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Advances in Low Vision Rehabilitation

By Samuel N. Markowitz, MD, FRCSC

Clinically, low vision (LV) is defined as an untreatable impairment that limits pertinent activities of daily living (ADL), and is one of the 10 most prevalent causes of disability.[1] Given current demographic changes, most cases of LV are caused by age-related eye diseases,[2] particularly, age-related macular degeneration (AMD). AMD is the leading cause of blindness in Canada and the United States (US) for people >65 years old and the second leading cause for those between the ages of 45 and 65 years.[3]

Despite past and recent advances in treatment for ocular diseases, many remain incurable and result in LV. Continuation of care mandates vision rehabilitation intervention as the only remaining option for such patients. LV rehabilitation (LVR) is a relatively new subspecialty in eye care that can assist LV patients with technology and techniques designed to enhance residual abilities required to perform vision-dependent tasks in a useful manner. This issue of *Ophthalmology Rounds* reviews recent advances in LVR. It summarizes the components of LVR and their applications, and discusses the assessment of residual functional vision such as reading ability and others. Finally, this article reviews how LVR interventions can be planned and implemented based on results from assessments. Interventions may include prescription and training in the use of LV devices (magnifiers, telescopes, selective transmission lenses, electronic devices, and computers), training and implementation of programs for rehabilitation of skills (reading, writing, driving, orientation, mobility, and other ADL), as well as counselling and social support.[4]

LVR is a multidisciplinary subspecialty and the best results are achieved with an interdisciplinary team approach. This concept is supported by the American Academy of Ophthalmology, the American Optometric Association, and the American Occupational Therapy Association, the US Association for Education and Rehabilitation of the Blind and Visually Impaired, and the International MD Support Group.[5] Accordingly, an ophthalmologist or optometrist provides the LVR assessment and identifies priority tasks for LVR. They also prescribe devices and devise treatment plans for LVR training sessions in specific skills identified during the assessment process. An occupational therapist (or certified LV therapist) performs additional functional and home evaluations. Subsequently, he/she provides rehabilitative training in the clinic or the patient's home according to the rehabilitation plan.

Programs

Recognition of and agreement with LVR by ophthalmologists and the public at large are critical for efforts to advance LVR as an integral part of the profession. Current evidence indicates an overwhelming positive response from colleagues and the public.

Recently, the introduction by the Ontario Health Insurance Plan[6] for reimbursement of LVR services provided by physicians, allows first-time comprehensive coverage. These provisions include 2 initial LV assessments within a 5-year period, as well as 10 follow-up assessments following the initial evaluation. Follow-up assessments can

Department of Ophthalmology and Vision Sciences, Faculty of Medicine, University of Toronto,
60 Murray St.
Suite 1-003
Toronto, ON M5G 1X5

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also be used for vision therapy and skill-training sessions. In addition, the assistive-device program financed by the Ontario Ministry of Health provides large subsidies to Ontario residents for the purchase of LV devices.

In a recent additional development, a Memorandum of Understanding among the Department of Ophthalmology at the University of Toronto and the Canadian National Institute for the Blind (CNIB) envisages close cooperation between staff, residents, and fellows in the practice of LVR. A 2-year pilot project currently in process is aimed at developing a multispecialty delivery-of-care template for emulation across the province. The LVR protocol development is centred on a satellite LVR clinic established on CNIB premises and the actual protocol is concentrating on LVR training for tasks such as reading and writing, among others.

Public concern is even greater when it involves the welfare of children; for example, a bill was introduced and passed in the US Senate this year, awarding funds to states for LVR services for children.[7] Accordingly, the funds are to be used to:

- provide comprehensive eye evaluation and LV assessment in children with LV aged 3-19 years, by an ophthalmologist or an optometrist
- access free LV devices with instruction and LVR training
- assure specific assistive technologies for these children
- offer instruction and consultation to parents and teachers related to the functional use of vision and the use of optical devices
- support orientation and mobility recommendations.

