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Case Report

Disorders Encountered Low Vision

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Abstract

In the practice of eye care "LOW VISION" has a specific meaning as defined by WHO". A person with low vision is one who has impairment of visual functioning even after treatment and/or standard refractive correction, and has a visual acuity of less than 6/18 to light perception, or a visual field of less than 10 degree from the point of fixation, but who uses, or is potentially able to use, vision for planning and/or execution of a task".

Keywords: Low Vision; WHO; Vision Rehabilitation

According to this, presenting visual acuity in the better eye

- Equal to or better than 6/18 is defined as mild or no visual impairment;
- Equal to or better than 6/60 and worse than 6/18 as moderate visual impairment (category 1);
- Equal to or better than 3/60 and worse than 6/60 as severe visual impairment (category 2); and
- Worse than 3/60 as blindness.

Blindness is also separated into 3 categories: visual acuity

- Worse than 3/60 (category 3),
- Worse than 1/60 (or counting finger at 1 meter) (category 4), and
- No light perception (category 5).

Based on this classification, those with moderate and severe visual impairment (visual acuity worse than 6/18 and equal to or better than 3/60) and those with a visual field less than or equal to 20 are defined as having low vision and require rehabilitation (Fontenot., *et al*. 2018; Şahlı and İdil 2019).

Vision rehabilitation as defined by American Optometric Association is the process of treatment and education that helps individuals who are visually disabled attain maximum function, a sense of well-being, a personally satisfying level of independence, and optimum quality of life. Low vision rehabilitation cannot restore the lost sight but helps the patient with his/her remaining vision to maintain an independent life style [6]. The low vision treatment must have evolved through an understanding the needs of the patient.

Globally, at least 2.2 billion people have a near or distance vision impairment. In at least 1 billion – or almost half – of these cases, vision impairment could have been prevented or has yet to be addressed [7]. In the United States, low vision is most commonly caused by age-related macular degeneration (AMD), glaucoma, and

diabetic retinopathy (DR). Low vision can also affect paediatric patients due to a variety of genetic or acquired diseases (Şahlı and İdil 2019, Shah., *et al*. 2018). Low vision in INDIA is commonly caused by Cataract, Glaucoma, Retinitis Pigmentosa, Optic atrophy, Myopic Macular degeneration, and Cortical blindness [1].

One of the acquired diseases for causing low vision in paediatric is Optic Atrophy. Optic atrophy is a pathological term referring to optic nerve shrinkage caused by the degeneration of retinal ganglion cell (RGC) axons [2]. Optic atrophy (optic neuropathy) is a term used to refer to the end stage of a number of conditions that can cause optic nerve damage [3]. No proven treatment exists to reverse optic atrophy related blindness. Early diagnosis and prompt treatment can help in compressive and toxic neuropathies. Neuro-protective agents like gingko biloba have been tried with anecdotal success [3].

Retinitis pigmentosa is a group of heritable disorders of the retinal pigment epithelium (RPE) and photoreceptors that cause progressive visual field constriction and nyctalopia [5]. RP can exhibit autosomal dominant, autosomal recessive, X-linked recessive and indeterminate patterns of inheritance. The association of RP with congenital sensorineural hearing loss in the absence of other systemic involvement is termed Usher's syndrome (Jerome P. Schartman., *et al*. 2022). Hearing loss in patients with Usher's syndrome is almost invariably nonprogressive. It is important to reassure patients with RP who have been hearing-impaired since birth that they are unlikely to become deaf, even though their vision may deteriorate.

Stargardt's disease is one of the commonest inherited retinal disorders, with a prevalence of 1 in 10,000. It is inherited as an autosomal recessive trait. Most cases present with central visual loss and there is typically macular atrophy and with yellow white flecks at the posterior pole, which are at the level of the retinal pigment epithelium (RPE) [6]. Autofluorescence (AF) imaging and fluorescein angiography can be helpful in the confirming the diagnosis. (Lois N, Halfyard AS, 2004).

