LIGHT UP

A Publication of
Vision Rehabilitation Centre

Newsletter 13

April 2004

World Health Organization
Collaborating Centre for
Prevention of Blindness

Meera & L.B. Deshpande Centre for
Sight Enhancement

Dr. P.R.K. Prasad Centre for
Rehabilitation of
Blind and Visually Impaired

L V Prasad Eye Institute
L V Prasad Marg, Banjara Hills,
Hyderabad, INDIA

Contents

Juvenile Macular Degeneration

by Dr Subhadra Jalali

What is juvenile macular degeneration?

There are several forms of macular degeneration that affect children, teenagers or adults that are known as early onset or juvenile macular degeneration. Many of these forms are hereditary and are more accurately called macular dystrophies, instead of degenerations. Juvenile macular degeneration is a type of central vision deterioration that occurs in young children.

Macular degeneration refers to an abnormality of that part of the eye, which is responsible for our sharpest central vision. This is the macula or the central region of the retina that enables us to read and to distinguish colors. The peripheral region of the retina enables us to distinguish light and dark and movement, but not the fine details. In macular degeneration, the light-sensing cells of the macula malfunction and may, over time, cease to work.

Symptoms:

1. Painless with gradually progressive, reduced visual acuity, usually in both eyes
2. Inability to see the centre of an object while retaining the ability to see the periphery (central scotoma)
3. Distortions in the appearance of objects (metamorphopsia)
4. Inability to match colours accurately
5. Inability to read and recognise faces at a distance
6. Slow adaptation to dark: After being exposed to sunlight, when persons with normal vision enter a dark place they need three minutes or so to adapt. However, persons with Stargardt's disease and other types of macular degeneration may take 8 - 10 minutes or even longer to adapt to the dark environment.

Types and causes of macular degeneration

There are three general types of hereditary macular degeneration. They are:

1. Early onset - birth to age seven: This type is dominantly inherited which means that both parents and their children can be affected (often called Best disease or Vitelliform macular degeneration). However, the severity of the vision loss can be different in different people and at different ages.

2. Middle onset - age five to twenty: This type is often called Stargardt's disease, juvenile macular dystrophy or fundus flavimaculatus. It is usually recessively inherited, which means that it can appear when few or no other family members have been affected. Rarely, this condition can be dominantly inherited. This is the commonest macular degeneration of the young seen in our Institute.

3. Late onset - in the thirties and forties: This type may have either a dominant or recessive mode of inheritance - often called Behr's, Sorsby's, Doyne's or honeycomb dystrophy.

What is a hereditary disease?

Each of us is the result of genes inherited from our parents. Sometimes an error occurs during replication of the genes as part of the reproductive process. Most of the errors are analogous to typos (or typographical mistakes) in printed text - a single wrong character appears in an instruction in a gene. The defective gene leads to a faulty component in the body. Since this defect can be accurately

copied in subsequent reproductive cycles, the defect in the body component becomes inheritable, making it a hereditary or genetic disease. Genetic diseases are not caused by an infectious agent, and cannot be transmitted through physical contact. Marrying blood relatives or marrying within close communities increases the risk of hereditary diseases among the children of such marriages.

What are the inheritance patterns in macular dystrophies?

These macular dystrophies can have different genetic patterns or ways of inheritance. Some may be passed on in a dominant pattern, others as a recessive pattern and still others as an x-linked trait carried by females and affecting males only. For many people with recessive inheritance patterns, it often comes as a surprise because they might never have known anyone in their family who was affected. In the case of a dominant pattern, generation after generation may be affected.

Diagnosis of juvenile macular degeneration

A comprehensive medical eye examination by an ophthalmologist is the first step towards diagnosing juvenile macular degeneration. When visual acuity is measured with a Snellen chart, it is usually decreased by at least 2 lines (e.g. 20/20 -> 20/80) if macular degeneration has occurred.

When viewing an Amsler grid, some straight lines appear wavy, and some patches of the grid may appear blank. Special tests such as fluorescein angiography, colour vision assessment and electroretinography may be necessary for an accurate diagnosis.

Can macular dystrophy lead to complete and total blindness?

It is reassuring to remember that the peripheral retina (side vision) usually functions normally in cases of juvenile macular degeneration and so it does not progress to total blindness. Many children with juvenile macular degeneration function very well, performing routine school tasks with only minimum extra help.

Treatment of macular degeneration

Glasses and low vision aids have been helpful in improving vision for patients with juvenile macular degeneration. Laser surgery or eye transplant surgeries are not effective.

Future trends and research

Scientists have been conducting research to improve the technology for gene identification and gene delivery. In 1997 the ABCR gene was discovered. Mutations in this gene are implicated in the causation of Stargardt's disease. Gene therapy could help to deliver active proteins to defective macular tissues. This future technology offers the greatest promise for a cure for these debilitating visual diseases.

Also being studied in this arena are the affected cells. Cell biologists begin to study in what ways is the cell affected because of a specific gene mutation. Also, what type of cell is affected, i.e., is it the rods and cone cells, or the layer of cells beneath them called the RPE cells.

