Age Related Macular Degeneration - An Overview

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Age related macular degeneration (AMD) is one of the leading causes of bilateral visual loss among the elderly across the globe. It accounts for 75% of legal blindness in individuals older than 50 years. On the basis of clinical features & histopathology AMD has been classified into Dry or Non-exudative type & Wet or Exudative/ Neovascular type. With aging, there is also an increasing risk of developing the more severe i.e. wet form of AMD (2% of AMD cases between the ages of 43 and 54 versus 29% of cases among people older than 75 years). Although far less common than the dry form of AMD (15%vs85%), the exudative AMD is responsible for most cases of severe visual loss. Differences in the prevalence of AMD based on gender, race, or geographic area have been observed. With increasing longevity & shift in demographic profile, prevalence of the disease is expected to rise dramatically in India also. It has been proposed that disease should be renamed to age related maculopathy (ARM), early & late forms with the term AMD reserved for the late forms of the disease & encompassing geographic atrophy & neovascular AMD. Early ARM represents changes predisposing to AMD.

The International Epidemiological Age- related Maculopathy study Group has defined early ARM as a degenerative disorder in persons >50 years characterized by the presence of any of the following

- Soft Drusen (>63µm). Soft indistinct are more pathognomonic than soft distinct & >125µm are still more significant.
- Areas of hyperpigmentation and/or hypopigmentation associated with drusen but excluding pigment surrounding small, hard drusen
- Visual acuity (VA) is not a criterion for diagnosis of ARM as advanced changes are sometimes seen with good VA due to sparing of the fixation.

The group has excluded small, hard drusens, & pigmentary changes surrounding them as (1) Hard drusen become almost constant finding in the fifth decade, (one or more hard drusen is seen in 95% of population aged over 43 years) and (2) number of diverse processes can cause pigment abnormalities. However eyes with both these characters can also proceed to AMD.

Chorio-retinal changes of ageing: With ageing there is loss of photoreceptors & retinal pigment epithelium (RPE), accumulation of lipofuscin in the RPE, & free radical mediated damage to the RPE cell structure & enzymes, and deposition of patches of abnormal basement membrane material called basal laminar deposits (BlamD) and basal linear deposits (BlinD). BlamD consist of membrano-granular material and foci of wide spaced collagen between the plasma membrane and basal lamina of the RPE. BlinD consist of vesicular material located in the inner collagenous zone of Bruch's membrane. The thickening of Bruchs membrane leads to reduction in the permeability which further compromises RPE & outer retinal layers most notably photoreceptors. The choroid shows decrease in the blood flow by laser Doppler flowmetery & this is due to decrease in the thickness of middle layers comprising of medium sized vessels. This throws the remaining larger vessels onto greater prominence accounting for the senile tigroid fundus. At what stage these diffuse changes of ageing become abnormal& pathological and hence predispose to AMD is not clear.

The diffuse deposition of extracellular material beneath the RPE though plays a vital role in the evolution of ARM, is very difficult to study clinically. Hence greater emphasis is laid on to the Drusens, which are localized collections of BlinD or localized detachment of BlamD.

Dry AMD: Clinical features & histopathogenesis

Drusens the hallmark of dry AMD can be classified into Hard and Soft types clinically. However, on the basis of light & electronic microscopy they can further be sub classified based on the type of accumulating material.

Hard Drusen: As small hard drusens are ubiquitously present over the age 40 they were thought not to be associated with increased risk of AMD in earlier studies. However 10 year follow up of enrolled population in Beaver Dam study has shown that large number of hard drusens predict higher incidence of soft drusen & pigmentary changes, both of which significantly increase the risk for the development of AMD. Clinically hard drusens are not visible in the fundus till they measure about 50µm, when they are seen as well defined, discrete, yellow white deposits external to RPE. On fundus fluorescein angiography (FFA) they are seen as pin point window defects even when as small as 25µm.

Large, Soft Drusen: These are >63µm, have ill defined

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borders & vary in size & shape. They have a tendency towards confluence. On the basis of pathogenesis these can be divided into three types

- Granular soft drusen: Clinically about 250µm (2x vein width) with a yellow solid appearance, there confluence resulting in sinuous shapes.
- Soft serous drusen and drusenoid pigment epithelial detachments (PEDs): Clinically larger than 500µm, may have pooled serous fluid, appearing blister like. Their further confluence leads to larger soft drusen that resemble serous PED. Both of the above show late fluorescence on FFA due to staining. Drusenoid PEDs are consistent with good vision although very large ones may cause distortion. However as they collapse, atrophy, rather than choroidal neo-vascularization (CNV) sets in & visual acuity then rapidly deteriorates.
- Soft membranous drusen: Clinically between 63-175µm (0.5-1.5 vein widths), appear paler & shallower than the granular drusens. They herald a high risk of development of CNV. On angiography they fluoresce later & less brightly than small hard drusens.

Regressing Drusens: All drusens may disappear in time, but this does not mean a return to normal state. Drusens begin to regress when the overlying RPE fails. Now they become whiter & harder due to inspissation of contents. Hypo & hyper pigmentation develops over the surface, margins become irregular & foci of calcification appear. Ultimately drusens fade leaving multifocal patches of RPE atrophy.

The risk of visual loss in subjects with drusens has been reported in some studies & varies from 23.5% over three years to much lower cumulative risk of 12.5 over 5 years. The cause of disparity between the two studies could be due to higher prevalence of soft drusens in the former.

Geographic atrophy: It is the end result of atrophic form of AMD. It is seen as a sharply delineated area of hypo or depigmentation in which choroidal vessels are more visible than the surroundings. It commences around the perimeter of fovea, & may spare the fixation for several years. Most cases occur in eyes with prominent drusens & develop as the drusens regress. However, it may follow collapse of an RPE detachment, especially the drusenoid type. It tends to be bilateral; in one series of 200 patients the fellow eye was affected in over 50% cases. Within the area of atrophy histologically there is absence of photoreceptors, RPE, & choriocapillaris. Long-standing cases also demonstrate choroidal atrophy, loss of medium sized vessels, and the larger choroidal vessels showing sclerosed appearance due to fibrous replacement of the media.

Geographic atrophy accounts for 12-21% of eyes with severe visual loss due to AMD, however the percentage with functional handicap is much more as a large area of atrophy even though sparing the fixation markedly reduces the reading speed. Reduced contrast sensitivity, paracentral scotomas along with reduced visual acuity in dim light add to the insult.

Neovascular (Exudative) AMD: Clinical features & histopathogenesis Overall 10% of AMD patients have the wet form of AMD. This includes CNV and associated manifestations like RPED, RPE tears, disciform scarring, and vitreous hemorrhage. Majority of AMD patients with vision, <20/200 have wet form of AMD. Most patients with CNV complain of blurred and/or distorted vision, central scotomas leading to difficulty in reading & recognizing faces. After noting the visual acuity, the scotomas should be mapped on an Amsler grid in order to have a fair idea of the patient's handicap & for planning low vision aids for the patient.

CNV: Clinically on slit lamp biomicroscopy CNV appears as grey-green elevation deep to retina with overlying neurosensory detachment, however, this characteristic appearance may not always be present in CNV due to AMD. In such scenarios the presence of CNV is indicated by any one of the following

- Subretinal blood or lipids
- RPED with or without overlying subretinal fluid.
- Occasionally a shallow neurosensory serous RD may be the only presenting sign of underlying CNV.

The CNV capillary network becomes more apparent after the atrophy of overlying RPE. CNV has been classified into classic and occult depending upon the angiographic appearance (described later). Depending upon its location CNV may be subfoveal, juxtafoveal (between 1&199µm from the centre of FAZ), or extrafoveal (>200µm from FAZ centre). Histologically CNV is growth of abnormal, fragile new vessels between the Bruchs membrane & RPE or between the latter & neurosensory retina. These vessels sprout from the chorio capillaries & proceed inwards through the defects in the Bruchs membrane

RPEDs: appear as sharply demarcated, dome shaped elevations of RPE. If filled with serous fluid they transilluminate. Three types of PEDs are seen & can be differentiated on the basis of their Angiographic pattern (described later)

- Drusenoid PED -does not have CNV
- Fibrovascular PED-is a form of occult CNV
- Serous PED-may or may not overlie CNV

Overlying serous RD, lipid & blood within or surrounding a PED implies the presence of CNV. Sub RPE blood is seen as green or dark red mound.

RPE tear: or rip occurs as a complication in serous or fibrovacular PED. It occurs at the border of attached & detached RPE due to stretching forces of the underlying





Fig.1: Geographic Atrophy

Fig.2: Classic CNV on FFA

fluid or from the contractile forces of the fibrovacular tissue. Clinically it is seen as area of hypopigmentation with hyperpigmented wavy border on one side due to rolling in of the free edge of torn RPE.

Massive sub retinal hemorrhage and breakthrough vitreous hemorrhage though unusual complications of AMD, are seen sometimes and result in sudden profound visual loss both central as well as peripheral.

Disciform Scar: is the last stage in the evolution of neovascular AMD just as geographic atrophy is in dry AMD. CNV is a fibrovascular tissue; however, the fibrous component is not readily appreciated in the early stages of CNV due to immaturity of the fibrous tissue & also due to the overwhelming signs like serous RD, subretinal lipids and/or blood, of the vascular component. When the fibrous tissue becomes apparent clinically then the fibrovascular complex is called disciform scar. Clinically it appears as white to yellow subretinal scar with intervening areas of hyperpigmentation. If the vascular component has died its own death then the scar does not grow, however, it can expand with neovascularization occurring along the edges.

Demographic Factors

Age: The incidence, prevalence, and progression of all forms of AMD increase with advancing age.

Gender: Female sex is associated with a slightly greater prevalence of AMD in many studies. Pooled data from the Beaver Dam Eye Study, Blue Mountains Eye Study, and the Rotterdam Study revealed no sex differences in AMD risk. However, recent analyses from the Blue Mountains Eye Study suggest that the 5-year incidence of neovascular AMD among women is double that of men (1.2% vs. 0.6%). *Race:* All forms of AMD are more prevalent in the white population than more darkly pigmented races leading to the belief that melanin may be protective against development of CNV. In the Baltimore Eye Survey, AMD accounted for 30% of bilateral blindness among whites and for 0% among blacks. The presence of melanin also seems to protect against

formation of RPE lipofuscin, a marker of cellular senescence and promoter of oxidative damage. Pooled data from large studies conducted in three continents has found no association between iris color and AMD.

Fig.3: ICG angiography picture

of CNV shown in Fig 2

Ocular Risk Factors: Weak association between hyperopia and early AMD have been suggested, but not with late AMD. Higher levels of ocular melanin may be protective against light induced oxidative damage to the retina; however till date literature is inconclusive about this. Similarly data regarding the relationship between cataract and AMD is inconsistent.

