2: Radiological features, management strategy and outcome in our series of ONG patients

Radiology and Management	Number
Tumour location	
Dodge I (prechiasmatic)	2
Dodge II (chiasmtic)	14
Dodge III (postchiasmatic)	4
Hydrocephalus	
Yes	8
Shunt	4
Resection	
GTR	2
STR	12
Partial decompression	6
Pathology	
Pilocytic astrocytoma	17
Diffuse astrocytoma	2
Anaplastic astrocytoma	1
Radiotherapy	
Yes	13
No	7
Chemotherapy	
Yes	1
Outcome	
Mortality	3
Vision	
Improvement	0
Stable	15
Deterioration	5
Recurrence	11
Resurgery for recurrence	2

GTR – Gross total resection, STR – Subtotal resection

Complications and mortality

The mean follows up period was 44 months. The overall 3-year survival rate was 85.71% (17/20). Three patients-two boys and one girl, died from tumour progression. All of these three patients had sporadic tumours, two Dodge III (Post chiasmatic) and one Dodge II (Chiasmatic) tumours. All of them had only STR of the tumour and all had undergone postoperative radiotherapy. All the three had evidence of tumour progression on follow up with raised intracranial pressure symptoms which resulted in mortality. One of these patients had an anaplastic astrocytoma on histology. Three patients developed postoperative infection with meningitis which responded well to appropriate antibiotics.

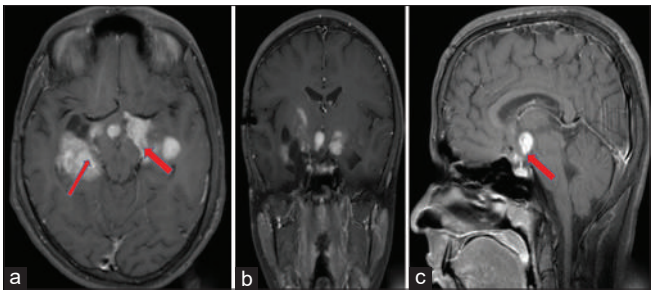


Figure 3: Axial contrast (a), coronal contrast (b) and sagittal contrast (c) magnetic resonance images of a Dodge III tumour with extensive spread along the hypothalamus and bilateral optic tracts (red arrows)

Recurrence

A total of 11 patients (55%) had tumour progression on MRI. Six of these recurrences were symptomatic with worsening of visual symptoms and raised intracranial pressure. Four of them underwent ventriculo peritoneal shunt and two underwent repeat surgery for further decompression [Table 2]. Both the pre chiasmatic tumours who underwent GTR remained clinically and radiologically stable at the end of nearly 4 years.

Visual outcome

VA outcome was uniformly poor in our series. None of the patients showed improvement following surgery, five deteriorated and fifteen remained static [Table 2]. Visual deterioration correlated with tumour progression in all these five patients. Interestingly, even patients (6/15; 40%) with stable VA showed a progression of field deficits on close follow up. Table 2 has no mention of visual field. So shift the reference to [Table 2] line 9.

Prognosticators

Optic atrophy on diagnosis was a definite prognosticator for visual deterioration and five of the thirteen patients with optic atrophy on admission deteriorated in vision. Vision was more likely to remain stable with chiasmatic lesions rather than pre or post chiasmatic lesions, but this observation lacked statistical significance. Age, sex, location of the tumour, extent of resection did not have statistically

Table 3: Predictors of visual outcome following surgery

	Improved (0)	Static (15)	Worsened (5)	Univariate analysis *P*
Age (years)				
<10 (9)	-	7	2	0.501
11-20 (8)	-	6	2	
>20 (03)	-	2	1	
Sex				
Male (11)	-	8	3	0.907
Female (9)	-	7	2	
Fundus				
Optic atrophy (13)		8	5	0.056
No atrophy (7)		7		
Location				
Dodge I (2)	-	2		0.334
Dodge II (14)	-	11	3	
Dodge III (4)	-	2	2	
Extent of resection**				
GTR (2)	-	2		0.512
STR (12)	-	10	2	
Partial decompression (6)	-	3	3	
Radiotherapy after first surgery				
Done (13)	-	10	3	0.378
Not done (7)	-	5	2	
Neuro fibromatosis				
NF I (4)	-	3	1	0.678
Sporadic (16)	-	12	4	

**Extent of removal was graded as Gross total removal (GTR) : 90-100%; Subtotal removal (STR); 25-90%; partial removal/ biopsy <25%. GTR – Gross total resection, STR – Subtotal resection, NF – Neurofibromatosis

significant correlation on vision or recurrence [Tables 3 and 4]. *P* value of sex with visual outcome-significant.

DISCUSSION

Epidemiology

OPGs are relatively rare neoplasms, comprising <1% of all CNS neoplasms in the general population and about 5% of central nervous system tumours in children.[1-9] The mean age at diagnosis ranges between 2.7 and 5.4 years and majority (80%–90%) of all patients belong to the first two decades of life.[10-12] Our study group had a mean age of 12.8 years, but we had nearly eight patients (40%) aged above 20 years. There is no gender predisposition and both men and females are equally affected. We did not notice any sex predilection in our series also.

OPGs are commonly associated with NF1 and appear in almost 30% of NF1 patients.[3] The mean age at diagnosis in NF1 patients is generally younger than generic patients. In a large series published recently by Nicolin *et al*. the mean age was 5.9 years, and there was a high proportion of Dodge II and III patients.[13] Bilateral OPGs are considered pathognomonic for NF1 and usually they tend to be located anteriorly.[14] Generally NF1 OPGs are considered relatively indolent tumours with less evidence of progression, an observation recently being challenged.[15] Surprisingly, the mean age of patients with NF1 (19.5 years) in our series was higher than that of sporadic OPG (11.12 years).

Location and classification

OPGs are low-grade glial neoplasms (WHO grade I) that originate from any location along the visual system such as the optic nerve, optic chiasm, optic tracts, and rarely, the optic radiation.[16] Various classification systems have been proposed for OPGs.[8,16,17] They were first classified in 1958 by Dodge and the old system is still widely followed. This system was modified in 2008 and where in each group was subdivided into further categories taking into account factors such as the existence of NF1 or leptomeningeal dissemination.[17] In Fred Epstein's classification proposed in 1985 the tumour component is rated anatomically (T1: One ON, T2: Both ONs, T3: OC, and T4: Hypothalamus/thalamus) and functionally (V0: Normal; V1: Impaired, one eye; V2: Impaired, both eyes, or blind, one eye; V3: Blind, one eye and impaired, one eye or field defect; and V4: Blind both eyes).[17] A comprehensive classification which takes into account additional parameters like age, NF, hydrocephalus, imaging features etc., has also been recently proposed by Shofty *et al*.[16] We adopted the earlier Dodge system in view of its ease in application.

Table 4: Predictors of recurrence following surgery

	Recurrence (11)	No recurrence (9)	Univariate analysis *P*
Age (years)			
<10 (9)	5	4	0.89
11-20 (8)	4	4	
>20 (3)	2	1	
Sex			
Male (11)	6	5	0.68
Female 09)	5	4	
Location			
Dodge I (2)	0	2	0.235
Dodge II (14)	9	5	
Dodge III (4)	2	2	
Extent of resection**			
GTR (2)	0	2	0.78
STR (12)	8	4	
Partial decompression (6)	3	3	
Radiotherapy after first surgery			
Done (13)	7	6	0.87
Not done (07)	4	3	
Neuro fibromatosis			
NF I (4)	2	2	0.97
Sporadic (16)	9	7	

**Extent of removal was graded as Gross total removal (GTR): 90-100%; Subtotal removal (STR); 25-90%; partial removal/ biopsy <25%. GTR – Gross total resection, STR – Subtotal resection, NF – Neurofibromatosis

Clinical presentation

Symptomatology can be variable depends on the location and extent of the lesion. Visual complaints dominate in almost all patients and neurological problems, such as headaches, vomiting, and seizures, are present in only 16% of patients.[12] Anterior optic nerve gliomas present with decreased VA, visual field cuts, nystagmus, and proptosis and the visual examination may reveal optic atrophy.[12] Posterior chiasmal tumours may produce endocrinological and hypothalamic symptoms like precocious puberty, obesity, short stature and somnolence.[16] Large tumours reaching up to the third ventricle may produce raised intracranial pressure symptoms due to hydrocephalus. Diencephalic syndrome (cachexia, macrocephaly, nystagmus, and visual deficit) is seen in 21% of infants with chiasmatic/ hypothalamic tumors.[18] The clinical manifestations in our series is almost similar to that mentioned in literature. One distinct difference was the relatively less incidence of hypothalamic disturbance in spite of the fact that majority of the tumours were Dodge II variety. One of our patients presented with an intratumoral bleed, an extremely uncommon finding in OPGs.

Radiology

OPGs are usually isointense on T1, hyperintense on T2, and show variable contrast enhancement. Tumours of the optic nerve appear as gross thickening of the nerve itself.[16,19] Differential diagnosis include other suprasellar tumours like craniopharyngiomas, pituitary adenomas, hypothalamic hamartomas etc., DTI tractography help in tracing the visual tract and is a useful adjunct for surgical planning.[20,21] Optical coherence tomography (OCT) has been shown to detect loss of retinal nerve fibre layer in patients with OPG.[22,23]

Pathology

PA accounts for the vast majority of these tumours and the rest include fibrillary and pilomyxoid astrocytomas (PMA), oligodendrogliomas, and gangliogliomas. PMA usually present in younger patients and show a more aggressive behaviour, and seeding through cerebrospinal fluid (CSF) pathways, higher mortality, decreased odd survival.[24,25] Malignant transformation of low-grade OPG is rarely seen; most frequently associated with irradiation.[26] In our series majority were PAs but we had one patient with a malignant anaplastic variant.

Natural history and prognosis

The natural history of OPGs is variable and not definite. Some tumours regress, some remain static and some grow and metastasize. Earlier considered benign, OPGs are increasingly being categorised as tumours with malignant potential.[27-29] Out of the 5% of low-grade gliomas that undergo leptomeningeal spread, OPGs represent ~ 50% and such spread is more commonly seen with sporadic OPG patients.[29] Patients with BRAF-KIAA 1549 fusion protein are known to have a better prognosis and molecular analysis for this marker protein may help in management decisions and surveillance.[15] None of the patients in our series exhibited spontaneous regression and none had leptomeningeal or CSF spread either.

Treatment

The exact timing of treatment is controversial. Patients with significant deficits, mass effect or hydrocephalus require immediate intervention. Treating an incidentally detected asymptomatic patients is controversial and is best managed by watchful expectancy and masterly inactivity.[12,30-32] For those with stable lesions, imaging and neuro-ophthalmological examinations (including visual field and OCT if available) needs to be done every 6 months. Treatment options include surgery, chemotherapy, and irradiation.

Surgery

For tumors with characteristic MRI appearance that are epicentered on the optic pathway, especially in an NF1 patient, no biopsy is required.[5,33] Biopsies are indicated if the tumor has an atypical appearance on MR, for an unusual age group (older than 10 years old or younger than

1-year-old), or with unusual clinical characteristics.[5,16] For Dodge I, isolated optic nerve gliomas surgery is best delayed and performed only when vision is severely compromised, when patient has intractable pain or progressive proptosis. Globe-sparing-resection of the lesion along with optic nerve is often carried out preferably through a transcranial approach. Surgical management of chiasmatic/hypothalamic tumours is challenging and controversial. Maximal safe resection especially of the exophytic multi compartmental lesion is the recommended surgical approach. Aim of surgery is to decompress the optic apparatus and open CSF pathways.[34-36] Ventriculoperitoneal shunt may be required is some cases if hydrocephalus persists.[37] Our surgical strategy too was directed along the above lines with an aim to preserve function at the expense of radical tumour clearance.

Chemotherapy

Chemotherapy is progressively being considered the first line of treatment in most cases of children with progressive OPG. "Gentle Chemotherapy" with vincristine and carboplatin was introduced in 1987 by Packer *et al*. and is now an accepted first-line treatment.[38] This regimen reported progression free survival rates (PFS) rates of 75% at 2 years and 68% at 3 years for chiasmatic tumours.[38] Weekly vinblastine is an option for patients with allergy to carboplatin.[39] Unfortunately, only one patient in our series received adjuvant chemotherapy.

Radiotherapy

Radiotherapy provides comparable results like chemotherapy and used to be the preferred approach before introduction of chemotherapy. With the usual dose of 45–60 Gy in 1.6–2 Gy fractions, 5-year PFS and overall survival rates are reported to be in the range of 82%–85% and 93%–94%, respectively.[40] Stereotactic radiosurgery, gamma knife and proton beam therapy are also reported to provide equally comparable results similar to conformal radiotherapy.[41-43]

Outcome

OPG outcome is generally favourable in terms of survival, but not so in terms of vision. Long-term survival rates are excellent, ranging between 80 and 96% in numerous series over the last 10 years.[4,13,30] The overall 10-year survival rate was 84%, and anterior tumors (nerve and chiasm) have a better 10-year survival of 95% then posterior tumors 76%.[44] In general, post treatment, 32% experience improvement in their VA, 40% stabilize, and 28% experience further deterioration.[45] Our mortality rate was higher in comparison to published literature. We had a 15% (3/20) mortality, all due to tumour regrowth. Our visual outcome, was not encouraging either, with vision remaining stable in fifteen patients (75%) and deteriorating in the rest. Eleven postoperative patients developed recurrence of whom six were symptomatic. Radiotherapy too did not seem to influence risk of recurrence was same in patients who underwent RT (53.84%) and who did not (57.14%).

Outcome prognosticators

As our series was small none of the parameters had a statistically significant influence on outcome prediction. However, we observed that presence of optic atrophy preoperatively and a Dodge III type of tumour had an adverse influence on outcome.

CONCLUSION

OPGs are uncommon with unpredictable outcome. Visual deficits are often irreversible and progressive. Curative resection is difficult and adjuvant therapy has variable response. Overall survival rates are good but progression free survival rates are suboptimal. The goal of visual control in OPG has not yet been reached. Further studies and research are needed to improve our current understanding about this enigmatic neoplasm.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflict of interest.

REFERENCES

1. Hidalgo ET, Orillac C, Wisoff JH. Optic pathway gliomas. In: Di Rocco C, Pang D, Rutka J, editors. Textbook of Pediatric Neurosurgery. Cham: Springer; 2018.
2. Shofty B, Ben-Sira L, Kesler A, Constantini S Optic pathway gliomas Adv Tech Stand Neurosurg 2015;42:123-46.
3. Binning MJ, Liu JK, Kestle JR, Brockmeyer DL, Walker ML. Optic pathway gliomas: A review. Neurosurg Focus 2007;23:E2.
4. Ahn Y, Cho BK, Kim SK, Chung YN, Lee CS, Kim IH, *et al*. Optic pathway glioma: Outcome and prognostic factors in a surgical series. Childs Nerv Syst 2006;22:1136-42.
5. Bommakanti K, Panigrahi M, Yarlagadda R, Sundaram C, Uppin MS, Purohit AK. Optic chiasmatic-hypothalamic gliomas: Is tissue diagnosis essential? Neurol India 2010;58:833-40.
6. Ganesh S, Gupta A, Sharma M, Bhuttan S. A case of neurofibromatosis 1 presenting with optic pathway glioma with an early onset and an aggressive course. Indian J Ophthalmol 2008;56:161-2.
7. Panigrahi M, Patel C, Chandrasekhar YB, Vooturi S. Management of optic pathway gliomas: Role of magnetic resonance imaging with diffusion-tensor imaging tractography in planning surgical resection. Int J Neurooncol 2019;2:3-6.
8. Dodge HW Jr., Love JG, Craig WM, Dockerty MB, Kearns TP, Holman CB, *et al*. Gliomas of the optic nerves. AMA Arch Neurol Psychiatry 1958;79:607-21.
9. Shamji MF, Benoit BG. Syndromic and sporadic pediatric optic pathway gliomas: Review of clinical and histopathological differences and treatment implications. Neurosurg Focus 2007;23:E3.
10. Grill J, Laithier V, Rodriguez D, Raquin MA, Pierre-Kahn A, Kalifa C. When do children with optic pathway tumours need treatment? An oncological perspective in 106 patients treated in a single centre. Eur J

Pediatr 2000;159:692-6.
11. Thiagalingam S, Flaherty M, Billson F, North K. Neurofibromatosis type 1 and optic pathway gliomas: Follow-up of 54 patients. Ophthalmology 2004;111:568-77.
12. Tow SL, Chandela S, Miller NR, Avellino AM. Long-term outcome in children with gliomas of the anterior visual pathway. Pediatr Neurol 2003;28:262-70.
13. Nicolin G, Parkin P, Mabbott D, Hargrave D, Bartels U, Tabori U, *et al*. Natural history and outcome of optic pathway gliomas in children. Pediatr Blood Cancer 2009;53:1231-7.
14. Chateil JF, Soussotte C, Pédespan JM, Brun M, Le Manh C, Diard F. MRI and clinical differences between optic pathway tumours in children with and without neurofibromatosis. Br J Radiol 2001;74:24-31.
15. Hawkins C, Walker E, Mohamed N, Zhang C, Jacob K, Shirinian M, *et al*. BRAF-KIAA1549 fusion predicts better clinical outcome in pediatric low-grade astrocytoma. Clin Cancer Res 2011;17:4790-8.
16. Shofty B, Constantini S, Bokstein F, Ram Z, Ben-Sira L, Freedman S, *et al*. Optic pathway gliomas in adults. Neurosurgery 2013;74:273-80.
17. Taylor T, Jaspan T, Milano G, Gregson R, Parker T, Ritzmann T, *et al*. Radiological classification of optic pathway gliomas: Experience of a modified functional classification system. Br J Radiol 2008;81:761-6.
18. Allen JC. Initial management of children with hypothalamic and thalamic tumors and the modifying role of neurofibromatosis-1. Pediatr Neurosurg 2000;32:154-62.
19. Shofty B, Constantini S, Bokstein F, Ram Z, Ben-Sira L, Freedman S, *et al*. Optic pathway gliomas in adults. Neurosurgery 2014;74:273-9.
20. Filippi CG, Bos A, Nickerson JP, Salmela MB, Koski CJ, Cauley KA. Magnetic resonance diffusion tensor imaging (MRDTI) of the optic nerve and optic radiations at 3T in children with neurofibromatosis type I (NF-1). Pediatr Radiol 2012;42:168-74.
21. Lober RM, Guzman R, Cheshier SH, Fredrick DR, Edwards MS, Yeom KW. Application of diffusion tensor tractography in pediatric optic pathway glioma. J Neurosurg Pediatr 2012;10:273-80.
22. Chang L, El-Dairi MA, Frempong TA, Burner EL, Bhatti MT, Young TL, *et al*. Optical coherence tomography in the evaluation of neurofibromatosis type-1 subjects with optic pathway gliomas. J AAPOS 2010;14:511-7.
23. Chang L, El-Dairi MA, Young TL, Bhatti MT. Retinal nerve fiber layer thickness in children with optic pathway gliomas. Am J Ophthalmol 2011;152:504-5.
24. Amatya VJ, Akazawa R, Sumimoto Y, Takeshima Y, Inai K. Clinicopathological and immunohistochemical features of three pilomyxoid astrocytomas: Comparative study with 11 pilocytic astrocytomas. Pathol Int 2009;59:80-5.
25. Komotar RJ, Burger PC, Carson BS, Brem H, Olivi A, Goldthwaite PT, *et al*. Pilocytic and pilomyxoid hypothalamic/chiasmatic astrocytomas. Neurosurgery 2004;54:72-9.
26. Zoeller GK, Brathwaite CD, Sandberg DI. Malignant transformation of an optic pathway glioma without prior radiation therapy. J Neurosurg Pediatr 2010;5:507-10.
27. Rozen WM, Joseph S, Lo PA. Spontaneous regression of low-grade gliomas in pediatric patients without neurofibromatosis. Pediatr Neurosurg 2008;44:324-8.
28. Piccirilli M, Lenzi J, Delfinis C, Trasimeni G, Salvati M, Raco A. Spontaneous regression of optic pathways gliomas in three patients with neurofibromatosis type I and critical review of the literature. Childs Nerv Syst 2006;22:1332-7.
29. Hoyt WF, Baghdassarian SA. Optic glioma of childhood. Natural history and rationale for conservative management. Br J Ophthalmol 1969;53:793-8.
30. Fouladi M, Wallace D, Langston JW, Mulhern R, Rose SR, Gajjar A, *et al*. Survival and functional outcome of children with hypothalamic/chiasmatic tumors. Cancer 2003;97:1084-92.
31. Listernick R, Ferner RE, Liu GT, Gutmann DH. Optic pathway gliomas in neurofibromatosis-1: Controversies and recommendations. Ann Neurol 2007;61:189-98.
32. Astrup J. Natural history and clinical management of optic pathway glioma. Br J Neurosurg 2003;17:327-35.
33. Leonard JR, Perry A, Rubin JB, King AA, Chicoine MR, Gutmann DH. The role of surgical biopsy in the diagnosis of glioma in individuals with neurofibromatosis-1. Neurology 2006;67:1509-1.
34. Wisoff JH, Abbott R, Epstein F. Surgical management of exophytic chiasmatic-hypothalamic tumors of childhood. J Neurosurg 1990;73:661-7.
35. Steinbok P, Hentschel S, Almqvist P, Cochrane DD, Poskitt K. Management of optic chiasmatic/hypothalamic astrocytomas in children. Can J Neurol Sci 2002;29:132-8.
36. Hoffman HJ, Humphreys RP, Drake JM, Rutka JT, Becker LE, Jenkin D, *et al*. Optic pathway/hypothalamic gliomas: A dilemma in management. Pediatr Neurosurg 1993;19:186-95.
37. Sawamura Y, Kamada K, Kamoshima Y, Yamaguchi S, Tajima T, Tsubaki J, *et al*. Role of surgery for optic pathway/hypothalamic astrocytomas in children. Neuro-Oncology 2008;10:725-33.
38. Packer RJ, Ater J, Allen J, Phillips P, Geyer R, Nicholson HS, *et al.* Carboplatin and vincristine chemotherapy for children with newly diagnosed progressive low-grade gliomas. J Neurosurg 1997;86:747-54.
39. Lafay-Cousin L, Holm S, Qaddoumi I, Nicolin G, Bartels U, Tabori U, *et al*. Weekly vinblastine in pediatric low-grade glioma patients with carboplatin allergic reaction. Cancer 2005;103:2636-42.
40. Erkal HS, Serin M Cakmak A. Management of optic pathway and chiasmatic-hypothalamic gliomas in children with radiation therapy. Radiother Oncol 1997;45:11-5
41. Hug EB, Muenter MW, Archambeau JO, DeVries A, Liwnicz B, Loredo LN, *et al.* Conformal proton radiation therapy for pediatric low-grade astrocytomas. Strahlenther Onkol 2002;178:10-7.
42. Kwon Y, Bae JS, Kim JM, Lee DH, Kim SY, Ahn JS, *et al.* Visual changes after gamma knife surgery for optic nerve tumors. Report of three cases. J Neurosurg 2005;102 Suppl: 143-6.
43. Merchant TE, Hua CH, Shukla H, Ying X, Nill S, Oelfke U. Proton versus photon radiotherapy for common pediatric brain tumors: Comparison of models of dose characteristics and their relationship to cognitive function. Pediatr Blood Cancer 2008;51:110-7.
44. Jenkin D, Angyalfi S, Becker L, Berry M, Buncic R, Chan H, *et al*. Optic glioma in children: Surveillance, resection, or irradiation? Int J Radiat Oncol Biol Phys 1993;25:215-25.
45. Fisher MJ, Loguidice M, Gutmann DH, Listernick R, Ferner RE, Ullrich NJ, *et al*. Visual outcomes in children with neurofibromatosis type 1-associated optic pathway glioma following chemotherapy: A multicenter retrospective analysis. Neuro-Oncology 2012;14:790-7.

Original Article

Efficacy of Trash To Treasure Retcam in Diabetic Retinopathy Screening

ABSTRACT

Background: Diabetes mellitus is a global burden and its prevalence has always been on an increase and screening is the only method by which early diagnosis and treatment can be achieved. The current screening tools are expensive, heavy and immovable. **Aims and Objectives:** To find out the efficacy in terms of sensitivity and specificity of the Trash To Treasure Retcam (T3 Retcam - An innovative, inexpensive, portable and light device) in comparison with direct ophthalmoscope and digital fundus camera (canon CF1). **Materials and Methods:** This was a hospital-based prospective study with a sample size of 80 patients calculated using Buderer's formula. The patients were ones who have diabetes for more than 5 years and have not more than NS-1 cataract status. They were dilated and examined using direct ophthalmoscope, digital fundus camera, and T3 Retcam. Each examination was carried out by three different consultants who were blinded to each other to reduce bias. Each of them was given a form in which they had to tick the particular finding as to yes or no according to its presence. A comparative analysis was done. **Results:** Sensitivity of the T3 Retcam was found to be 88.4% and 92% compared to Direct Ophthalmoscope and Canon funds camera, respectively. Specificity was found to be 100%. **Conclusion:** Trash To Treasure Retcam is a novel, inexpensive device made out of used material in the clinic/outpatient department. This device can help us detect diabetic retinopathy and can also be used as a major tool for screening in camps. It is comparable to the direct ophthalmoscope and digital cameras and yield good data for screening purposes.

Keywords: Carbon footprint, diabetic retinopathy screening, innovation, low-cost screening, smartphone ophthalmology

INTRODUCTION

Diabetes mellitus (DM) has established itself as a global burden in the light of microvascular and macrovascular complication and its prevalence has always been in the increase.[1] It is expected to rise from 285 million in 2010 to 439 million in 2030 globally.[2-5] Among this, around 35 million people have been diagnosed with diabetes in India. It is predicted to reach about 80 million by 2030.[3] Early detection and treatment of complications is imperative to reduce the morbidity caused by diabetes.[6] A well-known entity which comes with the complication of diabetes is diabetic retinopathy (DR). It is a potentially blinding disease in adults and is observed in both rural and urban settings in India.[7] The visual morbidity due to DR remains a significant health burden with 30% of them classified to have vision-threatening diabetic retinopathy.[5]

The progression of the disease is diagnosed through different stages ranging from Very Mild Non-Proliferative diabetic retinopathy (VMNPDR) to Moderate Non-Proliferative diabetic retinopathy (MODNPDR) stages, which are largely asymptomatic, to severe NPDR and proliferative diabetic retinopathy stages, which are potentially blinding if left untreated.[8] Vision loss can also be caused due to diabetic macular edema which can occur in addition to any of the

Prithvi Chandrakanth, K. S. Chandrakanth, A. K. Sheeja Vishwanathan[1], R. Nirupama[2], Tresa Mathew[3]

Departments of Comprehensive Ophthalmology, [1]Retina, [2]Cornea and [3]Glaucoma, Dr. Chandrakanth Malabar Nethralaya, Kozhikode, Kerala, India

Address for correspondence: Dr. Prithvi Chandrakanth, Chandradevi, Opp Super Petrol Pump, Behind QRS, Kannur Road, W. Naddakkavu, Kozhikode - 673 011, Kerala, India.
E-mail: dr.prithvi.chandrakanth@gmail.com

Submitted: 17-May-2020 **Accepted:** 23-Jun-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_60_20	

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Chandrakanth P, Chandrakanth KS, Sheeja Vishwanathan AK, Nirupama R, Mathew T. Efficacy of trash to treasure retcam in diabetic retinopathy screening. Kerala J Ophthalmol 2020;32:278-83.

above stages. DR has a fairly long asymptomatic period during which it can be easily diagnosed by fundus examination and imaging; hence, early detection and timely treatment are effective in reducing the risk of vision loss.[9-12] Dilated fundus examination using direct or indirect ophthalmoscopy by a trained ophthalmologist is ideal for screening DR.[13] Studies have suggested that retinal photography after dilatation is comparable to ophthalmoscopic examination for DR detection.[14,15] Fundus images traditionally are obtained with expensive and bulky tabletop machines which are usually operated by trained technicians in hospitals/clinical settings. Smartphones with their built-in camera technology and the ease and versatility of use have led to many ophthalmologists raising interest in their use in medical ophthalmic imaging. The portability, connectivity, storage capability, and the use of them in teleophthalmology have made them an attractive device for obtaining high-quality retina photographs in remote health centers/clinics and also in communities which currently receive no or suboptimal eye care facilities due to sociocultural and/or geographical barriers.[16] Comprehensive eye examination must be carried out at least annually to assess the DR grade in all patients with DM. A large number of patients do not receive the recommended quality of ophthalmic care.[17] This could mainly be due to fact that in India, majority of the population (72%) live in the rural areas and majority of the ophthalmologists are based in the urban centers. The ophthalmologist-to-population ratio is 1:107,000.[18-20] The nonavailability and high cost of the conventional tabletop fundus cameras further limit the screening process of DR.[21-23] Hence, to exploit the potential versatility of smartphones in diabetic retinopathy screening, various smartphone fundus imaging devices have been designed.[24-33] Trash To Treasure Retcam is a unique smartphone fundus imaging device which can be made by anyone in their hospital/clinic with just a used hand sanitizer bottle. This device couples a 20D lens with a smartphone in a stable and reliable manner and works on the principle of an indirect ophthalmoscope.[34]

The objective of this study was to evaluate the sensitivity and specificity of Trash To Treasure Retcam in diagnosing diabetic retinopathy and its usefulness in diabetic retinopathy screening.

