LOW VISION

Background

VISION impairment is prevalent. It is estimated that 285 million people are vision impaired globally, with 39 million blind. In Australia, more than 575,000 people over the age of 40 have vision loss, which represents more than 5% of the population in that age group. In 2010, it was estimated that the total economic cost of vision loss in Australia was $16.6 billion. While many causes of low vision can be prevented or treated, there remains a significant proportion that cannot. For these people, there are strategies and services that can provide assistance, and it is therefore important that access to these services is encouraged and facilitated by health practitioners. The term ‘legal blindness’ in Australia specifically refers to people who have best-corrected distance visual acuity (with spectacles or other refractive correction if required) of less than 6/60 in their better eye or a visual field constricted to 10 degrees of arc from fixation or less. People who are legally blind are unable to drive a car and may be eligible for assistance.
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for government benefits, such as a disability support pension or subsidised travel concessions. Low vision is often defined as visual acuity worse than 6/18 (20/70), but it may be thought of more generally as any vision impairment that causes a loss of quality of life and/or difficulties with activities of daily living. The WHO International Classification of Diseases (ICD-10) definitions of visual function are given in table 1. Owing to the variation in the levels of impairment that can exist, as well as individual adjustment strategies and personal goals, the management of low vision will vary from case to case. For example, patients with central field loss (such as in macular degeneration) will primarily have difficulty with reading and face recognition but will likely retain good mobility in the environment. They might benefit from optical or electronic magnification of print or eccentric fixation training (where the patient is trained to use their peripheral vision effectively). Conversely, a patient with glaucoma or retinitis pigmentosa will experience loss of peripheral vision, making mobility a challenge, whereas the ability to read may remain intact until late in the disease process. For this group, primary mobility aids — such as a long cane or a guide dog — might be useful.

People with vision loss are at an increased risk of falls, are more likely to be admitted to aged-care facilities and have a decreased life expectancy. In addition, vision impairment is a significant risk factor for depression and a decrease in quality of life. These medical comorbidities are important to consider and necessitate multidisciplinary care. The aim of this article is to provide an overview of the common causes of vision loss, the preliminary investigations and the management strategies available.

**Table 1: The WHO ICD-10 definitions of visual function, with corresponding visual acuity ranges in Australian (6/6) and US (20/20) scales.**

<table>
<thead>
<tr>
<th>Category</th>
<th>Presenting distance visual acuity</th>
<th>Note: These are best-corrected measures taken with optimal refractive correction</th>
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<tbody>
<tr>
<td>1: Mild or no visual impairment*</td>
<td>Worse than</td>
<td>6/18 20/70</td>
</tr>
<tr>
<td>2: Moderate visual impairment</td>
<td>6/18 20/70</td>
<td></td>
</tr>
<tr>
<td>3: Severe visual impairment**</td>
<td>6/20 20/200</td>
<td></td>
</tr>
<tr>
<td>4: Blindness</td>
<td>3/60 20/400</td>
<td></td>
</tr>
<tr>
<td>5: Blindness</td>
<td>1/60, or counts fingers (CF) at 1 metre 20/1200</td>
<td></td>
</tr>
<tr>
<td>6: Total blindness</td>
<td>No light perception Light perception</td>
<td></td>
</tr>
</tbody>
</table>

* The Australian standard for an unconditional private driving licence is visual acuity of 6/12 or better in the better-seeing eye, with a visual field of at least 110 degrees in the horizontal meridian (within 10 degrees above and below the horizontal midline). Patients with significant central field loss (oculoma) in the central 20 degrees of vision or diplopia are ineligible for an unconditional private driving licence. A conditional private driving licence might be granted under some circumstances. The requirements for driving heavy vehicles are more stringent. The criteria for conditional licences and heavy vehicles can be found in the Austrads Assessing Fitness to Drive for Commercial and Private Drivers guidelines.

** Eligibility criterion for Australian government support programs (disability support pensions, etc).

As the proportion of people over the age of 65 is expected to almost double in the next 40 years, medical practitioners will consult with an increasing number of people who require low-vision management.

**Figure 1: The pinhole test is a quick and effective way to differentiate between refractive and non-refractive causes of vision loss.**

Illustration of cataract surgery.

Epidemiology and aetiology

ONE of the challenges for low-vision service provision is the limited knowledge of how many individuals in Australia have vision impairment. Estimates are based on data that are almost 20 years old: the Melbourne Visual Impairment Project and the Blue Mountains Eye Study. Extrapolating the data from these studies, about 619,700 Australians have low vision, including 68,800 people who are blind. However, over the past decade, significant new treatment modalities are likely to have positively impacted on the number of people with severe vision impairment and blindness. A new study, the Australian National Eye Health Survey, has recently been launched to obtain up-to-date data and is expected to be completed by the end of 2016 (available on the Vision 2020 Australia website).

