PRESCRIBING FOR PATIENTS WITH GLAUCOMA AND RETINITIS PIGMENTOSA

AUTHOR

Hasan Minto: Brien Holden Vision Institute, Pakistan

PEER REVIEWERS

S May Ho: Brien Holden Vision Institute, Australia

Stephanie Schmiedecke Barbieri: University of the Incarnate Word

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AIMS

This module aims to outline the methods for prescribing vision correction for patients with glaucoma and retinitis pigmentosa via developing a:

- Systematic approach to vision assessment
- Protocol for visual enhancement

LEARNING OUTCOMES

After completing this module, students should be able to:

- Understand the functional implications of glaucoma and retinitis pigmentosa
- Design an optimum vision assessment strategy
- Implement a rehabilitation strategy

INTRODUCTION

This chapter includes a review of:

- Glaucoma, retinitis pigmentosa and their implications on visual function
- Management of low vision clients with glaucoma and retinitis pigmentosa
- Rehabilitation of low vision clients with glaucoma and retinitis pigmentosa
- Optical and non-optical low vision interventions

GLAUCOMA OVERVIEW

Glaucoma is a progressive disease of the optic nerve characteriseds by the loss of the retinal neurons and axons (nerve fibres) which can lead to a loss of vision, especially if left untreated. While increased intraocular pressure is the strongest risk factor for glaucoma, it is not the only factor for inducing neuropathy. There are many forms of glaucoma including primary and secondary glaucomas, open or closed angle glaucoma, high tension and low tension (normotensive) glaucoma. Glaucoma can also be congenital and affect children. These children generally have buphthalmic eyes with cloudy corneas.

Glaucoma is an important cause of vision loss. Glaucoma is estimated to account for 6.6% of blindness and 2.8% of vision impairment globally (Bourne et al, 2013).

Treatment options for glaucoma are limited in developing countries. Not all cases are suitable for surgery and laser treatment may not be available or suitable. Therapeutic treatment involves long term usage of medications which are expensive and may not be readily available. Compliance with medications is also an issue as there is no perceived benefit of using the treatment, in fact the potential side effects are often more apparent to the patient than the effectiveness of the treatment.

In early stages of the disease, patients are largely asymptomatic, earning it the moniker ‘silent’ or ‘sneak thief of sight’.

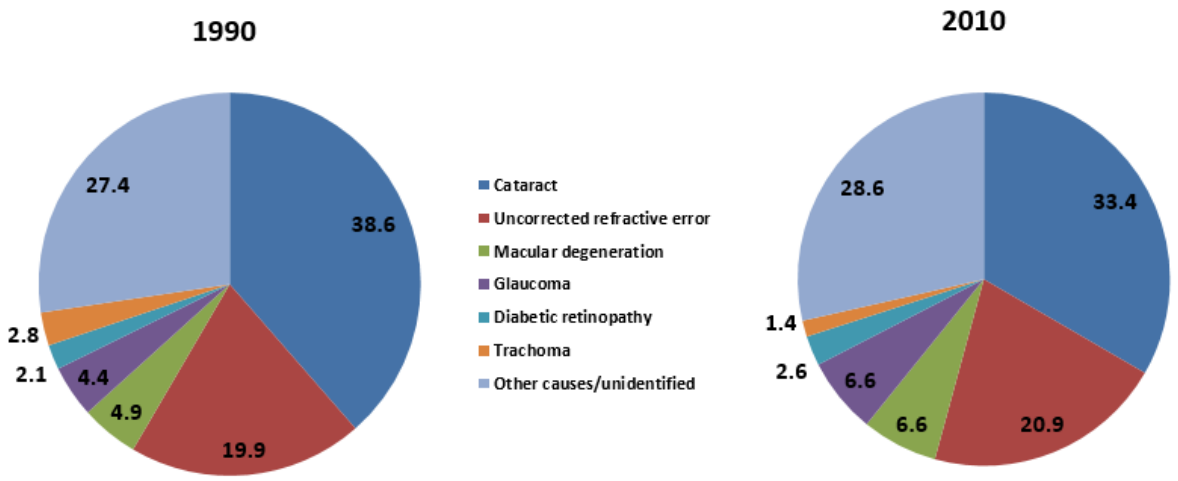

Figure 1: Global prevalence of blindness (Bourne et al, 2013)

