

Patient with Stargardt's Disease are Well Respond with Minimum Magnification - A Case Report

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Abstract: Stargardt's disease is a rare, inherited disorder of the retina which gradually loss of vision during childhood or adolescence. A case of 23 years old tribal female patient was referred from a medical college with the provisional diagnosis of Stargardt's disease to the low vision clinic. Since 4 years she was losing her vision slowly. Before that, she had no previous history of wearing spectacles. Even, she had no previous history of any systemic diseases related to the eye. On examination her distance visual acuity was 6/24 in RE and 6/24(p) in LE; near visual acuity was N12 in BE. No visual improvement with regular spectacle correction. Patient was prescribed 2.0X Dome magnifier for near work and distance visual acuity was significantly improved up to 6/9 by 1.0X spectacle mounted telescope. The yellow filter was advised for reduction of photophobia and fluorescent table lamp for brighter illumination of reading object.

Keywords: Stargardt's disease; dome magnifier; spectacle mounted telescope; yellow filter; fluorescent table lamp

1. Introduction

Stargardt's disease is a very rare and inherited disorder of the retina, causes vision loss during childhood or adolescence^[1]. The disease causes progressive damage or degeneration of the macula^{[1][2]}. There are no common symptoms of Stargardt's disease, but often gradual loss of central vision in both eyes, sometime gray, black, or hazy spots in the centre of their vision, or eyes may be more sensitive to bright light.^[1] There is no as such medical treatment for Stargardt's disease.^[1] People with Stargardt's disease are advice to wear dark glasses to protect bright light. Smoking restricts. Low-vision aids can be helpful for daily tasks from simple magnifier to telescope.^[1]

2. Case Report

A 23 years old tribal female presented to the low vision clinic referred from Tripura Medical College with the provisional diagnosis of Stargardt's disease. Her visual acuity was 6/24 in RE and 6/24(p) in her LE. The anterior segments were normal but on fundus examination typical features of Stargardt's disease found by medical college (figure 1).



Figure 1: Fundus of patient

After attending in low vision clinic, detail history was taken. She had no previous history of wearing glasses. Since 4 years she was gradually loose her vision. Even no previous history of any systemic diseases related to eye. She had problem with near vision related difficulties with newspaper reading, college books, difficulties in bright day light. She

had no difficulties in writing but comfortable with fluorescent light. She could not recognize faces, watching television, bus numbers in certain distance and had mobility problems in unfamiliar places. She could not tolerate bright light. But, she was no problem in her daily living at home including food identification, money identification (figure 2).

Diagnosis: ? Stargardt's disease											
Chief visual complaints: D/V(N) & photophobia.											
D/V Abnormalities	Yes	No	Board works in institute	Yes	No	Recognizing Faces	Yes	No	Watching TV	Yes	No
	✓						✓				✓
N/V Abnormalities	Yes	No	Writing Ability	Yes	No	Extra Illumination	Yes	No	Glare Affect	Yes	No
	✓			✓		✓			✓		✓
Colour Sensation	Yes	No	Food Identification	Yes	No	Mobility Problem	Yes	No	Currency Identification	Yes	No
	✓		✓			✓			✓		✓
Visual Acuity Assessment :											
Distance Visual Acuity											
By Snellen's Chart [Unaided]			Right Eye			Left Eye					
			6/24			6/24(P)					
By Snellen's Chart [Aided]											
By Log MAR Chart [Unaided]											
By Log MAR Chart [Aided]											
Near Visual Acuity											
Single Letter Chart [Unaided]			Right Eye			Left Eye					
			N/12			N/12					
Single Letter Chart [Aided]											
Crowded Letter Chart [Unaided]											
Crowded Letter Chart [Aided]											
Other Essentials Test:											
Contrast Sensitivity Test			Right Eye			Left Eye					
			V. Poor			V. Poor.					
Visual Field Test [Peripheral]			N			N					
Visual Field Test [Central]			N			N					
Colour Vision Test			N			N					
Glare Test			-			-					

Figure 3: Low Vision Evaluation Sheet

After taking complete history from the patient in low vision clinic, her distance visual acuity was 6/24 in right eye and 6/24(p) in left eye and near visual acuity was N12 from her comfortable reading distance in both eyes. Her refractive error correction done by objective method followed by subjective method was +0.75/-0.50X 90° in RE and +1.00/-0.75 X 90° in LE which was little bit clear but no useful improvement in visual acuity in her both eyes (figure 3A & 3B).