Table 1: Distribution of number of eyes

Eyes	Frequency (%)
Normal	23 (28.75)
Diabetic retinopathy	57 (71.25)

Table 2: Age group

Age	Frequency (%)
20-30	3 (3.75)
31-40	5 (6.25)
41-50	25 (31.25)
51-60	41 (51.25)
61-70	6 (7.5)

Table 3: Gender distribution

Gender	Male	Female
Frequency (%)	37 (46.25)	43 (53.75)

Aim

To find out the efficacy in terms of sensitivity and specificity of the Trash To Treasure Retcam in comparison with direct ophthalmoscope and digital fundus camera (canon CF1).

MATERIALS AND METHODS

The study was done in the retina clinic of a tertiary eye hospital. It was a cross-sectional, hospital-based, comparative study. Study participants were the patients coming to the outpatient department (OPD) with DM. The sample size (n) was calculated using Buderer's formula based on sensitivity $= z^2_{1-\alpha/2} \times S_N \times (1 - S_N)/L_2 \times$ Prevalence

where n = required sample size, S_N = anticipated sensitivity, S_p = anticipated specificity, α = size of the critical

Table 4: Distribution of diabetic retinopathy changes

	Total number (n)	Detected in both the devices (%)	Missed in canon CF1 (%)	Missed in T3R (%)
Microaneurysms	57	50 (87.71)	2 (3.50)	6 (10.52)
Dot and blot hemorrhage	50	50 (100)	-	-
Hard exudate	39	39 (100)	-	-
Soft exudate	24	24 (100)	-	-
Superficial hemorrhage	28	28 (100)	-	-
IRMA	9	9 (100)	-	-
NVE	7	7 (100)	-	-
NVD	4	4 (100)	-	-
Tractional bands	10	10 (100)	-	-
Laser marks	12	12 (100)	-	-
CSME	16	16 (100)	-	-

IRMA: Intra Retinal Microvascular Abnormality; NVE: Neovascularization Elsewhere; NVD: Neovascularization of the Disc; CSME: Clinically Significant Macular Edema

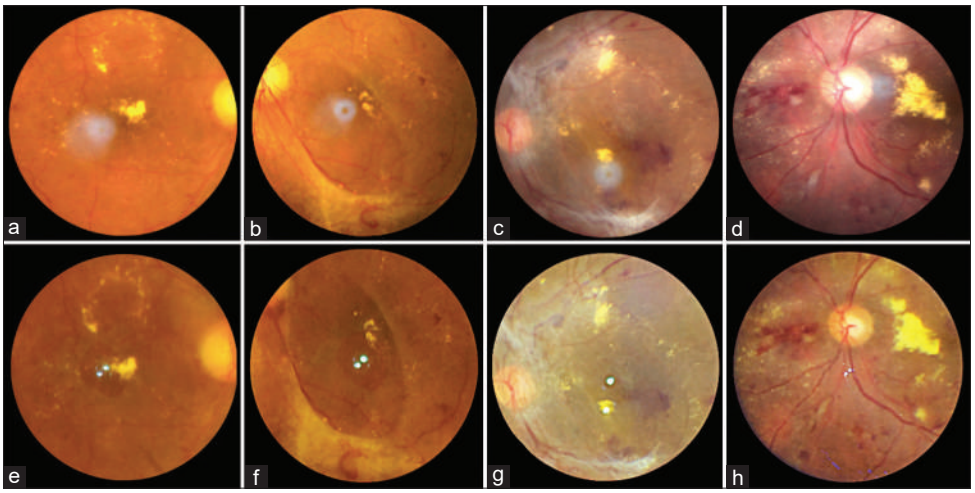


Figure 1: Images taken from Canon fundus camera (a-d). Images taken from the Trash To Treasure Retcam (e-h)

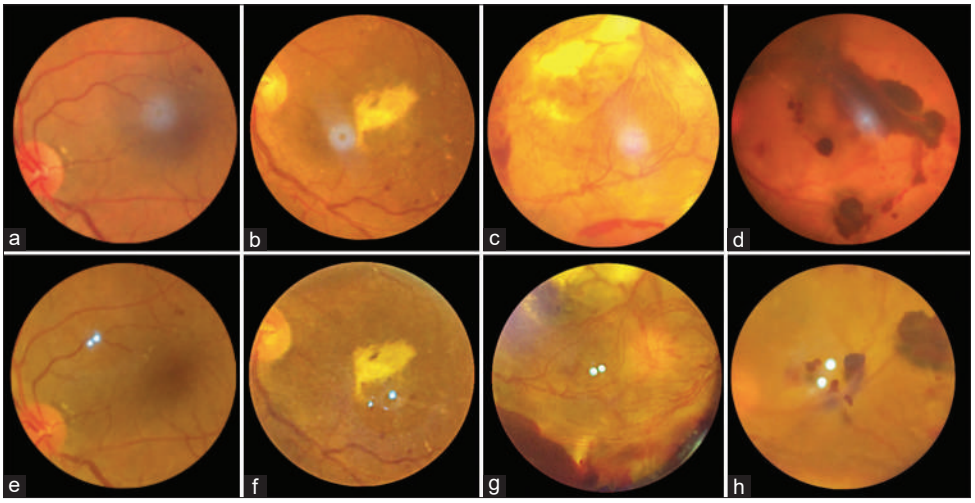


Figure 2: Images taken from Canon fundus camera (a-d). Images taken from the Trash To Treasure Retcam (e-h)

Table 5: ETDRS classification statistics

ETDRS grading	Direct ophthalmoscopy	CANON CF 1	T3 RETCAM
Normal	23	25	29
VMNPDR	7	5	1
MNPDR	14	14	14
MODNPDR	8	8	8
SNPDR	2	2	2
VSNPDR	6	6	6
EPDR	1	1	1
HRPDR	7	7	7
Post-PRP	12	12	12
CSME	16	16	16

PRP: Pan Retinal Photocoagulation, CSME: Clinically Significant Macular Edema, VMNPDR: Very Mild Non- Proliferative Diabetic Retinopathy, MNPDR: Mil Non-Proliferative Diabetic Retinopathy, MODNPDR: Moderate Non-Proliferative Diabetic Retinopathy; SNPDR: Severe Non-Proliferative Diabetic Retinopathy, VSNPDR: Very Severe Non-Proliferative Diabetic Retinopathy; EPDR: Early Proliferative Diabetic Retinopathy; HRPDR: High Risk Proliferative Diabetic Retinopathy

region ($1-\alpha$ is the confidence level), $z^2_{1-\alpha/2}$ = standard normal deviate corresponding to the specified size of the critical region (α), and L = absolute precision desired on either side (half-width of the confidence interval) of sensitivity or specificity assuming 80% sensitivity, 5% allowable error, prevalence 15%, the minimum sample size was calculated as 20 patients.

Sample and sampling

Inclusion criteria

i. Patients who are known diabetic for the past 5 years
ii. Age more than 18 years
iii. No contraindication to mydriasis
iv. No allergy to tropicamide.

Exclusion criteria

i. All patients with cataract, corneal pathology, or any media opacities

Table 6: Comparison and statistics of diabetic retinopathy diagnosis

	95% CI					*P*
	Sensitivity	Specificity	PPV	NPV	Kappa coefficient	
DO versus CCF	96.2 (88.4-100)	100	100	93.3 (80.1-100)	0.946	<0.001
DO versus T3R	88.4 (76.2-100)	100	100	82.2 (64.3-100)	0.843	
CCF versus T3R	92.0 (81.3-100)	100	100	88.00 (72.8-100)	0.896	

CI – Confidence interval, CCF – Carotid-cavernous fistula, PPV – Positive predictive value, NPV – Negative predictive value, DO - Direct ophthalmology

ii. Ocular infection.

Sample selection

By random selection, patients attending the OPD retina clinic were selected. A maximum of two patients were selected per day who fulfilled the above-mentioned inclusion and exclusion criteria. If there were more than two patients who fulfilled the criteria, the first two consecutive patients were taken up for the study.

Data collection tool/instrumentation

a. Pro forma
b. Details of measurement.

Informed and written consent were obtained. Detailed history of patients regarding name, age, sex, occupation, and address; drug history; any history of trauma or inflammation, presenting symptom, duration, and associated conditions; and medical history were recorded. Ocular examination was done to rule out any abnormality in the lids, conjunctiva, cornea, anterior chamber depth, iris, pupil, and lens. Gross systemic examination was done. Visual acuity and refraction were assessed using an illuminated Snellen's chart, and the WHO classification system was used to classify the best-corrected visual acuity and near vision with Jaegger's chart. Ocular examination was done to rule out any abnormality in the lids, conjunctiva, cornea, anterior chamber depth, iris, pupil, and lens with a torchlight. Slit-lamp biomicsroscopic examination was carried out to confirm the torchlight findings and to rule out conjunctival pathology, corneal pathology also anterior chamber dept. Dilatation of the eye was done by instilling eyedrop Itrop plus (tropicamide + phenylphrine) in the lower cul-de-sac. Stereoscopic fundus examination was done by slit lamp and + 90D lens or a ophthalmoscope and diagnosed by a senior consultant ophthalmologist. The patients were then taken up for fundus photography in the Canon CF 1 and the T3 Retcam which were operated by two different consultant ophthalmologists who were blinded to each other and were asked to tick the observational findings. Grading of diabetic retinopathy was done with Early Treatment Diabetic Retinopathy Study (ETDRS) grading system [Figures 1 and 2].

Statistical analysis

Details of data entry and storage/software: The data were entered and analyzed by SPSS version 24 (IBM, Armonk, New York, United States). The data were expressed as sensitivity, specificity, diagnostic accuracy, frequency, and percentages.

RESULTS

Demographics and baseline characteristics

We included 80 eyes in this study. There statistics shows that 23 eyes (28.75%) were found to be normal and 57 eyes (71.25%) had Diabetic retinopathy findings [Table 1]. Subjects in the age group of 51-60 years (51.25%) had the maximum number of patients with Diabetes Mellitus (DM) followed by those falling the age group of 41-50 years. Age group with the least subjects who had DM were those falling in 20-30 years [Table 2]. There were a total of 37 males (46.25%) and 43 Females (53.75%) included in this study [Table 3]. Direct Ophthalmoscope being the standard fundus examination technique, was used to examine each eye and with sign obtained were grouped into the Early Treatment Diabetic Retinopathy Study (ETDRS) classification. 23 eyes did not have any DR changes, 7 eye were diagnosed to have Very Mild Non proliferative Diabetic Retinopathy(VMNPDR), 14 were Mild Non Proliferative Diabetic Retinopathy (MNPDR), 8 were Moderate Non Proliferative Diabetic Retinopathy(MODNPDR), 2 were Severe Non Proliferative Diabetic Retinopathy(SNPDR), 6 were Very Severe Non Proliferative Diabetic Retinopathy, 1 was diagnosed as Early Proliferative Diabetic Retinopathy (EPDR), 7 were High Risk Non Proliferative Diabetic Retinopathy(HRPDR), 12 eyes had laser marks and were grouped under Post-Pan Retinal Photocoagulation Status(Post-PRP) and 16 were diagnosed to have Clinically Significant Macular Edema(CSME) [Tables 4 and 5].

Sensitivity and Specificity

After evaluating the eyes with both Canon Fundus Camera and Trash To Treasure Retcam the findings and diagnosis were compared with that of the Direct Ophthalmoscope and sensitivity and specificity were derived. The T3 Retcam had a sensitivity of 88.4% (95% Confidence Interval (CI) : 76.2 – 100) and a Specificity of 100% when compared to Direct

Ophthalmoscopy, Also a sensitivity of 92% (95% Confidence Interval (CI) : 81.3 - 100) and a Specificity of 100% when compared to The Canon CF1 Fundus Camera. The Positive Predictive Value, Negative Predictive Value, Kappa Coefficient and *P* Value were also calculated [Table 6].

DISCUSSION

Diabetic patients were selected and examinations and photographs were taken according to the method explained above. Demographic details show that there was a higher percentage of females (53.75) than males in this study and those with normal fundus comprised up to 28%. Diabetic retinopathy changes were more in individuals in the age group of 51–60 years. The diagnosis of Normal retina and Very Mild Non- Proliferative Diabetic Retinopathy(VMNPDR) depends upon the presence of the sign – Micro aneurysm. This sign was recognized in 57 cases through Direct Ophthalmoscopy and 50 cases were detected in both Canon CF1 and Trash To Treasure Retcam and hence was the limiting factor for these instrument as the misdiagnosed the VMNPDR cases as Normal cases. The number of cases missed by Canon cf1 was 2 and those with T3R was 6. All other diabetic retinopathy findings were detectable in both the devices. When each of the device was compared with each other, the CCF1 had a sensitivity of 96.2% compared to the DO and a specificity of 100%. The T3R had a sensitivity of 88.4% and a specificity of 100% with the DO. The T3R had a sensitivity of 92% with that of CCF1 and a specificity of 100%. When taking the statistics of the ETDRS classification, we can see that the T3R has misdiagnosed 6 patients as normal whereas they were diagnosed to have very mild NPDR by DO, which means that the T3R is not sensitive to the minute findings such as the microaneurysms and hence become the limitation of the T3R as it cannot pick the microaneurysms. The Trash To Treasure Retcam has been able to pick up larger findings/signs (Dot and Blot haemorrhage, hard exudate, soft exudate, superficial haemorrhage, IRMA, NVE, NVD, Tractional bands, Laser marks, CSME) which pose a threat to the vision. Although it misdiagnosed cases of VMNPDR group it has been able to diagnose sight threatening cases. Using Colour retinal photography has an advantage of being faster in diagnosing diabetic retinopathy with respect to easier acquisition, storage and transfer of retinal images. Studies have shown that Smartphone acquired images of diabetic retinopathy are comparable to those images taken by fundus cameras.[35-38] The Trash To Treasure Retcam can be valuable as a diagnostic tool with added benefit of carrying it anywhere and able to store and also share the picture. It can be used for tele ophthalmology and also for bedridden patient as well as for Retinopathy of Prematurity (ROP).

CONCLUSIONS

T3 Retcam is an inexpensive, quick, convenient, and portable device, with high sensitivity, specificity, and diagnostic accuracy for diabetic screening and helps reduce morbidity of patients who are at a high risk of vision loss especially in rural and camp settings.

Acknowledgment

- Mrs. Sree Devi Chandrakanth, Director, Dr. Chandrakanth Malabar Nethralaya, Kozhikode
- Miss. Grishma Chandrakanth, Director, Dr. Chandrakanth Malabar Nethralaya, Kozhikode.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. International Diabetes Federation, Diabetes Atlas. 6th ed.. Brussels, Belgium: International Diabetes Federation; 2014.
2. Shaw JE, Sicree RA, Zimmet PZ. Global estimates of the prevalence of diabetes for 2010 and 2030. Diabetes Res Clin Pract 2010;87:4-14.
3. Wild S, Roglic G, Green A, Sicree R, King H. Global prevalence of diabetes: Estimates for the year 2000 and projections for 2030. Diabetes Care 2004;27:1047-53.
4. Ding J, Wong TY. Current epidemiology of diabetic retinopathy and diabetic macular edema. Curr Diab Rep 2012;12:346-54.
5. Yau JW, Rogers SL, Kawasaki R, Lamoureux EL, Kowalski JW, Bek T, *et al*. Global prevalence and major risk factors of diabetic retinopathy. Diabetes Care 2012;35:556-64.
6. American Diabetes Association. Standards of medical care in diabetes–2013. Diabetes Care 2013;36 Suppl 1:S11-66.
7. Misra P, Upadhyay RP, Misra A, Anand K. A review of the epidemiology of diabetes in rural India. Diabetes Res Clin Pract 2011;92:303-11.
8. Sivaprasad S, Gupta B, Crosby-Nwaobi R, Evans J. Prevalence of diabetic retinopathy in various ethnic groups: A worldwide perspective. Surv Ophthalmol 2012;57:347-70.
9. Vashist P, Singh S, Gupta N, Saxena R. Role of early screening for diabetic retinopathy in patients with diabetes mellitus: An overview. Indian J Community Med 2011;36:247-52.
10. Rajalakshmi R, Amutha A, Ranjani H, Ali MK, Unnikrishnan R, Anjana RM, *et al*. Prevalence and risk factors for diabetic retinopathy in Asian Indians with young onset type 1 and type 2 diabetes. J Diabetes Complications 2014;28:291-7.
11. Early photocoagulation for diabetic retinopathy. ETDRS report number 9. Early Treatment Diabetic Retinopathy Study Research Group. Ophthalmol 1991;98:766-85.
12. Diabetic Retinopathy Study Research Group. Photocoagulation treatment of proliferative diabetic retinopathy. Clinical application of Diabetic Retinopathy Study (DRS) findings, DRS report number 8. Ophthalmology 1981;88:583-600.
13. Anjana RM, Pradeepa R, Deepa M, Datta M, Sudha V, Unnikrishnan R, *et al*. Prevalence of diabetes and prediabetes (impaired fasting glucose and/or impaired glucose tolerance) in urban and rural India: Phase I results of the Indian Council of Medical Research-India

DIABetes (ICMR-INDIAB) study. Diabetologia 2011;54:3022-7.
14. Hutchinson A, McIntosh A, Peters J, O'Keeffe C, Khunti K, Baker R, *et al*. Effectiveness of screening and monitoring tests for diabetic retinopathy – A systematic review. Diabet Med 2000;17:495-506.
15. Moss SE, Klein R, Kessler SD, Richie KA. Comparison between ophthalmoscopy and fundus photography in determining severity of diabetic retinopathy. Ophthalmology 1985;92:62-7.
16. Chow SP, Aiello LM, Cavallerano JD, Katalinic P, Hock K, Tolson A, *et al*. Comparison of nonmydriatic digital retinal imaging versus dilated ophthalmic examination for nondiabetic eye disease in persons with diabetes. Ophthalmology 2006;113:833-40.
17. American Diabetes Association. Standards of medical care in diabetes. Diabetes Care 2004;27 Suppl 1:S15-35.
18. Prathiba V, Rema M. Teleophthalmology: A model for eye care delivery in rural and underserved areas of India. Int J Family Med 2011;2011:683267. Available from: http://www.hindawi.com/journals/ijfm/2011/683267/. [Last accessed on 2012 Aug 18].
19. Kumar R. Ophthalmic manpower in India–need for a serious review. Int Ophthalmol 1993;17:269-75.
20. Samandar R, Kleefield S, Hammel J, Mehta M, Crone R. Privately funded quality health care in India: A sustainable and equitable model. Int J Qual Health Care 2001;13:283-8.
21. Rema M, Pradeepa R. Diabetic retinopathy: An Indian perspective. Indian J Med Res 2007;125:297-310.
22. Namperumalsamy P, Nirmalan PK, Ramasamy K. Developing a screening program to detect sight-threatening diabetic retinopathy in South India. Diabetes Care 2003;26:1831-5.
23. Mohan V, Prathiba V, Pradeepa R. Tele-diabetology to screen for diabetes and associated complications in rural India: The Chunampet rural diabetes prevention project model. J Diabetes Sci Technol 2014;8:256-61.
24. Teichman JC, Sher JH, Ahmed II. From iPhone to eyePhone: A technique for photodocumentation. Can J Ophthalmol 2011;46:284-6.
25. Lord RK, Shah VA, San Filippo AN, Krishna R. Novel uses of smartphones in ophthalmology. Ophthalmology 2010;117:1274-e3.
26. Myung D, Jais A, He L, Blumenkranz MS, Chang RT. 3D printed smartphone indirect lens adapter for rapid, high quality retinal imaging. J Mob Technol Med 2014;3:9-15.
27. Haddock L, Kim D, Mukai S. Simple, inexpensive technique for high-quality smartphone fundus photography in human and animal eyes. J Ophthalmol 2013;2013:1-5.
28. Sharma A, Subramaniam SD, Ramachandran KI, Lakshmikanthan C, Krishna S, Sundaramoorthy SK. Smartphone-based fundus camera device (MII Ret Cam) and technique with ability to image peripheral retina. Eur J Ophthalmol 2016;26:142-4.
29. Raju B, Raju NS, Akkara JD, Pathengay A. Do it yourself smartphone fundus camera-DIYretCAM. Indian J Ophthalmol 2016;64:663-7.
30. Shanmugam MP, Mishra DK, Madhukumar R, Ramanjulu R, Reddy SY, Rodrigues G. Fundus imaging with a mobile phone: A review of techniques. Indian J Ophthalmol 2014;62:960-2.
31. Raju B, Raju NS. Regarding fundus imaging with a mobile phone: A review of techniques. Indian J Ophthalmol 2015;63:170-1.
32. Nazari Khanamiri H, Nakatsuka A, El-Annan J. Smartphone Fundus Photography. J Vis Exp. 2017:55958.
33. Akkara J, Kuriakose A. The magic of three-dimensional printing in ophthalmology. Kerala J Ophthalmol 2018;30:209-15.
34. Chandrakanth P, Ravichandran R, Nischal NG, Subhashini M. Trash to treasure Retcam. Indian J Ophthalmol 2019;67:541-4.
35. Rajalakshmi R, Arulmalar S, Usha M, Prathiba V, Kareemuddin KS, Anjana RM, *et al*. Validation of smartphone based retinal photography for diabetic retinopathy screening. PLoS One 2015;10:e0138285.
36. Rachapelle S, Legood R, Alavi Y, Lindfield R, Sharma T, Kuper H, *et al*. The cost-utility of telemedicine to screen for diabetic retinopathy in India. Ophthalmology 2013;120:566-73.
37. Russo A, Morescalchi F, Costagliola C, Delcassi L, Semeraro F. Comparison of smartphone ophthalmoscopy with slit-lamp biomicroscopy for grading diabetic retinopathy. Am J Ophthalmol 2015;159:360-40.
38. Sengupta S, Sindal MD, Besirli CG, Upadhyaya S, Venkatesh R, Niziol LM, *et al*. Screening for vision-threatening diabetic retinopathy in South India: Comparing portable non-mydriatic and standard fundus cameras and clinical exam. Eye (Lond) 2018;32:375-83.

Basics of fluid air exchange in vitreoretinal surgery

FLUID AIR EXCHANGE

Fluid air exchange (FAE) is the substitution of fluid in the posterior cavity of eye with air with the aim to drain the subretinal fluid (SRF) and achieve a flat retina. FAE is followed by endolaser to the retinal breaks.

It is also performed in cases of an attached retina:

- After vitrectomy for vitreous hemorrhage, a partial/complete FAE helps in tamponading the retina and prevention of rebleed
- After silicone oil removal, few cycles of FAE are performed to remove the residual silicone oil droplets to remove the residual silicone oil droplets.

Technique of fluid air exchange

Three important things are to be ascertained before starting of FAE:[1-3]

1. Complete removal of vitreous at the edge of tears or posterior retinotomy (PR) is mandatory
2. Endocautery of edge of all the tears to facilitate in visualization and prevent excessive bleeding
3. Always check the air infusion pressure prior to starting it. It is generally kept at 30–35 mm Hg.

Flexible soft-tip cannulas are preferred over rigid cannulas for internal drainage of SRF because they can be angulated into the subretinal space and are less likely to damage the choroid.

Air infusion is then switched on.

Air is then insufflated, and the air bubble is enlarged to fill the anterior vitreous.

When the globe is partially filled with air, one may get double reflexes, and it is necessary to refocus the microscope and maneuver the extrusion cannula over the break/PR.

The extrusion cannula is kept over the break/PR [Figure 1] and SRF will be removed as the eye fills with air.

The retina may not flatten initially because fluid from the vitreous cavity often goes through retinal breaks or through the PR into the subretinal space.

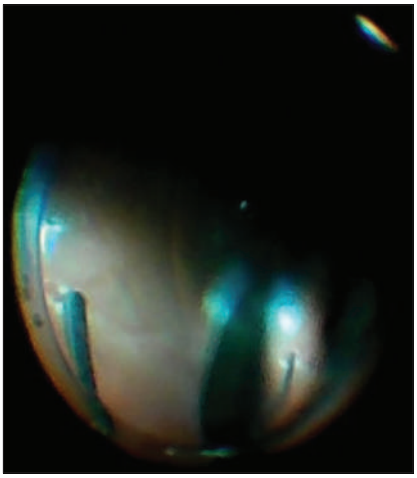


Figure 1: The extrusion cannula is kept over the retinotomy. The reflex from air is visible as it starts to fill the cavity

If difficulty is encountered in the initial part of the exchange and fluid cannot be aspirated while the silicone tube is in the subretinal space, the tube is withdrawn from the subretinal space and aspiration is applied in the vitreous cavity just anterior to the retinotomy. When the air level meets the PR, the silicone tip is gently slipped obliquely into the subretinal space, and aspiration is once more applied.

Care must be taken not to push the needle tip forcibly against the retinal pigment epithelium because choroidal hemorrhage may occur.

When the retina flattens posteriorly over the silicone tube, it is slowly withdrawn from the subretinal space while gently aspirating SRF.

At this point, all or most of the SRF has been removed.

Residual fluid can be aspirated over the optic nerve to complete the FAE. It is done by plunging the tip of the extrusion cannula into the fluid meniscus over the optic nerve [Figure 2].

A bright reflex disappears as the needle tip enters the fluid meniscus; this reflex reappears as the fluid level drops below the needle tip during aspiration.

Drainage from a peripheral break without liquid perfluorocarbon liquids

In rhegmatogenous retinal detachment (RRD), when the causative break is located at or posterior to the equator, endodrainage is usually done without perfluorocarbon liquids (PFCL).

© 2020 Kerala Journal of Ophthalmology | Published by Wolters Kluwer - Medknow

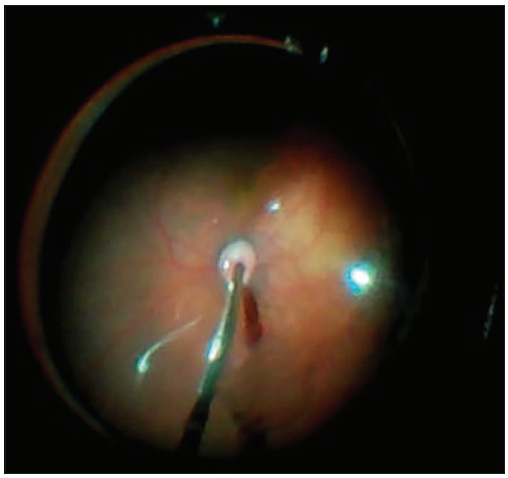


Figure 2: Residual fluid is aspirated by plunging the extrusion cannula into the fluid over the disc

Putting the soft tip cannula under the detached retina helps in decreasing the posterior fluid.