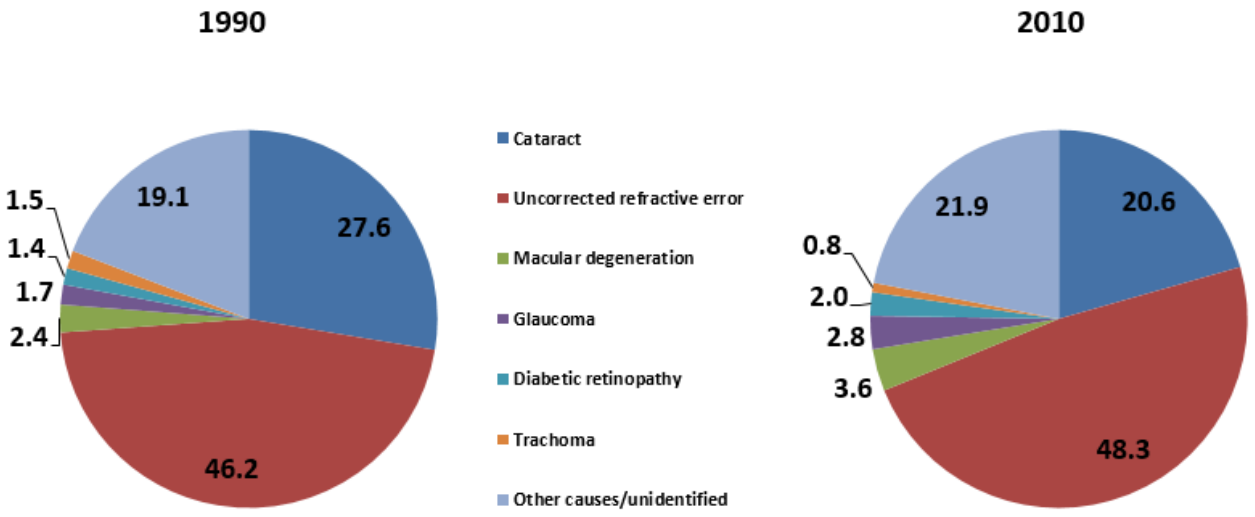

Figure 2: Global prevalence of vision impairment (Bourne et al, 2013)

Vision loss due to glaucoma arises due to the damage of the optic nerve and retinal nerve fibre layer. The peripheral visual field is lost first, typically in a pattern showing a nasal step. Para-central scotoma progressing to meet the physiological blind spot is also seen. Visual field loss is progressive.

