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A Critical understanding of Cone Rod Dystrophy (CRD) and its Ayurvedic interventions - A Case Report

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ABSTRACT

CRDs are Inherited Retinal Dystrophies that belong to the group of Pigmentary Retinopathies (Prevalence 1/40000). Is characterized by Primary Cone involvement or sometimes by concomitant loss of both Cones and Rods that explains the predominant symptoms of CRDs; Decreased Visual Acuity, Colour Vision Defects, Photophobia and Decreased Central Vision, later followed by progressive loss in Peripheral Vision and Night Blindness. Currently, there is no therapy that stops the progression of the disease or restores the vision and the visual prognosis is poor. The only Intervention of CRDs in contemporary science is using Low Vision Aids and Genetic Counselling. Hence Multidisciplinary Approach from Ayurvedic science plays a vital role in the restoration of vision. Though there is No direct reference available in our classical Ayurveda text, the different stages of CRD can be simulated with the conditions which are mentioned in *Dristigatagata Roga* like, *Pittavidagdha Dristi*, *Hrsvaja*, *Dhumadarshi* and *Vataja Timira*. Patient is thoroughly examined based on *Roga* and *Rogi Bala* and advocated multidisciplinary interventions. This helped this patient in improving the vision and as a prophylactic in arresting or delaying the progression of disease.

Key words: *Vataja Timira, Cone Rod Dystrophy, CRDs.*

INTRODUCTION

The cone rod dystrophies (CRDs) are a clinically and genetically heterogeneous group of progressive retinal disorders.[1] CRDs are characterized by retinal pigment deposits visible on fundus examination, predominantly localized to the macular region. CRDs reflect the opposite sequence of events to that of Rod cone dystrophies (RCDs).[2] The clinical course of CRDs is generally more severe and rapid than that of RCDs, leading to earlier legal blindness and disability. Prevalence of CRDs is estimated at 1/40,000.

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Functional signs and symptoms

- Decrease in the visual acuity is the earliest symptom
- Photophobia also occurs early
- Frequent dyschromatopsia
- Night blindness occurs later

Visual field

- Central scotoma appears first, preventing fluent reading
- Patchy losses of peripheral vision follow
- Severe loss of vision occurs earlier than in retinitis pigmentosa (RP)

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Fundus

- Normal looking macula or fine macular lesions and pallor of the optic disc may be the only signs at early stage.[3]
- Pigmentary deposits resembling bone spicules, frequently in macular area.
- Attenuation of the retinal vessels.
- Waxy pallor of the optic disc.
- Various degrees of retinal atrophy.

They occur in two forms Non Syndromic and Syndromic CRD. CRDs are most frequently non syndromic.

Non Syndromic CRD manifest in two stages; in the first stage the main symptoms is Reduced Visual Acuity with Deviated Gaze, Intense Photophobia, Variable degree of Dyschromatopsia and Night Blindness (not as prominent as Reduced Visual Acuity). Visual Field testing shows Central Scotoma sparing Periphery. Fundus shows Pigment deposits and various degree of Retinal Atrophy in the Macular Region. Optic disc shows Paleness with moderately Attenuated Retinal Vessels.

In the second stage Night Blindness becomes more apparent and the loss in the Peripheral Visual Field Progresses. In addition Visual Acuity continues to decrease, at this stage patients are Legally Blind (VA< 1/20).

Non syndromic CRDs are genetically heterogeneous (ten cloned genes and three loci have been identified so far). The four major causative genes involved in the pathogenesis of CRDs are ABCA4 (which causes Stargardt disease and also 30 to 60% of autosomal recessive CRDs), CRX *and* GUCY2D (which are responsible for many reported cases of autosomal dominant CRDs), and RPGR (which causes about 2/3 of X-linked RP and also an undetermined percentage of X-linked CRDs).[2]

Sushruta Samhita, one of the oldest textbook of Ayurveda has described 76 *Netra Rogas* and their detailed treatment comprising of both Medical and surgical methods. Among them *Dristigata Rogas*[4] like; *Pittavidagdha Dristi, Dhumadarsi, Vataja Timira, Hrsvajadya* clearly resembles CRD in their symptomology and different stages.

