Exploring the Role of Telemedicine in Low Vision Rehabilitation in Patients with Heredomacular Degeneration – A Novel Concept

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ABSTRACT

Background
Teleophthalmology is widely used for screening and diagnosis of many eye disorders. But its role in low vision rehabilitation is unexplored. The purpose of this study was to explore the feasibility of teleophthalmology for visual rehabilitation.

Methods
We prospectively enrolled 15 patients (age range 12-30 years, mean 16±3.2 years) with heredomacular degeneration with reduced central vision but healthy paracentral retina. We used visual stimulation and biofeedback with video game play. After the baseline investigations, the patients were given a compact disc with the video game and were trained to play the game. Subsequently, they did 40 hours of the visual training at home which was monitored using teleophthalmology by us at our tertiary institute. We used video chat and screen sharing software to guide the patients and monitor compliance. The primary outcome was feasibility of monitoring visual training remotely and secondary outcome was improvement in visual function.

Results
Seven patients returned for follow up after completing the training. The visual acuity improved from 0.74±0.29 to 0.66±0.32 logMAR (p=0.043). There was significant improvement in contrast sensitivity (p=0.023) and fixation stability (p=0.018). The vision related quality of life questionnaire showed improved scores. The ease of communication, patient comfort were high. The office visits were limited to two.

Conclusions
Our study showed preliminary evidence of benefit of teleophthalmology in visual rehabilitation and monitoring. The reduced visits would likely promote compliance and reduce economic burden of the rehabilitatory training.

Keywords: biofeedback, heredomacular degeneration, low vision rehabilitation, telerehabilitation, Teleophthalmology, video game training
INTRODUCTION

Teleophthalmology, by definition, is the integrated system of eye care delivery through digital medical equipment and telecommunications technology.¹ In the last 2 decades or so, there has been a rapid rise in e-health and remote health services.² The diagnosis in Ophthalmology being mainly a visual or image based diagnosis, it has lent itself very well to the concept of telemedicine. As a result, teleophthalmology has evolved as a major branch of telemedicine.² Also, the digital revolution and the tremendous advances in the visualization systems such as the fundus cameras has greatly helped in the evolution of teleophthalmology branch. The ease of capturing retinal and external eye images, portability of the instruments has made it easy to transport these visualization systems to remote areas. Thus, it has made medical care accessible to increasing number of patients.³

Teleophthalmology has till date focused on screening and monitoring of diseases. Several studies involving telemedicine based screening of diabetic retinopathy have documented increased number of patients being screened and identified with diabetic retinopathy changes, leading to decreasing visual impairment.³⁻⁷ Another area where teleophthalmology has been successful, is for screening and diagnosis of retinopathy of prematurity (ROP). Several studies have shown successful use of teleophthalmology for screening for ROP in remote neonatal intensive care units.²⁻⁸ This has led to reduction in the childhood blindness.⁸

However, the use of teleophthalmology for treatment or visual rehabilitation has not been explored. Pratibha et al⁶ and Murthy et al⁹ have hinted at the possibility of using teleophthalmology for low vision rehabilitation. Recently, Kozak et al¹⁰ have published a case report where teleophthalmology guided laser treatment has been done remotely. In a case report by Hall et al¹¹ removal of corneal foreign bodies was remotely supervised with teleophthalmology help. We propose the concept of telerehabilitation for low vision training in young patients with Stargardt disease.

The Concept:

Stargardt disease is the most common form of heredomacular degeneration (HMD) affecting young individuals in the first or second decade of life.¹² It can lead to severe visual impairment due to central visual loss. So far there is no treatment for this disease. Various optical, non-optical and/or assistive devices help the persons in recognizing faces, watch TV, see the black board, read fine prints, perform computer or mobile phone tasks.

As they lose the central vision, many of these young people use points that are located outside the central area of degeneration to fixate. This point is referred to as preferred retinal locus (PRL). The PRL may be at a considerable distance from the atrophic area resulting in eccentric fixation that is often unstable.¹³ Poor fixation stability is correlated with poor visual acuity and poor reading speed.¹⁴ One of the methods used to improve the fixation stability in such situations, is the bio-feedback training using the MP1 microperimeter (Nidek instruments Inc, Padova, Italy). It increases attention modulation and helps the brain to memorize the fixation location thereby providing an efficient PRL for visual tasks.¹⁵,¹⁶ Similar stimulation can be done with aggressive video game play. Action videogames have been shown to be effective in improving vision in adult patients with amblyopia.¹⁷,¹⁸ The hypothesis is that with visual stimulation and the biofeedback provided by the game when the target is correctly hit, induces the neurons located outside the area of degeneration to send stimuli to the cortical region in the brain. Plasticity of the brain allows this to function as new fixation. Gradually these connections are reinforced leading to a stable system, better fixation and resultant improved vision.¹⁹⁻²¹ We did a pilot study which showed possibility of vision
improvement in Stargardt disease with video game stimulation (manuscript under review). This visual rehabilitation can be monitored remotely with the help of teleophthalmology.