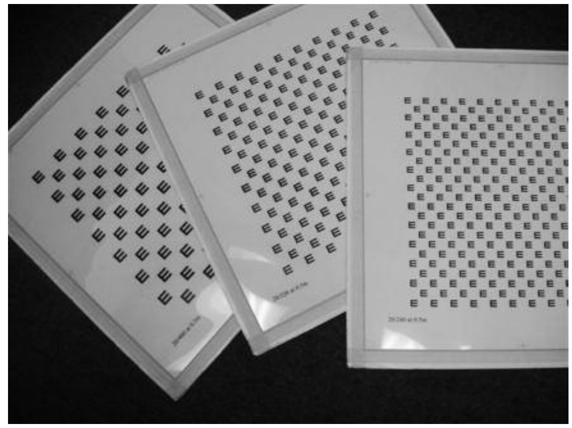
The widespread interest in LVR among patients, professionals, and the public is reflected in advances in all aspects of LVR, as detailed below.

Assessment

Current research advances support efforts to obtain accurate estimates of residual visual functions and functional vision, with developments in most aspects of the assessment process.

Residual visual function assessments begin with determining visual acuity (VA); in the presence of LV, the assessment of residual VA is a complex and demanding task essential for all successful LVR outcomes. Valuable information on all aspects of VA must be gathered systematically by using appropriate testing methods.[4] The Early Treatment for Diabetic Retinopathy Study (ETDRS) chart is the current accepted standard in clinical LV practice for assessment of residual VA levels. Recently, a new test was developed to detect residual VA more accurately, by estimating the potential visual acuity (PVA) under optimal viewing conditions, which goes beyond the ETDRS chart.[8] The PVA test measures resolution acuity after removing the limitations responsible for optical attenuation, such as defocus, diffraction, and poor oculomotor and cognitive skills. Best estimates of resolution acuity are the equivalent of PVA, and PVA charts are used routinely for VA assessment in the LV clinic to set targets for vision rehabilitation (Figure 1).

Figure 1: Potential visual acuity charts



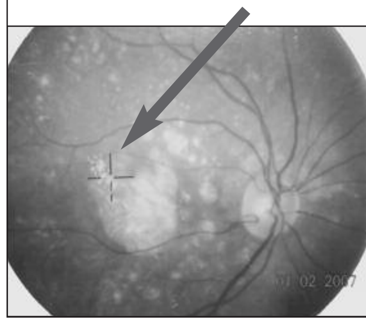
LVR approaches governing testing fields of vision are equally divided between the need to gain information about the size and characteristics of scotomata that reduce central vision, and the need to assess residual peripheral fields amenable to LVR. Perimetry methods produce scotoma records that may offer clues to scotoma displacement, a functional adaptation found in most LV patients. Automated and computerized perimetry, as well as tangent screen testing in those patients with acceptable fixation maintenance, provide valuable information on the location of scotoma displacement. Scotoma displacement indirectly points out the eccentric fixation locus used to maintain fixation during testing.[9,10] Microperimetry offers additional insight into the complex relationship between fixation location and scotomata. The "ring scotomata" are of great interest because a ring scotoma borders a fixation locus from at least 3 sides.[11,12] Ring scotomata present severe challenges to any LVR attempt. Usually the domain of laboratory scanning laser ophthalmoscopy, ring scotomata can now be identified in clinical practice with the Nidek microperimeter (MP)-1 instrument that combines perimetry and fundus imaging.[13]

Testing and assessing residual oculomotor efficiency is becoming more important as the understanding of LVR broadens; this assessment is performed in an increasing number of cases. A variety of tests are available to determine oculomotor control in LV patients. Global indices produced by automated perimetry offer a reasonable and accessible method for fixation-stability assessment. The MP-1 instrument automatically produces fixation-stability estimates for 2 circular fixation areas with arbitrary diameters set at 2° and 4°. Estimates produced in such cases are in line with estimates based on bivariate contour ellipse area calculations, and the estimates from these 2 different degree areas can assist in the assessment of outcomes from vision rehabilitation interventions.