Retinoschisis is a pathological and degenerative change in the retina, in which there is a separation of the retinal layers into external (choroidal) and internal (vitreal). It occurs as a juvenile X-linked form in children. (Handbook of retinal OCT, 2014). There are clinical manifestations in the early stages of retinoschisis, symptoms occur only with the development of complications in the form of decreased visual acuity, narrowing of the visual fields. The hereditary form manifests itself at the age of 10 [8].

Expected outcome of such disorders are usually rare so early diagnosis is the best defence that can be possible done by a comprehensive and multidisciplinary team approach, which includes Optometrists, Ophthalmologists, Rehabilitation workers, Health care workers, and Parents. If the specific treatment of the cause is initiated before the development, residual vision can be preserved.

Following cases, reports of low vision examination and management at tertiary level of some disorders related low vision to use their remaining visual capacity to its fullest and be somehow independent as possible to live a better life.

Case Reports

Case 1

A 11-year-old boy presented to our low vision department of our hospital (Vittala International Institute of Ophthalmology) with the complaint of diminished of vision for 5 years in both the eyes, OD>OS, as well as he was not comfortable in his daily living task. He was accompanied by his mother during the examination. He had previous history of diagnosed with bilateral partial optic atrophy and they advised to use glasses. He had history of wearing eyeglasses for 4 years which was broken 6 months back. The spectacle he was using of OD: +0.75dsph/-2.00dcyl X 180 and OS: +0.50dsph/-2.75dcyl X 180. He was born by Full Term Normal Vaginal Delivery (FTNVD) at 29 weeks with birth weight of 2000grams. There was no history that he received any oxygen therapy, admitted in NICU, and seizures. He was a third child among his siblings-who were apparently well. There is no evidence of consanguineous parentage. He had history of squint (EXOTROPIA) and nystagmus since birth, as noticed and informed by parents.

MRI was done in 2013 showed that there were changes in parieto-occipital area as he was diagnosed with hypoxic ischemic encephalopathy. There is a query that there might also be a neurological disease since childhood as report shows intra-cranial haemorrhage. There was no history of taking any ocular or systemic medication.

On examination at present, it was found that his aided vision was count finger close to face (CFCF) in OD and Count finger at 1.5 mt in OS. And on slit lamp examination, Anterior segments were normal. Then, retinoscopy was performed which was found as OD: -0.75dsph/-2.50dcyl X 20° and OS: -0.75dsph/-2.75dcyl X 180°. Subjective refraction was performed following retinoscope, the value was OD: -0.75dsph/-2.00dcyl X 180° with vision CFCF and OS: -0.50/-2.75 X 180°with vision CF at 1.5meters.

On ocular examination, we got

- Bilateral pendular nystagmus +
- AHP (Abnormal Head Posture): Chin Depression +
- HCRT (Hirschberg Test): 15° Right Exotropia

Homide (homatropine) refraction was advised further. The wet retinoscopy value was OD: -3.00dcyl at 30° and OS: -0.25dsph/-3.25dcyl at 180°which shows that patient had myopic astigmatism.

After that fundoscopy was done where we got, Foveal Reflex was present (FR+) but there was temporal disc pallor in both the eyes were present.

Diagnosis was made as

- OU: Myopic astigmatism with partial optic atrophy
- Divergent Concomitant Strabismus

So, for the plan of care:

- Glasses was prescribed (SR)
- The desirable LVA (Low-Vision Aids) trail was explained to the parents.
- And to review for LVA trail.

Second visit

He reported to our Low vision department as he was not comfortable with regular use of glasses and wanted to trail of low vision devices.

Low-vision assessment: On trial of distance and Near vision devices

Trail was done for both distance and near after magnification calculation. As his residual vision was quiet less, his response for the distance vision with low vision devices (telescope) was unsatisfactory. So, patient was prescribed glasses for the distance. Portable digital video magnifier 5 inches was prescribed for near with which his response was good and near visual acuity improved upto N8. Portable video magnifier provides better enhancement of contrast, proper magnification, comfortable working distance and wide field of view as compared to other devices. We recommended him to avoid excessive light, bold tip marker for writing, sunglasses/hat specially for outdoor activities. We taught about the techniques for proper handling like spotting, tracing, tracking and scanning method with the use of low-vision aids.

