



# Revolutionizing Low-Vision Tools: A Novel Approach for Stargardt Disease and Age-Related Macular Degeneration

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## ABSTRACT

This study explores the development of a new low-vision aid designed to enhance the quality of life for individuals with Stargardt disease and age-related macular degeneration (AMD). Both conditions lead to central vision loss, creating challenges in daily activities like reading, recognizing faces, and navigating public spaces. Traditional tools, such as bifocal monoculars and dome magnifiers, are often cumbersome and inefficient, particularly for school-aged children. This research aims to design an innovative, user-friendly visual aid that can support educational and social engagement, reducing the psychological impact of vision loss. While not a replacement for potential future treatments like gene therapy, this tool offers a significant improvement in accessibility and independence for those affected by these conditions. The development of new low-vision aid glasses designed for individuals with Stargardt disease and macular degeneration. These glasses are durable, rechargeable, and offer a convenient alternative to traditional tools like dome magnifiers and bifocal monoculars.

**KEYWORDS-** Low Vision Aid, Stargardt Disease, Age-Related Macular Degeneration (AMD), Central Vision Loss, Assistive Technology, Visual Impairment, Innovative Visual Aid, Dome Magnifiers, Bifocal Monoculars, School-Aged Children, Accessibility, User-Friendly Design, Educational Support, Social Engagement, Psychological Impact, Gene Therapy

## Introduction

Stargardt disease is a genetic eye condition that is inherited from generation to generation, while macular degeneration is an age-related eye disease. In macular degeneration, the majority of those affected are older adults, whereas Stargardt primarily impacts children and young adults. Despite these differences, both conditions lead to central vision loss, causing vision to become blurry or dark. Unfortunately, there is currently no cure for Stargardt disease. On the other hand, macular degeneration can be managed with treatments that vary depending on the type: wet AMD is treated with anti-VEGF drugs, while dry AMD has recently seen the introduction of medications like pegcetacoplan and avacincaptad pegol.

Hospitals and clinics often recommend traditional low vision aids, such as dome magnifiers and bifocal monoculars, to assist individuals with Stargardt disease and macular degeneration. These tools are vital, as early diagnosis is crucial in managing these progressive conditions. Although these diseases do not lead to complete blindness, individuals with Stargardt and macular degeneration often struggle to see objects clearly, though they may still perceive light. Stargardt, also known as juvenile macular dystrophy, can cause significant challenges for school-going children, making it difficult for them to see the blackboard, identify colors, or even recognize familiar faces in public settings.

For children, this impairment can severely affect their ability to engage in school activities and acquire knowledge. Traditional tools like dome magnifiers are helpful for reading, and bifocal monoculars can aid in viewing distant objects. However, using bifocal monoculars in a classroom setting is often cumbersome and time-consuming. This study proposes the design of a new, advanced low vision aid that aims to improve both the academic and social lives of individuals with Stargardt disease and macular degeneration.

The primary goal of this research is to provide clearer vision for those affected by these conditions. While this tool is not a substitute for a permanent cure, such as gene therapy, it holds the potential to significantly enhance the quality of life for those affected. By reducing indirect depression, stress, and social anxiety, this tool could make daily activities more manageable for those with vision impairments.

Here are a few scenarios where this low vision aid could prove invaluable:

- **Public Transportation:** Reading bus numbers or destination boards can be challenging for those with Stargardt or macular degeneration, often leading to missed buses. The time-consuming nature of adjusting bifocal monoculars makes this task even more difficult.
- **Classroom Learning:** Students with these conditions struggle to read content on blackboards or smartboards, making it hard to follow along with lessons and presentations.
- **Social Interactions:** Recognizing friends from a distance can be difficult, leading to potentially embarrassing situations where others may mistakenly think they are being ignored.
- **Cycling:** Riding a bicycle becomes hazardous when central vision is obstructed by a dark spot, making it hard to see traffic signs and signals.
- **Ordering Food:** Reading a menu board at a street food vendor can be a frustrating experience, particularly when it comes to selecting items or determining prices.

