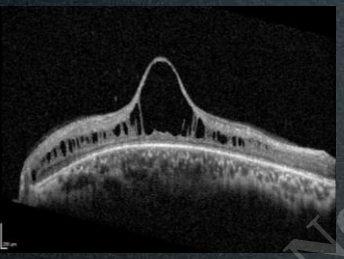
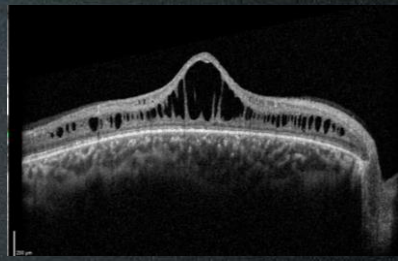
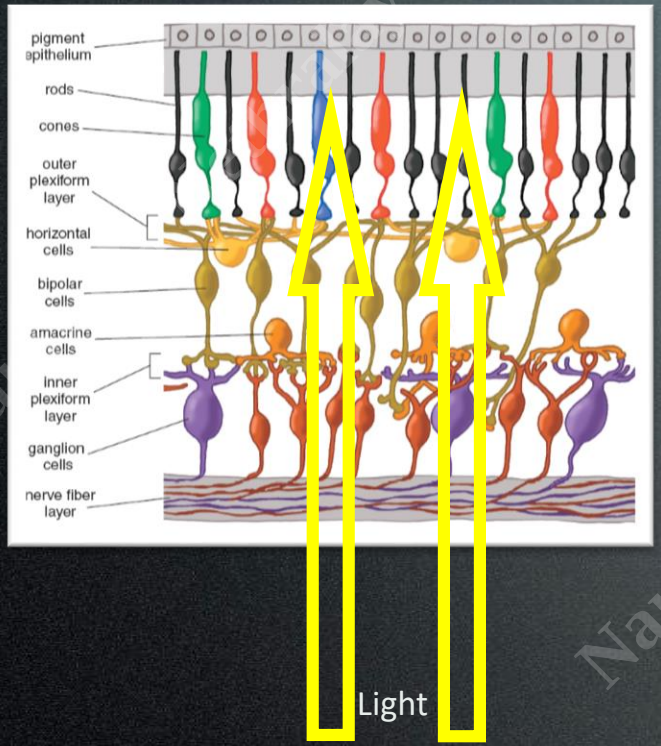


Retinal electrophysiology



Dr Poornachandra B

Consultant Vitreo Retinal Surgeon
Dept of Electrophysiology,
Retinal dystrophy & genetics clinic
Narayana Nethralya Eye Institute, Bangalore



Biochemical and ionic changes in photoreceptors and RPE

Electrical potentials

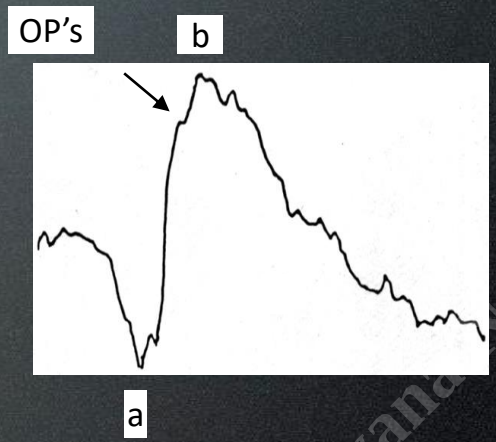
Recorded as Electroretinogram

Mass response of the retina to a luminance stimulus.....

Origin of ERG waveform

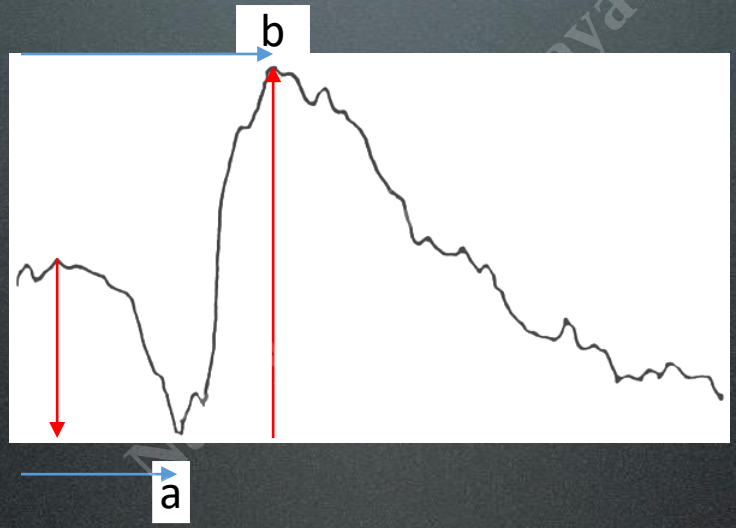
- a-wave : Photoreceptors
- b-wave : Muller cells

  Bipolar cells
- OP's : Amacrine cells



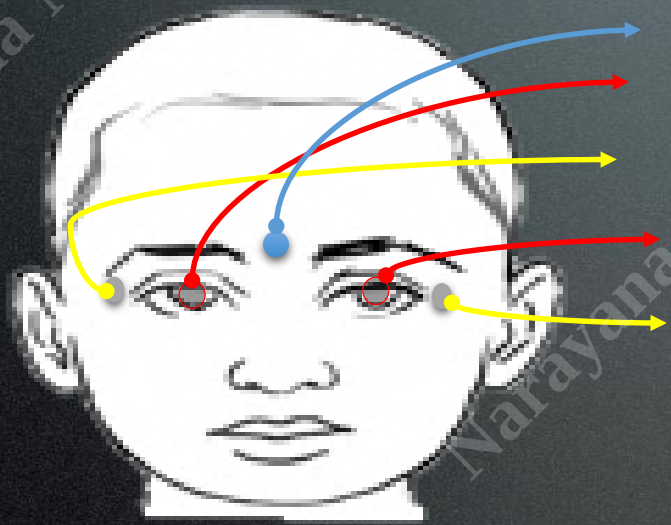
Implicit time

Amplitude



ERG Waveform

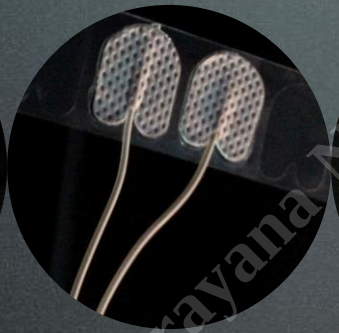
Electrode Placement



- Neutral
- Right Active (+) - Recording
- Right Reference (–)
- Left Active (+) - Recording
- Left Reference (–)

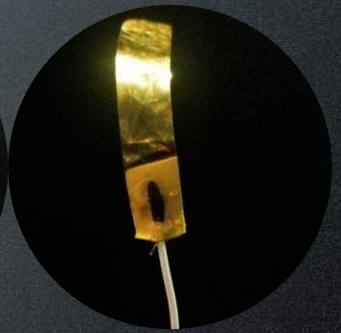
Types of Electrodes Used

Burian-Allen electrode



Skin electrodes

Jet electrode



Gold foil electrode

Dark adopted responses (20 mins of dark adaptation)

- Rod response

- Combined response

- Oscillatory potentials

Light adopted responses (10 mins of light adaptation)

- Single flash cone response

- 30 Hz flicker

ISCEV Recommendation

Nomenclature

(1) Dark-adapted 0.01 ERG (rod response)

(2) Dark-adapted 3.0 ERG (combined rod–cone response)

(3) Dark-adapted 3.0 oscillatory potentials

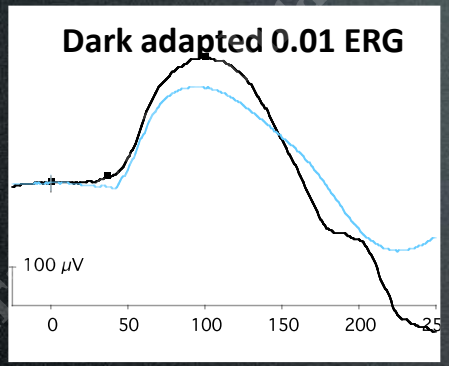
(4) Light-adapted 3.0 ERG (cone response)

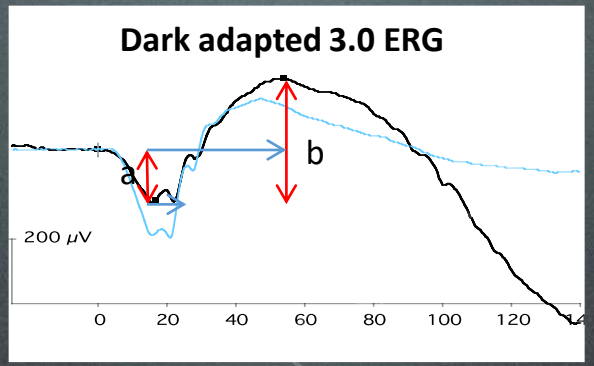
(5) Light-adapted 3.0 flicker (30 Hz flicker)

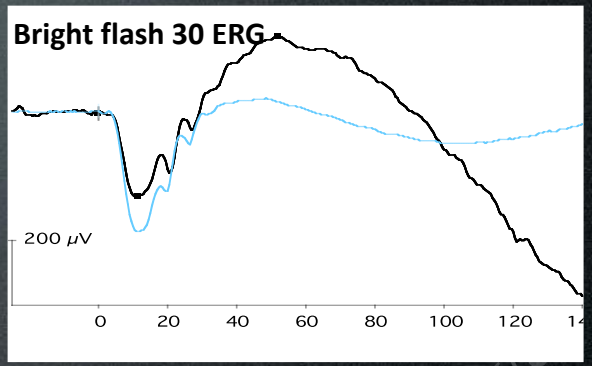
(6) An additional Dark-adapted 10.0 ERG or Dark-adapted 30.0 ERG response is recommended

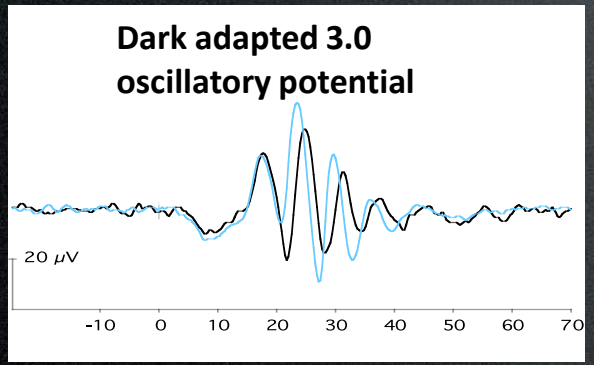
ISCEV: www.iscev.org

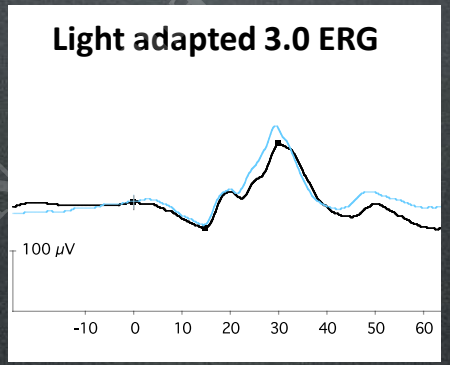
Normal Electroretinogram

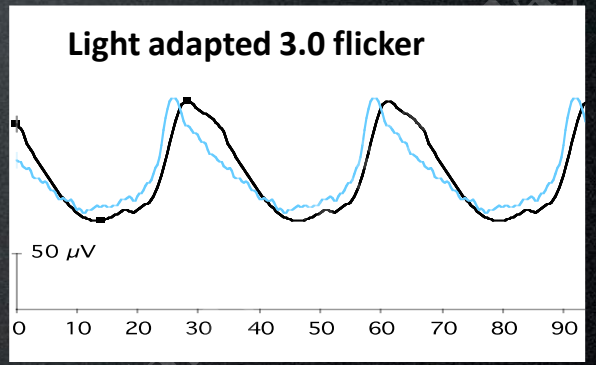








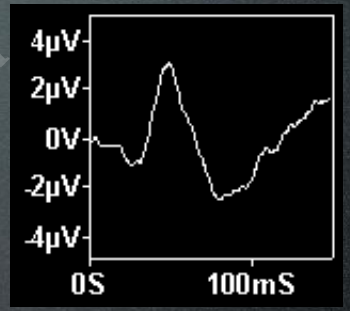




Non standard tests

- Red flash ERG- scotopic
- On- off ERG- Orange 200 ms, green background
- S-cone ERG- Blue 5 ms, Orange background

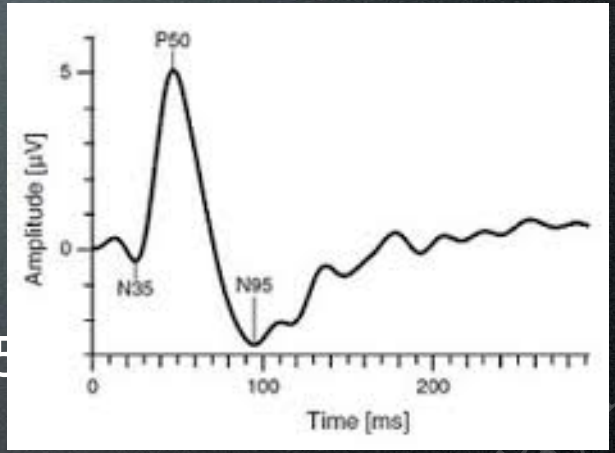
Pattern ERG



- Contrast response to a checkerboard
- No contribution from the peripheral retina
- P50 - macular function
- N95 - ganglion cell function

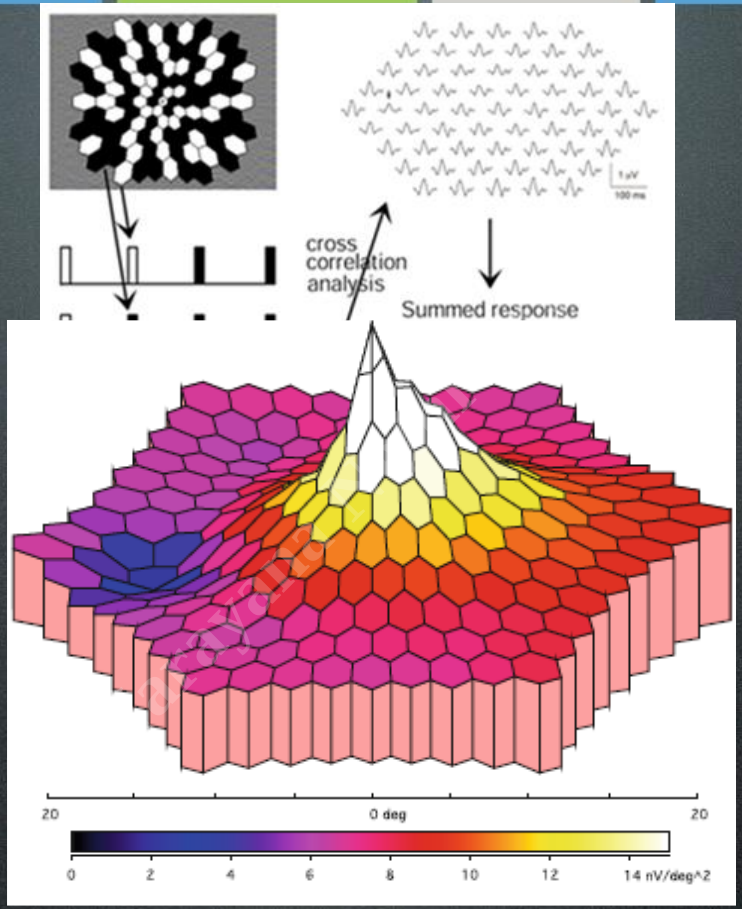
PERG

- Assessment of macular function -P50
- Assessment of retinal ganglion cells- N95
- Complements VEP- distinguishes between optic neuropathy and maculopathy
- Complements full field ERG- involvement or sparing of macula



Multifocal ERG

- MfERG allows for detection of small foci of retinal dysfunction
- The retina is stimulated with an assortment of 61 or 103 hexagonal elements with a central fixation point
- 50% of the elements are illuminated at one point of time.