Cardiovascular Factors: Cigarette smoking is third only to older age and family history as a significant risk factor for AMD. Smoking has been associated with an increased risk of developing AMD in most population-based studies. Smokers developing AMD 5 to 10 years before non-smokers. This may be due to effect of smoking on antioxidant metabolism and choroidal blood flow. Cigarette smoking is associated with higher rates of recurrent CNV after laser photocoagulation. However, the association between systemic hypertension and neovascular AMD is more persuasive. The visual benefits of laser photocoagulation of CNV do not extend to those with systemic hypertension suggesting an etiologic relationship to angiogenesis.

Light Exposure: Excessive exposure to light can damage the retina. Photooxidative damage, mediated by reactive oxygen intermediates (ROI), has been implicated as a culprit in the development of AMD. Overall, the data do not support a strong relation between ultraviolet exposure and AMD.

Dietary and Medication Factors: Data from the Age-Related Eye Disease Study (AREDS) suggests that



Fig.4: Colour, FFA & OCT picture of Wet AMD

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supplementation with very high doses of zinc; vitamin C, vitamin E, and ß-carotene provide a modest protective effect on progression to advanced neovascular AMD, in patients with extensive drusen or fellow eyes with neovascular AMD. This benefit did not extend to patients without AMD or with few drusen. Meanwhile, in the absence of convincing evidence, patients should be cautioned that nutrient supplements are not necessarily innocuous. For among male smokers, instance, ß-carotene supplementation increases the risk of lung cancer and mortality, whereas multivitamin supplementation increases overall cancer mortality. High serum levels of zinc are associated with Alzheimer's disease.

Evaluation of ARMD patients

Evaluation of ARMD patients includes slit lamp biomicroscopy with both macular noncontact and contact lenses. The latter technique facilitates detection of subtle signs, such as shallow sub-retinal fluid. Proper clinical examination supplemented with investigations help in complete evaluation of the disease and plan management.

Investigations that help in evaluation are:

- Fundus fluorescein angiography (FFA)
- Indocyanine green (ICG) angiography
- Optical coherence tomogram (OCT)
- Multifocal electroretinography (MERG)

Fundus fluorescein angiography: Two basic angiographic patterns for choroidal neovascular membranes (CNVM) were defined by the macular photocoagulation study (MPS).

- Classic CNVM present as discrete, early hyperfluorescence with late leakage of dye into the overlying neurosensory retinal detachment. A lacy pattern within the CNVM is most often not observed in exudative AMD.
- Occult CNVM are categorized into 2 basic forms, late leakage of undetermined source and fibrovascular PEDs.
 - Late leakage of undetermined source manifests as regions of stippled or ill-defined leakage into an overlying neurosensory retinal detachment without a distinct source focus that can be identified on the early frames of the angiogram.
 - Fibrovascular PEDs present as irregular elevation of RPE, which is associated with stippled leakage into an overlying neurosensory retinal detachment in the early and late frames of the angiograms.
 - Fibrovascular PEDs can be differentiated from serous PEDs, which show more rapid homogenous filling of the lesion in the early frames without leakage in the late frames of the angiogram. Serous PEDs typically show smooth and sharp

hyperfluorescent contours. Other causes of RPE elevation to differentiate from the entities listed above include hemorrhagic PEDs, RPE hyperplasia, and confluent soft drusen.

- Hemorrhagic PEDs present clinically with dark sub-RPE blood, which blocks choroidal fluorescence on angiography.
- RPE hyperplasia also will block choroidal fluorescence.
- RPE tears will present as regions of intense hyperfluorescence in the area bereft of RPE due to transmission of choroidal fluorescence and in the late stages as scleral staining. This is adjacent to sharply demarcated blockage of the choroidal fluorescence by the redundant scrolled RPE. CNVM may be associated with the RPE tear, causing leakage in addition to the above findings.
- Confluent soft drusen, which often present in the fovea, typically show cruciate pigment clumping. Confluent soft drusen will show angiographic findings that are similar to serous PEDs with homogenous pooling of dye and no leakage, but they typically exhibit only faint fluorescence.
- Disciform scarring shows diverse angiographic characteristics. Angiographically, the fibrotic component will block the choroidal fluorescence in the early frames of the angiogram, followed by late staining. Hyperpigmented regions as well as subretinal hemorrhage will block the choroidal fluorescence throughout the angiogram. Regions of active CNVM will show leakage.

Indocyanine green angiography

ICG facilitates the study of the choroidal circulation by better delineation of the choroidal circulation than fluorescein. Unlike fluorescein, ICG is strongly bound to plasma proteins, which prevents diffusion of the compound through the fenestrated choroidal capillaries and permits better delineation of choroidal details. ICG can facilitate visualization of choroidal vasculature and CNVM through hemorrhage.

ICG angiography can show CNVM as localized hot spots or as diffuse hyperfluorescent plaques. ICG could better reveal the occult CNVM with ICG angiography. ICG is particularly useful in delineating the other variant of AMD, the Polypoidal choroidal Vasculopathy (PCV). This disease thought to affect women in pigmented population, is now found in Asians with an almost equal predisposition among men and women. The patterns of ICG fluorescence seen in PCV are: early phase filling of larger choroidal blood vessel and the network of polyps arising from the large choroidal blood vessels can also be identified; in late phase, reversal of the pattern is noted causing hypo fluorescence in the center of the polyps. Late staining seen in CNVM is not noted in PCV. PCV in Indian population differ from those in other populations in being more common in males and in its macular location as compared to females and peripapillary location in western population.

Scanning laser ophthalmoscopy (SLO):

Confocal scanning angiography is a useful alternative to video-ICG angiography. Advantages compared to routine digital ICG videoangiography are, high image contrast, visualization of retinal vessels in the late phase, lower amount of light exposure and direct digital image acquisition. One group described the use of a 2-wavelength SLO to facilitate simultaneous recording of ICG and FA in 340 cases, two thirds of which had well-defined or occult choroidal neovascularization in ARMD. The angiograms are displayed as one combined red-green picture. They noted that this method allowed a precise comparison of the transit of both dyes through the circulation with perfect alignment of the critical retinal vascular landmarks provided by the fluorescein images onto the ICG angiogram.

Some studies have shown fundus auto fluorescence with SLO imaging provides a reliable means to delineate areas of geographic atrophy (GA). The automated image analysis allows more accurate detection and quantitative documentation of atrophic areas than manual outlining. This method will be useful in longitudinal natural history studies and for monitoring effects of future therapeutic interventions to slow down GA progression in patients with advanced atrophic AMD and other retinal diseases associated with outer retinal atrophy.

Optical coherence tomogram (OCT): OCT comes very handy in complete evaluation of AMD. Precise anatomical location of the CNVM is possible with OCT. It is possible to localize whether it is sub RPE (type 1), or sub neurosensory retina (type 2).

CNVM appear as highly reflective band in the retina pre-RPE or sub-RPE depending on the location associated with subretinal fluid which, appear as optically empty space between RPE band and sensory retina on OCT. Drusen may appear as areas of focal elevations of RPE, with shallow borders and no optical shadowing underneath. Subretinal hemorrhage when present, appear as hyper reflective with ill defined borders.

Emerging technologies that may help in screening and diagnosis include

- Spectral OCT: Gives good resolution.
- *Scanning laser ophthalmoscope with optical coherence tomography (SLO-OCT):* This takes coronal-like retinal

images.

These technologies will likely enable OCT scans to be confidently registered within their exact retinal location.

Multifocal electroretinography (MERG): This may be performed on the retina to evaluate the functional response of rods and cones. This test is not required for evaluation of AMD, but various authors have used it to follow the progression of the disease.

Management of AMD-To be continued in next Issue

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Management of Acute Central Chorioretinitis

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Active central chorioretinitis is a common clinical condition, requiring accurate and prompt diagnosis, appropriate investigations and early treatment for a good visual outcome.

Most central retinal lesions are a serious threat to vision. These may be focal, multifocal or diffuse, of which one specific presentation is a single, central retinal lesion. A white-yellow, large, elevated lesion at the macula may be the presenting feature of various pathologies :-

Active central choroiditis/chorioretinitis.

Solitary retinal haemangioma

Flat retinal astrocytoma

Central Serous Retinopathy (CSR)

Confluent patches of APMPPE (Acute Posterior Multifocal Pigment Placoid Epitheliopathy)

Confluent subretinal exudates at macula.

Best's vitelliform dystrophy

We describe two similar cases of active central choroiditis.

A 17 year old female presented with reduction of vision in RE for 2 months and LE for 2 years. Visual acuity on presentation was FC 1 feet and did not improve with pin hole. Pupillary reaction was normal in BE.

Slit lamp examination showed no KPs, flare or cells, and revealed a pinpoint opacity in the region of anterior lens capsule in RE. No vitreous haze or floaters were seen. LE was normal.

On fundus examination RE with + 90D lens a large pale white lesion, 1DD in size and hazy margins was seen adjacent to and slightly below the macula. The lesion appeared elevated and the surrounding retina appeared edematous (Fig.1).

Fundus of LE showed large confluent pigmented patches of disseminated healed choroiditis.

The patient was clinically diagnosed as a case of active central

State Institute of Ophthalmology, Allahabad choroiditis RE and old healed choroiditis LE.

FFA of the patient was performed. The right eye showed an early hypoflourescence in the area of the lesion. Later phases showed gradually increasing intensity fluorescence, beginning at the margins and then the inner portion of the lesion. The size remained the same.

The left eye FFA showed a combination of hyperflourescence, and blocked fluorescence (window defects).

She was started on IV methylprednisolone 250 mg QID for 3 days, then on oral Prednisolone 1 mg/kg body weight for 11 days, followed by steroid taper over the next 3 days. Oral Sulphamethoxazole 800 + Pyrimethamine 160 were given to be continued for 3 weeks.

On daily follow-up, vision RE improved to FC 3 ft, FC 3 m, 6/36 and 6/9 on days 2, 3, 4 and 7 respectively. At the end of 2 weeks, vision was 6/6 RE.

The lesion gradually resolved by decreasing in size and resorption of retinal edema around it. After 2 weeks, retinal edema also subsided completely.

Hematological and immunological profile of the patient was done. TORCH was positive for IgG and IgM Toxoplasma. Additional tests were done to rule out tubercular infection. She was thus diagnosed as ocular toxoplasmosis.

Discussion

Toxoplasma is one of the leading causes of infective

chorioretinitis, and can be congenital or acquired. Congenital toxoplasmosis manifests both as systemic and ocular lesions. Acquired infection that usually occurs in an immuno competent patient, is commonly asymptomatic. The parasite, on reaching the eye, establishes in the retina (as it has a predilection for neural tissues) and then secondarily involves the choroid.