Reflecting on my own school experience, I recall a time during a work experience exchange period when I was seated far from the front, despite everyone knowing that I had difficulty seeing, even with glasses. During a video presentation, a classmate made a sarcastic comment about my inability to see, prompting the entire class to laugh. If I had access to a low vision aid at that time, I could have participated fully without facing such ridicule.

This low vision aid has the potential to foster self-reliance and help prevent bullying among school-going children with Stargardt and macular degeneration. By normalizing the use of such aids, we can create a more inclusive environment for these students. Therefore, the importance of this innovation cannot be overstated.

## 2. LITERATURE REVIEW

### 2.1 Stargardt Disease

Stargardt disease, also known as juvenile macular dystrophy, was first described by Karl Stargardt in 1909. The prevalence of this genetic eye condition is estimated to be between 1:8,000 and 1:10,000. Stargardt primarily affects children and young adults, leading to central vision loss, similar to macular degeneration, though both are retinal eye diseases. In Stargardt disease, the photoreceptor cells responsible for sensing light gradually die, resulting in dark spots in the macula. This condition often goes unnoticed initially, as many individuals mistake the early symptoms for myopia. While central vision becomes increasingly blurry, peripheral vision remains unaffected. Some individuals may also experience difficulties distinguishing colors and adapting to changes in light levels. Stargardt typically causes vision to decline to 20/40, meaning that an individual with the condition can see at 20 feet what

someone with normal vision can see at 40 feet. Vision may eventually deteriorate to about 20/200, after which it generally stabilizes, though variations in progression can occur. Since Stargardt is a hereditary condition, individuals with a family history of the disease are at a higher risk. However, for the disease to manifest, the affected individual must inherit the gene mutation from both parents. The gene most commonly associated with Stargardt disease is ABCA4. Diagnosis of Stargardt disease involves a comprehensive retinal examination by an ophthalmologist. A characteristic sign of the disease is the presence of yellowish flecks, known as lipofuscin, which are fatty deposits under the macula. These deposits impair vision. Diagnostic tools such as optical coherence tomography (OCT) and electroretinography (ERG) are used to evaluate retinal health. Additionally, a specific test called fluorescein angiography can be performed, where a dye is injected into the patient's arm, allowing the ophthalmologist to photograph the retina's blood vessels and identify dark spots indicative of Stargardt disease. Genetic testing can further confirm the diagnosis and differentiate it from other forms of macular degeneration.

Currently, there is no cure for Stargardt disease, but ongoing research in gene therapy and drug therapy offers hope for future treatments



Fig 1: figure shows how a person with Stargardt sees the world.

## 2.2 Macular Degeneration

Macular degeneration, commonly referred to as age-related macular degeneration (AMD), is a retinal disease that leads to the deterioration of the macula, resulting in central vision loss. This condition predominantly affects individuals aged 50 and older and is the leading cause of vision loss within this age group. There are two primary types of AMD: dry AMD and wet AMD.

**Dry AMD** is the more prevalent form, accounting for approximately 80% of all AMD cases, meaning 8 out of 10 people with macular degeneration have this type. In dry AMD, the macula gradually thins with age, and drusen—small clumps of proteins—begin to accumulate. Individuals with dry AMD may exhibit signs such as drusen, pigment abnormalities, or geographic atrophy, all of which contribute to retinal cell loss and a gradual decline in vision.

**Wet AMD** is less common but far more severe. It is characterized by the abnormal growth of blood vessels beneath the retina. These vessels may leak blood or fluid, leading to rapid and significant vision loss. Several risk factors increase the likelihood of developing AMD, including smoking, a diet high in saturated fats, obesity, and high blood pressure. Genetics also play a crucial role, as individuals with a family history of AMD are at greater risk. Additional risk factors include heart disease, elevated cholesterol levels, and ethnicity, with white individuals being more prone to developing AMD.