Identification of presumed preferred retinal loci (PRL) is the core component of any modern LVR assessment. PRL identification offers the option to use best residual visual functions for rehabilitation interventions (Figure 2). Scotoma mapping as an indicator for PRL identification in LV patients is widespread and can be implemented with various field-testing methods.[10] When the Nidek MP-1 was introduced commercially, it became a common modality for direct PRL assessment. This instrument records fixation location during fixation attempts on a red cross projected in the middle of the viewing area. The instrument also records eye movements during fixation with an auto eye-tracking system that registers eye positions relative to an anatomical landmark (ie, a retinal blood vessel) and compensates for stimulus projection location. During this procedure, the auto-tracking system calculates horizontal and vertical eye movements relative to a reference frame at a rate of 25 Hz. The Nidek MP-1 instrument offers an accurate method for identification of the eccentric location of PRL in accordance with results from previous scanning laser ophthalmoscopy.[13]

Figure 2: **Preferred Retinal Locus (PRL)**

The PRL is the new "fovea"



- Best Visual acuity
- Best fixation stability
- Develops naturally
- Can be planed for another location
- Can be trained for better function
- Multiple-task related

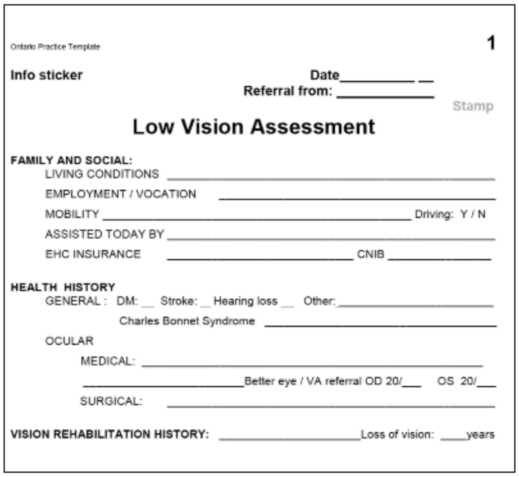
Functional vision assessment complements information collected from residual visual function determinations and is an integral part of any LVR assessment. Methods used for assessment of functional vision either document self-reported global visual abilities in specific domains of ADL or document actual performance with a certain task by measuring efficiency of performance, such as speed while reading.

Many assessment instruments are available, designed to assess domain-related visual abilities, and all are similar to the well-publicized 25-item National Eye Institute Visual Functioning Questionnaire (NEI VFQ-25).[14] These questionnaires are based on scores with rating scales and, as such, can underestimate rehabilitation effects if nonresponsive items are included in the string of questions presented to the patient. A new type of questionnaire was recently developed that provides more accurate estimates of visual abilities.[15] The Veteran Affairs Low Vision Visual Functioning Questionnaire (VA LV VFQ-48) was developed to serve as a patient evaluation tool and an outcome measure for vision rehabilitation. The VA LV VFQ-48 provides visual ability estimates in 4 domains (reading, mobility, visual information processing, and visual motor skills) and an overall visual ability score, all validated by Rasch analysis. The questionnaire offers a unique, easy to administer, and validated modality for obtaining functional visual ability estimates with all the required statistical calculations embedded in a standard spreadsheet questionnaire format; it is in the public domain and freely available to all.

Other methods used for assessment of functional vision document actual performance of a certain task and measure efficiency of performance, such as speed while reading. Evaluation of continuous-text print materials provides a more accurate measure of reading ability than single optotype reading. The Minnesota Low Vision Reading (MNRead) test acuity charts measure reading speed at different print sizes and determine the print size that supports the patient's maximal reading speed.[16] The recently introduced Colenbrander Mixed Contrast chart[17] combines testing of reading skills at high and low contrast. Scores from this chart for high contrast correlate well with ETDRS acuity and MNRead tests. The scores obtained with the Mixed Contrast charts for low contrast correlate well with the Pelli-Robson contrast tests. The mixed-contrast reading card provides an effective means to introduce contrast testing into general practice without the time-consuming use of separate tests for high and low contrast.