The most common causes of non-refractive low vision in Australians ages 40-49 are cataract (37%), age-related macular degeneration (AMD) (26%); glaucoma (8%); diabetic retinopathy (4%); and neuro-ophthalmic disorders, such as hemianopia post-stroke (5%). These eye conditions are age-related. Thus, vision impairment is more common among older people. As the proportion of people over the age of 65 is expected to almost double in the next 40 years, medical practitioners will consult with an increasing number of people who require low-vision management. While it is outside the scope of this article to elaborate on the pathophysiology of ocular disease in our population, it is relevant to remember that the type of vision loss (central vs peripheral) will vary among individuals, and this will affect the type of vision rehabilitation best suited to their needs.

The prevalence of severe vision impairment will diminish with time, as improved and additional treatment modalities for common eye diseases become available. For example, the use of intravitreal injections of the anti-vascular endothelial growth-factor (anti-VEGF) drugs ranibizumab (Lucentis) and aflibercept (Eylea) for neovascular AMD has improved outcomes for patients, decreasing the incidence of legal blindness by up to 50% over the past decade. However, not everyone responds equally to these treatments, and the presenting visual acuity has a major impact on the final visual outcome (ie, if the neovascularisation is detected too late, the outcomes are poorer). There are also a proportion of patients who will continue to lose vision despite timely treatment. As such, it is estimated that over half of patients with neovascular AMD may still end up with mild to moderate vision impairment (with visual acuity worse than the driving standard of 6/12).17

Initial investigations and diagnosis

THE initial goal when assessing a patient with low vision is to determine the cause of the vision loss, if not already known. The management will vary significantly depending on the cause of vision loss and the prognosis. For example, a patient who presents with moderate cataracts has an excellent prognosis for full vision recovery following cataract extraction, whereas a patient with a genetic retinal degenerative disease (such as retinitis pigmentosa) will likely have a poorer prognosis because there are no current treatment options. Referral to an ophthalmologist is required to diagnose and treat any active disease.

A useful test that can be used in a general medical setting is the pinhole test, which should be completed if the patient is unable to read letters smaller than the 6/12 acuity line while wearing their distance glasses. In this test, a pinhole occluder is held in front of the eye being examined, with the other eye occluded (see figure 1). The patient is then asked to read the letter acuity chart again. If the number of letters improves significantly (with the patient able to read at least an extra line), then it is likely the current visual status is due to refractive error. A referral can be made to an optometrist for refractive investigation and possible improvement in visual acuity. If, on the other hand, vision does not improve with the pinhole, then it is more likely the vision loss is due to pathology, and referral should be made to an optometrist or ophthalmologist for further assessment. Low-vision management is often required throughout the entire process of disease diagnosis and treatment, not just at the stage where the practice setting is of less importance.
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Management of low vision

It has been shown that low-vision rehabilitation can be effective at assisting patients to maximise their residual visual capabilities, although a systematic review in 2012 concluded that further study is required to provide high-quality evidence on outcomes.16 Unfortunately, low vision is often under-managed in Australia, with fewer than 15% of eligible people receiving low-vision rehabilitation services.4 This is despite many internationally recognised standards of care, such as the Kooyong low-vision model, being developed in Australia, and despite estimates that over 90% of people with vision impairment would benefit from such interventions.17 Part of this discrepancy between service availability and service provision is the lack of understanding of referral pathways and rehabilitation options for people with low vision.18,19

Low-vision management requires multidisciplinary professional services, which can be considered as follows:

• Initial disease diagnosis and medical or surgical management by an ophthalmologist.
• Disease monitoring and prescription of refractive correction (spectacles and contact lenses) and low-vision aids by an optometrist and orthoptist.
• Mobility training (including the use of primary mobility aids, such as a long cane or a guide dog) by an orientation and mobility instructor.
• Training in adaptive technology, strategies for improving activities of daily living and home adaptations (handrails on stairways, shower chairs, high-contrast strips on stairs) by an occupational therapist.
• Counselling and support services by a psychologist, psychiatrist or social worker.
• Management of coexisting medical conditions ensuring co-ordination of multidisciplinary care and issues relating to the increased chance of falls by a GP.

Optical devices and assistive technology

The basic principles of low-vision rehabilitation are to maximise the residual function a patient has and provide alternative options for information access and mobility as required. Low-vision care aims to make visual objects bigger, brighter and bolder — using magnification, improved task lighting and increased contrast, respectively.