Case 2

Another case of 13-year-old boy presented with the complaint of diminished vision in both the eyes. He was accompanied by his mother during the examination as he was deaf and dumb. In 2019, He was diagnosed with bilateral profound sensorineural hearing loss. He had previous history of trauma to right supra-orbital margin few months back. He had history of decreased vision during night time for past 2 years and it was even getting worse. He had history of wearing eyeglasses and the vision was not clear with the spectacle Rx of OD: -0.50dsph/-2.00dcyl at 10 and OS: -0.25dsph/-2.00dcyl at 170. There was no history that he received any oxygen therapy, admitted in NICU, and seizures. There was no evidence of consanguineous parentage. There was no history of taking any ocular or systemic medication.

On examination it was found that his unaided visual acuity was 1.0 logMAR and no improvement with the pin-hole. On slit lamp examination, Anterior segments were normal.

Fundoscopy was done where we got, cup-disc ratio was 0.2:1, foveal reflex was present (FR+) but there was disc hyperaemia, nasal blurring of disc margin and vitreous degeneration in both the eyes.

Diagnosis was made as

- OU: Retinitis Pigmentosa.

So, for the plan of care:

- VFT 30-2
- FFA
- ERG (Electro Retino-Gram)
- LVA trial

Second visit

Same child was reported to our department for ERG and LVA trial on second day. Where ERG shows extinguished response in both scotopic and photopic phases. So, there was significant rod and cone dystrophy.

Low-vision assessment: On trial of distance and near vision devices

Patient was prescribed, bifocal monocular telescope (6X 16) for distance vision. His response was good, and his visual acuity improved significantly upto 0.40 logMAR in both the eyes. He could walk and on searching things independently using the same device. Bifocal monocular telescope provides ultra-clear, fully coated optics, and brightness. Near Dome magnifier (4X 65) and digital video magnifier 5 inches for near visual task. He was comfortable using it for reading and near visual acuity improved upto N6. We recommend him to use table fluorescent lamp for near tasks and bigger marker for writing. We taught about the techniques for proper handling like spotting, tracing, tracking and scanning method with the use of low-vision aids.

Case 3

A case of 45-year-old woman presented to our low vision department after final diagnosis of B/L Stargardt's disease with the complaint of diminished of vision in both the eyes slowly and progressively. She was diagnosed at the age 12 years old. She had no previous history of wearing eyeglasses. She had recently diagnosed with diabetic mellitus. Her presenting unaided visual acuity was 1.0 logMAR for both the eyes and no improvement with the pin-hole. Her family tree suggests 2 siblings, she was first child to her parents where both siblings and even father was diagnosed with the same disease. She had problem with near vision tasks specially writing and reading books and newspaper of small prints. She also had difficulties in watching television, recognizing bus or auto number, and bright day light. She had mobility problem in outdoor and unfamiliar places. On examination by binocular slit lamp examination, the anterior segments were within normal limits but on fundus examination under mydriatics revealed typical features of Stargardt's disease.

So, for the plan of care:

- OCT
- FFA
- LVA trial

Figure 1: Fundus Photo a) OD, b) OS.

Figure 2: Fundus auto fluorescein a) OD, b) OS.

Low-vision assessment: On trial of distance and Near vision devices

A careful refraction was performed to rule out, if any refractive error was causing decrease in vision. There was no subjective acceptance. Then, Patient was prescribed, bifocal monocular telescope of 6X 16 for distance vision. His response was good, and his visual acuity improved significantly upto 0.10 logMAR in both the eyes. He could walk independently using the same device. For near visual task

- Dome magnifier (4X80)
- Portable digital video magnifier 5 inches.

He was comfortable using it for reading and near visual acuity improved upto N6. Dome magnifier provides better hand free magnification, unbreakable (Acrylic domes), and as well as fixed focus to the small prints. We recommend him to use table fluorescent lamp for near tasks, and sunglasses or use of hat for day-light/glare problem. We taught about the techniques for proper handling like spotting, tracing, tracking and scanning method with the use of low-vision aids.