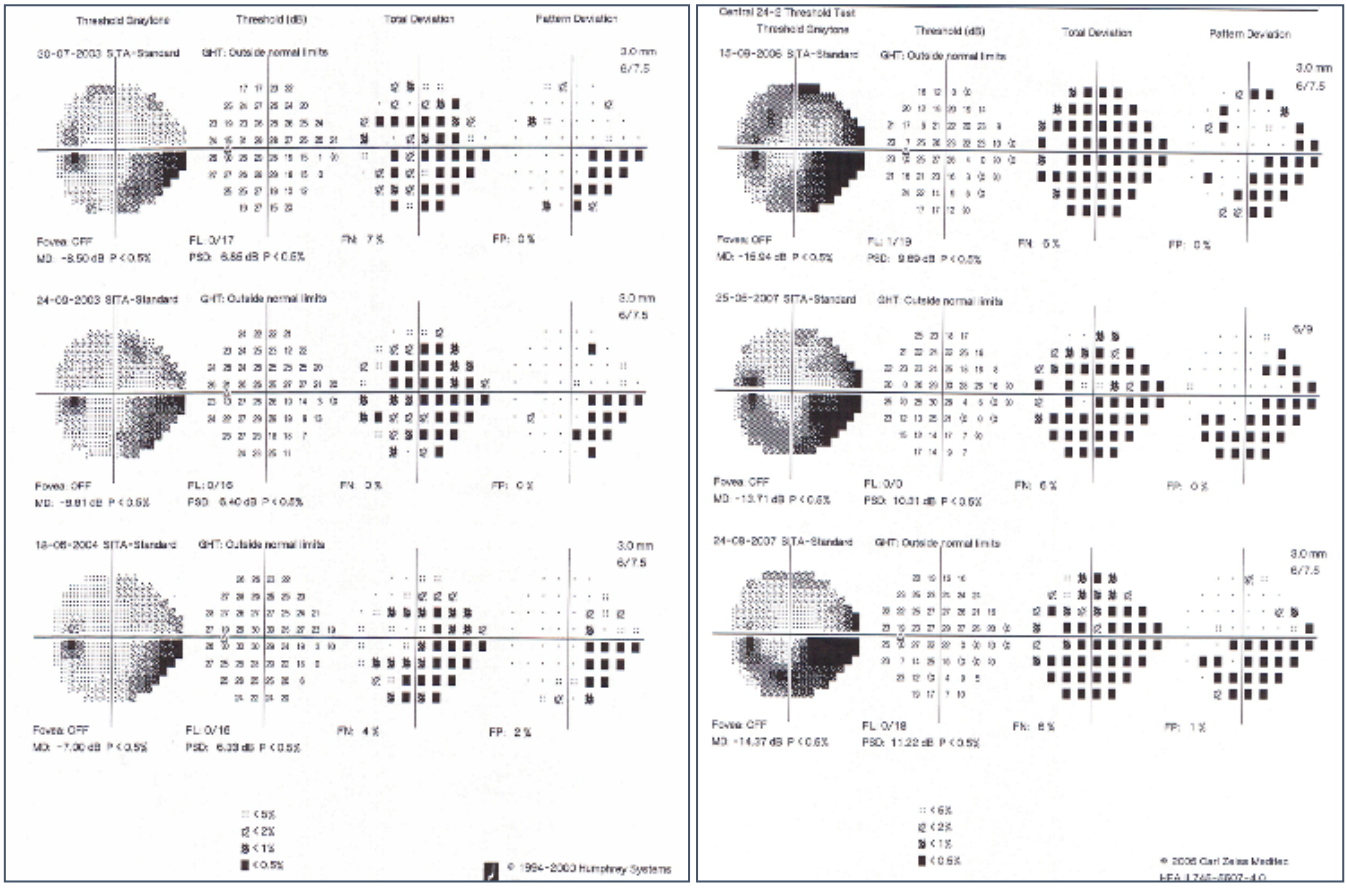

Figure 3: Visual fields in advanced glaucoma. Left: Nasal step. Right: Para-central scotoma progressing to meet physiological blind spot.

FUNCTIONAL IMPLICATIONS OF GLAUCOMA

Patients with glaucoma often show:

- Decreased peripheral vision
- Decreased contrast sensitivity
- Increased glare sensitivity
- Decreased visual acuity
- 20yr marker for Alzheimer's (Inoue et al. 2013)

As with other conditions, management of the patients with glaucoma focuses on maximizing residual vision and reducing visual symptoms.

BEHAVIOURAL MANIFESTATION OF VISION LOSS DUE TO GLAUCOMA

Loss of the peripheral visual field could result in:

- Adoption of unusual head postures to compensate for the field loss
- Difficulty in moving around safely, unable to negotiate obstacles
- Difficulty driving or loss of privilege to drive, as unable to detect vehicles or objects in areas of visual field loss
- Difficulty in reading – unable to track lines, losing place when reading
- Inability to locate objects placed in areas of field loss, i.e. cannot locate lost objects.

Decreased visual acuity could lead to:

- Difficulty in locating and reading road signs when driving
- Difficulty in driving
- Difficulty in reading and writing
- Difficulty recognising faces

Decreased contrast sensitvity can cause difficulty with various daily tasks, such as mobility tasks like negotiating steps and navigating from indoors to outdoors, reading materials with low contrast (e.g. newspapers), identifying coins and currency.

Increased sensitvity to glare can cause difficulty in mobility on sunny days or under bright indoor overhead lighting.

RETINITIS PIGMENTOSA OVERVIEW

Retinitis pigmentosa (RP) is a progressive genetic disease affecting the retinal photoreceptors and the retinal pigment epithelium. The diagnostic criteria for this disease include:

- Bilateral presentation
- Loss of peripheral vision
- Impaired dark adaptation
- Progressive loss of photoreceptor function

RP affects 1/3,000 to 1/5,000 patients and is found in people of all ages – typically starting in childhood or adolescence with symptoms of night blindness.

RP can be limited to the eye or manifest as part of a systemic disorder. It can also be part of a syndrome where other senses are involved such as deafness in Usher Syndrome. Diagnosis is made by ocular examination, electrodiagnosis (electroretinogram) and visual field assessment. Posterior subcapsular cataracts are common in all types of RP.

Vision loss is characteriseds by loss of night vision (nyctalopia), visual field constriction and reduced contrast sensitivity. In the forms of the disease where the macula is involved, there is a loss of visual acuity and eventually central vision. Mild loss of colour vision may also be found.

FUNCTIONAL IMPLICATIONS OF RETINITIS PIGMENTOSA

Patients with RP often show:

- Poor night vision
- Decreased peripheral vision
- Decreased contrast sensitivity
- Increased glare sensitivity
- Decreased visual acuity

Management of patients with RP focuses on maximisings residual vision and rehabilitation where possible.

BEHAVIORAL MANIFESTATION OF VISION LOSS DUE TO RP

Constriction of the visual field could result in:

- Adoption of unusual head postures to compensate for the field loss – e.g. face turn towards the area of loss
- Difficulty in moving around safely, unable to negotiate obstacles, bumps into things – 'clumsy'
- Difficulty driving, unable to detect vehicles or objects in areas of visual field loss
- Difficulty in reading – e.g. unable to track lines, losing place when reading
- Inability to locate objects placed in areas of field loss – e.g. cannot locate lost objects.

Visual acuity usually remains good until the late stages of the disease. Decreased night vision could lead to avoidance of activities at night time. Delayed adaptation moving from light to dark and vice versa.

Decreased contrast sensitvity can cause difficulty with negotiating steps, reading materials with low contrast (e.g. newspapers), identifying coins and currency.

Increased sensitvity to glare can cause difficulty in mobility on sunny days or under bright indoor overhead lighting.