Maneuvering the head position can be helpful.

Drainage with perfluorocarbon liquids

It is preferred in cases of bullous RRD or RRD that is associated with retinal breaks anterior to the equatorial zone.

It avoids the creation of PR.

The steps involved are:

- PFCL is injected up to the level of the posterior edge of the break
- Do not overfill as it risks the subretinal migration of PFCL
- The surface tension from the PFCL bubble displaces the SRF through the break
- Maneuvering the patients head might facilitate the SRF removal
- FAE with the soft-tip cannula positioned at the top of the PFCL bubble displaces peripheral (anterior) SRF through the break
- Once the fluid sandwiched between the PFCL and air is removed, the soft-tip cannula is then dipped into the remnant PFCL meniscus over the disc to completely remove it.

Posterior retinotomy

- The peripheral drainage retinotomy should be created in an area most accessible to the drainage instrument
- The best location is in the peripheral retina posterior to either the temporal or nasal sclerotomy
- This location allows direct access of the soft silicone tube of the silicone-tipped needle to the drainage retinotomy and points the silicone tube, when inserted into the retinotomy, toward the posterior pole
- If an encircling scleral buckle is present, the drainage retinotomy should not be made immediately posterior to the scleral buckle because, as the retina flattens, the retinotomy will become obscured by the buckle
- It is better to make the retinotomy over the posterior portion of the buckle, where it can be observed throughout the exchange.

Difficulties encountered in fluid air exchange

In pseudophakic eyes with posterior capsulotomy, intraocular lens (IOL) condensation may significantly impair visibility.

Tip: Coating the posterior surface of the IOL with a dispersive viscoelastic substance is the most efficient way to resolve this problem.

Corneal striate keratopathy or pupillary constriction can occur in the aphakic eye that can reduce visualization and interfere with FAE.

Tip: One can put dispersive viscoelastic substance behind corneal endothelium to improve visualization.

In the presence of ACIOL/large posterior capsule rent, air may try to come into the anterior chamber (AC) which will lead to impaired visualization.

Tip: AC may be filled with viscoelastic before starting of FAE which will push the iris back and prevent the air from coming into the AC.[4]

Decreased visibility can also occur when PFCL droplets are left behind which are seen as vapors.

Tip: Best option is to revert back to fluid and remove residual PFCL and drain SRF through the PR.

In cases of open globe injury, although rare, venous/pulmonary air embolisms have occurred.[5]

Tip: (a) Always check the infusion line placement into the vitreous cavity to avoid inadvertent suprachoroidal air infusion. (b) Perform a slow FAE with low infusion pressure. (c) Maintain stable blood pressure while preventing it to go too low.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Gaganjeet Singh Gujral
Vitreoretinal Surgeon and Consultant, Sharp Sight Group of Eye Hospitals, New Delhi, India

Address for correspondence: Dr. Gaganjeet Singh Gujral, Department of Vitreoretina, Sharp Sight Group of Eye Hospitals, New Delhi, India.
E-mail: docgsgujral@gmail.com

REFERENCES

1. Charles ST. Controlled drainage of subretinal and choroidal fluid. Retina 1985;5:233-4.
2. McCuen BW 2nd, Bessler M, Hickingbotham D, Isbey E 3rd. Automated fluid-gas exchange. Am J Ophthalmol 1983;95:717.
3. Berrod JP, Rozot P, Raspiller A, Thiery D. Fluid air exchange *in vitro* retinal surgery. Int Ophthalmol 1994;18:237-41.
4. Landers MB 3rd. Sodium hyaluronate (Healon) as an aid to internal fluid-gas exchange. Am J Ophthalmol 1982;94:557-9.
5. Lim LT, Somerville GM, Walker JD. Venous air embolism during air/fluid exchange: A potentially fatal complication. Arch Ophthalmol 2010;128:1618-9.

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

Submitted: 27-Jul-2020 **Accepted:** 29-Jul-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_111_20	

How to cite this article: Gujral GS. Basics of fluid air exchange in vitreoretinal surgery. Kerala J Ophthalmol 2020;32:284-6.

Case Report

Ocular Juvenile Xanthogranuloma

ABSTRACT

We present a case of a 3-year-old male child with a history of swelling in the left eye of 1-month duration who was seen elsewhere for this complaint and was started on topical steroids. Slit-lamp examination revealed a conjunctival swelling of size 2 mm × 2 mm, well-defined margins, slightly raised from the surface, had a pink hue without any surrounding conjunctival congestion. A clinical diagnosis of foreign body granuloma was made, and the child underwent excision biopsy. Histopathological examination revealed Juvenile Xanthogranuloma (JXG). JXG was first described by Adamson in 1905 in a child who had multiple skin nodules. He defined those lesions by the name congenital xanthoma multiplex. A case of JXG with iris involvement was presented for the first time in 1948 at the Ophthalmic Pathology Club gathering in Washington DC, and it was published 1 year later by Blank *et al.*

Keywords: Benign conjunctival lesion, ocular juvenile xanthogranuloma, Touton giant cells

INTRODUCTION

Juvenile xanthogranuloma (JXG) is considered to be a rare benign condition in which there is a proliferation of histiocytes.[1] It is seen in infants and small children. JXG is the commonest variety of non-Langerhan's cell histiocytosis, which is marked by histopathological evidence of Touton giant cells.[2] The skin is the most frequent site of JXG, where it is presented as a solitary nodular lesion. After skin, the eye is the next most common site of JXG, in which the iris being the most frequently involved tissue (68%). The disease can also involve orbit, eyelid, conjunctiva, choroid, retina, and optic disc.[3,4]

CASE REPORT

A 3-year-old systemically healthy male child presented to our outpatient department with a history of swelling in the left eye of a 1-month duration. The swelling was not associated with any pain, redness, photophobia, or diminution of vision. Slit-lamp biomicroscopic examination revealed a conjunctival swelling of size 2 mm × 2 mm, well-defined margins, slightly raised from the surface, had a pink hue without any surrounding conjunctival congestion. A clinical suspicion of foreign body granuloma was made, and the child was posted for excision biopsy. During an excisional biopsy, the lesion was limited to conjunctival tissue and had no scleral adhesions. The lesion was removed completely and intoto. Histopathological examination revealed ocular JXG.

DISCUSSION

JXG is most commonly seen in children <2 years, but older children can also get affected. In 2 cohorts studies of 174 and 30 JXG patients each, the mean age was found to be 3.3 years (median 1 year) and 4.3 years (median 1.3 years), respectively. Among children with cutaneous JXG, 0.3%–10% were found to have ocular disease where children younger than 2 years were at a higher risk.[5] Male:female ratio ranges between 1.1:1 and 1.4:1. There is a 0.75% rate of systemic involvement in those with cutaneous JXG.[6]

Shaji Hussain, Anu Mariya Paul

Department of Ophthalmology, Cornea Clinic, Al Salama Eye Hospital, Malappuram, Kerala, India

Address for correspondence: Dr. Shaji Hussain, Cornea Department, Cornea Clinic, Al Salama Eye Hospital, Perinthalmanna, Malappuram, Kerala, India.
E-mail: drshajihussain@yahoo.com

Submitted: 03-Dec-2019 **Revised:** 21-Jan-2020
Accepted: 23-Jan-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_84_19	

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Hussain S, Paul AM. Ocular juvenile xanthogranuloma. Kerala J Ophthalmol 2020;32:287-90.



Figure 1: (a) Conjunctival juvenile xanthogranuloma, (b) tissue excised in toto, (c) tissue in absolute alcohol and marked for pathology evaluation

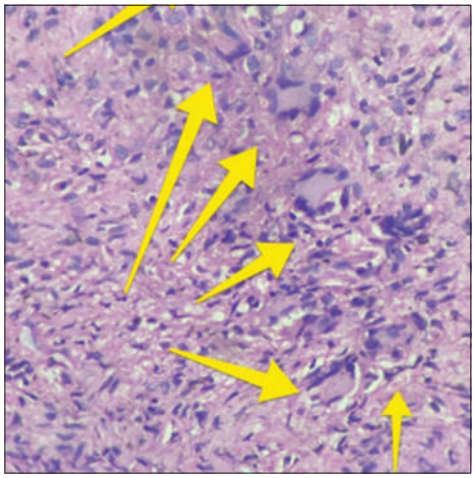


Figure 2: High power, multinucleated giant cells

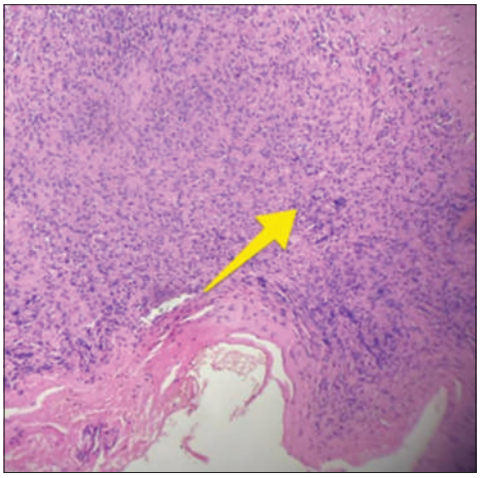


Figure 3: Low power, multinucleated giant cells

The skin is the most frequent site of JXG. Cutaneous lesions are seen in 75% of patients as the only clinical manifestation and may involve eyelids, head, neck, trunk, and upper and lower extremities.[5] Iris lesions are mostly unilateral, localized, yellow-colored, elevated, and vascularized. They may either appear as a thin layer diffused on the iris surface or as masses over the iris thus resulting in heterochromia.[3] These can lead to spontaneous hyphema and thus predispose to secondary glaucoma. Only 0.3%–10% of cutaneous JXG patients have ocular lesions.

After iris, the conjunctiva (19%) is the next common site. In the bulbar conjunctiva, these present as nodular lesions that are localized and yellow in color. The least common sites for JXG are the lungs, pericardium, salivary glands, viscera, central nervous system, kidneys, testes, ovaries, and bones.

Neurofibromatosis-1, Niemann–Pick disease, and urticaria pigmentosa are associated with JXG.[7] Patients having both JXG and neurofibromatosis type-1 (NF-1) are at 20–32 fold risk of developing juvenile myelomonocytic leukemia (JMML) than those having NF1 without JXG. Hence, all patients with NF1 and JXG must be evaluated for JMML development.[8]

Our patient underwent a complete systemic and ocular examination. There were not any evidence of systemic diseases. Excision biopsy was done under mild sedation to confirm the diagnosis.

The biopsy report revealed the following histopathological findings.

The pathogenesis in JXG is of reactive origin. Here, a histioxanthomatous reaction is thought to be evoked by a local tissue damage. Histopathological examination shows Touton giant cells, lymphocytes, epithelioid histiocytes, and eosinophils [Figures 1-6]. A Touton type of giant cell may be described as a multinucleated cell with a wreath of nuclei that surrounds a homogeneous cytoplasm, and in the periphery of the nuclei, there is a rim of foamy cytoplasm. Remember that Touton giant cells are not pathognomonic of JXG. Touton giant cells are rarely seen in early JXG, where small to intermediate-sized mononuclear histiocytes are found to display a compact sheet-like infiltrate. The cytoplasm will be eosinophilic and sparse to moderate in amount and may not contain lipid vacuoles or sometimes may have only fine vacuoles. The cells stain positive for CD68 and negative for CD1a, CD 20716, and S100 and have variable reactivity with factor XIIIa.[9]

JXG arising in the conjunctiva has close differential diagnoses like epibulbar dermoid (benign congenital lesions with choristomatous tissue occurring inferotemporal quadrant

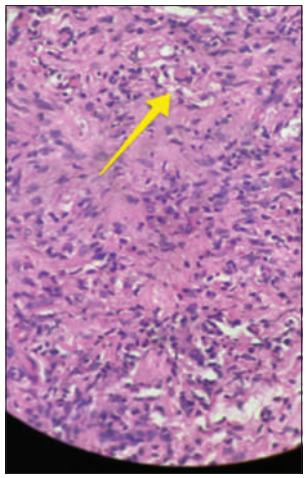


Figure 4: Fibrotic stroma with xanthomatous changes

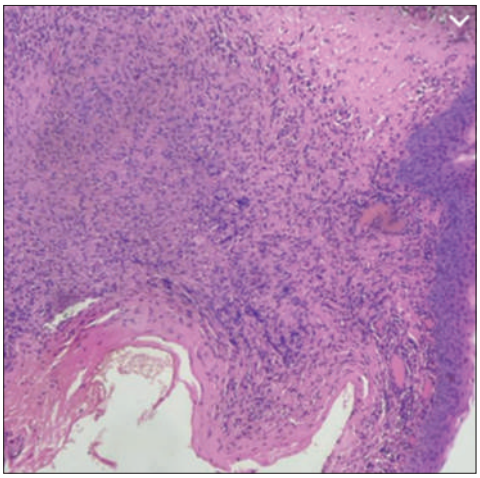


Figure 5: Scanner view, lesion showing spindle cell stroma with mild mixed inflammatory cells

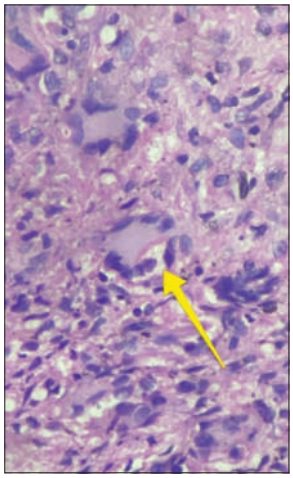


Figure 6: High power Touton giant cells

of the corneal limbus), dermolipoma (located near temporal fornix and composed of adipose tissue), and less frequently, phlyctenular keratoconjunctivitis, neurofibroma, fibrous histiocytoma (recurrent crop like lesions may spontaneously resolve in months), pterygium, pyogenic granuloma, and foreign body granuloma, as well as other primary and secondary inflammatory lesions such as Langerhans cell histiocytosis.

Excision biopsy is both diagnostic and therapeutic for lesions over the eyelid. Corticosteroids (topical and intralesional) may be tried over eyelid lesions alternatively. Limbal lesions have shown success with topical corticosteroid therapy. Cutaneous lesions may be observed for 1–5 years as they either regress or stabilize during this period. For conjunctival lesions, if topical treatment response is poor or in the contexts of poor compliance, periocular corticosteroid injection may be considered. In cases where there is no response to corticosteroids and in lesions which show atypical presentations, fine-needle aspiration cytology may be performed to exclude the possibility of a malignancy.

Low-dose ocular radiotherapy and systemic steroids may be considered in recalcitrant conditions. Extensive systemic JXG requires multimodal chemotherapy.[10]

JXG has a favorable prognosis in general. Cutaneous disease either stabilizes or regresses with time. In the eye, lid and conjunctival lesions have a benign course, whereas iris JXG can be vision-threatening due to its complications. Data analysis of 129 patients who underwent excisional biopsy showed no recurrence among 83% of the patients, 10% had recurrent lesions, and 7% developed an additional lesion near the original tumor site. Death was reported very rarely in systemic JXG. Unlike Langerhan's cell histiocytosis, systemic involvement is rare in JXG, thus explaining the favorable prognosis.[10]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient's bystanders have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published, and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Dehner LP. Juvenile xanthogranulomas in the first two decades of life: a clinicopathologic study of 174 cases with cutaneous and extracutaneous manifestations. Am J Surg Pathol 2003; 27:579–93.

2. American Academy of Ophthalmology. Juvenile xanthogranuloma. In: Basic and Clinical Science Course: Pediatric Ophthalmology and Strabismus. Sec. 6. San Francisco, CA: American Academy of Ophthalmology; 2017.
3. Samara WA, Khoo CT, Say EA, Saktanasate J, Eagle RC Jr., Shields JA, *et al*. Juvenile xanthogranuloma involving the eye and ocular adnexa: Tumor control, visual outcomes, and globe salvage in 30 patients. Ophthalmology 2015;122:2130-8.
4. Johnson TE, Alabiad C, Wei L, Davis JA. Extensive juvenile xanthogranuloma involving the orbit, sinuses, brain, and subtemporal fossa in a newborn. Ophthalmic Plast Reconstr Surg 2010;26:133-4.
5. Cypel TK, Zuker RM. Juvenile xanthogranuloma: Case report and review of the literature. Can J Plast Surg 2008;16:175-7.
6. Samuelov L, Kinori M, Chamlin SL, Wagner A, Kenner-Bell BM, Paller AS, *et al*. Risk of intraocular and other extracutaneous involvement in patients with cutaneous juvenile xanthogranuloma. Pediatr Dermatol 2018;35:329-35.
7. Cambiaghi S, Restano L, Caputo R. Juvenile xanthogranuloma associated with neurofibromatosis 1: 14 patients without evidence of hematologic malignancies. Pediatr Dermatol 2004;21:97-101.
8. Zvulunov A, Barak Y, Metzker A. Juvenile xanthogranuloma, neurofibromatosis, and juvenile chronic myelogenous leukemia. World statistical analysis. Arch Dermatol 1995;131:904-8.
9. Sonoda T, Hashimoto H, Enjoji M. Juvenile xanthogranuloma. Clinicopathologic analysis and immunohistochemical study of 57 patients. Cancer 1985;56:2280-6.
10. Janssen D, Harms D. Juvenile xanthogranuloma in childhood and adolescence: A clinicopathologic study of 129 patients from the Kiel pediatric tumor registry. Am J Surg Pathol 2005;29:21-8.

Case Report

Iridocorneal endothelial syndrome – A case of contradictions

ABSTRACT

Iridocorneal endothelial syndrome comprises a spectrum of disorders, which present with features of corneal edema, iris atrophy, peripheral anterior synechiae, and secondary glaucoma, in varying combinations. Clinical variants include Chandler's syndrome, progressive iris atrophy, and Cogan-Reese/iris nevus syndrome. Although classically described as a unilateral condition in young women, bilaterality, varied age of presentation, and male gender are also possibilities. Here, we report a case of bilateral progressive iris atrophy in an elderly male.

Keywords: Bilateral, iridocorneal endothelial syndrome, progressive iris atrophy, secondary glaucoma

INTRODUCTION

Iridocorneal endothelial (ICE) syndrome denotes a disorder in which the normal endothelium is altered, and this leads to varying degrees of corneal edema, iris atrophy, and secondary glaucoma. The altered endothelium migrates across the angle structures and the iris, and contraction of this tissue leads to the characteristic high peripheral anterior synechiae (PAS), iris changes, and secondary glaucoma.[1,2]

It includes three clinical variants:

1. Chandler's syndrome
2. Essential/progressive iris atrophy
3. Cogan-Reese/iris nevus syndrome.

It commonly presents unilaterally in women between the ages of 20 and 50 years.[3-5]

There have been a few isolated reports in literature of male preponderance as well as bilateral involvement.[1,4,6-8]

We report a case of bilateral progressive iris atrophy in an elderly male.

Submitted: 03-Mar-2020 **Revised:** 23-Mar-2020
Accepted: 18-Apr-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_26_20	

Cases presenting with bilateral involvement may be mistaken for posterior polymorphous dystrophy. Typical features should, however, direct the diagnosis toward ICE syndrome.

CASE REPORT

A 72-year-old male presented with a history of gradual, painless, and progressive decrease of vision in both eyes for 1 year. He had a recent onset of recurrent episodes of moderately intense eye pain, redness, colored haloes, and headache. There were no associated systemic illnesses or other relevant family, personal, or past histories.

Ocular examination revealed, band keratopathy, iris atrophy, corectopia and multiple PAS in all quadrants, in both eyes. There were stretch holes in the inferior quadrants in both eyes [Figures 1a, b and 2a, b].

Jincy Mariya Paul, Padma B. Prabhu, Charmaine B. Solomon

Department of Ophthalmology, Government Medical College, Kozhikode, Kerala, India

Address for correspondence: Dr. Jincy Mariya Paul, Department of Ophthalmology, Government Medical College, Kozhikode, Kerala, India.
E-mail: dr.jmpaul92@gmail.com

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Paul JM, Prabhu PB, Solomon CB. Iridocorneal endothelial syndrome – A case of contradictions. Kerala J Ophthalmol 2020;32:291-4.

Best-corrected visual acuity was 6/9 (right eye [OD]) and 6/24 (left eye [OS]) with elevated intraocular pressure (IOP) measured by applanation tonometry (OD = 28 mmHg and OS = 32 mmHg).

Fundus evaluation showed glaucomatous cupping in both eyes (cup-to-disc ratio; OD = 0.8 and OS = 0.9) [Figure 3a and b].

Field examination by Humphrey field analyzer showed severely depressed fields in both eyes, and specular microscopy showed typical ICE cells [Figure 4].

Routine blood investigations were within normal limits.

The patient was managed with augmented trabeculectomy, following control of IOP with medical management [Figure 5].

DISCUSSION

ICE syndrome is thought to be due to a replacement of the normal endothelial cells with cells having epithelial-like characteristic and migratory properties.[1,4]

The true etiology is not yet known with theories of viral infection being currently investigated.[9]

The endothelium shows a typical hammered silver or beaten-bronze appearance on slit-lamp examination.

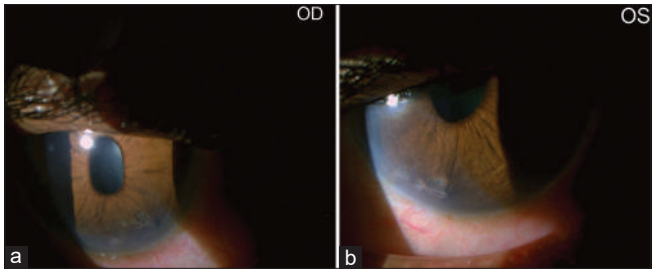


Figure 1: (a) Finding of corectopia in the right eye. (b) Peripheral anterior synechiae and stretch holes seen in the left eye

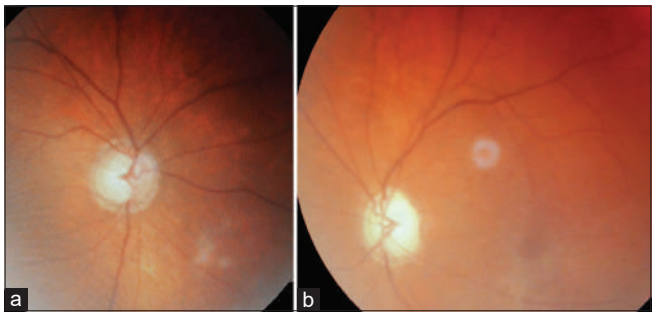


Figure 3: (a) Fundus photo showing glaucomatous cupping in the right eye. (b) Advanced cupping in the left eye

The variants are distinguished based on varied clinical findings.[2]

Chandler's syndrome

This is the most common subtype, and it typically presents with corneal edema with little or no iris changes.

Essential/progressive iris atrophy

Iris findings predominate in this subtype with typical findings such as polycoria, corectopia, iris hole formation, ectropion uveae, and iris atrophy.

Iris nevus/Cogan-Reese syndrome

In this variant of ICE syndrome, tan pedunculated nodules or diffuse pigmented lesions are found on the anterior surface of the iris, with iris atrophy being a less common finding.

Specular microscopy is an important diagnostic tool. ICE cells, which appear on a specular photomicrograph as dark, larger than normal endothelial cells, with a bright central spot, have been described as being pathognomonic for the disease.

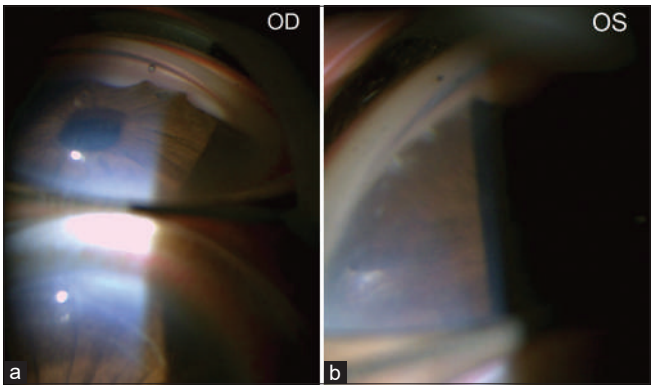


Figure 2: (a) Gonioscopy showing peripheral anterior synechiae in the right eye. (b) Peripheral anterior synechiae in the left eye

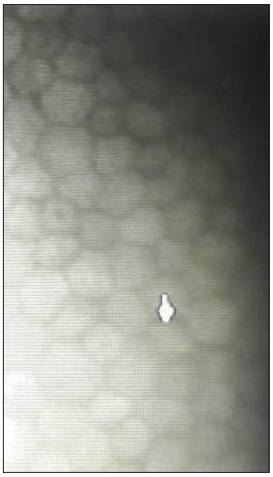


Figure 4: Specular microscopy showing iridocorneal endothelial cells

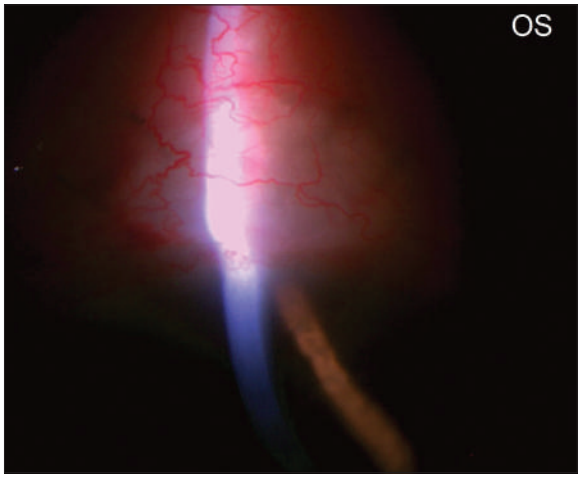


Figure 5: Functional filtering bleb seen postoperatively (left eye)

Malhotra *et al.* reported a male preponderance in his study, though only one bilateral case was reported.[1]

Sherrard *et al.*, in their series of 57 eyes with ICE syndrome, noted a lack of sex discrimination by the disease, with a male: female ratio of 47%:53%.[10]

Cases of bilateral ICE with the same variant presenting in both eyes or two different variants presenting in both eyes also have been reported.[1,4,6,8,11]

Posterior polymorphous dystrophy, iris hypoplasia, and Axenfeld–Rieger syndrome are a few of the commonly considered differential diagnoses. These can be differentiated as follows:

Posterior polymorphous dystrophy

It is corneal dystrophy, with autosomal dominant inheritance. Three patterns which are observed are endothelial vesicle-like lesions, band lesions, and diffuse deep stromal opacities. It rarely results in corneal edema or elevated IOP.

Iris hypoplasia

It is easily distinguished from ICE syndrome by the typical finding of a hypoplastic iris. It may appear like aniridia, but rudimentary iris often revealed on gonioscopy.

Axenfeld–Rieger syndrome

This is again an autosomal dominant inherited condition with similar findings to that of ICE syndrome but presents with glaucoma in early infancy or childhood.

Medical management with topical and systemic IOP-lowering agents as well as surgical management options such as glaucoma-filtering surgeries or glaucoma drainage devices may need to be tried.

In the case presented, the patient already suffered significant optic nerve head damage with depressed visual field from secondary glaucoma. He was managed with IOP-lowering agents initially to alleviate his symptoms of eye pain and corneal edema and then taken up for augmented trabeculectomy sequentially in both eyes to prevent further optic nerve head damage.