Frequently, the ocular presentation is that of focal

necrotising retinitis adjacent to a large atrophic scar, or a satellite lesion. Acquired cases may present as unilateral focal chorio retinitis with no pre-existing scar in either eye. Retinitis occurs due to bursting of cyst and release of



Fig. 1: Red free photograph RE showing active choroiditis lesion inferotemporal to macula.



Fig. 2: Initial arteriovenous phase of FFA RE showing blocked fluorescence

bradyzoites.

The lesion is usually round-oval, larger than IDD in size, a soft white fluffy infiltrate with surrounding retinal edema and subjacent choroiditis. Vitritis overlying the lesion is a hypersensitivity response to inflammation and gives the classical 'Headlight in fog' appearance. Perivascular inflammatory exudation occurs in areas

peripheral to the lesion. More than a third of lesions involve the macular area.

Even if there is evidence of old lesions in an immuno competent host, the disease seems to be activated only in one eye at a time. In more than half the patients, anterior uveitis also occurs.

As inflammation subsides, the lesion becomes less bright yellow, ultimately becoming atrophic with pigment heaping around its edges.

However, pigment clumping is not universally seen around all old lesions, hence is not a diagnostic sign. With time, vitreous haze also subsides and in case where fovea is uninvolved, vision improves greatly. However, intensive early treatment is required for a favourable visual outcome in lesions threatening the fovea.

Diagnostic difficulty may arise in cases of co-existent active and inactive lesions. Apart from the aforementioned

ophthalmoscopic features, an important diagnostic tool is FFA. FFA of the active lesion shows early hypoflourescence (fig. 2) due to relative blockage by associated retinal and choroidal edema and also due to delayed choroidal filling. Subsequent fluorescence occurs beginning at the margins an later filling the inner part of the lesion (Fig. 3). This causes hyperflourescence with fuzzy marings. Later phases show



Fig. 3: Late arteriovenous phase showing

hyperfluorescence of choroiditis lesion RE.

Fig. 4: Late FFA RE showing intense staining of the whole lesion.

persisting hyperflourescnece due to dye diffusion into surrounding space from the leaky capillaries (Fig. 4).

Old lesions show both hyperflourescent and hypoflourescent areas (Fig. 5, Fig. 6). Transmitted fluorescence is due to overlying RPE atrophy causing visibility of larger choroidal vessels. Blocked fluorescene occurs due to heaping up of pigment. Late marginal fluorescence is due to scleral staining.

Our patient showed a single lesion in the RE close to macula. No satellite lesions were seen in this eye. However, the other eye showed old choroiditis without any active lesion around the healed lesions. The patient improved on the usual treatment for toxoplasma.

Similar clinical and angiographic findings were seen in the case of an adult male who presented with gradual

> blurring of vision. He improved on oral prednisolone alone. His serial angiograms are shown in fig. 7(i), (ii), (iii).

The condition must be differentiated from other similar looking lesions as follows :

Solitary retinal haemangioma is usually associated with other systemic lesions and present by 2-3rd decade. On FFA early hyperflourescence occurs due to rich vascularity and late leakage due to

incompetent vessel walls.

Flat retinal astrocytoma is a benign, non vision threatening tumour and may be seen in cases of tuberous sclerosis. It may be a white, semi-translucent nodule, becoming more solid and white with time, and may even calcify.

CSR occurs typically in young males with blurring of vision, and on FFA gives a characteristic ink-dot or



Fig. 5: Red free photograph LE of same patient showing old healed patches of choroiditis.



Fig. 6: FFA LE showing window defect.



Fig. 7(i): Early hyperflourescence of lesion superonasal to macula.



Fig. 7(ii): Late intense hyperflourescence of lesion.



Fig. 7(iii): Diffuse staining of area surrounding lesion.

umbrella shaped hyperflourescence.

APMPPE present in 3-5 decades with subacute unilateral visual impairment, which soon affects the other eye. Associated anterior uveitis, mild vitritis, disc oedema and periphlebitis may be detected. FFA shows early hypoflourescence due to blocked choroidal flush. Late phases show hyperflourescence due to staining.

Confluent subretinal macular exudates may occur in vascular diseases, the fundus examination is likely to reveal haemorrhages, vascular dilatation, tortuosity and macular edema areas of exudates show blocked fluorescence on FA.

Bests Vitelliform dystrophy presents by 4-6 decades as a bilateral, symmetrical, round, yellow, elevated, subfoveal lesions, usually multiple. Visual loss is minimal, slight color vision defects may be seen.

FA shows central hypoflourescence surrounded by a small irregular hyperflourscence.

Hence we see that active central choroiditis is an important clinical entity and requires prompt diagnosis and early treatment for a good visual prognosis. FFA can be a very useful diagnosing tool for differentiating acute lesions from chronic ones and must be performed especially in eyes showing old lesions. The disease has a good prognosis if managed in time.

Semi-Soft Contact Lens Fitting in Special Situations

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Contact lens usage has increased markedly in our country in the past few years. Most of the increase has been in the soft contact lens segment. This has been possible because of improved materials and design of soft lenses, easier fitting parameters and greater patient acceptance of these lenses. Rigid gas permeable lenses (R.G.P) are still widely used and have a definite role to play. There are, however, some conditions of irregular corneal surfaces that specifically warrant the use of rigid gas permeable lenses.

Irregular corneal surfaces may be caused by trauma, corneal surgery and corneal diseases such as rosacea keratitis, herpes simplex stromal keratitis and anterior basement membrane dystrophies. Many of these conditions induce focal epithelial and stromal abnormalities which disrupt the regularity of the anterior refracting corneal surface causing irregular astigmatism and this induces excessive light scatter.

This article endeavours to provide an overview of R.G.P lens fitting in such special situations and is by no means an exhaustive review on the subject. Fitting such cases is comparatively difficult but gratifying results can be obtained by constant perseverance.

The corneas of patients with irregular astigmatism are no longer a simple toroidal refracting surface. Hence, spherocylinder spectacles cannot fully correct them. Rigid gas permeable (R.G.P) lenses provide these patients with their optimal obtainable vision because the tear lens formed between the corneal surface and the back surface of R.G.P contact lens optically neutralizes the corneal distortions. However patients may be reluctant to pursue contact lens fitting after trauma, refractive surgery or after they have just been diagnosed with corneal disease. Spherical base curve R.G.P contact lenses correct majority of the refractive astigmatism and provides an acceptable fit. Toric R.G.P contact lens should be considered only after ruling out the spherical design. Some of the more common conditions are being considered in this article.

Keratoconus

The diagnosis of Keratoconus is made on the basis of observing distortion of the mires in Keratometry, scissors reflex in retinoscopy and slit lamp signs of Fleischer's ring, Vogt's Striae and corneal thinning with ectasia.

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Table 1 : Corneal Conditions that induce Irregular Astigmatism.

Corneal Dystrophies

- Anterior Basement Membrane Dystrophy.
- Stromal Dystrophies.
- ➢ Keratoconous
- Keratoglobus.
- Pellucid Marginal Degeneration

Corneal Degenerations

- Salzmann's Nodular Degeneration.
- > Terrien's Marginal Degeneration.

Other Corneal Pathology

- > Herpes Keratitis
- > Healed Corneal Ulcers
- > Healed Corneal Lacerations and other Trauma.
- > Pterygium
- Permanent Contact Lens induced warpage

Surgically Induced

- Cataract Extraction
- Retinal Detachment
- Penetrating Keratoplasty
- Keratorefractive procedures
 - Radial Keratotomy (R.K)
 - Photo Refractive Keratectomy (P.R.K)
 - Automated Lamellar Keratoplasty (A.L.K)
 - Laser Assisted Situ Keratomileusis (LASIK)
 - Post- LASIK Keratectasia

Corneal topography helps to confirm the diagnosis and also to assess the morphological shape of cornea. The cone type can be classified as either central cone (nipple) or sagging cone (oval). In the central cone, the area of steepening is located very near the visual axis, is small in diameter and is uniformly surrounded by corneal flattening. A sagging cone is characterized by a large area of steepening located usually infero-temporally to the visual axis and may have a thinner zone of flattening peripheral to the cone in the same quadrant as the cone dislocation.





Fig.1(a): Post Corneal Tear Repair

Fg.1(b): Contact Lens Fitting in the same Patient

• Small lens diameter and optical zones.

Sagging Cone

- Allow greater apical touch.
- Attempt superior corneal alignment and lid attachment.
- Allow inferior edge stand off.
- Larger lens diameter and optic zone.

• Inferior lens decentration is common.

Nottingham was the first to appreciate that keratoconus is essentially a disease producing corneal thinning and then protrusion. In earlier days the management of the condition as described by Nottingham was mainly surgical but he had also mentioned using glass shells filled with gelatin. The first report of a powered contact lens used in keratoconus was by fick in 1888.

If keratoconus is mild with minimal corneal distortion and acceptable manifest refraction vision, patient can be managed with spectacles or soft contact lens. As the condition progresses, R.G.P contact lenses are indicated to achieve optimal vision.

Initial diagnostic lens should be chosen with a base curve similar to the steep keratometry. If the fluorescein pattern of the initial trial lens is of apical touch, apply lenses with a steeper base curve (usually in 0.10-or 0.20 mm step) until apical clearance is achieved and then perform an spherocylindrical over refraction. The equivalent sphere of over refraction is added to trial lens power to determine the power of the lens prescription.

In keratoconus patient peripheral curves need to be flattened to obtain adequate tear exchange so attention needs to be given on amount of edge clearance during follow-up visits. If bubbles or dimple veiling are observed beneath optic zone during fluorescein pattern evaluation, it indicates need for a smaller diameter or flatter peripheral curves.

Other type of Lens to Fit in Keratoconus

Scleral lens: The scleral contact lens rests over the sclera. Both front and back surfaces of scleral contact lens are optically finished. With the introduction of improved material (R.G.P material), corneal oxygenation has improved despite the greater thickness of these lenses.

Piggy-back: The term piggy-back refers to fitting of a R.G.P lens on to a soft contact lens, which then acts as a carrier. A soft contact lens of +4.00D or +5.00D power will provide an adequate lenticular bowl onto which the R.G.P lens is fitted. In special situations such as a very flat cornea (Post P.K), a high plus power soft lens can be used and if the cornea is too steep (Keratoconus), one may use a minus power soft contact lens.

The Rose K lens: An effective option to fit moderate to advanced keratoconus. The Rose K system uses R.G.P lenses of decreasing optic zone as the base curve steepens (eg.base curve 45.0D, optic zone 6.5 mm) and a computer generated peripheral system. Optic zone is significantly smaller so that it can saddle the cone. This minimizes mid-peripheral lens impingement and pooling of tears at the base of the cone. The mid-periphery has a consistent flattening from the optic zone to lens edge to provide suitable peripheral clearance.