<RIGHT>				VD: 0.0
SPH	CYL	AX		
-0.75	-0.25	106		
-1.00	0.00	180		
-1.00	0.00	180		
-1.00	0.00	180		
-1.00	0.00	180		
-1.00	0.00	180		
[
-1.00	0.00	180		
<LEFT>				
-0.75	-0.50	98		
-0.75	-0.50	92		
-0.75	-0.50	98		
-1.00	-0.50	76#		
-1.25	0.00	180		
[
-0.75	-0.25	98		

Figure 3 (A): Auto refractometer reading

Name: _____ Age: 23yr Sex: F

Mobile No. _____

C/O: ✓ Ref by Sriprava Medical College for Low vision evaluation
 D/V(N) ✓ cataract. + management.
 H/O: ✓ Phos. ✓ 2ant correction
 V/A ✓ 2ant correction
 O/E: ✓ 2ant correction

? Auto ref: -1.00
 -0.25 -0.50 106
 ✓ Advise for low vision evaluation

EYE	D. Sph	D. Cyl	AXIS	VISION
R.E.	+0.75	-0.50	90°	6/24
L.E.	+1.00	-0.75	90°	6/24
B.E.				
Near Add				+1.00
Lenses				

Advice: ✓ Reviewed after 45 days for LVA trial

Signature: _____
 09/12/2019

Figure 3 (B): First day prescription

Her extra-ocular motility and colour vision test was normal in range, even visual field tested by confrontation test was within normal limits but her contrast sensitivity were very poor in both eyes. After completion her evaluation and to overcome her boringness she was advised to attend after three days in low vision clinic.

After three days letter she came to the low vision clinic again. She was trailed by different low vision devices with different magnifications. But after using 2.0X dome magnifier she was able to read comfortably. And distance visual acuity was improved significantly up to 6/9 with 1.0X spectacle mounted telescope. She was also provided rehabilitation training to familiar with the prescribed devices (figure 4).

Current device used	Purpose	RE Vision	LE Vision
Distance	NO	---	---
Near	NO	---	---
Distance Low Vision Devices			
Low Vision Devices	Type (if any)	RE Vision	LE Vision
Multiple pin hole Spectacles	---	---	---
✓ Telescopes	Spect. Mounted (1.0X)/9	(10)/9	(10)/9
✓ Filters or Tints glass	yellow	---	---
Near Low Vision Devices			
Low Vision Devices	Power	RE Vision	LE Vision
Near Telescope	---	---	---
Hand magnifier	---	---	---
Stand magnifier	---	---	---
Spectacle magnifier	---	---	---
Self-illuminated magnifier	---	---	---
✓ If any other devices	Dome Magnifier (2.0X)	(6)	(6)
Non-Optical Devices			
Name of Non-Optical Devices (uses if any)	Yes	No	
✓ Reading Lamp	Fluorescent	---	---
Reading Table	---	---	---
Reading guide	---	---	---
Signature guide	---	---	---
Walking Stick	---	---	---
Large print book	---	---	---
Notex	---	---	---
Needle threader	---	---	---
If any other devices	---	---	---

Signature: _____
 13/12/2019

Figure 4: Prescription for Low Vision Device

She was also trail by different absorptive lenses, but best acceptance was yellow filter for daylight illumination by which she was more comfortable in day time. She had also advised to use fluorescent table light in the time of reading. She was advised to next follow up 45 days for low vision adjustment.

As per instruction she was again attend to the low vision clinic with prescribed devices on 48th day. She was adjusted with her prescribed devices comfortable and also asked to follow up every two months for low vision evaluation.

3. Discussion

Patient those who have no visual improvement with regular spectacles or contact lenses even surgical procedures or medical treatment, they need to correct with low vision devices. A patient have low vision, it depends on the reducing of visual acuity as well as the reduction in visual function, like decreased colour vision, contrast sensitivity, binocularity, abnormality in ocular motility or glare sensitivity.

In Stargardt’s disease, the cone cells deteriorate in function but the rod cells remain intact^[3]. Symptoms usually start at childhood or adolescence age. People with Stargardt’s disease start to experience difficulty with reading. The diagnosis is usually made as a result of deterioration in visual acuity and typical appearances around the macula on ophthalmoscopy. Low vision services improve the quality of life and mental state^[4].

4. Conclusion

Stargardt's disease patients are managed by small amount of magnification. Optical devices like dome magnifier or bar magnifier for reading purpose and telescope for distance visual performance are very much preferable for Stargardt's disease patient. Fluorescent table light brighter the reading objects and yellow filters also helpful to manage of glare and photophobia, as non-optical devices. To know the progression of visual function, a regular follow up should be done after certain interval.

References

- [1] www.nei.nih.gov/eye-conditions-and-diseases/stargardt-disease.jul10,2019.
- [2] Kanski's Clinical Ophthalmology A systemic approach, By Brad Bowling, 8th edition, pg-654, pp-917
- [3] Dhungel P, Joshi ND. Low vision management in a case of stargardt's disease. Adv Ophthalmol Vis Syst. 2015;2(1):11-13
- [4] Horowitz A, Reinhardt JP, Boerner k. The effect of rehabilitation on depression among visually disabled older adults. Aging Ment Health.2005;9(6):563-570