There are a number of near-optical magnification methods that can be used, such as a stand magnifier, a hand-held magnifier or high-powered spectacles (see figure 2). For distance, telescopes are available and are particularly useful for people who attend lectures, theatre or sporting events (see figure 3). Newer electronic magnification technologies, such as CCTV magnifiers (see figure 4), portable electronic vision-enhancement systems (see figure 5) and software on smartphones, tablets and computers can easily adjust magnification and contrast (see figure 6) and are becoming extremely popular (see box 1).

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Box 1: New technologies — the era of the smartphone

Standard low-vision aids are rapidly becoming supplemented and, in some cases, superseded by software available on mainstream electronic goods. For example, smartphones have baseline software capabilities to allow voice recognition, enlarge text, invert colour contrast settings, magnify images and convert text to speech. The in-built ‘reader’ mode on smartphones converts web pages to plain text, removing all visual noise and distractions. In addition to the baseline software, smartphones allow users to download applications that can turn the phone into a video magnifier (VisionAssist by Slinky Ware), scan barcodes on pantry items (GoScan by GS1 Australia) or convert books into audio programs (iBooks by Apple). Training on the use of electronic low-vision aids is freely available to patients through low-vision support agencies.
Mobility and activities of daily living

Referral to an orientation and mobility (O&M) instructor is essential if the patient has significant visual field loss because the instructor will be able to provide training in the use of any residual vision and supply aids for independent travel and safe mobility. O&M instructors also provide information on how people with vision loss should let others know about their vision loss and how they should be guided by a sighted person (see box 2). Improved mobility skills are an important factor in falls prevention and are especially important in people who are elderly or frail. Most O&M instructors are employed through low-vision agencies, such as Vision Australia or Guide Dogs Australia, and private instructors are also available.

In people with significant vision loss, or other contributing factors, a primary mobility aid is often required. A long cane is used to detect ground obstacles, stairs and drop-offs, with the user scanning the environment in an arc motion in front of them as they walk. A symbol cane is a shorter cane and is used as a secondary aid, mainly to signal to other people that the patient has vision impairment. This is used because vision loss is not obvious to observers, and the need for possible assistance is often not recognised. Support canes are used by people who have balance or physical restrictions and are usually white to signify that the user has a vision impairment. Ultrasonic guides, global positioning systems (GPS) and guide dogs are often provided to people who require additional mobility assistance.

‘Activities of daily living’ is a broad term that describes tasks that an individual must do to live a healthy, happy and productive life. These include personal hygiene (showering, toileting), cooking, social activities, participating in sports, and financial management. Many of these tasks are adversely impacted by vision loss, and rehabilitative training is often required. Occupational therapists are able to provide this training and, again, are often affiliated with organisations such as Guide Dogs Australia and Vision Australia.

Managing vision-related depression

A major comorbidity is depression, which affects about one-third of people with low vision and is often undetected or untreated.23,24 As such, the UK National Institute of Clinical Excellence recommends that all patients with vision loss be routinely screened for depression.25 Recognition of depressive symptoms and timely management have been shown to improve depression scores and quality of life for patients.26

Charles Bonnet syndrome

People with vision loss can sometimes experience visual hallucinations of a non-psychopathologic origin, known as Charles Bonnet syndrome (CBS).24,25 CBS is characterised by recurrent visual images that can vary in nature, ranging from basic imagery of lights or coloured patterns to complex hallucinations of objects, people or animals.26 Risk factors for CBS include bilateral vision impairment, increased age, social isolation, introversion, lower cognitive function, hearing impairment, history of stroke, and dim lighting.26,27 The prevalence of CBS is thought to vary with diagnosis; however, up to 40% of people with AMD experience CBS hallucinations.28

CBS is associated with negative emotional consequences and is often not reported to health practitioners by people who experience the condition because they are concerned that if they reveal their symptoms to family and friends, they will be thought to be ‘losing their mind’.27 Thus, it has been proposed that regular screening and monitoring of CBS should take place for any patient with vision loss to encourage timely diagnosis and appropriate management.28 There are questionnaires available to complete this screening, but patients will often respond to a broad question, such as the following: “Some people with certain types of vision loss have difficulty seeing certain things, and may even see things that are not really there. They may see things such as colourful patterns, animals, buildings, people or plants and trees. Have you ever seen anything similar to this?” If detected, the management for CBS is counselling and reassurance, with about one-third of patients no longer experiencing hallucinations after one year.29