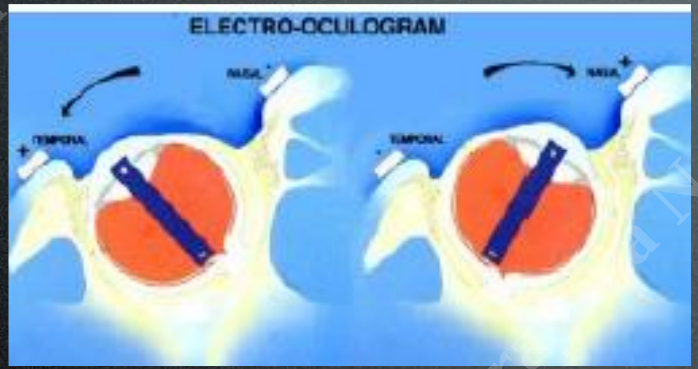


Indications

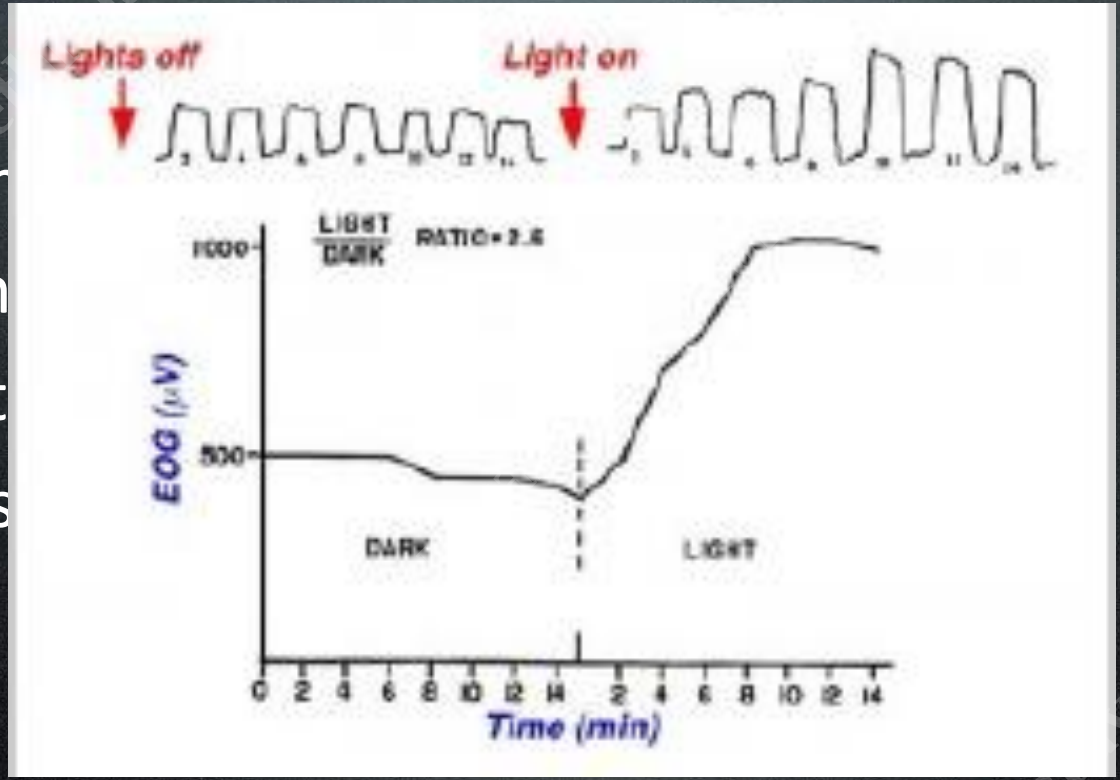
- Age-related macular degeneration
- Macular holes
- Hydroxychloroquine,Chloroquine, Ethambutol, Thiothixene toxicity
- Retinitis pigmentosa
- Branch retinal artery occlusion
- Fundus flavimaculatus, Stargardt’s disease
- Acute idiopathic blind spot enlargement

Electrooculogram (EOG)

- It measures the potential between the cornea and Bruch's membrane at the back of the eye.
- 5 millivolts



- Arder
- Norm
- Less t
- Best’s

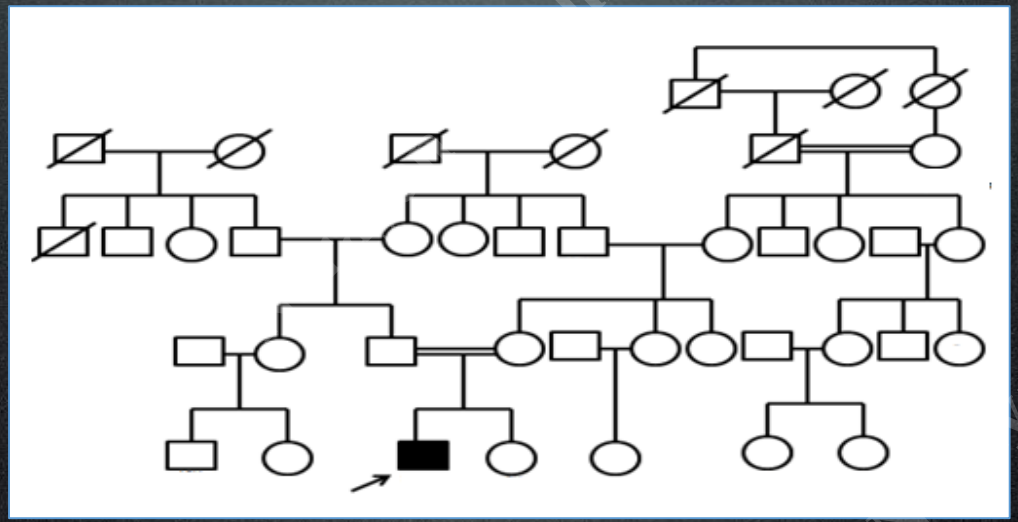


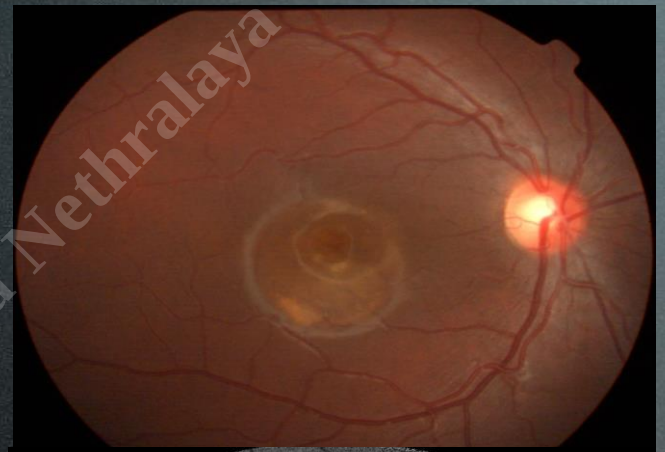
ERG - Reporting

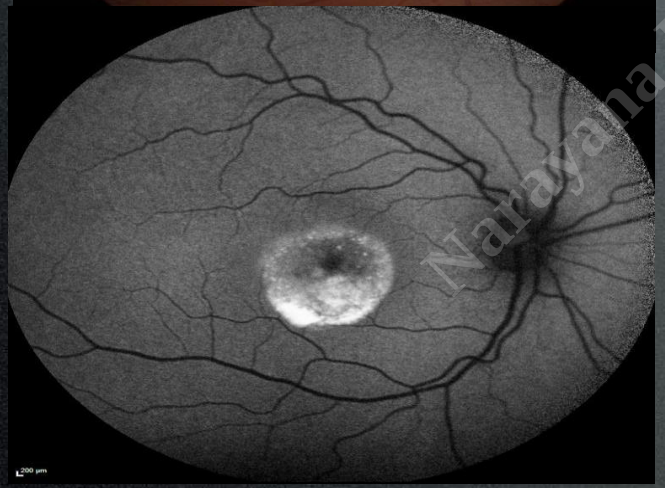
- Rods more affected
- Cones more affected
- Both affected : Rod - Cone / Cone -Rod
- Negative waveform