Hybrid lens: In this type of lens a soft rim is fused onto a hard central portion. Hence these lenses have the optics of a rigid lens with the comfort of a hydrogel lens.

Over all optic zone diameters are generally smaller in keratoconus and optic zone ranges from 7.00 mm in mild cases to 5.5 mm in advanced cases.

Keratoconus Fitting method based on Cone Morphology *Central Cone*

- Achieve faint apical touch or have apical clearance (vault apex)
- Aim for well-centered lens



Fig. 2(a): Topography Showing (a) Nipple Cone In Keratoconus



Fig.2 (b): Topography Showing Sagging (Oval) Cone in Keratoconus





Fig.3(a): Central Bearing and Inferior Edge Clearance

Fig.3(b): Apical Touch and three Point Fitting Relationship

The SoftPerm is one such lens. It is fitted like hydrogel lens. The lens is expensive, but useful for some patients with keratoconus.

Pellucid Marginal Degeneration

In pellucid marginal degeneration, the cornea thins near the inferior limbus, resulting in inferior corneal protrusion .In contrast to keratoconus, maximum corneal protrusion occurs just superior to rather than within the area of thining. Fleischer's ring and Vogt's Striae are not present with pellucid.

Keratometry reading often reveal high amount of against the rule toricity, and in pellucid the Gull-winged inferior areas of steepening are observed with videokeratography. Central corneal distortion is minimal for patient with pellucid. Therefore spectacles or toric soft contact lens may be prescribed to correct patients with against the rule astigmatism or oblique astigmatism. With the spherical R.G.P contact lens one can achieve most successful fit with a lid attachment fitting relationship. Most other R.G.P lens designs, including the back toric lens design, position too inferiorly in pellucid marginal degeneration. Piggyback design should be considered if other options fail to provide acceptable fit. post P.K corneas, Waring et al (1992) described 5 major shapes of cornea:-

1) Prolate shape- Steeper in center and flatter in the periphery.

2) Oblate shape- Flatter in center and steeper in the periphery.

3) Mixed Prolate and Oblate Shape- Described as regular astigmatic pattern.

4) Asymmetric Pattern- The distinguishing feature is two steep hemi-meridian, which are not 180 apart.

5) In Steep to Flat Pattern- The steepest hemimeridian is located 180° from the flattest hemi-meridian of the cornea.

Most eyes do well if fitted with spherical R.G.P contact lens. However, front surface torics or bi-toric R.G.P lenses are occasionally required.

A fitting relation that evenly distributes lens bearing is desired. If the lens is fitted too steep, lens adherence could lead to an acute red eye episode. One should avoid prescribing harsh bearing at the host-donor junction. Large overall diameters are often prescribed to optimize lens centration. Optic zone diameter is generally prescribed smaller than the size of donor lenticule. One should carefully note the area of fluorescein staining at follow-up visits to help determine any modifications to the lens design.

Piggyback and hybrid lenses may be options post operatively to minimize the lens discomfort and lens decent ration but full time use of this modality can limit the corneal oxygenation increasing the risk of neovascularization and graft rejection.

Post Keratorefractive Procedure Astigmatism

Post Penetrating Keratoplasty (PK) Astigmatism

Penetrating keratoplasty, particularly for Keratoconus and Corneal dystrophies now enjoys a very high success rate. However, irregular and regular corneal astigmatism occurs in an estimated 25% of eyes post PK due to surgical techniques and patient healing response. R.G.P contact lenses are often required to correct the astigmatism present after P.K. We can fit a post P.K cornea with R.G.P lenses approximately 4 months after the surgery.

Based on topography analysis in



Fig.4(a): RGP Lens Fitting In Keratectasia (Post LASIK)

Clinically significant irregular astigmatism may occur after refractive corneal surgery, particularly after radial



Fig. 4(b): Fluorescein Pattern in the same Patient

Topographic Patterns	
Shape	Suggested R.G.P Lens Design
Prolate	Standard spherical posterior curves, Aspheric, Bi-aspheric, Bi-toric, Keratoconus design
Oblate	Reverse geometry lens, Bi-toric
Mixed	Bi-toric
Asymmetric	Standard spherical posterior curves, Keratoconus design, Large diameter and optic zone in case of decentration.
Steep to Flat	Standard spherical posterior curve, Large diameter and optic zone in case of decentration, Aspheric periphery

Table 2: Design Recommendations for Post PKTopographic Patterns

keratotomy (R.K). Although R.K has become less common because of relatively poorer outcomes and induced irregular astigmatism, one may still come across a dissatisfied patient in clinical practice.

Although soft lenses can be fit for residual regular ametropia following keratorefractive procedures, R.G.P lenses are required to correct significant corneal irregularity if present.

When fitting R.G.P lenses after R.K, P.R.K, A.L.K or LASIK, the contact lens will vault the flatter central cornea resulting in significant fluorescein pooling and a plus powered lacrimal lens. This plus powered tear layer needs to be compensated with additional minus power in the R.G.P lens, ironically resulting in a lens power similar to patient's preoperative sphere power. A common problem with post R.K conventional R.G.P contact lens fitting is midperipheral displacement. Reverse geometry lenses are more suited for post R.K corneas as these lenses are more appropriate on cornea with large, excessively flat central zones, where stability is important. These lenses with the central optical portion of lens flatter than midperiphery are similar to the contour of the cornea after refractive surgery. Reverse geometry R.G.P lens typically requires secondary curves 3.00 to 5.00 D steeper than their central curves.

Reverse geometry lenses are rarely required after P.R.K because of smooth transition from the ablation zone to the unaltered corneal periphery. They may however be required after A.L.K and LASIK because of more abrupt

curvature changes surrounding the corneal cap from use of the microkeratome. A computerized videokeratography is critical to measure the mid-periphery cornea and the curvature changes across the knee of the transition zone.

Post-LASIK Keratectasia is a relatively new condition that has emerged in the past few years. In advanced keratectasia, astigmatism may often progress to a stage that spectacle correction may not be possible and RGP lens fitting is required. The fitting philosophy for keratectasia is similar to that of keratoconus and patients do well with a large diameter lens.

Scleral Lenses

Scleral lenses are recently getting popular as a mean of visual rehabilitation in very advanced Keratoconus and other conditions of irregular astigmatism. This helps to defer surgery when corneal R.G.P lenses prove inadequate.

Generally scleral lenses give a higher quality of vision than equivalent corneal lenses because of their flatter back optic zone radius and larger optic zone diameter. The use of gas permeable material for scleral lenses has an obvious attraction in that they might reduce adverse corneal changes.

Conclusion

We may need to alter our standard fitting strategies to successfully manage patients with distorted corneal surfaces and irregular astigmatism. Various trial lens fittings and numerous prescription changes are often required for a successful fit in these challenging corneas. Fluorescein pattern is a must to determine the lens to cornea relationship and to evaluate atypical area of touch and clearance. One should try to minimize areas of excessive touch and clearance when selecting parameters. Spherocylinder over refraction through the best-fitted diagnostic lens gives us the value of power of final lenses. R.G.P materials with high D.K value should be prescribed for these conditions and also for the patients fitted the piggyback designs. A regular follow-up of the patient is a must with careful examination of contact lens fitting to see if any modifications are required in the contact lens parameters.

Suggested Reading

- 1) The CLAO Guide to Basic Science and Clinical Practice.
- 2) Contact Lenses- A textbook for practitioner and students. Editors: Anthony J Philips, Janet Stone.
- A textbook of contact lens practice. Editors: M. Ruben and M. Guillon.
- 4) IACLE Modules

Vision 2020: An Overview

Harsh Goel DO, MS, Anil Tara MS

What is Vision 2020?

Vision 2020: the Right to Sight is a global initiative launched by the World Health Organization (WHO) and a Task Force of International Non-governmental Organizations to combat the gigantic problem of avoidable blindness in the world. Launched in Geneva on February 18, 1999 by the then Director General of WHO, Dr. Gro Harlem Brundtland, Vision 2020 envisages collaboration between different governments, WHO, International Agency for Prevention of Blindness (IAPB), various funding agencies, and other international, nongovernmental and private organizations that collaborate with the WHO in prevention and control of blindness.

The Rationale behind Vision 2020

The current estimate of global blindness, based on the 1997 global population, is nearly 45 million; in addition there are nearly 135 million with significantly disabling visual impairments classified as Low Vision. Thus, globally, nearly 180 million persons need rehabilitation services along with social and other support, to have a fair opportunity to lead a productive life. Over the decade, these numbers would have obviously increased.

Blindness and disabling Visual impairments are acquiring immense social, economic and developmental importance globally; this is because of the interplay of complex implications of not only humanitarian and quality of life considerations, but also of issues of equal opportunities and integration and participation of the visually disabled in the society. In the poorest of communities, blindness can sometimes make the difference between life and death, whereas in the developed world, the access to higher education, employment and social interaction are still a challenge for the blind. Despite these well known consequences, in a public health and socioeconomic perspective, 'killing diseases' steal a march over blindness and its prevention regardless of the costeffectiveness of interventions. What gets ignored is the direct cost of blindness, which, on very conservative estimates, is a staggering 6 billion US dollars annually in countries like the USA and India. Moreover, blindness

1/31, Venu Eye Institute, Sheikh Sarai Institutional Area, New Delhi-110 017 prevention is the most cost-effective of all public health interventions, and brings about a marked reduction in DALY (Disability Adjusted Life Years). To add to the problem, global ageing of populations, both in developed and developing countries, makes the world blindness increase by about 2 million people annually; *'whereas the developed countries first get rich and then old, the developing countries first get old, and then, if ever, rich'*. Most common blinding conditions are age-related, like cataract, glaucoma and macular degenerations. Population growth in some parts of the world contributes further to the scarcity of eye care resources. The final implications are that

- The number of blind is bound to increase.
- Despite best efforts and intentions, in the period preceding the launch of Vision 2020, blindness prevention activities were grossly inadequate.

Thus, this initiative was launched with the aim: "To intensify and accelerate present prevention of blindness activities so as to achieve the goal of eliminating avoidable blindness by the year 2020".

Components of Vision 2020

The three main components of this initiative are:

- Effective disease control
- Human resource development
- Infrastructure development
 Besides these, other important components are
- Use of appropriate and affordable technology
- Resource Mobilization

Globally, five conditions have been identified for immediate attention. These conditions are:

- Cataract
- Trachoma
- Onchocerciasis
- Childhood blindness
- Refractive Errors and Low Vision

These conditions have been chosen on the basis of their contribution to the burden of blindness, and the feasibility and affordability of the interventions to control them. However, each country will decide its priorities depending upon the magnitude of specific blinding conditions. With the change in the global scenario of causes of blindness, and identification of other major conditions as they have emerged over the period 1995-2002, *Glaucoma* and *Diabetic* *Retinopathy* have been added as emerging challenges to this list. According to the magnitude and intervention strategies, these conditions can be broadly classified into three groups as below:

Group I: Conditions like Cataract and Refractive Errors (including Low Vision) which are universal, occur everywhere, and can be successfully treated through good community intervention leading to a cost effective service delivery.