Finally by learning from what the cell biologists have discovered, and by what the doctors or clinicians have discovered from their patients, a precise story can be developed as to what has gone wrong with this particular degeneration. That is, an educated guess or hypothesis can be put forward as to the cause of this type of macular degeneration or dystrophy. Many patients say, "Will this type of research help me now?" There may not be any immediate results from this type of research for the current patient with macular degeneration but it is important that the researchers understand the causes before they can ultimately figure out the cure.

Website addresses: www.blindness.org

Age-related Macular Degeneration

by Taraprasad Das, MD, FRCS

Age-related macular degeneration (AMD) is a degenerative disorder affecting the macula. The macula is the most sensitive portion of the back of the eye, the retina. AMD is the one of the important causes for vision loss among people above 50 years of age. There are two forms of AMD - the dry (non-neovascular) form and the wet (neovascular) form, with dry AMD occurring more, even though vision loss is greater with the wet form.

Large soft drusen and pigmentary changes of the retina characterize the early stages of AMD. Choroidal neovascular membrane (CNV), pigment epithelial detachment (PED), and geographic atrophy or fibrous scarring of the macula characterize the late stages.

Epidemiology and risk factors: Several studies have shown that the risk of AMD rises with age. Other risk factors include gender (females), smoking, and high blood pressure. Studies from the USA (the Beaver Dam study), the Netherlands (the Rotterdam Study), and Australia (the Blue Mountain study) have shown that between 1.2% and 1.7% individuals above the age of 60 years suffer from AMD. We do not have a detailed study in India, though it is estimated that 1.1% individuals above the age of 70 years have AMD. It has also been shown that there is 12.4 % chance of development of CNV in patients with bilateral drusen. The risk of development of CNV in the fellow eye when the contralateral eye has CNV is 10% in one year, 28% in three years, and 42% in five years.

Symptoms and signs: The early symptom of AMD is distortion of vision. Straight lines look curved. Depending upon the type of AMD and its progression there may be painless reduction of vision and, finally, complete loss of central vision though peripheral or navigational vision remains largely intact. Severe visual loss is rare in 80% of patients with dry AMD except when there is geographic atrophy.

Distortion of central vision, increased glare sensitivity, decreased contrast sensitivity and decreased color vision are the main symptoms of dry AMD. On the other hand the main symptoms of wet AMD are sudden reduction in central visual acuity, central or paracentral scotoma. There is no pain in the eye at any time.

Detection: The distortion of vision and the central scotoma can be detected by Amsler's chart. A detailed dilated eye examination is needed to search for signs of AMD, such as drusen, pigment abnormalities, choroidal neovascular membrane (CNV), pigment epithelial detachment and disciform scar. Drusen are yellow-white deposits, closely associated with the development of CNV, which are the hallmark of wet AMD. Depending on the location in relation to the center of the macula (fovea) the CNV could be extrafoveal (away from the fovea), juxtafoveal (proximal to the fovea), and subfoveal (under the fovea). The location has an important bearing on the choice of treatment. The precise mapping of the CNV and many other lesions of AMD are done with the help of fluorescein and/or ICG angiography.

Treatment: At present there is no treatment for dry AMD, except for the use of low vision aids. Both surgery and laser photocoagulation are available for treatment of neovascular (wet) AMD. Surgery either involves removal of the sub-retinal membrane behind the macula (submacular surgery) or rotating the macula to a new healthy location (macular rotation surgery). Both have potential risks and not a good and predictable surgical prognosis. Hence, often surgery is not enthusiastically prescribed for treatment of wet AMD.

Photocoagulation is the mainstay of treatment for neovascular AMD. Trials (such as the Macular Photocoagulation Study, MPS and Treatment of AMD with Photodynamic therapy, TAP) have shown that while thermal laser is beneficial in extrafoveal and some justafoveal CNV, photodynamic therapy or PDT (cold laser) is beneficial for subfoveal CNV. The latter requires injection of a photosensitizing drug before the laser treatment. Currently only verteporphin has been approved for human use. Newer treatments include use of other photosensitizing and angiostatic drugs.

Low vision devices are still required in all patients with AMD despite treatment with any form of laser. In dry AMD this could be the only treatment. Today a variety of optical and non-optical devices are available, but the patient needs to have a check-up and counseling with low vision experts. We have found low vision devices to be of significant help for patients with AMD.

Prevention: There is no known method for prevention of AMD. Care for hypertension and quitting smoking may help to some extent. A recent study (Age Related Eye Disease Study - AREDS) has shown that vitamins C and E, antioxidants or beta carotene, and trace metals like zinc can delay the progression of AMD. Hence eating green leafy vegetables and carrots is recommended for all those above the age of 40. It is also essential for them to have a dilated eye examination at least once a year by a retina specialist.

Summary
The increasing longevity of people the world over puts an increasing number at risk of developing age-related eye diseases. In India the life expectancy is likely to be 73 years by the year 2021. Maintaining an active lifestyle and satisfactory functional ability late in life depends greatly on good vision. Eye care providers have a key role in ensuring that effective vision is preserved for as long as possible.