The new Ontario Practice Template for LVR[18] summarizes all assessment suggestions highlighted above and offers a protocol for possible therapeutic interventions, all in accordance with Ontario practice realities (Figure 3). The document is designed in a modular format with a Microsoft Word document preparation base. This allows the introduction of modifications to reflect a particular practice either in assessment methods or interventions; as well, it easily serves as a base, allowing possible changes to accommodate practice realities in other Canadian provinces or any other jurisdiction.

Figure 3: The Ontario Practice Template



Ontario Practice Template 1

Info sticker Date__________ __

Referral from: ____________

Stamp

Low Vision Assessment

FAMILY AND SOCIAL:

LIVING CONDITIONS ____

EMPLOYMENT / VOCATION ____________________________________

MOBILITY _______________________________________ Driving: Y / N

ASSISTED TODAY BY ____

EHC INSURANCE ________________________ CNIB _____________

HEALTH HISTORY

GENERAL : DM: __ Stroke: __ Hearing loss __ Other: ____________________

Charles Bonnet Syndrome ____________________________

OCULAR

MEDICAL: ____

____________________Better eye / VA referral OD 20/___ OS 20/___

SURGICAL: ____

VISION REHABILITATION HISTORY: ____________________Loss of vision: ____years

Interventions

Recent research leading to a better understanding of visual functions and functional vision has supported the development of new devices and strategies for LVR. Outcomes following an assessment of residual visual functions result in detection of functions that can be improved with the use of optical devices. Functional vision outcomes result in detection of skills that can be improved with training.

A sequential approach is recommended when using the various methods available for prescribing devices to improve residual visual functions. Initially, refractive errors are corrected, taking notice of eye dominance. This is followed by the stabilization of oculomotor function with the use of prisms for image relocation and the prescription of the best lighting conditions to reduce glare and improve contrast. The final step is the prescription of adequate magnification and field restitution devices.[4]

The use of prisms for image relocation in the presence of LV secondary to macular disease emerged recently as an important discussion topic in the LV literature. A recent prospective, randomized, double-blind study[19] confirmed previous unanimous observations[20] on the benefits of using prisms in such cases. Compliance with the use of prism glasses was high and an improvement in VA, from 20/210 to 20/100, was observed. One recent study[21] observed about 40% compliance and concluded that glasses with prisms were not more efficient than glasses without prisms. Data analysis for this study severely underestimated the benefits from wearing prism glasses by not making adjustments on compliance as required by proper statistical methods.[22] The general consensus among LVR practitioners is that the issue of using prisms for image relocation is a beneficial intervention in LV cases secondary to macular disease; furthermore, a continuation of the practice may be helpful and efficient for many clinical cases, and may be validated by further research.

Prisms for image relocation were included in the design of a new intraocular lens (IOL).[23] The investigational prototype IOL incorporates a Fresnel prism into the lens that redirects images onto the peripheral retina in a similar manner as spectacle glasses. The prismatic IOL was implanted in 2 patients in South Africa; both had bilateral AMD with VA <20/200 and postoperatively it was found that the central scotoma has resolved.

In recent years, interest in creating an intraocular telescope (IOT) device to improve distance vision remains unabated. The concept of an implantable miniature IOT and the benefits from such technology are still addressed in the literature as before in a 2005 editorial from the *Canadian Journal of Ophthalmology*.[24] Serious concerns still linger when considering this option for patients; these relate to possible permanent loss of peripheral fields of vision, loss of ability to use PRL, to the creation of aniseikonia, as well as the loss of access to the posterior pole for diagnosis and treatment of conditions that may develop in the future. The concept and benefits from such technology was also addressed in a letter to the Editor by North American leaders in LVR.[25] They suggested that benefits from the insertion of an IOT should be ascertained separately from benefits derived both from removal of the cataract at the time of surgery and from LVR training pre- and postsurgery.