Group II: Conditions like Vitamin A deficiency, Trachoma and Onchocerciasis which are focal in their distribution, affect the members of the poorest families in he poorest communities, start in childhood and can be both prevented and effectively treated inexpensively. Control measures for these conditions need to be integrated with existing primary health care programmes.

Group III: Conditions like Diabetic Retinopathy and Glaucoma which are increasing in magnitude, diagnostic and management strategies which are neither well defined nor cost-effective. These conditions, which are linked to life styles and are chronic thereby increasing the treatment costs, will acquire higher priority in societies where services for combating Group I conditions become effective.

Effective Disease Control *Cataract*

At nearly 50%, cataract is the leading cause of blindness globally; in numerical terms, there is a backlog of nearly 20 million unoperated cataract cases, whereas the global cataract surgical performance is only 10 million annually. The areas of concern, then, are *coverage* of cataract surgeries, *access* to these services and their *quality*. The projected targets of global cataract surgical rates are of 4000/ million population/ year translating into 32 million cataract surgeries globally by the year 2020.

Trachoma

An estimated 146 million people have active trachoma, for which appropriate antibiotic treatment is indicated; another 10.6 million adults with trichiasis/ entropion require surgical interventions. An estimated 6 million adults are blind due to trachomatous corneal scarring. Globally, trachoma is a major cause of blindness in Sub-Saharan Africa, China and the Middle-East. The **SAFE** strategy has been adopted to eliminate the blindness due to glaucoma, and has the following components:

- S: Surgery for Trichiasis/entropion by trained paramedical staff
- A: Antibiotic Treatment (Tetracycline Eye Ointment or oral Azithromycin) in children
- F: Facial Cleanliness (Personal Hygiene)
- E: Environmental improvement (Community sanitation)

The global Trachoma targets for the year 2020 envisage eliminating trichiasis/ entropion and reducing the prevalence of active trachoma infection to 5% of at risk population of 800 million, of whom 160 million would be children between 0-10 years.

Onchocerciasis

Onchocerciasis, which is endemic in 30 African countries and also occurs in a few foci in Yemen and six Latin American Countries, affects nearly 17 million people; out of these, 0.3-0.6 million are blind. The strategy is to establish National Onchocerciasis control programmes in all affected countries, and through development of effective surveillance systems, ensure that by 2020, no new case is reported from any of these affected countries.

Childhood Blindness

Globally, an estimated 1.5 million children are blind, out of which nearly 1.3 million are in Asia and Africa. The prevalence is 0.5-1/1000 children aged 0-15 years. At one child per minute, nearly 500,000 children go blind annually, many of whom die in childhood. The magnitude of blind years due to childhood blindness, at 75 million (number of blind X length of life) is second only to cataract. The causes of childhood blindness vary from place to place and change over time.

To eliminate Vitamin A deficiency disorders, the strategy adopted is to develop effective surveillance systems in affected countries, and achieve a nil incidence of Vitamin A blindness in all countries except in disaster situations.

In surgically treatable causes, cataract, glaucoma and Retinopathy of prematurity (ROP), services are to be developed to address these challenges.

Refractive Errors and Low Vision

Provision of appropriate spectacles and low vision devices has been identified as an important and integral part of eye care delivery. The strategies adopted are:

- Screening to identify the individuals in need.
- Refraction to evaluate the patient for his requirement of corrective devices.
- Manufacture or procurement of appropriate devices.
- Dispensing of these devices, ensuring correct prescription and a good fit.
- Follow-up services for repair and replacement as required.

Global Trends in Blindness

Despite a change in the profile of the leading causes of blindness worldwide between 1995 and 2002, many original factors have remained unchanged, namely, ageing, population growth and underdevelopment. The immediate, intermediate and long term implications of this trend will ensure that the global initiative of Vision 2020 remains relevant even after most of the current concerns have been addressed. With effective service delivery systems to tackle the current concerns getting into place and making the desired impact, other areas of concern will then be identified to be tackled by developing new public health approaches (operational research) to achieve the ultimate goals of the initiative, *i.e.*, high quality, equitable and comprehensive Eye Care services.

Immediate Implications

The immediate area of concern is the unfinished task of controlling blindness due to refractive errors, trachoma, Onchocerciasis, xerophthalmia and cataract. With effective interventional strategies, most of them are showing a downward trend, and the pressure only needs to be sustained to bring them under control.

Medium / Long term Implications

The changing global trends in life expectancy and life style are leading to life style related diseases; ophthalmic diseases are no different, and tend to be more chronic. Conditions like glaucoma and Diabetic Retinopathy have significant impact on health care expenditures, and the issues that need to be addressed are awareness about these conditions and compliance and adherence to their treatment schedules. Less avoidable are conditions like Age Related Macular Degenerations (ARMD) which are emerging as leading causes of blindness in many developed countries, and genetically determined conditions like Retinitis Pigmentosa. These changing trends emphasize the need to update projections related to health care needs and regional and country profiles of leading health care concerns. Conditions like Glaucoma and Diabetic Retinopathy need to be tackled more explicitly through an effective public health approach.

Human Resource Development At Community Level

Primary Health Care (PHC) is a fundamental concept of the WHO in improvement in health, and all the elements can contribute to blindness prevention. The PHC workers can participate in these activities in the control of blindness:

- Identification of blind and visually disabled adults and children in their own home.
- Assessment and diagnosis of patients with specific ocular conditions amenable to interventions by specialists.
- Referral for management and treatment.
- Follow-up and subsequent evaluation.

At Secondary and Tertiary Levels

The target is to enhance the number of

- Ophthalmologists per population two fold in sub-Saharan Africa and four fold in Asia
- Ophthalmic Medical Assistants/Ophthalmic Nurses four fold in Asia and sub-Saharan Africa
- Trained refractionists per population five fold
- Training all medical graduates in basic eye care
- 100% of Tertiary centres and at least 505 of secondary centres to have trained eye care Managers
- Similar targets for equipment maintenance technicians These targets are to be achieved by the year 2020.

Infra-structure and Appropriate Technology Development

The objective is to ensure universal coverage and access to services for the preservation of vision and restoration of sight. This will be achieved by ensuring availability of infra-structure in 955 of areas, and accessibility, utilization and coverage of eye care services in 90% areas by the year 2020.

For details of human resources planned at different levels, and the infra-structure needed and planned under the aegis of NPCB in India, the readers are advised to refer to the publications of NPCB.

Vision 2020: The Right to Sight in India

India was the first country in the world to launch the National Programme for Control of Blindness (NPCB) in 1976 aimed at reducing the prevalence of blindness. Vision 2020: The Right to Sight was launched in India on October 10-13, 2001 at Goa. The Government of India constituted a Working Group to prepare the Plan of Action and Strategies on the Vision 2020 initiatives in India. The draft Plan of Action was submitted in August 2002 and was approved as a document for future planning of NPCB in India.

The target diseases identified for Vision 2020 in India include

- Cataract
- Childhood Blindness
- Refractive Errors and Low Vision
- Corneal Blindness
- Diabetic Retinopathy
- Glaucoma
- Trachoma (Focal basis)

Strategies for Corneal Blindness

The major obstacle in tackling the issue of corneal blindness is the wide yawning gap between the requirement and the availability of donor corneas for corneal grafting. The recommendations to tackle this include:

- Strengthening of Hospital Cornea Retrieval Systems
- Assessment of persons needing corneal grafting

For Vitamin A supplementation to prevent Blinding Vitamin A Deficiency, focus on economically backward areas is needed, priority being slum populations, tribal regions, drought & flood prone areas and migrant populations.

Strategies for Glaucoma & Diabetic Retinopathy

These conditions need trained Human Resources, Infra-structure and System development.

Immediate Term

The major concern is the training of already qualified and working Ophthalmologists to handle these conditions. This can be done by training these ophthalmologists in a Comprehensive Eye Evaluation which should include:

- Slit lamp Biomicroscopy
- Applanation Tonometry
- Disc and Retinal Evaluation
- Gonioscopy

Intermediate Term

In the intermediate term, the Residency training programmes in Medical Colleges and other institutions offering speciality training in ophthalmology need to be strengthened, with emphasis on training in diagnosis and management of these conditions.

The second priority area is the training of Mid Level Ophthalmic Personnel in handling these conditions, again with emphasis on Basic Minimum Standards.

The next area of priority is the education of Non-Ophthalmic Physicians on the clinical profile of these conditions, follow-up care and referral systems.

Last but not the least, also needed is public education to enhance awareness about these conditions and the service delivery options available to handle them, along with the need for compliance and adherence to treatment schedules.

Long Term

The strategies in the immediate and intermediate term continue in the long term to provide high quality eye care services at all levels.

The Role of an Ophthalmologist in Vision 2020

The role of an ophthalmologist, as envisaged in Vision 2020, goes much beyond the stereotyped role of a clinician. At various levels, the ophthalmologist is expected to

contribute much more, which can be summarised as:

- Disease Control:
- Clinical care
- Planning
- Policy
- Awareness
- Resources
- Human Resource Development: Training of Ophthalmologists & Paramedical personnel
 - Curriculum Development
 - Education Materials
 - Direct Training
- Infra-structure & Appropriate Technology: Concepts and Development
 - Models
 - Equipment
 - Technology Transfer

Service Delivery Model

The model envisaged is in the form of a four-tiered Pyramid.

- Vision Centres, at the Primary level, each centre catering to the needs of a population of 50000, form the base of the pyramid.
- Service Centres, at the Secondary level, each catering to 500,000 populations form the next tier.
- Training Centres, at the Tertiary level, each catering to 5 million population come next.
- Centres of Excellence. At Advanced Tertiary Level, catering to 50 million population lie at the apex of the pyramid.

Strategic Approaches

For achieving the aims and objectives of the initiative effectively, the strategic approach includes:

- Advocacy
- Resource mobilization
- Program development
- Implementation
- Coordination

Advocacy is through

- 1. Follow-up on WHA resolution
- 2. Observing World Sight Day
- 3. Evidence-based Publications
- 4. Tool Kit
- 5. Branding of VISION 2020
- 6. Work with Governments

7. Eye Care Professionals

Resource mobilization through

- 1. Membership
- 2. Corporate Support
- 3. Restricted Program Grants
- 4. Professionals
- 5. National Plans
- 6. Global Projects

Program development through

- 1. Replicate successful models
- 2. Strengthen existing National Programs / Develop new programs
- 3. Coordination
- 4. Public-Private partnerships

Benefits of the Initiative

The benefits that will accrue from this initiative in the

long run include

- Blindness alleviation to 50 million
- Enhanced ophthalmic training
- Paramedical training
- Creation & up-gradation of facilities
- Access to modern technology

This will result in excellence in Eye Care delivered equitably, giving the Gift of Sight and a better world.