CASE REPORT

A 41 year old female approached with a C/O gradual diminished distant vision and distortion of vision since 15 years. Associated with blurred / smoky vision, diminished colour vision RE > LE since 10 years. Also C/O gradual diminished near vision and night vision and decreased dark adaptation since 3 years. Her medical history was significant for Diabetis Mellitus since 5years. There is no significant family history and consanguinity in the family. She approached allopathic hospital with same complaints and diagnosed as B/L Non syndromic CRD and was advised for fundus photography and OCT scan, which showed RPE mottling near macular region and disrupted RPE layer with loss of photoreceptor cells and foveal thinning (figure 1 and2) respectively with no medical intervention. Then she visited our Shalakya Eye OPD and was advised for admission and got admitted. During her presentation her blood glucose level was under control.

On examination the findings were as following

1. **Visual acuity:** Table 1
2. **Colour vision test:** Done with Ishiharas chart - Patient could only identify the color of book and few colors in the plates and she couldn't identify any number plates.
3. **Macular function test:** Done with Amslergrid chart - wavy Appearance of straight lines. BE with significant metamorphosia.
4. **Peripheral vision:** Though she is a diagnosed case of cone-rod dystrophy, her central vision remained unaffected.
5. **Ocular examination:** Table 2
6. **Slit - lamp Bi microscopic examination:** Table 3
7. **Fundus examination (by opthalmoscope):** Table 4

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Table 1: Visual acuity

VA	OD	OS	OU
DV	6/60	6/36	6/24P
NV	N_{10}	N_8	N_{6P}

Table 2: Ocular examination

Ocular structures		OD	OS
A	Eye lids Eye lashes	No abnormalities	No abnormalities
B.	Conjunctiva	No abnormalities	No abnormalities
C.	Cornea	Clear	Clear
	Anterior chamber	ND	ND
	Pupil	PERRLA	PERRLA
	Lens	Cataractous changes +	Cataractous changes +
D.	IOP	Digitally firm	Digitally firm

Table 3: Slit - lamp Bi microscopic examination

OD	OS
Early cataractous changes	Early cataractous changes

Table 4: Fundus examination (by opthalmoscope)

Examination	OD	OS
Fundal glow	Reduced	Reduced
Fundus	Salt pepper fundus	Salt pepper fundus
Media	Clear	Clear
Optic disc CDR	Pale 0.4	Pale 0.4
Macula	RPE mottling with few pigmentary deposits Dull FR	RPE mottling with few pigmentary deposits Dull FR
Retinal blood vessels	Attenuated	Attenuated
Peripheral Retina	Normal	Normal

Treatment intervention: Table 5

Patient was admitted in the hospital for a period of 45 days, the following treatment was employed;

Internally patient was advised with:

- *Patoladi Ghrita* 1tsp BD with milk
- Tab. *Saptamruta Loha* 1 BD

Table 5: Treatment intervention

Procedure	Drugs	Duration
Deepana pachana	*Chitrakadi Vati*	3 days
Snehapana	*Guggulu Tiktaka Gruta*	Till *Snehasidda Lakshana* (3days)
Virechana	*Trivrut Lehya*	1 day
Nasya karma	*Anutaila*	7days
Seka	*Guduchi* + *Patola* + *Punarnava*	7 days
Tarpana	*Patoladi Ghrita*	5days
Putapaka	*Ropana* drugs	2 days
Shiropichu	*Patoladi Ghrita*	7days
Tikta Ksheera Basti	*Guduchi* + *Patola*	8 days

Results after treatment:

Subject parameters

- Improvement in the blurriness of vision
- Increased sharpness of the vision

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- Improvement in dark adaptation

Objective parameters

- **Visual acuity:** Table 6
- **Fundus examination:** Stable Dystrophy

Table 6: Visual acuity

VA	OD	OS	OU
DV	6/36	6/24p	6/18P
NV	N_{10}	N_8	N_{6P}

OBSERVATIONS

Table 7: Showing effect of treatment on Visual acuity.

VA	BT		After Shodana		After tarpana putapaka		After Basti	
	OD	OS	OD	OS	OD	OS	OD	OS
DV	6/60	6/36	6/36	6/24	6/36	6/24	6/12	6/12
NV	N_{10}	N_8	N_{10}	N_8	N_{10}	N_8	N_{10}	N_8

VA - Visual acuity, BT - Before treatment, OD - Oculus dextrus, OS - Oculus sinister