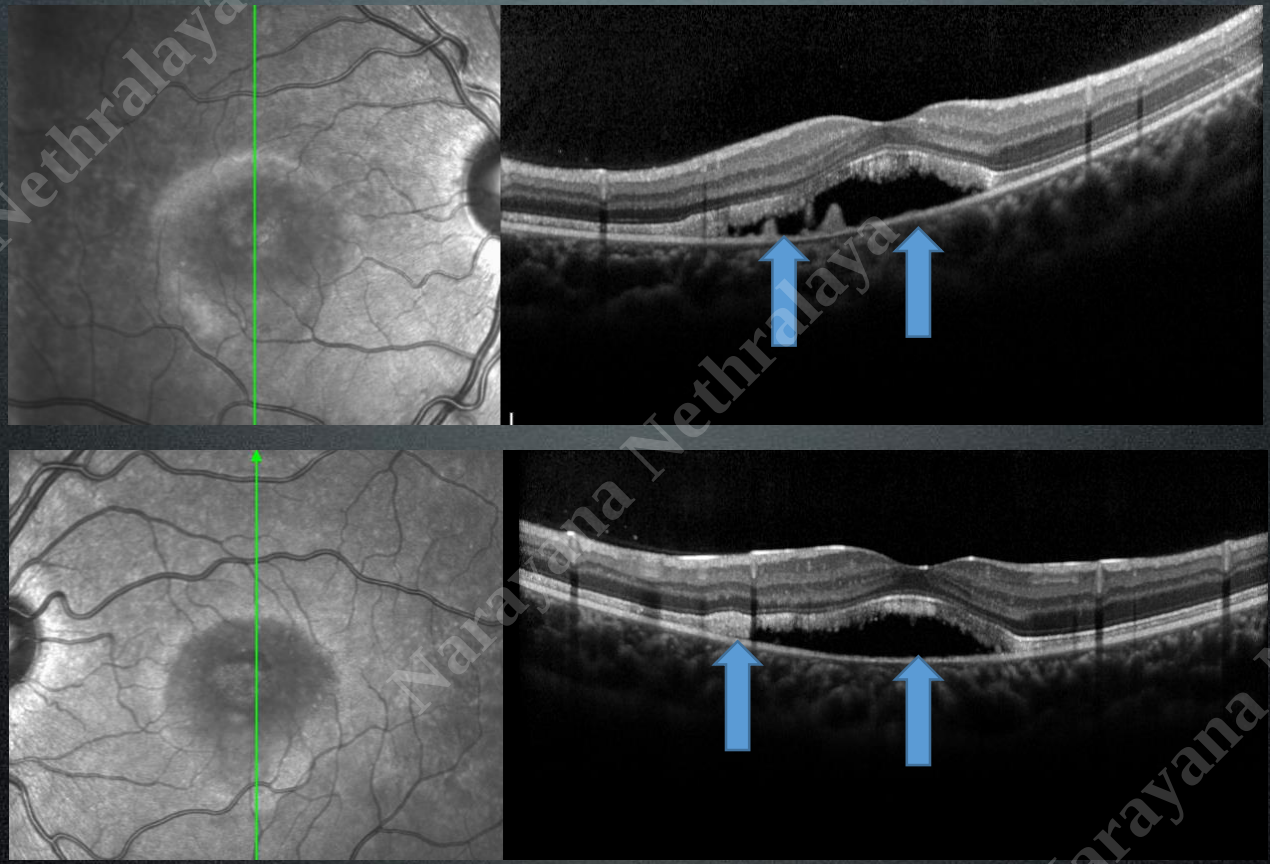
Case 1

- 11 year old male
- Blurred vision
- BCVA – 6/9 OU

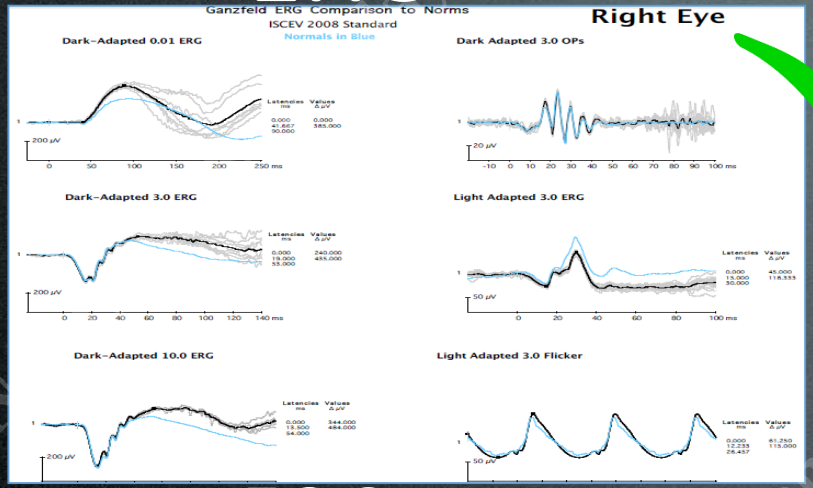


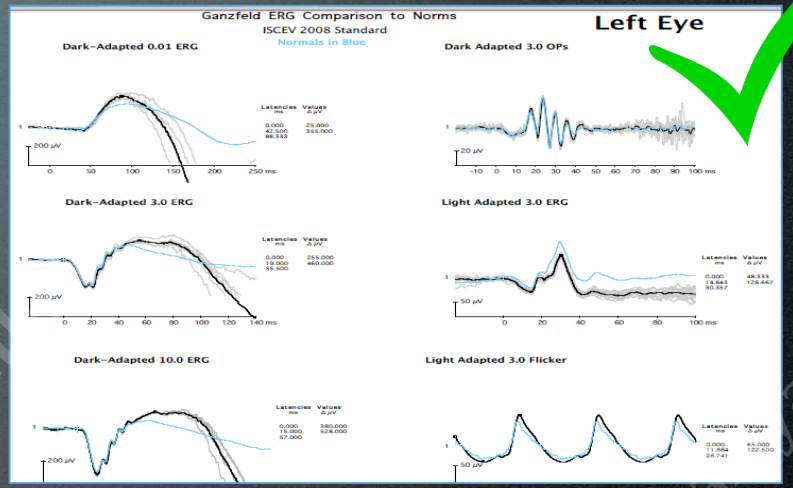




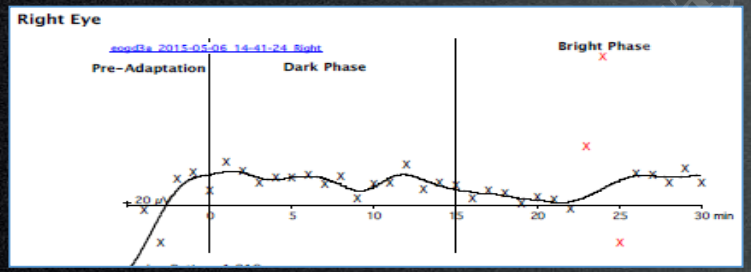


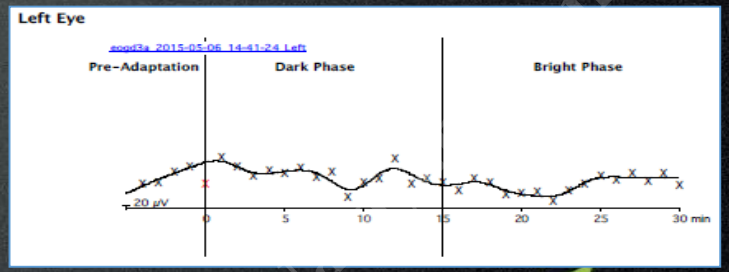
ERG

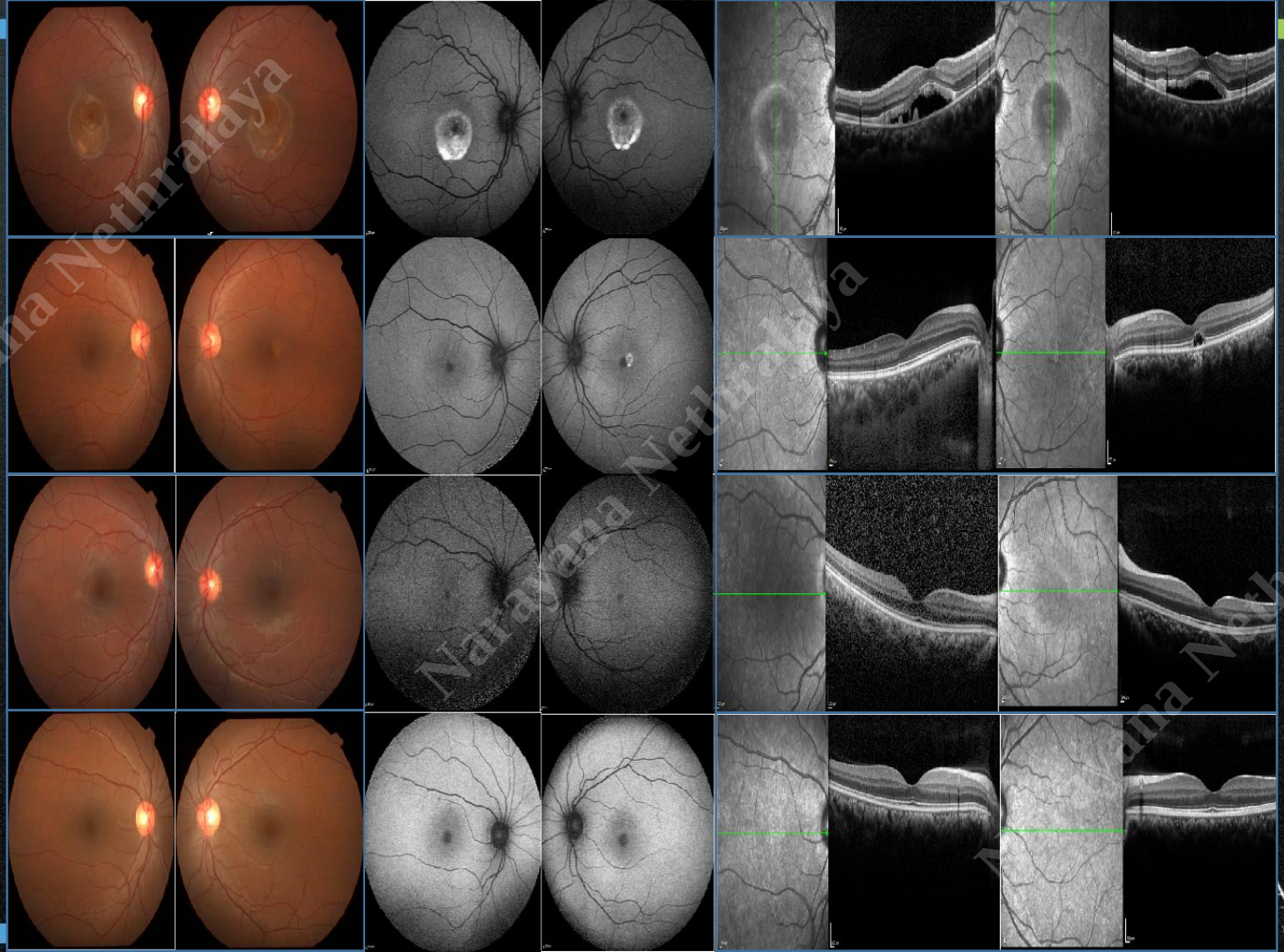




EOG







- The proband was found to have a heterozygous mutation in the *BEST1* gene.
- suggesting an Autosomal Dominant inheritance pattern

BEST MACULAR DYSTROPHY

BEST MACULAR DYSTROPHY

- AD, mutation in the gene BEST1(VMD2) ON 11q13
- Most common Mendelian macular dystrophies- 1 in 10, 000 individuals
- The macular lesions - characteristic of the disease "vitelliform" -egg-yolk-like appearance
- 20% develop CNVM in 1 eye

Miller sa,Bresnick GH,Chandra SR choroidal neovascular membrane in Best disease.Am j ophthalmol 1976;82(2);252-5

STAGES

- pre- vitelliform stage– subtle RPE chnages,EOG abnormal
- vitelliform stage – round,elevated yellow macular lesion described as classic ‘egg yolk’ appearance d/t accumalation of lipofuscin in RPE
- psuedohypopyon – at puberty, yellow material when break through RPE and accumlate in subretinal space with a fluid level formation
- Vitelliruptive-lesion breaks up with pigment clumping and early atrophic changes-’scrambled egg appearance’
- Atrophy stage-RPE atrophy
- CNVM-cnvm devlops with subretinal fibrous scar

- Visual acuity - 20/20 or better in eyes with undisturbed vitelliform lesions till first six decades of life
- visual loss occurs -when complicated by nodular fibrosis, choroidal neovascularization or central geographic atrophy
- Hyperopia with narraow angles may be present

Fishman GA,Baca W,Alexander KR,et al.visual acuity in patients with best vitelliform macular dystrophy.ophthalomolgy 1993:100(11);1665-70

Case 2

-10 year old male

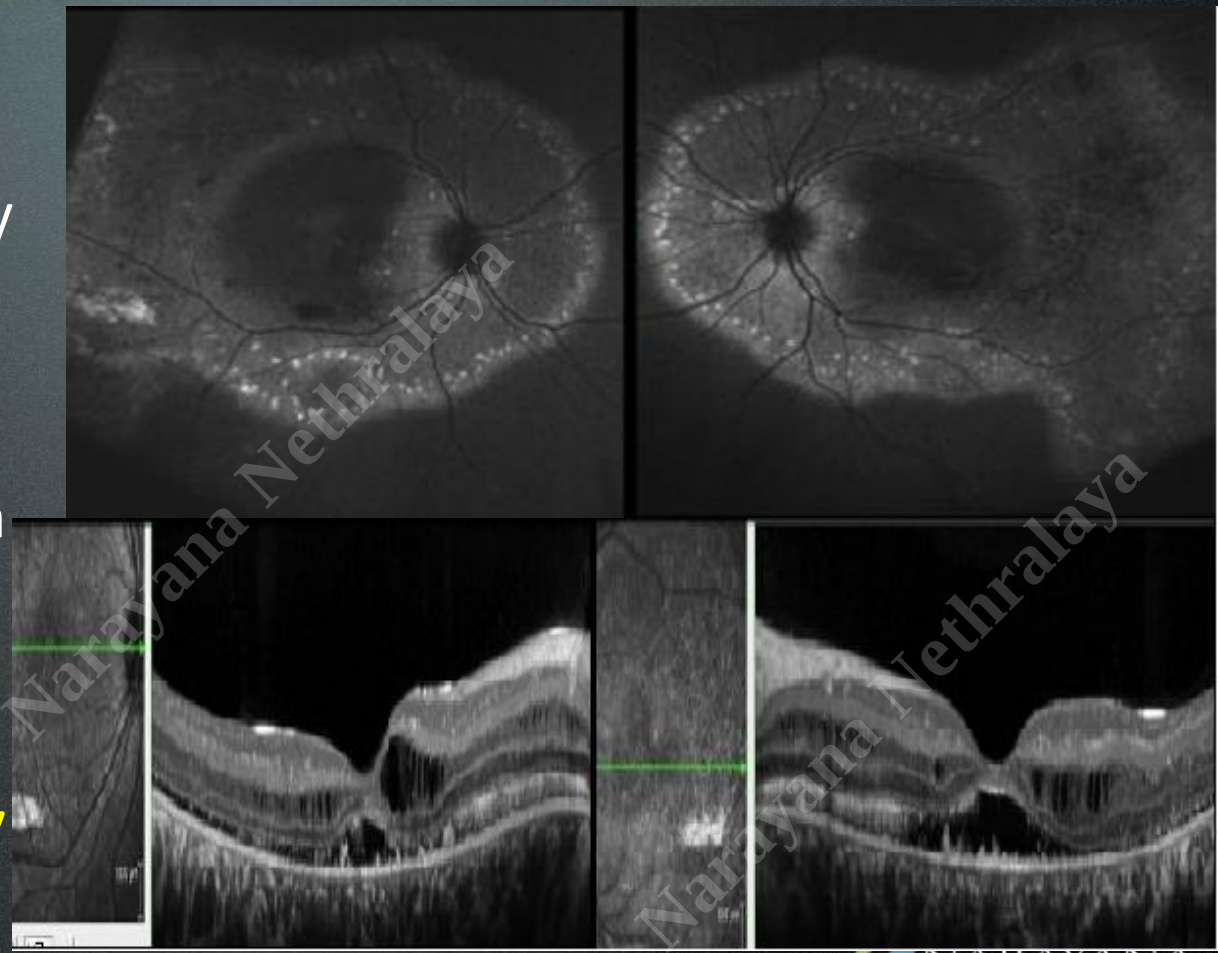
-Born out of consanguineous marriage

- Blurred vision, BCVA 6/18(p) OU, Hypermetropic refractions

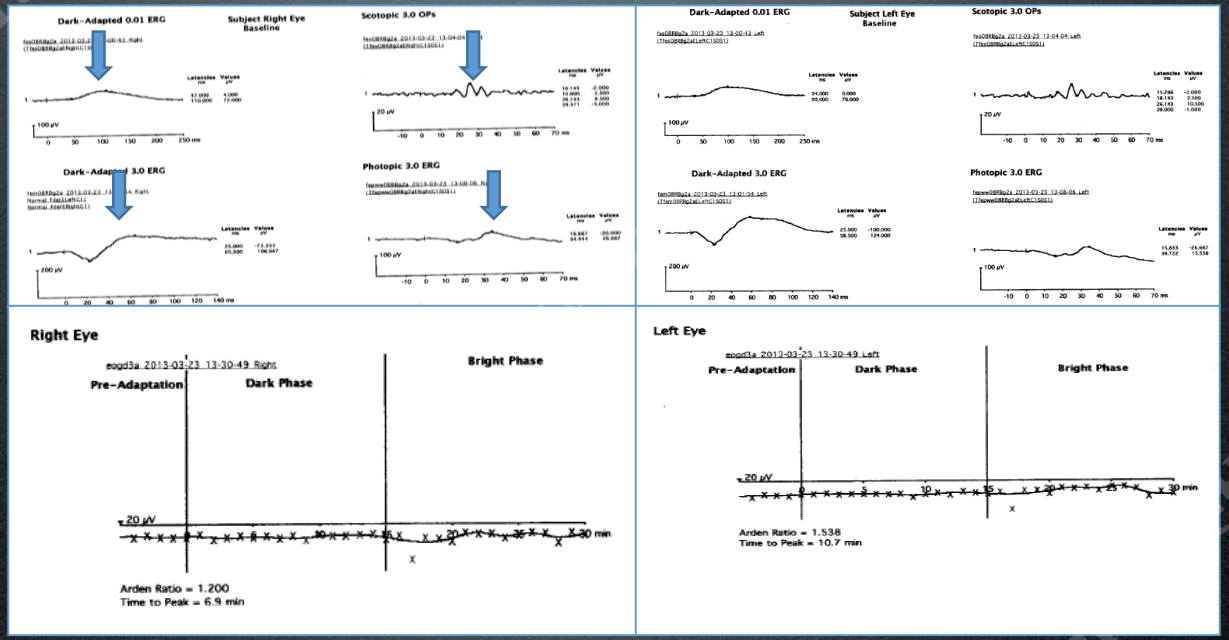
- Narrow angles,

- Sub-retinal lipofuscin deposits, predominantly outside the macula,
- Accumulation of fluid within and/or beneath the neurosensory retina

Autosomal recessive bestrophinopathy (ARB)



- Reduced ERG amplitudes
- Absence of light rise in EOG



Genetic test results

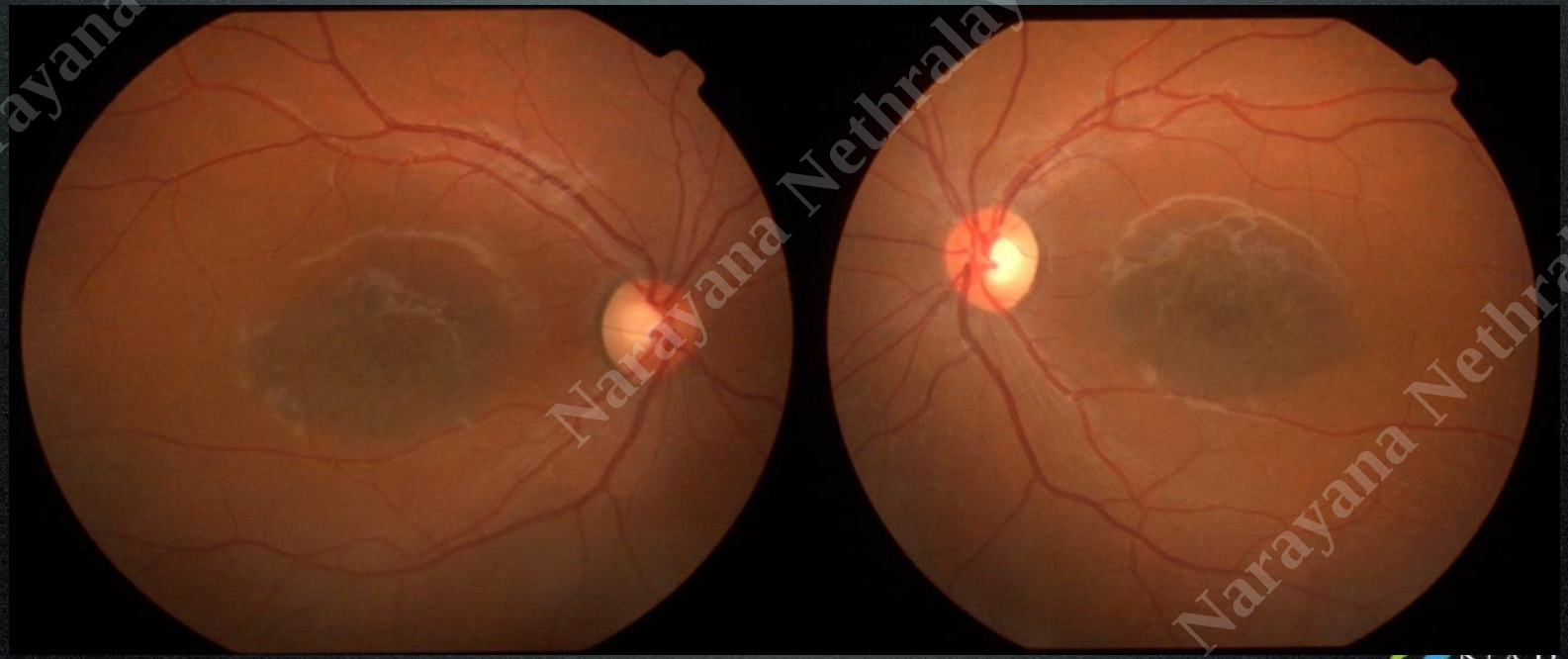
- The proband was identified with homozygous mutation in *BEST1* gene.
- The proband's unaffected sibling had normal genotype
- Unaffected parents were heterozygous carriers for this mutation.