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LASEK (Laser assisted sub epithelial keratectomy)

Sanjay Chaudhary MS

Why is lasik losing its glory

With the introduction of Lasik, PRK was immediately pushed to the background. The main reasons for lasik's popularity with the patients and the ophthalmologists were the quick recovery often termed the 'wow' factor, and the no corneal haze scenario. With the increasing numbers of lasik, problems about this procedure started showing up. As ophthalmologists got down to correcting higher degrees of myopia and greater degrees of tissue ablation, cases of corneal ectasia began to get reported from all over the world. Initially a safe standard was set as a residual corneal bed of 300 mic. However, in a normal corneal thickness of 530 mic, and after making an allowance for a 160 mic flap, 70 mic was just sufficient tissue to correct about 5 D of refractive error. Customized lasik and demands for prolate cornea, meant more ablation of the corneal tissue. This also meant that another one to two dioptre was reduced from our capacity to correct the refractive errors. And what if the cornea was slightly thin and in the range of 500 mic. Would this mean depriving the person from the benefits of lasik and a major loss of revenue for the ophthalmologist by increasing his rejections. Add to this the unpredictability of the flap thickness. It was demonstrated that the best of the microkeratomes, which talked of a 160 mic flap, would actually cut a flap in the range of 90 to 230 mic. Thus, with a lot of debate, the safe standard was set to 250 mic of residual corneal bed. And this gave the much-needed flexibility to the refractive surgeon to treat a mild to moderate myopia in routine clinical practice. So myopia of 1 to 8 could be handled in most of the situations. But what about the high myopes. Enterprising ophthalmologists started reducing the corneal bed further to 200 mic. As more and more cases of corneal ectasia got reported, there was a major concern for this man made complication even though the cases reported are few and far between. Doctors wanted to fell safe both for the patients and for themselves and this led to the transition to Lasek.

Lasek gains popularity

The more the residual stromal bed one leaves behind, the less the chances of ectasia was what makes lasek ride the crest of popularity.

Lasek has the following advantages to offer:

Chaudhary Eye Centre, Daryaganj, New Delhi 1. More residual corneal bed thickness, so less chances of ectasia

- 2. No flap to be lifted, so
- a. No flap related complications like buttonhole, partial cut, free caps, flap wrinkling, epithelial down growth etc.
- b. Another 90 mic of stroma available for corneal ablation thereby enhancing the limit of correction by approx. another 5 D. over lasik.
- c. Less costly because the expense of a keratome and blades eliminated.
- d. In a better position to handle thin cornea. A person with a corneal thickness of 490 mic may still have an option of a 6 D myopia correction.
- e. Large zone treatments for better corneal prolacity and in hyperopia are now possible. This essentially means that in lasik the flap size was a restricting factor to the size of treatment. And it was seldom possible to go over a 9 mm treatment. The flap size was further reduced in a flat cornea which was a catch situation in hyperopic patients who had a flat cornea, a small flap and a need for a large treatment zone. With lasek there is no limitation to the zone of treatment.

Surgical Considerations

Lasek involves the stripping of epithelium from the Bowman's membrane in a form of a hinged flap.

An epithelial trephine of the required size in placed on the centre of the cornea after anaesthetizing the eye with 0.5% proparacaine eye drops. A 4 mm segment of the trephine at the 12 o'clock is blunt. By placing on the cornea with pressure, the trephine cuts through the epithelium sparing the underlying stroma and the 12 0'clock epithelium. The diameter of the trephine could vary from 8.5 to 10.5 mm. A similar sized alcohol well is now centered on the cornea. 20% ethyl alcohol is filled in the well and kept in position for 60 sec. It is then removed with a cellulose sponge, the well taken off the eye and the epithelium washed with BSS. After waiting another minute to allow the alcohol to weaken the epithelial Bowman's adhesions, a micro hole is used to pick up the epithelium from the edges of the trephine marks. A hockey shaped spatula is now used to roll the epithelium slowly towards the hinge exposing a clean Bowman's to work on. Excimer laser is delivered to the cornea surface to make a correction for the refractive error. The corneal surface is washed thoroughly and scrapped to rid of the debris and the condensed plume.



Fig. 1: Delayed epithelial healing with corneal haze on 9th postop day after lasek with mitimycin-C. The eye was patched with an antibiotic and lubricating ointment and the defect healed over the next 3 days.

The epithelium is carefully rolled back with the help of an irrigating cannula. Because of the loose elasticity of the tissue, the replaced epithelium usually crosses over the natural edges to overlap some of the healthy epithelium. A bandage contact lens is now placed over the epithelium where it rests for the next five days. The patient is sent home after instilling a preservative free lubricating eye drop, an antibiotic and a NSAID eye drop. These are used for the next five days. Systemic antibiotic and strong pain killers are prescribed for the next three days.

Lasek with Mitomycin-C

Here, all the above steps are the same. After corneal ablation the treated area of the cornea is exposed to 0.02% Mitomycin-c for 30 seconds. The corneal surface is then thoroughly washed with BSS for a minute to remove all traces of Mitomycin and then the epithelium is reposited back. Mitomycin-C is useful in containing fibroblastic activity and thereby reducing and delaying the chances of corneal haze, more so when attempting to treat high myopia.

Follow-Up

The bandage contact lens is removed when the old epithelium is replaced by the new epithelium generated from the edges of the wound. This usually coincides on the fifth postoperative day. After removing the Bandage Contact Lens, FML eye drops are started to replace NSAID drops to contain the tissue edema and the subsequent fibroblastic reaction. In our clinical practice we use FML six times a day for a week, and then taper it off by a drop every week over the next six weeks. Antibiotic drops are used for two weeks and lubricating drops for at least two and a half month or more as per requirement.

Disadvantages of lasek

These are similar to PRK with minor modifications:

Postoperative pain: This is a major setback for lasek. The pain is intense on the day of lasik and reduces over the next 2 to 3 days. This may be accompanied with hyperemia, chemosis of the conjunctiva, and lid oedema. Strong analgesics and anti-inflammatory are required over the first 2 to 3 days. The intensity of pain is definitely less than what is encountered in PRK. This is also in sharp contrast to lasik where there is no pain and only an occasional irritation and watering on the day of the procedure.

Button-holing: Excess exposure to alcohol does help in easily picking up the flap, but the resultant chemical trauma to the epithelium results in greater tissue reaction, more haze, pain and tissue oedema. Less exposure to alcohol or less percentage of alcohol used prevents proper loosening up of the epithelium resulting in single or multiple buttonholing. Excessive breaks in the continuity of the epithelium make the flap useless and have to be discarded. The situation then mimics a PRK.

Blurred vision: The patient encounters blurred vision for a week. This is a result of epithelial haze as the new epithelium coming in from the sides replaces the old alcohol treated epithelium. The vision clears up in a week's time. This is in stark contrast to lasik where the patient has good vision within a few hours and has total clarity by next morning.

Corneal haze: This has become the most feared complication in a long-standing follow-up of Lasek. The haze is similar to the one encountered in PRK and can be graded from I to IV. The use of Mitomycin-c in our routine clinical practice for myopia of over 4 D seems to have helped



Fig. 2: The patient developed a gr. I corneal haze in the right eye and a gr. II haze in the left eye after one and a half year of simultaneous lasek with Mitomycin-c in both eyes

in the following ways:

- 1. Haze is usually not encountered in myopia of up to 7.0 D as compared with PRK where it could be encountered after 4 D.
- 2. Myopia of 8 to 12 usually results in Gr. I haze while 12 and above may result in Gr. II to III haze
- 3. There have been situations where even -18 D has had no haze and on the other hand, even a -5 developed a mild haze.
- 4. Haze usually develops after 6-9 months of the procedure.
- 5. Haze results in a regression of the refractive error and the degree of regression depends on he severity of haze.
- 6. Haze usually regresses spontaneously over a period of 2-3 years. Low dose topical steroids could assist resolution.
- 7. The regression of haze sometimes results in some reversal of regression of the refractive error and improvement in refractive error and vision.

The above observations are not a rule but an indication of the surgeon's experience with PRK of over 10 years and of lasek with and without Mitomycin on over 400 eyes in 4 years

Lasek in relation to PRK and Epi-lasik

PRK, Lasek and now Epi-lasik involve the removing or stripping of the epithelium from the Bowman's membrane. In PRK, it is mechanical scrapping, in Lasek it is alcohol assisted while in Epi-lasik, it is again separation with a blade. Since the three are essentially similar, they carry the same advantages and disadvantages with minor modifications. The postoperative pain has definitely reduced from PRK to Lasek to Epi-lasik. This is attributed to a healthier flap over the cornea and less alcohol injury. The rate of epithelial healing has improved favoring Epilasik. This has also reduced the blurring of vision to a shorter period. However, it postulates that the effect on corneal haze may be similar in Lasek and Epi-lasik. More experience is required to understand the long-term difference between the two. Epi-lasik is prone to buttonholing, and in such a situation, the entire epithelium has to be scrapped off to continue with the procedure, and this then becomes a PRK.

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Double Elevator Palsy

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Introduction and Clinical Features

Double Elevator palsy is a clinical entity, denoting a congenital limitation of active monocular elevation that is equal in abduction and adduction, on both versions and ductions, characterized by hypotropia of the involved eye, which increases on up gaze, the affected eye not elevating beyond the midline. It may be associated with a true or a pseudoptosis. Pseudoptosis occurs when the lid is drawn down, passively in the hypotropic eye, because of the fascial attachments between the superior rectus and the levator palpebrae superioris. It resolves when the paretic eye fixes in the primary position. A ptosis from a true levator weakness, does not resolve when the paretic eye fixes. In the presence of a preserved normal or a subnormal binocular vision, a compensating torticollis is evident. The patient elevates the chin. Rarely a patient fixes with the affected eye causing a large secondary hypotropia, in the non-paretic eye (if a secondary deviation is present, the non-paretic eye may be amblyopic).

Pathophysiology

The elevation inability, may occur on a paretic or a restrictive basis, and may be congenital or may be acquired. The term double elevator palsy was originally coined to reflect, what was then thought to be the basis for the disorder, namely, a congenital palsy of ipsilateral inferior oblique muscle and the superior rectus muscle, a concept that has since been abandoned. Because this mobility pattern is now known to be associated with a spectrum of restrictive and /or paretic pathophysiology, the term monoelevator deficiency is a more accurate descriptive term. The concomitant limited elevation may result from three disparate pathophysiologic disorders; inferior rectus restriction, a superior rectus paresis, and a supranuclear disturbance of monocular elevation. In truly paretic cases, a dual palsy of the inferior oblique muscle and the superior rectus does not occur; paresis of the superior rectus muscle (the dominant elevator of the globe) is sufficient to produce the clinical picture, as the inferior oblique muscle is not capable of elevating the eye without superior rectus participation, even in adduction. Recently, a group of observations has accumulated that indicated that the superior rectus was not paretic at all, and that double elevator palsy is due to a unilateral deficit in a nucleus functions to elevate one eye only, a unilateral center for upgaze.