The age of presentation, male gender, and bilaterality are all atypical in this case reported which shows the possibility of such a rare presentation in ICE syndromes.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Guided by my coauthors, Dr. Padma B. Prabhu and Dr. Charmaine B. Solomon, from the Department of Ophthalmology, Government Medical College, Kozhikode.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Malhotra C, Seth NG, Pandav SS, Jain AK, Kaushik S, Gupta A, *et al*. Iridocorneal endothelial syndrome: Evaluation of patient demographics and endothelial morphology by in vivo confocal microscopy in an Indian cohort. Indian J Ophthalmol. 2019;67:604–10.
2. Iridocorneal Endothelial Syndrome and Secondary Glaucoma - EyeWiki [Internet]. Available from: https://eyewiki.org/Iridocorneal_Endothelial_Syndrome_and_Secondary_Glaucoma. [cited 2020 Feb 3].
3. Shields MB. Progressive essential iris atrophy, Chandler's syndrome, and the iris nevus (Cogan-Reese) syndrome: a spectrum of disease. Surv Ophthalmol. 1979;24:3–20.
4. Liu YK, Wang IJ, Hu FR, Hung PT, Chang HW. Clinical and specular microscopic manifestations of iridocorneal endothelial syndrome. Jpn J Ophthalmol. 2001;45:281–7.
5. Le Q-H, Sun X-H, Xu J-J. In-vivo confocal microscopy of iridocorneal endothelial syndrome. Int Ophthalmol. 2009;29:11–8.
6. Hemady RK, Patel A, Blum S, Nirankari VS. Bilateral iridocorneal endothelial syndrome: case report and review of the literature. Cornea. 1994;13:368–72.
7. Kupfer C, Kaiser-Kupfer M, Kuwabara T. Progressive bilateral essential iris atrophy. Trans Am Ophthalmol Soc. 1976;74:341–56.
8. Huna R, Barak A, Melamed S. Bilateral iridocorneal endothelial syndrome presented as Cogan-Reese and Chandler's syndrome. J Glaucoma. 1996;5:60–2.
9. Unilateral Glaucoma: Background, Pathophysiology, Epidemiology. 2019. Available from: https://emedicine.medscape.com/article/1207362-

overview. [cited 2020 Mar 3].
10. Sherrard ES, Frangoulis MA, Muir MG. On the morphology of cells of posterior cornea in the iridocorneal endothelial syndrome. Cornea. 1991;10:233–43.
11. Kaiser-Kupfer M, Kuwabara T, Kupfer C. Progressive bilateral essential iris atrophy. Am J Ophthalmol. 1977;83:340–6.

Case Report

Scleral patch graft as a rescue for infectious scleritis

ABSTRACT

We report a case of a 68-year-old male who presented with redness and pain in the right eye with a previous history of pterygium surgery with mitomycin-C in the same eye 2 weeks before onset of symptoms. On examination, visual acuity in the right eye was counting fingers at 2 m improving to 6/24 with pinhole and the left eye was 6/18 improving to 6/12 with pinhole. Right eye showed signs of melting in the nasal sclera with avascularity and perilimbal corneal infiltrate. Scraping was done and culture showed growth of Fusarium species. The patient was started on antifungal drops, and a scleral patch graft was done with multilayered amniotic membrane graft with fibrin glue. On follow-up, the patient showed symptomatic improvement with vascularization of scleral graft and vision improved following cataract surgery.

Keywords: Amniotic membrane graft, *Fusarium*, infectious scleritis, sclera patch graft, scleral melt

INTRODUCTION

Scleral melting is a well-reported complication after pterygium excision,[1] especially with the adjunctive use of mitomycin-C[2] as a result of prolonged inhibition of wound healing with its antiproliferative action. Surgically induced necrotizing scleritis has been reported to occur after trabeculectomy, cataract extraction, retinal detachment surgery, and squint surgery. It presents as a focal area of intense inflammation of the sclera, adjacent to the site of previous scleral or limbal incision.[3]

Nearly 50% of scleritis cases are associated with collagen vascular disease, and an infectious etiology is uncommon. *Pseudomonas aeruginosa* is the most common organism associated with scleral and corneal melting. However, fungi are more commonly implicated in the tropical regions of India.[4]

Reinforcement of thin or perforated sclera is necessary, to prevent prolapse of ocular contents and secondary infection. Since sclera is avascular, it is well tolerated with minimal inflammatory reaction, and also, its natural curvature allows it to blend well with host sclera. On the other hand, lack of vascularity of the sclera will cause necrosis and sloughing and results in failure of scleral homografts.[5] Therefore, an overlying epithelial cover is essential to facilitate epithelialization and vascularization of the scleral graft and incorporation into host tissue.

We report a rare case of infectious scleritis with melting; postmitomycin-C treatment and through this report, we would like to emphasize the importance of early detection and early intervention in the management of infectious scleritis.

CASE REPORT

A 68-year-old male presented with a history of redness and pain in the right eye. The clinical records of the patient showed that he had previously undergone pterygium excision

Hilda K. Nixon, Lilan Bhat, David Pudukadan, Neena Chris William

Little Flower Hospital and Research Centre, Angamaly, Kerala, India

Address for correspondence: Dr. Hilda K. Nixon,
C/o M. T Joseph, #40, Meleppuram House, Haritha Nagar,
Potta P. O., Chalakudy, Thrissur District - 680 722, Kerala, India.
E-mail: hilda24ophthal@gmail.com

Submitted: 21-Mar-2020 **Accepted:** 23-Apr-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_35_20	

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Nixon HK, Bhat L, Pudukadan D, William NC. Scleral patch graft as a rescue for infectious scleritis. Kerala J Ophthalmol 2020;32:295-8.

using adjunctive mitomycin-C in the right eye 2 weeks before onset of symptoms. the patient was on topical prednisolone acetate 1% drops 6 times per day and moxifloxacin 0.05% 4 times/day. He also underwent trabeculectomy in the left eye.

On examination, visual acuity in the right eye was counting fingers at 2 m improving to 6/24 with pinhole and the left eye was 6/18 improving to 6/12 with pinhole.

Right eye showed lid edema with mucopurulent discharge, intense congestion with engorged episcleral vessels in the nasal quadrant. Nasal sclera (previous surgical site) showed avascularity and signs of melting measuring 5 mm × 3 mm with the absence of overlying conjunctiva [Figure 1] with perilimbal corneal infiltrate adjacent to the area of thinning. Keratic precipitates were seen over endothelium along with anterior chamber cells (Grade 3) and flare seen. Lens showed nuclear sclerosis Grade 3 with posterior subcapsular cataract. Anterior vitreous phase was normal, and on fundus examination, media were hazy due to cataract. B scan of the right eye showed choroidal detachment in all quadrants with attached retina and no signs of vitritis [Figure 2]. Left eye showed well-formed bleb with moderate vascularization with patent peripheral iridotomy. Lens showed nuclear sclerosis Grade 2 with posterior subcapsular cataract. Fundus showed glaucomatous cupping with cup/disc of 0.8:1 and normal macula. Intraocular pressure was 4 mmHg in the right eye and 14 mmHg in the left eye using applanation tonometry.

The patient was admitted, gentle scraping was done, and empirical antimicrobial treatment was started. KOH showed fungal hyphae. In culture, *Fusarium* species was identified with moderate growth. The patient was admitted and started on the right eye natamycin 5% 1 hourly, voriconazole 1% 1 hourly, fluconazole 0.1% 1 hourly, amphotericin B 1 hourly, dorzolamide and timolol combination twice daily, moxifloxacin 4 times daily, and lubricants and systemic ketoconazole 200 mg twice daily. In the left eye, his topical antiglaucoma medications were continued. However, the patient showed worsening of symptoms with the progression of scleral thinning. The risk and benefits of surgical intervention were explained to the patient. Patient underwent right eye scleral patch graft and multilayered amniotic membrane graft (AMG) with fibrin glue [Figure 3].

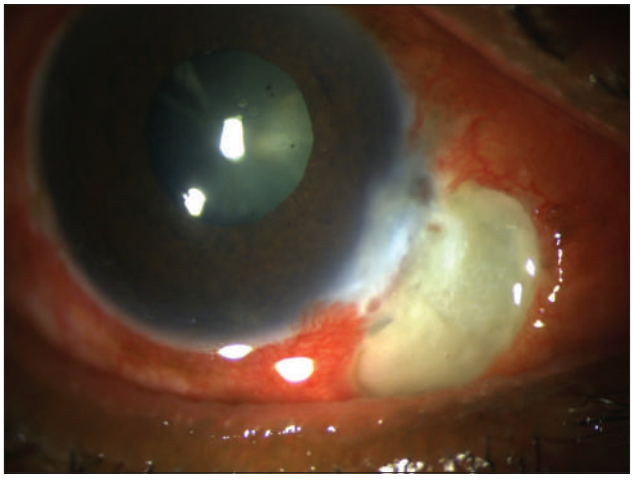


Figure 1: Scleral melt at the site of pterygium excision with perilimbal corneal infiltrate

At 1 week follow-up, the patient was symptomatically better. Corneal peripheral infiltrate decreased, and scleral patch graft was well attached. Graft edges showed signs of vascularization [Figure 4]. Overlying amniotic membrane was intact, and his medications were continued.

At 1 month of followup, corneal infiltrate resolved, with no signs of anterior uveitis. Scleral patch graft showed increased vascularity and overlying AMG slowly disintegrated. Antifungal medications were slowly tapered and stopped after 3 months.

At 6 months, the patient showed remarkable improvement with conjunctivalization of the scleral graft and scarring of the perilimbal corneal and anterior stroma [Figure 5]. There was resolution of choroidal detachment on B scan.

The follow-up examination at 1 year revealed that the scleral patch graft was successful in maintaining the integrity of the globe and cataract surgery (phacoemulsification) with intraocular lens implantation done. Postoperative best-corrected visual acuity (BCVA) vision improved to 6/9.

At 3 years of follow-up, BCVA of 6/9 was maintained in the right eye and the scleral patch graft was healthy and vascularized [Figure 6].

DISCUSSION

An infective scleritis after any ophthalmic surgery is a serious threat to vision and integrity of the globe. Infectious scleritis is often misdiagnosed and initially managed as autoimmune

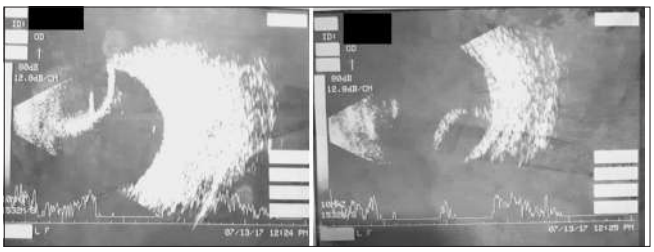


Figure 2: B scan of right eye showing choroidal detachment

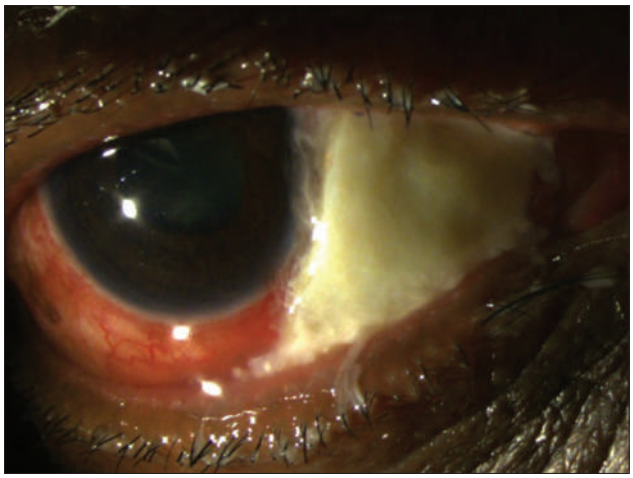


Figure 3: Scleral patch graft with overlying amniotic membrane graft

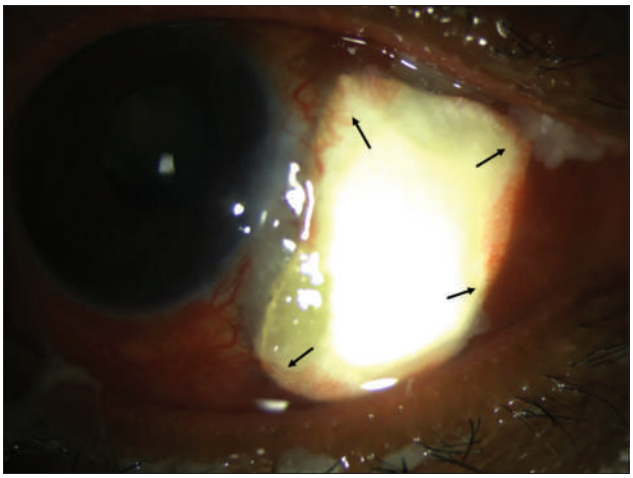


Figure 4: Scleral graft showing signs of vascularization in the periphery

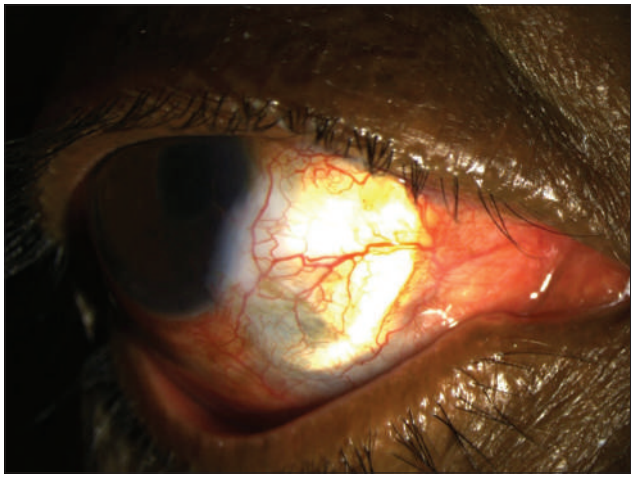


Figure 5: Total vascularization of scleral graft at 6 months

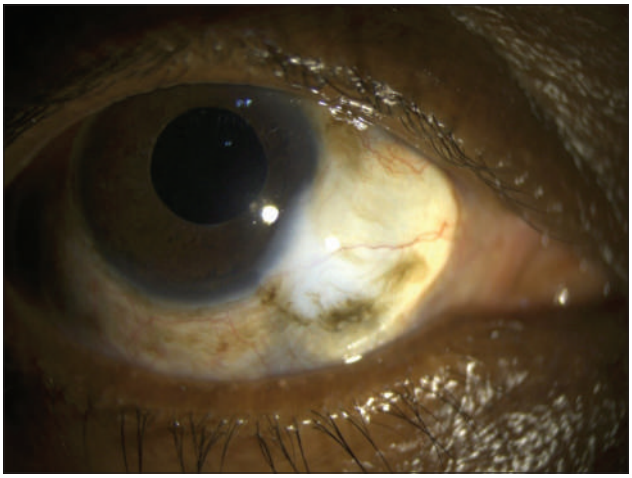


Figure 6: Scleral patch graft at 3 years of follow-up

scleritis. However, in all such cases, an underlying immune etiology should be ruled out. Thus, the overall visual outcome is worsened both due to delay in diagnosis and due to aggressive nature of associated microbes. A complete microbiological workup is needed to rule out any infectious etiology.

In our patient, culture report showed *Fusarium* species. The use of intraoperative mitomycin-C on bare sclera, aggressive cautery, and postoperative topical corticosteroids following pterygium excision could have been the cause of rapid progression of scleritis with secondary infection. Fungal scleritis responded to the treatment following surgical removal of the debris and necrotic tissue, discontinuation of corticosteroids, and aggressive antifungal therapy, both systemic and topical.

Debridement of the necrotic tissue helps in reduction of the infective load. A scleral patch graft has proven to be an effective method of closing the scleral defect.[6]

The scleral patch graft was successful in our case in restoring the anatomic integrity of the globe and facilitating visual outcome. Prior medical management with prompt surgical intervention and close follow-up is required to prevent recurrence and further devastating complications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Alsagoff Z, Tan DT, Chee SP. Necrotising scleritis after bare sclera excision of pterygium. Br J Ophthalmol 2000;84:1050-2.
2. Lu L, Xu S, Ge S, Shao C, Wang Z, Weng X, *et al*. Tailored treatment for the management of scleral necrosis following pterygium excision. Exp Ther Med 2017;13:845-50.
3. Gokhale NS, Samant R. Surgically induced necrotizing scleritis after pterygium surgery. Indian J Ophthalmol 2007;55:144-6.
4. Pradhan ZS, Jacob P. Infectious scleritis: Clinical spectrum and management outcomes in India. Indian J Ophthalmol 2013;61:590-3.
5. Lee JS, Shin MK, Park JH, Park YM, Song M. Autologous advanced tenon grafting combined with conjunctival flap in scleromalacia after pterygium excision. J Ophthalmol 2015;2015:547276.
6. Sangwan VS, Jain V, Gupta P. Structural and functional outcome of scleral patch graft. Eye (Lond) 2007;21:930-5.

Case Report

Primary conjunctival non-Hodgkin's marginal zone lymphoma

ABSTRACT

A 41-year-old male patient presented with conjunctival mass in both eyes since 5 years with no history of pain or visual disturbance and nonprogressive. On examination, bilateral, pinkish, freely mobile, nontender conjunctival mass noticed on both sides of cornea, with rest of the findings being normal. No lymphadenopathy. No history of systemic illness. Surgical excision biopsy with histopathological examination of conjunctival mass of both eyes was done. Report showed low-grade Extranodal non-Hodgkin's marginal zone lymphoma. As the isolated ophthalmic features of lymphoma is as such rare, there are high chances of misdiagnosing the condition with the other ocular conditions. Suspicious presentation requires a surgical excision biopsy.

Keywords: Conjunctiva, extranodal, immunohistochemistry, salmon pink

INTRODUCTION

Lymphoma is a malignant lymphoproliferative tumor that can involve the conjunctiva. Approximately 5%–15% of all extranodal lymphomas are found in the ocular adnexal region, with approximately 25% of those involving the conjunctiva. The most common subtype of conjunctival lymphoma is extranodal marginal zone lymphoma (EMZL) (80%).[1] Even though low-grade non-Hodgkin's marginal zone lymphoma are slow growing, and mimics other ocular conditions, it is important to diagnose the condition early to prevent the systemic spread. In our case, the mass was present since 5 years without systemic spread, which was diagnosed by excision biopsy.

CASE REPORT

A 41-year-old male patient presented with complaints of conjunctival mass in both eyes since 5 years. It was non-progressive. Negative history of pain, redness, itching, visual disturbance, trauma or any other systemic illness. On ocular examination, both eyes showed salmon pinkish mass on either side of cornea within palpebral fissure area, freely mobile, nontender with rest of the anterior segment findings being normal [Figures 1 and 2]. The posterior segment of both eyes was normal. No lymphadenopathy. Ultrasound B-scan was normal in both eyes. The patient was advised tapering dose of topical steroids, on follow-up, the condition was the same. The patient was taken for surgical excision biopsy with histopathological examination of both eyes [Figure 3]. Biopsy report showed non-Hodgkin's Lymphoma and required for immunohistochemistry for further grading. The patient was referred to systemic oncologist where tests to rule out systemic involvement were done which showed normal results. Immunohistochemistry was done, report showed positive for CD 3, CD20, CD5, Bcl 2 and negative for CD2, cyclin D1 and reported as EMZL. Patient was referred to ocular oncology center for radiation therapy, but the patient was not affordable for further treatment. Patient is been followed

Nareddy Prasanna

Department of Ophthalmology, Santhiram Medical College, Kurnool, Andhra Pradesh, India

Address for correspondence: Dr. Nareddy Prasanna, Flat No. 306, Block 4, B.K. Singh Towers, Seetharama Raju Nagar, Near K.C. Canal, Kurnool - 518 002, Andhra Pradesh, India.
E-mail: prasannan.08@gmail.com

Submitted: 31-Mar-2020 **Accepted:** 13-Apr-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_37_20	

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Prasanna N. Primary conjunctival non-Hodgkin's marginal zone lymphoma. Kerala J Ophthalmol 2020;32:299-301.

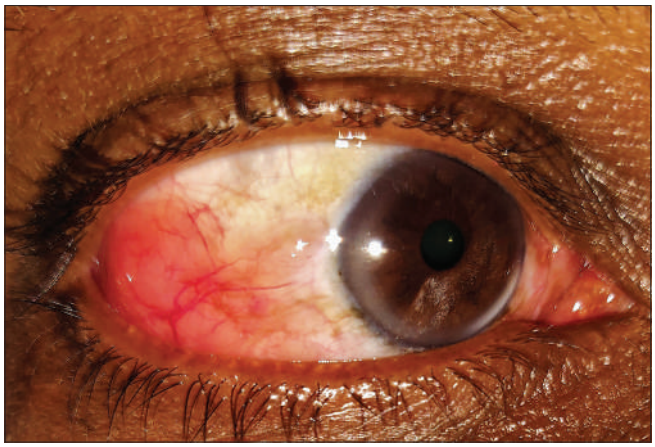


Figure 1: Right eye-pinkish subconjunctival mass temporally

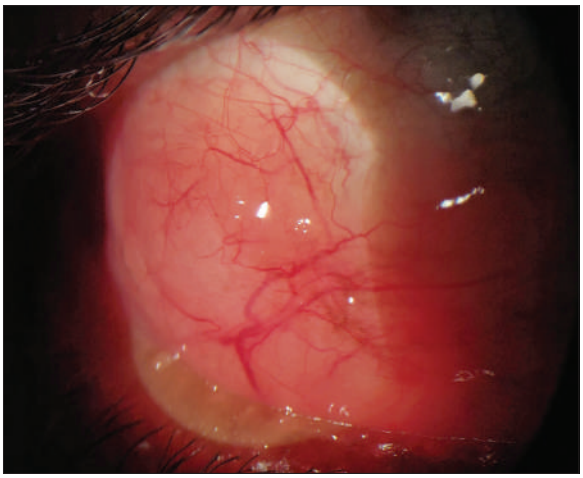


Figure 2: Localized subconjunctival mass, freely mobile, nontender

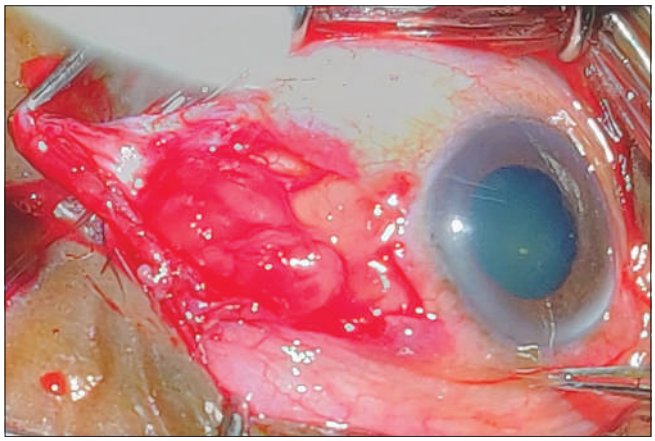


Figure 3: Intraoperative excision of subconjunctival mass by delineating the surrounding structures

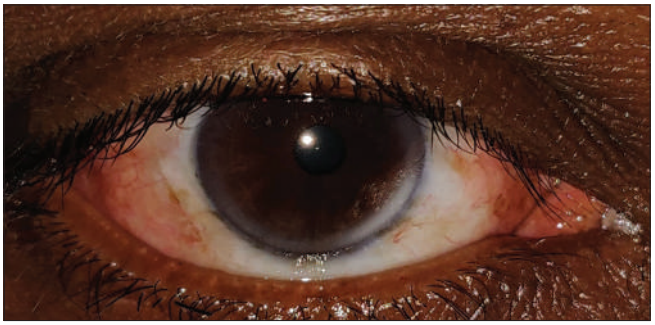


Figure 4: Eye during follow-up period

since 1 year and shows no signs of recurrence and yet to be followed further [Figure 4].

DISCUSSION

The etiology of conjunctival lymphoma remains elusive. Conjunctival lymphoma appears to develop as a result of chronic inflammation triggered by endogenous or exogenous antigens, leading to sustained proliferation that permits the potential for genetic mutations leading to eventual monoclonal B or T lymphocyte populations.[1] Although low-grade conjunctival lymphomas tends to be slow-growing, unilateral and indolent, systemic metastasis is seen in approximately 20% of patients; risk of metastasis increases in patients with a bilateral presentation.[2] While conjunctival lymphoma is primarily seen in the elderly, it has been described in individuals as young as 33 months. Chronic antigenic stimulation from pathogens including *Helicobacter pylori*, Chlamydia psittaci, and hepatitis C has been associated with conjunctival EMZL. Autoimmune disorders such as Sjögren syndrome, systemic lupus erythematosus, and Hashimoto thyroiditis have also been implicated in the pathogenesis of conjunctival EMZL.[2] In our case, where lymphoma is confined to the conjunctiva which is a rare presentation, there are high chances of misdiagnosing the condition with other ocular conditions such as benign lymphoid hyperplasia, nodular episcleritis, and delay in the diagnosis may lead to systemic spread, it is important to have a thorough slit lamp examination and warrants surgical excision biopsy with histopathological examination for definitive diagnosis. And detailed systemic evaluation to rule out metastasis. 5 year survival rate of lymphoma depends mainly on subtype which ranges from 97% (extranodal marginal zone Bcell lymphoma) to 9% (mantle cell lymphoma).[3] Overall, prognosis is generally good, with 90% of patients not experiencing progression or recurrence during 1 year follow-up period.[4] Low-grade lymphomas have the lowest progression, followed by higher grade diffuse large B-cell and mantle cell lymphomas.[5]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. The patient has given his consent for his images and other clinical information to be reported in the journal.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Tanenbaum RE, Galor A, Dubovy SR, Karp CL. Classification, diagnosis, and management of conjunctival lymphoma. Eye Vis (Lond) 2019;6:22.
2. Yumori JW, Jakirlic N, Van K. Conjunctival lymphoma: A teaching case report. Optom Educ 2019;44:3.
3. Available from: https://eyewiki.aao.org/Conjunctival_lymphoma. [Last accessed on 2020 Jan 08].
4. Kirkegaard MM, Coupland SE, Prause JU, Heegaard S. Malignant lymphoma of the conjunctiva. Surv Ophthalmol 2015;60:444-58.
5. Baldini L, Blini M, Guffanti A, Fossati V, Colombi M, La Targia ML, *et al*. Treatment and prognosis in a series of primary extranodal lymphomas of the ocular adnexa. Ann Oncol 1998;9:779-81.

Photo Essay

A challenging case of refractory childhood glaucoma associated with bilateral congenital ectropion uveae

ABSTRACT

Congenital ectropion uveae (CEU) is a rare disorder, resulting from the proliferation of iris pigment epithelium on the anterior surface of the iris. We report the case of a 3-year-old boy, who was recently diagnosed with glaucoma, on treatment with two topical antiglaucoma medications and referred to us for further evaluation and management. His visual acuity, at presentation, was 6/7.5 (Cardiff) in both his eyes. Slit-lamp examination of both his eyes revealed the presence of ectropion uveae, with intraocular pressure (IOP) of 32 mm Hg. Gonioscopy in both eyes showed anterior insertion of iris. On Fundoscopy, both eyes showed glaucomatous disc changes. He had no systemic associations. Owing to inadequate IOP control, despite optimal medical therapy, he underwent trabeculectomy with intra-operative application of Mitomycin-C. However, the IOP reduction post-trabeculectomy was inadequate, and he subsequently required a bleb needling, which also failed. Hence, he underwent Ahmed glaucoma valve implantation in his right eye, with a favorable final outcome. IOP in the right eye was brought under control following multiple surgical interventions and additional medical treatment. CEU in children can be associated with refractory childhood glaucoma. It is important to evaluate for glaucoma in CEU cases to prevent blindness, especially in young patients and improve their overall prognosis.

Keywords: Ahmed glaucoma valve, bilateral congenital ectropion uveae, refractory childhood glaucoma

INTRODUCTION

Congenital ectropion uveae (CEU) is a rare, nonprogressive neural crest cells' disorder resulting from proliferation of iris pigment epithelium on the anterior surface of the iris.[1,2] Usually unilateral, the condition is frequently associated with ipsilateral glaucoma due to iridotrabecular dysgenesis.[1-4] CEU is reported with various systemic associations of which neurofibromatosis is the most common disorder.[1] Filtering surgery often becomes the effective treatment modality for necessary reduction of intraocular pressure (IOP).[3-5] This report describes a rare and challenging case of bilateral CEU presenting in early childhood with unilateral refractory glaucoma without any associated systemic disease.