- Primarily of recognizing choroidal neovascularization and hastening its regression with anti-VEGF therapy
- caution against playing contact sports
- Protective eyewear is recommended for all sports

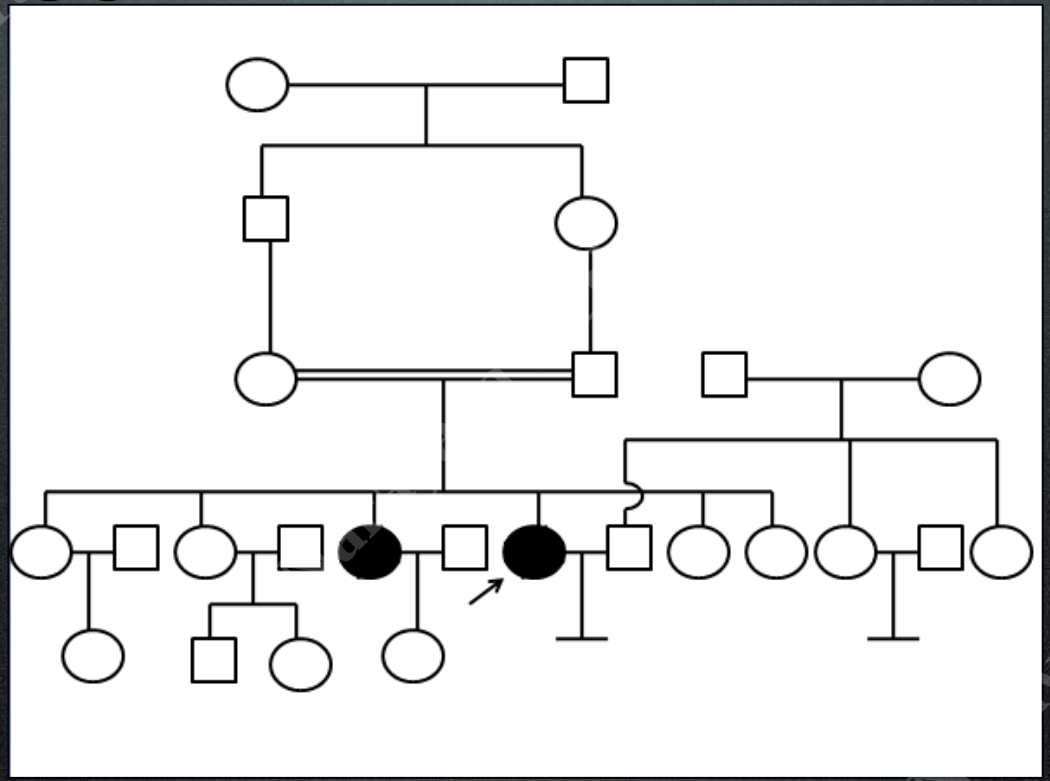
Case 3

10 years old female

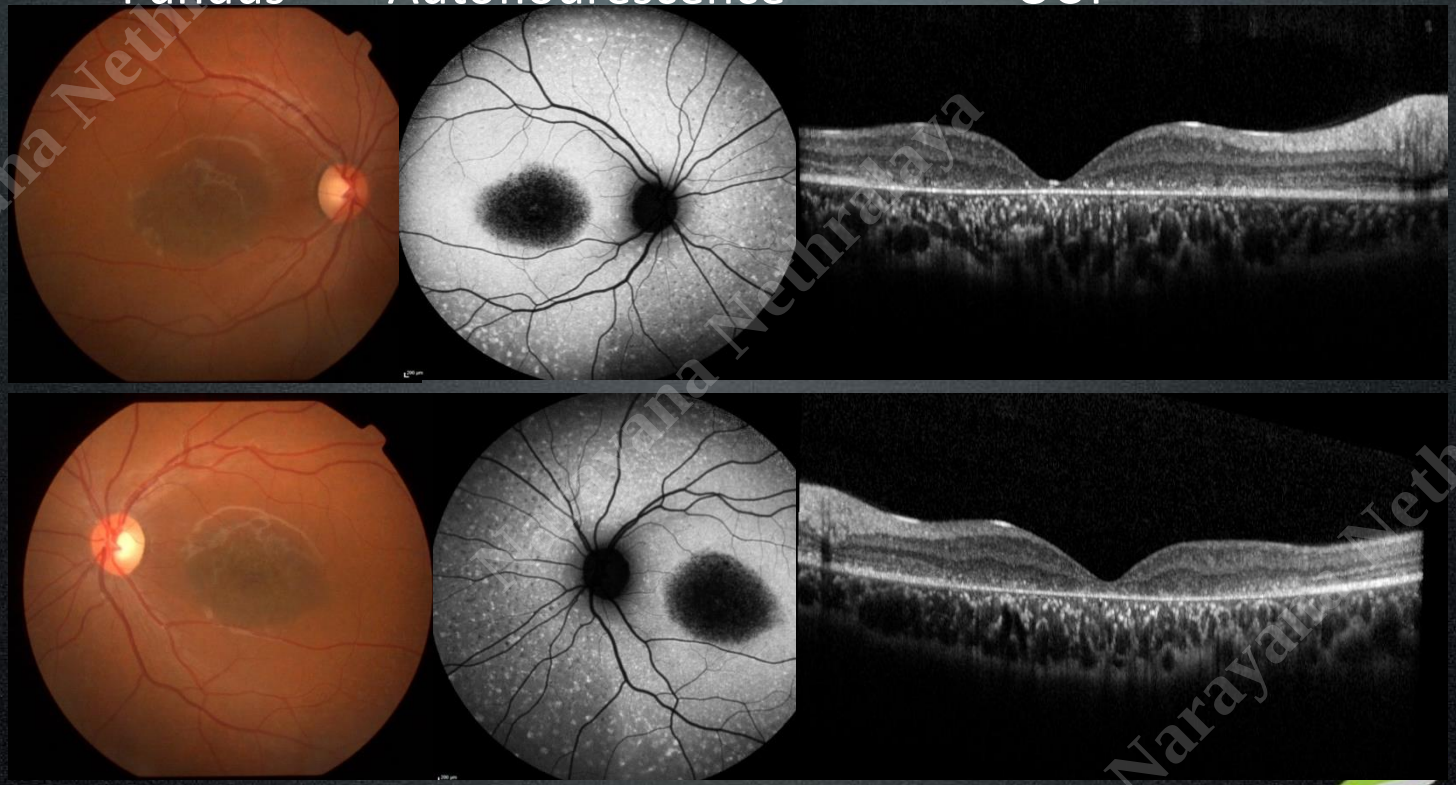
VA-BE 6/60

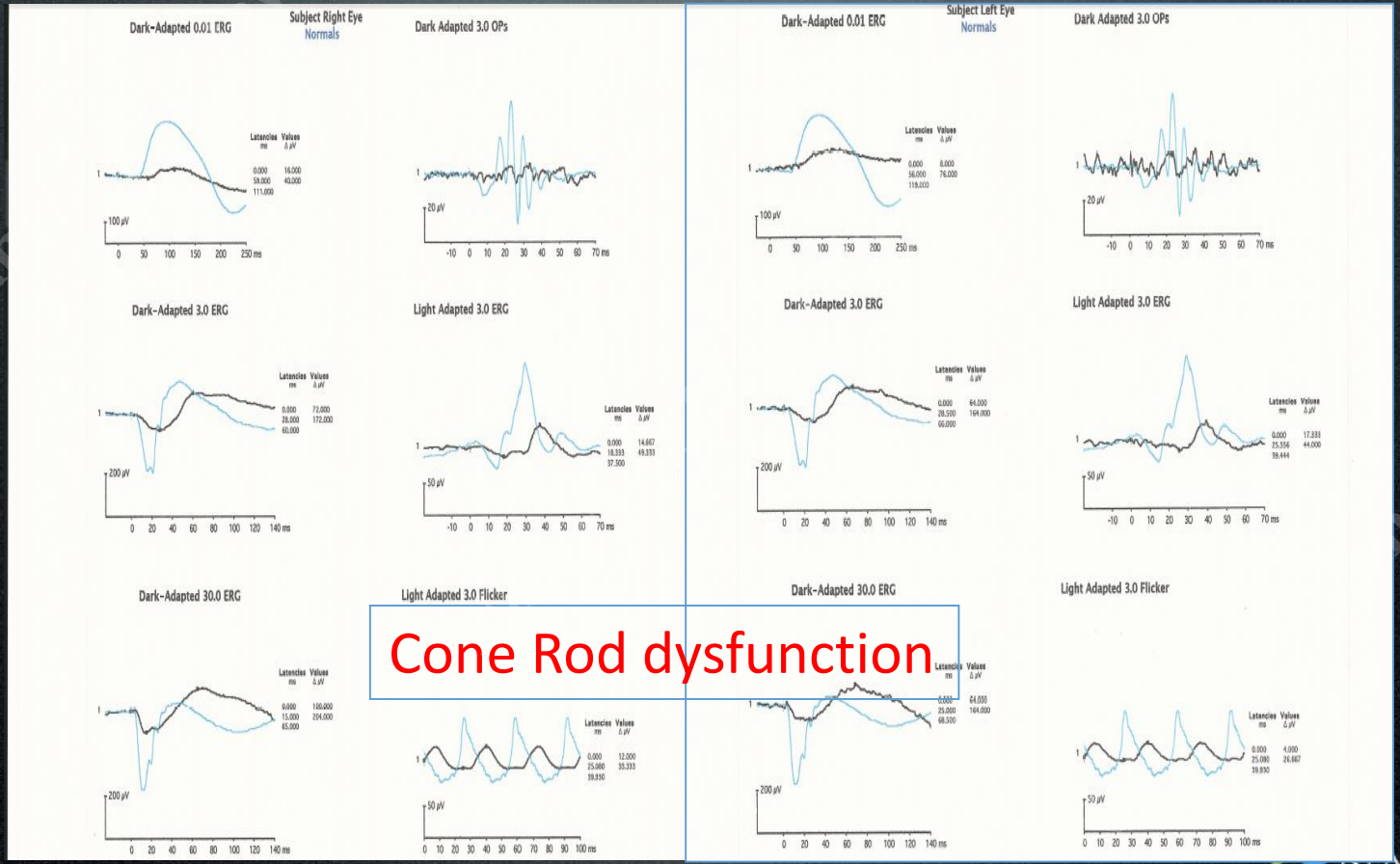


Pedigree



- Anterior Segment examination OU - WNL
- Fundus Autoflourescence OCT





Cone Rod dysfunction

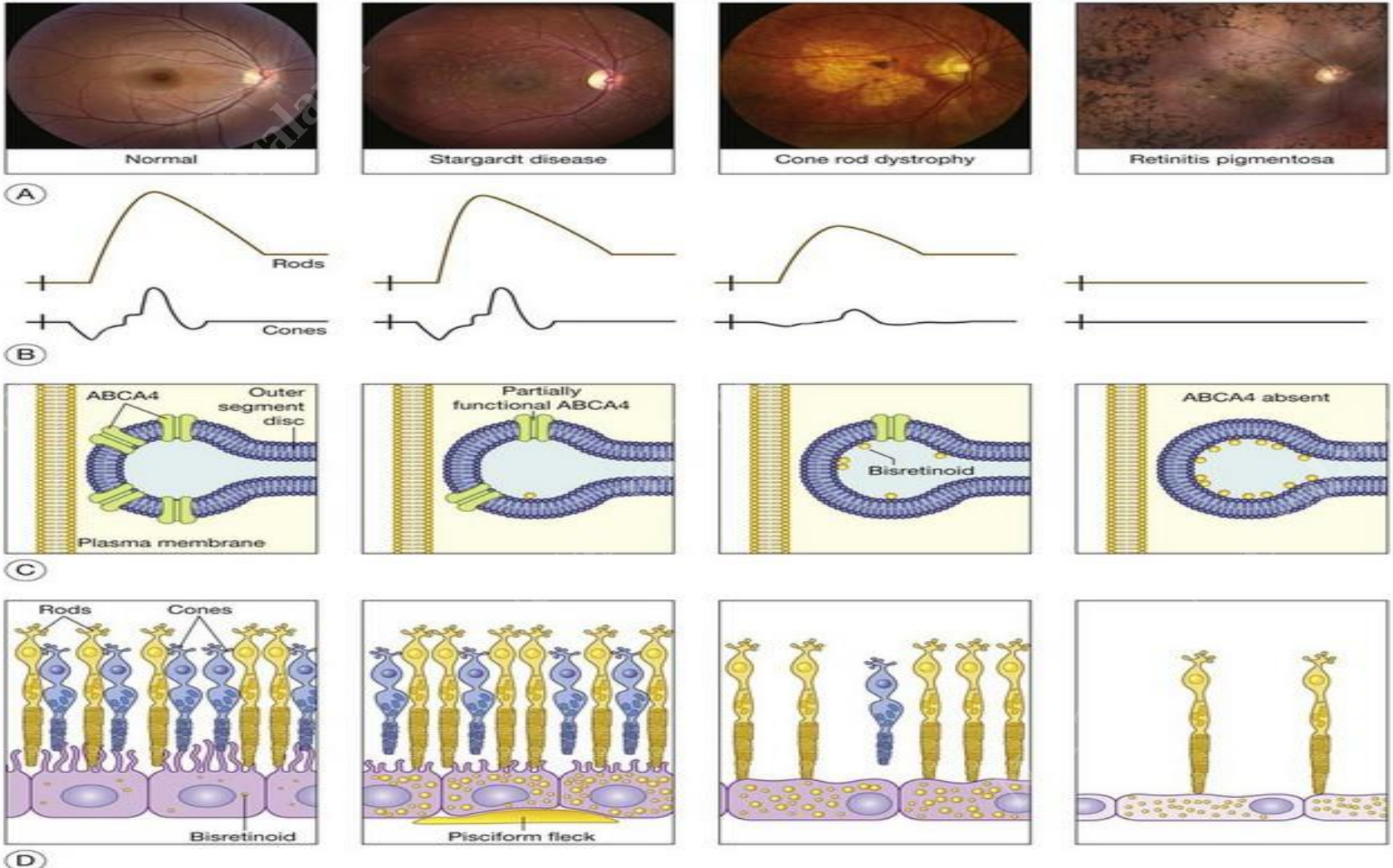
- Type 3 Stargardt’s

Type 1 –Normal full field ERG - Progression nil

Type 2 – Only Cones affected -20 % chance of progression

Type 3 – Cone and rodes affected -80 % chance of progreesion

- Characterised by accumulation of lipofuschin in the RPE cells specifically -A2E(bisretinoid)which is the major component of lipofuschin
- Also on the inner leaflet of the photoreceptor outer-segment disc membranes.