Low Vision Rehabilitation Services - Macular Degeneration

by Mallineni Sharmila M.R.Sc MS

Low vision is a significant reduction of visual function that cannot be corrected by ordinary glasses, contact lenses, medical treatment and/or surgery. It is found in people of all ages, adversely affecting daily activities like reading, cooking, taking medication and watching television.

Low vision rehabilitation does not affect the physical condition of the eye; it cannot make the disease better or worse. The goal is to learn how to use the remaining healthy vision as effectively as possible. For people with macular degeneration, low vision devices can help them with functional vision tasks. This does not only mean helping them to see better, but also how to function better in everyday life. The counselling is aimed at addressing their lifestyle and work needs, hobbies, social and recreational needs, financial and personal needs.

The low vision specialists have a vast array of devices designed to help the visually impaired see and function better. These include magnifiers such as spectacle magnifier, hand magnifier, pocket magnifier, stand magnifier and telescopes, as well as electronic devices such as closed-circuit television.

Non-optical devices do not require lenses but they help improve viewing conditions through better lighting and improved contrast. Some of the non-optical devices are:

- Reading stands: They help maintain a suitable working distance for optical devices and for comfortable posture
- Overhead reading lamp: It improves contrast and provides focussed illumination
- Felt tip pen - the black ink improves contrast
- Notex: It enables a person to differentiate between currency of various denominations.
- Letter writer/bold line notebook: It improves contrast and helps people to write in a straight line.

How to use peripheral vision to see better
A very useful technique for people with macular degeneration is to intentionally look slightly off-center, a little bit away from what they want to look at. This works especially well for seeing food, watching television or recognizing faces. This method of seeing places the blind spot out of the way, pretending that the center of what they want to see is slightly over to the side or up or down. Everybody can find an ideal

place to look at, by experimenting and learning to make it work for them.

Every patient is different; a comprehensive low vision assessment will identify the patient's individual needs, problems and functional implications.

A Letter from a participant of LAP program

Dear Dr Sarfaraz Khan,

Just a few lines to thank you for the opportunity you gave me to attend the just concluded Low Vision Awareness Programme. It literally improved, without any devices - optical/non optical - my own very low vision concerning this much neglected ophthalmological problem! This very delicate subject and its management was put across to us in a very simple and practical way. It made me aware of the immense potential that low vision patients have to improve their lot, and to carry on a near normal life!

My gratitude extends to your associates, Ms Beula Christy and Ms Sharmila, both extremely competent professionals, fully committed to their mission in rehabilitating the visually handicapped.

Caetano de Loiola Pereira

NEWS

KIT FOR LOW VISION TOOLS

March 2004 witnessed the grand completion of the project 'Development of teaching learning materials for teachers to assist children with low vision.' The kit consists of materials such as handbook, assistive technology guide, vision stimulation material, as well as optical and non-optical devices to assist teachers in the management of low vision in classrooms. The Dr Richard Charles and Esther Lee Foundation has provided a grant for the project, through the International Council for Education of People with Visual Impairment (ICEVI). The center is also planning teacher preparation programs to train them in educational intervention techniques for helping children with low vision.

Resource Centre for Information Technology

Long-time supporters of LVPEI, the Deshpande family and Sunita and Praveen K Gottipalli, have also pledged their support for IT resource centre for the Vision Rehabilitation Centres. The centre will have software technology to enable the visually impaired to 'read' the computer monitor and access information regarding educational and employment opportunities.

Low Vision Awareness Program

October 01-03, 2004

Short term fellowship program in Low Vision Care

Three month program for ophthalmologists and optometrists commencing January 1, April 1, July 1 and October 1, 2004.

For more details contact to:

Dr. Sarfaraz Ali Khan
Director
VISION REHABILITATION CENTRES
L.V. Prasad Eye Institute
L.V. Prasad Marg, Banjara Hills
Hyderabad - 500 034
Ph: (091) 040 2354 2790, 2360 8262,
Fax :(091) 040 2354 8271
email : sarfaraz@lvpei.org

You can help the Vision Rehabilitation Centres of the L.V. Prasad Eye Institute to several ways: to discover basic causes and treatment strategies for eye disease through research, restore the vision ofan indigent patient, or and help expand the frontiers of ophthalmology. Contributions to the Hyderabad Eye Institute or the Hyderabad Eye Research Foundation are tax deductible.

(Donations above Rs. 250 are exempt under Section 80G of Income Tax Act, 1961 for the Hyderabad Eye Institute and under section 35(i) (ii) for the Hyderabad Eye Research Foundation). For more information, please contact :

You Can Make A Difference

VISION REHABILITATION CENTRES
L.V. Prasad Eye Institute
L.V. Prasad Marg, Banjara Hills
Hyderabad - 500 034
Ph: (091) 040 2354 2790, 2360 8262, Fax :(091) 040 2354 8271
email : sarfaraz@lvpei.org
Web : www.lvpei.org