CASE REPORT

A 3-year-old male child was referred for glaucoma management. He was on topical Dorzolamide and Timolol fixed drug combination eye drops for elevated IOP in both eyes.

On examination, visual acuity was 6/7.5 both eyes (Cardiff). There were few prominent nerves noticed in otherwise clear cornea with normal cell morphology in both eyes. Anterior chamber (AC) was normal in depth in both eyes. Iris showed a loss of normal pattern with ectropion uveae present in the entire 360° around pupillary border in the right eye and around 270°–300° in the left eye sparing inferonasal quadrant. This ectropion uveae extended to the mid periphery of the iris and was separated from an otherwise smooth and cryptless, atrophic rest of the iris by a definite demarcation border. IOP measured 32 mm Hg in both eyes. Both pupils were circular with right pupil more

Submitted: 09-May-2020 **Revised:** 03-Jun-2020
Accepted: 06-Jun-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_54_20	

V. K. Harshavardhan, Meena Menon
Glaucoma Services, Sankara Eye Hospital, Bengaluru, Karnataka, India

Address for correspondence: Dr. Meena Menon, Sankara Eye Hospital, Bengaluru, Karnataka, India.
E-mail: meenagm@gmail.com

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Harshavardhan VK, Menon M. A challenging case of refractory childhood glaucoma associated with bilateral congenital ectropion uveae. Kerala J Ophthalmol 2020;32:302-4.

dilated than the left pupil. Pupillary reaction was sluggish in both eyes [Figures 1 and 2]. Gonioscopy revealed anterior insertion of iris over trabecular meshwork in both eyes. On fundoscopy, the cup-disc ratio was 0.8:1 with neuroretinal rim thinning superiorly and inferiorly in the right eye. In left eye, cup-disc ratio was 0.6:1 with vertically oval cup with early neuroretinal rim thinning inferiorly. Other fundus details were within normal limits in both eyes. There were no signs of any systemic association.

Initially, the patient was advised to commence additional topical Travoprost eyedrops in both eyes along with Dorzolamide and Timolol eyedrops. The IOP was controlled in the left eye. However, IOP was still high in the right eye (28 mm Hg) for which he underwent Trabeculectomy surgery with intraoperative application of 0.4 mg/mL Mitomycin C for 2 minutes. The supero-nasal triangular trabeculectomy flap was closed with single apical suture [Figure 3]. Postoperative medications included tapering doses of topical Prednisolone acetate 1% for 5 weeks, topical Moxifloxacin 0.5%, and Homatropine hydrobromide for 2 weeks each.

The IOP was still uncontrolled in the right eye for which he underwent bleb needling along with subconjunctival injection of 0.1 mL of 5-fluorouracil, 7 months posttrabeculectomy. This procedure failed to bring the IOP down to satisfactory level. The patient further underwent Ahmed glaucoma valve (AGV-FP8 model New World Medical, Rancho Cucamonga, CA, USA) implantation with tube entry into anterior chamber, 3 months after bleb needling procedure [Figure 4]. The procedureinvolved corneal traction suture placement, securing the implant with in supero-temporal quadrant using two 9-0 Nylon sutures and covering the tube with donor scleral patch graft. Postoperative medications included tapering doses of topical Prednisolone acetate 1% for 5 weeks, topical Moxifloxacin 0.5%, and Homatropine hydrobromide for 2 weeks each. At the last follow up, 1 year after AGV implantation, IOP in the right eye was under control with additional medical treatment.

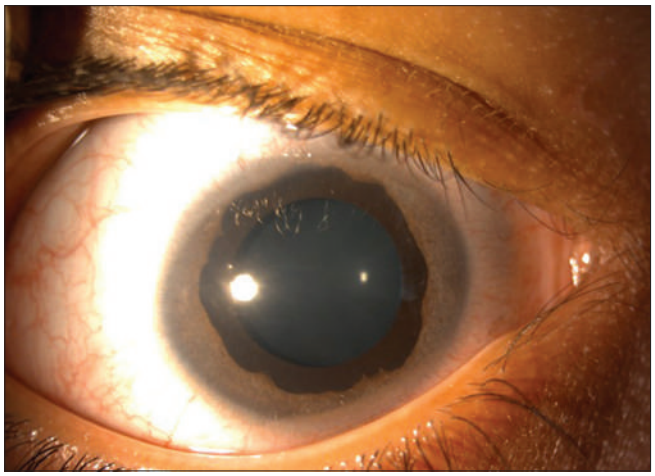


Figure 1: Slit-lamp photograph of the right eye

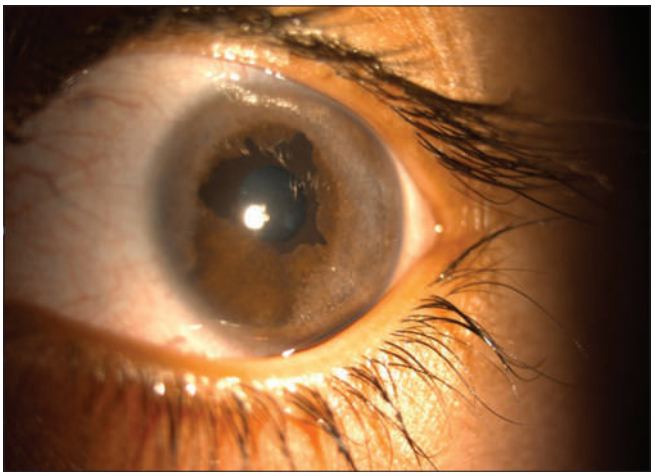


Figure 2: Slit-lamp photograph of the left eye

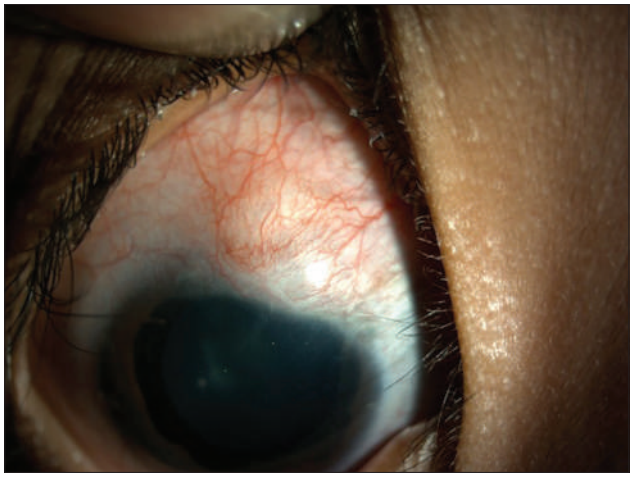


Figure 3: Filtering bleb in the right eye

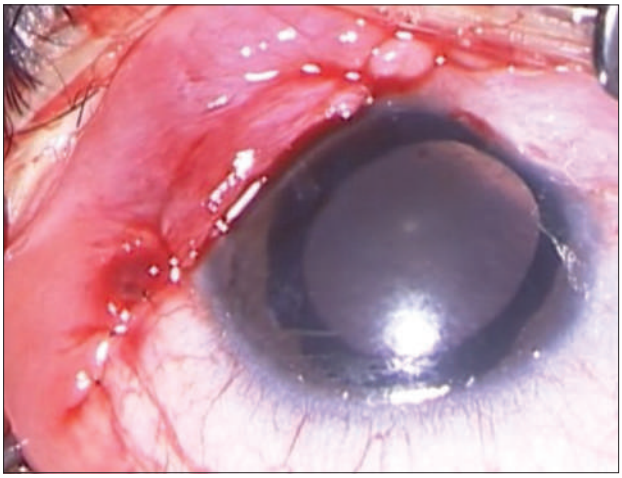


Figure 4: Ahmed glaucoma valve implantation in the right eye with tube entry into anterior chamber. Distal end of tube is seen superotemparally

DISCUSSION

CEU is a disorder resulting from proliferation of iris pigment epithelium on the anterior surface of iris. CEU can present either in early childhood or in adolescence.[1,2,6] CEU is unilateral in most of the cases, and there are only few bilateral cases described in the literature.[4,5] Ritch *et al*.[1] found glaucoma in seven out of eight cases of CEU and Dowling *et al*.[2] in eight of nine cases, highlighting the frequent association of glaucoma with CEU. It is important to evaluate all the patients with CEU for glaucoma. CEU is also associated with prominent corneal nerves.[5,6] The most common association is neurofibromatosis, though other ocular and systemic associations such as facial hemihypertropy, Prader-Willi syndrome, chromosomal abnormalities, and Rieger's syndrome have been described.[1]

In this case, there was a bilateral CEU, prominent corneal nerves and glaucoma, one eye severely affected than the other. The severely affected eye was particularly refractory to treat. The greater extent of CEU may also be the indicator of poor prognosis. There was no systemic association found.

Glaucoma in CEU may be in part due to developmental abnormalities of the angle structures as well as progressive angle closure secondary to abnormal endothelial cell membranes.[7] Most CEU cases only temporarily respond to medical management often necessitating surgical treatment.[2-5] Wang *et al*.[7] performed glaucoma drainage device surgery in their series of cases and noted early encapsulation of the plates which often required additional glaucoma surgery (cycloablation or trabeculectomy with mitomycin C) within 4 months of tube implantation.

In this case, the better eye was treated satisfactorily with medical management alone, whereas the severely affected eye was associated with relatively refractory glaucoma which required multiple intraocular surgeries. Target IOP was achieved in the right eye after multiple surgeries along with additional medical treatment and is maintaining the same at the last follow-up, 1 year after the last surgery.

CONCLUSION

CEU is an ocular abnormality, often missed especially when it is in milder form. However, its frequent association with glaucoma calls for a careful examination and early surgical intervention if required, to preserve good vision. This rare form of refractory glaucoma may require multiple surgeries and warrants close monitoring of intraocular pressures.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Ritch R, Forbes M, Hetherington J Jr., Harrison R, Podos SM. Congenital ectropion uveae with glaucoma. Ophthalmology 1984;91:326-31.
2. Dowling JL Jr., Albert DM, Nelson LB, Walton DS. Primary glaucoma associated with iridotrabecular dysgenesis and ectropion uveae. Ophthalmology 1985;92:912-21.
3. Bansal A, Luck J. Primary iris pigment epithelial hyperplasia and glaucoma. Br J Ophthalmol 2002;86:352-3.
4. Hertzberg R. Congenital ectropion uveae and glaucoma. Aust N Z J Ophthalmol 1985;13:45-8.
5. Sethi H, Pal N, Dada T. Bilateral juvenile glaucoma with iridotrabecular dysgenesis, congenital ectropion uveae, and thickened corneal nerves. Eye 2005;19:1347-9.
6. Mandal AK. Late-onset unilateral primary developmental glaucoma associated with iridotrabecular dysgenesis, congenital ectropion uveae and thickened corneal nerves: A new neural crest syndrome? Ophthalmic Surg Lasers 1999;30:567-70.
7. Wang GM, Thuente D, Bohnsack BL. Angle closure glaucoma in congenital ectropion uvea. Am J Ophthalmol Case Rep 2018;10:215-20.

Eye grader-smartphone app for clinical grading scales in ophthalmology

ABSTRACT

Clinical grading scales help us to objectively document clinical findings. Even though there are several good clinical grading scales, it is not feasible to memorize all of them. It would be very convenient to have all this information at your fingertips. Noting the absence of any existing app, the authors programmed a smartphone app with all the necessary grading systems. This app should be useful for all ophthalmic clinicians to quickly lookup these grading systems and helps in objective documentation.

Keywords: Clinical grading, clinical scales, eye grader, ophthalmic app, smartphone app

INTRODUCTION

Clinical grades and scales help in the standardization of documentation and have several advantages.[1] They are essential in measuring, recording, and monitoring changes in clinical signs. These help in objective documentation of the magnitude or severity of clinical findings. Most of them are numeric grades or scales and have four or five-step scales of severity. Bailey *et al*. discussed how qualitative and quantitative grading systems can be improved with finer scaling to enhance the ability to detect smaller degrees of change.[1]

There are several clinical signs in ophthalmology and several grading systems for each of them. It is practically impossible to memorize all the grading systems, and this difficulty leads to the poor utility of these otherwise useful clinical instruments. Often, grading scales are ignored, or approximate measures are used, and sometimes, they are misremembered or wrongly interpreted. This leads to confusion in the documentation, and a lot of valuable clinical information is lost, especially when communicating between multiple clinicians.

Smartphones are now ubiquitous and have been driving innovation in several fields, including ophthalmology.[2] We had previously discussed several innovative apps for ophthalmologists[3] and for visually impaired persons.[4] We decided to make a smartphone app, "Eye Grader"[5] which provides and offline, searchable database of clinical grading scales which can be readily accessed by a busy ophthalmologist, optometrist or eye care personnel so that objective documentation of the clinical findings can be easily done and quickly interpreted.

CLINICAL GRADING SCALE SYSTEMS

Proper use of grading scales improves the standard of record-keeping which results in improved medicolegal evidence. It helps in standardization, which leads to reduced

Submitted: 02-Nov-2020 **Accepted:** 03-Nov-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_176_20	

John Davis Akkara[1,2,3], Anju Kuriakose[4,5]

[1]Department of Ophthalmology, Sri Ramachandra Institute of Higher Education and Research, Departments of [4]Ophthalmology and [5]Retina, Aravind Eye Hospital, Chennai, Tamil Nadu, [2]Department of Glaucoma, Westend Eye Hospital, Cochin, [3]Department of Glaucoma, Little Flower Hospital, Angamaly, Kerala, India

Address for correspondence: Dr. John Davis Akkara, Department of Glaucoma, Westend Eye Hospital, Chitoor Road, Kacheripady, Cochin - 682 018, Kerala, India.
E-mail: johndavisakkara@gmail.com

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Akkara JD, Kuriakose A. Eye grader-smartphone app for clinical grading scales in ophthalmology. Kerala J Ophthalmol 2020;32:305-10.

inter- and intraobserver variability. This is especially relevant when patient care is by multiple clinicians. It helps in precise monitoring and earlier detection of disease progression and the effectiveness of interventions. Last but not least, it helps in better doctor-patient communication of the disease status.

Several studies have looked at clinical grading scales and their reliability. Chong *et al*. evaluated Verbal Descriptor Scales, Photographic Matching Scale, and Continuous Matching Scale for anterior segment findings such as conjunctival staining and bulbar redness and found good reliability.[6] Schulze *et al*. noted that perceived redness is based on chromaticity and vessel-based components.[7] Later, they investigated the effect of reference anchors in arranging images of bulbar redness.[8] They also investigated cross-calibrated reference scales for bulbar redness.[9] Efron proposed grading scales for the severity of eight common complications of contact lens wear.[10] Efron *et al*. also validated scales for contact lens complications.[11] Murphy *et al*. investigated interobserver agreement of Cornea and Contact Lens Research Unit grading scale for bulbar conjunctival hyperemia.[12] MacKinven *et al*. studied a modified scale with decimal increments for palpebral conjunctiva redness and roughness with good results on the agreement.[13] Park *et al*. established that image analysis with the Contrast-Limited Adaptive Histogram Equalization algorithm was useful for objectively grading conjunctival redness compared to three other methods they tested.[14] Wolffsohn used computer image analysis on printed grading scales of the anterior segment and identified that increments followed a quadratic than a linear function and that image analysis was up to 35 times more repeatable that subjective grading.[15] Rahman *et al*. published their work on training field graders for trachoma using conjunctival photographs.[16]

Huang *et al*. validated a new pterygium grading scale with five parameters for research and clinical trials.[17] Bonini *et al*. proposed a clinical grading system for vernal keratoconjunctivitis based on the severity of disease.[18] Zhong *et al*. proposed a grading system for limbal dermoid with postoperative prognostic value.[19] Ong *et al*. validated a semi-quantitative tool for clinical assessment of cicatrizing conjunctivitis.[20]

Sook Chun and Park studied the reliability of four grading systems for corneal staining and noted that the National Eye

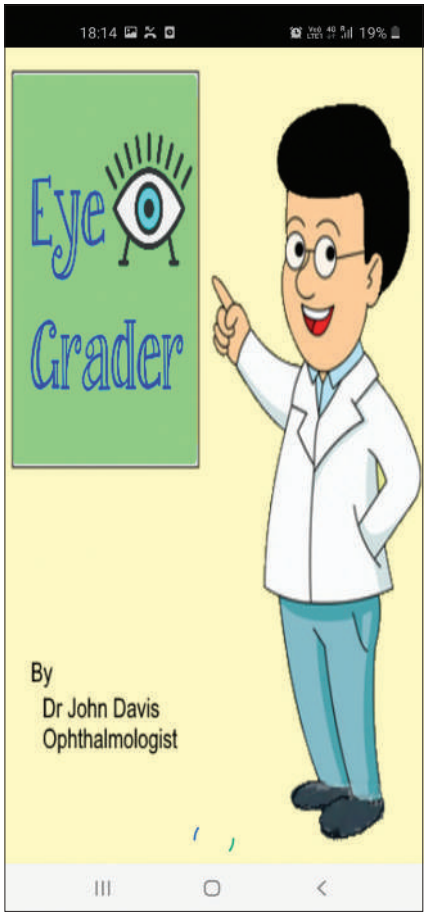


Figure 1: Splash screen of eye grader showing the logo

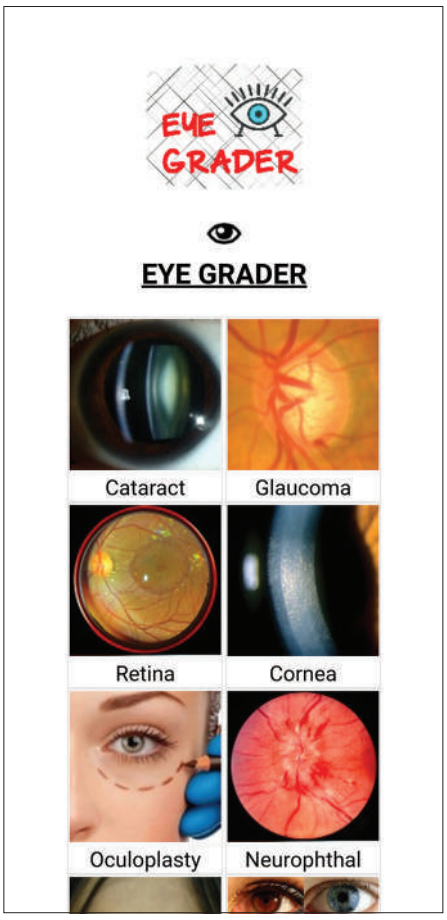


Figure 2: Page showing the grading systems grouped according to sub-specialties

Institute system had the best reliability and repeatability.[21] Dundas *et al*. used a photographic grading scale for corneal staining and noted low interobserver variability and suggested that decimal increments can be used.[22]

Bell *et al*. evaluated the grading of Relative Afferent Pupillary Defect (RAPD) by swinging flashlight test and compared with neutral density filters and found good correlation.[23] Yotharak and Aui-Aree also found good correlation between the similar clinical plus scale for RAPD (used in Thailand) with neutral density filters.[24] They suggest that the swinging flashlight grading can be converted into log units if necessary.

Taylor and West proposed a system for slit lamp grading of lens opacities with standard photographs.[25] Gali *et al*. published a comprehensive summary of cataract grading systems[26], including the use of deep learning and artificial intelligence.[27]

Hornbeak *et al*. evaluated a new 9-step scale for vitreous haze compared to the existing 6-step scale.[28] Xu *et al*. proposed and validated a clinical grading system for the experimental autoimmune uveitis animal model for laboratories.[29] Seddon *et al*. validated a 5-level clinical classification system for age-related maculopathy (ARM) known as the clinical ARM staging system.[30] Sliesoraityte *et al*. proposed a grading system for optical coherence tomography scans for cystic macular lesions in usher syndrome patients.[31] Ohno-Matsui *et al*. proposed a classification for myopic Maculopathy.[32] Bhardwaj *et al*. proposed a grading system for retinal hemorrhages in abusive head trauma.[33] Lee *et al*. proposed a grading system for Sunset Glow Fundus in Vogt-Koyanagi-Harada disease.[34] Gangaputra *et al*. from the Fundus Photograph Reading Center at University of Wisconsin compared standardized clinical classification with fundus photograph reading for diabetic retinopathy and diabetic macular edema.[35]

Wells *et al*. evaluated the Moorfields bleb grading system and the Indiana bleb appearance grading scale and noted good clinical reproducibility and minor deficiencies, which could be improved.[36] Bouremel *et al*. mathematically modeled blebs using bleb height, area, and pressure measured in rabbit eyes and other *in vitro* methods.[37]

Barrio-Barrio *et al*. compared vision, inflammation, strabismus, and appearance (VISA), and the European Group of Graves' Orbitopathy classifications for Graves' Ophthalmopathy.[38]

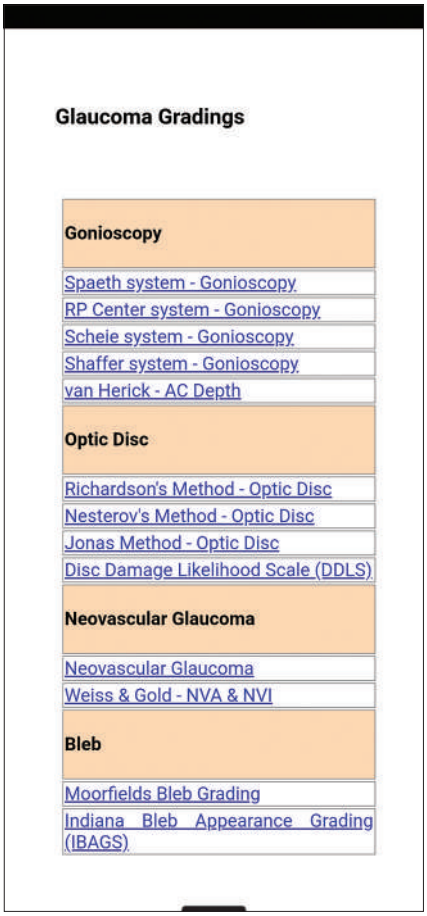


Figure 3: Page showing clickable list of grading systems in glaucoma

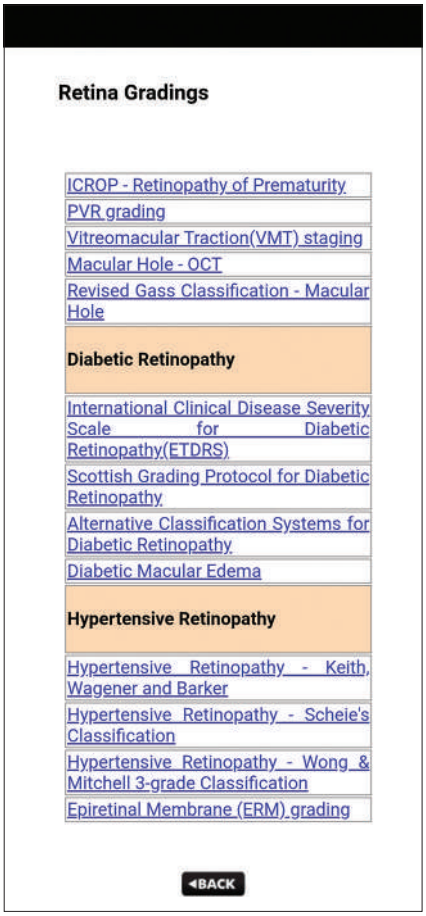


Figure 4: Page showing clickable list of grading systems in the retina

Peter Dolman reviewed the methods of grading severity of thyroid eye disease focusing on VISA.[39]

MAKING OF "EYE GRADER" APP

With the help of ophthalmology postgraduate residents preparing for their examinations, the authors collected several clinical grading scales most relevant to practicing ophthalmologists and formatted the data for uniformity and accuracy. Using the app-building tool PhoneGap, the authors compiled the collected information into an Android-based smartphone App "Eye Grader"[5] and uploaded it on the Google Play store. The iPhone version was delayed due to the higher cost involved. As of October 2020, the app had around 3000 downloads from around 80 countries, with the maximum users being from India, the USA, Mexico, Egypt, and Jordan. Any suggestions for improving the app can be sent to the author, preferably at eyedoctorengineer@gmail.com.

USING "EYE GRADER" APP

The "Eye Grader" App can be searched for and downloaded from the Google Play store [Figure 1]. (https://play.google.com/store/apps/details?id = com.fundazone.eyegrader). When opening the app, it opens to a page with sections for Cataract, Glaucoma, Retina, Cornea, Oculoplasty, Neuro-ophthalmology, Squint, and Others [Figure 2]. These sections have the corresponding grading systems related to the sub-specialty [Figures 3 and 4]. In addition, there is a search box where you can type and search for any word in the name of the grading scale [Figure 5]. On opening a particular grading scale, it shows the grading system along with any images where needed [Figures 6 and 7].

CONCLUSION

The eye grader app is a free tool for ophthalmologists to help in objective documentation of clinical signs using standardized clinical grading scales. Validation of the usefulness and widespread use of this app would allow better communication between ophthalmic personnel of patient's clinical condition.