Normal
Stargardt disease
Cone rod dystrophy
Retinitis pigmentosa
A
Rods
Cones
B
ABCA4
Outer segment disc
Plasma membrane
Partially functional ABCA4
Bisretinoid
ABCA4 absent
C
Rods
Cones
Bisretinoid
Pisciform fleck
D

- Fovea- Normal or non specific mottling.
- Later stages may show oval area of foveal atrophy -

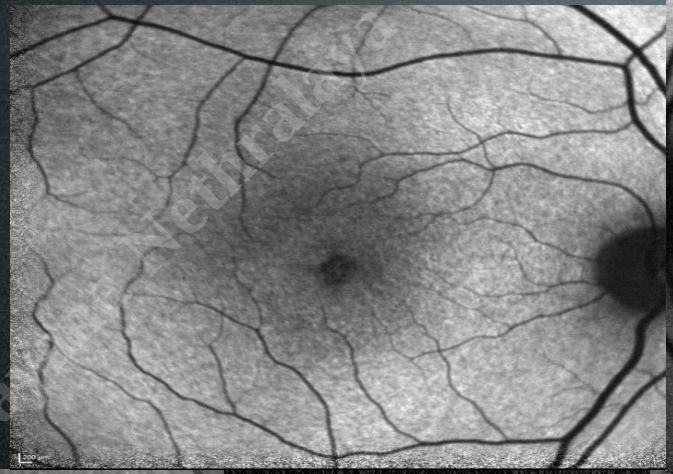
  ‘beaten bronze ’ appearance
- Varying degree of yellow

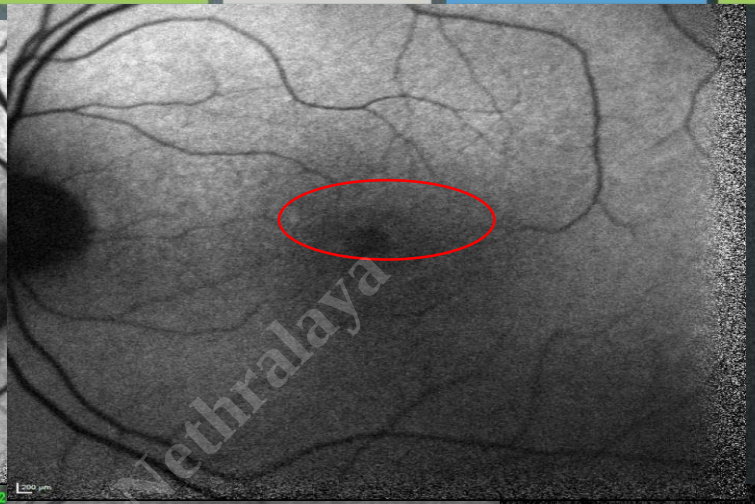
  white flecks
- Can have

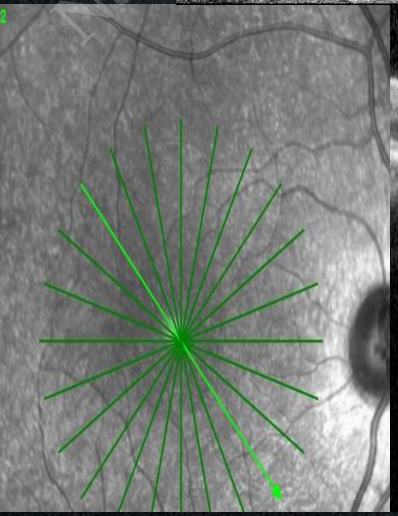
Bull’s eye configuration

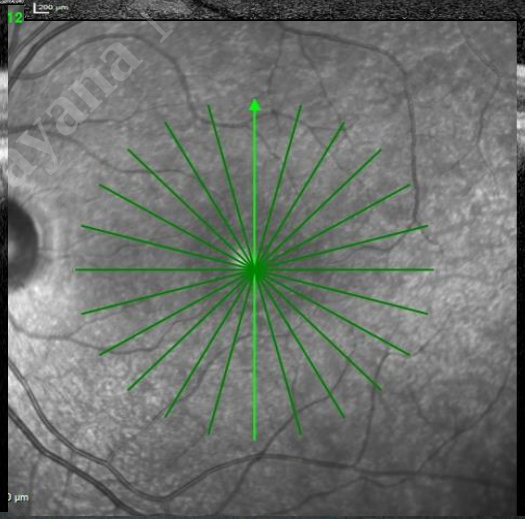
Case 4

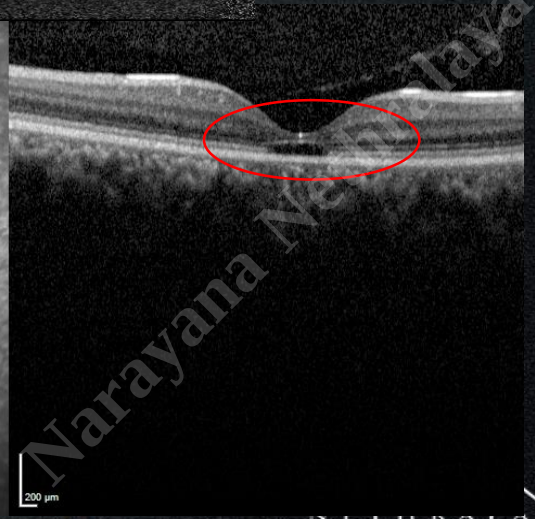
- 22 year /Male, c/o Diminution of vision in both eye since childhood
- BCVA OU: 6/36 ,N 18
- H/O photophobia
- Both eye Fundus was normal

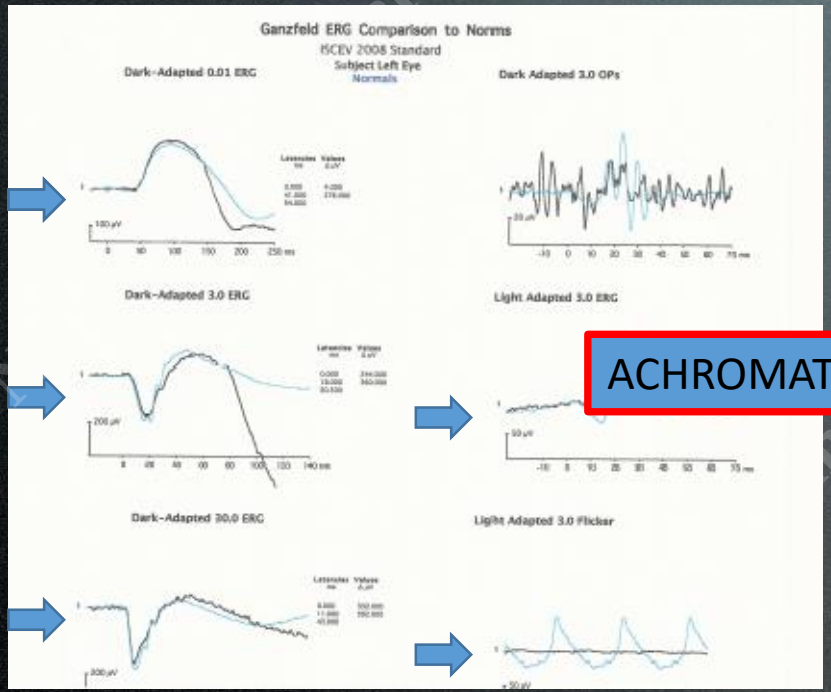




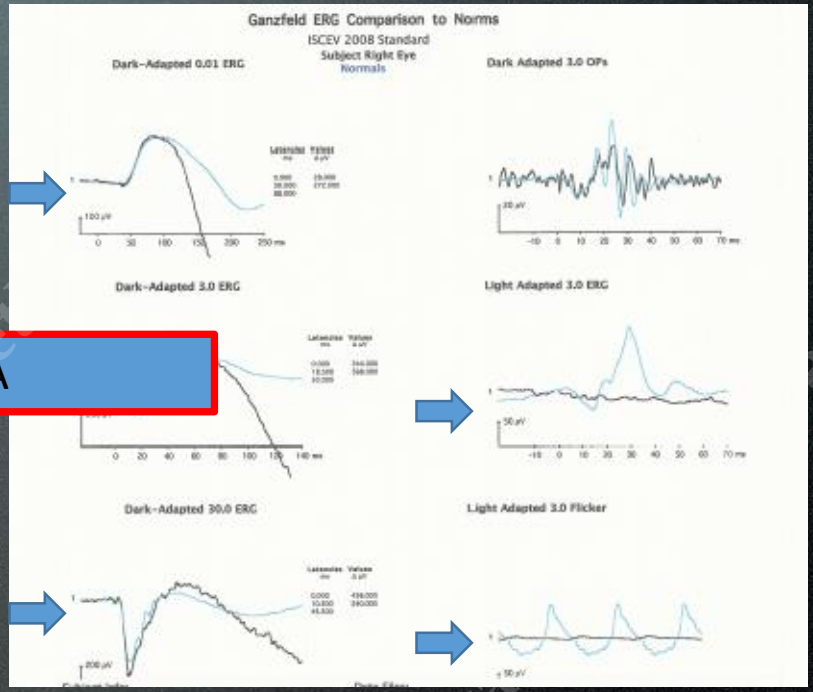








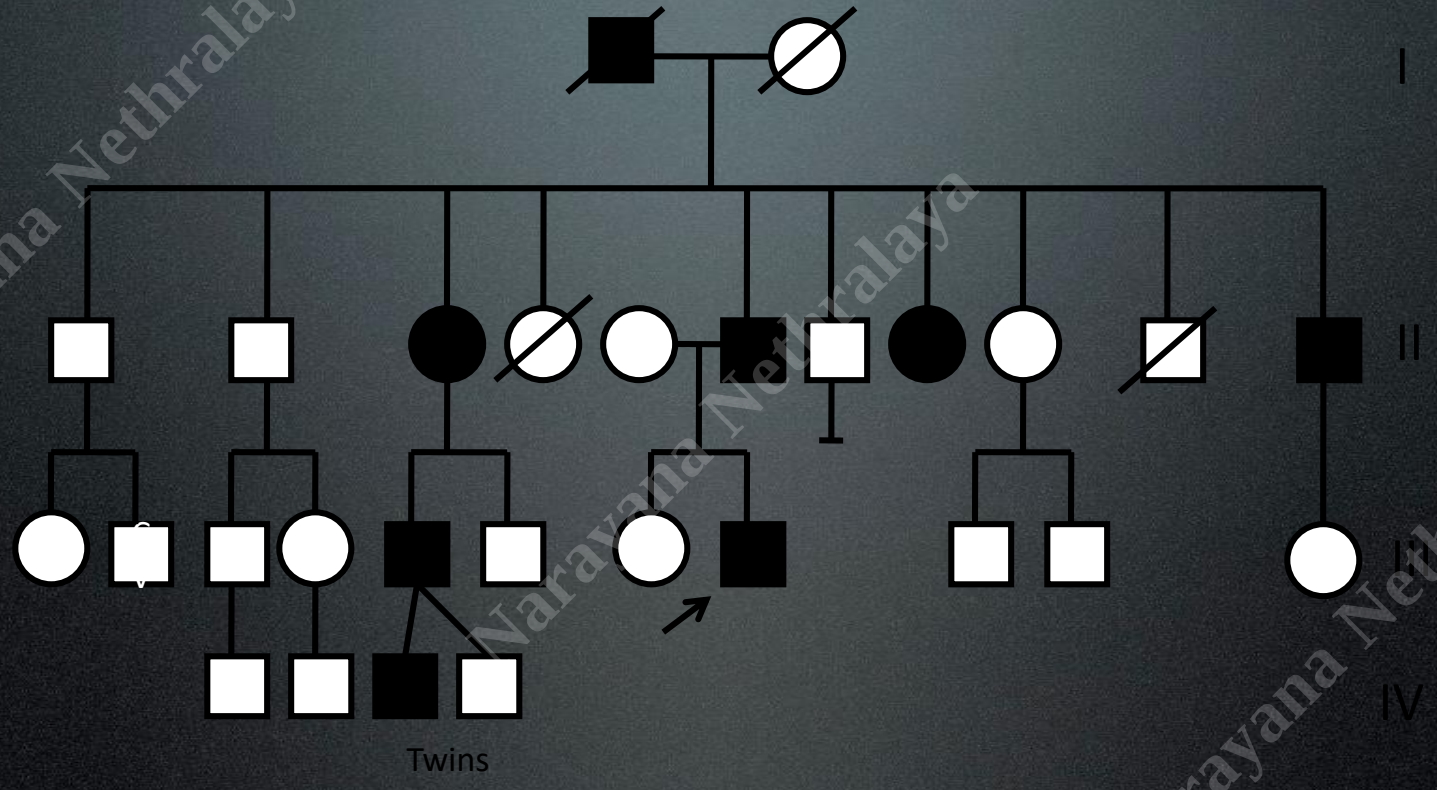
ACHROMATOPISA



CONE DYSTROPHY

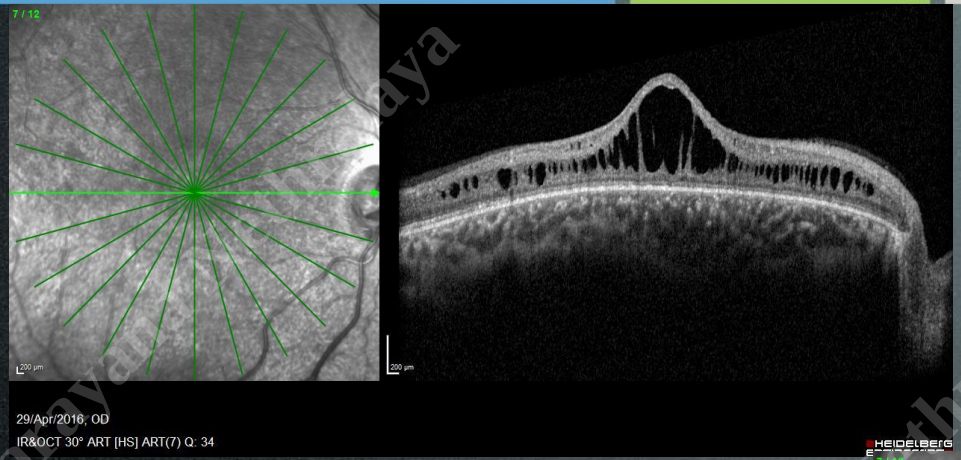
CLASSIFICATION

- Early onset, Stationary
  - Complete and incomplete achromatopsia
  - Blue cone monochromatism
- Later onset, Progressive
  - Cone dystrophy
  - cone-rod dystrophy

