

Case Series: Tinted and Light Adaptive Contact Lenses as a Therapeutic Method in Siblings with Complete Achromatopsia

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BACKGROUND

Achromatopsia, a rare autosomal recessive inherited retinal disorder, presents with absent color vision, severe photophobia, nystagmus, reduced visual acuity and reduced contrast sensitivity. Tinted lenses can reduce the effect of debilitating photophobia and improve visual function in these patients.¹ Herein we report on two brothers who had improved visual comfort with custom soft red tinted lenses. The benefits of light adaptive contact lenses for achromatopsia were also explored.

CASE 1

A 21 yo Hispanic male presented with a chief complaint of significant photophobia, and reduced visual acuity. He is currently a college student.

Past ocular history included a diagnosis of complete achromatopsia and an unknown ocular surgery in Chile at a young age (year unknown). The patient also had a history of corneal gas permeable lens wear with tinted lenses OU for 2 years, but had discontinued wear for 3 months due to pain, redness, and tearing with lenses. His medical history was unremarkable and he denied being on any medications.

Pertinent Findings

DVAsc: OD 20/200 OS 20/150

NVAsc: OD 20/125 OS 20/150

CTsc: Distance 2XP Near 4XP

EOMs: Full with mild nystagmus on left gaze OU

HRR Color Plates: Failed all plates OU

Anterior Segment: Unremarkable OU

Iris: Brown, flat, and intact OU

| Eye | Manifest Refraction | DVA |
|-----|---------------------|--------|
| OD | -2.00-2.00x180 | 20/100 |
| OS | -2.25-1.50x155 | 20/60 |

| Contact Lenses Dispensed | | | | | | |
|--------------------------|----------------|-----|------|--------------|--------------------------|--------|
| Eye | CL Power | BC | Diam | Tint Zone | Brand | DVA |
| OD | -2.00-1.75x180 | 8.6 | 14.3 | 10.5mm (red) | Orion BioMed Therapeutic | 20/100 |
| OS | -2.00-1.25x160 | 8.6 | 14.3 | 10.5mm (red) | Orion BioMed Therapeutic | 20/60 |

*NVA OU with contact lenses: 20/40 and reduced nystagmus



Figure 1. Patient 1 wearing Orion BioMed Therapeutic lenses OU with little to no cosmetic appearance of red tint

CASE 2

A 19 yo Hispanic male presented for a contact lens fit with complaints of severe photophobia and reduced VA. Ocular history included a diagnosis of complete achromatopsia. Medical history was unremarkable and the patient did not take any medications. Pt reported no previous contact lens wear. The patient currently works as a barber full time.

Pertinent Findings

DVAsc: OD 20/125 OS 20/80

NVAsc: OD 20/125 OS 20/80

CTsc : Distance 2 XP Near 5 XP

EOMs: Full with mild nystagmus OU

HRR Color Plates: Failed all plates OU

Anterior Segment: Unremarkable OU

Iris: Brown, flat, and intact OU

| Eye | Manifest Refraction | DVA |
|-----|---------------------|--------|
| OD | -2.50-1.50x045 | 20/125 |
| OS | -1.75-1.25x150 | 20/60 |

| Contact Lenses Dispensed after First Visit | | | | | |
|--|----------|-----|------|--|--------|
| Eye | CL Power | BC | Diam | Brand | DVA |
| OD | -3.25 DS | 8.4 | 14.0 | Johnson Vision Acuvue Oasys with Transitions | 20/100 |
| OS | -2.25 DS | 8.4 | 14.0 | Johnson Vision Acuvue Oasys with Transitions | 20/60 |

| Contact Lenses Dispensed at Follow-Up | | | | | | |
|---------------------------------------|----------------|-----|------|--------------|--------------------------|--------|
| Eye | CL Power | BC | Diam | Tint Zone | Brand | DVA |
| OD | -3.25 DS | 8.5 | 14.3 | 10.5mm (red) | Orion BioMed Therapeutic | 20/100 |
| OS | -1.50-1.25x150 | 8.5 | 14.3 | 10.5mm (red) | Orion BioMed Therapeutic | 20/80 |

*NVA OU with contact lenses: 20/50 and reduced nystagmus

Trial light adaptive contact lenses were dispensed at the initial visit to investigate improvements in visual comfort. Vision was minimally improved in the light adaptive lenses with mild improvement in visual comfort indoors. The patient however did not appreciate a significant difference in photophobia while outdoors. Upon comparison, the patient strongly preferred the custom soft red tinted lenses for vision and visual comfort from photophobia.



Figure 2. Patient 2 wearing no lens OD and Orion BioMed Therapeutic red tinted lens OS displaying little to no cosmetic difference between the two eyes

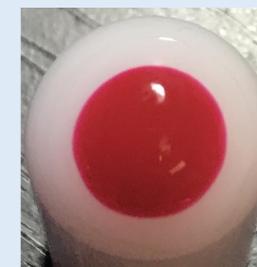


Figure 3. Sample Orion BioMed Therapeutic custom soft lens with center red tint zone. As seen in Figures 1 and 2, the red tint had little to no effect on iris color when worn on the eye in the two brown eyed patients.

DISCUSSION AND FUTURE CONSIDERATIONS

Patients with achromatopsia are typically managed by using tinted lenses to limit retinal light exposure and mitigate symptoms. Tinted contact lenses and cut off filters do this by transmitting light at wavelengths between 400nm and 480nm thereby reducing photophobia.¹ Red tinted contact lenses have been most successful in improving photophobic symptoms in patients with cone disorders.² Our cases supported previous reports of the benefit of tinted lenses, specifically red tinted lenses, in relieving photophobia.

Both patients preferred red tinted lenses in both eyes to have the most reduction in photophobia. The patient in Case 1 reported significantly increased visual comfort with the red tinted lenses both indoors and outdoors. While the patient in Case 2 appreciated some benefit of improved visual comfort indoors with the light adaptive lenses, the greatest reduction in photophobia was experienced with the red tinted lenses, particularly while outdoors. Low vision services were recommended.

To our knowledge, this is the first case report of management of photophobia with tinted lenses and light adaptive lenses in siblings with achromatopsia. Future investigations on the impact of light adaptive lenses on milder forms of photophobia are warranted.

CONCLUSION

Red tinted contact lenses are viable therapeutic and visual corrective options for enhancing the quality of life of patients coping with achromatopsia. Light adaptive contact lenses may be a promising and visually appealing option for managing visual comfort in patients with mild light sensitivity.

REFERENCES

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