Acknowledgment

The author would like thank to Dr Jyothi Rajasekhar and other ophthalmology postgraduate students at little

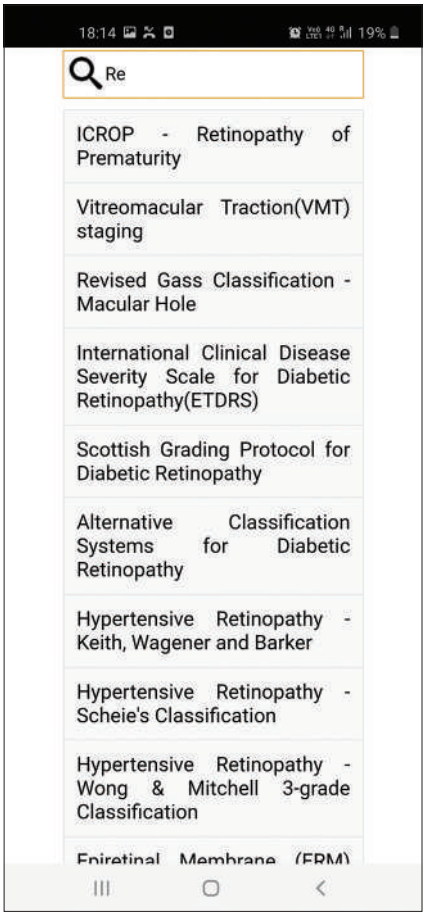


Figure 5: Search box being used

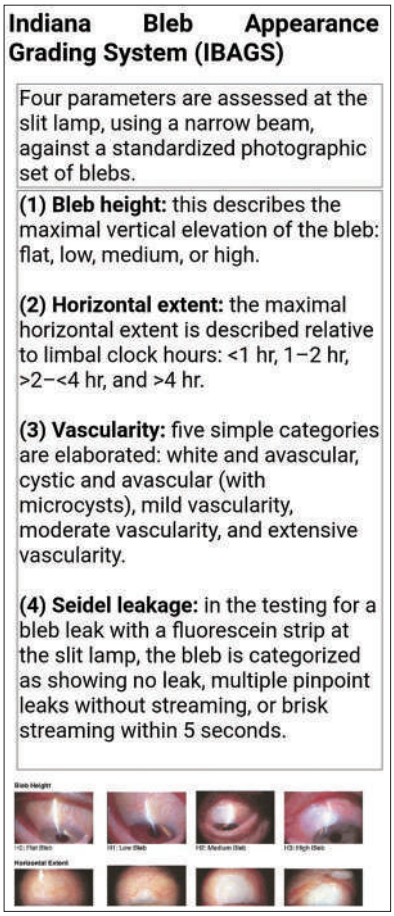


Figure 6: Page showing Indiana bleb appearance grading system

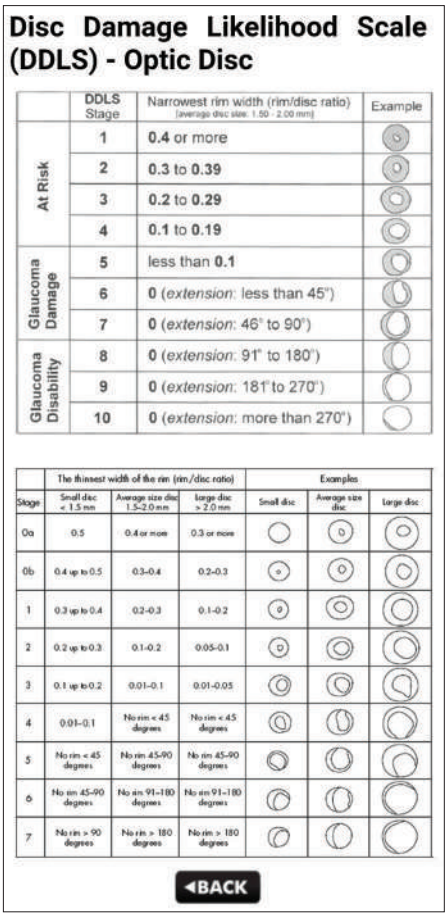


Figure 7: Page showing disc damage likelihood score

flower hospital and research centre, Angamaly, Kerala, for collecting the grading scale information that was used for creating the eye grader app and for expressing the need for such an app.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Bailey IL, Bullimore MA, Raasch TW, Taylor HR. Clinical grading and the effects of scaling. Invest Ophthalmol Vis Sci 1991;32:422-32.
2. Akkara JD. Commentary: Dawn of smartphones in frugal ophthalmic innovation. Indian J Ophthalmol 2018;66:1619.
3. Akkara J, Kuriakose A. Innovative smartphone apps for ophthalmologists. Kerala J Ophthalmol 2018;30:138-44.
4. Akkara J, Kuriakose A. Smartphone apps for visually impaired persons. Kerala J Ophthalmol 2019;31:242-8.
5. Eye Grader – Apps on Google Play. Available from: https://play.google.com/store/apps/details?id=com.fundazone.eyegrader&hl=en_IN. [Last acessed on 2020 May 12].
6. Chong T, Simpson T, Fonn D. The repeatability of discrete and continuous anterior segment grading scales. Optom Vis Sci 2000;77:244-51.
7. Schulze MM, Hutchings N, Simpson TL. The perceived bulbar redness of clinical grading scales. Optom Vis Sci 2009;86:E1250-8.
8. Schulze MM, Hutchings N, Simpson TL. The conversion of bulbar redness grades using psychophysical scaling. Optom Vis Sci 2010;87:159-67.
9. Schulze MM, Hutchings N, Simpson TL. Grading bulbar redness using cross-calibrated clinical grading scales. Invest Ophthalmol Vis Sci 2011;52:5812-7.
10. Efron N. Grading scales for contact lens complications. Ophthalmic Physiol Opt 1998;18:182-6.
11. Efron N, Morgan PB, Katsara SS. Validation of grading scales for contact lens complications. Ophthalmic Physiol Opt 2001;21:17-29.
12. Murphy PJ, Lau JS, Sim MM, Woods RL. How red is a white eye? Clinical grading of normal conjunctival hyperaemia. Eye (Lond) 2007;21:633-8.
13. MacKinven J, McGuinness CL, Pascal E, Woods RL. Clinical grading of the upper palpebral conjunctiva of non-contact lens wearers. Optom Vis Sci 2001;78:13-8.
14. Park IK, Chun YS, Kim KG, Yang HK, Hwang JM. New clinical grading scales and objective measurement for conjunctival injection. Invest Ophthalmol Vis Sci 2013;54:5249-57.
15. Wolffsohn JS. Incremental nature of anterior eye grading scales determined by objective image analysis. Br J Ophthalmol 2004;88:1434-8.
16. Rahman SA, Yu SN, Amza A, Gebreselassie S, Kadri B, Baido N, *et al*. Reliability of trachoma clinical grading Assessing grading of marginal cases. PLoS Negl Trop Dis 2014;8:e2840.
17. Huang P, Huang J, Tepelus T, Maram J, Sadda S, Lee OL. Validity of a new comprehensive pterygia grading scale for use in clinical research and clinical trial. Int Ophthalmol 2018;38:2303-11.
18. Bonini S, Sacchetti M, Mantelli F, Lambiase A. Clinical grading of vernal keratoconjunctivitis. Curr Opin Allergy Clin Immunol 2007;7:436-41.
19. Zhong J, Deng Y, Zhang P, Li S, Huang H, Wang B, *et al*. New grading system for limbal dermoid: A retrospective analysis of 261 cases over a 10-year period. Cornea 2018;37:66-71.
20. Ong HS, Minassian D, Rauz S, Mehta JS, Dart JK. Validation of a clinical assessment tool for cicatrising conjunctivitis. Ocul Surf 2020;18:121-9.
21. Sook Chun Y, Park IK. Reliability of 4 clinical grading systems for corneal staining. Am J Ophthalmol 2014;157:1097-102.
22. Dundas M, Walker A, Woods RL. Clinical grading of corneal staining of non-contact lens wearers. Ophthalmic Physiol Opt 2001;21:30-5.
23. Bell RA, Waggoner PM, Boyd WM, Akers RE, Yee CE. Clinical grading of relative afferent pupillary defects. Arch Ophthalmol 1993;111:938-42.
24. Yotharak P, Aui-Aree N. Correlation between clinical grading and quantification by neutral density filter of relative afferent pupillary defect (RAPD). J Med Assoc Thail Chotmaihet Thangphaet 2012;95 Suppl 4:S92-5.
25. Taylor HR, West SK. The clinical grading of lens opacities. Aust N Z J Ophthalmol 1989;17:81-6.
26. Gali HE, Sella R, Afshari NA. Cataract grading systems: A review of past and present. Curr Opin Ophthalmol 2019;30:13-8.
27. Akkara JD, Kuriakose A. Role of artificial intelligence and machine learning in ophthalmology. Kerala J Ophthalmol 2019;31:150.
28. Hornbeak DM, Payal A, Pistilli M, Biswas J, Ganesh SK, Gupta V, *et al*. Interobserver agreement in clinical grading of vitreous haze using alternative grading scales. Ophthalmology 2014;121:1643-8.
29. Xu H, Koch P, Chen M, Lau A, Reid DM, Forrester JV. A clinical grading system for retinal inflammation in the chronic model of experimental autoimmune uveoretinitis using digital fundus images. Exp Eye Res 2008;87:319-26.
30. Seddon JM, Sharma S, Adelman RA. Evaluation of the clinical age-related maculopathy staging system. Ophthalmology 2006;113:260-6.
31. Sliesoraityte I, Peto T, Mohand-Said S, Sahel JA. Novel grading system for quantification of cystic macular lesions in Usher syndrome. Orphanet J Rare Dis 2015;10:157.

32. Ohno-Matsui K, Kawasaki R, Jonas JB, Cheung CM, Saw SM, Verhoeven VJ, *et al*. International photographic classification and grading system for myopic maculopathy. Am J Ophthalmol 2015;159:877-83.e7.
33. Bhardwaj G, Jacobs MB, Martin FJ, Donaldson C, Moran KT, Vollmer-Conna U, *et al*. Grading system for retinal hemorrhages in abusive head trauma: Clinical description and reliability study. J AAPOS 2014;18:523-8.
34. Lee EK, Lee SY, Yu HG. A clinical grading system based on ultra-wide field retinal imaging for sunset glow fundus in Vogt-Koyanagi-Harada disease. Graefes Arch Clin Exp Ophthalmol 2015;253:359-68.
35. Gangaputra S, Lovato JF, Hubbard L, Davis MD, Esser BA, Ambrosius WT, *et al*. Comparison of standardized clinical classification with fundus photograph grading for the assessment of diabetic retinopathy and diabetic macular edema severity. Retina 2013;33:1393-9.
36. Wells AP, Ashraff NN, Hall RC, Purdie G. Comparison of two clinical Bleb grading systems. Ophthalmology 2006;113:77-83.
37. Bouremel Y, Lee RMH, Eames I, Brocchini S, Khaw PT. Novel approaches to model effects of subconjunctival blebs on flow pressure to improve clinical grading systems after glaucoma drainage surgery. PLoS One 2019;14:e0221715.
38. Barrio-Barrio J, Sabater AL, Bonet-Farriol E, Velázquez-Villoria Á, Galofré JC. Graves' Ophthalmopathy: VISA versus EUGOGO classification, assessment, and management. J Ophthalmol 2015;2015:249125.
39. Dolman PJ. Grading severity and activity in thyroid eye disease. Ophthal Plast Reconstr Surg 2018;34 4S Suppl 1:S34-40.

Unilateral abducent nerve palsy in a diabetic patient with treated cerebellopontine angle tumor

ABSTRACT

Isolated abducent nerve palsy in a diabetic patient with a history of treatment for cerebellopontine angle tumor poses challenges with regard to clinical diagnosis and management. This case discusses the challenges in clinching the diagnosis clinically, the need for imaging, and the guidelines for management.

Keywords: Cerebellopontine angle tumor, cerebellopontine angle meningioma, diabetic mononeuropathy, meningioma recurrence, unilateral abducent nerve palsy

INTRODUCTION

Cerebellopontine angle (CPA) tumors are usually slow-growing masses and are diagnosed based on the characteristic symptomatology and brain imaging findings. Mononeuropathy presenting as unilateral abducent nerve palsy is a relatively common occurrence in long-standing diabetes mellitus. In a scenario in which a diabetic patient with a history of treatment for CPA tumor presents with unilateral abducent nerve palsy, clinical decision-making becomes difficult. This clinical query section discusses pertinent points regarding the diagnosis, brain imaging, and line of management in one such case.

PANELISTS

1. Dr. M V Francis, Chief Consultant, Teresa Eye and Migraine Center, Cherthala, Alleppey, Kerala
2. Dr. Thomas Arun Varghese, Consultant in Glaucoma and Neuro-ophthalmology, Alphonsa Eye Hospitals, Pala, Thodupuzha, Kerala
   St Joseph Eye Hospital, Kanjirapally, Kerala
   Alexander Eye Centre, Ernakulam, Kerala
3. Dr. Natasha Radhakrishnan, Professor, Amrita Institute of Medical Sciences, Kochi, Kerala.

Submitted: 24-Oct-2020 **Accepted:** 25-Oct-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_166_20	

CASE

A 54-year-old female presented with doubling of images on looking toward the left side for the past 1-week duration. She was diagnosed of diabetes mellitus (Type-2), 35 years back and was on oral hypoglycemic agents for the past 20 years. For the past 15 years, she is on insulin injections, but her diabetic status remains poorly controlled. She was diagnosed of hypertension and hyperlipidemia 30 years back and is on oral medications since then. She was diagnosed of meningioma of left CPA 3 years back following investigations for "ringing sensation" in the left ear. She underwent gamma knife irradiation for the same and had relief of symptoms after the treatment. She has had two episodes of binocular double vision 7 and 5 years back, for which she was treated conservatively and was symptom free after 6 months. There was no history of trauma, headache, ringing in the ears,

Sanitha Sathyan[1,2]

[1]Department of Ophthalmology, Chaithanya Eye Institute, Kochi, [2]Vettam Eye Clinic, Mulanthuruthy, Ernakulam, Kerala, India

Address for correspondence: Dr. Sanitha Sathyan, Vettam Eye Clinic, Perumpilly, Mulanthuruthy, Ernakulam - 682 314, Kerala, India.
E-mail: dr.sanitha@gmail.com

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Sathyan S. Unilateral abducent nerve palsy in a diabetic patient with treated cerebellopontine angle tumor. Kerala J Ophthalmol 2020;32:311-4.

vertigo, nausea/vomiting or blurred vision associated with the current episode.

Now, she presented with doubling of images on looking toward the left side and occasionally in the straight gaze. The double vision is more for distance and no doubling is reported while reading or watching television.

On examination, her unaided visual acuity was 6/12 in both the eyes. With –1.25 D cylinder at 70° and + 2.50 D near addition, she is improving to 6/6, N6. There is no abnormal posturing of the head, ptosis, or nystagmus. Corneal reflex test is central. There is limitation of abduction in the left eye (−2), and the pupils are normal. Saccades and pursuits are normal except for limitation of abduction in the left eye. Corneal sensation: Normal, color vision (Ishihara isochromatic test plates, Farnsworth D 15 test: normal, and there is no red desaturation in the left eye. Humphrey visual fields 30-2: Normal. Diplopia charting: no diplopia elicited in any position of gaze, including levoversion. Anterior segment examination shows grade: 1 nuclear sclerosis in both eyes, dilated fundus: disc normal, mild nonproliferative diabetic retinopathy changes present, and no clinically significant macular edema. General examination, neurological examination, and other systems: Normal.

Magnetic resonance imaging (MRI) brain (3.0 Tesla) done in 2017, post gamma knife radiation therapy, revealed a well-defined extra-axial dural mass of intermediated signal in T1-weighted image (WI) and T2-W1 and hyperintense signal in fluid-attenuated inversion recovery measuring 2.5 cm × 1.5 cm × 2 cm, involving the posterior margin of the petrous temporal bone on the left side [Figure 1]. The lesion showed a cerebrospinal fluid (CSF) cleft around it and was intending the pons and cerebellum on the left side and was covering the medial end of the external auditory meatus on the left side. No extension to the external auditory meatus, hydrocephalus, or encasement of the basilar artery was noted. These features were suggestive of meningioma of the left CPA.

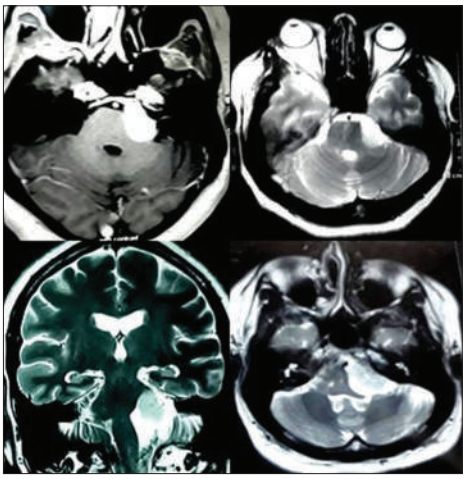


Figure 1: Magnetic resonance imaging of the brain showing the lesion in the cerebellopontine angle

Three follow-up MRIs done at 6-monthly intervals showed no significant changes in the size and morphology of the left cerebellopontine angle meningioma.

Queries

1. What is the possible diagnosis? Left abducent nerve paresis due to diabetic mononeuropathy or due to worsening of CPA meningioma?

Dr. M V Francis

Vasculopathic cranial neuropathy is the most common cause of isolated unilateral sixth nerve deficit in individuals over age 50. Extracting all the pivotal points from the history, my tentative diagnosis is left abducent nerve palsy due to diabetic mononeuropathy. Salient diagnostic points are as follows:

- Typical age of presentation and the presence of long-standing diabetes mellitus, which is poorly controlled
- Two more additional vascular risk factors in the form of long-standing systemic hypertension and hyperlipidemia
- Two episodes of binocular diplopia in the past with an interval of 2 years, with complete recovery within 6 months of conservative treatment
- No other Vestibulo-cocchlear symptoms suggestive of worsening of CPA meningioma, with no significant change in size and morphology as per the follow-up MRIs
- Her first diagnostic symptom at diagnosis was tinnitus, and a recurrence or further growth will be usually present with tinnitus or hearing loss. However, no symptoms suggestive of CPA tumor recurrence such as paresthesia in the distribution of trigeminal nerve, facial twitching, reduced corneal sensation, or Brun nystagmus, occurred during the present episode. The combination of these factors indicates that worsening of CPA tumor is unlikely in this patient.

Dr. Thomas Arun Varghese

Isolated abducent nerve palsy in a person with systemic comorbidities is usually attributed to vascular risk factors. However, in patients with other intracranial or ocular pathology, pointing out the exact cause becomes pertinent.

A diabetic patient with poor control is a high-risk candidate for acquiring a mononeuropathy. In a large Indian study on isolated abducent nerve palsies, 54.70% were attributed to

vasculopathic factors after ruling out other causes through appropriate imaging.[1] In a patient with a history of CPA tumor treated with gamma knife irradiation 3 years prior, recurrence of the tumor is a definite possibility which can only be ruled out by a repeat neuroimaging. Clinical features suggestive of recurrence would be recent-onset involvement of other cranial nerves such as reduction in corneal sensation, which was previously normal, nystagmus, or mild facial palsy.

Some of the hypotheses regarding relapsing and remitting course of abducent nerve palsy are the following:

1. Compression, sometimes intermittent, by a mass causing demyelination followed by remyelination[2]
2. Neuropraxia: Segmental demyelination often triggered by trivial trauma in an already-stretched nerve
3. Thickening and hyalinization of nutrient vessels, leading to focal ischemic demyelination followed by remyelination. This is the likely mechanism in vasculopathic cases.[3]

As can be understood from mechanisms (1) and (2), mass lesions can also cause abducent nerve palsy, which usually recovers. Meningiomas, which are slow-growing tumors, can cause gradual stretching of the nerve. The past history of abducent nerve palsy 7 and 5 years back, may have been due to neuropraxia of a stretched nerve which had recovered.

Dr. Natasha Radhakrishnan

The most likely diagnosis appears to be diabetic mononeuropathy in view of the uncontrolled blood sugars and the previous two episodes which recovered on conservative management. The points against worsening of CPA tumor are the lack of characteristic symptoms such as deafness and tinnitus. Furthermore, corneal sensation is normal and there is no evidence of papilledema. However, given the history of treatment for meningioma, I would repeat imaging to confirm that the tumor has not increased in size.

Queries

2. What is your approach toward suspected diabetic mononeuropathy? Will you offer intravenous steroids to the patient in view of a possible diabetic mononeuropathy or choose to observe the patient?

Dr. M V Francis

As we know, symptomatic diplopia in diabetic mononeuropathy will improve on its own within 3–6 months, like the past two attacks in the patient. There is no need of any intervention at present, and only observation is enough with careful 2-weekly or monthly follow-up. The most important factor in management is strict control of diabetes mellitus by a physician/diabetologist/endocrinologist. Intravenous steroids are not indicated as her diabetes is poorly controlled. With proper control of diabetes, recovery will be almost complete within 3–6 months. Furthermore, the patient is to be taught self-monitoring of her hearing with finger rubbing test. In case of any suspicion, an otorhinolaryngological reference and audiometry are to be done.

Dr. Thomas Arun Varghese

As the mechanism is usually ischemia, I will observe the patient and liaise with the treating physician to improve the control of diabetes mellitus. I usually monitor the patient weekly for the first 2 weeks and will look for any other new-onset neurological involvement. If the patient is stable or improving, further follow-ups may be done once fortnightly. Symptomatic measures such as occlusion are advised during this period.

Dr. Natasha Radhakrishnan

I will not consider intravenous steroids as diabetic mononeuropathy is a self-limiting condition. I would ask the patient to achieve better control of diabetes mellitus and would follow-up the patient monthly. Monitoring of pupils, corneal sensation, fundus, and development of any other cranial nerve palsy will be done at each visit, even if the diagnosis is diabetic mononeuropathy. I expect full recovery in 3–4 months.

Queries

3. What is your approach toward suspected CPA tumors? Will you ask for imaging directly or refer to a neurologist before imaging? If you ask for imaging, what will be your investigation of choice?

Dr. M V Francis

If worsening of CPA lesion is suspected with very early symptoms and signs discussed above or if abduction deficit is persisting after 3 months of observation and good control of vascular risk factors, I will image this patient on my own to confirm my suspicion of worsening of CPA tumor. If the imaging indicates worsening of the tumor, I will refer her to the same center where gamma knife stereotactic radiosurgery was done previously. My choice of imaging will be in discussion with the radiologist/neuroradiologist as radiographic investigations of the skull base and posterior fossa (PF) can be challenging. Standard computed tomography sequences have poor resolution in this region due to signal degradation at soft tissue-to-bone interfaces. Magnetic resonance images are not hampered by this limitation. Contrast-enhanced MRI is the most sensitive investigation in the evaluation of CPA lesions. Ideally, imaging must include high resolution (1.5/3 Tesla), T2 weighted, three dimensional MRI. This sequence is currently the MRI technique of choice

for CSF cisternography for visualizing the CSF spaces at the skull base and thus the cranial nerves, the Meckel's caves, the internal auditory canals, and the fluid within the inner ear structures.

Dr. Thomas Arun Varghese

When CPA mass is suspected due to other symptoms and signs in addition to abducent nerve palsy, I will request for MRI brain with contrast with a detailed letter to the radiologist. Referral to the neurologist will be based on the report.

In majority of cases of recurrent abducent nerve palsy, there is a definite cause such as compressive[4,5] (aneurysm, tumor) or vasospasm.[5] Appropriate and sometimes repeat imaging will pick up the cause in most cases.

Dr. Natasha Radhakrishnan

In a patient with suspected diabetic mononeuropathy, if I need to do an MRI to rule out intracranial pathology, I would order an MRI myself. However, in this patient with the history of a meningioma and gamma knife surgery, I would liaise with the treating neurologist or neurosurgeon before ordering MRI. MRI brain with contrast would be my preferred imaging modality.

SUMMARY

In this case, all the panelists have agreed upon the first provisional diagnosis as diabetic mononeuropathy, due to its high prevalence, absence of characteristic symptoms associated with CPA tumor, and documented stable lesion in serial MRIs. However, due to the multiple clinical factors involved, two of the panelists would also suggest an MRI brain at the presentation itself to rule out the possible increase in tumor size and subsequent compression. One of the panelists would observe the patient closely for any symptom/sign indicative of cerebellopontine tumor. He would suggest imaging only if any such symptoms/signs appear or if abduction deficit is persisting after 3 months of observation and good control of vascular risk factors.

MRI of the brain is the preferred imaging modality and the specifications would be decided in consultation with a radiologist/neurologist/neurosurgeon. All the panelists would manage isolated diabetic neuropathy cases with observation and close follow-ups. This is to rule out new-onset symptoms/cranial nerve involvements, and monocular occlusion will be offered for symptomatic diplopia. Control of vascular risk factors such as diabetes mellitus, hypertension, and hyperlipidemia would be done in consultation with the physician/endocrinologist and it is expected that the isolated diabetic abducent nerve palsy would resolve within 6 months.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initial s will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Nair AG, Ambika S, Noronha VO, Gandhi RA. The diagnostic yield of neuroimaging in sixth nerve palsy-Sankara Nethralaya Abducens Palsy Study (SNAPS): Report 1. Indian Journal of Ophthalmology. 2014;62:1008.
2. Blumenthal EZ, Gomori JM, Dotan S. Recurrent abducens nerve palsy caused by dolichoectasia of the cavernous internal carotid artery. Am J Ophthalmol 1997;124:255-7.
3. Asbury AK, Aldredge H, Hershberg R, Fisher CM. Oculomotor palsy in diabetes mellitus: A clinico-pathological study. Brain 1970;93:555-66.
4. Nguyen DQ, Perera L, Kyle G. Recurrent isolated sixth nerve palsy secondary to an intracavernous carotid artery aneurysm. Eye (Lond) 2006;20:1416-7.
5. Chan JW, Albreston J. Causes of isolated recurrent ipsilateral abducent nerve palsy in older adults: A case series and review of literature. Clin Ophthalmol 2015;9:373-7.

Sir Harold Ridley and the intraocular foreign body that made history

The operation theater register of St Thomas' Hospital, London, in the 1949–1950 period, contains two entries which are part of history now. Both described surgeries done on the left eye of a 45-year-old female by a certain L. Ridley. There was nothing unusual about the first entry, which was on November 29, 1949, and simply read "Extracapsular ext"[1,2] [Figure 1a].

The second entry on February 8, 1950, however, was very unusual as the procedure is mentioned as "Lenticular Graft"[1,2] [Figure 1b]. No such entry had ever been made in any operation theater register before. What it meant to the hospital and what it gave to the world is clarified by a decorative plaque [Figure 2] installed 50 years later at the hospital, which reads *"The first intraocular lens (IOL) for the treatment of cataract was implanted by Mr. Harold Ridley FRS at St Thomas' Hospital on 8th February 1950."*[3]

However, for the surgeon himself, the seemingly usual first date and entry was the milestone date as he would clarify later in his publications and interviews and vehemently vouched for by his faithful nurse and operation theater assistant Ms. Doreen Ogg. She would explain later that in fact the first implantation of an IOL was performed on November 29, 1949. In written records, the operation on that date was referred to only as "Extracapsular ext." Mention of an IOL was purposely omitted to retain the secrecy as requested by the surgeon.

Ms. Ogg also remembers vividly that Dr. Harold Ridley and the opticians who helped him design the IOL were totally disappointed when the patient had a postoperative myopia of about -18 D. She recalls that a second surgery was done months later, in which the first IOL was removed and the second one implanted. Hence, this must have been the "Lenticular Graft" on 8th February. Thus, both the register entries are historic – World's first IOL Implantation happened, in all probability on November 29, 1949, and the first IOL exchange on February 8, 1950.[1,4]

The IOL was a warchild. It was born out of the 2nd World War and it created another world war in the ophthalmic world, a war which left its inventor badly bruised and battered, a war in which he was ridiculed, abused, and threatened with litigations, a war which saw many of the big names in ophthalmology join forces against him.

The ophthalmic world was not ready to accept the IOL. However, as Victor Hugo had said "Nothing is more powerful than an idea whose time has come." The way this particular idea emerged and how it found the right person at the right time is a story of startling coincidences.

To begin with, the inventor of the IOL, Sir Nicholas Harold Lloyd Ridley (July 10, 1906–May 25, 2001), was never short of ideas. He was born as the elder of the two sons to Nicholas Charles Ridley and his wife Margaret Parker in Kibworth, Leicestershire, in England. At the age of 7 years, the shy-stammering boy once told his mother that he wanted to be an inventor.[3] He was always making new toys and all sorts of things in his childhood. In spite of not being a great communicator, he asked the right questions more often than not and ended up finding practical solutions. This very same nature was in display when during his house surgency at the St Thomas' Hospital, London; he encountered a young skilled work man whose career was brought to an abrupt halt by a traumatic cataract and intraocular foreign body in one eye. The man's work was such that binocular vision was essential. The young Ridley brooded over it and posed a question to two people whom he adored and worshipped most at that time, his father, who was then a consultant ophthalmic surgeon to the Royal Navy and his beloved teacher and mentor in the

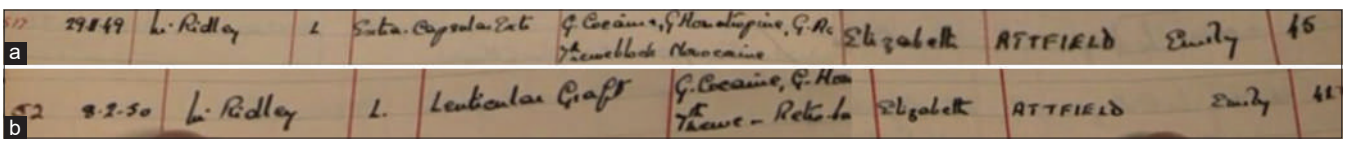


Figure 1: (a and b) Reproduced with permission Original Entry in the OT register of St Thomas' Hospital on November 29, 1949 and February 8, 1950 (Source: A video of the logbook from 1949 showing summary of Sir Harold Ridley operations from 1949 and 1950 supplied by Prof. David Spalton in the ESCRS Video Collection (https://player.escrs.org/online-museum/logbook-from-1949-showing-summary-of-ridley-operations)



Figure 2: Plaque commemorating the world's first intraocular implantation at, St Thomas' Hospital, London, (Source: Wikipedia https://en.wikipedia.org/wiki/Harold_Ridley_(ophthalmologist))

Department of Ophthalmology of St Thomas' hospital, Mr. A Cyril Hudson. "How nice it would be to put a new lens in his eye?"

The senior Ridley said it was impossible and dangerous. The answer of his teacher Mr. Hudson reflected the collective ophthalmic mindset at that time "We take foreign bodies out of eyes. We don't put them in."[3] And, that was the end of the matter or so thought the senior men. But, unknown to them or even the questioner himself, the embers of the flame were residing deep inside that questioning mind waiting to be fanned. The fanning came eventually a decade later when Harold Ridley was working as a full-time ophthalmic surgeon at the famous Moorfields Eye Hospital, London.