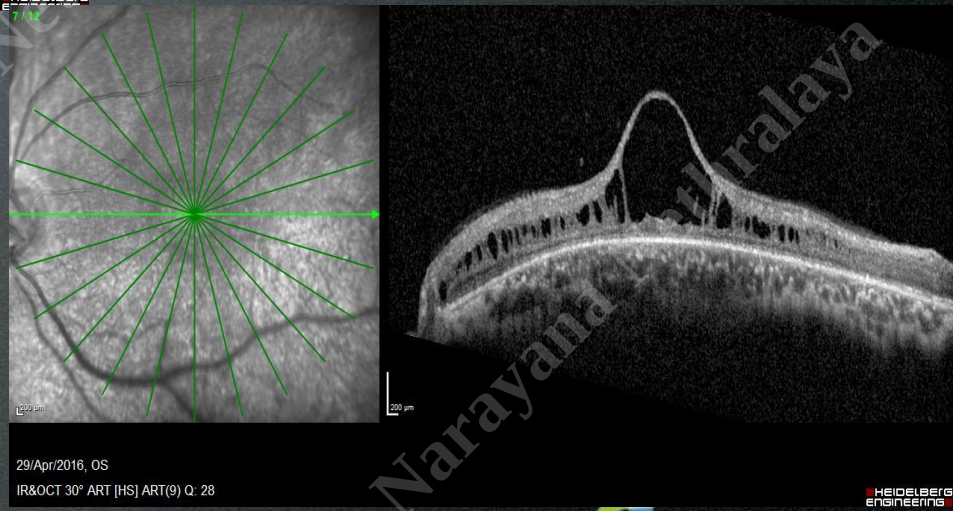


Case 5

- 8 year male
- Blurred vision
- OU vision – 6/36, normal anterior segment



X-LINKED JUVENILE RETINOSCHISIS



X-LINKED JUVENILE RETINOSCHISIS

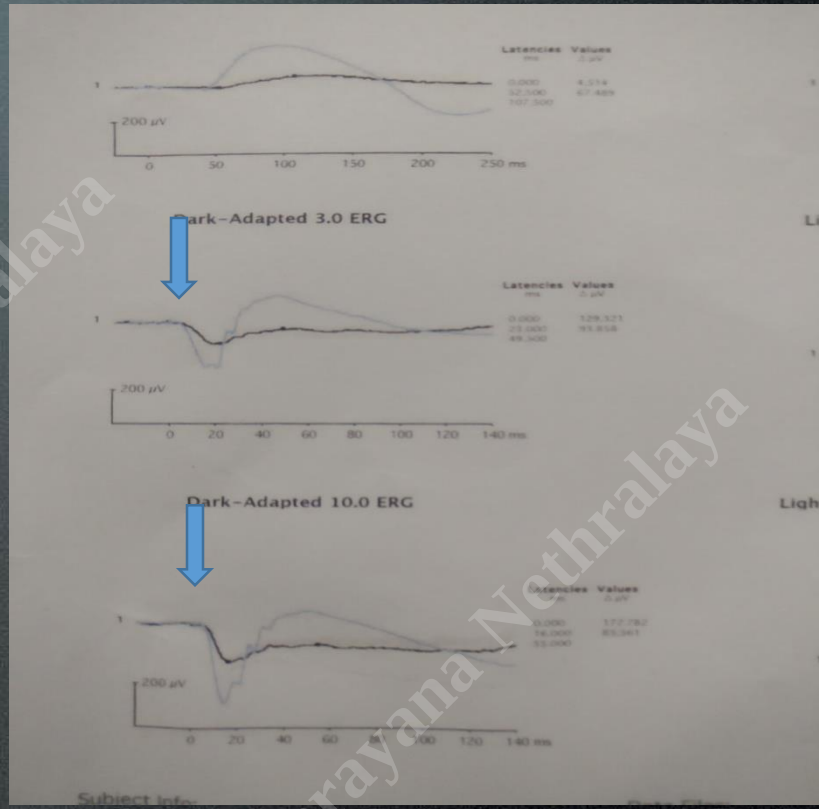
- Most common juenvile onset retinal degeneration in males-prevalance between 1 in 15000 and 1 in 30000

Cart wheel appearance

Vitreous veils

ERG

- Diagnostic with the typical wave form being “negative”
- The “b” wave amplitudes are reduced , “a” wave amplitudes are maintained at a near normal level
- b/a ratio is < 1.0



MANAGEMENT

- Genetic counselling
- Pharmacological –topical dorzolamide
- Laser-as adjuvant or preventive treatment
- Treatment – Surgery is recommended for vitreous hemorrhage and retinal detachments

Genead MA,Fishman GA,Walia S.efficacy of sustained topical dorzolamide therapy for cystic macular lesions in pateints with x-linked retinoschisis Arch Ophthalmol 2010;128:190-7

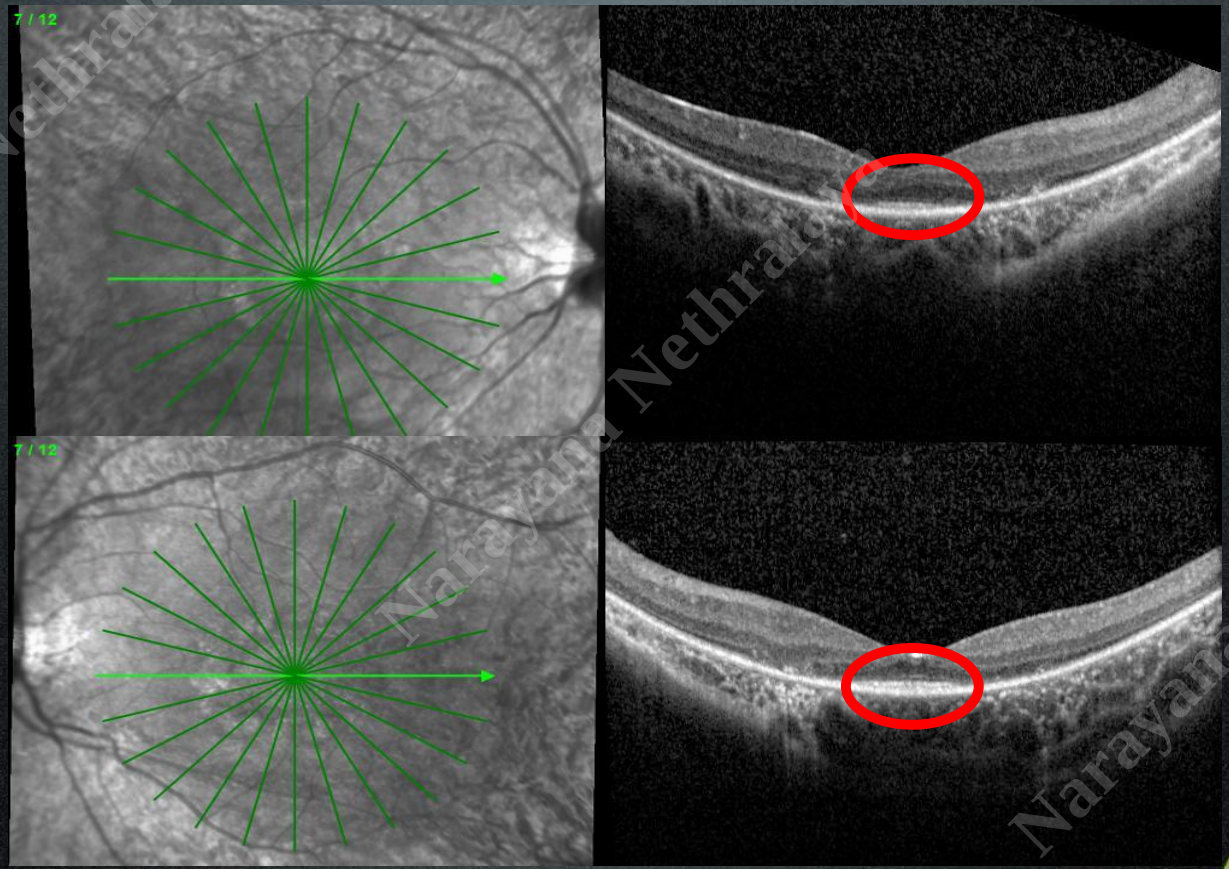
Case 6

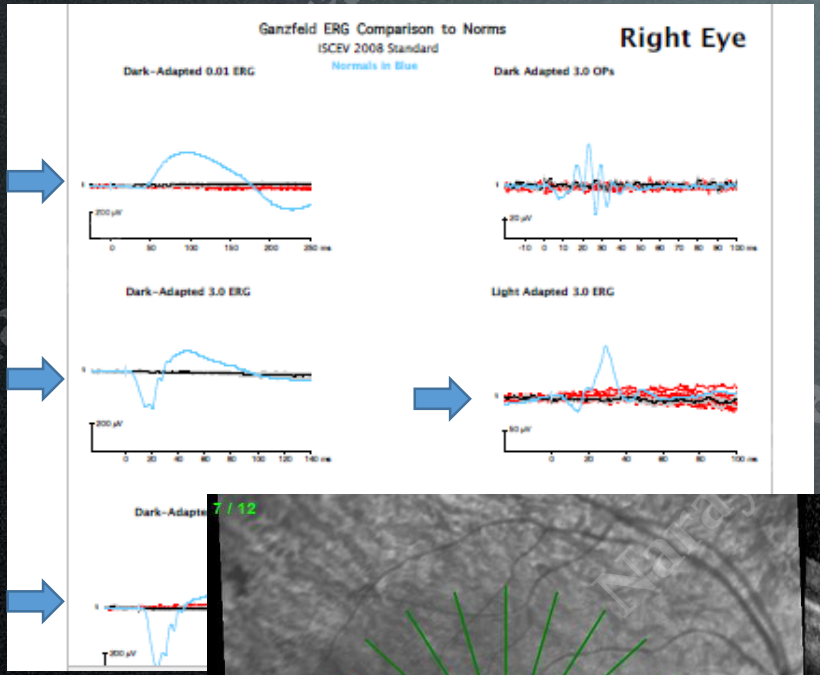
•**VISION:**

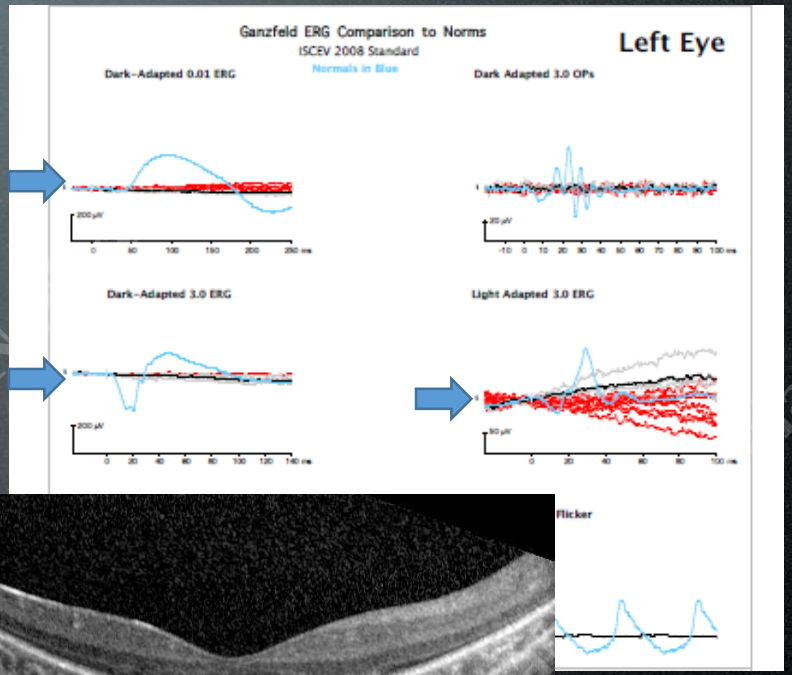
VISION/DISTANCE		VISION/NEAR	
RE	LE	RE	LE
6/9	6/9	N6	N6