Neurophysiology and Neuroanatomy

An intact Bell's phenomenon (the oculocephalic maneuver), suggests supranuclear disturbance at the level of the pretectum. An absent bell's would, be indicative of either inferior rectus muscle restriction or superior oblique muscle palsy. Saccadic velocity analysis, forced duction testing, and active force generation may distinguish the two. A supranuclear disturbance is inferred, if restriction is absent and the eyes are orthotropic in the primary position. This clinical entity may be considered separate from the classical teaching of double elevator palsy; there is an absence of active elevation of one eye, both voluntary and automatic, except for the bell's sign, which is intact. No deviation is seen in the primary position. The lesion would be located slightly above the superior limit of ipsilateral portion of the, third nerve nucleus, including the fibers that connect the intermediate contralateral, center of vertical movements to the subnucleus of the elevator muscles. Most frequently, the etiology is vascular, but tumors, or a degenerative process, also may cause the anomaly. The neuroanatomic substrate, of unilateral supranuclear upgaze deficiency is controversial. It has been attributed to either lesions of the contralateral pretectum, or lesions involving the upgaze efferents from the ipsilateral rostral interstitial nucleus of the medial longitudinal fasciculus. The most recent evidence incriminates a lesion of the vertical saccadic burst or pause neurons of the rostral interstitial nucleus of the medial fasciculus.

Clinical Sub-Types

The entity may be divided into 2 broad clinical types;

1. With Inferior rectus muscle restriction - diagnosed by a positive forced duction in elevation, normal force generations (no muscle paralysis), and normal elevation saccades. Patient's with inferior rectus restriction often have an extra/ deeper lower eyelid fold, especially apparent on attempted upgaze.

2. With elevator weakness (with or without a secondary inferior rectus stricture) - characterized by reduced elevation force generation, and reduced saccadic velocities for elevation of the affected eye, bell's phenomenon if preserved is indicative of a supranuclear cause.

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Clinical Examination

A thorough clinical evaluation distinguishes Monocular elevator deficiency caused bv primary superior rectus palsy; supranuclear causes of superior rectus weakness; and primary and secondary inferior rectus restriction. Primary superior rectus palsy is characterized by a forced duction testing demonstrating no restriction to a full upward rotation and slowed upward saccades, both below and above the midline; the bell's sign is absent. Patients with primary inferior rectus restriction / fibrosis may not be hypotropic in the



generation testing is useful to verify the degree of muscle paresis. Vertical saccadic velocity studies can differentiate between inferior rectus restriction, superior rectus paresis, and supranuclear monocular elevator deficiency. Electrooculography and scleral search coil testing are methods objective of measuring saccadic velocity. A Hess chart test, documents the severity of hypotropia and binocular visual field testing documents diplopia free areas. Acquired DEP is characterized by the acute onset of diplopia in the primary position and upgaze, usually not associated with

primary gaze. The force duction testing demonstrates restriction to upward rotation because of a tight inferior rectus preventing further upgaze. A secondary contracture of the inferior rectus may occur with SR palsy or with supranuclear disorders. Hypotropia and ptosis are usually present. Forced duction testing reveals a tight inferior rectus. An exaggerated infra orbital lid crease may be seen on attempted upgaze. Supranuclear monocular elevator deficiency which is usually congenital, is characterized by intact vertical saccadic velocity below midline, but abnormal or absent velocity above the midline, monocular absence of vertical eye movements, in superior fields of gaze and no resistance to upward rotation by force duction testing. Bell's phenomenon is present, indicating an intact oculomotor nerve, fasciculus and nucleus. The Binocularity is tested by Worth four dot testing; and stereopsis tests should be done both in forced primary gaze and down gaze. Many patients of double elevator palsy would fuse in down gaze, but suppress or develop diplopia in primary position. The amount of vertical strabismus and any horizontal deviation may be recorded with the prism cover tests. The eyes may be orthotropic in primary position, or a primary or secondary deviation may be present. A characteristic feature of monocular elevator deficiency is that the amount of attempted elevation does not change on adduction or abduction. Version and duction testing should be done in all nine diagnostic positions to identify any relative restrictions of gaze. Park's 3-step test and Bielshowsky's head tilt test are used to rule out a superior oblique palsy. Forced duction testing and active force blepharoptosis. Bell's phenomenon is usually preserved, the patient often assume a chin up position to maintain binocularity and diplopia.

Indications for Treatment

Include a significant vertical deviation in primary position with or without ptosis, and an abnormal toticollis. A deviation causing suppression and amblyopia, or in acquired cases- diplopia, also requires treatment. A marked compensatory head posture, may resolve to surgery. A ptosis which interferes with fixation in the primary position, or is cosmetically poor may require correction too. Traditionally the strabismus is corrected prior too the ptosis, although, both may be done at the same sitting. The goal of the surgery is to improve the position of the affected eye in primary gaze, thereby increasing the field of binocular single vision.

Surgical Techniques

The type of surgery depends on the cause of the elevation deficit. It is important to evaluate preoperatively for the presence of an upgaze saccade and to perform forced ductions at the time of surgery to make the correct procedural choice. In cases of pseudo-double elevator deficiency with a restrictive etiology, with normal upgaze saccade, recess the ipsilateral inferior rectus muscle, usually around 5-6 mm, depending on the size of the hypotropia. A conjunctival recession may be added. In cases of residual hypotropia, an ipsilateral superior rectus muscle resection may be needed. Cases with lack of upgaze saccades combined with a weak superior rectus muscle on forced generation testing, indicates true double elevator palsy. In these cases, a recession of the ipsilateral inferior rectus alone will not correct the hypotropia. A full tendon transposition of both horizontal recti towards the extremities of the insertion of the superior rectus is performed. The reinsertions should be almost parallel to the limbus, their superior extremities close to the extremities of the superior rectus insertion and slightly further from the limbus, whereas the inferior extremity should be slightly nearer the limbus than the superior rectus muscle insertion. Some prefer the partial tendon transfer (Hummelsheim) instead of the full tendon (Knapp) transposition to avoid the complication of anterior segment ischemia. A ciliary blood vessel sparing technique is another option. If combined with an inferior rectus recession, one should wait a period of at least 4 months between the two surgeries. In severe cases of hypotropia, consider adding a recession of the contralateral superior rectus muscle. A recent technique of augmented Knapp procedure with superior posterior fixation sutures (foster) is the preferred treatment. A recession of the ipsilateral Inferior Rectus muscle may sometimes cause a limitation of active depression, leading to intractable diplopia in down gaze, in susceptible patients; the contralateral

Superior rectus muscle should be recessed in such situations.

Whatever the procedure, there is always a possibility of under correction. A thorough patient counseling, and oculoplastic evaluation for treating coexisting ptosis is essential. The goal of the surgery is to improve the position of the affected eye in primary gaze, thereby increasing the field of binocular singular vision. It should be clear that surgery will not cure the monocular elevator deficiency, and the affected eye will still not elevate as well as the other eye.

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Gonioscopy Techniques for Narrow Angles

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On routine gonioscopy, the angle structures may not be identified in eyes with a steep iris configuration and a narrow angle. A steep approach to the angle makes examination difficult in such cases and the examiner has to use special maneuvers to look over the iris. This technique of manipulating the goniolens to visualize over a steep iris (over the hill view) is known as dynamic or manipulative gonioscopy. The second technique of indentation gonioscopy involves the use of a Zeiss type corneal indirect gonioscope to indent the cornea and thereby open up the angle, in order to differentiate between appositional or synechial closure. A new technique for objective estimation of the angle has also been developed, which uses a reticule mounted on the slit lamp oculars to measure the distance between the iris insertion and the Schwalbe's line. This technique is known as Biometric





Fig. 1.3: Narrow angle recess on normal view

Fig. 1.2: Manipulation of Gonioscope towards angle being viewed

Fig. 1.4: Opening up of angle recess on manipulation

Gonioscopy. Another method is by using the technique of sclerotic scatter on slit lamp biomicroscopy. The angle structures can sometimes be seen and identified more accurately than with direct illumination.

Manipulative Gonioscopy

The anterior chamber angle width should be initially evaluated in primary gaze with all types of goniolenses positioned centrally on the cornea. With the scleral type lenses care is to be taken to avoid indentation of the peripheral cornea, as this can narrow the angle. Manipulation is of value in studying angle anatomy in narrow iridocorneal angles. A more tangential viewing of the angle aids in identification of angle structures obscured by a convex iris. This can be achieved in Goldmann type lenses by simply asking the patient to look in the direction of the mirror or moving the mirror towards the angle being

Glaucoma Research Lab Dr. R.P. Center for Ophthalmic Sciences, AIIMS, New Delhi - 29 viewed (Figure 1.1- 1.4). Once angle structures are seen, the slit beam may be narrowed to identify the corneal wedge and interpret the findings.

The examiner should report the normal angle view in primary gaze and then document the opening of the angle on manipulation by asking the patient to look into the mirror of the gonioscope, opposite to the angle being examined. For example if you are examining the inferior angle with a Goldmann single mirror gonioscope, the mirror is positioned superiorly. After viewing the angle in primary gaze, you now ask the patient to look upwards, ie. towards the mirror. This allows a better view of the inferior angle as the examiner can look over the iris and into the angle. If the patient looks in the direction opposite of the mirror, the angle appears narrower and vice versa.

This manipulative maneuver can be facilitated by shifting the fixation light, which is positioned in front of the other eye, in the same direction, as that of the goniomirror being viewed. The examiner can produce a similar effect by moving the lens towards the part of the angle to be examined (eg. By displacing a goniolens inferiorly when examining the inferior quadrant).

Fig. 1.5: Indentation / Pressure Gonioscopy pushes aqueous into angle and opens up the angle recess

Indentation Gonioscopy

With the corneal type goniolenses, that have a small diameter, the central cornea may be indented to force the aqueous out and artificially widen the angle. Because the smaller radius of curvature allows these lenses to come into direct contact with the anterior corneal surface, central depression of the cornea will displace aqueous humor peripherally and the iris root posteriorly (Figure 1.5-1.7). This technique is also known as pressure or dynamic gonioscopy. When the iridocorneal angle is optically narrow, indentation gonioscopy facilitates the identification of angle structures. Should the angle be closed, indentation helps differentiate appositional from synechial angle closure. This is important as synechial closure is irreversible, while appositional closure can be reversed.