To diagnose AMD, an ophthalmologist will conduct a comprehensive eye examination. This may include the Amsler grid test, which helps identify blurry or blank spots in the patient's field of vision. The ophthalmologist



will then examine the retina using a special lens and light, often followed by imaging tests such as optical coherence tomography (OCT) and fluorescein angiography, to assess the condition of the macula and detect abnormalities. Treatment options for AMD differ depending on whether the patient has dry or wet AMD. Dry AMD currently has no definitive treatment, though certain vitamins and minerals may slow its progression. In contrast, wet AMD can be managed with medications such as anti-VEGF drugs, which inhibit the growth of abnormal blood vessels, and newer treatments like pegcetacoplan and avacincaptad pegol.

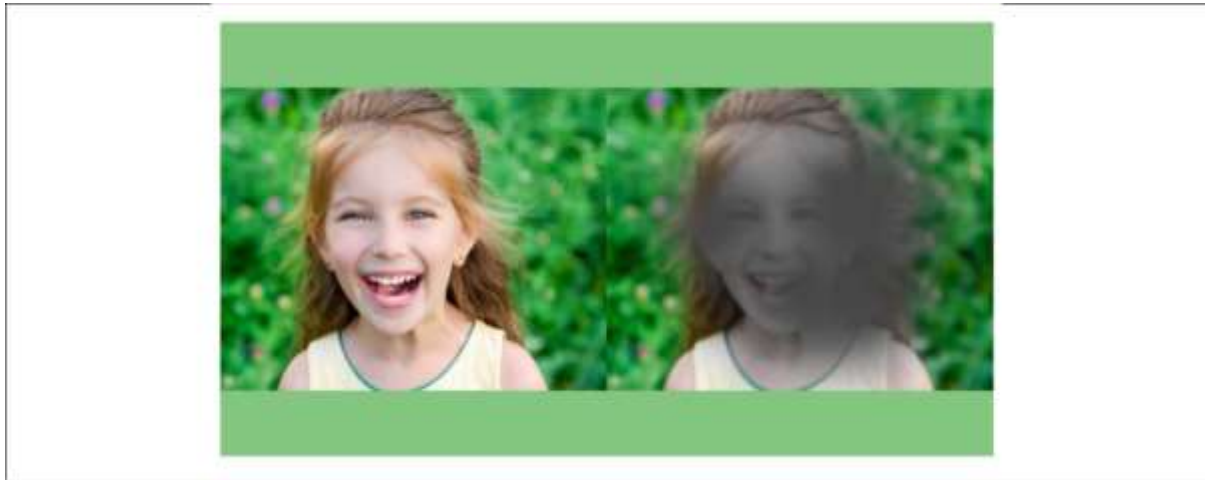


Fig 3: how a person with Macular degeneration sees the world.

## Geographic Atrophy and Treatments for AMD

Geographic atrophy is a significant condition associated with dry AMD, where the degeneration of retinal cells leads to irreversible vision loss. Currently, the only treatment option for geographic atrophy involves delaying its progression. Two medications, pegcetacoplan and avacincaptad, are administered via injections into the eye to help slow down the vision loss associated with this condition. Alongside these treatments, certain vitamins and minerals have been shown to be beneficial for managing dry AMD. These include vitamin C, vitamin E, lutein, zeaxanthin, zinc, and copper. However, it's important to consult with a healthcare provider before taking any supplements, as certain substances, like beta carotene, can increase the risk of lung cancer in smokers.

In addition to medication, lifestyle changes such as eating a diet rich in green leafy vegetables, fruits, vegetables, and fish can also be beneficial for individuals with AMD. For wet AMD, where abnormal blood vessels grow beneath the retina, anti-VEGF drugs are used to slow the leakage of blood vessels. Another treatment option for wet AMD is laser surgery, which can be used to reduce the abnormal blood vessels in the retina.

## Genetics of Central Vision Loss

Central vision loss, particularly in diseases like Stargardt, is often inherited in an autosomal recessive manner, meaning that both parents must carry and pass on the defective gene for a child to be affected. The primary gene associated with Stargardt disease is **ABCA4**, located on chromosome 1. Identified by the National Human Genome Research Institute in 1997, the ABCA4 gene plays a crucial role in the visual cycle. It is responsible for transporting Vitamin A derivatives across the disc membranes of photoreceptor cells in the retina, which is essential for clearing toxic byproducts and preventing their accumulation.