Fig. 1: Oculus dextrus

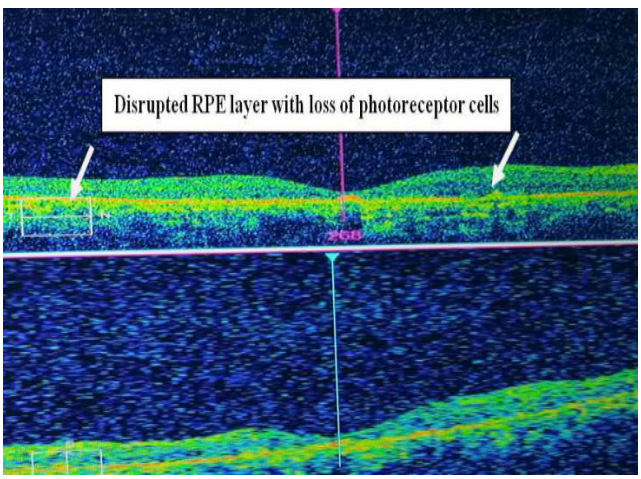
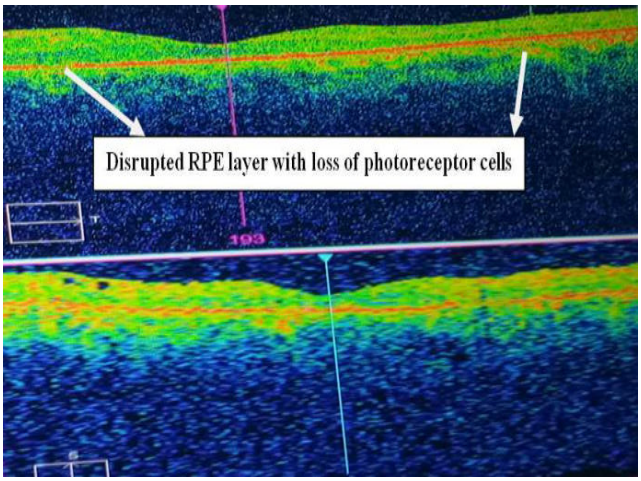


Fig. 2: Oculus sinister



Follow up : Patient showed marked improvement with her symptoms in subsequent follow-ups.

DISCUSSION

CRDs present first as a macular disease or as a diffuse retinopathy with predominance of the macular involvement. Non syndromic CRDs are genetically heterogeneous. Three Mendelian types of inheritance have been reported. Today, there are 13 genes responsible for non syndromic CRDs (10 cloned, 3 mapped) are detected.

Ayurvedic interpretation of CRDs by its signs and symptoms can be done with different conditions mentioned in *Dristipatalagata Rogas* like*; Pittavidagdha Dristi, Hrsvaja, Dhumadarshi* and *Vataja Timira.*

Its Manifestation simulates the predominance of *Pitta Dosha* followed by *Vata*.

In *Pittavidagdhadristi* due to *Pittakara Achakshushya Ahara Sevana*, vitiated *Dosha* gets lodged in *Dristi Mandala (Pitta* as *Asraya* in *Rakta,* simultaneously vitiates *Rakta* by exhibiting *Margavarodha* due to *Sanga* in *Raktavaha Srotas) (1st* and *2nd Patala*) and shows *Timira Lakshana* (blurred vision) in day time. When it reaches 3rd *Patala* person end up with complete day blindness,[5] which simulate early and late stages of CRD respectively.

Further *Madhukosa* adds '*Varne Patalantaragata Dosa Linga Bhava*' i.e. disturbed color vision

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(dyschromatopsia) due to *Doshas* seating in between the *Patalas*, as a speciality of *Pitta Vidagdha Dristi.*

Dhumadarshi manifested due to vitiated *Pitta*, exhibit in the form of reduced form sense and smoky vision in day time.[6]

Person with *Hriswajadya* experience difficulty in visualizing objects during day time due to vitiated *Pitta* and if *Dosa* gets lodged in the middle of *Dristimandala*, object will appear smaller than their normal size.[7] This signifies with Metamorphosia due to the involvement of macula in the early stages of CRD.

When vitiated *Pitta* (along with *Rakta*) vitiates *Vata* (by *Avarana*) and gets lodged in *Dristimandala*, objects appears to be hazy (*Vataja Timira*). *Acharya Vagbhata* adds *Vyaviddha Darsana* (wavy appearance of straight line) as one of the manifestation of this condition.[8] Due to *Avruta Rakta* blood circulation to retina gets hampered lead to paleness of disc, eventually attenuation of vessels.