MATERIALS AND METHODS

We prospectively enrolled 15 patients in this study. The study conformed to the tenets of the Declaration of Helsinki and was approved by the institutional review board and ethics committee. Written informed consent was obtained from all the participants and also from their guardians in case of minors.

Patients with Stargardt disease and no other health issues were recruited for this study. The diagnosis of Stargardt disease was clinical and was confirmed by fundus photo, autofluorescence, optical coherence tomography (OCT) and full field electro-retinography (ERG). The fundus showed a well demarcated area of degeneration at the macula with beaten bronze appearance and hypoaufotluorescence. The surrounding retina was normal or had a few flecks. The optic nerve head was normal. OCT revealed thinning of the retina with atrophy of the photoreceptors at the fovea. The ERG showed normal photopic and scotopic responses in all the patients.

The patients underwent baseline tests which included logMAR visual acuity recording for distance and near, visual evoked potentials (VEP), stereopsis (assessed by Randot Stereo test, Stereo Optical Co., Inc., Chicago, Illinois), contrast sensitivity measurements (assessed by Pelli-Robson test). The retinal sensitivity and fixation were analyzed with microperimetry on scanning laser ophthalmoscopy optical coherence tomography (SLO/OCT) (Optos PLC, Washington DC).

All patients were advised to play action videogame (Call of Duty 4 Modern Warfare, developed by Infinity Ward and published by Activision) for 1 hour daily in each eye with alternate patching. The participants played the game at a distance between 33 and 40 cms from the screen. A participant with minimum visual acuity of better than or equal to 1.0 LogMAR can play comfortably at 33 cms. The video game had 3 levels of magnification and resolutions viz. video mode 720x576, 1024x768 and 1920x1080 pixels. In case of difficulty, the resolution and magnification could be increased. The playing distance was kept constant as far as possible. They were given 2 training sessions of 15 minutes at the baseline office visit. The availability of internet enabled device for video chat was confirmed from the patient and attendant. Patients were advised to download a screen sharing software and a video chat software. A day and time was fixed for video chat each week. Patients were given the timetable for the training and telecommunication. Patients were asked to maintain a log book recording the sessions of training, duration and difficulties faced. Patients were then sent home.

During the teleconsultation, a link was established between the patient who was at his residence and the authors at the teleophthalmology department of their institute using the broadband satellite connection. After greeting, the patients were asked a set of questions each time to confirm the number of sessions completed, the duration of training completed as well as any difficulties faced during the training and any improvements noted. The level of the game currently being played was noted down. A screen sharing software (V see or Skype) was turned on and patients were asked to play the game live for about 10-15 mins. The screen sharing software showed the game being played on the patient's screen and an external device with another video chat software operated by the patient's attendant showed the patient in action. Thus, it was possible to monitor the way the training was undergoing. The compliance was monitored weekly.

At the end of 40 hours, (20 hours each eye), all patients were reassessed in the office and all the tests were repeated. All patients were administered a previously validated low vision questionnaire before and at the
end of the training, which specifically tests visually impaired young adults on their level of impairment based on the difficulty in doing daily vision based tasks. (Appendix)

**Statistical Analysis**

The data were processed in Microsoft Excel 2013 (Microsoft Corp., Redmond, Washington, USA), and statistical analyses were carried out using the statistical software IBM SPSS Statistics for Windows (IBM Corp., Released 2013. Version 21.0. Armonk, New York, USA). Mean and standard deviation were calculated for continuous variables. Students’ paired t test was used for comparison and p value of < 0.05 was considered to be significant.

**RESULTS**

The study was conducted between September 2016 and September 2017. Fifteen patients...
in the age range of 13-30 years (mean 18.5±3.2 years, median 16 years) were included. There were 7 patients who were fully compliant and came back for follow up tests. These were included in the final analysis. These included 5 males and 2 females. All the patients were diagnosed clinically to have Stargardt disease. Healthy paracentral and peripheral retina was confirmed on fundus examination, autofluorescence and electroretinography. The visual acuity was tested with the corrected refractive error. For all the participants, the difference between present glasses (if any) and current refraction was within 0.50 DS (Spherical equivalent). There was no change in the visual acuity with present glasses in use and current refraction.