ASSESSMENT

A careful history focusing on the patient's symptoms and activities is important. The history should be goal oriented, it is necessary to ascertain what the patient would like to be able to see and to do. In addition to measuring visual acuities, visual field assessment and contrast sensitivity tests should be undertaken. Glare sensitivity should be tested if the equipment is available.

VISUAL ACUITY ASSESSMENT

Visual acuities can be measured using the usual LogMAR charts or flip charts, varying the test distance as necessary depending on the patient's visual acuity.

In cases where the visual acuity is very poor, the Berkeley Rudimentary Vision Test (BRVT) can be employed. The BRVT uses three pairs of hinged cards, 25 cm square. It is a simple and efficient test of spatial vision that extends the range of measurement from the limits of the letter chart up to light perception.

VISUAL FIELD ASSESSMENT

There are several methods of perimetry that can be employed. In low vision patients with glaucoma, the purpose is to assess functional loss and not threshold loss for monitoring disease progression. If using automated perimetry, the Esterman binocular field on the Humphrey's Field Analyser with a testing pattern of 120 points extending over 150° bitemporally is the method of choice to assess functional vision loss. This will give an idea of the extent of loss of peripheral vision and may give a more real-life functional assessment of loss.

Goldmann perimetry is also a valuable tool in estimating the functional visual field as it offers a kinetic method of assesing the visual field. Some patients with low vision respond better to kinetic testing.

The Bjerrum or tangent screen offers an economical albeit gross method to assess the peripheral visual field. Other options such as a disc perimeter can also be employed. For patients with advanced vision loss, the high contrast variable sized test and fixation targets may make these tests easier to use.

The results of visual field assessment can be used for advising the patient on mobility strategies and their eligibility to drive.

Ranges of visual field loss
(Visual standards, International Council of Ophthalmology, 2002)

Range of normal vision:
60° visual field radius or more
Mild field restriction
A loss of peripheral vision such that only 40-50° of the visual field remains
Moderate field restriction
The visual field is reduced to between 10-30° degrees
Severe field restriction
The visual field is 10° or less

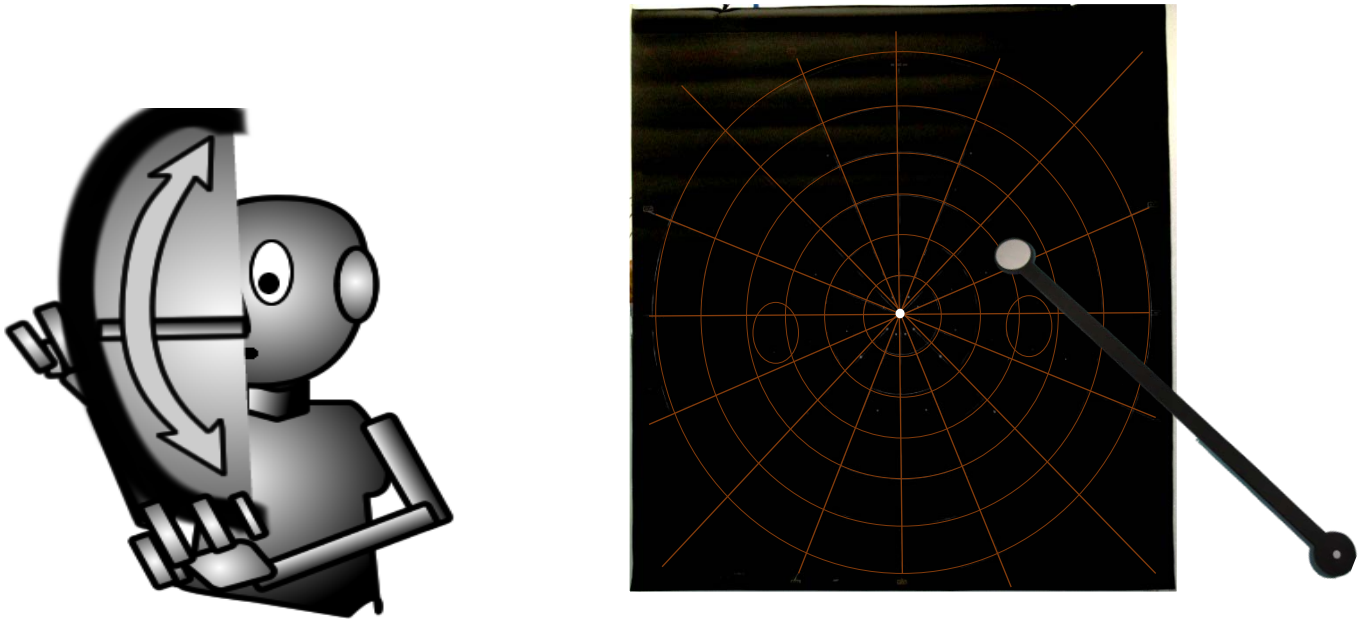

Figure 4: Visual field testing. (Left) demonstration of handheld arc perimetry testing superior and inferior field in free space. (right) tangent screen measuring central field