Mutations in the ABCA4 gene impair this process, leading to the buildup of toxic substances like lipofuscin, particularly A2E, which contributes to retinal damage. Gene therapy is a promising approach to correct these genetic defects, especially before significant cell loss occurs. Additionally, drug therapy and stem cell therapy are being explored as potential treatments. Drug therapy focuses on early intervention to slow the progression of the disease, while stem cell therapy aims to replace lost retinal cells with healthy ones, potentially restoring some visual function.

The **heterogeneity of mutations** in the ABCA4 gene is notable, with approximately 154 different mutations identified in individuals with Stargardt disease. The variability in the disease's presentation can be influenced by a combination of genetic, environmental, and individual factors.

Another condition, **Stargardt-like macular degeneration (STGD3)**, shares similarities with Stargardt disease but is caused by a mutation in the **ELOVL4 gene**. This gene is involved in the production of long-chain fatty acids essential for retinal health. Mutations in ELOVL4 disrupt this process, leading to retinal degeneration and central vision loss. STGD3 typically begins in adolescence or early adulthood and is linked to a specific region on chromosome 6. Research in mice has shown that disruptions in the ELOVL4 gene can result in photoreceptor cell damage, highlighting the importance of this gene in maintaining normal vision.

Understanding the genetic basis of these conditions provides valuable insights into potential treatments and the development of therapies that could one day halt or reverse the progression of vision loss in affected individuals.