It was in 1938 that Ridley got the coveted appointment as a full surgeon and permanent consultant at Moorfields Eye Hospital.[5] He had plenty of experience both professional and worldly under his belt by then, which included 6 months as casualty officer, 1 year of general surgery, 6 months in ophthalmology under Mr. A C Hudson, all at St Thomas' Hospital, followed by 1 year as ship's surgeon in various vessels and then an 18-month formal ophthalmology residency training at Moorfields in 1934–1935.

THE FORGOTTEN GOGGLES AND A HISTORIC FOREIGN BODY

As Dr. Ridley began spreading his wings at the Moorfield's, eighty miles away another young man, having obtained his wings 1 year back was beginning to make good use of it. This was Flight Lieutenant Gordon Mouse Cleaver of the Royal Air Force 601 Squadron. A set of curious coincidences starting with the outbreak of the Second World War in 1939 contrived to place Dr. Ridley in the flight lieutenants "flight path." On August 13, 1940, the German Air force launched one of the fiercest aerial attacks on Britain , code named "Eagle Attack,". The objective was to "Wipe out the British air Force from the sky"[4] and what resulted was one of the fiercest battles in the history of aerial warfare. The 601 Squadron was in the thick of things. On August 14, 1940, Gordon Cleaver took off into the battle in the "Hurricane" that was normally assigned to him. He survived the morning sortie and returned by noon. He was just entering the officer's mess for refreshments when he was called back for another "scramble" based on the radar warning. His plane, it seemed, was not ready and by pulling the choke Cleaver ended up flooding and incapacitating the engine. He was immediately allotted a brand new Hurricane by the commander and ordered to catch up. In his haste and confusion, he forgot the air man's goggles. He finished the combat, but while returning to base was ambushed by German fighter planes. The canopy of his cockpit made of Perspex (Commercial Polymethyl methacrylate [PMMA]) shattered under the enemy fire. Without the protection of his goggles, he was blinded instantly as multiple fragments of the Perspex penetrated into both his eyes. The brave pilot maintained control and flipped the plane upside down enabling him to just fall off from the plane and parachute down to safety.

The first words spoken by Cleaver to his friend and mate Jack Riddle when the latter visited him at the military hospital were."Jack, tell them all to wear their goggles."[5] Evidently, this was not an isolated problem.

Eventually, Cleaver was referred to Moorfield's Eye Hospital and this was where he came under Dr. Ridley's care. He was under treatment at Moorfields for several years during which about 18 surgeries were done on his eye and face to preserve some vision and several of them were performed by Dr. Harold Ridley. Many of these were for the removal of the Perspex pieces that had gone inside his eyes or embedded in the ocular coats. Eventually, one of the Cleaver's eyes (left) could be saved, and in spite of a large iris hole through which one Perspex piece had gone in and hit his lens and one or two pieces still inside, he was able to return to civilian life with reasonably good vision. The other eye was permanently blinded. This was when Ridley made the historic observation that the acrylic pieces in cleaver's eye did not seem to elicit any sort of inflammatory reaction. He observed the same phenomenon in several other pilots' eyes with similar foreign bodies. The old flame of an idea in his mind was rekindled. Was it the Eureka moment?

A FRUITFUL DETOUR

The flames were soon doused as he found his plate full for the next 6–7 years by other pursuits in life. May 1941 turned out to be bitter sweet for Dr. Ridley, sweet because of his marriage to Elisabeth Weather hill and his entry into temporary military service as Major, Royal Army Medical Corps. The bitter part was that he found himself posted in Ghana, West Africa. Ridley was deeply distressed by this as he knew his surgical experience was going to be wasted there as it was not a place of any war action.[5] He also believed, perhaps rightly that Sir Stewart Duke Elder who was a very high ranking official in the Medical Corps at that time was behind this. He put 18 months of service in Ghana followed by a short period of service in Calcutta, India, and then finally in Rangoon, Burma. These years in the wilderness also saw significant contributions to ophthalmology coming from the irrepressible Ridley and included first of its kind papers on Onchocerciasis and Nutritional amblyopia based on his vast experiences in Ghana and Burma, respectively. The fundus drawing painted by Ridley showing the posterior pole changes in Onchocerciasis came to be known as the "Ridley Fundus." He was given an early release from the army in 1945 and brought back to Britain to make use of his unparalleled experience in tropical ophthalmology to treat the refugees and prisoners returning after the war. Ridley was appointed honorary ophthalmologist, Ministry of Defense, and resumed duties with an affiliation at both St Thomas and Moorfields, performing surgeries in both hospitals.[5]

Dr. Ridley's inventive and enthusiastic mind found a new passion at this time, Electronics. With the help of Marconi's Wireless Telegraph Company, he became the first person to televise eye operations, first in black and white (1948) and later in color. He did pioneering work in electronic ophthalmoscopy which helped later in establishing principles of confocal microscopy and scanning laser ophthalmoscopy.[4]

THE FINAL CATALYST AND THE FIRE

In 1930, a young house surgeon had innocently asked his father and teacher about the feasibility of putting an artificial lens inside the human eye. 17 years later, history turned the tables on the questioner. Major Harold Ridley found himself at the receiving end of his old heretical question. It came from a medical student (Stephen Perry) who after watching Ridley perform a cataract surgery remarked. "It is a pity that you can't replace the cataract with a clear lens."[5] Ridley decided that it was time to change the answer. The old flame was back. There was no holding back of the idea now. Its time had come. This time Ridley knew exactly what material he was going to use for the lens. Gordon Cleaver and the other pilots whom he treated had taught him that. The 8-year follow-up of the Perspex (PMMA) pieces in Cleaver's and the other Pilot's eyes was to Ridley the preclinical study of his material.[4] However, he was acutely aware of the general sentiment of ophthalmologists toward foreign bodies inside the eye and wanted to keep things under wrap till he had concrete results.

One evening in 1948, a rather secretive meeting took place between two men in the back seat of a Bentley Car parked in a side road of Cavendish Square, London. One was the owner of the car, Dr. Harold Ridley and the other his friend John Pike, an optical scientist at Rayner and Keeler, a leading optical company in England. The cure of aphakia was perhaps established in the next 30 min in that car when the two men discussed and finalized the nitty-gritty and major principles of the first IOL implantation surgery.[4] Other materials such as glass and quartz were also considered for the implant and rejected. As for the site of implantation, the more accessible anterior chamber was considered and rejected. In Ridley's own words, he wanted the implant to be "just where the nature had placed a biconvex lens throughout the animal kingdom." "Extracapsular extraction was preferred to the more fashionable intracapsular because it produced a stronger eye with the intact posterior capsule acting as a bulkhead within the globe."[5] John Pike roped in his friend Dr. John Holt at the Imperial Chemical Industries to make some pure high quality Perspex, as the commercially available one was not pure. John Holt obliged and the material was made, which he named the Perspex clinical quality (CQ). Allvar Gullstrand's work and other known formulae helped them to fashion a lens with what they thought was the desired optical effect. The three of them Dr. Ridley, Dr. John Holt and John Pike agreed to forgo any financial reward to prevent colleagues from alleging it was a commercial venture and did not even apply for a patent.

The next step was to find a volunteer for the procedure. It had to be a patient with unilateral cataract who was able to work quite well with the other eye and for whom the loss of function in one eye would not be so bad. The patient had to understand the risks of the untried operation and be prepared for the possible loss of the defective eye. It took Ridley about 1 year to get the right candidate.[3,4,6] The patient was a 45-year-old hospital nurse named Elizabeth Attfield who had a cataract in her left eye. The lens was a simple disc with a peripheral groove, exquisitely manufactured by Rayner and Keeler from Perspex CQ, and given to Ridley at the cost of 18 Shillings (<1 pound).

The operation for which Dr. Ridley had been preparing, which he might have rehearsed a hundred times in his mind, was

finally carried out in secrecy at the St Thomas' Hospital in the afternoon of November 29, 1949, and history was made. The confusion regarding the dates and why the plaque in St Thomas mentions 8th February as the historic date and still believed by many as the actual date have already been mentioned in the beginning of this article. As per the latter theory, the entire exercise was a 2-stage procedure and the IOL implantation happened in the second stage on February 8, 1950.

Ridley and John Pike had been a little too accurate in copying the radii of curvature of the human lens to their implant with the result that the first two patients were rendered highly myopic. However, they made the necessary adjustments with the result that the subsequent implants only produced anisometropia in the range of 2–3 diopters[7] [Figure 3].

THE WRONG RIDLEY AND AN OPHTHALMIC WORLD WAR

The 2nd, 3rd, and 4th implantations were done in Moorfield's hospital. A total of 8 such operations were done till 1951. Dr. Ridley wanted to keep everything secret for about 2 years by which he thought he would be sure about the results. However, another curious coincidence forced his hand. One of his implant patients, delighted after the surgery wanted to meet Dr. Ridley outside the hospital to give him a fee.[3] He looked up Ridley in the phone book, made an appointment, and presented himself at the office of "Dr. Ridley." The problem was it was the wrong Ridley. It was the office of Dr. Frederick Ridley, another ophthalmologist. This happened as "F" came before "H" in the directory and the patient had selected the number of the first Dr. Ridley for fixing up the appointment. The news was out. So, to establish priority, he published about the IOL in the "St Thomas' Hospital Reports," which was a low profile, not widely read journal. Subsequently, he published reports in the Lancet and also in BJO.



Figure 3: Ridley Posterior Chamber intraocular lens (1949–1960) (Source: https://rayner.com/about-us/history/)

(Dr. Fredrick Ridley introduced the wet pack [soak in 10% NaOH for 1 h at 30°C, store in 0.1% NaOH, neutralize on use with 0.5% NaHCO3, rinse with saline] method of IOL sterilization which was used from 1957 to 1978)

Arguably Dr. Ridley's IOL work cannot be claimed to have adhered to the present day ethical research principles, but then those were not well established at that time. However, one mistake was that the dean and the head of research of Moorfields Eye Hospital who also happened to be one of the most powerful men in British Ophthalmology at that time was kept in the dark about these implantations and that was a certain Sir Stewart Duke Elder. There was a row over it in and Ridley is reported to have virtually told Duke Elder that he did not see any reason to inform him about what he was doing. He was never forgiven for that and thus ensued one of the bitter battles in ophthalmology.

In July 1951, Ridley drove to the Oxford Ophthalmological Conference with high hopes of presenting his work there in a lecture allotted to him. In his car was his wife and two of his IOL implant patients with good postoperative vision. One had 20/20 unaided. He also had prepared a cine film of one of his implant operations.

Dr. Ridley's lecture was well prepared and presented, but the response was something for which he was not prepared at all. Barring a few greatly impressed people, most from USA and Britain showed little interest and there were some very hostile reactions. Sir Stewart Duke Elder and the prominent ophthalmologists repeatedly refused to even look at the patients. The film was never shown. The session was foreclosed by the deputy master and Dr. Ridley and his wife drove off quietly, missing the dinner arranged. His attempt at getting his work recognized at the US also met with a similar fate when he had to face an even more hostile reaction at the 57th session of the American Academy of Ophthalmology at Chicago in 1952. The comments of Derrik Veil the then Editor in Chief of the AJO who incidentally was a close friend of Sir Duke Elder were especially scathing. However, there were also people like Warren Reese who enthusiastically collected one Lens from Ridley, flew back to Philadelphia in his private plane, and implanted the same the next morning at Wills Eye Hospital, thus becoming the first implant surgeon in the United States.[4]

Dr. Ridley had hoped that his Oxford presentation would be a land mark day in the history of Ophthalmology. To quote David Apple, "Indeed, it was a landmark day, but not in the way he expected. Instead, it was the beginning of more than 30 years of personal trials and tribulations that led to health problems that plagued him for the rest of his life. Worse,

it was the beginning of an unconscionable delay in the implementation of this new procedure, a delay that deprived an entire generation of patients of the benefits of the IOL."[4]

Ridley did about 1000 IOL implantations with reasonable success. But, many of them had complications related to the instability and poor fixation of the rather heavy lens and uveitis in a few due to the cetrimide solution used for sterilization of the lenses. Eventually, 15%–20% of these lenses had to be explanted.[8] He was forced to abandon the procedure due to severe opposition, ostracization from the mainstream ophthalmology, fear of litigations, and report of a plethora of complications happening around the world due to poor quality IOLs and suboptimal surgery by many. The quality of some of these lenses even led to the joke that they would have made better IUDs than IOLs.[6] Many ophthalmologists who could never reconcile with the absolutely heretical idea of "putting things into the eye" viewed the IOL as a "time bomb." Had Dr. Ridley access to things such as viscoelastics, operating microscopes, and capsulorhexis principles, the story would have played out differently.

Anyway, there were people like Ridley's favourite protégé Peter Choyce, Edward Epstein, Binkhorst, and Fyodrov to take the IOL story forward through the first-generation anterior chamber IOLs (1952–1962), iris-supported lenses (1953–1973), modern anterior chamber lenses (1963 onward), and finally back to modern posterior chamber IOLs from 1975 onward. In 1979, when almost simultaneously John Pierce of UK and Peter Sherring of US came up with the posterior chamber IOL design of today, it was the Ridley lens which was back again, this time with 2 haptics for fixation which made the central disc smaller and much lighter.[9] Its implantation place also returned to where Ridley had planned 30 years back, i.e., "where nature had placed a biconvex lens throughout the animal kingdom." By 1980, posterior Chamber intraocular lenses got the Food and Drug Administration approval and finally Dr. Harold Ridley was vindicated in all respects.

RECOGNITION FINALLY

For 30 years after his historic IOL implantation, Dr. Harold Ridley received more of ridicule, abuse, and professional ostracism than recognition. There were times when he had to take medication for depression. But, things began to change after 1980. He is reported to have said at European Intraocular Implant Lens Council in 1986 "All you people have enjoyed your implant work, I'm sure, I suffered from it." But from then on, at last in the last 15 years of his life, he began receiving the honor and recognition which was overdue. These included -1.Election to the Royal Society, London, in 1986 2.Doctor of Humane Letters, (DHL) conferred in 1989, by the Medical University of South Carolina, USA. 3.Publication of a detailed biography by David J Apple in the Survey of Ophthalmology in 1996(6)4.The Gullstrand Medal conferred by the Swedish Society of Medicine in 1992, 5.Being honoured at the annual meeting of the American Society of Cataract and Refractive Surgery in Seattle,Washington in April 1999. And finally to cap it all knighthood was conferred on him by Queen Elizabeth in February 2000 [Figure 4]. He was 92 years old and partly deaf at the time of his knighting. After the ceremony when he was asked what did the queen tell him, Sir Harold Ridley answered with a smile, "I couldn't hear a damn thing!"[4]

HISTORY FILLING UP SOME DOTTED LINES

In 1940, multiple pieces of PMMA from an air plane canopy plunged a young pilot's world into darkness, but illuminated the mind of a young inventor. Forty-seven years later another PMMA piece, this time deliberately placed into the now retired pilot's eye brought him back into the world of light. This beautiful quirk of history was effected, when Dr. Eric Arnott, in 1987, did cataract surgery on Gordon Mouse Cleaver's only functional eye and implanted an IOL. And, the surgeon reported that there was still a piece of the original Perspex buried in the Cornea.

Two years later, it was the turn of the inventor to benefit from his own invention and he chose to receive it at exactly the same venue where he made the invention. Dr. Harold Ridley successfully underwent cataract surgery with IOL implantation in both his eyes at his favorite St Thomas' Hospital in 1989 and 1990.[4] He asked for GA for both



Figure 4: Investiture of knighthood on Harold Ridley by Queen Elizabeth II, February 9, 2000 (Source: Apple[1], David J., Werner, Liliana, & Werner, Leonardo P. Arquivos Brasileiros de Oftalmologia 2002;65:137-40. https://doi.org/10.1590/S0004-27492002000100025)

surgeries and explained to the surgeon Michel Falcon "Well my boy if I am awake, I will be telling you what to do all the time."[9] He would jokingly say about it later. "I am the only man to have invented his own operation."[10]

HUMANITARIAN SERVICES

The man who, as a little child had sat in the lap of the great Florence Nightingale (was a friend of his mother), who had spent 5 productive years in the service of the underprivileged people of the third world countries, whose life partner had grown up in India, will not be left far behind in humanitarian activities. He established the Ridley foundation in 1967 financed by his inherited assets to promote ophthalmological services in underprivileged countries and to help needy people especially nurses and their families in dire financial situations. Dr. Ridley along with his wife and another nurse used to make several visits to underdeveloped countries, training the local surgeons there in cataract surgery and rendering other help.

EPITAPH

Dr. Ridley was very specific about how he should be remembered. He said in an interview in his inimitable style.[3] "I am going to have on my tombstone: "He cured Aphakia." People are then going to say 'Who was Aphakia?". Well, he did not get a tombstone with that epitaph. But, the millions of intraocular lenses happily sitting in innumerable eyes all over the world literally enlightening and empowering the owners will be proclaiming exactly that silently.

Acknowledgement

I would like to place on record my sincere thanks to Dr Sobha Joseph, Consultant Ophthalmologist Birmingham Heartlands Hospital and Solihull Hospital, London who helped me immensely in getting relevant materials for this article. My sincere thanks also to Professor Dr David Spalton who gave permission to use a screen shot of Dr Ridley's surgical log book from the video he had supplied to the ESCRS Video Collection.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

C. Biju John

Department Of Ophthalmology, Regional Institute of Ophthalmology, Government Medical College, Thiruvananthapuram, Kerala, India

Address for correspondence: Dr. C. Biju John, Regional Institute of Ophthalmology, Government Medical College, Thiruvananthapuram, Kerala, India.
E-mail: bijujc@gmail.com

REFERENCES

1. Trivedi RH, Apple DJ, Pandey SK, Werner L, Izak AM, Vasavada AR, *et al*. Sir Nicholas Harold Ridley. He changed the world, so that we might better see it. Indian J Ophthalmol 2003;51:211-6.
2. Williams H. Casanova and the Spitfire Pilots Ophthalmology Documentaries. Available from: http://www.rcophth.ac.uk/about/rcophth/museum/#. [Last accessed on 2020 Sep 12].
3. Sir Harold Ridley-Science Museum Tribute 2. Available from: https://youtu.be/T7kj7GnCf9I. [Last accessed on 2020 Sep 14].
4. Apple DJ. Sir Nicholas Harold Lloyd Ridley. Biogr Mems Fell R Soc 2007;53:285-307.
5. Apple DJ. Harold Ridley and his fight for sight. He Changed the World so that we May Better See it; Slack Books; Slack Incorporated; 1st edition (September 10, 2006).
6. Apple DJ, Sims J. Harold Ridley and the Invention of the intraocular lens. Surv Ophthalmol 1996;40:279-92.
7. Williams HP. Sir Harold Ridley's vision. Br J Ophthalmol 2001;85:1022-3.
8. Hoffer KJ. The Evolution of the Intraocular Lens. Cataract Surgery. 3rd ed.. PART VI – Intraocular Lenses. Ch. 35. Elsevier Inc. 2010
9. Bhattacharya D. Tribute to Sir Harold Ridley (Video). Available from: https://www.youtube.com/watch?v=RkpssUDW2_Ehttps://www.youtube.com/watch?v=RkpssUDW2_E. [Last accessed on 2020 Sep 12].
10. Cataract Pioneer Sir Harold Ridley to be Honoured with Plaque: BBC News article on 18 February, 2012. Available from: https://www.bbc.com/news/uk-england-leicestershire-17076063. [Last accessed on 2020 Sep 09].

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

Submitted: 18-Oct-2020 **Accepted:** 19-Oct-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_160_20	

How to cite this article: John CB. Sir Harold Ridley and the intraocular foreign body that made history. Kerala J Ophthalmol 2020;32:315-20.

PROSPECTIVE, RANDOMIZED, EYE-TO-EYE COMPARISON OF A NEW SILICONE COREAL SHIELD VERSUS CONVENTIONAL BANDAGE CONTACT LENS AFTER PHOTOREFRACTIVE KEPATECTOMY

Hirabayashi KE, Sales C, Slade SG, Manche EE. Prospective, randomized, eye-to-eye comparison of a new silicone corneal shield versus conventional bandage contact lens after photorefractive keratectomy. J Cataract Refract Surg 2019;45:1782-8 Q 2019 ASCRS and ESCRS.

Photorefractive keratectomy (PRK) is a commonly performed refractive surgery that has equivalent safety and efficacy when compared to laser *in situ* keratomileusis (LASIK) in treating myopia. Although PRK avoids potential flap-related complications, one disadvantage of PRK is the postoperative healing phase, which is more prolonged and uncomfortable compared with LASIK. Various options for decreasing pain after PRK include the use of bandage contact lenses (BCL), topical anesthetics, topical nonsteroidal anti-inflammatory agents, and oral analgesics.

A contact lens that can offer quicker visual rehabilitation and improved comfort would be a game-changer for PRK. Silicone hydrogel lenses are generally preferred because they have high oxygen permeability. However, there is no definite consensus as to which lens is the superior choice. There is a new silicone corneal shield made by NexisVision which has a thin soft periphery and a thicker and slightly stiffer central optic design to improve postoperative vision. It is composed of a bimodulus, silicone, elastomer material that is highly permeable to oxygen, especially in the area overlying the limbus, to facilitate reepithelialization. The Dk of the material is more than 300, but the shield is thicker than most contact lenses, making its Dk/t 110. It is impermeable to water, which helps minimize the development of corneal edema.

The aim of this study is to compare the new silicone corneal shield with a conventional silicone hydrogel BCL after PRK in terms of uncorrected distance visual acuity (UDVA) and patient-reported outcomes.

This is a prospective randomized cohort study that enrolled 25 participants (50 eyes) undergoing myopic PRK from May 2012 to April 2013. Inclusion criteria included less than 11.0 D of myopia with less than 3.50 D of astigmatism. The use of rigid gas-permeable contact lenses previously was an exclusion criterion. Patients were randomized by ocular dominance to receive either the new silicone corneal shield or a conventional BCL in their dominant eye and the other intervention in the contralateral eye. The UDVA and patient-reported outcomes were measured preoperatively, 1 h after surgery, and at 1, 2, 3, 4, and 7 days postoperatively.

Surgery was done by experienced refractive surgeons. An epithelial scrubber or an alcohol well was used to remove the epithelium. A 6.0-mm optical treatment zone was selected, and Mitomycin C (MMC) 0.02% was applied after ablation for 20 s in all cases. Postoperative medications were topical moxifloxacin 0.5%, topical prednisolone 0.125%, and topical diclofenac 0.1% and oral ibuprofen. A postoperative questionnaire was used to assess patient-reported outcomes.

The new silicone corneal shield group had statistically significant superior mean logMAR UDVA than the conventional BCL group 1 h after the surgery and at 1, 2, 3, 4, and 7 days postoperatively. Eyes treated with the new silicone corneal shield also achieved epithelial closure more frequently by 3 days postoperatively compared with eyes treated with the BCL. However, these patients reported increased symptoms of pain, foreign body sensation, and eyelid heaviness with the shield. Potential explanation is that the new silicone corneal shield is nearly 4 times thicker than a standard BCL, and it has lower absolute oxygen transmissibility. The bulkiness of the lens and the relative hypoxia it induces might explain the increased discomfort experienced as compared with a conventional BCL.

The strengths of this study include its prospective, randomized, contralateral, and controlled nature and the masked nature of the study. The limitations include the lack of validation for the questionnaire and the modest size of the study population.

In summary, postoperative recovery remains the largest drawback of PRK. Although the Nexis silicone corneal shield provides a quicker visual recovery, it is associated with increased symptoms of pain, light sensitivity, foreign body sensation, and tearing when compared with a conventional BCL.

ASSESSMENT OF FACTORS ASSOCIATED WITH POSTOPERATIVE PAIN AFTER PHOTOREFRACTIVE KERATECTOMY

Palochak CM, Santamaria J, Justin GA, Apsey DA, Caldwell MC, Steigleman WA, *et al*. Assessment of factors associated with postoperative pain after photorefractive keratectomy. Cornea 2020;39:1215-20.

Photorefractive keratectomy (PRK) is a refractive surgery procedure that uses an excimer laser to reshape the cornea to treat refractive error in patients with myopia, astigmatism, and hyperopia. This correction can reduce or eliminate a patient's need for glasses or contact lenses. PRK reshapes the cornea without the creation of a stromal flap, giving it unique advantages over flap-based procedures, such as laser *in situ* keratomileusis.

However, PRK has a noted disadvantage of postoperative discomfort and pain. PRK involves manually removing the epithelium, creating an epithelial defect in the ablation zone that contributes to the increased pain experienced by patients postoperatively. Multiple studies have shown that this pain is typically most severe during the first day with peak subjective pain reported at 24 h. For some, this pain can be severe and difficult to control in the early postoperative period. Optimal management of this pain has been extensively researched and debated, with studies proposing a wide array of topical and oral nonsteroidal anti-inflammatory drugs, topical anesthetics, bandage contact lens, or a combination of these modalities.

This study aims to determine what factors contribute to postoperative pain in PRK. This retrospective chart review evaluated patients who underwent PRK in 2016. Anonymized data collected included patient gender, age, and season at the time of surgery, ablation depth, surgeon status (attending vs. resident), topical tetracaine use, and subjective pain scores at postoperative days (PODs) 1 and 7. Average pain scores and amount of pain medication taken were analyzed for each of the previously mentioned variables.

All surgeries were performed by or with the direct supervision by a cornea-trained military ophthalmologist following the same standardized protocol. Postoperative medications included topical moxifloxacin 0.5% drops 4 times a day, fluorometholone acetate drops 4 times a day, preservative-free artificial tears as needed, and oral analgesics. Patients were also given an unopened bottle of topical tetracaine hydrochloride 0.5% with instructions to use only in the event of severe pain that was preventing adequate rest. Patients were followed as per standard protocol on PODs 1 and 7 with assessment of their pain, number of tetracaine drops used, and number of pain pills consumed, verified, and recorded by a trained ophthalmic technician.

Postoperative pain was significantly elevated in patients who used tetracaine on POD 1 and POD 7. These patients were also more likely to take oral pain medication than those who did not use topical tetracaine. Surgeon status, season, gender, and ablation depth showed no significant differences in subjective pain scores.

Limitations of this study are that it was retrospective in nature and that it only recorded pain data from PODs 1 and 7. Furthermore, the 10-point scale was based on an established onsite system that may not be equivalent to more detailed methods and questionnaires for quantifying and qualifying pain.

To conclude, most treated patients have minimal postoperative pain, allowing PRK to continue to be a good and effective option for military patients. Furthermore, oral pain medication should be evaluated to assess efficacy and safety in inhibiting ocular pain after PRK. This research study can be added to the growing body of evidence that PRK has its unique role in the military medical setting.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

S Reshmi
Sreepury, Balussery PO, Calicut - 673 612, Kerala, India.
E-mail: reshmisreepadmam@gmail.com

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

Submitted: 17-Sep-2020 **Accepted:** 18-Sep-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_144_20	

How to cite this article: Reshmi S. Journal Review. Kerala J Ophthalmol 2020;32:321-2.

© 2020 Kerala Journal of Ophthalmology | Published by Wolters Kluwer - Medknow

Insights into IOLMaster®: A useful tool for cataract surgeons

INTRODUCTION

The IOL Master (Carl Zeiss Meditec AG, Goeschwitzer Strasse 51-52 07745 Jena, Germany) was first launched in 1999 and revolutionized ocular biometry principles due to its superior repeatability, reproducibility, and accuracy in results.[1,2] The principle involved is Michelson's partial coherence interferometry to measure axial length (AL).[3] It also uses the innovative lateral slit illumination of the crystalline lens and the cornea to measure anterior chamber depth (ACD) and autokeratometry to determine corneal curvature. Currently, it is the gold standard for preoperative computation of intraocular lens (IOL) power in patients undergoing cataract surgery.[4] Being a noncontact noninvasive technique, it avoids the use for topical anesthesia, making it particularly profitable in studying ocular growth, myopia, and interventions in both children and adults.[5] Since then, several other instruments have been introduced. They all utilize the same Michelson optical low-coherence interferometry to determine the AL and a range of other technologies to measure other components.