CONTRAST SENSITIVITY ASSESSMENT

Contrast sensitivity can be assessed in several ways. Tests can be designed as low contrast visual acuity charts (varying letter size), or with one symbol at and several contrast levels (e.g. Pelli-Robson)

- Plotting the contrast sensitivity curve (VISTECH® chart)
- High spatial frequency vs low spatial frequency charts
- Lea symbols
- Pelli-Robson
- Hiding Heidi (for children)

GLARE SENSITIVITY ASSESSMENT

Glare sensitivity can be assessed using a Brightness Acuity Tester (BAT). Visual acuity is measured pre- and during exposure to a known glare source (BAT). The BAT has three settings simulating direct overhead sunlight, partly cloudy day and bright overhead commercial lighting. The difference between the visual acuity pre- and during exposure shows the effect of the different types of glare on the patient.

Figure 5: Brightness Acuity Tester

LOW VISION MANAGEMENT OF PATIENTS WITH GLAUCOMA AND RETINITIS PIGMENTOSA

The general principle of management of low vision patients is to enhance the residual visual function by increasing the visual information reaching the brain through an impaired eye by:

- Sharpening the retinal image
- Increasing the size of the image
- Improving the contrast
- Displacing the image out of the scotoma
- Visual training

SHARPENING THE RETINAL IMAGE

Refraction

Optimizing vision is important. A good refraction should be performed and appropriate spectacles/optical correction prescribed, even if visual acuities are low.

Large steps and bracketing techniques should be employed to find the endpoint of refraction efficiently.

INCREASING THE SIZE OF THE RETINAL IMAGE

Image size at the retina can be increased by employing:

- Relative distance magnification
- Relative size magnification
- Optical magnification
- Electronic magnification

Relative distance magnification

Image size can be increased by simply approaching the object, ie reducing viewing distance to increase the viewing angle. For example, one could sit closer to the television, hold reading materials at a closer distance or use a reading stand.

Relative size magnification

Increasing the size of the object viewed will result in a corresponding increase in retinal image size. Examples of this are large print books, using thick felt tip pen for writing rather than a conventional pen.

Optical magnification

Optical devices ranging from simple hand held magnifiers to telescopes can be used to enlarge the image size. The topic of optical magnification is covered elsewhere in the course.

Electronic magnification

Image size can also be enlarged using electronic means such as

- Projectors
- Multimedia
- CCTV
- Software programs

Where available, electronic devices offer a very good option for magnification. Miniaturisations of electronics means that these devices are becoming more portable. The drawback is the high costs associated, although these are now also decreasing, making them an increasingly more viable option.

IMPROVING CONTRAST

Contrast sensitivity is often reduced in people with either glaucoma or RP. Several strategies can be used to improve contrast including:

- Increase illumination
- Decrease glare
- Filters
- Skillful use of color contrast

Increasing task illumination can improve contrast. However, care must be taken not to introduce disability glare while increasing contrast.

Decrease glare by using matt surfaces or reverse contrast when reading e.g. white print on black paper. Using typoscopes made from a black card will also help to reduce glare from white paper, in addition to helping the reader keep their place when reading.

Coloured filters such as coloured overlays can be used on reading materials. Yellow overlays can provide better contrast for reading black on white print.

Colour contrast can also be employed e.g. using coloured plates on white tablecloth or placemat, white rice on dark coloured plate, black coffee in white cup (see figure 6).

Figure 6: Methods to improve contrast for everyday tasks

DISPLACING THE IMAGE OUT OF A SCOTOMA

Eccentric viewing

Patients can be trained to view eccentrically by:

- turning their face to the area of field loss, or
- moving their eye instead of their head so that they keep a normal head position.

Use of Prisms

Prisms can be used to move objects into the field of view. This can help persons with constricted visual fields to be more ‘aware’ of objects in the periphery. Shifting the the image will also eliminate the need for inefficient and unusual neck movements to compensate for field loss. It is essential to correctly measure the amount of prism required and it is critical to position the prisms correctly on the spectacle lenses. Stick-on Fresnel prisms, which can be cut to size, is a useful way to trial the positioning of the prism. The base of the prism should be pointed towards the direction of the field loss.

Then, at each follow-up appointment, if they state that the prism is getting in their way, continue to cut back at each visit until it is not. This is reinforcement that they are scanning.

Fresnel prism treatment

- Install 25 prism diopter base out R and L
- Prism scanning and training at home for 2 weeks.
- The image will be displaced and the extent depends on the amount of prism prescribed. The patient will have to be trained to reach for objects or people as their positions will not be where they anticipate them to be.
- Some diplopia may also be experienced.
- The prisms will also cause a reduction in contrast and visual acuity.
- Outdoor adaptation is also required as moving around with prism lenses will be disconcerting.
- At follow-up, move the prism an additional 3 mm temporal to encourage further scanning. Whenever the prism is getting in the patient's way, we move the prism until it's no longer in their direct view.
- The final placement of the prism is based on the desire of the patient to continue to use prism for detection.