Case 4

A 10-year-old boy presented to our low vision department of our hospital with the complaints of diminished of vision for distance since many years in both the eyes and history of nystagmus since childhood as noticed and informed by parents. He was accompanied by his father during the examination. He had previous history of diagnosed with bilateral esotropia. He had no history of wearing eyeglasses. He was born by Full Term Normal Vaginal Delivery (FTNVD) at 29 weeks with birth weight of 2300 grams. There was no history that he received any oxygen therapy, admitted in NICU, and seizures. He was a second child among his siblings among which elder brother also has similar issues. There is no evidence of consanguineous parentage.

On examination it was found that his unaided visual acuity was count finger at 3 meter in OD and no improvement with the pin-hole whereas in OS unaided visual acuity was 1.0log MAR and 0.70 log MAR improvement with the pin-hole for distance. Near unaided visual acuity was N12 at 10 cm in OD and N6 at 10 cm in OS. On slit lamp examination, Anterior segments were normal. Cycloplegic refraction was done to try with subjective correction. The wet retinoscopy was taken which was OD: +2.25dsph/-1.50dcyl X 180° and OS: +1.75dsph/-0.75dcyl X 180.

Fundoscopy was done where we got, retinoschisis with foveoschisis and there was to treatable lesions in the periphery. There was split in inner retina whereas outer retina was within the normal limit.

Diagnosis was made as

- OU: Retinoschisis with foveoschisis.

So, for the plan of care:

- OCT
- LVA trial

Figure 3: Fundus photo: OD, b) OS.

Figure 4: OCT a) OD, b) OS.

Second visit

Same child was reported to our department for LVA trial on second day.

Low-vision assessment: On trial of distance and near vision devices

Patient was prescribed, bifocal monocular telescope (10X42) for distance vision. His response was good, and his visual acuity improved significantly upto 0.30 log MAR in both the eyes. He could walk and on searching things independently using the same device. Bifocal monocular telescope provides ultra-clear, fully coated optics, and brightness. Near Dome magnifier (4X65) for near visual task. He was comfortable using it for reading and near visual acuity improved upto N6 at 30 cm. We recommend him to use table fluorescent lamp for near tasks. We taught about the techniques for proper handling like spotting, tracing, tracking and scanning method with the use of low-vision aids.

Discussion and Conclusion

Low vision is equivalent to visual impairment [6]. Low vision in a person due to many disorders should be known by every Ophthalmologist and Optometrist that the impairment can affect daily life activities. Thus, the main objective to provide best low vision aids to the patient to enhance their residual vision. Low vision devices were the most significant solution for performing visual skills (Suneel, dixit., *et al*. 2022). That might be an optical and non-optical device. The needs of the patient can be evaluated in different manner like: providing best refractive correction, simple and portable electronic magnification, determination of proper illumination for near tasks, glare management, visual field enhancement, sighting and mobility techniques, and additional care.

The common disorders that encountered in low vision, affecting the general population. A person with Optic atrophy presented to our department required more counselling. Under normal circumstances there was relatively increased in visual acuity with the magnification mostly for near. Yellow filter and sunglasses were helping him at bight illumination. Exploring non-optical devices really helped them with psychological aspects. People with Stargardt's disease start to experience difficulty with reading and fine handwork. They do not usually have a problem with bumping into objects when moving around. They may however experience difficulties in adjusting to light. Low vision services may be very helpful for patients with RP, who experience multiple visual disabilities, including reduced contrast sensitivity, poor dark adaptation, glare and poor color vision. These disabilities may lead to difficulties with walking, recognizing objects, searching in cabinets, preparing food and other activities of daily living. For dark-adaptation difficulties, a simple penlight is useful for searching in dark cabinets or finding a keyhole at night (Jerome P. Schartman., *et al*. 2022).

Low vision services improve the quality of life and mental state (Horowitz A., *et al*. 2005). Thus, the clinical trials of providing such low vision aids.

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