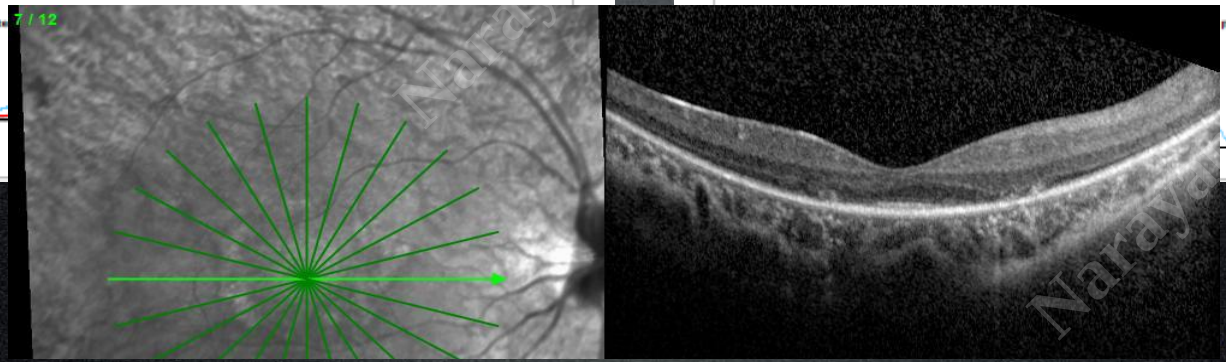
•**COMPLAINTS:**

• Night vision problem and history suggestive of tunnel vision

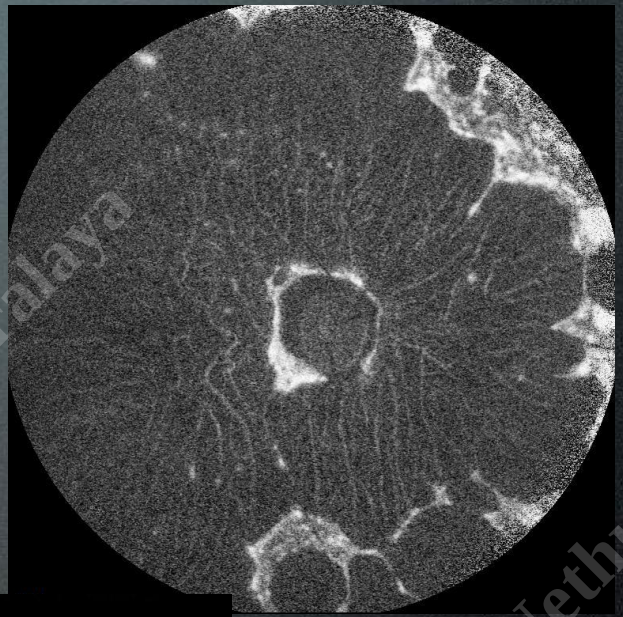


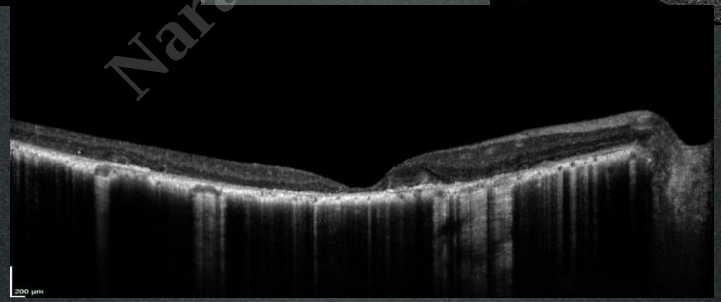




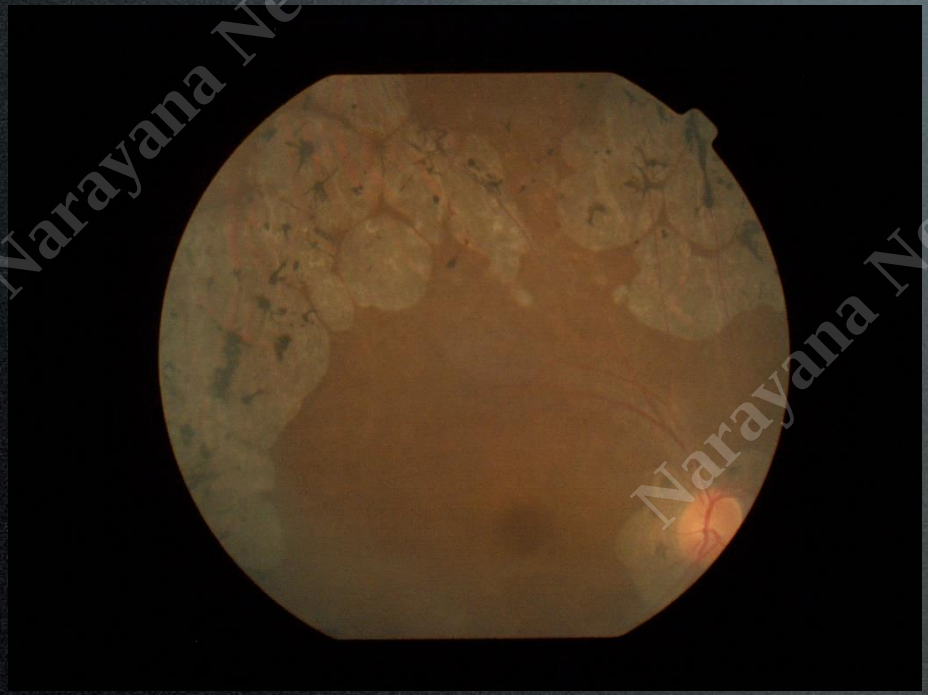


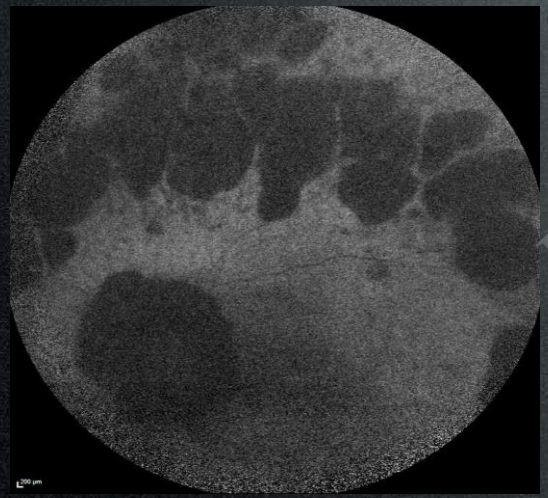
Choroideremia

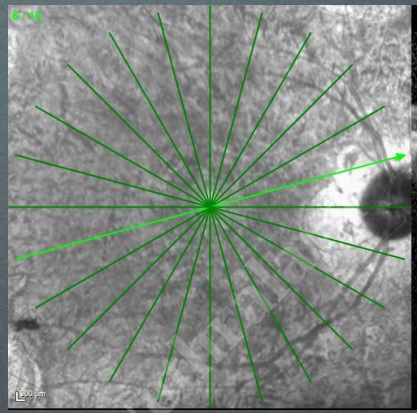


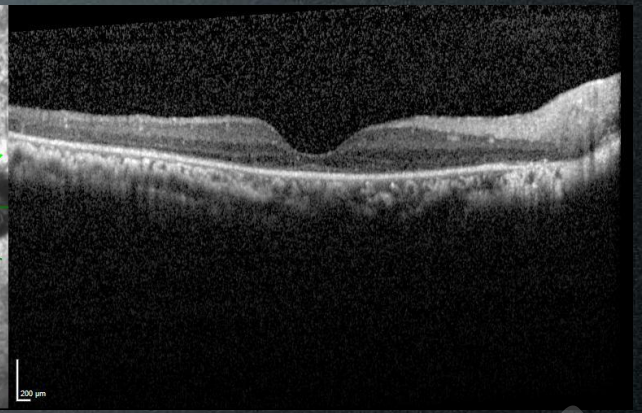


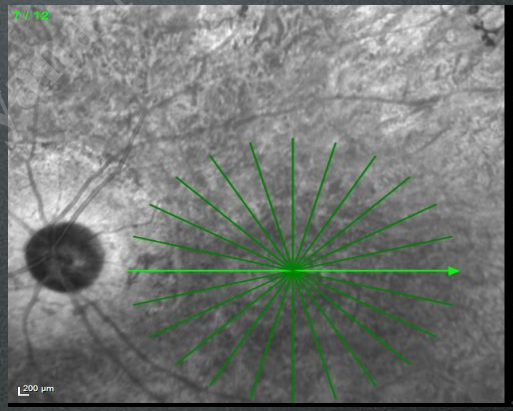
Gyrate atrophy

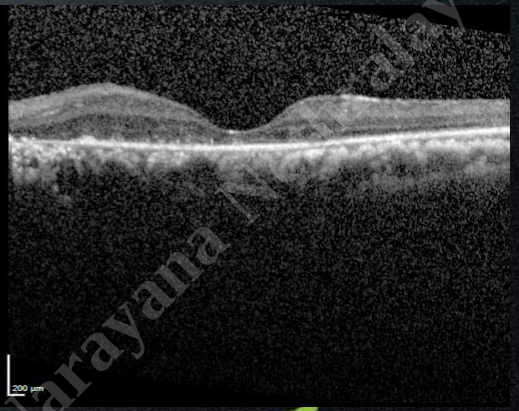






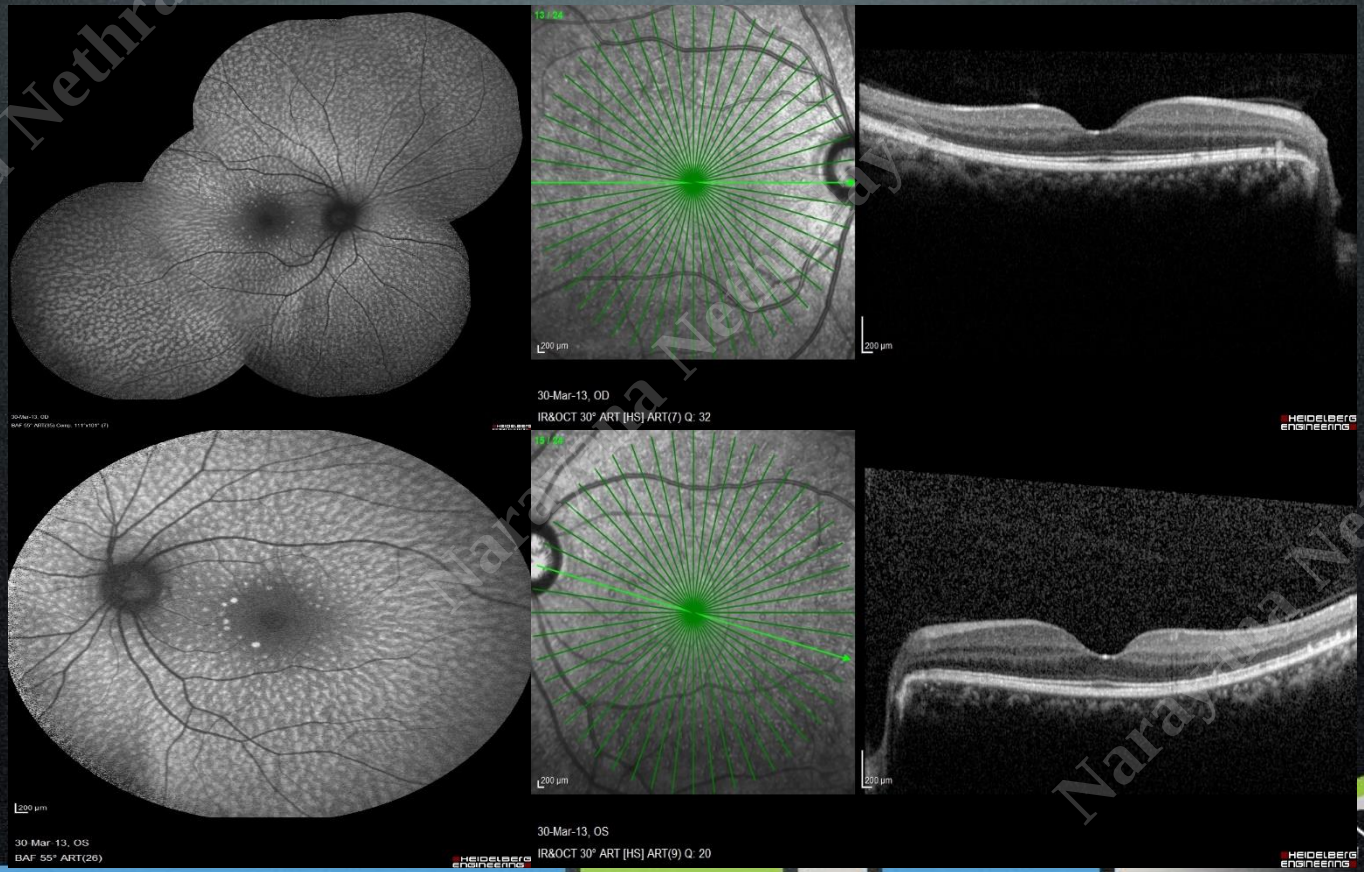






- Deficient OAT (Ornithine Amino Transferase)
- Increased Serum Ornithine level
- Dietary management
  - Arginine deficient diet
  - Vitamin B6 500 – 100 mg/day