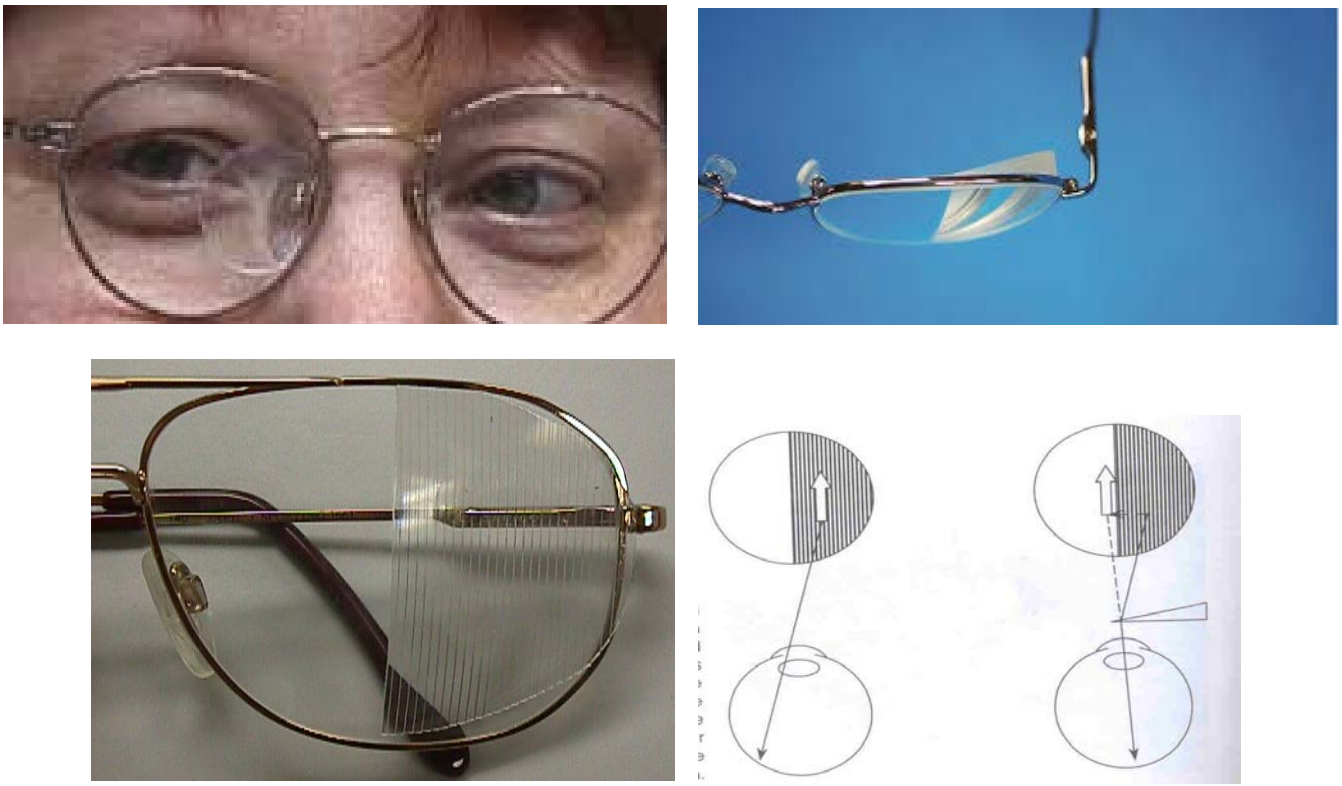

Figure 7: Fresnel and conventional prisms

FIELD ENHANCERS

Where the disease is advanced, the visual field can become severely constricted. The visual field can be artificially expanded by using, for example:

- Reversed telescope
- Fresnel prism
- Mirror system
- Concave lenses

Viewing through a reversed telescope would effectively be rendering it into a miniscope where the object is minified, thereby fitting more into the field of view (See Fig 8 below). The concept is that if images are minified, more information can be seen at one time in the same field.

A major disadvantage is since objects are twice as small, detail is lost. Its usefulness is limited, however, by the visual acuity of the patient, which if reduced will restrict what the patient can resolve visually. This method is useful for short spotting tasks. The reversed telescopes can be hand held or spectacle mounted (Fig 9).

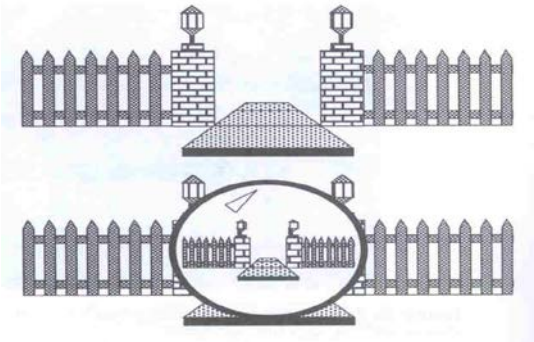

Figure 8: Viewing through a reversed telescope

Figure 9: Spectacle mounted reverse telescopes.