Indenting the central cornea however causes folds in the central cornea, which can distort the angle details. The angle structures may be identified by using a narrow slit beam and applying and withdrawing pressure in repeated sequence. Also sliding the lens towards the angle reduces the folds and improves view. Extreme gaze in the direction of the mirror, and lifting of the goniolens while pressing inward can aid maximal displacement of the aqueous peripherally when angle structures are not readily apparent despite indentation. Indentation is not possible with the Goldmann type gonioscopes as they have a diameter greater than the corneal diameter and tend to narrow the angle on pressure. Indentation is difficult when the intraocular pressure is higher than 40mmHg.

When no angle structure is directly visible before indentation, the closure of the angle can be due to three reasons ;

- 1. Synechial
- 2. Appositional
- 3. Optical (apparent closure of the angle due to steep curvature of the peripheral iris, impairing visibility of

angle structures)

When no angle structure is directly visible before indentation, four things can happen on indentation (Figure 1.8 a):

- A) The iris moves peripherally backwards, assumes a concave configuration and the angle recess widens. This represents an appositional closure with a suspicion of a relative pupillary block (Figure 1.8 b).
- B) The iris moves peripherally backwards, but the

Fig. 1.6: Opening of angle recess on pressure gonioscopy

Fig. 1.7: Indentation Gonioscopy being performed with Posner Lens

Fig. 1.8(a) : Indentation gonioscopy in an apparently closed angle

Fig. 1.8(b): If the angle opens up, it was a case with appositional closure

Fig. 1.8(c): Indentation reveals synechial Irido corneal adhesions (PAS)

Fig. 1.8(d): A thick lens does not allow the chamber to deepen or the iris to move backwards

periphery of the iris bulges out and does not assume a concave configuration. This represents an anteriorly displaced ciliary body and iris root, typically seen in plateau iris.

- C) The angle widens but iris strands remain attached to the outer wall of the angle. This represents organic synechial closure of the angle (Figure 1.8 c).
- D) The iris moves only slightly and evenly backward, but retains a convex profile. This can occur due to an anteriorly displaced lens or a large diameter lens (Figure 1.8 d).

Biometric Gonioscopy

This is a new method for objective measurement of the anterior chamber angle proposed by Congdon et al (Ophthalmology 1999;106:2161-7). In this method gonioscopic measurements are performed with the help of a special reticule. The anterior chamber angle is viewed under the following conditions on a Haag-Stret 900 BM slit lamp :- ambient lighting from a small side lamp is used to provide only an indirect illumination with a total magnification of x16, power of 6W, middle filter setting and a slit lamp beam of 4mm length and 1mm width. The reticule is mounted on a slit lamp x 10 ocular and ruled in

0.1 mm units, which is used to measure the distance between the insertion of the iris and the Schwalbe's line. Measurements are recorded separately in the superior, inferior, nasal and temporal quadrants. If the angle is closed a measurement of 0 is recorded while an occludable angle is defined as one with an average measurement of 0.25 mm or less for the four quadrants.

This method correlates well with other measures of the anterior chamber angle like conventional gonioscopy and Scheimpflug photography, shows a much higher degree of interobserver reliability than conventional gonioscopy and can be readily learned and performed by an inexperienced observer. Hence it

offers a definite advantage over conventional gonioscopy which is purely a subjective technique and has a long learning curve.

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Confocal Scanning Laser Ophthalmoscopy (HRT II) Glaucoma Application

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Glaucomatous Optic Neuropathy (GON) continues to baffle the ophthalmologists as the more we know about it, more and more new influencing factors emerge. Intraocular Pressure (IOP) remains the only measurable parameter that can be pharmaco-modulated and now we know that controlling it may not be sufficient to arrest visual field loss. The human retina comprises of approximately 1.2 -1.5 million neurons that bundle together to emerge as the optic nerve. GON causes gradual irreversible loss of these neurons and some studies suggest that nearly 40% neurons may have been lost when the disease clinically manifests as visual field defects and it may take 4-5 years of gradual nerve fiber loss for this. It is therefore critical that objective means of measuring the health of Retinal Nerve Fiber Layer (RNFL) be devised so that pre-clinical NFL loss and its sub-clinical progression can be diagnosed and treated in time.

The concept

The Scanning Laser Ophthalmoscope (SLO) [1] is a device which allows the visualisation of the fundus of the eye. A low power laser is scanned in a faster fashion over the retina. The reflected light is descanned, detected by a photodiode and the digitised image is stored in a computer. This technique has been used has a method of ophthalmic diagnosis over the past decade. Confocal laser scanning ophthalmoscopes (CSLO) use a low-power laser combined with special confocal optics to image the retina and the optic disc in three dimensions. The laser scans across the retina in x, y, and z directions at a number of different depth planes to obtain several separate two-dimensional images of the retinal structure. Computer software is used to construct a topographical three-dimensional image from these individual two-dimensional images. Analysis of each three-dimensional image involves the calculation of several topographic optic disc (OD) parameters (cup area, cup shape, cup volume, cup/disc area ratio, rim volume, rim area, retinal height, retinal cross-sectional area, retinal nerve fiber layer thickness and cross-sectional area). In the CSLO [2] a small pinhole is placed in front of the photodiode on a conjugate plane to the retina. By moving the pinhole it is possible to select light reflected from different focal planes in the retina and so produce tomographic images. The clinical importance of tomographic imaging of the retina is high, as many diseases affect the deeper structures in the retina and so are difficult to analyse with conventional 2D imaging techniques such as the fundus camera.

Heidelberg Retina Tomograph II (HRT II) is a Confocal laser scanning ophthalmoscope that can be used to measure the Retinal Nerve Fiber Layer (RNFL) thickness at the Optic Disc in the Glaucoma application. It uses a 670 nm low power laser and acquires 3 separate image series where each image comprises of 16-64 planes and each plane itself is formed of 384*384 pixels. Each pixel is 10u in size and the image quality is measured as standard deviation. (Fig.1)

The Principle

It is observed that Papillomacular Bundle (PMB) remains in tact even during very advanced stages of GON and it usually lies between 350-356 deg of the Optic Nerve Head (ONH) (Fig.2). Its thickness is 50 um and this is where the HRT II defines the reference plane (Fig.3). The component above this considered the Neuro-retinal Rim (NRR) and the component below is the cup. Based on this premise, various stereo-metric parameters are calculated and the five important ones are

- Rim Area
- Rim Volume
- Height Variation Contour
- Mean RNFL Thickness
- Cup Shape Measure

Recording

First the camera is adjusted by cleaning the chinrest, front fixation and the objective. The chinrest and camera head are adjusted on medium position and camera head is moved to the left or right. Expected refraction is adjusted on the objective and if required, astigmatism correction lenses are applied. Camera takes two images, a topography image and a reflectance image (Fig4). The quality of the image is determined by the displayed image quality display.

Using 6-8 points (Fig.4) a contour line is marked to

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Fig. 2: Papillo-macular Bundle

Fig. 1: Computing the disc topography

delineate the ONH. This contour line drawn at the first examination is automatically transferred to all subsequent examinations and the stereo-metric parameters so calculated will form the baseline for the future comparisons and therefore must be taken with great care. Only 6-8 points should be taken since more points make it difficult to control shape. The scleral ring is frequently apparent as a depressed (pale) band. The contour line has to be placed at the inner edge of the scleral ring. The depressed band can be identified as a 'valley' using the interactive height profile (Fig.4).

HRT II Printout (Fig.5)

Section 1- Patient examination & data Section 2 -

- Red = Cup
- Blue = Sloping Rim
- Green = Stable Rim

Section 3 - Stereo-metric parameters (Top5)

- Rim Area
- Rim Volume
- Cup Shape Measure

Fig. 4: Topography image (Left), Reflectance Image (Right)-Contour Line

Height Variation Contour

- Mean RNFL Thickness
- Standard Deviation
- < 10 Excellent</p>
- 10-20 Very Good
- 20-30 Good
- ◆ 30-40 Acceptable

• 40-50 Probably Cataract *Section 4* - Reflectance image

Compared to Normal Database with Simple Classification

- Normal (Green)
- Borderline (Yellow)
- Outside Normal (Red)

Section 5- Retinal surface height profile

• Along Contour Line (Green)

• Reference Plane (Red) set 50 microns below Papillomacular bundle

Section 6- Moorfield's regression analysis

- Column = Total ONH
- Cup (Red) v Rim (Green)
- Age Dependent Confidence Intervals
 - a) "Within Normal Limits" (Green)% Rim ³ 95% Limit
 - b) "Borderline" (Yellow)% Rim between 95% and 99.9% Limits
- c) "Outside Normal Limits" (Red)

% Rim lower than 99.9%

Interpreting Progression

Glaucoma is а progressive disease and there is significant individual variability which makes labeling an eye glaucomatous after one single test hazardous. Therefore proven progression of the disease becomes critical to the diagnosis and management. The base line measurements are extremely since those important parameters alone are taken for

Fig. 5: HRT II Printout-Sections1, 2, 3,4,5,6 (See text)

further retesting. Therefore the image quality (as ascertained by Standard Deviation) should be good and it should be ensured that ONH is centered, illumination is even, refractive error is incorporated and eye movements are minimal. It is claimed that disc changes are more frequent than field changes. Progression requires 3 consecutive readings (Baseline + 3 follow-up) to perform a Topographic Change Analysis (TCA).

TCA can be done by two methods:

- i) Change Probability Maps
- ii) Stereo-metric Parameters
- (i) Change probability maps (Fig.6) are independent of the reference plane and the contour line and are calculated automatically comparing mean topography images.

Red signifies "Significant" Depression

Green signifies "Significant" Elevation

Fig. 7: Change Probability Maps

Change is calculated by local change in surface height measured in microns at the location selected

A height change is considered significant

- If it is repeated in at least two (better: three) consecutive follow-up examinations,
- If it is region of at least 20 connected super-pixels.
- (ii) Parametric change is evaluated in the follow-up diagram that plots normalised streo-metric values vs. time. If average normalised parametric values decrease by more than -0.05 significant in 2 consecutive

examinations it is deemed "suspected" and if it appears in 3 consecutive examinations it is considered "confirmed" progression (Fig.7).

Frequency of Examinations

Time determines the speed of progression and repeated examinations can not detect disease. High risk cases on basis of race, age, family history and raised IOP should undergo a 6 monthly examination and other patients may be followed up annually. Examinations may be done more frequently for the first 18

months in patients showing signs of clinical progression so as to start detecting statistical "change".

Conclusion

HRT II is an objective means of assessing the thickness of the RNFL at the ONH. It utilizes an ingenious concept of defining a reference plane at the thickest part of the ONH and thus virtually eliminates the subjective concept of cup, NRR and optic disc. The change analysis is user independent and records a change only when it is statistically tenable. This makes it a useful adjunct to a scientific glaucoma practice.

References

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