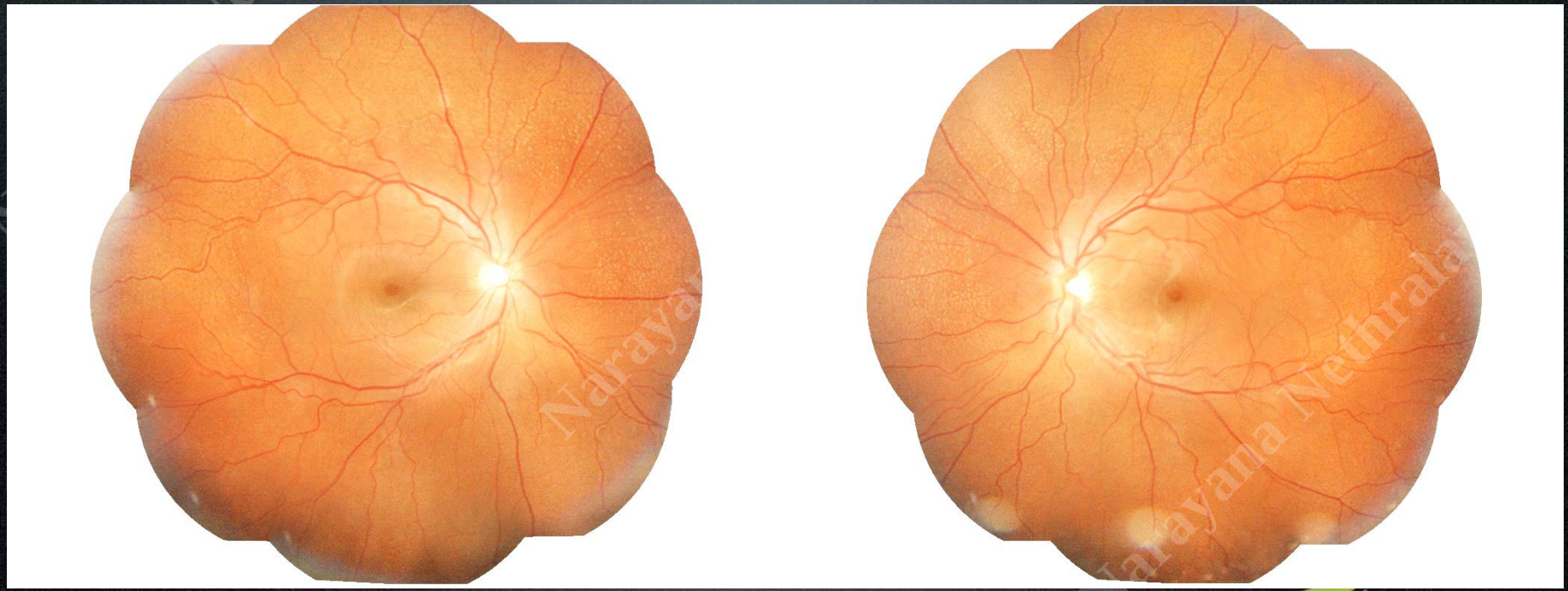
BENIGN FLECKED RETINA

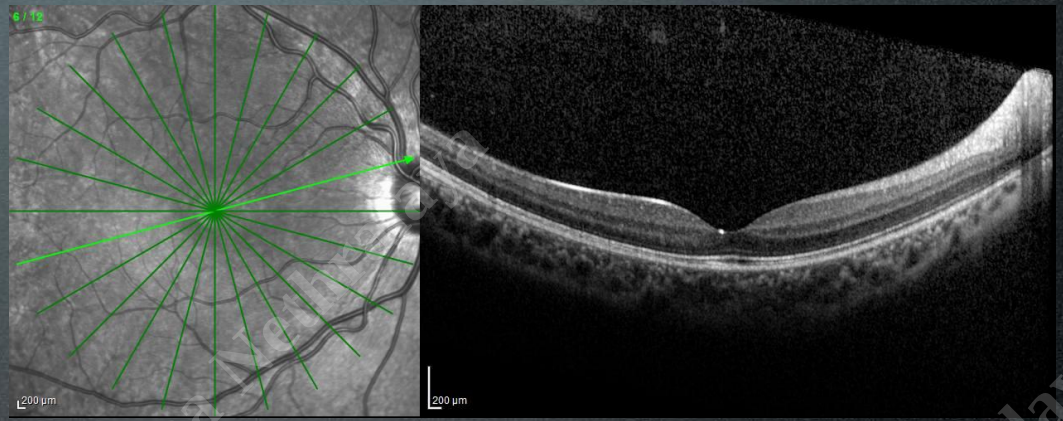


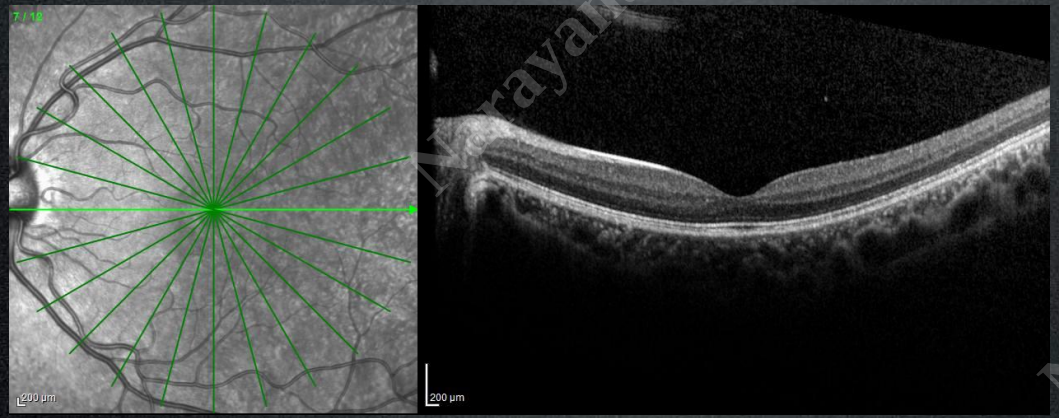
Case 7

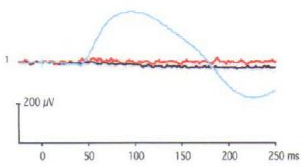
H/O night vision problem

BCVA – 6/6, N6

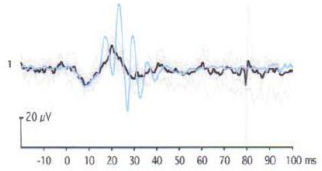




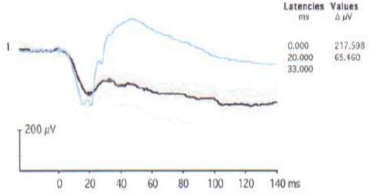




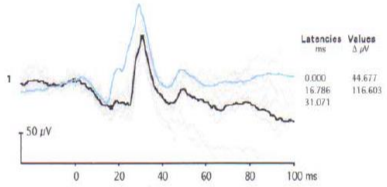
Dark-Adapted 3.0 ERG



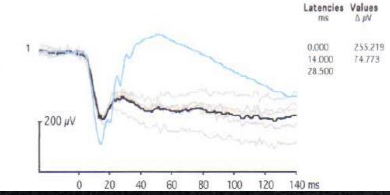
Light Adapted 3.0 ERG

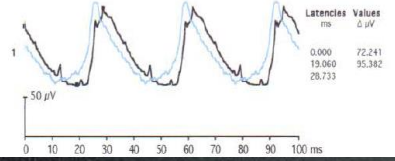


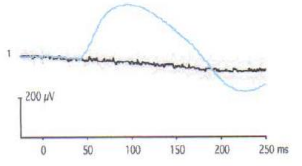
Dark-Adapted 10.0 ERG



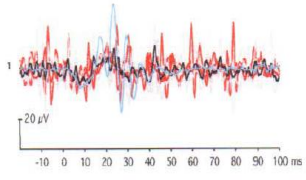
Light Adapted 3.0 Flicker



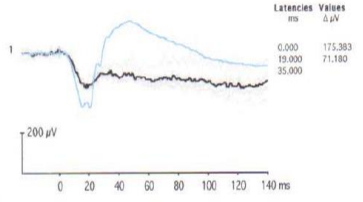




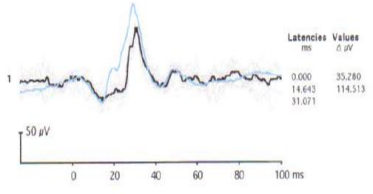
Dark-Adapted 3.0 ERG



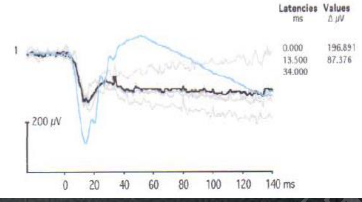
Light Adapted 3.0 ERG

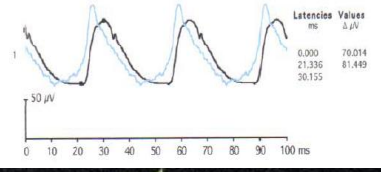


Dark-Adapted 10.0 ERG

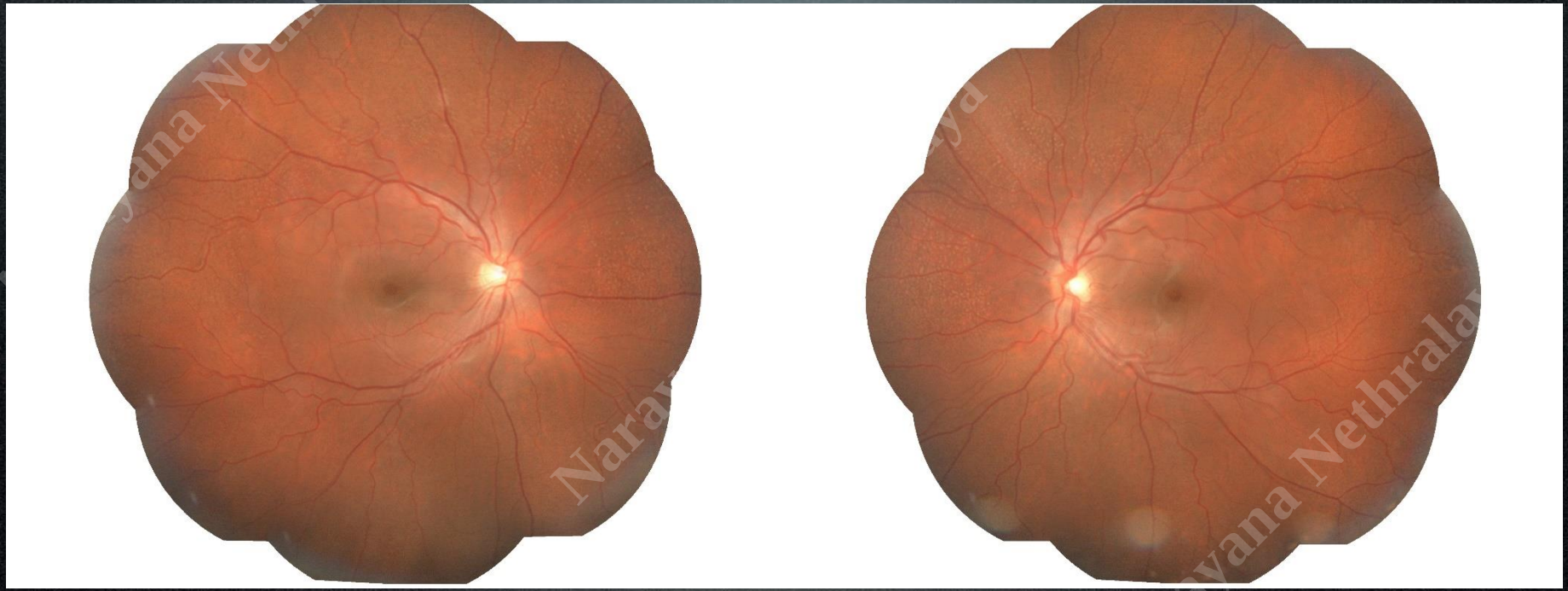


Light Adapted 3.0 Flicker





Fundus albipuctatus



Congenital stationary night blindness

Case – 8

26 year old male

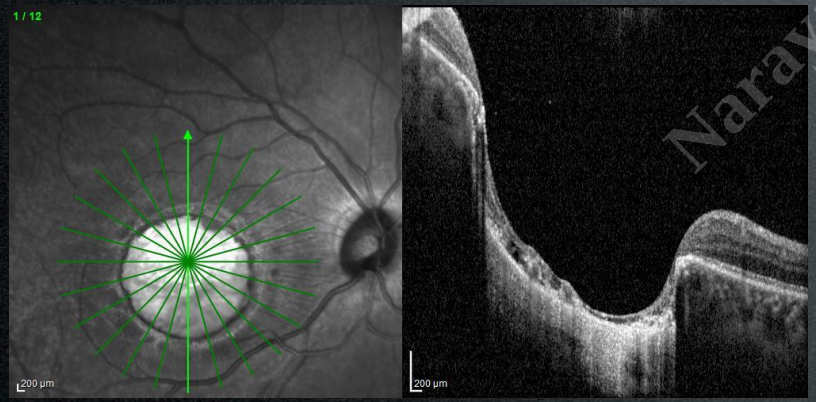
Low vision since chldhood

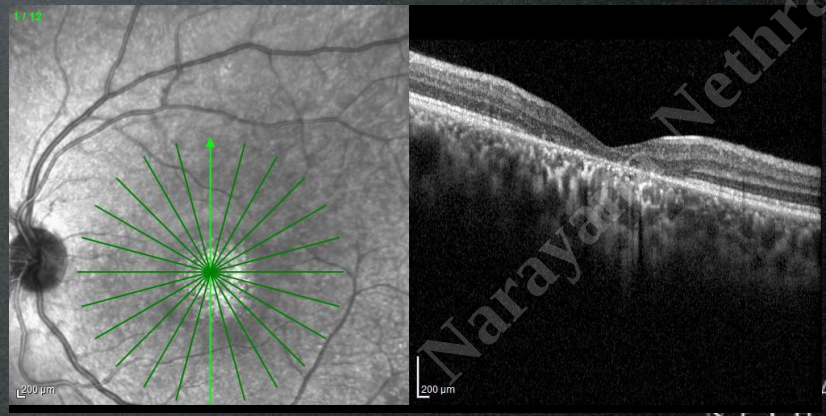
•**VISION**:

VISION/DISTANCE		VISION/NEAR	
RE	LE	RE	LE
6/24	6/24P	N6	N6

• **COMPLAINTS**:

• Blurring of vision for distance





ID: 1290693
Full Name: Sachin Sumanth
Date of Birth: 24/06/96

EDI

Vera™ Science 6.4.5

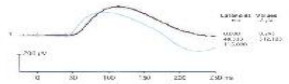
Ganzfeld ERG Comparison to Norms

ISCEV 2015 Standard

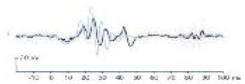
Normals in Blue

Right Eye

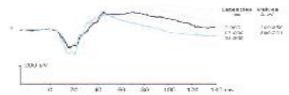
Dark Adapted 0.01 ERG



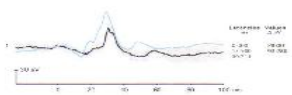
Dark Adapted 3.0 OPs



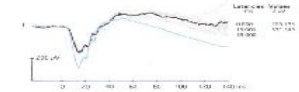
Dark-Adapted 3.0 ERG



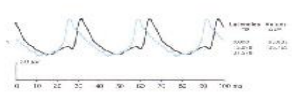
Light Adapted 3.0 ERG



Dark-Adapted 10.0 ERG



Light Adapted 3.0 Flicker



Subject Info:

ID:	1290693
First Name:	Sachin
Last Name:	Sumanth
Date of Birth:	24 June 1996
Sex:	Male
Subject Group:	Unassigned

Data Files:

FileName: fes08RBg2a 1290693 2017-09-09 12-50-35 Right
Comments: None.
Pupil Size:
Refraction:
Visual Acuity:

FileName: fepww08RBg2a 1290693 2017-09-09 12-59-44 Right
Comments: None.
Pupil Size:
Refraction:
Visual Acuity:

FileName: feo08RBg2a 1290693 2017-09-09 12-54-21 Right
Comments: None.
Pupil Size:
Refraction:
Visual Acuity:

FileName: DA10x00NN 1290693 2017-09-09 12-57-14 Right
Comments: None.
Pupil Size:
Refraction:
Visual Acuity:

ID: 1290693
Full Name: Sachin Sumanth
Date of Birth: 24/06/96

EDI

Vera™ Science 6.4.5

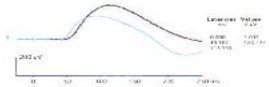
Ganzfeld ERG Comparison to Norms

ISCEV 2015 Standard

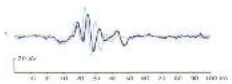
Normals in Blue

Left Eye

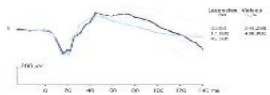
Dark-Adapted 0.01 ERG



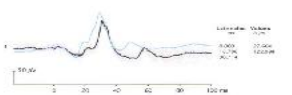
Dark Adapted 3.0 OPs



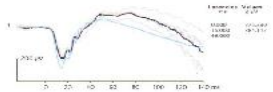
Dark-Adapted 3.0 ERG



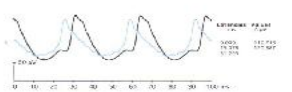
Light Adapted 3.0 ERG



Dark-Adapted 10.0 ERG



Light Adapted 3.0 Flicker



Subject Info:

ID:	1290693
First Name:	Sachin
Last Name:	Sumanth
Date of Birth:	24 June 1996
Sex:	Male
Subject Group:	Unassigned

Data Files:

FileName: DA10x00NN 1290693 2017-09-09 12-57-14 Left
Comments: None.
Pupil Size:
Refraction:
Visual Acuity:

FileName: fes08RBg2a 1290693 2017-09-09 12-50-35 Left
Comments: None.
Pupil Size:
Refraction:
Visual Acuity:

FileName: fepww08RBg2a 1290693 2017-09-09 12-59-44 Left
Comments: None.
Pupil Size:
Refraction:
Visual Acuity:

FileName: feo08RBg2a 1290693 2017-09-09 12-54-21 Left
Comments: None.
Pupil Size:
Refraction:
Visual Acuity:

NARAYANA NETHRALAYA
your faith shall heal you

THANK YOU

NARAYANA NETHRALAYA
your faith shall heal you