Viewing through a high minus lens held at arms length (Fig 10) also produces a similar effect. This is more economical than using a reversed telescope. The increase in visual field is equivalent to the inverse of magnification.

Figure 10: Viewing through a high minus “minifier” lens.

Mirrors can also be used to shift images into the field of view (Fig 11). They act in a similar fashion to prisms.

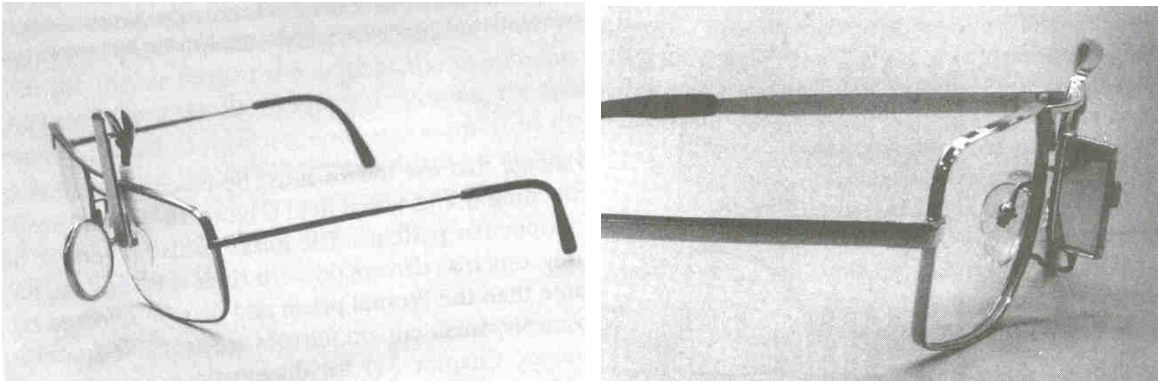

Figure 11: Viewing with a mirror to shift an image

VISUAL SKILLS TRAINING

Various visual skills training including fixation techniques, tracking techniques and scanning techniques can be taught to low vision patients with either glaucoma or RP to compensate for the loss of visual field.

- Training peripheral vision to notice details
- Scanning and tracking techniques
- Co-ordination of visual and tactual skills
- Develop scanning and searching techniques.

LIGHTING

Appropriate illumination is required for patients with RP and should be adjusted according to the tasks they wish to perform. While bright lighting is required, care should be taken not to introduce too much glare. An angle poise lamp (Fig 12) fitted with a dimmer (rheostat) switch to control the amount of lighting offers a good and portable solution to tasks requiring good illumination. The patient can adjust both the angle of the light and amount of light to maximises task illumination and reduce glare.

Figure 12: Angle-poise light and stand

Impaired scotopic vision means restriction of mobility in the dark. A widebeam handheld (Wide Angle Mobility Light, WAML) or head mounted torch can be used to illuminate the way when walking in dark areas or going out at night. Small torches are also handy when searching for objects in cupboards or locating key holes.

The ITT Pocketscope is an infrared night vision device designed for use by RP patients for mobility purposes. This device is very expensive and has a small field of view. In a study comparing the use of the Pocketscope and the WAML, patients with RP showed a preference for the WAML for mobility purposes.

FILTERS

Patients with either glaucoma or RP often suffer from glare and reduced contrast sensitivity. Filters can be used to reduce glare and improve contrast. The CPF Medical Filter lenses developed by Corning Glass can be used to reduce glare and improve contrast in patients with glaucoma. In addition, using tints can also help to reduce adaptation time between moving from light to dark conditions and vice versa. These tints are photochromic and cut out blue light and absorb ultraviolet radiation. CPF 511 which is a medium range filter providing moderate blue light filtering may be useful.