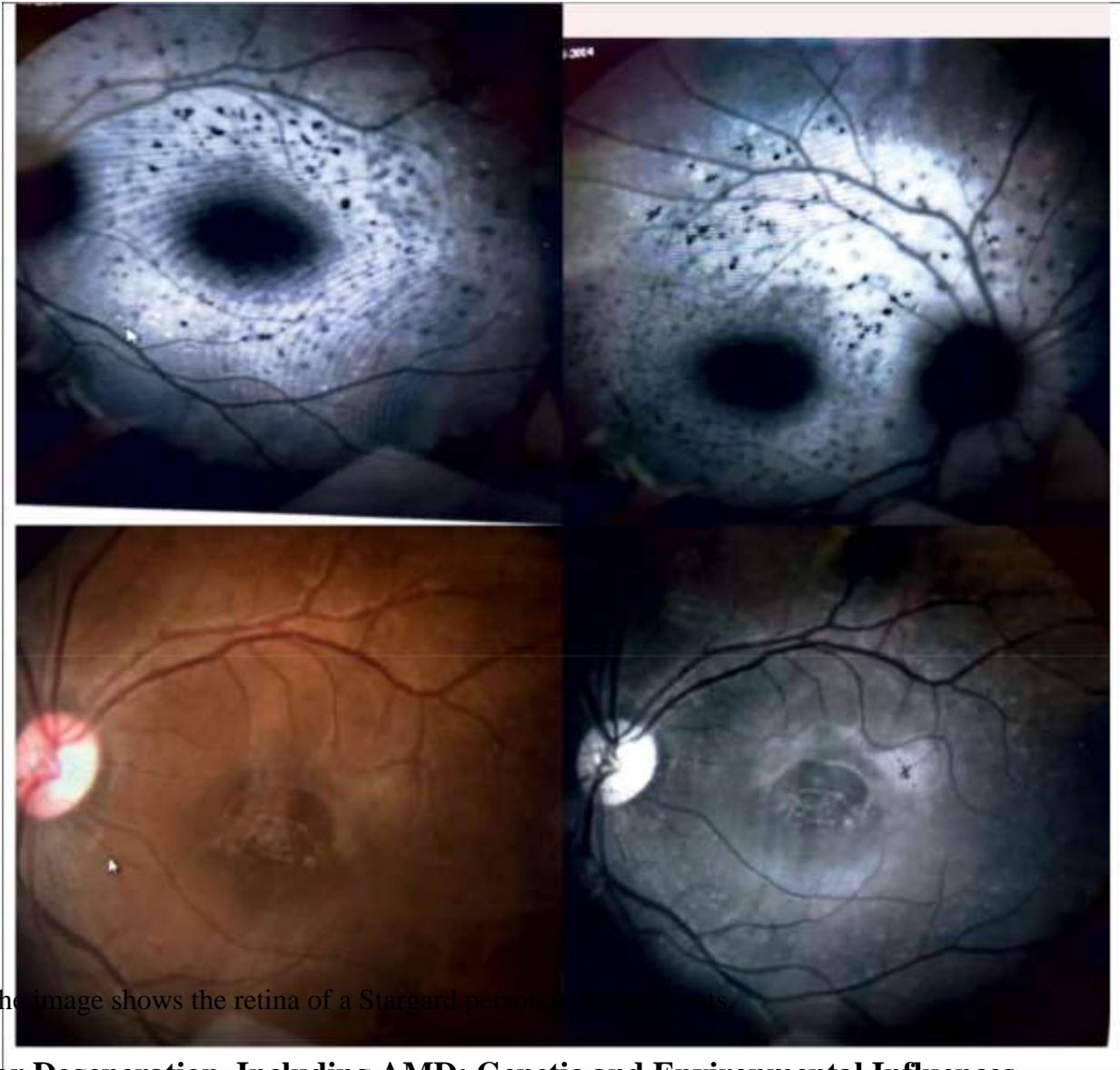


Fig 4; The image shows the retina of a Stargardt person with dark spots.

### Macular Degeneration, Including AMD: Genetic and Environmental Influences

Macular degeneration, particularly age-related macular degeneration (AMD), is influenced by a combination of genetic and environmental factors. Understanding the genetic basis of conditions like Stargardt disease (STGD3) and AMD is crucial for advancing research and developing potential treatments. Genetic mutations, such as those in the **ABCA4** gene for Stargardt disease and **ELOVL4** for STGD3, play a significant role in these conditions, but lifestyle factors like diet, exercise, and smoking habits also contribute to their progression.

## Diagnostic Tests for Macular Degeneration and Stargardt Disease

### 1. Fundus Photography:

The fundus, or rear surface of the eye, includes the retina, macula, optic disc, fovea, and blood vessels. Fundus photography uses a specialized camera to capture images of the back of the eye through the pupil. These images help eye doctors diagnose, monitor, and treat retinal diseases, including AMD and Stargardt disease.

### 2. Optical Coherence Tomography (OCT):

OCT allows ophthalmologists to see the distinct layers of the retina, helping them map and measure the thickness of these layers. This test is crucial for diagnosing and treating retinal diseases such as AMD and diabetic eye disease. OCT angiography (OCTA) is a variation of this test that images the blood vessels in and under the retina, similar to fluorescein angiography but faster and without the need for dye.

### 3. Electroretinogram (ERG):

An ERG measures the electrical activity of the retina in response to light, confirming retinal diseases. This test is vital for diagnosing conditions like Stargardt disease and AMD.

## Preventive Measures for Stargardt Disease and AMD

To slow down the progression of retinal diseases like Stargardt disease and AMD, certain preventive measures can be taken:

### 1. Mediterranean Diet:

Adhering to a Mediterranean diet, which includes high intakes of vegetables, fruits, legumes, grains, and nuts, with moderate consumption of fish, poultry, dairy, and red wine, is associated with a significantly lower risk of developing AMD and slowing its progression. The diet also emphasizes the use of olive oil over butter and limits red meat consumption.

### 2. Supplements:

AREDS (Age-Related Eye Disease Study) supplements, containing vitamins C and E, zinc, copper, lutein, and zeaxanthin, have been shown to slow down the progression of intermediate-stage AMD. Lutein and zeaxanthin, in particular, filter out harsh light and are associated with a reduced risk of advanced AMD.