Box 2: Hints for working with people with vision impairment

1. Identify yourself when you enter a room, including introducing anyone who is with you.
2. Ask if the person requires any assistance to move through the environment (ie, ‘I am Dr Smith, and we are going to go into my consultation room now. It is down a hallway with some chairs and obstacles — can I give you some assistance?’).
3. If the person would like to use you as a sighted guide to navigate, first touch the back of your hand against the back of the vision-impaired person’s hand. This allows the person to gauge your position and find your arm.
4. The patient will then slide their hand just above the elbow of the guide and grip so that their fingers are on the inside of the guide’s arm.
5. The guide should keep their arms beside their body in a relaxed, vertical position.
6. The guide will walk slightly ahead of the patient at all times.
7. Walk smoothly and warn the patient of any upcoming obstacles, including stairs or doors that open towards them.
8. Keep your consulting room organised and free of obstacles. If there are hazardous items in the path of the patient, it is usually best to offer to guide them past the area.

Figure 4: A CCTV magnifier.

Figure 5: Portable electronic vision-enhancement systems.
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Specialist referrals and support

THE majority of low-vision services in Australia are provided by non-for-profit organisations, such as Vision Australia, Guide Dogs Australia, VisAbility and the Royal Society for the Blind. These organisations provide a wide range of low-vision support services, including the following:

- Mobility training with orientation and mobility instructors — which may include the use of primary mobility aids, such as a long cane or a guide dog.
- Training in the use of electronic mobility aids, such as GPS and auditory equipment.
- Special education programs, youth camps and school holiday programs.
- Counselling.
- Adaptive technology (ie, large-print clocks, text-to-speech software for computers).

In addition to these organisations, a number of low-vision support agencies have been formed in Australia, which can be for general vision loss (ie, Blind Citizens Association of Australia) or disease-specific (ie, Retina Australia, Macular Disease Foundation, Glaucoma Australia, Diabetes Australia, Keratoconus Australia). These agencies also play a significant role in advocacy to government and policymakers. A list of the major low-vision groups can be found on the Vision 2020 Australia website.

Vision restoration therapies

DESpite significant advances in the treatment of ocular disease, there will always be patients who experience irreversible vision loss. As such, there are currently a number of research projects around the globe that aim to restore vision — an idea that was previously thought to be science fiction but is now showing true promise in early clinical trials. There are four main areas of research in the field of vision restoration: stem cells, gene therapy, optogenetics and vision prostheses (‘bionic eyes’).

Stem cell therapy aims to restore cells that no longer function within the eye, such as the implantation of replacement retinal pigment epithelial cells. The advent of techniques to produce induced pluripotent stem cells has vastly improved the potential for this therapy, and recent clinical trials have shown the safety and possibly efficacy of stem cells in the treatment of severe AMD.

Gene therapy is targeted at monogenic disorders that result in severe vision loss, such as retinitis pigmentosa, and has been shown to be effective in clinical trials of Leber’s congenital amaurosis.

Optogenetics is currently in the early stages of development but is an exciting treatment modality that aims to use gene therapy to confer light-sensitive properties on residual cells in a damaged retina (when the photoreceptors have been lost).

Electronically generated visual prostheses, or bionic eyes, aim to restore visual function by stimulating the residual visual pathway in patients. The device consists of a camera to capture images, an external processing unit and implanted electrodes, which can be in the eye itself or further back in the pathway in the optic nerve or visual cortex. Bionic eye technology is the most commercially advanced of the vision restoration treatments, with an FDA-approved device available for sale in the US and Europe (the Argus II implant from Second Sight Medical Products, California) and a number of other devices currently being developed. The world’s first suprachoroidal implant, being developed by Bionic Vision Australia, recently completed a proof of concept study, showing safety and basic efficacy.

A comprehensive review issue on vision restoration treatments was recently published in Translational Vision Science & Technology.

Case study

MARY, a 78-year-old woman, presents with a sudden loss of vision in her right eye. Apart from routine bilateral cataract surgery five years ago, she does not have any relevant ocular history. She reports that her mother lost vision in her 70s from a bleed but is unsure of the actual diagnosis.

The ophthalmologist comments on her visual acuity of 6/60 in the left eye and an R 6/60 L 6/38, with no improvement when diverse aids are employed.

From this conversation, it is determined that Mary would benefit from low-vision aids, mobility training and depression management. She is referred to a psychologist, who begins working with her on the emotional distress she is experiencing from the vision loss. A referral is also made to a multidisciplinary low-vision organisation, which arranges a home assessment with an occupational therapist. Subsequent modifications include a walking clock, large-print labels for cooking supplies and a large-button telephone (see figure 7). Contrast edging is applied to her stairs to ensure easier detection, and a rail is installed at her front steps to reduce the risk of falls. Mary is offered a cane to use when walking outside to provide further stability, but she elects not to use one at this time.