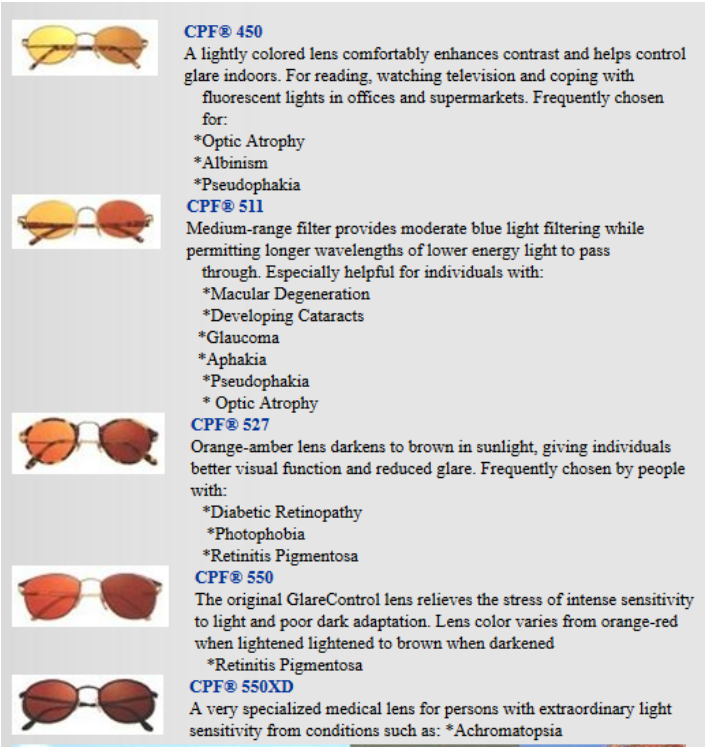

Figure 13: CPF Medical filter lenses to reduce glare
http://www.michiganoptical.com/CPF-Medical-Filter-Lenses.html

GLARE PROTECTION

Wearing a cap or sunshade is a practical and affordable option for glare protection (Fig 14).

Figure 14: Use of a cap to reduce glare

OTHER SERVICES

Patients with RP have complex visual needs and should be referred to other professionals for ancillary care. This should include referral to:

- Orientation and mobility instructors
- Counselling
- Occupational therapist – activities of daily living advice
- Audiometry – due to the possibility of Usher's Syndrome.

REHABILITATION - STEPS TOWARDS INDEPENDENT LIVING

Some basic factors that should be considered and implemented to promote independent living for people with low vision include:

1. **Self awareness**

   This refers to person's feelings about their vision e.g. whether they consider themselves as being blind, having some residual but not normal vision or as having normal vision. It is helpful to explain to the client the difference between normal vision, low vision and blindness, if the residual vision is usable for everyday activities. It is also important that the client's family and friends be made aware of the results of any vision assessment so that they can understand the challenges faced by the client and what assistance may be required and what considerations need to be made.

   Suggestions to the client include:

   - Implications of restricted visual fields, effects of poor contrast or a problem with colour vision, if necessary
   - Enhancement of his/her visual functioning

     For example – Working in the best light; moving closer to objects to see them better; using objects with good contrast; allowing plenty of time for fixation

2. **Self confidence**

   Refers to whether the available vision is used to find out about the environment and what activities are taking place, or if the person waits to be told. A loss of vision can lead to a loss of confidence in using remaining vision for obtaining information. It is important to use positive messages during the rehabilitation process to encourage the client to use their remaining vision effectively to manage their activities of daily living.

3. **Environmental awareness**

   Clients should be encouraged to look closely at objects in the environment and be provided with an audio description of the objects and activity taking place around them. This will help them to paint a picture of the environment they are in.

4. **Lighting**

   Clients with either glaucoma or RP can face problems when there is either too much or conversely, too little light. This can be from either natural or artificial light both inside and outside. The direction and amount of light are important points to consider for best visual function. Clients should be encouraged to use good lighting or sit near a window when working indoors, however, they should avoid facing the window directly to reduce glare.

   When outdoors, clients should be encouraged to move to different positions to adjust the amount of light from direct sunlight to shade, or from a shady to bright position as required. Hats, parasols or sun shades can provide relief from overhead glare from the sun.

5. **Contrast**

   Clients with either glaucoma or RP often have reduced contrast sensitivity. Contrast can be increased simply by using a dark background for light objects and vice versa, e.g. white rice on a black plate, pouring black coffee into a white cup.

   Clients should be provided with advice on this simple solution.

6. **Colour vision**

   Colour can also be used to increase visibility of objects where vision is poor, e.g. using navy blue buttons on a white shirt. Again this is an inexpensive intervention and clients should be encouraged to trial what colour contrasts work best for them.

CONCLUSION

The success of rehabilitation of low vision patients with glaucoma and retinitis pigmentosa can be limited by the level of poor vision, as both diseases can affect both contrast sensitivity and visual fields.

There are many forms of RP and associated conditions which can affect patient outcomes. Issues to be considered include mental disabilities and other disabilities presenting as part of syndromes with RP as a manifestation. Glaucoma patients can also benefit from orientation and mobility assistance, depending on the advancement of their field loss. To summarises, clients with RP and glaucoma will benefit from multidisciplinary interventions - both diseases can be managed with a team approach.

The key to success is conducting a good clinical assessment and understanding the functional implications of the vision loss. It is important to communicate the limitations to the patients as well. As the nature of the vision loss and needs of patients are variable, individualised programs must be developed for people with low vision due to glaucoma and RP. These steps and a high level of motivation from the clients will increase the rate of success in rehabilitation.

Key considerations include:

- Limited success rates
- High level of client motivation is required
- Good clinical assessment is necessary
- Understanding functional implication is critical
- Individualized instructional programs must be developed

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