### 3. Exercise and Weight Loss:

Maintaining a healthy BMI and waist-to-hip ratio (WHR) is important, as increased BMI is linked to both early and late AMD. Regular physical activity, especially three hours per week of low to moderate exercise, is associated with a lower risk of developing late AMD.

### 4. Sleep:

Poor sleep quality, including sleep apnea and insomnia, increases the risk of developing AMD. Shorter sleep duration (less than 6 hours) is particularly associated with an increased risk of wet AMD, while longer sleep duration may reduce this risk.

### 5. Avoid Smoking:

Smoking is the leading environmental cause of AMD, significantly increasing the risk of developing the condition. Quitting smoking is one of the most effective preventive measures against AMD. These preventive



measures, combined with an understanding of the genetic factors involved in conditions like Stargardt disease and AMD, can help manage and reduce the risk of vision loss

## Traditional Low-Vision Aids

Traditional low-vision aids play a crucial role in enhancing the quality of life for individuals with vision impairments such as Stargardt disease and age-related macular degeneration (AMD). Some of the commonly used aids include:

1. **Sunglasses:** Sunglasses protect the eyes from harmful UV rays, reducing glare and preventing further damage to the retina.
2. **Blue Light Glasses:** These glasses are designed to protect the eyes from the blue light emitted by screens, such as those on laptops, smartphones, and other digital devices, reducing eye strain and discomfort.
3. **Yellow-Tinted Lenses:** For individuals with AMD, yellow-tinted lenses can enhance contrast and improve visual clarity, making daily activities like reading easier.
4. **Anti-Glare Coating:** Glasses with anti-glare coatings help reduce reflections and improve visual comfort, especially in bright environments.
5. **Polycarbonate Lenses:** These lenses are impact-resistant and provide protection against eye injuries, making them ideal for individuals who are at risk of accidents or falls due to low vision.
6. **Bifocal Glasses:** Bifocals offer dual vision correction, allowing individuals to see both near and distant objects clearly, which is essential for activities like reading and driving.
7. **Magnifying Glasses:** Magnifying glasses can enlarge text and images, making it easier for individuals with low vision to read and perform other close-up tasks.
8. **High-Powered Lenses:** These lenses provide stronger magnification than standard lenses, aiding in tasks that require detailed vision.
9. **Telescopic Glasses:** Telescopic glasses incorporate small telescopes within the lenses to magnify distant objects, improving the ability to see far-away details, such as street signs or presentations.

These traditional aids, while effective, often have limitations, such as being cumbersome or insufficient for prolonged use. As a result, there is a growing need for more advanced and user-friendly low-vision aids that can better support individuals in their daily lives.

## FOR READING DOME MAGNIFIER AND

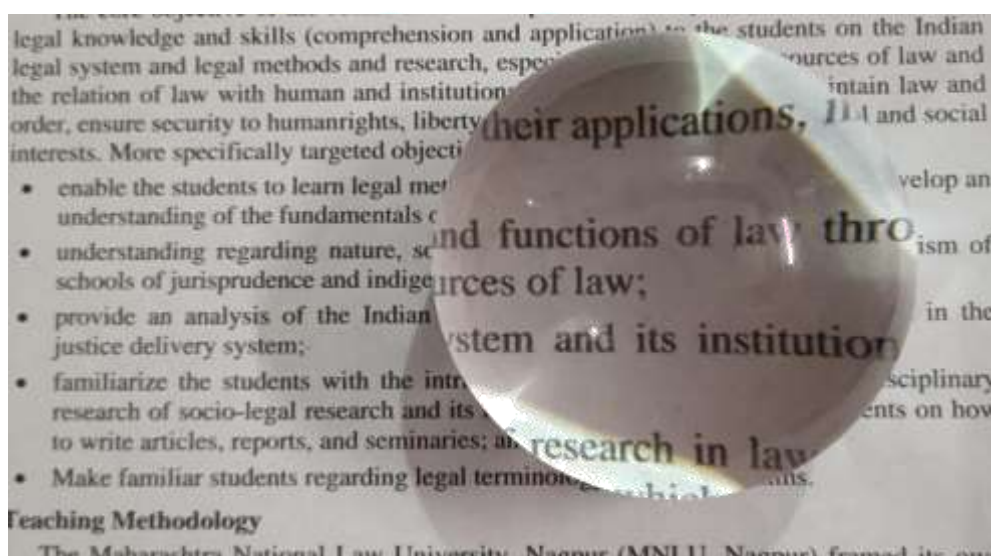


Fig 5 Image of a dome magnifier.