Recently, surgical variations of an IOT were proposed. One study[26] explored the feasibility and utility of an IOT created at the time of cataract surgery in patients with AMD. The technique involves a telescopic device created between a plus lens in spectacle glasses and minus power at the lens plane with an IOL, while using lower amounts of magnification. All results reported from this study were positive. Another recently published study[27] proposes creating an IOT at the time of cataract surgery with 2 IOLs. The plus lens of the IOT is created by an IOL in the anterior chamber and the minus lens is created by an IOL in the posterior chamber to produce lower amounts of magnification. Most results reported from this study were also positive and both studies suggest that low magnification (30%) is probably responsible for most of the positive results observed.

Brain plasticity was proposed recently as responsible for vision restitution. Researchers discovered that cortical neurons deprived of stimuli emerging from the corresponding areas of the retina begin responding to visual input from other noncorresponding spots on the retina.[28] This mechanism is believed to underlie the improvement in vision when using prisms for image relocation to the peripheral retina and when training for eccentric viewing in AMD cases. Brain plasticity also seems to be responsible for vision restitution in older children with anisometropic amblyopia. The method reported for this achievement involves macular stimulation of the amblyopic eye with telescopic magnification.[29] Improvement in VA following the training was observed, in addition to the improvements observed following preliminary traditional interventions.

Intense research efforts worldwide strive to develop vision substitution methods and devices.[30] The direction for research in developing a visual prosthesis varies between those aiming to utilize stimulation of residual retinal cells, and those who aim for the optic nerve fibres or for cortical targets. Few studies are *in vivo* and the majority are still at the laboratory level. The probability of a breakthrough demonstrating significant results is still remote. One such breakthrough study[31] indicated that a subretinal microphotodiode array device was durable and well tolerated by the feline retina 5 years postimplantation. Another study[32] revealed that shapes could be conveyed by visual-to-auditory sensory substitution activating the lateral occipital complex.

A variety of methods and protocols were advocated in the past for use in practical vision rehabilitation training related to the various domains defining LV, such as reading, writing, eccentric viewing training, driving, orientation and mobility, space and page navigation, home environment evaluation and modifications, education and counselling, and many others. They vary accordingly to the time and place when and where they were proposed.[33]

In the past, many studies scrutinized the impact of LVR protocols; however, none offered compelling evidence about positive benefits on patients. Now, however, the landmark Veterans Affairs Low Vision Intervention Trial (LOVIT),[34] published last year, offers solid support for LVR and is based on solid scientific methodology with an additional insight into the positive effect size of the interventions. The multicentre, prospective, randomized study, demonstrated significant improvements following the implementation of a 10-hour LVR-training program in a cohort of LV subjects. The interventions described in the study included correction of refractive errors, education on the eye disease, prescription of LV devices, addressing the rehabilitation goals selected by patients (reading at near and distance, spot-checking at far and intermediate distances, and glare control), as well as vision therapy aimed at teaching the use of prescribed devices together with strategies for more effective use of residual vision, all in accordance with modern standards of practice. The authors concluded that interventions provided significant improvements in all domains of vision and were characterized by a large-effect size in each case. In LVR, positive evidence-based outcomes, as in this study, are critical, not only for further developing and establishing the concept of LVR, but also by demonstrating to patients and the public the benefits of such interventions.

Current developments offer a glimpse of the future, and based on advances witnessed in LVR in the last few years, the future looks very promising. Innovations in basic and applied sciences promise to deliver devices and techniques that will further enhance LVR programs, as well as provide solutions to LV patients for whom today there is no help. In the LVR community of practitioners, all are looking to the future and hoping that some day the vision problems for these patients will be addressed and solved, and that this future is not too far away.

***Dr. Markowitz** is Associate Professor and Director, Low Vision Rehabilitation Program, Department of Ophthalmology, University of Toronto, Staff Member, Toronto Western Hospital, University Health Network, and Courtesy Staff, Trillium Health Centre, Mississauga, Ontario.*

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