A low-vision aid assessment is carried out by an optometrist, who determines that Mary would benefit from near-task lighting, achieved with a floor lamp placed next to her reading chair. Mary couldn’t page 28

Special education programs, youth camps and school holiday programs.
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begin to read her mail and the local church newsletter again, making her feel more connected and improving her social participation. The optometrist also recommends some specialised embroidery kits for low vision, which use larger holes and high-contrast threads, and this is also well-received. Low-vision care is a long-term process, and so Mary is regularly reviewed to monitor her progress and to educate her on new progress in low-vision aid technology.

### How to Treat Quiz

**Low Vision — 24 April 2015**

<table>
<thead>
<tr>
<th>Quiz Question</th>
<th>Correct Answers</th>
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</thead>
<tbody>
<tr>
<td>1. Which THREE statements regarding the background of low vision are correct?</td>
<td>a) More than 5% of the Australian population aged over 40 has vision loss. b) The term ‘legal blindness’ in Australia specifically refers to people who have best-corrected distance visual acuity (with spectacles or other refractive correction if required) of less than 6/60 in their better eye. c) The treatment of all patients presenting with low vision is standard. d) Low vision is any visual impairment that causes a loss of quality of life and/or difficulties with activities of daily living.</td>
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<tr>
<td>2. Which THREE statements regarding the epidemiology of low vision are correct?</td>
<td>a) Knowledge is limited regarding how many individuals in Australia have vision impairment. b) The most common cause of non-refractive low vision in Australians aged over 40 is diabetic retinopathy. c) Vision impairment is more common among older people. d) The prevalence of severe vision impairment will diminish with time, as improved and additional treatment modalities for common eye diseases become available.</td>
</tr>
<tr>
<td>3. Which THREE statements regarding the investigation of low vision are correct?</td>
<td>a) The pinhole test is a quick and effective way to differentiate between refractive and non-refractive causes of vision loss. b) If vision improves significantly on the pinhole test, then it is likely the current visual status is due to refractive error. c) If vision does not improve on the pinhole test, then it is likely the current visual status is due to refractive error.</td>
</tr>
<tr>
<td>4. Which THREE statements regarding the management of low vision are correct?</td>
<td>a) Estimates indicate 10% of people with vision impairment could be helped using the Kooyong low-vision model. b) Fewer than 15% of eligible people in Australia are receiving low-vision rehabilitation services. c) Part of this discrepancy between service availability and service provision is the lack of understanding of referral pathways and rehabilitation options.</td>
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<tr>
<td>5. Which TWO statements regarding optical devices and assistive technology are correct?</td>
<td>a) The basic principles of low-vision rehabilitation are to maximise residual function and provide alternative options for information access and mobility. b) Near-optical magnification methods include telescopes for sporting events.</td>
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<tr>
<td>6. Which TWO statements regarding mobility and activities of daily living are correct?</td>
<td>a) An orientation and mobility instructor (O&amp;M) will be able to provide training in the use of any residual vision and supply aids for independent travel and safe mobility. b) Improved mobility skills are not essential in patients with low vision but may appeal to some.</td>
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<tr>
<td>7. Which TWO statements regarding vision-related depression are correct?</td>
<td>a) Vision-related depression is often undetected and untreated. b) Vision-related depression occurs in fewer than 10% of those with low vision.</td>
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<tr>
<td>8. Which THREE statements regarding Charles Bonnet syndrome (CBS) are correct?</td>
<td>a) CBS describes visual hallucinations of a psychopathologic origin occurring in people with low vision. b) CBS may be able to provide training in the use of any residual vision and supply aids for independent travel and safe mobility. c) Recognition of depressive symptoms and timely management have been shown to improve depression scores and quality of life for patients.</td>
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### Conclusion

LOW vision is a complex condition that can be the result of various disease pathologies or injury. Two patients will be alike, and management strategies will vary significantly. However, there are key factors that should always be considered: the diagnosis, the prognosis, the patient’s understanding of their condition, depression, risk of falls and quality of life. In Australia, we have access to world-class support agencies and low-vision resources, but these are significantly under-utilised. Appropriate and timely referral of people with low vision to such services will maximise their rehabilitation and provide improved outcomes for those with vision loss.

**INSTRUCTIONS**

Complete this quiz online and fill in the GP evaluation form to earn 2 CPD or PDP points. We no longer accept quizzes by post or fax.

The mark required to obtain points is 80%. Please note that some questions have more than one correct answer.

**GO ONLINE TO COMPLETE THE QUIZ**