A Dome magnifier is used to see the text in large it helps read text like books and newspapers. It needs a little bit of practice to use it once you are fluent it is relaxing for the eyes There is a digital version of it called the digital magnifier on that device you can change the text colour and font.the only limitation is that it is expensive.

### BIFOCAL MONOCULAR

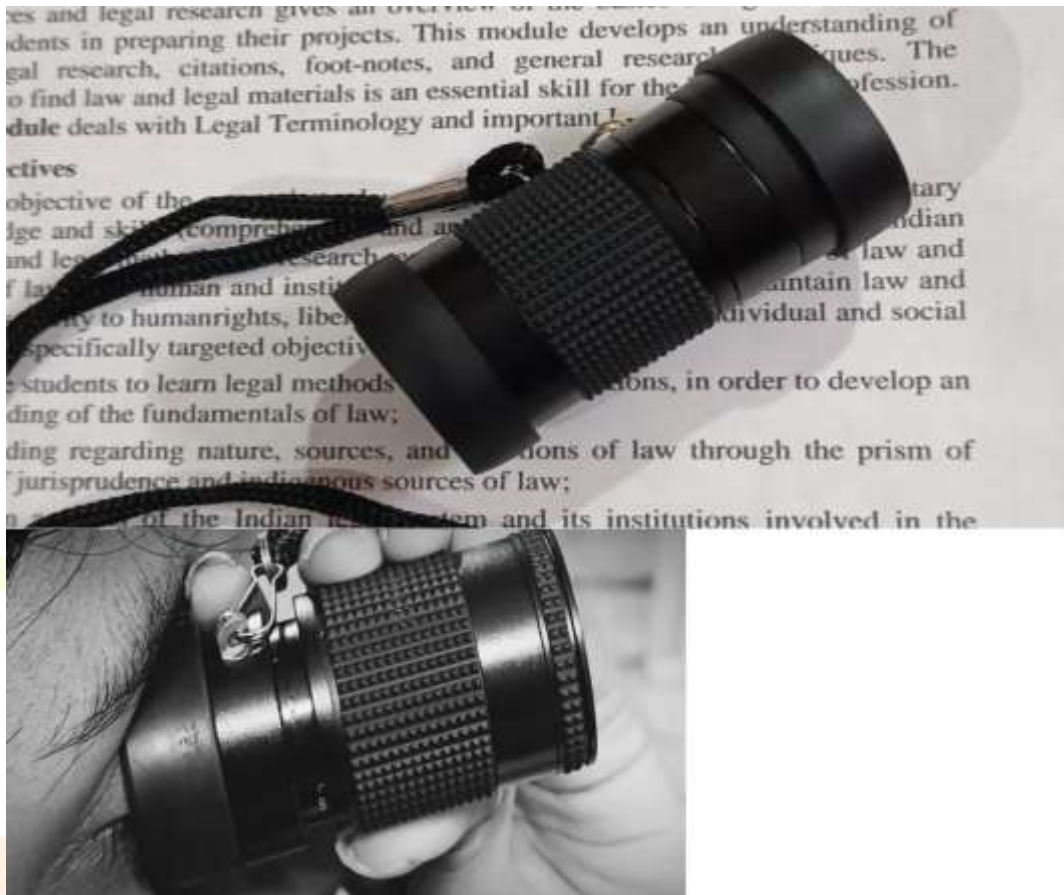


Fig 7 image shows the bifocal monocular near eyes.