Hence main pathology involved in the manifestation of these conditions is *Agnimandya, Ama, Margavarodha, Sanga* and *Avarana* .These should be kept in mind while dealing with CRDs.

Deepana, Pachana and *Anulomana* was done to relieve *Agnimandhya* both at *Koshta* (gastro intestinal) and *Dhatu* (tissue) level and to bring *Vata Doshanulomana* (homeostasis of *Vata*). Having *Samagni* is the base for being healthy and to have proper metabolism and absorption of the drug.

Shareera Shodhana was done in the form of *Virechana*, as it is *Sroto Shuddhikara* and *Indriya Samprasadakara.*

Shiro Virechana was done with *Anutaila* owing to its properties of *Indriyasroto Pravesa* i.e. permeating into minute channels and *Margavishodhana* (removes obstruction). *Seka* and *Vidalaka* was done with *Chakshushya Dravyas* as a *Netra Amahara Kriyas*.

Tarpana was done with *Patoladi Grutha*[9] followed by *Ropana Putapaka* owing to their *Tikta Rasa* and *Chakshushya* properties helps to control vitiated *Pitta Dosa.*

Tikta Ksheera Basti was done both as *Shamana* and *Shodhana* line of treatment to alleviate *Raktavruta Vata Samprapti* and to access retinal circulation.[10]

The ingredients were having Immunomodulatory, Adaptogenic, Antioxidant and Neuroprotective activities. Drugs having Lutein and zeaxanthin were selected for treatment. Ghee which was given internally having *Chakshushya*, *Rasayana* properties along with omega - 3 fatty acids, essential vitamins and minerals, was also added to the combination for its targeted and synergistic action on ocular tissue.

CONCLUSION

Restoration of vision and arresting the progression of the disease was the objective of treatment in this case. The treatment modalities employed were efficacious in slowing down the degenerative process and helping patients to cope with the social and psychological impact of blindness. As a prophylactic treatment a proper screening of patients by *Chakshu Visharada*'s at regular interval with proper intervention of *Kriyakalpa*, genetic counselling, lifestyle modifications, *Pathyapathya* along with oral medicines at appropriate time will definitely retard the progression of the disease and maintains the retinal function.

REFERENCES

1. Michaelides M, Bloch-Zupan A, Holder GE, *et.al.* An autosomal recessive cone rod dystrophy associated with amelogenesis imperfect *Journal of Medical Genetics* 2004; 41:468-473.
2. Christian P Hamel. Article Cone rod dystrophies Orphanet Journal of Rare Diseases, 2007, Volume 2, Number 1, Page 1.
3. Brad Bowling. Kanski's Clinical Ophthalmology a systemic approach, hereditary fundus dystrophies, edition 8; 15/652-653.
4. Susruta, Nagarjuna, Dalhana commentator, Sushruta Samhita, Uttaratantra , Drstigata roga Adhyaya 7, Reprint, Chaukhamba Sanskrit Sansthan, Varanasi,2014,71.

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5. Susruta, Nagarjuna, Dalhana commentator, Sushruta Samhita, Uttaratantra , Drstigata roga Adhyaya 7/34-35 sloka, Reprint, Chaukhamba Sanskrit Sansthan, Varanasi,2014,71.
6. Susruta, Nagarjuna, Dalhana commentator, Sushruta Samhita, Uttaratantra , Drstigata roga Adhyaya 7/39 sloka , Reprint, Chaukhamba Sanskrit Sansthan, Varanasi,2014,71.
7. Susruta, Nagarjuna, Dalhana commentator, Sushruta Samhita, Uttaratantra , Drstigata roga Adhyaya 7/40 sloka, Reprint, Chaukhamba Sanskrit Sansthan, Varanasi,2014,71.
8. Susruta, Nagarjuna, Dalhana commentator, Sushruta Samhita, Uttaratantra , Drstigata roga Adhyaya 7/18-19, Reprint, Chaukhamba Sanskrit Sansthan, Varanasi,2014,71.
9. Vagbhata, Arunadatta, commentator, Ashtanga Hridaya, uttara sthana, Timira Pratisheda Adhyaya 13/6-9, Reprint, Chaukhamba Sanskrit Sansthan, Varanasi, 2011,818.
10. Agnivesha, Charaka, Drudabala, Chakrapani datta commentator, charaka samhita, chikitsa sthana, vatarakta chikitsa adhyaya 29/88, Reprint, Chaukhamba Sanskrit Sansthan, Varanasi, 2011,629.

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