The other instruments include:

1. Lenstar (Haag-Streit, Mason, OH, USA)[6,7]
2. Oculus Pentacam® AXL
3. AL Scan (Nidek, Inc., Fremont, CA, USA)
4. Aladdin (Topcon Corp., Oakland, NJ, USA)[4]
5. Galilei G6 (Ziemer Ophthalmic Systems AG, Switzerland),[8] and
6. OA-2000/OA-1000 (Tomey Corp., Phoenix, AZ, USA).[9]

STUDY DEVICES

IOLMaster 500

The IOLMaster 500 is a noncontact, high-resolution biometric device that measures AL using the principle of partial coherence interferometry, keratometry using reflected light spots on the surface of the cornea, ACD from optical sections of the lens and the cornea, and white-to-white distance based on a scleral and iris image.

IOLMaster 700

The IOLMaster 700 is another noninvasive, noncontact, high-resolution biometric device that employs swept-source technology (laser with a tunable wavelength centered on 1055 nm) and generates ultrasound B-scan images to determine the biometric data of the eye. These B-scans are displayed as a full-length optical coherence tomography (OCT) image showing anatomical details on a longitudinal section through the entire eye. Thus, unusual ocular features, such as a tilt or decentration of the crystalline lens, can be detected. It measures AL, ACD, central cornea thickness, and lens thickness using swept-source OCT technology, corneal curvature using reflected light spots on the surface of the cornea, pupil diameter, visual axis (line of sight), and white-to-white measurement based on a scleral and iris image. The device acquires multiple measurements for each of the parameters in a single measurement-capture process. Axial measurements are based on swept-source frequency-domain OCT enabling a 44-mm scan depth with 22-mm tissue resolution. Note that corneal and lens thickness has been added to the already-existing measurement modes of the IOLMaster 500,

Bharat Gurnani, Kirandeep Kaur, Poulomi Das
Cataract, Cornea and Refractive Services, Aravind Eye Hospital, Pondicherry, India

Address for correspondence: Dr. Kirandeep Kaur,
Fellow Pediatric Ophthalmology and Squint Services, Aravind Eye Hospital, Pondicherry, India.
E-mail: beingkirandeep@gmail.com

Submitted: 14-May-2020 **Accepted:** 27-May-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	Quick Response Code
DOI: 10.4103/kjo.kjo_57_20	

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Gurnani B, Kaur K, Das P. Insights into IOLMaster®: A useful tool for cataract surgeons. Kerala J Ophthalmol 2020;32:323-8.

and the technology used to measure ACD is different. The Sweptsource OCT machine scans sequentially through a series of wavelengths to enhance the interference pattern, which is then decoded with Fourier transformation into an Ascan trace. Multiple adjacent A-scans can be combined to form a B-scan image. The advantages of swept-source OCT include a deeper range of imaging into the eye, less sensitivity reduction with depth, and faster scanning speeds [Figure 1].

Lenstar LS 900

The Lenstar LS 900 is another noninvasive, noncontact, biometric device that measures AL, ACD, central corneal thickness, and lens thickness using the principle of partial coherence interferometry, corneal curvature using reflected light spots on the surface of the cornea, pupil diameter, visual axis (line of sight), and white-to-white measurement based on a scleral and iris image. All the above axial measurements are obtained by optical low coherence reflectometry using a broadband light source (20–30 nm) with a center wavelength of 820 mm. From a single measurement, all of these mentioned parameters can be assessed. The Lenstar LS 900 allows the user to view a clearly defined display represented in the style of an immersion A-scan. The measurement calipers can be adjusted if necessary, in the presence of artifacts, for example, vitreous floaters.[10-13]

COMPONENTS OF IOLMASTER[14,15] [FIGURES 2a AND b, 3a AND b]

1. Instrument table IT 3 L
2. Holding bar for securing the IOLMaster on the instrument table
3. Printer
4. Keyboard support
5. Narrow holding bracket for securing the IOLMaster on the keyboard support
6. Paper pads for patient chinrest
7. Power isolation transformer for connection of external accessory units
8. Network isolator
9. Software option A plus
10. Software option B
11. Connecting cable for coupling with PC.

The various parts are highlighted in the line diagrams.

PATIENT POSITIONING[14,15]

Adjust the instrument table, the headrest, and IOLMaster according to the patient's height so that they can sit comfortably in a stable position. The whole procedure is explained in detail to the patient. It is necessary not to move the head or blink during the procedure. Let the patient blink before starting the measurement. They should fix steadily to the fixation light. The eyelid or eyelashes must not obscure light spots seen on the screen. If necessary, the patient should be asked to open the eye widely or assist them manually to keep it open. Measurement started with a centered and focused beam. If the signal quality is worse, then decentering and defocussing should be done and try again. A total of 3–5 measurements can be done per eye. If IOLMaster does not display a mean value or inconsistent reading is measured, find any posterior segment pathology or verify each measurement trace. Eventually then, zoom in and shift manually the measurement cursor for obtaining a correct signal [Figure 4].

USES OF IOLMASTER[14,15][FIGURES 5-10]

1. AL measurements
2. Keratometry (corneal curvature)
3. ACD measurements
4. White-to-white diameter (WTW) (optional)
5. Calculation of IOL power.

Description

Axial length

The measurement is based on the patented interference optical method known as partial coherence interferometry.



Figure 1: Gross image of IOLMaster 700

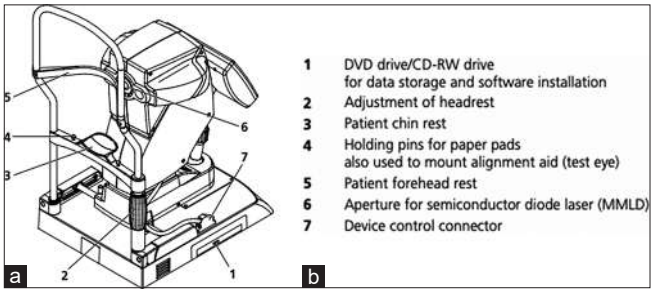


Figure 2: (a) Various components of IOLMaster, (b) description of various components of IOLMaster

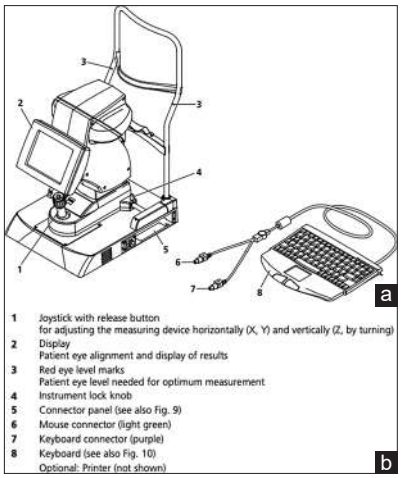


Figure 3: (a) Lateral view image of IOLMaster showing various components, (b) description of various components of image 3a



Figure 4: Image depicting patient positioned comfortably in front of IOL Master

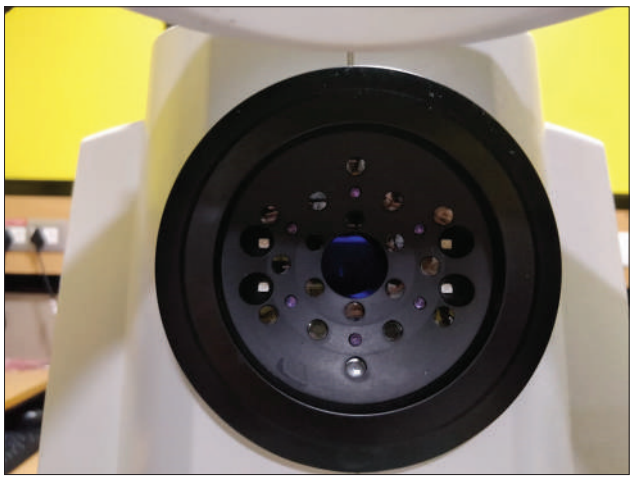


Figure 5: Head-on view of IOLMaster 700

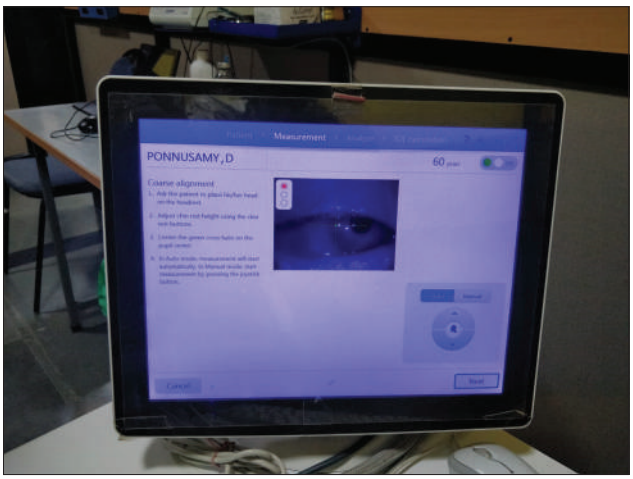


Figure 6: Coarse alignment with incorrect readings

ALM modes:

1. Phakic
2. Pseudophakic
3. Aphakic
4. Silicone-filled eyed.

Keratometry

It is determined by measuring the distance between reflected light images projected onto the cornea.

The IOLMaster requires three keratometry readings for the final calculation. The IOLMaster reflects six points of light, arranged in a 2.3-mm diameter hexagonal pattern (measured by digital calipers) from the air/tear film interface. The instrument's internal software measures the separation of opposite pairs of lights objectively and hence, the toroidal surface curvatures are calculated.

Anterior chamber depth

It is measured as the distance between the optical sections of the crystalline lens and the cornea produced by lateral slit illumination.

When ACD mode is turned on, the lateral slit lamp is automatically turned on. This illumination thus produced subjectively appears to be very bright to the patients.

The ACD can be measured only on phakic eyes. If ACD measurements are performed on pseudophakic eyes, it will result in measuring errors and/or incorrect readings. The readings for pseudophakic eyes do not reflect the ACD correctly.

Eventually, five ACD readings will be taken together with the calculated mean.

White-to-white diameter

It is determined from the image of the iris.



Figure 7: Fine alignment with correct keratometry and anterior chamber volume. Also, the signal-to-noise ratio is within normal limit



Figure 8: Another example of the fine alignment of IOLMaster when a patient is being evaluated



Figure 9: How to measure correctly white-to-white distance

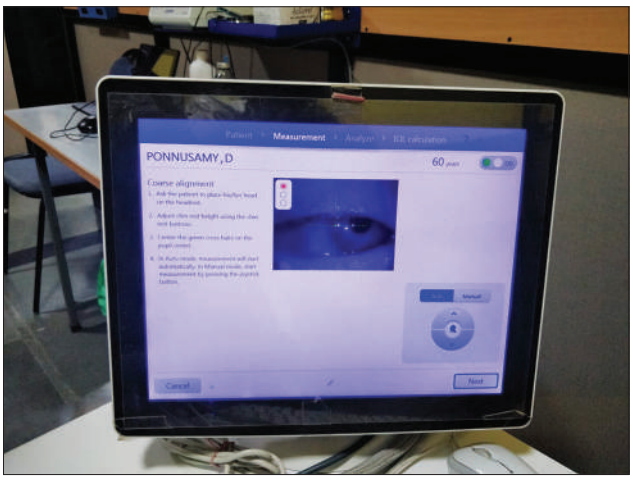


Figure 10: All the measurements done by IOLMaster with fine alignment

Method of measurement:

1. The patient is asked to look at the fixation point
2. The point of focus is iris and not the spotlight
3. The validity of the WTW determination depends on this check of the correct recognition of the iris edge image
4. The WTW is the horizontal diameter of the iris
5. In addition to the measured WTW value, the deviation of the visual axis from the center of the iris (X, Y) will also be displayed.

Intraocular lens power calculation

1. Depending on the IOL calculation formula, once all the measurements have been taken, options can be generated for IOL to be implanted
2. Click on the appropriate option to select the desired formula
3. The IOL Haggis, Hoffer Q, Holladay, SRK 2, and SRKT formulas are implanted as standard
4. After refractive corneal surgery, the Haggis-L formulae may be selected.

Another important entity to be kept in mind during the measurement is SNR (signal to noise) ratio. In that:

a. Red indicates incorrect value
b. Yellow indicates uncertain value/borderline
c. Green indicates good value.

ADVANTAGES OF IOLMASTER

1. Easy to learn and quick time measurements can be done
2. Noncontact measurements
3. Noninvasive
4. Ultra-high-resolution biometry
5. Extensive integrated safety features which make measurements less cumbersome
6. It has the advantage of imaging fovea in case of posterior staphyloma
7. It gives the true refractive length rather than anatomical AL

8. The accuracy of IOLMaster is 0.02 μm which is operator dependent
9. Highly ametropic patients with refractive error can wear glasses while sitting on the IOLMaster which aids in fixation.

LIMITATIONS

1. In patients with media opacities such as corneal opacities, dense cataract, NS iv grade, and posterior polar cataract, it cannot measure AL
2. In case of vitreous hemorrhage, again AL cannot be measured
3. Difficulty in measuring AL in infants, small children, and mentally disabled patients.

INTRAOCULAR LENS POWER CALCULATION WITH ULTRASOUND (A-SCAN) BIOMETRY

The three major aspects of intraocular lens power calculation are:[16]

a. Biometry
b. IOL power calculation formulae
c. Clinical variables – IOL power calculations in special situations.

BIOMETRY INCLUDES:[16]

- AL measurement
- Keratometry (K-reading) – corneal power
- Effective IOL position.

A-SCAN BIOMETRY[16]

While performing A-scan biometry, a thin parallel sound beam is emitted from the probe tip at a given frequency of approximately 10 MHz, with an echo bouncing back into the tip of the probe as the sound beam strikes each interface.

An interface is defined as the junction between any two media of different densities and velocities.

In the eye, these interfaces can be divided into:

a. Anterior corneal surface
b. Aqueous/anterior lens surface
c. Posterior lens capsule/anterior vitreous
d. Posterior vitreous/retinal surface
e. Choroid/anterior scleral surface.

KERATOMETRY[16]

The process of measuring the curvature of the cornea is called keratometry.

By definition, a keratometer (ophthalmometer) is a noninvasive diagnostic instrument for assessing the curvature of the anterior surface of the cornea. It is used particularly for assessing axis and the extent of astigmatism [Figure 11].

KERATOMETRY CAN BE PERFORMED BY:

a. Manual topography
b. Autokeratometer
c. IOLMaster/Lenstar 900.

A-scan measurement techniques can be divided into:[16]

Contact technique

Applanation method

By the applanation biometry method, an ultrasound probe is placed directly on the cornea which is attached to a slit lamp.

Handheld method

After explaining the procedure to the patient under topical anesthesia, clean the probe. The probe is placed on the patient's cornea which is attached to a device that delivers adjustable sound waves. The measurements are displayed as spikes on the screen of the monitor. The appearance of the spikes and the distance between them can be correlated to structures within the eye and the distance between them. The operator while performing the handheld method aims the probe toward the macula of the eye. The alignment with the optical axis is indicated by high lens spikes and a high retina spike on the scan graph [Figure 12a-d].

Immersion technique

Immersion A-scan biometry

The immersion technique employs the use of a Prager Scleral Shell (ESI, Inc, 2915 Everest lane N, Plymouth, Mn 55447). The immersion technique is accomplished by placing a small scleral shell between the eyelids of the patient, filling it with saline and then immersing the probe into the fluid, being careful to avoid contact with the cornea. It is more



Figure 11: Mid-level ophthalmic personnel performing keratometry on the patient

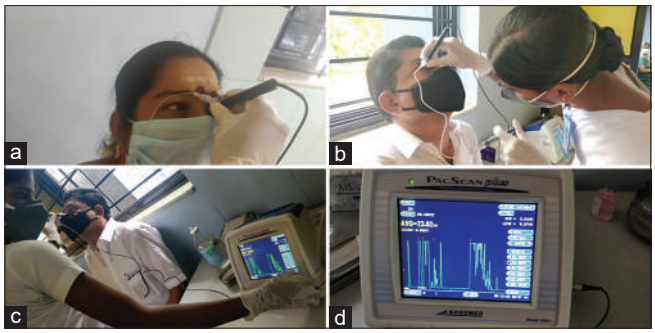


Figure 12: (a) Image of the patient depicting the positioning of the probe on the ocular surface, (b) handheld method of A scan – the probe is placed on the patient's cornea which is attached to a device that delivers adjustable sound waves, (c) display monitor on which readings are noted for axial length and keratometry, (d) the alignment with the optical axis is indicated by high lens spikes and a high retina spike on the scan graph

accurate than contact method because corneal compression is avoided. The eyes measured with this method are, on an average, 0.1–0.3 mm longer.

INTRAOCULAR LENS POWER CALCULATION FORMULAE[16]

- Theoretical formulae – Binkhorst, Modified Binkhorst, Colenbrander–Hoffer, Gill's, Clayman's, and Fyodorov formulae
- Regression formulae – SRK-I, SRK-II, Modified SRK-II, Holladay-I, Holladay-II, and Hoffer-Q
- Most commonly used- SRK II formula: $P = A - 2.5L - 0.9K$.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Sheng H, Bottjer CA, Bullimore MA. Ocular component measurement using the Zeiss IOLMaster. Optom Vis Sci 2004;81:27-34.
2. Santodomingo-Rubido J, Mallen EA, Gilmartin B, Wolffsohn JS. A new non-contact optical device for ocular biometry. Br J Ophthalmol 2002;86:458-46.
3. Drexler W, Findl O, Menapace R, Rainer G, Vass C, Hitzenberger CK, *et al*. Partial coherence interferometry: A novel approach to biometry in cataract surgery. Am J Ophthalmol 1998;126:524-34.
4. Mandal P, Berrow EJ, Naroo SA, Wolffsohn JS, Uthoff D, Holland D, *et al*. Validity and repeatability of the Aladdin ocular biometer. Br J Ophthalmol 2014;98:256-8.
5. Santodomingo-Rubido J, Villa-Collar C, Gilmartin B, Gutiérrez-Ortega R. Myopia control with orthokeratology contact lenses in Spain: Refractive and biometric changes. Invest Ophthalmol Vis Sci 2012;53:5060-5.
6. Buckhurst PJ, Wolffsohn JS, Shah S, Naroo SA, Davies LN, Berrow EJ, *et al*. A new optical low coherence reflectometry device for ocular biometry in cataract patients. Br J Ophthalmol 2009;93:949-53.
7. Rohrer K, Frueh BE, Walti R, Clemetson IA, Tappeiner C, Goldblum D, *et al*. Comparison and evaluation of ocular biometry using a new noncontact optical low-coherence reflectometer. Ophthalmology 2009;116:2087-92.
8. Shirayama M, Wang L, Weikert MP, Koch DD. Comparison of corneal powers obtained from 4 different devices. Am J Ophthalmol 2009;148:528-35.e1.
9. Goebels SC, Seitz B, Langenbucher A. Comparison of the new biometerOA-1000 with IOLMaster and Tomey AL-3000. Curr Eye Res 2013;38:910-6.
10. Kunert KS, Peter M, Blum M, Haigis W, Sekundo W, Schütze J, *et al*. Repeatability and agreement in optical biometry of a new swept-source optical coherence tomography-based bio-meter versus partial coherence interferometry and optical low-coherence reflectometry. J Cataract Refract Surg 2016;42:76-83.
11. Srivannaboon S, Chirapapaisan C, Chonpimai P, Loket S. Clinical comparison of a new swept-source optical coherence tomography-based optical bio-meter and a time-domain optical coherence tomography-based optical bio-meter. J Cataract Refract Surg 2015;41:2224-32.
12. Hoffer KJ, Hoffmann PC, Savini G. Comparison of a new optical biometer using swept-source optical coherence tomography and a biometer using optical low-coherence reflectometry. J Cataract Refract Surg 2016;42:1165-72.
13. Arriola-Villalobos P, Almendral-Gomez J, Garzon N, Ruiz-Medrano J, Fernández-Pérez C, Martínez-de-la-Casa JM, *et al*. Agreement and clinical comparison between a new swept-source optical coherence tomography-based optical biometer and an optical low-coherence reflectometry biometer. Eye (Lond) 2017;31:437-42.
14. Joel H. Emerson and Kelly Tompkins, Clinical Application Specialists. IOLMaster a Practical Operation Guide. Carl Zeiss Meditec; 2009.
15. IOLMaster with Advanced Technology Software Version 5.4. User Manual, 2008.
16. Khurana AK Theory and Practice of Optics and Refraction 3rd ed, 2013, 274-82.

Comment on "Role of mitomycin C in the management of ocular surface squamous neoplasia"

Dear Editor,

We read the article by Menaka *et al*.[1] with great interest and at the outset, we must congratulate the authors for the important analysis. However, we have few important observations and suggestions to make.

Firstly, the authors have mentioned that "A prospective study was conducted for a period of 1 year at a tertiary care hospital in Chennai." It will great if the authors mention the time period of the study (year) because during this corona pandemic most of the prospective studies are at halt and if done during this period, the importance of hand hygiene, wearing mask, and social distancing should be highlighted in the study. This needs clarification.

Secondly, the authors have included patients above the age of 40 years only and all systemic comorbidities were excluded. It would be good to know why young patients were excluded, as they form a good chunk of ocular surface squamous neoplasia (OSSN) cases and also there will be less systemic comorbidities.[2]

Thirdly, it will be great to know, whether all patients underwent human immunodeficiency virus TRI-DOT test or they were excluded based on history alone. Moreover, whether anterior segment optical coherence tomography was a part of routine evaluation as it is a very good tool to assess the depth of OSSN apart from ultrasound biomicroscopy.[3] This will be interesting to know for the readers.

Lastly, the authors have used mitomycin C (MMC) for two cycles at an interval of 1 week. At our center, we are using three cycles of MMC 0.04% four times/day for 1-week on and 1-week off regimen along with topical steroids and lubricants.[4]

We totally agree with the authors that the use of topical 0.04% MMC has shown good clinical results without any serious side effects with a very less recurrence rate at the end of 1-year follow-up when used as alternate 7-day courses. More number of large-sample multicentric studies with a longer follow-up period are warranted to substantiate the role of MMC.

Acknowledgments

The authors would like to thank the management of Aravind Eye Hospital and Post Graduate Institute of Ophthalmology, Puducherry.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Bharat Gurnani, Kirandeep Kaur[1]

Departments of Cornea and Refractive Services and [1]Pediatric Ophthalmology and Strabismus, Aravind Eye Hospital and Post Graduate Institute of Ophthalmology, Puducherry, India

Address for correspondence: Dr. Kirandeep Kaur, Aravind Eye Hospital and Post Graduate Institute of Ophthalmology, Puducherry - 605 007, India.
E-mail: beingkirandeep@gmail.com

REFERENCES

1. Menaka C, Perumal V, Shankar R. Role of mitomycin C in the management of ocular surface squamous neoplasia. Kerala J Ophthalmol 2020;32:143-7.
2. Pe'er J. Ocular surface squamous neoplasia. Ophthalmol Clin North Am 2005;18:1-13.
3. Thomas BJ, Galor A, Nanji AA, El Sayyad F, Wang J, Dubovy SR, *et al*. Ultra high-resolution anterior segment optical coherence tomography in

the diagnosis and management of ocular surface squamous neoplasia. Ocul Surf 2014;12:46-58.
4. Chen C, Louis D, Dodd T, Muecke J. Mitomycin C as an adjunct in the treatment of localised ocular surface squamous neoplasia. Br J Ophthalmol 2004;88:17-8.

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

Submitted: 26-Aug-2020 **Accepted:** 27-Aug-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	**Quick Response Code**
DOI: 10.4103/kjo.kjo_126_20	

How to cite this article: Gurnani B, Kaur K. Comment on "Role of mitomycin C in the management of ocular surface squamous neoplasia". Kerala J Ophthalmol 2020;32:329-30.

© 2020 Kerala Journal of Ophthalmology | Published by Wolters Kluwer - Medknow

Comment on ' Comparison of the scleral tunnel constructed with crescent versus razor blade using the anterior segment optical coherence tomography"

Dear Sir,

We agree that manual small-incision cataract surgery (MSICS) has emerged as the most relied surgery for cataract in developing countries like India.[1] MSICS has come a long way, with self-sealing wounds, capsulorhexis, and in the bag placement of intraocular lens.[2] We read the article by Murthy and Sudhakar[3] with great interest, and we would like to congratulate the authors for bringing out this novel study.

However, we have some important observations, and we would like the authors to throw light on these. First, in the abstract, one case of premature entry has been mentioned, and the same has not been highlighted in the result section. It would be great if authors can give details of this. It will be good to know how much was the baseline incision length constructed.

Second, the tunnel construction has been described with blade versus crescent. As per the conventional teaching, angulated instruments (such as the crescent knife and keratome) are used for MSICS triplanar wound construction. In the study, the authors have described scleral tunnels using razor blade, which is a much sharper and straight instrument. The same has been described in literature.[4] We would like to know if this is regularly followed for routine cases and how they fashioned self-sealing incisions with the straight blade. Moreover, the blade being a sharp instrument might be associated with higher complication rates of premature entries and Descemet membrane detachment (DMD). It will be great if authors can share their experience and challenges faced, especially in young, less experienced hands.

It is an interesting finding that the time taken to construct tunnels with a crescent knife (32.9 s) was more as compared to that of a blade (27.3 s). It would be interesting to know how standardization was done for recording the time accurately.

The authors have mentioned that tunnels made with blade were deeper as compared to crescent. It will be good to have details about the depth of tunnel in the cases that required suturing or if any intraoperative positive pressure or iris prolapse was experienced in these cases. This can help in enhancing the understanding of readers that suturing was required because of problem with depth or because of inadequate tunnel width (measured from the scleral incision to the corneal entry point), if this was measured.

It will also be good if the authors can highlight details of the patient who had DMD. The details of the depth of external and internal tunnel in this case will be very important. It would be great to know that DMD was detected intraoperatively or was diagnosed on the postoperative day 1, based on ocular coherence tomography.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Kirandeep Kaur[1,2], Bharat Gurnani[1,2]

[1]Department of Cataract, Cornea and Refractive Services-Bharat Gurnani, [2]Department of Pediatric and Squint Services-Kirandeep, Aravind Eye Hospital, Puducherry, India.

Address for correspondence: Dr. Bharat Gurnani, Aravind Eye Hospital, Thavalakuppam, Cuddalore main road, Pondicherry, India.
E-mail: drgurnanibharat25@gmail.com

REFERENCES

1. Gogate PM, Deshpande M, Wormald RP. Is manual small incision cataract surgery affordable in the developing countries? A cost comparison with extracapsular cataract extraction. Br J Ophthalmol 2003;87:843-6.
2. Gurung R, Hennig A. Small incision cataract surgery: tips for avoiding surgical complications. Community Eye Health 2008;21:4-5.
3. Murthy SR, Sudhakar P. Comparison of the scleral tunnel constructed with crescent versus razor blade using the anterior segment optical coherence tomography. Kerala J Ophthalmol 2020;32:166-70.
4. Olu ME. Wound geometry as it relates to tunnel valvular competence in manual small incision cataract surgery. Niger J Ophthalmol 2014;22:1-6.

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

Submitted: 29-Aug-2020 **Accepted:** 01-Sep-2020 **Published:** 23-Dec-2020

Access this article online	
Website: www.kjophthal.com	**Quick Response Code**
DOI: 10.4103/kjo.kjo_130_20	

How to cite this article: Kaur K, Gurnani B. Comment on ' Comparison of the scleral tunnel constructed with crescent versus razor blade using the anterior segment optical coherence tomography". Kerala J Ophthalmol 2020;32:331-2.

© 2020 Kerala Journal of Ophthalmology | Published by Wolters Kluwer - Medknow