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The bifocal monocular is expensive and heavy but useful to see far away objects this has two lenses soft side is for the eyes and the focal length of the lens is for the looking side. adjusting it is time-consuming and is not possible in public as it is uncomfortable.

## **Revolutionizing Low Vision Aids: Designing Practical Glasses for Stargardt and Macular Degeneration**

Traditional low vision aids like dome magnifiers are convenient for reading, while bifocal monoculars represent a significant innovation in visual assistance. However, using these devices in public can be uncomfortable and time-consuming, as they require constant adjustment of the focal length. To address these challenges, I have designed a new type of glasses tailored to school-going students, incorporating basic features that align with school codes of conduct. These glasses are not only more durable and cost-effective but also provide greater convenience for users, aiming to improve both educational experiences and social interactions.

Inspired by the innovative designs of the Chinese company Superhexa, these glasses offer a practical solution for those with Stargardt disease and macular degeneration. Below is a rough diagram (Fig 8) that illustrates the future design of these low vision aid glasses.

### **Key Features of the Glasses:**

#### **Control System for Lenses:**

**Zooming Capability:** The glasses are equipped with variable focus lenses that utilize electro-active technology, allowing for electronic adjustment of focal length. This enables in-and-out zooming without the need for manual adjustments.

**Actuators:** Tiny motors or piezoelectric components are integrated into the design to move the lenses and change the zoom level smoothly.

**Microcontroller:** A microcontroller manages user inputs and operates the zooming mechanism, ensuring a seamless experience.

#### **Blue Light Filtering:**

The lenses are coated with a blue light filter to block or reduce harmful blue light, protecting the eyes during prolonged use.

#### **Touch Control Interface:**

**Touch Sensors:** Capacitive touch sensors are embedded in the lens or frame to detect user inputs. The interface is simple and intuitive, allowing users to control the zoom by swiping or tapping.

**Embedded System:** A compact, energy-efficient microprocessor processes touch inputs and adjusts the lenses accordingly.

### Rechargeable Battery:

The glasses are powered by a long-lasting, rechargeable battery, ensuring convenient and uninterrupted use throughout the day.

These glasses are designed with the needs of school-going children in mind. The zooming feature is calibrated to stay within the natural range of the human eye, ensuring that the product remains safe and compliant with regulations. Additionally, the battery is engineered to provide extended usage, minimizing the need for frequent recharging.

To bring this innovative product to market, the idea can be sold to companies that specialize in research and development. By targeting hospitals and clinics, these companies can effectively reach those who would benefit most from this low vision aid, while also enhancing their reputation and goodwill. The development team must keep in mind the primary goal: to create a practical, reliable, and accessible solution for young students with visual impairments, enabling them to thrive in their educational and social environments.



Fig 8. Rough design of future low vision eye aid.

### CONCLUSION

A new innovation in low vision aid glasses has been developed to enhance the quality of life for individuals with Stargardt disease and macular degeneration. These glasses are designed to be durable, rechargeable, and easy to use, significantly improving traditional assistive tools such as dome magnifiers and bifocal monoculars. The primary goal of this study is to provide a comprehensive analysis comparing the effectiveness of these innovative glasses with traditional low vision aids. Traditional tools often present challenges, especially in public settings, where their use can be cumbersome and impractical. In contrast, these new low vision glasses offer convenience, enabling users to carry and wear them like regular glasses,



eliminating the need to juggle multiple devices. For young adults and teenagers, particularly those in school, these glasses represent a significant advancement. They allow for seamless transitions between activities such as reading, writing, and other academic tasks, providing a "two-in-one" solution that enhances both functionality and ease of use. Users can engage in everyday activities with greater independence and confidence, as these glasses simplify the process of managing visual impairments. Future studies will focus on systematically analyzing the performance and reliability of these low vision glasses. By examining their effectiveness in real-world settings, researchers aim to publish findings that will further validate the practicality and benefits of this innovative solution for those living with Stargardt disease and macular degeneration

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**NOTE FIGURES 4,5,6,7 AND 8 ARE FROM AUTHOR S PERSONAL COLLECTION.**