The pre-training mean distance visual acuity was 0.74±0.29 logMAR (range 0.20 to 1.0 logMAR). After training, it improved to 0.66±0.32 logMAR (p=0.043). There was significant improvement in contrast sensitivity (1.30±0.25 to 1.47±0.18, p=0.023) and in fixation stability measured by the log of bivariate contour ellipse area (6.6±0.54 to 5.69±0.75, p=0.018). The retinal sensitivity improved by 1.7±3.39dB (p=0.86). There was improvement in stereopsis. The 1 degree checker size pattern visual evoked potentials showed mild improvement in amplitudes as well as latencies (p=0.17, p=0.75). Similarly, the 15 degree checker size also showed improvement but was not statistically significant (p=0.34, p=0.07). Near visual acuity before the training, ranged from 0.32 to 0.6 logMAR (mean 0.44). And after the training, it improved to 0.35 logMAR (range 0.20 to 0.50 log MAR) (p= 0.60), but did not reach statistical significance. The details are given in table 1.

Subjectively, patients noted some improvement in daily tasks with improvement in questionnaire scores. Almost, all the patients reported improvement in distance vision tasks. They all reported improvement in identifying persons and improved scores on the task of walking alone along a corridor without bumping into objects or persons. All of them also reported better scores in locating dropped objects, reading bus numbers and other details such as bus destination. Cases 4 and 5 also reported improvement in reading textbook at an arm’s distance and identifying colors.

All the patients were comfortable with using the technology for communication. They could successfully connect and communicate with us regularly during the training. The rehabilitation could be successfully monitored in all the patients with telecommunication.

**DISCUSSION**

This was a feasibility study undertaken to test the possibility of using teleophthalmology for visual rehabilitation. We successfully demonstrated the application of real time teleophthalmology in visual training and monitoring. Traditionally, teleophthalmology involves acquisition of images which are stored and then transferred to a remotely placed specialist. This is followed by further interaction with the patient and formation of a referral plan for appropriate treatment. In another modification of this system, automated imaging systems with built in pattern recognition algorithms or machine learning software are used to screen large sections of population.  

Although, the main application of teleophthalmology has been in diagnosis of diseases, recently a few reports have evaluated the feasibility of using teleophthalmology for therapeutic purposes. Teleophthalmology guided retinal photocoagulation has been reported wherein images of the patients retina were transferred from the base hospital to the specialist situated in another hospital in another country, a plan for retinal photocoagulation was made and transferred to base hospital where the laser photocoagulation was performed for 10 patients with results comparable to conventional methods. Scan images, surgical plans and 3D models have been transferred previously using telemedicine in other specialties. In another report, removal of a
A corneal foreign body was remotely supervised by teleophthalmology means.\textsuperscript{11}

This is probably the first report of application of teleophthalmology for visual rehabilitation. Previously, Pratibha et al\textsuperscript{6} and Murthy et al\textsuperscript{9} have hinted at the possibility of low vision rehabilitation using teleophthalmology, mainly for patients with diabetic retinopathy. The main advantages of this telerehabilitation were reduced visits to the hospital. Only 2 visits were needed in this study whereas the number of visits would have been nearly 20 for each session of video game training. Also, the rehabilitation and training could be done in the more natural environment of patient's familiar surroundings. This would help in correctly assessing the visual needs of the patient and directly addressing them in a more customized manner. We selected patients with Stargardt disease for this study for 3 reasons. First, they have a healthy retina surrounding the area of atrophy at the macula which is amenable to stimulation and can lead to better fixation stability and visual improvement. Secondly, these young patients have more brain plasticity and would show better results. Thirdly, the young patients are more technology savvy. They would be more interested in playing a video game. They would also be more at home using newer technologies of telecommunications such as video chat or screen sharing.

In conclusion, we have explored the novel concept of visual rehabilitation using teleophthalmology. The concept of visual stimulation using video game for patients with heredomacular degeneration is also a new concept. To the best of our knowledge, this is the first study using teleophthalmology for visual rehabilitation. A pubmed search did not reveal any articles pertaining to this subject. The study demonstrates that it is feasible to use teleophthalmology for visual rehabilitation.

Appendix

The L V Prasad low vision questionnaire validated and published by Gothwal, et al.\textsuperscript{24}

REFERENCES


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Dr. Ratra is an accomplished vitreoretinal surgeon. She also practices medical retina and treats many patients with macular degenerative conditions. She has a special interest in developing newer and simpler methods for visual rehabilitation of young patients with heredomacular degeneration.